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Cardiology in the Young

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Cardiology in the Young

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1. Arrhythmia/electrophysiology

O26
Initial experience of ICE integrated with electro-anatomic mapping CARTOSOUND® for cardiac ablations in children.

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Introduction: CartoSound®(EAM-ICE) intra-cardiac echocardiography(ICE) integrated with electro-anatomic mapping(EAM) is a new tool that has emerged in the field of pediatric electrophysiology. The combination of both technologies allows defining the arrhythmia propagation maps for different arrhythmia substrates while shows direct visualisation of cardiac structures and electrode tip. This report aims to describe our initial experience with CartoSound® for cardiac ablations in paediatric patients, with and without congenital heart defects(CHD).
Methods: The clinical and electrophysiologic data of the first 30 paediatric patients (<18 y old) in whom we used CartoSound@ (EAM-ICE) system for cardiac ablation was reviewed. The procedure was done using ALARA Fluro-protocol. A 3-dimensional shape of the heart was created using 8Fr SoundStar® Probe. The ICE catheter was introduced via femoral vein access in 28 patients. In 2 cases, the ICE catheter was placed in the oesophagus.
Results: The median age was 13,4 y (5-17,4) with 16 females. Arrhythmia substrates comprised 20 patients with WPW syndrome, 4 AVNRT, 3 VT and 3 focal atrial tachycardia. Twenty-one patients had a structurally normal heart, and 9 had CHD. Procedure duration was 200±100 min(median ± SD), and fluoroscopy time was 10± 3 min. 3D Cardiac anatomy acquisition took 45 ± 30 min. ICE image quality was excellent in all cases. In all 7 patients with left-sided substrates, the transseptal puncture was guided by ICE with minimal fluoroscopy without tracheal intubation. Cryoablation was used in 2 patients, one with a parahisian pathway and another one with AVNRT in a fenestrated TCPC. The other 28 cases, radiofrequency was used. Non irrigated tip catheters were used in 26, and only two irrigated tip catheters were used for 2 pathways inside the coronary sinus. All applications were performed with direct visualisation of the ablation catheter tip in contact with the endocardium. Correlation between ablation efficacy and the cardiac contact with catheter tip was excellent. Acute success during ablation was achieved in 27/30 patients. No complications were reported.
Conclusions: CartoSound®(EAM-ICE) is an excellent tool for guiding cardiac catheter ablation in children. The demonstration of catheter position and direct visualisation of catheter tip contact during ablation lesions correlates well with ablation success.

O27
Outcomes of paediatric pacing in univentricular versus complex biventricular congenital heart disease

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Introduction: Pacing in a univentricular circulation has been associated with worsened outcomes and survival. We sought to investigate how the outcomes of pacing in children with a univentricular circulation compare to the outcomes of pacing in complex biventricular congenital heart disease. In addition, we aimed to identify predictors of adverse outcomes for the two groups.
Methods: We performed a retrospective review of medical records and the pacemaker database at our institution of all paediatric patients with major congenital heart disease who underwent pacemaker implantation under the age of 18 years between November 1994 and October 2017. The primary outcome was to compare the incidence of adverse events between the paced patients with a complex biventricular circulation and those with a univentricular circulation. The secondary outcome was to identify predictors of adverse events.
Results: 89 patients were included; 19 (21%) with a univentricular circulation and 70 (79%) with a complex biventricular circulation. 96% of pacemaker systems were epicardial. The incidence of adverse outcome was similar between the two groups (univentricular 31.6%, biventricular 22.9%). Death and transplantation were the most serious adverse outcomes. 5 (5.6%) patients died and 2

(2.2%) underwent heart transplantation. Most adverse events occurred within the first 8 years after pacemaker implantation. Univariate analysis identified 5 predictors of adverse outcomes in the patients with a biventricular circulation but none in the univentricular group. The predictors of adverse outcome in the biventricular circulation were the right morphologic ventricle as the systemic ventricle, age at first congenital heart disease operation, number of congenital heart disease operations and female gender. Lastly, of the patients who received a high percentage ventricular pacing, the non-apical lead position was associated with a higher risk of an adverse outcome with an estimated hazard ratio of 3.39 (95% CI 1.02-11.26, $p = 0.034$).
Conclusions: Children with a pacemaker and a complex biventricular circulation have similar adverse outcomes and survival to the children with a pacemaker and a univentricular circulation. This study identified specific predictors of adverse outcomes. The only modifiable predictor was the epicardial lead position on the paced ventricle, stressing the importance of apical placement.

O28
Cardiac abnormalities in an exclusively paediatric congenital myotonic dystrophy type 1 cohort

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Introduction: Myotonic dystrophy type 1 (DM1) is the most prevalent inherited neuromuscular dystrophy in adults. It is an autosomal dominant multisystem disease involving myotonia, progressive muscle weakness and cardiac abnormalities. Whilst the latter are well-defined in adults, there is scarce published data in paediatric population. This study aimed to investigate the yield and progression of cardiac disease in paediatric DM1 patients, focusing on congenital DM1 (cDM1).
Methods: A retrospective observational study was conducted of DM1 paediatric patients referred to our centre between December 2000 and November 2020. They were classified into cDM1 and non-cDM1 based on age of onset of symptoms and severity of disease. Genetic confirmation was routinely performed. Patients were systematically evaluated including medical and family history, 12-lead-ECG, signal-averaged-ECG, echocardiogram and ambulatory ECG monitoring. Electrocardiographic abnormalities were defined following current guidelines.
Results: We included 56 (83.6%) patients with cDM1and 11 (13.4%) with non-cDM1. Median follow-up time of cDM1 patients was 8.0 years [IQR 3.25-11.0], 33 (58.9%) were females and 54 (96.4%) had maternally inherited cDM1. 49 (87.5%) and 44 (78.6%) cDM1 patients had baseline and follow-up 12-lead-ECG, respectively. 43 (87.8%) presented ECG abnormalities during follow-up. The prevalence of cardiac conduction disease was 46.9% (n=23) being first degree AVB the most common form (42.9%). The abnormality with the highest increase in prevalence from baseline to follow-up ECG was low QRS voltage (16.7%). One patient (1.8%) underwent a pacemaker implantation after syncopal events in the context of second degree AV block Mobitz type I and II and following an electrophysiological study that confirmed cardiac conduction disease. There were four (7.1%) deaths [median age 8.5 years (IQR 6.25–16.75)], one due to progression of disease and three sudden and unexplained, and an acute electric event could not be ruled out.
Conclusions: This is the largest study that has comprehensively evaluated cardiac abnormalities and the first that has demonstrated progressive ECG disturbances in a paediatric cDM1 cohort. We have been able to identify a high yield of electrocardiographic abnormalities in these patients although no ventricular arrhythmias were documented. These findings stress the importance of continued clinical evaluation of paediatric cDM1 throughout childhood, even when asymptomatic.

Baseline information	Baseline ECG (n=51)	Follow-up ECG (n=46)
Median age at the ECG (years)	5.2 [1.8 – 10.5]	12.5 [7.2 – 16.5]
1st degree AV block, n (%)	12 (23.5)	15 (32.6)
Nonspecific intraventricular conduction disturbance	12 (23.5)	14 (30.3)
Right BBB, n (%)	3 (5.8)	0 (0)
Left BBB, n (%)	0 (0)	0 (0)
Left Anterior Fascicular Block, n (%)	2 (3.9)	5 (10.9)
QRS axis		
Normal, n (%)	21 (41.2)	18 (39.1)
Left axis deviation, n (%)	7 (13.7)	10 (21.7)
Right axis deviation, n (%)	12 (23.5)	9 (19.6)
Superior axis deviation, n (%)	5 (9.8)	5 (10.9)
Indeterminate axis, n (%)	6 (11.7)	4 (8.7)
Low QRS voltage, n (%)	13 (25.5)	19 (41.3)
Poor R-wave progression, n (%)	1 (2.0)	7 (15.2)
Abnormal repolarisation (Flat/inverted T waves)	12 (23.5)	16 (34.8)
ECG abnormalities	44 (86.3)	44 (95.7)

O29
Performance of pacemaker leads in alternative lead positions after tricuspid valve replacement.

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Introduction: Bradycardic arrhythmias requiring pacemaker implantation are still a common complication after tricuspid valve replacement (TVR). Leaving the pacemaker lead in an extravalvular position may help to prevent prosthesis dysfunction. This study was aimed to examine the mid-term outcome of paravalvular or single coronary sinus leads with respect to lead survival and prosthesis dysfunction in patients after TVR.
Methods: A retrospective case control study of patients with TVR and necessity for ventricular pacing was conducted. Consecutive patients from the database of the Leipzig Heart Center were included. Data of the paravalvular lead group (PVG) and coronary sinus lead group (CSG) were compared to a control group with conventional transvalvular lead positioning (TVG).
Results: 80 patients with TVR and cardiac pacemaker (TVG n=13, PVG n= 40, CSG n=27) were included. The mean follow-up was 2.8 years. The rate of lead revisions (TVG 15.4%, PVG 2.5%, CSG 7.5%) was lower in PVG but without significance (p=0.286). The CSG demonstrated significantly higher pacing thresholds at

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follow-up (1.4 V/0.8ms) than TVG (0.5 V/0.4ms), (p=0.004). However, the deterioration of threshold amplitudes during follow-up was similar in CSG (7.4%) and PVG (7.5%) compared with controls (7.7%). Function of TV prosthesis regarding development of stenosis or regurgitation showed a similarity between PVG and CSG compared to control(regurgitation PVG p= 0.692, CSG p=1; stenosis PVG p=0.586, CSG 0.69).
Conclusions: Paravalvular positioning of pacemaker leads seems to represent a reasonable alternative to the conventional transvalvular lead positioning with regard to function of the lead and tricuspid valve prosthesis.

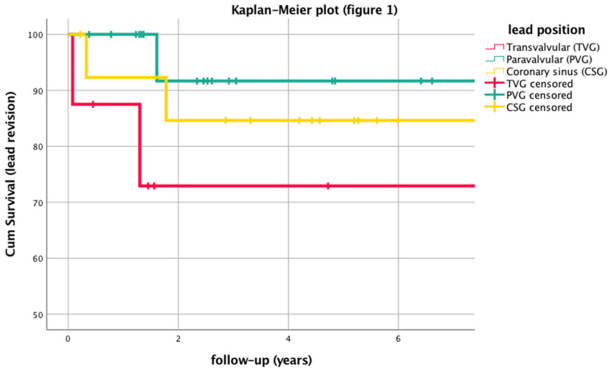

Figure 1 Kaplan-Meier plot for the number of lead revision at short-to-mind- follow-up between TVG, PVG and CSG.

O30
Results of the SIDECAR (S-IcD registry in European paediatriC and Adult patients with congenital heaRt defects) project.

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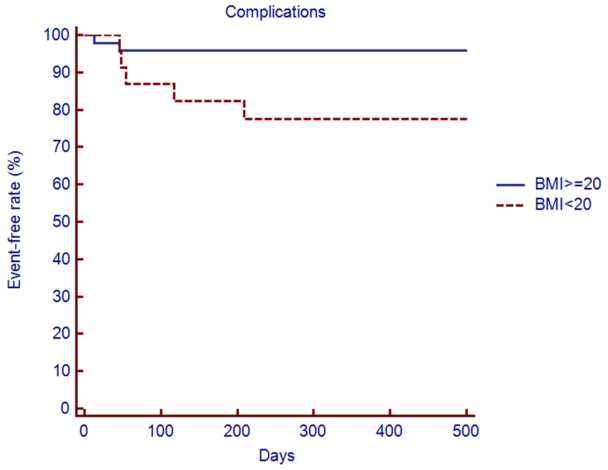
Introduction: Use of the subcutaneous implantable cardioverter-defibrillator (S-ICD) to prevent sudden cardiac death is increasing. Few data exist on S-ICD in young patients. We report the results from a multicenter European registry of paediatric and young adult patients who underwent S-ICD implantation.
Methods: Observational, ambispective, non-randomized, standard-of-care study on S-ICD implantation/follow-up in young patients with inherited arrhythmias (IA), idiopathic ventricular fibrillation (IVF), cardiomyopathies, and congenital heart defects (CHD). P<0.05 is significant.
Results: 78 patients (52% cardiomyopathies, 19% IVF, 16% CHD, 13% IA), 45% females, mean age 16±5 years (80% <18 years), with body mass index (BMI) 22±4, ejection fraction (EF) 54±17, underwent S-ICD implantation, two thirds of them for primary prevention. A two-incision procedure was used in 82% of patients, a standard procedure (three surgical incisions) in the remaining ones. Devices were implanted in subcutaneous (60%), intermuscular (25%) e submuscular (15%) sites. Shock/conditional zone were programmed at median bpm 240/200. No intraoperative complications occurred. Over a 21-month median follow-up (25th–75th percentiles, 7–38 months), 18% of patients received appropriate shocks (1st shock effective in 77% of patients, 3rd shock in 8%), and 14% inappropriate shocks (supraventricular tachycardias, SVT, 40%, T-wave oversensing, TWO, 30%). New programming (TWO) or proper drug therapy (SVT) avoided further inappropriate episodes. Device-related complications requiring surgical intervention (skin erosions, pocket infections) occurred in 10% of patients, mostly (85%) in subcutaneous implanted devices. A significant higher risk of complications was seen in patients who underwent standard procedures [hazard ratio (HR) 4.3, 95% confidence interval (CI) 0.5-34; P = 0.038] and those with BMI <20 (HR 5.1, 95% CI 1–24; P = 0.031). Non-device related death occurred in 3% of patients, heart-transplantation in 7%, shift to transvenous pacing in 3%.
Conclusions: These results of a multicenter European paediatric registry suggest that S-ICD is safe and effective. Improvement of implantation techniques and BMI>20 are associated with better outcome.



O51
Risk factors for sudden death in childhood – a comparison between non-syndrome hypertrophic cardiomyopathy (HCM) and Noonan-associated HCM

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Introduction: Risk-stratification for primary prophylactic ICD-implantation in patients with hypertrophic cardiomyopathy (HCM) is quantified by risk for sudden death (SCD) during five year follow-up. 22-30% of cases of HCM presenting in childhood

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are associated with Noonan or related syndromes where risk-factors have not been studied.
Methods: A Swedish national cohort of patients with HCM presenting <19yrs of age, mean follow-up 10.6yrs, contains 107 patients with familial and sporadic HCM without syndrome-association (sarc-HCM), and 40 with HCM associated with Noonan, Leopard or Cardio-Facio-Cutaneous syndromes (syndr-HCM). Individual proposed risk-factors were assessed with ROC-curve analysis (C-statistic) in the two groups, and cut-offs compared.
Results: Median age at diagnosis were 10.8yrs for sarc-HCM, and 0.20yrs in syndr-HCM. Age at SCD (n=30) were 15.6 [IQR 11.3-23.8] and 14.4 [12.3-41.8] yrs respectively. Some phenotypic risk-factors at diagnosis were the same in sarc-HCM and syndr-HCM: ECG risk-score C-statistic 0.83 [0.75-0.92;p<0.001] and 0.80 [0.63-0.97;p=0.032]; Detroit Z-score maximal wall thickness 0.78 [0.68-0.88;p<0.001] and 0.85 [0.67-1.0;p=0.024]. Other differered: Presence of LV outflow-tract obstruction at rest at diagnosis was a significant risk-factor in sarc-HCM 0.68 [0.56-0.80; p=0.004] but not in syndr-HCM 0.32 [0.05-0.59;p=0.19]. For SCD occurring during first five yrs of follow-up ECG risk-score >5 at diagnosis had C-statistic of 0.87 [0.80-0.95], sensitivity of 100 [66-100]%, specificity 74 [64-83]%, positive predictive value (PPV) 27.3 [13.3-45.5]%, and negative predictive value (NPV) 100 [95-100]% for sarc-HCM. Corresponding values for Detroit-Z-score>4.5 was C-statistic 0.78 [0.67-0.90], sensitivity 86 [42-100]%, specificity 66 [56-76]%, PPV 16.2 [6.2-32.0]%, NPV 98 [91-100]%. ESC2014 guidelines are non-significant, p=0.50, with sensitivity of 67 [30-93]%, specificity of 47 [37-58]%, PPV 10.7 [4.0-21.9]%, NPV 94 [83-99]%. For syndr-HCM there are few early end-points and wide confidence intervals, ECG risk-score >5 had sensitivity of 100 [29-100]%, specificity 64 [44-81]%, PPV 23.1 [5.0-53.8]%, NPV 100 [81-100]% over first 10 years follow-up; Detroit Z-score>6 was optimal cut-off for wall thickness with sensitivity 100 [16-100]%, specificity 92 [78-93]%, PPV 40 [5.3-85]%, and NPV 100 [89-100]%.
Conclusions: Among phenotypic risk-markers, ECG risk-score >5 points and Detroit Z-score of maximal wall thickness >4.5 are the best risk-markers common to sarc-HCM and syndr-HCM, but in syndr-HCM a higher Detroit Z-score cut-off of >6 performs better.

O52
Robotic Navigation shows superior long-term outcomes in Pediatric Atrioventricular (Nodal) Tachycardia ablation compared to Manual RF and Cryoablation

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Introduction: Catheter ablation (CA) is frequently used as a first-choice treatment for tachyarrhythmia in pediatric patients. The currently available CA techniques differ in manner of catheter steering and energy sources. There are no studies comparing long-term outcomes between available CA techniques in pediatric patients with atrioventricular reentry tachycardia (AVRT) or atrioventricular nodal reentry tachycardia (AVNRT) mechanisms. The aim of this study was to compare procedural parameters and outcomes of remote magnetic navigation-guided radiofrequency (RF) ablation (RMN), manual-guided RF ablation (MAN) and manual-guided cryoablation (Cryo).
Methods: This single-center, retrospective study included all first consecutive CA procedures for AVRT or AVNRT mechanisms performed in pediatric patients with no structural heart disease from January 2008 until June 2019. Three study groups were defined by the ablation technique used: RMN, MAN or Cryo. Primary outcome was recurrence of tachyarrhythmia and/or pre-excitation on ECG. Baseline clinical parameters, procedure times and complication rates were also evaluated.
Results: In total, we included 223 patients, aged 13.8 ± 2.8 years, with a mean weight of 55.6 ± 14 kilograms. In total, 108 procedures were performed using RMN, 76 using MAN and 39 using Cryo. RMN had the lowest recurrence rates at a mean follow-up of 5.5 ± 2.9 years (AVRT ablation: 4% vs. 16% vs. 55%, P<0.001; AVNRT ablation: 8% vs. 8% vs. 36%, P=0.008; for RMN vs. MAN vs. Cryo respectively). In AVRT ablation, procedure and fluor times were comparable between groups. However, in AVNRT ablation, RMN and MAN had significantly lower fluoroscopy times compared to Cryo (10 (IQR 7-14) vs. 9 (IQR 6-26) vs. 15 (IQR 10-22) minutes respectively, P=0.040). Procedure times were shortest in MAN ablation (101 (IQR 87-121) vs. 88 (IQR 62-99) vs. 120 (IQR 88-143) minutes respectively, P=0.018). We observed minor complications in 3 patients (1%), which were comparable between groups.
Conclusions: In pediatric patients with no structural heart disease who underwent AV(N)RT ablation, RMN has the most favorable long-term outcomes, in addition to favorable fluoroscopy and procedure times.

O53
SUDDEN CARDIAC ARREST AND ITS WARNING SIGNS AND SYMPTOMS IN A PEDIATRIC POPULATION

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Introduction: It's important to identify patients at risk for sudden cardiac arrest (SCA) since it's often lethal. The aim of the study was to investigate patients admitted to the pediatric cardiology ward for SCA or warning symptoms of SCA defined as ventricular tachycardia or syncope.
Methods: We retrospectively reviewed the charts of patients admitted since January 2006 to July 2020. A negative outcome was defined as death or relapse of SCA, syncope or ventricular tachycardia.
Results: We selected 78 patients, 45 males and 33 females, aged between 1 month and 27 years admitted for SCA (27%), syncope (34,5%) and ventricular tachycardia (38,5%). We defined pre work-up risk assessment with 5 criteria: positive family history for sudden cardiac death or channelopathies (12 patients;15%), known underlying cardiac disease (31 pts; 40%), experience of previous cardiac symptoms (25, 32%), ongoing antiarrhythmic treatment (16, 21%), exercise-related event (24, 31%). Most patients (66%) had 0 or 1 known risk factor. Discharge diagnosis were heterogeneous: ventricular tachycardia in otherwise healthy heart 16, failing congenital heart disease 16, reflex neurocardiogenic syncope 15, channelopathies 11, cardiomyopathies 7, non-cardiogenic event 5, acute myocarditis 4, bradyarrhythmia 3, rhabdomyoma 1. At follow-up there were 20 negative outcomes (2 deaths and 18 relapses), 4 patients were dismissed, 53 patients are still on follow-up with no relapse. Sex, age and the presence of an underlying cardiac disease did not impact on outcome at

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follow up. There was no significant association between a negative outcome and higher pre work-up risk. A history of antiarrhythmic drug use was a predictor for a negative outcome (p=0,0005). Patients with a negative outcome were more often admitted with a diagnosis or SCA or ventricular tachycardia than syncope (p<0,001) and required more often some non-pharmacological intervention (ECV, RFA, ICD or PM implantation) (p=0,0002). *Conclusions:* Comparing the risk assessment with the discharge diagnosis there was some degree of mismatch: in 9 patients (11,5%) the discharge diagnosis was less serious than the risk criteria would have suggested and in 9 patients (11,5%), despite a low pre work up risk, a serious condition was diagnosed. In 60 patients (77%) the risk assessment matched the post-test risk.

O54

The duration of antiarrhythmic medication for supraventricular tachycardia in infants can be shortened

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Introduction: Supraventricular tachycardia (SVT) is the most common arrhythmia in infancy. Prophylactic antiarrhythmic medication (AM) is used to prevent the recurrent SVT which may lead to heart failure in infants, if unrecognized. The choice and duration of AM varies, and lack of evidence has favoured longer treatment. We compared recurrence of SVT with shorter AM to standard prophylaxis of 12 months.
Methods: This multicentre cohort study included all infants with SVT aged less than 12 months from the five university hospitals between 2005 and 2017. The infants with SVT were divided into two groups according to the current national recommendation on secondary prevention. Those diagnosed between 2005 and 2012 had AM for 12 months (group 1) and those between 2013 and 2017 for 6 months (group 2). We used Kaplan-Meier analysis to express recurrence-free survival, and Cox regression model in multivariate analysis. All clinical factors related to risk for the recurrence of SVT and those with statistical differences in univariate analysis were included into multivariate analysis.
Results: A total of 277 infants had SVT (group 1 n = 181 and group 2 n = 96). Median duration of AM was 366 days (interquartile range, IQR 347.5-408.5) in group 1 and 217 days (IQR 182-308, $p < 0.001$) in group 2. Propranolol was the most common AM used (group 1 92% and group 2 95%, $p < 0.001$) and as monotherapy it was successful in over 50 % in both groups. Propranolol combined with flecainide or amiodarone were the most frequent combination therapies. There were no statistically significant differences between two groups in recurrence-free survival ($p = 0.536$), when age, sex, Wolff-Parkinson-White syndrome (WPW), antenatal arrhythmia and antenatal AM were included in Cox regression multivariate analysis for recurrence-free survival for SVT in the 2-year follow-up.
Conclusions: Shortening of prophylactic AM duration in infants with SVT from 12 months to 6 months does not lead to more frequent recurrence of SVT.

O55

The role of the electrocardiographic phenotype in risk stratification for sudden cardiac death in childhood hypertrophic cardiomyopathy

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Introduction: The 12-lead electrocardiogram (ECG) is routinely performed in children with hypertrophic cardiomyopathy (HCM). An ECG risk score has been suggested as a useful tool for risk stratification, but this has not been independently validated. This aim of this study was to describe the ECG phenotype of childhood HCM in a large, international, multi-centre cohort and investigate its role in risk prediction for arrhythmic events.
Methods: Data from 356 childhood HCM patients with a mean age of 10.1 years (± 4.5) were collected from a retrospective,

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multi-centre international cohort (International Paediatric Hypertrophic Cardiomyopathy Consortium).
Results: Three hundred and forty-seven (97.5%) patients had ECG abnormalities at baseline, most commonly repolarisation abnormalities (n= 277, 77.8%); left ventricular hypertrophy (n = 240, 67.6%); abnormal QRS axis (n=126, 35.4%); or QT prolongation (n=131, 36.8%). Over a median follow up of 3.9 years (IQR 2.0-7.7), 25 (7%) had an arrhythmic event, with an overall annual event rate of 1.38 (95% CI 0.93-2.04). No ECG variables were associated with 5-year arrhythmic event on univariable or multivariable analysis. The ECG risk score threshold of >5 had modest discriminatory ability (C-index 0.60 (95% CI 0.484-0.715)), with corresponding negative and positive predictive values of 6.7% and 96.9%.
Conclusions: In a large, international, multi-centre cohort of childhood HCM, ECG abnormalities were common and varied. No ECG characteristic, either in isolation or combined in the previously described ECG risk score, was associated with 5-year SCD risk. This suggests that the role of baseline ECG phenotype in improving risk stratification in childhood HCM is limited.

Cox regression analysis for arrhythmic outcome within 5 years

	Univariate Cox regression		Multivariate penalized regression
Variable	HR (95% CI)	p-value	Lasso estimates
Clinical risk factors			
Heart failure (NYHA >1)	0.62 (0.18-2.14)	0.446	
Family History of SCD	1.08 (0.25- 4.73)	0.918	
Unexplained syncope	2.33 (0.76 – 7.14)	0.140	
NSVT	2.71 (0.77-9.53)	0.120	
MWT (mm)	1.09 (1.03-1.15)	0.002	1.063
MWT z-score	1.07 (1.01 – 1.13)	0.002	
LA diameter (mm)	1.05 (1.00 – 1.10)	0.067	1.014
LA diameter z-score	1.03 (0.84 – 1.26)	0.788	
Maximal LVOT gradient (mmHg)	1.00 (0.99 – 1.02)	0.519	1.001
ECG risk factors			
Pathological Q-waves	0.65 (0.24 – 1.75)	0.394	
Giant inverted T-waves	0.58 (0.77 -4.39)	0.600	
Giant positive T-waves	1.60 (0.59 – 4.33)	0.354	
Pathological T-wave inversion-any lead	1.85 (0.65 – 5.256)	0.247	
Pathological T-wave inversion limb leads	2.42 (0.85 – 6.88)	0.097	
Pathological T-wave inversion precordial leads	1.51 (0.58 – 3.90)	0.400	
ST-segment depression >2mm	1.18 (0.67 – 2.07)	0.562	
ST elevation	2.06 (0.79 – 5.34)	0.138	
Dominant S wave in V4	1.46 (0.56 – 3.79)	0.436	
Limb-lead QRS-sum, mV	1.05 (0.97 – 1.14)	0.198	
Chest-lead QRS-sum, mV	1.00 (0.98 – 1.02)	0.950	
12-lead QRS-sum, mV	1.00 (0.99 – 1.01)	0.849	
12-lead product, mV	1.03 (0.94 – 1.13)	0.576	
Sokolow-Lyon, mm	1.00 (0.98-1.01)	0.939	
QTc >440 ms	1.51(0.48-3.91)	0.400	
Left bundle branch block	1.73 (0.23 – 13.10)	0.595	
Right bundle branch block	1.96 (0.25-14.87)	0.515	
Low QRS voltages	0.00 (0, ∞)	0.998	
Pseudonecrosis	0.89 (0.29 – 2.75)	0.846	
Pseudo-STEMI	2.37 (0.91 – 6.14)	0.076	
Total Risk score	1.11 (0.97 – 1.28)	0.114	
Risk score > 5	2.07 (0.77 – 5.60)	0.142	

2. Basic science, Genetics

O46
Cardiovascular outcomes in patients with Noonan syndrome enrolled in NordiNet® International Outcome Study (IOS) and ANSWER Program

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Introduction: Noonan syndrome (NS) is associated with short stature and cardiovascular (CV) abnormalities. Recombinant human growth hormone (rhGH) increases acceleration of height velocity and height standard deviation score (SDS), but has raised concern about the potential progression of ventricular hypertrophy. Although CV outcome data in patients with NS receiving rhGH are sparse, findings suggest that CV event rates are generally low, and that the left ventricular wall thickness remains normal. This analysis supplements this body of evidence with data from two non-interventional, multicentre studies: NordiNet® IOS (NCT00960128) and ANSWER Program (NCT01009905).
Methods: NordiNet® IOS and ANSWER evaluated long-term effectiveness and safety of Norditropin® (somatropin; Novo Nordisk A/S) as prescribed by physicians in a real-world clinical setting. CV outcomes were evaluated in rhGH-treated patients with NS (n=412) from the full analysis set.
Results: Baseline characteristics [% or mean (SD)] (all patients): female, 29.1%; age, 9.48 (3.92) years; height SDS, –2.65 (0.95); weight SDS, –2.03 (1.31); bone age/chronological age ratio, 0.83 (0.19); insulin-like growth factor-I (IGF-I) SDS, –1.13 (1.62); GH dose at baseline, 43.9 (13.7) µg/kg/day; GH-naïve at baseline, 68.4%; duration of treatment follow-up, 3.1 (2.6) years; GH dose during childhood, 46.6 (13.6) µg/kg/day. Genotype data were available for 61 patients from ANSWER. Variants (one patient could have >1 variant) were reported in: *PTPN11* (n=56), *RAF1* (n=5), *KRAS* (n=2), *SOS1* (n=2) and *SHOC2* (n=1). The most common CV comorbidities reported at baseline (Table 1) were pulmonary valve stenosis (PVS) and atrial septal defect. Twenty-one non-serious adverse reactions were reported in 15 patients; most common reactions: headache (six reactions in six patients) and arthralgia (five reactions in three patients). Two serious adverse reactions (brain neoplasm and metastases to spine) were observed in one patient. Five CV complications were reported following rhGH start: unspecified CV disease (three patients), PVS (one patient) and ruptured abdominal aortic aneurysm (one patient).
Conclusions: The data suggest that rhGH was well tolerated in patients with NS, with no new left ventricular hypertrophy reported over the duration of the study. Further prospective studies to systematically assess the effects of rhGH on the CV system in patients with NS are warranted.

Diagnosis	Baseline*	After GHT start
Aortic valve disorder (unspecified)	1	–
Atrial septal defect	4	–
Atrioventricular septal defect	1	–
Benign and innocent cardiac murmurs	1	–
Cardiac arrest with successful resuscitation	1	–
Cardiac disease	2	–
Cardiac murmur (unspecified)	3	–
Cardiomegaly	1	–
Cardiovascular disease (unspecified)	1	–
Cardiovascular disorder originating in the perinatal period (unspecified)	1	–
Coarctation of aorta	2	–
Congenital malformation of heart (unspecified)	3	–
Congenital pulmonary valve stenosis	6	–
Mitral valve disease (unspecified)	1	–
Other hypertrophic cardiomyopathy	3	–
Other pulmonary valve disorders	1	–
Pulmonary valve stenosis	12	1
Ruptured abdominal aortic aneurysm	–	1
Stenosis of pulmonary artery	1	–
Tetralogy of Fallot	1	–
Unspecified cardiovascular disease	–	3
Ventricular septal defect	2	–

*A patient can have more than one diagnosis in a given period.

GHT, growth hormone therapy

O47
Familial recurrence patterns in congenitally corrected transposition of the great arteries: an international study.

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Introduction: Congenitally corrected transposition of the great arteries (ccTGA) is a rare disease of unknown aetiology. We aimed to better understand familial recurrence pattern.
Methods: An international, multicentre, retrospective cohort study was conducted in 31 tertiary hospitals in 6 countries between 1990 and 2018, leading to investigate 1043 unrelated ccTGA probands.
Results: Atrioventricular block at diagnosis and laterality defects were observed in 35.4% and 29.9%, respectively. ccTGA associated with primary ciliary dyskinesia in 10 patients. Parental consanguinity was noted in 3.4% cases. A congenital heart defect was diagnosed in 81 relatives from 69 families, 58% of them being first-degree relatives, including 28 siblings. The most prevalent defects in relatives were dextro-transposition of the great arteries (d-TGA: 28.4%), laterality defects (13.6%) and ccTGA (11.1%); 36 new familial clusters were described, including 8 pedigrees with concordant familial aggregation of ccTGA, 19 pedigrees with familial co-segregation of ccTGA and d-TGA and 9 familial co-segregation of ccTGA and laterality defects. In one family there was co-segregation of ccTGA, d-TGA and heterotaxy syndrome in 3 distinct relatives.
Conclusions: ccTGA is not always a sporadic congenital heart defect. Familial clusters as well as evidence of an association between ccTGA, d-TGA, laterality defects and in some cases primary ciliary dyskinesia strongly suggest a common pathogenetic pathway involving laterality genes in the pathophysiology of ccTGA.

O48

Fetal autopsy after second trimester elective termination of pregnancy. A single center experience and correlation with prenatal imaging.

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Introduction: Background. Fetal autopsy rates are decreasing in Western countries, even though post-mortem examination gives important information about the detection of fetal malformations and possible correlation with clinical diagnosis.
The aim of this a prospective study was to carry out an evaluation of fetal autopsies in a single Veneto Region referral center during the second trimester elective termination of pregnancy (TOP) and to compare in vivo imaging and autopsy findings to evaluate the degree of agreement.
Methods: The study group consisted of 248 autopsies performed after the second trimester TOP from June 2013 to August 2018 at the Cardiovascular Pathology Unit of the University Hospital of Padua. Cases imaging and autopsy findings were available and compared in 157 (63%). Cases were classified into four

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categories according to the degree of agreement between prenatal imaging and pathology, as follow: I concordant, II partial concordant, III discrepant (the organ of malformation was concordant but not the defined malformation), IV discordant.
Results: Fifty-three fetuses were normal (53/248, 23,4%), 195 presented with malformations, gestation weeks (GW), ranging from 13 to23 GW, median of 22 GW, and maternal age between 17-47 years, median 34 years. Congenital heart disease (CHD) were detected in 73 out of 195 (37%), central nervous system malformation were present in 50/196 (25%) fetuses. Of 73 cases with CHD, 40 (55%) were isolated anomalies, the others were associated extracardiac other malformation. Of 73 cases of CHD, 29 (40%) were diagnosed with third level echocardiography. The table reported in detail the categories of concordance in different organs.
Conclusions: In our series, the most frequent malformation was represented by CHD 71 out of 195 (37%), at difference from the literature showing CNS was the organ most frequently involved. Higher rate of concordance was present in CHD compared to CNS malformations. Despite the decrease in the request for fetal autopsies, this study confirms the role of the autopsy as a complementary tool in reaching a detailed diagnosis of fetal malformations.

Affected Organs	Category I	Category II	Category III	Category IV	Tot
	N %	N %	N %	N %	
CHD	53 (73)	10 (14)	3 (4)	7 (9)	73
CNS	28 (56)	8 (16)	10 (20)	4 (8)	50
GU system	8 (40)	8 (40)	3 (15)	1 (5)	20
Others	-	7 (50)	2 (14)	5 (36)	14
Tot	89 (57)	33 (21)	18 (11)	17 (11)	157

O49
Genetic Variant Burden in Pediatric Myocarditis Predicts Outcome

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Introduction: Myocarditis is one of the most common causes leading to heart failure in children and a possible genetic background has been postulated. We sought to characterize the clinical and genetic characteristics in patients with myocarditis ≤18 years of age to predict outcome.
Methods: A cohort of 42 patients (MYCPEDIG) with biopsy-proven myocarditis underwent genetic testing with targeted panel sequencing of cardiomyopathy-associated genes. MYCPEDIG patients were divided into subgroups according to the phenotype of dilated cardiomyopathy (DCM) at presentation, resulting in 22 patients without DCM (MYC-NonDCM) and 20 patients with DCM (MYC-DCM).
Results: MYC-DCM patients (median age 1.4 years) were younger than MYC-NonDCM patients (median age 16.1 years; $p<0.001$) and event-free survival was lower ($p=0.003$). At least one pathogenic genetic variant was identified in 9/42 patients (22%), 8 of them were heterozygous, and 7/9 were in MYC-DCM. Variants were found in functional groups of genes typically involved in primary DCM. Rare variant enrichment analysis revealed significant accumulation of high impact disease variants in MYC-DCM versus healthy individuals ($p=0.002$). We then compared the MYCPEDIG subgroups with a cohort of patients with primary DCM. The number of variants were higher ($p=0.006$) and event-free survival was lower ($p=0.008$) in MYC-DCM patients compared to MYC-NonDCM and primary DCM.
Conclusions: We report heterozygous pathogenic genetic variants in biopsy-proven childhood myocarditis. Myocarditis patients with DCM phenotype were characterized by early-onset heart failure, significant enrichment of genetic variants, and poor outcome. These findings support genetic evaluation in children newly diagnosed with myocarditis and DCM phenotype.

O50
Heart transplanted children have major immunological differences regarding T-cells compared with kidney transplanted children

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Introduction: Our institution faced an increased number of post transplant lymphoproliferative lymphoma (PTLD) among heart transplanted children. As the immune system has a crucial influence on the susceptibility to develop lymphoma, we decided to investigate the immune system in heart transplanted children.
Methods: A prospective case control, cross section study for one year was performed. 36 children who had underwent heart transplantation were compared to 34 age- and sex-matched kidney transplanted children and 33 healthy age and sex-matched subjects. Subpopulations of lymphocytes in blood were determined by flow cytometry. The analysis included T-cells, and subtypes, B-cells,

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monocytes, recent thymic emigrants and T-cell receptor excision circles (TREC).
Results: Heart transplanted children had lower levels of TREC, T-cells, all subpopulations of naive T-cells, recent thymic emigrants, mononuclear cells, and B-cells than the healthy control group ($p < 0.05$) and kidney transplanted children ($p < 0.05$). The immunosuppressive regimen at kidney transplantation in children comprise the same drugs but at slightly lower doses compared to heart transplantation. However, kidney transplanted children had higher levels of T-cells, subpopulations of T-cells, naive cytotoxic T-cells, lymphocytes, mononuclear cells and B-cells compared to the healthy control group ($p < 0.05$). Within the T-cell population heart transplanted children had lower proportions of naive T-cells, and recent thymic emigrants but higher proportions of T-follicular helper cells and effector T-cells compared to the healthy control group and kidney transplanted children ($p < 0.05$)
Conclusions: Heart transplanted children had significantly lower T-cells, B-cells and TREC than the healthy control group and kidney transplanted children. In contrary, kidney transplanted children had higher levels of Tcells despite the immune suppression. Therefore, it is plausible that the immune dysfunction after heart transplantation has other explanations than the immune suppressive treatment.

O71

Human cardiac progenitors cell-seeded collagen patches improve right ventricular contractility: an innovative approach to treat right heart failure ?

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Introduction: Right ventricular (RV) failure is a major concern in grown-up congenital heart patients. Conventional treatments yield poor results calling for innovative approach to restore overloaded RV function
Methods: A repaired Tetralogy of Fallot pig model was used. After 5 months of a combined overloaded RV, a patch containing 10^7 human embryonic stem cell-derived NKX2.5+ cardiac progenitors was engineered and applied on the epicardium of whole RV free wall of 4 pigs (treated group). A control group (n=4) received a cell-free (3) or no patch (1). RV function was assessed by echocardiography using standard (indexed TAPSE, FAC, S' wave) and strain (free wall and global longitudinal) parameters before and 2 months after cell graft. The fate of progenitors was tracked using human and cardiac specific antibodies.
Results: 2 months after graft, indexed RV diameter significantly decreased in treated group whereas increased in control (from 27.2 to 22.2 mm/m and from 23.5 to 28.2 mm/m, p=0.038, respectively), telediastolic RV area and tricuspid diameter slightly decreased. The free wall thickness significantly increased in treated group *vs* control (from 7.8 to 12 mm vs from 7.9 to 8.3 mm, p<.001, respectively). In treated group, RV free wall contractility assessed by strain significantly improved *vs* control (respectively from -13.4 to -15.9% vs -18.9 to -12.9%, p<.001), the global longitudinal strain evolving in the same way. The global function was maintained in treated group while decreased in control. Differentiating TnT+ and actinin+ human cardiac progenitors were found in the patch and within the myocardium. Proliferating small Ki-67+ TnT+ differentiated fetal-like pig myocytes, as well as Nfkb+ and Oct4+ pig cells were observed. Transfection of *REL-A* (activated Nfkb) into neonatal mouse cardiomyocytes reprogrammed them into OCT4+ cells which then re-differentiated into cardiac myocytes.
Conclusions: Human cardiac progenitor while differentiating into cardiomyocytes degrade the extracellular matrix and invade the fibrosis. This likely induces inflammation. Inflammatory cells secrete ILs and Tnfa which activate Nfkb pathway in surrounding pig cardiomyocytes. This leads to cell reprogramming and re-differentiation into fetal like proliferating cardiomyocytes. Altogether, this contributes to maintain the RV adaptation to overload and to improve free wall contractility.

O72

Non-invasive imaging of the healing process of atrial septal defect percutaneous occluders: a proof of concept study

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Introduction: After percutaneous implantation of an atrial septal defect (ASD) occluder, a complex healing process leads to the device coverage within several months. However, an unexplained incomplete device coverage increases the risk of device related complications such as thrombosis or infectious endocarditis. The aim of the study was to non-invasively assess the device coverage process of ASD occluders in a chronic sheep model using micro-CT technology.
Methods: After percutaneous creation of an ASD, 8 ewes were implanted with a 16-mm Nit-Occlud ASD-R occluder (PFM medical, Germany) and were followed for 1 month (N=3) and 3 months (N =5) before sacrifice. After heart explant, the device coverage was assessed using micro-CT and was compared to histological analysis used as the gold standard for healing evaluation. The micro-CT raw image 3D reconstruction was performed and followed by a measurement of the coverage surface and thickness for each device.
Results: ASD creation and device closure was successful in 100% animals without complications. Following sacrifice, macroscopic assessment of devices showed that the coverage was complete for the left-side disk regardless of the duration of the follow-up and variable for the right-side disk, depending on the protrusion of this disk. 3D reconstruction from micro-CT raw images analysis allowed an accurate evaluation of device coverage of each disk and was overall well correlated to histology slices (Figure). Surface calculation from micro-CT images showed that the median surface of coverage was 93±8% for the left-side disk and 55±31% for the right-side disk. A mapping of the thickness was obtained, showing a coverage thicker in the center of the device.

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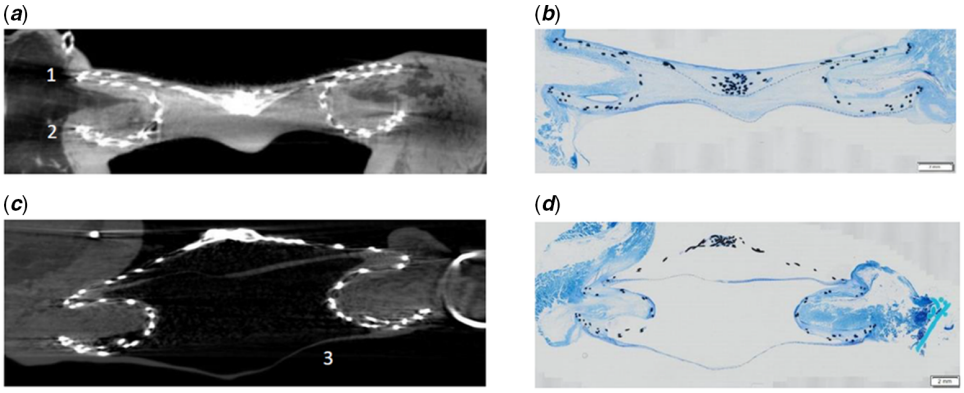

Figure: Micro-CT (A and C) and histology (B and D) Slices of 2 ASD occluders at 3 months follow-up. The upper disk iss the right-side disk (1) and the lower disk is the left-side one (2). The metal wores are white on the micro-CT images and black on the histology slices. The tissue Covvering the device appears in grey (3)

Conclusions: This preliminary study made the proof of concept that micro-CT is a reliable tool to assess the coverage of intra-cardiac occluders in vitro. The translation to clinical practice is challenging but would allow an individual follow-up to avoid thrombotic or infective complications.

O73
Proteomics in HCM in the young

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Introduction: Proteomic profiling in young hypertrophic cardiomyopathy (HCM) patients could lead to improved risk prediction and novel insights in the pathophysiological cardiovascular disease-progression.
Methods: Plasma levels of 184 cardiovascular disease-associated proteins were assessed by two proximity extension assays (Proseek Multiplex CVD-2 and CVD-3, Olink Bioscience, Uppsala, Sweden) in 30 young index-patients with familial HCM (23 male, 7 female, mean-age 19.8 years) from 23 unrelated families, and age and gender matched healthy controls (n=60). Data on echocardiography and electrocardiogram were available in all participants. Outcome data on cardiac arrests were recorded during 5 years follow-up.
Results: Three of the proteins were associated with an increased odds of HCM compared to healthy controls in Bonferroni corrected (p=0.00027) logistic regression models adjusting for age and sex. In order of significance, these included Selectin Glycoprotein Ligand 1 (PSGL-1, Odds ratio (OR) per standard deviation (SD) increase 7.87, 95 % CI (3.05-20.3), p=0.00002, Polymeric Immunoglobulin Receptor (PIgR, OR 3.39, 95 % CI (1.85-6.20) p =0.00007) and Paraoxonase (PON3, OR 0.30, 95 % CI (0.15-0.56) p=0 .0002). In addition, 50 of the proteins were nominally associated with HCM. In the patients with HCM, 7 had a history of cardiac arrest or suffered a cardiac arrest during follow-up. Higher levels of the proinflammatory PSGL and PIgR was associated with a higher, but non-significant, odds to suffer cardiac arrest (OR per SD increase 1.95, 95% CI (0.70-5.44), p=0.20, and 2.62; 95% CI (0.88-7.80), p=0.08, respectively). In contrast, higher levels of cardioprotective PON3 was associated with lower, but non-significant, odds for cardiac arrest (OR 0.44; 95% CI (0.16-1.20), p=0.11. In these secondary analyses a nominal p-value (p=0.05) was considered statistically significant.
Conclusions: Our proteomics approach identified 3 plasma proteins involved in neutrophil recruitment and migration (PSGL1), transmembrane-transport (PIgR), and cholesterol metabolism (PON3) that were closely associated with HCM compared to healthy controls. These 3 proteins also seemed to be suggestive prognostic markers for cardiac arrest, but the statistical power was low due to the small study sample and few events. Our findings encourage further large-scale proteomics studies in HCM patients in order to identify novel underlying mechanism and relevant risk markers.

O74
Relation between N-terminal pro B-type natriuretic peptide (NT-proBNP) and disease severity in paediatric hypertrophic cardiomyopathy

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Introduction: N-terminal pro B-type natriuretic peptide (NT-proBNP) is associated with increased risk of mortality and heart failure related adverse events in adults with hypertrophic cardiomyopathy (HCM). Elevated NT-proBNP levels have been correlated with multiple subjective and objective parameters of HCM severity including dyspnoea and left ventricular maximal wall thickness (LVMWT). However, robust prognostic markers in adults may not be reliable for children with HCM in whom disease severity assessment is challenging. No studies have yet evaluated utility of NT-proBNP in children with HCM. Thus, the objective

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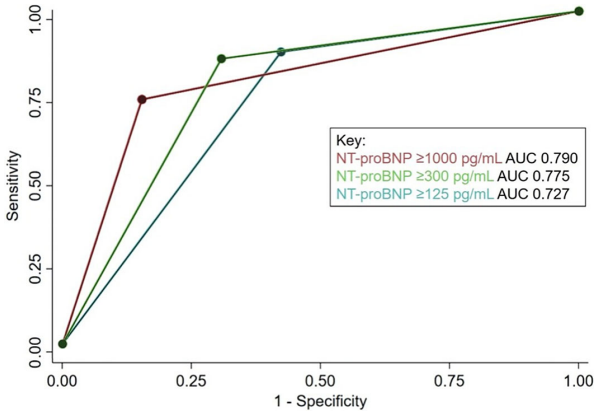
of this study was to assess associations of NT-proBNP with conventional markers of disease severity and predictive ability of NT-proBNP in a paediatric HCM cohort.
Methods: Plasma NT-proBNP levels were measured in eighty consecutive patients [23 (28.8%) females; median age: 12.3 years (interquartile range (IQR): 6.4-16.0); 37 (46.3%) sarcomeric aetiology]. Contemporaneous data from conventional clinical evaluation was used to establish disease severity including electrocardiography, echocardiography, tissue Doppler imaging, magnetic resonance imaging (MRI) and cardiopulmonary exercise testing.
Results: Median NT-proBNP concentration was 1104.5 pg/mL (range: 20–11206 pg/mL and IQR: 108.5–2613.5 pg/mL). NT-proBNP levels correlated with: QTc (ρ= 0.445, $p<0.01$); septal thickness z-score (ρ=0.618, $p<0.001$); MLVWT z-score (ρ=0.582, $p<0.001$); lateral S' (ρ=–0.668, $p<0.001$); septal E/E' (ρ=0.609, $p<0.001$); MRI MWT (ρ=0.773, $p<0.001$; indexed LV mass (ρ=0.576, $p<0.001$) and peak systolic blood pressure (ρ=–0.605, $p<0.001$). There were weak associations between NT-proBNP and aetiology or subjective symptoms including palpitations and chest pain ($p>0.05$). NT-proBNP levels were higher in patients who were: female; dyspnoeic (defined as Ross/NYHA Class ≥II); prescribed cardioactive medication and had an implantable cardioverter defibrillator ($p<0.05$). Lateral S' (β=–0.306, $p=0.001$) and MLVWT (β=0.217, $p=0.013$) were independent predictors of NT-proBNP in multivariate analysis. At a cut-off point of 300 pg/ml, NT-proBNP had a positive predictive value of 84% and a negative predictive value of 72% for predicting septal E/E'>10 (AUC=0.775 ($p<0.001$)) (See Figure 1).
Conclusions: NT-proBNP levels correlate with parameters of disease severity in paediatric HCM including measures of diastolic dysfunction (septal E/E') and systolic dysfunction (lateral S'). NT-proBNP measurement may be an effective adjunct for assessing disease severity in children, particularly when conventional clinical evaluation is challenging.



Receiver operator characteristic (ROC) curves of NT-proBNP cut-offs for predicting septal E/E'<10, a marker of diastoic dysfunction. In children with hypertrohic cardiomyopathy. True-positive rate (sensitivity), false positive rate (1-specificity) and ara uder the curve (AUC) are displayed for the following cuts-offs: NT-proBNP ≥1000 pg/ml (red), NT-roBNP ≥300 pg/ml (green) and NT-proBNP ≥125 pg/ml (blue).

O75

The Prdm16csp1/wt mouse mutant confirms a critical role of Prdm16 for heart function

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Introduction: Mutations in the gene transcriptional regulator PR domain containing 16 (*PRDM16*) were found to cause cardiomyopathy (CMP). However, the role of the PRDM16 protein in the mammalian heart has not been studied so far. In order to establish a murine model for the *PRDM16* associated CMP, we analyze heterozygous *Prdm16*$^{csp1/wt}$ mice, carrying the c.888-3C>A splice-acceptor site mutation in the *Prdm16* gene.
Methods: The impact of the c.888-3C>A mutation on splicing was tested with endpoint PCR and quantitative PCR (qPCR) using total RNA isolated from cardiac tissue. The cardiac phenotype was evaluated using echocardiography, electrocardiography and body composition analysis. The concentrations of natriuretic peptides were measured in plasma and heart tissue by qPCR and competitive ELISA. Furthermore, heart sections were stained for fibrosis with Picro-Sirius Red and tissue organization with H&E. Left ventricular ultrastructure was studied with transmission electron microscopy (TEM). The expression of selected sarcomere and mitochondrial genes was evaluated by qPCR and Western Blot.
Results: The c.888-3C>A mutation causes aberrant splicing of the *Prdm16* transcript possibly leading to reduced Prdm16 protein levels Heterozygous animals demonstrate with reduced body weights and heart hypotrophy. Moreover, *Prdm16*$^{csp1/wt}$ mice demonstrate a significant reduction in cardiac performance, as indicated by decreased stroke volume, ejection fraction and cardiac output. Interestingly, female mice present a more severe cardiac phenotype compared to their male littermates. In *Prdm16*$^{csp1/wt}$ mice, levels of the cardiac stress marker BNP and ANP were mildly elevated or unaffected, respectively. Histological analysis revealed normal cardiac tissue structure with absence of cardiac fibrosis. Ultrastructural analysis demonstrated in *Prdm16*$^{csp1/wt}$ heart tissue normal sarcomere structure and mildly elevated mitochondrial content. Expression analysis shows normal composition of the sarcomere, whereas selected markers of the mitochondrial electron transport chain (ETC) were diminished in females.
Conclusions: Heterozygous *Prdm16* deactivation in *Prdm16*$^{csp1/wt}$ mice results in significant reduction of cardiac performance and heart hypotrophy. Degradation of selected ETC proteins suggest a critical role for cardiomyocyte metabolism. Further analysis will explore the metabolic changes due to *Prdm16* deactivation.

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3. End-stage heart and lung disease

O21

Long-term survival of newly diagnosed children with pulmonary hypertension in the current treatment era. Results from the TOPP-1 registry

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Introduction: Pulmonary arterial hypertension (PAH) is a progressive, devastating pulmonary vascular disease, with a poor prognosis -especially in childhood- characterized by right ventricular failure and death. Although treatment strategies have evolved in the last 20 years, today the disease is still uncurable. Evaluation of outcome of PAH in patient cohorts is often obscured by the inclusion of prevalent patients, leading to survival bias. This study aims to describe outcome and risk factors in a large, worldwide cohort of pediatric patients, exclusively newly diagnosed with PAH.
Methods: This study, using data from the Tracking-Outcomes–in-Pediatric-Pulmonary-Hypertension (TOPP-1) registry, describes the occurrence of individual outcomes defined as death or (heart-)lung transplantation (LT). Children, > 3 months and < 18 years of age, newly diagnosed with PH between 2008-2015 were included (diagnosis confirmed by cardiac catheterization within 3 month of enrollment, incident patients). Patient characteristics, including functional class and hemodynamics, as well as outcome parameters death, transplantation and Potts shunt, were collected. Analyses included event rate calculations, KM curves and uni- and multivariate Cox regression analyses.
Results: In total, 269 children were included (90% with PAH) with a median follow up time of 2,6 years. Fortyfive children died (17%), 12 underwent LTx (4%), where 69% of these children (n=31) died within 1 year from diagnosis. Event rate for death/LTx was 9 per 100 patient years. Five years transplant/Potts free survival was 75%, but strongly depended on etiology. Younger age at diagnosis and male sex were independent risk factors for adverse outcome. Functional state and hemodynamic variables at diagnosis were associated with outcome with different weights over different subgroups of PAH (idiopathic PAH, PAH-associated congenital heart disease with either repaired or unrepaired shunts)
Conclusions: In the current treatment era, 5-years transplant/Potts free survival for children newly-diagnosed with PAH is 75%, with the highest event rate for death in the first year after diagnosis. Survival rate strongly depends on associated conditions: children with idiopathic PAH and those with PAH associated with no shunt or repaired shunt had significantly worse outcome compared to those with open shunts. Young age and male sex were independent risk factors for adverse outcome.

4. Fetal cardiology

O11

Can Z-scores from the normal fetal heart be applied to simple transposition of the great arteries?

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Introduction: Z-scores are an effective means of describing the size of the fetal great arteries in relation to a gestation-specific normal range. There is a paucity of data whether z-scores derived from the normally connected heart can be reliably applied to fetuses with transposition of the great arteries and intact ventricular septum (TGA). This is important for the assessment of associated lesions, for example, coarctation of the aorta. Our hypothesis was z scores of normal fetuses can be used for fetuses with TGA.
Methods: Fetal echocardiograms of all fetuses with isolated TGA between 2013 and 2018 were retrospectively reviewed. The arterial duct, distal transverse aortic arch (DTAA), aortic (AoV) and pulmonary valves (PV) were measured according to z-scores from >7000 fetuses recently published from our unit. Interobserver variability was assessed on 23 scans.
Results: One hundred and thirty-six fetal echocardiograms from 45 singleton fetuses with TGA with intact ventricular septum were reviewed. No additional findings were identified after birth. Median gestational age at birth was 38.4weeks (IQR:38-38.7) and mean birth weight was 3.227±0.366kg. The aortic valve and DTAA were significantly larger in fetuses with TGA than normal from 19-36 weeks ($p<0.0001$, $p<0.003$ respectively) and z-scores increased with advanced gestation ($p<0.0001$). The PV and ductal size were significantly smaller than normal at 19-23 weeks' gestation ($p<0.03$, $p<0.04$), but z-scores were significantly different with advanced gestational age ($p<0.0001$), increased beyond 34 weeks (Fig.1). At 34-36 weeks the AoV, DTAA, PV

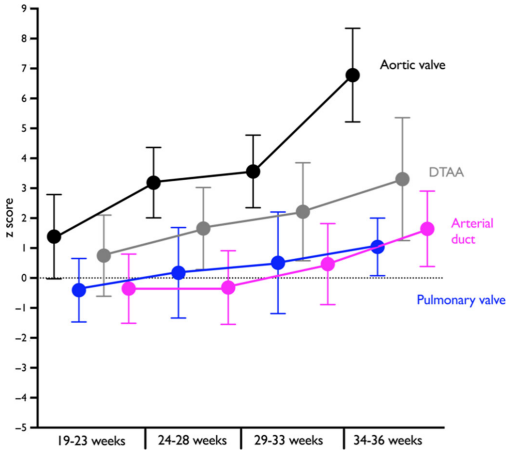

Fig. 1 Fetal z-scores of aortic and pulmonary valves, distal transverse aortic arch (DTAA) and arterial duct of 45 fetuses with isolated TGA

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and arterial duct were a mean of 2.9, 1.7, 0.8, and 0.8 mm larger than mean value in the normally connected heart. Interobserver correlation coefficient was good for all measures: AoV 0.87, DTAA 0.87, arterial duct 0.77, PV 0.69.
Conclusions: Outflow tract reference ranges for normal fetuses cannot be used to assess fetuses with TGA with the aortic valve and DTAA being the most different. This may reflect different loading conditions. TGA specific reference ranges and z scores would be most appropriate in affected fetuses.

O12
Congenital vascular rings: retrospective view from prenatal diagnosis to pediatric outcome in 61 cases

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Introduction: Anomalies of the aortic arch (vascular rings) are uncommon, presenting a broad spectrum of symptoms and causing significant tracheoesophageal compression in some cases. The aim of this study is to describe prenatal and postnatal characteristics, outcome and management of vascular rings in two tertiary centers.
Methods: Retrospective, descriptive, observational study including all cases diagnosed with vascular ring (double aortic arch (DAA) or right aortic arch (RAA) with left ductus arteriosus and/or aberrant left subclavian artery (ALSA), both without associated cardiac abnormalities) in two Fetal Cardiac Units from 2006 to 2020. Clinical records were reviewed for prenatal/postnatal sonographic features, genetic studies, symptoms, imaging studies performed, surgery and follow-up.
Results: Vascular ring was identified on 61 fetuses at a median gestational age of 22 weeks (93,4% were RAA and 6,6% DAA). Left ductus was found in all cases of RAA and an ALSA was seen in 57,4% of them. Prenatal genetic screening was performed on 57,3% of fetuses, being normal in all cases except for one 22q11.2 deletion syndrome (in whom legal interruption of pregnancy was performed). After a median follow-up of 5,2 years (SD 3), 53,3% of patients had developed symptoms, although only 18,3% of infants had symptoms before 6 months of age. Imaging studies were performed in 28 patients (46,6%), being normal in only 10%. Tracheal compression <50% was identified in 14/28 patients (half of them without any symptoms) and 4 patients had tracheal compression >50% (all were symptomatic). Regarding fibrobronchoscopy, it was performed in 11 patients (18,3%), finding compression in all cases: <50% in 7/11 cases and >50% in 4/11 cases. Spirometry was done in 10 patients, 4 were normal. Surgery was performed in 12 patients (20%), 10 of them with symptoms that resolved after the procedure.
Conclusions: Detection of vascular rings has increased in recent years. As there is no clear correlation between clinical and radiological findings, multidisciplinary management seems reasonable, performing imaging and/or respiratory studies, and maintaining follow-up throughout pediatric life.

O13
Growth of the pulmonary arteries in fetuses with tetralogy of Fallot – preliminary results

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Introduction: Postnatal treatment strategy in patients with tetralogy of Fallot (TOF) depends on the size of pulmonary valve and pulmonary arteries, source and net volume of pulmonary blood flow. Our aim was to examine the growth trends of pulmonary arteries in fetuses with TOF, to define their modifying variables and assess the impact on postnatal management.
Methods: We retrospectively reviewed echocardiographic studies of 116 fetuses diagnosed with TOF between 2011 and (2020), excluding cases with pulmonary atresia and/or multiple aorto-pulmonary collaterals. Measurements of the pulmonary valve (vPA), main and branch pulmonary arteries (MPA, RPA and LPA), together with the maximum velocity across vPA (Vmax-vPA) were taken from the first and last prenatal study. The presence and direction of flow through the ductus arteriosus (DA) was also recorded. Growth rates of vascular structures were expressed as the changes in absolute values (-a) and Z-scores (-z) indexed over time.
Results: Growth rates of measured structures are listed in table below. Growth of vPA was better in fetuses with right-to-left DA flow compared to these with left-to-right DA flow (p=0.01) or no duct present (NS). Growth rates of MPA, RPA, and LPA did not differ significantly between groups. In fetuses with right-to-left DA flow Z-scores of measured structures remain relatively stable throughout pregnancy. Among the possible predictors of the need of pulmonary flow augmentation the 1st month of life, statistically significant were: left-to-right or bidirectional DA flow, and smaller vPA diameter in the last fetal study.
Conclusions: Growth of the pulmonary arteries in fetuses with TOF is well preserved, probably due to relatively small volume of pulmonary blood flow throughout pregnancy. The direction of DA flow does not impact the growth of pulmonary arteries (contrary to vPA), however predicts the need of prostaglandin administration or intervention to augment the pulmonary blood flow in the newborn. These observations are clinically important, however further studies with higher numbers of patients are needed to support suggested trends and correlations.

	Parameter	All	R-L DA flow	L-R/bid. DA flow	No DA present
[mm/week]	vPA-a	0.19±0.14	0.21±0.10	0.12±0.21	0.18±0.15
	MPA-a	0.22±0.17	0.20±0.13	0.23±0.22	0.26±0.23
	RPA-a	0.12±0.12	0.12±0.11	0.11±0.13	0.16±0.12
	LPA-a	0.13±0.09	0.13±0.09	0.13±0.11	0.15±0.12
[Z-score/week]	vPA-z	0.00±0.25	0.01±0.20	-0.06±0.37	0.00±0.30
	MPA-z	0.03±0.28	0.00±0.16	0.04±0.37	0.11±0.38
	RPA-z	0.02±0.20	0.00±0.20	0.00±0.21	0.08±0.23
	LPA-z	0.03±0.18	0.02±0.17	0.03±0.19	0.05±0.21

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O14

Impact of prenatal diagnosis of coarctation on short and long term cardiovascular outcome

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Introduction: Prenatal diagnosis of congenital heart disease has become increasingly more frequent. We investigated the cardiovascular impact of prenatal diagnosis on pre-operative course and short-, mid-, and long-term follow up of surgically corrected neonatal coarctation.
Methods: We retrospectively reviewed 102 patients with isolated neonatal coarctation corrected surgically in our center between 1999 and 2019 with more than one year follow up and who maintained regular follow-up by our group. Cases of coarctation associated with any other significant heart disease were excluded. We collected data regarding presence of prenatal diagnosis, complicated pre-operative course (shock, need for ventilatory or inotropic support, multiorgan failure), prostaglandin infusion, age at surgery, post-operative complications, presence of acute renal injury, days at ICU, hospital stay, residual coarctation, persistent hypertension requiring medication at 1 and 6 months post-operative. Long term follow-up data included persistent hypertension, indexed left ventricular mass and need for reintervention were collected. A long-term negative composite outcome of persistent hypertension or Left Ventricular hypertrophy or need for reintervention was evaluated. Statistical analysis was performed with R v3.5.3. For binomial variables chi-squared tests were used, for continuous variables we used logistic regression tests.
Results: We identified 102 cases of simple neonatal coarctation. Of these 33 (32.3%) had prenatal diagnosis. In cases with prenatal diagnosis there was a statistically significant lower incidence of complicated neonatal course (3,0% vs 30,4%; p<0,01), and lower incidence of shock, multiorgan failure or need for ventilatory support (table 1). Age at surgery was significantly lower in the prenatal diagnosis group (5,1 days vs 14,9 days, p < 0.001). At mid- and long-term follow up, there were no significant differences between the groups with or without prenatal diagnosis, on rates of late reintervention, hypertension, or prevalence of LV hypertrophy (table 1), p = NS. Similar findings were seen for presence of neonatal complicated course, which did not influence the occurrence of any of the aforementioned late complications.
Conclusions: Despite playing a significant role in minimizing the severity of the neonatal presentation, prenatal diagnosis of coarctation or a less severe neonatal course did not influence mid- and long-term cardiovascular outcomes of neonates with coarctation.

		With prenatal diagnosis (%)	Without prenatal diagnosis (%)	p-value
Pre and early postoperative	Complicated neonatal course	1 (3,0%)	21 (30,4%)	**0.004**
	Shock	1 (3,0%)	21 (30,4%)	**0.004**
	Multiorgan failure	1 (3,0%)	14 (20,3%)	**0.045**
	Pre-operative ventilation	1 (3,0%)	14 (20,3%)	**0.045**
	PGE1 infusion	28 (84,8%)	46 (66,7%)	0.091
	Post-op AKI (KDIGO 2 or 3)	3 (9,1%)	5 (7,2%)	1
	Post-operative complications (≥3)	2 (6,1%)	9 (13,0%)	0.47
	Prolonged ICU stay (≥3 days)	10 (30,3%)	32 (48,4%)	0.10
Mid-term follow-up	Late hypertension (at 6 months post-operative)	17 (53,1%)	42 (64,6%)	0.385
	Reintervention (<1y)	1 (3,0%)	1 (1,4%)	1
Long-term follow-up	Persistent late hypertension	8 (25,8%)	8 (13,3%)	0.234
	Need for reintervention (cumulative)	8 (24,2%)	17 (24,6%)	1
	LV hypertrophy	2 (6,0%)	5 (7,2%)	1

O15

Impact of regional teaching programme and introduction of three vessels and tracheal view on prenatal detection of major congenital heart disease

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Introduction: Prenatal cardiac diagnosis allows opportunity for comprehensive counselling and can result in improved long –term outcomes of newborns with Congenital Heart disease (CHD)
*Aim:*To compare prenatal detection rates of cardiac anomalies in the region during prenatal screening based on the five FASP views. We aimed to compare detection rates between 2009-2013 and 2013-2019 investigating trends and variation within the region.
Methods: We retrospectively reviewed the data in two cohorts of patients who needed cardiac interventions (either cardiac catheter or surgery) in their first year of life at Birmingham Children's Hospital. Cohort A included infants born between 1/1/2009 until 30/6/2014 and cohort B included patients who were born between 1/1/2014 to 31/12/2019. We then compared the results both temporally and geographically. Patients with patent ductus arteriosus, isolated atrial septal defects, acquired heart disease and arrhythmias were excluded.
Results: 2077 patients were included (cohort A n=1039, cohort B n=1038). Overall antenatal diagnosis rate was 27.91% (n=290) in cohort A and 43.73% (n=454) in cohort B. Antenatal detection rates for specific lesions shown in table 1. The regional antenatal detection rates varied from 30% to 60% although in smaller hospitals the overall numbers were small affecting results. Single ventricle detection rates in cohort B varied between 78-93% in regions that had more than 3 cases. VSD, AS and PS were the lowest antenatal detection rates in cohort B (24.71%, 15.55% and 18.84% respectively) Detection rate of cardiac abnormalities with an abnormal 3VT view were significantly increased in the later era. Vascular ring data was only collected for cohort B with an antenatal detection rate of 74.2%. The target for >90% antenatal detection of TGA with intact ventricular septum and HLHS was reached by 2019.
Conclusions: There has been a significant improvement in prenatal detection of major congenital heart disease in our region. This is likely due to the combination of adding the 3VT view to the anomaly scan (2015) but also due to an intense educational programme provided by the Tiny Tickers Charity (2014). Further study is needed to assess differences in morbidity and mortality outcomes.

Lesion	Fetal echocardiographic view	Cohort A	Cohort B	Significant difference p value<0.05
Single ventricle	4 chamber view	Total 82 AN 65 (79.3%)	Total 198 AN 164 (83.8%)	P=0.47
Atrioventricular septal defect (AVSD)	4 chamber view	Total 74 AN30 (40.5%)	Total 103 AN 69 (67%)	**P=0.00012**
Ventricular septal defect (VSD)	4 chamber view and outlet view	Total 139 AN 10 (7.2%)	Total 178 AN 44 (24.7%)	**P=0.00052**
Aortic stenosis (AS)	Outlet view	Total 30 AN 6 (20%)	Total 45 AN 7 (15.6%)	P=0.45
Pulmonary atresia/stenosis (PA/PS)	Outlet view and 3 vessel view	Total 69 AN 13 (18.8%)	Total 70 AN 13 (18.6%)	P=0.85
Conotruncal defects	Outlet view and 3 vessel view	Total 161 AN 46 (28.6%)	Total 144 AN 79 (54.9%)	**P=0.0002**
Transposition of the great arteries (TGA)	Outlet view and 3 vessel view	Total 91 AN 25 (27.5%)	Total 105 AN 76 (72.4%)	**P<0.0001**
Aortic arch lesions	Arch view and 3 vessel view	Total 52 AN 7 (13.5%)	Total 112 AN 41 (36.6%)	**P<0.00001**

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O81

Left Ventricular Isovolumetric Relaxation Time improves detection of fetal Long QT Syndrome

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Introduction: Detection of fetal Long-QT syndrome (fLQTS) is challenging, but LQTS causes significant fetal and neonatal mortality. A fetal heart rate(FHR) <3rd percentile for gestational age(GA) is specific for diagnosing fLQTS, but has low sensitivity, (<50%). A retrospective analysis demonstrated a prolonged normalized left ventricular isovolumetric relaxation time (N-LVIRT) in fLQTS[1]. We aimed to prospectively evaluate the diagnostic accuracy of N-LVIRT for fLQTS.
Methods: Multicenter prospective case series of fetuses with a possible diagnosis of fLQTS. Serial fetal echocardiograms were performed/subject. LVIRT, cycle length (CL) and FHR were measured using pulsed Doppler over the mitral and aortic valves. Cut-offs for N-LVIRT (LVIRT/CL as %) were determined for GA periods: ≤20 weeks; 21-30 weeks; ≥31weeks. One measurement/fetus/GA was assessed based on previously published data.[1]
Results: (See Table) 53 fetuses with 135 echocardiograms were included (19 with familial/de novo LQTS (Group1), 18 familial LQTS negative controls (Group2), and 16 with other diagnoses (Group3). The median (range) FHR (beats/min for Groups1, 2, and 3 were 130 (69-162), 140 (125-170) and 122 (81-170) respectively. The sensitivity and specificity of N-LVIRT for fLQTS overall were, 73,3% and 74,2% respectively, and the postive predictive value was 57,9%, with a negative predictive value of 85,2%.

The N-LVIRT was 100% sensitive for LQTS ≤20 weeks. The N-LVIRT was 90% specific for LQTS ≥31 weeks. All false negatives were cases of familial LQTS, the majority with a known mild phenotype. De-novo LQTS (n=2) and symptomatic cases were correctly identified.
Conclusions: In this cohort N-LVIRT had a better sensitivity for fLQTS than FHR alone.

N-LVIRT may be helpful in the excluding and detection of fLQTS, especially when the phenotype is severe.

1) Clur et al. 2018

	True Negatives	True Positives	False Negatives	False Positives	Total	Sensitivy (%)	Specificity (%)	Postive Predictive Value (%)	Negative Predictive Value (%)
≤20 weeks	10	7	0	7	24	100	58,8	50	100
21-30 weeks,	18	10	2	7	37	83,3	72	58,8	90
≥30 weeks	18	5	6	2	31	45,5	90	71,4	75
TOTAL	46	22	8	16	92	73,3	74,2	57,9	85,4

O82

Persistent left superior vena cava does not predict postnatally confirmed coarctation of the aorta

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Introduction: Persistent left superior vena cava (PLSVC) is a common form of anomalous systemic venous return observed in 0.3% of the population with structurally normal hearts and in 4–8% of patients with congenital heart disease; there is a particular association with Coarctation of the aorta (CoA). Since the 3 vessel view was included in the screening programme in the UK, PLSVC is now more frequently diagnosed antenatally. We therefore aimed to investigate the relationship between foetal suspicion of CoA and antenatally diagnosed PLSVC.
Methods: All neonates with prenatally suspected CoA were identified between 1st January 2016 to 1st October 2020; antenatal and postnatal electronic medical records of 136 neonates (after exclusion) were then reviewed. Chi-square analysis was used to determine if the observed frequencies were significant.
Results: Of the 136 neonates with antenatal suspicion of CoA, 28 (20%) also had antenatal diagnosis of PLSVC. Postnatally, 53 (39%) neonates had confirmed CoA. Of these, 12/53 (22%) also had PLSVC compared to 16/83 (19%) in the group where CoA was postnatally excluded. A chi-square test of independence showed that there was no significant association between postnatal confirmed diagnosis of CoA and antenatally suspected PLSVC, X^2 (1, N = 136) = 0.22, p = 0.64.
Conclusions: Antenatally diagnosed PLSVC was not more common in postnatally confirmed CoA and is therefore not a useful predictor of postnatal confirmation of antenatally suspected CoA. Interestingly, the incidence was higher than expected with 10% expected in the postnatally confirmed CoA group, and 0.3% in the postnatally disproved CoA (normal) population. PLSVC leads to an enlarged coronary sinus which can cause mild left ventricular inflow restriction in the foetus. This can result in mild ventricular disproportion, which then raises the suspicion of discrete CoA.

O83

Postnatal management and outcome after fetal aortic valvuloplasty

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Introduction: Critical aortic stenosis (CAS) may lead to a hypoplastic left heart syndrome (HLHS) after birth. Fetal aortic valvuloplasty (FAV) is performed to improve chances for a biventricular repair which is also dependent on postnatal treatment strategies. The aim of this study is to assess postnatal treatments and overall survival after successful fetal aortic valvuloplasty.
Methods: The local fetal cardiac intervention (FCI) database was retrospectively reviewed for all fetuses who underwent FAV in our center since 2001. Patient's charts were analyzed for postnatal management and outcome.
Results: Since 2001 a total of 128 FAVs was performed in 102 patients. There were 15 IUDs, 11 procedure related. Six patients were liveborn after unsuccessful FAV and all became univentricular. There were 4 neonatal deaths before treatment. 77 neonates underwent further management at 12 different European centers: 17 underwent univentricular (UV) repair (3 died), 2 had hybrid (one died) and 58 underwent a biventricular (BV) repair. In the BV group 15 patients were initially managed with aortic balloon valvuloplasty (5 died), 3 underwent surgical valvotomy (1 died), 32 underwent Ross-Konno surgery (5 died), 3 had different surgical therapies (2 died) and 5 patients didn't need postnatal treatment (all alive). Four patients in the BV group underwent UV conversion and all died. Freedom from mortality including the two patients with hybrid repair was as follows (BV vs. UV): 28d: 95% vs. 100%; 1 year: 76.9% vs. 78.7%; 3 years: 76.9% vs. 67.4%.

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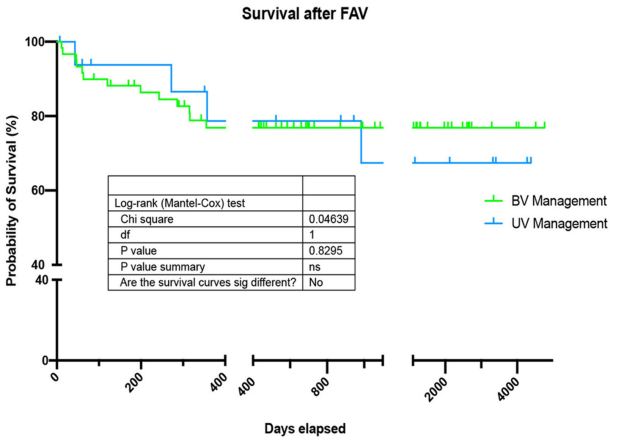
Freedom from mortality was not significantly different between the two groups (P = 0.8295). Follow up durations were similar in both groups (median follow up 1.70 years UV vs. 1.84 years BV).
Conclusions: Patients after successful FAV still carry significant risk of mortality regardless of postnatal management. Mortality was not different between the UV and the BV group. Most patients received an aortic valve replacement with an acceptable mortality. Overall, there were differences in postnatal management strategies between centers.



O84
The Innovative Use of HoloLens2 in the fetal cardiac servcies during COVID-19 pandemic

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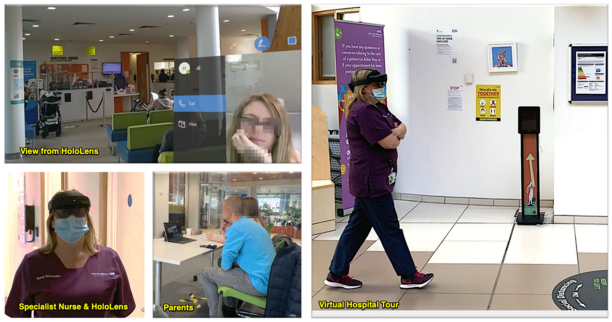
Introduction: COVID-19 prevent expectant parents of babies with congenital heart disease (CHD) from visiting cardiac surgical units prior to the birth of their baby who may be separated early for treatment. Pre-recorded video-clips or telephony consultation are generic and cannot deliver individual needs, especially in complex care pathways. We demonstrate our pilot experience using HoloLens 2 (Microsoft, Washington, USA) to deliver bespoke one-to-one virtual tour of the hospital and acute cardiac service environment.
Methods: Families with antenatal diagnosis of significant CHD were offered the virtual tour in their third trimester. Parents and Fetal Nurse Specialists communicate via Microsoft Teams (Microsoft 365, Washington, USA) on portable electronic devices. HoloLens was deployed by the Nurse Specialists to provide one-to-one audio-visual experience, with opportunity to interact and raise questions. All parents participated remotely from the hospital site. The first 5 sets of parents had company of a second Nurse Specialist during the tour, in case of adverse psychological reaction. Due to zero negative event, the presence of the 2nd Nurse was optional subsequently. All were asked to complete a questionnaire.
Results: Over the 3-month period, 25 sets of parents participated in the quality improvement survey. Respond rate was 96%. 85% said the virtual visit met or exceeded their expectations for preparedness, 80% said they liked that it allows them to 'visit' the hospital without physical attendance and associated costs of childcare/leave from work/travel time, 80% said it is a great use of technology, 70% liked that they were able to interact with the nurse specialist during the visit, 80% would recommend a virtual visit to other families and only 10% said they would have preferred to visit the hospital in person.
Conclusions: The innovative use of HoloLens in cardiac fetal services is proven useful for the family, enabling preparedness for the arrival and early separation of their babies. Its deployment met the needs to continue high quality service in critical time. Whilst not intended to replace physical consultation; however, this technology proves to be an effective option for parents once the pandemic has passed.



O85
FETAL HETEROTAXY: SHOULD WE STILL CATEGORIZE?

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Introduction: Heterotaxy is usually stratified in right isomerism (RI) and left isomerism (LI), classically associated with abdominal, cardiac and venous abnormalities.
Methods: We analyzed 66 fetal specimens with heterotaxy divided in 5 groups : classic LI (CLI: left bronchopulmonary isomerism, bilaterally absent pectinate muscles, polysplenia, n=23), non-classic LI (NCLI: one discordant feature, n=16), classic RI (CRI: right bronchopulmonary isomerism, bilateral extent of pectinate muscles extent to the crux, asplenia, n=12), non-classic RI (NCRI: one discordant feature, n=7), totally discordant features (n=8).
Results: Non-classic patterns were found in 47% (37% RI, 41% LI). Among non-classic patterns, the status of the spleen was highly variable (NCLI with asplenia 38%, NCRI with single spleen 83%), while the bronchial status was almost always concordant. Status of liver, gallbladder, stomach was highly variable in all categories. Intestinal malrotation was present in 83% CRI, 65% CLI, 44% NCLI, 43% NCRI. Extent of pectinate muscles was highly variable in NCLI (bilaterally left 56%, normal 38%, bilaterally right 6%), and was bilaterally right in 86% of NCRI only. Interrupted inferior caval vein was seen in LI only; total extracardiac anomalous pulmonary venous connection was seen in RI only, ipsilateral pulmonary venous connection in LI only. A common atrioventricular (AV) junction was constant in RI vs 64% in LI, with a complete AV canal in 100% RI, 56% LI. Functionally univentricular hearts were found in 68% RI, 44% LI, predominantly right in all groups (65%). Ventriculo-arterial connections were abnormal in 100% RI, 39% LI. Double outlet right ventricle was the most frequent type of abnormal VA connections in all categories. Pulmonary atresia and stenosis were frequent in RI (68%), rare in LI. Aortic arch hypoplasia and coarctation were found almost exclusively in LI (30% LI, 5% RI). Right-sided aortic arch was found in 63% RI, 11% LI.

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Conclusions: Heterotaxy and isomerism are not synonymous. Extent of pectinate muscles is not always symmetrical (14%) and cannot therefore be considered the hallmark of heterotaxy. Because heterotaxy includes both abnormal symmetry and random organization of organs, each anatomic feature should be analysed individually rather than to categorize patients in right and left isomerism.

5. Adult congenital heart disease

O6
Adults with congenitally corrected transposition of the great arteries are burdened with significant morbidity and mortality

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Introduction: Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital heart disease (CHD). Contemporary data regarding the outcome in adults with ccTGA is scarce. The aim of this study was to describe a contemporary group of adults with ccTGA and evaluate predictors of outcome.
Methods: In this retrospective, single center analysis all patients with the diagnosis of ccTGA and an age over 16 years, who were treated at our hospital during the time period 2006-2018 were included. Time point of inclusion was the first appointment during the study period, if the patient was already 16 years of age or older or the first appointment after the 16th birthday, if the pat. reached this age threshold during the study period. Clinical data, including survival status, morbidities, hospitalisations, need of operations or interventions, as well as need for pacemaker, were retrieved from hospital records. The primary endpoint was death from any cause.
Results: Altogether, 100 patients were included, 96 with a systemic right ventricle, 3 with anatomical repair und 1 palliated with aorto-pulmonary shunt. Mean age at baseline was 32.3 ± 15.8 years, while median follow up was 6.5 years (IQR 2.6-12.8). There were 11 deaths, 10 due to cardiac causes, 1 due to suicide. Three patients underwent heart transplantation, two of them died. During the follow-up, 67 patients were hospitalized, 52 due to heart failure. On univariate Cox analysis, age at baseline (hazard ratio (HR): 1.043, CI 95%: 1.002-1.085, p=0.039), tricuspid regurgitation (HR: 2.652, CI 95%: 1.548-4.542, p<0.001), systemic ventricular function (HR: 3.486, CI 95%: 1.658-7.327, p=0.001), NYHA class (HR: 4.566, CI 95%: 2.194-9.504, p<0.001), and presence of sinus rhythm (HR: 0.252, CI 95%: 0.073-0.872, p=0.030) were predictors of the primary endpoint. On multivariable analysis, only tricuspid regurgitation (HR: 2.324, CI 95%: 1.128-4.789, p=0.022), and NYHA class (HR: 3.052, CI 95%: 1.459-6.382, p=0.003) remained as independent predictors.
Conclusions: Even young adults with ccTGA are burdened with significant morbidity and mortality. Predictors for death are the severity of tricuspid regurgitation and functional class. Close surveillance is mandatory.

O7
Cardiac MRI study on the coronary arteries (CA) late after neonatal arterial switch operation (ASO) for transposition of the great arteries (TGA)

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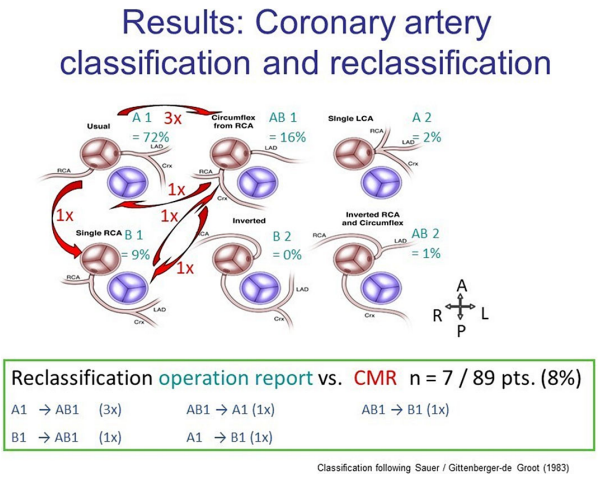
Introduction: Long-term follow-up after ASO can be complicated by myocardial ischemia due to stenosis, kinking or stretching of the transferred CA.
Methods: In a monocentric prospective study, 92 unselected patients (age 18-29, mean 23 y) underwent assessment of insertion/proximal course of the CA by coronary MR-angiography (MRA) as well as rate/extent of stress induced myocardial ischemia by dobutamine stress MRI (DSMR). 89/92 (97%) underwent CMR at rest (coronary MRA, cine), 85/92 (92%) DSMR (cine, perfusion); followed by late gadolinium enhancement (LGE).
Results: Coronary MRA: 87/89 (98%) presented without stenosis/kinking of the proximal CA. 2/89 (2%) were pathologic: 1x known prox. LAD occlusion (collateralized via RCA; coronary type AB1); 1x single right coronary ostium (reclassified from AB1→B1). CA classification: 7 pts. (8%) were reclassified compared to the operation report (Image). LGE: no ischemic scar, 1x intramyocardial fibrosis. DSMR: 79/85 (93%) reached target heart rate; 4x premature termination. 3/85 (3.5%) DSMR pathologic: 1 pat. with known prox. LAD occlusion (AB 1 with functionally single ostium, collateralized via RCA) had chest pain and hypokinesia under max. stress. 1 pat with single right coronary ostium (reclassified AB1→B1) had complex VES and stress induced ischemia during DSMR, chest pain during school sports → invasive CA angiography without stenosis. 1 pat had strong chest pain during DSMR due to dobutamine induced coronary spasm (A1, uneventful medical history) invasive CA angiography not pathologic.
Conclusions: CMR with coronary angiography is feasible to assess proximal courses of the transferred CA noninvasively and without ionizing radiation in grownups after neonatal ASO. In addition to other non-invasive diagnostic procedures, we suggest to routinely perform CMR at age 14-16 y in all patients after neonatal ASO. DSMR represents the best possible form of simulation of a physiologic adrenergic exercise with increased cardiac workload and movement of the CA. Due to the high incidence of normal results (96,5%), routine DSMR in asymptomatic young adults after neonatal ASO does not seem necessary. In case of abnormal coronary status or complications, pathologic anamnestic events or routine diagnostics and before starting with competitive sports, DSMR should be considered as a safe and meaningful additional diagnostic method.



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O8

Fontan Associated Liver Disease (FALD) Service in the North West of England

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Introduction: The Fontan procedure is a complex open-heart surgery associated with FALD. There is no agreed standard in how best to assess the severity of liver damage (fibrosis stage) in these patients at neither a national nor international level. Furthermore, scoring systems used in other liver diseases have not been validated in FALD. We performed a retrospective analysis of our FALD cohort to establish what proportion of patients would be classified as having cirrhosis F4. By risk stratifying in this way, we aim to inform service delivery, development, and thus surveillance strategies for FALD.
Methods: In the North West, there are approximately 200 Fontan patients, of which 71 have been referred to the Hepatology FALD service. We collected data retrospectively on these 71 patients and classified liver cirrhosis based on the following: platelet count (< 150 x 109/L), AST/ALT ratio (> 1.0), AST to Platelet Ratio Index (APRI) (≥ 1.0), Enhanced Liver Fibrosis (ELF) score > 9.8 and FibroScan ≥ 11.5 kPa.
Results: 71 patients were referred to the FALD service. All completed initial sets of data are presented. 32 (45%) were male and 39 (55%) female. The mean (±SD) age was 29 (±8) years. The mean (±SD) year of Fontan procedure was 1998 (±8), with the most common type being the total cavopulmonary connection (TCPC) procedure. 20 (37%) had platelets < 150 x 10^9/L (n=54). 28 (56%) had an AST/ALT ratio > 1.0 (n=50). 2 (4%) had an APRI score ≥ 1.0 (n=49). 11 (31%) had an ELF score > 9.8 (n=36). 38 (90%) had a Fibroscan stiffness ≥ 11.5kPa (n=42). 24 patients had data for all five parameters: no patient fulfilled all parameters; two fulfilled four parameters and 11 fulfilled three parameters.
Conclusions: With an increasing number of Fontan patients surviving longer, the proportion of patients with FALD is increasing. Studies demonstrate that conventional models of non-invasive assessment are unreliable in this cohort, and new thresholds or models are needed. This will enable correct risk stratification of patients, allowing screening for advanced liver disease, its complications (varices, hepatocellular carcinoma) and optimal timing of referral for combined heart and liver transplant.

O9

Rest and exercise hemodynamic characteristics of bicuspid and tricuspid valves after valve sparing aortic root replacement.

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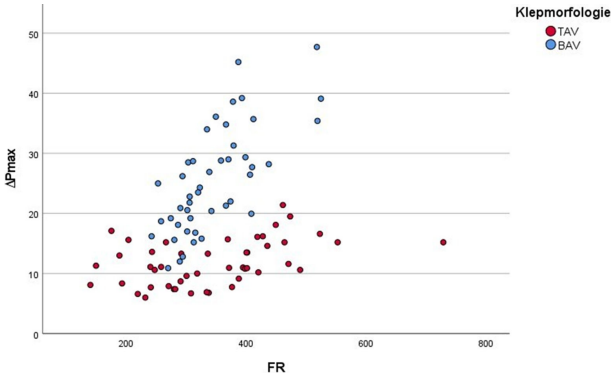
Introduction: Valve sparing root replacement (VSRR) has become the treatment of choice in patients with aortic root aneurysm and non-calcified aortic valves, demonstrating stable valve function and few long-term complications compared to valve replacement. Only a few studies however assessed the haemodynamics of the preserved aortic valve during exercise, and the differences between bicuspid (BAV) and tricuspid aortic valves (TAV).
Methods: Sixteen patients (TAV, n=9; BAV, n=7) that previously underwent a VSRR with reimplantation were investigated with transthoracic echocardiography in a reclining bicycle exercise protocol, starting at 45W, with 10W incremental steps every 2 minutes. Heart rate and blood pressure were continuously measured. Maximum (ΔPmax) and mean (ΔPmean) gradients across the aortic valve, velocity time integrals and ejection time in the left ventricular outflow tract were obtained by continuous-wave and pulsed-wave Doppler at rest and at each increment in effort. Aortic valve area (AVA) was calculated. Mean age of the patients was 50±9 years, median time after VSRR was 3(2-4) years. Exclusion criteria were ΔPmax at rest of ≥20mmHg, aortic incompetence ≥ 2/4, depressed left ventricle, and being physically unfit to perform exercise.
Results: With increasing exercise the ΔPmax in TAV increased from 7±2mmHg at rest to 18±2mmHg at maximal exercise, and in BAV from 18±6mmHg to 41±10mmHg, significantly different between the 2 valve types at each step-up. AVA increased from 2.9±0.9cm^2 to 3.6±0.2cm^2 in TAV, and from 2.2±0.4cm^2 to 2.5±0.6cm^2 in BAV patients, not significant between valve types. However, when transvalvular gradients and AVA were related to flow rate, which is considered a better representation of cardiac effort, ΔPmax and ΔPmean were significantly higher (figure), and AVA significantly lower in BAV compared to TAV at each flow rate increase. In multivariable analysis, BAV valve type (p=0.001) and increasing flow rate (p=0.002), were independent predictors of higher ΔPmax.
Conclusions: In this echocardiographic study, graded exercise led to a significantly higher increase in ΔPmax and ΔPmean after VSRR in BAV compared to TAV. In BAV patients a reduced valve opening was demontrated during exercise. The clinical relevance of these findings in view of premature BAV failure after VSRR has to be evaluated in long-term follow-up.



O10

Transition care for congenital heart disease in the COVID-19: Adaptations & improvisations in a supra regional CHD service in the United Kingdom

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Introduction: The COVID-19 pandemic resulted in prioritisation of healthcare resources to cope with the surge in infected patients,

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leading to suspension of routine clinical services, including Transition Care Services (TCS). In these unprecendented times, our TCS decided to adapt and improvise so that we could continue with the transition process. We present our experience of the last few months of COVID-19.

Methods: The TCS is well established across the North West, North Wales & Isle of Man Congenital Heart Disease Network with the 2 weekly Transition Clinics in two major Children's Hospitals in Liverpool & Manchester. The team consists of 1 ACHD cardiologist, 3 paediatric cardiologists and 6 clinical specialist nurses. With the sudden shut down there was the expected pressure on the wait lists and with no clear end in sight, our Network Transition Service decided to go "fully" virtual as soon as we could set up this platform.

We improvised 2 different models:

1. Initially we established "Fully Virtual Clinics" on the NHS virtual platform "Attend Anywhere", whereby the patient & family, the adult team and the paediatric team could "log-in" and conduct a virtual clinic. Each clinic had an initial 30 minutes of "Team Huddle" to review patient data. Based on the status and recent investigations, future-plans were made.
2. Once guidelines were eased, we moved to a "Hybrid Clinic" model, whereby the patient would attend the children's hospital, have investigations and be seen by the paediatric team. The adult team would remotely log in, on "Attend Anywhere".

Results: Over eighteen-week period during the pandemic, 106 patients were booked in for initially full virtual & subsequently hybrid clinics. 81 attended their appointments. 17 did not attend and 8 cancelled their appointments.

Conclusions: These models proved a big success, with good feedback from patients/families. Virtual clinics were particularly popular with youngsters. It took away the need to travel, helped maintaining social-distancing and reduced the risk of COVID-19 in this vulnerable group. To our knowledge, there have been no studies in the UK looking at the effect of the COVID-19 on the provision of TCS. This experience has been critical for us to provide our TCS in the future.

6. Imaging/functional assessment

O25

Epicardial fat volume assessed by MRI in adolescents: Associations with obesity and cardiovascular risk factors

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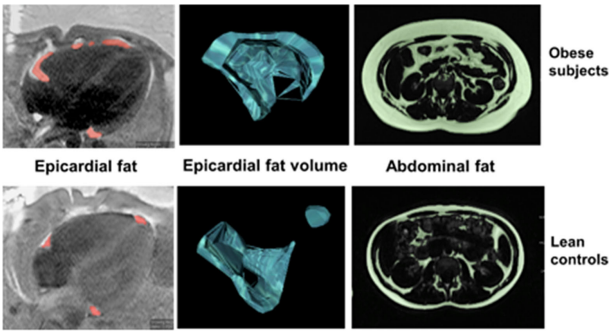
Introduction: Epicardial adipose tissue (EAT) is a metabolically active ectopic visceral fat depot, located between the myocardium and the visceral layer of the pericardium. EAT is associated with metabolic syndrome (MS) and coronary artery disease in adults. EAT thickness measured by echocardiography is increased in obese youth, but total EAT burden and its correlation with cardiovascular risk factors have not been studied.

Methods: We performed a cross-sectional study including 48 pubertal adolescents (24 obese and 24 lean subjects, aged 13.6 ±1.5 yr). EAT volume (using 3D phase-sensitive inversion recovery), as well as total, visceral, and subcutaneous abdominal adipose tissue volumes (using a 3D volume interpolated breath-hold enhanced imaging sequence) were obtained by magnetic resonance imaging. Anthropometrical parameters, blood pressure (BP), fasting serum triglycerides, total, low- and high-density lipoprotein (HDL-C) cholesterol, glucose and insulin levels were measured.

Results: Obese adolescents had higher EAT volume compared to lean controls (49.6±18.0 vs 17.6±6.7 cm^3, $p<.0005$). They also had significantly increased total and visceral abdominal fat volumes, systolic BP, serum triglycerides and insulin levels, and decreased HDL-C concentration. EAT volume was significantly associated with anthropometrical indexes and cardiovascular risk factors (using Pearson correlation coefficient, $p<0.05$): waist circumference ($r=0.827$), systolic BP ($r=0.316$), triglycerides ($r=0.534$), HDL-C levels ($r= -0.477$), fasting insulin level ($r=0.583$). Metabolic syndrome was present in 25% of obese adolescents. EAT volume was significantly higher in obese adolescents with MS compared to those without MS (63.5±21.4 vs 44.9±14.6 cm^3, $p=.026$), and this difference subsisted after controlling for visceral abdominal adipose tissue volume.

Conclusions: EAT volume, which is known to contribute to atherogenesis in adults, is increased in obese adolescents, and is associated with abdominal visceral fat, cardiovascular risk factors and MS. Excessive EAT early in life may contribute to the development of premature cardiometabolic disease.



O41

Machine-learning based phenogrouping of echocardiographic data can predict the risk of death or heart transplant in pediatric dilated cardiomyopathy

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Introduction: Pediatric dilated cardiomyopathy (DCM) affects left ventricular (LV) function with a high risk of death or heart transplantation (DoT). However, the relation of LV regional function

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and inefficiency to clinical outcomes is underexplored. We aimed to investigate the relationship of regional LV mechanics, global LV systolic and diastolic function and clinical parameters to the outcomes of DoT in children with DCM, using an interpretable unsupervised machine learning approach.
Methods: DCM was defined by a LV end-diastolic dimension z-score>2 and LV ejection fraction (EF) <55%. Echocardiographic and clinical data from healthy and DCM children were retrospectively analyzed. Specifically, aortic, mitral and pulmonary vein Doppler velocity patterns were analyzed over the entire cardiac cycle; together with longitudinal strain curves sampled at 6 LV wall segments from the 4ch apical view. In addition, age, body surface area (BSA), sex and heart failure medications were included. We used unsupervised multiple kernel learning and k-means clustering to reduce data dimensionality, position patients based on complex information similarity and find homogeneous groups of patients with similar echocardiographic phenotype.
Results: A total of 25 heathy (44% male; age = 8.23 ± 6.02 years) and 47 DCM (53% male; age = 4.09 ± 5.5 years) children were analyzed. Clustering on the six first dimensions of the low-dimensional space resulted in five clusters (Cl) (Figure A), with significant proportions of the composite outcome of DoT ($Cl_1 = Cl_2 = Cl_3$ = 0%, Cl_4 = 50%, Cl_5 = 89%; p<0.01). The two first clusters only included healthy subjects while all DCM subjects were in Cl_3-Cl_5. The group with the highest proportion of DoT (Cl_5) comprised the oldest and most frequently medicated subjects (Figure B), while the group with the second highest proportion of DoT (Cl_4) compromised patients with the lowest LVEF and GLS, higher LVEDD z-score (Figure B), reduced and delayed peak aortic outflow velocity, longer IVRT and severely impaired basal LV strain (Figure C).
Conclusions: Our results serve as a proof of concept that interpretable machine-learning can be useful to explore and understand which regional and global echo parameters, in relation to clinical parameters, are associated with a higher risk of DoT in pediatric DCM.

O42

Determinants of oxygen uptake and prognostic factors in cardiopulmonary exercise test (CPET) in patients with Fontan surgery.

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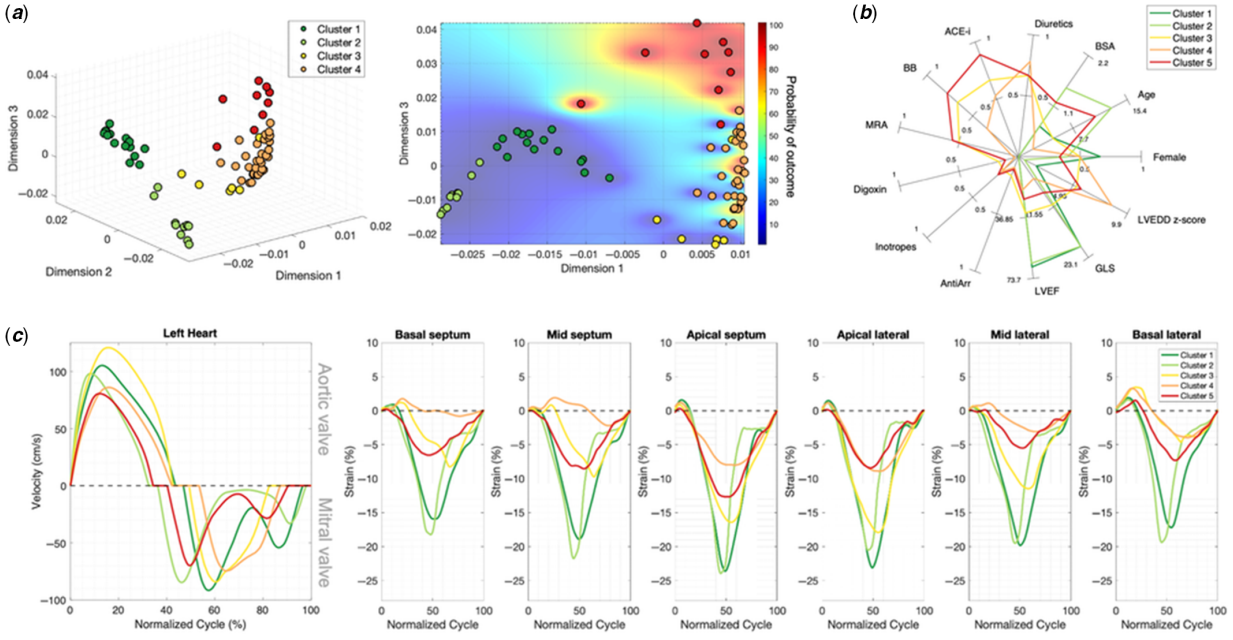
Introduction: CPET allows quantification of functional capacity of patients with Fontan. Their maximum oxygen consumption (VO2max) is significantly lower than expected, and research has been carried out on its determinants.
Methods: A retrospective, unicentric, cross-sectional, descriptive study was carried out on 57 patients with Fontan, who had undergone incremental CPET with cycloergometer between 2010-2020 (table). Determinants of VO2max were analyzed using linear regression model; and determinants of clinical deterioration (defined as functional class III-IV, need for admission or re-interventions, liver failure or nodules suggesting malignancy, ventricular dysfunction, severe atrioventricular valve failure, obstruction of the Fontan circuit and/or significant arteriovenous or venous collaterals), using binary logistic regression model. Variables with statistically significant association in the univariate analysis were included in the multivariate analysis.
Results: The median VO2max was 23(18-28)ml/kg/min and the median oxygen consumption at anaerobic threshold (VO2AT) was 13.6(11-17)ml/kg/min, the median CO2 equivalents at anaerobic threshold (VE/VCO2) was 32(29-35) and the median maximum expected heart rate was 84%(72-88). Chronotropic insufficiency was 38%(22). Significant desaturation to effort was registered in 29%(15). The profile of the oxygen pulse curve was "flattened" in 58% (33). In the univariate analysis, the variables significantly related



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to VO2max were: age at the time of CPET, sex, BMI, years of Fontan evolution, intracardiac Fontan, VO2AT, VE/VCo2 and chronotropic insufficiency. The multiple linear regression model that best fitted the relationship between VO2max and independent variables (correlation coefficient 0.73) included sex (correlation index 3.35; p=0.02), BMI (-0.27; p=0.02), chronotropic failure (-2.79; p=0.01) and VO2AT (0.92; p<0.0001). In the univariate analysis of the prognostic CPET variables related to an unfavorable clinical situation, significance was obtained with chronotropic insufficiency(p=0.003), VO2max (p=0.099) and VO2AT(p=0.089). In multivariate analysis, chronotropic insufficiency maintains its association with unfavourable clinical situation (P=0.017, OR=4.65(1.3-16.5).

Conclusions: In conclusion, together with the anthropometric parameters universally related to VO2max (age, sex and BMI), chronotropic insufficiency and VO2AT are the main determinants of functional capacity in patients with Fontan. Moreover, chronotropic insufficiency is closely related with unfavourable clinical evolution. Our data would support the intensive treatment of chronotropic insufficiency in order to improve the quality of life and the clinical situation of patients with Fontan.

Continuous clinical variables	Median (interquartile range)	
Age at CPET (years)	23 (13-42)	
BMI (kg/m2)	21.3 (18.8-25.3)	
Age at Fontan surgery (years)	8.8 (2-20)	
Average follow-up time from Fontan surgery (years)	12 (3-36)	
Categorical clinical variables		**Proportion N=57**
Gender (male)		35 (61.5%)
Early Fontan (< 8 years)		22 (38.6 %)
Surgical technique	intracardiac	14 (24.6%)
	extracardiac	43 (75.4%)
Single ventricle anatomy	left	30 (52.6%)
	right	5 (8.8%)
	Double ventricular muscle mass sor single AV valve	22 (39.6 %)
Beta-blocker treatment		17 (29.8%)
Pacemaker holder		7 (12.3%)
Atrial arrhythmias		9 (15.7%)
Unfavourable baseline clinical situation		22 (38.6 %)
	Ventricular echocardiographic dysfunction	6 (10.5%)
	Obstruction of the Fontan circuit (stent/thrombosis/stenosis/dilation)	10 (17.5%)
	Liver failure	2 (3%)
	Protein-Losing Enteropathy	1 (1.7%)
	Plastic bronchitis	1 (1.7%)
	Cerebrovascular accident	1 (1.7%)
	Ventricular tachycardia (ICD)	1 (1.7%)

Table. Demographic, anthropometric and clinical variables.

O43

Determinants of adverse response to exercise in treated aortic coarctation patients

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Introduction: Aortic coarctation is associated with several sequelae after treatment, including abnormal responses to exercise. We investigated determinants of adverse outcomes on exercise testing.

Methods: Asymptomatic patients with successfully treated aortic coarctation (residual isthmic Doppler gradient ≤ 20mmHg) or with borderline gradient (>20≤25mmHg) were prospectively evaluated with exercise testing and exercise echocardiography. Age at evaluation ranged from 8-40 years (mean 20.6). Exclusion criteria included other significant anomalies. Exercise was performed on a treadmill with a Bruce protocol. Isthmic Doppler gradient and flow pattern was assessed within 30 seconds of peak exercise. Adverse exercise outcome was defined by a composite endpoint consisting of exercise hypertension, isthmic diastolic flow on peak exercise Doppler, or ischemic changes. Clinical, physiological and morphological (MR) data were correlated with exercise test results. Statistical analysis was performed with Stata v13. For binomial variables chi-squared tests were used, for continuous variables we used t-test or Wilcoxon rank sum test. Multivariable logistic regression models were built, and the best models chosen using ROC curves.

Results: Forty-one patients were evaluated. Twelve (29%) reached the endpoint, which did not correlate with age, sex, BMI, type of treatment, or indexed LV mass. The endpoint was strongly associated (p<0.01) with higher baseline office systolic BP (mean 140.0mmHg (95% CI 131.3-148.7) vs. 120.7mmHg (115.2-126.2) for those not reaching the endpoint); with a borderline isthmic Doppler gradient at rest; with a higher Doppler gradient at peak exercise (mean 47.2mmHg (37.2-57.2) vs. 30.8mmHg (26.0-35.6)); and with a lower cardiac MR ratio of narrowest diameter of aortic arch/aortic diameter at diaphragm level (0.71 (0.64-0.79) vs. 0.94 (0.86-1.03)). Multivariable logistic regression, after adjusting for confounders, showed that the ratio of narrowest aortic arch segment diameter/aortic diameter at the diaphragm was the single best predictor of adverse exercise outcome (p<0.01, AUC = 0.9167) with an optimal cut-off point of 0.87.

Conclusions: Treated aortic coarctation patients have a high prevalence of abnormal exercise responses. Persistent aortic hypoplasia determined by a ratio of narrowest aortic arch segment/aorta at the diaphragm <0.87 by MR was found to be the best predictor of adverse outcomes during exercise.

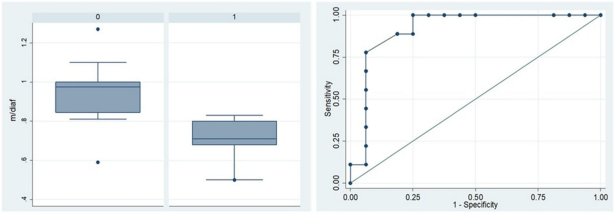

Figure 1 Left: ratio of narrowest diameter of aortic/diameter of the aorta at diaphrag, level (m/diaf) plotted against abnormal responmse to exercise (0=no, 1=yes). Right: ROC curve for m/diaf against abnormal response to exercise (AUC=0.9167). * - abnormal response to exercise = composite endpoint: hypertensive response, isthmic diastolic flow on exercise, significant ST-T changes.*

O44

Decline of exercise capacity in patients after total cavopulmonary connection in longitudinal follow-up

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Children's Heart Centre, Second Faculty of Medicine, Charles University in Prague and Motol University Hospital (1), Department of Rehabilitation and Sports Medicine, Second Faculty of Medicine, Charles University in Prague and Motol University Hospital (2)

Introduction: Fontan circulation is associated with suboptimal long-term outcome due to chronic venous congestion. Cardiopulmonary exercise

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test (CPET) is a sensitive non-invasive method of functional assessment in patients after surgical repair of congenital heart disease. Our study aims to analyse exercise capacity of patients after total cavopulmonary connection (TCPC) in long-term follow up.
Methods: A retrospective longitudinal study of all patients with TCPC, who were operated during childhood, with analysis of available CPET data.
Results: All consecutive patients (N=354, 40.6% female) who underwent TCPC between 1992 and 2016 in our country were identified from the institutional database. N=288 (81.4%) patients (40.0% female) underwent at least one CPET. True maximal stress test data were acquired in N=258/288 (89.6%) patients. Individual trajectories of VO2peak were studied in 206 patients who had at least 2 valid VO2peak values throughout follow-up. The underlying systemic ventricle morphology included 106 patients with morphologically left ventricle (LV), 88 patients with morphologically right ventricle (RV) and 12 patients with undetermined ventricle. The trajectory of VO2peak expressed as mean (SEM) in all TCPC patients was 37.0 (1.1) ml/min/kg - 0.39 (0.1)*age, compared to national reference values for healthy peers, where VO2peak (SEM) equals 50.4 (0.2) ml/min/kg - 0.40*age. The steepness of decline in exercise capacity over time was not dependent on ventricle morphology (p=0.23).
Conclusions: TCPC patients have significantly reduced exercise capacity. The steepness of decline of VO2peak did not differ from the natural steepness of decline in general population. Single ventricular morphology had no influence on the exercise capacity dynamics. Supported by grant provided by MHCZ, 00064203

O45
Advanced imaging and new cardiac biomarkers in long-term follow-up after childhood cancer

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Introduction: Pathological findings of ejection fraction (EF), shortening fraction (FS) and standard heart failure biomarkers (troponin T and NTproBNP) during follow-up after childhood cancer have been associated to irreversible cardiac damage, therefore more sensitive parameters for the detection of subclinical functional cardiac changes are needed. Aim of our investigation was to evaluate strain imaging values by echocardiography and new biomarkers for heart failure with preserved ejection fraction (HFpEF) in children, adolescents and young adults in follow-up after childhood cancer.
Methods: Prospective study in 50 childhood cancer survivors [median 16.2 years (IQR 14-18.5)], at median follow- up of 13 years (IQR 10-15). In addition to standard echo and laboratory parameters for heart failure, strain measurements and the following new biomarkers were obtained and compared to 50 healthy controls: myocardial inflammation (IL-6), extracellular matrix remodeling (CITP: C-terminal telopeptide of type-I collagen; PIIINP: intact N-terminal propeptide of type III procollagen) and other heart failure biomarkers (galectin 3, sST2: solutable ST2, GDF 15: growth differentiation factor 15).
Results: No significant differences in EF, FS, troponin T and NT proBNP, IL-6 and sST2 were found between study and control group. Instead, advanced imaging parameters showed significant differences between both groups [global longitudinal strain (-15.9% vs -20.4%, p<0.0001), global circumferential strain (-14.3 vs -20.3%, p < 0.0001)], detecting 66% (GLS) and 76% (GCS) pathological values in contrast to 20% (EF) and 16% (FS) for standard parameters. Markers for disturbances of extracellular matrix remodeling (CITP, PIIINP, each p < 0.0001), galectin 3 (p 0.01) and GDF 15 (p < 0.0001) were significantly different between the groups.
Conclusions: Standard echo and laboratory parameters used during cardiac evaluation in follow-up after childhood cancer seem to be less sensitive in detecting early remodeling processes in contrast to strain imaging and newer cardiac biomarkers used in HFpEF. Especially the detection of myocardial remodeling processes due to disturbed collagen turnover at an early stage might give the opportunity to begin heart failure treatment earlier with the potential to delay its negative influence on cardiac function.

O61
Minor Pressure Drops within the Fontan-Anastomosis in Patients with Total Cavopulmonary Connection (TCPC) by 4D-MR Pressure Mapping

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Introduction: Pressure measurement in TCPC patients is a domain of cardiac catheterization. 4D-MR flow pressure mapping offers an alternative for assessment of even minor pressure differences using Navier-Stokes and Pressure Poisson equations. The scope of this study was to define flow characteristics over the Fontan-anastomosis and to measure even minor pressure differences between caval veins and pulmonary arteries.
Methods: Twelve patients (mean age 16.1 ± 6.1 years) with clinically uncompromised TCPC and 18 mm extracardiac conduit were studied by 4D-MR flow pressure mapping. Postprocessing was performed by MEVISFlow software (Fraunhofer Mevis, Berlin). Pressure differences between superior vena cava (SVC) and extracardiac conduit (C) to both pulmonary arteries (RPA, LPA) were assessed. For this purpose, anatomically correct centerlines were created on predefined vessel masks and maximal pressure drop was taken from time resolved pressure curves along these centerlines. Vortices and direction of flow within the anastomosis were registered by flow pathlines from 4D MR flow maps (see image attached).
Results: Median [minimim und maximum] pressure drop between SVC and RPA was 0.63 mmHg [0.21 – 1.10], between C and RPA 0.55 mmHg [0.29 – 2.2], between SVC and LPA 0.63mmHg [0.27 – 1.2] and between C and LPA 0.45 [0.24 – 1,72]. Pressure drop between SVC-LPA and C-LPA was significantly different (p value < 0,04), compared to SVC-RPA and C-RPA (p < 0,13).Vortices in the Fontan-anastomosis were registered in 3 from the included 12 patients, representing the three highest pressure drops in the whole study group.
Conclusions: Even minor pressure drops over the Fontan-anastomosis in venous low-pressure circulation can be assessed by 4D-MR flow pressure mapping. These values are below the order of conventional cardiac catheterization. Pressure drops can be detected in even clinically uncompromised TCPC patients. Pressure drops from SVC into LPA are slightly higher than from extracardiac conduit into LPA.

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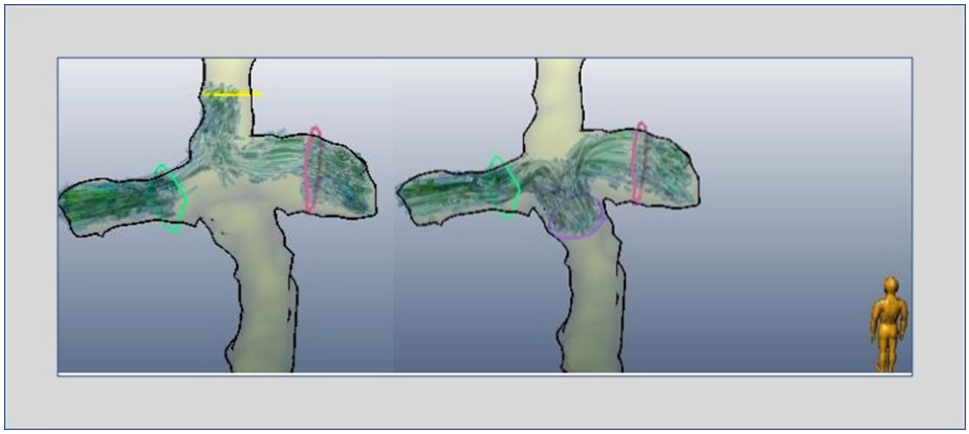

O62

Pulmonary valve replacement in repaired Tetralogy of Fallot: are we using all the right parameters for the indications?

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Introduction: Chronic pulmonary regurgitation (PR) is a common consequence of Tetralogy of Fallot repair (rToF), leading to right ventricular (RV) dilation, dysfunction and ventricular arrhythmias. Thus, pulmonary valve replacement (PVR) is useful to reduce RV dimensions and preserve RV function. RV dilation and RV dysfunction are the most important MRI parameters considered for PVR indication in asymptomatic rToF patients. However, it is still unclear whether additional clinical and/or MRI parameters can be relevant in determining PVR indication. Our aim was to investigate clinical and MRI determinants of PVR indication in our large cohort of rToF children and adolescents.
Methods: All rToF patients who underwent MRI evaluation at Bambino Gesù Children Hospital between 2007 and 2020 were included. Demographic, surgical data and MRI parameters (biventricular volumes and function, pulmonary and tricuspid regurgitation, pulmonary branch and/or right ventricle outflow stenosis) before PVR (if performed) were collected.
Results: 342 patients (60% males, 41,5% aged>18 yrs) aged 18.7 ±8.4 (range 3-54) were included. Mean age at repair was 1,2 ± 1,5 years. In most cases (85%) a transannular patch was performed, of which 177 (52%) patients had a transventricular approach. At multivariate analysis adjusted for age at repair, male gender and transventricular approach were risk factors for RVEDVi>160 ml/m^2, whereas at multivariate analysis for RV dysfunction, male gender and age at repair resulted independent predictors. rToF patients who had PVR were more frequently man, had a transannular patch and a transventricular approach in higher prevalence and were younger at MRI exam compared to the others (Table 1). At multivariate analysis adjusted for age at repair and presence of pulmonary stenosis, male gender (O.R. 2,399, 95% IC 1,502-3,830, p<0.0001), transannular patch (O.R. 6,759, 95% IC 2,961-15,430, p<0.0001), transventricular approach (O.R. 1,695, 95% IC 1,066-2,694, p=0.026), and RV obstruction (O.R. 0,306, 95% IC 0.107-0.881, p=0.028) resulted independent determinants for PVR indication.
Conclusions: Male gender is a risk factor for both significant RV dilation (RVEDVi>160 ml/m^2) and dysfunction; therefore, it should be included in PVR indication. The transventricular approach should be taken into consideration as one of determinants for PVR indication in rToF.

	PVR (n=146)	Not PVR (n=196)	p value
Gender (male, %)	105 (72)	102 (52)	<0.0001
Transanular Patch	138 (95)	153 (78)	<0.0001
Transventricular approach	87 (60)	90 (46)	0.016
Age>18 yrs	52 (36)	90 (46)	0.060
Age at correction	1,14±1,4	1,2±1,5	0.686
Age at cardio MRI	16,6±6,5	20±9	<0.0001
Time between surgery and MRI	15±6	19±8	<0.0001
BMI	20±4	22±4	0.001
RVEDVi>160	78 (53)	11 (5)	<0.0001
RVEDV	230±68	189±52	<0.0001
RVEDVi	144±33	128±31	<0.0001
RVESV	112±41	89±35	<0.0001
RVESVi>80	70 (45)	8 (4)	<0.0001
RVESVi	68±22	57±20	<0.0001
RVEF<45%	22 (15)	14 (7)	0.021
RVEF	53±6	55±6	<0.001
LVEDV	136±39	111±26	<0.0001
LVEDVi	85±14	75±11	<0.0001
LVESV	59±22	46±14	<0.0001
LVESVi	37±10	31±7	<0.0001
LVEF	56±6	58±6	0.001
LVEF<55%	59 (40)	48 (24)	0.002
PR%	39±16	39±15	0.990

O63

The oxygen uptake efficiency slope: a reliable surrogate parameter for exercise capacity in healthy and cardiac children?

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Introduction: Cardio-pulmonary exercise test (CPET) provides accurate evaluation of physical capacity and disease severity in children with congenital heart disease (CHD). However, full participation to obtain optimal measure of $VO2_{max}$ may be difficult. As an alternative, the oxygen uptake efficiency slope (OUES) is a reproducible and reliable parameter measured during CPET, which does not require a maximal exercise to be interpretable. This study aimed to evaluate the OUES of a large cohort of children with CHD, in comparison with healthy controls. We also intended to identify, in this specific population, the clinical and CPET variables associated with the OUES.
Methods: This cross-sectional study was carried out between November 2010 and September 2015 in two tertiary care pediatric and congenital cardiology centers.

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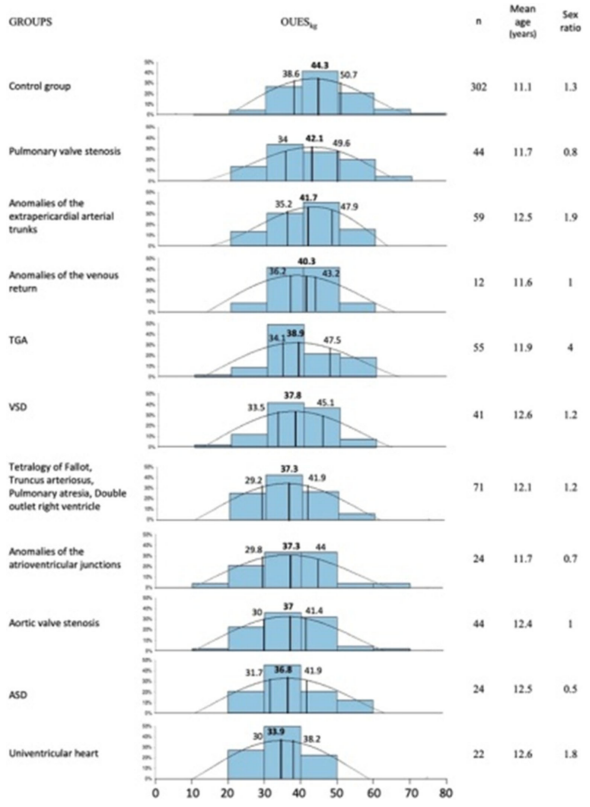
Results: 709 children were included (407 CHD and 302 healthy controls). The association of clinical characteristics with weight-normalized OUES ($OUES_{kg}$) was studied using a multivariable analysis. The mean $OUES_{kg}$ was significantly lower in CHD than in healthy controls (38.6±8.5 and 43.9±8.5; P<0.001, respectively), especially in the most severe CHD. The $OUES_{kg}$ correlated with $VO2_{max}$ (r=0.85, P<0.001), with cut-off values for normal exercise capacity of 38.4 in boys and 31.0 in girls. The decrease of $OUES_{kg}$ was associated with increased age, increased BMI, number of cardiac catheter or surgical procedures, female gender and decreased FVC (Z-score).
Conclusions: The OUES is significantly impaired in children with CHD and strongly correlates with $VO2_{max}$. The OUES has the same clinical determinants as $VO2_{max}$ and therefore may be of interest in submaximal exercise.



O64

The VE/VCO2 slope: a useful tool to evaluate the physiological status of children with congenital heart disease

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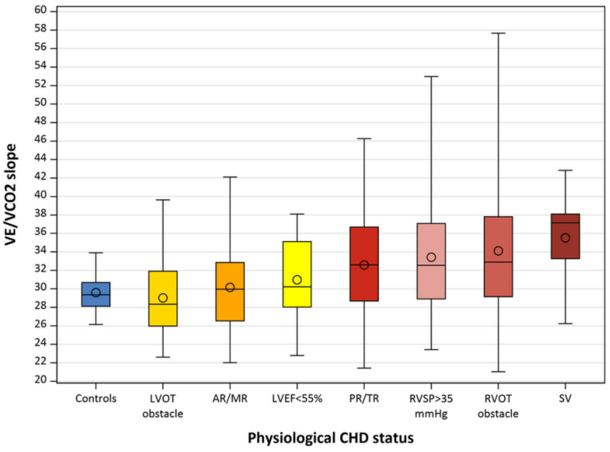
Introduction: Cardio-pulmonary exercise test (CPET) is becoming a key examination to assess physical capacity and disease severity in paediatric cardiology. The VE/VCO_2 slope has been increasingly used as a surrogate marker for morbidity and mortality in adult heart failure, pulmonary arterial hypertension and for adult patients with CHD. Nevertheless, the use of the VE/VCO_2 slope in children remains limited in the absence of reference values and clearly identified clinical determinants. This study aimed to compare the VE/VCO_2 slope in a paediatric cohort with CHD to that of age- and gender-adjustedhealthy controls. We also intended to identify the clinical and CPET variables associated with VE/VCO_2 slope in this population.
Methods: This cross-sectional study was carried out between November 2010 and September 2015 in two tertiary care paediatric cardiology reference centres.
Results: A total of 700 children were enrolled (399 CHD and 301 healthy controls). The mean VE/VCO_2 slope was significantly higher in CHD than in healthy subjects (31.6±4.8 vs. 29.3±4.8; P<0.001). The VE/VCO_2 slope was higher in children with significant pulmonary regurgitation, tricuspid regurgitation, right ventricular hypertension and right ventricle outflow tract (RVOT) obstacle. In the CHD group, VE/VCO_2 slope increase was associated with BMI, the presence of a RVOT obstacle, the number of cardiac catheter procedures, as well as low age, FVC, tidal volume, and PetCO2.
Conclusions: Increased VE/VCO_2 slope was predominantly in children with single ventricle and/or residual right heart abnormalities suggesting that maldistribution of pulmonary blood flow during exercise is an important CHD-unique determinant of VE/VCO_2 slope.



O65

Tricuspid insufficiency reduction and preload recruitment during submaximal exercise-CMR in patients with unoperated Ebstein and tricuspid dysplasia

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Introduction: Adolescents and adults with native Ebstein's anomaly represent the benign part of the Ebstein spectrum that survived infancy without surgery; in this population exercise capacity is crucial to guide further treatment and to evaluate the need for surgery. We analyzed the pathophysiology of this population by CMR at rest and during submaximal exercise.
Methods: We used a CMR protocol, established in our unit for submaximal exercise (about 24% peak exercise; JACC Cardiovasc Imaging 2014;7(3):314-5). We analyzed 12 unoperated patients (9 Ebstein, 3 tricuspid dysplasia, age-range 12-61 years). We

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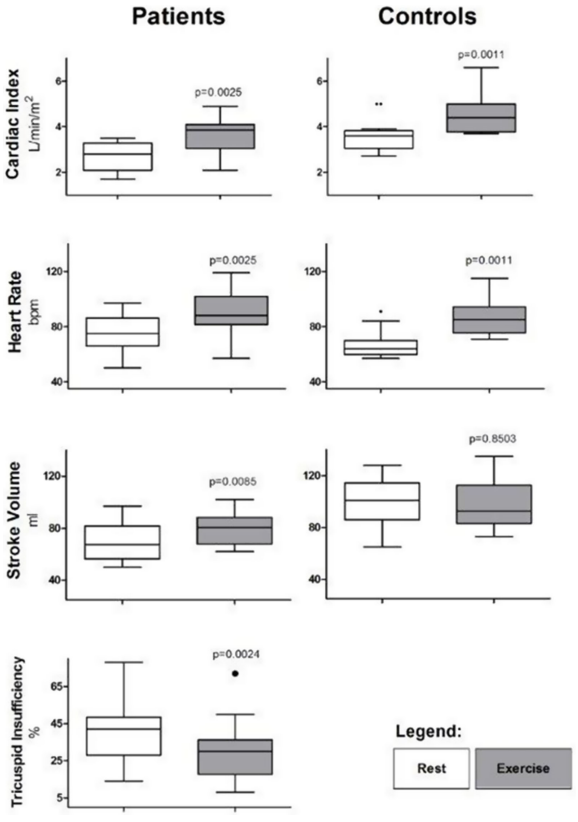
measured ventricular volume and flow at rest and during exercise, including direct measurement of tricuspid insufficiency (TI) by tricuspid valve flow. We performed control examinations in 14 healthy subjects.
Results: Our patients had significant TI at rest which decreased during exercise (TI at rest and during exercise was 41% ± 18 and 30% ± (17), respectively, p=0.0024); concomitantly, the LV-preload increased significantly as shown by increase in LVEDV (5% increase, p=0.0024) and SV. Cardiac index (CI) and aortic stroke volume (SV) at rest were significantly higher in controls (p=0.0005 and 0.0010) while heart rate at rest (HR) was similar in the two groups (p=0.0774). During exercise CI increased significantly and similarly by controls and patients (22% and 25% respectively). The patients had a mean CI of 2.69 ± 0.63 L/min/m2 at rest, and 3.61 ± 0.78 during exercise (p=0.0025). Controls had mean CI of 3.59 ± 0.72 L/min/m2 at rest, 4.58 ± 0.84 during exercise (p=0.0011). HR increased similarly in the two groups (16% increase in patients, p=0.0025; 23% in controls, p=0.0011). SV increased in the patient group (11% increase, p=0.0085) while remained stable in the control group (1% decrease, p=0.8503).
Conclusions: Results confirm high reproducibility of our submaximal exercise protocol. During submaximal exercise, patients with Ebstein and significant TI by valve dysplasia improved their cardiovascular efficiency by decreasing TI and recruitment of volume, as proven by an increase in LVEDV and SV. Thereby, increase in CI during submaximal exercise did not differ from normal controls.

O86



Individual differences in brain development and cognitive outcome in infants with Congenital Heart Disease

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Introduction: Infants with Congenital Heart Disease (CHD) are at risk of neurodevelopmental impairments, the origins of which are currently unclear. The aim of this study was to characterise the relationship between neonatal brain development, cerebral oxygen delivery and neurodevelopmental outcome in infants with CHD.
Methods: Sixty-six infants with serious or critical CHD underwent brain MRI prior to surgery on a 3T MRI scanner situated on the neonatal unit at St Thomas' Hospital, London. T2-weighted images were segmented into brain regions using a neonatal-specific algorithm. We generated normative curves of typical volumetric brain development using a data-driven technique applied to 219 healthy infants from the Developing Human Connectome Project (dHCP). Atypicality indices, representing the degree of positive or negative deviation of a regional volume from the normative mean for a given gestational age, sex and postnatal age, were calculated for each infant with CHD. Extreme deviations from typical brain development were taken as atypicality indices >±2.6. Cerebral oxygen delivery (CDO_2) was calculated from phase contrast angiography in 53 infants with CHD. Cognitive and motor abilities were assessed at 22 months (N=44) using the Bayley-III. We assessed the relationship between atypicality indices, CDO_2 and cognitive and motor outcome. We also examined whether CDO_2 was associated with neurodevelopmental outcome through the mediating effect of regional brain volumes.
Results: Extreme deviations in development were identified in extracerebral CSF, ventricles and subcortical brain structures. Negative atypicality indices in bilateral caudate nuclei and thalami and left lentiform nucleus were associated with both reduced neonatal CDO_2 and poorer cognitive abilities at 22 months. There was a significant indirect relationship between CDO_2 and cognition through the mediating effect of lower bilateral caudate and thalami atypicality indices (Figure 1).
Conclusions: Lower cognitive abilities in toddlers with CHD were associated with smaller caudate nuclei, thalami and left lentiform volumes prior to cardiac surgery. The aetiology of poor cognition may encompass poor cerebral oxygen delivery leading to impaired caudate and thalamus growth. Interventions to improve cerebral oxygen delivery may promote early brain growth and improve cognitive outcomes in infants with CHD.

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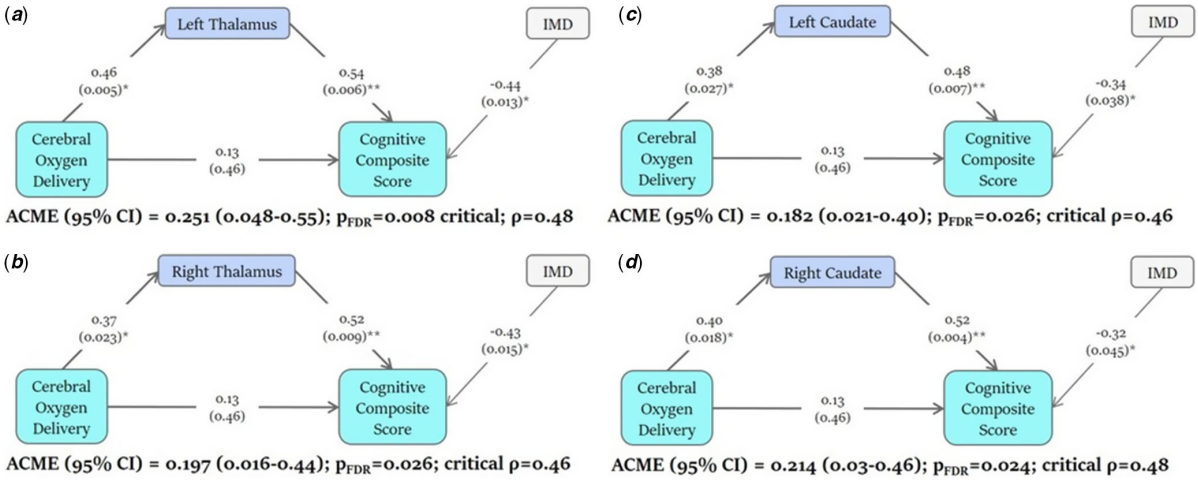

Figure 1. Path diagrams showing the indirect relationship between Cerebral Oxygen Delivery and Cognitive Composite Score mediated by (A) left thalamus (B) right thalamus (C) left caudate nucleus (D) right caudate nucleus. Standardised regression coefficients are reported; numbers in brackets show p-value *p<0.05 **p<0.01. ACME, Average Causal Mediation Effect; IMD, Index of Multiple Deprivation

O87

Are hepatic and cardiac fibrosis two sides of the medal in Fontan-associated multi-organ disease?

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Introduction: Fontan patients are at risk of developing liver fibrosis and impaired cardiac function maybe related diffuse myocardial fibrosis. This study aims to investigate Magnetic Resonance (MR) T1 mapping and extracellular volume fraction (ECV) of the myocardium and liver in Fontan operated adolescents. We hypothesized that MR biomarkers of diffuse fibrosis in the heart and liver are increased in Fontan adolescents and that myocardial and hepatic findings are correlated.
Methods: Myocardial and hepatic native T1 times and ECV were prospectively measured in a Fontan cohort using a modified Look-Locker inversion recovery (MOLLI) sequence from respectively cardiac short axis views and a separate axial MOLLI sequence over the liver. In addition, myocardial late gadolinium enhancement was performed. Findings were compared and correlated to cardiac MR volumetrics and the results were compared to an age matched control group.
Results: Forty-six patients aged 16.7±0.6 years and 15 healthy individuals aged 19.2±1.2 years were enrolled. MR performed in 35 patients revealed slightly increased myocardial native T1 times in the lateral wall 1013± 48ms versus in controls 992 ±26 ms (p=0.04) while hepatic T1 times were elevated in the Fontan group, 774±44 ms versus 632±52 ms in controls (p<0.001). The ECV values were comparable between the groups in the myocardium; in Fontan patients, 24.4±4.3% versus 23.5±2.7% in controls (p=0.6), while significantly increased I the liver, 47.4 ±5.0% in Fontan versus 34.6±3.8 % in controls, (p<0.001). Focal myocardial fibrosis LGE was only found in one patient. There was no significant difference regarding ventricular morphology (p=0.4-0.9) and no correlation between T1 times or ECV in heart and liver (R=0.23-0.36, p=0.06-0.2). Myocardial ECV correlated positively with ventricular stroke volume (p=0.001, R=0.55). T1 correlated weakly with ventricular end diastolic and end systolic volume (p=0.04, R= 0.40 for both).
Conclusions: In this young cohort of Fontan adolescents there was MR evidence of diffuse myocardial fibrosis and significantly increased MR markers suggestive of diffuse hepatic fibrosis and/or congestion. There was no correlation between the hepatic and myocardial MR markers but ventricular stroke volume seems linked to increased myocardial ECV.

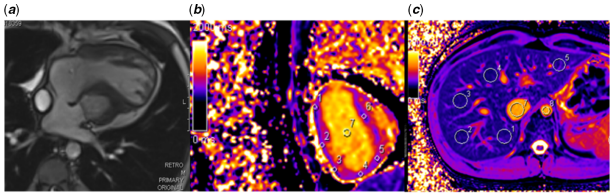

Figure 1. MR in an adolescent with hypoplastic left heart syndrome and Fontan palliation a) Cardiac 4-chamber view b) T1 mapping with regions of interest in the heart, short axis view and c) in the liver, axial view.

O88

Noninvasive versus Invasive Assessment before Total Cavopulmonary Connection in Single Ventricle Patients: a Multi-center Study

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Introduction: Prior to total cavopulmonary connection (TCPC), careful cardiac and pulmonary vascular assessment of patients with single ventricle (SV) physiology is mandatory but there is still no consensus whether noninvasive or invasive assessment should be used, particularly in patients with acceptable clinical course and

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hemodynamics at previous (Glenn) surgery. Furthermore, transcatheter therapy of e.g. pulmonary artery branch stenosis or aortopulmonary collaterals is sometimes used as complement to pre-TCPC hemodynamic assessment. We assessed in these patients the utility of pre-TCPC catheterization compared to noninvasive assessment.

Methods: All patients with SV born 2004 to 2016 with TCPC at the Skåne University Hospital in Lund, Sweden, and pre-TCPC assessment with either computed tomography (CT) or magnetic resonance imaging (MRI) ("Noninvasive" group; n=22) were one-to-one matched for SV morphology (hypoplastic left heart syndrome, tricuspid atresia, double inlet left ventricle and pulmonary atresia with intact ventricular septum), surgical age, clinical status and hemodynamics with patients with either diagnostic catheterization ("Dx-cath"; n=22) or combined diagnostic-interventional catheterization ("Interv-cath"; n=22) from the Queen Silvia Children's Hospital, Gothenburg, Sweden (n=17) and Wilhelmina Children's Hospital, Utrecht, Netherlands (n=27). The hemodynamic data (mean pulmonary artery and transpulmonary pressure gradient) was obtained either intraoperatively immediately after Glenn anastomosis in the Noninvasive group or via pre-TCPC catheterization in the other 2 groups. All interventions in the Interv-cath group were pursued on indications that are not eligible for pre-TCPC interventions in Lund. The groups were compared for several adverse outcomes during the 1st year after TCPC.

Results: There were no statistically significant differences (p>0.1) between the three pre-TCPC assessment groups regarding pleural drainage >30 days, duration of hospital stay >30 days, reoperations and catheterization within 12 months after TCPC (Table). One patient in the Dx-cath group died on day 8 post-TCPC due to Fontan failure, and one in the same group underwent heart transplantation during the first year post-TCPC.

Conclusions: In clinically stable SV patients, pre-TCPC catheterization does not seem to provide further benefit on early post-TCPC outcome, as compared to noninvasive assessment.

Table.

	Noninvasive (n=22)	Diagnostic (n=22)	Interventional (n=22)	P-value
Pleural drainage >30 days	2 (9.1%)	2 (9.1%)	5 (22.7%)	0.35
Hospital stay >30 days	2 (9.1%)	6 (27.3%)	6 (27.3%)	0.22
Htx or Htx listing within 12 months	0 (0%)	1 (4.5%)	0 (0.0%)	0.36
Death within 12 months	0 (0%)	1 (4.5%)	0 (0.0%)	0.40
Operations within 12 months	1 (4.5%)	1 (4.5%)	3 (13.6%)	0.74
Cath within 12 months	0 (0.0%)	3 (13.6%)	5 (22.7%)	0.11

O89

3D statistical shape models of the paediatric heart and the anatomical remodelling due to obesity

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Introduction: 3D statistical shape modelling is a new method of analysing cardiac magnetic resonance (CMR) images, through which the 3D cardiac morphology can be modelled and compared. In adult cohorts, specific cardiac morphologies (for example, with increased sphericities) were strongly associated with known cardiovascular risk factors and (sub)clinical disease processes. We applied this method on a paediatric cohort to build an atlas of the shape of the paediatric left ventricle (LV), to describe the range of its natural morphological variability and to identify early deviations associated with obesity that could signal increased disease risk.

Methods: CMR images from 2631 Dutch children without cardiac disease (10.1 (SD 0.6) years, 51% girls) from the Generation R study were manually segmented, and 3D LV meshes were automatically fitted to the segmentations to reconstruct the LV morphology. A statistical shape model was computed from the meshes with a principal component analysis (PCA) to derive a set of modes of variation over the mean shape. We assessed the unique cardiac morphology of obesity by identifying the PCA modes that were associated with a subject's BMI (continuous variable), but not with other physical characteristics, including age, gender and height.

Results: The 20 first PCA modes described over 90% of the variability within the population, capturing both the variability in conventional metrics (LV length, width or wall thickness) and novel 3D remodelling patterns. Three shape modes were uniquely correlated with BMI (Fig. 1). These modes were related to LV conicity, apico-basal axis tilting, and antero-inferoseptal convexity.

Conclusions: We present the first morphological atlas of the paediatric heart, based on a large cohort of healthy children. In addition, we identified unique anatomical features related to obesity: obese children present with a more spherical shaped heart, with increased volume in the apical segments and anteroseptal convexity. These findings suggest that the child's heart adapts to obesity by increasing the apical volume to cope with additional workload.

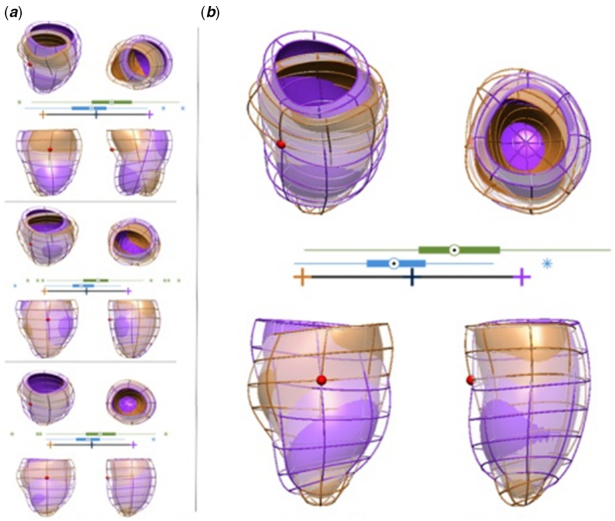

Fig1. LV shape modes (A) and their linear combination (B) that are specifically related to obesity, showing that hearts of obese children (purple overlay) are more spherical, with increased volume in the apical segments and anteroseptal convexity compared to children with low BMI (orange overlay). The red sphere indicates the location of the right ventricle. Distributions of high BMI (top, green) and low BMI (bottom, blue) young heart shape modes are provided in boxplots.

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O90

Exercise dependent changes in ventricular-arterial coupling and aortopulmonary collateral flow in Fontan patients: a real-time CMR study

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Introduction: Inefficient ventricular-arterial (V-A) coupling has been described in Fontan patients that may result in adverse haemodynamics. A varying amount of aortopulmonary collateral (APC) flow is also frequently present that increases volume load of the single ventricle. The aim of the study was to assess changes in V-A coupling and APC flow during exercise CMR.
Methods: Eighteen Fontan patients (mean age 24 ± 3 years, 4 females) and 14 healthy controls (mean age 23 ± 4 years, 5 females, p=0.17) underwent exercise CMR using a recumbent bicycle ergometer. Short-axis ventricular volumetry and 2D flow measurements in the ascending aorta (AAO), inferior vena cava (IVC) and superior vena cava (SVC) were assessed using real-time cine and phase-contrast sequences at baseline, 15, 30, 45 and 60 watts work load. Measures of arterial elastance Ea (endsystolic pressure/stroke volume), ventricular elastance Ees (endsystolic pressure/endsystolic volume) and V-A coupling (Ea/Ees) were assessed. APC flow was quantified as AAO – (SVC + IVC).
Results: Ea did not differ between the two groups and remained unchanged during exercise in both groups while the increase in Ees during exercise was significantly lower in Fontan patients (1.1 ± 0.5 to 1.5 ± 0.8 vs. 1.2 ± 0.4 to 2.2 ± 1.0 mmHg/ml/m^2, p=0.04). V-A coupling was significantly impaired in Fontan patients at baseline (0.7 ± 0.2 vs. 0.5 ± 0.1, p=0.04). Despite significant improvement during exercise V-A coupling was still impaired compared to controls (0.5 ± 0.2 vs. 0.3 ± 0.1, p=0.001). An attenuated increase in heart rate and cardiac output was observed in the Fontan group. Absolute APC flow in Fontan patients did not change during exercise even at maximum work load (0.7 ± 0.5 to 0.8 ± 0.7 l/min/m^2, p=0.82).
Conclusions: Already at rest, Fontan patients showed inefficient V-A coupling that was aggravated during exercise due to a restricted increase in ventricular contractility while APC flow remained unchanged. These results demonstrate the importance of a limited functional reserve of the single ventricle but no further increase in volume load during exercise.

7. Interventional cardiology

O1

Direct paratracheal lymphosclerosis for plastic bronchitis after Fontan: percutaneous versus endoscopic transtracheal technique.

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Introduction: Plastic bronchitis (PB) after Fontan palliation results from abnormal mediastinal lacteals leaking into the bronchial tree. Itkin et al. showed the possibility to embolize these abnormal lymphatic vessels via cannulation of the ductus thoracicus, which is a complex and demanding technique, especially when absent/hypoplastic/stenotic. We report on a new technique comparing two possible accesses: percutaneous and endoscopic.
Methods: To assess efficacy of embolizing abnormal mediastinal lymphatic vessels and leaks in Fontan patients with PB by direct access to the pathological region, shortcutting the ductus thoracicus. Experience in 2 patients.
Results: Mediastinal lymphatic anatomy and function was investigated by inguinal intranodal gadolinium based MR lymphangiography. In patient (1), paratracheal dilated lymph vessels were punctured directly percutaneously with a 22G Chiba needle through the intercostal space under fluoroscopic and cone-beam computerized tomographical (XperGuide®, Philips) and ultrasound guidance (Fig 1, A-B). Good intralymphatic position was ascertained by injecting water soluble contrast with drainage to abnormal lacteals; after flushing with glucose 5%, occlusion was obtained by injection of 5-10 cc of a mixture of lipiodol/n-BCA N-butyl cyanoacrylate (Histoacryl®) 4/1 under fluoroscopic guidance (DAB 25.34 Gy/cm2). There was a total remission of PB, now 27 months of follow-up. In patient (2), all 4 abnormal hypertrophic mediastinal lymfnodes (paratracheal, but also bronchal and subcarinal) were punctured with a 22G ViziShot EBUS-TBNA needle under direct guidance and visualization of an EBUS EndoBronchial UltraSound-guided bronchoscope (Fig (1), C-D). After flushing with 0.5 ml glucose 5%, occlusion was obtained by injecting 10 cc of a mixture of lipiodol/n-BCA 5/1 under fluoroscopic control (DAB 15.2 Gy/cm2). There was a total remission of PB, now 14 months of follow-up.
Conclusions: Direct paratracheal lymphocclusion is effective and obviates the need for transductal access. When comparing both direct puncture techniques, we find the trans-tracheal EBUS puncture more elegant having easier and safer access, and a more complete occlusion with less radiation than the percutaneous puncture.

O2

Ductal Stenting in Infants with Duct-Dependent Pulmonary Blood Flow results in adequate pulmonary artery growth

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Introduction: Infants with duct-dependent pulmonary blood flow often undergo palliation with a systemic-to-pulmonary artery shunt. However, this procedure is associated with postprocedural instability and potential morbidity and mortality. Stenting of the ductus arteriosus (DA) is an emerging alternative; however, up to now with limited data. Thus, the aim of the study was to determine potential risk factors for reintervention and mortality following stenting the DA.
Methods: All patients who underwent stenting of the DA between 2013 and 2020 were included in the study. A classification scheme

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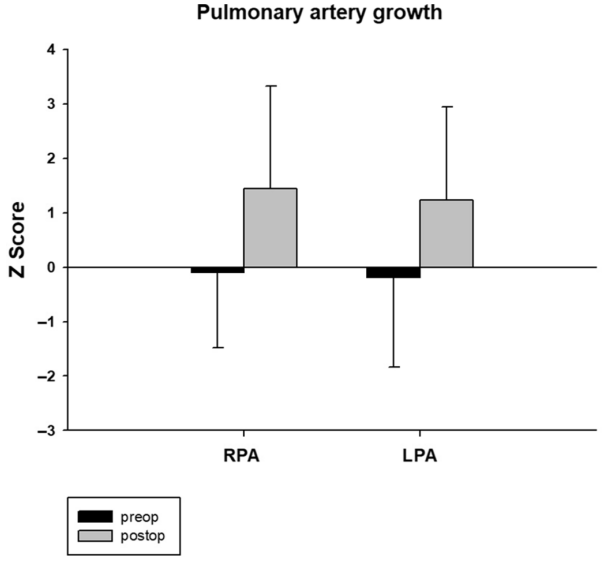
of duct type was developed: Type 1 – straight, type 2 – curved, type 3 – one turn, type 4 – multiple turns. The diameters of the left and right pulmonary artery (LPA; RPA) were evaluated as z-scores prior to DA stenting and prior to next step surgery (NSS). Failure of implantation was defined as an emergency NSS performed up to 9 days after the intervention. Time zero for outcome analyzes was the time of DA stenting.
Results: 76 patients were included in the study. Median age and weight at DA stenting were 7 days [IQR, 4;14 days] and 3022 g [IQR, 2.677-3.455g], respectively. Univentricular physiology was present in 39 %. Survival was 95% ± 2.7% and 90% ± 3.6% at 60 days and 1 year, respectively. Freedom from reintervention (FOR) was 80% ± 5.2 % at 30 days. There was no difference in mortality or FOR between univentricular and biventricular hearts. Complex DA anatomy (Type IV, n= 22) was a risk factor for failure of implantation (p=0.004) and for reintervention (p=0.008). The z-score of the RPA increased from -0.34 [IQR, -1.06-0.69] to 1.32 [IQR, 0.595-2.835] (p<0.001), whereas the z-score of the LPA increased from -0.18 [IQR, -0.78-0.67] to 1.16 [IQR, 0.14-2.365] (p<0.001). There was no difference in growth of LPA/RPA and DA type between univentricular and biventricular hearts. RPA and LPA exhibited symmetric growth with a RPA/LPA ratio of 1.1.
Conclusions: DA stenting yields symmetrical pulmonary artery growth. Complex DA morphology, however, is a risk factor for failure and reintervention. Further studies need to compare these results with the surgical approach.



O3
PDA stenting in infants with duct-dependent pulmonary circulation: Single center experience.

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Cairo University (1)

Introduction: Cyanotic congenital heart disease with duct-dependent pulmonary circulation is a life threatening condition. Percutaneous patent ductus arteriosus (PDA) stenting provides an alternative temporary bridge towards later surgical repair.
Methods: This is a retrospective cohort study including all infants with ductal-dependent pulmonary circulation palliated with PDA stenting from January 2011 to November 2019
Results: The study included 181 patients (60.8% males) with a median age of 20 days (2-120 days range) and a median weight of 3 kg (range 1.8 – 5.5 kg). 72.4% of patients had a tortuous PDA with a mean length of 15 mm and a mean diameter of 3.07 mm at the pulmonary end. 66.4% of patients had a univentricular physiology. The median oxygen saturation before stenting was 71% (with some patients on prostaglandin) compared to 88% post-stenting. Retrograde femoral approach was used in 158 (87.2%) patients, axillary approach in 19 (10.5%) while the carotid access was used in 4 (2.3%) patients. The most commonly used stent sizes were 3.5X24 & 4X16 followed by 4X19. The procedure was successful in 125 (69%) patients, failed in 56 (31%) patients with acute stent displacement in 9 patients, three of which requiring surgery. The median fluoroscopy time was 23.65 minutes & the median radiation dose was 38 Gyc/cm2. The success rate was 100%, 75% and 61% in cases with carotid, femoral and axillary approaches respectively. Stenting in patients with straight PDA (90%) was more successful than in those with tortuous PDAs (67.2%).
Conclusions: percutaneous PDA stenting improves oxygenation in infants with ductal-dependent pulmonary circulation and provides an alternative to palliative surgical shunts with reasonable success rates, which can save lives especially in countries where surgical waiting lists are too long.

O4
Percutaneous atrioseptostomy for left ventricle unloading in patients on veno-arterial extracorporeal membrane oxygenation support

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Introduction: Veno-arterial extracorporeal membrane oxygenation (VA-ECMO) is most commonly considered in paediatric patients presenting with cardiogenic shock. However, left cardiac cavities are not properly unloaded, causing left ventricular dilation with lung congestion and pulmonary edema. Unloading left cavities could facilitate left ventricular recovery. Percutaneous atrial septostomy (PAS) could be a good left ventricle (LV) unloading strategy.
Methods: We retrospectively registered paediatric patients admitted from 2015 to 2020 in our hospital with cardiogenic shock requiring VA-ECMO in which PAS was performed to unload left cavities.
Results: 16 patients were included, 62.5% (10/16) female, with a median age of 6.8 years (10 days-15 years). 15 patients underwent PAS with atrial septal stenting and one patient, who was a neonate with a small ventricular septal defect, underwent just atrial septostomy. 8 patients were diagnosed with myocarditis and 8 patients had dilated cardiomyopathy secondary to different aetiologies. In our hospital, we perform PAS in all patients with severe ventricular dysfunction requiring VA-ECMO to facilitate left ventricular recovery. For that reason, the median time between the initiation of ECMO support and the stent placement was only 5.8 hours (3-12.9 hours). Endomyocardial biopsy for diagnosis was performed in 11 patients. Atrial pressure before and after the procedure was registered in 8 patients, in which there was a significant decrease with a median -16.5 mmHg (CI -23 -5.5 mmHg p 0.02). No complications were reported in any patient. 2 patients required stent dilation for clinical or

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radiological findings suggestive of pulmonary edema. Interestingly, there was no significant gradient across the stent, but both patients had high biventricular filling pressures. 8 patients could be weanned of ECMO with LV recovery, 6 patients required heart transplant and 2 patients died during the ECMO support. Regarding patients who recovered LV function, they currently have a left to right shunt across the atrial stent and 5 of them have mild RV dilation, none of them requiring any intervention to date.
Conclusions: VA ECMO needs decompression of left cavities to facilitate left ventricular recovery. Performing a PAS with atrial septal stenting could be a safe and effective option in paediatric patients.

PATIENT DEMOGRAPHIC DATA		
Gender	Male	6
	Female	10
Age (years)		6,8 (10 days -15 years)
Weight (kg)		17,5 (2,5-77)
Diagnosis	Myocarditis	8
	Genetic DCM	4
	Idiopathic DCM	4
Outcome	Recovery	8
	Transplant	6
	Death	2
ECMO (days)		18 (7-78)
Time admission-ECMO (hours)		11,5 (3-288)
Time ECMO-stent (hours)		5,8 (3-12,9)

CARDIAC CATHETERIZATION DATA		
Vascular access	Double access	9
	Single access	7
Septal length		28 mm (11-49 mm)
Stent placement		15
Stent	PG 19x10	7
	PG 25x10	1
	CP 34 mm	5
	CP 28 mm	2
Stent diameter		11 mm (10-20 mm)
Endomyocardial byopsy		11
Stent dilation		2
Procedure duration (min)	Stent and byopsy	101 (68-147)
	Stent	67 (29-137)
Fluoroscopy time (min)	Stent and byopsy	34 (17-63)
	Stent	19 (10-43)

O5

Trans-catheter atrial septal defect closure with the new GORE® Cardioform ASD occluder: First European experience

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Introduction: This perspective, observational study evaluated safety and efficacy of the GORE® Cardioform ASD Occluder (WL Gore & Associates, Flagstaff, AZ), compliant and potentially innovative prosthesis recently approved for closure of ostium secundum atrial septal defects (ASD).
Methods: Between January and June 2020, 43 unselected patients with -significant ASD were submitted to transcatheter closure with GORE® Cardioform ASD Occluder at two high-volume Italian Pediatric Cardiology centers. Primary endpoints were procedural success and safety. Secondary endpoints were closure rate and clinical safety at 1-month follow-up.
Results: Patients' age and weight were 8.2 ± 3.9 years (range 3–21, median 9.9) and 29.6 ± 15.3 kg (range 16–57, median 33.3), respectively. ASD diameter was 16.6 ± 4.5 mm (median 10), resulting in QP/QS of 1.7 ± 0.7 (median 1.6). Seventeen pts. (39.5%) were considered "surgical" candidates due to challenging septum morphology, ASD rim deficiency or ASD diameter/patient weight ratio ≥ 1.2. Device placement was successfully achieved in all but one patient (97.7%), in whom it embolized early after deployment, resulting in rescue surgical repair. No cross-over with different devices was recorded. Median procedure and fluoroscopy times were 40 and 6.8 min, respectively. Major adverse events were recorded in 7.0% (3 pts). Complete closure rate was 78.5% at discharge, rising to 100% at 6 months evaluation, without cardiac or extra-cardiac adverse events. "Challenging" procedures were more time-consuming but as effective and safe as the "simple" ones.
Conclusions: The GORE® Cardioform ASD Occluder device was highly effective and versatile in closure of ASDs with different anatomy and size, even in challenging settings.

O66

Transcatheter PAD closure in premature infants: a new technique to ease access to the PAD with particular interest to the tricuspid valve.

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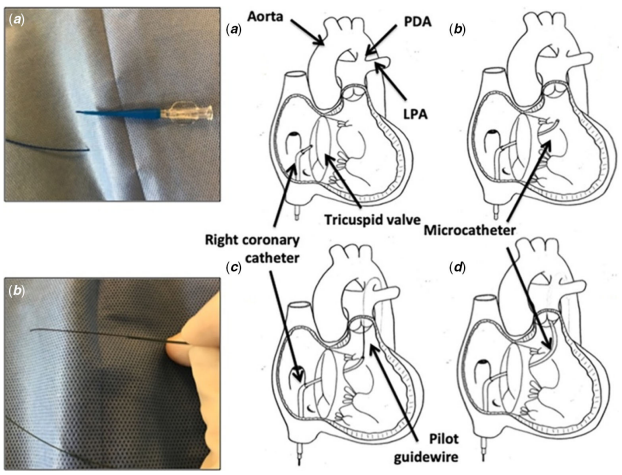
Introduction: Transcatheter patent arterial duct closure (TPAD) in premature infants has been shown to be feasible. Since our early procedures of TPAD closure in premature infants at Necker Enfants Malades hospital, we have changed our technique several times to advance the guidewire through the right heart in order to avoid tricuspid valve damage. We describe our most recent technique and results in a cohort of 33 patients.
Methods: All premature infants below 2 kg who underwent TPAD closure with this new technique were included. Demographic data, procedural data, outcome and procedural complications were reviewed, with particular attention to the occurrence of tricuspid regurgitation.
Results: Between May 2019 and May 2020, 33 patients were included. Median gestational age was 25 weeks. Median birth weight and procedure weight were 690g (IQR 620-785) and 1160g (IQR 1030-1300), respectively. Median age at procedure was 35 (30-46) days. PAD anatomy was evaluated on TTE only. Median PAD diameter was 3 (2.5-3.2) mm. Success rate was 100% (defined as successful closure without residual shunt). One patient had a renal vein thrombosis, which fully resolved with low molecular weight heparin anticoagulation. No stenosis of the left pulmonary artery or of the aorta was seen, or tricuspid regurgitation. One patient died of a superior caval vein obstruction with bilateral chylothorax related to a central catheter thrombosis 56 days after the procedure, unrelated to the catheter procedure.
Conclusions: In this prospective study, we describe a new technique to avoid tricuspid valve damage and facilitate delivery of the PAD device.



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O67

Simplified procedure for percutaneous closure of patent ductus arteriosus in low weight newborns

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Introduction: Transcatheter closure of patent ductus arteriosus (PDA) in extremely low birth weight infants has become an alternative to surgical ligation. Several procedure-related major adverse events have been reported in the literature. We report our experience with a simplified percutaneous approach.
Methods: Between 0october 2017 and November 2020, 70 infants weighing less than 3000 grams attempted device closure of a hemodynamically significant, left-to-right shunting PDA. All procedures were done via an antegrade route, under general anesthesia, transthoracic echocardiography and fluoroscopic guidance. A 4-French Terumo sheath was inserted in the femoral vein under echo-guidance. We used neither heparin nor contrast injection, to minimize the risk of cerebral hemorrhage and kidney injury in this vulnerable population. Tricuspid valve was crossed gently with the 4-French TorqVue^{TM} LP delivery system and a 0.014 floppy wire placed in the descending aorta, allowing placement of either an Amplatzer Piccolo Occluder (APO) or an Amplatzer Vascular Plug-II (AVPII) device according to manufacturer's instructions for use. Device selection was based on echocardiographic measures only.
Results: The mean procedural body weight was 1539±625 grams (extreme 740-3000). APO, AVPII and the new Lifetech Konar-MF^{TM} VSD Occluder were used in 67, 2 and 1 infants, respectively. All devices were successfully implanted, without any residual shunt at day-1 assessment. Procedure-related death, tricuspid valve regurgitation, cardiac tamponade, device embolization or vascular injury were not observed. Mean fluoroscopic time and mean radiation dose were 5.1±8.9min was 43 ±52cGy.cm^2 respectively, that are two to five times lower than reported in previous published reports. Three (4.3%) patients experienced delayed complications represented by severe left pulmonary artery stenosis in 2 patients at 5 and 6 months follow-up, and acquired coarctation of the aorta in 1 patient at 6 months follow-up, all managed by surgical retrieval of the device. Five infants died before hospital discharge because of complications of prematurity; none of these deaths were related to the procedure.
Conclusions: We report a new and simplified transcatheter approach to safely and efficiently close the PDA in low body weight infants. Our technique allows a shorter procedural time with a lower radiation exposure compared to previous reports.

O68

Impact of COVID-19 on Congenital Heart Disease interventional cardiology: a Survey on behalf of Italian Society of Congenital Heart Disease (SICP)

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Introduction: Among all western countries, Italy was the first to be strongly affected by COVID-19 pandemic, and one of the first to apply the "hard" lockdown. In order to limit the in-hospital infections and to re-distribute the healthcare professionals, all healthcare elective activities were reduced or cancelled, and among them, cardiac percutaneous interventions in Pediatric and Adult Congenital Heart Disease (ACHD) patients were limited to urgent or emergent ones. The aim of this paper is to describe the impact of COVID-19 pandemic on Pediatric and ACHD cath lab activity during the so called "hard lockdown" in Italy.
Methods: On behalf of the interventional working group of the Italian Society of Pediatric Cardiology, 11 out of 12 Italian Institutions with a dedicated Invasive Cardiology Unit in Congenital Heart Disease actively participated to the survey. The institutions were distributed over all the national territory, 5 in the northern regions, 3 in the central and 3 in the southern ones. The data from each center were collected using a self-completion questionnaire containing 41 multiple choices questions.
Results: Most of the hospitals were affected by the COVID-19 pandemic, either actively, with direct management of infected patients, or passively due to decrease of routinely clinical activities. The majority of institutions stated a change in the cath lab usual workflow plan and accessibility. Most of the centers had to cease at least temporarily the Cath lab practice, and the overall reduction of procedures number ranged between 50% and 75%. This reduction was more pronounced for teenagers and ACHD compared to neonates and children. Interestingly, there was an evident discrepancy in the management of the lock-down, irrespective of the number of COVID-19 positive cases registered, with higher reduction in Southern Italy compared to the most affected Regions
Conclusions: COVID-19 pandemic has significantly affected the activity of 11 different pediatric cardiology and ACHD units all over Italy. Cath lab services were deeply impacted due to decline

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in outpatient clinic consultations and referrals from local hospitals. ACHD cath lab procedures suffered the biggest drop. However, overall activity reduction was not consistent with the severity of outbreak in the different Italian regions.

Geographic Area	Regions	N° of Units
NORTH	Piedmont	1
	Lombardy	2
	Veneto	2
CENTRE	Emilia-Romagna	1
	Tuscany	1
	Lazio	1
SOUTH	Campania	1
	Puglia	1
	Sicily	1

ACTIVITY	RATE OF REDUCTION			
	Low (0-25%)	Moderate (25-50%)	High (50-75%)	Very High (>75%)
Outpatient Clinic Consultations	0	3 (27%)	2 (18%)	6 (54%)
Hospital Admissions	1 (10%)	3 (27%)	4 (36%)	3 (27%)
Urgent cases referrals	4 (36%)	2 (18%)	3 (27%)	2 (18%)
Cath Lab procedures	0	5 (45%)	2 (18%)	4 (36%)

Patient's age	Level of Cath Lab procedures reduction
Neonatal	Mild-Moderate
Paediatric	Moderate
Teenagers	Severe
Adults	Severe

O69

Ultrasound-guided basilic vein approach for cardiac catheterization and cardiovascular interventions in pediatric population

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Introduction: Femoral and internal jugular venous approaches are used the most frequently by pediatric interventionalists. In some cases, however, these large veins cannot be accessed due to or the anatomy of the lesion requires an alternative venous access. At our institution we decided to use the basilic vein for diagnostic and interventional procedures whenever anatomically possible and technically feasible. The basilic vein is a superficial vein of the forearm and arm that terminates in brachial vein or in the axillary vein thus providing a quick access to the superior vena cava. This study presents our experience with ultrasound-guided basilic approach in the pediatric population.
Methods: Ultrasound-guided basilic vein approach was performed successfully in 50 patients in years 2016-2017. There were 19 female and 31 male patients, aged from 0 to 18 years, weighting from 3 to 75 kg, median 18.1 kg. There were 36 patients with functionally univentricular heart, 12 patients with other congenital cardiac defects, 1 patient with pulmonary hypertension due to liver disease, one with left brachiocephalic vein stenosis. Procedures were performed under general anesthesia. Basilic vein (45 right, 5 left) was punctured in lower 1/3 of the forearm under ultrasound guidance (L17-7io linear transducer, Philips Epiq 7c ultrasound system).
Results: The rationale for basilic vein approach was access to pulmonary arteries in patients after Glenn anastomosis (36 cases), thrombosis of femoral and iliac veins (11 cases), anatomical procedure planning (6 cases), need for accessory venous access (1 case), patient's request to avoid groin vessels (1 case). The sheath size was 4F (24 cases), 5F (19 cases), 6F (6 cases), 7F (1 case). Procedures performed included diagnostic catheterization (48 cases), pulmonary artery balloon angioplasty (10 cases), left brachiocephalic vein angioplasty (3 cases), subclavian vein angioplasty (1 case), systemicto-pulmonary collateral vein closure (1 case), pulmonary allograft angioplasty (1 case) and placement of right ventricle rapid pacing lead (1 case). There were no access-related complications reported.
Conclusions: Ultrasound-guided basilic vein approach provides an attractive alternative for other venous accesses. As a superficial venous access it is safe and easy to control. It allows to perform diagnostic and interventional procedures even in small children.

O70

French multi center survey on percutaneous treatment of native aortic coarctation in children

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Introduction: Percutaneous treatment of native coarctation is a Gold standard in adult patients and in operated children with recoarctation. Less evidence exists for percutaneous treatment of native coarctation in children. We report the French multicenter experience of percutaneous treatment of native coarctation in children
Methods: All the patients with native coarctation who received percutaneous treatment between 1 and 18 years of age were included in this multcentric retrospective survey. 9 centers participated. Demographic data, reason for intervention, procedural data, local and general complications and follow-up data including need for reintervention were recorded.
Results: 133 patients were included. Mean age at diagnosis was 9.3 (0.1 -18 years). Aortic valve was bicuspid in 59 patients and associated lesions were presented in 8. Indication for treatment was hypertension in 109 patients, exercise hypertension in 8 and other reasons in 16 patients. Mean age at cardiac catheterisation was 11.5 +/-3.9 years. Mean weight at intervention was 41 +/-18 kg. Mean invasive peak to peak gradient was 28 +/-27 mmHg. 120 patients had stent implantation. 5 ballon diltation alone and 8 balloon dilatation followed by stent implantation. There were 76 non covered stent used and 44 covered stents implanted. Post intervention mean residual gradient was 3 mmHg. There were 3 associated lesions treatment: 1 PDA closure, 1 aortic valvuloplasty and 1 left SVC occlusion. 8 patients had serious post procedural arterial lesions (3 pseudoaneurysm, 2 dissections, 2 stenosis and 1 occlusion) 2 needed surgical treatment for femoral lesions. 22% of patients required reintervention. 2 had surgical treatment of aortic arch lesion. 28 patients had a second transcatheter intervention: 14 had balloon dilatation and 14 had stent implantation. Risks factors for reintervention or residual hypertension will be presented. 25% of the

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patients remained hypertensive with 15 patients (11%) on anti hypertensive medication. Mean follow-up was 4.9 +/-4.7 years.
Conclusions: Percutaneous treatment of native coarctation in children was safe and sustainable. There were no aortic perprocedural complication. 6% of patients had serious femoral artery lesion with 2 surgical treatment required. Reintervention rate was 22% and consisted in transcatheter intervention in 28 and surgery in 2 patients. 25 % patients remained hypertensive.

O76
Preliminary experience with Optical Coherence Tomographic (OCT) evaluation of pulmonary venous stenosis in children

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Introduction: Pediatric pulmonary venous stenosis is a complex cardiovascular disorder affecting children for which there are limited treatment options. Current treatment strategies include surgical reconstruction and/or re-implantation of the affected vessel as well as catheter based strategies including balloon and stent angioplasty. Evaluation of disease severity and success of treatment is currently limited to cross sectional imaging assessment including echocardiography, computed tomography and invasive angiography. Here we present the first descriptions of Optical Coherence Tomography (OCT) in the evaluation of pulmonary venous stenosis in the pediatric population.
Methods: Institutional Review Board (IRB) approval was sought for permission to evaluate patients using this established technology in this novel substrate. 8 patients were prospectively recruited to OCT evaluation at the time of elective cardiac catheterisation for the evaluation of both native vessel disease as well as post procedural evaluation of balloon and stent angioplasty. Imaging data was acquired using a DragonFly Optis Imaging Catheter via a 4Fr venous sheath or a 5F or 6Fr Guide catheters, using the Abbott Cardiovascular OCT platform. Data was analysed using SPSS.
Results: 8 patients were evaluated using OCT. The median age was 3 years. The median weight was 10.6kg. 3 patients had congenital heart disease associated with pulmonary venous stenosis. 5 patients had pulmonary venous stenosis associated with premature birth and chronic lung disease. All patients had balloon angioplasty and/or stent angioplasty performed and were evaluated with OCT both before and after. Evaluation of cutting balloon angioplasty was also performed. There were no complications of performing OCT. Imaging results are presented and described.
Conclusions: OCT is a novel imaging modality that has limited utility to date in pediatric cardiology practice. This invasive form of intravascular assessment can be carried out at the time of routine cardiac catheterisation for pulmonary venous assessment and intervention. It is a technically feasible catheter based evaluation yielding high definition surface and subsurface anatomy and tissue profiling which presents the opportunity to further understand the underlying disease processes as well as the defining the potential success of various interventions.

O77
Frequency and heatmap distribution of frame fractures at medium-term follow-up of the GORE Cardioform ASD Device

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Introduction: The latest device approved by the FDA for atrial septal defect (ASD) closure is the GORE® Cardioform ASD occluder (GCA). Despite a very low incidence of complications having a direct impact on patient wellbeing, a significant amount of Frame Fractures (FFs) have been observed in the GCA at short to medium-term follow-up, being described in almost one-third of the devices implanted during clinical trials. Here we evaluate the frequency and characteristics of Gore Cardioform ASD Device Frame Fractures and explore potential associations in patient and procedural practice variables.
Methods: We performed a retrospective single-center chart review of 54 patients who received a GCA between January 2017 and December 2019, and selected 35 patients in whom a 6-months fluoroscopic follow-up was performed. We evaluated the number and type of fractures based on the fluoroscopic assessment and devised a simple nomenclature system based on three-dimensional (3D) reconstruction of the device's fluoroscopic images. We also evaluated the association between the positions and number of fractures and patient and procedural characteristics.
Results: The FF incidence in our population was 42.9% at a 6-months follow-up, and the factors associated with these events were the size of the defect and the size of the device implanted. FFs were more frequently located in a central position in the right atrial disc, and by 3D evaluation, the most common anatomical position for fractures was the anteroinferior quadrant of the right atrial disc. At the time of manuscript submission, no echocardiographic or clinical sequelae were noted in our population.
Conclusions: The incidence of fractures in our population was higher than the observed in published literature, albeit using more intense image scrutiny than that required other cited studies. We noted that the central portion of the anteroinferior quadrant of the right atrial disc was the most susceptible to FF and found that the size of the defect and the device were the only statistically associated factors.

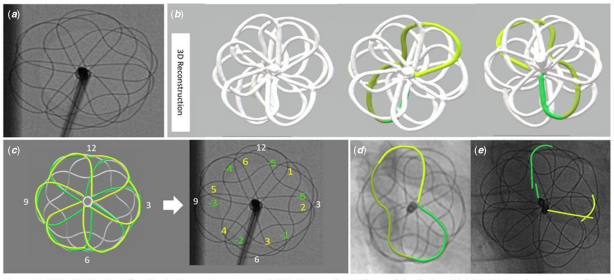

Image 1. Wire distribution after the deployment process, and delimitation of atrial faces of the device. (A) A 3D rotational angiogram of the device in a plastic support system during the deployment process to mimic anatomical conditions was performed. (B) A 3D reconstruction of the rotational angiogram was performed, evaluating the wire distribution sequence between the conformed "discs," determining the right atrial (light green) and the left atrial (dark green) discs, and defining which wire segments belonged to which side. (C) Once the distribution of the different wire segments was confirmed, a virtual 3D model was allowed us to translate these findings into a number system that can be used to determine the atrial position of the fractures in a 2D view. (D-E) examples of the 3D position of wires and fractures on a 2D view. All these processes were made based on the right atrial view as the baseline observations.

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O78

The Gore ASSURED Clinical Study: Pivotal + Continued Access Results in 568 patients Treated with the Gore Cardioform Atrial Septal Occluder

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Introduction: Previously-approved ASD Closure devices have known limitations. Device erosion has been associated with the AMPLATZER®Septal Occluder in patients with retro-aortic rim deficiency (<5mm), while defects ≥18 mm are too large for the GORE®CARDIOFORM Septal Occluder. The GORE®CARDIOFORM ASD Occluder (GCA), a hybrid of the approved devices, was designed to expand the eligible ASD population. The purpose of the Gore ASSURED Trial was to assess clinical performance of the GCA for transcatheter closure of atrial septal defect (ASD). We present the data from the Pivotal phase and Continued Access phase (CAP) of the ASSURED trial.
Methods: One hundred twenty-five ASD patients were enrolled in a prospective, multicenter registry, and 443 patients were enrolled in the CAP. Descriptive clinical endpoints included: Technical Implant Success, Closure Success, Serious Adverse Events (SAE), Clinically Significant New Arrhythmia, and Wire Frame Fracture. Procedural outcomes and adverse events were adjudicated by an Echocardiography Core Lab and Independent Data Review Board respectively.
Results: PIVOTAL TRIAL RESULTS: Median subject age was 12.3 years (range 2.9 - 84.7), with 72% of patients ≤ 18 years old. Median ASD stop-flow diameter was 17.0 mm (8.0 - 30.0), with 43% ≥18 mm. 57% of subjects had a deficient retro-aortic rim. 30% had both diameter ≥18mm and deficient rim. 120/125 (96%) had Technical Implant Success, though 3 devices were removed within 24 hours. 112/117 returned for the scheduled 6-month evaluation. All 112 had Closure Success. 6/125(5%) had SAE. 6/125 (4.8%) had Clinically Significant New Arrhythmia.Wire Frame Fractures occurred in 37/104 (36%), without any associated clinical sequelae, residual shunt or device instability. ****** The 6-month results from the CAP of the trial will be available for presentation in time for the AEPC meeting******
Conclusions: The GCA performed effectively and safely in the Pivotal Trial cohort, and led to FDA approval (and CE Mark) based on this data. The CAP data (443 additional patients) will be available for presentation at AEPC 2021.

O79

Wire frame fractuers after percutaneous closure of atrial septal defect with GSO device

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Introduction: Percutaneous closure ofatrial septal defect (ASD) is a widespread procedure with high chance of success. In this study we evaluated the effectiveness and the incidence of wire-frame fracture (WFF) in mid-term follow up after transcatheter closure of PFO and ASD with the GORE Cardioform Septal Occluder (GSO) in pediatric patients.
Methods: Single-center, prospective mid-term follow-up study of all children and adolescents with a GSO (≥12 months). Periprocedural data and follow-up data were evaluated including clinical, echocardiographic findings and X-rays fluoroscopy to assess the GSO's wire-frame morphology.
Results: Seventy-seven consecutive patients were enrolled with mean age of 10 ☐ 3.9 and mean weight of 37.7 ☐ 17.3 kg at implantation. ASD anatomy included 8 PFO, 60 single and 9 multi-fenestrated defects. Nine patients had small retro-aortic rims (≤3 mm). Follow-up period was 3.1 ☐ 1.3 years. Fluoroscopies were available in 60 children. WFF was detected in 1 GSO 20 mm (8 analyzed-12.5%), 6 GSO 25 mm (26 analyzed -23%), 15 GSO 30 mm (26 analyzed -58%). Three fractures (all in GSO 30 mm) compromised the outer perimeter of the device. Incidence of fracture in GSO 30 mm was higher ($p = 0.001$). A multivariate analysis including patient age, ASD diameter, ASD stretched diameter, GSO size and ASD/GSO ratio confirmed that GSO diameter is the independent factor related to device fracture ($p = 0.013$; $F = 6.7$). Devices appeared stably fixed and well aligned to both sides of the atrial septum. All patients were all asymptomatic at follow up. Residual trivial shunt was present in 4 patients (5%). No further treatment was required.
Conclusions: GSO is safe and effective for ASD closure and it seems to offer reliable mechanical durability. Device diameter strongly correlates with incidence of WFF, without any clinical sequelae during follow up.

O80

Initial Management of Pulmonary Atresia/Intact Ventricular Septum in a Developing Country: Valve Mechanical Perforation in the Era of CTO Hardware

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Introduction: The heterogeneous morphology and hemodynamic implications of pulmonary atresia with intact ventricular septum (PAIVS) keep on challenging pediatric cardiologists in clinical decision-making. In selected patients with favorable anatomy for biventricular repair, transcatheter valve mechanical perforation (TVMP) is an acceptable yet challenging alternative to radiofrequency to establish right ventricle decompression and antegrade flow. We aimed to evaluate and compare the safety, feasibility, and efficiency of two TVMP techniques.
Methods: Clinical data of neonates with PAIVS who underwent an attempt for TVMP between 2009 and 2019 were retrospectively reviewed. Included neonates had membranous total atresia, well-developed infundibulum with no RV coronary artery dependent circulation, and were considered likely candidates for biventricular repair. Patients were divided into two groups according to perforation technique: using the stiff end of a percutaneous transluminal coronary angioplasty (PTCA) ordinary 0.014" wire (group A) and subsequently with the floppy tip of a chronic total occlusion (CTO) guidewire (group B). The technical aspects, procedural and discharge outcomes of both groups were compared.
Results: A total of 19 antegrade TVMP procedures (Group A, n=10, and Group B, n=9) were attempted in 18 neonates with an overall success rate of 73.7% and no procedure-related mortality. Groups' analysis showed that the introduction of CTO hardware maximized procedure success rates ($p=0.002$) with zero failure and misperforations ($p=0.022$). The significant drop in perforation time ($p<0.001$) and irradiation ($p=0.006$) allowed additional ductal stenting during the same setting, optimizing

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patients' outcomes and shortening overall hospital stay. Discharged patients had room air mean oxygen saturation of 91.4% (± 5.5) with no evidence of heart failure.
Conclusions: In selected cases of PAIVS, TVMP using CTO wires is a safer, highly efficient, and simplified alternative to other mechanical perforation techniques. It substantially revolutionized the management of PAIVS in our center optimizing short-term prognosis

Table 1. Group comparsion, n=19

		Total n=19	Group A n=10	Group B n=9	p-value
TVMP success	*N (%)*	14 (73.7)	5 (50.0)	9 (100.0)	**0.022[a]**
TVMP-related complications	*N (%)*	5 (26.3)	5 (50.0)	--	**0.022[a]**
Perforation time (min)	*Median (range)*	17 (4-120)	45 (15-120)	10 (4-18)	**<0.001[b]**
Overall SI-SO (min)	*M ± SD (range)*	123.1 ± 51.1 (50-240)	129.4 ± 45.2 (80-240)	116.7 ± 58.4 (50-215)	0.611[c]
Overall fluoroscopy time (min)	*M ± SD (range)*	35.3 ± 18.7 (7.1-65)	46.5 ± 12.3 (23.2-59.5)	24.1 ± 17.7 (7.1-65)	**0.006[c]**
LOS from perforation attempt date (days), *n=16*	*Median (range)*	6 (3-17)	12 (3-17)	5 (3-9)	0.121[b]
Intrahospital mortality, *n=18*	*N (%)*	2 (11.1)	1	1	--

TVMP= Transcatheter valve mechanical perforation; SI-SO= sheath in-to-sheath out; LOS = length of stay.
Group A: TVMP with the stiff end of a percutaneous transluminal coronary angioplasty (PTCA) ordinary 0.014" wire;
Group B: TVMP with the floppy tip of a 0.014" chronic total occlusion (CTO) wire.
[a]: Fisher's exact test; [b]: Mann-Whitney U test; [c]: T-test
Bold values are significant p-values

8. Surgery and Intensive care

O16
35 Years Follow up of Survival and Re-Intervention of Valved Conduits Implanted in Patients with Congenital Heart Disease.

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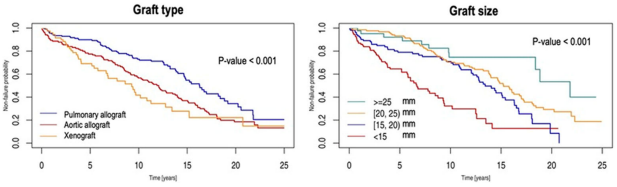
Introduction: The aim of this study was to evaluate long-term survival and risk factors for re-intervention in conduits used for right ventricular outflow tract reconstruction in the young.
Methods: The study included 468 consecutive patients who underwent first valved conduit implantation between 1981 and 2016 at a median age of 2.9 years and were followed-up for up to 35 (median 12.2) years. A total of 673 conduits were implanted during the primary and re-implantation procedures. Valved conduit types and numbers were recorded. The endpoints were death from any cause and graft failure defined as need for re-intervention, respectively.
Results: Total follow-up was 5665 patient-years. Overall mortality was 17.7% (83 patients) with a patient's survival rate of 83.8, 82.0, and 79.4 % at 10, 15, and 20 years after first conduit implantation. Early surgical era (P<0.001) and the number of previous graft failures (P<0.001) were significant independent predictors of death. During the observation period, a total of 264 conduit re-interventions were necessary for 216 (46.2 %) patients. Freedom from first conduit failure differed significantly by graft type (pulmonary allograft superior to aortic allograft or xenograft, P=0.006) and graft size (P<0.001). Risk of any implanted graft failure was 1.54 higher in case of a reimplanted graft as compared to the primary conduit (P=0.015).
Conclusions: In young patients with congenital heart disease, valved conduits provide satisfactory long-term results. Better initial conduit survival is associated with larger graft size and with the use of pulmonary allografts as compared to aortic allografts and xenografts. Repeated conduit implantations are risk factor for both death and worse graft survival.

Supported by grant MH CZ - DRO, 00064203

Figure: Freedom from conduit re-intervention.



O17
Histological analysis of subaortic obstruction in neonates with mitral stenosis, coarctation and ventricular septal defect in Shone Complex

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Introduction: The morphological spectrum of Shone Complex is well described. In his original publication, Shone described the protrusion of subaortic muscular tissue ("bulge") of the left ventricular outflow tract, or "sub-aortic stenosis" respectively in all of his eight patients. Yet it is still unknown why some of the patients develop subaortic obstruction. Moreover the origin of this muscular "bulge" is unclear.
Methods: Six heart specimens with muscular subaortic obstruction besides mitral valve stenosis, coarctation of the aorta and ventricular septal defect were identified in the anatomical collection of the German Heart Center (1974-1992). Histological analysis was performed in transmural biopsies of the subaortic "bulge" in the superior-anterior left ventricular outflow tract, and left ventricular posterior wall, as well as of the anterior right ventricular wall. Two biopsies of the infundibular septum from patients with Double outlet right ventricle (DORV) served as controls.
Results: Mean age at death was 15 days (5 to 33 days), and of the controls 30 and 40 days, respectively. All myocardial specimens showed unspecific regressive changes and intercellular fibrosis of varying degree. In addition, the muscular subaortic stenotic tissue biopsies from patients with Shone Complex demonstrated marked disarray of cardiomyocytes. By contrast, both control specimens showed regular arrangement of the muscular tissue.
Conclusions: Our results of the histological analysis of subaortic obstructive muscular tissue reveal the pattern of hypertrophy as demonstrated in hypertrophic cardiomyopathy, and opposed to the regular arrangement of cardiomyocytes in two controls. The latter findings confirm that the subaortic "bulge" in patients with Shone complex is not a displaced infundibular septum. Further research including embryologic studies are necessary to elucidate the origin of the muscular subvalvular obstruction in patients with Shone complex.

O18
Is Earlier Repair of Total Anomalous Pulmonary Venous Connection (TAPVC) associated with better outcome?

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Introduction: Total Anomalous Pulmonary Venous Connection (TAPVC) is a rare but heterogeneous anomaly, characterized by failure of the pulmonary veins to connect to the left atrium.

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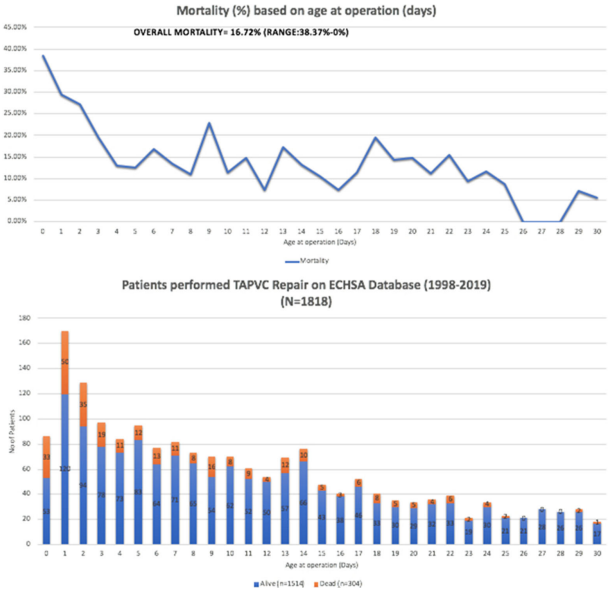
The degree of pulmonary venous obstruction (PVO) determines the clinical presentation and the necessary surgical management. Unobstructed TAPVC is typically asymptomatic, may be diagnosed weeks or months after birth, and can be dealt electively, but, usually these patients undergo repair soon after diagnosis. TAPVC with high grade anatomic obstruction presents at birth with catastrophic symptoms of severe hypoxemia and circulatory collapse, unresponsive to stabilizing maneuvers, necessitating "super urgent" surgery. Most neonates present with significant but variable degrees of PVO. The aim of this study is to define the relationship of precise timing of surgery with hospital mortality in neonates.
Methods: The ECHSA Congenital Database was interrogated to identify all patients who have undergone TAPVC repair as primary procedure between 12/1999 until 12/2019. Demographic data, age at operation, type of TAPVC, associated risk factors, operative variables and outcome were retrieved and analyzed.
Results: For all 3881 patients, with TAPVC identified, repair at mean age 9.38 months was associated with hospital mortality (HM) of 10.02%. For the neonatal subgroup of 1818 patients, operated at mean age 10.9 days, HM was 16.72%. Mortality varied significantly according to day of life at operation, from 38.4% on day 0 (4.73% of all neonates) to 7.62%, during days 20-30 of life (16.9% of neonates).
Conclusions: Despite the standard recommendation for immediate surgery for neonates diagnosed with TAPVC, in reality, this occurs only in a small percentage of patients. Most neonates are diagnosed and operated later, with mortality diminishing with advancing neonatal age (see figure). Early operation seems to be a marker for more severe disease. Analysis of other risk factors is underway.



O19

Long-term outcome after single ventricle palliation: A Swedish nationwide cohort study

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Introduction: To examine long-term survival after surgery for univentricular heart defects.
Methods: All children in Sweden operated due to univentricular heart defects before 18 years of age from 1994 to 2019 were included. Medical records were analyzed and survival cross-checked against the National Population Registry allowing for reliable and complete follow-up. Primary outcome was all-cause mortality.
Results: 778 patients (39% female) had univentricular heart surgery. Mean follow up time was 11.3 ± 8.4 years (range 0-26.7 years). 10 patients (1.3%) were lost to follow up due to emigration. 176 deaths (22.6%) occurred. Main diagnoses were; hypoplastic left heart syndrome 268 patients (34.4%), tricuspid atresia 107 (13.7%), double inlet left ventricle 90 (11.5%), atrio-ventricular septal defect 89 (11.4%), double outlet right ventricle 73 (9.4%), pulmonary atresia with intact ventricular septum 60 (7.7%), and miscellanous defects 91 (11.7%). 83 (10.7%) patients had an extracardiac defect, syndrome and/or chromosomal aberration. Atrial isomerism was diagnosed in 56 patients (7.2%). Right ventricular dominance was found in 432 (55.5%) patients, left in 303 (38.9%), biventricular in 40 (5.1%) and indeterminate morphology in 3 (0.4%). Stage I palliation was performed in 716 patients with 30-day mortality in 53 (7.4%) and late mortality in 60 (8.4%). Bidirectional cavopulmonary connection was performed in 645 patients, with 30-day mortality in 3 (0.5%) and late mortality in 26 (4%). Total cavo-pulmonary connection was performed in 547 patients with 30-day mortality in 1 (0.4%) and late mortality in 23 (4.2%). Survival at ten years of age in patients with right ventricular dominance was significantly lower as compared to patients with left or biventricular dominance (65.5% vs 89.4%, $p<0.001$), but similar in patients with and without atrial isomerism. At follow-up, 11 patients were alive with stage I palliation, 50 with bidirectional cavo-pulmonary connection and 508 with total cavo-pulmonary anastomosis. Heart transplantation was performed in 28 patients (3.6%).
Conclusions: Overall long-term survival was 77.4% with a significantly higher survival in patients with left ventricular dominance as compared to patients with right ventricular dominance (89.4% vs 65.5%, $p<0.001$). Heart transplantation was performed in 3.6% of all patients.

O20

Impact of performing superior cavopulmonary connection at 3 months rather than 6 months in patients undergoing Norwood palliation

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Introduction: There is increasing evidence that performing the superior cavopulmonary connection (SCPC) at 3 months reduces mortality by shortening the at risk 'interstage' period. It also off-loads the ventricle and reduces coronary steal thereby hopefully preserving ventricular function. This study reviews the change to early SCPC at our institution that occurred as part of a review of our Norwood programme.
Methods: Patients who had undergone the Norwood procedure (NP) from 01/01/2005 to 01/12/2019 were included and divided into two eras: standard SCPC (era 1, 2005-2014) and early SCPC (era 2, 2015-2019). Demographics, mortality (interstage, early and late post-SCPC) and data on post-operative course and

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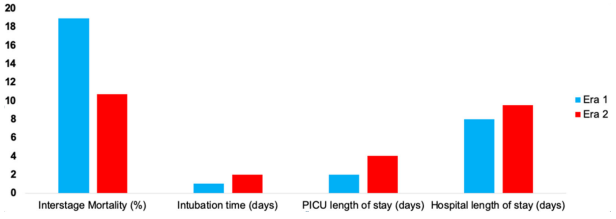
complications were recorded. Results are expressed as median (interquartile range) and mean (standard deviation) as appropriate.
Results: In era 1, 174 patients underwent NP with 107 survivors to SCPC and 33 patients with 25 survivors in era 2. There were no significant differences in the demographics of the patients undergoing NP. Interstage mortality was less in the second era but not statistically significant (18.9% vs 10.7%). The median age in days at pre-SCPC imaging and SCPC was significantly different in the two eras ($p < 0.005$): 105 (89-124) vs 71 (52.3-82.5) and 178.5 (141.8-212.3) vs 109 (95.5-121) and patients were significantly lighter at SCPC ($P<0.05$): 6.36kg (1.25) vs 5.0kg (0.7). Intubation time and PICU stay were statistically longer (but probably of limited clinical significance): 1 (1-2) vs 2 (1-3) days and 2 (3-3.5) vs 4 (3-6) days respectively ($P<0.05$). Overall hospital stay was not significantly different between the two groups. There was no significant difference in early or late mortality, nor in the rates of diagnostic or interventional catheter, post-operative MRI/CT imaging or stroke.
Conclusions: Early review of our change in practice has seen a fall in interstage mortality from 18.9% to 10.7%. Although not statistically significant we feel this is an important gain in this high risk group, and although there are minor increases in ventilation and PICU stay, there is no increase in post-SCPC mortality or complications. This has given us confidence to continue this strategy and we will continue to monitor this group.



O31
Major aorto-pulmonary collateral arteries after arterial switch operation in transposition of the great arteries: impact and management

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Introduction: Major aorto-pulmonary collateral arteries (MAPCA) are frequent in patients with simple transposition of the great arteries (TGA). However, only few studies describe post-operative management. We analyzed predictive factors of hemodynamically significant MAPCAs (hs-MAPCA), their clinical impact and we reported treatment and outcome after embolization of these collaterals.
Methods: All neonates admitted with simple TGA or TGA with isolated VSD, between January 2014 and December 2019 in our center, were retrospectively reviewed and divided in 2 groups according to the presence of hs-MAPCAs. Demographic data at birth, surgical data, pre and postoperative data, the need for post-operative catheterization, and short-term outcome were reported.
Results: We included 222 patients. Twenty-six had hs-MAPCA diagnosed after arterial switch operation (ASO) on TTE and confirmed on angiography. All of them were successfully embolized, with a mean of 1.9 +/- 0.77 MAPCA closure. Mean delay between ASO and catheterization was 6.4 +/- 6.2 days. There were no complications after embolization. Average time between catheterization and extubation was 2.9 +/- 3.3 days. Birth weight, gestational age and prenatal diagnosis rate were similar in the two groups. There were no higher need for an atrioseptostomy ($p=0.82$) nor prostaglandin infusion ($p=0.56$) in the MAPCA group. Mean age at ASO was 6.1 +/- 2.3 days in the MAPCA group and 6.7 +/- 9.8 days in the other group ($p=0.78$). Mean by-pass time was similar in both groups: 134.9+/-22 minutes in the MAPCA group vs 136.4 +/- 39.3 minutes in the other group. There was significantly increased pulmonary venous return (noticed by the surgeon) during ASO (55% vs 7%, $p<0.001$), and higher left arterial pressure (LAP) in the MAPCA group (13.1+/-4.3 vs 8.7+/-2.5 mmHg). The MAPCA group had a more complex postoperative course, with a longer mean ICU stay (13.2 vs 6.6 days, $p=0.082$), mean duration of inotropic support (8.9 vs 5.1 days, $p=0.036$) and mean hospital stay (24.2 vs 14 days, $p=0.017$).
Conclusions: We found no predictive factor in preoperative course to identify patients with hs-MAPCA. However, increased pulmonary venous return during ASO and high LAP may help to early diagnose hs-MAPCAS, and treat them as quickly as possible in order to ease postoperative course.

O32
Outflow tract rotation for complex transposition of great arteries, review of a single center series.

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Introduction: When transposition of the great arteries (TGA) is associated with ventricular septal defect (VSD) and pulmonary valve stenosis (PS), Rastelli or Bex-Nikaidoh (posterior aortic translocation) procedures are usually performed. Complications include right and left ventricular outflow tract (LVOT) obstruction and necessity for further pulmonary conduit replacement. A modification of a novel surgical technique, initially described by Yamagishi, currently known as outflow tract rotation (OTR), has been used at our center in selected cases. OTR allows for conservation of both native valves, replaced in their physiological position by a half-turn rotation of the truncal block. We sought to report our experience with this technique.
Methods: We retrospectively reviewed all patients that underwent OTR at our center, performed by the same surgeon, and described their characteristics and outcome.
Results: Patients characteristics and outcome are included in Table 1. A total of 13 patients, aged 4 days to 11 years, underwent OTR between 2016 and 2020. Pre-operative patient selection was performed in multidisciplinary cardiology team after echocardiography, and CT or MRI 3D reconstruction, and included: TGA/VSD/PS anatomy, TGA with inlet VSD, and TGA with major

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root size discrepancy. Intra-operative anatomy was carefully inspected to confirm the adequacy of OTR. Pulmonary valve (PV) was conserved in 10 patients. Two patients with inlet VSD required pacemaker implant for complete atrioventricular block. Short and medium term outcome were excellent, with no mortality, and no need for reoperation after a mean follow up of 18,7 months.
Conclusions: OTR is the most physiological surgical option in TGA/PS/VSD, when PV is unsuitable for arterial switch but acceptable in the pulmonary position. It allows for a free and straight LVOT, wall growth potential of pulmonary annulus, without the need for conduit replacement. It seems to be a safe and reproducible technique in expert hands, with excellent short and medium term outcomes. Further studies are needed to assess long term outcome, but prospects are encouraging considering excellent results of Tetralogy of Fallot with preserved dysplastic pulmonary valve with mild stenosis and/or regurgitation. Application of OTR to other situations, including TGA with great discrepancy in aortic and pulmonary root sizes, warrants further research.

Table 1 : Demographics, diagnosis, short term outcome and follow up of our serie of patients who underwent outflow tract rotation procedure

Patient n°	Age, months	Weight, kg	Diagnosis	XCT, min	CPB, min	Perioperative Complication	ICU LOS, days	Follow up, months	Outcome
1	27	9.4	TGA/VSD/PS	128	210		4	26	Alive, no reintervention
2	48	11.8	TGA/multiple VSD/PS	180	285		7	35	Alive, no reintervention
3	2	3.4	TGA/VSD/PS	150	180		10	48	Alive, no reintervention
4	81	18.0	TGA/VSD/PS	150	300		4	44	Alive, no reintervention
5	< 1	3.8	TGA/VSD/PS	150	185		10	12	Alive, no reintervention
6	< 1	3.2	TGA/VSD/PS/ TV straddling	135	181		40	25	Alive, no reintervention
7	120	18.9	DORV/side by side GV/ VSD/sub PS	155	174	AVB, PM implantation	15	13	Alive, no reintervention
8	10	4.0	DORV/inlet VSD	160	210		25	11	Alive, no reintervention
9	19	10.0	TGA/VSD/PS	130	156		4	10	Alive, no reintervention
10	75	19.0	DORV/TGA/VSD/PS	155	230		9	2	Alive, no reintervention
11	108	29.8	DORV/TGA/inlet VSD/PS	174	224	AVB, PM implantation	11	13	Alive, no reintervention
12	7	4.2	DORV/VSD/TV straddling/ root size discrepancy	190	225		8	2	Alive, no reintervention
13	132	25,40	DORV/VSD	153	170		8	2	Alive, no reintervention

AVB : atrioventricular block, CPB : cardiopulmonary bypass, DORV : double outlet right ventricle, GV : great vessels, LOS : length of stay, PM : pacemaker, PS : pulmonary stenosis, TGA : transposition of the great arteries, TV : tricuspid valve, VSD : ventricular septal defect, XCT : cross-clamp time

O33

On the importance of discrimination between critical and non-critical aortic stenosis when comparing results after treatment in the neonatal period

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Introduction: Comparison of results after surgery or catheter treatment of isolated valvular aortic stenosis in the neonate is often difficult due to different inclusion criteria as to the definition of critical valvular aortic stenosis. We studied long-term survival and reinterventions in a complete national cohort using strict criteria when defining critical and non-critical aortic stenosis.
Methods: All neonates in Sweden with isolated valvular aortic stenosis treated before 30 days of age, from Jan 1st 1994 to Dec 31st 2018 were included. Patient files were analyzed, and survival was cross-checked against the Swedish National Population Registry as of Nov 15th 2019 allowing for reliable and complete data. Diagnosis was confirmed by reviewing echo-studies and reports. Critical valvular aortic stenosis was defined as valvular stenosis with duct-dependent systemic circulation or depressed left ventricular function (fractional shortening ≤ 27%). Indication for treatment of non-critical stenosis was Doppler mean gradient >50mmHg. Primary outcome was all cause mortality or heart transplantation (Htx), secondary outcomes were need of reintervention and aortic valve replacement.
Results: 107 patients were identified (85 boys, 22 girls). Median gestational age was 40 weeks (26-42). 65 patients had critical stenosis and 42 non-critical stenosis. All medical files were retrieved, with no patient lost to follow-up. Median follow-up time was 13.5 years. Initial treatment was surgical valvotomy (95), primary Ross (2), balloon dilatation (8) and closed transventricular valvotomy (2). Median age at treatment was 6 days (0-26). No 30-day mortality occurred, and long-term transplant-free survival was 93.5%. In the critical group six patients (9%) died or had a Htx, vs one (2%) in the non-critical group. Event free survival was 43% vs 69% (p=0.002), in critical vs non critical stenosis. Median time from the initial treatment to reintervention was 3.6 months vs 3.9 years respectively (p=0.008).
Conclusions: Surgical valvotomy as initial treatment of isolated valvular aortic stenosis in the neonate can be performed with excellent results even in critical forms. However, the results remain better for patients without duct dependency and/or LV depression which underlines the importance of using strict definitions of critical vs non-critical stenosis when comparing different treatment strategies.

O34

modified Blalock Taussig shunt versus restrictive right ventricle to pulmonary artery connection in tetralogy of Fallot patients

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Introduction: We aim to compare early, and midterm outcomes of Tetralogy of Fallot (TOF) and TOF with pulmonary atresia (TOF-PA) who underwent prior palliation in the first year of life using modified Blalock Taussig Shunt (mBTS) or palliative right ventricle to pulmonary artery connection (RVPAc).
Methods: We performed a monocentric retrospective review of 198 patients (93 TOF, and 105 TOF-PA) who had prior palliation between 1996 to 2016. Patients with Major aortopulmonary collaterals were excluded. Median follow-up was 11.0 [CI95%:5.8-16.2] years after palliation.
Results: Ninety-five patients underwent mBTS, and 103 had an RVPAc. Nakata index before palliation was 128.5 (80-169.5) mm2/m2 and 100 (71-146) mm2/m2 respectively in mBTS and RVPAc groups (p=0.001). Weight at palliation was 3.01 (2.7-3.4) kg and similar between mBTS and RVPAc patients (p=0.1). The complete repair was conducted on 175 patients. Among them, 23 mBTS vs 16 RVPAc (p=0.004) were repaired using a pulmonary valve conduit. Eleven patients died before the repair (5.7%) with no differences between both groups (p=0,43). Interstage reintervention rate was 37 (18.7%) with no difference regarding the type of palliation (p=0.5). The delay

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between the palliation and the repair was shorter in RVPAc compared to mBTS (10.7 (6.1-31) months versus 6.5 (4.8-9.9) months (p<0.01).
Conclusions: RVPAc provide a better and faster pulmonary artery growth in our population and led to limit the use of RVPA conduit at the repair. We found no significant differences between mBTS and RVPAc for mortality and interstage reintervention rate.

O35
Long-term survival and freedom from coronary artery reintervention after arterial switch operation for transposition of the great arteries.
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Introduction: The aim of this study was to compare long-term survival after arterial switch operation (ASO) for transposition of the great arteries and Taussig-Bing anomaly with the background general population and to evaluate freedom from coronary artery reinterventions with emphasis on coronary artery patterns and anomalies.
Methods: This single-centre nation-wide retrospective study included consecutive children who underwent ASO between 1990 and 2016. Long-term outcomes were obtained by cross-mapping individual data from the institutional database with the National Death Registry and the National Registry of Cardiovascular Interventions for adults. To compare overall survival after ASO with the background population, a control group of children matched for sex and age was randomly retrieved at a 1:10 ratio from the National Death Registry.
Results: A total of 605 patients underwent ASO. Early mortality was 3.3% and late mortality 1.7% during a median follow-up of 10 (IQR 5 – 16) years. The probability of overall survival at 15, 20 and 25 years after ASO was 94.9%, 94.9% and 94.9% as compared to 99.7%, 99.5% and 99.5% survival in the background population (HR 15.6, 95% CI, 8.9 – 27.5, p < 0.001). The only independent multivariable predictors of a worse overall survival rate were an intramural coronary artery (HR, 5.2; 95% CI 1.8 – 15.2, p = 0.002) and the period of ASO 1990 – 1999 (HR, 4.6; 95% CI 1.5 – 13.6, p < 0.001). Fourteen patients (2.3%) required 16 coronary artery reoperations. Freedom from coronary artery reintervention at 10, 15, 20 and 25 years after ASO was 98%, 97%, 96% and 96%, respectively. The only independent multivariable predictor associated with a higher hazard for coronary artery reintervention was an intramural coronary artery (HR, 33.9; 95% CI 11.8 – 97.5, p < 0.001).
Conclusions: Long-term survival after ASO is excellent. The only independent predictors associated with worse survival were an early surgical period and the presence of an intramural coronary artery. Coronary artery reinterventions are rare at 25 years after ASO. The presence of an intramural coronary artery was the only independent predictor associated with a higher risk of coronary artery reinterventions, regardless of the surgical period.

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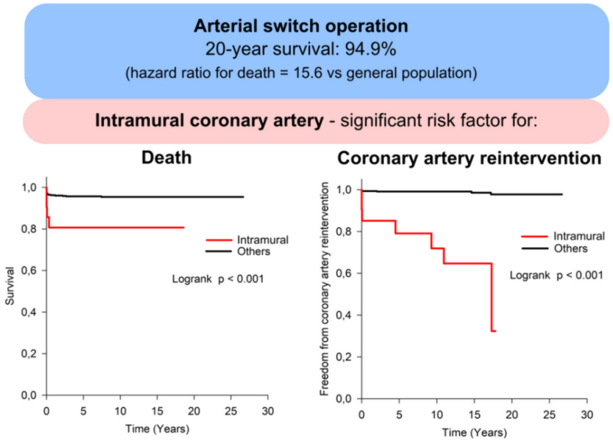

9. Heart failure

O22
Cardiovascular disorders and congenital portosystemic shunt, an unrecognized association: Report on a large paediatric series in a tertiary center.
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Bicetre University Hospital (1), Institut Mutualiste Montsouris (2), Swiss Pediatric Liver Center, University Hospital Geneva (3)

Introduction: Congenital portosystemic shunts (CPSS) are rare and underdiagnosed abdominal vascular malformations. Their relationship with cardiovascular (CV) disorders are poorly reported. The objectives of this study were to describe CV disorders associated with CPSS in a large cohort in order to propose adequate diagnosis and management.
Methods: Among 168 children referred for a CPSS between 1996 and 2019, those presenting at least one clinically significant CV disease including congenital heart diseases (CHD), heart failure (HF), pulmonary hypertension (PH) and/or hepatopulmonary syndrome (HPS) were included. CV disorders were detected using echocardiography complemented by right-sided heart catheterization and/or contrast-enhanced echocardiography and/or lung perfusion radionuclide when appropriate.
Results: 46/168 children with CPSS (27,4%) had one or more CV disorders. CPSS consisted in 7 malformative ductus venosus, 22 extrahepatic shunts, 11 intrahepatic shunts and 6 miscellaneous; 36 patients required endovascular or surgical CPSS closure, including 24 for CV indications. CHD was present in 28 patients (16,6 %); 7 had heterotaxy (4,1%). HF occurred in 21 neonates, diagnosed prenatally in 7, including 13 with CHD. Nine presented with transient PH and/or respiratory distress, mainly related to CHD. HF decreased after cardiac surgery in 10 with CHD but also spontaneously before the age of 3 years in 8 except one without CHD. In 11 patients (mean age :11±5.2 years-old) CPSS were diagnosed as part of a work up for fixed PH (mean pressures 46 ±15.1mmHg, pulmonary vascular resistances 7±2.5UW/m^2).

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One of them died before CPSS closure, while PH remained stable despite CPSS closure in 10 patients, of which two then required lung transplantation. In 6 patients (median age :5.5 years-old), hypoxia (finger pulse oximetry : 70 to 95%) revealed a HPS which resolved within 2 years of CPSS closure.
Conclusions: CV disorders are frequently associated with CPSS including CHD with/without heterotaxy. HF is found in fetuses and neonates, regressing in most cases. Later, HPS and PH may develop. After CPSS closure, HPS is reversible while PH may only stabilize. Perinatal HF, complex CHD, heterotaxy, unusual course of CHD, HPS and PH should raise the possibility of CPSS. Conversely CV complications should be searched for in patients with CPSS.

10. Psychosocial

O36
A controversial topic: HrQoL in twins with congenital heart defect and without- First preliminary results of the "Same Same, but different?" project

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Introduction: Health-related quality of life (HrQoL) is a topic on which contradictory literature in the field of children with congenital heart defects is existing. This may be because this construct is dependent on various external influences such as socioeconomic aspects, environmental and family influences the children live. To exclude these influences, this study aims to determine HrQoL in children with congenital heart defects and their twin sibling without.
Methods: Out of the "Same Same, but different?" project 27 pairs of twins (32 girls (59%), 9.8±4.4 years, one with congenital heart defect and the other without) filled in the KINDL-R questionnaire measuring the self-reported health-related quality of life (HrQoL) in children. The KINDL-R consists of 6 sub-dimensions and a total HrQoL score which are calculated to scores between 0-100 (the higher the better). Statistical analyses were conducted using paired-sample student's t-test by using the software SPSS V.25 (SPSS Inc., Chicago, Illinois, USA).
Results: The directly twin sibling comparison using paired-sample t-test showed no significant difference between the twins neither in the total sore nor in subdimensions (total HrQoL: mean difference (MD) 0.74, p=.697; physical well-being: MD 0.78, p=.781; emotional well-being: MD 3.58, p=.140; self-esteem: MD 1.22, p=.811; family: MD 0.59, p=.805; friends: MD 1.19, p=.724; school: MD 0.74, p=.830). In a comparison with the KINDL-R reference values matched by age and sex, there was no significant difference for both groups, the children with and without CHD, respectively.
Conclusions: That means both showed normal HrQoL and at the same time that in twins the congenital heart defect seems to have no greater influence on their self-reported HrQoL. Maybe in a twin relationship, there is a close connection; that means if one is on a high level of HrQoL the twin is it as well, and the other way around if HrQoL is low.

O38
Cognitive functioning in children with congenital heart defects

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Introduction: Risk for children with congenital heart defects (CHD) to develop lower cognitive functions compared to healthy children has been reported. However, few studies look at specific factors related to the differences on cognitive functioning. This study compares cognitive functioning in children with congenital heart defects in relation to severity of the heart defect, the child´s age and the socioeconomic status of the family (SES).
Methods: 227 children with CHD were tested using the Wechsler intelligence scales to determine Full Scale IQ (FSIQ), and indexes for Verbal Comprehension (VCI), Perceptual Reasoning (PRI), Working Memory (WMI) and Processing Speed (PSI). We analyzed FSIQ, VCI, PRI, WMI, and PSI in relation to age (3-, 5-, 9-, and 15-year-olds), severity of the diagnosis (mild, moderate, and severe), and SES (low, medium, and high).
Results: Children with severe CHD had significantly lower FSIQ, VCI and PCI but not WMI and PSI than children with mild CHD. The 9-years old children had significantly lower FSIQ and VCI compared to the 3-year olds. There was no significantly difference on the PRI, WMI and PSI within the different age groups. Children from families with low SES had significantly lower FSIQ, VCI, PRI, WMI and PSI. No interaction between severity of diagnosis, age and SES was found for FSIQ or any of the cognitive indexes.
Conclusions: Severity of the heart defect and SES of the family had significant main effects on cognitive functioning for children with CHD. These factors are important and should be considered when planning interventions and follow-up programs for children with CHD and their families.

O39
Parental anxiety among families of children with congenital heart diseases

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Introduction: The coronavirus disease 2019 (COVID-19) pandemic has swept across the world, causing disruption in all aspects of human life. Our aim was to access the psychoemotional impact the pandemic on families of patients with congenital heart diseases (CHD).
Methods: This is cross-sectional observational study with patient reported outcomes by online questionnaire. The questionnaire included demographic, medical, social and anxiety (Generalized Anxiety Disorder 7-item; GAD-7) sections and was disseminated via online forums and discussion groups bringing together patients of children with CHD. The statistical analysis was caried out using Wizard Pro 2.0.0beta238. Categorical variables were expressed as percentages, continuous variables as mean ± standard deviation or median (minimum-maximum) dependently on the distribution. Appropriate tests were employed and $p<0.05$ was considered statistically significant.
Results: From 30. April till 6. May 2020, 243 eligible, complete responses were collected. Parents were 34±5 years of age, and

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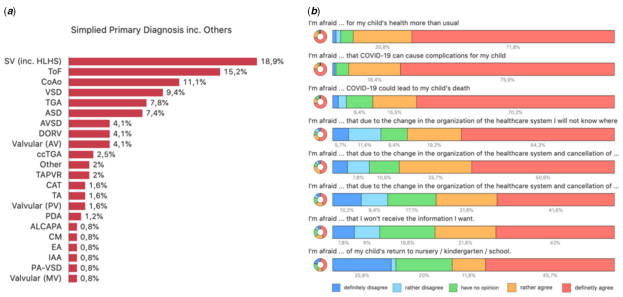
children's age was 4 (0–18) years. Distribution of CHD is represented in Figure 1A. Figure 1B shows degrees of parental concern regarding various aspects of medical as well as daily matters. A vast majority of guardians agreed that they would feel more secure if they knew when scheduled visits (93%) and procedures (80%) would resume, and if children with CHD are actually in high-risk group (94%) and how to protect them (96%). Interestingly there was no correlation of parental degree of anxiety and child's age (0.272), CHD complexity (p=0.855), whether the child had undergone cardiac surgery or catheterization (p=0.350) or awaits such (p=0.982). Conversely there was a strong correlation between the level of concern and the degree of anxiety (p<0.001) as well as on what would make them feel more secure (p<0.001). Our study design allows for no assumptions on the causal matter of this correlations.
Conclusions: The COVID-19 pandemic caused high levels anxiety among parents of chronically ill chirdnen like those with congenital heart diseases. Much of this is correlated to uncertainty regarding both the new disease itself and the resultant reorganization of healthcare system. A strong expectation for clear and timely communication regarding the new risks, safety measures and new plans of providing care was expressed.



11. Morphology

O23

The particular anatomy of ventricular septal defect in patients with Down syndrome and complete atrioventricular septal defect

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Introduction: Complete atrioventricular septal defect (CAVSD) is associated with Down syndrome (DS) in almost half of cases, and is due in this setting to an abnormal development of the vestibular spine, derived from the posterior second heart field (SHF). Clinical studies have shown that Tetralogy of Fallot(TF), an outflow tract defect involving the anterior SHF, is more frequent in CAVSD patients with DS. We therefore decided to look for an outlet extension of the VSD in DS and non-DS.
Methods: We reviewed retrospectively all 208 consecutive patients hospitalized in our unit with the diagnosis of CAVSD between 01/01/2017 and 09/12/2019. All echocardiographic examinations were screened by 2 pediatric cardiologists (ZB /LH).We excluded 46 patients whose VSD anatomical type could not be determined. We used the classification as agreed upon by ICD-11 to determine the VSD anatomical type in the 162 patients included.
Results: Among the cohort, 101/162 had DS (62.7%). An outlet extension of the VSD, with anterior malalignment of the outlet septum, was found in 88/101 (87.1%) DS vs 4/61 (6.5%) non-DS (p<0.0001). Associated TF was found in 12 DS vs 1 in non-DS (p=ns). Ventricles were balanced in 126 patients, 23 had a hypoplastic left ventricle (LV), 13 had a hypoplastic right ventricle (RV). There was no significant difference regarding balanced or not ventricles between DS and non-DS. However, the hypoplastic ventricle was always the LV in non-DS, and was the RV in 62% of DS (p=0.006).
Conclusions: This echocardiographic study describes for the first time that outlet extension of the VSD is the rule in CAVSD with DS and that hypoplastic RV occurs only in DS. This seems to confirm the involvement of the two parts of the second heart field in the morphogenesis of CAVSD in DS patients. The presence of a malaligned outlet septum in a fetus with CAVSD should raise awareness of the high risk of associated DS.

O24

Virtual reality for surgical planning, training and patient engagement in the field of congenital heart disease: a single centre experience

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Introduction: The treatment of congenital heart disease (CHD) is complex and often requires a personalised approach involving several surgical interventions. Despite advancements in imaging, understanding CHD in 3D can still be a major challenge, especially for atypical cases spanning multiple pathologies. Virtual reality (VR) is a rapidly growing technology allowing the user to be completely immersed within a simulated environment. At our centre, we pioneered the use of VR for CHD to: (i) aid surgical planning (ii) enhance teaching and (iii) improve patient communication.
Methods: A VR application has been designed in house specifically to meet these aims over the last year at our centre. The VR software allows the user to import multiple models of patient-specific anatomies reconstructed from medical images. The application uses a commercial headset with controllers to allow the user to interact with the virtual heart. Different tools were implemented to help the planning (measuring, cutting, device interaction), improve teaching (labelling, ultrasound simulators, multi-user experiences) and patient engagement (labelling, colours, splitting). The development was a collaborative process between clinicans, engineers and educators, allowing for tailored tools to be developed to address the clinical/educational needs.
Results: In clinical practice, our VR app was successfully used for planning more than 20 cases, including: (i) double-outlet right ventricle repairs (ii) aortic root replacements for Marfan syndrome (iii) percutaneous pulmonary valve implantations (iv) stenting of pulmonary arteries. In an educational setting, VR has been routinely added to a specialised cardiac morphology course for healthcare professionals, over 90% of whom found it "extremely/very useful" in aiding their understanding of CHD. In addition, it was implemented in an undergraduate course during COVID-19 to support remote learning, with the use of online multi-user teaching. Over the course of two years, more than 100 students were taught using VR. Finally, our app was found to be

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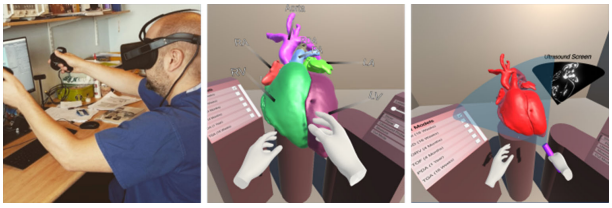
particularly engaging by patients and their families in understanding the complexity of CHD during public events held at our Centre.
Conclusions: Our preliminary research with in-house developed VR software showed how this can be used in facilitating the understanding of CHD for treatment planning, education and communication.



12. Preventive

O56
Cardiopulmonary function testing in 4-6-year-old children: use of outdoor testing

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Introduction: Cardiopulmonary exercise testing (CPET) in very young children (4-6y) represents a challenge. Requirements for a representative treadmill testing are good motor skills and a high motivation. Most studies investigating CPET have therefore been limited to older children (> 8 y). However, knowledge of the performance of small children is essential for evaluating cardiorespiratory fitness in that age-group. Thus, this study strives to compare the modified Bruce protocol endorsed by the DGPK with a new age-appropriate incremental CPET during natural movement running outdoors, using a mobile device.
Methods: A group of twenty-two 4 – 6-year-old healthy children was tested indoors using the modified Bruce protocol. The results were then compared with a self-paced incremental running test, using a mobile CPET device in an outdoor park. The speeds for the incremental running test were described as 1) slow walking, 2) slow running, 3) regular running, 4) running with full speed as long as possible. An experienced running coach accompanied each child during the test for encouragement and for sustaining pace during each step.
Results: The children were able to reach significantly higher values for most of the CPET variables during the outdoor test. None of the children reached maximum exertion indoors defined as RER ≥1.1 (mean RER 0.98). In comparison, mean RER outdoors was 1,10. Even though mean exercise time outdoors (6,57min) was significantly shorter than on the treadmill (11,20min), CPET values were significantly higher for the following CPET parameters: VO_{2peak}, peak velocity, RER, VE_{max}, O_2pulse, heart rate and breath rate. The submaximal parameter OUES was comparable between both tests.
Conclusions: Testing very young children outdoors, using a mobile device, is a new alternative to treadmill testing. With a significantly shorter test duration, significantly higher values for almost all cardiopulmonary variables can be achieved without losing the ability to determine VT1 and VT2. It avoids common treadmill problems and allows for individualized exercise testing with the aim of a standardized exercise time with an individual protocol instead of a standardized protocol with an individual exercise time, thus allowing for better comparability.

O57
False Negative Pulse Oximetry Screening; Data from a Swedish national population-based study 2014-2019

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Introduction: Since 2013, routine pulse oximetry screening (POS) covers all maternity units in Sweden. We performed a population based retrospective analysis of the false negative rate of routine POS for critical congential heart disease (CCHD) on a national level. Our aims were to determine the proportion of all newborn infants with CCHD who were diagnosed prenatally and postnatally before discharge. We wished to study the clinical consequences of a missed diagnosis of CCHD for a better understanding of the limitations of prevailing screening method and in the context of increasing prenatal detection rates and decreasing duration of postnatal stay.
Methods: All infants with CCHD between 2014-2019 were included in the study. CCHD was defined as requiring surgery and/or catheterization, or who died without such treatment within the first month of life. Cases were identified in local surgery databases, the Swedish Registry of Congenital Heart Disease (Swedcon) and the Surveillance Register of Birth Defects. Case note review was performed in each case.
Results: So far, 332 infants with CCHD, operated in Gothenburg have been reviewed only by searching local surgery and catheter procedure records. Preliminary data give a total prenatal detection rate of 35%, the lowest detection rate was in aortic coarctation (CoA) (13%). Of 67 neonates with CoA without a prenatal diagnosis, 49 (73%) had a false negative screen. Additional CCHD without a prenatal diagnosis with a false negative screen on POS include hypoplastic left heart syndrome (n=1), truncus arteriosus/CoA (n=2), pulmonary stenosis (n=6), aortic stenosis (n=6), unbalanced atrioventricular septal defect (n=1), total anomalous pulmonary venous return (n=1), cor triatriatum (n=2), aortic atresia/CoA (n=1) and arch hypoplasia (n=3). None of the neonates born with transposition without prenatal diagnosis had a false negative screen. Postoperative mortality was 8% (n=28). In 10 cases, death occurred within 30 days.
Conclusions: Preliminary data show that the prenatal detection rate in our referral region is still not satisfying. The sensitivity of POS to left heart obstructive defects (aortic stenosis, arch hypoplasia, CoA) is low to moderate, as for pulmonary stenosis. Data collections of the total cohort continues and the clinical consequences of a negative screen will be further studied.

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O58

Growth of the Aortic Root in Children and Young Adults with Marfan Syndrome

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Introduction: Marfan syndrome (MFS) causes severe aortic root dilatation, which may start during early childhood. Accordingly, close clinical follow up of aortic diameters in pediatric MFS patients is warranted as aortic root replacement early in life may be necessary. Nevertheless, published data on the aortic root growth in children with MFS are limited. Therefore, the primary aim of our study is to gain insight in the growth of the aortic root of children and young adults with MFS. Furthermore, the effect of sex and *FBN1*-mutationtype (dominant negative (DN) *FBN1*-mutations versus haploinsufficient (HI) *FBN1*-mutations) on aortic root growth was assessed.
Methods: Aortic root dimensions of 97 MFS patients between 0-20 years and 30 controls were serially assessed with echocardiography. Age-related trends were analysed using a linear mixed-effect-model. Additionally, only including MFS patients, trends were allowed to differ by sex, aortic root replacement, or *FBN1*-mutationtype.
Results: Patients with MFS had a significantly larger aortic root diameter at all ages compared to controls, which became more pronounced from the age of eight years. In the MFS-cohort, males had a significantly greater aortic root diameter than females at all ages. There was no difference in aortic root growth between children with DN *FBN1*-mutations or HI *FBN1*-mutations. Eleven children (11%) underwent aortic root replacement (mean aortic root diameter of 45.6 ± 3.3 mm, mean age of 16.5 ± 1.8 years). Compared to MFS patients without aortic root surgery, these children had a significantly greater aortic root diameter from a very early age.
Conclusions: The present study provides clinically useful longitudinal growth charts on aortic root growth in children and young adults with MFS. Furthermore, the study shows that children needing prophylactic aortic root replacement during childhood can be identified at a very young age. Accordingly, our growth charts can help clinicians in decision-making with regard to follow-up and prophylactic therapy. No differences in aortic root growth is observed among HI *FBN1*-mutation and DN *FBN1*-mutation carriers in children and young adults.

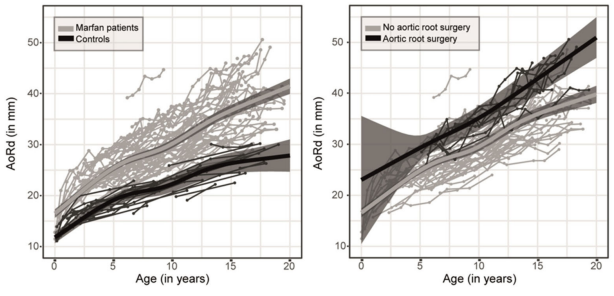

Figure1: Left panel: Aortic root diameter (AoRD) versus age (in years) for each individual Marfan patient (grey) and control subject (black). Dots are individual measurements and lines are individual trends. Mean progression of the aortic root diameter (thick grey or black line) and 95% confidence intervals are depicted as well (grey area). Right panel: Progression of the aortic root diameter (im mm) versus age (in years) in the aortic-root-surgery group (black) and no-surgery group (grey). 95% confidence intervals are depicted (grey area).

O59

Multimodal assessment of vascular and ventricular function in children and adults with a history of bicuspid aortic valves

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Introduction: Bicuspid aortic valve (BAV) is the most common congenital cardiac anomaly, existing in 1-2% of the population. Patients with BAV have been suggested to have increased aortic stiffness and abnormal diastolic function. In this study, a multimodal approach was used to investigate cardiovascular function, the relation between arterial and cardiac function as well as their age-related changes in patients with BAV compared to controls.
Methods: Patients with BAV and healthy controls were recruited for the study. Cardiac function (including tissue Doppler and 4-dimensional imaging), aortic elasticity and thickness of the common carotid intima media (cIMT) were analysed with ultrasound. Central blood pressure, central augmentation index (cAIx) and pulse wave velocity (PWV) were measured (SpygmoCor XCEL). Digital pulse wave analysis (DPA) was used to assess aging index (AI). Endothelial function (reactive hyperemia index, RHI) and peripheral Aix (pAIx) was measured (EndoPAT). Laser doppler measured microcirculatory flow at baseline and its response to Acetylcholine (endothelium-dependent) and Sodium-Nitroprusside (endothelium-independent; Periflux5000). Statistical analyses were performed using Mann-Whitney U test, linear regression analysis to correct for covariates and Pearson's correlation coefficient.
Results: Fourty-seven patients with BAV and 84 controls age between 8-65 years were included in the study. Diastolic function, and left ventricular mass normalized to body surface area [LVMi] were significantly abnormal in the BAV subjects compared to controls, while systolic function was preserved. Ascending aortic stiffness, cIMT and pulse wave reflection (cAIx, pAIx and AI) as well as central blood pressure all were significantly increased in patients with BAV. However, PWV was significantly decreased. Further, there was no difference in endothelial function or microcirculation. All parameters of arterial stiffness had moderate-strong correlations with diastolic dysfunction and age. In the BAV group, ascending aortic elasticity parametrs had the stronges correlations with both diastolic dysfunction and age (for stiffness index $r=0.6$, and $r=0.76$, both $p<0.001$).
Conclusions: Patients with BAV have increased proximal arterial stiffness and wave reflection which correlates with diastolic dysfunction and age. However, PWV and cIMT are not increased, and endothelial function is not impaired. This suggests that the mechanism of arterial and cardiac stiffening is different from patients with acquired heart diseases.

O60

Ventilatory and metabolic exercise characterization of young competitive athletes with congenital heart diseases

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Introduction: Although physical activity is recognized as an important intervention in patients with congenital heart disease (CHD),

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data on effect of competitive sport are scant. We aim to assess outcome of patients with CHD enrolled in competitive sport and to characterize their cardiopulmonary exercise response.
Methods: Patients with CHD followed at the sport medicine department, who had undergone serial cardiopulmonary test (CPET) between 2011 and 2019, were included. Patients involved in competitive sports were matched 1:1 for age (± 2 years), gender, BMI and anatomic diagnosis with patients practicing physical activity at recreational level, that was assumed as control group. Ventilatory and metabolic exercise parameters were compared between the two groups.
Results: 111 patients, 73 males (66%), were analyzed. Twenty-nine patients (27%) were practicing sport at competitive level. 60 % of whom had CHD of moderate or great complexity. Median age was 14 (13-16) years and 13 (12-15) years for competitive and recreational athletes respectively (p=0.17). Percentage of maximum work achieved and peak oxygen consumption (VO2) were significantly higher in patients involved in competitive sports as compared with the matched control group: 100 (93-115) % vs 88 (69-100) % and 38 (33-44) vs 42 (36-42) ml/min/Kg, p=0.0007 and p=0.03 respectively. Percentage of predicted peak VO2, maximal ventilation/minute (VE max), O2 pulse, ventilatory efficiency (VE/VCO2 slope), VO2/Work slope were comparable between the two matched groups.(figure) Median follow up of 2.1 (max 7) years for the group of competitive athletes was uneventful.
Conclusions: These preliminary data suggest that CHD patients, including those with complex anatomies, can safely practice sport at competitive level. Apart from percentage of maximum work achieved and peak VO2, all other exercise parameters were comparable suggesting a ceiling effect of training level and cardiovascular performance in patients with CHD. These preliminary data suggest a positive effect of systematic training in this subset of patients.

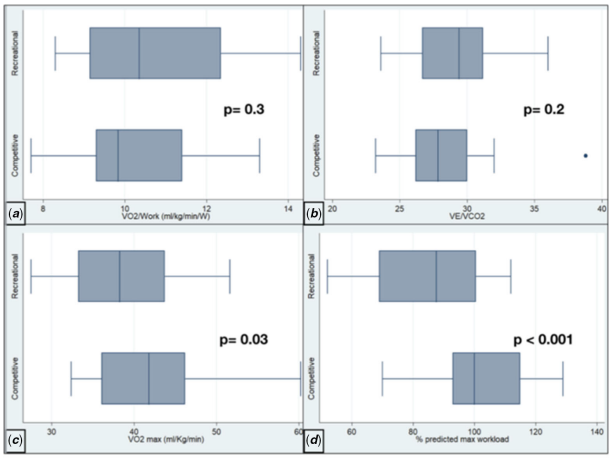

Figure: Box plot illustrating comparison of VO2/Work (panel A), VE/VCO2 (panel B), VO2 max (panel C), % of maximum predicted workload (panel D).

13. Nursing

O37
Factors perceived by young adults with congenital heart disease to affect continuing follow-up care after transfer

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Introduction: The growing population of young persons with congenital heart disease (CHD) needs appropriate follow-up care across their life-spectrum to safeguard future health. Among these patients, discontinuation of care is highly prevalent and associated with adverse outcomes, such as increased morbidity and need for urgent interventions. To develop preventive strategies for discontinuation, factors affecting continuing follow-up care from the patients' perspective should be explored. Settings characterized by a low prevalence of discontinuation of CHD care provide insights into facilitating factors for continuing care. The present study aimed to identify and explore factors perceived by young adults with CHD to affect continuing follow-up care.
Methods: This qualitative study included participants from seven Swedish university hospitals. All participants received follow-up at paediatric cardiology clinics and were transferred to adult healthcare facilities at 18 years of age. Both young persons with continued and discontinued follow-up were included. Sixteen interviews with young people aged 27y-29y were conducted a decade after their transfer to adult care. Interviews were transcribed verbatim and analysed with qualitative content analysis.
Results: Three main categories were identified describing factors perceived by participants to affect continuing follow-up: 1. Motivation for follow-up care; 2. Participation in care and sense of connectedness with a healthcare provider (HCP); 3. Accessibility of care. (Figure 1). Multiple factors affected the participant's choice to continue or discontinue follow-up care. Their choice was often related to perceptions of CHD and follow-up care needs. Finding personal motivation was an important facilitator, as well as the interpersonal relationships with HCP´s and a sense of belonging to the clinic. Behaviours and attitudes of HCP´s were described as barriers and differences between paediatric and adult care were raised. Participants without follow-up care stressed the importance of encouragement and support in order to return to follow-up care.
Conclusions: To provide holistic care for this patient population, sufficient competencies among HCP´s is required to carefully consider the specific health care needs of young people and in particular remaining transitional needs after transfer. Factors on three levels: patient-, hospital- and healthcare-system level were described by participants, indicating the need for holistic approaches to prevent discontinuation of follow-up care.

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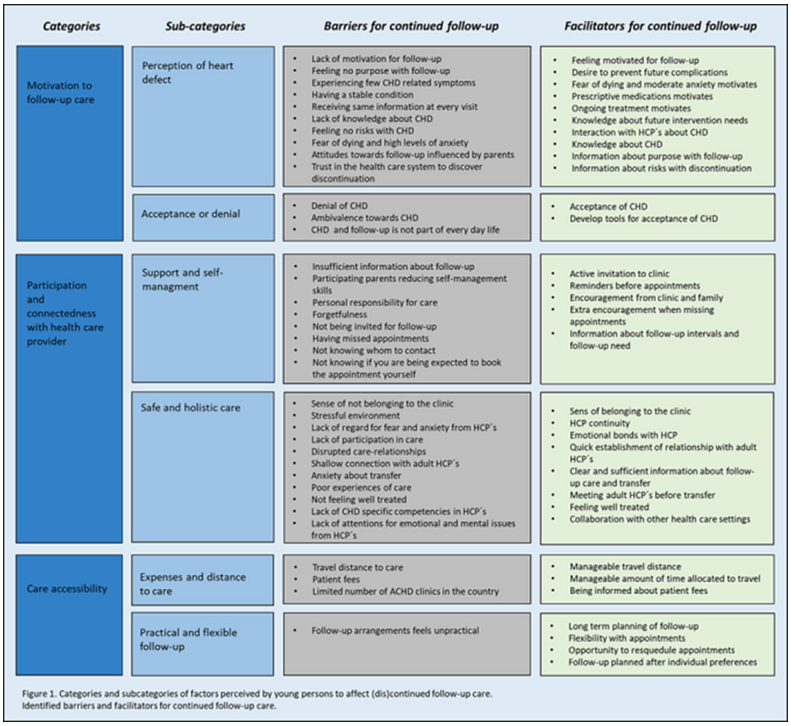

Figure 1. Categories and subcategories of factors perceived by young persons to affect (dis)continued follow-up care.
Identified barriers and facilitators for continued follow-up care.

O40
Families and Congenital Heart Disease: Family Adaptation in Three Groups of Families

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Introduction: Congenital Heart Disease (CHD) is one of the most common birth defects. One group of children at increased risk for CHD are children with Down syndrome (DS); between 40 to 50% of children with DS have CHD. There is growing evidence that while some families have difficulty adapting to their child's diagnosis of CHD and/or DS, other families adapt well and some thrive. The purpose of this study, guided by the Resiliency Model of Family Stress, Adjustment and Adaptation, was to explore family adaptation in three groups of families: (1) families of children with CHD (2) families of children with DSand (3) families of children with CHD and DS.
Methods: Sixty parents of children who have been diagnosed with either CHD, DS, or both CHD and DS were interviewed (20 interviews per group). Clinicians and group leaders of support groups were sent an Invitation to Participate that they could share with eligible families. The main goal of the interview was to give parents the opportunity to share their family's story. Most interviews lasted 60-90 minutes. The interviews were transcribed verbatim, then they were coded using coding categories based on the Resiliency Model. Within and across group comparisons were made between the three groups of families. For this presentation, the focus will be on family challenges.
Results: While families in the three groups experienced some of the same challenges, there were unique challenges for each group. Moreover, there were across group differences in the number of challenges faced and the type of support available to families to address these challenges; families with children who have both CHD and DS typically experienced the most challenges.
Conclusions: Families of children with CHD, DS, and both CHD and DS face a wide variety of challenges. For some families, these challenges make it difficult for the family to adapt successfully to the child's diagnosis. There is currently a need for research on how family variables and other social determinants of health intersect to influence child, parent and family outcomes. Findings from this research could be used to develop condition-specific family interventions.

Oral YIA
Arrhythmia/electrophysiology

O-Y4
Atenolol Should Not Be the ß-blocker of Choice for Symptomatic Children with Catecholaminergic Polymorphic Ventricular Tachycardia

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Introduction: Children with catecholaminergic polymorphic ventricular tachycardia (CPVT) are at risk for malignant ventricular arrhythmias during exercise and emotions, which may lead to arrhythmic events such as sudden cardiac death (SCD). Symptomatic patients are at particular risk for the reoccurrence of arrhythmic events. Beta-blockers are the cornerstone of therapy in patients with CPVT. However, studies comparing the efficacy of different types of beta-blockers are scarce. We aimed to determine the efficacy of different types of beta-blockers in reducing the risk for recurrent arrhythmic events in a large cohort of symptomatic children with CPVT.
Methods: Data were derived from the International CPVT Registry, a large retrospective observational cohort study. We included symptomatic children aged <19 years who were carrier of a *RYR2* variant and who were prescribed a beta-blocker. The primary endpoint was the occurrence of an arrhythmic event (AE), defined as SCD, aborted cardiac arrest, appropriate ICD discharge or syncope. Time-dependent Cox-regression analyses were used to compare the occurrence of AEs between different beta-blockers corrected for possible confounders with nadolol as reference group.
Results: We included 267 children treated with a beta-blocker. One hundred five (39.3%) children were first treated with nadolol, 64 (24.0%) with propranolol, 43 (16.1%) with atenolol, 26 (9.7%) with metoprolol and 21 (7.9%) bisoprolol. Age at initiation of beta-blocker differed between the groups, with the youngest mean age in propranolol and highest in bisoprolol and metoprolol (10±4 years in propranolol, 13±4 years in bisoprolol and nadolol, overall-p=0.023). Sex, the proportion of probands and the proportion of patients treated with flecainide, left cardiac sympathetic denervation and an ICD were equally distributed among all groups. In total 86 (32.2%) children had an AE. The AE-rate was significantly higher in patients treated with atenolol compared to nadolol (hazard ratio (HR) 2.15, 95% confidence interval (CI) 1.05-4.40, p=0.036, Table). There were no significant differences in the AE-rate in patients treated with bisoprolol (HR 2.08, 95% CI 0.92-4.71), metoprolol (HR 1.79, 95% CI 0.82-3.92), and propranolol (HR 1.55, 95% CI 0.84-2.86) compared with nadolol.
Conclusions: Atenolol is associated with a higher risk for a subsequent arrhythmic event in symptomatic children with CPVT compared to nadolol.

Table. Adjusted hazard ratios for the occurrence of AEs in CPVT patients treated with different beta-blockers types relative to nadolol.

	HR	Lower limit 95%CI	Upper limit 95%CI	P-value
atenolol	2,15	1,05	4,40	0,036
bisoprolol	2,08	0,92	4,71	0,079
metoprolol	1,79	0,82	3,92	0,142
propranolol	1,55	0,84	2,86	0,157

HR=Hazard ratio, CI=confidence interval

O-Y6
Contact Force Guided Radiofrequency Current Application at Developing Myocardium: Lesion Size and Coronary Artery Involvement

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Introduction: Catheter contact is one key determinant of lesion size in radiofrequency catheter ablation (RFA). Monitoring of contact force (CF) during RFA has been shown to improve efficacy of RFA in experimental settings as well as in adult patients. Value of CF monitoring in pediatric patients has not been systematically studied yet.
Methods: RFA with continuous CF monitoring was performed in 24 piglets (median weight 18.5 kg) using a 7F TactiCath Quartz RF ablation catheter (Abott, Abbott Park, Illinois, USA). A total of 7 lesions were induced in each animal applying low (10-20 g) or high (40-60 g) CF. RF energy was delivered with a target temperature of 65 °C at 30 W for 30 seconds. Coronary angiography was performed prior and immediately after RF application. Animals were assigned to repeat coronary angiography followed by heart removal after 48 h (n=12) or 6 months (n=12). Lesions with surrounding myocardium were excised, fixated and stained. Lesion volumes were measured by microscopic planimetry.
Results: A total of 148/172(86%) of applied lesions were identified in the explanted hearts. Only in the subset of lesions at the AV annulus 6 month after ablation, lesion size and proportion of transmural lesions were higher in the high CF group while CF had no impact on lesion size and extension in all lesions after 48 h as well as in the atrial and ventricular lesions after 6 months. Additional parameters as Lesion-Size-Index and Force-Time-Integral were also not related to lesion size. Coronary artery damage was not related to catheter CF and was present in 2 animals after 48 h and in 1 after 6 months.
Conclusions: In our experimental setting in piglets lesion size was not related to catheter CF. Transmural extension of the RF lesions involving the layers of the coronary arteries was frequently noted irrespective of CF. Coronary artery narrowing was present in 3/24 animals. According to these findings it may be speculated that even lower CF during RF ablation in infants and toddlers may be equally effective and less traumatic than applied in adults. Impact of CF monitoring during conventional RF ablation in children requires further investigations.

Basic science, Genetics

O-Y2
Can Regional Differences in Expression of Cardiomyopathy-related Proteins Explain the Clinical Phenotype: A Pilot Study

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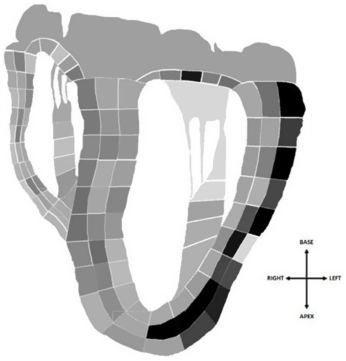
Introduction: Recognised gene mutations poorly explain regional phenotypic differences in the myocardium of patients developing cardiomyopathy. Understanding the mechanisms driving these patterns, which often begin during childhood, may offer clues to innovate new treatment and diagnostic strategies. Previous proteomic studies have typically analysed single, small tissue samples obtained from a cardiac chamber or cell culture. Developing a novel approach, we aim to describe regional differences in the expression of important cardiomyopathy-associated proteins, with high resolution in different axes across each ventricular wall.
Methods: Continuous samples were obtained from 4-chamber cross-sections of bovine myocardium. Proteins from each were solubilised, extracted and digested, before analysis by mass spectrometry using a 'hypothesis-free' approach. Multivariate analysis was applied, to make unbiased comparisons between samples at whole-proteome level. Twenty-eight cardiomyopathy-associated proteins were selected and compared between samples by relative abundance. Multiple correlation analysis described variation from endocardium-to-epicardium, apex-to-base and between each ventricular free-wall. Relative intensity maps were additionally generated.
Results: One-hundred and twenty-two samples of ventricular myocardium were analysed over 128 hours, generating 278 GB of data. 1,017 unique proteins were consistently detected among intra-sample repeats. Their relative expression conformed to three distinct regional patterns, varying predominantly from epicardial to endocardial layers. Regional variations in abundance were demonstrated across all selected proteins. Eleven disease-associated proteins, including Myomesin-1 and Actin alpha-1, were enriched within the ventricular septum ($p<0.05$). Likewise, eight proteins were specifically enriched within the right ventricular epicardial wall ($p<0.05$). Interestingly, some proteins were most abundant within regions associated with their corresponding cardiomyopathy. Mutations in the Desmoglein-2 gene, for example, are associated with a more left-ventricular dominant phenotype of arrhythmogenic cardiomyopathy (AVC). Unlike other AVC-related proteins, Desmoglein-2 was significantly more abundance within the left ventricular free-wall (figure).
Conclusions: This novel approach describes considerable and detailed variation in the regional abundance of 28 proteins implicated in three major cardiomyopathies. Such variation questions the interpretation of previous cardiac proteomic studies, which typically assume random tissue samples to be representative of the wider myocardium. Application of this approach to disease models at different stages, may offer new insights into development of a cardiomyopathy phenotype in populations of genotype-positive children and adolescents.



O-Y5
Mitochondria-activated myocardial progenitor cell (MITO cell) transplantation therapy in a mouse model of ischemia and reperfusion

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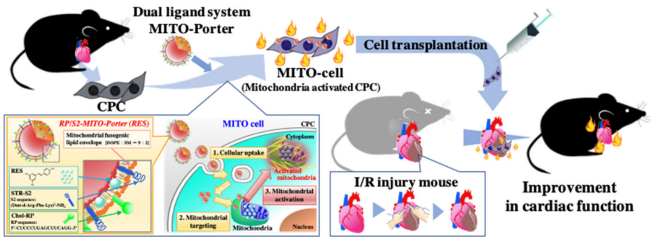
Introduction: Myocardial stem cell transplantation has been clinically tested for use in the treatment of heart failure, and its usefulness has been recognized. However, it is difficult to maintain a sustained cardiac stem cell transplantation effect, to overcome in the transplantation of cells into the heart. We hypothesized that enhancing the mitochondrial function of transplanted cells could increase the survival rate, proliferation rate, and engraftment rate of cells after transplantation. We previously reported on the development of a drug delivery system targeting mitochondria (MITO-Porter). A MITO-Porter could efficiently deliver a drug encapsulated in the MITO-Porter to the mitochondria of the cells. We succeeded in preparing mitochondria activated cardiac stem cells (MITO cells) by the mitochondrial delivery of an antioxidant in transplanted cells using a MITO-Porter system. In this study, in order to prove the therapeutic effect of MITO-cell transplantation, a myocardial ischemia reperfusion model mouse was prepared, and the therapeutic effect of myocardial stem cell transplantation therapy using such MITO-cell on the ischemic myocardium was examined.
Methods: On the basis of isolated mouse CPCs, MITO cells were created using MITO-Porter. The transplantation of CPCs and MITO cell into the heart of the mice were conducted after myocardial ischemia/reperfusion, and the therapeutic effect was determined 30 days later. Cardiac functions were evaluated by echocardiography and the determination of the extent of formation of histological fibrosis.
Results: The MITO cells that were transplanted into an ischemic myocardium showed an increase in postoperative weight gain compared to the non-transplanted group and the CPC group (PBS group +0.2 g (+1%), CPC transplantation +1.5g (+7.4%), MITO cell group +2.3g (+12.4%), PBS vs MITO cell $p<0.05$). It was confirmed that cardiac function was improved (PBS group 23±1.8%, CPC group 30±1.2% and MITO cell group 33±2.3% (PBS vs CPC $p<0.05$, PBS vs MITO cell $p<0.01$)), as evidenced by echocardiography and the formation of histological fibrosis was suppressed.
Conclusions: The transplantation of MITO cells to the ischemic myocardium showed a stronger transplantation effect compared to conventional CPC transplantation.



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Fetal cardiology

O-Y1
Association between assisted reproductive technology and major congenital heart disease in Japan

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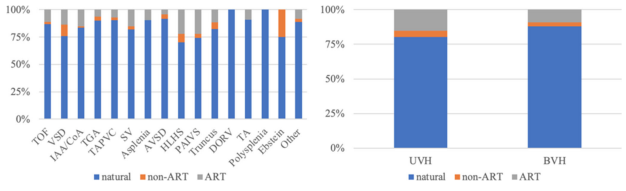
Introduction: Association between assisted reproductive technology (ART), which began 40 years ago, has spread all over the world. And 1 in 16.7Japanese infants is conceived through ART. Because of reproductive aging and advanced medical system, Japan has the highest number of ART implementations in the world.

Infants conceived through ART have a high frequency of congenital anomalies including congenital heart disease (CHD). However, there are few reports on the frequency of CHD in infants conceived through ART in Japan, so we performed this study.
Methods: Major CHD (a lesion that requires surgery or intervention catheter in the first year of life; MCHD) cases diagnosed in 592 infants (male=318) at two Japanese pediatric heart centers from January 2013 to July 2020, were reviewed retrospectively. The infants' and parents' characteristics, and types of fertility treatment were evaluated.
Results: Gestational age was 38.3±1.9 weeks. Maternal and paternal ages were 32±5 and 34±6 years, respectively. One hundred fifty patients (25%) had chromosome or gene abnormalities, or syndrome. Sixty-two patients (10%) and 20 patients (3%) were conceived through ART and non-ART fertility treatments, respectively. One hundred thirty-one patients (22%) had univentricular heart defect (UVH). Compared with the proportion of ART (5.5%) in all births in Japan, the proportion of ART (10%) in MCHD was high. In addition, the proportion of ART in UVH was 20/131 (15%), while the proportion of ART in biventricular heart defect was 42/461 (9%) ($p<0.05$).Among them, the highest proportion of ART in hypoplastic left heart syndrome and pulmonary atresia with intact ventricular septum was 6/27 (22%) and 6/27 (22%), respectively.
Conclusions: Infants conceived through ART in Japan are expected to increase. But some infants with MCHD conceived through ART cannot be diagnosed with CHD before birth even now. Since infants conceived through ART might have a high incidence of complex CHD and UVH, they need more careful fetal follow-up than infants conceived naturally.



Heart failure

O-Y3
Hepatic Venous Oxygen Saturation As a Novel Marker for Fontan Associated Liver Disorder

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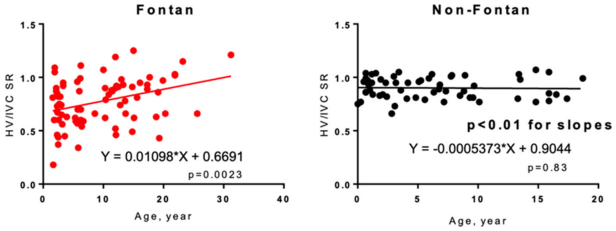
Introduction: Fontan associated liver disorder (FALD) is one of critical complications after Fontan procedure. Despite advancement of imaging and molecular technologies, prospective studies failed to identify sensitive biomarker to detect subclinical FALD, where its prevalence is estimated more than 80% in 10 years. As the main source of liver blood supply changes from portal vein (low oxygen saturation:SO2) to hepatic artery (high SO2) with the progression of hepatic fibrosis, we hypothesized that the hepatic venous SO2 increased with the development of liver fibrosis.
Methods: During catheterization, hemodynamics as well as hepatic circulation property including trans-hepatic pressure were assessed in consecutive 117 Fontan and 86 non-Fontan patients. Multiple markers for liver fibrosis were measured and their relationship with hemodynamic properties was analyzed.
Results: As compared with non-Fontan patients, Fontan patients had low cardiac output (CI), high central venous, hepatic venous, and hepatic wedge pressures (p values for all, <0.001), whereas transhepatic pressure was similar. As hepatic venous (HV) as well as inferior venous cava (IVC) SO2 were dependent on arterial SO2 and CI, SO2 ratio of HV/IVC (SR-HV/IVC), which was independent of them, was analyzed. The SR-HV/IVC in the Fontan patients was markedly lower than non-Fontan patients, suggesting unfavorable liver perfusion in the Fontan patients. Interestingly, although the SR-HV/IVC in the non-Fontan patients was consistent regardless of their age (SO2 HV/IVC= 0.90-0.001*age, p=0.83), it became markedly low at 1year of Fontan procedure in the Fontan patients, with subsequent increase to the level of non-Fontan patients after 10 years of Fontan procedure (SO2 HV/IVC= 0.67+0.01*age, p=0.0023), in compliance with the reported prevalence of subclinical FALD. Importantly, SR-HV/IVC in the Fontan patients was negatively correlated with platelet counts (p=0.026) and albumin/IgG ratio (p=0.0035), and positively correlated with serum levels of total-bilirubin (p=0.037) and hyaluronic acid (p=0.055).
Conclusions: SR-HV/IVC, which supposed to reflect pathological feature of hepatic vascular remodeling, is a novel biomarker for subclinical FALD. Prospective studies to investigate utility of SR-HV/IVC to guide prevention of FALD would be warranted.



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Oral PH/HF/Htx
End-stage heart and lung disease

O-P1
Pulmonary artery banding for ventricular rehabilitation in infants with dilated cardiomyopathy. Early results in a single center experience.

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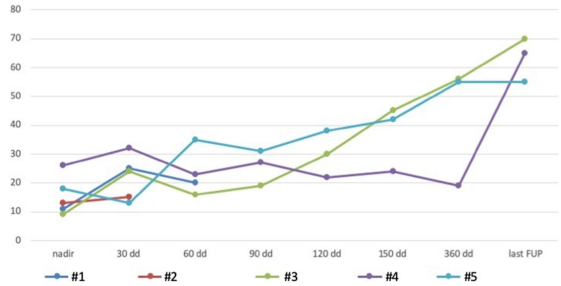
Introduction: Surgical reversible central pulmonary artery banding (PAB) has been proposed as a bridge to heart transplant (HT) or recovery for pediatric end-stage heart failure (ESHF) due to left ventricle dilative cardiomyopathy (LV-DCM). Ventriculo-ventricular interaction is assumed to benefit LV failure.
Methods: We conducted a single-centre prospective clinical study from September 2015 to September 2018 including patients admitted to our Institution for severe HF/cardiogenic shock without congenital heart disease. Inclusion criteria were: age<4 years; severe LV dysfunction due to DCM (EF<30%, LVEDD z-score>4.5) refractory to inotropic and anti-congestive therapy; preserved right ventricular function; candidate for mechanical circulatory support (MCS) and HT. After PAB procedure, all patients underwent periodic echocardiogram monitoring to assess myocardial recovery. Primary outcomes were: survival of study population, reduction of the need for MCS and HT, improvement of LV shape and function.
Results: We selected 5 patients (median age 8.6 months; median weight 7.7 kg). At a median follow up of 22.4 months all them were alive. 1 patient didn't respond to PAB procedure. He required atrial septostomy in conjunction with PAB and MCS for 12 months, with final successful HT. 4 patients responded to PAB procedure. One of them experienced severe acute HF during pneumonia 3 months after PAB and required emergent MCS in another Institution and successful HT within 21 days. The remaining 3 patients are doing well at home. All they required elective percutaneous partial de-banding after 18.5, 4.8 and 10.7 months respectively. At the last follow up (median 16.9 months), they all presented with normal EF value (increased from 17.7±8.5% to 63.3±7.6%, p=0.03), decreased LVEDD z-score from 11.4 ±2.1 to 1.97±3.14 (p=0.01), significant decreased or disappearance of mitral valve regurgitation. All they have been removed from HT list due to reduction in the functional status to Ross class I.
Conclusions: PAB is an effective alternative to MCS in selected infants for bridging to transplant or recovery in ESHF due to LV-DCM. The early postoperative period is difficult and requires strict follow-up and experience, long hospitalization and frequent hospital admissions. Further experience and research are required to identify responders and non-responders to this approach.



Heart failure

O-P2
Clinical presentation and long-term outcomes of infantile hypertrophic cardiomyopathy: A European multi-center study

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Introduction: Children presenting with infantile hypertrophic cardiomyopathy (HCM) are reported to have poor outcomes but prognostic factors are poorly understood. The aim of this study was to describe the aetiology, phenotype and outcomes of infantile HCM in a well-characterised multicentre European cohort.
Methods: Anonymized, non-invasive clinical data were collected from 301 children diagnosed with infantile HCM presenting to 16 European centers.
Results: Underlying etiology was: non-syndromic (n=138, 45.6%), RASopathy (n=101, 33.6%) or Inborn Error of Metabolism (IEM) (n=49, 16.3%). The most common reasons for presentation were symptoms (n=77, 30.6%), which were more prevalent in those with syndromic disease (n=62, 61.5% p<0.001), and an isolated murmur (n=75, 29.8%). Sixty-nine (24%) had a family history of HCM, which was more frequent in those with non-syndromic HCM (n=48, 37.5%, p< 0.001). One hundred and thirty-three (44%) had one or more co-morbidities. Over a median follow up of 4.1 years, 50 (17%) underwent one or more surgical interventions; 15 (5%) had a major arrhythmic event (6 in the first year of life); and 48 (16%) died, with an overall 5-year survival of 85%. Predictors of all-cause mortality were an underlying diagnosis of IEM (HR 4.4, p=0.070), cardiac symptoms (HR 3.2, p=0.005) and impaired LV systolic function (HR 3.0, p=0.028).
Conclusions: This large, multi-centre study of infantile HCM describes a complex cohort of patients with a diverse phenotypic spectrum and clinical course. Although overall outcomes were poor, this was largely related to underlying aetiology. Those with non-syndromic disease had more favourable outcomes.

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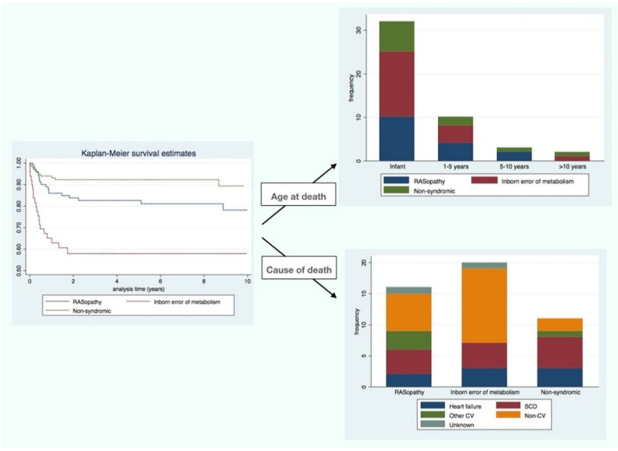

O-P3

Impact of diastolic pulmonary arterial pressure on prognostic assessment in children with pulmonary arterial hypertension

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Introduction: Pulmonary arterial hypertension is defined by the value of mean pulmonary arterial pressure (mPAP) and mPAP-derived pulmonary vascular resistance index (PVRI). Furthermore, these invasively assessed hemodynamic variables and its changes during acute vasoreactivity testing (AVT) serve for risk stratification in pediatric idiopathic/heritable pulmonary arterial hypertension (IPAH/HPAH). Recently, diastolic PAP (dPAP) was proposed as an alternative marker compared to mPAP in the hemodynamic prognostic assessment of PAH patients. We therefore purposed to compare baseline values of dPAP, dPAP/dSAP ratio, dPAP-derived PVRI, and its changes during AVT with corresponding mPAP-derived variables for the prognostication of death and lung transplantation in children with IPAH/HPAH.

Methods: We conducted a retrospective analysis of the TOPP (Tracking Outcomes and Practice in Pediatric Pulmonary Hypertension) registry. Of 686 registered children with confirmed pulmonary hypertension, 331 children (48%) were diagnosed with IPAH/HPAH, 246 of them had evaluable AVT. Statistical analysis was performed using multivariate Cox regression, adjusted for age at diagnosis and gender.

Results: During the observation period, 45 children (18.3%) died, and 13 (5.3%) received lung transplantation. Mean time from inclusion to death was 1.66 ± 1.7 years, mean time from inclusion to transplantation was 3.34 ± 2.1 years. At baseline, dPAP and mPAP (HR 1.0, p= 0.22 vs. p=0.08), and dPAP/dSAP (HR 2.1; p=0.09) were not significantly related to time to death/transplantation. Baseline mPAP/mSAP (HR 3.2; p=0.01), and more strongly, baseline dPAP- and mPAP-derived PVRI were associated with time to death/transplantation (HR 1.0, p=0.002 vs. p<0.001). At maximum response, percentage reduction of dPAP and mPAP (HR 0.97, p<0.001 vs. p=0.001), and reduction of dPAP/dSAP and mPAP/mSAP (HR 0.98, p<0.001 vs. p=0.001) were associated with time to death/transplantation. No predictive value was found for percentage reduction of PVRI during AVT, neither dPAP- nor mPAP-derived (p>0.05 for both).

Conclusions: According to our results, the assessment of dPAP and dPAP-derived hemodynamic parameters and its changes during AVT provide a useful adjunct to the catheterization protocol and could be used alternatively for prognostication of children with IPAH/HPAH. Values of maximum reduction during AVT, whether dPAP- or mPAP-derived, represent a better predictor of death/transplantation than corresponding baseline hemodynamic values, except for PVRI.

O-P4

Prediction Model for a Severe Disease Course in Children with Suspected Myocarditis – "MYKKE-Score"

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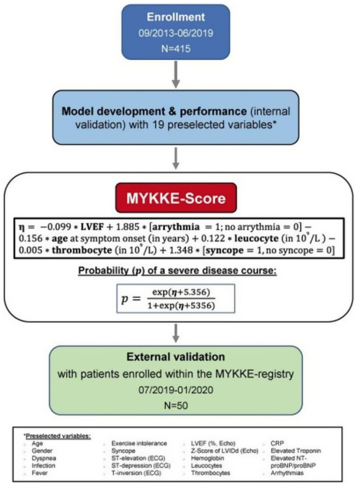
Introduction: As myocarditis is one cause of severe heart failure, the purpose of this study was to develop a validated model for the prediction of a severe disease course (SDC) in children with suspected myocarditis.
Methods: Symptoms and diagnostic parameters at admission where prospectively recorded from 22 centers within the MYKKE registry. A SDC was defined by inotropic therapy, malignant arrhythmia, mechanical circulatory support, heart transplantation and/or death during first admission or follow-up. After preselection of baselines characteristics, multiple imputation and logistic regression were performed to develop the MYKKE-Score, followed by a validation with new patients (see Figure).
Results: 415 patients included between September 2013 and June 2019 were employed (65% male, median age 13.6 (1.5-16.3) years, 43% experienced SDC). The final model with an AUC of 0.909 (0.900-0.943) included six covariates and revealed decreased odds for a SDC by an increase of the left ventricular ejection fraction (OR 0.906; p<0.001). The odds were 6.585 times higher for patients with documented arrhythmias and 5.144 times for patients with syncope (p<0.001, respectively). An increase of leucocytes increased (OR 1.130; p=0.004), and an increase of thrombocytes decreased the odds (OR 0.995; p=0.001). A further risk decrease was observed for increasing age (OR 0.856; p<0.001). The validation model consisted of 50 patients (age: 13.1 (1.4-16.0) years, 58% male, 40% with SDC) with an AUC of 0.797 (0.647-0.921).
Conclusions: The model of the MYKKE-Score enables a risk stratification of pediatric patients with suspected myocarditis and consequently might support clinicians to perform an individualized diagnostic and therapeutic management.



Poster
1. Arrhythmia/electrophysiology

P1
A rare case of neonatal long QT type 2 with atrioventricular heart block

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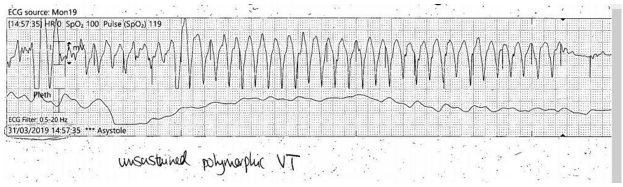
Introduction: 2:1 atrioventricular heart block is a rare association with long QT. Patients in this subset usually present in the neonatal or early infancy period and the mortality rate has previously been reported at 50%.

We present the case study of an infant who antenatally was noted to be bradycardic and aged 48 hours of life was noted to have a prolonged QT with 2:1 block.
Methods: A 2 day old infant was born to fit and healthy parents, he was their first baby following two previous miscarriages. Antenatally there had been concerns regarding bradycardia which was diagnosed as non-conducted atrial ectopics.

He was noted on the postnatal ward to be bradycardic with a prolonged QT and 2:1 block. He was admitted to the neonatal unit and shortly after this, developed non sustained broad complex ventricular tachycardia. He was commenced on isopranaline and transferred to his local tertiary cardiology unit.

The patient underwent pacemaker insertion on day 6 of life and commenced propranolol. The patient continued to have frequent episodes of Torsades de Pointes. Mexilitine and magnesium supplements were therefore commenced with resolution of the tachycardia. Genetics confirmed a likely pathogenic de novo KCNH2 mutation.
Results: Due to the age of this patient and his high risk presentation, he was discussed with several international cardiologists for advice regarding a sympathectomy. The smallest weight that this has been performed at is 13kg but this remains a treatment option in the future. For now our patient is well controlled on nadolol and has a loop recorder in situ which has not identified any further arrhythmias.
Conclusions: Long QT syndrome is a familial disease which is inherited in an autosomal mode of transmission or via a de novo mutation. The presence of a very long QT interval, 2:1 atrioventricular block, T wave alternans or congenital deafness imply a higher risk of sudden cardiac death. Beta blockers are the standard therapy to reduce the risk of life threatening arrhythmias but increase the risk of AV block, therefore many of these patients receive pacemakers and betablocker therapy. Due to this rare occurrence and high mortality, international guidelines are needed.



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P2

Accuracy of current accessory pathway localization algorithms in pediatric Wolf-Parkinson-White syndrome

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Introduction: Wolf Parkinson White (WPW) syndrome represents a major indication for electrophysiology study (EPS) and ablation therapy in pediatric patients. To enhance efficacy and reduce duration of EPS and burden through radiation, several algorithms were developed to predict accessory pathway (AP) location from 12-lead electrocardiogram (ECG). We aimed to assess accuracy determining AP localization from 12-lead resting ECG tracings using 10 published algorithms in pediatric Wolff-Parkinson-White syndrome. Furthermore, to evaluate accuracy of these algorithms in predicting AP localization in maximum preexcited ECG tracings from electrophysiologic study.
Methods: We used 10 AP determination algorithms as published in the literature. To assure the most reliable results the assessing pediatric cardiologist was blinded to AP location as confirmed by elcetrophysiologic study. The exact AP location, and laterality (i.e. left, right or septal location) were assessed according to each algorithm.
Results: Accuracy of predictive algorithms was stable throughout different age groups. The D`Avila, Boersma and Xie algorithms yielded highest accuracy to determine exact AP location and from 12-lead resting ECG tracings (57.7%, 53.8% and 53.8% respectively). In ECG tracings from electrophysiologic study the highest accuracy rates for exact AP localization were obtained using the Boersma algorithm (55.6%), while the algorithms by Boersma, Milstein, Iturralde and D`Avila showed equal accuracy rates for laterality (50%).
Conclusions: To predict AP localization in pediatric Wolff-Parkinson-White syndrome using 12-lead resting ECG tracings yields 58% accuracy for exact AP localization and 77% for laterality. We found the D`Avila, Boersma and Xie algorithms to be most accurate in our pediatric cohort.

Table 1: Accuracy rates for prediction of exact AP localization and laterality from 12-lead resting ECG tracings.

	Resting ECG			
	Exact		Laterality	
	%	Cohens K	%	Cohens K
Taguchi	50.0	0.328 (0.106-0.556)	57.7	0.341 (0.071 - 0.601)
Boersma	53,8	.436 (.236 - .638)	65,4	.477 (.222 - .749)
Arruda	34,6	.193 (.056 - .346)	46,2	.222 (-.041 - .461)
Iturralde	42,3	.304 (.105 - .492)	57,7	.356 (.120 - .617)
D`Avila	57,7	.445 (.252 .634)	76,9	.644 (.408 - .887)
Chiang	19,2	.103 (-.053 - .270)	34,6	.100 (-.104 - .312)
Fitzpatrick	34,6	.267 (.096 - .449)	42,3	.211 (.005 - .435)
Xie	53,8	.368 (.131 - .614)	65,4	.439 (.143 - .692)
Milstein	46,2	.139 (-.049 - .361)	46,2	.085 (-.125 - .318)
Lindsay	30,8	.203 (.026 - .369)	38,5	.151 (-.057 - .354)

Leg.: ECG = electrocardiogram. Values are given as percent and Cohens K with Confidence interval in brackets.

P3

Ajmaline provocation test in screening of Brugada syndrome in the Paediatric population-a single centre experience

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Introduction: BrS is an inherited condition with a dominant trait and shows age and sex-related penetrance. Diagnosis is made in patients with spontaneous ST-segment elevation with type 1 morphology or after provocative drug test with sodium channel blockers ie. ajmaline. Once diagnosed, the probands' family are advised to undergo clinical screening with ECG and provocative test if deemed necessary. We aim to review our paediatric population attending RBHSC who had undergone an ajmaline test, indications, positivity rate and the age of children who had a positive test.
Methods: Demographics of consecutive children who had undergone the provocative test between December 2010 and October 2018 were studied retrospectively. The children were reviewed at the clinic prior with standard and high chest lead ECG performed before planning for the provocative test. The test was performed in the theatre with advanced cardioresuscitation after obtaining parents' consent, supervised and led by Paediatric Cardiologist with Anaesthesia cover being present. A Paediatric Cardiac Physiologist supervised continuous ECG monitoring throughout the test. Ajmaline dose was calculated based on the childrens' weight of total dose 1mg/kg or (max 50mg) given over five minutes. Test stopped early if positive criteria were met or if there was any evidence of conduction problem.
Results: In total 38 children had the ajmaline provocative test. 58% were male. Mean age at the time of test was 10±2 years. 95% had family history of BrS followed by only 5% with family history of SUD/SADs. 75% of the proband in the family was diagnosed BrS after provocative test. The ajmaline test positivity rate was 40% and of these eight were male. Only 33%, five children were identified as having SCN5A mutation had a positive ajmaline test.
Conclusions: In our cohort, 40% of the children were diagnosed with BrS. No serious side effects were recorded. Children with negative ajmaline provocative test were followed till their puberty age of least 16 before discharge. All children remain alive and well to date. We performed the test for the right indication. We now have delayed arranging the test till their post puberty age as now seems to be the current consensus view.

P4

An Extremely Rare case of Long QT 15 in Paediatrics, the CALM before the storm

Jennifer Shortland (1), Richard Ferguson (1), Cecilia Gonzalez (1) Bristol Royal Children's Hospital (1)

Introduction: Long QT syndrome is a arrhythmogenic syndrome characterised by a prolongation of the QT interval on the electrocardiogram and is associated with life threatening arrhythmias. Patients are usually identified after presenting with syncope or after resuscitation of a cardiac arrest, often triggered by emotion or adrenaline or following family screening.

Long QT 15 is an extremely rare type of long QT Syndrome linked to a heterozygous mutation in the calmodulin gene CALM2 on chromozome 2p21. Patients usually present in infancy

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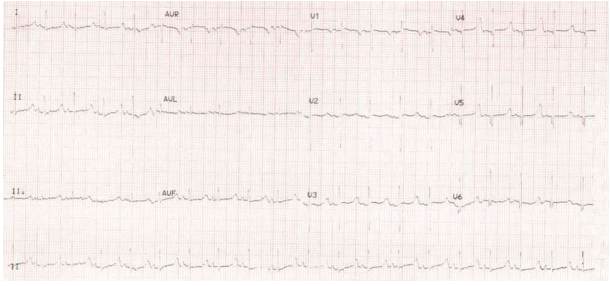
with ventricular arrythmias, T wave alternans and profound QT prolongation with a more severe phenotype than other long QT syndromes.
Methods: We performed a retrospective case review on our cohort of long QT patients within our tertiary institution. We were able to identify only one patient under the age of 16 who were genotype positive for a CALM2 mutation and analysed the patient's presentation, treatment and outcomes.
Results: Our patient presented aged 2 after a witnessed collapse at home. She received cardiopulmonary resuscitation at home by her parents. ROSC was achieved following DC cardioversion by paramedics. ECGs in her local hospital on presentation demonstrated a prolonged QTc >500ms. Following stabilisation the patient was commenced on Nadolol and remains clinically well with no neurological sequelae.

After review of the scarce literature, our patient was discussed with national and international ICC cardiologists and the decision was made to insert a loop recorder and commence Mexiletine.
Conclusions: Long QT syndrome is a life threatening arrhythmogenic condition. Mutations in the calmodulin genes CALM1, CALM2 and CALM3 result in LQT14, LQT15 and LQT16 respectively which make up <1% of patients with congenital long QT. Whilst there are variations in gene penetrance and modifier genes resulting in some patients with Long QT having a normal QT interval, patients with calmodulin mutations often present earlier with more severe symptoms. In adults the management of patients who present following a cardiac arrest includes betablockers and insertion of an ICD however management of infants brings more complex challenges as the complications associated with ICD insertion in infants has to be considered.



P5
Arrhythmic event detection of a wireless ambulatory monitoring system (WAMS) compared with 24-hour holter recording in pediatric patients.

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Introduction: Detecting the presence of cardiac arrhythmias in pediatric patients is a frequent need in pediatric cardiology consultations, although its detection is not always easy. This study has as objective to compare the performance diagnostic between the monitoring with an WAMS and the traditional monitoring with 24-hour Holter system.
Methods: Prospective single-center observational and analytical cohort study that includes pediatric patients under follow-up in the Pediatric Cardiology Unit, between November 1, 2019 and November 30, 2020. Patients with structural cardiological diseases, arrhythmogenic disorders, susceptible symptoms of cardiac origin and/or with systemic diseases with possible cardiovascular involvement were included. The included patients underwent both the conventional 24-hour holter-ecg and SMAI (Nuubo®) (figure 1). Informed consent was requested
Results: 27 studies have been carried out in 24 patients. The mean age of the patients was 8.67 years (median 7.38 years). The mean study time with the WAMS system was 287.25 hours (DT 226.78), and 21.26 hours (DT2.10) with the conventional Holter. The reasons for the study were: six patients with structural heart disease and cardiomyopathy (mean age: 10.6 years (DT 4.32 years)), 10 patients with arrhythmogenic primary diseases (mean age 6.9 years, (DT 4.32 years)), seven patients with symptoms of possible cardiological origin (six with palpitations and one with presyncope with exercise) with a mean age of 11.05 years (SD 2.28 years) and a female patient with a diagnosis of tuberous sclerosis with cardiac involvement. The age difference between the different groups, as well as the distribution by sex, was statistically significant. The most common electrocardiographic alteration was non-pathological sinus tachycardia in 100% of patients, followed by supraventricular ectopics (8 with WAMS, 6 with conventional Holter), ventricular ectopics (6 WAMS 6 conventional Holter), sinus pauses (4 WAMS 2 conventional Holter), ventricular tachycardia spells (2 WAMS, 1 conventional Holter) and supraventricular spells (2 WAMS, 1 conventional Holter).
Conclusions: The WAMS in this study has allowed the identification of 1 supraventricular tachycardia (in a patient with palpitations symptoms) and 1 ventricular one (in a patient with noncompaction cardiomyopathy), which had not been diagnosed in the conventional 24-hour registry.

P6
Atrial standstill in a newborn patient (case report)

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Introduction: Introduction: Atrial standstill (AS) is the absence of mechanical and electrical atrial activity. According to literature data, AS is rarely reported in pediatric population.
Methods: Methods: Here we present a case of diagnosed AS in a newborn patient. The patient C. was disclosed signs of heart failure (HF) at the age of 1 months. Holter monitoring showed idioventricular rhythm, episodes of ventricular tachycardia up to 207

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beats/min. The child was prescribed amiodarone as an antiarrhythmic therapy (AAT) for HF stopping. With AAT the bradycardia with heart rate 86 beats/min was recorded.

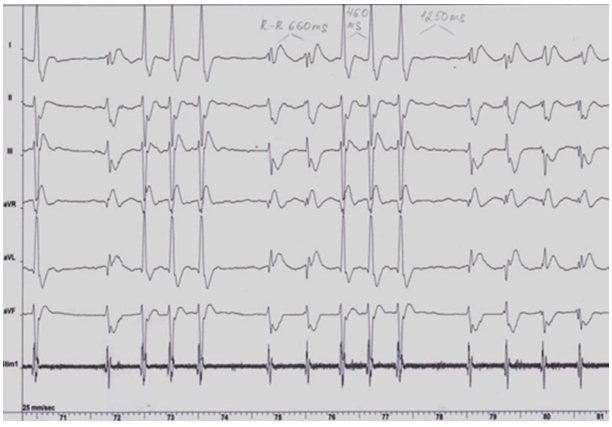
Transesophageal electrophysiological study (EPS) at the age of 3 months disclosed polymorphic idioventricular rhythm with heart rate 46–119 beats/min. Atrial commissure (A wave) was not recorded (Fig.1). According to echocardiography (Echo) left atrium volume was 314% of norm, right atrium volume – 250% of norm, ejection fraction (b) – 50%. Mechanical activity was not recorded.

Accounting decrease of average heart rate AAT was discontinued. Single chamber epicardial pacemaker was implanted in the VVIR mode with basic heart rate 80/180 beats/min. The surgery was performed by sub-xyphoid approach without sternotomy.

The patient was prescribed warfarin to prevent thrombus formation. The therapy with captopril and spironolactone continued. The patient was released from hospital in stable satisfactory state under the supervision of the cardiologist. During the year the state is stable without negative changes.

Results: Results: HF signs and electrocardiogram (ECG) non-sinus tachycardia were the first AS manifestations that required AAT. AS was diagnosed in a 3-month-old patient. AS was defined by the absence of P waves in surface and intracavitary ECGs, the absence of electrical activity at atrial pacing, and also the absence of mechanical activity during Echo.

Conclusions: Conclusion: Early diagnostics and pacemaker implantation allow avoiding unmotivated AAT and minimization of HF signs. Accounting high risk of progressive arrhythmogenic cardiomyopathy in children of early age it is necessary to define indications for AAT or pacemaker implantation in time.



P7

Biochemical markers of myocardial injury after radiofrequency ablation in children and adolescent

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Introduction: Radiofrequency (RF) ablation plays a key role in management of cardiac arrhythmias both in children and adults. The RF energy generates a lesion in the heart, leading to the formation of a localized cardiac muscle necrosis.

The aim of this study was to analyze potential diagnostic and prognostic role of biomarkers of myocardial injury in terms of catheter treatment of tachyarrhythmias in children. In future some of these biomarkers may be an effective tool in control of RF ablation parameters and, hence, provide hidher effic.

In subsequent studies using the correlation between the level of serum enzymes and the degree of myocardial damage admit to predict catheter ablation efficiency and monitoring the parameters of radiofrequency exposure to prevent the development of complications after catheter treatment.

Methods: The study included 58 children (24 females, 34 males), mean age at the moment of the procedure was 14,7 years (range 7–17 years) with heart rhythm disturbances (Wolf-Parkinson-White syndrome n = 37, atrioventricular nodal reentrant tachycardia n = 11, ventricular tachycardia n = 5, atrial tachycardia n = 5). Catheter ablation was performed during a period from July to October 2019. The level of the serum markers: myoglobin, creatine kinase MB (CK-MB), C-reactive protein (CRP), interleukin-8 (IL-8), tumor necrosis factor-alpha (TNF-alpha), heart-type fatty acid binding proteins (H-FABP), metalloproteinase-2 (MMP-2), metalloproteinase-9 (MMP-9), was determined before, 2 hours and 5 days after catheter ablation.

Results: The level of myoglobin ($p < 0.05$), MMP-9 ($p < 0.05$), CK-MB ($p < 0.05$), H-FABP ($p < 0.05$) was increased in 2 hours after radiofrequency ablation with a further decrease on the fifth day after treatment. Statistically significant changes in other serum markers were not identified.

Conclusions: Myoglobin, MMP-9, CK-MB, H-FABP have the potential to be clinically useful in evaluating myocardial damage volume after radiofrequency ablation

P8

Brugada syndrome in children less than 2 years of age

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Introduction: Symptomatic Brugada syndrome (BrS) presenting before age of 2 years is known but rare. The larger published series of children with BrS have very few children in this age group. We present 3 children presenting before 2 years of age with subsequent confirmation of diagnosis of BrS.

Methods: Case 1: A 19-month-old girl presented with sustained monomorphic ventricular tachycardia (mVT) during febrile illness. It was treated with intravenous (IV) Amiodarone in another hospital. Her ECG's intermittently showed Type 1 Brugada pattern. A non-tranvenous (NTV)ICD implanted for secondary prevention. A SCN5A mutation has been identified with family screening identifying more individuals with same mutation.

Case 2: A 19-month-old boy presented with ventricular fibrillation (VF) arrest in sleep during a viral illness and was successfully resuscitated. The baseline ECG was normal. Ajmaline challenge provoked Type 1 Brugada pattern. A NTV ICD was implanted for secondary prevention. A SCN5A mutation has been identified.

Case 3: A 16-month-old girl presented with haemodynamically stable monomorphic VT during a febrile illness and was treated with IV Amiodarone. Baseline ECG was normal apart from prolonged PR interval at 192 msec. She had further VT during same febrile illness. Ajmaline challenge was positive for BrS with induction of self-resolving polymorphic VT. A NTV ICD was implanted and she has had further appropriate therapies. She was commenced on oral Quinidine therapy. A SCN5A mutation has been confirmed.

Results: All 3 children below 2 years of age presented with VT/VF during viral illness with documented fever in 2. Interestingly 2 children had mVT which was treated with IV Amiodarone prior to the establishment of the diagnosis. 2 had normal baseline ECG's even during illness but positive Ajmaline challenge. 1 had transient

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spontaneous Brugada pattern during fever. All had confirmed SCN5A mutation and had ICD implanted for secondary prevention. 1 required treatment with Quinidine due to recurrent VT.
Conclusions: Brugada syndrome can present in children less than 2 years with malignant arrhythmias. Children presenting with VT/VF in this age should undergo Ajmaline challenge and genetic testing should be considered if no clear cause is identified for arrhythmia even if baseline ECG is normal.

P9
CLINICAL CHARACTERISTICS AND RISK FOR ARRHYTHMIC EVENTS IN PATIENTS UNDER 12 YEARS OF AGE DIAGNOSED WITH BRUGADA SYNDROME

Daniela Righi 1, Luigina Porco (1), Camilla Calvieri (1), Corrado Di Mambro (1), Simone Paglia (1), Anwar Baban (1), Massimo Stefano Silvetti (1), Mario Gnazzo (1), Antonio Novelli (1), Fabrizio Drago (1)
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Introduction: Brugada syndrome (BrS) is an inheritable disease with an increased risk of sudden cardiac death. Although several score systems have been proposed, the management of children with BrS has been inconsistently described. This study aims to identify the characteristics, outcome and risk factors associated with cardiovascular events in children < 12 years with BrS.
Methods: In this single-centre retrospective study, children < 12 years with spontaneous or drug/fever-induced type 1 Brugada ECG pattern were enrolled. Mean follow-up was 3.97 years.
Results: No significant differences were observed between patients with spontaneous and drug/fever-induced type 1 Brugada ECG pattern in terms of episodes of syncope, first-degree atrioventricular block, premature beats, non-malignant and malignant arrhythmic events (AEs). Female patients experienced more episodes of syncope ($p=0.035$) and malignant AEs ($p=0.035$) than male patients. A higher incidence of malignant AEs was observed in patients with syncope ($p=0.005$) and with non-malignant AEs ($p=0.047$). Malignant and non-malignant AEs were significantly more frequent in patients with positive electrophysiological study ($p=0.008$). Seven patients received an implantable loop recorder, 3 patients a cardioverter-defibrillator and 1 patient a pacemaker.
Conclusions: A spontaneous type 1 Brugada ECG pattern is not associated with a higher incidence of syncope episodes, first-degree atrioventricular block, premature beats, non-malignant and malignant AEs. Syncope events or non-malignant AEs are correlated to an increased incidence of malignant AEs. In addition, females seem to experience more frequently malignant AEs and episodes of syncope compared to males and SCN5A mutations are associated with the occurrence of malignant AEs.

P10
Co-existance of acute myocarditis and Arrhythmogenic Right Ventricular Cardiomyopathy as a cause of ventricular tachycardia – Case report

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Introduction: Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inherited, life-threatening disease and also a significant cause of premature death. It is characterized by the gradual replacement of myocardium by fibrous or fatty tissue in the right ventricle, accompanied by an increased risk of ventricular arrhythmia. Myocarditis is defined as a disease characterized by myocardial inflammation associated with myocyte necrosis. It is often associated with various types of ECG abnormalities including ventricular tachycardias.
Methods: We report a case of an 18-year-old man, with a history of chest pain and palpitation during physical effort, with no previous history of cardiac disease. Two years earlier he was hospitalized in the pediatric cardiology department for the first time. The symptoms were an episode of chest pain accompanying with palpitation and dizziness (without loss of consciousness) when playing a soccer match. He was diagnosed with ventricular tachycardia accompanied by LBBB morphology (left bundle branch block) which was alleviated with Amiodaron. Laboratory tests and cardiac magnetic resonance imaging were suggesting active myocardial inflammation, however, due to transthoracic echocardiography (dilated right ventricle with moderately reduced systolic function) and family history (sister of the father has a diagnosis of genetically confirmed ARVC), the ARVC was suspected.
Results: After examinations and additional testing, including follow-up CMR, the diagnosis of ARVC was established by a combination of characteristic abnormalities using a scoring system. 5 months after the first syndromes and impaired inflammation process, the ICD implantation was performed. Because of ventricular arrhythmias and pacemaker mediated tachycardia, the patient underwent ablation.
Conclusions: Myocarditis, as well as arrhythmogenic right ventricular cardiomyopathy, are life-threatening and largely unrecognized in children and young adults. The association between acute myocarditis and ARVC may increase the risks of ventricular tachycardia and sudden cardiac death. It is very important to complete the RV assessment through a CMR study in the context of ventricular arrhythmias.

P11
Confidence in paediatric ECG interpretation among junior doctors in a UK district general hospital

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Alder Hey children hospital (1)

Introduction: Formal training in paediatric ECG interpretation is rarely undertaken and reading them often fills junior doctors with fear.
Methods: An online survey of doctors in training - of all grades - currently working in paediatrics at a UK district general hospital was conducted. 10 questions related to confidence in paediatric ECG interpretation and the documentation of findings were completed.
Results: 19 junior doctors were emailed the link to the online survey, of whom 14 doctors participated. Of these 10 reported a requirement to assess paediatric ECG's at least weekly in their current clinical role. 93% of trainees had not received any formal training in paediatric ECG interpretation. No trainees were 'very confident' in the interpretation of a neonatal, pre-school or primary school aged child ECG. 36% of trainees did not write their findings on the ECG or in the notes. All of the trainees felt that they would benefit from further training.
Conclusions: There is currently limited undergraduate and postgraduate training for paediatric ECG interpretation resulting in varied knowledge and confidence among trainees. This small study shows that further training is not only a necessity – but would be welcomed by trainees.

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P12

Cryoballoon ablation of atrial fibrillation in adults with congenital heart disease – Siginificance of additional RF ablation of atrial tachycardia.

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Introduction: Prevalence of atrial fibrillation (AF) is increasing in adult patients with congenital heart disease (CHD). Experience using the 2nd generation cryoballoon to achieve pulmonary vein isolation (PVI) in adult CHD patients with AF is limited. Aim of the present study was to assess the value of PVI by cryoballoon in adult CHD patients with AF and to evaluate the significance of additional RF ablation of atrial macro-reentrant tachycardia (AT).
Methods: Retrospective data analysis, all patients with CHD and AF and pulmonary vein ablation using a Cryoballoon (Arctic Front Advance™, 28 mm, Medtronic) from January 2017 through November 2020 were included.
Results: A total of 12 adult patients with various types of CHD were included. Median age at the time of ablation was 57.5 years (IQR 51 – 61.5) years. 6/12 (50%) patients had had RF ablation of right atrial AT before. Median procedure duration was 242.5 (IQR 219 – 285) min. Median fluoroscopy time during EPS was 14.2 (IQR 12.6 – 23.7) min and median freeze time was 32 (IQR 28 – 35) min. Procedural success (defined as isolation of all pulmonary vein ostia) was achieved in all patients. In 1 subject, additional RF ablation of the lower right pulmonary vein was required. Additional RF ablation of left atrial AT was performed in 2/12 (17%) subjects and of right atrial reentrant tachycardia was performed in 5/12 (42%) patients, respectively. Median follow up was 26 months (IQR 19 – 32 months). Recurrence of AT and/or AF was observed in 6/12 (50%) patients. Recurrence of AT and/or AF was associated with cryoballoon use only. Patients with additional RF ablation of either right or left atrial macro-reentrant tachycardia were less likely to have recurrence of AF (p=0.021). Femoral artery injury requiring surgical repair occurred in one patient. No other major complications were observed.
Conclusions: Initial results after PVI using the cryoballoon plus additional RF ablation of AT was excellent (83% success) during medium-term follow-up. Success of AF ablation was unsatisfactory in all patients who had no additional AT ablation. Ablation of any AT in these patients is recommended.

P13

Electric Remodeling after percutaneous ASD occlusion with GCA device

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Introduction: Trans-catheter closure of ostium secundum atrial septal defect (ASD) is considered nowadays as the first-line treatment**.** Despite many studies were focused on cardiac mechanical remodeling, less is known on the electrical atrio-ventricular remodeling after ASD device implantation.

The aim of the study was to compare cardiac electrocardiographic changes in patients undergone to ASD closure with Gore Cardioform ASD (GCA) Occluder before and after closure.
Methods: We enrolled 15 consecutive pediatric patients (population mean age was 8.7±2.6 years) with hemodynamically significant ASD for transcatheter closure with the GCA device. We perfomed twelve-lead surface EKG the day before the procedure, 24 hours after, 1 month and 6 months after closure. In addition, an Holter-EKG was performed one and six months after the procedure. We focused on atrial changes by analysing type of cardiac rhythm, P-wave dispersion and PR interval at short- and mid-term follow up. Second, electric ventricular changes were considered, such as QTc interval and its dispersion. In addition, we performed EKG-Holter 24 hours at short and mid-term follow-up, in order to evaluate evidence of tachyarrhythmias or bradyarrhythmias (SVT, atrioventricular blocks).
Results: By analysing all twelve-leads surface EKG, we demonstrated the reduction of P wave dispersion at 24 hours after procedure (P=0.003), confirmed at 6 months after as well (P=0.04)(mean ΔP 0.028 s, 0.017 s, and 0.022 s, respectively).

This trend is also recorded at the ventricular level, as confirmed by the reduction in the mean dispersion of the Qtc interval (pre-procedure mean ΔQtc 39.8 s vs post-procedure mean ΔQTc 27.8 s (P = 0.002). The atrioventricular conduction showed a transient, insignificant prolongation after 24 hours with subsequent normalization after 6 months. A junctional rhythm was detected in one patient, which then presented transient episode of paroxysmal supraventricular tachycardia in the seventh postoperative day. This single arrhythmic complication was effectively treated pharmacologically with Flecainide for eighy weeks, without recidives after the withrawal.
Conclusions: This study showed an improvement in terms of intracardiac conduction homogeneity, which appears to be stable at 6 months after percutaneous procedure. Neither hemodynamically significant tachyarrhthmias nor bradyarrhythmias enveloped at short- and mid-term follow up.

P14

Epicardial cryotherapy of ventricular tachycardia as hybrid procedure in congenital heart disease

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Introduction: Because of prior surgical procedures ventricular tachycardia (VT) in congenital heart disease is of different mechanism, occurs in even younger age groups and often needs different approaches, especially since epicardial ablation is handicapped by post surgery adhesions.
Methods: We report the case of a 21-year-old patient with recurrent VT requiring electric cardioversion. He had previous surgical repair of congenital heart disease, truncus arteriosus communis A1, lately he got bioprosthetic aortic valve replacement in 2016. Shortly after this he developed VTs. Imaging examinations (CT angiography, MRI) showed an intramural diverticle from the aortic sinus towards the right ventricle (RV). A first attempt of 3D activation mapping showed a macroreentrant RV-VT in which the diverticle was involved. Radiofrequency ablation of the VT in the RV in 2016 was not successfull. Under antiarrhythmic medication his course was stable for 3 years, unfortunately frequency of VTs now increased.
Results: Therefore another electrophysiological study (EPS) was done. First a noninvasive body surface mapping was performed showing the course of the VT, arising from the aortic root, leading

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right through the diverticle structure to proceed in the lateral wall of the R.V. In the following endocardial EPS the activation map during VT (CL 440ms) missed almost half of cycle length in the area of the diverticle, so there was no endocardial target to terminate the macro reentry mechanism. To reach the diverticle a hybrid procedure was planned. Due to pulmonary homograft stenosis surgical valve replacement was necessary anyway. After sternotomy the VT was induced by epicardial pacing. The cryoablation probe was placed along the epicardial site of the diverticle structure. The tool was cooled by Argon gas down to -80°C, the VT terminated after application time of 30s, cryoenergy was administered for a total time of 4min. Afterwards the VT was non inducible. Surgery then was completed with pulmonary valve replacement. Follow up was uneventful with freedom from VT for more than 6 months.
Conclusions: Epicardial ablation for VT therapy also in patients with congenital heart disease can be essential. In specific cases the effort of open surgery in combination with epicardial cryotherapy or radiofrequency ablation should be considered.

P15
Exercise induced ventricular arrythmia in a patient with pectus excavatum: A rare case report
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Queen Silvia Childrenäs hospita, Cardiology Department (1)

Introduction: Pectus excavatum has been implicated as a cause of ventricular arrythmia in four case reports previously. We report a case of ventricular arrythmia which resolved after surgical correction of the chest deformity.
Methods: (Case report. Please see below)
Results: A 15-year-old boy with severe pectus excavatum was referred for cardiac evaluation by his surgeon. He was asymptomatic and trained judo intensely. The only discomfort he could recall was up to 10 second long episodes of mild chest pain at rest which occurred once a month in the last 4-5 months.

Echocardiography showed compression of the right ventricle but was otherwise unremarkable. ECG showed incomplete right bundle block. During exercise test he developed ectopic ventricular beats which presented as four paired, one triplet as well as frequent single contractions which were often seen as fusion beats. 24-hour ambulatory ECG monitoring showed 80 polymorphic ventricular contractions of which 13 were paired and one was a triplet. Furthermore over 800 broad QRS complexes were recorded which were interpreted as most-likely being fusion beats. MRI scanning confirmed a severe pectus excavatum with displacement of the heart and severe compression of the right ventricle. There were no signs of cardiomyopathy or other significant findings. Haller index measured by a CT scan was 4,2.

The patient underwent a Nuss procedure for correction of his chest deformity after which repeat holter registrations at 3, 5 and 19 months post-operatively were free of paired or triple ventricular beats and showed only scarce, isolated ventricular ectopic beats. The patient's mild symptom of chest pain was also resolved.
Conclusions: Patients with pectus excavatum resulting in cardiac displacement and compression of the heart can have potentially life-threatening arrythmias and should be referred for cardiac evaluation with echocardiography, exercise test and holter monitoring regardless of presence or absence of symptoms as subsequent surgical correction may resolve the arrythmia.

P16
Feasibility of zerofluoroscopy cryoablation of parahisian accessory pathways in children
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Introduction: Cryoablation is an established and safe procedure to eliminate parahisian accessory pathways (APs). Due to the adjacent conduction system ablation remains challenging. The aim of our study was to investigate the feasibility of zerofluoroscopy cryoablation in the pediatric population.
Methods: Ten cryoablation procedures of parahisian accessory pathways (40% retrograde, 20% antegrade and 40% antegrade and retrograde) were performed in nine patients. The median age was 12 years with a range of 6 – 17 years, 78% females. Fluoroscopy was used during three procedures with a median time of 0.7 minutes (range 0.2 – 4.5 minutes) to enhance catheter positions in these cases. The Ensite NavX system was used for 3D electroanatomical mapping, a 7 Fr steerable Freezor Xtra catheter with a 6 mm tip was used for cryoablation. At first the tip was cooled to -30 °Celsius. If safe elimination was seen, the tip was further cooled down to a temperature of -80 °Celsius. Breeze-thaw-breeze cycles of 240 seconds were applied as long as no temporary atrioventricular block occured.
Results: The acute success rate was 100%. The recurrence rate was 33% with preexcitation shown again in the electrocardiogram of three patients and one patient complained of tachycardias. Two patients underwent a second ablation with no recurrence so far. During the median follow-up of 330 days (range 2-1095 days) the total success rate was 80%. No procedure-related complications occurred.
Conclusions: Cryoablation is a safe method to eliminate parahisian accessory pathways in children. It has shown a good overall midterm success rate and can be performed without fluoroscopy in the majority of cases.

P17
Features of arrhythmia recurrences after radiofrequency ablation in preschool children
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Introduction: Introduction: It is well known that the risk of radiofrequency ablation (RFA) complications in children is inversely proportional to the patient's age and weight.
Methods: Methods: 240 RFA procedures were performed to 209 preschool patients with average age 4,5±2,3 years and average weight 19±5,8 kilos. 56% of children were with WPW syndrome, 21,5% - with atrial ectopic tachycardia, 4,8% - with atrioventricular nodal reentry tachycardia (AVNRT), 17,2% - with ventricular arrhythmia. Antiarrhythmic refractoriness and sings of tachycardia-induced arrhythmogenic cardiomyopathy were the indications for RFA in all patients.
Results: Results: 3 (1,4%) patients had "major" complications presented by injury of mitral valve with left-sided accessory pathway. 44 (21%) patients had recurrences of tachycardia and preexcitation. 20 children had immediate recurrences appeared during first 8 days

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after RFA. 24 patients had long term recurrences during first 6 months after RFA. The repeated RFA was performed to 40 patients. Additional RFA was performed to the patients after ineffective repeated RFA (n=2) and with repeated recurrences. The total efficiency including recurrences and repeated RFA was 91,3%.

To assess potential predictors of arrhythmia recurrences after successful RFA we compared patients with and without recurrences: by age, nosological arrhythmia type, parameters of radiofrequency (RF) exposure, localization of an arrhythmogenic zone. Only the highest RF energy had statistically significant differences in patients with (22,50 W; IQR: 20,00 - 30,00) and without recurrences (30,00 W; IQR: 25,00 - 40,00) (p=0,003). Regression analysis showed association of recurrences with parameters of RF exposure (p=0,039; OR 0,894; 95% CI OR 0,804-0,994). Decrease of the highest RF energy in 1 W increases risk of recurrences of 10,06%. This fact is associated with the aim of pediatric electrophysiologists to use sparing RFA parameters for minimization of complications.

Conclusions: Conclusion: Thus, the use of minimum effective parameters of RF exposure in small children decreases the risk of RFA complications but increases the risk of arrhythmia recurrences.

P18

Heart rate and heart rate variability during surgical stages to completed Total Cavopulmonary connection (TCPC)

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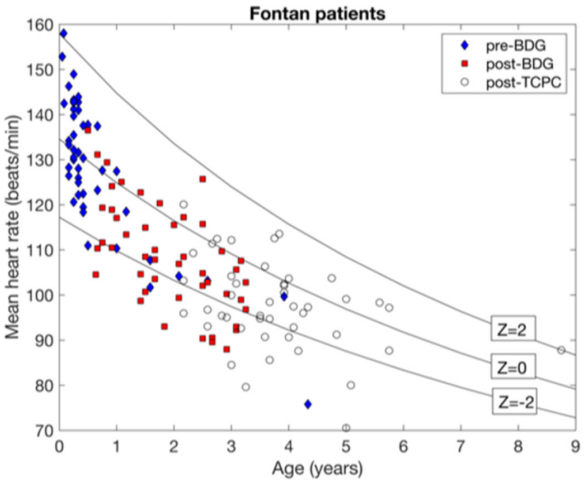
Introduction: Arrhythmia is related to heart rate variability (HRV), which reflects autonomic nervous regulation of the heart. We hypothesised that autonomic nervous ganglia, located at the junction of the superior vena cava´s entrance to the heart, may be affected during the bidirectional Glenn procedure (BDG), resulting in reduced HRV. Our aim was to investigate changes in heart rate (HR) and HRV in a cohort of children with univentricular heart defects, undergoing stepwise surgical procedures towards total cavopulmonary connection (TCPC), and compare these results with healthy controls of the same mean age.
Methods: HR and HRV was analysed from 24-hour Holter ECG recordings acquired from 47 patients before BDG, 47 patients after BDG, 45 patients after TCPC and 38 healthy controls. HRV was assessed by spectral and Poincaré methods. Age-related z-scores were calculated and compared using linear mixed effects modeling.
Results: In 6% (3/47) of the pre-BDG-, 30% (14/47) of the post-BDG- and 31% (14/45) of the post-TCPC- recordings, patients showed significant bradycardia (RR interval >2 Z-score). Total power HRV was significantly lower in patients before BDG when compared to healthy controls. Compared to healthy controls; patients before BDG had no significant difference in HR. Patients operated with BDG had significantly slower HR's (longer RR intervals) and reduced total power HRV compared to healthy controls. Patients post TCPC showed longer RR intervals and reduced HRV compared with healthy controls. In patients after TCPC HRV was decreased compared to before TCPC.
Conclusions: The main findings of this study were that a significant reduction in HR occurred in children with univentricular heart defects after BDG. Furthermore, reduction of HRV was found after complete TCPC surgery compared to pre TCPC. Our results indicate that autonomic regulation of cardiac rhythm is affected both after BDG and after TCPC. This may be reflected as, and contribute to, postoperative arrhythmic events.



P19

Histiocytoid cardiomyopathy management at the era of extracorporeal membrane assistance

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Introduction: Histiocytoid cardiomyopathy is a rare life-threatening pediatric condition characterized by incessant ventricular tachyarrhythmia, dilated cardiomyopathy but also sudden cardiac death in over 20% of cases. Management is challenging and aggressive therapy as catheter ablation, surgical resection and even heart transplantation is reported in the literature.
Methods: Over the last decade, 4 patients were referred to our institution for incessant ventricular arrythmia consistent with histiocytoid cardiomyopathy (HC) diagnosis. We retrospectively reviewed clinical data in order to highlight the management and document natural history.
Results: Mean patient age at diagnosis was 7 months [1.5-16], 3 were girls. Initial clinical presentation was incessant ventricular tachycardia associated with dilated cardiomyopathy in 3, 1 presented with resuscitated sudden cardiac arrest (SCA). Ventricular arrythmia were sustained rapid ventricular tachycardia with right bundle branch block pattern and superior axis (figure 1) or inferior axis and multiple PVCs with right bundle branch block pattern and inferior axis. Despite association of anti-arrhythmic drugs including amiodarone, hemodynamic deterioration required intubation for mechanical ventilatory support in 3 and extracorporeal membrane assistance (ECMO) in 2 patients. The patient diagnosed after SCA have implanted with an cardioverter defibrillator (ICD) at 9 months old. The first patient of this case series diagnosed 10 years ago, for who ECMO was not initiated, died of terminal cardiac

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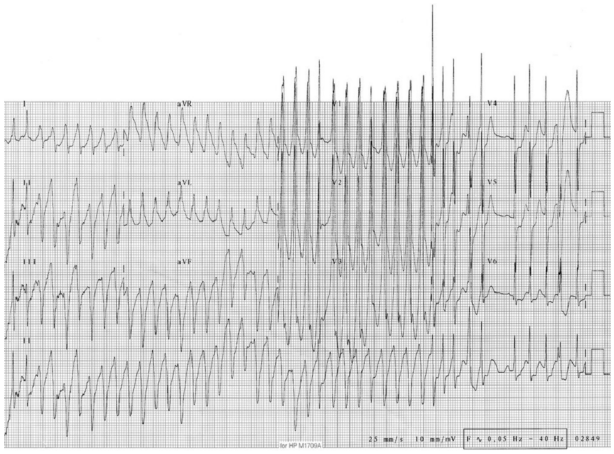
failure despite rate control. Arrythmia burden decreased for the 3 others patients, allowing to slowly tapped down medication.
Conclusions: HC is known to be a rare severe condition affecting mostly girls under 2 years old. Initial management can be dreadful. Still, aggressive therapy such as ECMO and ICD can allow to overcome the acute phase. Based on this small case series and report in the literature (1), secondary natural history seams favorable.

1: Villain et al. Incessant idiopathic ventricular tachycardia in infants. Arch Mal Coeur Vaiss. 1990 May; 85(5):665-71.



P20

How Safe Are Children With Covid-19 From Cardiac Risks? Pediatric Risk Assessment, insights from Echocardiography and Electrocardiography

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Introduction: Approximately 40 million individuals worldwide have been infected with SARS-CoV-2, the virus responsible for the novel coronavirus disease-2019 (COVID-19). Despite the current literature about the cardiac effects of COVID-19 in children, more information is required. We aimed to determine both cardiovascular and arrhythmia assessment via electrocardiographic and echocardiographic parameters.
Methods: We evaluated seventy children who were hospitalized with COVID-19 infections through laboratory findings, electrocardiography (ECG), and transthoracic echocardiography (TTE).
Results: We observed significantly increased levels of Tp-Te, Tp-Te/QT, and Tp-Te/QTc compared with the control group. 25 of 70 (35.7%) patients had fragmented QRS(fQRS) without increased troponin levels. On the other hand, none of the patients had pathologic corrected QT(QTc) prolongation during the illness or its treatment. On TTE, 20 patients had mild mitral insufficiency, among whom only five had systolic dysfunction (ejection fraction <55%). There was no significant difference between the patient and control groups, except for isovolumic relaxation time(IVRT) in terms of mean systolic and diastolic function parameters. IVRT was significantly lower than in the control group.
Conclusions: Despite all the adult studies, the effects of COVID-19 on myocardial function are not well established in children. The thought that children are less affected by the illness may be a misconception.

P21

Improving paediatric and neonatal ECG interpretations – a single centre quality Improvement project

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Introduction: General paediatricians are often the first physicians to assess children with possible cardiac pathology. Because paediatricians read paediatric ECGs, it is essential that they interpret these investigations correctly.
Aims and Objectives: To improve the accuracy of interpretation of paediatric and neonatal ECG amongst paediatric junior doctors; and to improve the documentation of paediatric ECG analysis.
Methods: Single centre study with a number of components. Firstly, retrospective review of notes of cases that had an ECG between March 2018 and September 2018 in paediatric ward and NICU. Secondly, semi-structured questionnaire to determine clinicians' perceived confidence interpreting ECGs and training needs and ECG quiz to assess current competence. Thirdly, QI project with three PDSA cycles run in three two monthly blocks between September 2018 and March 2019. Interventions with each cycle: A-Intervention 1: Implementing an easy ECG data collection sheet (ECG proforma) to be filled in with each ECG. B-Intervention 2: Teaching package on paediatric ECG interpretation method with introduction of interpretation tool. C-Intervention 3: Introduction of Crib Card for easy reference to be attached to new ECG interpretation proforma. Fourthly, clinical notes were audited (snapshot audit in January and February 2019) and the semi-structured survey was repeated post interventions to measure outcome.
Results: 15/54 case notes (28%) had ECG interpretations documented and completed in the initial epoch compared to 36/39 case notes (92%) in the second epoch ($p<0.001$). Accuracy in completion of 8 primary parameters in ECG analysis (rate, rhythm, axis, PR interval, QTc, P wave, QRS complex, T wave) also improved from (3/15) 20% of cases pre-interventions to (36/36) 100% post-interventions ($p=0.01$). (16/20) 80% of trainees reported being confident in interpreting paediatric ECG post-interventions compared to (10/20) 50% pre-interventions ($p=0.35$). Perceived training needs reported by (6/20) 30% of trainees post-interventions in comparison to (14/20) 70% pre-interventions ($p=0.06$).
Conclusions: An ECG teaching package with introduction of ECG proforma and reference crib card has significantly improved the accuracy and the documentation of paediatric ECG interpretation.

Measured Outcomes	Pre-interventions	Post-interventions	P-value
Documentation of ECG analysis	15/54 (28%)	36/39 (92%)	<0.001
Accuracy of interpretations of ECG analysis	3/15 (20%)	36/36 (100%)	0.01
Number of participants in survey	20/24 (83%)	20/24 (83%)	
Confident in ECG interpretations	10/20 (50%)	16/20 (80%)	0.35
Perceived training needs among trainees	14/20 (70%)	6/20 (30%)	0.06

Measured outcomes of QI pre- and post-interventions

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P22

Inherited arrhythmias in children with infectious diseases

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Introduction: The connection of a number of inherited arrhythmias with febrile body temperature is proved. Due to connection between fever and clinical manifestations (including ECG changes) of inherited arrhythmias there are additional opportunities for diagnostics of these life-threatening arrhythmias in infectious patients (including Covid -19).
Methods: 3584 ECGs of children with infectious diseases (average age 8.5±5.3 years old; boys - 57.5%, girls - 42.5%) were analyzed. Patients (pts) with QTc>440 ms or QTc<320 ms, complete right bundle branch block, left bundle branch block or its branches, atrioventricular block, ST elevation in the right precordial leads were given additional examination depending on the intended diagnosis (inherited arrhythmias): daily 12-channel Holter ECG monitoring, stress test, echocardiography. The family history was also clarified (cases of sudden cardiac death, syncope). The diagnosis was made on the basis of generally accepted diagnostic criteria and confirmed by molecular genetic analysis.
Results: ECG changes, which are typical for Brugada syndrome (BrS), type 1, were detected in 2 pts (0.05%). Long QT syndrome (LQTS) was detected in 2 pts too. Mutations in the SCN5A gene (exon 16 Arg893Cys, R878H) were identified in pts with BrS and in the KCNQ1 (exon 9 Trp379Ter) with LQTS. In pts with LQTS, sinus tachycardia was registered with the background of increased body temperature, which allowed to reveal long QT interval. 1 pt with LQTS is a female athlete. 1pt with BrS had been previously observed by a cardiologist in connection with grade I atrioventricular block. An increase in body temperature leads to disruption of the sodium ion channels which underlie the development of the BrS. In the case of LQTS, in our study, the increase in the QTc is most likely due to a change in heart rate rather than a direct effect of an increase in body temperature on the ion channels.
Conclusions: 1. BrS (type 1) was detected in 2 pts (0.05%) and LQTS in 2 pts (0.05%) at first. We consider that when taking an ECG from pts with fibril body temperature, this percentage may be higher. 2. ECG registration in pts with fever (including athletes) raises the probability of timely inherited arrhythmias diagnosis.

P23

Levothyroxine medication during pregnancy is associated with a lower mean heart rate in newborn infants

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Introduction: In clinical practice, occasional bradycardia below 80 beats per minute (bpm) seems to be common in newborns. In this study we retrospectively analyzed findings on 24-hour ambulatory ECG (Holter) monitoring of newborn infants whose main reason for referral to Holter monitoring was bradycardia.
Methods: Findings on Holter recordings performed in Tampere University Hospital in 2011 – 2017 to term-born infants up to 15 days of age due to bradycardia were gathered from patient records. Additional patient data were collected from patient records to assess factors affecting the rhythm and the heart rate. A total of 139 Holter recordings performed due to neonatal bradycardia were included in the analysis. The association between mean heart rate and pre- and postnatal factors was assessed by linear regression analysis.
Results: The mean (SD) age during the Holter monitoring was 6.4 (±1.7) days. The median (min-max) duration of the recordings was 19 (18-25) hours. The mean heart rate was 127 (±10) bpm and the minimum and the maximum 75 (±7) bpm and 200 (±14) bpm, respectively. In the linear regression analysis, the mean heart rate was positively associated with age in days ($p<0.001$) and female sex ($p=0.035$), whereas a negative association was found between the mean heart rate and maternal levothyroxine medication during pregnancy ($p=0.017$). Each consecutive day of age and female sex raised the mean heart rate 3 bpm whereas maternal levothyroxine medication accounted for a reduction of 6 bpm in the mean heart rate. Interestingly, only 12 mothers of these infants were under levothyroxine treatment. Umbilical cord blood thyroid-stimulating hormone (TSH) levels of the newborns showed no association with the maternal levothyroxine medication.
Conclusions: These results show that future studies on newborn heart rate should take maternal levothyroxine medication into account. Additional studies with larger sample sizes are needed to evaluate the duration of the effect and the effect size of maternal levothyroxine medication on the heart rate of a newborn.

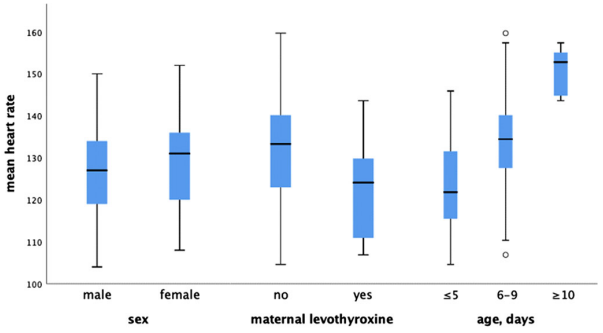

Figure 1. Factors associated with mean heart rate on Holter recording

P24

Longitudinal ECG changes following repair of tetralogy of Fallot

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Introduction: Patients with repaired tetralogy of Fallot (rTOF) carry residual sequelae including pulmonary regurgitation and dyssynchrony, putting them at risk for right ventricular dysfunction, arrhythmias and an increased risk of sudden cardiac death. ECG changes including QRS duration, QTc, QRS fragmentation and dispersion have been shown to increase risk of adverse events in rTOF and other diseases. We sought to describe these ECG changes longitudinally in rTOF.
Methods: Retrospective chart review of patients undergoing surgery at our institution 20000101-20181231. ECGs were analyzed by a single observer at 6 timepoints: pre-repair, discharge from

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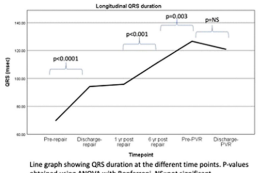
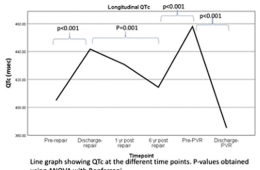
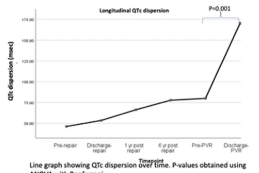
repair, 1 and 6 years post repair as well as pre-pulmonary valve replacement (PVR) and at discharge from PVR. Patients with less than 3 ECGs were excluded. Significant QRS fragmentation was defined as >=moderate. Data is presented as median (IQR).
Results: 94 patients were included with a median age of 11.9(9.5) years. Of these, 66% (n=62) were male and 19% (n=18) had a prior shunt. Repair was performed at a median age of 8.8(2.2) months with 36% repaired with valve sparing surgery, 32% transannular patch (TAP) and 32% with TAP and monocusp valve reconstruction. During this period 47% (n=44) underwent reoperation and 35% (n=33) underwent PVR at median ages of 6.4 and 7.9 years respectively. There was a significant increase in QRS from pre-repair to discharge from repair (p<0.0001), from 1 to 6 years post repair (<0.001) and from 6 year post repair to pre-PVR (p=0.003) but not from pre-PVR to discharge from PVR (see Figure). QTc increased from pre-repair to discharge-repair (p<0.001). QTc then decreased to the 6 yr post repair (p=0.001) and increased at the pre-PVR timepoint (p<0.001) with a significant decrease at discharge PVR (p<0.001). QRS dispersion increased from discharge-repair to pre-PVR (p=0.001) but didn't change with PVR, whereas QTc dispersion increased from pre-PVR to discharge PVR (p=0.001). Signficant QRS fragmentation increased following repair and through 6 year postop but decreased following PVR (see Figure).
Conclusions: Patients with rTOF have a significant burden of adverse ECG changes in follow up. While QTc and fragmentation improve early after PVR, other ECG changes persist.



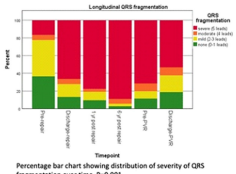
Line graph showing QRS duration at the different time points. P-values obtained using ANOVA with Bonferroni. NS=not significant



Line graph showing QTc at the different time points. P-values obtained using ANOVA with Bonferroni.



Line graph showing QTc dispersion over time. P-values obtained using ANOVA with Bonferroni.



Percentage bar chart showing distribution of severity of QRS fragmentation over time. P=0.001

P25

Not every child with palpitation has cardiac arrhythmia. Outcome of children referred with palpitation to local paediatric cardiology clinic

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Introduction: In adults, palpitations occasionally herald serious underlying cardiac events. However, palpitations in children typically arise from physiologic stimuli, rather than life-threatening causes. In addition, children with serious arrhythmias may report no palpitations.
Aim of Study: To review outcomes of children with palpitation who were referred to a clinic conducted by a paediatrician with expertise in cardiology.
Methods: A retrospective review of children referred by general practitioners and general paediatricians for assessment of palpitation to a paediatrician with expertise in cardiology. Hospital records of all children under 17 years of age who attended the clinic between April 2018 and October 2019 were reviewed.
Results: 38 patients referred over 18 months. 16 /38 (47%) referred by general practitioners and 18/38 (53%) referred by general paediatricians. 22/38(58%) females and 16/38 (42 %) males. Median age at referral 14 years. 2 /38 patients (5%) had family history of inherited cardiac conditions. One had a family history of Wolff-Parkinson-White syndrome, the other had a family history of dilated cardiomyopathy. 18/38(47%) had associated symptoms such as presyncope episodes and chest pain with palpitation. 1/38 (3%) had systolic murmur whilst 37/38 (97%) had normal cardiovascular examination. 3/38 (8%) received medications that may cause palpitation as a side-effect. 38/38(100%) of patients underwent 12 lead ECG. Of them, 35/38(92%)had normal sinus rhythm and 3/38(8%) had physiologic paediatric findings as junctional rhythm and incomplete right bundle branch block.100% of patients had 24-hour ECG monitoring with normal results. 31/38 (82%) had a trans-thoracic echocardiography. Of them, 21/31 (68%) had normal scan,8/31 (26%) had trivial tricuspid regurgitation, 1/31 (3%)had trivial mitral regurgitation and 1/31 (3%)had trivial pulmonary regurgitation. These findings considered physiologic. No patients (0%) had diagnosis of arrhythmias or any significant cardiac abnormality. 29/38(76%) were discharged, 9/38(24%) had follow-up.
Conclusions: Palpitation in children are more often benign than not. They could be managed in general paediatric setting unless they have history of syncope, congenital heart disease or cardiac surgery as they are at higher risk of having arrhythmia.

Parameter	Findings	Number	%
Sex	Male	16	42%
	Female	22	58%
Family History	+ve	2	5%
	-ve	36	95%
Symptomatology	Palpitations only	20	52%
	Associated presyncope	8	21%
	Associated chest pain	9	24%
	Associated vacant episodes	1	3%
Co-morbidities	Corrected TAPVD	1	3%
	No co-morbidities	37	98%
CVS Examination	Systolic murmur	1	3%
	Normal examination	37	97%
12 Lead ECG	Normal Sinus Rhythm	35	92%
	Incomplete RBBB	2	5%
	Junctional Rhythm	1	3%
Echocardiography	Normal	21	68%
	Trivial TR	8	26%
	Trivial MR	1	3%
	Trivial PR	1	3%
Outcome	Discharged	29	76%
	Local follow-up	6	16%
	f/u with adult cardiology	3	8%

Outcome of children referred with palpitation to local cardiology clinic

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P26

Out-of-hospital paediatric cardiorespiratory arrest – 13-year experience

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Introduction: Sudden cardiac arrest is defined as unexplained collapse without previous symptoms and corresponds to 25-30% of cases of sudden death in paediatrics. The success of cardiopulmonary resuscitation depends on cardiorespiratory recovery and survival without neurological sequelae. The prompt and efficient start of basic life support improves survival.
Methods: The authors characterize the extra-hospital cardiorespiratory arrest of cardiac aetiology of patients admitted to a Paediatric Intensive Care Unit, from January 2007 to December 2019 through retrospective analysis of clinical processes.
Results: Among 5357 admissions, there were 35 admissions caused by cardiorespiratory arrest: 19 respiratory aetiology (7 asphyxia, 8 drowning, 4 respiratory insufficiency), 3 neurologic causes, 1 death with unknown aetiology and 12 primary cardiac aetiology. Twelve children had cardiac aetiology for cardiorespiratory arrest, mean age of 11,4 years. The onset of basic life support ranged from 1 to 40 minutes. Nine patients required defibrillation. Two patients had relevant family history. Five cases had an aetiological diagnosis (three congenital heart diseases and one metabolic disease) and four had previous unrelated symptomatology. In seven cases, the cardiac arrest occurred during physical activity. The echocardiogram revealed the underlying diagnosis in five children and the electrocardiogram was diagnostic in one case. Genetic investigation was positive in four cases and three families were screened for the identified mutation. In two children, metabolic investigation was performed, although inconclusive. There were seven deaths. Concerning the five survivors, four had early basic life support.
Conclusions: Out-of-hospital cardiorespiratory arrest usually has a negative outcome. Early basic life support reduces morbidity and mortality. To prevent sudden death, it is essential to investigate treatable etiologies. A systematized approach is fundamental to improve patient care.

P27

Outcomes and Reoccurrence of Atrial Tachycardia Ablations in Adults with Congenital Heart Disease: A single centre perspective

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Introduction: Atrial arrhythmias are prevalent in Adult Congenital Heart Disease (ACHD) patients, are often difficult to control medically and have high recurrence rates with the majority occurring in the first year after ablation. In addition to the associated mortality, incessant arrythmias in this group cause a significant symptomatic morbidity. Reflecting these challenges, the 2020 ESC ACHD guidelines include formal recommendations on ablation for arrhythmia management. This study aimed to look at our experience of ablation procedures for atrial tachycardias in ACHD patients.
Methods: We conducted a retrospective study of all electrophysiology (EP) procedures in a ACHD department over a 25-month period (September 2018 – October 2020). Inclusion criteria were ACHD patients undergoing ablation for atrial arrythmias. Exclusion criteria were patients with a structurally normal heart, EP diagnostic studies, Ventricular Tachycardia (VT) studies and ablations. Data collection was from Electronic Patient Records. Outcome measures were success of procedure, complications and documented recurrence after ablation.
Results: 205 ablations were performed in total over the study time period. 108 procedures were excluded for the following reasons: 100 patients had normal cardiac anatomy, 8 were diagnostic or VT studies. The 97 ablations included were from 83 different patients. Median patient age was 44.5 years (range 19-79). Underlying cardiac structure included septal defects, Ebstein's, tetralogy of Fallot, Fontan, transposition and congenitally corrected transposition, AVSD, coarctation, pulmonary atresia and cyanotic single ventricle pathology. Procedural complications included one arterial femoral thrombosis. Three patients died in the follow up period for reasons unrelated to the ablation. There were 24 reoccurrences of arrythmias of which 7 were in atrio-pulmonary Fontan patients. The rate of recurrence in the whole cohort was 25% which is lower than previous published studies. Recurrence rate in the AP-Fontan patients was significantly higher at 58% (P= 0.004). There was no correlation with the more than one tachycardia circuit seen at the ablation and the risk of recurrence.
Conclusions: Ablations for atrial arrythmias are safe with increasing success rates and low complication rates. There is a risk of arrythmia recurrence and this risk is significantly higher in the patients with an AP Fontan circulation.

P28

QTc interval prolongation in paediatric patients admitted due to SARS-CoV-2 infection and their relationship with antiviral drugs

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Introduction: Many antiviral agents such as hydroxychloroquine have been studied to treat COVID-19, without being broadly accepted. Multiple side effects have been described, with QTc interval prolongation being one of the most worrisome. Literature on electrocardiographic alterations in COVID-19 is scarce. There aren't large samples of paediatric patients receiving hydroxychloroquine beyond Covid-19 to establish its relationship with electrocardiographic abnormalities. This study aims to describe QTc prolongation in relation to hydroxychloroquine and its association with other antivirals: lopinavir/ritonavir, remdesivir and azithromycin.
Methods: COVID-19 cases were detected by Polymerase Chain Reaction from nasopharyngeal aspirate and matched at a 1:2 ratio according to age and sex with controls not exposed to study drugs nor infected by COVID-19. Electrocardiograms, collected

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prospectively, were evaluated manually by the same person. QT intervals were calculated in 3 different beats and corrected with the Bazett formula. Electrocardiographic cut-off points were determined: before treatment, within 72 hours of the start and after more than 72 hours. Data were compared by using one-way ANOVA.
Results: 11 out of 48 paediatric patients admitted due to COVID-19 from March to July 2020, received antiviral therapy (22.9%) based on clinical evidence at the time; median age 9 years (IQR 10.5), 54.5% were male. Among the main underlying pathologies, congenital heart diseases (36.4%) and malignant haematological diseases (27.3%) stood out; 5 had received treatments potentially causing QTc prolongation. 10 patients (90.9%) received hydroxychloroquine, mostly in association with azithromycin (80%). 3 patients received lopinavir/ritonavir and one remdesivir. The mean of the baseline QTc interval was 418.5ms (407.4-429.6, 95%CI), before 72 hours was 424.6ms (398.1-451.2). A prolongation occurred after 72 hours: 439.7ms (408.5-470.9) but was not significant (p=0.253). 2 patients had long QTc interval before starting the treatment, and 4 after 72 hours. No patient presented arrhythmias.
Conclusions: A small proportion of patients received antiviral drugs. All had underlying diseases and a great proportion were taking drugs with an effect on QTc interval; this could contribute to QTc prolong. QTc prolongation occurred after 72 hours under treatment. Although only one patient had a QTc interval longer than 500ms (treatment was stopped afterwards) and none presented arrhythmias, QTc monitoring is advised.

P29
Rhythm and conduction disturbances early after transcatheter closure of perimembraneous VSD

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Introduction: Herein we present our experience focusing on the arrhythmias associated with transcatheter perimembranous ventricular septal defect (PmVSD) closure.
Methods: The medical records of patients undergoing transcatheter PmVSD closure between 2014 to 2019 were retrospectively analyzed. A total of 64 patients were selected for transcatheter VSD closure. These patients were taken to the catheterization laboratory and left ventriculography was performed. Six patients who had high pulmonary arterial pressure or were not suitable for transcatheter closure were eliminated. The remaining 58 patients had a PmVSD closure procedure.
Results: In total 58 devices including 17 Amplatzer duct occluder, 4 Amplatzer duct occluder II, 12 CERA perimembraneous VSD occluders, 8 Nit-Occlud Lê VSD coil and 17 Konar-MF device were used in 58 patients. Mean age of the patients was 8.41±4.46 (range 0.8-17) years. Insignificant and temporary rhythm disturbances including sinus tachycardia, short-term supraventricular tachycardia, ventricular or supraventricular ectopic beats and right bundle branch block were seen in 17.2% (10/58) of the patients during the procedure. Three patients (5 %) had more severe rhythm problems. Two patients had intermittant junctional rhythm immediately after *releasing* the device. In one of these patients, junctional rhythm regressed completely one day after the VSD closure procedure with Konar-MF and did not recur during the 1-year follow-up. However junctional rhythm persisted in other patient and CERA asymmetric device was surgically removed on the 8th day due to *dizziness* and fatigue while *walking* (chronotropic incompetence). Another patient whose defect was closed using ADO II developed left bundle branch block one day after the procedure. Echocardiography showed the device moving to the right side during the systole and to the original position during diastole similar to accordion bands. After 1 month of follow-up, the device settled more properly and left bundle branch block was improved. No patient had complete trioventricular block (CAVB) during or after the procedure
Conclusions: Although the incidence of CAVB decreases due to the preference of softer and more flexible devices for VSD closure, many rhythm anomalies may occur due to the location of the perimembranous defect. Therefore close monitoring is mandatory in terms of arrhythmia after transcatheter PmVSD closure.

P30
Temporary transvenous pacing in infancy

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Introduction: The approach to cardiac pacing in newborns depends on multiple factors including cardiac rhythm, structural heart disease and patient size. In infants with cardiac failure secondary to bradycardia, first line treatment includes medical management with isoprotenerol. Some patients require a longer term pacing strategy. Permanent epicardial pacing may be more suitable in smaller patients given concerns that early permanent transvenous lead implantation can result in longer term complications such as cardiac valve damage and venous occlusion. However, an epicardial system could result in unacceptable risk in low birth weight infants. It has been the strategy in our centre to stabilise the sick infant with a temporary transvenous pacing lead and external pacemaker generator and then, if ongoing pacing is indicated, convert to a permanent epicardial pacing system.
Methods: A retrospective review of temporary transvenous pacing interventions in children less than six months of age performed at a tertiary paediatric cardiology centre. Patients identified from an electronic patient management system and clinical notes reviewed.
Results: Three infants (two males) requiring temporary transvenous pacing were identified. Two with structurally normal hearts had congenital complete heart block secondary to maternal autoimmune disease. The third, VSD, had a family history of complete heart block and presented at two months following an out of hospital arrest. Age and weight at time of temporary pacing wire insertion were 1 day weighing 1700g (Patient 1), 8 days weighing 1400g (Patient 2) and 72 days weighing 5000g (Patient 3). The right femoral vein (Patient 1) and right subclavian vein were accessed. A 2Fr (Patient 1) or 5Fr temporary pacing wire was deployed in the right ventricle and attached to an external pacemaker generator. Complications included incorporation of the tip of the temporary pacing lead into the ventricular muscle, that did not required intervention (Patients 1 and 3) and pacemaker infection (Patient 2).
Conclusions: Temporary transvenous pacemaker lead insertion to stabilise the sick newborn prior to insertion of a permanent epicardial pacemaker system is a recognised and effective life saving strategy in bradycardic, haemodynamically unstable patients. We recommend removal of the temporary lead at the earliest safest opportunity to reduce associated complications.

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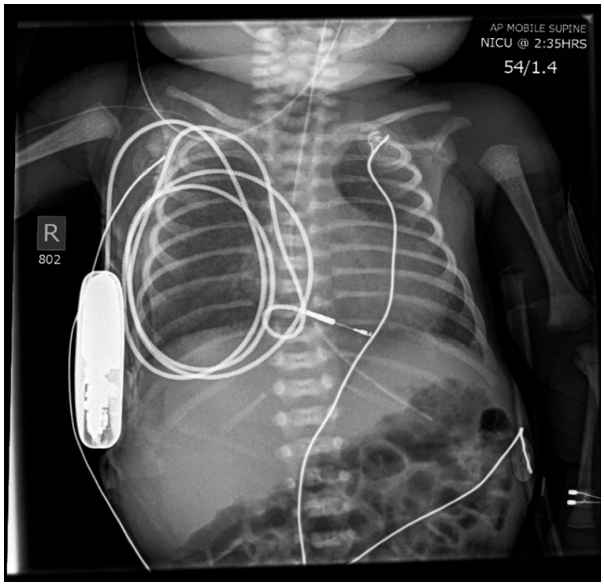

P31
THE ECG NUUBO® VEST FILLING GAPS IN PEDIATRIC ARRHYTHMIAS DIAGNOSIS

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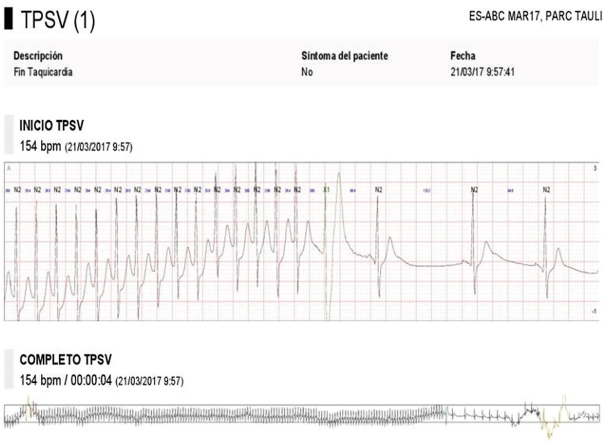
Introduction: Arrhythmias in pediatrics can be difficult in their identification and diagnosis the episodes of short duration, such as the difficulty in the communication of symptoms by the pediatric patient. The use of the 24-hour Holter offers limited performance, due to the rare occurrence of symptoms and their short time of use. The appearance of new wireless devices such as the NUUBO® allows continuous recording up to 12 weeks. We review the initial experience of our center with this device.
Methods: Wireless device with 3-channel ECU from NUUBO®. Retrospective review of all pediatric patients to whom monitoring with the NUUBO® device was indicated (August 2016-November 2019).
Results: The NUUBO® device was used in 30 consecutive patients. Median age of 9,5 years. Age ranges between (4 months and 17 years). With a mean follow-up time of 14 days (range of 7 - 21 days). 30 patients without structural heart disease with the following indications; palpitations, presyncopes/syncopes, pharmacological monitoring and BRUE. In 13 patients pathological events were documented. 8 patients with supraventricular paroxysmal tachycardia. 2 patients with WPW syndrome. 1 patient with non sustained Ventricular Tachycardia. 1 patient with frequent premature ventricular contractions in the context of presyncope. 1 patient with Second Degree AVBlock Mobitz type I with a sinus pause of 2.8 seconds was documented. The rest of the 17 patients did not present events or symptoms during the monitoring.
Conclusions: The monitoring with the NUUBO® system proves to be a useful tool in the diagnosis of pediatric arrhythmias, with a excellent diagnostic profitability as presented in this series



P32
The Leipzig Apple Heart Rhythm Study in Children

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Introduction: Currently, most supraventricular tachycardias are approachable by curative therapies like catheter ablation. Yet, there is still a group of very young children that has to be treated with antiarrhythmic drugs. It remains the physicians task to perfom routine check-ups using a standard 12- lead ECG and Holter monitoring. In the mean time parents remain relatively left alone with the worries about possible relapses of the tachycardia, which can only be resolved by visiting a specialized pediatric cardiologist. As Smartwatches like the Apple Watch gained increasing spread, the possibility to use a Smartwatch and record an iECG in case of suspected arrhythmia may help to provide valuable information to the treating physician, without having the patient to come to the hospital and necessary treatment adjustments would not be delayed until the next visit.Currently, the Apple Watch provides an FDA approved iECG function. Yet, the Apple Watch ECG is only evaluated for users from 22 years of age. The aim of this study was therefore to evaluate the performance of the Apple Watch`s 1- lead ECG in comparison to a standard 12- lead ECG in pediatric patients.
Methods: Consequtive patients treated in the Departement for Pediatric Cardiology, University of Leipzig, Heart Center are included into the study. All patients receive a patients history and baseline physical examination. Standard 12- lead ECG and 1 lead iECG using the Apple Watch will be performed. Overall 200 patients will be included into the study. ECG´s will be pseudonomyzed and analyzed blinded to the cardiologist. Cardiac rhyhtm will be classified and amplitudes and timing intervals will be analyzed for comparability.
Results: As this study is currently ongoing, actually the first 50 patients are recruited during a period of 4 weeks, these are preliminary results. The recruitment period will be finished in february 2020. First results show very good iECG quality with excellent matching to the amplitudes and timing intervals to the standard 12- lead ECG. ($p<0.01$).
Conclusions: Presumably, the modified lead 1 from the Apple Watch iECG corresponds seems to correspond well to lead 1 in the standard 12- lead ECG in children.

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P33

The prevalence of sinus tachycardia in Duchenne muscular dystrophy patients by Holter ECG monitoring

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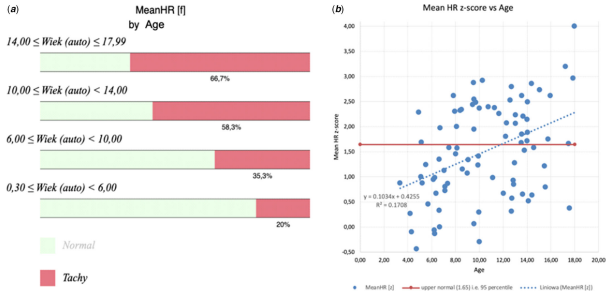
Introduction: Duchenne muscular dystrophy (DMD), a chromosome X-linked dystrophinopathy, affects not only skeletal and respiratory muscles but also the cardiac muscle, resulting in heart failure among the leading causes of death. Heart rhythm abnormalities evolve throughout patients' life, beginning with sinus tachycardia. This may be preceding cardiomyopathy by even 7 years. Although this phenomenon is known for more than 60 years its exact pathophysiology and the relationship with the development of heart failure remain unclear.
Methods: This is cross-sectional observational study presenting results of Holter ECG monitoring conducted in years 2017-2019 in patients with genetically confirmed DMD diagnosis. Heart rate, as well as presence of ventricular and supraventricular arrhythmias were assessed (using 24-hour count threshold of 10 and 200 respectively). Average heart rate was normalized for sex and age (z-score) using the reference data by Salameh et al. Statistical analysis was performed using Wizard Pro 1.9.38 (Evan Miller, Chicago, IL). All data are reported as mean ± standard deviation.
Results: Eighty-sixty studies in 67 patients aged 10.5±3.8 (3.4 – 18.0) were analysed. The recording time was 21.5±1.2 (19.1 – 24.0). The minimum and maximum HR were 61.8±7.9 (41.0 – 90.0) and 157.2±17.0 (122.0 – 197.0) respectively. The average HR was 100.4±10.3 (78.0 – 124.0) and the calculated z-score 1.5 ±0.9 (-0.4 – 4.0). In 40 (46.5%) instances sinus tachycardia was diagnosed. The tachycardia prevalence was dependent on age (chi2, p=0.032, Figure 1A) and becoming more severe with age (Pearson, r=0.413, p < 0.001, Figure 1B). Only benign arrhythmias were found. Singleton supraventricular paroxysmal beats were found in 33 (35%) studies, mean n 527±442., and singleton ventricular paroxysmal beats 6 (7%) studies, mean n 236±219. One case of transient AVB II Wenckebacha type was noted.
Conclusions: While severe arrhythmias are rare among the patients with DMD, sinus tachycardia is very common. It remains unclear whether it is a compensatory measure in the setting of heart failure as the growing with age prevalence would suggest or is it caused by progressive with the disease autonomous disfunction. Also unknown is the prognostic value of this finding. Longitudinal observations and multimodality analysis are key to elucidation of this phenomenon.



P34

The triangle of Koch may act as part of the slow pathway during atrioventricular nodal reentrant tachycardia

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Introduction: It has been considered that the posterior nodal extension (PNE) acted as the slow pathway during atrioventricular nodal reentrant tachycardia (AVNRT), and the posterior edge of the PNE was the target of catheter ablation in patients with AVNRT. However, optical mapping techniques revealed that conduction delay occurred between the ostium of the coronary sinus (CS os) and the tricuspid annulus (TA). Based on the results, it was speculated that in addition to PNE, the triangle of Koch might act as part of the slow pathway during AVNRT. The present study investigated the conduction property of the triangle of Koch in children to verify the hypothesis.
Methods: All the consecutive children who were referred to Kindai University hospital for the catheter treatment of arrhythmia between March 2016 and October 2019 were enrolled. Children with underlying diseases, including congenital heart diseases, were excluded. In advance to catheter ablation, the activation map of the right atrium (RA) during RA appendage pacing was created using 3D navigation system (CARTOIII®), and the area between CS os and TA was equally divided into three parts (from CS os to TA, zone 1 to zone 3 respectively). The activation potentials at the His recording site (His), CS os, zone 1-3 were recorded, and the conduction velocities from the pacing site (P) to each region were calculated and compared.
Results: A total of 31 children were enrolled. Their median age was 12 years (range 1 – 17), and 14 of them demonstrated dual AV node physiology. The atrial activation at zone 3 occurred 26.2 ± 12.6 ms later than that at His. The tendency of gradual conduction delay from CS os towards zone 3 was observed in all children, and multiple comparison tests revealed a significant difference in conduction velocities between P-His and P-zone 3 (0.84 ± 0.24 mm/ms vs 0.69 ± 0.17 mm/ms, P<0.001).
Conclusions: The results suggested that all the children had a slow conduction zone in the triangle of Koch, and that the triangle of Koch possibly acted as part of the slow pathway during AVNRT in children.

2. Basic science, Genetics

P35

Adaptive response to hypoxia in neonates with congenital cardiac defects: Role of myocardial expression of HIF-1 and EPO

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Introduction: Hypoxia Inducible Factor (HIF)-1 is the main mediator expressed in response to hypoxia. HIF-1 leads to the induction of a variety of adaptive genes, among others erythropoietin (EPO). EPO belongs to the interleukin (IL)-6 cytokine family. IL-6 is a cytokine that has been shown to cause myocardial injury in the setting of neonatal cardiac operations. In contrast, EPO has cardio-protective effects in vitro and in vivo. However, the role

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of the adaptive response to hypoxia in neonates with cardiac defect on post-operative outcome has not been investigated that far.
Methods: 15 neonates (mean age:8,4 days (1-26 days)(minimum-maximum) with congenital cardiac disease were investigated. A myocardial biopsy was taken from the right atrial appendage during cardiac operation, before connection to cardiopulmonary bypass. Expression of mRNA coding for HIF-1, EPO and IL-6 was studied by RT-PCR and normalized for 18S-mRNA expression. Data were analyzed with respect to preoperative oxygen saturation and clinical outcome variables.
Results: All patients were hypoxemic prior to surgery (SaO_2< 90%). mRNA coding for HIF-1, EPO and IL-6 was detected in all neonates. Levels of mRNA coding for HIF-1 and EPO correlated negatively with each others (p<0,0001). Expression of IL-6-mRNA but not that of HIF-1 correlated with preoperative SaO_2 (p<0,1). Neonates with worse postopertaive outcome showed higher myocardial expression of IL6-mRNA and lower expression of EPO-mRNA.
Conclusions: Our study shows that HIF-1, EPO and IL-6 are expressed at mRNA level in the myocardium of neonates born with congenital cardiac defect. The strong inverse relationship between mRNA-expression of HIF-1 and EPO suggests a retro-control of EPO expression by HIF-1 in neonatal myocardium. Our results suggest that EPO offers organ protection after neonatal cardiac surgery while IL-6 harms. Increasing EPO expression before and during cardiac surgery might therefore improve postoperative outcome in this fragile population

P36
Can meteorological change impact the incidence of Kawasaki disease?

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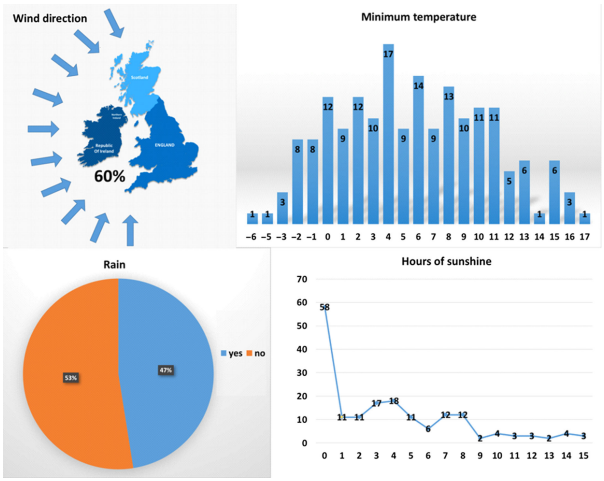
Introduction: Kawasaki disease (KD) is an acute febrile vasculitis that is the commonest cause of acquired heart disease in children. Suggestions of its association with different pathogens and genetic predisposition have been made. We have previously shown seasonal variation and tropospheric wind patterns are important in the aetiology. In this study, we sought to analyse different weather parameters and association with the incidence of KD.
Methods: A recent British Paediatric Surveillance Unit study identified a cohort of patients with KD. Of these data all consecutive children from 2013 in the UK were sub-analysed. Location and date of presentations of symptoms were prospectively reported. Using meteorological data from the National Meteorological Archive, minimum and maximum temperature, hours of sun, presence of rain or snow, wind orientation and speed at 24 hours prior to the presentation of symptoms were obtained. Statistical analysis was performed using SPSSv20.
Results: 180 patients were included in the study. 85% of cases happened when the minimum temperature was between -2 to 11oC. The number of patients according to maximum temperature did not follow any particular pattern. A significantly lower number of cases were found to correlate with reduced hours of sunshine (p=0.005), days when fog or overcast sky led to no sunshine accounted for 32% of the cases. The percentage of patients affected was not different depending on the presence of rain (47% rainy days, 53% dry days). 60% of cases presented when the wind was blowing from the west or the southern areas. 62% Kawasaki patients started symptoms when wind speed was less than 5 knots (9 km/h) and 93% when less than 10 knots (18 km/h). High velocity wind accounted for very little number of cases (7%). Findings are plotted in figure 1.
Conclusions: Low velocity wind directed from the south and the west, minimum temperature between -2 and 11oC, reduced hours of sunshine all relate to a greater incidence of patients presenting with Kawasaki symptoms in the UK. The presence of rain and maximum temperature appears not to impact the outbreak of this disease. These factors might help us to identify causal agents in this increasingly common disease.



P37
Cardiac involvement in Pediatric Multisystem Inflammatory Syndrome (PIMS) triggered by pandemic COVID19

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Introduction: We want to present the cardiac implication in PMIS, a new and serious SARS COV2 pandemic entity, that has similar characteristics with Kawasaki disease(KD) or Toxic Shock Syndrome and may develop a severe and potential fatal Macrophage Activation Syndrome(MAS), targeting major organs.
Methods: During pandemic, 7 patients were admitted into our clinic with prolonged unremitted fever and different clinical onset, 4 female and 3 males, with ages between 4 mo and 16 yo. All patients underwent clinical examination, ECG, serial Echocardiography, abdominal ultrasound, Cardio-pulmonary X ray or CT, complex lab tests. Two tested positive for COVID antibodies.

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Results: Two patients developed incomplete Kawasaki-like disease with slight dilatation of the coronary arteries and thrombocytosis over 1 million/mm3, treated with IGIV and Aspirin. PIMS with severe MAS, a rare complication was present in 5 patients admitted for fever and: diarrhea(1), rash(2), aseptic meningitis(1), encephalitis, seizures and coma(1), ground glass pneumonia(5), generalized purpura(1), appendicitis(2), ascites(1), colonic abscesses(1), arthritis mimicking Systemic Juvenile Arthritis(1), symptoms presented in different combinations, targeting 2 or 3 organs. Hepato-splenomegaly was palpable in all 5. Cardiac involvement was present at the beginning in 2 cases: pericarditis(2), mitral insufficiency(1), mitral and aortic insufficiency(1), and rapidly after admittance in 3 cases: cardiac tamponade and severe hypotension(1), pericarditis(1), myocarditis(1). Cardiac biomarkers: NT-proBNP and Troponins T and I were elevated in all cases, with more sensitive Troponin I. All 5 patients had: elevated CRP, Ferritin, D-Dimers, liver enzymes, triglycerides, low fibrinogen, falling ESR, leucopenia and thrombocytopenia(one patient with 5000/mm3 thrombocyte), abnormal coagulation. Interleukin 6(IL6) was high in all 7 patients, but in PIMS with MAS, the cytokine storm syndrome, was confirmed by highly elevated soluble IL2 receptor. This patients were promptly treated with IV corticotherapy, anticoagulants, IGIV and antibiotics with favorable evolution.
Conclusions: Cardiac tamponade, pericarditis, myocarditis, mitral and aortic insufficiency and Kawasaki-like disease with coronary artery dilatation were found in PIMS patients. Only two cases were positive for COVID Antibodies. Rapid recognition of PIMS and aggressive treatment of MAS prevent fatalities and determined a favorable evolution.

P38

Cardiac myosin binding protein-C variants in paediatric-onset hypertrophic cardiomyopathy: natural history and clinical outcomes

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Introduction: Variants in the cardiac myosin-binding protein C gene (*MYBPC3*) are a common cause of hypertrophic cardiomyopathy (HCM) in adults and have been associated with late-onset disease, but there are limited data on their role in paediatric-onset HCM. The objective of this study was to describe the natural history and clinical outcomes in a large cohort of children with HCM and pathogenic/likely pathogenic (P/LP) *MYBPC3* variants.
Methods: Longitudinal data from 62 consecutive patients diagnosed with HCM under 18 years of age and carrying at least one P/LP *MYBPC3* variant were collected from a single specialist referral centre. The primary patient outcome was a major adverse cardiac event (MACE).
Results: Median age at diagnosis was 10 (IQR: 2-14) years, with twelve patients (19.4%) diagnosed in infancy. Forty-seven (75%) were male and 31 (50%) were probands. Median length of follow-up was 3.1 (IQR: 1.6-6.9) years. Nine patients (14.5%) experienced a MACE during follow-up and five (8%) died. Twenty patients (32.3%) had evidence of ventricular arrhythmia, including 6 patients (9.7%) presenting with out-of-hospital cardiac arrest. Five year freedom from MACE for those with a single or two *MYBPC3* variants was 95.2% (95% CI: 78.6-98.5) and 68.4% (95% CI: 40.6-88.9), respectively (hazard ratio 4.65, 95% CI: 1.16-18.66, p=0.03). None of the twelve patients diagnosed during infancy experienced MACE during follow-up.
Conclusions: MYBPC3 variants can cause childhood-onset disease, which is frequently associated with life-threatening ventricular arrhythmia. Clinical outcomes in this cohort vary substantially from aetiologically and genetically mixed paediatric HCM cohorts described previously, highlighting the importance of identifying specific genetic subtypes for the clinical management of childhood HCM.

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Cardiac tumours in children: a referral centre experience

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Introduction: Cardiac tumours are extremely rare in children and may be primarily benign or malignant, or secondary to a metastatic process. Although more than 90% are benign, tumours may cause inflow/outflow obstruction or even be involved in embolization processes. Haemodynamic instability due to arrhythmias and cardiac tamponade or sudden cardiac death may be other related clinical findings. Although definitive diagnosis requires histological analysis, cardiac magnetic resonance findings permits a fairly accurate tumour type diagnosis. Dependent on tumour type, management may be conservative. However, surgical resection is advocated for symptomatic patients, and those with cardiac dysfunction and a higher risk of embolization.
Methods: The authors present the cases of cardiac tumours followed up at a Portuguese Paediatric Cardiology Referral Centre from January 2005 to December 2019. Diagnosis were made with echocardiography and/or cardiac magnetic resonance.
Results: During this period, 15 children were diagnosed with cardiac tumours, all benign. Rhabdomyomas were the most common type (nine, 56% associated with tuberous sclerosis), followed by fibromas (three), teratoma (two) and myxoma (one). Four patients with rhabdomyomas had prenatal diagnosis, all asymptomatic during follow-up. The remaining cases were diagnosed at a median age of 4 years. The reason for referral was a heart murmur in four patients, evaluation in the setting of tuberous sclerosis in three (asymptomatic), syncope (fibroma), acute pericarditis with pericardial effusion (teratoma) and an incidental image on chest X-Ray (fibroma). All rhabdomyomas derived from the ventricles or interventricular septum, excepting one which was in the left atrium. The three fibromas were in the left ventricle, the myxoma in the right atrium and the teratomas in the pericardium. Two fibromas were surgically resected, one due to left ventricular outflow tract obstruction and the other due to mitral valve impairment. The both pericardial teratomas were also resected as they were compressed the right atrium. There was no mortality among the patients. During follow-up one patient with tuberous sclerosis and rhabdomyomas developed frequent ventricular and atrial premature contractions and one patient with teratoma developed recurrent pericarditis.
Conclusions: All cardiac tumors in recent years have been benign, which is in line with the literature, with few comorbidities and no mortality.

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Catecholaminergic polymorphic ventricular tachycardia in a patient with Turner Syndrome

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Introduction: Patients with Turner Syndrome (TS) carry a higher risk for sudden cardiac death (SCD) than healthy age matched women, independently from the presence of congenital cardiac disease. Higher cardiac arrhythmogenic potential with, among others, long QT-interval and QT-wave dispersion has been reported in patients with TS that may be responsible for SCD. There is no evidence yet between the CPVT and the TS.
Methods: Clinical case

We report on the case of a 14 year-old adolescent with TS (XO) who had regular follow-up. Her treatment consisted of growth hormone. Previous cardiologic examinations were normal. She was re-evaluated after a period of 3 years. She complained about exercise intolerance. She never experienced syncope or palpitations. The clinical examination was normal apart of the short stature and overweight. ECG und echocardiography were normal. Stress ECG was performed on egometric bicycle. Under a load of 1,4 Watts/kg, HR rose up to 178/min. Polymorphic ventricular extrasystoles followed by several episodes of bilateral ventricular tachycardia (VT), a short episode of supra-ventricular re-entry tachycardia (SVT) and ventricular fibrillation occurred. Exercise was immediately interrupted. Patient converted spontaneously in a stable sinus rhythm. A treatment with the b-blocker Nadolol was introduced and titrated up to a dosage of 1,6 mg/kg/d. Magnesium supplementation and spironolactone were introduced. Under treatment, inconstant prolonged QTc interval at rest was observed (460-480 msec.), whereas stress ECG and Holter-ECG were normal with normal QTc measured 4 minutes after stress termination (435-455 msec.). Genetic testing shows a mosaic RYR2 mutation (c.13516G>C).
Results: Discussion

At the best of our knowledge, this is the first patient with TS with CPVT associated with a typical mutation of the gene RYR2 reported. The QT interval in CPVT patients is typically within the normal range. However, in some cases showing a RYR2 mutation, prolonged QT interval in the absence of genetically confirmed long QT gene mutation has been described.
Conclusions: This original report of CPVT in a girl with TS confirms higher cardiac arrhythmogenic potential in XO patients. This must be taken into account for the follow-up plan that should include regular stress test in these patients.

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Clinical and genetic profile of left ventricular non-compaction cardiomyopathy in children - own experience.

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Introduction: Left ventricular non-compaction (LVNC) is a genetically and clinically heterogeneous cardiomyopathy. Clinical presentation vary from mild cases to severe symptoms of heart failure, thromboembolic events and arrhythmias. Echocardiography and cardiovascular magnetic resonance (CMR) are important diagnostic tools. So far, pathogenic variants in 15 genes have been described as the cause of LVNC. The aim of study was a retrospective analysis of the clinical and genetic profile in children diagnosed with isolated LVNC in the years 2003-2019.
Methods: The study included 30 children, mean age 11.2years with suspicion of LVNC. The familial history, NYHA class, clinical symptoms, ECG, echocardiography, CMR were analyzed. The echocardiographic and CMR criteria for LVNC was the ratio of non-compacted to compacted layer >2 in systole and >2.3 in diastole, respectively. All patients underwent molecular analysis using the next generation sequencing (NGS) technique.
Results: The echocardiographic criteria for LVNC was fulfilled in 29 children, one patient had NC/C ratio 1.9. CMR study was performed in 27 pts, among which echocardiographic diagnosis of LVNC was confirmed in 24 pts, in 2 children NC/C ratio was borderline (2.0-2.1), and in 1 child LVNC was not confirmed. The spectrum of clinical symptoms included: heart failure in 7pts, ventricular arrhythmias in 8, supraventricular arrhythmias in 4, third degree atrioventricular block in 5, WPW syndrome in 2 and sinus bradycardia in 4pts. Pathogenic variants in genes correlated with LVNC were detected in 17 (56.7%) children. In 3 of them there was a mutation in the HCN4 gene, in 2 in MYH7, in 2 in TTN and in 2 in the RBM20 gene. In the remaining 8 patients, pathogenic variants were found in subsequent genes (TAZ, ACTN2, DES, ACTC1, EYA4, KCNQ1, PRDM16, HCCS).
Conclusions: Significant clinical problems in children with LVNC were heart failure, ventricular or supraventricular arrhythmias and atrioventricular conduction disorders. The molecular aetiology of LVNC has been confirmed in over half of the studied children. Our research indicate a good correlation between echocardiographic and CMR study but the diagnosis of LVNC remains a challenge due to the lack of standardized criteria for the pediatric population.

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Clinical features and outcomes of patients with heterotaxy syndromes in Western Australia

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Introduction: Heterotaxy syndromes encompass left and right atrial isomerism (LAI and RAI respectively) and are associated with variable cardiac and non-cardiac anomalies influencing outcome. RAI is usually associated with complex congenital heart disease (CHD), early mortality with surgical intervention and low survival rates. LAI is less commonly associated with complex CHD but are at greater risk of atrioventricular block. The objective of this study was to review the clinical features and outcomes of patients over a sixteen-year period diagnosed with LAI or RAI in Western Australia (WA).
Methods: A retrospective review was performed of patients diagnosed with heterotaxy between 2003 and 2018; representing all cases in WA via the only tertiary centre for cardiology in the state.
Results: 22 patients were diagnosed with LAI (12 patients, 55%) or RAI (10 patients, 45%). Common cardiac associations are outlined in Figure 1.The majority of RAI cases (80%) were antenatally diagnosed. All patients with RAI had complex CHD. Asplenia was diagnosed in 8 patients (80%). Most RAI patients (90%) underwent surgical interventions including repair of total anomalous

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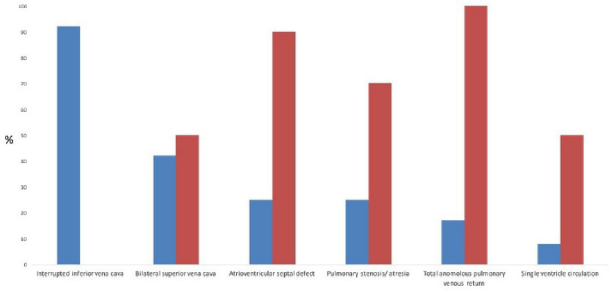
pulmonary venous drainage (50%), Blalock-Taussig Shunt (BTS, 50%) and Bidirectional Cavo-pulmonary Shunt (30%). Of the RAI cohort, only 2 (20%) patients reached Fontan completion. Four (40%) patients with RAI are deceased. 3 had residual pulmonary venous stenosis with mortality occurring before 4 months. Survival for patients with LAI was 92%. 50% patients with LAI required cardiac surgery including atrial septal defect closure (25%), aortic arch repair (17%), BTS (17%) or complete atrioventricular septal defect repair (8%). 25% developed complete heart block with pacemaker placement (17%). Extracardiac manifestations included polysplenia (83%), malrotation (42%) and biliary atresia (17%).
Conclusions: The outcomes of patients with RAI continues to be poor, with 60% survival and only 20% making it to Fontan completion. By comparison patients with LAI have significantly lower morbidity and mortality. The management of heterotaxy continues to be challenging due to widely associated cardiac and extracardiac manifestations.



P43
Clinical profile and outcome of restrictive cardiomyopathy in children: a single center experience.
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Introduction: Restrictive cardiomyopathy (RCM) is a rare in childhood primary disease of the myocardium. It is characterized by increased myocardial stiffness and rapid rise in ventricular filling pressures reflected in both the systemic and pulmonary circulations. The genetic basis of RCM remains largely unknown although mutations in sarcomeric genes have been identified as causative in a subset of affected patients. The aim of study was to evaluate the clinical course, molecular etiology and outcome in our group of children with RCM.
Methods: Retrospective analysis of 11pts (6 males,5 females), mean age 9yrs with RCM diagnosed from 2004 to 2017. Mean follow-up was 6yrs. Patients demographics, clinical symptoms, family history of cardiomyopathy, echocardiography, ECG, 24-h Holter ECG, cardiac catheterization and genetic tests results were analyzed. Familial evaluation was performed on first-degree relatives.
Results: In the analyzed group, mean age at the diagnosis of RCM was 3yrs. Five patients (45%) had a family history of cardiomyopathy and sudden cardiac death associated with RCM (n=4) and DCM (n=1). At the time of diagnosis in 9 children symptoms of heart failure were present. During follow-up, in 5pts arrhythmic events occured: 4 children had recurrent episodes of ventricular tachycardia, in 1pt resuscitated cardiac arrest appeared because of a ventricular fibrillation and cardioverter-defibrillator was implanted. In 7pts progression of heart failure symptoms was observed, in 2 also features of pulmonary hypertension. Five patients (45%) were qualified for heart transplantation. Three children had a heart transplant, in one patient sudden cardiac death occurred while waiting on the transplant list. In 7pts genetic tests were performed. Sarcomere protein gene mutations were identified in six patients (86%): in 4pts in the cardiac troponin I gene *(TNNI3*), in 1 in the filamin-C *(FLNC)* and in 1pt in the four genes: tropomyosin alpha-1 *(TPM1),* desmin (*DES*), titin (*TTN*), desmoglein-2 *(DSG2).*
Conclusions: RCM has a poor prognosis in children therefore pediatric patients should be considered for early cardiac transplantation. Sarcomere protein gene mutations are an important cause of idiopathic RCM in childhood. The identification of RCM in a child should be an indication for screening in family members.

P44
Congenital heart disease: a restrospective analysis from a tertiary referral centre
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Introduction: Congenital heart disease is the most prevalent congenital malformation contributing to perinatal mortality. The aim of this study was to characterize its incidence and to establish survival in the first year of life.
Methods: Retrospective analysis of cases born in a central maternity hospital, diagnosed up to 72 hours after birth, during a period of 16 years. Results considered statiscally significant if $p < 0.05$.
Results: Of the 47198 neonates born during the study period, 297 had congenital heart disease, 16% associated with syndromes or extra-cardiac disease. The incidence was 6:1000. Left to right shunt lesions accounted for 211, followed by cyanotic (n = 46), non-cyanotic obstructive (n = 31) and miscellaneous (n = 9). Coarctation of the aorta was positively correlated to gestational diabetes ($p = 0.014$). Prenatal diagnosis contributed to 26%, mostly in the cyanotics, itself related to mortality ($p < 0.001$). Atrial septal defect were found more commonly in females ($p = 0.02$). Mortality due to heart disease was 3,4%. Overall survival of cyanotic disease cases was 88%, 81% and 78% at 28 days, 6 months and 1 year of age, respectively. For the miscellaneous cases was 91%, 91%, 90%, the obstructive cases was 97%, 97%, 97% and for the left to right shunt diseases was 99%, 98%, 98%, respectively.
Conclusions: The incidence of congenital heart disease was 6:1000, mostly left to right shunt lesions. Heart disease accounted for only half of deaths. Cyanotic diseases carry a higher nonspecific mortality rate, usually progressive during the first year of life.

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Differential secretion of PF4 and platelet derived TGF-β by in children with congenital cardiac defects

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Introduction: Platelet-derived TGF-βis a cytokine that regulated fibrosis and has been implicated in cardiac remodeling and dysfunction secondary to ventricular pressure overload. The role of platelet-derived TGF-bin myocardial remodeling in children with congenital cardiac defect has not been investigated yet. Therefore, we aimed to investigate platelet secretion of TGF-bin children with congenital cardiac disease and pressure overload of the right ventricle.
Methods: Ex vivo activation of thrombocytes was performed in 11 infants with native large ventricular septum defect (VSD) (n= 5) or tetralogy of Fallot (TOF) (n= 6). TGF-band PF4, a marker of platelet activation, were measured. Ten age-matched otherwise healthy children in whom blood was withdrawn during scheduled INT surgery served as controls.
Results: TGF-b-and PF4-secretion was not different among patients and was similar in patients and in controls. Control subjects showed the highest ratio TGF-b/PF4 (3.9 ± 0,7 (mean ± SEM)), in comparison to patients with VSD (2,6 ± 1,17) and patients with Tetralogy of Fallot (2,52 ± 0,69). There was no significant difference between both patient groups but between patients and controls (p= 0.0014).
Conclusions: Our results show differential secretion of TGF-bin children with congenital cardiac defects in comparison to healthy children. Lower TGF-b/PF4 ratio in patients than in healthy controls suggests exhaustion of TGF-b secretion that might be secondary to chronic platelet stimulation by intracardiac turbulent flow and might impact the mechanisms of myocardial remodeling in this context of congenital cardiac disease.

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Experimental model for deep hypothermic cardiocirculatory arrest with peripheral cardiopulmonary bypass in swine.

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Introduction: Children with congenital heart disease usually undergo cardiac surgery, often performed under cardiopulmonary bypass (CPB) with deep hypothermic circulatory arrest (DHCA). Existing animal models, instrumental for the study of CPB/DHCA in this population, usually involve sternotomy and cardioplegia. Our aim was to develop a CPB model with DHCA in infant pigs, avoiding sternotomy and cardioplegia.
Methods: Seventeen female piglets (8.33 ± 0.82 kg), separated into anesthesia controls (n=7), and intervention group (CPB/DHCA, n=10), were intubated and mechanically ventilated. Isoflurane and propofol anesthesia, analgesia, fluids and antibiotics were provided. ECG, temperature, hemodynamic, tissue perfusion and respiratory parameters were recorded. CPB cannulation was performed through the right external jugular vein and right common carotid artery, the animals were then cooled to 22°C, and arrested for 75 min. After the cardiac arrest, the animals were rewarmed (36°C), separated from bypass and anesthesia was maintained for 4 hours with inotropic support. Control animals were placed under the same anesthetic and ventilatory regime, but no CPB or DHCA was performed.
Results: CPB/DHCA animals recovered spontaneous circulation and survived the required 4 hours of postoperative anesthesia.
Conclusions: This less invasive model successfully recreated a CPB/DHCA procedure with good perfusion indicators, without sternotomy, central cannulation or cardioplegia. It avoids technical challenges, complications, and stress factors (to the animal and the researcher) inherent to the sternotomy and central cannulation CPB.

P47

FISH for identification and visualization of microorganisms in heart valve tissue derived by cardiac biopsy in culture-negative IE – is it feasible?

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Introduction: Identification of the causative organism in infective endocarditis (IE) can be difficult, especially in right-sided endocarditis, but it is crucial for patient treatment and survival. FISH (Fluoreszenz in situ Hybridisierung) is a molecular, culture-independent technique that allows to identify and visualize microorganisms within tissue and to recognize their morphology, number and activity. We analysed the diagnostic benefit of FISH/PCR by comparing its results to those of standard diagnostic tests in two complex cases with IE.
Methods: In two patients, where balloon valvuloplasty was performed to treat severe pulmonary stenosis and signs of right ventricular failure, tissue specimens were obtained by cardiac biopsy, because of clinical signs and suspicious ultrasound imaging for IE. FISH/PCR samples were fixed in FISH fixation solution in the cath lab and sent to the Biofilmcenter, Deutsches Herzzentrum Berlin. In each case additional blood culture were taken in cath lab from pulmonary artery. In total more than 12 in each Patient. Patient 1: 21 Years female. Pulmonary atresia and VSD. Melody valve 2009. No improvement after 3 weeks of therapy. Blood cultures sterile. Biopsy from Melody™ Pulmonary Valve. Detection of Streptococcus salivarius. Patient 2: 22 Years male. Ross procedure 2015. Blood cultures sterile. Biopsy from pulmonary homograft. Detection of Staphylococcus epidermidis.
Results: In two cases of culture negative IE, a single causative microorganisms could be identified; no mixed infection was detected by FISH/PCR. In our second case S. epidermidis (part of regular skin flora) was detected - in blood culture contamination would be supposed. Thus, we were able to identify microorganisms in cases where standard diagnostic tests failed to provide sufficient results.

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Conclusions: In complex, apparently negative cases where under antibiotic treatment, bacteria organized in biofilms can be metabolically less active but viable. These populations are often missed in culture. However, an approach combining histopathological and molecular techniques (FISH/PCR) may lead to a definite final diagnosis which may guide targeted antibiotic therapy.

P48

Hereditary hypertrophic cardiomyopathy in children and young adults - the value of reevaluating and expanding gene panel analyses

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Introduction: Sudden cardiac death (SCD) and early onset cardiomyopathy (CM) in the young will always lead to suspicion of an underlying genetic disorder. Incited by the rapid advances in genetic testing for disease we have revisited families, which previously tested "gene-negative" for familial predominantly pediatric CM, in hopes of finding a causative gene variant.
Methods: 10 different families with non-syndromic pediatric CM or hypertrophic cardiomyopathy (HCM) with severe disease progression and/or heredity for HCM/CM related SCD with "gene-negative" results were included. The index patient underwent genetic testing with a recently updated gene panel for CM and SCD. In case of failure to detect a pathogenic variant in a relevant gene, the index patient and both parents underwent clinical (i.e. partial) exome sequencing (trio-exome) in order to catch pathogenic variants linked to the disease in genes that were not included in the CM panel.
Results: The mean age at clinical presentation of the 10 index cases was 12.5 years (boys 13,4 years, n=8; girls 9 years, n=2) and the family history burden was 33 HCM/CM cases including 9 HCM-related SCD and one heart transplantation. In 5 (50%) families we identified a genetic variant classified as pathogenic or likely pathogenic, in accordance with the ACMG criteria, in *MYH7* (n=2), *RBM20*, *ALPK3* and *PGM1*, respectively, and genetic variants of unknown significance (VUS) segregating with the disease in an additional 3 (30%) families, in *MYBPC3*, *ABCC9* and *FLNC*, respectively.
Conclusions: Our results show the importance of renewed thorough clinical assessment and the necessity to challenge previous genetic test results with more comprehensive updated gene panels or exome sequencing if the initial test failed to identify a causative gene for early onset CM or SCD in children. In pediatric cardiomyopathy cases when the gene panel still fails to detect a causative variant, a trio exome sequencing strategy might resolve some unexplained cases, especially if a multisystemic condition is clinically missed.

P49

Hypertrophic Cardiomyopathy in the first year of life: experience from a tertiary referral centre

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Introduction: Hypertrophic cardiomyopathy (HCM) in the first year of life is an extremely rare disorder affecting 3,6 per 1 million children. In this age group, prognosis is considered poor when compared with older ages, with a mortality rate that can reach 89%, especially when presenting with congestive heart failure.
Methods: The authors present a retrospective analysis of 14 cases of HCM diagnosed over the past 20 years during the first year of life and followed up at a portuguese paediatric cardiology tertiary referral centre. Half of this population was diagnosed during the neonatal period. Secondary causes of HCM such as gestational diabetes or systemic arterial hypertension were excluded from the study.
Results: Five patients presented with signs of congestive heart failure (3 neonates), although heart murmur was the most common finding. Six had concentric hypertrophy. Of these, 3 had left ventricular outflow tract obstruction. Sarcomeric HCM was identified in 4 cases, all of which with positive family history (29%). There was 1 case of Noonan Syndrome with associated pulmonary valve stenosis and atrial septal defect. In 5 cases (36%), the left ventricle hypertrophy resolved by the second year of life (1 of them with sarcomeric HCM genotype). In the remaining cases, one evolved into left ventricular noncompaction and another developed a severe restrictive pattern, the latter, 16 years of age, recently underwent a heart transplant. One of the sarcomeric HCM cases had a subcutaneous implantable cardioverter defibrillator implanted at the age of 8 years for secondary prevention (aborted sudden cardiac death). Only 1 child died (neonatal onset group) at three months of age with rapidly progressive biventricular hypertrophy of unknown cause (ongoing necropsy – figure 1), with refractory congestive heart failure whilst awaiting cardiac transplantation.
Conclusions: The four above-mentioned cases with a more complicated clinical course had early onset congestive heart failure symptoms (plus the Noonan syndrome case), whereas, none of the patients in which left ventricle hypertrophy resolved spontaneously presented with congestive cardiac failure. A third of the sample had family history of sarcomeric HCM (29%), highlighting the importance of early follow-up in this group of patients.

A 3-month-old female infant died from biventricular hypertrophic cardiomyopathy of unknown cause.
The images show a post-mortem pathology exam of her left ventricle.
LVOT Left ventricle outflow tract; *MV* Mitral valve; *IVS* Interventricular septum; *LVPW* Left ventricle posterior wall.

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P50

Inhomogeneous occurrence of congenital heart disease: clustering vs seasonality

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Introduction: Isolated congenital heart defects are a group of diseases with a complex etiology. Although some are associated with a genetic inheritance for most of them no clear inheritance pattern has been discovered. There are anecdotal reports of cluster presentation and some studies show seasonal trends with peaks in summer months for hypoplastic left heart syndrome. The understanding of the patterns of occurrence of congenital heart defects may give an insight on etiopathogenesis as cluster presentation and seasonality may suggest the possible involvement of environmental components.
Methods: We conducted a retrospective analysis of the inpatient database from the regional reference center for paediatric cardiology in the Emilia-Romagna region of Italy. We recorded diagnosis, date of birth and gestational age for each patient referred to our ward in the years 2000-2019. Exclusion criteria were polimalformative syndromes and genetic diseases. Seasonality was explored with the Walter and Elwood test while cluster presentation (defined as 2 or more occurrences of a disease in a period of less than 1 month or less than 2/3 of the mean time between births whichever the shorter) was analyzed with Grimson's method for temporal clustering.
Results: A seasonal occurrence was found in cor triatriatum (June and July, p<0,001), Ebstein anomaly (August and September, p<0,001), Total anomalous pulmonary connection (July p<0,001) and Transposition of the great arteries (February and July, p<0,05) but not for coronary abnormalities, Pulmonary atresia, tricuspid atresia, Coarctation of the aorta, Atrial septal defect, Ventricular septal defect, Hypoplastic left heart syndrome, aortic arch interruption, Tetralogy of Fallot, Truncus arteriosus and Double outlet right ventricle. Only Tetralogy of Fallot, Double outlet right ventricle and transposition of the great arteries show a significant <img71/>cluster presentation (details in table 1).
Conclusions: There is a significant seasonal pattern in the presentation of different congenital heart diseases with preponderance in summer months. TGA, TDF and DORV show a significant cluster presentation. Further studies are warranted to identify potential environmental factor(s) in the pathogenesis.

Disease	N. of clusters	N. of cases in cluster	Mean cluster duration (days)	Mean time between cases in cluster (days)	Mean Time between non cluster cases and clusters (days)	P
Transposition of great arteries	40/69 (58%)	3,1 ± 1,5	27 ± 18	8,6 ± 5,4	85,7 ± 52,3	<0,001
Tetralogy of Fallot	66/89 (74%)	3,1 ± 1,3	18,9 ± 10,3	6,3 ± 3,2	65,1 ± 36,8	<0,001
Double-Outlet right ventricle	11/35 (31%)	2,4 ± 0,9	20,8 ± 12,6	8,8 ± 12,6	165,4 ± 120,1	0,04

P51

Investigation of DEL22 frequency with FISH method in conotruncal heart anomaly patients

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Introduction: Conotruncal heart defects (CTHDs) represent 10-15% of congenital heart diseases and are often associated with genetic syndromes with deletion of the 22nd chromosome (Del22) in the case of DiGeorge velocardiofacial syndrome. Classical conotruncal heart defects include tetralogy of fallot (TOF), pulmonary atresia with ventricular septal defect (PA-VSD), truncus arteriosus (TA) and discontinuous aortic arch (IAA).
Methods: The aim of our study was to investigate the prevalence of 22q11.2 deletion by FISH method in children who were followed up due to CTHDs. In 104 cases with CTHDs, 22q11.2 region was examined for deletion by FISH analysis using DiGeorge-VCFS TUPLE1 probe, a specific probe for this region.
Results: In patients with CTHDs; in the FISH analysis in group 1 with isolated cardiopathy Del22 was detected in 3 cases, in group 2 with cardiopathy + dysmorphism in 2 cases, in group 3 with cardiopathy + immunodeficiency + dysmorphism in 2 cases and in group 4 with cardiopathy + immunodeficiency in 1 cases Del22 was detected with FISH analysis. Del22 was detected in 8 (7.5%) of 104 patients with congenital heart anomalies.
Conclusions: In conclusion, our findings are consistent with the literature. The clinical presentation of Del22 has been found to be highly variable. In addition, various organ systems may accompany this syndrome. For early intervention and disease management, it is important to diagnose the deletion as early as possible. Since the seriousness of cardiac anomalies shortens the life of the patients, we recommend performing Del22 screening with FISH for selective conotruncal anomalies in addition to chromosome analysis.

P52

Non-compaction cardiomyopathy, sick sinus disease and aortic dilatation: too much for a single diagnosis?

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Introduction: HCN4 mutations had been reported to be associated with a broad spectrum of phenotypes primarily sinus bradycardia (SB) and sick sinus syndrome (SSS). More recently, only few cases have also been described to have non-compaction cardiomyopathy (NCC) and aortic dilation (AD). We report 3 family members with this combined cardiac phenotype in the context of a pathogenic variant in *HCN4* gene.
Methods: Patients underwent systematic evaluation including medical history; physical examination; 12-lead-ECG; echocardiography; and ambulatory ECG monitoring. Exercise testing and cardiac magnetic resonance imaging (cMRI) were performed when clinically indicated. Genetic testing was performed after obtaining written informed consent.
Results: The proband presented with mild pulmonary stenosis on a dysplastic pulmonary valve at birth, and remained under regular cardiac follow-up. He was found to have SB, left ventricular (LV) hypertrabeculation and aortic root and ascending AD at the age of 16, and referred to tertiary Centre. He was found to be heterozygous for a pathogenic variant in *HCN4* gene [p.Gly482Arg (NM_005477.2:c.144G>A)]. cMRI confirmed LVNNC and proximal AD. Ambulatory ECG monitoring showed significant bradycardia, multiple asymptomatic and prolonged sinus pauses, ventricular couplets and triplets. Exercise testing revealed good chronotropic response, rare supraventricular ectopics, isolated multifocal ventricular ectopics and occasional bidirectional ventricular couplets. In view of these findings, young age and potential for bradycardic arrest and malignant arrhythmias, a transvenous dual-chamber implantable cardioverter defibrillator

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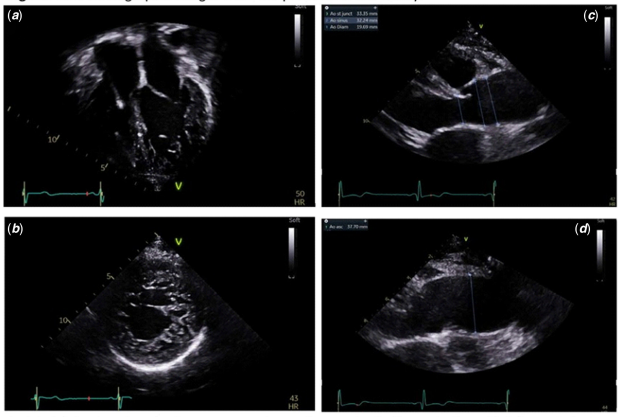
(ICD) was inserted in primary prevention and he was started on a beta-blocker. Cascade screening in the family identified that his sister and mum were heterozygous for the same pathogenic variant, both presenting with SB and LVNCC. Additionally, his sister also had mild AD.
Conclusions: Our results confirm the recent reported association between the combined clinical phenotype of SSS, NCC and AD, and the pathogenic variant p.Gly482Arg on *HCN4* gene. Our family presented with a fully penetrant phenotype with variable expressivity. Our findings highlight the importance of suspecting *HCN4* gene mutations when this diverse phenotype is encountered in clinical practice. More accurate risk stratification regarding arrhythmic burden, aortic disease progression and thresholds for elective surgery and progression of cardiac muscle disease will only be possible when larger cohorts are described.

Figure. Echocardiographic images from the proband of the family.



A. Apical 4-chamber view showing non compaction cardiomyopathy. **B.** Short axis view with left ventricular non compaction cardiomyopathy. **C, D.** Long-axis view showing aortic root dilatation (C) and ascending aortic dilatation (D).

P53

Oxygen uptake kinetics and local muscle oxygenation during submaximal exercise in children after the Fontan procedure compared to healthy peers

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Introduction: Oxygen consumption and muscle oxygenation during submaximal exercise in children with univentricular heart (UVH) have been poorly investigated. This study compares the oxygen uptake kinetics and the local tissue oxygenation after the Fontan procedure compared to healthy peers during a submaximal constant load test (CLT).
Methods: 18 UVH and 16 healthy control children performed a CLT and a maximal incremental ramp exercise test (CPET). 13 patients had dominant RV, 5 patients dominant LV. CLT was performed at 30% of the maximal exercise capacity during 6 minutes using cycle ergometry. Near infrared spectroscopy (NIRS) was used at m. Vastus Lateralis to measure local tissue oxygenation index (TOI) in both tests. The oxygen uptake kinetics (VO2) was measured during CLT and tau (63% of steady state level) was calculated in both groups. Correlations between VO2, TOI and other parameters were investigated.
Results: UVH patients had lower VO2peak (29±8 vs. 46±12 ml/min/kg, P <0.01), peakload (72±19 vs. 133±67 W, P<0.001) and maximal heart rate (HR) (168±13 vs. 193±12 bpm, P<0.001) compared with the controls. In CLT, HR was higher from 2 mins onwards in UVH patients. Higher tau-value and thus slower VO2-kinetics was found in the patient group (31.3±6.3 vs. 25.1 ±5.7, P<0.01). The TOI was lower at the onset of exercise (59.9±4.26 vs. 67.6±5.5s, P< 0.001) in UVH. The TOI decreased steeply in both groups immediately after start of the exercise, but even more steep in the patient group. TOI remained lower in UVH children. Slower VO2 kinetics in CLT was correlated with lower VO2peak, lower rest TOI and lower TOI correlated with lower VO2peak.
Conclusions: During submaximal exercise at 30% of VO2peak, slower VO2-kinetics, higher HR-response and different local TOI-patterns are determined. They correlated with a decreased maximal exercise performance in UVH children. This reflects a mismatch between O2-delivery and O2-demand of the working muscle from the onset of a low-level exercise. A good correlation was found between VO2peak and TOI and between VO2peak and VO2-kinetics of CLT, this means that the constant load test is useful whenever patients are not able to perform maximal CPET or for inbetween evaluation.

P54

Pathological insights into a case of propionyl acidemia with dilated cardiomyopathy

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Introduction: Propionic acidemia (PA) is a congenital metabolic disorder that typically develops in the neonatal period with a metabolic crisis, but some patients are presented as late-onset cardiomyopathy. We experienced a 14-year-old case of PA with dilated cardiomyopathy (DCM), and cardiac pathology was similar to mitochondrial cardiomyopathy. Thus, secondary mitochondrial cardiomyopathy is strongly suspected in pathophysiologic mechanisms.
Methods: A case report
Results: **Case presentation**: The case is a 14-year-old female who was born on a cesarean section as the second child of nonconsanguineous parents. Her neonatal period was uneventful. Mild mental retardation and epilepsy were noted in childhood. At the age of 14 years, she was diagnosed as DCM with end-diastolic left ventricular diameter of 65.0 mm and left ventricular ejection fraction of 35% by echocardiography. The acylcarnitine analysis of dried spot blood showed an increase in propionyl carnitine with 14.616 nmol/mL (normal range; <3.6), propionyl carnitine/acetylcarnitine ratio with 0.426 (normal range; <0.25). Genetic analysis identified a compound heterozygous mutation in the PCCB gene and she was diagnosed with PA.
Pathologic study: Endomyocardial biopsy found hypertrophy of cardiomyocytes and vacuoles positive with Periodic acid-Schiff (PAS) staining and negative with PAS-diastase staining in light microscopy. Electron microscopy showed an increase in the

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number of mitochondria within myofibrils, some larger mitochondria and atypical distribution of cristae that was similar to mitochondrial cardiomyopathy [Figure]. Immunohistopathologic analysis with respiratory chain enzyme antibodies showed mildly decreased signal of both Complex I and IV as a ration of Complex II (35% and 24% of normal, respectively).
Conclusions: In cardiac complication with PA, there would be three pathophysiologic mechanisms. 1) Secondary carnitine deficiency due to accumulation of propionyl-CoA, 2) Energetic deficiency due todeficiency of substrate in TCA cycle, and 3) Increased reactive oxygen species due to the inhibition of the oxidative phosphorylation in mitochondria by propionyl-CoA. In fact, multiple deficiencies of respiratory chain complexes were found in cardiac cells of patients with PA. Thus, our pathologic study would strongly support these hypotheses. In conclusion, our case suggests that cardiac complications from PA result in secondary mitochondrial cardiomyopathy.

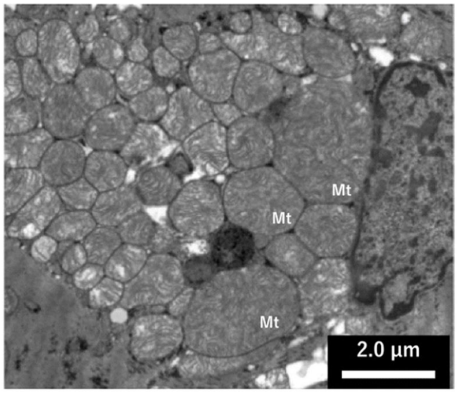

Figure. Electron micrograph of myocardium in propionic acidemia: Increased and some large mitochondria with abnormal cristae (Mt) was found.

P55
Pediatric genetic cardiomyopathies in the French South-East Region: genetic profile and genotype-phenotype correlations
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Introduction: Genetic cardiomyopathies affect 1-1.5 children per 100,000 each year. Five subtypes of cardiomyopathy exist, dilated cardiomyopathy (DCM) and hypertrophic cardiomyopathy (HCM) being the most common. This study aimed to establish the genetic profile of cardiomyopathies in children in the French South-East Region and describe the genotype-phenotype correlations.
Methods: Retrospective analysis of patients < 18 years diagnosed with cardiomyopathy between 2005 and 2019 was performed. Mitochondrial and other secondary causes were excluded. Completion of the gene panel according to current scientific data was realized for incomplete panels. Exome sequencing was offered for patients without identified mutation.
Results: Eighty-three children were included. The median age at diagnosis was 1.2 [0.3-10.5] years. Thirty-one patients (37%) had HCM, 22 patients (27%) had DCM, 10 patients (12%) had left ventricular non-compaction and 6 patients (7.2%) had restrictive cardiomyopathy. Median follow-up was 2 [0,3-7] years. Nine (11%) patients died during the study. Genetic investigations had been performed in 66(80%) patients, with a mutation found in 60% of them. *MYH7* (20% of cases, often associated with HCM), *MYBPC3* (10%) and *TTNT2* (7.5%) were the most frequent involved genes. Further analysis (panel of 64 genes) was performed in 14 patients, with negative or incomplete panel or in those not yet tested, and results are still pending. Exome sequencing analysis was performed in 3 patients with a negative complete panel as well as in their non-affected parents. Variants were found in genes *TCF7L1, MYOM2* and *SMARCD1*. Genotype-phenotype correlation did not show significant differences in clinical evolution nor in survival curves between patients with an identified mutation and those without. In CMH, patients with an identified mutation had increased myocardial septal thickness mesured on the trans-thoracic echocardiography, compared to those without (p=0.05).
Conclusions: In our pediatric population, a pathogenic mutation was found so far in 60% of the tested cardiomyopathies. Exome sequencing identified some interesting variants. Further research needs to be performed to confirm the pathogenicity of these mutations

P56
Phenotypic, clinical and mutation profile of paediatric patients with RASopathy-associated cardiac disease
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Introduction: RASopathies are caused by genetic mutations affecting the RAS-MAPK cell-signalling pathway. The most frequent syndromes are Noonan (NS), Leopard (LS), Costello (CS), Cardiofaciocutaneous (CFC) and Neurofibromatosis type-1. They can be transmitted to the offspring, thus Genetic Counselling play an important role. The aim of this study is to describe the cohort of paediatric patients diagnosed with any Rasopathy and cardiopathy in a Paediatric Cardiology Service, national reference in Inherited Cardiovascular Diseases (ICVD). Investigation also hunts to analyse genetic mutations, genotype-phenotype relationship, comorbidities and cardiac natural history.
Methods: An observational cross-sectional study is presented, including patients who were followed-up by the ICVD Unit in the first half of 2019, meeting the requirements of the hospital´s Ethics Committee. Data were collected from medical records. The statistical analysis was performed using Stata/IC version 15.1.
Results: 34 patients (0-23 year-old) were included, 56% females. 74% had NS, 15% LS, 3% CS and 8% other syndromes. Genetic tests were performed with a mean age of 5.6 year-old. The most frequent mutations were detected in PTPN11 (67%), RAF1 (9%), SOS1 (7%) and RIT1 (7%) genes, resulting the most recurrent in p.Asn308Asp (PTPN11). 77% of mutations were de novo and 20% inherited. 68% of patients presented valvulopathies, 55% pulmonary stenosis (PS), 50% hypertrophic cardiomyopathy

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(HCM), 41% atrial septal defects, 21% arrhythmias and 15% aortic disease. More than a third of patients required any cardiac intervention (similar number of surgeries and catheterizations), with a mean age of 9.5 year-old. All patients with mutation in PTPN11 were diagnosed with NS, and their most prevalent heart disease was PS (60%), followed by HCM (40%) and coronary disease (in cases with mutation in p.Gln510Arg). On the other hand, patients with RAF1 mutations presented with HCM more frequently. Most repeatedly associated pathologies were dysmorphic (91%), endocrinological (56%) and neurobehavioral (56%) disorders.
Conclusions: Most frequent cardiopathies in this paediatric cohort with Rasopathies are PS and HMC. There is a correlation between heart disease and specific genetic mutations, and the same occurs with the comorbidities that may affect overall prognosis.

Genetic tests and multidisciplinary management are essential for the better diagnosis and management of these patients.

P57
Prospective randomized study: metoprolol protects cavity size and diastolic function over six-year follow-up in hypertrophic cardiomyopathy

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Introduction: Cohort studies on pediatric HCM-patients have reported that beta-blocker therapy has a dose-related protective effect against cardiac mortality, suggesting that prophylactic beta-blocker therapy might benefit symptom-free patients. We have recently reported from a randomized study that exercise performance deteriorates significantly over time in un-treated patients, but not in metoprolol-treated patients.
Methods: Previously un-diagnosed cases of HCM were identified by family screening. Patients without symptoms and risk-factors were randomized to either life-style advice and observation (Obs-group; n=15), or to life-style advice plus introduction of metoprolol 3.8 [3.5-4.3] mg/kg therapy (Bbl-group; n=12). Median age was 18 [IQR 14-26] yrs, with a median follow-up of 6.0 [2.0-8.0] yrs. Detailed ultrasound examinations were performed at rest before start of study and yearly thereafter; diastolic function was also examined at 5 and 10 min after a maximal exercise-test on bicycle ergometer. Echocardiographic examinations were coded and re-measured blindly.
Results: At last follow-up septum-to-cavity ratio (sepcavr) had increased in Obs-group to 0.30 [0.28-0.31], p=008, whereas it had decreased to 0.23 [0.23-0.24], p=0.0001, in Bbl-group; p=0.00005 for inter-group comparison. Last left ventricular-wall-to-cavity ratios were 0.25 [0.22-0.28] and 0.20 [0.18-0.22] respectively, p=0.0013. Already after two years LVEDD Z-score fell by -0.24 in Obs-group and improved by +0.74 [0.36-1.05] in Bbl-group, p=0.00008. At last follow-up all patients were fully grown with LVEDD 4.5 [4.4-4.6] cm in Obs-group and 4.8 [4.7-5.1] cm in Bbl-group (p=0.0028), and 3D LV-volumes of 41.0 [38.9-47.1] versus 51.0 [44.3-54.1] ml/m^2 BSA (p=0.006). The difference was not secondary to bradycardia as calculated cardiac output at rest was also significantly different, 1.51 [1.36-1.69] and 1.90 [1.67-2.05] L/min respectively (p=0.008). Diastolic septal e-velocities reduced slowly overtime both at rest (-1.46 cm/s), and 5 min after exercise (-1.1cm/s), in Obs-group, whereas they did not in Bbl-group (+0.6cm/s) and (+0.4cm/s); p=0.0037 and p=0.05 respectively compared to Obs-group.
Conclusions: Metoprolol-therapy with adequate beta-blockade is associated with a beneficial cardiac re-modelling with increase in cavity-size and reduced wall-to-cavity ratios compared to controls. A reduced LV-cavity size is a recognized risk-factor for heart-failure death and we find that left ventricular cavity size and diastolic function slowly deteriorates in untreated patients.

P58
Pulmonary embolism in childhood

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Introduction: In recent years, a progressive increase in pulmonary thromboembolism (PTE) incidence is observed. This is related with both more frequent recognition and increased survival of patients with underlying predisposing conditions (neoplasia, orthopedic interventions, central venous lines, hormonal and contraceptive medication, and obesity).
Methods: Retrospective analysis of etiologic and predisposing factors, clinical presentation, diagnostic workup and management in childhood PTE.
Results: For a period of 10 years, 12 children with proven community-acquired PTE were treated in our tertiary care center (age from 2 months to 17.6 years, weight from 3 kg to 118 kg). The most common predisposing factors were prolonged immobilization, vasculitis, childbirth or use of contraceptives, congenital or acquired thrombophilia. In 6 patients, congenital thrombophilia was demonstrated, i.e. homo- or heterozygotes for factor V Leiden mutation, factor II G20210A, MTHFR, PAI. Three of them had combination of two or more mutations. Two children had both antithrombin III and protein C deficiency. Peripheral venous thromboembolism was demonstrated in 5 cases. The rest probably had in situ PTE. The most common clinical manifestations were chest pain and dyspnea. In all patients, the diagnosis was confirmed by echocardiography and computed tomography. In 40% of the patients, direct or indirect evidence of pulmonary hypertension were found at presentation. The D-dimers were elevated in only 60% of cases of massive PTE. In 4 children, local fibrinolysis with recombinant tissue plasminogen activator was performed, preceded by transcatheter thrombus fragmentation and thrombus aspiration in 3 of them. Conservative treatment included standard therapy, i.e. unfractionated or low molecular weight heparin, followed by acenocoumarol. The homozygous carriers of mutations associated with thrombophilia remained on lifelong prophylaxis with indirect anticoagulant. In 11 cases, thrombotic formations resorbed without recurrences. One infant died at 2 months of age with right atrial thrombus, PTE and sepsis.
Conclusions: PTE in childhood has a specific etiology, clinical manifestations and favorable evolution, if treated in a timely manner. Currently, recommendations for behavior in the pediatric population are extrapolated from those for adult patients, but in recent years specific diagnostic and therapeutic strategies, that are applicable in children, are under development.

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P59

Rapidly growing cardiac tumour in an infant

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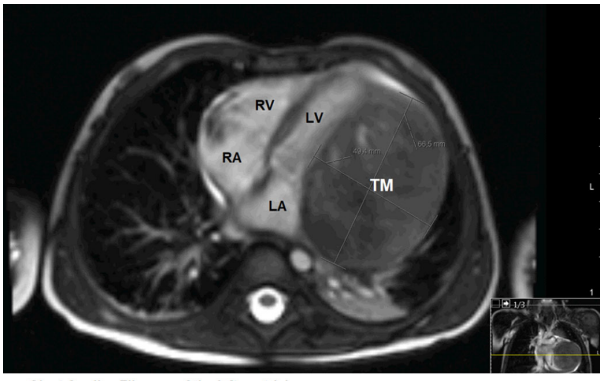
Introduction: Cardiac tumours in children are exceptional findings and are usually benign. The most commonly found primary tumours in children are rhabdomyomas mainly diagnosed in the context of tuberous sclerosis. As a rule they tend to regress spontaneously and only require surgery if symptomatic. Cardiac fibromas, unlike rhabdomyomas, are more aggressive, tend to present with arrhythmias and rarely regress.
Methods: We report the case of a 3 month old female infant referred to our Department with a heart murmur.
Results: She was thriving and asymptomatic from a cardiovascular point of view. The pre and post-natal history was uneventful and there were no concerns regarding family history. The transthoracic echocardiogram (TTecho) revealed a large oval, vascularized mass adherent to the free wall of the LV, abutting on the posterior leaflet of the mitral valve causing turbulent inflow, but no obstruction to the LV outflow. A cardiac MRI showed a 50x40x40mm mass, whose radiological features were suggestive of a fibroma without a clear myocardial cleavage plan. Follow up showed a progressively increasing mass, occupying most of the left ventricular cavity. The most recent cardiac MRI at 14 months of age showed the tumour mass dimensions to be 67x49x54mm. Despite this finding, the infant remains asymptomatic and thriving. A 24 hour Holter did not identify any rhythm problems.
Conclusions: Considering the progressive nature of this lesion, the diagnosis of a fibroma is most likely. Based on our patient´s clinical stability, we opted for a wait-and-see approach. However, close follow-up is mandatory to search for possible complications, namely, inflow/outflow obstruction, cardiac failure, coronary artery obstruction and, particularly in this type of tumour, arrhythmias. Due to the infiltrative nature of the tumour, complete surgical resectability is unlikely.



Giant Cardiac Fibroma of the left ventricle.
TM **Tumour Mass;** ***LA*** **Left atrium;** ***RA*** **Right atrium;** ***LV*** **Left ventricle;** ***RV*** **Right ventricle**

P60

Recovery kinetics of gas exchange parameters and heart rate after maximal exercise in children with repaired Tetralogy of Fallot compared to controls

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Introduction: Few studies demonstrate delayed recovery after exercise in children and adults with congenital heart disease. In this study we evaluate the recovery kinetics of children with repaired Tetralogy of Fallot (rToF) in comparison to healthy peers and investigate correlations with ventricular function and QRS duration.
Methods: Forty-five children after rToF and forty-five matched controls performed a maximal incremental cardiopulmonary exercise test. In the subsequent 6 minute recovery period, patterns of VO_2, VCO_2 and HR were analysed. Half-life time ($T_{1/2}$) of the exponential decay and percentage drop per minute (Recmin) were compared between groups. In the rToF group, correlations were examined between the recovery parameters and QRS-duration and ventricular function, described by fractional shortening (FS) and tricuspid annular plane systolic excursion (TAPSE).
Results: Exercise performance was reduced in the rToF group (VO_2peak: 34.46 ± 8.14 vs. 42.77 ± 8.14 ml/min/kg; p<0,001 and maximal load: 112.2 ± 42.4 vs. 149.9 ± 65.7 W; p<0,001). Maximal HR (174 ± 13.8 vs 192 ± 9.4 bpm; p<0,001) was lower in the rToF patients, due to chronotropic incompetence. Recovery of VO_2 and VCO_2 was delayed in rToF patients, half-life time values were higher compared to controls ($T_{1/2}VO_2$ 52.5 ±11.3 s vs. 44.3 ± 10.5 s; p=0.001 and $T_{1/2}VCO_2$ 68.3 ± 13.8 s vs. 59.4 ± 12.1 s; p=0.002) and percentage drop from maximal value was slower at each minute of recovery (p<0.05). Correlations were found with FS ($T_{1/2}VO_2$: r=-0.52; p<0.001; Rec1min-VO_2: r=-0.64, p<0.001; Rec1min-VCO_2: r=-0.37, p=0.012) and zTAPSE ($T_{1/2}VO_2$: r=-0.51; p<0.001; Rec1min-VO_2: r=-0.57, p<0.001; $T_{1/2}VCO_2$: r=-0.47; p=0.001; Rec1min-VCO_2: r=-0.51, p<0.001). QRS-duration correlated with maximal exercise parameters (Predicted%VO_2 r=-0.42; p=0.005 and Predicted%Load r=-0.39; p=0.010), but not with recovery. No difference was found in HR recovery between patients and controls.
Conclusions: Besides a lower exercise tolerance, rToF patients have a prolonged recovery of VO_2 and VCO_2. Whereas QRS-duration is correlated with maximal exercise capacity, ventricular function is related to recovery after maximal exercise. No difference can be observed in HR recovery. This suggests that an inadequate stroke volume adaptation, influenced by impaired ventricular function, plays a determining role in the recovery phase.

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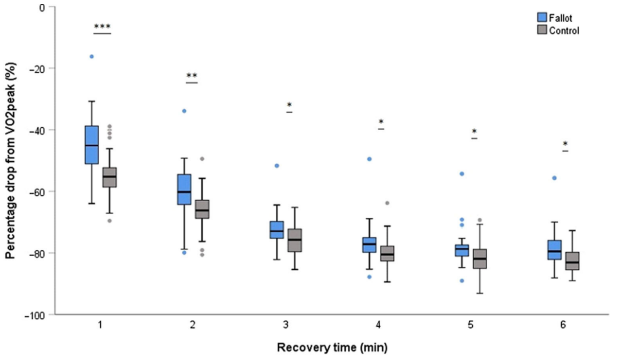

Recovery of VO_2 after maximal exercise, expressed as percentage drop from VO_2peak, at minute one to six in recovery, in children with repaired Tetralogy of Fallot (blue boxes) and healthy controls (grey boxes). *, ** and *** indicate statistically significant difference respectively at p<0,05, p<0,01 and p<0,001

P61

Successful treatment of life threatening lymphatic disorder by MEK-inhibition in a girl with Noonan syndrome and PTPN11 mutation

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Introduction: RASopathies due to germline mutations in genes of the RAS/MAPK signaling pathway may be associated with severe lymphangiopathies. In patients with gain of function mutations in genes of this pathway a therapeutical option may be MEK-inhibition. We report our experience with the MEK-inhibitor trametinib in a patient with *PTPN11* mutation and life threatening lymphatic disorder.

Methods: Our patient presented in the neonatal period with fetal hydrops and recurrent chylothoraces. Cardiac evaluation showed pulmonary and aortic valve stenosis. Genetic testing confirmed Noonan syndrome (heterozygous *PTPN11* mutation ENST00000351677:c.854T>C; p.F285S). Patch-enlargement of the RVOT was performed in infancy, reconstruction of the aortic valve at the age of 3 years. Following aortic and pulmonary valve replacement at the age of 5.4 yrs. bilateral chylothoraces developed refractory to conservative therapy. Despite left pleurodesis the respiratory situation deteriorated requiring tracheotomy and intermittent ventilation. Progressive lymphedema of the left chest wall resulted in dyspnea requiring continuous ventilation, oxygen supplementation and maximal diuretic therapy. At this stage palliative treatment was discussed with the parents. As a last option we proposed an attempt of treatment by MEK-inhibition with trametinib, which was approved by the parents and consented by our ethical committee.

Results: Trametinib was started at the age of 7.18 yrs. at a dose of 0.01 mg/kg/day. Following initiation of treatment our patient required a significant increase of diuretics due to fluid retention and increasing edema. Sepsis with positive blood cultures for streptococci required repeat hospitalization 6 weeks later. During this admission trametinib was increased to 0.02mg/kg. About this time continuous clinical improvement occurred resulting in cessation of oxygen supplementation, progressive weaning from mechanical ventilation and significant reduction of diuretics. 3-4 months following initiation of treatment the chest wall edema had decreased significantly and there was reappearance of ventilation to the left lung. Following 9 months of treatment NYHA classification status has improved from IV to II and the girl remains without side effects.

Conclusions: Our experience confirms two previous reports that MEK-inhibition with trametinib can be considered as a treatment option in patients with RASopathies and life threatening lymphatic disorders resistant to conventional treatment.

P62

The Roles of mir-101, mir-1183, and mir-1299 in Acute Rheumatic Fever and Rheumatic Heart Disease

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Introduction: Acute rheumatic fever (ARF), which commonly causes carditis in children, sometimes evolves into rheumatic heart disease (RHD). It is critical to have biomarkers for diagnostic and prognostic purposes. We aimed to investigate the expressions of miR-101, miR-1183, and miR-1299 in children with ARF and RHD and determine their potential as diagnostic biomarkers.

Methods: This cross-sectional study was conducted on 69 pediatric ARF patients and 27 gender and age-matched controls. The clinical findings were noted from medical records, echocardiography was repeated, and plasma miR-101, miR-1183, and miR-1299 expressions were studied using real-time PCR.

Results: MiR-101 and miR-1183 expressions were lower in children with ARF than the controls ($p<0.05$). The miR-1183 expression was lower in patients had a family history than the controls ($p<0.05$). Whereas fifteen patients had only arthritis and 54 (17 of them mild, 21 of them moderate and16 of them serious) patients had associated carditis during the ARF acute attack. No differences were detected between the ARF patients had isolated arthritis and the controls for the miRNAs ($p>0.05$). The expression of miR-1183 was lower in the ARF patients had carditis than the controls ($p<0.05$). During acute attack ARF, miR-101 and miR-1183 were significantly lower in the patients had moderate-serious carditis than the controls ($p<0.05$). RHD was observed in the 36 children, during the last control (after the mean 3.7± 2.4 years follow-up period). MiR-101, miR-1183, and miR-1299 expressions were detected as lower in children with RHD than the controls. However the differences were found statistically insignificant ($p>0.05$). The performances of miR-101 and miR-1183 were evaluated by ROC analysis, as possible biomarkers to be used on the diagnosis of ARF and RHD. The AUC (area under the curve) values for these miRNAs on the ARF diagnosis were found statistically significant ($p<0.05$).

Conclusions: Significantly lower plasma expressions of miR-101 and miR-1183 in ARF patients, especially in cases with moderate-severe carditis, might indicate the potential use of these miRNAs as diagnostic biomarkers. However, further studies are needed for lightening whether these miRNAs might be helpful using as biomarkers.

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3. End-stage heart and lung disease

P63
Cell-free DNA after pediatric heart transplantation in a national Swedish study: Rejections, PTLD, infections & more

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Introduction: Heart transplantation (HTx) comes with a higher risk for rejection compared with other solid organs. Graft surveillance largely depends on scheduled endomyocardial biopsies (EMB) as the gold standard due to the lack of reliable biomarkers. Donor-derived cell-free DNA (dd-cfDNA) has gained attention as a potential marker to assess organ function after HTx.
Methods: In a collaboration between the two pediatric heart centers performing heart transplantations in Sweden, a prospective cohort study is conducted since 2016. Patients are followed with parallel measurements of dd-cfDNA and EMB during one year after HTx. 35 single-nucleotide polymorphisms (SNP) were chosen to distinguish between recipient and donor. After a targeted preamplification-step, digital PCR is conducted.
Results: A total of 29 consecutive patients were included. 5 patients died on the waiting list. 19 patients could be followed for one year after HTx, generating 160 EMB and 181 blood samples. dd-cfDNA levels were high on the first sample after HTx reflecting reperfusion injury, declining to very low levels (below 0,02%) in stable patients. Dd-cfDNA showed good correlation to clinical events of the patients such as rejection, infection and post-transplant lympho-proliferative disease (PTLD).
Conclusions: We constructed a technically robust method to measure cell-free DNA after HTx. Results are promising with respect to establishing a threshold for the exclusion of rejection requiring treatment (high negative predictive value). High levels of dd-cfDNA can, however, have multiple causes and cannot distinguish between various clinical events including rejection and infection.

P64
Determinants of quality of life in children with pulmonary arterial hypertension

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Introduction: Pulmonary arterial hypertension (PAH) is a progressive disease with elevation of pulmonary vascular resistance, right ventricular failure and premature death. Introduction of early combination therapy improved PAH patient survival. As next step clinical trials are focusing on pediatric PAH patients functional status assessment. Our aim was to evaluate quality of life (physical, emotional, social, and school) in children with PAH using a validated Pediatric Quality of Life (PedsQL) survey.
Methods: Parents of 27 PAH children and patients (age: 9.2+/- 4.9 years) with appropriate developmental capacity completed the PedsQL questionnaire. PedsQL 4.0TM is a validated quality of life (QOL) survey that assesses four domains: physical (1), emotional (2), social (3) and school functioning (4). Demographic information, data on WHO PAH clinical and functional classification, follow-up echocardiographic parameters and treatment information were collected.
Results: PedQLs survey was completed by 27 parents and 15 patients: eight patients were aged 2–4 years; five were aged 5–7 years; eight were aged 8–12 years, and six were aged 13–18 years. Seventeen patients were classified as WHO Group I pulmonary arterial hypertension, three WHO Group III PH due to lung disease, and seven WHO Group V. Fourteen patients had WHO functional class (FC) 1, thirteen had FC 2 or more. PAH patients had significantly lower QOL scores than healthy children in all domains (1-4) on both parent and self-reported. We have found that patients with FC 1 had significantly higher (parents and patients reported) physical activity (95 vs. 50 and 82 vs. 55 QOL score, $p<0.05$). In our study population, clinical parameters like left ventricular systolic diameter, systolic eccentricity index, number of PAH specific drugs had no impact on QOL scores. Patients with decreased TAPSE (<-2 Z score) had significantly lower QOL scores in every patient reported domain (93 vs. 64; 80 vs. 60; 90 vs. 67; 95 vs. 50 QOL score, $p<0.05$) and in the parent reported cognitive domain (75 vs. 30 QOL score, $p<0.05$).
Conclusions: Assessment of the QOL in pediatric PAH patients revealed functional impairment in this patient group. Based on self-reported information WHO FC is a reliable marker of QOL impairment in our pediatric PAH patients.

P65
Forns Index for diagnostics of hepatopathy in children with Fontan circulation

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Introduction: Earlier studies have shown that adults with Fontan circulation suffer from hepatic fibrosis, but there are few studies conducted regarding hepatic complications in children with Fontan circulation. Yet, early detection of hepatic fibrosis is the key to prevent further development of liver damage. Diagnostic tools to detect fibrosis early are lacking; biopsies are highly intrusive, radiology often miss early stages, and no individual laboratory parameters can accurately detect fibrosis. Forns Index (FI) has been developed to detect fibrosis in adult patients with hepatitis C, but recent studies on adult patients with Fontan circulation indicate that FI can be used in this population as well. This study aimed to investigate if FI could be used as a non-invasive indicator of hepatopathy in children and adolescents after Total Cavo Pulmonary Connection (TCPC).
Methods: Fasting blood sample were collected in a group of 35 patients with TCPC, 5-18 years of age. Analysis of platelet count, gamma-glutamyl transferase (GGT), and cholesterol was performed and FI calculated. Patients' results were compared to reference laboratory parameters. Mean FI for patients and the reference values were compared using t-test. Analysis was performed; both on group level and in subgroups stratifies by age; 5-7, 8-12, and 13-18 years old.

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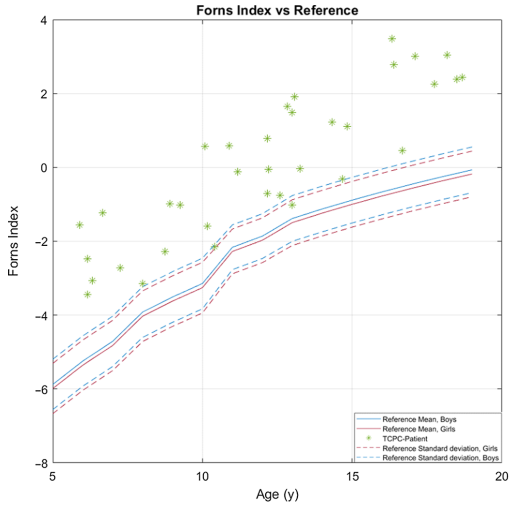
Results: The FI for patients was elevated compared to the references both at group level and in subgroup analysis (Fig 1). Specific analysis of the 6 patients with the highest FI scores showed that they were between 16-18 years old, five were male, and 4 had a right systemic ventricle.
Conclusions: Forns Index shows the potential to be used as a non-invasive screening method for hepatopathy in children and adolescents after TCPC. Larger studies are needed to correlate the index values with incipient fibrosis verified by other methods, also to determine cut-off levels for hepatopathy in patients < 18 years of age.



P67
The effect of ozone in an experimental autoimmune myocarditis model

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Introduction: Myocarditis is an inflammatory disease of the myocardium characterized by myocyte damage and necrosis, which can lead to mortality and morbidity in childhood. Ozone is a medical agent used in the treatment of various diseases due to its anti-inflammatory and immunomodulator effects. The aim of this study was to investigate the possible effects of ozone on myocarditis.
Methods: An experimental autoimmune myocarditis (EAM) model was created in 6-8 week-old male Balb/c mice. A total of 49 mice were divided into seven groups. On the 1st and 7th days, an equal amount of pig cardiac myosin and complete Freund's adjuvant mixture was administered subcutaneously to the mice in which myocarditis would be developed. In order to investigate the effectiveness of ozone, 20, 30, 40, 50, and 60 μg/mL concentrations of ozone were administered intraperitoneally every day from the 8th to 22nd day. The mice were sacrificed on the 22nd day. First and last body weights, and heart weight of all mice were measured. Serum troponin T, TNF-α, IL-1β, IL-6, IL-10, and TGF- β1 levels were measured in intracardiac blood samples. The heart tissues removed were subjected to histologic examination.
Results: There was no difference between the groups in terms of first and last body weights ($p>0.05$). It was observed that mice in all groups, gained weight during the follow-up period ($p=0.003$). Weight gain and heart weight were lower in the EAM group compared with the other groups ($p<0.004$). The ratio of heart weight/last body weight was increased in the EAM group ($p<0.001$). Troponin T, TNF-α, IL-1β, and IL-6 levels were higher in the EAM group than the control group ($p<0.005$). Troponin T levels were significantly decreased in the groups that received 50 and 60 μg/mL concentrations of ozone ($p=0.001$). TNF-α and IL-1β levels were lower in the ozone-treated groups ($p<0.001$). Histologically, discoloration, inflammation, and fibrosis levels were significantly higher in the EAM group than the other groups ($p<0.001$).
Conclusions: Our results show that the anti-inflammatory and immunomodulating effect of ozone can be promising in the treatment of myocarditis.

P68
Vasodilator therapy for pulmonary hypertension in children: a Swedish national study of patient characteristics and current treatment strategies

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Introduction: Pulmonary vasodilator therapy is uncommon and still often an off-label treatment for pulmonary hypertension in children. The aim of this study was to assess patient characteristics and current strategies for vasodilator prescription to young children in Sweden.
Methods: This nationwide registry-based study includes all children, below seven years of age at time of inclusion, who were prescribed pulmonary vasodilator therapy during 2007-2017. Information on prescriptions was retrieved from the Prescribed Drug Register. All cases were categorized according to the WHO classification for diagnosis of pulmonary hypertension. Characteristics for known and unknown factors on all children with a prescribed vasodilator were retrieved by linkage to the Swedish Medical Birth Register, the National Patient register and the Death Register. Descriptive statistics were used to describe type of drug treatment, combinations of therapy, type of cardiac disease and comorbidities.
Results: In total, 233 children were prescribed a pulmonary vasodilator as single or combination therapy. Sildenafil was most commonly prescribed (N=224 children) followed by bosentan (N=29), iloprost (N=14), macitentan (N=4), treprostinil (N=2) and riociguat (N=2). Over the study period, the prescription rate for sildenafil tripled. Monotherapy was most common, 87% (N=203), while 13% (N=20) had combination therapy. Bronchopulmonary dysplasia (BPD; N=82, 35%) and congenital

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heart disease (CHD; N=60, 26%) were the most common associated conditions. Overall, 8% (N=18) had Down syndrome. Concomitant treatments, with for example diuretics, were common. Cardiac catheterization had been performed in 39% (N=91). Pulmonary vasodilator treatment was initiated before 1 year of age in 61% (N=143). Overall mortality was 13% (N=30) during the study period.
Conclusions: Pediatric prescriptions of sildenafil increased in Sweden during the study period. Most patients had monotherapy, but dual and triple vasodilator therapy were also used to some extent. Children with pulmonary hypertension associated with CHD or BPD were the largest groups to receive therapy. Despite treatment, mortality was high and additional pediatric studies are needed for a better understanding of underlying pathologies and treatment effects.

4. Fetal cardiology

P69
Analysis of the diagnostic value of prenatal echocardiography for the detection of vascular rings

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Introduction: In this study we compared diagnostic value of the complex and individual application of all special echocardiography views and signs for vascular rings (VR).
Methods: The study comprised of 128 fetuses receiving prenatal echocardiography from 2004 to 2017. The study group consisted of 68 fetuses with prenatal suspicion of VR. The control group was included 60 fetuses with various congenital heart defects combined with the right aortic arch. All special ultrasound views were used for the diagnosis of vascular rings – the 3-vessel and trachea view, the aortic arch long-axis view, the transverse view of the thorax and upper abdomen, demonstrating the position of aorta.
Results: Primary prenatal echocardiography with complex of all special views for VR diagnosis has high diagnostic value: clinical utility index – 93,3, sensitivity – 95,7 %, specifity – 84,6 %, positive predictive value – 95,7 %, negative predictive value – 84,6 %, positive likelihood ratio – 6,22, negative likelihood ratio – 0,05. Isolated use the transverse view of the upper abdomen, demonstrating the abnormal abdominal aorta (median or right relative to the spine) has an average level of clinical utility index (83,3): sensitivity – 93,6 %, specifity – 46,2 %, positive predictive value – 86,3 %, negative predictive value – 66,7 %, positive likelihood ratio – 1,74, negative likelihood ratio – 0,14. U-shaped angle of ascent of the aortic arch and arterial ductus on the three vessels and trachea view with isolated use has a low level of clinical utility index (76,7): sensitivity – 85,1 %, specifity – 46,2 %, positive predictive value – 85,1 %, negative predictive value – 46,2 %, positive likelihood ratio – 1,58, negative likelihood ratio – 0,32. Isolated assessment of tracheal location has a good level of clinical utility index (90,0): sensitivsty – 89,4 %, specifity – 92,3 %, positive predictive value – 97,7 %, negative predictive value – 70,6 %, positive likelihood ratio – 11,62, negative likelihood ratio – 0,12.
Conclusions: The complex of all special echocardiography views and sings is valuable for diagnostics of vascular rings. If it is not possible to obtain all the necessary views, the most useful is the assessment of the position of the trachea.

P70
Antenatal echocardiographic predictors of urgent baloon atrial septectomy (bas) in neonates with d-tga: can we rely on them?

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Introduction: Profound hypoxia caused by inadequate intracardiac mixing due to a restrictive foramen ovale (FO) is a potentially life-thereatening complication in neonates with transposition of the great arteries (dTGA). An urgent balloon atrial septectomy (BAS) is a procedure of choice in severe cases, but dependent on the availability of a 24-hour interventional cardiology facility. The prenatal identification of predictors for an urgent BAS at birth would help in programming the delivery of these babies with a ready cath lab service in order to minimize the risk of hypoxic damage.
Methods: We performed a retrospective study of all patients with a prenatal diagnosis of dTGA cared for in our Center from 2013 to 2019. The following fetal echocardiographic parameters obtained less than two weeks before delivery were recorded: size and appearance of FO (restrictive or aneurismatic), septum primum excursion (max flap angle), diameters of the atria, size and shunting of the ductus arteriosus. BAS was defined as urgent if performed within 12 hours from birth in neonates with restrictive foramen ovale and severe hypoxemia not improved by prostaglandin E1 infusion.
Results: 76 subjects (54 males) were included during the study period. A total of 55 patients (72%) underwent BAS before surgical correction, while 21 (28%) necessitated an urgent BAS for severe hypoxemia at birth. Restrictive and aneurysmatic appearance of FO did not show sufficient sensitivity for screening of neonatal emergencies (Se16%/Sp80% and Se23%/Sp94%, respectively). By combining of both characteristics the accuracy raised to Se20% and Sp72%, while the presence of at least one of the two increased sensitivity to 80% while lowering specificity to 30%. Moreover, none of the other echocardiographic parameters studied showed a significant correlation with the need for urgent BAS (table 1).
Conclusions: Antenatal evaluation of the foramen ovale in fetuses with dTGA is still challenging and, based on our experience, poorly predicts the need of urgent BAS. A combination of echocardiographic indices improves the sensitivity, but still not enough to be considered a successful screening tool. Additional studies are needed to create a more complex index taking into account other parameters to further improve Sp and Se

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Table 1.		
Logistic regression analysis	Odds Ratio	p-value
Foramen ovale diameter	0,87	0,57 (NS)
Maximum angle of septum primum	0,98	0,36 (NS)
Right atrium diameter	1,2	0,23 (NS)
Left atrium diameter	0,98	0,92 (NS)
Ductus arteriosus diameter	0,72	0,30 (NS)
	Sensitivity	Specificity
Restrictive FO	16% (CI 08%-27%)	80% (CI 69%-89%)
Aneurismatic FO	50% (CI 31%-69%)	38% (CI 21%-58%)
Restrictive AND Aneurismatic FO	20% (CI 11%-34%)	27% (CI 16%-41%)
Restrictive FO OR Aneurismatic FO	80% (CI 58%-93%)	33% (CI 16%-56%)

P71

Case presentation: Heart block of unknown cause in fetus requiring a pacemaker insertion – the implication for counselling.

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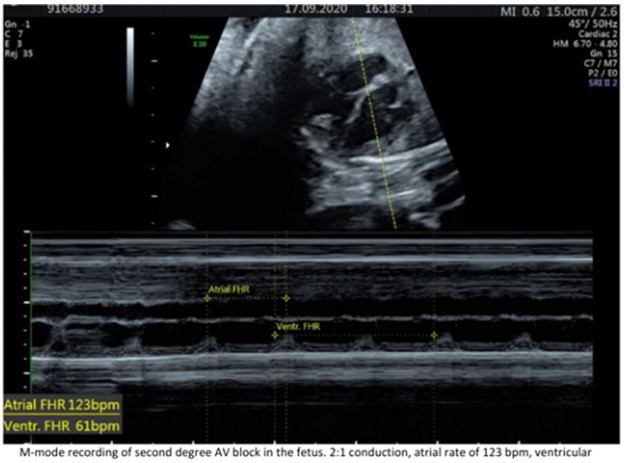
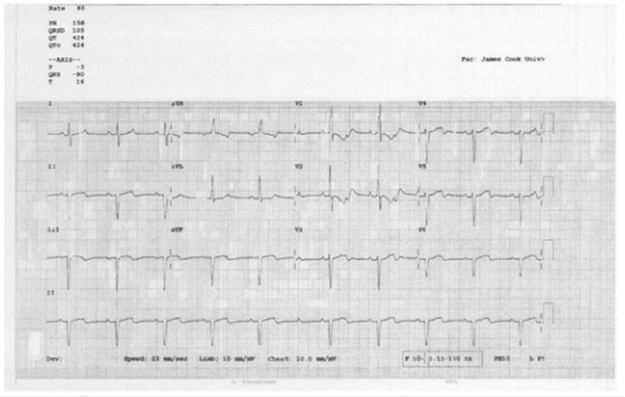
The Newcastle upon Tyne Hospitals NHS Foundation Trust (1)

Introduction: Congenital heart block is usually related to maternal anti-Ro/SSA and anti-La/SSB antibodies or abnormal conduction in congenital cardiac anomaly, such as the left isomerism, congenitally corrected transposition of the great arteries or double inlet left ventricle. Counselling should aim to give the prognosis and the neonatal outcome. This is challenging, particularly in rare cases with unknown etiology.
Methods: We present a fetus with normal heart and second-degree heart block, in which the mother had negative anti-Ro/SSA and anti-La/SSB antibodies. The baby required a pacemaker insertion at two days of life. Thirty-two years old G2 P2 woman at 28 w.g. was referred for a fetal echocardiogram with concerns regarding fetal arrhythmia. The ultrasound revealed a structurally normal heart, at an atrial rate of 136 bpm, and a ventricular rate of 48 bpm. The cardiac function was normal, and there was no evidence of hydrops. Differential diagnosis included complete or second-degree AV block with 3:1 conduction. Maternal blood sample for anti-Ro/SSA and anti-La/SSB antibodies was obtained. The result was negative. During counselling, parents were informed that the cause of heart block is unknown, and the baby would likely require a pacemaker insertion immediately after birth. Besides, long QT syndrome had to be excluded. Subsequent scans showed a second-degree AV block with 2:1 conduction, ventricular rate of 62bpm. We informed parents that the heart rate is acceptable, and that timing of a pacemaker insertion remained unpredictable. At 39 w.g., she underwent an elective caesarean section. A female baby was born in good condition weighing 3220 grams. An ECG confirmed diagnosis of second-degree AV block 2:1, ventricular rate of 62 bpm. On day two of life, the baby deteriorated, and an ECG revealed second degree AV block with 3:1conduction, ventricular rate of 39 bpm.
Results: We proceeded with an epicardial ventricular permanent pacemaker insertion. The baby remains clinically stable. On the pacemaker check, the underlying rhythm remains at 50 bpm—no evidence of long QT syndrome in subsequent ECG's.
Conclusions: This is an atypical case of congenital heart block, which proved challenging. This will impact counselling as it was difficult to predict the need for pacemaker insertion.



M-mode recording of second degree AV block in the fetus. 2:1 conduction, atrial rate of 123 bpm, ventricular rate of 61 bpm.


Postnatal 12 lead ECG confirming second degree AV block with 2:1 conduction.

P72

Common arterial trunk – the prenatal pattern of development in human

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Introduction: Common arterial trunk (CAT) is a complex cyanotic congenital heart disease with mixed surgical outcomes and guarded prognosis. The current knowledge, based on animal models, and neonatal, surgical or autopsy series, does not reflect fetal epidemiology and morphological characteristics. The aim of the study was to elucidate the prenatal development of CAT.
Methods: Of the 49 cases of CAT diagnosed, the recordings of the most recent cases (since 2016) were reviewed. The measurements and z-scores were noted according to the published convention [Schneider 2005, Pasquini 2007]. Common arterial valve and trunk sizes was normalized based on aortic valve and ascending aorta reference data. Data is given as mean +/- standard. T-test was used to compare the studied sample with a normal population. $P<0.05$ was considered significant.

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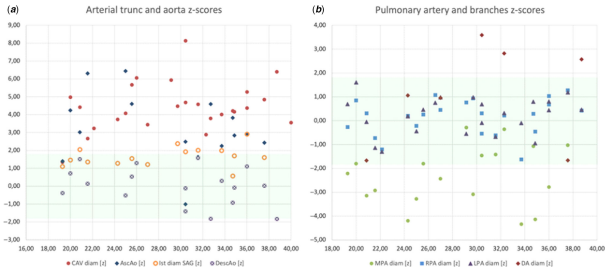
Results: Twelve cases and 28 studies were included (one twin). The gestational age (GA) at the first visit was 27.2 +/- 6.0 weeks. Of seven cases genetically tested, 43% were abnormal. There was one termination of pregnancy and one intra-uterine demise, one neonatal death with the remnant undergoing surgery. There were two cases of II/III type and two with an interrupted aortic arch, the rest were type I. The truncal valve cusps were variously thickened in 75% percent of the fetuses, and this was associated with truncal valve stenosis and regurgitation. The arterial duct was present in 33%.Z-scores were (p – compared to normal): CAV 4.4 ± 1.4 (enlarged, <0.001), AscAo 2.6 ± 3.0 (enlarged, <0.001), IstSAG 1.7 ± 0.6 (larger <0.001), DescAo 0.0 ± 1.1 (0.982), MPA -2.3 ± 1.3 (diminished <0.001), RPA 0.1 ± 0.8 (0.419), LPA 0.2 ± 0.8 (0.257), DA 1.1 ± 2.1 (0.176). Cardiac chambers and atrioventricular valves dimensions were normal. There was no significant change in the structure size (z-scores) compared second and third trimester (e.c. CAV 4.0 ± 1.4 vs 4.8 ± 1.3, p=0.168).
Conclusions: The truncal valve and artery are enlarged already in fetal life, while the main pulmonary artery, if present (type I), is diminutive. The isthmus was enlarged in cases without the ductal arch. The normalized dimensions of the structures did not change throughout the second half of the pregnancy.



P73
Defining normal values of fetal myocardial performance indexes and inflow times – a single-center, retrospective study

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Introduction: Myocardial Performance Index (MPI, Tei index) is the most commonly utilized measure of fetal global myocardial function. Diastolic function remains difficult to assess due to different prenatal physiology, yet it may be indirectly estimated from ventricular inflow times. Our aim was to define normal values of MPIs and inflow times based on the data from a large retrospective cohort.
Methods: We retrospectively reviewed normal fetal echocardiographic studies (n=12494) performed in a single center between 2011 and 2019. Values of AV-closure-to-open times, ejection times and heart rate (FHR) for both ventricles were retrieved to calculate MPIs, inflow times (LVIT, RVIT) and inflow times indexed over heart cycle length (LVIT%, RVIT%). Data were later analyzed to define normal ranges across different gestational ages (GA) and FHR values if appropriate.
Results: MPI values remained independent of FHR and showed a slight, yet significant tendency to increase with advancing GA: LV MPI from 0.36±0.06 in 11th week to 0.43±0.07 in 39th week (RV MPI: 0.36±0.08 and 0.42±0.11 respectively). IT and IT% values followed linear regression and showed significant correlation with both GA (positive) and FHR (inverse) – see table below. Because of relatively wide variance of normal values, data were used to create percentile tables instead of providing with regression equations.
Conclusions: MPI remains a useful method to assess fetal global myocardial function, as it shows similar values for both ventricles and remains relatively stable throughout advancing pregnancy and with different FHR. Inflow times may serve as a reflection of diastolic function, yet have to be interpreted together with GA and FHR values. Created nomograms may serve as a reference to evaluate and monitor fetal ventricular function in different clinical situations.

Parameter	GA β-coefficient	GA r	FHR β-coefficient	FHR r	Range of values 11-39th week
LVIT [ms]	0.03	0.32	–0.80	–0.82	139-200 ms SD 13-22 ms
LVIT%	0.043	0.22	–0.477	–0.49	0.38-0.45 SD 0.03
RVIT [ms]	–0.03	0.24	–0.74	–0.73	135-184 ms SD 13-21 ms
RVIT%	–0.057	0.08	–0.368	–0.35	0.38-0.42 SD 0.03

P74
Descending aortic flow characterization by cardiovascular magnetic resonance

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Introduction: Neonatal coarctation of the aorta (CoA) is associated with significant risk for severe complications if not diagnosed in time. Prenatal diagnosis by fetal echocardiography remains difficult with low detection rates in most centers. We postulated that altered flow through an anomalous aortic arch might lead to flow disturbance in the fetal descending aorta (DAo) with subsequent changes in aortic distensibility. The latter has been shown to be impaired in CoA. We therefore assessed the feasibility and prognostic utility of fetal cardiovascular magnetic resonance (CMR) imaging in improving prenatal diagnosis of CoA.
Methods: This prospective study included eight fetuses (gestational week 31–38) with echocardiography-based prenatal suspicion of CoA and five healthy controls of similar gestational age. Phase-contrast fetal CMR at 1.5T Aera (Siemens, Erlangen, Germany) gated by Doppler ultrasound (Hamburg, Germany) provided quantification of blood flow and DAo area change between maximum and minimum area over the cardiac cycle.
Results: Three of the eight (37 %) fetuses with prenatal suspicion of CoA developed coarctation and underwent CoA repair during the first week of life. In one patient CoA was confirmed within the first hours after birth due to a hypoplastic isthmus with a slightly

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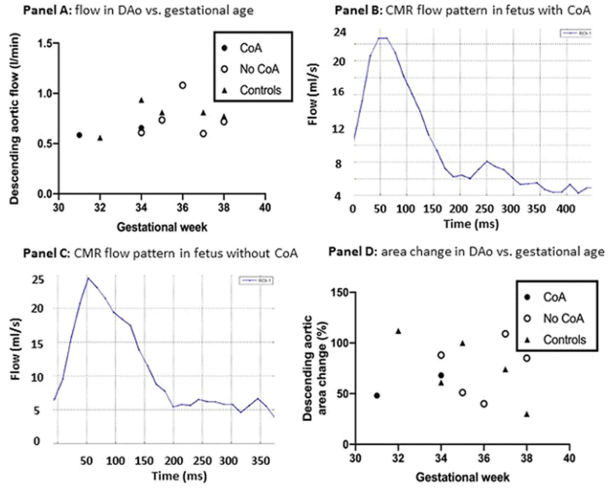
abnormal flow pattern. The other two patients developed the typical diastolic isthmus flow pattern more than 24 hours postpartum, at almost complete closure of the arterial duct. In one fetus CMR flow analysis was inconclusive for flow curve shape with no area change over the cardiac cycle. However, flow volume was adequate for gestational age. As it was unclear whether this was related to error in image acquisition or to pathology, this fetus was excluded from further analysis. Total DAo flow indicated correlation with gestational age, but not with postpartum diagnosis (Figure 1A). DAo flow patterns did not differ between groups (Figure 1B–C). DAo area change was not related to gestational age, nor did it correlate with postnatal CoA diagnosis (Figure 1D).
Conclusions: Assessment of fetal descending aortic flow and area change using fetal CMR is feasible. Further studies are needed to investigate whether CMR-based flow characterization can improve fetal CoA diagnosis.



P75
Early Fetal Cardiac Screening: Detection of Congenital Heart Disease

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Introduction: Routine fetal cardiac screening forms part of the UK Fetal Anomaly Screening Programme between 18 to 20 weeks + 6 days gestation. Higher frequency transducers allowing for better image resolution, training and improved operator experience have enabled fetal echocardiography to be performed and confirmation of congenital heart disease (CHD) in the late first and early second trimester of pregnancy. Early diagnosis of CHD will allow parents to make informed decisions regarding the pregnancy and to allow for planning for further management. Prenatal cardiac diagnosis also allows for additional fetal evaluation for extracardiac anomalies and genetic assessment in the form of invasive testing. The objective of this review was to evaluate the feasibility and utility of early fetal echocardiography for detection of CHD in high risk pregnancies.
Methods: Review of patients referred for fetal echocardiography performed between 13-17 weeks' gestation at a single tertiary fetal cardiac unit over a three year period (January 2015 to December 2018). Indications for referral, fetal cardiac findings, extracardiac findings, genetic results as well as outcome of pregnancies were reviewed.
Results: 86 patients had early scans. 42 with raised Nuchal Translucency (NT) and 44 for previous seriously affected pregnancy. Nine patients with cardiac anomalies were picked up in early screening, 4 of which had normalised at the 18-20 week scan. There were no missed cardiac diagnoses. In the raised NT group with cardiac abnormalities, 3 patients had abnormal karyotype and two of these pregnancies were terminated. There were no patients with genetic abnormalities in the group referred due to previous seriously affected pregnancy.
Conclusions: Early fetal echocardiography is feasible and accurate, providing earlier cardiac diagnosis in affected pregnancies and reassurance when a normal fetal echocardiogram is found. The quality of the images is of sufficient resolution to enable confident diagnosis of congenital heart disease. It should be considered in addition to, but not instead of routine fetal cardiac assessment at 18-20 weeks to maximise detection and accuracy of diagnosis. With more training and experience, it would be expected that early fetal cardiac screening will become the routine in many fetal medicine units.

P76
Early prenatal detection of isolated ascending aorta dilatation, a single center experience

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Introduction: Objectives: to report our experience in management of isolated ascending aorta (AscAo) dilation detected early during fetal life.
Methods: Background: aortic root dilation is usually described in fetuses affected by genetic disorders of the connective tissue and it is usually associated with other intra or extra-cardiac anomalies. To the best of our knowledge we reported the only case of early isolated fetal AscAo dilation which resulted positive at the genetic testing for Loeys Dietz Syndrome after birth (Viassolo et al. Prenatal Diagnosis 2006). We report other 3 cases of isolated AscAo dilation detected during the II trimester of gestation. None of them had associated intra or extracardiac anomalies.
Results: Case 1: at 20 weeks of gestation the AscAo was dilated (5.8 mm, z score +4). At 28 weeks of gestation AscAo was measured 7.2 mm (z score +2.5). The baby was born at term and he presented a normal phenotype. At the echocardiogram performed at 6 months of age the aortic root showed normal dimension. The genetic testing identified heterozygous mutation in ABCC9 gene that is associated with Cantù syndrome. Case 2: at 19 weeks of gestation the AscAo was dilated (6 mm, z score +4). At last prenatal ultrasound evaluation the AscAo measured 10 mm (z score +2.5). The baby born at term, he presented a normal phenotype; at echo evaluation we showed a progressively normalization of the aortic root measurements. The genetic analysis did not show any evidence of collagenopathy. Case 3: at 20 weeks of gestation the AscAo was dilated (6.5 mm, z score +5). At 36 weeks of gestation the AscAo measured 10 mm (z score +3). After birth the aortic root measurements progressively normalized at echocardiographic evaluation. She presented also ankle hyperlaxity. The genetic analysis did not show any evidence of collagenopathy.
Conclusions: Conclusions: on the basis of our experience the early detection of isolated AscAo dilation has a good short-term outcome. However the parental counseling remain challenging for the fetal cardiologist because this prenatal finding could also be the early marker of a genetic disorder and may have the indication to test the implicated genes.

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P77

Feasibility of non-gated dynamic fetal cardiac MRI for identification of fetal cardiovascular anatomy

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Introduction: We sought to evaluate the feasibility of identifying the fetal cardiac and thoracic vascular structures with non-gated dynamic balanced steady-state free precession MRI sequences.
Methods: We retrospectively assessed the visibility of cardiovascular anatomy in 66 fetuses without suspicion of congenital heart defect (range 21-38 weeks). Non-gated dynamic balanced steady-state free precession (SSFP) sequences were acquired in three thoracic planes (axial, coronal and sagittal). The segmental anatomy was defined in consensus reading by an experienced paediatric cardiologist and radiologist. An imaging score was created by giving one point to each visualized structure. Basic diagnostic structures included the atria, ventricles, systemic veins, right and left ventricular outflow tracts (RVOT/LVOT), aortic arch, descending aorta (DAO), ductus arteriosus and thymus (12 points); advanced diagnostic features included the atrioventricular (AV) valves, pulmonary arteries and veins, supraaortic arteries and trachea (maximum score 21 points). Image quality was rated from 0 (poor) to 2 (good). The influence of gestational age (GA), field strength, placenta position, and maternal panniculus on image quality and imaging score were tested.
Results: 34 scans were performed at 1.5 T, 32 at 3 T. Cardiac position, atria and ventricles were seen in all 66 fetuses. Basic diagnosis (>12 points) was achieved in 60 (90%) cases, with visualization of the IVC and SVC in 65 (98%) and 63 (95%), RVOT in 62 (94%), LVOT in 61 (92%), aortic arch in 60 (91%), DAO in 64 (97%), and ductus arteriosus in 59 (89%) fetuses. The AV valves were recognised in 55 (83%), the pulmonary arteries in 35 (53%), at least one pulmonary vein in 46 (70%), the supraaortic arteries in 42 (64%), and the trachea in 59 (89%) fetuses. The mean imaging score was 16.8 +/- 3.7. Maternal panniculus (r -0.3; p 0.01) and GA (r 0.6; p < 0.001) correlated with imaging score. Image quality was better on 1.5 T than 3T (p 0.04) while the total imaging score showed no significant difference.
Conclusions: Fetal heart MRI with a non-gated SSFP enables recognition of basic cardiovascular anatomy. Advanced diagnostics may be limited by thick maternal panniculus, lower GA and higher field strength.

P78

Fetal Isolated Atrial Septal Aneurysm: Does it matter?

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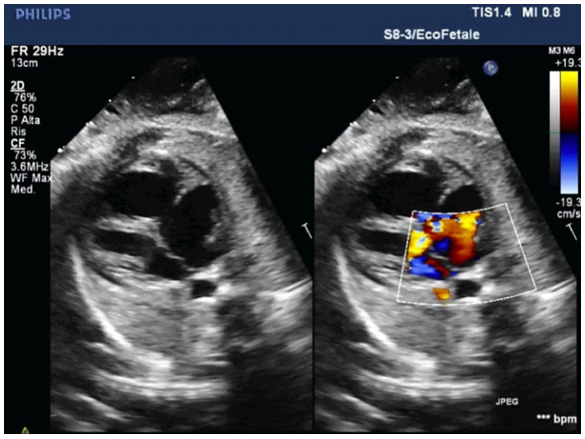
Introduction:
Objectives: to describe the prenatal diagnosis and the postnatal outcome of isolated atrial septal aneurysm (ASA).
Background: few case series have been published regarding fetal isolated ASA and its impact on heart development and on prognosis.
Methods: Methods: we included all consecutive fetuses diagnosed with isolated ASA between January 2015 and December 2019. ASA was defined as a large redundant structure that herniated more than 50% across the length of the left atrium. Only the last fetal scan was considered.
Results: Results: 40 cases were identified with a prevalence of 2.3%. Mean gestational age was 34 (±2.5 weeks).16/40 (40%) fetuses presented left to right size discrepancy both at the level of ventricles and great vessels, 4/40 only of the ventricles. 7/40 (2%) showed retrograde flow in the aortic arch. Acceleration of flow through one or more pulmonary veins and/or through the mitral valve due to ASA was detected in 12/40 (30%). 11/40 (27%) had some degree of tricuspid regurgitation. Among fetuses with irregular heart rhythm (8/40, 20%) none had major arrhythmia except one who developed an atrial flutter at 38 weeks of gestation. The arrhythmia required an urgent cesarean section and external cardioversion. All other patients were in good clinical condition at birth and did not need of any treatment except 4 that developed neonatal aortic coarctation (4/40, 10%). None developed persistent pulmonary hypertension or low cardiac output syndrome. All fetuses with premature atrial beats in utero developed a regular sinus rhythm within 3 months of life. At a mean follow up of 9 months (±11), 2/40 (5%) had a large atrial septal defect and 22/40 (55%) had a patent foramen ovale (PFO).
Conclusions: Conclusions: prenatal diagnosis of isolated ASA is not uncommon and it is frequently associated with size disproportion both at the level of the ventricles and of the great arteries. In our experience four patients developed aortic coarctation. ASA may also represent a mechanical stimulus for generation of premature atrial beats but it is rarely associated with major arrhythmias. Short term outcome was good for all liveborn infants even if most of them showed a PFO at last scan.



P79

Fetal predictors for Aortic Coarctation in patients with disproportion of cardiac structures.

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Introduction: Despite advances in prenatal ultrasound and due to fetal physiology the prenatal diagnosis of Aortic Coarctation (CoA) is still a challenge. The most common sign of diagnosis is Disproportion of cardiac chambers and vessels, in spite of its low specificity. The aim of this study is to evaluate the prenatal signs in the CoA and an intermediate follow-up.

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Methods: Retrospective study of fetal echocardiograms with cardiac asymmetry in third trimester between 2011 until 2020. We analyzed the following parameters: diameter of cardiac valves, great vessels, aortic isthmus and ductus; right (RV) and left ventricular (LV) length, the flow in the isthmus (normal, prolonged, reverse) and presence of restrictive foramen ovale (RFO). Ratios of RV and LV, aortic and pulmonary valve, mitral and tricuspid valve (MV:TV ratio), and ductal and isthmus diameter ratio (D:I ratio) were calculated. These measures were compared between those with CoA after birth (CoA group) and those without (no CoA group). Follow-up was until October 2020 and analyzed outcomes (mitral stenosis, bicuspid aortic valve and ventricular septal defects).
Results: Of the 107 subjects, 33 were in CoA and 74 in no CoA group. CoA group had a significantly smaller mitral valve (p=0,025), higher MV:TV and D:I ratio (p=0,034 and p<0,005) compared to no CoA group. No significantly differences in ventricular ratio and great vessels ratio were demonstrated. The ROC curves showed that D:I ratio had the highest AUC (0,88). Almost no RFO was detected in the CoA group (only 2 cases of 38, p<0,005). We studied the isthmus flow in 52 subjects (only in the last 3 years). Higher proportion of normal flow in the CoA group (22) was detected. In the intermediate follow-up, 16 aortic bicuspid valve, 19 ventricular septal defects, 12 mild-moderate and 1 severe mitral stenosis were detected.
Conclusions: CoA is still requiring a lot of parameters for its accurate diagnosis. This study shows that ratio ductus:isthmus, the absence of RFO and a normal isthmus flow are the most significant parameters in the fetal cardiac asymmetry for CoA. The presence of RFO in the no CoA group may explain the different etiology in these asymmetries.

P80

Fetal ventricular tachycardia as a rare prenatal presentation of congenital LQT3 syndrome.

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Introduction: Fetal arrhytmias concern 1-2% of pregnancies. Among them, ventricular tachycardias (VT) are exceptional and, if they are an expression of a long QT syndrome (LQTS), they can cause fetal loss.
Methods: F.D. 38-y-old was referred to our Service for fetal echocardiography at 32 weeks of pregnancy because of fetal tachyarrhythmia. Ultrasound showed several episodes of VT with ventricular rates between 280 and 350 bpm, with a structurally normal heart. Maternal treatment with Sotalol 160 mg/die didn't achieve great results. Due to initial fetal heart failure, an emergency caesarean was performed at 33 weeks of gestation, and Z.E. was born in initial stable hemodynamic conditions. ECG presented an arrhythmic storm with polymorphic VT and several phases of torsade de pointe (TdP). Brief moments in sinus rhythm showed a QTc interval of 700 msec with dysmorphic T waves, strongly evocative of a LQTS type 3.
Results: Neither MgSO4 nor Lidocaine, at low doses in consideration of prematurity, or Esmolol could control arrhythmias. When hemodynamic conditions became critical, we decided to try with Lidocaine at maximal therapeutic doses, gaining the conversion to SR immediately. Orally treatment with Mexiletine at 10 mg/kg/die and Propranolol at 5 mg/kg/die was started instead of intravenous drugs, obtaining definitive stabilization of baby's rhythm (SR with QTc 450-480 msec). Genetics confirmed a malignant heterozygotic mutation of SCN5A, responsible of LQTS3, associated with p.R1047L variant of gene KCNH2, which, according to literature, is associated with an increased risk of TdP. After 4 months hospitalisation he suffered a sudden cardiac arrest, promptly resuscitated, so a definitive ICD implantation was performed using a Medtronic DVF C31D with an epicardial electrode on the right ventricle. The baby was at home asymptomatic for 1 year, then, when he was 17-moths-old, arrhythmia storms reappeared again, so a left cardiac sympathetic denervation (LCSD) with removal of the first four ganglia was performed with surgical approach (left thoracotomy), with no arrhythmic recurrences so far.
Conclusions: In presence of fetal VT, LQTS must always be sought, especially type 2 and 3, as they are often responsible for early onset malignant arrhythmias, require specific treatments and carry a poor prognosis.

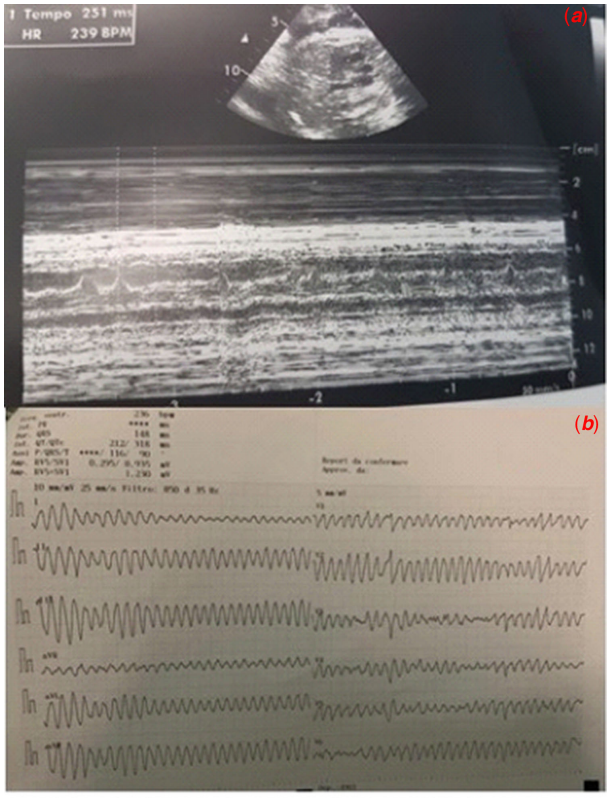

Figure 1-A: M-MODE fetal ultrasuonds showing high rates VT; 1-B: neonatal ECG showing TdP

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P81

Impact and precision of prenatal diagnosis in common arterial trunk

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Introduction: Outcomes of common arterial trunk (CAT) depend mainly on truncal valve function, coronary artery abnormalities and presence of interrupted aortic arch. Little is known about the impact and precision of prenatal diagnosis of CAT in the current era.

Methods: Retrospective analysis of all diagnosed CAT patients with pre-and postnatal diagnosis or fetal autopsy confirmation from 2011-2019 in a single tertiary center. Postoperative mortality and morbidity was analyzed. Cohen's Kappa statistic was used to measure agreement of prenatal diagnosis with postnatal assessment regarding anatomic subtypes according to Van Praagh and truncal valve function.

Results: 84 patients (62 live born with prenatal diagnosis/16 live born with postnatal diagnosis/ 6 terminations of pregnancy with fetopsy) met inclusion criteria. There was no difference in postoperative mortality for CAT patients with or without prenatal diagnosis (log rank p=0.87). CAT patients with prenatal diagnosis underwent earlier intervention (p=<0.001), had shorter intubation time (p=0.047) and shorter global hospital stay (p=0.01). Diagnostic precision for prenatal diagnosis of CAT subtypes was moderate (Kappa κ= 0.43) with a pre-and postnatal discordance in 19%. Notably, no fetus had a prenatal diagnosis of CAT Type 3, and only approximately half of patients with CAT Type 4 (8/17) were diagnosed prenatally. Prenatal evaluation of truncal valve function tended to underestimate the presence of insufficiency, since 21 (62%) of 34 fetuses with no prenatally visible truncal regurgitation finally had some degree of insufficiency after birth (κ=0.09). Prenatal assessment of severity of valvular regurgitation showed only a slight agreement with postnatal evaluation: 5 fetuses had significant insufficiency prenatally, while 12 had moderate to severe truncal regurgitation after delivery (κ=0.19). Concordance for the diagnosis of truncal valve stenosis was moderate with 4 of 5 postnatally confirmed significant stenoses having been diagnosed prenatally (κ=0.38).

Conclusions: Prenatal diagnosis of CAT has no impact on postoperative mortality but tends to favorably influence morbidity related to initial surgery. However, the precision of prenatal diagnosis for aortic arch anatomy and truncal valve function has to be improved. Anatomic subtypes 3 and 4 remain largely underdiagnosed during fetal life with potential negative impact on neonatal management strategies and on functional outcomes.

P82

Initial survey of detection rate for prenatal diagnosis of congenital heart malformations in Japan

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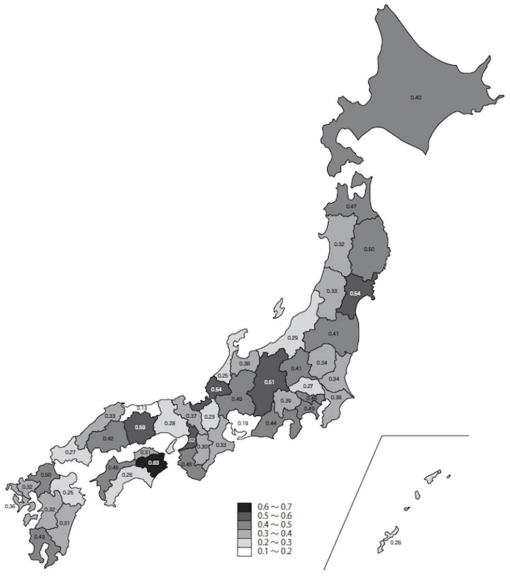
Introduction: Investigation of detection rates (DR) for prenatal diagnosis of moderate to severe congenital heart diseases is essential for evaluation of effectiveness of the fetal cardiac screening system.

Methods: This is an initial survey of DR by database analysis of national registry by national clinical database for congenital heart surgeries (Japan CardioVascular Surgery Database: JCVSD) during 2013-2017 in Japan. Subjects are neonates or infants with moderate or severe congenital heart diseases who received heart surgeries until the age of one. Ventricular septal defect, Patent ductus arteriosus and atrial septal defect are excluded from the subjects. The number of cases of prenatal diagnosis, DR, types of congenital heart diseases are analyzed.

Results: DR is 0.41 (3425 cases of fetal diagnosis in 8339 cases of the subjects) and increased 2 percentage every year in the five-years period. No significant relationship between DR and population in each prefecture is found. DR is more than 0.50 in 6 prefectures and is less than 0.30 in 11 prefectures of 47 prefectures. DR is 0.67 in single ventricle including hypoplastic left heart syndrome, 0.42 in transposition of great arteries and 0.41 in tetralogy of Fallot or double outlet from the right ventricle while that is 0.36 in atrioventricular septal defect and 0.29 in coarcation of the aorta or Interrupted aortic arch, respectively. DR in isolated total or partial anomalous pulmonary venous connection is 0.13.

Conclusions: Detection rate is still low and common national system for fetal cardiac screening is required in Japan. (This work was supported by JSPS KAKENHI Grant Number 18H06207/ 19K21310.)



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P83

Long term outcome in 39 fetal cases with rhabdomyomas

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Introduction: Rhabdomyomas (R) are more common cardiac tumors detectable in utero. Counselling regards both possibility of association to tuberous sclerosis (TS) and prediction of hemodynamic complications. The aim of this study was to analyse retrospectively the long term outcome of our cases.
Methods: Between Jan.1987 and Dec. 2019 39 fetuses (0. 60% of 6500 studied by fetal echocardiography) had cardiac masses, suggestive of R, at 21-36 w.g. and were followed-up in utero and postnatally for a median period of 11 yrs (1 – 33 yrs).
Results: The diagnosis (dg) was made before 24 w.g. in 13 fetuses, 3 with 1-2 small noduli and 10 with 1-2 medium-large masses: 8 opted for termination of pregnancy (TP) and had a postmortem histological dg. of R. Three fetuses diagnosed at 27 and 28 w. with multiple masses opted also for TP (in another country), after positive MRI findings for TS. The remaining fetuses were diagnosed at 26-36 w.g. All presented multiple masses with partial inflow and/or outflow obstruction in half of them. Two fetuses had mothers with TS, one with a large intrapericardial mass. One fetus presented Bourneville syndrome- with polycystic kidneys of adult type.

Growth in utero: The size of R progressed usually till 35-36 w.g. Diagnosis of TSC was made in 26 (78% of investigated cases) by MRI findings in utero or after birth and by genetic tests.

Long term outcome: none of the live born cases needed immediate cardiosurgery, while one child developed a severe aortic stenosis, after a progressive regression of a large mass involving aortic valve and was successfully operated at 10 yrs. The cardiac R progressively regressed over time, 2 on Everolimus therapy; 8 have extrasystolic arrhythmia, 2 with short episodic tachycadia. All but one TS cases have seizures and 3 were operated for astrogliomas. Minor renal, dermatological and ocular signs are present in third of them. The case with Bourneville syndrome is on dialysis.
Conclusions: Cardiac R show a big variability in presentation and growth in utero and at the long term outcome, with regression of the masses but with a relevant association to TS that conditions the clinical state.

P84

Outcomes following fetal diagnosis of Ebstein anomaly/ Tricuspid valve dysplasia in Bristol Royal Hospital for Children

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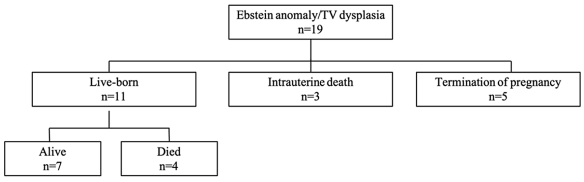
Bristol Royal Hospital for Children (1)

Introduction: Ebstein anomaly/tricuspid valve (TV) dysplasia diagnosed in fetal life carries a high risk of perinatal mortality. Predictors associated with worse outcome include gestational age (GA) <32 weeks at diagnosis, reduced/absent pulmonary flow, retrograde duct flow, pulmonary valve regurgitation (PR) and hydrops. (1,2) We aimed to review the mortality in this cohort of patients and some of the most important predictors of poorer outcome.
Methods: We reviewed retrospectively all the clinical information and echocardiographic parameters of the patients with fetal diagnosis of Ebstein anomaly/TV dysplasia in Bristol Royal Hospital for Children between 01/03/2013 and 01/11/2020.
Results: There were 19 fetuses diagnosed with Ebstein anomaly/ TV dysplasia, at a median 20.8 weeks GA. All but one was diagnosed before 32 weeks GA, one was diagnosed at 37 weeks GA following an abnormal presentation scan. There were 5 terminations of pregnancies and 3 intrauterine deaths. There were 11 live-born, of whom 2 died within an hour after birth, 2 died before discharge at around 2 months of age, and 7 are alive. Among the 7 survivals, 2 had tricuspid valve replacement with RV to PA reconstruction and 1 had TV repair with Glenn anastomosis before discharge, all of them developed complete heart block after surgery. 4 are well without any intervention at median 1 year of follow up (0.4-6.3 years). There were 12 fetuses diagnosed with absent/reduced pulmonary flow of whom 4 are alive (3 had surgery before discharge), 4 died and there were 4 terminations of pregnancy. All of the fetuses with absent/reduced PA flow had retrograde ductal flow. PR was present in two, 1 had hydrops but survived requiring surgery, 1 was born in poor condition and died within an hour after birth. There was frequent atrial ectopic beats in 1, and intermittent faster heart rate in 1, who had AVRT postnatally.
Conclusions: The prognosis following fetal diagnosis of Ebstein anomaly/TV dysplasia is poor. Only 36% of the live-born fetuses are well without any intervention. The overall mortality rate was 50% in our centre, comparable with other series in the literature.



P85

Prenatal diagnosis of persistent left and bilateral superior vena cava and association with extracardiac malformations

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Introduction: Advanced fetal echocardiography has increased the prenatal detection of variants of systemic venous drainage. Persistent left superior vena cava (PLSVC) can be associated with an increased risk of congenital heart disease (CHD) and laterality disturbances but may also be a variant of normal. Our aim was to investigate whether the detection of bilateral SVCs (BSVCs) was associated with the presence of extra-cardiac anomalies (ECA) or genetic abnormalities
Methods: A retrospective review of all cases of PLSVC and BSVCs detected prenatally between October 2014 and 2019 in a tertiary referral centre in Scotland. Patient notes and ultrasound reports were reviewed for ECAs; genetic abnormalities and postnatal outcome. The presence of PLSVC was diagnosed based on a referral for an abnormal three vessel/tracheal view or because a dilated coronary sinus was identified in the four-chamber view.
Results: 61 cases of PLSVC/BSVC (N=61/792, 7.7%) were identified; isolated PLSVC/BSVC represented 27 cases (44.3%). 12 fetuses had an isolated PLSVC; only one had an associated ECA – bilateral ventriculomegaly. 4/15 fetuses with an isolated

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BSVC had at least one ECA, +/- a genetic abnormality. One fetus had a small cerebellum and ventriculomegaly and a deletion on chromosome 6 and 7. The second fetus was diagnosed with duodenal atresia, anal atresia and a horseshoe kidney. The third case had a skeletal dysplasia with no genetic abnormality. The last fetus had hearing loss and trisomy 21. In the group with associated CHD (N=34/61, 55.7%), twelve fetuses (35.3%) were diagnosed with extracardiac malformations with or without abnormal genetic tests. In the latter population, BSVCs was observed in 66.7% (N=8/12). Presence of BSVC resulted in a positive predictive value of 26.3% for the co-existence of an ECA. After excluding cases of pregnancy termination and stillbirths the survival rate in fetuses with BSVCs and extracardiac conditions was 66.7%.
Conclusions: The presence of isolated PLSVC is uncommonly associated with ECAs, however the presence of isolated BSVCs increases the risk of an associated ECA or genetic abnormality. Therefore, detection of BSVC during fetal echocardiography warrants detailed assessment by a fetal medicine specialist.

P86

Simple TGA: Comparison of outcomes with standby transport for prenatally diagnosed patients. Experience at a single tertiary cardiac centre

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Introduction: Transposition of great arteries (TGA) is the second commonest congenital cardiac defect and is increasingly diagnosed prenatally. To improve postnatal outcomes we introduced a perinatal policy to offer options of normal delivery (SVD) versus caesarean section (CS), to have the transport team and ambulance on standby for prenatally diagnosed TGA. We review our experience and compared outcomes.
Methods: We retrospectively reviewed our cardiac and prenatal databases for patients diagnosed with simple TGA born between 01/01/2017 - 30/06/2020. We excluded patients with significant ventricular septal defects or outflow tract obstructions. Patients were grouped into prenatally diagnosed before (A), after (B) the perinatal policy, and postnatally diagnosed (C). We analysed and compared the outcomes between groups A&B, and groups B&C.
Results: All groups were closely gender, weight and gestation matched. Comparing groups A&B there was no statistically significant difference in mortality, age at arterial switch or post operative complications. In group B there were statistically significant higher CS rates, earlier age of arrival to the cardiac centre, better saturations at arrival and reduced total length of stay. Comparing groups B&C, there was no statistical difference in saturations on arrival, intubation rates, mortality, complications or total length of stay. Group C had statistically significantly lower CS rate, were older at age of arrival and therefore older at septostomy.
Conclusions: Our new perinatal policy reduced the time between birth and arrival at our cardiac centre, improved saturations, but resulted in significantly increased CS rate. Although there was no significant change in complications, mortality or total length of hospital stay we feel this has enabled us to overcome geographical distance from the neonatal unit. A larger population sample may have resulted in slight improvement in immediate neonatal outcomes. For offsite neonatal units timely transfer is of paramount importance. The ability to forward plan impacts cardiac intensive care resources and families' experience. With the introduction of elective section in this group of patients, we have streamlined and improved the process.

Table 1: Prenatal diagnosis comparison Groups A versus B

	Group A: Jan 2017 - June 2018 Antenatal (n=18)	Group B: July 2018 - June 2020 Antenatal (n=16)	P value
Gender	11 males, 7 females	13 males, 3 females	
Weight (Kg)	Median 3.34 Range 3 - 4.05	Median 3.9 Range: 2.41 - 3.99	
Premature <36/40	NIL	NIL	
Delivery methods (CS= caesarean section; NVD = normal vaginal delivery)	10 CS; 8 NVD	13 CS; 3 NVD	P = 0.04
Age at arrival to cardiac centre (hours)	Range: 01:26 – 05:10 Median: 02:40	Range: 01:09 – 03:29 Median: 01:39	P= 0.0049
SATs at presentation (%) [median, range]	55 (20-80)	68 (40-82)	P=0.02
Mortality pre Arterial switch	1 of 18	0	
Mortality post Arterial switch	0 of 17	3 of 16	NS (P=0.3)
Septostomy	16 of 18	14 of 16	NS
Age at septostomy (hours)	Median: 1.5 Range: 0 - 24	Median: 3 Range: 2 – 8	NS (P=0.2)
Age of switch (days)	Median: 12 Range: 8 - 32	Median: 15 Range: 7 – 23	NS (P=0.5)
Post arterial switch complications	6 of 17	9 of 16	NS (P=0.3)
Intubation at delivery	16 of 18	16 of 16	NS (P=0.5)
Total length of hospital stay (days)	Median: 24 Range: 15 - 52	Median: 33 Range: 12- 49	P = 0.03

Table 2: Comparison between Groups B versus C

	Group B: July 2018 - June 2020 Antenatal (n=16)	Group C: Jan 2017 - June 2020 Postnatal (n = 17)	P value
Gender	13 Males, 3 females	14 Males, 3 females	
Weight (Kg)	Median 3.9 Range: 2.41 - 3.99	Median 3.15 Range: 2.14 - 3.86	NS (P=0.1)
Premature <36/40	NIL	1 of 16	NS
Delivery methods (CS= caesarean section; NVD = normal vaginal delivery)	13 CS; 3 NVD	6 CS; 11 NVD	P = 0.00001
Age at arrival to cardiac centre (hours)	Range: 01:09 – 03:29 Median: 01:39	Range: 05:45 – 20:26 Median: 11:38	P=0.00001
SATs at presentation (%) [median, range]	68 (40-82)	70 (40-96)	NS
Mortality pre Arterial switch	NIL	NIL	
Mortality post Arterial switch	3 of 16	NIL	NS (P = 0.1)
Septostomy	14 of 16	15 of 17	NS
Age at septostomy (hours)	Median: 3 Range: 2 – 8	Median – 9 Range: 1 - 54	P=0.01
Age of switch (days)	Median: 15 Range: 7 – 23	Range: 2 - 52 Median: 14	NS
Post arterial switch complications	9 of 16	4 of 17	P = 0.08
Intubation for transfer	16 of 16	16 of 17	NS
Total length of hospital stay (days)	Median: 33 Range: 12- 49	Median: 25 Range: 9 – 49	NS (P=0.09)

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5. Adult congenital heart disease

P87
morbidity and mortality for surgery of adult congenital heart diseases: 10 years-experience

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Introduction: Adult congenital heart disease surgery has particular characteristics that differentiate it from congenital heart surgery in children and cardiac surgery for acquired diseases in adults. Our objective is to assess the immediate and 6-month results of adult congenital heart disease surgery in terms of morbidity and mortality.
Methods: It is a mono-centric, descriptive and retrospective study carried out over a period of ten years (2008 to 2018). All adult patients who have undergone a surgical correction of a congenital heart disease are included. A total of 130 patients were identified, divided into 2 groups: Group 1 comprising 52 patients who have cyanogenic congenital heart disease. Group 2 comprising 78 patients who have non-cyanogenic congenital heart disease
Results: Mean age was 32 years old [18 - 76], with an F/M sex ratio of 1.5. Hypertension was the most common comorbidity (20%). Group 1 : atrial septal defect (ASD) in 40 patients (51.3%), Aortic Coarctation in 15 patients (19.2%), Atrioventricular septal defect (AVSD) in 12 patients (15.4%), ventricular septal defect (VSD) and Patent ductus arteriosus (PDA) in 4 patients every one and subvalvular aortic stenosis in 2 patients. Group 2 : Tetralogy of Fallot is the most frequent (53 %; n = 28). Pulmonary atresia with open septum (PAOS) and Single ventricle defect was noted in 9 and 6 patients respectively.

Before surgery, 46% of our patients were symptomatic of dyspnea, 16 % had supraventricular arrhythmias and the mean LVEF was 65.7 ± 10.4%. 85% of operations take place under Cardiopulmonary bypass, were pallaitive in 13 % of cases and redo-surgery in 32% of cases. The mean duration of cardiopulmonary bypass and aortic clamping were respectively 70 min [35- 286 min] and 48 minutes [17-180 min] in Group 1 ; 50 min [20-250min] and 30 minutes [15-176] in Groupe 2. The mean hospital stay was 36 days in group 1 and 22 days in group 2 with a statistically significant difference (p <0.001). In-hospital mortality was 5% and in-hospital morbidity was 31%. At 6-12 months of follow-up, functional improvement was observed in 79% of patients (table).
Conclusions: Our study confirms the safety and immediate good results of Adult Congenital Herat Disease surgery

		Before Surgery	After Surgery	P
Dyspnea	NYHA I NYHA II NYHA III	70(54%) 47(36%) 13(10%)	117(90%) 13(10%) 0(0%)	<0,001 0,001 0,004
Palpitations		35(27%)	8(6%)	0,002
Heart Pain		15(12%)	8(6%)	0,295
ECG	Rythme sinusal	109(84%)	105(80%)	0,799
	Atrial fibrillation	17(13%)	24(18%)	0,585
	Flutter	3(2%)	0(0%)	0,558
TTE	Sys PAP	46,3±14,1	33,3±7,3	<0,001
	RV diameter	41,0±5,8	31,4±7,1	<0,001
	RA surface	28,6±7,4	22,1±5,2	<0,001

Table : evolution of clinical and para-clinical parameters after surgery

P88
21 years follow-up of liver function in patients after Fontan operation (longitudinal study)

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Introduction: The aim of this longitudinal study was to analyse the influence of long-term venous congestion on the liver function and quality of life in repeatedly examined individuals who underwent total cavopulmonary connection (TCPC) in childhood.
Methods: 46 patients (25 men, 21 women) underwent TCPC at median (IQR) age of 4 (3-6) years and were repeatedly examined during 3 subsequent cross-sectional studies at 5 (3-6), 13 (11-14) and 21 (20-22) years after TCPC, respectively. Cardiac function was evaluated by echocardiography and liver function by a comprehensive panel of biochemical and hematological examinations. In addition to those, elastographic examination of liver stiffness, direct biochemical marker of liver fibrosis (ELF) and subjective quality of life (SF-36 questionnaire) were evaluated in Study 3. Repeated measures ANOVA was used to process longitudinal data.
Results: Between Studies 1, 2 and 3 we found progressive impairment of both excretory and synthetic liver function, particularly significant increase in levels of gamma glutamyl transferase ($p < 0.001$), bilirubin ($p = 0.001$) and biochemical markers of hepatic impairment the Fibrosis-4 (FIB-4) index ($p < 0.001$) as well as Forns index ($p < 0.001$) and significant decrease in levels of prealbumin ($p < 0.001$). An ELF index was abnormal (>7.7) in 41/43 (95%) patients. Abnormal elastography (>7.2 kPa) was revealed in all 39 (100%) examined subjects. At last study semiquantitative echocardiographic evaluation revealed good ventricular function in 38/46 (83%), slightly reduced in 6/46 (13%) and moderately reduced in 2/46 (4%) examined subjects. 8/46 patients (17%) rated their health as excellent, 23/46 (50%) as very good, 11/46 (24%) as good, 4/46 (9%) as fair and none as poor.
Conclusions: Progressive impairment of liver function occurs during long term follow-up in patients after TCPC despite preserved ventricular function and good quality of life. Liver dysfunction is a sensitive marker of end organ damage in Fontan circulation.

P89
6p25 deletion syndrome diagnosed in a 52-year-old patient in the first visit to an adult congenital heart disease clinic: a case report

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Introduction: Genetic disorders can affect patients´ health but affection of relatives or transmission to offspring must be taken into account. Congenital heart defects, even mild ones, can associate a genetic origin, so genetic test and counselling may be recommended. A consensus paper has been recently published for this

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purpose. However, there are a great number of adults with congenital heart disease that are not followed in specific clinics, so, probably, they would never receive genetic diagnosis or counselling if needed.
Methods: We report the case of a 52-year-old patient to our adult congenital heart disease clinic, followed in several clinics (general cardiology, cardiac surgery, cardiac interventionalist) for decades, in whom genetic origin was suspected in the first visit, so, after consultation to genetics service, diagnosis of 6p25.3 deletion syndrome was stablished.
Results: A 52-year-old man with history of atrial septal defect surgically close 18 years ago, aortic valve and ascendent aorta replacement with mitral and tricuspid annuloplasty 8 years ago, severe perivalvular aortic leak with severely dilated left ventricle, pending for percutaneous closure and descendent aorta dissection was referred to our adult congenital heart disease clinic. He also had hypertension, dyslipidemia, short stature, mental retardation, hearing impairment and marked kyphoscoliosis. His treatment was omeprazole, ivabradine, furosemide, carvedilol, rosuvastatin, acenocumarol, amlodipine and valsartan. Due to extracardiac findings, consultation to genetics service was done and genetic test was performed (array comparative genomic hybridization (aCGH)). A pathogenic terminal deletion of 2,4 Mb in the band 6p25.3p25.2 that altered several genes, including FOXC1, was found.
Conclusions: Congenital heart disease, even mild ones, can be associated to genetic disorders. This can affect to patients but their relatives and offspring too, so it is important not to miss the diagnosis. Despite of been followed by other clinicians, even during decades, sometimes it is not suspected and genetic testing is not perform. Because of complexity and possible relation to a genetic disorder with the implications it has, we think adult patients with congenital heart disease, even those with mild defects, especially when extracardiac findings are present, should be evaluated in a specific clinic, in close collaboration with genetics services.

P90

Acute vasodilator response testing in the adult Fontan circulation using non-invasive 4D flow MRI – a proof of principle study

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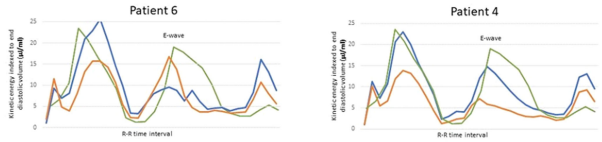
Introduction: A recent meta-analysis has shown the benefit of vasodilator therapy in Fontan patients, with an increase in 6 minute walk time and reduction in NYHA class. Acute vasodilator response is often tested in the catheter laboratory prior to commencing treatment, to determine those likely to benefit. This is invasive and often involves a general anaesthetic which in itself has vasodilatory properties. We aimed to assess the potential for *non-invasive* response using 4D flow MRI during Oxygen inhalation.
Methods: Six Fontan patients with at most only mildly reduced ventricular function, no symptoms and minimal or no atrioventricular valve were prospectively recruited and underwent a cardiac MRI (3T trio, Siemens, Germany). Ventricular anatomical imaging and 4D flow MRI were acquired at baseline and during inhalation of Oxygen 15L/min via a non-rebreath mask after a steady state was achieved. Data was compared with 6 age matched healthy volunteers who underwent 4D flow MRI at baseline only. 4D flow MRI data was analysed using PIE medical imaging software, CASS, The Netherlands and research software MASS.
Results: All patients tolerated the MRI scan well. The underlying diagnosis was tricuspid atresia ± ventricular septal defect in 4/6, pulmonary atresia with intact septum 1/6, unbalanced atrioventricular septal defect 1/6. 5/6 had extracardiac total cavopulmonary connection and 1/6 had an atrial Fontan. On 4D flow MRI assessment, 3 patients (Patient 1, 2 and 6) showed improved cardiac function with improved preload during oxygen administration by improved mitral inflow, improved maximum E-wave kinetic energy and improved kinetic energy flow profile. Patient 5 showed no change and patient 3 had equivocal results. Patient 4, however, showed a decrease in preload and cardiac function.
Conclusions: Using Oxygen administration as a vasodilator to assess increased pulmonary venous return as a marker for positive acute vasodilator response would provide pre-treatment assessment in a more physiological state – the awake patient. This proof of concept study showed that it is well tolerated and has shown changes in some well patients with the Fontan circulation. Further assessment is required in patients with AV-valve regurgitation and reduced cardiac function, as well as determining the reliability compared to invasive vasodilator therapy.

Patient	Age	Baseline Ejection fraction	Ejection fraction change with O2	Mitral inflow change with O2	max E-wave KE change with O2	KE flow curve change with O2
1	20	52	∞	↑	↑	↑
2	19	53	∞	↑	↑	↑
3	20	53	∞	↓	∞	↑
4	27	61	↓	↓	↓	↓
5 (AP)	31	49	∞	∞	∞	∞
6	21	43	∞	↑	↑	↑

Patient 1-6 changes with oxygen administration; KE = kinetic energy, AP = AP Fontan



Patient 6 shows improved kinetic energy profile with improved diastolic E-wave kinetic energy, Patient 4 shows worsened kinetic energy profile; Blue = baseline; Orange = on oxygen therapy; green = average kinetic energy profile of 6 age matched healthy volunteers

P91

Appropriate heart rate response during exercise in Fontan patients

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Introduction: Fontan patients have a reduced maximal heart rate and many have suggested this to be a limiting factor for physical performance. Recent research has shown that limiting factor for exercise capacity is impaired diastolic filling and stroke volume rather than heart rate. The present study was made to evaluate relationship between heart rate response against workload and oxygen uptake during exercise in Fontan patients.
Methods: Young Fontan patients (n = 27, age 14.4 ± 3.1 years) and healthy matched controls (n = 25, age 13.6 ± 3.5 years) underwent cardiopulmonary exercise testing on bicycle ergometer to exhaustion. Heart rate, workload and oxygen uptake were recorded at start (rest), midpoint of test as measure of submaximal effort (mid) and maximal effort (max). Heart rate in relation to workload and oxygen uptake respectively was analysed. Heart rate recovery was recorded until ten minutes after test.

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Results: Heart rate at midpoint and maximal effort (171 ± 14 vs. 191 ± 10 beats per minute, $p < 0.001$) was lower for patients than controls. Heart rate recovery was similar between groups. Heart rate in relation to workload was higher for patients than controls both at midpoint and maximal effort (76.3 ± 13.6 vs 67.1 ± 14.3 $HR \cdot w^{-1} \cdot kg^{-1}$, $p < 0.05$) (Fig). Heart rate in relation to oxygen uptake was similar between the groups throughout the test. Oxygen pulse, indirect measure of stroke volume, was reduced at maximal effort in patients compared to controls (6.6 ± 1.1 vs. 7.5 ± 1.4 $ml \cdot beat^{-1} \cdot m^{-2}$, $p < 0.05$). Oxygen pulse increased significantly less from midpoint to maximal effort for patients than controls ($p < 0.05$).
Conclusions: Heart rate response during exercise is increased in relation to workload in Fontan patients compared with controls. At higher workloads, Fontan patients seem to have reduced ability to increase heart rate and also oxygen pulse. Limitation of heart rate response at maximal effort can be an autonomic protective measure to maintain high stroke volume and cardiac output. Reduced ability to increase stroke volume at higher heart rates may be a more limiting factor for cardiac output than chronotropic incompetence per se.

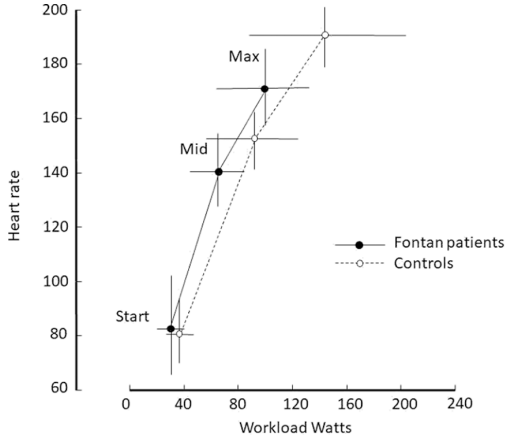

Fig. Heart rate vs. workload (watts) at start, midpoint (Mid) and maximal (Max) effort in Fontan patients and matched controls. Mean ± 1SD.

P92
Clinical characteristics, prevalence and prognosis of infective endocarditis in patients with congenital heart disease

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Introduction: Infective endocarditis (IE) is considered as a life threatening complication in patients with congenital heart disease (CHD) because of it high mortality. IE clinical chracteristics and orognosis in the population of CHD remain little studied.

Purpose: We aimed to determine the incidence, clinical features and evolutive outcomes of IE in patients with CHD.
Methods: A retrospective single centre review of patients who presented IE (defined by the modified Duke criteria) admitted on the cardiology B department of Fattouma Bourguiba University Hospital between january 1998 and march 2017. Prevalence, clinical carachteristics and outcomes were studied.
Results: Among the 235 patients included, 10.2% (24 patients) had CHD. The most frequent CHD was ventricular septal defect (30%).Patients with CHD had lower mean age (24±10 years vs 38±17 years, $p<0.0001$). there was no significant difference for sex. Comorbidity in patients with CHD was low. The most comon valves involved were tricuspid and aortic valve (19% vs 5.44% - 14.3% vs 12.4%,$p<0.0001$ respectively). Half of blood-culture were négative. Among culture-positive patients, staphyloccocus species was the most frequently implicated causative pathogen. Detection of vegetation on trans-thoracic and trans-oesophageal echocardiography was not significantly different (91.3% vs 81.3%, $p=0.186$ and 100% vs 91.8%, $p=0.3$ respectively). Heart failure, embolic events, cerebro-vascular complications and in-hospital mortality were similar compared to patients without CHD. Population with CHD had higher reccurence rate of IE (33.3% vs 9.1%, $p=0.023$).
Conclusions: IE in patients with CHD occurs at young age. It implicates most commonly the right heart and the aortic valve. Its prognosis is not different from IE in the general population but has higher reccurence rate.

P93
Ebstein's anomaly in adult congenital heart disease: 12 year single centre experience

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Introduction: The diagnosis of Ebstein's anomaly (EA) of the Tricispid Valve (TV) occurs across a spectrum of ages and clinical presentation varies from incidental finding to arrythmias and symptomatic heart failure. In this retrospective study we reviewed records of our adult patients with a diagnosis of Ebstein's, to look at our practice.
Methods: We identified 42 patients with a diagnosis of Ebstein's, from 2008-2020. The range was 24-81years and three of the patients had sadly deceased. Data collected included age at diagnosis, surgical history and arrythmias.
Results: Age at diagnosis was available for 42/42 patients. 29 patients were diagnosed in childhood and 13 patients in adulthood, of which 7 were diagnosed over the age of 50 years. The commonest presentation in adulthood was due to palpitations/arrhythmias. 27 patients had an episode of arrhythmia documented in adult age (incidence 76%), mostly atrial tachycardias (including AF/flutter), ventricular tachycardia and complete heart block. 16 patients had pacemakers and/or ICDs. No documented arrythmias in patients under 34 years of age. 31 patients had cardiac surgery in adult age, with seven having had more than one surgery. Incidence of surgery in paediatric & adults was the same. Six patients had a cone repair with no post-operative deaths. Of the three deceased, one died in post-operative period following a TV repair and Glen-shunt procedure a few years back, one died from bowel cancer six years after TV surgery and one was unoperated at the time of death which was due to severe cyanotic Ebstein's anomaly.
Conclusions: Ebstein's anomaly is complex condition, majority will have heart rhythm issues. We believe ours is one of the largest Ebstein's cohort of adult patients being managed in a congenital centre in the UK. We have demonstrated good surgical outcomes including the newer technique of cone repair. Longer-term data in this heterogeneous group of with multi-centric collaboration is important.

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P94

How many patients after Norwood procedure reach adulthood?

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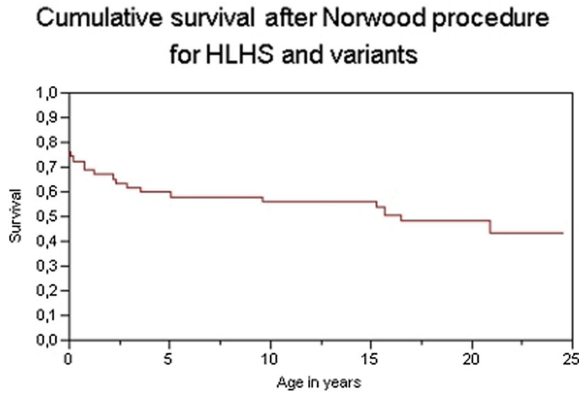
Introduction: With the development of the Norwood procedure in the 1980s, survival became possible for infants with hypoplastic left heart syndrome (HLHS) and related single right ventricle anomalies. Since then, many studies have analyzed early and post Fontan outcomes for this emerging population of children. However, data pertaining to survival and morbidity after patients reach adulthood is limited. We sought to determine cumulative transplant-free survival for patients with left heart hypoplasia reaching adulthood and their late post Fontan morbidity.
Methods: Retrospective observational study of patients with HLHS and variants born between 1995 and 2002, who underwent initial Norwood procedure at our institution, was conducted.
Results: The Norwood procedure was performed in 55 neonates with postoperative mortality of 27%. Total pre-Fontan mortality was 38% and 1 patient underwent transplantation after bidirectional Glenn. There was one early and six late deaths (13%) after fenestrated Fontan procedure during median follow-up of 14,9 years (2,0-19,9 years). Transplant-free survival to 18 years was 44% (n=24). There was trend for higher mortality in patients after fenestration closure (p=0,078). Freedom from reintervention at 5, 10 and 15 years after Fontan completion was 45%, 19% and 6%, respectively. Late morbidity after Fontan procedure was significant for liver cirrhosis (19%), symptomatic dysrhythmias (13%), protein losing enteropathy (10%), right ventricular ejection fraction <40% (10%), symptomatic thromboembolism (10%), hemoptysis (10%) and plastic bronchitis (6%). New York Heart Association functional class of survivors at last check up was I in 21%, II in 67% and III in 12%. Ten patients (42% of adult survivors) were free of any major adverse clinical event.
Conclusions: In one of the first single center cohorts of patients after Norwood procedure reaching adulthood with a Fontan circulation the survival was 44%. Almost all survivors underwent cardiovascular reintervention and more than half of them suffered from significant adverse cardiovascular event.



P95

Is it a mass, is it an aneurysm, it is a diverticulum!

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Introduction: Congenital diverticulae of the aortic sinus are extremely rare. They are often latent if they remain un-ruptured. Consequently, they are typically only found incidentally by transthoracic echocardiography (TTE). The most common differential diagnosis is aortic-sinus aneurysm. If ruptured, both equally manifest acutely as a surgical emergency. We present a case of varying differential diagnoses given by multiple imaging modalities and highlight how these guided the changing management plan of the patient.
Methods: A hypertensive asymptomatic forty-six-year-old male was referred for an TTE for an incidental murmur. The TTE revealed a mobile cavity-like structure in the LVOT that was suggestive of prolapsing right coronary cusp (RCC) aneurysm with no LVOT obstruction, a competent tri-leaflet aortic-valve with good ventricular size and function. Additional diagnostic imaging via trans-oesophageal echocardiography further detailed this large aneurysmal cavity (23mmx22mm) attached to the RCC, that prolapsed into the LVOT in diastole and collapsed in systole. A CT scan offered a differential diagnosis of a large LVOT cystic mass. On these grounds, the aortic team accepted for surgical correction. But, on further discussions at the adult congenital heart disease cardiac surgical meeting, uncertainties were raised regarding the differential diagnosis and its functional significance.
Results: The patient underwent a cardiac MRI scan, which reported a diverticulum attached to the RCC, extending into the LVOT (13mmx23mm) with a narrow connection to the RCC of 2mm. Tissue characterisation confirmed this was not a cystic mass and signal characteristics were similar to that of blood flow, perfusing similarly to the aortic sinus, in keeping with diverticulum pathology. A stress TTE demonstrated no significant LVOT obstruction, with no electrophysiological abnormalities or symptomology. On re-discussion it was agreed that regardless of the exact pathology, conservative management with regular follow-up would be most appropriate.
Conclusions: This case highlights the importance of multimodality imaging and functional testing in delineating a rare congenital anomaly and permitting detailed quantification for optimal patient management, avoiding surgery in an otherwise well and asymptomatic individual. It is also appreciated that the use of aneurysm and diverticulum are close by definition and thus have likely been used as synonyms for describing this unique pathology.

P96

Late outcome of Tetralogy of Fallot with or without pulmonary atresia repaired without using a right ventricle to pulmonary artery conduct

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Introduction: To describe the outcomes in infancy after repair with right ventricle to pulmonary artery connection technique

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(RVPAc) in tetralogy of Fallot with pulmonary stenosis (TOF-PS) or with pulmonary atresia (TOF-PA) without MAPCAs.
Methods: We retrospectively reviewed patients with TOF-PS and TOF-PA without MAPCAs who underwent complete repair using RVPAc between 1995 and 2016 in our center. Median follow-up was 10.0 [CI95%:6.4-14.1] years after repair.
Results: Four hundred and ninety-four patients were included into two main anatomical groups: TOF-PS (n=412, 83.4%), and patients with TOF-PA (n=82, 16.6%). The median weight at repair was 6.2kg (5.30kg-8.30kg). Overall mortality was 8 (1.6%), with 4 early deaths (0.8 %). Survival at 10 years after repair was 99% in TOF-PS and 94% in TOF-PA (p=0.12). Weight at the repair was a risk factor of mortality (HR 0.4 $IC_{95\%}$ [0.25-0.98], p=0.04). Ninety-three patients (18.8 %) underwent reintervention (surgical or percutaneous). Freedom from reintervention at 10 years after repair was 87.9%, for TOF-PS, and 65.1% for TOF-PA (p=0.002). In multivariate analysis, the absence of main pulmonary artery (HR 0.4 $IC_{95\%}$ [0.2-0.8], p=0.009), prior palliative surgery (HR 2.2 $IC_{95\%}$ [1.4-3.5], p<0.001), and pulmonary artery stenosis (HR 1.9 $IC_{95\%}$ [1.2 -3.0] p=0.007) were reintervention risk factors. However, Nakata index before the complete repair was not a risk factor of reintervention(p=0.7) or mortality (p=0.68)
Conclusions: Surgical repair of TOF remains a low risk procedure with low reintervention in infancy. Prior palliative surgery influences the risk of reintervention after the complete repair. Prior palliative surgery should be then avoided as much as possible.

P97

Less is more in ccTGA?

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Introduction: Congenitally Corrected Transposition of the Great Arteries (ccTGA) is a rare congenital condition in which there is both atrio-ventricular and ventriculo-arterial discordance. Associated cardiac lesions include atrial septal defects (ASD), ventricular septal defects (VSD) and pulmonary stenosis (PS).
Methods: This case exhibits the unique clinical management of a fifty-four year-old male with unrepaired ccTGA, VSD and PS since birth. It highlights the advancement of cardiac medicine and technology throughout his care and how this has impacted his changing diagnosis and clinical management. We also highlight a change in the new 2020 ESC ACHD guidelines for this patient cohort.
Results: This patient presented at three weeks, with differential cyanosis and a heart murmur. Based on clinical signs alone, he was given a provisional diagnosis of TGA with a Patent Ductus Arteriosus. No further follow-up was arranged. A cardiac catheter at the age of six re-diagnosed him to ccTGA with irreversible pulmonary arterial hypertension. He was deemed unsuitable for surgical repair/palliation. His first transthoracic echocardiogram was performed at fourteen, which further re-diagnosed him to ccTGA, VSD and PS anatomy. Although cyanosis and finger clubbing were revealed, the patient remained well. Into adulthood, he developed secondary erythrocytosis that was managed by regular venesection, until this was queried by his cardiologist. Now in his 6^{th} decade, he remains active with no symptoms. His latest echocardiogram revealed good ventricular function, an anterior mal-aligned VSD, leading to severe pulmonary outflow obstruction (Vmax – 4.5m/sec), systemic RVH, and importantly minimal systemic tricuspid valve regurgitation. This is due to his physiology reflecting loading conditions of a surgical PA band, which is still commonly used to favorably shift the septum contralaterally to reduce TV annular size and thus regurgitation severity, whilst also protecting pulmonary vasculature from systemic hemodynamics in the presence of VSDs.
Conclusions: Correct diagnosis was likely delayed due to presentation predating availability of a transthoracic echocardiogram. Latest ESC ACHD guidelines no longer recommend anatomical double-switch repairs. This case displays that in some ccTGA patients, "less is more", and supports the notion of moving away from anatomically repairing all ccTGA anatomies as this unrepaired patient remains symptom free into adult-life.

P98

Medical non-cardiac critically ill patients with congenital heart anomalies in intensive care unit at a second level hospital: Report of 2 cases

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Introduction: Most of congenital heart patients reach adulthood and nowadays non-cardiac causes of death outnumbered cardiac ones. In critical situations, initial stabilization is mandatory but anatomy and/or physiology must be taken into account in these cases. Clinicians in second level hospitals think they will never attend these patients and nothing can be done without advanced therapies, as extracorporeal membrane oxygenator (ECMO).
Methods: We report 2 cases admitted in our intensive care unit (ICU) due to non-surgical non-cardiac cause during 16 months. Diagnosis at admission, cardiac diagnosis, treatment, and evolution were described.
Results: Case 1: 35-year-old woman admitted due to bilateral pneumococcal and influenza A pneumonia with adult respiratory distress syndrome. She had tricuspid and right ventricle hypoplasia, restrictive ventricular septal defect and pulmonary branches stenosis palliated with Fontan operation and multiple surgical and percutaneous interventions on sildenafil 60 mg per day. She was in failing-Fontan stage (chronic desaturation, chronic atrial fibrillation, protein losing enteropathy with some episodes of congestion and hypoalbuminemia) but she had refused any study or interventions. There were no other ICU beds available but our institution, so she was admitted at our hospital. She was intubated with the least Positive end-expiratory pressure (PEEP) that permitted oxygenation similar to patient´s basal one (5 mmHg) and was on dobutamine, norepinephrine, terlipressin, albumin and furosemide in perfusion during 4 days, when she could be transferred to an ECMO and congenital heart centre. She was discharge after 3 months (21 days on veno-venous ECMO). Case 2: 58-year-old man admitted due to intermediate-high risk pulmonary embolism with desaturation and cardiac biomarkers augmented. He had essential thrombocythemia and situs inversus totalis (dextroapex), so echocardiographic evaluation of right ventricle was more difficult than usual. He was on low-molecular weight heparin. He was discharge home uneventfully after 10 days.
Conclusions: Although rare, general intensivists of non-tertiary hospital will probably have to attend these patients in the future, even the more complicated cases. Because of differences from other

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cardiopathies, prompt consultation and multidisciplinary approach will be necessary. Despite the lack of advanced therapies, it is possible to stabilize and then transfer these patients, allowing them to survive

P99

Pregnancy outcomes among 14 patients with tetralogy of Fallot

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Introduction: Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease and shows an excellent long-term survival after correction. The outcome of pregnancy for women with TOF has not been well studied to date.
Methods: This was a retrospective study. 14 women with corrected TOF and a history of pregnancy who were referred to our Centre for Congenital Heart Disease between December 2005 and December 2020 were included. Information on each completed pregnancy was obtained using medical records. Obstetric and cardiovascular complications and offspring events were investigated. Cardiovascular Magnetic Resonance Imaging (MRI) examinations for the assessment of biventricular function and volumes and valve competence before and after pregnancy were compared where available.
Results: A total of 14 pregnant women with operated TOF were identified. No obstetric complications were observed. Spontaneous fetal loss was documented in one patient. Cardiovascular complications were observed in two patients (14%), who presented with palpitations. An arrhythmia could be documented only in one of these patients, consistent with atrial tachycardia requiring multiple electrical cardioversions and non-responsive to a cavo-tricuspid isthmus ablation. 6 patients underwent cardiovascular MRI before and after the pregnancy. Significant worsening of biventricular function and dilatation, mainly affecting the right ventricle with exacerbation of pulmonary valve dysfunction after pregnancy was detected only in one patient (17%), without associated symptoms. In one woman MRI showed worsening of left ventricular function after pregnancy. This examination was performed during atrial tachyarrhythmia and a further assessment after electrical cardioversion was not possible due to claustrophobia. No significant change was observed in the remaining 4 patients.
Conclusions: Corrected TOF appears to be associated with good maternal outcomes in our cohort of patients. MRI is an useful tool in the follow-up of this setting of patients, as it provides accurate and reproducible measures of biventricular volumes and function and allows quantification of pulmonary regurgitation. Our population was relatively small. Larger studies with a systematic clinical and imaging assessment - approach are required in order to adequately identify prognostic predictors in this cohortof patients.

P100

Prevalence of obstructive and restrictive lung pattern in adults with congenital heart disease

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Introduction: Pulmonary function testing became of more and more interest, also in patients with congenital heart diseases (CHDs). The parameters collected are forced vital capacity (FVC), forced expiratory volume within the first second (FEV1) and its ratio. But residual volume, tested via bodyplethysmography, and total lung capacity (TLC, calculated) are more precise in detecting possible obstruction in patients. This study gives first impressions in the results of a large-scale study that investigates CHD patients' lung volumes via spirometry and bodyplethysmography.
Methods: From April 2018 to October 2019 a total of 81 adults with various CHDs (27.2 ± 7.9 years, 45.7% female) underwent spirometry and bodyplethysmography. Current reference data (Global Lung Initiative 2012 for lung function and Stocks et al., 1995 for TLC) was used to identify possible restrictions (z-scores < -1.64 in FEV1/FVC, FVC, and lower limit of normal in TLC).
Results: Of all patients, 58% had normal lung values. A sole restriction was seen in 14 patients (17.3%), a sole obstruction in 16 (19.8%) and both in four (4.9%). Nine of the 16 patients with obstruction had a normal z-score in FEV1/FVC, only bodyplethysmographie identified their obstruction. All nine patients had a restrictive pattern in FVC.
Conclusions: Almost half of the tested cohort of adult CHD patients have a remarkable lung function. Screening with spirometry is urgently needed. Bodyplethismography is compulsory to classify obstruction and restriction in an accurate manner.

P101

Serial cardiopulmonary exercise testing in adult congenital heart disease patients. The rise and fall of peak predicted VO2 max.

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Introduction: Serial Cardio-Pulmonary Exercise Testing (CPET) is an informative tool in patients with Adult Congenital Heart Disease (ACHD). In patients who require intervention, it is often perceived that this will make people 'feel better', but there is very little evidence for improvement in peak predicted VO2 max. Here we look at the changes in serial peak predicted VO2 max performed as part of routine clinical care.
Methods: Patients who had undergone serial CPET testing under the North West Congenital Heart Disease Service were identified via a database search. CPET reports and clinical notes were reviewed for patient demographic details and CPET results.
Results: 119 patients were retrospectively identified as having serial CPET tests; 2 of these had missing data. 117 patients were included in the final analysis. Patient age ranged between 21-75 years (median 31 years). 55 (48%) patients were male. There was a wide mix of pathology, the most common being tetralogy of Fallot (34), pulmonary valve disease (15), aortic valve disease/outflow obstruction 13, transposition of the great arteries (17) and atrioventricular septal defect (9). There were 7 patients with a palliative Fontan circulation. There were low numbers of patients (13%) achieving a percentage of predicted peak VO2 >85%. 17% of patients who had a serial CPET test had a decline of >10% in their percentage predicted peak VO2. 47% of patients who had intervention experienced a decline in percentage predicted peak VO2.

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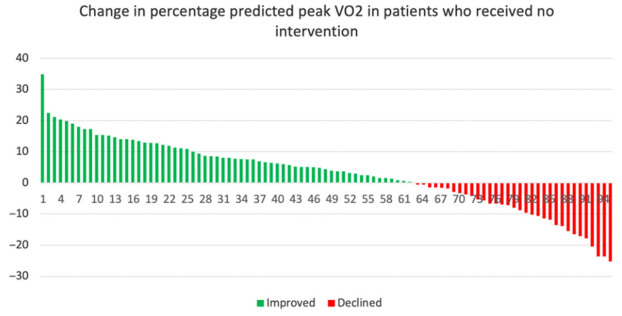
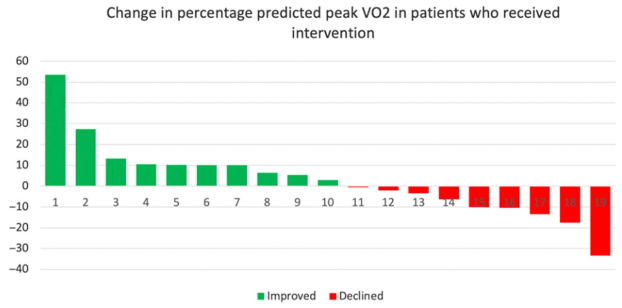
The median duration between tests was 1492 days in those who had an improvement in peak VO2 and 1308 days in those who had a decline. The median time between intervention and the second CPET was 759 days.
Conclusions: A considerable proportion of patients have a large decline in peak predicted VO2 max between tests, regardless of whether intervention is performed or not. It may be beneficial to perform CPET soon after complete recovery from intervention to get a better understanding of how exercise capacity recovers from this, as it may provide important information for the consent process.





P102
Severe Candida tropicalis prosthetic valve endocarditis and life-threatening pulmonary hemorrhage rescued by ECMO of a patient with repaired dTGA

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Introduction: Candida infective endocarditis is a severe and rare complication in patients with congenital heart disease with very high hospital mortality rates (>50%). Survival of a candida endocarditis and massive intrapulmonary bleeding treated by support of extracorporeal membrane oxygenation has so far not been reported.
Methods: Case report
Results: A 27 year old woman with repaired dTGA pulmonal stenosis and Rastelli operation who developed a severe Candida tropicalis prosthetic endocarditis of her pulmonary conduit one year after implantation. Intraoperatively she developed massive intrapulmonary bleeding which could not be stopped. As a rescue therapy she was placed on arterio– venous ECMO with blocked and disconnected endotracheal tube to induce a pulmonary clotting. Two days after surgery a first bronchoscopy was performed to remove the clots and pressure support ventilation was consecutively started with extracorporeal membrane oxygenation support over a period of 20 days. The patient has been discharged from hospital and continues on long-term fluconazole therapy for candida endocarditis prophylaxis.
Conclusions: Candida prosthetic endocarditis and massive intrapulmonary bleeding is not necessarily a contradindication for extracorporeal membrane oxygenation support. With antifungal therapy and surgical replacement survival can be ensured until lung and heart function adequately recover allowing decannulation, removal of all catheters and eventually negative Candida antigen titers. Extracorporeal membrane oxygenation (ECMO) is applied in patients with acute refractory respiratory or cardiac failure that is deemed reversible. Bleeding is the most frequent complication during ECMO support. Severe pre-existing bleeding has been considered a contraindication for ECMO application. Nevertheless, there are cases of successful ECMO application in patients with multiple trauma and hemorrhagic shock. We successfully applied ECMO in a 27-year-old woman with acute life-threatening pulmonary hemorrhage and severe Candida tropicalis prosthetic endocarditis of a pulmonary conduit.

P103
Using machine learning algorithms to predict exercise capacity in patients with congenital heart disease – future or fallacy?

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Introduction: We encounter algorithms of machine learning not only in our everyday lives, they have also made their way into the medical field. In patients with congenital heart disease (CHD) exercise capacity an important predictor of morbidity and mortality. This study aims if machine learning algorithms from simple baseline parameters, instead of performing a cardiopulmonary exercise test (CPET), can predict this parameter satisfactory.
Methods: From 5653 CPETs performed during the last ten years we abstracted, year of birth, age, sex, CHD diagnosis, forced vital capacity, forced expiratory volume in the first second, presence of pacemaker, use of betablockers, and presence of surgery as well as peak oxygen uptake. Peak oxygen uptake was categorized as normal (>80%predicted reference values), moderate impaired (80-60%predicted) and severely impaired (<60%predicted). Afterwards a random forest classifier, based on machine learning algorithms learned from 500 classification trees, was performed to predict categories of peak oxygen uptake. Combining analysis with repeated 5-fold cross validation modelto optimize algorithm of prediction. The performance of random forest classifier was evaluated by the out of bag error rate.
Results: The overall prediction accuracy for random forest classier model is 62.4%, based on out-of-bag error. The prediction accuracy for peak oxygen uptake of normal, moderate impaired, and severely impaired category was 78.5%, 46.7%, 49.8%, respectively. The feature importance plot (Figure 1) showed that CHD diagnosis had highest value in mean decrease accuracy, which means exclusion of CHD diagnosis brings biggest impact on decreasing accuracy of prediction. Gender, forced expiratory volume in the

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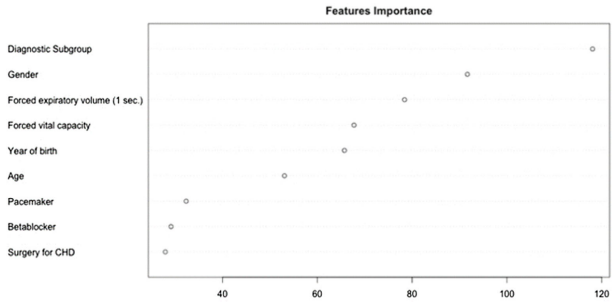
first second, forced vital capacity and year of birth followed in their importance for prediction accuracy.
Conclusions: Patients with CHD that had normal exercise capacity could be determined with a relatively good accuracy. This has an economic benefit, as unnecessary CPETs on patients can be avoided or postponed. In case of doubt, however, a CPET should still be performed.



P104

'But I thought I had a Bicuspid Aortic valve?' Challenges in imaging the elusive Unicuspid Aortic Valve.

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Introduction: Unicuspid aortic valve (UAV) is a rare congenital anomaly present in 0.02% of adults. This may be anatomically described as unicommissural or acommissural depending on the presence of lateral attachments to the aorta. Clinically, this is a significant distinction, as haemodynamically significant stenosis often presents later in the unicommissural form. Distinguishing a UAV from a bicuspid aortic valve (BAV) on transthoracic echocardiography (TTE) can be very difficult. We present the case of a young man under surveillance for asymptomatic BAV whose anatomy was subsequently demonstrated to be that of unicommissural UAV.
Methods: A 21-year old Caucasian male presented to the adult congenital clinic with progressive exertional dyspnoea, non-anginal chest pain and increasing frequency of palpitations, in the absence of significant pre-syncope or syncope. Follow-up TTE was synonymous with previous paediatric surveillance; demonstrating a dysplastic valve that appeared bicuspid with adequate fused left /non-coronary cusp exertion and a fixed right coronary cusp. There was progressive disease with moderate stenosis and severe regurgitation. Subsequent cross-sectional imaging demonstrated a UAV, no significant aortopathy and normal coronary arrangement. Aortic valve replacement was undertaken on prognostic grounds. Intra-operative trans-oesophageal echocardiography demonstrated a functionally unicommissural UAV, which was confirmed on visual inspection. A mechanical prosthesis was implanted. The patient recovered well and was discharged at day five with no electrophysiological or haemodynamic complications. At 4-week follow up he remained symptom-free with a competent prosthesis demonstrated echocardiographically.
Results: Although TTE criteria exist to diagnose and heighten the index of suspicion of a UAV, imaging limitations in the adult patient and planar complexities of the UAV still make this a difficult diagnosis. Lack of symptomatology whilst under paediatric management may explain why cross-sectional imaging was unprovoked, and hence the exact morphology remained unknown.
Conclusions: This case highlights the accelerated valve dysfunction seen in patients with UAV. Recognition of the UAV is important in order to guide the most appropriate timing of follow-up and to better inform our patients. Due to the difficulty diagnosing UAV on TTE there should be a low threshold for considering cross-sectional imaging when there is diagnostic uncertainty or where there is more rapid valve dysfunction than expected.

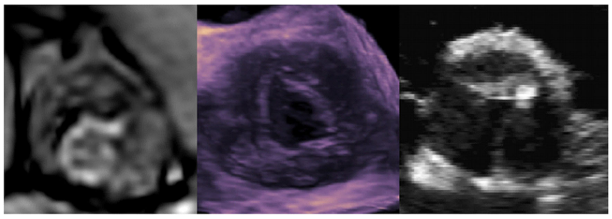

Figure 1. Functionally unicommissural unicuspid aortic valve with RCC/NCC and RCC/LCC raphe. Left panel: CT with RCA ostium seen anterioly. Middle panel: 3D TOE reconstruction. Right panel: TTE PSAX.

6. Imaging/functional assessment

Ventricular speckle tracking echocardiography values in children after pediatric cardiac surgery

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Introduction: Speckle tracking echocardiography (STE) ϵ analysis is gaining consensus in children, but its use in pediatric cardiac surgery has been poorly investigated so far. We aimed to evaluate the STE ϵ response after pediatric cardiac surgery.
Methods: We prospectively enrolled 101 children (mean 2.03±3.42 years; range 0-15.16 years) undergoing cardiac surgery from May 2016 to May 2019 at a Single Institution. Echocardiographic examinations were performed at 4 different post-operative intervals: Time-1: 24-36h (n=90), Time-2: 3-5 days (n=66), Time-3: 6-9 days (n=49), and Time-4: at discharge (mean of 16.5±7.4 days after surgery, range 9.4-47.79 days; n=50). The following parameters have been evaluated: longitudinal left ventricular (LV) ϵ (in 4, 2 and 3 chamber view, and LV global longitudinal -LVGL-ϵ), and right ventricular global longitudinal strain (RVGLϵ). 792 age-matched normal subjects (mean: 7.57±10.63 years) were used as controls.
Results: All 255 examinations were performed at different post-operative times. At each sample time, all STE evaluated parameters were significantly lower than in controls (p all <0.0001). As expected, the lowest ϵ values were found at Time-1, with a significant trend to recover thereafter ($p < 0.002$ to 0.01). However, values at discharged were all still lower than in normal

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subjects (p <0.0001 for all the ϵ values analyzed). When strain values were analyzed as percentiles, at Time-1 subjects with a percentile < 5th for LVGLϵ and RVGLϵ, respectively were 58.4% and 40%. At Time-4 38.7% still had LVGL values <5th percentile. LV ejection fraction (LVEF) had strong correlation with all LV strain values (P=0.0001) and with GLRV (p=0.008). LVEF had trends similar to strain values, but a more rapid recover (e.g. at Time-3 in no cases LVEF was <45%).
Conclusions: All STE ventricular ϵ parameters decreased after pediatric cardiac surgery. A recover during days has been observed, however ϵ values at discharged were significantly lower than in normal subjects. The recovery of ϵ values is slower compared to LVEF, thus ϵ values may indicate also subclinical damages not detectable with conventional markers.

P105
2D speckle tracking echocardiography reference values for left and right ventricular global longitudinal strain in healthy young adults

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Introduction: Two-dimensional (2D) speckle tracking echocardiography (STE) serves as a diagnosis tool for myocardial deformation. We studied the distribution of global longitudinal strain (GLS) of the left (LV) and right ventricle (RV) to establish reference values for healthy young adults. This facilitates the further implementation of STE in clinical practices.
Methods: We analyzed echocardiograms of 90 sports or health science students without any pathologies (20.6±2.5 years of age, range 18-32, BMI of 21.69±2.29, 72% females) in EchoPAC. Using a 6-segment model (free wall and septum approach), we assessed the right ventricular global longitudinal strain (RVGLS) in apical 4-chamber view. Additionally, also using a 6-segment model, we assessed the left ventricular global longitudinal strain (LVGLS) in apical 4-chamber (LVGLS-4C), 2- chamber (LVGLS-2C) as well as in apical long-axis view (LVGLS-LA). On average, 73% (66 out of 90 participants) of all images per view had sufficient quality to conduct STE as echocardiograms with more than one erroneous tracking segment in EchoPAC had to be excluded.
Results: The following average GLS values were measured: RVGLS (n=57, -23.55±2.30%), LVGLS-4C (n=87, - 19.60 ±2.14%), LVGLS-2C (n=71, -20.95±2.54%), LVGLS-LA (n=48, -20.69±2.46%). To evaluate differences between males and females, we employed Student's t-test. For RVGLS, females (n=42, -24.11±2.20%) exhibited significantly higher absolute values than males (n=15, -21.99±1.87%, p=.002). Likewise, females' absolute LVGLS-4C (n=63, -20.07±2.08%) was significantly higher than males' (n=24, -18.37±1.82%, p=.001). Absolute LVGLS-2C was also significantly higher in females (n=53, -21.49±2.38% vs. n=18, -19.36±2.41%, p=.002). However, females' LVGLS-LA (n=38, - 20.98±2.39%) versus males' (n=10, -19.61±2.56%, p=.119) was not significantly different. LVGLS-4C and LVGLS-2C correlate significantly (r=.56, p<.001) with a mean bias of -1.54±2.22 (Bland- Altman), limits of agreement were -5.89 to 2.82, with no proportional bias in linear regression (p=.088). LVGLS-4C and LVGLS-LA measurements also correlate significantly (r=.58, p<.001) with a mean bias of -0.84±2.09, limits of agreement were -4.95 to 3.27, with no proportional bias (p=.102).
Conclusions: We provide reference values for both the left and right ventricle of young adults. This can support the detection of subclinical changes in myocardial deformation during clinical assessment and routine follow- ups. Further research should substantiate STE's effectiveness in the early prevention of cardiopulmonary pathologies.

Global longitudinal strain by view, gender and age groups in years

	Total							18-21				22 and older			
	Both gender			Male		Female		Male		Female		Male		Female	
View	Mean±2SD	Min	Max	n	Mean±2SD	n	Mean±2SD	n	Mean±2SD	n	Mean±2SD	n	Mean±2SD	n	Mean±2SD	n
RVGLS	-23.6±4.6	-29.0	-18.8	(57)	-22.0±3.7	(15)	-24.1±4.4	(42)	-22.2 ±3.7	(11)	-24.1±4.4	(36)	-21.5±4.1	(4)	-24.2±4.8	(6)
LVGLS-4C	-19.6±4.3	-25.6	-14.1	(87)	-18.4±3.6	(24)	-20.1±4.2	(63)	-18.4±3.9	(18)	-20.2±4.1	(51)	-18.2±3.0	(6)	-19.4±4.2	(12)
LVGLS-2C	-21.0±5.1	-27.0	-15.0	(71)	-19.4±4.8	(18)	-21.5±4.8	(53)	-19.7±4.8	(14)	-21.6±4.7	(42)	-18.3±4.8	(4)	-21.1±4.9	(11)
LVGLS-LA	-20.7±4.9	-27.6	-15.9	(48)	-19.6±5.1	(10)	-21.0±4.8	(38)	-19.3±5.3	(8)	-21.0±5.0	(33)	-20.9	(2)	-20.8±3.6	(5)

Global longitudinal strain (GLS) reported in %; SD = standard deviation; n = number of volunteers; RVGLS = right ventricular GLS in apical 4-chamber view; LVGLS-4C = left ventricular GLS in apical 4-chamber view; LVGLS-2C = left ventricular GLS in apical 2-chamber view; LVGLS-LA = left ventricular GLS in apical long-axis view.

P106
3 dimensional echocardiographic evaluation of right ventricular function in pediatric sickle cell disease population

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Introduction: Sickle cell disease (SCD) is characterized by chronic hemolytic anemia and intermittent vaso-occlusive events associated with cardiac abnormalities. The aim of the study was to assess 3 dimensional (3D) echocardiographic of right ventricle (RV) volumes and function in a pediatric SCD population.
Methods: Eighteen patients with SCD aged 4 to 17 years old (mean age: 8.0 ± 4 years, 56% male, body surface area (BSA) 1.0 ± 0.35) and 18 healthy controls matched for age, gender and BSA were prospectively included and compared. Echocardiograms were performed using a commercially available ultrasound Philipps EPIQ 9 system using matrix X5-1 transducer. 3D indexed RV volumes and ejection fraction (3D-RVEF) were obtained using full volume acquisitions. RV free wall strain, tricuspid S-wave, tricuspid annular plane systolic excursion (TAPSE), indexed cardiac output, systolic pulmonary pressure (sPAP) and hemoglobin were assessed. Data were analyzed with TomtecArena© software (v2.3, Germany).
Results: Cardiac output was significantly higher in SCD children (4.5 vs 3.6 l/min/m², p=0.025), as sPAP (24.9 vs 21.9 mmHg, p=0.015), 3D-RV diastolic volume (58.1 vs 47.5 ml/m², p=0.025) and 3D-RV systolic volume (28.8 vs 21.4 ml/m², p=0.005). 3D-RVEF and RV free wall strain were significantly lower in SCD compared to control population (respectively 51.9 vs 56.3 %, p=0.018; -28.6 vs -32, p=0.017). There were no difference regarding TAPSE and doppler S-wave. Mean hemoglobin in SCD population was 9.6 ± 1.7 g/dl.
Conclusions: Despite normal RV systolic function parameters, 3D-RVEF and RV free wall strain are lower in children with SCD. Chronic anemia generating volume overload and vaso-occlusive events could explain these findings.

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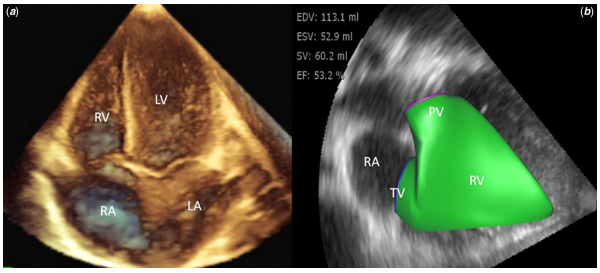

P107

A case report of Leigh syndrome diagnosed by endomyocardial biopsy

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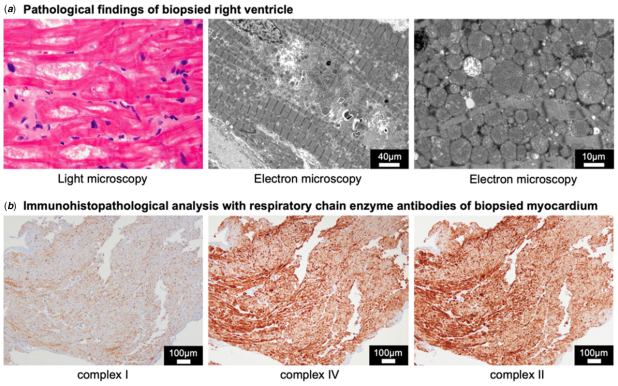
Introduction: Leigh syndrome is a neurodegenerative disorder caused by mitochondrial dysfunction with both phenotypic and genetic heterogeneity. Mitochondrial impairments are usually demonstrated by skeletal muscle biopsy. We report a case of Leigh syndrome diagnosed by endomyocardial biopsy (EMB), not by skeletal muscle biopsy.

Methods: A 17-year-old boy was admitted to our hospital to investigate the cause of hypertrophic cardiomyopathy. Delayed motor development appeared in 7 months. He developed metabolic acidosis triggered by an infection with elevated lactate and pyruvate values in serum and cerebrospinal fluid when he was 1 year old. T2-weighted imaging revealed bilateral hyperintensity in midbrain and dorsal pons. Biopsied skeletal muscle did not show evidence of mitochondrial disease. Hypertrophic cardiomyopathy, bilateral putamen hyperintensity in T2-weighted imaging and a lactate peak in the right basal ganglia in single voxel spectroscopy, and a convulsive seizure appeared at the age of 12, 15, and 16, respectively.

Results: EMB was performed by cardiac catheterization and biopsy samples were obtained at the ventricular septum in the right ventricle. Light microscopy of the biopsied myocardium showed cytoplasmic vacuolization and electron microscopy showed a marked proliferation of mitochondria within myofibrils. Respiratory chain enzyme activity of the biopsied myocardium and cultured fibroblasts obtained from the biopsied skin showed decreased activity of complex I. Immunohistopathological analysis with respiratory chain enzyme antibodies of biopsied myocardium showed decreased signal intensity of complex I compared with complex II (22% of normal) and normal signal intensity of complex IV compared with complex II (116% of normal). Genetic testing revealed an m.14453 A > G mutation on the MT-ND6 gene, with a heteroplasmy rate of the blood sample 60% and the cardiac tissue 83%. He was finally diagnosed with Leigh syndrome and mitochondrial cardiomyopathy. Administration of 5-aminolevulinic acid was initiated, which reduced the frequency of seizures.

Conclusions: EMB led to the diagnosis of Leigh syndrome. Efforts to find and conduct the biopsy of affected organs are important to diagnose mitochondrial disease. EMB is a useful diagnostic method when there is a difficulty in diagnosing mitochondrial disease by skeletal muscle biopsy.



P108

A cervical arch presenting as a pulsatile neck mass

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Introduction: An 11 year-old boy was referred to our paediatric cardiology service to investigate a large pulsatile mass in his right neck. He was asymptomatic with no breathing or swallowing difficulties. He had a known diagnosis of 22q11 deletion. Examination confirmed a large mass with moderate pulsatility in the anterior triangle of the neck with associated bruit. Echocardiogram showed normal intra-cardiac anatomy, bilateral superior venae cavae and a dilated structure in the suprasternal views suggestive of an aneurysm. A cardiac CT was performed to investigate further.

Methods: An awake non-ECG gated, contrast-enhanced computed tomography (CT) scan was performed with a kV of 80 and a reference mAs of 136 on a Siemens Force scanner.

Results: Cardiac CT demonstrated a right aortic arch extending high into the right side of the neck (figure 1). It had an elongated course 4cm superior to the level of the clavicles, with an acute apex before forming the descending aorta. The first branch was the left common carotid artery, the second was the right common carotid artery, the third was the right subclavian artery and the fourth was an aberrant left subclavian artery.

Conclusions: Cervical arches are a very rare cause of a pulsatile neck mass. 1 During embryogenesis, the great vessels develop from the six branchial arches. Arch III normally forms the carotid system and arch IV the aortic arch. It is thought that a cervical arch forms when there is abnormal regression of the fourth arch, leading to the third arch forming the aortic arch. Cervical arches may be asymptomatic and present with a pulsatile mass in the neck. 2 Symptoms usually arise due to compression of adjacent structures, such as the trachea, oesophagus or the brachial plexus, or due to dilatation, coarctation or dissection of the aorta. 3 Despite displaying the conventional

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substrate for a vascular ring, the distorted arch anatomy resulted in a patent airway and oesophagus. Further intervention was not felt to be indicated and he was advised to refrain from contact sports due to the risk of trauma to the aorta.

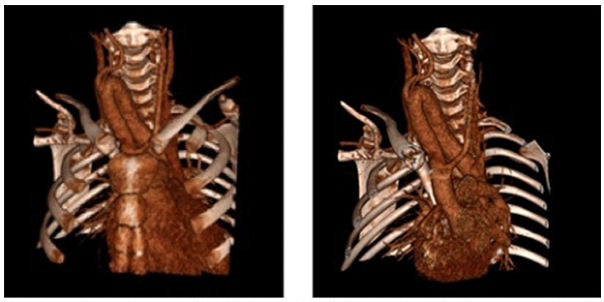

Figure 1. Left panel shows a coronal view of the chest with bony landmark structures in place demonstrating the cervical arch extending high into the right side of the neck. The right panel demonstrates the same view with sternum cut away to show the elongation of the arch structures and the arch branching pattern.

P109

A new model for accurate non-invasive prediction of transpulmonary peak-to-peak gradients

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Introduction: The threshold for intervention in congenital heart disease with right-sided obstructive lesions depends on the haemodynamic measurements obtained during the procedure. Therefore, the accurate non-invasive estimation of the stenosis is critical to avoid unnecessary invasive procedures. However, current non-invasive estimations lack precision. We sought to create a predictive model to fine tune these indications.
Methods: Retrospective review of all consecutive catheterizations for right-sided lesions in a single paediatric cardiology department from 2016-2019. Data was gathered on demographics. Echocardiographic measurements included antegrade and tricuspid regurgitation peak instantaneous and mean estimated gradients, and end-systolic left ventricle eccentricity index (defined as the ratio between the anteriorinferior and septal-posterolateral cavity dimensions at the mid-ventricular level). Invasive measurements were peak-to-peak gradient and right to left ventricle pressure ratio. Statistical correlation and linear regression modelling were done using SPSS 20.
Results: We included 64 patients (56% male). Mean age was 8.3 years (SD 6.7), median weight was 22 (interquartile range 39). The main obstruction was located at the valve in 31 (48.4%) patients, branch pulmonary arteries in 15, right-sided conduit in 11 and supravalvular in 5. Diagnoses were pulmonary valve stenosis or atresia with intact ventricular septum (33 pts), operated tetralogy of Fallot and variants (17 pts), transposition of the great arteries (13 pts) and truncus arteriosus (6 pts). All five echocardiographic measurements had a significant ($p < 0.05$) but poor to moderate correlation (r value between 0.429 and 0.615) with invasive haemodynamic measurements. In the univariate analysis, echocardiography had a low predictive value of the invasive haemodynamics. In the subgroup analysis of the valvular obstruction patients, however, we were able to create a multivariate linear regression model that predicted accurately the peak-to-peak invasive gradient, using all five echocardiographic measurements, ($r2 = 0.974$, $p = 0.014$). Associations in the other subgroups were not as strong.
Conclusions: In our sample, accurate peak-to-peak gradient prediction was possible using a regression model in the valvular obstruction subgroup. A larger prospective study is required to validate the usefulness of our model to plan invasive catheterization in patients with right-sided obstructive lesions.

P110

A single centre review of the cardiac investigational burden of patients undergoing the Fontan Palliation

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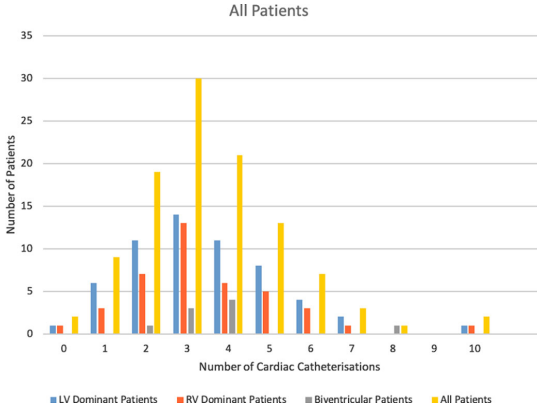
Introduction: Patients undergoing single ventricle palliation who survive to Fontan often require multiple cardiac investigations. We aimed to quantify the burden of cardiac investigations on patients whom underwent the Fontan palliation procedure.
Methods: All patients who underwent the Fontan palliation procedure from November 2004 to December 2012 had their medical record analysed and the number of echocardiograms, cardiac MRIs and cardiac catheterisation procedures recorded.
Results: 133 patients underwent the Fontan palliation procedure in the time period. 26 patients were excluded with incomplete data, thus 107 patients were analysed. 58 patients had morphological left ventricles, 40 had morphological right ventricles and 9 patients had a combination of both morphological ventricles. 1 patient died on post op day 6, from complications related to ECLS. The remaining 106 patients have ongoing follow up with either paediatric or adult congenital cardiology. There were 68 male and 39 female patients. The median age at Fontan was 50 months (range 16 to 198 months). The median weight at Fontan was 16.25kg (range 12.2 to 66.2kg). For LV patients the range for echocardiography was 3 to 46 studies with a median of 21 and for catheterisation the range was 0 to 10 with a median of 3. For RV patients the range for echo was 1 to 52 studies with a median of 27 and for catheterisation the range was 0 to 10 with a median of 3. For biventricular patients the range for echo was 14 to 38 studies with a median of 27 and for catheterisation was 0 to 8 with a median of 4. For all patients, the median number of MRI scans was 1 (range 0 to 4). More MRIs were performed in the later years.
Conclusions: Patients who are treated with the Fontan surgery require an extensive number of investigations. This data is useful to demonstrate the potential resource utilisation of patients in our unit and is useful for patient and parental counselling when advising on the Fontan palliation pathway.



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P111
ACUTE PEDIATRIC MYOCARDITIS BEFORE AND DURING SARS-CoV-2 PANDEMIC

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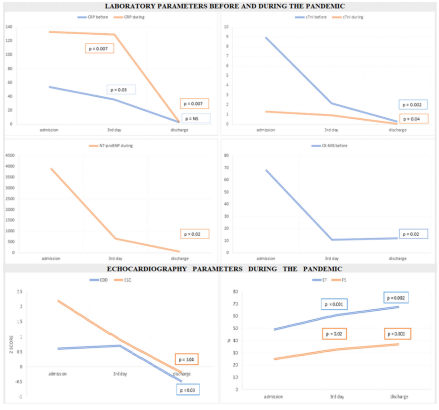
Introduction: Acute myocarditis (AM) is an inflammatory disease of the myocardium. Myocardial injury in COVID-19 could appear as a result of the direct virus attacking, or viral-induced myocardial inflammation as a consequence of the aggressive immune response. The aim of our study is a comparative analysis of the difference between children with AM before and during the SARS-CoV-2 pandemic.
Methods: The retrospective analysis included all patients treated in our Institute with a diagnosis of AM from January 2018 to November 2020.
Results: 24 patients were included in the study; in all patients (7/24) treated from April to November 2020, the infection was caused by SARS-CoV-2 (2 PCR, 5 serological tests of IgM antibodies). All patients with AM during the pandemic were older than 7 years. They were more likely to have abdominal pain (p=0.014), headache (p=0.003), cutaneous rash (p=0.003), conjunctivitis (p=0.003), while fulminant myocarditis was more commonly registered before the pandemic (p=0.04). A multisystem inflammatory syndrome in children associated with COVID-19 was present in 6 patients. Patients with AM in the pandemic had significantly lower values of troponin I (cTnI) (p=0.012), and platelets (p<0.001), but higher values of serum creatinine (p=0.013) and CRP (p=0.04) compared with patients before the pandemic. In the group of patients during the pandemic, a significant CRP reduction (p=0.007) was observed on the day of discharge compared to admission value. In the group of patients before the pandemic, cTnI values were significantly reduced (p=0.002). A significant recovery of systolic function was registered on the third in-hospital day in the group of patients presented during the pandemic (EF p=0.001; FS p=0.019); improvement was not observed in the group before COVID-19 pandemic. Adverse events were observed frequently in patients before the pandemic (p=0.04; 3 died, and 4 dilated cardiomyopathy).
Conclusions: In contrast to patients before the pandemic, in patients with AM during the COVID-19 pandemic, significantly higher values of inflammatory parameters, polymorphic clinical presentation as well as prompt recovery of LV function after applied therapy noticed in patients during SARS-CoV-2 pandemic underlie a possible new spectrum inflammatory disease with consequence viral-related myocardial inflammation with favorable prognosis.



P112
Admission serological markers can be used to predict significant cardiac involvement in children with paediatric multisystem inflammatory syndrome

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Leeds Children's Hospital (1), Leeds Congenital Heart Unit (2), Sheffield Children's Hospital (3)

Introduction: Paediatric multisystem inflammatory syndrome (PIMS) began to present in April 2020 midway through the covid-19 pandemic. Occurring 2-4 weeks after initial covid-19 infection, patients presented with persistent fever, evidence of inflammation and single or multiorgan dysfunction[1]. The Yorkshire and Humber congenital heart disease network is made up of the Leeds congenital heart unit and 18 peripheral hospitals[2]. With limited local paediatric cardiology availability, the vast majority of children presenting with PIMS required transfer to Leeds. This presentation aims to describe the cohort of children that were seen within the network as well as to identify any markers of significant cardiac involvement which could be used to reduce the frequency of unnecessary interhospital transfers.
Methods: This was a retrospective case notes review of all patients treated within the Yorkshire and Humber network with symptoms of PIMS between 1st May and 30th November 2020. Patients were classified as to whether or not they had significant cardiac involvement (defined as at least one of: inotrope requirement, ejection fraction <50%, pericardial effusion, coronary artery changes and significant ECG abnormalities). Cardiac markers were analysed at presentation and throughout the hospital admission including plasma NT pro-BNP, LDH, CRP, d-dimer and troponin. Statistical tests (Fisher's exact test for categorical variables, t-test for continuous variables) were used to identify which factors were indicative of significant cardiac involvement (SCI).
Results: 22 patients met the inclusion criteria (Table 1). 14/22 patients (63.6%) were judged to have SCI. Markers that were found to be indicative of SCI included CRP and plasma NT pro-BNP (Table 2). Furthermore, when using a threshold of 2000ng/L, plasma NT pro-BNP was found to be 71% sensitive and 80% specific for SCI. In addition, when combined with a CRP threshold of 100mg/L, there was a positive predictive value of 85% and negative predictive value of 75%.
Conclusions: PIMS is an important new syndrome affecting paediatric patients across the Yorkshire and Humber region. A significant proportion of the affected patients have cardiac involvement and require management in a specialist centre. Early identification of these patients using serological markers facilitates rapid treatment preventing long term sequelae whilst also reducing unnecessary interhospital transfers.

	n	%
Male	16	72.73
BAME	12	54.55
	Range	Median
Age (years)	0.3-15.5	6.40
Weight (centile)	11-99.6	83.00

Table 1: demographics of the patient cohort presenting within the Yorkshire and Humber Paediatric Cardiology Network with paediatric multi-inflammatory syndrome BAME = black and ethnic minority

	SCI	No SCI	p=
CRP (mg/L)	178	97	0.035
LDH (IU/L)	283	399	0.0267
hsTNI (ng/L)	1172	9	0.28
Plasma NT- pro BNP (ng/L)	9564	1726	0.041
D-dimer (ng/ml)	5317	1633	0.47
25 OH- vitamin D (nmol/L)	32	52	0.15
Ferritin (ug/L)	933	330	0.23

Table 2: mean serological marker levels on admission. CRP and Plasma NT- pro BNP were significantly higher in the patients with significant cardiac involvement than those without. CRP: c-reactive protein; LDH: lactate dehydrogenase; hsTNI: troponin; BNP: brain natriuretic peptide

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P113

Aortic morphology and distensibility influence clinical outcomes after aortic coarctation treatment

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Introduction: Aortic coarctation is a systemic vascular disease that predisposes patients to several comorbidities, even after successful treatment. Our aim was to investigate the role of aortic morphology and distensibility on relevant outcomes in patients with treated aortic coarctation.
Methods: Asymptomatic adolescents and young adults with treated aortic coarctation with no gradient or with borderline gradients (>20 and ≤25mmHg) were prospectively evaluated. Echocardiography was performed at rest and peak exercise. Pulse wave velocity (PWV) was evaluated between the right carotid and right radial arteries. Aortic hypoplasia was calculated as the ratio of narrowest diameter of the aortic arch to aortic diameter at the diaphragm level (AoArch/AoDiaphr) by cardiac MR. Univariate and multivariate linear regression models were used to evaluate the impact of PWV, AoArch/AoDiaphr, and isthmic Doppler gradient at rest (gradr) and peak exercise (gradp), on the following variables: systolic blood pressure (SBP) at rest and peak exercise, ambulatory 24-hour SBP, and indexed left ventricular mass (ILVM).
Results: Our sample comprised 43 patients (60.5% male), with a mean age of 21.2 years (range 12–40), at a mean of 14.6 years (range 0.6–34.0) after aortic coarctation treatment. Office SBP correlated with PWV (β=2.9, p=0.011), AoArch/AoDiaphr (β.=-44.1, p=0.01), gradr (β=0.8, p=0.042), and gradp (β=0.5, p=0.001). Multivariable analysis identified PWV (p=0.006) and gradp (p=0.001) as the strongest determinants of office SBP (adjusted R^2=0.36). PWV was significantly associated with mean nocturnal SBP (β=2.3, p=0.031), but not with mean 24-hour ambulatory SBP (β=1.8, p=0.052). Peak exercise SBP was determined by gradp (β=1.0, p=0.002). Patients in the group with borderline gradient had a higher SBP at rest and with exercise (p=0.001 and p=0.014, respectively). Finally, ILVM was inversely associated with AoArch/AoDiaphr (β=-50.5, p=0.033).
Conclusions: Aortic coarctation outcomes at follow-up are influenced by morphological and functional aortic properties with variable impact. Higher SBP at rest (office visits and night time) is related to a lower distensibility. Higher exercise SBP is dependent of higher isthmic gradients during exercise. The degree of aortic arch hypoplasia is the major determinant of ventricular hypertrophy. These results have implications on the type of routine evaluation.

P114

Assessment of left ventricular function using two-dimensional (2D) strain imaging in patients with beta-thalassemia major.

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Introduction: Iron-mediated cardiomyopathy is the leading cause of death in patients with beta-thalassemia major (BTM). Traditional echocardiography is unable to detect neither the early stages of iron overload cardiomyopathy nor myocardial iron deposition. We aimed to evaluate myocardial function in children with beta-thalassemia major using standard echocardiography technique and strain rate imaging.
Methods: Thirty asymptomatic children with BTM and 30 healthy control subjects were prospectively enrolled. All children underwent standard echocardiography and subsequent offline analysis to assess left ventricular (LV) longitudinal mechanics using speckle-tracking. Systolic (Sm), early diastolic (Em), and late diastolic (Am) (Em/Am) myocardial velocities at basal lateral and septal left ventricular (LV) segments, strain (S), and strain rate (SR) in basal, mid and apical LV, systolic function of right ventricular (RV) were measured in both patients and control group.
Results: Patients and healthy control group had normal standard parameters of systolic, diastolic LV function and systolic RV function. Patients with BTM showed a significant reduction of basal and median LV longitudinal strain from the three and four-chamber apical views and global strain (P<0.001, P =0 .04 and P=0.003) respectively). There was no significant difference in strain from two-chamber view and in LV apical side.
Conclusions: Echocardiography, using strain, could predict clinically silent myocardial dysfunction in the early phases of the disease to optimize therapeutic strategies.

P115

Assessment of phase characteristics of left atrioventricular coupling in infants and children by three-dimensional echocardiography

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Introduction: Pediatric volume and phase characteristics of the left atrioventricular (AV) coupling can give insight into heart function, especially the diastolic function. Noninvasive assessment of those parameters is possible by three-dimensional echocardiography (3DE). We sought to assess phase characteristics of atrioventricular coupling in a large cohort of healthy newborns, infants and children and establish reference values.
Methods: 3DE-data sets of 345 healthy children (0 days-18years) where analyzed by TomTec 4D LV-Analysis software 3.1. Volume-time curves of the left atrium (LA) and left ventricle (LV) as well as their time derivatives were generated. Conduit volume (LV stroke volume – LA emptying volume) and its contribution to LV stroke volume was calculated. Moreover, we investigated LV and LA peak filling and peak ejection rates (LVPFR, LVPER, LAPFR, LAPER) and time to those peaks normalized to R-R-interval (LVPFRt, LVPERt, LAPFRt, LAPERt).
Results: Measuring parameters of AV coupling in infants and children by the time derivatives of volume-time curves assessed by 3DE was feasible. Conduit volume contributed most to LV stroke volume with 69.8% (±6.8% std. deviation) and was not correlated to heart rate. LVPERt and LAPFRt were poorly correlated to heart rate, LVPERt being shorter than LAPFRt. LVPFRt and LAPERt correlated with heart rate with decreasing time until peak filling and peak ejection with decreasing heart rate. LVPFRt and LAPERt were virtually identical.
Conclusions: Conduit volume contributed with almost 70% most to LV stroke volume in infants and children. In adults, studies showed contributions of conduit volume to LV stroke volume of 40-55%, decreasing with age. This shows that pediatric reference values are needed to use conduit volume as a possible marker for diastolic

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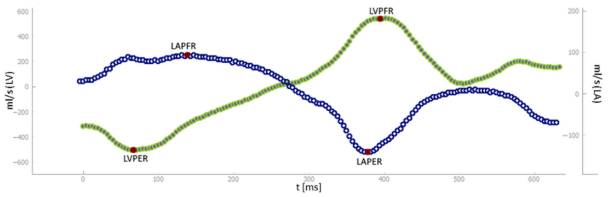
function. Our results show a close coupling between LV and LA during diastole and a prolongation of the diastole after LVPFRt with decreasing heart rate. Our pediatric reference values for phase characteristics of AV coupling obtained by 3DE can provide the basis for further noninvasive evaluation of the diastolic function of the left heart.

Supported by Fördergemeinschaft Deutsche Kinderherzzentren e.V.

Figure 1 Example of time derivatives of volume-time curves of the left ventrivle and left atrium



LV = left ventricle; LA = left atrium; PER = peak ejection rate, PFR = peak filling rate

P116

Association of preoperative mixed venous oxygen saturation with postoperative pulmonary hypertension with TOF/MAPCAs

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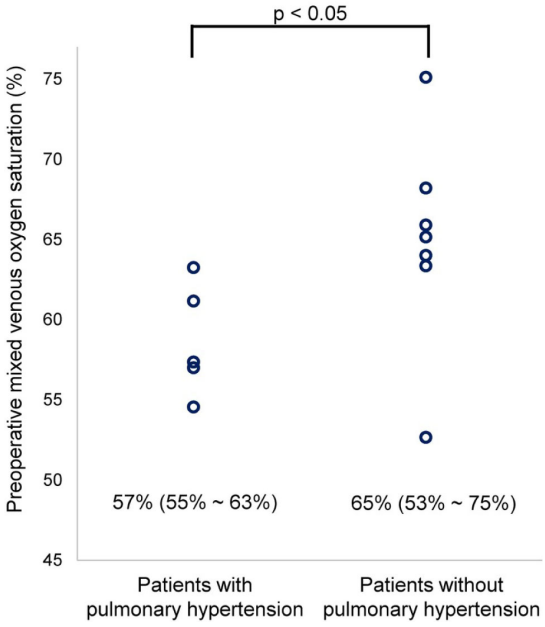
Introduction: In patients with tetralogy of Fallot with major aorto-pulmonary collaterals (TOF/MAPCAs), segmental pulmonary hypertension is not uncommon; however, its pathophysiology remains to be elucidated. This study aimed to identify preoperative hemodynamic factors associated with pulmonary hypertension after intracardiac repair in patients with TOF/MAPCAs.
Methods: This study included 12 patients with TOF/MAPCAs, who underwent unifocalization and intracardiac repair from 2009 at our institution. The hemodynamic measurements of pre-operative and postoperative cardiac catheterization were collected. Five patients with residual peripheral pulmonary stenosis under-went surgical or percutaneous reintervention on the stenosis, and the hemodynamic measurements after the reinterventions were included in this study. Patients were considered to have pul-monary hypertension if the mean pulmonary pressure measured postoperatively at any peripheral pulmonary artery was $\geq$ 20 mmHg. The data were compared between the patients with and without postoperative pulmonary hypertension. A p value of <0.05 was considered statistically significant.
Results: Intracardiac repair was performed at the age of 2.1 years (range, 0.5-28.2 years). Unifocalization was performed separately prior to the intracardiac repair in 2 patients and concomitantly in 10 patients. Postoperative cardiac catheterization, performed 1.2 years (range, 0.2-8.7 years) after intracardiac repair, revealed that 5 patients had pulmonary hypertension. Patients with postopera-tive pulmonary hypertension had significantly lower preoperative mixed venous oxygen saturation than those without pulmonary hypertension (57% [55%-63%] vs. 65% [53%-75%], $p < 0.05$; Figure). No significant differences were found between the groups in terms of age at intracardiac repair, preoperative arterial oxygen saturation (83% [69%-86%] vs 83% [75%-90%], p = 0.53), and preoperative peripheral pulmonary arterial mean pressure (mmHg; right, 20 [11-36] vs. 17 [8-37], p = 0.74: left, 16 [10-18] vs. 18 [8-24], p = 0.57).
Conclusions: Lower preoperative mixed venous oxygen saturation was associated with pulmonary hypertension after intracardiac repair in patients with TOF/MAPCAs. Pulmonary circulation via MAPCAs is usually characterized by high total resistance. Low mixed venous oxygen saturation reflects decreased cardiac output, which further aggravates pulmonary vascular hypoplasia. Low mixed venous oxygen saturation is possibly associated with inadequate size and arborization of pulmonary vasculature and, hence, may predict postoperative pulmonary hypertension.



P117

B-lines lung ultrasound assessment of pulmonary congestion by handheld device in critically-ill children

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Introduction: Pulmonary congestion has impact on morbidity and mortality of critically-ill children. Chest radiograph is traditionally used to assess; however, it was partially subjective and put patient on radiation. B-lines can be imaged by lung ultrasound (LUS) as parallel vertical white lines from pleura. Quantity of B-lines cor-responds to an increase in extravascular lung water or congestion. Our aim is to assess correlation for congestion among B-lines, clinical score, and radiographic score.
Methods: A prospective cohort of children; age 1 month-15 years, who were admitted to PICU of University Hospital with pre-sumed pulmonary congestion from May 2019 to January 2020 were enrolled. Patients with known lung conditions which might interfere B-lines interpretation were excluded. Demographic data and pre-specified clinical parameters, modified Ross score, inten-sivist clinical matrices, and radiographic RALE score were assessed. A 12 MHz linear array handheld ultrasound system; connected to 9.7" Android tablet, was used to image B-lines on 8 regions of both

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anterolateral chest regions. The strategies to manage pulmonary congestion included fluid restriction, diuretic, and inotropic drug. All parameters were determined at pre- and post-interventions.
Results: A total of 38 children; mean age of 4.9 ± 5 years (SD) were included. The majority has cardiovascular diseases. Scanning found mean B-lines 43.8 ±15.6 (SD). After intervention, there was a decrease of mean B-lines to 28.9 ±12.4 (SD) (p value <0.01). The B-lines assessment for pulmonary congestion was well correlated with RALE score (r= 0.59, p value <0.01) and modified RALE score (r=0.61, p value <0.01). Amount of B-lines significantly associated with higher modified Ross score (p value = 0.001) and higher intensivist clinical score (p value = 0.004). B-lines assessment demonstrated good agreement for intraobserver and interobserver intraclass correlation coefficient 0.99 (95% CI 0.98-0.99) and 0.92 (95% CI 0.86-0.95) respectively. B-lines at least 5 at lateral lung zone indicated pulmonary congestion with ROC analysis (81.5% sensitivity and 81.8% specificity, AUC 0.928).
Conclusions: B-lines bedside LUS with handheld device correlated well with pulmonary congestion and congestive heart failure. A reduction of B-lines correlated with clinical improvement after interventions. This technique can be useful in pediatric critical care setting.

P118
Can ECHO Simulation Support Paediatric and Neonatal Specialists to Diagnose Acute Presentations of Congenital Heart Disease? A Systematic Review

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Introduction: Across Europe, recent policy has developed the role of local paediatrician-delivered echocardiography. Thanks to high commitment to learning, agreement of international standards, strong local networks and the EACVI accreditation process, such scans are typically performed with high quality. Nevertheless, the volume of acute and complex cases encountered in local hospitals is naturally lower than within tertiary cardiology units. A training gap is therefore foreseen to support these professionals, both to initially develop their skills in this area and to maintain their skills in the years following accreditation.
Methods: A systematic literature review was performed using the PubMed database, EMBASE and Google Scholar. Three search-term groups were developed, each summarized by the terms 'congenital heart disease', 'echocardiography' and 'simulation'. All peer-reviewed articles were included, without date, language or geographic restrictions. Studies were assessed for relevance i) to congenital echocardiography simulation for paediatricians and neonatologists and ii) to outcome measures assessing diagnostic accuracy. Study quality was further assessed using the CEBM 'Level of Evidence' tool.
Results: The search identified 406 studies, of which 7 met pre-agreed inclusion criteria. Five papers were highly relevant to simulation of congenital heart disease outside of a tertiary setting, whilst two others solely demonstrated content and construct validity. Learners in most studies were classed as 'novices' to congenital echo, limiting inferences to paediatricians who have already reached accreditation-equivalent standard. Four papers assessed clinically-relevant outcome measures before and after simulation training. These documented improvements in factual and applied knowledge, probe kinematics, quality of ECHO images, and diagnostic accuracy of new simulated cases. No paper was sufficiently powered to assess effects upon subsequent clinical practice.
Conclusions: Current evidence supports the validity of ECHO simulators as training and assessment tools for diagnosis of complex congenital heart disease, irrespective of previous learner experience. Potential for use within pre-planned learning sessions is also demonstrated, dramatically reducing the cost of real patient demonstrators and offering rare experience in scanning acute and complex lesions. Although simulation has clear benefits in ECHO performance for novices, the effect on more experienced practitioners is not specifically tested. Further investigation is planned to assess the role of simulation in this group.

P119
Cardiac dyssynchrony in patients with Paediatric Inflammatory Multisystem Syndrome Temporarily associated with SARS-CoV-2 (PIMS-TS)

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Introduction: Paediatric Inflammatory Multisystem Syndrome Temporarily associated with SARS-CoV-2 (PIMS-TS) is commonly associated with cardiovascular compromise. We have previously described the time course and magnitude of left ventricular (LV) systolic dysfunction in children and young adults with PIMS-TS. However, it remains unknown if this inflammatory process can cause LV dyssynchrony. We aim to establish whether paediatric patients with PIMS TS develop LV dyssynchrony as assessed by echocardiography.
Methods: Comprehensive transthoracic echocardiography in 10 PIMS-TS patients was performed during the acute stage of the initial illness when LV systolic function (3D Ejection Fraction (EF)) was worst and then again at six months post PIMS-TS. At both time points, we compared: 3D EF, LV fractional shortening (FS) and global longitudinal strain (GLS). Intraventricular LV dyssynchrony was assessed by M mode, PW tissue Doppler Velocities (TDI), 2-D speckle tracking and 3D echocardiography, while the interventricular dyssynchrony was also assessed by TDI at both time points.
Results: Any improvement in 3D-LV EF at six months post illness (57.8±5.5 %) vs acute phase (51.8± 9.9%) was not statistically significant (p=0.166), whereas the LV FS (29.9± 9.5% vs 36.5± 12.5%, p=0.043) and GLS (-13.8±1.9% vs -18.6±3.1%, p=0.005) were significantly lower during the acute phase of the illness compared to six months later. Regarding dyssynchrony, none of the measures differed at follow up compared with acute phase; the septal to posterior wall motion delay assessed by M-mode (46.1±2.7msec vs 38.6±2.1msec, p=0.417), the basal septal to basal lateral peak velocity delay assessed by TDI (23.2±1.9.msec vs 24±1.9msec, p=0.930), the 2D speckle tracking-derived strain delay index was 1.1±1.2% at the time of the worst LV systolic performance and 0.62±0.26% at 6 months in the recovery period (p=0.219). The 3D echocardiography demonstrated that the 3D systolic dyssynchrony index (SDI) remained similar throughout the follow up period (3.04±1.23% at baseline vs 3.22±1.25% at 6 months, p=0.466).
Conclusions: Despite the fact that in patients with PIMS TS cardiac involvement show a decline on LV systolic performance, this does not appear to be associated with LV dyssynchrony as assessed by echocardiography. We recommend larger patient cohort studies to investigate this further.

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P120

Cardiac function among pediatric cancer survivors exposed to cardiotoxic treatments– LV longitudinal strain should be added to screening methods

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Introduction: In many heart diseases the left ventricular (LV) longitudinal strain detects a decreased cardiac systolic function even with the LV ejection fraction (EF) remaining normal. Majority of the childhood cancer survivors (CCS) are exposed to cardiotoxic treatments and are besides often presented with numerous modifiable cardiovascular risk factors (e.g. obesity, hypertension and dyslipidemia), causing LV hypertrophy with preserved EF. Our aim was to evaluate whether an assessment of the LV longitudinal strain would increase the sensitivity for the detection of cardiac dysfunction after treatment for childhood cancer.
Methods: Clinical and echocardiographic data of 95 long-term CCS and their controls from two previous crosssectional studies were studied. As inclusion criteria, first, all the study patients were exposed to cardiotoxic treatments e.g. anthracyclines and/or high-dose therapy with autologous stem cell rescue and, second, were followed up > 5 years. LV longitudinal strain from the four-chamber view was assessed with a speckle-tracking based method (Qlab). The LV EF and LV systolic mass index (LVMSi) were measured with three-dimensional echocardiography (3DE).
Results: Ninety CCSs and 86 controls with an adequate four-chamber view were included. The mean (±SD) age of the former was 16.0 ± 5.0 years and median (range) follow-up time after cancer treatment 8.1 (5.0 -27.4) years. Study subjects were divided into three groups, S1: CCSs with an abnormal Qlab LV longitudinal strain < -17.5%, (n = 10); S2: CCSs with a normal Qlab LV longitudinal strain > -17.5%, (n = 80), and C3: the controls, (n = 86). 70% in group S1 had normal 3DE LV EF despite the decreased LV longitudinal strain. Group S1 survivors had higher LVMSi than controls (38.0 ± 8.3 vs 30.9 ± 7.0 g/m2.7, P = .012). The multivariable linear model analysis demonstrated that, follow-up time (p = 0.027), sex (p=0.020) and BMI (p =0.002) were significant predictors for strain.
Conclusions: The LV EF alone is not an adequate method for the detection of cardiotoxicity among the CCSs. Therefore, especially for those with modifiable cardiovascular risk factors, LV hypertrophy and often normal LV EF, the LV longitudinal strain should be added to the panel of screening methods.

P121

Cardiac function and its perinatal determinants in former extremely low birthweight infants in late childhood

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Introduction: Extremely Low Birthweight (ELBW) infants suffer from adverse cardiovascular outcomes in later life, including an increased incidence in heart failure. Less is known on perinatal risk factors for these outcomes.
Methods: Within the PREMATCH study, perinatal characteristics were documented in 93 ELBW children and 87 controls and cardiac function was phenotyped at age 11 by echocardiography. First, we compared echocardiographic function between cases and controls. We subsequently investigated perinatal risk factors for adverse cardiac outcome amongst ELBW children. Parameters were adjusted for age and/or body surface area (BSA), when appropriate.
Results: Most cardiac dimensions were significantly smaller in ELBW children compared to controls, even when corrected for BSA. Regarding left ventricular (LV) systolic function, ELBW children had a similar LV ejection fraction but lower LVs' (9.3 vs. 10.5cm/s, *p=0.005*). LV diastolic function in ELBW children exhibited a similar E-wave, A-wave and E/A, a slightly higher E/e' (5.5 vs. 5.2, *p=0.053*) and a higher LVa' (5.9 vs. 6.4cm/s, *p=0.026*), despite similar LVe'. Left atrial sizes were similar between both groups. Right ventricular (RV) function in ELBW children was characterised by a slightly higher fractional area change (47.4 vs. 45.5%, *p=0.028*) despite lower RVs' (12.6 vs. 13.3cm/s, *p=0.019*) and higher tricuspid regurgitation (TR) pressure gradient (18.4 vs. 16.3mmHg, *p=0.002*).

Amongst ELBW children, lower LV mass and interventricular septum thickness (IVSd) were associated with ibuprofen exposure (ρ=-0.25, *p=0.026*; ρ=-0.31, *p=0.005*). LVs' correlated with birthweight and gestational age (ρ=0.38, *p=0.006*; ρ=0.22, *p=0.051*) and inversely with bronchopulmonary dysplasia (BPD), intraventricular hemorrhage, longer ventilation therapy and time to full enteral feeding (ρ=-0.27, *p=0.016*; ρ=-0.26, *p=0.024*; ρ=-0.23, *p=0.051*; ρ=-0.24, *p=0.041*). TR pressure gradient associated with longer hospitalisation and higher discharge weight (ρ=0.28, *p=0.012*; ρ=0.28, *p=0.021*). RVs' was higher in children who received ibuprofen (ρ=0.23, *p=0.044*). No associations existed between RV function and BPD.
Conclusions: At age 11, ELBW children have morphologically smaller hearts, with similar LV and RV systolic hemodynamics despite a subtly altered contractility and RV pressure and without evidence of LV diastolic dysfunction. Cardiac function is influenced by identifiable perinatal factors that may assist the prediction of cardiovascular outcome in ELBW children.

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Echocardiographic outcomes for ELBW children and controls. *Results are displayed as raw or unadjusted values (standard deviation; SD) and adjusted mean differences (confidence interval; CI) where appropriate. Adjusted mean difference (AMD) is the difference between z-scores of cases and controls. EF: ejection fraction; FS: fractional shortening; ESV: end-systolic volume; EDV: end-diastolic volume; SV: stroke volume; LVs': tissue Doppler imaging (TDI) of the mitral annulus during systole; DT: deceleration time; IVRT: isovolumic relaxation time; LVe': TDI of the mitral annulus during early diastole; LVa': TDI of the mitral annulus during atrial diastole; LVIDd: left ventricular (LV) inner dimension; IVSd: interventricular septum thickness; LVPWd: LV posterior wall thickness; LALD2ch: left atrial (LA) long axis dimension in 2-chamber view; LALD4ch: LA long axis dimension in 4-chamber view; RVs': TDI of the right ventricular (RV) wall during systole; RVFAC: RV fractional area change; RVAs: RV area during systole; RVAd: RV area during diastole; TR_{PG}: tricuspid regurgitation pressure gradient; PAAT: pulmonary artery acceleration time; RVminor: RV basal diameter minor axis.*

	Unadjusted values (SD)			Z-scores (CI)	
	ELBW	**Controls**	**p**	**AMD**	**p**
Heart rate (bpm)	73 (13)	70 (11)	0.28	-	-
Left ventricle – systolic function					
EF (%)	57.9 (4.4)	57.4 (4.1)	0.5	0.09 (-0.21 to 0.39)	0.55
FS (%)	38.4 (7.5)	37.2 (5.8)	0.322	-	-
ESV (ml)	29 (7.5)	35 (9.5)	**<0.001**	-0.23 (-0.5 to 0.04)	0.1
EDV (ml)	69 (15.5)	81 (18.5)	**<0.001**	-0.22 (-0.5 to 0.07)	0.13
SV (ml)	40 (9.1)	46 (10.1)	**<0.001**	-0.21 (-0.56 to 0.13)	0.22
LVs' (cm/s)	9.3 (3.9)	10.5 (2)	**0.005**	-	-
Left ventricle – diastolic function					
E-wave velocity (cm/s)	0.97 (0.15)	0.94 (0.12)	0.12	0.12 (-0.12 to 0.37)	0.32
A-wave velocity (cm/s)	0.47 (0.11)	0.45 (0.09)	0.21	0.16 (-0.1 to 0.41)	0.22
E/A	2.2 (0.63)	2.16 (0.46)	0.65	0.02 (-0.27 to 0.3)	0.91
DT (ms)	133 (21)	138 (23)	0.11	-	-
IVRT (ms)	58 (8.2)	58 (8.9)	0.66	-	-
LVe' (cm/s)	17.9 (2.3)	18.5 (3)	0.2	-	-
LVa' (cm/s)	5.9 (1.4)	6.4 (1.3)	**0.03**	-	-
E/e'	5.5 (0.01)	5.2 (0.01)	0.05	-	-
Left heart – morphology					
LV mass (g)	75 (19)	84 (20)	**0.004**	-0.1 (-0.45 to 0.23)	0.53
LVIDd (cm)	4.04 (0.34)	4.2 (0.31)	**0.003**	-0.21 (-0.42 to 0)	**0.045**
IVSd (cm)	0.66 (0.09)	0.71 (0.09)	**0.004**	-0.16 (-0.32 to 0)	**0.049**
LVPWd (cm)	0.65 (0.08)	0.66 (0.08)	0.34	0.05 (-0.12 to 0.22)	0.55
LALD2ch (cm)	3.85 (0.53)	3.99 (0.48)	0.11	-0.1 (-0.44 to 0.25)	0.58
LALD4ch (cm)	4.24 (0.4)	4.38 (0.39)	**0.031**	-0.05 (-0.31 to 0.22)	0.73
Right ventricle – systolic function					
RVs' (cm/s)	12.7 (1.5)	13.3 (1.6)	**0.019**	-	-
RVFAC (%)	47.4 (0.07)	45.5 (0.06)	**0.028**	-	-
RVAs (cm^2)	8.0 (2.3)	9.0 (1.8)	**0.003**	-0.63 (-0.92 to -0.34)	**<0.001**
RVAd (cm^2)	14.9 (3)	16.4 (2.5)	**<0.001**	-0.64 (-0.92 to -0.36)	**<0.001**
TR_{PG} (mmHg)	18.4 (4)	16.3 (3.1)	**0.002**	-	-
PAAT (ms)	124 (20.8)	126 (20.7)	0.38	-	-
Right heart – morphology					
RVminor (cm)	3.12 (0.31)	3.32 (0.33)	**<0.001**	-0.46 (-0.66 to -0.26)	**<0.001**

P122
Cardiac function in male pediatric patients with classic Fabry disease

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Introduction: Fabry disease (FD) is an X-linked lysosomal storage disorder of the glycosphingolipid metabolic pathway. Progressive cardiac involvement is characterized by left ventricular hypertrophy (LVH) and is the leading cause of death in FD patients. However, in pediatric patients with early stages of the disease, LVH is generally not present. Strain analysis by 2D speckle-tracking echocardiography (2DSTE) may be a sensitive tool to detect preclinical myocardial involvement before LVH, yet its usefulness has not been assessed in pediatric patients.
Methods: We assessed the echocardiographic images and electrocardiograms (ECGs) of sixteen genetically confirmed male pediatric patients with classic FD. All examinations were performed below the age of nineteen and patients were compared with sixteen healthy controls that were matched for age and gender. Cardiac parameters were derived from conventional echocardiographic measurements of function and dimensions, tissue Doppler imaging (TDI), global and segmental 2DSTE tracings, and electrocardiograms.
Results: Conventional echocardiographic parameters revealed no significant changes in the cardiac function or dimensions of our sixteen patients (median age 15.3 years, range 10.5 - 17.7) compared to controls (median age 15.3 years, range 10.7 - 18.0), and no cases of LVH were observed. One patient was reported to have aortic root dilation with asymptomatic aortic regurgitation. TDI of the left ventricle (LV) revealed no significant changes, though TDI of the right ventricle (RV) revealed a subclinically lowered median tricuspid annular early diastolic (E') velocity for patients compared to controls (12.4 cm/s versus 15.3 cm/s, $p < 0.01$). 2DSTE analyses revealed no differences in global or segmental myocardial systolic function. Compared to controls, heart rates derived from ECGs were significantly lower in patients (61 bpm versus 68 bpm, $p = 0.04$), though bradycardias, other arrhythmias, or conduction disorders were not observed.
Conclusions: The LV myocardial systolic function in male pediatric patients with classic FD is likely unimpaired. Subclinical changes in the tricuspid annular E' velocities may be useful in the early detection of RV involvement. Furthermore, subclinically lowered heart rates and asymptomatic aortic regurgitation can be observed in FD during childhood.

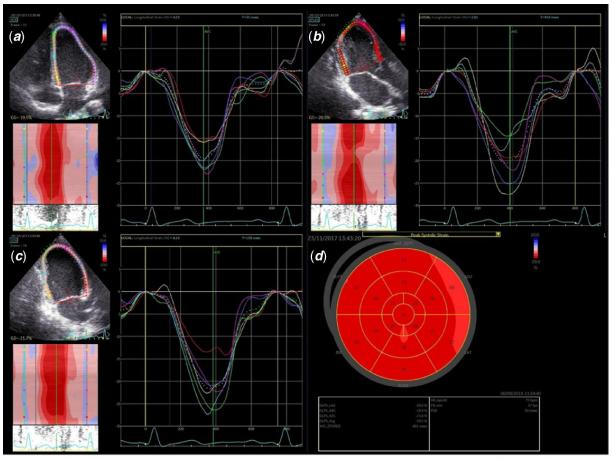

Figure 1. Longitudinal strain curves of a patient with classic Fabry disease of the apical four-chamber view (**A**), apical three-chamber view (**B**), and apical two-chamber view (**C**). The bull's-eye plot shows the average systolic strain per segment (**D**).

P123
Characterization of Fontan associated liver disease with non-contrast enhanced magnetic resonance imaging

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Introduction: The Fontan circulation is a palliative strategy for single ventricle congenital heart defects. It results in an obligate elevation in systemic venous pressure, which provides a predisposed environment for chronic hepatic injury. Reliable non-invasive methods to monitor FALD have not been described. Non-contrast enhanced MRI techniques are potentially robust methods to characterize liver fibrosis. The study aimed to evaluate non-contrast enhanced MRI sequences for diagnosis and grading of Fontan associated liver disease (FALD).
Methods: Fontan patients (n=29) and healthy volunteers (n=11) underwent a non-contrast enhanced magnetic resonance imaging

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(MRI) protocol at 1.5 Tesla with T_1- and T_2-mapping and apparent diffusion coefficient (ADC) mapping. The entire liver was scanned and segmented to provide tissue characterization for each anatomic segment and for the left and right lobe. Fontan patients also underwent routine FALD screening including abdominal ultrasound and laboratory assessment. MRI parameters were compared within and between patients and healthy subjects, respectively.

Results: T_1 relaxation times were higher in Fontan patients compared to healthy volunteers (left lobe: 735 (IQR 705-764) ms vs. 588 (IQR 579-603) ms, $p<0.001$; right lobe: 745 (IQR 715-784) ms vs. 586 (IQR 555-602) ms, $p<0.001$). The same was observed for T_2 relaxation times (left lobe: 60 (IQR 58-62) ms vs. 56 (IQR 56-58) ms, $p=0.003$; right lobe 63 (IQR 59-64) ms vs. 58 (IQR 56-60) ms, $p=0.002$). Left liver lobe ADC was lower in Fontan patients compared to healthy volunteers (1.10 (IQR 1.06-1.18) x 10^{-3} mm^2/s vs. 1.23 (IQR 1.19-1.29) x 10^{-3} mm^2/s, $p<0.001$). There was no difference for the right liver lobe (1.07 (IQR 1.02-1.09) x 10^{-3} mm^2/s vs. 1.09 (IQR 1.06-1.13) x 10^{-3} mm^2/s, $p=0.280$). Prediction of moderate to severe FALD was only achieved with the T_2 relaxation time in segment VII or right lobe T_2 relaxation time.

Conclusions: Non-contrast enhanced MRI methods are able to visualize regional differences in tissue characteristics in healthy volunteers and Fontan patients. T_1- and T_2-relaxation times were prolonged in Fontan patients suggestive of fibrosis or congestive hepatopathy, while reduced ADC might reflect impaired microperfusion. These methods have promising clinical potential for detection of liver abnormalities in Fontan patients.

P124

Churg-Strauss Syndrome in a paediatric patient presenting with myocarditis: role of 2D strain echocardiography in directing management.

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Introduction: Churg–Strauss syndrome (CSS) is an eosinophil-rich necrotizing vasculitis of small-to-medium blood vessels that affects many organs including cardiac, pulmonary, renal, nervous, and vascular systems. It is rarely reported in paediatric patients and its etiology remains unknown. Myocarditis in the paediatric population is very rare. We present a case of a paediatric patients diagnosed with CSS presenting with myocarditis in whom 2D strain echocardiography was essential in guiding the management.

Methods: A 13 year old girl presented with lethargy, calf pain and chest pain. Blood tests showed raised troponin I (peak at 2593ng/L), eosinophilia and raised CRP. She was previously diagnosed with asthma. An echocardiogram showed pericardial effusion and moderately impaired left ventricular function. She underwent extensive rheumatology work up and the diagnosis of Churg-Strauss syndrome was made. Cardiomyopathy screen was also performed to exclude a primarily heart muscle disease.

Results: The initial echocardiogram showed left ventricular hypertrophy with normal left ventricular size and moderately impaired left ventricular systolic function (left ventricular ejection fraction (LVEF) 45%). Global longitudinal strain (GLS) was reduced at -12% (Fig 1a). There was mild-moderate global pericardial effusion without haemodynamic compromise. She was treated with steroids, azathioprine and cyclophosphamide infusion, with improvement of the ventricular function (normal with conventional 2D parameters) and GLS (-17% vs -12% pre-treatment) (Fig 1b). The steroids dose was then reduced. A repeat echocardiogram 4 weeks later showed reduction of the GLS (-13%) despite normal values on conventional 2D parameters, with LVEF of 56% and fractional shortening (FS) of 29% (Fig 1c). She remained asymptomatic. The treatment was then optimized with subsequent improvement in the GLS (-18%) (Fig 1d). On last echo and on weaning dose of steroids, her LV systolic function remains satisfactory with GLS of -18%, LVEF 53% and FS 32%.

Conclusions: CSS is rare in the paediatric population. The prevalence of myocarditis in this setting is unknown but is thought to be extremely rare. We present a case where 2D strain echocardiography, being superior to conventional 2D left ventricular function measurements, played a pivotal role in the management and follow up of our patient.

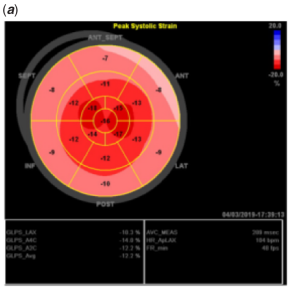

Fig 1a: at presentation

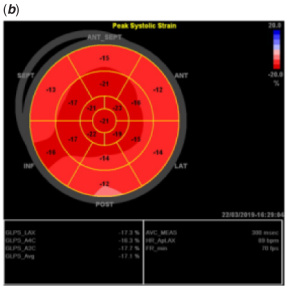

Fig 1b: on treatment

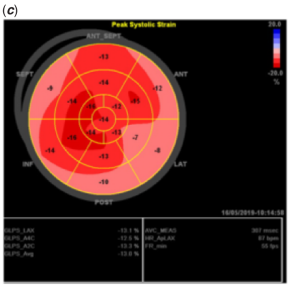

Fig 1c: with reduced steroids dose

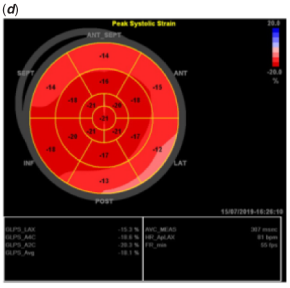

Fig 1d: on increased steroids dose

P125

Classification of congenital heart disease associated with pulmonary arterial hypertension (PAH-CHD) within the TOPP registry

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Introduction: PAH-CHD is a heterogenous group of different pathophysiologic conditions. Previously, a clinical classification of congenital systemic-to-pulmonary shunts associated with pulmonary hypertension was suggested for adult patients and considered feasible for pediatric patients, but showed several limitations not allowing to classify all pediatric PAH-CHD patients. We thought to analyze all patients enrolled in the TOPP registry (**T**racking **O**utcomes and **P**ractice in **P**ediatric Pulmonary Hypertension) using a modified classification.

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Methods: TOPP cohort was screened and all patients with PAH-CHD reviewed. Hemodynamic and clinical data were reviewed and individually classified according to predefined definitions (Table 1) by 2 independent investigators, in case of disagreement decision was reached with a 3rd investigator. If the third investigator cannot classify the patient was not classified. Survival curves were calculated for each group; idiopathic PAH was used for comparison.
Results: There were 563 patients in the registry, 205 fulfilled inclusion criteria of PAH-CHD and categorized to groups A-E respectively: 39 (19.0%), 27 (13.2%), 73 (35.6%), 59 (28.8%), 7 (3.4%) and a mean age at diagnosis of 8.2, 6.3, 5.7, 8.6, 1.9 years. mPAP[mmHg]/PVRi[WUxm2], range: 67.5 (33-110)/20.4 (4.4-73.7); 60.1 (26-96)/10.7 (3.2-23.8); 57 (29-126)/14.7 (3.3-62.5); 53.1 (25-143)/13.3 (4.4-22.2); 49.9 (32-77)/17.8 (3.3-64.3). The majority of patients where in Functional Class II and III, independent of CHD group. Survival was worse for group C and similar to idiopathic PH. 6 patients were categorized as "Group 5", 24 patients were unable to be classified due to lack of reported data.
Conclusions: Most patients were grouped in C (with a significant proportion of ASDs). Higher mean age at diagnosis were found for groups A and D. Patients in group A (despite higher mPAP and PVRi) and B tend to have better outcome whereas C and D had a similar survival as idiopathic PAH. This modified classification improves the accuracy of diagnosis but still has limitations due to the heterogeneity of patients.

A	Patients with previous left-right shunt, presents with right-left shunt and saturation ≤92% at diagnosis (includes patients <0.5–1 years of age with Eisenmenger physiology who have never had left-right shunt)
B	Patients with left-right shunt, saturation >92 % (independent of operability) but PVRi >3WU (TOPP inclusion criteria)
C	Patients with "coincidental shunts" e.g. small VSD and/or PDA (includes all paediatric patients with ASD)
D	Patients with operated CHD without significant haemodynamic residual lesions
E	Patients with TGA following neonatal arterial switch
Clinical specifications outside of ABCD(E) classification	
	WHO Group 5 Patients with (segmental) TOF/MAPCAs or Scimitar (complex CHD) as per new NICE classification

P126
Complete atrioventricular septal defect – echocardiographic assessment of imbalance

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Introduction: By 10-15% of the cases with complete atrioventricular septal defect (AVSD), there is ventricular imbalance, most often with right ventricular dominance. A very important and unsolved problem by the unbalanced AVSD with borderline left ventricle are the anatomical and physiological characteristics that can identify the patients suitable for two-ventricle correction from the cases which are indicated for single-ventricle palliation. The purpose of the study is to identify the echocardiographic parameters which distinguish the balanced from the unbalanced forms of AVSD and which aid in determining the type of surgical intervention.
Methods: We conducted a retrospective analysis of the data of 64 patients with AVSD (9 of them with unbalanced AVSD with single-ventricle palliation), operated at the Pediatric Cardiology Department of the National Heart Hospital, Sofia, Bulgaria, from 01.01.2014 to 31.12.2019. We measured a group of predefined echocardiographic parameters on stored echocardiographic images.
Results: The statistically significant echocardiographic measurements, which identify the balanced from the unbalanced forms of complete AVSD, are the ratio between the mitral and the tricuspid valve, the ventricular inflow angle, and the indexed ventricular septal defect ($p<0.001$). The parameters for which p is close to the critical value of 0.05 are the z-scores of the tricuspid and of the mitral valve, and the z-score of the left ventricle. The following echocardiographic measurements did not show any significant differences between patients with two-ventricle correction and single-ventricle palliation: the ratio between the long axis of the two ventricles, the ratio between the diastolic diameters of the two ventricles, the atrioventricular septal angle, the atrioventricular valve index, and the modified atrioventricular valve index.
Conclusions: Following a protocol with predefined echocardiographic parameters by the preoperative evaluation of patients with complete AVSD will contribute to their correct indication for two-ventricle correction or for single-ventricle palliation.

P127
Coronary CT Angiography in Asymptomatic Patients with Transposition of the Great Arteries After Arterial Switch Operation

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Introduction: In patients with transposition of the great arteries (TGA) after arterial switch operation (ASO) neoaortic dilatation, pulmonary artery stenosis and coronary disorders are the most common complications. The need for routine screening for anomalies of the orifice regions of the reimplanted coronary arteries as the most dangerous long term complication in patients after the ASO still remains debatable. The aim of our study was to present the results of coronary computed tomography angiography (CCTA) in asymptomatic patients with TGA after an ASO.
Methods: We reviewed 21 CCTA scans performed after ASO randomly selected from asymptomatic patients in Gottsegen Gyorgy Hungarian Institute of Cardiology in 2020. Retrospective ECG gated CT acquisitions were performed with a 256-slice CT scanner (Revolution GE Medical Systems, Milwaukee, USA) with intravenous contrast enhancement. The analysis included measurements of aortic and pulmonary diameters, multiplanar reconstructions of the main coronary arteries, localization of the origin of the coronary arteries in the neoaortic sinus. In order to normalize our results base as per pediatric age and/or body size data were expressed as Standard Deviation Score (SDS).
Results: 10.2 (+ 3.5) years after the ASO aortic annulus dilation averaged 2.7 SDS according to normal aortic diameter and 1.5 SDS according to normal pulmonary annulus. Mean left pulmonary artery (LPA) diameter was -1.9 (+/- 1.6) SDS, mean right pulmonary artery (RPA) diameter was -1.8 (+1.7) SDS. 8/21 patients had hypoplastic (- 2 SDS) LPA and 9/21 had hypoplastic RPA. Localization of the origin of the coronary arteries in the neoaortic sinus (determined by the angle between the connecting axis of the aortic and pulmonary valves and the orifice of the coronary arteries) were LCA 38.6+10.5° and RCA 64.5+10.2° respectively. Potentially critical (<23°) LCA angulation was found in 5/21, while potentially critical LCA first centimeter angle <23° in 4/21 patients. Ellipticity-index (coronary height-width ratio) was 1.3+0.1 in LCA and 1.2+0.1 in RCA.
Conclusions: CCTA performed in asymptomatic patients with TGA after ASO provides useful information for postoperative

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follow-up. The frequency of coronary anomalies is high in this patient group. The role of potentially dangerous anatomical features and their impact on late complications should been evaluated with follow-up studies.

P128

CT angiography or cardiac MRI for detection of coronary artery aneurysms in Kawasaki disease

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Introduction: Kawasaki disease (KD) is an acute vasculitis that mainly affects the coronary arteries. This inflammation can cause coronary artery aneurysms (CAAs). Cardiac assessment consists of risk stratification for the development of myocardial ischemia, based on *Z* score (luminal diameter of the coronary artery corrected for body surface area). Echocardiography is the primary imaging modality in KD but has several important limitations. Coronary computed tomographic angiography (cCTA) and Cardiac MRI (CMR) are non-invasive imaging modalities and of additional value for assessment of CAAs with a higher diagnostic yield compared to echocardiography. The objective of this single center study is to determine the difference of coronary artery assessment of cCTA versus CMR children with KD.
Methods: Out of 965 KD patients from our database, a total of 111 cCTAs (104 patients) and 311 CMR (225 patients) have been performed since 2010. For comparison we identified 54 KD patients who had undergone both cCTA and CMR.
Results: CMR missed 50% of the CAAs identified by cCTA and thereby only identified 8 patients with CAAs compared to 14 patients by cCTA. For comparison we identified 54 KD patients who had undergone both cCTA and CMR
Conclusions: Our single center study in KD demonstrates that cCTA is the better modality to detect CAAs as it is capable to detect more CAAs than CMR.

P129

Do parents of children with congenital heart disease really know their children's disease?

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Introduction: Due to the diagnostic-therapeutic advances that have occurred in the last 3 decades, the patients with congenital heart disease (CHD) are a growing population. Although most of these patients present residual defects, thus turning them into a chronic population. The knowledge of the disease by parents of children with CHD is a factor in their health promotion, and with an impact on health-related behaviour and quality of life of these patients in terms of health. The objective of the present study is to evaluate how the perception of disease is and the degree of knowledge of it in parents of children with CHD.
Methods: Descriptive cross-sectional study conducted over a period of 6 months in a Tertiary Centre of Pediatric Cardiology, which involves 130 parents of children with CHD (ranging from 3 months to 17 years; 42 % girls). It is carried out using an anonymous questionnaire with 15 items, which can be grouped into 4 domains: name and nature of the heart disease, impact of the disease in the quality of life, perception of illness and its social impact
Results: 76% of parents of children with CHD know the name of their disease. However 46% of them do not understand it. 45% of parents believe that the CHD of their children has a cure. 89% of them believe that their illness does not pose a problem in terms of of social relations, referring to 80% that they can make a daily life similar to healthy children of the same age. Only 6% of parents believe that their children are not feeling well in general health terms.
Conclusions: Parents of children with CHD present important knowledge gaps in their children's disease. It is necessary to intensify the education of the families of these patients, to ensure the understanding of the disease and that in this way, they can impart this knowledge to their children to improve health-related behaviour.

P130

Early and easy diagnosis of Marfan Syndrome in children: Utility of AV valve prolapse at primary consultation

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Introduction: Clinical diagnosis of Marfan syndrome (MFS) in children still faces difficulties, because Ghent criteria often do not appropriately identify children with MFS. We use the tricuspid and mitral valve prolapse (TVP, MVP) as a surrogate marker for the "hypermobile" heart in MFS patients and evaluate this as an early diagnostic tool.
Methods: In our pediatric Marfan clinic, we diagnosed MFS in 182 patients (9,3±6,1y). We retrospectively analysed data and 2D-echocardiograms at the first two visits in our clinic, evaluated the presence of TVP and MVP and the genetic or clinical diagnosis of MFS according to Ghent criteria.
Results: In our cohort at baseline 99 patients showed TVP (9,4 ±5,8y), 80 patients MVP (10,0±5,6y) and 46 both TVP and MVP (9,5±5,5y). The specificity was 88, 91 and 97%, leading to a positive predictive value (PPV) of diagnosing a MFS of 73, 78 an 91%. The negative predictive value (NPV) for MFS when not having TVP and MVP was 76%.
Conclusions: The combination of a TVP and MVP in pediatric patients at the first two visits is significantly associated with the later diagnosis of MFS (specificity 97%, PPV 91%). The absence of TVP and MVP makes the diagnosis MFS unlikely (NPV 76%). In conclusion the presence of a TVP and MVP can be used as an additional early diagnostic predictor for MFS in children.

	MFS positive (n)	MFS negative (n)	Sensitivity %	Specificity %	Positive predictive value %	Negative predictive value %	p-value
TVP +	73	26	46	88	73	70	<0,0001
TVP -	83	199					
MVP +	63	17	50	91	78	73	<0,0001
MVP -	62	172					
TVP+/MVP+	42	4	47	97	91	76	<0,0001
TVP-/MVP-	47	153					

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P131

Early clinical manifestation analysis of patients with critical congenital heart disease

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Introduction: Critical congenital heart diseases (CCHD) represent a group of defects with early and severe clinical presentations requiring urgent operative or transcatheter intervention or a fatality may occur. Early diagnosis is mandatory, defining the prognosis and outcome of treatment. The purpose of the current study is to analyze early clinical manifestation and the options for diagnosis of CCHD in the region of north-east Bulgaria.
Methods: 272 newborns with CCHD were retrospectively studied for 25 years, from 1987 to 2011. Patients were divided into three separate groups: obstructive lesions of the systemic circulation (OLSC) - 103 patients, obstructive lesions of the pulmonary circulations (OLPC) – 64 patients and non-obstructive, mixing cardiopathies (NOMC) – 105 children.
Results: The prevalence of CCHD is 12.5% of all congenital heart diseases. The most common early clinical sign is cyanosis - in 65% of the cases, which is in support of the use of the pulse-oximetry screening test. In 21% of the children, there is no audible cardiac murmur. Not surprisingly, newborns with OLPC manifest at the earliest stage – 1.8 days, and the latest clinical manifestation has the newborns with OLSC – 4.5 days ($p<0,05$). Clinical manifestation is presented in 44.4% of patients during the first day of life, and in 81.7% during the next 4 days. The medium age of diagnosis is 8.5 days. The earliest diagnosis is in patients with OLPC - 4.2 days, followed by OLSC – 9.5 days and NOMC – 10 days ($p<0.05$). Until the 4th day after birth, 54% of CCHD can be diagnosed, and 27.7% of newborns with manifested clinical signs remain undiagnosed. Most often newborns are diagnosed in Children's hospitals – 60%, and only in 30% of cases in Neonatology Departments. 15,8% of newborns are discharged without being diagnosed, and in 22% early diagnosis was incorrect, especially in patients with OLSC and NOMC.
Conclusions: The diagnosis of CCHD often is delayed, especially in patients with OLSC and some NOMC. Screening programs for early diagnosis and improvement in prognosis are needed.

P132

Echocardiographic assessment of infants born premature with catheter laboratory diagnosed pulmonary hypertension

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Introduction: Neonates born premature has an increased risk of developing bronchopulmonary dysplasia (BPD) and pulmonary hypertension (PHT) in respect of dull term babies. There is no clear consensus on when and which parameters to use as a screening for PHT in this population.
Methods: We retrospectively studied echocardiographic parameters of PHT in 17 children born prematurely (gestational age < 36 +6w) who underwent right heart catheterization and had a demonstration of PHT.
Results: The 17 infants where studied at 3,3 + 3 years of age had Gestational Age of 28,6 + 3,9 weeks and birth weight of 943 + 504g. 47% suffered from intrauterine growth retardation and 71% from BPD. Atrial septal defect (ASD) was seen in 88%, ventricular septal defect (VSD) in 31%. One had positive family history of PHT. No correlation between the CATH grade of PHT and prenatal, postnatal and cardiac condition were seen. A moderate to good positive correlation in catheter systolic ($R=0,7$, $p=0,006$), mean ($R=0,63$, $p=0,01$) and diastolic pulmonary pressure ($R=0,58$, $p=0,03$) with echography estimation of PAPs was seen. The grade of dilation of inferior vena cava (IVC) showed a strong positive correlation with catheter PAPs ($R=0,79$, $p<0,003$), very strong with PAPm ($R=0,96$, $p<0,001$) and PAPd ($R=0,97$, $p<0,001$). Right ventricular FAC correlates negatively moderately with PAPm ($R=-0,58$, $pp=0,02$) and strongly with PAPd ($R=-0,76$, $p= 0,007$). The grade of D-shaping of the IV septum correlates moderately with PAPd ($R=0,60$ $p=0,039$). The echography estimate of PAPs shows a strong negative correlation with FAC ($R=-0,76$, $p=0,001$) and a moderate positive correlation with RV/LV ratio ($R=0,50$, $p=0,004$) and the shape of the IV septum ($R=0,57$, $p=0,02$). No correlation was seen with TAPSE and TDI tricuspid s' wave speed with any of the pressures. There is a strong negative correlation between FAC and RV dilatation ($R=-0,77$, $p=0,003$).
Conclusions: Echocardiographic signs of PHT show a good correlation with catheter laboratory measurements of PAP. A screening strategy for assessment of PHT using echocardiography in infants with chronic lung disease and/or history of prematurity should be routinely performed even in asymptomatic child. Further studies will be necessary to confirm its validity.

P133

evaluation of exercise related change of TAPSE and TAPSE velocity in healthy teenagers

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Introduction: The investigation of the rise rate of TAPSE (Tricuspid Annular Plain Systolic Excursion) under physical activity may provide further information for adequately determining right ventricular function. We performed echocardiographic assessment during exercise stress testing of cardiopulmonary healthy children. We determined the increase of TAPSE and calculated the velocity of TAPSE (mm/s).
Methods: A physical stress (semi suspine bycicle stress echocardiography) test was performed on a total of 23 children including 11 boys and 12 girls, aged between 6 and 17 years. All patients were cardiopulmonally healthy. Participants started at 20 W, with an increase of 0.5W/kgBw and a maximal increase of 20W per stage every 2 minutes. A two-dimensional echocardiography-supported M-mode of the tricuspid valve was carried out using the four-chamber view, while determining the heart rate via ECG. TAPSE and its time were measured at the end of each exercise level. The TAPSE velocity was calculated as mm/s.
Results: With increasing exercise load a constantly heart rate increased up to the end of the end of exercise from 83/min ±13.89 up to 143.25/min±29.44. TAPSE increased from 20.22 ±4.92 mm to a value of 30.25±5.8 mm at the end of exercise (Load level 6) with a plateau from level 4 onwards. The TAPSE velocity changed from 67.03±16.06 mm/s to a value of 130.38 ±20.89 mm/s with a similar plateau formation. There was a more significant (< 0.05) correlation between exercise and TAPSE velocity than with TAPSE as such.

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Conclusions: We suggest to consider the rate of increase of TAPSE velocity as a new function parameter to study right ventricular function. TAPSE alone, on the other hand, correlates poorly with heart rate/ exercise. Thus, the rate of TAPSE velocity should be the preferred parameter to study right ventricular function.

P134

Evidence of accelerating atherosclerosis among children, adolescents and young adults' survivors for childhood malignancies

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Introduction: Cardiotoxicity following treatment of malignancies in all ages has been proven to be the second after early death from the primary disease most serious complication of our existing oncology available treatments. Early and late adverse events of the mechanical or electrical function of the heart are mostly researched. Little is known about the accelerating atherosclerotic process that is seen in increasing number among the surviving populations. The study aims to measure the incidence of subclinical accelerating atherosclerosis in a population of survivors from childhood malignancies
Methods: We examined 105 adolescents-adults, survivors from childhood malignancies. Males:57 Females:48, age range :08-35 years, mean 17.6 y. Divided in 3 age groups: Children: 24, Adolescents: 47, adults 34. From them suffered initially from Leukaemia's (44), CNS tumours: (25), Lymphomas (16), Neuroblastomas (12), Others (8). Years from completion of treatment were: 5.5 years to 25 years with a mean of completion of treatment: 7 years and 8 months. A double-blind study of compering more than six classic and non, apart from the male gender, risk factors (RF) for atherosclerotic Cardiac and Vascular Disease (ACVD) by clinical and laboratory tests, detected by a first research team. Then another team blinded to the findings of the first, conducted measurements of the Intima Media Thickness (IMT) of the internal carotid artery by high frequency resolution ultrasonography. We followed the proposed by the Association of European Pediatric and Congenital Cardiology (AEPC) guidelines regarding IMT calculation. A similar age, gender and BMI control study with no detected risk factors ACVD was examined to detect their IMT calculations. This group served as control to our study group.
Results: A pathologic calculation of IMT in29.4% was proportionally found in all groups. The leukaemia had:20%, CNS tumours:19.04% lymphomas: 18.75%, neuroblastomas:18.18%, miscellaneous group: 16.6% survivors. These corelated with the oldest age population of survivors, the number of years after treatment completion and the BMI of the individuals.
Conclusions: This study must alert physicians that follow-up these patients on the way and methods that must be used to detect early accelerating ACVD among survivors suffered childhood malignancies.

P135

exercise related change of TAPSE and the tricuspid annular movement velocity in M-mode echocardiography in patients with corrected tetralogy of fallot

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Introduction: There are poorly predictive parameters for an irreversibly right heart failure due to regurgitation and right ventricular (RV) dilatation common after Fallot repair. Tricuspid annular plane systolic excursion (TAPSE) has been proven as a good corelative to RV function. The aim of the study was to investigate the changes of the TAPSE and the TAPSE Velocity in echocardiographic assessment during exercise stress testing in these patients compared to healthy subjects.
Methods: 16 patients (12 male, mean age 13.13±6.46 years) with asymptomatic impaired RV systolic function and 15 healthy subjects (5 male, mean age 14±6.79) were enrolled. All subjects underwent a semi suspine bycicle stress echocardiography (sBEE), up to 6 levels of resistance increasing every 2 minutes and 2 minutes of recovery. All participants started at 20 Watt, the increase was calculated with 0.5W/kg Bodyweight with a maximal increase of 20W per stage. At two minutes at every stage TAPSE and TAPSE Velocity were measured. The mean value of five measurements at each stage was used. Differences between groups were tested using Mann-Whitney-U Test with a critical value of U=70. Statistical significance was assumed for $p < 0.05$. Data analysis was performed with SPSS.
Results: Data of both groups for TAPSE and Gradient of TAPSE at rest, stage 7, peak load and recovery showed normal distribution. There was no significant difference in the amount of stages reached by both groups (mean 5.38±1.11 vs. 5.80±1.38). The increase of both parameters from rest to peak load was significant higher in the healthy group (TAPSE: healthy group 8.49±4.66 mm, patients 4.43±3.01 mm, U-value 66; TAPSE Velocity: healthy group 7.96±2.93 mm/s, patients 3.14±1.57 mm/s, U-value 11) with TAPSE Velocity being the more sensitive parameter. (Cohens d TAPSE=1.03, Gradient of TAPSE=2.05) The peak load stage showed the highest difference between groups in TAPSE and Gradient of TAPSE. Thus, the statistical significance of difference of TAPSE Velocity versus TAPSE alone increased with exercise in both groups.
Conclusions: Increase of TAPSE and TAPSE Velocity during sBEE was significant lower in patients after Fallot repair. The more useful parameter to differentiage normal right ventricular performance is TAPSE Velocity.

P136

Exercise Stress Echocardiography in paediatric patients; a single centre experience

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Introduction: Exercise Stress Echocardiography (ESE) is currently applied to paediatric patients mainly to detect myocardial ischemia and very few data is available for congenital heart diseases (CHDs).

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The aim of this study is to describe the current application of ESE in our Paediatric Cardiology Department.
Methods: Data from patients who underwent ESE were retrospectively analysed. Basal and under stress echocardiographic parameters, electrocardiograms, changing in blood pressure and symptoms were reviewed, as well as clinical management plans formulated based on ESEs results.
Results: Nineteen patients (median age 15 years) were included. Fifteen had CHDs and 10 had been previously treated with surgery; one for pulmonary atresia and intact ventricular septum (PA IVS) plus tricuspid valve (TV) dysplasia, 4 had had arterial switch operation (ASO) and 1 Rastelli operation for D-Transposition of Great Arteries (TGA), 2 had been previously treated for Tetralogy of Fallot (TOF), 1 for aortic coarctation (ACo) and subaortic membrane resection and 1 for atrioventricular septal defect (AVSD) Fallot type. Three had undergone percutaneous procedures (2 aortic balloon valvuloplasty in bicuspid aortic valve (BAV) and 1 stent implantation in ACo) and 2 were simply under follow-up (1 for BAV and 1 for left cor triatriatum). Among the other patients, 3 of them were evaluated for potential myocardial ischemia, and 1 was affected by hypertrophic cardiomyopathy (HCM). Exercise was maximal in 10 patients and 2 of them had symptoms at the peak of exercise. It was stopped beforehand in 8 patients because of muscle fatigue and in 1 for dyspnea. No arrhythmia was detected. Clinical management changed in 9 of them (47%) as result of the ESE. Three patients received indication for percutaneous interventions; 1 for aortic balloon valvuloplasty, 1 for stent dilation and 1 for pulmonary valve replacement (PVR); 2 for surgery (1 for aortic valve replacement and 1 for subaortic membrane resection), 3 for further imaging including cardiac computed tomography (CT) or magnetic resonance (CMR); 1 for restriction from intense physical activities.
Conclusions: ESE is a feasible and promising technique in paediatric cardiology, particularly in children with congenital heart diseases, and could influence significantly clinical management plans.

Native diagnosis	Indication for ESE	ESE	Final decision
1 PA IVS+ Ebstein anomaly	TV gradient	Negative	Follow up
2 BAV with aortic regurgitation	Aortic gradient; left ventricular (LV) function	Negative	Follow up
3 HCM	LV obstruction	Negative	Follow up
4 Coronary abnormalities	Ischemia	Negative	Follow up
5 D-TGA, ACo, absence of left coronary artery	Ischemia, hypertension	Negative	Follow up
6 BAV	Transvalvular gradient	Severe aortic stenosis (AS): max/mean gradient=85/42 mmHg	*Aortic balloon valvuloplasty*
7 AVSD TOF type	Right ventricle (RV) stenosis	Significant RV stenosis; max gradient= 65 mmHg	*CMR*
8 D-TGA	Ischemia	Negative	Follow up
9 ACo, BAV, subaortic membrane	Transvalvular gradient	Severe obstruction: max/mean gradient=60/36 mmHg + hypotension	*Subaortic membrane resection*
10 ACo	Re-coarctation and hypertension	Re-coarctation: max gradient 108 mmHg+hypertension	*Stent dilation*
11 Coronary abnormalities	Competitive sports activities	Negative	Follow up
12 TOF	PR and RV function	Pulmonary artery systolic pressure >80 mmHg	*CMR*
13 Left cor triatriatum	Transmembrane gradient	Increased gradient: max/mean =16/10 mmHg without symptoms	*Exercise restriction*
14 TOF	PR and RV function	Persistent RV dysfunction+ symptoms	*PVR*
15 BAV with severe AS	Ischemia	Negative	Follow up
16 Chest pain	Ischemia	Rule out myocardial bridge	*Cardiac CT*
17 D-TGA	Ischemia	Negative	Follow up
18 D-TGA	Competitive sport activities	Negative	Follow up
19 BAV	Transvalvular gradient	Severe AS: max/mean gradient=120/47 mmHg	*Aortic valve surgery*

P137
Extremely rare case of almost complete absence of myocardium of the left ventricle in the newborn.

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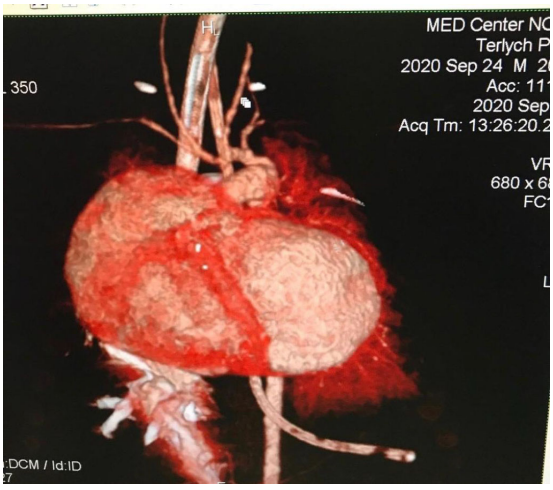
Introduction: We present an extremely rare case of absence of the myocardium of the left ventricle in a newborn. There is a similar pathology of the right ventricle Uhl Anomaly described, however there is a complete lack of information regarding the absence of the left one.
Methods: A newborn child was admitted to the neonatal Intensive Care Unit with the clinical symptoms of respiratory distress. Child form the fourth pregnancy, born on the 37-th week of gestation by physiological delivery with the birthweight of 3400 gr, APGAR score 6\7 points. Mother was diagnosed with gestational diabetes during pregnancy and was receiving insulin (12 week). Shortly after birth child presented with respiratory distress and required administration of oxygen. On examination: severe general state, pale skin, increased work of breathing with intercostal retractions, nasal flaring, RR – 72/min, HR – 168 b/min, SpO2 – 94%. On auscultation of the lungs and heart - bronchial breathing, 4\6 systolic murmur along the left lower sternal border with palpable thrill. Abdomen was soft on palpation, liver +2 cm. A congenital heart defect was suspected, the child was placed on artificial respiration and prostaglandin and furosemide infusion was administered.
Results: Paraclinical examination. ECG revealed signs of the left ventricular hypertrophy, Sever cardiomegaly on chest roentgenography. On 2D Echocardiogram: enlargement of the left ventricle, thinning of the left ventricular wall, common atrium, hypoplastic mitral valve with severe insufficiency. Mild tricuspid insufficiency. MPA 9 mm, RPA – 3,5 mm, LPA – 4 mm, Severe hypoplasia of aortic arch with interruption. PDA – 9 mm were visualized. Computer tomography was performed: agenesia of the ascending aorta, dilated ductus arteriosus that fills descending aorta, common atrium, mitral valve agenesia, enlarged ventricles, thinning of the left ventricular wall, right ventricle hypertrophy, expressed lung congestion were visualised.
Conclusions: Due to the complicated anatomy, which included extreme thinning of the myocardium wall and mitral valve agenesia, surgical correction is impossible, therefore palliative care is indicated in this particular case.



P138
Giant right atrium, what does it hide? Case presentation.

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Introduction: Malformation of the right atrium is a rare cardiac abnormality and usually it is reported as isolated malformation in the literature.
Methods: We present a newborn patient with prenatal diagnosis of right atrial dilatation.

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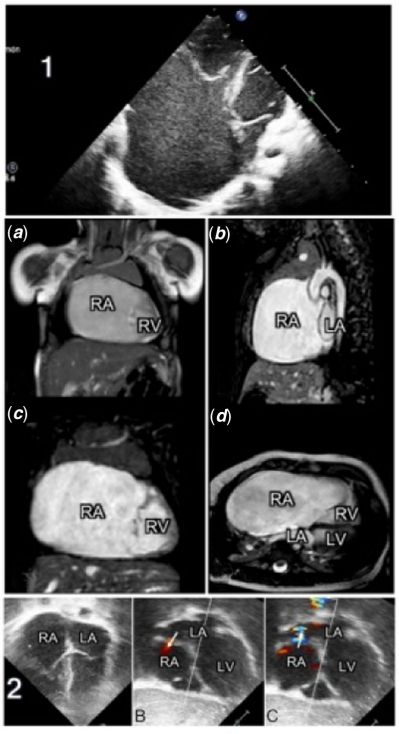
Results: Post-delivery, patient was asymptomatic and the diagnosis was confirmed by transthoracic echocardiography (Figure). There were no signs of vascular nor airway compression. During follow up, there was a progressive enlargement of the right atrium and the tricuspid valve regurgitation progressed from mild at birth to moderate-severe. Cardiovascular Magnetic Resonance Imaging (MRI) was performed at six months (Figure) showing cardiomegaly at the expense of the right atrium with normal right and left ventricular volumes and function. Due to the risk of thrombosis and arrhythmias[1], surgical treatment was decided at the age of 18 months. Intraoperative findings confirmed the diagnosis of giant right atrial aneurysm and absence of right atrioventricular sulcus. The right coronary artery could not be identified and only small collateral of epicardial vessels were seen over the right ventricle. Extensive right atrium wall resection and De Vega tricuspid valve annuloplasty were performed. Two years later, the patient remained asymptomatic. Echocardiography study demonstrated a residual stiffness of the right atrium and suggested several indirect signs of elevated right atrial pressures. The right atrium had a globular shape (Figure 3) with an interatrial septum bulging from right to left and bidirectional shunt across the foramen oval. Evaluation of the inferior vein cava demonstrated reversal 'a' wave with normal diameter. The residual tricuspid valve regurgitation was only mild and doppler evaluation excluded the presence of pulmonary artery hypertension. Shall we actually reduce the risk of thrombosis and arrhythmias we do not know, but follow-up evaluation is mandatory.
Conclusions: Sometimes, as seems in our case, right atrial dilatation hides a problem outside of itself. In spite of surgical right atrium plasty in order to avoid risk factors (arrhythmia, thrombosis), problems derived from underlying right ventricle restriction could appear in follow-up.



1. Neonatal echocardiography, 4-chamber plane.

A-D. MRI at 6 months. Cardiomegaly with a normal size of the right ventricle.

2. Postsurgical echocardiography. Convex interatrial septum on the left and bidirectional shunt in oval foramen.

P139

Haemodynamic impact and prognostic relevance of right ventricular pressure load in patients after repair of Tetralogy of Fallot

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Introduction: Right ventricular outflow tract (RVOT) stenosis after repair of Tetralogy of Fallot (TOF) has been linked with favourable RV remodelling but adverse outcomes. The aim of our study was to assess the haemodynamic impact and prognostic relevance of RV pressure load in this population.
Methods: 337 TOF patients (mean age 17.8 ± 8.0 years) were included in a prospective CMR multicentre study. Myocardial strain was quantified by feature tracking technique at study entry. Follow-up including the need for pulmonary valve replacement (PVR) was assessed. The combined endpoint consisted of ventricular tachycardia (VT) and cardiac death.
Results: Echocardiographic RVOT peak gradient was significantly associated with smaller RV volumes and less pulmonary regurgitation (PR), but lower biventricular longitudinal strain. During a follow-up of 10.1 (0.1-12.9) years the primary endpoint was reached in 19/301 patients (cardiac death n=6, sustained VT n=2, non-sustained VT n=11). A higher RVOT gradient was associated with the combined outcome (HR 1.03, CI 1.00-1.06, p=0.026) and a cut-off gradient of ≥26 mmHg was predictive for cardiovascular events (HR 3.97, CI 1.58-9.96, p=0.003). In patients with PR ≥ 25%, a mild residual RVOT gradient (15-30 mmHg) was not associated with a lower risk for PVR.
Conclusions: A higher RVOT gradient reduced PR and RV dimensions but negatively affected biventricular function. Mildly increased pressure gradients did not protect from PVR and were already associated with adverse outcome, which may have implications for both the initial surgical strategy and the indication for reintervention in TOF patients.

P140

Imaging Modalities Employed in Delineation of Sinus Venous Defects and Outcome Over Last Decade

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Introduction: Sinus venosus defect (SVD) is often associated with partial anomalous pulmonary venous drainage (PAPVD). The diagnosis can be challenging and may be delay due to the underlying anatomy. We review the mode of diagnosis and surgical outcome in children with sinus venosus defects at our centre.

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Methods: Retrospective review of patients with a diagnosis of SVD and PAPVD underwent surgical repair, between February 2008 to January 2019. Medical records were reviewed to obtain clinical, demographic and outcome data.
Results: 37 children underwent surgical repair for SVD with PAPVD. Median age at time of diagnosis was 4.2 years (range 0.5 to 15.5 years old). In 32 of 37 (86%) patients, SVD was diagnosed and PAPVD was suspected or diagnosed on by two-dimensional transthoracic echocardiogram including modified subcostal sagittal-oblique bicaval view. In five of 37 patients, the final diagnosis of SVD and PAPVD were achieved by transoesophageal echocardiogram, diagnostic cardiac catheterization, cardiac computed tomography and intraoperatively. The median age at the time of surgical was 5.2 years (range 1.6 to 15.8 years old). 31 patients underwent a double patch repair, four patients underwent a Warden repair and two patients underwent a single patch repair. Of the four patients who underwent a Warden repair two patients had a high insertion of right sided anomalous pulmonary vein into the superior caval vein, one patient had bilateral superior caval veins and one patient had right lower pulmonary vein insertion into the right atrium/superior caval vein junction. There was no mortality, reoperation, residual shunt and pulmonary vein obstruction reported. One patient developed superior caval vein obstruction underwent successful balloon dilatation subsequently. One patient developed atrial flutter two months postoperatively required cardioversion with resumed of sinus rhythm since.
Conclusions: Complementary cardiac imaging modalities improved diagnosis of sinus venosus defect and anomalous pulmonary venous drainage. There is a wide variation in the pattern of anomalous pulmonary venous connection in association with sinus venosus defects. Surgical treatment is associated with an excellent outcome and minimal complications. With progressive evolution in congenital cardiac catheterization, a move to transcatheter closure has been shown to be suitable in selected cases.

P141

Insights into Fluid-Structure Interaction Computational Modeling in Repaired Coarctation of the Aorta: Assessing the Impact of Treatment Modality

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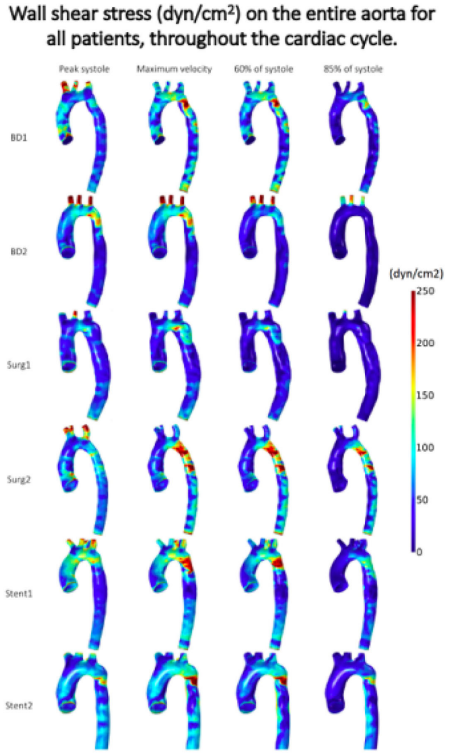
Introduction: Short term results of coarctation of the aorta (CoA) treatment with surgery, balloon dilation (BD) or stent implantation are excellent. Long-term cardiovascular morbidity and mortality remain high. Treatment modality is not associated with differences in major vascular outcomes. The purpose of our pilot, prospective study was to determine the feasibility of patient-specific Fluid structure interaction (FSI) modeling in CoA and to explore its potential in detecting more subtle vascular function differences between treatment modalities.
Methods: We created a FSI model based on patient-specific anatomy and functional data, followed by the computation of hemodynamic and biomechanical biomarkers including velocity and direction of flow, vorticity, pressure, wall shear stress (WSS) and compliance. We selected two CoA patients per treatment type (total 6). The FSI model used the Arbitrary Lagrangian-Eulerian formulation with Navier- Stokes equations for blood flow and the linear elastic equation for the aortic wall. The aortic anatomy, stiffness and velocity waveforms were based on patient-specific data derived from CMR.
Results: Peak velocities at the intervention site were 2.5 m/s for stent, 1.8 m/s for surgery and 1.6 m/s for BD patients. Qualitatively, flow vortices were more marked in the stent than in the surgery and BD groups. The area-averaged WSS were 157 and 168 dyn/cm2 for the stent patients, 60 and 180 dyn/cm2 for the surgery patients and 90 to 115 dyn/cm2 for the BD patients. The two BD patients had the lowest area of luminal surface subjected to elevated WSS (260 and 330 mm2) compared to the two surgery (530 and 1150 mm2) and two stent patients (910 and 1050 mm2). The aortic compliance was higher for the BD patients (1.64 and 2.26 mm) compared to surgery (0.67 and 1.47 mm) and stent (1.12 and 0.6 mm) patients.
Conclusions: Our study is the first to demonstrate feasibility of patient-specific FSI computational modeling in patients with CoA treated with different treatment modalities. Our small sample did not allow a quantitative comparison between treatment modalities but provided qualitive and quantitative data which may become a useful adjunct tool in assessing the post-treatment hemodynamics and vascular function outcomes in patients with CoA.



Legend: BD = patient treated with balloon dilation; Surg = patient treated with surgery; stent = patient treated with stent implantation

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P142

Interrupted Aortic Arch with Aortopulmonary Window. Multidetector computed tomography in the diagnosis and surgical strategy.

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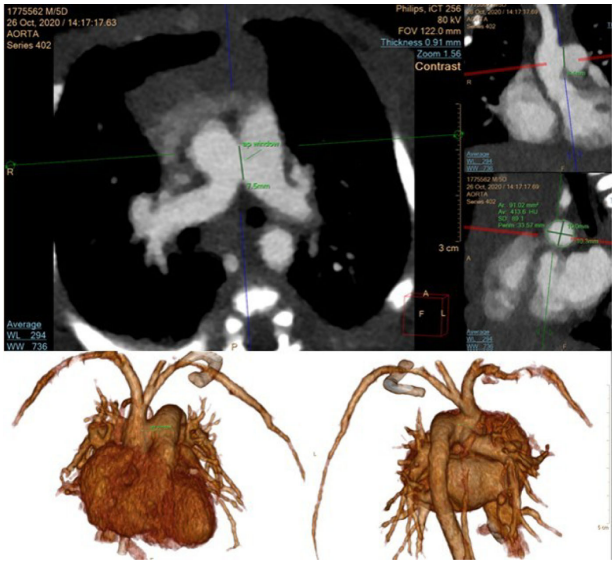
Introduction: Interrupted aortic arch (IAA) is a rare congenital malformation that usually is associated with ventricular septal defect, whereas aortopulmonary window (APW) is commonly associated with an intact ventricular septum and normal aortic arch.
Methods: We present a patient with an Interrupted Aortic Arch Type A associated with APW. We remark the key role of multidetector computed tomography (MDCT) to decide the best surgical approach.
Results: A three-day old newborn was admitted to our hospital with tachypnoea and intake difficulties. Chest X-ray showed increased cardio-thoracic index and pulmonary vascular markings. Echocardiography detected a discontinuity in the aorta just beyond the left subclavian artery, an atrial septal defect, a large defect connecting the ascending aorta and the proximal main pulmonary artery, and a patent ductus arteriosus that continuing with the descending aorta. Prostaglandin E1 infusion was started to maintain ductal patency. A CT-angiography was performed in a 256-MDCT scanner, using a low dose axial protocol, defining more clearly the lesions, depicting the associated lesions, and determining the accurately the distance in the interrupted aorta. A single-stage repair of both APW/IAA reconstruction was performed. The ductus arteriosus was closed and transected making sure to remove all the ductal tissue. APW was repaired through a trans-window approach using a heterologous bovine pericardium patch. Finally, the thoracic descending aorta was widely mobilized upward and anteriorly close to the APW and the aortic arch was reconstructed with a direct end-to-side anastomosis. Neither obstruction of the aortic arch nor pulmonary artery, have been observed in the regular echocardiography follow-up.
Conclusions: The combination of APW/IAA is rare, and the experience with this combination of lesions is limited. The surgical management of APW/IAA is necessarily complex, and a single-stage reconstruction is currently the preferred approach in neonates. The higher risk of aortic arch obstruction in APW/IAA type A could be related to inadequate mobilization of the descending aorta at the time of repair. MDCT is an image technique that demonstrated its usefulness in complex anatomies, as in our case, and provided a fast and non-invasive way for accurate morphological evaluation, obtaining a more reliable diagnosis and allowing the best surgical repair.



P143

Is experienced pregnancy in repaired tetralogy of Fallot related to diffuse myocardial fibrosis?

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Introduction: Most women with repaired Fallot tetralogy (rToF) have accomplished a successful pregnancy, while the cardiac consequences of this status are not well defined. Myocardial fibrosis is linked to adverse clinical outcomes in rToF patients. This study investigates Magnetic resonance (MR) T1 mapping, extracellular volume fraction (ECV) and ventricular function to uncover possible differences related to previous fulfilled pregnancy in rToF women.
Methods: In a prospective cross-sectional study, women with rToF, were invited. CMR including T1 mapping/ ECV and late Gd enhancement (LGE) as well as functional, volumetric and flow data were performed. The results were compared according to clinical parameters between the Fallot subgroups with different parity and to healthy age matched controls.
Results: Fifty out of 56 women, median age 36 (range 21-67) years, 15 nulliparous, performed CMR compared to 31 controls median age 41.9 (24-64) years, 15 women. T1 times/ECV in the left ventricle (LV) in rToF women vs female controls were 1248±61 ms/ 25.8±2.9 % vs 1255±40 ms/26.8±3.1%, p=0.7 and p=0.3 respectively. Right ventricle (RV) T1 times were 1385±124 ms and ECV 37.7±5.4%. The number of pregnancies was not associated with LV T1/ECV p= 0.9 for both or with RV T1/ ECV p= 0.4

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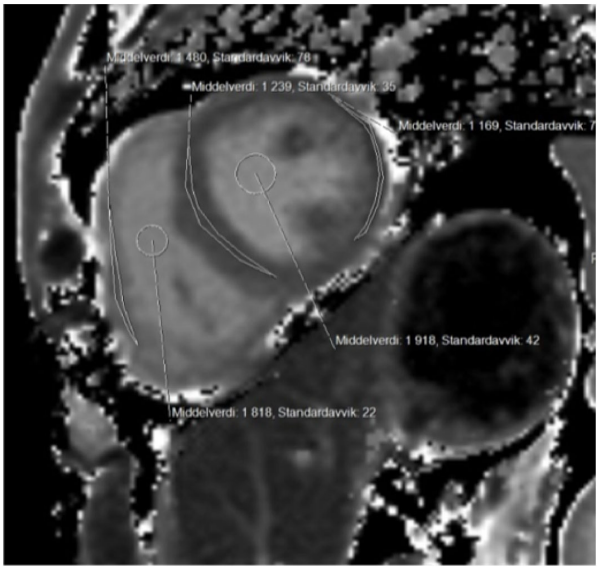
and p=0.6 respectively. In the pregnancy group, indexed LVmass was 43±10 vs non pregnant 38±6 g/m2 p=0.03 while RV ejection fraction was 49±7 % vs 53±6 %, p=0.04. There was no significant difference between groups regarding; LGE in RV/septum present in 27/46 (58%) of patients (p=0.3) or pulmonary regurgitation >10 m/s, median 25 (range 11-42) % in 20/47 (42%) of patients (p=0.9). Indexed LV end-diastolic and end-systolic volumes correlated with LVECV, R= 0.5 p=0.003, and R=0.4 p= 0.005, while RV stroke volume correlated with LV ECV R=0.3 p=0.03 but not to RV ECV R=0.05 p= 0.05.
Conclusions: Women repaired for tetralogy of Fallot have normal LV native T1 or ECV however they show evidence of increased RV markers suggestive of diffuse fibrosis. LV and RV markers were partially correlated to volumetric measurements but they were not influenced by the condition of having fulfilled pregnancy(ies) in rToF women.



Native T1 map with regions of interest in the right and left ventricular walls and in the respective bloodpool, in a woman without experienced pregnancy

P144
Is Left Ventricular diastolic dysfunction an early sign of accelerated atherosclerosis in pediatric survivors for childhood malignancies

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Introduction: Many studies up to date have proven that cardiotoxicity following treatment of childhood malignancies have many adverse events on the Cardiovascular System (CVS). Most common investigated area is the induced cardiomyopathy that presents mostly as heart failure. Little is known about the acceleration of atherosclerotic cardiac disease (ACVD) among this population. Our aim is to study if echocardiographic easy to obtain in clinical follow-up, functional markers can be linked to early ACVD among these patients.
Methods: We examined 105 survivors from childhood malignancies. Males: 57, Females:48. Age range: 8-35 years, mean age 17.7 years. Divided in 3 age groups: Children: 24, Adolescents: 47, adults 34. They initially suffered from Leukemia's (44), CNS tumors: (25), Lymphomas (16), Neuroblastomas (12), Miscellaneous (8). Time elapsed from completion of treatment: 5.5 to 25 years. Mean time of completion treatment: 7 years and 8 months. We used a double-blinded study model. Firstly, we conducted measurements of the Intima Media Thickness (IMT) of the internal carotid artery by high frequency resolution ultrasonography. Following the proposed by the Association of European Pediatric and Congenital Cardiology (AEPC) guidelines regarding IMT calculation. Then, a second research team, blinded to the IMT measurements, studied their diastolic function of the Left Ventricle (LV) by Echocardiography -2D(Echo-2D) and Tissue Doppler Index techniques.
Results: All individuals were clinical free of symptoms and had a normal sinus rhythm during both IMT and Echo-2D examinations. They also had normal Ejection Fraction and Shortening Fraction calculated by Simpson technique. In total we detected 10.48% of abnormal LV diastolic disfunction. These consisted with 1. increased isovolumetric relaxation time, 2. reduced systolic pulmonary venous flow,3. E<A in mitral doppler inflow,4. Pathological septal e' and lateral e' waves,5.velosity through the tricuspid "normal" regurgitation > 2.8 m/s, 6. Left Atrial volume index> 34ml/m^2. This were mostly seen in Lymphomas12.5%, Miscellaneous:12.5%, CNS tumors:12%, Leukemia's: 11.36%. All these subjects had pathological IMT measurements.
Conclusions: Although our studied cohort is small shows that diastolic disfunction of the LV detected by simple Echo-2D can be an early sing of accelerating ACVD among survivors from childhood malignancies.

P145
Left ventricle longitudinal strain alterations in asymptomatic or mildly symptomatic pediatric patients with recent SARS-CoV-2 infection

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Introduction: Evidence suggests that, compared with adult patients, clinical manifestations of children's COVID-19 may be less severe. However, multiple reports have raised concern about the so called pediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2 (PIMS-TS) which resembles other inflammatory conditions (i.e. Kawasaki disease, toxic shock). Patients affected by PIMS-TS showed cardiac involvement with myocardial injury, reduced left ventricle systolic function and coronary artery abnormalities, and in some cases, need for inotropes/vasopressors and extracorporeal life support (ECLS). Little is known regarding cardiac involvement in pediatric patients with SARS-CoV-2 infection and none or only mild symptoms of disease.

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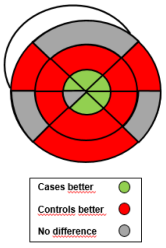
Methods: We analyzed 52 pediatric patients (29males, 56%) with diagnosis of SARS-CoV-2 infection based on either PCR analysis of nasopharingeal swab (NPS), or serological finding of IgG on blood sample and asymptomatic (23%) or only mildly symptomatic (77%) for COVID-19. Patients underwent transthoracic echocardiogram (TTE) after a median time of 3.6 months from diagnosis and negative NPS for SARS-CoV-2. Offline analysis with GE EchoPAC software to measure global longitudinal strain (GLS) of the LV using 2D speckle tracking imaging. Therefore, we compared the results with an age-matched group of 32 controls (18males, 56%).
Results: Cases and controls were similar regarding age and gender. LV biplane EF was significantly lower in the cases group, although still in the normal range (62.4±4.1% vs. 65.2±5.5%, p=0.012). TAPSE and LV-GLS were comparable between the two groups. GLS analysis showed significant strain reduction of the LV mid-wall segments and of the basal anterior, posterior and septal inferior segments among cases compared to controls. On the other hand, apical segments showed higher deformation in cases compared to controls. Furthermore, in the case group there were 14 subjects (27%) with a strain below 16% (mean value minus 2.5 SD) in at least 2 segments.
Conclusions: SARS-CoV-2 infection may affect LV deformation in asymptomatic or only mildly symptomatic children, showing a peculiar pattern with lower longitudinal strain in all mid-wall segments of LV compared to control subjects. The clinical significance of this findings is unclear and follow-up is needed to verify the reversibility of this alterations.

	Cases (n=52)	Controls (n=32)	p
Gender	29 males (56%)	18 males (56%)	0,966
Age	7.5±4.7 years	8±4.9 years	0,673

	Cases (n=52)	Controls (n=32)	p
TAPSE	20.1±3	19.8±3.4	0.822
LVEF (%)	62.4±4.1	65.2±5.5	0.012*
GLS (%)	-21.9±2.4	-22.6±2.5	0.208



P146
Lung ultrasound evaluation of postoperative lung injury in patients with congenital heart disease.

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Introduction: Congenital heart disease reparative surgery almost invariably requires extracorporeal circulation which is often responsible for reversible lung injury in the postoperative period. These patients thus necessitate intensive monitoring of the pulmonary function. The best pulmonary function index is the oxygen administration required to obtain a satisfactory blood oxygen saturation that varies depending on the heart disease and the corrective stage. Fluid balance control is an esier to interpret indirect index. Lung disease manifests itself as pulmonary congestion, pleural effusion, atelectasias and pneumothorax; these are evaluated with imaging techniques such as chest X-ray (CXR) or lung sonography (LUS). The purpose of the present study is to evaluate which imaging technique better evaluates lung congestion and its correlation with oxygen requirement.

Methods: Retrospective study of 53 consecutive patients (55% M age 0-17 years) that underwent surgical intervention for congenital heart disease. All patients were studied with LUS and CXR in the same day by two experimenters blind to the other's result. Pulmonary congestion was rated with a semiquantitative score for both methods. 27 patients repeated the exams on the following day and the difference in clinical and imaging parameters were recorded. Oxygen requirement and daily fluid balance were recorded.
Results: Our data shows that oxygen requirement correlates weakly with pulmonary congestion diagnosed with CXR (rho=0,34, p=0,01) and moderately with LUS (rho=0,53, p<0,001). Oxygen requirement changes in the longitudinal evaluation correlate very strongly (rho=0,83, p<0,001) with modifications in pulmonary congestion demonstrated with LUS while showing no correlation (rho=-0,14, NS) at all with the same modifications demonstrated with CXR. Fluid balance correlates strongly (rho=0,72, p<0,001) with modifications in pulmonary congestion demonstrated with LUS while showing no correlation (rho=-0,15, NS) at all with the same modifications demonstrated with CXR.
Conclusions: Lung ultrasound is a valuable bed side aide in the monitoring of pulmonary congestion after heart surgery for congenital heart disease especially for the possibility of serial evaluation. Our data shows that serial evaluations of pulmonary congestion with LUS correlate with the clinical parameters of reversible lung injury. Serial LUS allows for tailoring of oxygen administration and diuretic treatment to opromise the treatment of lung disfunction.

	Bilancio idrico pro kg media ± DS	Versamento ECO mediana (ri)	Congestione ECO mediana (ri)	Versamento RX mediana (ri)	Congestione RX mediana (ri)	FiO2 (%) media ± DS
T0		0 (0-4)	4 (2-6)	0 (0-1)	0,5 (0-1)	35 ± 16
T24		0 (0-4)	4 (2-5)	0 (0-1)	0 (0-1)	31 ± 12
Delta	-4,8 ± 22 mL	0,2 (0-3)	-0,5 (-2-2)	-0,2 (-1-1)	-0,6 (-1-1)	-4,6 ± 13

Correlazione	p	ρ	Significatività
Versamento RX (T0) - FiO2 (T0)	0,62	0,07	N.S.
Versamento ECO (T0) - FiO2 (T0)	0,85	0,027	N.S.
Versamento RX (T0) - Versamento ECO (T0)	<0,001	0,46	S (moderato)
Congestione RX (T0) - FiO2 (T0)	0,01	0,34	S (debole)
Congestione ECO (T0) - FiO2 (T0)	<0,001	0,53	S (moderato)
Congestione RX (T0) - Congestione ECO (T0)	0,03	0,30	S (debole)
ΔVersamento RX – ΔFiO2	0,14	0,30	N.S.
ΔVersamento ECO – ΔFiO2	0,99	-0,001	N.S.
ΔVersamento ECO – ΔVersamento RX	0,17	-0,27	N.S.
ΔCongestione RX – ΔFiO2	0,49	-0,14	N.S.
ΔCongestione ECO – ΔFiO2	<0,001	0,83	S (molto forte)
ΔCongestione ECO – ΔCongestione RX	0,24	-0,23	N.S.
Bilancio idrico – ΔFiO2	0,0003	0,66	S (forte)
Bilancio idrico – ΔCongestione RX	0,46	-0,15	N.S.
Bilancio idrico – ΔCongestione ECO	<0,001	0,72	S (forte)
Bilancio idrico – ΔVersamento RX	0,14	0,29	N.S.
Bilancio idrico – ΔVersamento ECO	0,99	-0,0008	N.S.

P147
Management of children with giant coronary artery aneurysms secondary to Kawasaki disease within a national referral centre

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Introduction: Kawasaki disease (KD) is an inflammatory disorder of young children associated with coronary artery aneurysms in up to one third of untreated patients. From the recent British surveillance study, 1.6% of these were giant aneurysms (GCAA), defined

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as z score >/= 10. Previously rare, these are becoming more common, especially in infants. As the incidence of KD doubles every 10 years it is now the commonest cause of acquired heart disease in children. In light of recent published guidance, we discuss our recent experience of the ongoing management of these children.
Methods: Over a period of 18 months, four infants presented with GCAA, 3 boys and 1 girl. Age at presentation was between 14 months and 4 years and days of fever before diagnosis were 15, 17, 20, 23. No children had more than 3 associated symptoms. Echocardiography was performed at diagnosis; all demonstrated involvement of both main coronaries - Z scores ranging from +11.5 to +44. Two had significant effusions, 1 had moderate aortic valve regurgitation, 3 had mild ventricular dysfunction. All acutely received 2 doses of intravenous immunoglobulin, high dose aspirin, intravenous methylprednisolone and 1 also received infliximab.
Results: By 6 months follow up, all demonstrated multiple GCAA on angiography but were otherwise well. One child's GCAA has returned to z score <10, but the rest were still giant. Maintenance medication included low dose aspirin with low molecular weight heparin in the 3 infants and warfarin in the older child. All had undergone magnetic resonance imaging (MRI) perfusion scans - all of which showed perfusion defect on adenosine stress. All children were asymptomatic; however, we increased anti Xa therapeutic range to 0.7-1.0 u/l. To date, none have had Electrocardiographic changes of ischaemia.
Conclusions: Infants with few clinical features of KD are at increased risk of GCAA. Careful monitoring with increased range of anticoagulation is essential to prevent complications. Management of these high-risk children requires a multi-disciplinary team approach to reduce risk of major adverse coronary events. We recommend that children with giant coronary artery aneurysms undergo a stress MRI to assess coronary perfusion to gauge risk of an acute coronary event.

P148
Marfan Syndrome: comprehensive cardiovascular characterization

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Introduction: Marfan syndrome (MFS) is an autosomal-dominant connective tissue disorder, which is commonly caused by mutations in FBN1. The cardiovascular phenotype is characterized by aortic root dilation, mitral valve prolapse as well as decreased left ventricular (LV) systolic and diastolic cardiac function. It was the aim of this study to characterize arterial and cardiac function in children and adults MFS.
Methods: Multimodal cardiovascular assessment included echocardiography, ascending aortic distensibility, common carotid intima media thickness [cIMT], parameters of wave reflection (central [cAIx75] and peripheral [pAIx75] augmentation index corrected to a heart rate of 75/minute, aging index [AI]), carotid-femoral pulse wave velocity [cfPWV]), and endothelial function (EndoPAT). Multivariable linear regression (correcting for e.g. age, sex, medication, prior aortic root replacement), and correlation analyses were performed.
Results: We included 20 MFS patients and 67 controls (age 9-49 years). Ascending aortic stiffness, pulse wave reflection (cAIx75, pAIx75 and AI) and cfPWV were significantly increased in patients with MFS. By contrast, cIMT and cIMT/dimension ratio were significantly decreased in MFS. There was no evidence of endothelial dysfunction. Diastolic and systolic function were significantly impaired, but did not correlate with vascular parameters in MFS. These findings were consistent with an increased arterio-ventricular coupling ratio in MFS ($B=0.3$, $p<0.001$). Lastly, normalized LV volume and mass were significantly larger compared to controls, and correlated moderately ($r=0.6$, $p<0.008$) with cfPWV.
Conclusions: MFS is associated with both general central arterial stiffening, and cardiac dysfunction, but the extents of each appear to be unrelated to each other. As cfPWV is associated with LV mass and volume in MFS, it may be suitable for monitoring overall treatment effects in patients with MFS. CIMT is decreased in MFS, supporting that the mechanism of arterial stiffening is different from patients with atherosclerotic changes.

P149
MASSIVE DILATATION OF ASCENDING AORTA IN A PATIENT WITH PSEUDOXANTHOMA ELASTICUM

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Introduction: Pathologic arterial calcifications in childhood occur in the generalized arterial calcification of infancy (GACI) or in the pseudoxanthoma elasticum (PXE); both caused by mutations in ENPP1 and ABCC6, respectively. Affected children could present severe cardiac failure, refractory arterial hypertension, and myocardial ischemia.
Methods: We present a patient with early onset of generalized arterial calcification who showed typical clinical involvement of the GACI, but with a mutation in the gene ABBC6, to which was added a massive fusiform aneurysm of the ascending aorta, a complication described for the first time.
Results: A newborn presented in the early neonatal period with severe arterial hypertension. After ruling out aortic coarctation and renal disease, antihypertensive therapy was initiated with limited response, requiring multiple treatment drugs. An X-ray detected calcification on limb arteries, and the kidney ultrasound showed calcification in renal and interlobar arteries. On suspicion of idiopathic intra-arterial calcification, etidronate and vitamin-D treatment was started. The echocardiography detected a patent ductus arteriosus which was closed percutaneously, and the angiography detected a mild fusiform dilation of the ascending aorta. Molecular analysis detected a mutation of the ABCC6 gene {(c.3421C>T p.(Arg1141*)}, inherited from his father who is a homozygous carrier of this mutation, making the diagnosis of PXE. On regular echocardiography follow-up, the increasing in aorta diameter was progressive, and at the age of nine-years old it became severe, with a diameter of 65 mm on CT scan. An elective surgery was performed with interposition of a 24 mm vascular Dacron® prosthesis between the aorta above the commissures to the brachycephalic trunk.
Conclusions: We present a patient with PXE/GACI manifestations and a fusiform aneurysmatic dilatation of the ascending thoracic aorta, a previously unknown complication in those pathologies. Contrary to other genetic connective tissue diseases (Marfan's, Ehlers–Danlos), aneurysms in PXE/GACI patients are sparse and usually located in intracranial arteries, coronary arteries, and

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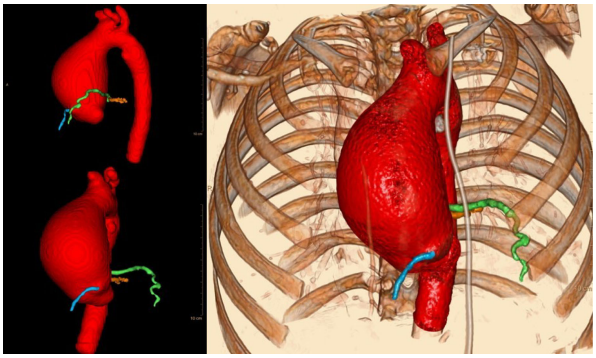
abdominal aorta. PXE/GACI are in a closely related spectrum of disease and the clinical presentation of this disorder is not yet completely understood. There is an overlap between PEX and GACI in clinical phenotypes, and in the genotype as well.



P150

Measured EAT and LV diastolic disfunction an early markers of accelerating Cardiovascular Disease in childhood malignancies survivors?

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Introduction: Adverse effects of oncology treatment on the cardiovascular system has recently become significant interesting. Partly to the increasing rates of survivors, mostly among those presenting in childhood and partly by invention of new drugs in which their late effects as still unknown. New indexes, easy to apply in everyday clinical work are vital, in early detection of any forms of cardiotoxicity. Echocardiography has a key role in detecting cardiotoxicity induced cardiomyopathy, similar indexes have to be established focusing in detecting accelerated atherosclerotic cardiovascular disease (ACVD) among survivors. We aim to examine if combining the use of measured epicardial adipose tissue (EAT) and diastolic disfunction (DD) of LV can serve as early markers of ACVD
Methods: 105 survivors from childhood malignancies were examined. Males: 58, Females:47. Age range: 9-36 years, mean age 17.7 years. Divided in 3 age groups: Children: 24, Adolescents: 47, adults 34. They initially suffered from Leukaemia's (45), CNS tumours: (21), Lymphomas (16), Neuroblastomas (11), Miscellaneous (12). Time elapsed from completion of treatment: 5.5 to 25 years. Mean time of completion treatment: 8 years and 4 months. Firstly, conducted measurements of Intima Media Thickness (IMT) of the internal carotid artery by high frequency resolution ultrasonography. Following guidelines by the Association of European Pediatric and Congenital Cardiology (AEPC). A second team, blinded to the IMT measurements, calculated: 1. The EAT, 2. Their DD of LV by Echo-2Dand Tissue Doppler Index techniques.
Results: We calculated 15% with increased EAT, when compared to a similar age, gender and BMI to our cohort, control population. We detected 23% of abnormal LV diastolic disfunction. These consisted with 1. increased isovolumetric relaxation time, 2. reduced systolic pulmonary venous flow,3. E<A in mitral doppler inflow,4. Pathological septal e' and lateral e' waves,5.velosity through the tricuspid "normal" regurgitation > 2.8 m/s, 6. Left Atrial volume index> 34ml/m2. Pathological combination EAT+DD with pathological IMT was found in 20%.
Conclusions: Although a primary study in this field we show a trend of detecting early subclinical ACVD among survivors from childhood malignancies.

P151

Metastatic involvement of the heart by a genian embryonal Rhabdomyosarcoma in a teenage female

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Introduction: To present a very rare case of genian embryonal Rhabdomyosarcoma with cardiac metastases in a young teenage female.
Methods: A 15 yo girl was referred to our clinic for a mass biopsy in the left genian area, that appeared 5 months ago, presenting a slow growth. A CT scan of the region revealed the mass extension incorporating the carotid artery. No metastases in lung or bones were seen. Two syncopes were mentioned by her mother.
Results: Embryonal Rhabdomyosarcoma was diagnosed. Complex investigations were performed in the Oncology Dept. including cardiac examination, ECG, Echocardiography, and thoracic and abdominal CT. No murmur was audible, but she was tachycardic 138b/min, with weak peripheral pulses and hypotension 90/50mmHg. The ECG showed low voltage in almost all leads. Echocardiography revealed cardiac tamponade with massive pericardial effusion of 3 cm all around the heart, with collapsed right atrium and ventricle and "swinging heart". Multiple tumoral masses attached to the visceral pericardium of the right and left ventricle were detected. The biggest was 4/5 cm, located on the visceral pericardium of the left ventricle, invading and infiltrating the myocardial wall. The interventricular septum was thick and irregular. A mass of 1.8/1.2 cm, adherent to the pulmonary cusp, but mobile with cusp was detected. Systolic dysfunction, gr I mitral insufficiency and speckle tracking with GLS of −6% was found. An emergency pericardial window was performed and 600 ml hemorrhagic liquid was drained, with tumoral cells. Gr III systolic murmur was audible in the upper left sternal border. Bilateral pleural

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massive effusion developed the next day. Emergency thoracentesis was performed. Ventricular fibrillation(VF) electrically converted to sinus rhythm with cardiac arrest occurred, successfully resuscitated. Amiodarone was initiated because of recurrence of VF. Pancreatic reaction developed after Amiodarone. Despite guidelines, where implantable cardioverter defibrillator(ICD) is not indicated if life expectancy is under 1 year, placing an ICD is recommended to prolong life expectancy as shown in the literature and to stop antiarrhythmic medication.
Conclusions: Extremely rare in children, Rhabdomyosarcoma can produce cardiac metastases. Oncologic treatment is mandatory under the protection of an ICD. Complex team is necessary to collaborate.

P152
modeling of mitral valve annulus from 3 dimensional transthoracic echocardiography in healthy and repaired atrioventricular septal defect children.
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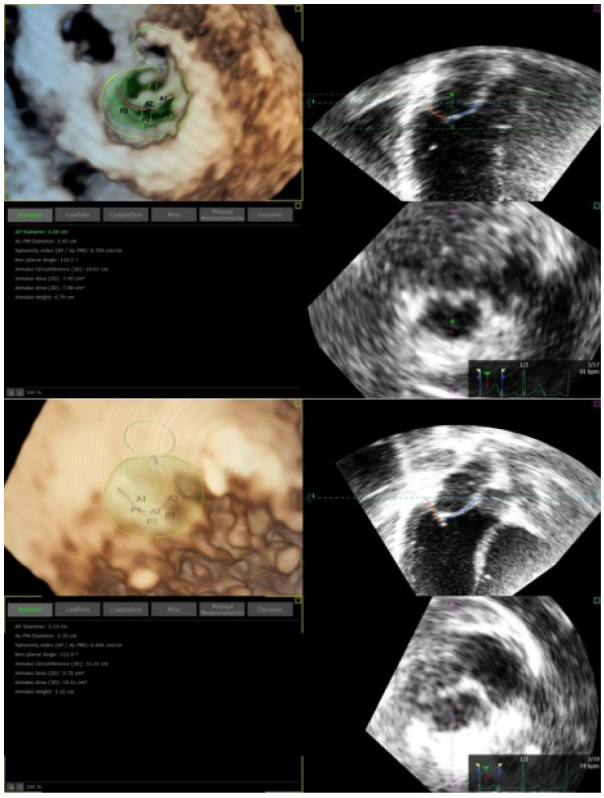
Introduction: Understanding of cardiac structure has been improved using 3D echocardiography. The anatomy and geometry of the mitral valve annulus (MVA) has been previously described with advanced imaging techniques, allowing for a better comprehension of valve dysfunction and providing significant information for the surgical repair. We aimed to assess MVA morphology using 3D transthoracic echocardiography (3DTTE) in patients with repaired atrioventricular septal defect (AVSD), and to compare this morphology with a control group of healthy children.
Methods: 20 patients with repaired AVSD and 20 healthy children were included. 3DTTE full volume and 3D zoom acquisitions were performed (Philips EPIQ7) from apical 4 chambers view. MVA modelization was obtained in all patients using TOMTEC™ 4D MV-ASSESSMENT© software (Fig. 1). Annular area (2D & 3D), circumference and height were calculated, anterolateral-posteromedian (ALPM) and anteroposterior (AP) diameters were measured. Non-planar angle (NPA), sphericity index and the angle between the aortic annulus and AP axe (AAoAP angle) were also calculated. All measurements were indexed to body surface area (BSA).
Results: The two groups were matched for sex, age and BSA. Median age was 5 years and median weight was 16.5 kg. MVA modelization was successfully obtained in all patients from both groups. No difference was found in indexed MVA height, area and diameters between the two groups. However, NPA was significantly more pronounced in patients with a repaired AVSD rather than normal valve (144,5° vs 132,9° respectively; $p<0.01$). A higher sphericity index of MVA was also found in patients with repaired AVSD rather than the control group (0,99 vs 0,85 respectively; $p=0,01$) reflecting a more circular shape of MVA in these patients.
Conclusions: The specific and exclusive shape of the MVA in patients with repaired AVSD was revealed in the patients studied, resulting in a loss of the saddle shape structure. Those results could help in the comprehension of anatomic changes of MVA geometry occurring after AVSD surgery.



P153
MRI STUDY OF THE RV AND LV VOLUME NORMAL GROWTH RELATIVE TO BODY SIZE: THE FALLACY OF BSA INDEXED VOLUME
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Introduction: Indication of surgical or percutaneous treatment often includes RV and LV volume measurements, usually adjusted for body surface area (BSA) by division (Indexed value).
Methods: To study the most appropriate way to index LV and RV size for somatic growth, end-diastolic LV volume (LVEDV) and RV volume (REDV) were measured on MRI performed in 158 and 128 normal patients respectively (age 0.1-18 years). The usual method of indexation (ventricular volume/BSA) was compared with a growth model (LV volume = $A \star BSA^{\wedge B}$) based on physiologically driven methodologies for indexing cardiovascular dimensions (*J Appl Physiol* 2005;99:445–457).
Results: The growth model gives the best model of indexation, with the highest R value and the lowest standard deviation (STD), while the Z-score obtained using this method has a normal distribution and no relationship with age (R value =0). On the contrary, the indexed value has not a normal distribution, and the indexed mean value and the Z-score obtained using this

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method remain correlated with age (R value 0.17 and 0.22, p< 0.05) (see figure).

Conclusions: Adjustment of LV and RV volume for body size by dividing by BSA is incorrect because it assumes a linear relationship, while the growth model using BSA raised to a specific power (B) is the good method for ventricular volume indexation, with a power similar to value 1.3 obtained by echo measurements (*Circ Cardiovasc Imaging 2017 Nov; 10(11): e006979.*) Adjustement of LV and RV volume Our data provide normal values of RV and LV volume adjusted for growth, with Z-score of LVEDV and RVEDV in a patient = (measured volume – (A * BSA $^{\wedge B}$)/STD.

Model	volume (ml) - BSA (m^2)	A	B	R	STD	Z Score vs Age R Value
Growth model	LVEDV = A * BSA $^{\wedge B}$	73,8	1,228	0,91	14,2	0,004
Indexed value	LVEDV = A * BSA	80,6		0,83	16,3	0,17
Growth model	RVEDV = A * BSA $^{\wedge B}$	73,26	1,269	0,9	16,3	0,001
Indexed value	RVEDV A * BSA	82,04		0,82	19	0,22

P154

Muscle function during, and rate of recovery after isotonic exercise is altered and decline with age in children with total cavopulmonary connection

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Introduction: Patients palliated with Total Cavopulmonary Connection (TCPC) have a lower muscle mass and a lower exercise capacity. This study aimed to assess the oxidative metabolic function of the calf muscle during and after heel raises in young patients with TCPC compared to healthy peers.

Methods: Forty-three patients with TCPC, aged 6–18 years, and 43 age and sex matched control subjects participated. Near infrared spectroscopy was used for measuring the dynamic balance of oxygen metabolism in the medial portion of the m. gastrocnemius during isotonic heel raises until exhaustion and subsequent rest period.

Results: During exercise lower rise of deoxygenated haemoglobin (5.13±2.99 arbitrary units (au) *vs.* 7.75±4.15au, p=0.001) and lower increase velocity of the slope of change in total haemoglobin was seen for the patients compared to controls (0.004±0.015au vs 0.016±0.01au, p=0.001). Post exercise slower initial increase in tissue saturation index (0.144±0.11au vs 0.249±0.226au, p=0.007) and a longer half-time to max hyperaemic state was found (23.7±11.4s vs 16.8±7.5s, p=0.001). All differences persisted in subgroup analyses of adolescents (13-18y) but not in the children (6-12y) group. No differences could be seen in number of heel raises performed, in drop of peripheral arterial saturation or in drop of tissue oxygenation index during exercise between patients and controls.

Conclusions: Young patients with TCPC had an altered metabolism in calf muscle during exercise and required longer time to recover even though muscle endurance capacity did not differ compared to control subject. These differences were only seen in the adolescents which could indicate a declining function with age.

P155

NeoDoppler - Continous cerebral blood flow monitoring during cardiac surgery and interventions in children with congenital heart disease

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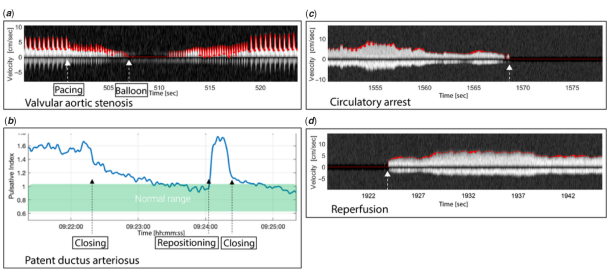
Introduction: Neurodevelopmental impairment is common after surgery for congenital heart disease (CHD). White matter injuries are most common brain damage, present in 20% before and 42% after surgery. Cerebral protection during surgery and catheter interventions is a primary goal. Our aim was to study the clinical potential of the NeoDoppler system to detect cerebral hemodynamic changes during catheter-based interventions and surgery in infants with CHD.

Methods: NeoDoppler is a novel, non-invasive ultrasound system based on plane wave transmissions to monitor cerebral blood flow continuously. The probe frequency is 7.8 MHz, the framerate 300 fps, and the beam covers a wide cylindric area (10/35 mm width/depth). The NeoDoppler probe was attached to the anterior fontanelle of infants during cardiac interventions (n=14) and surgery (n=10). Maximum velocity (Vmax), end diastolic velocity (ED), mean velocity (Vmean) and pulsatility index (PI) were recorded during the procedures.

Results: NeoDoppler provided high quality continuous Doppler data on the cerebral blood flow in several depths simultaneously in all 24 infants. Median monitoring time was 101 min (range 30-342 min), median age 3 months (range 0.10-8 months). The NeoDoppler system was able to rapidly detect alterations in cerebral blood flow in different patients and procedures. Examples are shown in figure 1, demonstrating the hemodynamic changes during balloon valvuloplasty of an aortic stenosis (1a), device closure of a patent ductus arteriosus (1b) and accidental occlusion of the arterial cannula – circulatory arrest (1c) followed by resolution of occlusion – reperfusion (1d) during surgery with selective cerebral perfusion.

Conclusions: By monitoring the cerebral circulation continuously using NeoDoppler, the hemodynamic effects of different procedures and events can be measured directly and instantly. There is a need for better neuroprotective strategies in CHD patients, and improved monitoring during surgery or interventions may potentially contribute toward preventing brain injuries.



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P156

Non-invasive diagnosis of constrictive pericarditis in children: report of two cases and literature review

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Introduction: Constrictive pericarditis is a rare treatable cause of paediatric diastolic heart failure. Early identification of constrictive pericarditis is essential. We present two paediatric cases of constrictive pericarditis in an 11 and a 14-year-old and discuss aspects of advanced imaging and diagnostic challenges
Methods: We applied Mayo clinic echocardiographic criteria to support clinically suspected diagnosis of constrictive pericarditis. In both cases, diagnosis was consolidated by cardiac magnetic resonance imaging.
Results: Patient 1 is an 11-year-old boy presenting with breathlessness, lethargy and abdominal distention several weeks after recovering from a viral illness. Clinical assessment revealed generalised oedema, ascites, pericardial and pleural effusion (anasarca), and remained symptomatic after pericardial and pleural drainage. Patient 2 is a 14-year-old boy who complained of abdominal pain and swelling and found to have ascites with hepatomegaly. He had past medical history of acute lymphoblastic leukaemia (ALL) diagnosed and treated between the ages of three and six (combined chemotherapy COG0331 protocol), and he remained in remission at the time of presentation. We could demonstrate echocardiographic evidence of constriction including diastolic septal "shudder", respiratory ventricular septal shift and variation in mitral inflow E velocity, ratio of medial mitral annular E' to lateral E' and hepatic vein expiratory diastolic reversal ratio. Laboratory investigations ruled out infective or autoimmune disease and demonstrated normal plasma albumin levels in both cases. In both patients, cardiac magnetic resonance imaging (C-MRI) allowed to confirm and consolidate the diagnosis; invasive haemodynamic assessment by cardiac catheterisation was thus avoided. Both patients underwent surgical pericardiectomy with histological confirmation of diagnosis, followed by a rapid and complete resolution of symptoms. One year after surgery, both patients remain asymptomatic with echocardiographic and C-MRI evidence of normalisation of cardiac chambers size; subclinical diastolic function impairment persists in patient 2.
Conclusions: Aetiology of pericardial constriction is heterogeneous: pure constriction is relatively uncommon whilst mixed constriction/restriction picture is seen more often. We emphasize the importance of clinical suspicion guiding the diagnostic search, and value of comprehensive echocardiographic assessment in children presenting with unexplained right-sided heart failure. Cardiac MRI proves a valuable diagnostic tool and allowed to avoid invasive investigations.

P157

Outcomes of Cardiac Function with Tissue Doppler Imaging in Infants After Neonatal Asphyxia

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Introduction: It is known that two-thirds neonates with asphyxia develop varying degrees of myocardial dysfunction as a result of transient myocardial ischemia. Myocardial immaturity in the neonatal period and myocardial regenerative capacity are known. Less studied is the late period of asphyxial post-syndrome and the adaptation of the myocardium over time. The aim of our study was to analyze left ventricular (LV) function in neonates with perinatal asphyxia in dynamics.
Methods: We examined neonates with perinatal asphyxia, at first week of birth, 6 and 12 weeks old. Exclusion criteria was association of primary congenital heart disease -CHD (patent ductus arteriosus was not considered CHD at this age). We used traditional echocardiography to measure M mode fractional shortening (FS) and ejection fraction (EF), Simpson EF, lateral mitral E/A wave and tissue Doppler imaging (TDI) velocities (peak E'/A' wave ratio), peak E/E' wave ratio.
Results: From 49 neonates included, 56,25% were boys, 27 arrived at 6 weeks assessment and 15 at 12 weeks assessment. Mean EF at first week of birth was 60.30% (std. deviation-SD 7.93), at 6 weeks old 60.84% (SD 6.5), and 66.79% (SD 3.76) at 12 weeks old. Mean Simpson EF at birth 57.96% (SD 6.89), at 6 weeks 60.42% (SD 6.23), at 12 weeks 63.33% (SD 6.04). Mean FS at birth 29.63% (SD 5.59), at 6 weeks 30.44% (SD 4.64), at 12 weeks 34.99% (SD 3.1). Lateral mitral E/A wave mean ratio at birth was 0.94 (SD 0.25), at 6 weeks 1.39 (SD 1.5), at 12 weeks 1.52 (SD 1.79). Mean lateral TDI E'/A' at birth was 1.07 (SD 0.94), at 6 weeks 1.07 (SD 0.3), at 12 weeks 1.16 (SD 0.19). Mean TDI E/E' velocities at birth was 8.44 (SD 3.28), at 6 weeks 8.75 (SD 2.64), at 12 weeks 9.41 (SD 2.94).
Conclusions: Our serial asessment showed improvement of echocardiographic parameters of LV function at 12 weeks old, for todlers with neonatal asphyxia. TDI modality for the assesment of myocardial performance, utility in detecting myocardial dysfunction and the interpretation of the obtained values must be done in the context of the clinical situation.

P158

Pediatric Cororary Artery Stent, In-stent Restenosis Assessment and Timing for Reintervention

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Introduction: We present a patient who successfully underwent a Left Coronary Artery (LCA) Drug Eluting Stent (DES) implantation at 7 months of age. 16 years after stent deployment, the patient experienced ventricular fibrillation requiring urgent cardiac catheterization and surgical intervention. Retrospectively, we suggest other means to assess coronary artery stent status and timing for reintervention as an appropriate strategy to avoid future risks.
Methods: A male patient, weighing 6 kg, had surgery at the age of 7 months for anomalous origin of the LCA from the Pulmonary Artery (ALCAPA). LCA stenosis detected postoperatively was dealt with unsuccessful balloon angioplasty and by placement of a sirolimus coated 2.25 x 8 mm Cypher endovascular coronary artery stent. This patient was followed annually with multiple diagnostic studies: coronary angiograms, ECG's, exercise stress

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tests, and cardiac MRI stress tests. None of these demonstrated evidence of significant in-stent restenosis, including a catheterization performed 15 years after stent implantation, correlating with patient's asymptomatic clinical status. After consulting the adult cardiology group, we decided no action was required at that moment

Results: Almost a year after this cardiac catheterization, he presented to the emergency department in ventricular fibrillation arrest. Angiography showed moderate in stent stenosis and an outgrown reference LMCA compared to the stent. A fractional flow reserve study was suggestive of physiological narrowing across the stent. Optical coherence Tomography (OCT) specified the stenosis of approximately 70%. He was taken to the operating room for osteoplasty of the proximal LMCA and implantation of a subcutaneous defibrillator.

Conclusions: Percutaneous coronary intervention (PCI) with stent implantation in infants and children remains rare. Effective detection and quantification of in-stent restenosis and timing for re-intervention in the absence of clinical symptoms is still debated. Retrospectively, it was deemed that another imaging modality, likeOCT should have been used in addition to the annual diagnostic studies. To avoid an extended delay of treatment and this serious event, we believe a preemptive approach for LCA stent expansion had to be attempted, despite the lack of clinical symptomology.

P159

PET/CT role in the diagnosis of Infective Endocarditis in children with Congenital Heart Disease. Is the best thing since the slide bread.

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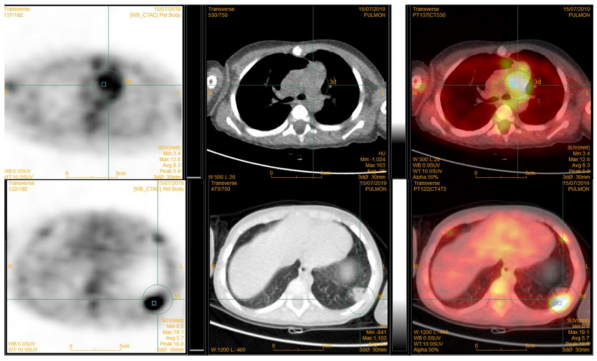
Introduction: The diagnosis of Infective Endocarditis (IE) in Congenital Heart Disease (CHD) involves greater difficulty due to its atypical clinical presentation, and the lower sensitivity/specificity of the echocardiography. The 18F-FDG-PET/CT has shown the value in the IE diagnosis and management in these patients.

Methods: We report five patients with repaired CHD, presented with fever, where TTE/TEE was doubtful in the diagnosis of IE.

Results: **Case#1:** A 4-years-old girl with D-TGA, VSD and PS, intervened with Rastelli procedure and Bovine Jugular Vein (BJV) graft, presented with sepsis by S.aureus. PET/CT detected hypermetabolic activity in the pulmonary graft. One week after the correct treatment, the fever restarted with negative blood cultures but persistent accumulation of FDG in the graft. The final diagnosis was "Relapse of the IE". **Case#2**: A 5-years-old boy intervened of LVOTO with neonatal Norwood procedure, and biventricular reconversion at the 9 months with Rastelli and BJV graft. Presented persistent fever without conclusive TTE/TEE. PET/CT detected hypermetabolic activity in BJV. After 8 weeks PET/CT activity persisted and the graft obstruction got worse, requiring surgical conduit replacement. **Case#3:** An 8-years-old boy intervened of TOF, presented LPB stenosis requiring stent implantation. Presented fever, negative blood cultures and inconclusive TTE/TEE. The PET/TC demonstrated FDG accumulation in the stent zone. **Case#4:** An 11-months-old boy intervened of TAC I presented persistent fever. The blood cultures demonstrated S.epidermidis, and the conventional CT a pediculated image in BJV. The PET/TC showed inflammatory activity in BJV and a pulmonary embolic lesion. **Case#5:** A 13-years-old boy intervened of TAC II with progressive obstruction in the BJV graft. Six weeks after a percutaneous implantation of Transcatheter Pulmonary Valve, presented septic shock. Fever persisted with negative blood cultures and normal echocardiography. The PET/CT detected hypermetabolic activity near to the pulmonary graft.

Conclusions: PET-CT is reported to have a high sensitivity (91%) and specificity (97%) for the diagnosis of IE of prosthetic valves and intracardiac devices, that makes it especially useful in cardiac patients. In our series, PET/CT reduced the rate of misdiagnosed IE; four of five initially classified as "Possible IE" were reclassified to "Definitive IE", and one "Rejected IE" as "Possible IE".



P160

Pre-surgical simulation of AV-valve interventions in AVSD patients - a feasibility study

6. Imaging/functional assessment

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Introduction: Patients with atrio-ventricular septum defect (AVSD) usually undergo biventricular repair with septation and valvoplastic surgery. In some patients, the atrioventricular (AV) valves are invariably abnormal. The surgical outcome in terms of the resulting AV-valve function can be difficult to predict due to the complex nature of the intervention. Therefore, we aim to step-wise test the feasibility of patient-specific AV-valve computer modeling with the goal to provide clinically integrated predictive functional testing of septation and valvoplastic surgery. This is the first study where fluid-structural interaction (FSI) is being used in simulation of AV valve function.

Methods: A simplified computer model with realistic material properties of two coapting AV-valve leaflets and related chordae was constructed to representatively test the feasibility of simulated FSI of AV-leaflets, using SIMULIA software (SIMULIA Dassault Systèmes, France). Next, a simplified but patient-specific anatomic model of the AV-valves in a 5-month-old AVSD patient was constructed in Creo Parametric (PTC, Boston, USA) from ultrasound imaging for actual testing of AV-valve interventions in the next step.

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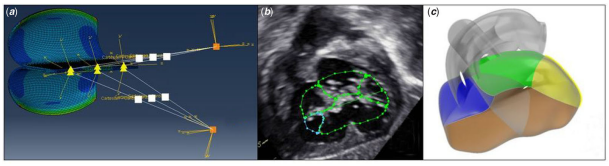
Results: The simplified computer model of the leaflets and chordae successfully interacted and coapted under physiological loading (Panel A). A patient-specific anatomic model of the AV-valves in an AVSD patient was successfully created based on projections obtained from ultrasound images. This means that ultrasound imaging (Panel B) may be a sufficient basis for anatomical modeling (Panel C) and will therefore be used in the forthcoming steps with FSI simulation of septation and valve reconstruction.
Conclusions: We have shown that it is fundamentally feasible to simulate patient-specific AV-valve interactions with commercially available software. Clinically ubiquitous ultrasound imaging may be sufficient to create patient specific computer models required for simulation of septation and valvoplastic surgery. The predictive capabilities of the proposed method remain to be demonstrated.



Panel A: A simplified conceptual computer simulation of two coapting leaflets suspended with chordae.
Panel B: Ultrasound image of the leaflets in the atrioventricular (AV) plane of a patient with atrioventricular septum defect (AVSD).
Panel C: Apical view of a simplified 3D model with patient specific AVSD anatomy created from ultrasound data. This will be used in the forthcoming steps with fluid-structural interaction (FSI) simulations.

P161

Preoperative echocardiographic misdiagnosis of coronary anatomy in transposition of the great arteries (dTGA): is there an impact on surgical outcome?

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Introduction: Nowadays arterial switch operation (ASO) is the preferred approach for surgical correction of D-TGA. The success of the above intervention is mainly determined by surgical translocation of coronary arteries (CA) with potential morbidity and mortality due to myocardial ischemia. The association between CA abnormalities and increased mortality is well understood, but few studies investigated whether preoperative misdiagnosis of CA anatomy impacts on surgical outcome. Echocardiography is the preferred method for the diagnosis of CA anatomy in D-TGA patients and the aim of our study is to determine its diagnostic accuracy and whether misdiagnosis affects the outcome of these patients.
Methods: Retrospective study of all patients who underwent ASO for TGA in our center from 2011 to 2019. Preoperative echocardiography data was compared with intraoperative diagnosis and patients with misdiagnosis where compared with patients with a correct diagnosis for mortality, major postoperative morbidity and CA alteration at cardiac catheterization after 2 years from operation where available. Yacoub classification for CA variants was used.
Results: In the study period 97 (69 males=71%) neonates underwent ASO. The most common coronary pattern was A (65=67%). 33% of patients had other variants comprising pattern B (11), pattern D (15), pattern E (5) and inverted coronary arteries (1). Diagnostic accuracy was 91% in pattern A and decreased to 80% for pattern E, 60% for pattern D and 9% for pattern B. The only inverted coronary pattern diagnosis was correctly identified. No statistically significant differences where identified in mortality, intraoperative times, short term (delayed sternal closure, prolonged mechanical ventilation, reintubation, ECMO, arrhythmias, sepsis) and long term morbidity (abnormalities at catheterization). We observed only a slight increase in coronary artery abnormalities rate in misdiagnosed patients (27% vs 11%) that did not reach statistical significance. Detailed results are displayed in table 1.
Conclusions: We did not find any significant impact of misdiagnosis on mortality, short and long term morbidity in our study. The diagnostic accuracy in our study was higher than previously reported. Our results suggest that correct identification of coronary artery pattern does not affect outcomes of ASO and thus more invasive studies are not routinely recommended.

Table 1.	Correct diagnosis	Misdiagnosis	P value
Mortality	2 (3%)	0 (0%)	0,43
Extracorporeal circulation time	147 ± 7 min	164 ± 8 min	0,29
Cross-clamp time	100 ± 4 min	108 ± 3 min	0,39
Surgical complication	11 (17%)	3 (17%)	0,93
Mechanical ventilation time	30 ± 5 hours	35 ± 4 hours	0,50
Delayed sternal closure	6 (8,2%)	2 (9,1%)	0,90
ECMO	2 (3%)	0 (0%)	0,43
PICU stay	4,1 ± 0,3 days	3,7 ± 0,3 days	0,42
Post-operative hospital stay	11,8 ± 0,9 days	10,3 ± 0,8 days	0,35
Arrhythmias	8 (12,5%)	3 (15%)	0,77
Reintervention	5 (6,8%)	1 (4,3%)	0,66
Sepsis	6 (9,23%)	2 (10%)	0,92
Coronary abnormalities at 2 y catheterization	5 (27,8%)	6 (10,2%)	0,06
Non coronary abnormalities at 2 y catheterization	11 (18%)	2 (11%)	0,45

P162

Protein C deficiency presenting with a mass in the right ventricle in a neonate

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Introduction: 1-month old asymptomatic boy presented to us with loud systolic murmur. Echocardiogram showed a small mid muscular VSD with restrictive flow, but there was a mass in mid right heart that was swimming towards the right ventricle outflow tract. The mass that was pedunculated to the free wall of right heart, was not obstructive and well formed. It measured 14mm by 10 mm, non-friable, has similar echogenic signal of the heart muscle. There was a small non-obstructive friable mass in the left pulmonary artery.
Methods: History revealed that the mother had Protein C deficiency few years ago and was treated with Enoxaparin. The mother was also diagnosed with anti phospholipid antibodies. Her first child was completely normal. No genetic testing was performed and the mother was on no medications on presentation. We treated with Heparin infusion, followed by LMWH. We documented clear shrinking in the size of the RV mass, and complete resolution of the LPA clot after 2 weeks from starting the anticoagulation. However, all our Haematology tests came back non-conclusive so far. This is the Protein C/levels, but no genetics results yet.
Results: Protein C binds to the endothelial cell surface protein thrombomodulin and is converted to APC by thrombin. The APC molecule then interacts with protein S to inactivate two critical coagulation cofactors, factors Va and VIIIa 1.

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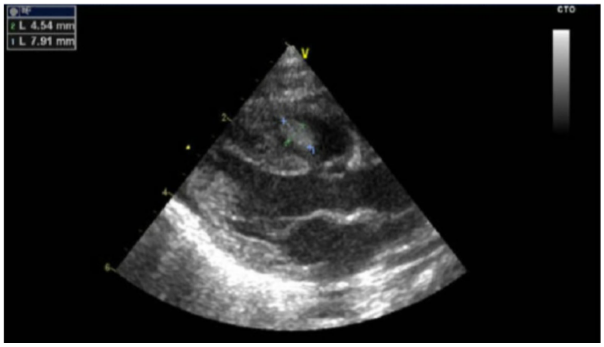
Conclusions: Deficiency of protein C is usually transmitted in an autosomal dominant manner. In the heterozygous state the typical manifestations are recurrent venous thrombosis and associated pulmonary embolism 2. The homozygous state, which is rare, shows itself as fulminant neonatal intravascular coagulation 2. Absence of protein C activity despite normal amounts of the protein is due to the presence of a dysfunctional protein C molecule 3. Cardiac intraventricular thrombus is most commonly associated with an area of myocardial infarction. Left ventricular thrombi can also develop in congestive heart failure and cardiomyopathies. It is unusual for cardiac intraventricular thrombus to form in such environment of the right ventricle when there is a VSD jet nearby. We find no previous report of this happening in protein C deficiency in Paediatrics.



P163

Regression of coronary arteries dilatation 6 months after multisystem inflammatory syndrome in children (MIS-C)

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Introduction: Multisystem inflammatory syndrome in children (MIS-C) is a rare complication of SARS-CoV-2 infection, with an incidence of about 1:100'000 children. According to published case series, between 10% and 40% of MIS-C develop coronary artery modifications, mainly hyperechogenicity, with a lower incidence of aneurysm. Evolution and outcome of coronary artery aneurysm post MIS-C is unknown.
Methods: We report the case of a 10-year old male with medium left anterior descending coronary artery (LAD) aneurysm (diameter of 6.2 mm, z-score +7.9) and small right coronary artery (RCA) aneurysm (z-score +2.9) detected one week after his hospital admission for hypotensif shock in the context of MIS-C and positive serologies for SARS-CoV-2. He didn't meet diagnosis criteria for Kawasaki disease. He was treated with 2 g/kg immunoglobulin (administered after coronary artery dilatation was observed, as the recognition and definition of MIS-C was contemporary with our case), corticosteroids and anakinra. He rapidly normalized his initial mild LV dysfunction and cardiac enzymes elevation.
Results: Since discharge, the patient was treated with antiplatelet therapy (100 mg aspirin daily) and carefully followed up in outpatient cardiology. On echocardiography, coronary artery dimensions progressively regressed, prompting a control computed tomography (CT) 6 months after MIS-C episode. CT confirmed LAD and RCA dimension near-normalization, compared to the fusiform dilatations 6 months ago : LAD maximal diameter of 3.7 mm (z-score +2.3), RAD maximal diameter of 4 mm (zscore +1.8). Moreover, no coronary stenosis was observed.
Conclusions: Coronary artery aneurysm in the context of MIS-C probably represents a post-infectious vasculitis. This case illustrates a regression of coronary artery dilatation after a few months. Further research is needed to assess if this finding reflects a generalisable outcome and to study the effect of medical treatment on the evolution of coronary artery dilatation post MIS-C.

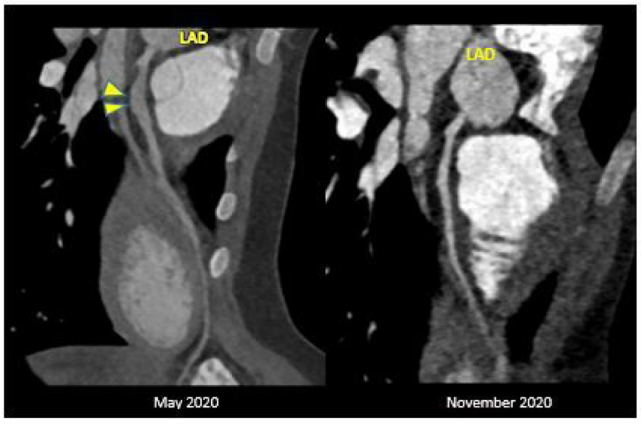

Figure 1: Curved multiplanar reconstruction of the LAD and comparison between May 2020 (aneurysm, z-score +7.9) and November 2020 (ectasia, z-score 2.3)

P164

Relation between blood concentration of pulmonary hypertension drugs and hemodynamics

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Introduction: The efficacy of pulmonary hypertension drug in congenital heart disease (CHD) is widely known. Japanese circulation Society guideline recommend the usage and dosage of pulmonary hypertension drugs in pediatric CHD patients. However, the pharmacokinetics and hemodynamic effects of these drugs are unknown.We examined the blood concentration of pulmonary hypertension drug and its effect on hemodynamics in pediatric CHD.
Methods: Fifteen CHD patients who administrated pulmonary hypertension drug were enrolled. The age of patients was 3.7 years (1.0-12.5), and the administration period was 385 days (80-1772). We measured the mean pulmonary artery pressure (mPAP), transpulmonary pressure (TPG), pulmonary vascular resistance (Rp)

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and pulmonary blood flow (Qp) before and after administration. The drug dosages were Sildenafil (S) 2.6mg/kg /day (1.1-3.5), Tadalafil (T) 0.9mg/kg /day (0.7-1.1), Bosentan (B) 5.3mg/kg/day (2.1-5.4), Ambrisentan (A) 0.1mg/kg/day (0.9-1.7), and Macitentan (M) 0.1mg/kg/day. Monotherapy and combination therapy were administrated to 5 and 9 patients, respectively. A drug blood level below -1SD from the interview form was defined as a low level. Cases in which all drugs were at low level were classified as low group, and the others were classified as non-low group.
Results: The frequencies of low level in each drug were S: 4/7 (57%), T: 1/6 (17%), B:1/3 (33%), A: 5/6 (83%) and M: 1 /1 (100%).3 patients(monotherapy: 2, combination: 1) were classified to low group and 11 patients (monotherapy: 3, combination: 8) to non-low group. Changes in mPAP, TPG, Rp, and Qp in the low group were 4.5 (-5 to 10) mmHg, 6 (-1 to 12) mmHg, 1.2 (-0.7 to 3.5) U.m2, and -0.15 (-0.6 to 0.5) L/min/m2, respectively. In the non-low group, changes in mPAP, TPG, Rp, and Qp were 4.5 (-15 to 15.5) mmHg, 2 (-14 to 17.5) mmHg, 0.5 (-1.2 to 2.2) U.m2, and +0.4 (-01.3 to 1.7) L/min/m2, respectively. None of the hemodynamic parameters were different between the two groups.
Conclusions: In some cases, the drug blood drug level is low even when the recommended dosages in the guidelines were administered. There was no difference in hemodynamic changes due to blood drug levels.

P165
Reliability of echocardiography parameters in patients with a systemic right ventricle: A prospective multicentre study

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Introduction: Systemic right ventricle (RV) is a rare and complex form of congenital heart disease (CHD) with a prognosis related to RV dysfunction and impaired physical capacity. Routine follow-up relies on cardiac ultrasound, however the prognostic value of echocardiography parameters remains under debate. Real-life patient follow-up involves different ultrasound systems. We aimed to evaluate echocardiography parameters' reliability in systemic RV, in terms of reproducibility, using a vendor-independent software, and in terms prediction of physical capacity impairment.
Methods: Adult patients with D-transposition of the great artery (d-TGA) who underwent atrial switch or with congenitally corrected TGA (cc-TGA) were included in this multicentre prospective study. Current ultrasound parameters were analysed using TomTec-Arena version 40 (TOMTEC Imaging Systems GmbH, Unterschleissheim, Germany)™ software. Intraclass correlation coefficients (ICC) assessed inter- and intraobserver reliability. Associations between the most reproducible echocardiography parameters and exercise capacity (peak VO2, VE/VCO2 slope) were explored.
Results: A total of 47 patients were included in the study (85% d-TGA, median age 36.4 ± 8 years). Conventional and 2D strain echocardiography parameters indicated the existence of a RV dysfunction (TAPSE 12.8±3.1 mm; RV free wall longitudinal 2D strain 13.6±3.9 %). Good reproducibility (ICC>0.75) for both intra and interobserver variability was observed in 8 RV echocardiography parameters. Among them, the TAPSE and the RV free wall longitudinal 2D strain, using TomTec-Arena™ RV speckle tracking software algorithm, were associated with peak VO2 (r=0.4 and 0.26, respectively).
Conclusions: Using a vendor-independent software, TAPSE and RV free wall longitudinal 2D strain were the most reliable echocardiography parameters in this prospective study, which attempted to reflect real-life follow-up of patients with systemic RV.

P166
Right circumflex aorta with aberrant left subclavian artery and aortic coarctation in an infant: a case report

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Introduction: Right circumflex aortic arch (RCAA) is a rare aortic arch anomaly caused by retroesophageal crossing of the aorta to the left side. A vascular ring is formed when a left ductal ligament connects the descending aorta to the pulmonary artery. Its association with aortic coarctation is unusual.
Methods: Case presentation of an infant with a vascular ring due to a RCAA with an aberrant left subclavian artery, and coarctation.
Results: 6-month-old baby referred from an outside hospital with a history of aortic coarctation and a vascular ring. At twelve weeks of age, she presented with signs of coarctation and was taken to the operating room where was found to have a RCAA. No coarctation was viewed from the left thoracotomy. Resection of the ductal ligament was done to relieve the vascular ring. Postoperatively, a computed tomography (CT) scan confirmed the RCAA and a coarctation in the middle of the transverse arch. Two days later, she underwent coarctation repair with good recover. One month after, she presented with weak femoral pulses. Echocardiogram showed a residual coarctation of the aorta and left ventricular hypertrophy. The patient was brought to our institution and underwent a CT angiography, which confirmed a right aortic arch crossing the midline behind the esophagus, above the carina, with an independent origin of the supra-aortic vessels as follows: left carotid, right carotid, right subclavian, and an aberrant left subclavian artery arising from a retroesophageal diverticulum (Figure). Additionally, aortic arch coarctation between the origin of the right carotid and right subclavian artery was seen. Balloon angioplasty and stent implantation was done with improvement of the blood pressure in the lower extremities. She continued her follow-up asymptomatic with symmetrical pulses.

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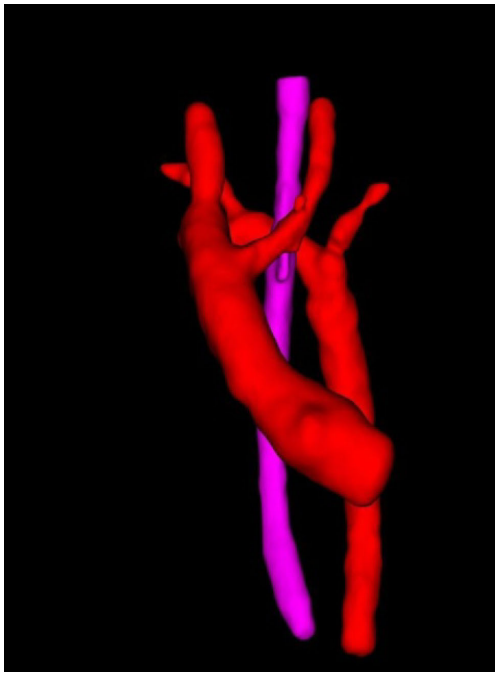
Conclusions: The association of RCAA with an aberrant left subclavian artery and coarctation is a rare anomaly. It is essential to recognise it in a timely fashion, look for other vascular and cardiac associations due to the complications that can arise from the vascular ring. Depicting the anatomy in cross-sectional imaging for an accurate diagnosis guides the management and helps the surgeons for optimal repair.



P167

Risk of late pulmonary hypertension in extremely premature babies

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Introduction: Persistent pulmonary hypertension (PHT) is a common complication in premature babies whether or not in association with bronchopulmonary dysplasia. Unfortunately, due to the difficulties in the diagnosis of PHT in premature babies little is known about the true prevalence of this condition and its evolution during the growth of the baby, a notion that is useful in the proposal of screening programs and eventual initiation of a treatment for such population.
Methods: In this prospective study we enrolled all premature babies born at < 30 weeks of gestational age at S. Orsola – Malpighi hospital in Bologna. Exclusion criteria were the presence of diaphragmatic ernia, congenital heart disease other than ASD and PDA and genetic disease. All patients were evaluated with echocardiography at 10 days of life, 32 and 40 W of post-conceptional age and at 3, 6 and 12 weeks of corrected age. Presence of PHT was defined as the presence of any of the following findings: estimated right ventricular systolic pressure (RVSP) > 40 mmHg, any cardiac shunt with bidirectional or right-to-left flow, RV/LV diameter in systole in short axis > 1, Exentricity index > 1,4 or End-dyastolic RVarea/End-dyastolic LVarea in 4 chamber view > 1.
Results: 30 patients (56% males) were included in the study. The prevalence of echocardiographic parameters compatible with pulmonary hypertension was 83,3% at birth, 20% at 32 weeks, 50% at 40 weeks, 0% at 3 and 6 moths and 80% at 12 months of corrected age. All patients were asymptomatic during the follow-up.
Conclusions: The prevalence of PHT shows a bimodal curve with a first peak after birth, probably due to a delayed reduction in pulmonary vascular resistance and a second peak at about 12 months of corrected age. This late recurrence suggests that if a screening for PHT in preterm babies is to be successful should be performed at about this age to minimize false positive and negative results.

P168

Safety of ambulatory sedation in cardiovascular magnetic resonance and computed tomography of complex congenital heart disease in infants and children

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Introduction: In infants and young children good image quality in cardiovascular magnetic resonance (CMR) and computed tomography (CT) may generally be achieved by using sedation or general anesthesia to avoid motion artifacts. The aim of this study was to determine the safety of ambulatory sedation in an outpatient setting for CMR and CT as a feasible alternative to in-hospital management.
Methods: We analyzed 102 consecutive examinations with ambulatory sedation between 2017 and 2020. All patients had a congenital heart disease (CHD) and received a cardiac examination by CMR or CT. We investigated the kind of CHD, vital parameters, applied sedatives, adverse events during or after ambulatory sedation and overall success rate.
Results: Eighty-four patients under 6 years of age (median 29, range 1 to 70 months; 37% female) were finally analyzed. Sixty-five percent were classified as ASA class 4 (American Society of Anesthesiologists Risk Classification), 26% as ASA class 3 and 8% as ASA class 2. Ambulatory sedation was performed by using midazolam, propofol or combinations with s-ketamine and/or clonidine. The average sedation time for CMR (71 patients) was 105 minutes, which was longer than for CT (13 patients) with a sedation time of 91 minutes. The average durations for the CMR and CT examinations were 60 and 21 minutes, respectively. Oxygen saturation before sedation was median 96% (range 54 to 100%). One patient (age 1.5 months; ASA 2) was admitted for in-hospital observation due to unexpected severe airway obstruction caused by bronchial secretions. Another patient (age 17 months; ASA 4) was admitted to ICU after receiving CPR due to airway obstruction and cyanosis during induction of sedation. Both patients were discharged after one and 3 days, respectively. All other patients were sent home after an observational period of a few hours on the same day.
Conclusions: Sedation of infants and young children with CHD for CMR or CT can be carried out routinely in an outpatient setting. In-hospital backup should be available for unexpected events at all times.

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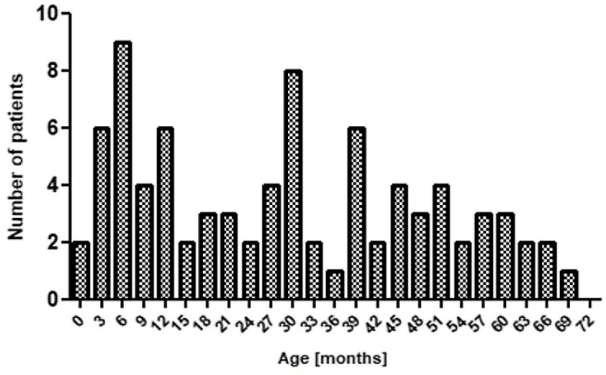

Figure 1. Age distribution of the patients

P169

Safety of intrahospital transport for MR or CT scans in ventilated pediatric intensive care patients with congenital heart disease

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Introduction: MR or CT scans are often required in the treatment of pediatric intensive care patients. Therefore, an intrahospital transport is needed, where continuous ventilation of the patient must be maintained. Literature considers intrahospital transport safe regarding changes in hemodynamics or adverse events (AE). As those studies cover inhomogeneous patient groups, we analyzed the safety for ventilated pediatric patients with congenital heart disease (CHD) focusing on differences between manual and mechanical ventilation during transport and examination.
Methods: Retrospective monocentric study covering a 10 years' period in a tertiary cardiac center for CHD. Sixtyone critically ill ventilator-dependent patients, median 2 (0 – 37) months, underwent intrahospital transport from the ICU to the department of radiology back. Fifty patients got ventilated manually and 11 patients got ventilated with a high-end MR-conditional mobile ventilator. The data include vital parameters and blood gas data before leaving and after returning to the ICU, as well as catecholamine support, occurrence of AE and total duration of transport and examination.
Results: In both groups we found small changes of vital parameters or blood gas data. There was a slight increase in lactate levels from median 1.8 (0.3 – 12.2) to 1.9 (0.5 – 19.0) mmol/L in the manually and a small decrease from median 1.1 (0.5 – 8.7) to 1.0 (0.5 – 6.2) mmol/L in the mechanically ventilated group. The partial arterial pressure of carbon dioxide showed an unremarkable decrease from median 49.5 (30 – 75) to 48.0 (28 – 89) mmHg in the manually and from 53.0 (34 – 70) to 48.0 (37 – 66) mmHg in the mechanically ventilated group. The aforementioned changes were statistically not significant. The median transport time including scan was significantly higher in the manually ventilated group (p-value 0.02). A total of 11 AE was recorded, 9 (20%) of them in the manually and 2 (18%) in the mechanically ventilated group with a drop of the mean arterial pressure and an increase in catecholamine support.
Conclusions: We consider both manual ventilation and mechanical ventilation for intrahospital transport safe for pediatric intensive care patients with CHD. Using a mechanical ventilator might have the advantage to react faster to changes in hemodynamics.

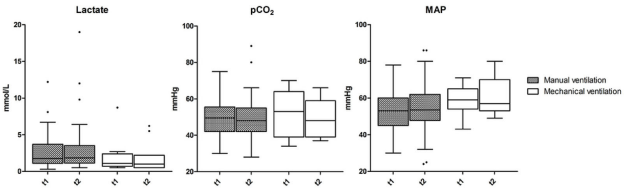

Figure: Box plot (Tukey). Lactate, partial pressure of carbon dioxide (pCO_2) and mean arterial blood pressure (MAP) before (t1) and after (t2) transport and examination. Manual ventilation (n=50), mechanical ventilation (n=11).

P170

TGA repaired with Neonatal Arterial Switch: heart changes observed at school stage with morphometric and functional echocardiographic parameters

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Introduction: The increase in the survival of patients with transposition of the great arteries (TGA) repaired with arterial switch has produced greater morbidity in the long term, so current efforts are focused on improving it by early diagnosis of possible complications. Although echocardiography is the most commonly used image diagnosis method in children, the echocardiographic study of cardiac functionality parameters in children operated on congenital heart defects is very scarce and with heterogeneous results. The present study seeks to detect and describe in children operated on for arterial switch, echocardiographic changes that could affect the prognosis, also seeking to reflect on their possible causes.
Methods: Prospective observational case-control study in patients aged 1 to 5 years with TGA operated without complications in the neonatal period, and without subsequent incidents. Morphometric and functional echocardiographic parameters were analysed in 21 patients and compared with 52 healthy controls with similar characteristics.
Results: Echocardiographic findings: Morphometry: the left atrium was smaller (sfericity 1,4 vs 1,2;*p=0,048*; volume 8,7 vs 10,9 ml; *p=0,015*) and right ventricle more globulous (basal sfericity 1,5 vs 1,8;*p=0,016*). Functionality: Systole parameters were lower, in RV (FAC 51 vs 58%;*p=0,006*; TAPSE 13 vs 20 mm;*p=0,001*; s' 7 vs 12 cm/s;*p=0,001*) and LV (MAPSE 11 vs 13 mm; *p=0,001*; VFC 0,154 vs 0,17;*p=0,006*). Septal parameters showed lower values also (SAPSE 9 vs 12 mm;*p=0,001*; s'5 vs 7 cm/s; *p=0,001).* Diastolic parameters showed inferior values in TDI-PW (RV e' 12 vs 15 cm/s;*p= 0,001,* a' 5 vs 10 cm/s;*p= 0,001*; LV lateral e' 14 vs 16 cm/s;*p=0,017,* a' 4 vs 7 cm/s;*p=0,001*) and higher RV transvalvular Doppler velocity (E 82 vs 67 cm/s; *p=0,001*; A 58 vs 47 cm/s;*p= 0,004*) resulting in a higher E/e' ratio (6.7 vs 4.5;*p=0,001*). STE study showed a decrease in the longitudinal deformation of the apical septal myocardium (-23% vs -27%;*p=0,005*).
Conclusions: After a few years of the arterial switch, morphometric and functional echocardiographic changes can be detected, especially in the RV. Presurgical systemic overload could be an important factor in its origin. Knowing the long-term-effect of these changes on the prognosis of patients will require more studies.

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	TGA	Controls	p
	(n=21)	*(n=52)*	
HF (bpm)	93 ± 21	100 ± 16	0,082
RA sphericity	1,1 ± 0,18	1,0 ± 0,15	0,061
RV basal sphericity	1,5 ± 0,34	1,8 ± 0,26	0,016
LA sphericity	1,4 ± 0,25	1,2 ± 0,22	0,048
LV basal sphericity	1,6 ± 0,37	1,72 ± 0,18	0,227
FAC(%)	51 ± 7	58 ± 7	0,006
TAPSE(mm)	13 ± 2	20 ± 4	0,001
RV s'(cm/s)	7 ± 1	12 ± 2	0,001
Tricuspid E(cm/s)	82 ± 11	67 ± 14	0,001
Tricuspid A(cm/s)	58 ± 14	47 ± 12	0,004
RV e'(cm/s)	12 ± 2	15 ± 3	0,001
RV a'(cm/s)	5 ± 2	10 ± 3	0,001
RV E/e'	6,7 ± 1,6	4,5 ± 1,3	0,001
EF Simpson(%)	63 ± 5	64 ± 6	0,734
MAPSE(mm)	11 ± 2	13 ± 2	0,001
LV s' lateral(cm/s)	7 ± 1	7 ± 1	0,069
VFC¥	0,154 ± 0,05	0,17 ± 0,04	0,006
Mitral E(cm/s)	96 ± 24	101 ± 13	0,289
Mitral A(cm/s)	56 ± 16	65 ± 13	0,003
LV e' lateral(cm/s)	14 ± 2	16 ± 3	0,017
LV a' lateral(cm/s)	4 ± 1,4	7 ± 1,7	0,001
SAPSE(mm)	9 ± 1	12 ± 2	0,001
Septal s' (cm/s)	5 ± 1	7 ± 1	0,001
TCIV septal(ms)	76 ± 18	62 ± 11	0,001

P171

The diagnostic value of 4Dflow CMR in the clinical evaluation of children with congenital heart defects

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Introduction: Clinical blood flow measurements using short free breathing 4Dflow CMR in 3-dimensions over time are now possible for clinical evaluation of children with congenital heart defects (CHD). From one single measurement, several different types of images can be reconstructed that might be useful in the diagnostic evaluation. 4Dflow also offers new advanced hemodynamic measures such as wall-shear stress, pulse-wave velocity, pressure difference, turbulent kinetic energy, as well as eccentricity that have all been suggested as risk markers for patients with congenital diseases. The aim of this work was to study the value of short free breathing 4Dflow CMR for diagnostic evaluation of children with CHD in our clinic.
Methods: Patients with CHD referred for CMR were consecutively included. As a complement to conventional clinical examination, a very short (~9 min) free breathing 4Dflow measurement was added. 4Dflow-data was collected from a large volume, including the heart and the great vessels. For each patient, the improved diagnostic value of 4Dflow was determined and comparison with conventional 2Dflow was performed.
Results: 4Dflow was successfully performed with high image quality in all patients that could lay still during the examination (down to 5 years without sedation and 2 years with sedation). 4Dflow strengthened the diagnosis in all cases and in some cases, even revealed new diagnostic information of importance for the patient handling (one example is shown in Fig.1). The unique possibility to screen blood flow within the whole vasculature and entire cardiac cycle enabled retrospective reconstruction of images in preferred positions and angulations for improved diagnostics, e.g. in patients with stenotic jets for peak flow estimation at the vena cava and insufficient valves for regurgitation estimation in areas of less disturbed flow. Also, 4Dflow agreed well with conventional 2Dflow (CVpeak<0.2% and CVflow<10%; linear regression: 4Dflowpeak=2Dflowpeak+2.5 with R^2peak=0.95 and 4Dflowflow=0.9★2Dflowflow-0.3 with R^2flow=0.99).
Conclusions: For children with CHD, short free-breathing 4Dflow presents visualizations of hemodynamic important flows that improves the clinical decision-making and offers reliable hemodynamic measures in agreement with conventional 2Dflow. Moreover, the possibility to freely choose the position and angulation of the slice retrospectively for analysis was shown to improve the diagnostic flow measurement.

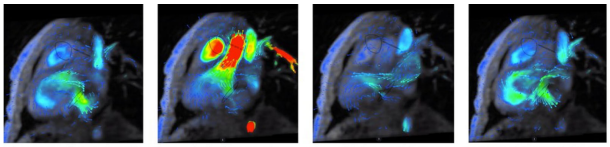

Fig.1: 4Dflow visualization of the heart of a 4- year old boy with severe neonatal heart failure. Early CMR displayed an aneurysm-like structure (ALS) anterior of the left ventricle, initially determined to be an aneurysm. In a follow-up CMR, with better visualization of the ALS by 4Dflow, the pattern was shown to be associated with a double chambered left ventricle with good contraction and emptying of blood from the structure but without effect on the elliptical shape or function of the main left ventricle. The clinical decision-making was facilitated by this information, and with an almost complete emptying, there was no need for operation at this juncture. Hence, 4Dflow improved the diagnostic value in this patient.

P172

The impact of heart failure on retinal vascular density assessed by optical coherence tomography angiography in children with dilated cardiomyopathy.

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Introduction: Dilated cardiomyopathy (DCM) is a significant cause of acute or chronic heart failure (CHF), sudden cardiac death and the most common indication for heart transplant in childhood. In response to reduction of the LV function, individual organs, including the eye and its structures, receive an insufficient supply of oxygen in relation to demand. It is assumed that the condition of retinal microcirculation is an easily measurable substitute for coronary circulation and can be equivalent to that of blood vessels throughout the body.The aim of study was to assess retinal vessel density (VD) in the superficial capillary plexus layer (SP) and deep capillary plexus layer (DP) in children with CHF due to DCM using optical coherence tomography angiography (OCTA).
Methods: Thirty children, mean age 9.9±3.57yrs with CHF and LVEF ≤ 55% lasting more than six months were enrolled. The control group consisted of 30 children without CHF (mean age 11.27±3.33yrs) matched for age and gender against the study group. In all participants transthoracic echocardiography and

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blood serum for NT-proBNP analysis were performed. All children underwent OCTA with evaluation of the foveal avascular zone (FAZ), whole superficial vessel density (wsVD), foveal superficial vessel density (fsVD), parafoveal superficial vessel density (psVD), whole deep vessel density (wdVD), foveal deep vessel density (fdVD) and parafoveal deep vessel density (pdVD).
Results: Retinal VD in SP in the study group were significantly lower in children with CHF as compared to the controls: wsVD (46.2%vs.49.83%,p<0.05), fsVD (18.07%vs.24.15%, p<0.05), psVD (49.24%vs.52.51%,p<0.05). No significant differences in DP retinal VD and FAZ were found between the groups. The fsVD were correlated with the age of the study participants. There were no correlations between retinal VD in both plexuses and sex, NT-proBNP, and LVEF.
Conclusions: In children with CHF in the course of DCM as compared to the control group significant decrease in SP retinal VD was observed. The results of our study indicate that measurements of the OCTA may be a useful diagnostic method in children with CHF, but it is necessary to further studies in larger group of participants and long-term observation of these patients.

P173
The impact of the genetic syndrome on the outcome of Tetralogy of Fallot

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Introduction: Tetralogy of Fallot (ToF) is the most common cyanotic congenital heart defect (CHD) and it is often associated to genetic syndromes and extracardiac anomalies. Little is known on the impact of the most frequent syndromes associated to ToF on its outcome, although the long-term consequence of the chronic pulmonary regurgitation due to the repair has been extensively studied for its impact on the survival in this population. Therefore, our aim was to assess the correlation among clinical (ventricular arrhythmias, QRS duration), surgical and cardiac magnetic resonance imaging (MRI) data in syndromic repaired ToF (rToF) and compared their outcome to rToF patients without syndrome.
Methods: Fifty-four syndromic rToF patients were selected and compared with ones without syndrome selected from our cohort matched for year of birth, age at MRI, sex and BSA. Demographic, surgery, echocardiographic, MRI, ECG and Holter data were collected for each patient.
Results: Del22 and Trisomy 21 were the syndrome more frequent in our cohort (42% and 31% respectively). The palliation was more frequent in the syndromic group (25% vs 9,6%, p=0.038), whereas a patch infundibular in the non-syndromic one (13,5% vs 1,9%, p=0.027). However, no significant differences were found between the two groups in MRI parameters, except for higher values of LVEDVi in the patients without syndrome (78±19 vs 83 ±14, p=0.018). Nevertheless, syndromic patients was younger at PVR (15,9 yrs±5,6 vs 19,5 yrs ±6, p=0.049). Significant differences were found between patients with del22 and Trisomy on LV EF, pulmonary regurgitation, RV systolic pressure, QRS and QTc duration (see table 1).
Conclusions: The presence of the syndrome don't seem to have a significant impact on the arrhythmias and MRI parameters. However, the syndromic patients are younger at PVR. The presence of Down syndrome may have an negative impact major on LVEF, pulmonary regurgitation and RV systolic pressure.

	del22 syndrome	21 trisomy	p value
LVEF (%)	56±7	51±9	0.042
MPA RF (%)	40±16	51±9	0.043
RVSP (mmHg)	37±10	54±10	0.011
QRS (msec)	133±26	115±22	0.021
QTc (msec)	421±36	398±26	0.021

Table 1. Main significant differences between del22 and Trisomy 21 syndromes. LVEF: Left ventricle ejection fraction; MPA RF: Main pulmonary regurgitation fraction; RVSP: Right ventricle systolic pressure.

P174
The role of cardiovascular magnetic resonance in evaluation of idiopathic ventricular arrhythmia in children

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Introduction: The utility of advanced non-invasive imaging methods in the evaluation of idiopathic ventricular arrhythmia (VA) in children has not yet been analyzed. The aim of the study was to assess the role of cardiovascular magnetic resonance (CMR) in the diagnosis of idiopathic VA in children.
Methods: A single-center retrospective study enrolled consecutive children with VA and coinciding equivocal baseline tests, referred for CMR for further evaluation of disease etiology. All patients underwent a 3.0T scanning involving balanced steady state free precession cine images as well as dark-blood T2W images and assessment of late gadolinium enhancement (LGE).
Results: Ventricular arrhythmia was the reason for 20.5% (n=80) out of all the CMR referrals (n=390). The mean age of the studied group was 13.1±3.6 years and 60% were male. In 26% of patients (n=21) CMR revealed cardiac abnormalities, of which 20% (n=16) were not suspected on prior echocardiography. The main findings included: non-ischemic ventricular scars (n=8), criteria for arrhythmogenic right ventricular cardiomyopathy (n=6), single or multiple left ventricular clefts (n=4) and active myocarditis (n=3). There was no difference in left and right ventricular volumes, ejection fraction or mass between children with and without abnormal CMR findings. LGE was present in 57% of patients with abnormal findings. Univariate predictors of abnormal CMR result included abnormalities in echocardiography and severe VA (combination of >10% of 24h VA burden and/or presence of ventricular tachycardia and/or polymorphic VA).
Conclusions: CMR provides valuable clinical information in many cases of idiopathic ventricular arrhythmia in children, mainly due to its potential to detect the fibrosis and assessment of right ventricle.

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P175

Tricuspid valve atresia with transposition of the great arteries, abnormalities of the aortic arch and double "arterial duct" – a rare combination

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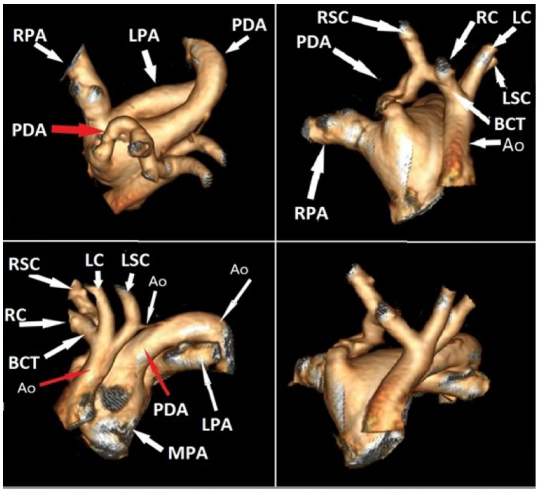
Introduction: Tricuspid atresia (TA) represents 1 – 5% of all congenital heart diseases(CHD). TA associated with transposition of the great arteries (TGA) is rare an its association with aortic arch abnormalities is Known, particularly with a restrictive ventricular septal defect (VSD), as a result of too little systemic flow.
Methods: Case report based on detailed analysis of medical records.
Results: We present a case of a 2- month -old baby boy, born at term, by vaginal labour, with a birth weigth of 3.18 kg. His Apgar 1' and 5' was. 9. His fetal echocardiography was able to diagnose a complex CHD - TA type II-B with a restrictive ventricular septal defect (VSD) and interrupted aortic arch. His postnatal transthoracic 2D echocardiogram showed: Situs solitus, univentricular atrioventricular connection due to atresia of the right atrioventricular valve; Ventriculoarterial discordance; restrictive VSD, very dilated main pulmonary artery ; Hypoplastic ascending aorta with severe hypoplasia of the transverse arch and isthmus, a huge arterial duct maintaining systemic circulation. In order to clarify, his aortic arch anatomy an angiotomography was done, which showed a left aortic arch with severe preductal aortic coarctation, and two "arterial ducts". One to the left arising from its usual place from the pulmonary artery to descending, and another to the right arising from the right subclavian artery to the main pulmonary artery. On his 24th day, of life he underwent to a hybrid procedure for banding his pulmonary arteries and stenting his arterial duct stent The angiotomography anatomical findings were confirmed by open surgery. The baby is doing well, with good weight gain, awaiting for his next step for the planned treatment, a Damus – Kaye – Stansel surgery and aortic arch reconstruction.
Conclusions: The description of this rare and complex congenital heart disease confirm the importance of an earlier diagnosis and also the use of multiple image modality to define precisely the anatomy for planning which allow a better management strategy.



Figures 1,2,3 and 4: Reconstruction from angiotomography before hybrid procedure. Ao, aorta; PDA, patent ductus arteriosus; RPA, right pulmonary artery; LPA, left pulmonary artery; MPA, main pulmonary artery; RSC, right subclavian artery; LSC, left subclavian artery; BCT, brachiocephalic trunk; RC, right carotid artery; LC, left carotid artery.

P176

Volumetric ECG-gated computed tomography in the assessment of coronary arteries in neonates and infants with heart diseases

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Introduction: Computed tomography angiography (CTA) is one of the imaging methods used in the assessment of complex congenital heart diseases (HD) but detailed visualization of coronary arteries (CA) in neonates and infants is a challenge. We aimed to evaluate CA image quality on volumetric, 320-row, ECG-gated CTA in neonates and infants with congenital or acquired HD.
Methods: CA image quality was assessed in 4-point scale and compared between the group with good image quality of all CA segments and group with at least one non-diagnostic CA segment. Statistical analysis was performed to define predictors of good image quality.
Results: A total of 110 CTA performed in 37 girls and 73 boys; 37 neonates and 64 infants of mean age 3.5 ± 3.08 months, mean body weight 5.31 ± 1.97 kg, with the mean HR 134 ± 21 beats/min. All examinations were performed free breathing, in 30% no sedation was used. The mean CA score was 2.2 ± 0.74 points. The orifices of left CA were visible in 100% of CTA, orifices of right CA in 96%, all segments in 45%. Patients with non-diagnostic segments were younger (2.75 ± 2.79 months), of lower body weight (4.93 ± 2.05 kg), and faster HR (137.75 ± 20.22 beats/min) ($p < 0.05$). Age and body weight significantly influenced CA image quality (OR 1.20, 95% CI: 1.05 – 1.37, $p = 0.004$ and OR 1.25, 95% CI: 1.02 – 1.53, $p = 0.024$, respectively), HR showed only a trend (OR 0.98, 95% CI: 0.96-1.00, $p = 0.057$). Sedation significantly influenced image quality. Radiation doses showed no significant differences and were similar to reported in literature and to suggested pediatric diagnostic reference levels. The optimal cut-off points for good quality CA image for age, body weight and HR were 0.53 months, > 4.85 kilograms, and ≤ 130/min, respectively. Body weight was the strongest predictor of good image quality.
Conclusions: ECG-triggered CTA allows for good visibility of the CA in neonates and infants with HD. Children aged > 15 days, with body weight > 4.85kg and HR <130/min are suitable candidates for good quality computed tomography coronarography.

P177

"When things are not as expected" – a rare case of persistent respiratory distress in a newborn:

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Introduction: Respiratory distress is common in neonates and it occurs in up to 7% of neonates. Multiple conditions can present with features of respiratory distress including transient tachypnoea of the newborn. Pneumonia and meconium aspiration syndrome can cause persistent pulmonary hypertension of the newborn. Primary cardiac problems rarely present with respiratory distress immediately after birth. We present a term baby with antenatal cardiac diagnosis which, although on it is own non-significant, but it turned out to be associated with serious airway compromise.

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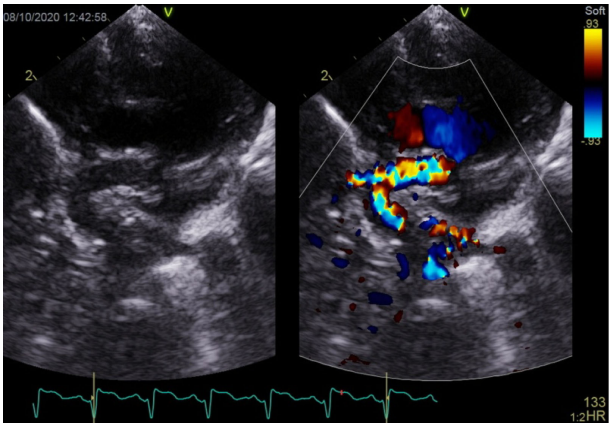
Methods: The clinical data including history, examinataion and images of a term baby were reviewed. The baby presented to the neonatal unit with respiratory distress. She had an antenatal diagnosis of bilateral superior vena cavae (SVC), with left SVC draining to coronary sinus (CS), small muscular ventricular septal defect (VSD) and unilateral renal agenesis. Antenatal plan was for outpatient follow-up as she was expected to be cardiovascularly stable. Chest X ray showed the heart in the right chest. Baby failed to progress and could not be weaned from respiratory support.
Results: In view of the progressive respiratory symptoms which were not fitting with the prenatal diagnosis the baby was transferred to a tertiary centre for assessment. Echocardiography confirmed antenatal findings however, in addition there was evidence of a left pulmonary artery (LPA) sling (figure 1) which was confirmed on computed tomography (CT) Bronchoscopy and CT confirmed complete tracheal rings with distal tracheal and bronchial stenosis. Baby was then transferred to the National Tracheal Service for surgical intervention.
Conclusions: LPA sling is a rare congenital anomaly which can be associated with airway malformations causing respiratory distress. Although it has been described prenatally, LPA sling is rarely diagnosed before birth. This case highlights that if differences are identified prenatally, there may be other serious problems that cannot be diagnosed on prenatal scans. It is important to perform a thorough assessment of the neonate particularly if the clinical condition does not match the prenatal diagnosis.



7. Interventional cardiology

A case series of disconnected pulmonary arteries; anatomical and technical considerations

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Introduction: Disconnected pulmonary arteries are frequently fed by an aberrant ductus (PDA) from the proximal truncus brachiocephalicus. After ductal closure, the disconnected artery cannot be echocardiographically or radiologically visualized. Pulmonary venous-wedge angiography can delineate anatomy. In early childhood the duct can be recanalised, facilitating growth to allow for future re-implantation. We present four cases and illustrate anatomical and technical considerations.
Methods: A cases series is presented.
Results: Patient 1: infant, 5 months, ex-dysmature, 22q11 deletion, with left lung hypoplasia and right-descending arch. The left pulmonary artery (LPA) was discontinuous and had a small connection to the truncus brachiocephalicus, which was stented with two overlapping Onyx stents (4,5x15mm and 5x15mm).Re-implantation followed 11 months later. During 1 year follow-up the anastomosis remained unobstructed. Patient 2: infant, 9 months, discontinuous RPA hair thin connection to the truncus brachiocephalicus. Balloon dilation with subsequent stenting with a Cook Formula 5x20mm. 6 months later the RPA was surgically re-implanted using homograft material. 8 months later, balloon angioplasty of the anastomosis was needed. Patient 3: 4d old neonate after resuscitation at home. She showed right-sided pneumothorax, and a yet unclarified phenotype of this lung. The RPA could not be visualized on CT, but this suggested a PDA-ampulla at the proximal truncus brachiocephalicus. RPA was confirmed by pulmonary venous wedge-angiography. Subsequent power-injection into the aortic ampulla revealed a miniscule vessel, connecting the RPA, which was stented using two overlapping Rebel stents (3,5x8mm and 3,5x12mm). The patient is currently awaiting surgery. Patient 4 was diagnosed with "absent" LPA, in the presence of a right arch and a hypoplastic left lung. CT at the age of 13 years suggested a small ampulla at the proximal truncus brachiocephalicus. A hypoplastic LPA could be revealed by pulmonary wedge-angiography, after transseptal puncture. Recanalisation was not possible.
Conclusions: In conclusion, disconnected pulmonary arteries can be misdiagnosed as "absent". They are a distinct anatomic entity with perfusion by a duct-like vessel, typically from the proximal truncus brachiocephalicus. If antegrade angiography is unsuccessful, pulmonary venous-wedge angiography can be performed. Prompt diagnosis is crucial, since early canalization allows for growth and subsequent re-implantation.

P178
Immediate results of arterial duct stenting in duct-dependent pulmonary flow and comparison with Blalock-Taussig shunt in a Brazilian Hospital

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Introduction: Patients with duct-dependent pulmonary circulation still a challenge, particularly in neonates who are unsuitable for primary repair. These patients require palliative procedure, either surgically by construction of systemic- to- pulmonary artery shunt (Blalock-Taussig shunt-BTS) or percutaneously by cardiac catheterization with stent implantation in the arterial duct. Objective: To evaluate the efficacy of stenting arterial duct versus BTS in patients with duct-dependent pulmonary circulation
Methods: Retrospective, longitudinal and unicentric study was performed that included all 27 patients with duct-dependent pulmonary circulation, admitted to our Hospital between November 2016 to October 2020. They were divided into two groups. **G1 (Surgery for BTS)** and **G2 (Duct stenting)**. BTS initially was our first choice as a palliative procedure.
Results: G1 – 14 patients (52%) vs G2 – 13 patients (48%). A median age in **G1** was 15 days (1d – 4mo) vs 14 days (1d – 3mo) in **G2.** There was male predominance in both groups, in **G1** - 10 pts (71.5%) vs 8pts (61.5%) in **G2.** In **G1**, the most common diagnosis was tricuspid atresia (TA) with PS, 5pts (35.7%), followed by TOF – 4pts (28.5%), PA with VSD 3pts (21.4%) and PA/IVS, 2pts (14.3%). In **G2**, the most common diagnosis was PA/IVS 5 pts (38.5%), TOF and PA/VSD 2pts each

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(25%). Others diagnosis were Critical PS, DORV and DILV with PS. The average for pulmonary mechanical ventilation time was 5 days in **G1** vs 1.8 days and hospital stay was 17.5 days vs 10.3 days in **G2**. All patients in **G1** presented complications, the most commom was sepsis in 5pts (35.7%). There were 6 deaths (42.8%) in **G1**. In **G2**, all procedures were successful, 8 pts (61.5%) had evolution without any complication, there was no death. Sepsis, HF and post-extubation laryngitis were rarely observed.
Conclusions: Arterial duct stenting did not present acute serious complications, neither surgical crossover, and should be considered as a safe alternative to maintain pulmonary blood flow. Blalock -Taussig shunt in our institution has a high rate of hospital mortality and morbidities, including prolonged time of pulmonary mechanical ventilation and hospital stay.

P179

10-Zig Cheatham-Platinum stents in congenital heart disease: a new tool to widen the spectrum of potentially treatable disease.

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Introduction: This case series details our experience with the new 10-zig Cheatham-Platinum stents in congenital heart disease (CHD).
Methods: Between 2015 and 2020 there were nineteen patients. Six females, median weight 84 kg (25-81 kg), median age 22 years (10-61 years). Thirteen males, median weight 86 kg (69-119 kg), median age 44 years (22-69 years). MHRA approval was required for use of the stents.
Results: Ten, 10-zig CP stents were used in eight patients in the RVOT site (two also had implantation of Sapien venous valves). Median weight 84 kg (25-112 kg), median age 27 years (10-69 years). Indications included; pulmonary homograft stenosis after Ross procedure (n=4), post PA/VSD repair (n=1), RVOT lesions following Tetralogy of Fallot repair (n=5). Stent lengths were 5-6 cm. Median sheath size 20 Fr (6-26 Fr). Median final stent diameter 20 mm (16-29 mm). Median fluoroscopy time 41 mins (31-131 mins), procedure time 164 mins (152-361 mins). There were no residual RVOT gradients. There were two stent fractures and one femoral vein intimal injury conservatively managed. There was one parenchymal lung injury. At a median follow up of 15 months (4- 60 months) there were no significant complications. Nine 10-zig stents were used in nine patients with aortic coarctation. Median weight 88 kg (71-119 kg). Median age 44 years (22-61 years). Indications included native (n= 8) and homograft/re-coarctation (n= 1). Stent lengths were 5-7 cm. Median sheath size 20 Fr (16-20 Fr). Median final stent diameter 21 mm (14-25 mm). Median fluoroscopy time 36 mins (11-215 mins), procedure time 164 mins (95-225 mins). Six stents required flaring or further dilation at the time of the procedure. Median systolic BP pre-intervention 160 mmHg (110-180 mmHg). Post intervention 132 mmHg (114-140 mmHg). Median peak systolic gradient pre intervention 33mmHg (15-60 mmHg), post intervention 3 mmHg (0-4 mmHg). There was one femoral artery spasm. At a median follow up of 14 months (1-60 months) there were no significant complications.
Conclusions: This is the largest case series documenting an extended range of applications for the 10-zig Cheatham- Platinum stents. This demonstrates their safe and effective use in paediatric and adult populations.

P180

A Rare Case of Pulmonary Arteriovenous Malformation Diagnosed While Examining Polycythemia

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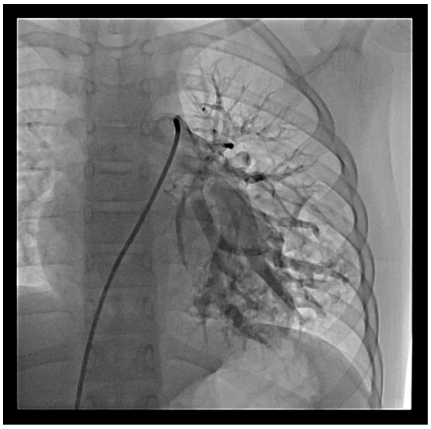
Introduction: Pulmonary arteriovenous malformations (PAVMs) are abnormal direct connections between the pulmonary artery and pulmonary vein which may result in a right to left shunt. Clinical signs vary according to the amount of shunt in proportion to the number and size of the fistulae. Patients may present with cyanosis.
Methods: A 4-year-old Syrian female patient has been followed by family medicine with the diagnosis of polycythemia. The patient referred to pediatric cardiology unit because of clubbing and decreased level of oxygen saturation. Physical examination showed a well-developed, nondysmorfhic child. Blood pressure was within normal range for age, heart rate was 110 beats/min, respiratory rate was 28 breaths/min. There was bluish discoloration of her oral mucosa. The cardiovascular examination revealed a regular rhythm with a normal S1 and S2. There were no murmurs, rubs or gallops. Her abdomen was soft and nondistended, with no organomegaly or mass. Her extremities were warm with no edema. However, there was clubbing of her fingers and toes. Pulse oximetry showed the oxygen saturation to be 75% breathing room air. Hemoglobin was 18,7 g/dL, and Hematocrit 57%. On chest X-ray a hyperdense area with irregular contours on medial basal region of the left lung was seen. Her electrocardiographic, and echocardiographic examination findings, and all routine laboratory analyses yielded normal results. Thoracic CT angiography was performed on the patient and pulmonary arteriovenous malformation (PAVM) was detected.
Results: Catheter angiography was performed. In the selective contrast injection into the left pulmonary artery, it was observed that there were at least three major PAVMs(Figure) The fistulas were closed with 3 vasculer plugs, sized 10, 12, and 14 mm, respectively. Transcutaneous oxygen saturation raised immediately to 96% after the procedure. No post procedural complication was seen, and the patient was discharged one day after the procedure
Conclusions: PAVM should come to mind as a rare cause of hypoxemia and cyanosis. Even if physical examination, chest X-ray and echocardiography are normal, patients should be evaluated with chest CT angiography. When the diagnosis of this very rare disease is delayed, heart failure may develop. In treatment, transcatheter embolization can be successfully applied.



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P181
Atrial flow regulator for drug resistant pulmonary hypertension in children

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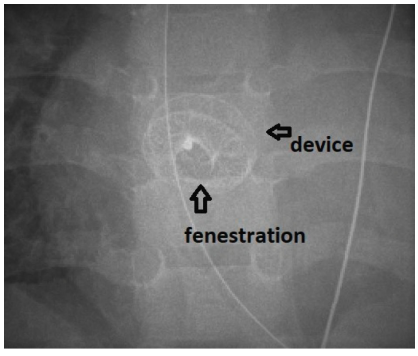
MD (1), PhD (2)

Introduction: Pulmonary arterial hypertension (PAH) is a rare but life threatening condition in children. Compared to adults, right heart failure is a late presentation and episodes of severe PAH with syncope are more common in children. In drug resistant patients with progressive PAH, elevated atrial pressure and low cardiac output, atrial septostomy should be highly considered as well as in patients with persistent PAH-related syncopes despite full medical treatment. Treatment options for a permanent atrial septostomy are creating and stenting an atrial septum defect with risk of stent migration and occlusion, or implantation of a custom made fenestrated ASD device complicated by a high occlusion rate. In adults the recently introduced Occlutech Atrial Flow Regulator® is used with success for pulmonary hypertension.
Methods: Case report
Results: This case describes the successful implantation of a 6 mm fenestration Occlutech Atrial Flow Regulator® in compassionate use in a young child with idiopathic pulmonary hypertension and recurrent syncope despite targeted combination therapy. The device placement and deployment were a lot easier than stent implantation. Clinical results were satisfying with disappearance of clinical symptoms.
Conclusions: Implantation of the AFR device is proven to be a safe and effective percutaneous procedure for treatment of severe and drug-resistant pulmonary hypertension in adults. The small size AFR devices are not yet CE approved and data of the use in children are scarce. In this case, we show the successful percutaneous implantation of an AFR device in a small child with idiopathic pulmonary hypertension and syncope with subsequent complete relief of symptoms.



P182
Cardiac mechanical remodeling after ASD closure with Gore® Cardioform ASD Occluder in children

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Introduction: Nowadays, trans-catheter closure of ostium secundum atrial septal defect (ASD) is considered as the first-line treatment**.** Previous studies demonstrated that ASD devices may impact on cardiac mechanics. The GORE® CARDIOFORM ASD Occluder (GCA, WL Gore & Associates, Flagstaff, AZ) is a recently approved double-disc, soft, conformable device, potentially innovative compared to other self-centering devices. It combines high softness and anatomic compliance with the potential to close defects as large as 35 mm. This study aims to evaluate the changes in atrial and ventricular mechanical properties after implantation of GCA device.
Methods: Between January and June 2020, 16 consecutive pediatric patients with hemodynamically-significant ASD were enrolled to undergo transcatheter closure with the GCA device. Standard transthoracic echocardiography (TTE) was performed the day before the procedure, 24 hours, 1 and 6 months after ASD percutaneous closure. Atrial and ventricular 2D speckle tracking analysis was performed off-line by the use of dedicated software. Left ventricle (LV) longitudinal, radial and circumferential strain (S) and strain rate (SR), right ventricle (RV), right and left atria (RA, LA) longitudinal S and SR were assessed from apical and short axis views.
Results: The population mean age was 8.7±2.6 years, and mean defect diameter was 14.9±4.6 mm. The rate of successful closure was 87.5% (14/16) at 24 hours and 100% at 6 months (0/14). One patient experienced transient supraventricular tachycardia. Longitudinal, circumferential and radial S and SR analysis of the LV, and longitudinal S and SR analysis of the RV and RA didn't show any significant difference between time 0 and 6 months after ASD closure, although transient modifications were found at 24 hours and at 1 month. A reduction of LA septal segmental function was found after the procedure, with a gradual LA global function recovery at 6 months. (Table 1)
Conclusions: This study shows that myocardial deformation remains unaltered 6 months after GCA implantation, differently from previously reported studies after surgical ASD closure and after Amplatzer ASD device implantation. These data highlight the softness and compliance of this new device towards the myocardial walls.

Table 1 Strain (%) e Strain rate (1/sec) 2D speckle-tracking analysis.

		T0	24 h	1 mese	6 mesi®
LV	Average§	−22,1±1,8	−21,5±1,7	−22,0±1,8	−21,5±1,1
L-S	Bas AS	−19,9±3,6	−17,7±3,0*	−17,0±3,4*	−18,4±2,2
	Mid AS	−20,7±3,3	−19,9±3,1	−20,2±3,6	−20,7±2,7
	Bas PS	−19,0±1,6	−18,4±2,6	−19,3±2,6	−18,2±1,3*
	Mid PS	−20,6±2,8	−20,6±1,7	−21,0±2,5	−21,1±2,2
LV	Average§	−1,43±0,16	−1,44±0,21	−1,38±0,18	−1,32±0,09*
L-SR	Bas AS	−1,20±0,29	−1,23±0,17	−1,13±0,27	−1,27±0,23
	Mid AS	−1,35±0,24	−1,38±0,29	−1,23±0,22	−1,25±0,19
	Bas PS	−1,28±0,30	−1,17±0,22	−1,15±0,19	−1,14±0,23
	Mid PS	−1,30±0,17	−1,38±0,19	−1,27±0,19	−1,22±0,14
LV	Average§	−25,1±4,8	−26,7±4,1	−24,5±3,7	−23,8±2,7
C-S	Bas average	−25,1±4,0	−25,1±4,1	−23,7±5,0	−24,4±5,5
	Bas AS	−27,0±8,4	−27,0±6,0	−27,1±6,8	−29,3±4,4
	Bas PS	−25,3±5,4	−29,5±7,8	−25,2±8,0	−29,7±7,0*
LV	Average§	−1,95±0,29	−2,27±0,51*	−1,79±0,43	−1,78±0,34*
C-SR	Bas average	−2,13±0,6	−2,4±0,78	−1,59±1,0*	−1,82±0,49
	Bas AS	−2,22±0,81	−2,25±0,65	−1,66±1,05*	−2,07±0,48
	Bas PS	−1,84±1,1	−2,34±0,96	−1,6±1,02	−2,08±0,61

LV	Average§	38,7±11,6	33,6±11	37,9±10,7	37,7±1,7
R-S	Bas average	42,5±16,9	37,7±17,8	47,3±20,1	45,6±16,5
	Bas AS	39,0±20,1	34,0±16,3	37,3±17,6	38,0±16,9
	Bas PS	44,3±16,5	33,6+17,4*	47,3±25,4	45,6±20,5
LV	Average§	2,33±0,55	2,42±0,68	2,08±0,61	2,21±0,59
R-SR	Bas average	2,35±0,61	2,67±0,97	2,36±0,81	2,08±0,42
	Bas AS	2,40±1,05	2,38±0,99	2,12±0,73	1,80±0,50
	Bas PS	2,60±0,86	2,58±1,02	2,38±1,35	2,07±0,56
Bas rot (°)		−7,3±2,5	−6,8±2,7	−5,0±2,6*	−6,2±2,4
Ap rot (°)		8,1±3,6	8,5±3,3	6,1±4,5	6,9±2,6
Twist		10,7±3,9	11,9±4,5	8,6±3,4	9,5±2,3
RV L-S		−32,5±4,1	−27,5+4,8*	−29,2±3,7	−31,6±2,2
RV L-SR		−2,14±0,61	−1,83±0,41	−1,73±0,28	−1,91±0,20
LA L-S		53,3±14,7	36,6+9,2*	36,5+10,1*	43,5±10,1
LA L-SR		2,48±0,66	1,97+0,44*	1,87+0,45*	2,13±0,46
RA L-S		84,0±24,0	74,1±23,9	68,3+17,8*	76,9±27,0
RA L-SR		3,61±1,11	3,31±1,08	3,45±0,65	3,35±1,17

LV: left ventricle; RV: right ventricle; LA: left atrium; RA: right atrium; L: longitudinal; C: circumferential; R: radial; Bas: basal; Ap: apical; AS: anterior septum; PS: posterior septum; Rot: rotation. * P value < 0,05 vs T0 (paired Student T test). ®Data available for 14/16 patients. §Average value of 18 longitudinal segments.

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P183

Closure of Aortopulmonary Window using Amplatzer Duct Occlude I: two cases and literature review

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Introduction: Aortopulmonary window (APW) is a rare defect between ascending aorta and pulmonary artery above the semilunar valves. Surgical approach has been the standard treatment over the previous decade, however, increasing experience in transcatheter therapies provides an alternative lower-risk technique to close the lesion.
Methods: We present two cases of transcatheter device occlusion of APW in a 3-month-old infants, after a multidisciplinary discussion with surgeons. Patient 1 was a 3-month-old male infant (4.5 kg) with clinical signs of cardiac failure and loud pan-systolic murmur during intercurrent RSV bronchiolitis. Echocardiogram showed a 4mm-restrictive APW with left to right shunt and significant left ventricle dilatation requiring diuretic therapy. On hemodynamic assessment, right-sided pressures were 60% systemic pressures. Patient 2 was a 3-month female infant (6kg) referred for a continuous murmur and feeding intolerance. Echocardiogram showed a 3.3 mm-AP and a small ductus arteriosus with left heart overload. The procedure was through femoral artery and vein access under general anesthesia. Dimensions of the defect were estimated on anterior-posterior and lateral views of ascending aortography and an 8-6 Amplatz duct occluder device was selected. The defect was crossed from pulmonary artery with a 0.035 inch Terumo wire. The guidewire was exchanged within a 5 Fr Catheter to an extra stiff Cook 035 inch wire to deliver the Torqvue delivery sheath and then the device, with check aortography to confirm position. In the second patient, the duct was occluded with a 5-4 Amplatz duct occlude II Additional Sizes retrogradely.
Results: Good position and no residual leak were confirmed on echocardiogram. Both patients were started on aspirin for six months and they have been followed-up for 12 and 5 months respectively with good outcomes
Conclusions: Even in early infancy transcatheter device closure of APW is a safe procedure and should be preferred in cases of favorable anatomy.

P184

Coronary artery compression during intention to treat pulmonary homograft dysfunction with percutaneous pulmonary valve implant

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Introduction: Percutaneous pulmonary valve implant (PPVI) has been developed as a nonsurgical alternative for the treatment of congenital or acquired pulmonary valve pathologies. The latest ESC guidelines don't provide any recommendations about the treatment of pulmonary homograft dysfunction. The aim of this report is to share our opinion about the management of this particular case.
Methods: A 40-years-old male patient who underwent a Ross procedure at the age of 23 years with implantation of a 24 mm pulmonary homograft presented at our Institute after 17 years. Clinically stable and asymptomatic. The Echocardiographic evaluation detected an anterograde supravalvular mean gradient of 61 mmHg with associated severe valvular insufficiency and right ventricle dilatation and dysfunction. Cardiac magnetic resonance imaging with 3D reconstruction allowed a better anatomic evaluation and showed normal pulmonary arteries branches and a minimum stenosis diameter of 10x11 mm. After extensive discussion with the congenital and GUCH team the patient was named eligible for PPVI.
Results: The procedure consisted in progressive balloon dilatation of the stenotic homograft and simultaneous selective angiography of the left main coronary artery. A first dilatation performed with a semicompliant 20x40 mm balloon did not result in any coronary compression, second inflation with a non-compliant 20x30 mm balloon determined a clear disappearance of the "waist" and obstruction of the proximal anterior descending coronary artery. Since this finding represents an absolute contraindication to PPVI the patient was scheduled for elective surgery. At 17 days, the patient underwent successful substitution of the dysfunctional homograft with a pulmonary homograft 26 mm and substitution of the dilated ascending aorta with a Jotec 26 mm prothesis.
Conclusions: PPVI is a safe and highly effective procedure, but still challenging in some particular anatomic features. Patients who have undergone Ross operation are generally at high risk of coronary compression during PPVI due to both the orthotopic position of the pulmonary homograft and the re-implanted coronary arteries. This justifies the need of adequate imaging techniques and balloon characteristics in order to assess the better treatment and avoid any possible complications.

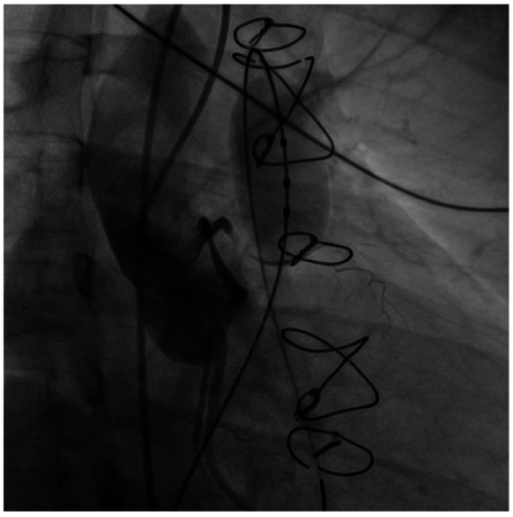

Figure 1. Coronary angiography during inflation with non-compliant 20x30 mm balloon with evidence of clear obstruction of the proximal anterior descending coronary artery.

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Coronary artery fistulas after pediatric heart transplantation

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Introduction: Coronary artery fistulas (CAF) are a rare entity. Although after heart transplant are found more frequently.

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Typically, CAF are draining into the right ventricle. Non-cameral coronary artery fistulas (NC-CAF), are even less frequent, so there is little described about them, especially in children.
Methods: We retrospectively revised our center's database of heart transplant patients registered between 2008-2020. Those with at least one coronoriography were included. The presence of CAF between the coronary arteries and cardiac or extracardiac structures, its distribution, degree and evolution were analyzed, as well as the characteristics of transplant procedure (receptor's sex, age at transplantation, indication, induction agent), immunosuppressant treatments and its potential relationship with the CAF.
Results: A total of 30 patients were identified. 11 girls and 19 boys. Median age at heart transplant was of 8,5 years, with median follow up of 5,3 years. On its first coronary angiography, we found at least one NC-CAF in thirteen of our patients. Nine of them had just one fistula, two presented two fistulas, and two had three or more. According to its flow, 52,6% were small, 26,3% medium and 21% large. Fistulas originated from coronary arteries of the graft, and most of them ended on the ipsilateral lung field, except two draining directly to the pulmonary artery and two to systemic arteries. On the second coronarography (median 22 months after first one) just six of them still had the fistulas. All of them small. The reason for heart transplant was 74% Cardiomyopathy, 23% congenital heart defect and 3% retransplantation.
Conclusions: Coronary artery fistulas are not an uncommon anomaly in patients undergoing heart transplantation. In our series, most NC-CAF drain to the pulmonary parenchyma, but we report the first two cases in pediatrics of NC-CAF draining to systemic arteries. All fistulas showed involutional tendency over time. Main pathophysiological hypothesis for fistula formation is great post-transplant pro-inflammatory state. No statistically significant relationship between NC-CAF and any of the studied factors was found. Neither any relationship with mortality nor loss of the graft

Table 1. Characteristics of the patients.

Case	Age at HTx (years)	Etiology	Blood type	Age donor	Isquemia T (min)	Induction Drug	Immunosuppressant treatment	T Tx-1st Cath (years)	Nº fistulas 1st	Grade	Origin	Destination	2nd Cath (T after tx)	Evolution	Follow-Up Years	Nº Cath total
1	15	CONG	A+	16	190	BAS	TC/MM/COR	2,5	1	Small	LAD	Lung field	No	No control	11,3	1
2	13	CMOP	0+	32	245	ATG	TC/EV/COR	0,7	1	Small	Circ	Lung field	1	Same	8,9	5
3	6	CMOP	0+	23	255	ATG	TC/EV/COR	0,5	1	Small	LAD	Lung field	0,5	Same	8,7	7
4	10	CMOP	0+	1,5	210	ATG	TC/MM/EV/COR	1	1	Moderate	RCA	Lung field	5	Same	8,3	2
5	13	CMOP	A+	16	253	BAS	TC/MM/COR	0,5	1	Moderate	LAD	Lung field	3	Same	5,1	2
6	2	CMOP	B+	28	206	ATG	TC/MM/COR	0,9	2	Small Moderate	RCA Thoracic	Lung field LAD	1	Same Regressed	4,84	2
7	14	CONG	A+	U	194	ATG	TC/MM/COR	1,25	1	Small	LAD	Lung field	0,8	Regressed	4,38	2
8	6	CONG	A+	9	U	BAS	TC/MM/EV/COR	2	3	Moderate Small Large	Circ RCA LAD	Lung field Lung field RPA	No	No control	4,15	1
9	14	CMOP	A+	38	270	BAS	TAC/MM/COR	0,4	2	Small Small	LAD Circ	Lung field Lung field	2,2	Same Regressed	2,75	2
10	8	CMOP	0+	U	220	BAS	TC/MM/COR	0,75	1	Small	RCA	Lung field	No	No control	2,3	1
11	6	CMOP	0+	14	165	BAS	TC/MM/COR	1,6	1	Small	LAD	MPA	No	No control	1,72	1
12	1	CMOP	AB+	3	U	ATG	TC/MM/COR	0,5	> 3	Large Large Large	CD CI Thoracic	Lung field Lung field LAD	No	No control	0,65	1
13	1,6	CMOP	A+	0,4	242	BAS	TC/MM	7	1	Moderate	LAD	Lung Field	No	No control	7	1

CMOP: Cardiomyopathy; CONG: Congenital; U: Unknown; BAS : Basiliximab; ATG: Thymoglobulin; TC: Tacrolimus; MM: Mycophenolate; EV : Everolimus; COR: Corticosteroids; LAD: Left anterior descending; RCA: Right Coronary Artery ; Circ: Circumflex; RPA: Right Pulmonary Artery; MPA: Main Pulmonary Artery

P186
Covered Stent implantation into the Right Ventricular Outflow Tract in Infants with TOF/PA+VSD and Diminutive Pulmonary Arteries - A game changer?

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Introduction: Right ventricular outflow tract (RVOT) stenting is a palliative treatment option in symptomatic infants with tetralogy of Fallot (ToF) or in patients with pulmonary atresia and ventricular septal defect (PA+VSD). We present our experience in patients who were treated with covered coronary stent grafts - BeGraft (Bentley InnoMed, Hechingen, Germany) in the RVOT.
Methods: Between 11/2017 and 04/2020, the BeGraft stent was used to widen the RVOT in 14 patients. Indications for the procedure, clinical data at the time of the procedure were noted. Efficacy and safety of the catheter intervention were assessed, additional procedures performed at the time of stent implantation or later and follow-up data were collected.
Results: The study group consists of 14 patients (8 males, PA+VSD - 4, ToF - 10), 6 patients with arterial duct dependent pulmonary circulation. The median age of the patients was 25 (3-306) days, the median body weight 3,2 (1,89-6,5) kg and the median height 50,5 (42,0-74,0) cm. Eight patients were newborns, the rest was treated in the first year of life. Diameters of implanted stents were as follow: 3,5x12,0mm (6, 3),5x16,0mm (2, 4),0x12,0mm (1, 4),5x18,0mm (1, 4),5x16,0mm (2) and 5,0x16,0mm (2). All BeGraft stents were implanted successfully. The median time of palliation was 156 (28-835) days. Five patients required stent re-dilation(s). Eight patients required additional stent implantation to relieve a proximal obstruction in RVOT. There were two complications: RVOT perforation - 1 patient, stent embolization to aorta - 1 patient. All stents were patent during follow-up time, and we observed a significant increase in the diameter of the pulmonary artery and its branches after the procedure ($p<0,02$). Eight patients had corrective surgery performed. Cardiac surgeons evaluated the removal of the implanted stents as complete and easy in all cases.
Conclusions: Stenting of the RVOT with BeGraft stents in young patients with diminutive pulmonary arteries not being amenable for primary surgical correction was safe and effective. This provides a durable method of palliation with a possibility of further redilatation. Utilization of the BeGraft stents may facilitate surgical removal of the implanted stent during the surgical correction.

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Creation of interatrial communication in children with low body weight – is needle trans-septal puncture safe and effective?

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Introduction: Atrial septostomy in patients with intact interatrial septum requires radiofrequency puncture or needle trans-septal puncture. The aim of the study was to evaluate the feasibility, efficacy and safety of needle trans-septal puncture (TSP) in creation of interatrial communication in children with body weight below 15 kg.
Methods: Between 2017-2019, 14 procedures of TSP were performed in 12 patients (3 girls, 9 boys), at median age of 8,8 months (0,03-48,5 months), with median body weigh 6,1 kg (3-13,9kg). In 2 patients procedure was repeated after 4 and 7,3 months due to fenestration closure. The indications for the procedure were: elevated pressure in the left atrium (LA) secondary to left ventricular diastolic dysfunction and/or mitral valve pathology – 12 procedures (patients with aortic stenosis after: pre- and/or postnatal balloon valvuloplasty, hybrid procedure and percutaneous conversion to biventricular circulation or Ross-Conno operation; dilative cardiomyopathy) and congenital heart disease associated with severe cyanosis due to inadequate mixing – 2 procedures. Percutaneous TSP was performed uneventfully in all cases, from the femoral approach (11), or hepatic approach (3), using a Brockenbrough technique with a needle length of 56 cm or 71 cm, under TEE guidance (11), or TTE guidance (3). TSP was followed by static balloon septostomy - cutting balloons, angioplasty balloons (14), and additional stent implantation (1).
Results: Twelve procedures of atrioseptostomy were successful (86%), in one procedure it was not possible to cross the septum with balloon greater than 2 mm, in one atrioseptostomy was ineffective due to floppy septum (TGA, critical condition, resuscitation during the procedure, death). Median diameter of created interatrial communication was 4 mm (3-5mm), median pressure gradient between left and right atrium was 5 mm Hg (2-9 mm Hg), median LA pressure after procedure was 14 mm Hg (6-18 mm Hg). No complications related to TSP occurred. Thrombus formation in the LA during static septostomy in 1 patient was successfully treated with heparin.
Conclusions: TSP in children with low body weight is technically demanding, but safe and effective procedure.

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Device closure of left lower pulmonary artery to left atrial fistula using vascular plug

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Introduction: Left side pulmonary artery to left atrial fistula is relatively rare in comparison to right side.Significant right left shunt is clinically manifested as effort intolerance,cyansois and pan-digital clubbing, Whenever anatomy is suitable device closure is should be considered as first option. A 27-year-old male weighing 47 kg presented effort intolerance, cyanosis and history of recurrent seizures. The oxygen saturation was 87% in room air. In addition to routine tests, Computed enhanced tomography of pulmonary artery angiogram showed the left pulmonary artery to left tortuous fistula with aneurysmal sac.The narrowest diameter of tortuous course was 1cm just before the aneurysmal sac.The plan was device closure under local anaesthesia after informed consent the patient was not willing for surgery because of multiple haemangioma and recurrent seizure
Methods: From right femoral approach,A 035x260cm Terumo wire (Terumo, Tokyo, Japan) was taken across the fistula from the left pulmonary artery through the sac far into upper right pulmonary vein.A compatible 8Fr sheath with its dilator was tried to reach up to the sac but was not possible.The dilator was exchanged with a 5Fr multipurpose diagnostic catheter and sheath was parked desired placed.An Amplatzer vascular plug of size 20mm(St Jude Medical, Minnesota, USA) was deployed without any residual shunt.
Results: The fistula was closed completely without any residual shunt.The arterial oxygen saturation (SpO2) increased to 98%. The patient was discharged on and aspirin on the third post-procedure
Conclusions: A 40yrs old male presented with effort intolerance, central cyanosis, pandigital clubbing and recurrent seizure. He had large left pulmonary artery to left atrial fistula causing significant right to left shunt. It was closed under local anaesthesia using Amplatzer vascular plug.

P189

Early complications after arterial duct stenting (ADS) in newborns with ductdependent pulmonary circulation- initial single centre experience.

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Introduction: ADS in newborns with critical CHD with duct-depending pulmonary circulation is an alternative to surgical treatment. Especially while gaining experience, ADS can also lead to life-threatening complications such as stent thrombosis or suboptimal position (incomplete covering of the aortic part of arterial duct or stent protrusion in pulmonary artery). We present three cases that needed medical, interventional and/or surgical treatment in the first 48 post procedural hours due to acute occlusion of the implanted stent.
Methods: For a period of 3 years (2016-2019), 12 interventions in duct-dependent pulmonary circulations were planned and ADS was performed in 8 newborns, mean age 10,3 days and mean weight 3005g (range from 2100g to 3860g). All patients were diagnosed with complex congenital heart defects - 6 with pulmonary atresia and 2 with severe pulmonary stenosis. The diameter of used coronary stents was 3 to 4.5 mm and the length between 9 and 28 mm. The size of the stent was chosen depending on the body weight and the length of the arterial duct. Systemic anticoagulant treatment was applied to all patients.
Results: Thrombosis of the stent occurred to 2 patients. In first case, the thrombosis of ADS, presented immediately after implantation, was successfully overcome with application of local and systemic fibrinolysis. In second case the thrombosis occurred on the second post procedural day. Systemic fibrinolysis was ineffective and urgent surgical implantation of BT-shunt was performed. Incomplete covering of the aortic part of PDA was detected in 1 patient, which led to folding of the uncovered part the duct, acute occlusion and critical hypoxemia. In setting of emergency, urgent reimplantation of a new stent was performed, but child expired by the end of the procedure.
Conclusions: Early complications during ADS in setting of learning curve could be overcome with proactive team approach including on time use of all assets of fibrinolytic medications, different stent sizes for immediate interventional procedures and surgical stand-by.

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Embolization of anomalous vessels in children: management using "off-label" devices

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Introduction: The management of anomalous vessels, such as coronary artery fistulae (CAF) and aortopulmonary collaterals (APC) has become increasingly accessible to transcatheter closure (TCC) due to the advent of novel devices. The authors describe the cases where three different types of anomalous vascular anomalies were embolized using "off-label" devices for this purpose.
Methods: Case 1: a post ventricular septal defect surgical closure acquired fistula between the right sinus of Valsalva and the right ventricle was embolized in a 10 years old child with a 5mm x 6mm Amplatzer® Duct Occluder Additional Sizes (ADO II AS) (image 1A' and 1A''); Case 2: a 5mm x 4mm ADO II AS device was used to embolize a congenital fistula arising from the left sinus of Valsalva and draining into the right atrium (image 1B' and 1B'') in a 23 months old boy; Case 3: a 3mm x 4mm ADO II was used to embolize an arterial fistula between the descending aorta and the right pulmonary branch (image 1C' and 1C'') in a 4 months old girl.
Results: In all 3 patients, angiograms showed good position of the device and complete vessel embolization with minimal residual shunting. No complication occurred during or post-procedure.
Conclusions: Though the ADO II and II AS are designed to treat patent ductus arteriosus, our experience illustrates that they can be used safely in non-ductal position like CAF and APC. Compare to other devices used to close such fistulae they have several advantages, including a high rate of occlusion with a single device, improved control over the placement and release of the device and the use of low-profile delivery catheters. Longer-term follow up in a larger cohort of patients will be required to establish long-term efficacy and device safety.

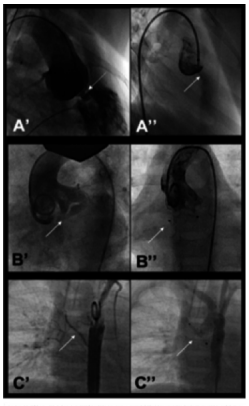

Image 1: A' Ascending aortic angiography in the right anterior oblique view shows a fistula between the right sinus of Valsalva and the right ventricle (white arrow); B' Fistula arising from the left sinus of Valsalva and draining into the right atrium (white arrow); C' Collateral between the descending aorta and the right pulmonary branch (white arrow); A'', B'', C'' Post-procedure angiogram showing the device (white arrow) in a good position with no residual shunt.

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Frequency of thyroid dysfunction in pediatric patients with CHD exposed to iodinated contrast media – a long-term observational study

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Introduction: The thyroid gland of patients with congenital heart disease may be exposed to large doses of iodine from various sources. We assessed the thyroid response after iodine exposure during conventional angiography in cardiac catheterization and angiographic computer tomography in childhood.
Methods: Retrospective mid- to long-term follow-up of 104 individuals (24% neonates, 51% infants, 25% children) with a median age and body weight of 104 days [0-8 yrs] and 5.3 kg [1.6-20]. Serum levels of thyroid-stimulating hormone, free triiodthyronine and free thyroxine were evaluated at baseline and after excess iodine. We also assessed risk factors that may affect thyroid dysfunction.
Results: Baseline thyroidal levels were within normal range in all patients. The mean cumulative iodinate contrast load was 6.6±1.6 mL/kg. In fact, 75% had experienced more than one event involving iodine exposure, whose median frequency was 3 times per patient [1-12]. During the median 3-year follow-up period [0.5-10], the incidence of thyroid dysfunction was 15.4% (n=16). Those patients developed acquired hypothyroidism (transient n=14, long-lasting n=2 [both died]) with 10 of them requiring temporary replacement therapy for transient thyroid dysfunction, while 4 patients recovered spontaneously. 88 individuals (84.6%) remained euthyroid. Repeated cardiac interventions, use of drugs that interfere with the thyroid and treatment in the intensive care unit at the index date were strong predictors for acquired thyroid dysfunction.
Conclusions: The incidence of acquired hypothyroidism after iodine excess was 15.4%. However, most patients developed only transient hypothyroidism. Systemic iodine exposure seems to be clinically and metabolically well tolerated during long-term follow-up.

P192

GORE® Cardioform ASD Occluder Experience in Transcatheter Closure of "Complex" Atrial Septal Defects

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Introduction: Trans-catheter ASD closure is still challenging in "complex" clinical or anatomic settings. This study evaluated feasibility and results of percutaneous closure of "complex" ASDs with the GSO-ASD, a novel and compliant device, in a tertiary referral centre. The aim of the study is to evaluate safety and efficacy of the GORE® Cardioform ASD Occluder (GSO-ASD, WL Gore & Associates, Flagstaff, AZ) in closure of "complex" ostium secundum atrial septal defects (ASD).
Methods: Between January 2020 and November 2020, 52 patients with significant ASD were submitted to trans-catheter closure with the ASD-GSO at our Institution. They were classified as "complex" (n= 25, Group I) or "simple" ASD (n= 27, Group II) on the basis of clinical or anatomic characteristics, that anticipate them as plain "surgical" candidates. Procedural success and safety as well as early results were compared between groups.
Results: Age and weight at the procedure were significantly different in the two groups (7,8 ± 5,9 vs 19,2 ± 18,9 ys, p<0.0004 and 28 ± 15 vs 45 ± 23 kg, p<0.003, respectively) as were ASD

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diameter (19,8 ± 4,6 vs 16 ± 3,3 mm, p<0.002), ASD diameter/ patient weight (0.86 ± 0,35 vs 0,47 ± 0,26, p<0.00004), QP/QS (2 ± 0,78 vs 1,3 ± 0,3, p<0.00005), procedure and fluoroscopy times (68,5 ± 30 vs 46 ± 19 min, p<0.004 and 14,2 ± 9 vs 6,6 ± 5,3 min, p<0.0005, respectively) and complication rate (0 vs 28%, p<0.002). However, successful closure did not significantly differ between the groups either at hospital discharge (96 vs 100%, p=NS) or 1 month-follow-up (96 vs 100%, p=NS).
Conclusions: Percutaneous closure of "challenging" ASDs with ASD-GSO device is effective and safe, although still significantly more demanding as compared to "simple" ASD.

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Initial and Mid-Term Results After Percutaneous Pulmonary Valve Implantation: Comparing Melody and Sapien Valves

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Introduction: Melody and Sapien percutaneous pulmonary valves are available nowadays in most European countries to treat right ventricular outflow tracts (RVOT). Although these valves are widely used, few studies compare both of them.
Methods: Retrospective analysis of percutaneous pulmonary valve implantations performed in our centre. Comparison of initial and mit-term results between Melody and Sapien valve.
Results: In a 7 year period, 56 valves (44 Melody/ 12 Sapien) were implanted in 56 patients (26.1 years old (SD 10.63) and 62 Kg (SD 14.9)). Tetralogy of Fallot and pulmonary atresia with VSD were the main congenital heart disease, present in 64.2%. The indication for valve implantation was pulmonary stenosis with pulmonary regurgitation (PR) in 78.6% and isolated PR in 21.4%. RVOT anatomy was transannular patch in 22 cases (39.3%), 13 bioprothesis (23.2%), 11 conduits (19.6%) and 8 native RVOT (14.3%). The intervention was performed trough femoral access in 48 cases (85.7%) and by yugular access in 8 cases (14.3%). We performed prestenting in 42 Melodys (95.5%) and 6 Sapien (50%). Pre-stent and valve were implanted in the same procedure in 35 Melodys (81.4%) and in no Sapien. Immediate complications appeared in 11 pacientes (19.6%) (8 Melodys (18.2%) and 3 Sapien (25%) (p=0.598)). Mean follow-up was of 2.8 years (SD 2.14): 2.9 (SD 2.33) for Melody and 2.2 for Sapien (SD 1.14) (p=0.301). In the follow-up 1 Melody patient (2.4%) and 2 Sapien (18.2%) developed moderate PR (p=0.03). Five Melody patients (12.5%) and 1 Sapien (10%) presented with a gradient over 40 mmHg (p=0.756). The incidence of endocarditis was similar in both groups 12.5% in Melody and 10% in Sapien (p=0.764). The rate of reintervention was of 6.8% (3 patients) in the Melody group versus no interventions in the Sapien group (p=0.352)
Conclusions: Percutaneous pulmonary valve implantation is an effective alternative to surgery. In our series we have a similar rate of success with both valves. In the follow-up, the rate of valve dysfunction was higher in the Sapien group. The number of reinterventions and infective endocartidits was higher in the Melody group although the difference with Sapien was not statistically significant.

P194
Long-term follow-up of percutaneous secundum-type atrial septal defect closure using Amplatzer septal occluder since 1995

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Introduction: The goal of this single-centre study was to evaluate long-term results of percutaneous closure of secundum-type atrial septal defect (ASD II) using Amplatzer septal occluders.
Methods: Between September 1995 and Oktober 2012, 803 patients underwent a percutaneous closure of ASD II in our institution. This group of patients was followed up from 1 month to 23 years (median 10 years).
Results: The mean stretched defect diameter was 15,9 ± 4,8mm. There were no deaths during the study. Complete closure rate at 10-years follow up was 99%. The rate of major complications was 0,5%. 1 device embolisation, 1 thrombus formation at the occluder surface and 1 cardiac erosion in perprocedural or short postprocedural course was experienced. Only 1 late complication of infectious endocarditis at the region of implanted occluder and aortic valve was detected.
Conclusions: Percutaneous closure of secundum-type ASD II using Amplatzer septal occluder is a safe and effective procedure accounting very low incidence of major complications in long-term follow-up.

P195
Management and follow-up of pulmonary atresia with intact ventricular septum. A tertiary center's experience in the last 40 years

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Introduction: Pulmonary atresia with intact ventricular septum (PAIVS) comprises a wide spectrum of pathology whose severity depends on the development of the right ventricle and the tricuspid valve. The therapeutic strategy in the neonatal period can be directed to uni or biventricular physiology, and is defined according to the size of the RV and the presence of coronary circulation right ventricle dependent. The objective of our study is to describe the casuistry of PAIVS in our center, its therapeutic approach in the neonatal period, as well as its management and its follow-up in the short and long term.
Methods: A retrospective analysis of patients with PAIVS with active follow-up in our center was carried out, from 1970 to January 2020. The percentage of patients with prenatal diagnosis, the severity defined according to the size of the RV and the tricuspid valve at birth, the neonatal approach, the therapeutic strategy and the short and long-term evolution are analyzed.
Results: 34 cases of APSI are analyzed. 30% were diagnosed in the fetal period, increasing to 50% considering only the last 10 years. We classified the patients into 3 groups according to their target therapy: group A formed by patients with univentricular physiology (11.7%), group B with ventricular and a half physiology (5.9%) and group C with biventricular physiology (82.3%). The tricuspid regurgitation severity at birth is significantly lower in biventricular cases that require additional pulmonary flow, compared to those that do not. Group C patients diagnosed before 2000 underwent surgical valvulotomy(35%), while those diagnosed after 2001 underwent percutaneous valvuloplasty(65%), being both techniques equally effective.
Conclusions: According to our series, the characteristics that best determine the target physiology are: the existence of coronary circulation dependent on the right ventricle, the right ventricle development, the tricuspid valve z-score and the tricuspid regurgitation severity. The main long-term complications are impaired right ventricle diastolic function and severe pulmonary regurgitation, with no differences depending on the decompression technique used.

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P196

Melody valve in mitral position in very young children with atrio-ventricular septum defect and severe mitral valve dysfunction.

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Introduction: The availability of prosthetic valves in infants with atrio-ventricular valve annulus <15mm is very limited. Off-label use for surgical implantation of a folded or trimmed Melody valve is an option for mitral valve dysfunction in this population. Moreover, as the Melody is expandable by percutaneous catheter balloon dilatation, it even avoids early re-intervention and allows growth until adult size prosthetic valve replacement is possible.
Methods: The Melody is implanted via surgical access by left atriotomy. The native valve is excised; the annulus is measured with Hegar stifts. The stent is folded at both extremities to reduce final length. A pericardial strip is attached in the middle or at the edge of the stent as suture ring; the valve is shrinked manually and the band is sutured in the mitral annulus; stay suture in LV if ventricular protrusion. Thereafter the valve is dilated with a balloon. Finally the stent is fixed on the interatrial septum. (Fig.1) During growth balloon dilation was performed through the interatrial septum; stable position of guide wire was assured by apical snare from retrograde transaortic access.
Results: We describe 3 similar AVSD patients (Table 1), in which the implantation of a Melody valve was life-saving as they developed all severe mitral dysfunction (stenosis MS or regurgitation MR) after prior AVSD surgery with valvuloplasty. None of them had known genetic syndromes or major comorbidities. Our results show 100% of mechanical success, no mortality nor early valve replacement; no endocarditis, LVOT obstruction nor inflow obstruction. At last check-up, none of them had MR, MS mean gradients, respectively 7, 12 and 5mmHg, without retrograde pulmonary hypertension.
Conclusions: Melody valve in mitral position in infants functions well down to 10mm of mitral annulus. LVOT obstruction can be avoided by positioning the stent completely in the atrium, without obstruction of pulmonary venous flow. Expansion up to 14-16mm in small infants is associated with higher incidence of AVB.

	Age (months)	Weight (kg)	Indication	Strip position	Inner size (mm)	Compli - cation	Later dilation	Follow-up (age, months)
P1	16	9.5	MR	middle	16	AVB3 < PM	at 17M: balloon 20 mm	66
P2	5	6.3	MR	middle	10		at 10M: balloon 14 mm	30
P3	4	4.1	MS	V end	14	AVB3 < PM		6

Table 1: patient characteristics

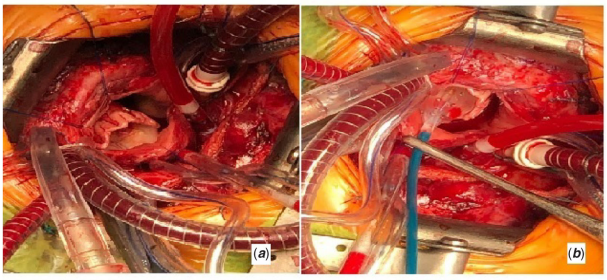

Fig. 1, surgical view, left atriotomy. A: Folded and shrinked Melody stent, getting attached into the mitral annulus by a pericardial band; B: balloon dilatation of the valve.

P197

Melody valve in rigid frame conduits: A single-institution retrospective analysis from a medium sized pediatric heart center in central europe

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Introduction: Transcatheter pulmonary valve replacement (TPVR) with the Melody valve is a well-established treatment approach for patients with dysfunctional right ventricular outflow tract in congenital heart disease. Debate continuous regarding size of implanted valve and optimal timing of implantation to minimize adverse events such as endocarditis or early explantation.
Methods: This is a retrospective single center analysis of all TPVR with Melody valve at our institution between 2010 and 2019. We reviewed the pre- and postprocedural echo-, cMRI- and catheterization parameters as available. We analyzed the patients according to age (children ≤12 years, adolescent 13-18 years,adults ≥18years) and for time dependent variables affecting the primary outcome parameters (freedom from endocarditis, reintervention or explantation). All patients had a rigid frame environment previous the TPVR (either stented bioprosthetic valve or prestenting).
Results: The Melody valve was successfully implanted in 60 patients. Mean FU-time was 5.57 and 4.39 years in the pediatric respectively adult cohort. Majority of underlying conduit types were rigid frame conduits with 47 % (diameter 20 to 27 mm) with mean post peak-to-peak gradient of 15,6 mmHg (3-35 mmHg). Melody valve durability and function were excellent in the adult group (5-year freedom from events 97,4%) and in all groups competence of the melody valve was sustained over the surveillance period. Pediatric patients demonstrated shorter freedom from any events compared to adults (HR=10.258, p=0.041). A delivery size system >18 mm was associated with extended freedom from any event (HR 0.133; P=0.026). In contrast to previous studies, higher postinterventional peak gradient was not associated with worse outcome. Furthermore, the underlying conduit-type did not influence Melody valve function.
Conclusions: Our data demonstrates the feasibility of performing the TPVR in a lower volume center in regards to percutaneous valve implantation with similar results compared to high volume centers. Risk factors for shorter freedom from any event were pediatric patient and 18 mm Melody valve. This is consistent with recently published data and should be taken into account before implantation in this age group. As we utilized quite frequently rigid frame conduits such as Hancock-Conduits, our analysis confirms the excellent long term function of the Melody valve in this setting.

P198

Multi-institutional U.S. experience of the Occlutech Atrial Flow Regulator in children and adults with congenital and acquired heart disease

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Introduction: The creation of an atrial communication with long term patency is a desirable scenario in several cardiovascular disease phenotypes, most notably pulmonary arterial hypertension, disorders of increased left ventricular filling and increased cavopulmonary pressures in patients with a Fontan type circulation. The atrial flow regulator (AFR) device by Occlutech is a fenestrated device that facilitates the long term patency of a native or de-novo shunt within the heart and/or its related vessels. Here we detail the US multi-institutional experience with the Occlutech™ atrial flow regulator (AFR) in children and adults with congenital and acquired heart disease.
Methods: Data was prospectively collected in 6 institutions after Food & Drug Administration (FDA) approval under the humanitarian use device (HUD) classification. We included adult and pediatric patients with congenital heart disease and/or pulmonary hypertensive disorders who were on maximal medical therapy and required treatment for ongoing symptoms and/or clinical deterioration. Data was analysed using SPSS.
Results: 15 patients had an AFR implanted at 6 specialist centres in the United States for a variety of indications. Implantation was technically successful in all patients. There were no device or procedure related complications reported. There was symptomatic improvement across all groups of patients. Fenestration patency rates were satisfactory on short term follow up. 3 patients died remote from the procedure, 2 from evolution of their cardiovascular disease.
Conclusions: Compassionate use of the AFR device in children and adults with congenital & acquired heart disease and/or pulmonary arterial hypertension is technically feasible, produces beneficial results and potentially improves survival. Widespread uptake of this technique and treatment at specialist centres has the potential to provide symptomatic and survival benefit to a variety of complex patients with currently limited treatment options and indeterminate prognosis.

P199

Neonatal percutaneous valvuloplasty in severe and critical congenital aortic stenosis: evolution and worse outcome predictive factors.

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Introduction: Neonatal aortic stenosis (AS) treatment aiming to improve its high morbimortality is still challenging. The standard treatment in new-borns is percutaneous valvuloplasty (PV), with very variable results. The present work's objective is to review the clinical and echocardiographic evolution of AS having undergone neonatal PV and to identify predictive factors for negative outcomes.
Methods: It is a retrospective study including severe and critical AS diagnosed perinatally and treated with PV in the neonatal period. Standard descriptive statistical analysis is used for clinical and echocardiographic items, the Fisher's exact or Mann-Whitney U tests and lineal regression are used for the worse outcome predictive factors (Ross surgery, valve replacement, transplantation or death). The multivariate analysis is calculated with the step-by-step method.
Results: 22 patients are obtained, 5 being critical AS. There are 3 cases of cardiogenic shock at birth with a left ventricular ejection fraction median of 70 (range 25-87)%. PV is performed at a median of 42 (range 1-60) days of life. 3 of 6 patients prenatally diagnosed had undergone successful foetal valvuloplasty. The PV effectivity in severe AS is of 12 of 16, and in critical AS, 4 of 6 cases. 37.50% of cases develop aortic regurgitation, none being severe. After follow-up of 4 years, 5 patients need a second PV, 3 need surgical commissurotomy, and Ross surgery is performed on 3 patients. Global survival rate is 90.91% (2 deaths after commissurotomy and none after Ross surgeries). Regarding worse outcome predictive factors, the univariate analysis demonstrates statistical differences in lower birth weight ($p<0.01$), critical AS ($p<0.01$) and worse AS grade ($p=0.01$), aortic systolic gradient at birth ($p=0.04$), aortic annulus diameter ($p=0.03$), unicuspid or bicuspid valves ($p=0.03$) and S' value of the left ventricle at birth ($p<0.01$). When these variables are included in a multivariate analysis statistical significance is found in worse AS grade ($p=0.04$) and lower birth weight ($p=0.03$) variables.
Conclusions: AS is a complex pathology with variable evolution, in this study 39.00% of cases undergo a new intervention of any type after a neonatal PV. In the present work, unfavourable outcomes are independently associated with worse AS grade and lower birth weight.

	Mann-Withney U or Fisher F test	Multivariate analysis (step by step method)	
	Significance p value	OR [Confidence Interval 95%]	Significance p-value
Aortic stenosis (AS) grade	0.01	0.69 [0.02; 1.36]	0.04
Critical AS	< 0.01		
Unicuspid or bicuspid valve	0.03	0.06 [-0.30; 0.41]	0.74
Left ventricle ejection fraction at birth	0.09		
Left ventricle S' at birth	< 0.01		
Aortic systolic gradient at birth	0.04	0.00 [-0.01; 0.01]	0.94
Aortic regurgitation at birth	0.50		
Fibroelastosis grade at birth	0.10		
Mitral annulus z-score at birth	1.00		
Aortic annulus z-score at birth	0.03	-0.02 [-0.09; 0.05]	0.51
Inotropic score at birth	1.00		
Birth weight	< 0.01	-0.16 [-0.30; -0.02]	0.03
Aortic medium gradient post-percutaneous valvuloplasty (PV)	0.36		
Aortic peak gradient post-PV	0.44		
Left ventricular ejection fraction post-PV	< 0.01	0.01 [-0.01; 0.03]	0.36
Left ventricle S' post-PV	0.18		

Table 1. Worse ouctome predictive factors analysis

P200

Our initial experience with the BeGraft® aortic stent graft for treatment of aortic coarctation

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Introduction: Herein,we present our initial experience of a new covered stent "BeGraft® aortic stent graft" in children and adults.
Methods: The study included 9 consecutive patients (5 women, 4 men; mean age 16±3 years; range 13 to 21 years) who underwent stenting for *aortic coarctation(coA)* with Begraft® aortic stent. This stent system consists of a Cobalt-chromium (CoCr) alloy which is covered with a ~200 ePTFE tubing and pre-mounted semicompliant expandable balloon. The immediate hemodynamic and procedural results were reviewed. Four patients had native, 5 patient had recurrent coarctation.
Results: Between November 2018 and November 2019, 9 patients with CoA underwent endovascular stent implantation using Begraft® aortic stent graft at our institution. The median follow-up period was 5 months. Eight of 9 patients had successful

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implantation. The one failure was related to stent migration to the abdominal aorta. The median diameter of the coarcted aortic segment was 6.1 mm. The mean stent length and balloon diameter were 37.3±7.2 mm and 15.5±3.5 mm respectively. The decrease in mean peak-to peak systolic gradient from 34.1 to 8.8 mmHg and systolic blood pressure declined from 138±10.3 mmHg to 118±12.6 mmHg following stenting. Median balloon/ coarcted segment ratio was 2.7. A 21-year-old patient who had previously undergone coarctation surgery had a stent migration to the abdominal aorta 1 day after the procedure. It was assured that the stent did not obstruct any aortic branch and it was left in the abdominal aorta. As the reason for migration, we suggest that the anatomy of the aortic arch is impaired due to previous surgery, and fixation of the stent may be impaired when the balloon is retrieved after stent implantation.
Conclusions: Our early results indicate that Begraft aortic stent implantation is safe and very effective in reducing coarctation gradient both in native and recoarctated cases. There are 2 important points associated with the procedure. First the balloon must be fully inflated to *ensure the apposition* of the *stent* against the *aortic wall*, second during the removal of balloon catheter make sure that the balloon is completely deflated and separated from the stent to prevent inadvertent displacement to the stent.

P201

Outcomes of balloon pulmonary valvuloplasty in children with Noonan syndrome: a 36-year single centre study.

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Introduction:
Background Pulmonary stenosis (PS) is the most common congenital heart lesion in Noonan syndrome (NS). The response to balloon pulmonary valvuloplasty (BPV) is typically poorer than in non-syndromic patients. The extent to which this is the case is not well understood and neither are the features that predispose to reintervention.
Objectives To assess the short- and long-term outcomes of BPV in children with NS.
Methods: Retrospective study of all patients with NS who underwent BPV at a quaternary referral institution between 1985–2020. We divided patients into 2 groups: SPS+ve – those with supravalvar pulmonary stenosis in addition to valvar PS, and SPS-ve those with isolated valvar PS.
Results: 55 patients with NS underwent BPV at a median of 246 (IQR 108-374) days of age, 28 (51%) of them in the SPS+ve group. The pre-procedural peak invasive gradient was 46 (IQR 34-68) mmHg, and 44 (IQR 35-48) mmHg in groups SPS+ve and SPS-ve respectively (p>0.05). Post procedure gradient decreased to 26 (IQR 21-40) mmHg in the SPS+ve group and 24 (IQR 17-32) mmHg in the SPS-ve group with no significant difference in post-procedural gradients between groups. Post-procedural gradient conferred no significance in reintervention rates. 22 (40%) patients underwent reintervention at a median of 207 (IQR 89-581) days post intervention in the SPS+ve group and 401 (IQR 361-945) days in the SPS-ve group (p>0.05). Of these, 17 (77%) were in the SPS+ve group (P<0.005). 14 patients (11 in the SPS+ve group) required surgical reintervention and 8 (6 in the SPS+ve group) required further BPV (P<0.005). 3 of these patients subsequently required surgery. At one-year post intervention, 12 patients had undergone surgery and 1 repeat BPV. At 5 years post intervention, a further 2 patients had undergone surgery and 7 had repeat BPV.
Conclusions: We present the largest cohort of patients with Noonan syndrome undergoing balloon pulmonary valvuloplasty hitherto described. Although BPV is often successful, the re-intervention rates are high. Supravalvular pulmonary stenosis is an independent risk factor for re-intervention. However, long-term outcomes with BPV are satisfactory and it still remains the intervention of choice in this difficult substrate.

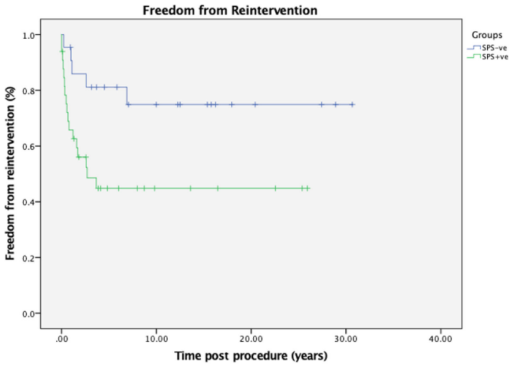

Figure 1: Kaplan Meier of freedom from reintervention.

P202

Percutaneous Closure of Ventricular Septal Defects in Pediatrics: Key Parameters Affecting Patient Radiation Exposure

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Introduction: Transcatheter ventricular septal defect (VSD) closure is gaining worldwide popularity despite its complexity. Until this date, reports on key factors affecting radiation exposure in children are still scarce. This study is the first to comprehensively analyze the impact of all relevant parameters on pediatric patients' radiation exposure during VSD closures.
Methods: Between March 2016 and August 2019, all pediatric VSD cases percutaneously treated at a reference center for interventional congenital cardiology and equipped with a single-plane Innova 2100 X-ray unit were retrospectively reviewed. Multiple linear regression was performed to investigate the impact of all morphological, clinical, technical, and procedural parameters on patients' radiation exposure assessed using total air kerma area product (PKA,T).
Results: A total of 85 patients were included in this study and 82.4% of them had perimembranous defects. The device was successfully implanted in 96.5% of cases. The procedure lasted for a median of 60 min with a median $P_{KA,T}$ of 19.6 Gy.cm^2 (range, 1.1 to 244.8 Gy.cm (2)). Patients' weight (B = 1.679, p=0.01), number of operators (B = 1.561, p=0.02), device positioning complexity (B = 2.381, p=0.002), and procedural incidents (B = 2.096, p=0.008) significantly increased $P_{KA,T}$. Patients' age (B = 1.053, p=0.784), device design (B = -1.216, p=0.780) and approach of delivery (B = -1.119, p=0.511) did not significantly affect $P_{KA,T}$.
Conclusions: Radiation exposure in pediatric patients undergoing percutaneous VSD closure was highly variable. A higher patient's weight, numbers of operators, and complexity in device positioning, as well as procedural incidents, were identified as key factors that increased patient dose in this kind of intervention.

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P203

Percutaneous closure procedure with ADO II device an effective alternative treatment for Mitral Valve Perforation.

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M Mercedes Dittler (1)

Introduction: Transcatheter mitral valve procedures are increasingly used as an alternative to surgery for symptomatic patients at high surgical risk. Transesophageal echocardiography (TEE) especially three-dimensional (3D) echocardiography allows evaluating MV in real time playing a key role guiding those procedures. We present a case of mitral regurgitation after surgery for recurrent subaortic stenosis that was successfully closed percutaneously.
Methods: CLINICAL CASE A 16-year-old patient diagnosed with incomplete form of Shone´s complex was initially undergoing for aortic coarctation. At 5-month-old the ventricular septal defect was closed and a thick subaortic fibromuscular ridge was excised. During clinic follow-up a recurrent subaortic stenosis appeared, so at 2 years old he underwent a reoperation performing an excision of the fibrous ridge with septal myectomy. Echocardiography follow-up showed a new mitral regurgitation. For years his condition remained stable, but recently he started with atrial fibrillation requiring anticoagulation and electrical cardioversion. 2-D and 3-D TEE demonstrated a perforation at the base of the anterior mitral leaflet, was elliptical in shape measuring 3mm x 6mm, so we decided to close percutaneously.
Results: By an anterograde approach through a transseptal puncture the mitral orifice couldn´t be crossed through probably. By a retrograde approach the defect was crossed with a 5F left Judkins catheter and a floppy guidewire. We interchanged the floppy guidewire with a stiff wire and the device was advanced and deployed across the perforation through the 5F delivery catheter with an Amplatzer Duct Occluder II 3mm × 4mm device. The distal disk of the device was opened in the left atrium, the whole system was pulled gently to obtain a complete apposition of the distal disk over the atrial face of the mitral leaflet, the proximal disk was opened in the left ventricle. The size and placement of the device were considered successful, so the delivery system was unscrewed. A small regurgitant jet through the device with no interference in leaflet function was seen.
Conclusions: DISCUSSION We presented the first experience using the ADO II device to close mitral valve perforation. Percutaneous therapy might have a role as an effective alternative treatment to open surgical repair in selected patients.

P204

Percutaneous embolization of a large patent ductus arteriosus in a child with significant pulmonary arterial hypertension

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Introduction: The conventional devices available for transcatheter closure (TCC) of a patent ductus arteriosus (PDA) may have limitations, especially in small children with a large tubular PDA and significant pulmonary arterial hypertension (PHT). To overcome some of these limitations, "off label" devices have been used. The authors describe a case of a child with a large PDA and PHT where an Amplatzer® Vascular Plug II device (AVP II), originally designed for peripheral vascular embolization, was successfully used.
Methods: A 13.5Kg, 3 years old male with a large, nonrestrictive PDA with evident left-sided volume overload was referred to our hospital for TCC. He had signs of congestive cardiac failure and was on diuretic and vasodilator therapy. Aortic and pulmonary pressures were measured. An aortogram was done to define the anatomy and dimensions of the PDA (image 1A). An AVP II was delivered anterogradely via 9Fr sheath.
Results: According to the Krichenko classification, the PDA was considered type C (tubular) PDA, measuring 7mm at the pulmonary end, 13 mm at the aortic ampulla and 10mm in length. The mean pulmonary artery pressure was 43mmHg and the aortic mean pressure was 63mmHg. Initially, an Amplatzer® Duct Occluder (ADO) 14mmx12mm device was used, but it was not delivered due to the device´s instability and a moderate residual shunt (image 1B). Due to the PDA´s characteristics and the concomitant high pulmonary pressure, an AVP II 18mmx14mm device was successfully delivered. The final aortogram (image 1C) showed a well-placed device and complete closure of the defect. There was no increase in flow velocity in the left pulmonary artery or the aortic isthmus. The procedure was complication-free.
Conclusions: This case illustrates that "off-label" devices can safely be used in the percutaneous closure of haemodynamically significant PDA in small, low-weight-for-age, children. The anterograde route permits the use of large-calibre catheters, such as the one used in our case.

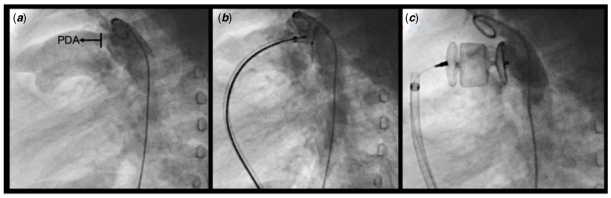

Image 1: A – Initial lateral aortogram shows a type C (tubular) PDA (patent ductus arteriosus), measuring 7mm at the pulmonary end, 13 mm at the aortic ampulla and 10mm in length; B – A 14mmx12mm Amplatzer® Duct Occluder was employed but caused a moderate residual shunt; C - The Amplatzer® Vascular Plug II device (AVP II) is well seated in the PDA with no residual shunting, the distal retention disc sits within the aortic ampulla with no protrusion into the aorta.

P205

Percutaneous pulmonary valve implantation: 10 years of experience

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Introduction: Percutaneous valve implantation (PPVI) in the right ventricular outflow tract (RVOT) is the treatment of choice for patients > 25 kg with pulmonary stenosis (PS) and/or severe regurgitation (PR). There are 2 principal devices: Melody® for anuli between 18-22 mm and Sapien® for anuli between 23 and 29 mm.
Methods: We retrospectively analyzed the procedure of PPVI performed bsince 2008 to the 2019 in our Center.
Results: Since 2008, 58 percutaneous pulmonary valve (PV) implantations were performed: 32 Melody® (18 mm in 8 patients, 20 mm in 9 and 22mm in 15) and 26 SapienXT (23 mm in 2 patients, 26 mm in 6, 29 mm in 18). Indications were PS (13 patients-20%), PR (30 patients-47%) and both (21 patients-

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33%). All patients underwent to cardiac-MR or, when contraindicated, cardia-CT before procedure. In 15 patients RVOT was reinforced by a conduit, in 41 it was widened by a mono-cuspid patch. One patient had a biological valve. In 57/58 cases a pre-stenting was performed. In 22 cases (38%) pre-stenting was performed 6 months before valve implantation: 4/32 for Melody® and 19/26 for Sapien®. In 17 patients (29%) the procedure was associated to an angioplasty of one or both pulmonary branches: in 7 patients (12%) it was followed by stent implantation (41% of them had stenosis). Mean procedure duration was 120 ± 40 min; DAP was 48 ± 30 Gy*cmq; medium od contrast employed (MOC) was 234 ± 90 cc. At a median follow-up of 30 months (IQR 0.5-10 years), 54% of patients had no residual PS or PR. Mild PR was present in 19% while in 2% it was moderates. 21% of patients had mild PS. One patient was lost at follow-up and one had a heart transplantation because of STEMI due to device related pulmonary trunk compression. One patient required an AICD for ventricular tachycardia/fibrillation. Stent embolization occurred in 5 patients, all of whom underwent succesful stent replacement.
Conclusions: PPVI is a feasible procedure with few AE. Valve functioning is preserved at medium-term follow-up. Completion of learning curve (10 patients per valve type) optimizes procedure, reduces exposure time and of the MOC employed.

P206

Percutaneous treatment for stenosis at the bifurcation of pulmonary branches involving the distal pulmonary trunk: what role for stenting?

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Introduction: The residual stenosis of the pulmonary arteries (PAs) after right outflow (RVOT) surgery represents an issue difficult to manage due to the peculiar anatomical, physio-pathological and surgical variability of the different cases. The therapeutic approach must be individualized.
Methods: We retrospectively analyzed the case treated for stenosis involving the bifurcation of PAs between the 2015 and 2018. We recorded invasive data and follow-up data, including clinical variables and echocardiographic parameters.
Results: Seven patients underwent catheterization for pulmonary bifurcation complex stenosis. Treatment flow-chart included pulmonary trunk and/or PAs angioplasty (first step), angioplasty with kissing balloon technique (second) and stenting of pulmonary bifurcation (third). All patients underwent previous surgical repair (4 tetralogy of Fallot, 2 D-transposition of the large arteries, 1 Truncus Arteriosus A4 type). Material employed for RVOT reconstruction was: Contegra (2 patients), Goretex (1), monocuspid Homograft (2). In 5/7 cases stenosis involved both PAs and preferentially a branch in the others. Median age at procedure was 9.3 years (IQR 3-29). At the beginning of catheterization, the overall mean gradient across RVOT was 60□23 mmHg with mean right ventricle-aorta ratio (RV/Ao) of 0.90□0.18. First and second step were performed in all patients. One patient successfully responded to kissing balloon: initial mean gradient across RVOT 100 mmHg with mean VDx/Ao of 0.8; final gradient 30 mmHg with VDx/Ao of 0.6. The post second step overall mean gradient across RVOT was 43□16 mmHg (p=0.29 vs baseline) with VDx/Ao of 0.77□0.22 (p=0.09 vs baseline). Third step was performed in 5 patients; in one it was impossible to perform due to his young age (three years) and the complexity of stenosis. Mean procedure duration and DAP were respectively 160□30 min and 35□25 Gy*cm (2). After stenting mean gradient across PAs was 19□14 mmHg (p=0.01) and VDx/Ao ratio was 0.45□0.17. After median follow up of 2 years (IQR 6-48 months), 1 patient underwent stent dilation 3 years after. In the others 5 there was a stable normalization of RV pressure.
Conclusions: Stenting of the pulmonary bifurcation is a complex technique, but it can be feasible and effective: it should be considered when angioplasty of the pulmonary branches alone is ineffective.

P207

Persistent Pulmonary Hypertension of the Newborn- Not always what it seems like

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Introduction: Persistent Pulmonary Hypertension (PHT) of the newborn is mostly due to pulmonary problems. PHT and heart failure as a result of pulmonary sequestration (PS) in the newborn is extremely rare, and there is no clear recommended therapeutic strategy.
Methods: We present a preterm baby born with 34+3 SSW, birth weight 1430 g. Prenatal diagnostic showed a growth restriction and polyhydramnion, but no further examination was done. After birth the boy needed mechanical ventilation and surfactant application. At the 2nd day of life the situation was complicated due to pulmonary hypertension and heart failure with detection of a perimembranous ventricular septal defect (VSD), a big atrial septal defect and most important a big left sided intralobar lung sequestration. At this age, PHT is most probably attributable to the PS.
Results: In our patient catheter-intervention was considered as being less perilous compared to surgery. Via Seldinger technique a 3 Fr lock was placed to the right groin whilst the patient was sedated and intubated. The PS feeding vessel was localized by angiography; it measured proximal 2,5 increasing to a distal diameter of 3,5 mm. A 3 Fr cobra catheter could be sufficiently advanced into the vessel as to implant 3 micro-helices (Helix ev 4 mm x 10 cm and 3 mm x 8 cm, Hydrocoil 10 3 mm/10 cm)There was only a little residual shunt afterwards and no coil protrusion into the aorta. In the echocardiographic follow-up evaluations the vessels-flow diminished and PHT reduced. However the VSD gained in hemodynamic relevance. Furthermore unfortunately in the meantime the returned karyogram revealed a trisomie 18. Since clinical situation deteriorated we decided in accordance to the parents to go for compassionate care.
Conclusions: PS is rare, but needs to be considered in PHT of the newborn. Diagnosis can be made echocardiografically and even in preterm neonates catheter intervention can be applied save and efficient without serious side effects especially if the patient isn´t eligible for surgery. However the best therapeutic option in these patients needs an interdisciplinary approach on an individual basis;

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habing had the results of the karyogram prior probably might have had influenced the therapeutic approach.

P208

Preliminary Experience with the New AmplatzerTM TrevisioTM Delivery System in Transcatheter Atrial Septal Defect Closures in Children

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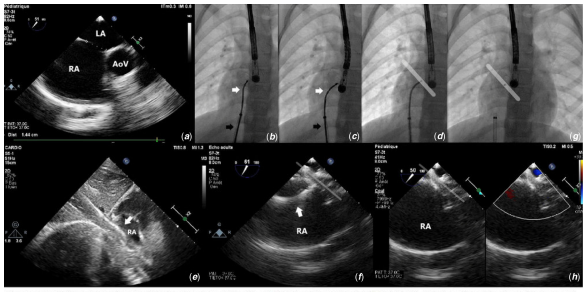
Introduction: Amplatzer™ Septal occluder remains the most widely used device with proven long-term efficacy and safety in adult and pediatric patients. In some cases, implantation success may be limited by complex atrial septal defect (ASD) anatomies. The Amplatzer™ Trevisio™ intravascular delivery system (ATIDS) is a novel delivery system designed for accurate and facilitated implantation of Amplatzer™ devices with no published clinical reports so far. We aimed to evaluate the safety, efficacy, and technical advantages of ATIDS in percutaneous ASD closure in children.
Methods: In September 2020, 9 children with anatomically challenging ASDs underwent attempted transcatheter closure using ATIDS to deliver ASO. All interventions were performed under general anesthesia, trans-oesophageal echocardiography (TOE), and fluoroscopic guidance. Standard safety, immediate, and 60-days outcomes were prospectively assessed.
Results: The median age was 8.1 (5.1 to 16.9) years and the median bodyweight was 30 (18 to 63) kg. Six patients had isolated secundum-type ASDs with absent anterosuperior rims including one with an aneurysmal septum. Three patients had unclassical defects associated with complex congenital heart anomalies. Eight devices were delivered from the femoral vein and the jugular vein was accessed in one patient with interrupted inferior vena cava and azygos continuation**.** All implantations were successful. The shape, position, and orientation of the ASO were identical before and after release on TOE and fluoroscopy. There was no device embolization or serious complication following closure. Complete shunt closure was confirmed on follow-up.
Conclusions: We report the first clinical experience with AITDS in transcatheter ASD pediatric closures. Safety and efficacy were witnessed in our case-series. The major advantage of reduced-tension deployment and reliable precision in device positioning is highly beneficial in challenging anatomies and unusual access.



Pre-procedural 2-D and color Doppler TOE showing a large ASD with a deficient aortic margin and dilated RA (A). Note how the cable adapts (black arrows) and positions the deployed device into its final definitive position once the sheath is rotated clockwise and the cable is gently pushed forward (B-D). The flexible tip of the Trevisio™ delivery wire is clearly visualized on TTE (E), TOE (F), and fluoroscopy (G) resulting in less tension on the device and more favorable device orientation with identical post-release device position and orientation (H). *AoV - aortic valve; LA - left atrium, RA - right atrium.*

P209

Pulmonary artery rehabilitation by stenting of the Right Ventricular Outflow Tract in Tetralogy of Fallot with Pulmonary Stenosis and MAPCA's

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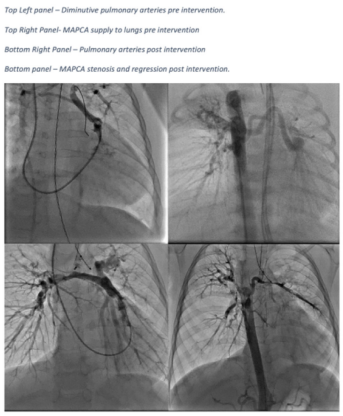
Introduction: **Objectives** To assess the role of right ventricular outflow tract (RVOT) stenting has in pulmonary artery (PA) rehabilitation in Tetralogy of Fallot with pulmonary stenosis and major aorto-pulmonary collateral arteries (TOF/PS/MAPCA's).
Background The management of a subset of infants with TOF/PS/MAPCA's requires a staged approach to management. Importantly, rehabilitation of diminutive native pulmonary arteries allowing for future corrective surgery is often a critical first step. This has traditionally been done using a surgical aorto-pulmonary shunt. Our centre has demonstrated excellent results in pulmonary artery growth following RVOT stenting in simple tetralogy of Fallot. We share our experience of pulmonary artery growth with RVOT stenting in TOF/PS/MAPCA's.
Methods: Retrospective review of all patients with TOF/PS/MAPCA's undergoing RVOT stenting as their primary intervention over a 9-year period.
Results: Ten patients (Seven antenatal diagnoses) underwent right ventricular outflow tract stent insertion at a median age of 61 days (interquartile range (IQR) 8.3-156 days). Two had concomitant MAPCA occlusion and two had concomitant ductal stenting. Median weight at stent deployment was 4.2kg (IQR3.2-5.7kg)**.** Oxygen saturations improved from a median of 79% (IQR76 -80%) to 92% (IQR90-95%), p=0.012. At subsequent angiogram the median Nakata index for right and left pulmonary arteries (RPA, LPA) increased from 45.75 (IQR 34.1 – 119) to 106.7 (IQR 77.6-178.8), p<0.05. RPA Z-score increased from -3.51 (IQR-4.59 to -2.80) to -1.17 (IQR-2.26-0.16) p<0.05. The LPA Z score increased from -2.07 (IQR-3.72-0.15) to a median of 0.24 (IQR-1.09-1.84) p<0.05. Nine patients underwent a median of 1 further catheterisations (IQR 1-2) as an elective procedure. Five patients underwent complete anatomical repair. One patient required unifocalisation to a flow limiting conduit with the VSD remaining open.
Conclusions: RVOT stenting, although often requiring further elective reintervention, is a useful procedure in the subset of patients with TOF/PS/MAPCA's, where native pulmonary arterial growth is required to facilitate repair. We demonstrate one of the largest cohorts of patients with this anatomy undergoing this palliation.



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P210

Pulmonary atresia with Intact Ventricular Septum- factors affecting choice of initial intervention and outcomes – single centre experience

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Introduction: Pulmonary atresia with intact ventricular septum (PA/IVS) accounts for 3% of all congenital heart disease. The morphology of the right ventricle and coronary artery anatomy are considered when deciding on whether an initial Radio Frequency Perforation (RFP) of pulmonary valve or a Blalock Taussig (BT) shunt would be most suitable to facilitate pulmonary blood flow.
Methods: The aim of the study was to consider which measures can be best used to predict which patients with PA/IVS can undergo RFP rather than BT shunt insertion and achieve good functional outcomes. A retrospective review of cases over an 11-year period (2008 -2019) was carried out. Data was collected from electronic patient record (EPR) and imaging modalities like echocardiography and angiography. Data collected included Tricuspid valve (TV) annulus Z score (Detroit) and ventricular morphology, if right ventricle dependent coronary circulation (RVDCC). Outcome measures included post intervention ICU stay, complications, 30-day mortality, need for re-intervention and final outcomes.
Results: In total 28 cases were reviewed. 17 had a RFP and 11 underwent surgical BT shunt. The patients with tripartite RV; all had RFP as initial intervention. 4 patients needed additional procedure post RFV to augment pulmonary flow. There was 1 complication in this group with perforation of the PA with pericardial tamponade which was successfully treated. Both the RFV and BT shunt group had no difference in 30-day mortality (1 in each group). Patients with RFV were more likely not to need PICU stay as compared to the BT shunt group. Of the RFV group 12 patients had biventricular, 3 had 1.5 ventricle and 2 had single ventricle outcome while in the BT shunt group 9 had single ventricle, 1had 1.5 ventricle and 1 patient achieved biventricular outcome.
Conclusions: In our cohort we found RFP was predominantly done in those with a tripartite RV with a higher TV Z- score and this intervention resulted in no or a much shorter PICU stay when compared to surgical BT shunt insertion group. The data also illustrated that majority of these patients achieve biventricular outcome as compared to those who have BT shunt insertion.

P211

Radiofrequency perforation of the pulmonary valve in PA-IVS – report of twelve years of experience from Gothenburg, Sweden

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Introduction: Pulmonary atresia with intact ventricular septum (PA-IVS) is a rare and heterogeneous congenital heart defect that still represents a challenge to pediatric cardiac surgeons and cardiologists. In selected cases it is possible to enable survival through transcutaneous treatment. The objective was to investigate the outcome of patients with PA-IVS selected for interventional technique based on radiofrequency perforation of the pulmonary valve, with regard to survival, need for reintervention and modality of final repair.
Methods: A retrospective, descriptive study based on medical reports, the initial echocardiographic examination and on angiocardiography in all children with PA-IVS treated with interventional technique at Queen Silvia's Children hospital, Gothenburg, between 2007 and 2019.
Results: 16 children were offered transcutaneous treatment, 1.3 per year, compared to the total incidence of PAIVS in Sweden of about 4–5 live births each year. All 16 interventions were successful. In 8/16 patients an additional source for pulmonary blood flow was needed, a modified BT-shunt in 4 cases, and insertion of a stent in the arterial duct in 4 cases. Four patients, in average 5 years old (0.7 – 10) did not need any reintervention or surgery. Five patients had percutaneous reinterventions only and in seven cases surgery could be postponed to an average of 1.7 (range 0.5 – 4.6) years of age. After a mean follow-up period of 3.6 (0.1 – 12) years, there was no mortality. Four patients suffered from complications; stroke, necrotizing enterocolitis, circulatory failure and perforation. At the end of the follow up time 14 individuals had biventricular circulation, one had 1,5 ventricular circulation and one had not yet reached definite repair.
Conclusions: Interventional treatment for PA-IVS can replace surgery as first line treatment outside of high-volume centers. In four of 16 cases there was no need for reintervention or surgery.

P212

Short and medium term outcomes of ductus stenting in PDA dependent pulmonary circulation: A single center experience

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Introduction: The revolution in the field of coronary stents in recent years and a better understanding of the ductus anatomy has led to a significant improvement in patent ductus arteriousus (PDA) stenting. Ductus stenting is now considered to be the preferred approach in many centers for bridging patients with ductus dependent pulmonary circulation to the next surgery. Aims: to evaluate the outcome of PDA stenting in patients with ductus dependent pulmonary circulation and to assess the complications caused by procedure.
Methods: A retrospective single center study. Patients undergoing PDA stenting were recruited to evaluate short and medium term outcomes. McGoon ratio, Nakata index and the total lower lobe index (TLLI) were used to assess the growth of the pulmonary arteries at closest time prior to the next planned surgery. Event survival was defined as freedom of death, ECMO and need of surgical creation of modified Blalock Taussig shunt (mBT) at the time of the next planned operation.
Results: All 69 patients underwent PDA-stenting between 2011 and 2019. Event survival was achieved in 60 patients (87%). Pulmonary parameters measured by Nakata, McGoon and TTLI showed a significant development of the pulmonary arteries in all these cases. Diagnosis, complications, mortality and need of surgical mBT Shunt are presented in image1. 3 hospital deaths were reported 1, 7, and 47 days after the Procedure. Two of them were twin birth and Preterm (30,33 gestational age) with infant respiratory distress syndrome (IRDS). One was born with low birth weight. Familial factor V Leiden mutation was documented in 2 cases with thrombotic events. 32 reinterventions were required (dilatation of stents in 26 and additionally stents implantation in 6).

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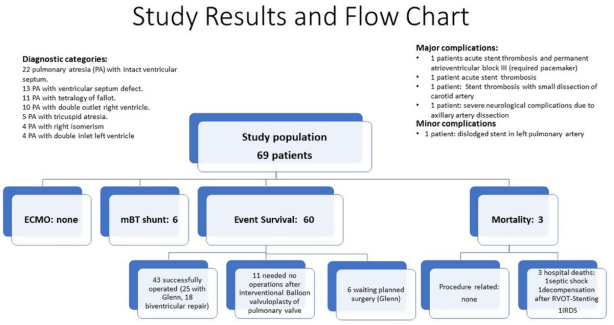
Conclusions: Stenting of PDA in ductus dependent pulmonary circulation provides a sufficient pulmonary perfusion. Preterm birth and twin pregnancy can have a negative impact on the outcome due to IRDS. Thrombophilia screen is recommended before the intervention to minimize thrombotic complications in high risk patients.



P213

Spontaneous Coronary Artery Dissection in a 30-year-old Otherwise Healthy Post Partum Female at Perpetual Succour Hospital Cebu: A Case Report

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Perpetual Succour Hospital Cebu (1)

Introduction: Spontaneous coronary artery dissection (SCAD) is a rare cause of acute coronary syndrome (ACSSTEMI). Literature highlights its low incidence and it is estimated to only comprise 0.2% of all coronary angiography cases. It is commonly reported among peripartum patients without risk factors for atherosclerosis. Its diagnosis is a challenge because its symptoms such as dyspnea, chest pain and orthopnea may present in the late stage of normal pregnancy and early puerperium. SCAD carries with it a high mortality rate if not recognized immediately, thus prompt recognition of the presentation and appropriate therapy must be determined early. This paper presents a case of Spontaneous Coronary Artery Dissection presenting as ACS NSTEMI in a 30year old postpartum woman with no known comorbidities and a seemingly unremarkable pregnancy.
Methods: We present a case of a 30-year-old postpartum woman who presented with chest pain at 13 days postpartum and was managed as a case of ACS STEMI. 2D-echocardiography with doppler was initially done to rule out common causes of symptomatology.
Results: Patient was subsequently diagnosed with Spontaneous Coronary Artery (LAD) dissection via coronary angiogram. She eventually underwent Percutaneous Coronary Intervention with application of 2 stents and was discharged improved after 2 days.
Conclusions: SCAD is an uncommon, fatal disease which occurs in young healthy subjects, usually peripartum women and still remains to be a rare cause of ACS STEMI. Thus, it should be considered in any young pregnant woman who present with angina or dyspnea with no risk factors for coronary artery disease and myocardial ischemia. SCAD has a high mortality rate if not recognized early. Although, various reports prefer conservative management as the initial approach, in the presence of features such as cardiogenic shock and unstable cardiac status like the case presented, Percutaneous Coronary Intervention remains the reperfusion strategy of choice.

P214

The rescue snared wire technique for challenging transcatheter pulmonary valve implantation

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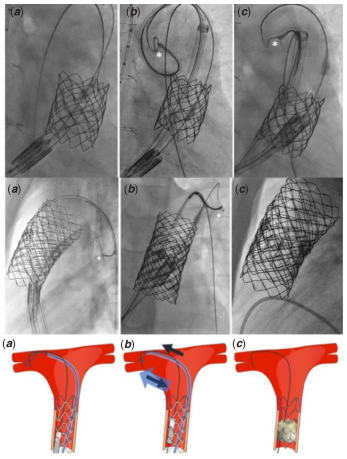
Introduction: Transcatheter pulmonary valve implantation (TPVI) is an effective treatment for right ventricular outflow tract (RVOT) dysfunction. The intervention is technically demanding and various technical tips have been described to facilitate the Sapien transcatheter heart valve (THV) delivery in the pulmonary position. We report two cases with complex RVOT anatomies and successful TPVI after the application of the snared wire technique (SWT).
Methods: Case1: A 62-year-old woman was referred to our Institution due to right ventricular outflow tract (RVOT) dysfunction, severe pulmonary regurgitation and right ventricle (RV) dilation as a result of prior surgical commissurotomy of congenital pulmonary valve stenosis. The patient was symptomatic for worsening dyspnea and reduced exercise tolerance. Cardiac MRI showed favourable RVOT anatomy and the patient was scheduled for RVOT stenting and subsequent TPVI. Case 2: A 15-year-old boy with history of critical pulmonary stenosis, and previous pulmonary valve balloon dilation at birth, was referred due to RVOT dysfunction, free pulmonary regurgitation and dilation of the RV. Cardiac MRI imaging showed large RVOT with short landing zone. For this reason, he underwent first coronary artery compression and pre-stenting of the RVOT with 3 CP stents and after 9 months was scheduled for TPVI.
Results: Procedure: Both patients underwent TPVI with implantation of an Edwards Sapien XT 29 mm THV. The advancement of the delivery system within the pre-stented RVOT was challenging and during these maneuvers the optimal position of the Lunderqvist guidewire was lost resulting in a lack of support to complete the deployment. A 16 Fr Cook long-sheath was then introduced through a second venous access, a Gooseneck Snare and a guiding catheter were advanced inside the long sheaths and the guidewire tip was snared and slightly pulled, creating a veno-venous rail inside the main pulmonary trunk, thus giving support to the delivery system, which was further advanced inside the pre-stented RVOT. After removing the 16 Fr sheath and snaring catheter, the THV was deployed in the proposed landing zone with excellent angiographic result.
Conclusions: The SWT represents a useful procedural tool and may support interventionists to overcome the pitfalls of TPVI in very complex RVOT anatomies.



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P215

The use of HeartNavigator for cardiac and peripheral vascular interventions in paediatric catheterization laboratory

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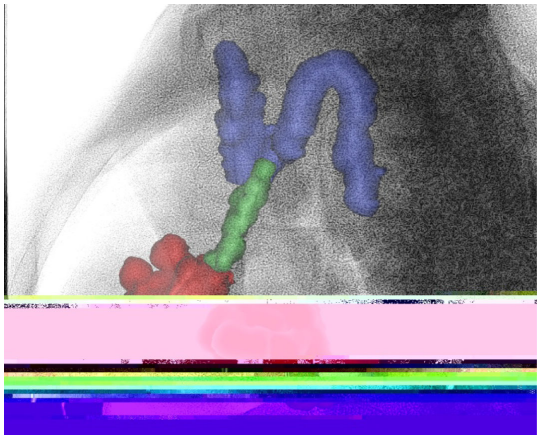
Cardiovascular Interventions Laboratory, The Children's Memorial Health Institute, Warsaw, Poland (1), Department of Perinatal Cardiology and Congenital Anomalies, The Centre of Postgraduate Medical Education, Warsaw, Poland (2), Department of Descriptive and Clinical Anatomy, The Medical University of Warsaw, Poland (3), Department of Imaging, The Children's Memorial Health Institute, Warsaw, Poland (4)

Introduction: HeartNavigator (HN, Philips Healthcare) software facilitates the use of three-dimensional (3D) roadmaps based on preregistered Computed Tomography (CT) datasets overlaid on live fluoroscopy, for real time support and guidance of the most complex trans-catheter interventions. HN is most often used in TAVI, but it can be very helpful for other cardiac and non-cardiac catheter interventions. Single paediatric referral centre experience with HeartNavigator in treatment of children, the patients of Cardiovascular Interventional Laboratory is presented.

Methods: Thirty eight consecutive patients, age median 40 mths (0-206), body weight median15.75 kg (1.6-58), with pre-existing CT imaging suitable for HeartNavigator assisted procedures were included in the study. CT datasets were segmented according to the type of procedure to visualize anatomical structures and landmarks. In 34 patients HeartNavigator was used for interventional (89%) and in 4 patients (11%) for diagnostic procedures. Cardiac anomalies were the reason for investigation in 31/38 patients, non-cardiac in 7/38 patients.

Results: HeartNavigator planning (choice of vascular access, best working projection, planning of interventional equipment etc.), guidance of procedures and judgement of anatomical results allowed for successful diagnostic cardiac catheterisation in complex post-surgical anatomy (4 patients) and for different cardiac interventions (27 patients) (precise stent implantations: CoAo/reCoAo - 8 patients, pulmonary arteries stenosis - 5 patients, RVOTO - 3 patients, SVC post thrombotic severe narrowing - 1 patient, ductus arteriosus -2 patients, Blalock-Taussig shunt -1 patient, restrictive TAPVD - 2 patients, balloon angioplasty of pulmonary arteries - 2 patients, Blalock-Taussig shunt -1 patient, severe pulmonary vein stenosis -1 patient and embolisation of PAVM - 1 patient). In the group of non-cardiac anomalies HeartNavigator guidance was used in shunt occlusion in Abernethy syndrome (congenital extrahepatic portosystemic shunt) in 2 patients, balloon angioplasty of portal vein stenosis after liver transplantation (transhepatic approach) in 4 patients and embolisation of vascular liver tumor in 1 patient. In all cases HeartNavigator was found successful in reaching the goal of procedures, shortening the time of procedures, decreasing the radiation dose and contrast volume.

Conclusions: Heart Navigator allows for precise planning and live image guidance of cardiac and peripheral cardiovascular interventions in paediatric patients.



P216

Transcatheter closure of large patent ductus arteriosus with pulmonary arterial hypertension in infants: experience with the Lifetech KONAR-MF device.

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Introduction: Device closure of large hypertensive patent ductus arteriosus (PDA) may be challenging in infants. We report our initial experience with the new Lifetech multifunctional occluder device (Konar-MF^{TM}).

Methods: Between November 2019 and October 2020, 5 infants with a hemodynamically significant, left-to-right shunting, hypertensive PDA underwent an attempted transcatheter closure using the new Konar-MF^{TM}. All implantations were performed via an antegrade route under general anaesthesia, transthoracic echocardiography and fluoroscopic guidance. Prospective follow-up was done until December 2020.

Results: The median age was 5.9 (1.8-11.4) months and the median body weight was 4.7 (3.2-7.8) kg. All 5 patients had a large, unrestrictive PDA with systemic pulmonary arterial hypertension. In 1 patient, PDA was associated with tetralogy of Fallot. PDA were either tubular (Krichenko type C: 3 patients) or window-type (Krichenko type B: 2 patients), with a mean diameter of 4.5 ±0.8mm (3.6 to 5.5mm) and a mean length of 4.3±0.7mm (3.5 to 5.0mm). Two patients had a previous failed attempt of a first device implantation (Amplatzer Duct Occluder-1 8/6), whilst Konar-MF^{TM} was implanted straightaway in 4 patients. All devices were successfully and rapidly implanted (median fluoroscopy time: 5.6min, PDS: 59.0 cGy.cm (2), Air Kerma: 7.0 mGy), without any device-related complication. There was no residual shunt 2 weeks after device implantation in all patients. The tetralogy of Fallot patient kept a severe, intractable pulmonary arterial hypertension and died in-hospital from an acute pulmonary hypertensive crisis 3

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months after the procedure despite maximal medical management. The 4 remaining patients are alive and asymptomatic. Over a median follow-up of 3.5 months (3-14 months), complete closure was achieved in all patients. Tricuspid valve regurgitation, haemolysis, pulmonary artery stenosis, acquired coarctation of the aorta and device embolization were not observed.
Conclusions: Off-label use of the new Konar-MF™ VSD occluder to percutaneously close large hypertensive PDA in young infants is feasible. Although this has to be confirmed in bigger case series, this high conformability device appears to be a safe and effective alternative for large PDA closure in selected infants.

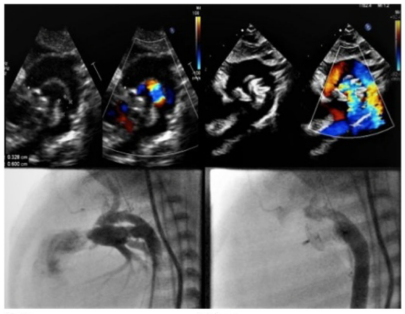

Fig 3 Fig 4

Fig1-3 : Echographic (1) and angiographic (2) view of a large non restrictive patent ductus arteriosus

Fig 2-4 : Echographic and angiographic view of Konar-MFO device closing the ductus without residual shunt or any pulmonary or aortic obstruction

P217

Up to ten years single center experience with the Melody transcatheter valve in the right ventricular outflow tract

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Introduction: The percutaneous pulmonary valve implantation (PPVI) has become an established alternative to surgical procedures over the last 12 years, with expanded indications to native right ventricular outflow tracts (RVOT) and failed bioprotheses. The objective was to summarize the experience with PPVI in our institution over the last ten years.
Methods: 101 patients from 1/2009 to 3/2019 underwent catheterization for PPVI. In 86 patients the valve was successfully implanted, 15 procedures had to be abandoned mainly due to coronary issues. The primary diagnoses included conotruncal malformations like Tetralogy of Fallot (TOF), double-outlet-right-ventricle (DORV), pulmonary atresia with VSD and major-aorto-pulmonary-collaterals (PA/VSD/MAPCA) and common arterial trunk (CAT) in two third of the cases, followed by right ventricular outflow tract obstructions (RVOTO) after Ross-surgery in 23 cases and other lesions. 72 Patients had previously implanted RVPA-Conduits, 7 patients had RVOT patch plasty, 6 patients came for valve-in-valve implantation, 2 underwent arterial switch operation and 1 patient had a pulmonary valve from autologeous pericardium. Leading lesion was RVOT obstruction in all but 11 patients who presented with severe regurgitation. 9 patients have had previous RVOT stenting in younger age to establish an adequate landing zone. Coronary testing and prestenting was performed in every patient.
Results: All but one procedures could be carried out successfully and without any coronary compromise. Major complications occurred in 4 (5%) cases (conduit rupture n=1, pulmonary embolism n=1, balloon rupture n=1 and ventricular fibrillation n=1). RVOT gradient decreased significantly from a median of 32mmHg (10-76) to 11mmHg (1-29), p>0.0001, and valve competence could be restored in all patients. In the follow-up period from 6 to 126 months 6 valves were explanted because of endocarditis (n=4), complex RVOTO (n=1) and acute thrombotic stenosis (n=1). Subacute bacterial endocarditis occurred in 5 patients with 1 patient managed medically. Reinterventions were necessary in 13 patients (redilatation or valve-in-valve implantations).
Conclusions: PPVI can be performed safely with low morbidity and mortality with a durability comparable to surgical valve implants for the reduction of the gradient across the pulmonary valve and the elimination of the pulmonary regurgitation.

P218

Ventricular Septal Defect Closure with Nit-Occlud® Lê VSD Device – Five Years' Experience

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Introduction: Transcatheter closure is an alternative to surgery for ventricular septal defect (VSD) occlusion. The aim of the study was to evaluate immediate and midterm results after VSD closure with Nit-Occlud® Lê VSD coil.
Methods: Retrospective analysis included 30 patients with VSD referred for closure during the period from October 2015 to December 2020.
Results: At the time of intervention, the mean age of patients and body weight were 7.5 ± 5.6 years and 29.3 ± 19.1 kg. Majority of the defects had perimembranous location (24/30), with the left-side diameter of 7.8 ± 3.2 mm and effective right side diameter of 3.5 ± 0.9 mm. Four defects had muscular and two outlet subaortic location, with effective diameter of 3.2 ± 1.0 mm and 6.0 mm ± 2.8 mm, respectively. Ventricular fibrilation, device embloization, contrast allergy and haemolysis developed once in different patients and were successfully treated. The coil was successfully placed in 25/30 (83.3%) patients. Majority of the devices were 10x6 mm (11/25) and 12x8 mm (8/25) in size. Two patients required implantation of second device. Follow-up period was 2.1 ± 1.4 years. Complete VSD closure was achieved in 48% cases immediately after intervention, in 74% during 2.1 ± 1.6 months after procedure, and in 82% over midterm follow-up. Remaining patients had trivial residual defect. During the follow-up, approximately one third of patients developed trace/mild aortic (0+ (IQR 0-0.5) vs. 0+ (IQR 0-0), p = 0.034) and mitral (0 + (IQR 0-0.5) vs. 0+ (IQR 0-0), p = 0.011) valve regurgitation, and approximately half of patients acquired trace/mild tricuspid regurgitation (1+ (IQR 0.5-1) vs. 0.5+ (IQR 0-1), p = 0.105). Standardized (z-score) left ventricular end-diastolic diameter (0.15 ± 0.37 vs. 0.92 ± 0.82, p = 0.005) and left atrium dimension (0.47 ± 0.58 vs. 1.89 ± 1.11, p = 0.005), as well as left atrium aortic root ratio (1.2 ± 0.1 vs. 1.4 ± 0.2, p = 0.005) showed significant decrease over follow-up related to period before intervention (Figure 1).
Conclusions: Intervention with Nit-Occlud® Lê VSD coil showed appropriate results regarding VSD closure rate, complications and chamber remodeling. Introduction of this device represents step forward in treatment of patients with VSD.

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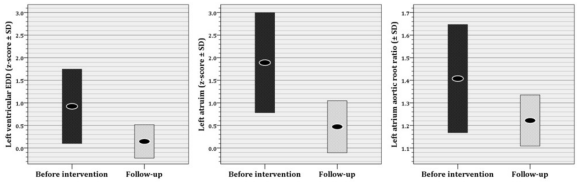

Figure 1. Echocardiographic findings before intervention and during follow-up

8. Surgery and Intensive care

P219

Anatomical autologous vein patch coronary angioplasty in children provides excellent relief in the medium to long-term

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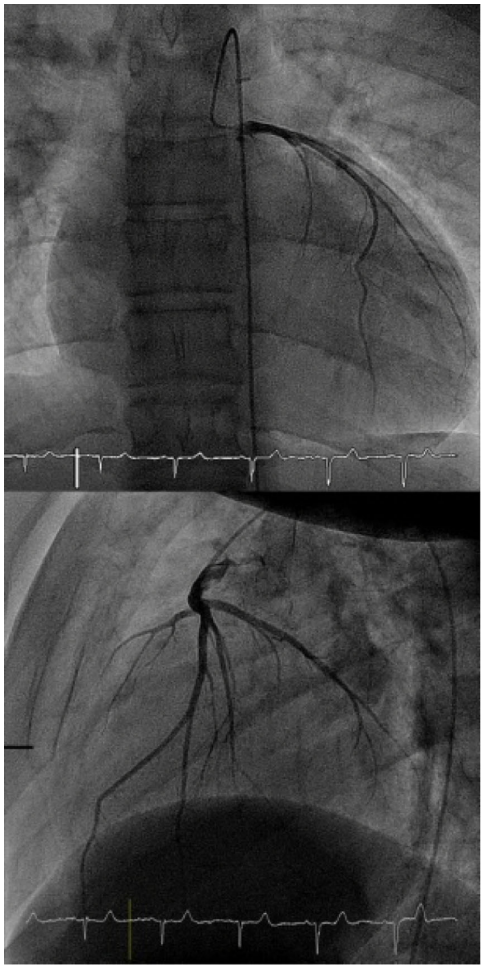
Introduction: Coronary insufficiency due to proximal coronary stenosis is increasingly being encountered. Indications, timing and the method of reconstruction are evolving. We have pursued an anatomical reconstruction of osteal and short segment proximal coronary stenosis in children using an autologous saphenous vein patch. Medium term outcome are presented.
Methods: 9 consecutive children (undergoing 10 operations) operated upon by a single surgeon (RP) for proximal coronary patch plasty (2002-19) were analyzed. Median age at coronary reconstruction was 2.6(0.2-160) months. Diagnosis included d-TGA receiving arterial switch operation (ASO)(6), Kawasaki disease(1), Takayasu arteritis(1) and idiopathic left main coronary atresia(1). In 3/6 with d-TGA, coronary reconstruction was performed during the ASO for intramural sub-commissural LCA; in the remaining 3, it was performed secondarily 22 days to 6 years post ASO. The only patient receiving patch plasty of the RCA suffered from Kawasaki disease induced aneurysm and stenosis. While autologous pericardium and xenopericardium was used in initial few cases, autologous saphenous vein patch has been used exclusively since 2009 across all age groups.
Results: All children survived the operation and are alive at a median follow-up of 7.7 years (1mo-17years). Perioperative course was uneventful in all cases. Median ventilation time and hospital stay were 2(1-8) and 19.5(9-27) days respectively. One patient with a primary LCA pericardial patch plasty needed revision with an autologous vein patch 3 months postoperatively. At follow-up, all patients were in NYHA class I (8) or II (1), and showed normal LVEF with discrete septo-basal dyskinesia in 2 cases. In all but one, angiography revealed unobstructed coronary arteries. Attached figure demonstrate angiogram 16.5 years after such a plasty. One documented patent RCA vein patch in Kawasaki disease closed during follow-up without impacting cardiac function. Cardiac MRI showed no significant perfusion deficit in any of the children.
Conclusions: Discrete coronary insufficiency in children undergoing neonatal coronary button transfer is likely to present more often in the future, thanks to high contemporary success rate of these operations. An anatomical vein patch angioplasty provides excellent prospect of restoring normal coronary flow, while at the same time keeping the option of a future coronary bypass operation open.



P220

Cardiac manifestations in the national cohort of Multisystem Inflammatory Syndrome in Children (MIS-C) in Sweden

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Introduction: The Covid-19 pandemic has been associated with an upsurge of cases of hyperinflammation in children. Named "Multisystem Inflammatory Syndrome in Children (MIS-C)" in the USA and parts of Europe and "Pediatric Inflammatory Multisystem Syndrome temporally associated with Covid-19 (PIMS-TS)" in the UK, the first cases were reported from Europe and the USA in late spring, a few weeks after the culmination of the first wave. Symptoms range from fever and high inflammatory markers via a Kawasaki-like syndrome to multiple organ failure with need of intensive care and ECMO. The Swedish national cohort of children with MIS-C is special in that it is based on a national register with early, very high coverage and validity in a country with a different approach to the pandemic. We describe here the cardiac manifestations of MIS-C in children in Sweden.
Methods: Cases were identified in the local hospitals and, after informed consent, reported to national registers. A uniform diagnostic, management and follow-up pathway was adopted early-

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on. Through retrospective study of electronic health records including ECG and echocardiography, study of intensive care and ECMO data, the cardiac manifestations could be identified.
Results: The study awaits consent fron the Swedish Ethical Review Authority. Results can therefore only be presented if authorization is given in time for the annual AEPC meeting 2021.
Conclusions: The development of the national consensus diagnostic, management and follow-up guidelines for MIS-C has resulted in the successful uniform case management in Sweden. The cases described in this study illustrate the broad clinical spectrum of MIS-C in a country with a different approach to the pandemic.

P221
Clinical outcomes of severe Ebstein anomaly required surgical intervention in neonates

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Introduction: Symptomatic neonates with Ebstein anomaly are generally at high mortality rates and the management of them remains challenging due to congestive heart failure and cyanosis.
Methods: We retrospectively reviewed the records of 21 neonates identified with Ebstein anomaly in our hospital between 2006 and 2019.
Results: Early surgical intervention was required for 15 of the 21 patients in the neonatal period. GOSE (Great Ormond Street Ebstein echocardiogram) score was 0.5 - 1.3 (1.1 ± 0.4) and TRPG (tricuspid regurgitant pressure gradient) was 12 - 89 (34 ± 21) mmHg. Median age at intervention was 11 (5 - 30) days. Among 11 patients with inadequate right ventricle function (TRPG < 40 mmHg), 9 patients without antegrade flow from the right ventricle underwent the Starnes procedure. 2 patients with adequate right ventricle function underwent tricuspid valve repair. And the remaining 4 patients with antegrade flow underwent a shunt alone or ductus ligation for minimally invasive. Total survival was 66.7% (14/21). It was lower in inadequate right ventricle function without antegrade flow cases (44.4 % (4/9)). Early death was 3 patients in inadequate right ventricle function with severe lung hypoplasia and fetal hydrops. In the cases of late death, 3 of the 4 patients had chromosomal abnormalities.
Conclusions: The survival of neonatal Ebstein anomaly required early surgical intervention was lower in cases with inadequate right ventricle. It was possibly caused due to complication of the lung hypoplasia. And the cause of late death concerned with immunodeficiency and lymphatic disorders of the chromosomal abnormalities.

P222
Coarctation of the aorta repair in adults: Twenty- year experiance

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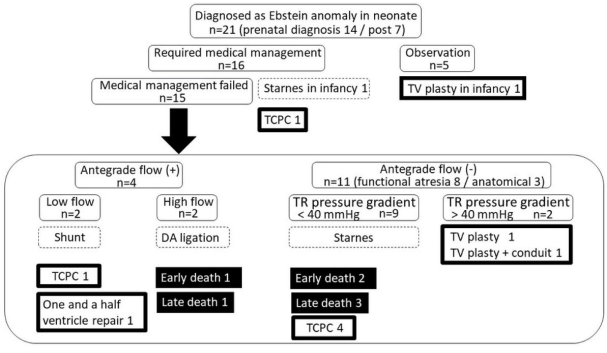
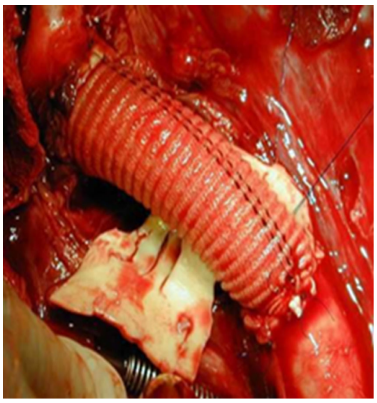
Introduction: Aortic coarctation accounts for 5 to 8% of the congenital heart diseases. Adult patients showing such a disease have an unfavorable prognosis with an expected mortality of 75% at 43 years old. Two therapeutic options are available: surgery and endovascular treatment. The aim of this work is to report the experience of our center in the diagnosis and treatment of aortic coarctation in adults and compare our results with those of the literature.
Methods: We reported a retrospective, monocentric observational study of 57 patients treated in the Cardiovascular Surgery Service and the Functional Exploration and Cardiac Resuscitation Service at La Rabta Hospital over a period of 20 years, from the 1st January 2000 until June 31, 2020.
Results: The median age of our patients was 22 years. The sex ratio was 1.42. All patients were hypertensive. The median of the transstenotic gradient was 60mmHg. Surgical repair consisted mainly of resection-anastomosis in 44% of the cases. seven patients benefited from the placement of a stent. The final gradient was 10 mmHg ± 5. No deaths were observed at 30 days. After an average follow-up of 70 months, residual hypertension was found in seven of them. And no case of recoarctation has been observed. The factors associated with the persistence of hypertension sought in the univariate analysis were: age (p = 0.110), the preoperative transisthmic gradient (p = 0.097), the postoperative transisthmic gradient (p = 0.115), severe hypertension (p=0,119), developed collateral circulation (p = 0.07), diaphragm shape (p = 0.07), costal notches (p = 0.1), bicuspid aortic valve (p = 0.15).
Conclusions: The adult aortic coarctation is a life threatening disease. The gold standard treatment remains the surgical correction but in selected patients the endo-vascular repair is a valid option. The decision making process should be based on a multidisciplinary approach taking in account the benefit/risk ratio for each available treatment





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P223

Differential anti-inflammatory response to cardiac surgery in children after total cavo-pulmonary connection versus biventricular repair

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Introduction: To examine whether cardiac surgery for univentricular palliation (UV) results in a different pattern of systemic inflammatory response compared to that for biventricular repair (BV).
Methods: In 20 children (median age 39,5 months) undergoing either (UV) n=12 or BV (n=8), plasma levels of the inflammatory cytokines TNF-a, IL-6, IL-10 and IL-12 and of procalcitonin (PCT), were measured before, during surgery and up to postoperative day (POD) 5.
Results: Epidemiologic, operative and outcome variables were similar in both groups but post-operative central venous pressure that was higher in UV. In the whole cohort, the inflammatory response was characterized by an early important and significant increase of IL-6 and IL-10 that reached their peak values either at the end of cardiopulmonary bypass (IL-10) or 4 hours postoperatively (IL-6), respectively and by a significant decrease of TNF-a and IL-12 levels after connection to extracorporeal circuit, followed by a bi-phasic significant increase first during ECC and in the early po period and second at POD 5. Patients after UV showed a shift of the cytokine balance with a predominance of the anti-inflammatory response in the early post-operative period as compared with patients with BV. Levels of PCT were similar in both groups.
Conclusions: After UV there is a shift in the balance between pro- and anti-inflammatory cytokine release towards an anti-inflammatory response. We suggest that this might be related to the hemodynamic particularities in patients after UV with higher central venous pressures and liver congestion.

P224

Drug treatment at paediatric cardiology ward and in intensive care

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Introduction: Off label use of medicines in children is common and the use of clinical support systems are widely used. The aim of this study was to assess the number of on- and off label drugs and the level support for off label-drugs used at paediatric cardiac wards and intensive care units in Sweden.
Methods: All in-hospital paediatric patients treated for cardiac conditions at a cardiac ward or intensive care units were included in the study. Data was retrieved during one day for each patient. Retrieved data included demographic information of the patient, type of drug, indication, dose and route of administration. Drugs were stratified according to; on-label according to the documented product information or off-label. Off label drugs were stratified according to use of clinical support systems (national database on drugs; ePed.se, local guidelines or other published clinical experience).
Results: Overall 28 patients were included in the study. The number of drugs used in these patients were 65 for a total of drug treatments was 233. All over 175 (75%) of drug treatments were used off label, where indication for heart failure and pain/abstinence were most common. A majority of the off-label drug-treatments were supported by clinical shared decision systems, most often the national database on drugs (ePed.se). A total of 16 drug-treatments (7%) were used without any support.
Conclusions: Off label use of drugs is still common in Swedish paediatric cardiac care. Other shared clinical support systems are widely used, but 7% of all drug-treatments did not use any support for decision of chosen drug, dose or indication.

P225

Early bidirectional cavopulmonary connection (BCPC) prior to 3 months of age is a useful strategy in selected single ventricle patients

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Introduction: Traditionally, BCPC is performed beyond 3 months of age. We sought to describe our outcomes for BCPC prior to 3 months, primarily in the setting of clinical deterioration.
Methods: All patients undergoing BCPC at ≤ 90 days of age at our institution between 1 January 2000 and 31 December 2017 were included. Patient data were collected from local databases, and follow-up for those who reached Fontan was obtained from a binational registry.
Results: Early BCPC was performed in 41 pts at median age of 82 days (range 54-90) and mean weight of 4.9 kg (SD 0.8). Primary diagnosis was HLHS 73% (30 pts), tricuspid atresia 10% (4 pts), Shone's complex 7% (3 pts) and other 10% (4 pts). Indications for early BCPC were desaturation 49% (20 pts), atrioventricular (AV) valve regurgitation 17% (7 pts), ventricular dysfunction 17% (7 pts), elective 12% (5 pts) and other 5% (2 pts). Two pts (5%) were supported on ECMO prior to BCPC, and 5 (12%) were mechanically ventilated. Concomitant procedures were undertaken in 41% (17 pts: 7 pulmonary artery augmentations, 6 AV valve repairs, 4 septectomies and 4 arch or Damus re-interventions). By univariable analysis, the 14 pts undergoing early BCPC for ventricular dysfunction or AV valve regurgitation had higher hospital mortality (21% vs 0%, $\chi 2$ p=0.01). Hospital survival amongst patients requiring postoperative ECMO was 50% (2 pts). Survival to hospital discharge was achieved in 38 pts (93%), with median (IQR) ventilation time, ICU stay and hospital stay of 20 hrs (13-39), 2 days (1-3) and 12 days (7-29) respectively. There were 2 late deaths, 1 and 4 years post-BCPC. At last follow-up, 27 pts (66%) had undergone Fontan completion after 4.2 years (SD 1.3) post-BCPC. Kaplan-Meier survival at 30 days, 1 year and 5 years was 98% (84-99%), 93% (79-98%) and 86% (70-94%) respectively (figure). The youngest long-term survivor underwent BCPC at 61 days.

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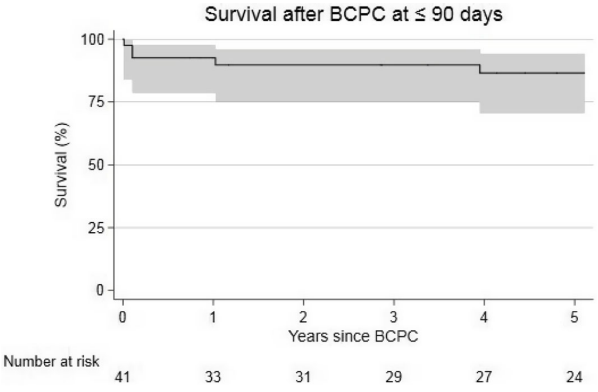
Conclusions: Early BCPC can achieve long-term survival in selected single ventricle patients, especially in the setting of inadequate pulmonary blood flow. Despite higher mortality, it remains a valuable strategy in patients with ventricular dysfunction or AV valve regurgitation.



P226

Early graft failure in the RVOT – think of TBC!

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Introduction: Recent results from our lab revealed the general pathomechanisms of Contegra graft failure: elastica degeneration and intima hyperplasia. Here, we present a special case with a rather unusual pathological aspect. A girl with pulmonary atresia and ventricular septal defect needed re-operation only 1.25 years after primary implantation of a Contegra® conduit. What was the reason for such an early graft failure?
Methods: Medical history of a Russian girl was analyzed retrospectively. Furthermore, a specimen intraoperatively taken from the RVOT was examined histologically with standard dyes. Based on our microscopic findings, TBC was suspected coincidentally. Ziehl-Neelsen staining and PCR were performed to confirm the diagnosis.
Results: The girl was born with pulmonary atresia and ventricular septal defect in 2015. She was treated with a surgical BT shunt in Russia within the first month of life. One year later, also in Russia, corrective surgery was performed with implantation of a Contegra conduit. Perioperatively, the child needed tracheostomy due to a subglottic stenosis. In 2017, she was presented in Germany with suprasystemic pressure in the right ventricle caused by severe conduit stenosis. The conduit was replaced and a remaining atrial septal defect was closed by direct suture. Histological examination showed extensive inflammatory response in intimal and medial layers. Giant cells were identified and raised the suspicion of TBC infection. Ziehl-Neelsen staining confirmed the differential diagnosis as well as PCR. Genome sequencing by hybridization was used for detection of mycobacterium tuberculosis.
Conclusions: This case shows that every patient's anamnesis should be taken accurately. If the duration of implantation is remarkably short or prior operations were performed in a tertiary referral hospital abroad, one should think of even more and rare differential diagnoses. The elementary medical care differs in many countries. As the child was treated in Germany where TBC is not as common, no one expected such a diagnosis in this girl. Fortunately, a pathologist recognized specific hints (giant cells) during microscopy and confirmed his suspicion with adequate further diagnostics. TBC infection aggravated the extensive alterations, i.e. elastica degeneration and intima hyperplasia in this infant, and certainly influenced the child's outcome.

P227

Evaluation of different approaches for atrial septal defect closure

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Introduction: Transcatheter closure of atrial septal defects (ASD) and patent foramen ovale (PFO) is the therapeutic goldstandard. However, current devices consist of a lot of foreign material and are stiff. The underlying mechanism of fixation relies on compression. Thus, there is a need for alternative materials and closure techniques. Aim of the study was evaluation of approaches with flexible patch materials and tissue adhesives in an *in vitro* burst pressure setup and a beating heart model (BHM).
Methods: ASD and PFO were created in epicardial porcine slices. A burst pressure setup and a BHM were developed to determine pressures which can be withstand after closure. The defects were closed with novel and established approaches: Sutures in combination with Poly(glycerol sebacate urethane) (PGSU) patches, a biocompatible and biodegradable elastomer (I), pericardial patches (PP) treated with glutaraldehyde (II) and without treatment (III), fibrin glue alone (IV), Tachosil® patches (V) and Histoacryl® glue alone (VI) and in combination with CardioCel® (VII). The maximum burst pressure (MBP) was determined.
Results: Sutured PGSU patches, treated and untreated PP showed sufficient tensile strength and withstood supraphysiological burst pressures after PFO (MBP of 217,2±6,2 mmHg for PGSU patches and 250±0 mmHg for treated and untreated PP; n=18) and ASD closure (MBP of 138,2±26,1 mmHg for PGSU patches and 221,8 ±9,0 mmHg for treated PP; n=18). In contrast, fibrin glue alone achieved MBP of 62,7±5,5 mmHg after PFO (n=24) and Tachosil® patches achieved MBP of 67±9,5 mmHg after PFO (n=18) and 50,7±6,2 mmHg (n=18) after ASD closure. Histoacryl® glue coated CardioCel® patches for ASD and Histoacryl® alone for PFO closure showed a MBP of 237,7±2,8 (n=15) and 250±0 mmHg (n=15). Furthermore, MBP for closure of PFOs with fibrin glue alone and ASDs with Tachosil® patches were 20,3±2,9 mmHg (n=3) and 80,7±11,8 mmHg (n=3) in the BHM, respectively.
Conclusions: We present a reliable burst pressure setup and a BHM for evaluation of closure approaches for PFO and ASD. Treated and untreated PP showed no difference in MBP. All approaches showed sufficient adhesive strength and therefore have potential to be used for the development of novel closure approaches for PFO and ASD closure.

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P228

Evolving surgical strategy in repair of interrupted aortic arch with left ventricular outflow tract obstruction

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Introduction: Surgical repair of interrupted aortic arch (IAA) remains challenging and associated with significant morbidity and need for reintervention, especially on the left ventricular outflow tract (LVOT). This study focused on the follow-up results after primary repair, with specific focus on LVOT obstruction (LVOTO).
Methods: Demographic, procedural, and outcome data were retrospectively collected for 28 consecutive patients who underwent IAA repair between 2004 and 2020 at our institution. The perioperative course and need for reinterventions (reoperations and catheter-based interventions) were reviewed, and clinical follow-up analysed.
Results: The cohort consisted of patients with type B (71%), type A (25%), and type C (4%) interruption. Associated cardiac anomalies were present in all patients, most commonly a ventricular septal defect (93%) and LVOTO (57%). Median age at operation was 12 days (range 5-86 days). Thirteen patients underwent IAA repair with LVOT intervention: 3 patients (11%) underwent a LVOT myectomy, 4 (14%) a LV bypass procedure (Yasui), and 6 (21%) a pulmonary autograft aortic root replacement (Ross-Konno procedure). No LVOT intervention was performed in 15 patients (54%). All patients survived the operation (median ICU stay 9 days). There was one late death from a non-cardiac cause during follow-up (median 7.3 years, range 0.7-14.8 years). All patients show good functional capacity (NYHA class I-II) at time of latest visit. Freedom from any reintervention was 64% at 1 year and 36% at 5 years. Most reinterventions were performed for residual arch stenosis and replacement of the right ventricle-pulmonary artery (RV-PA) homograft. Reintervention for LVOTO was indicated in 4 patients (14%): one aortic root augmentation, a myectomy, and two Ross-Konno procedures.
Conclusions: IAA repair can be performed with good early outcomes and result in good functional capacity at follow-up. In the case of concomitant LVOTO, we prefer a primary repair. In these patients we abandoned Yasui repair in favour of Ross-Konno to avoid complex intracardiac baffling and implantation of a substernal extracardiac conduit. A Ross-Konno procedure performed during neonatal life may reduce the risk of subsequent LVOT interventions, although an increased risk for right-sided interventions on the RV-PA homograft should be considered.

P229

Fate of significant left atrioventricular valve residual gradient after atrioventricular canal repair

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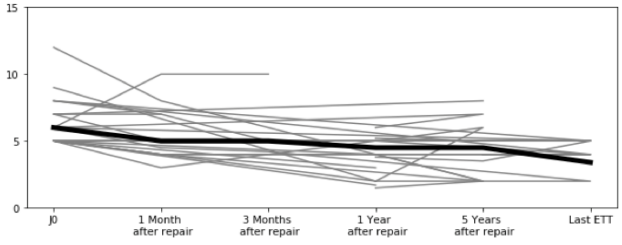
Introduction: Reoperation for left atrioventricular valve (LAVV) dysfunction after complete repair is a well-known situation. Although regurgitation is the main cause, few studies review the fate of postoperative residual stenosis. Clinical and echographic outcomes of patients with significant early postoperative gradient of left atrioventricular valve after atrioventricular septal defect (AVSD) repair
Methods: Between January 2000 and December 2019, 887 with an AVSD repaired were reviewed. Forty-one of them presented a LAVV mean gradient of more or equal than 5 mmHg at the first postoperative echo. The follow up postoperative data were obtained from hospital records and cardiologists correspondences. We studied the postoperative mean gradient of the LAVV at discharge, one and tree months, one and five years after the repair. The median follow up was 51 months (3 – 123 months).
Results: Among the 41 patient included 15 had a complete AVSD (cAVSD) and 26 had a partial AVSD p(AVSD). The median age and weight at repair were 4.5 (2.4 – 107.5) months, 4.8 (4-19) kg for the complete AVSD (cAVSD) and 50.8 (4.2 – 200.4) months, 15 (4.7 – 38.6) kg for the partial AVSD (pAVSD) respectively. 6 (40) cAVSD were diagnosed with Down syndrome and 5 (33.3) had a pulmonary artery banding before the repair. There was no reoperation for LAVV stenosis neither for regurgitation. Patients remained asymptomatic with normal range of pulmonary pressure. One patient was reoperated for left ventricular outflow tract obstruction. The median (range) LAVV gradient measured were the following at first postoperative echo, discharge, one and 3 months, one an, last follow-up: 5 (3 – 10), 4.5 (2 – 5), 3.4 (1.5 – 6) (figure 1). The difference between the first postoperative echo gradient and the last one was not significant ($p = 0.47$)
Conclusions: Significant LAVV gradient after AVSD is not uncommon. Overall trend is the stability of the gradient with normal pulmonary pressure or even decreasing. Isolated LAVV mean gradient between 5 to 10 mmHg doesn't seem to be an indication for reclamping. A moderate gradient due to the zone of apposition closure and commissural annuloplasty is acceptable when needed to obtain a competent LAVV



P230

Functional long-term results after right ventricle to pulmonary artery connection with a valved conduit in patients weighing less than 10 kilograms

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Introduction: There are several options for right ventricle to pulmonary artery (RV-PA) connection. Commercially available

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xenograft conduits such as bovine jugular vein conduits (Contegra®) or porcine valved conduits (Hancock®) show a limited durability. This is of major concern especially for small children. The aim of our study was to analyse long-term functional results for RV-PA connection in children weighing less than 10 kg.

Methods: All patients who underwent a RV-PA connection using a Contegra® (Cont) or Hancock® (HC) conduit were reviewed. Patients weighing 10 kg or less at time of implantation were included in the study. Function of the conduit was analysed reviewing serial echocardiographies. Primary endpoint was conduit reintervention (surgical or interventional). Secondary endpoints were moderate stenosis (defined as peak gradient ≥ 40 mmHg or peak velocity ≥3m/s) and conduit regurgitation (defined as more than grade 3/4).

Results: A total of 161 conduits (Cont n= 83; HC n= 78) were implanted between 1994 and 2019. There was no significant difference between the groups regarding conduit size, body weight and diagnosis. The median follow-up was 8 years [IQR 3 – 16] and is complete for 93% of the patients. Eighty-eight patients (54%) reached the primary endpoint. Cumulative incidence for the primary endpoint at 5 and 10 years was 49±6% and 81±5% for HC and 28±5% and 47±8% for Cont, respectively (p=0.002, Figure 1). Cumulative incidence of moderate conduit stenosis at 5 and 10 years was 58±6 % and72±5 % for HC and 44±6 % and 61±7% for Cont, respectively (n=0.59). Cumulative incidence of moderate conduit insufficiency at 5 and 10 years was 23±5 % and 36±5 % for HC and 52±6 % and 63±6 %for Cont, respectively (p<0.001). There were 3 endocarditis and 1 conduit thrombosis in patients with Cont and 1 endocarditis and 4 conduit thrombosis in patients with HC.

Conclusions: A conduit insufficiency occurred earlier in Contegra® conduits, compared to Hancock® conduits but the incidence of conduit stenosis was similar. A conduit reintervention was delayed longer in Contegra®. This shows that a moderate insufficiency is tolerated for a longer period than a stenosis.

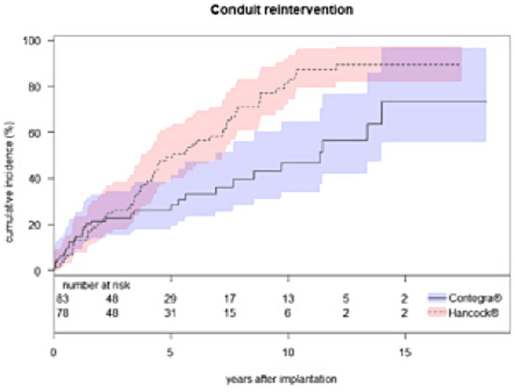

Figure 1. Cumulative incidence of conduit reintervention (p<0.001)

P231

Growth and nutrition in hypoplastic left heart syndrome. A growing problem.

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Introduction: Hypoplastic left heart syndrome (HLHS) accounts for 2-3% of all congenital heart disease. Improved growth in patients with HLHS is positively associated with outcome with lower risk of complications and reduced hospital stay. We reviewed growth in patients surgically palliated for HLHS and identified time points where faltering growth was more apparent.

Methods: Data was collected from a single centre to include all patients surgically palliated for HLHS over a 5-year period (21/07/2015-01/06/2020). Patients were divided into conventional palliation (Norwood, superior cavopulmonary connection (SCPC), Fontan) and hybrid 4-stage palliation (Hybrid procedure, Norwood, SCPC, Fontan). Weight-for-age Z-scores (WAZ) at different time points, the lowest post-operative WAZ, days to regain pre-operative weight, rates of necrotising enterocolitis (NEC) and total parental nutrition (TPN) were collected and analysed.

Results: Among 43 patients, 32 survived and were included in the analysis. In the conventional palliation group the most significant WAZ drop occurred between birth through to discharge with a WAZ different of -1.6 (95% CI -0.9 to -2.4) and -1.8 (95% CI -1.3 to -2.2) in inpatient interstage and discharge interstage groups respectively. Figure 1 illustrates the WAZ at each time point. The maximal immediate post-operative WAZ drop was greater following the Norwood compared to SCPC with a mean of 45 days (inpatient interstage) and 30 days (discharged interstage) to regain pre-operative weights following the Norwood. Patients that underwent the hybrid palliation demonstrated an immediate post-operative WAZ drop of -2.5 (95% CI -0.5 to -4.4) following the hybrid procedure with a mean of 31 days to regain pre-operative weight and -1.9 (95% CI +0.2 to -4.1) following their subsequent Norwood with a mean of 21 days to regain their pre-operative weight. Rates of NEC were 41% and TPN use 59%.

Conclusions: Poor growth is most apparent from birth through to discharge with maximal weight loss occurring following the 1st intervention with increased weight loss seen in the hybrid palliation group with long periods to regain pre-operative weight. Nutritional strategies should be focused on this early time period to improve growth with an aim to improve outcomes in this difficult cohort of patients.

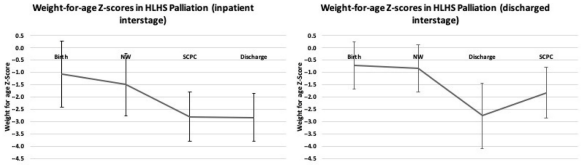

Figure 1: Two graphs to demonstrate mean weight-for-age Z-scores at birth, Norwood, superior cavopulmonary connection and discharge in patients that underwent surgical palliation for hypoplastic left heart syndrome. Patients divided into inpatient interstage and discharged interstage. HLH = hypoplastic left heart. NW = Norwood 1, SCPC = superior cavopulmonary connection

P232

Higher epinephrine dose and deep hypothermic circulatory arrest are adversely related with hemodynamics early after pediatric cardiopulmonary bypass

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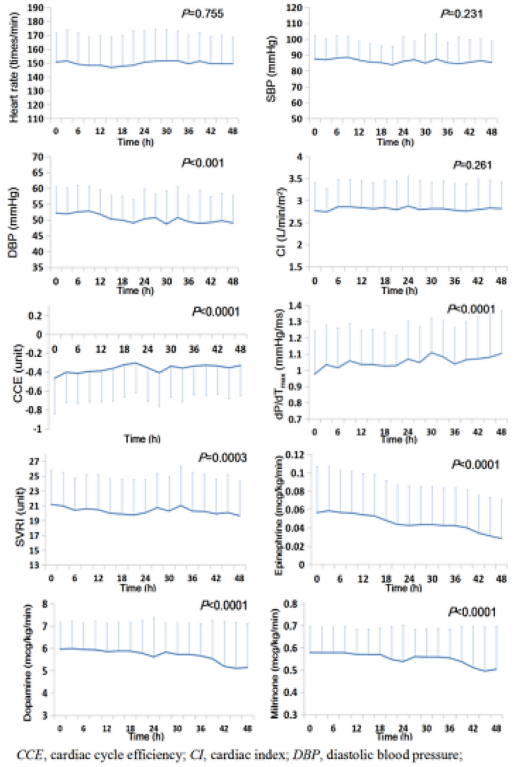
Introduction: The pressure recording analytical method (PRAM) is a non-invasive, sensitive and continuous hemodynamic and myocardial performance monitoring technique based on beat-to-beat arterial pressure waveform, but it has not been used in children with congenital heart disease (CHD) after cardiopulmonary bypass (CPB). Using PRAM, we examined the temporal trends and adverse factors of blood pressure (BP), cardiac index (CI), cardiac cycle efficiency (CCE), dP/dTmax, and systematic vascular resistance index (SVRI) in clinical management of these children.
Methods: Ninety-one children (aged 186256 days) were monitored during the first 48 hours in the CICU. Hemodynamic variables, inotropic and vasoactive drug doses and serum lactate were recorded 3-hourly. NT-proBNP was measured daily.
Results: CCE and dP/dTmax gradually increased (Ps<0.0001), while SVRI, diastolic BP and inotrope dosages decreased (Ps<0.0001) over time. No significant change was found in CI, systolic BP and heart rate (Ps≥0.231) (Figure 1). Patients undergoing deep hypothermic circulatory arrest (DHCA) had significantly higher heart rate and lower CCE (Ps≤0.006) over time. Multivariate analyses indicated that epinephrine dose significantly correlated with systolic BP, CI, CCE, dP/dTmax after polynomial transformation, with the peaks ranging from 0.075 to 0.097. CCE, but not CI and other parameters, significantly correlated with NT-proBNP (P<0.0001).
Conclusions: Systemic hemodynamic and myocardial status gradually improved in the first 48 hours after CPB without the 'classic' nadir at 9 to 12 hours. Cardiac performance was worse in patients undergoing DHCA. Higher doses of epinephrine>0.08-0.10 mcg/kg/min were adversely associated with systemic hemodynamics and myocardial mechanics and efficiency. CCE, rather than CI or other parameters, was most consistent and sensitive to reflect the patient's hemodynamic status.

Figure. Temporal trends of systemic hemodynamic and myocardial status and doses of inotropic and vasoactive drugs during the first 48 hours after CPB (n=91).



CCE, cardiac cycle efficiency; *CI*, cardiac index; *DBP*, diastolic blood pressure; *dP/dT*$_{max}$, maximal left ventricular pressure rise rate; *SBP*, systolic blood pressure; *SVRI*, systematic vascular resistance

P233

Influence of bronchus on the growth of MAPCAs after unifocalization

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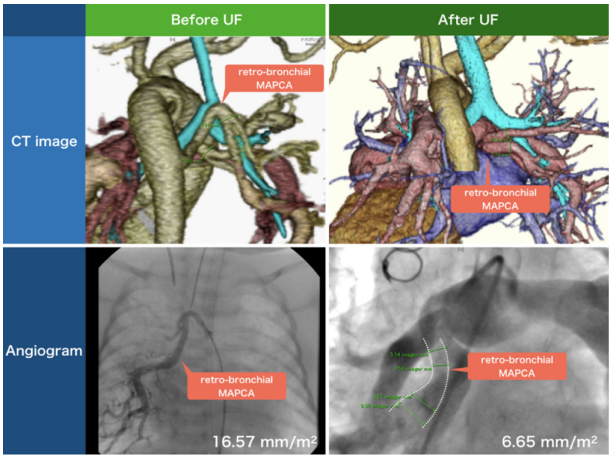
Introduction: Unifocalization(UF) of MAPCAs during infancy has become a standard surgical treatment. However, some MAPCAs need additional surgical or catheter interventions after UF due to their stenosis and poor growth. We hypothesize that the design of UF influences the vascular growths. In this study, we examine the growth of MAPCAs based on their passing route related to the bronchus.
Methods: Five patients with pulmonary atresia/VSD/MAPCA, who underwent UF and subsequent definitive repair in our institute from 2008 to 2020, were included in this study. Prior to surgical interventions, both angiography and CT scan was routinely performed to clarify the pulmonary circulation, as well as the relationships of MAPCAs and bronchus. These exams revealed peculiar MAPCAs which direct to the pulmonary hilum passing behind the bronchus(we defined them as retro-bronchial MAPCAs; rbMAPCAs). We evaluated the vascular growth of rbMAPCAs, non-rbMAPCAs and the native pulmonary artery(PA) by examining serial angiograms before and after UF.
Results: The angiogram before UF(age; 42(24-76)days, body weight; 3.2(2.7-4.2)kg) demonstrated the following data; All 5 patients had the native PA(one had a non-confluent left PA), the diameter of original unilateral PA, rbMAPCA and non-rbMAPCA was 19.94±6.64, 20.72±5.36, 19.34±7.22 (mm/m2), respectively(p=0.9171). In all patients, UF was completed with a placement of modified BT shunt through median sternotomy in a single stage(age; 1.6(1.0-2.5)months, body weight; 2.9(2.7-4.5)kg). The latest angiograms, which were performed 3.0(1.0-10.0) years after UF completion and 0.96(0.50-6.9) years after definitive repair, demonstrated a smaller diameter of rbMAPCA at peri-bronchial region(3.84±2.84mm/m (2)) compared to the native unilateral PAs(16.11±5.46mm/m (2), p=0.0043) and non-rbMAPCA(10.13±4.44mm/m (2), p=0.0103).
Conclusions: RbMAPCAs tend to be stenosed at the point where they cross the bronchus and emerge to the middle mediastinum, if they are unifocalized in situ. In unifocalization of MAPCAs, translocation of rbMAPCAs anterior to the bronchus brings a similar arrangement to the normal pulmonary artery and can be a solution to avoid future stenosis.



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P234

International survey of the use of arginine vasopressin after cardiopulmonary bypass in pediatric patients with hypoplastic left heart syndrome

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Introduction: Management after cardiopulmonary bypass (CPB) surgery remains challenging for pediatric patients with congenital heart disease (CHD). There is increasing interest in the use of arginine vasopressin (AVP) due to its unique effect of increasing systemic blood pressure without increasing heart rate or myocardial oxygen demand. Data to support the use of AVP is limited to small retrospective studies and anecdotal evidence. This study aims to characterize the prevalence, perceptions, and practice of AVP use in the postoperative management of pediatric hypoplastic left heart syndrome (HLHS) patients in intensive care units worldwide.
Methods: We performed a cross-sectional international survey between November 2017 and January 2018, targeted at neonatal, pediatric, and cardiac intensive care unit directors. Questions aimed at assessing use and perceptions on AVP safety, indications for use, benefits, advantages, and disadvantages. Participants were recruited through the following international societies: European Society of Pediatric Neonatal Intensive Care (ESPNIC), Association for European Pediatric and Congenital Cardiology (AEPC), and Pediatric Cardiac Intensive Care Society (PCICS). Responses were anonymous, except for the respondents' country of practice.
Results: Sixty-two directors from 21 countries and five continents responded. Half used AVP after Stage 1 palliation surgeries, of which 90% used it in subsequent interventions. Only 27.2% reported standardized institutional practice. Frequent indications for use of AVP included vasoplegia (90.6%), refractory shock (68.8%), and hemodynamic instability (75%). Most common dosage range was 0.0003 to 0.0006 U/kg/min. Amongst non-users, reasons for not using AVP included lack of availability (29.6%), concern about potential adverse effects (20.6%), absence of clear indication for use (22.2%), and absence of evidence suggesting improved outcomes (14.8%). Alternate drugs used instead of vasopressin included norepinephrine (71.8%) and phenylephrine (12.5%). The most common perceived disadvantages cited by both groups included extreme vasoconstriction and increased myocardial afterload.
Conclusions: Despite the paucity of data to support the routine use of AVP in the post-operative management of neonates with HLHS, survey responses demonstrate that its use is widespread. Availability appears as the primary limitation to its use. The results highlight the importance of further research into the effects, risks, and benefits of AVP, to protocolize its use.

P235

Long-Term Outcome following Surgical Repair of Absent Pulmonary Valve Syndrome

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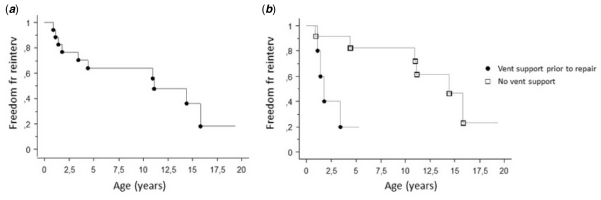
Introduction: Absent pulmonary valve syndrome (APVS) is a very rare congenital heart lesion associated with pulmonary artery dilatation, airway compression and sometimes need for respiratory support prior to surgical repair. There are only a few published studies on long-term outcome.
Methods: We included all consecutive patients at our institution with surgical repair for APVS during 1993-2018. Lund is since 1993 a tertiary referral center for pediatric cardiac surgery. Data are shown as median and range.
Results: In total, 17 patients (11 girls) were identified. Five (29%) patients needed mechanical ventilation (MV) prior to repair. The median age and weight at repair were 0.4 (0.1-2)years and 6.1 (2.5-13.7)kg, respectively, and included valved conduit in 14 (82 %) patients and transannular patch in the remaining 3 patients. Reduction pulmonary arterioplasty was performed in all 5 patients with preoperative MV and in 2 other patients (41%). Preoperative MV and longer preoperative ICU stay were associated with earlier repair, longer postoperative ICU stay and longer hospital stay ($p<0.01$ for all). Surgical or transcatheter reinterventions were needed in 10 (59%) patients at the age of 3.9 (0.9-15.8) years, the majority (60%) being performed by the age of 5 years (Figure/ Panel A). The latter subgroup included all patients with MV prior to repair (logrank p: 0.008 vs non-ventilator subgroup; Figure/ Panel B). Reoperation was required in 8 (47%) patients at the age of 2.6 (0.9-11.1) years and consisted of conduit replacement in 7 patients and conduit implantation in 1 patient who had earlier repair with transannular patch. Transcatheter intervention was performed in 4 patients at the age of 13.8 (4.4-15.8) years and included pulmonary valve replacement in 3 patients and left pulmonary artery stenting in 1 patient. There were 2 deaths, occurring at 2.4 and 13.9 years, respectively, after repair, none with preoperative MV. There was no death among cases with surgical repair after 2000.
Conclusions: Long-time survival of patients with APVS receiving surgical repair in our institution is very good but early reinterventions are common. Preoperative ventilator dependency is associated with earlier repair, longer postoperative ward and earlier reintervention but does not seem to lead to increased mortality.



P236

Low mortality but high morbidity define the outcome of truncus arteriosus repair at 280 patient-years of follow-up

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Children's Heart Centre, Children's Research Centre, University Children's Hospital Zurich, Switzerland. (1)

Introduction: Truncus arteriosus communis presents significant challenges to repair due to the neonatal age as well as the morphological substrate of the truncal valve and the neo-RVOT.
Methods: 29 consecutive children (18 males) undergoing truncus repair (2004-19) were retrospectively analyzed. Median age and weight were 27[IQR 14-37] days and 3.5[IQR 2.9-4] kg respectively. There were 15 Van Praagh Type I, 9 Type II and 5 Type IV (with arch interruption) morphologies. 1 pre-term child (1.9 kg) underwent bilateral PA banding. 21(72.4%) underwent RVOT

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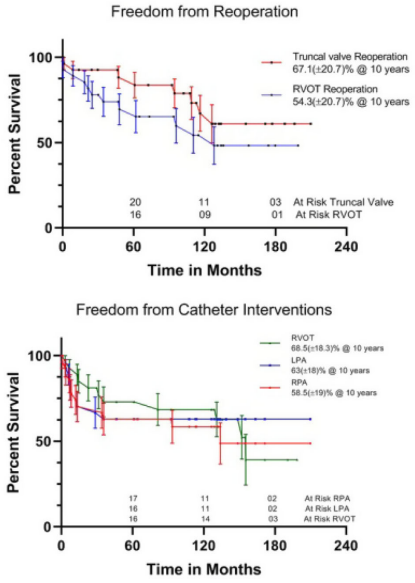
reconstruction with a conduit (8 or 12 mm Contegra graft in 20 and a Shellhigh conduit in 1) while 8(2.6%) received a conduit-less reconstruction using a monocusp. A concomitant truncal valve repair was undertaken in 2(7%) patients. Median follow-up was 10.5[IQR 5.3-12.9] years.
Results: 2(6.8%) children suffered in-hospital death. There were no late deaths, resulting in an overall survival of 93[95% CI 83-100]% at 10 years. 21 children underwent 83 reoperations and catheter interventions; individual Kaplan Meier survival are shown in the attached Fig. These include 12/29 children undergoing 19 pulmonary valve replacements and 3/29 children undergoing 4 truncal valve replacements after the primary repair. 5 patients underwent more than 1 reoperations and 8 patients more than1 catheter interventions. At follow-up, 4 truncal valves showed moderate or severe regurgitation; 2 had peak gradients of 48 and 58 mm Hg. RVOT construct showed moderate or severe regurgitation in 13; peak gradient being a median of 28 (6-74) mm Hg.
Conclusions: Long-term survival after surgical repair of Truncus arteriosus is good. However, the survivors suffer significant number of reoperations and catheter interventions on the truncal valve as well as the RVOT-pulmonary artery complex before reaching adulthood. Future efforts needs to be directed to reduce the long-term morbidity.



P237

Management of an aortic dissection in a 9-year old boy with Loeys-Dietz-Syndrome

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Introduction: Loeys-Dietz syndrome (LDS) is an autosomal-dominant connective tissue disorder characterized by aortic aneurysm, generalized arterial tortuosity, hypertelorism and split uvula/palate. The vascular lesions tend to be aggressive with a high incidence of aortic adverse events even at younger age (especially LDS type 2). Early and extensive surgical strategy is occasionally needed to prolong the patient's life expectancy.
Methods: A 9-year old boy (26kg, 130cm) with known diagnosis of LDS 2 (TGFBR 2-mutation) and dilated aortic root (31mm, Z +4.4) on Losartan-therapy presented with acute back-and abdominal pain. The diagnosis of Stanford type B dissection was made by computed tomographic angiography (CTA) starting from A. subclavia sinistra with a distal reentry at the level of Truncus coeliacus. Lacking any end organ malperfusion an aggressive blood pressure lowering therapy was initiated. Frequent follow-up CTA revealed rapid dilatation of the abdominal aorta from 27x26 to 38x35mm within 8 days. Emergency surgical treatment was decided starting with a valve sparing aortic root replacement and additional arch replacement (frozen elephant-trunc technique); a thoracic-abdominal aortic replacement (Dacron 20mm) followed two days later (Figure 1). The patient was discharged after 6 weeks of hospitalisation with Irbesartan, Atenolol and antiplatelet therapy.

The story went on and unfortunately 3 months later the boy presented with asymptomatic progressive aneurysm of both subclavian arteries and right-sided dissection (right 12mm to 23mm; left: 14mm to 22mm). This was successfully resected and replaced by an rightsided interponat. Only 2 month later a implantation of a protheto-biiliacal, alloplastic aortic prothesis with reinsertion of mesenteric artery and both renal arteries was mandatory due to ongoing aneurysm formation (21mm to 39mm) of the abdominal aorta. Subsequently the left subclavian artery needed replacement.
Results: 18 month after first surgery and finally complete replacement of the entire aorta the meanwhile 11 yo boy is asymptomatic without any neurological impairment.
Conclusions: LDS 2 can present with severe vascular findings and dramatic deterioration even in younger age. In the presented case an early and aggressive surgical approach with replacement of the entire aorta has been successful. However the underlying disease is not healed and may cause further vascular events and challenge future management.

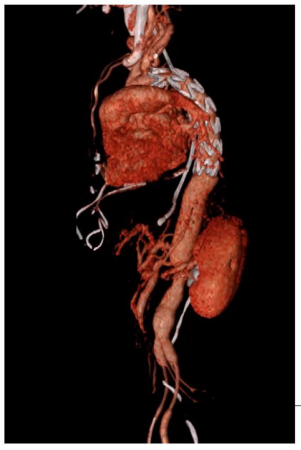

Figure 1: CTA after valve sparing aortic root replacement, aortic arch elephant trunc replacement and thoracic-abdominal aortic replacement.

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P238

Midterm follow up total cavopulmonary connection with atrioventricular valve insufficiency in children: single centre experience

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Ukrainian Children`s Cardiac Center (1), Shupyc National Medical Academy of Postgraduate Education (2)

Introduction: Children with single ventricle physiology (SV) and moderate/severe atrioventricular valve insufficiency (AVVI) may have increased risk compared to those without attendant AVVI. The presence of significant insufficiency systemic atrioventricular valves (AVV) negative impact on survival and the quality of life. Obiective:to estimate midterm follow-up in children with SV and AVVI after simultaneous surgical valve correction together with total cavopulmonary connection (TCPC).
Methods: From 2005 to 2015 years in our centre the TCPC was performed in 132 patients. Twenty four (18%) of then had significant (moderate or severe) insufficiency of systemic AVV. Median age was 8.8 years (range 4.6 to 17.8 years), median body weight was 28.8 kg (range 13.8 to 66 kg). By anatomy of SV 5 (20.8%) patiens had DILV, 4 (16.7%) - TA, 3 (12.5%) - unbalanced AVSD, 3 (12.5%) - hypoplasia MV, LV, 3 (12.5%) - DORV, 2 (8.3%) - TGA, 2 (8.3%)-heterotaxy syndrome, 2 (8.3%) - CCTGA. For 12 (50%) patiens there were perfomed surgical correction AVV (8 patiens (33.3%) - combination of annuloplasty and commissuroplasty, 4 (16.7%) patiens-leaflets repair,4 (16.7%) patiensclosure AVV. Eight patients (33.3%) patiens didn`t have any intervensions for AVV.
Results: Median follow-up was 5±3.1 years. Three (11 %) patients had recurrence of AVV already at the hospital stage,12 (50%)- in the midterm follow-up. Risk factors progression were tricuspid valve, common AVV, treatment without valve ring stabilization. One patient (4 %) received repeated plasty of AVV one year after correction. Freedom from AVV reoperation at 5 years was 92 %.
Conclusions: Patients with SV and AVVI before need an individual approach to treatment choice. Significant insufficiency requires surgical correction AVV.

P239

Neonatal complex arch reconstruction using ducal patency for lower body perfusion: Long-term growth and function

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Introduction: In suitable neonatal morphologies, we have pursued a modified technique of enlarging the roof of the distal arch and extended resection (without CPB) by capitalizing on the neonatal ductal perfusion. This study evaluates the long-term outcome of this approach.
Methods: 11 consecutive children operated upon by 2 authors (2006-2019) were included. Median age and weight were 5±3 days and 3.3±0.6 kg respectively. Associated malformations included ASD (10), VSD (7), complete AVSD (2) and others (4). Left posterior muscle-sparing thoracotomy was performed. The roof of the arch intervening L Carotid-L Subclavian arteries was enlarged, using a non-ischaemic clamping (45±11 min); upper body perfusion through the brachiocephalic trunk and lower body perfusion through the patent duct. After adequate reperfusion, a standard resection of coarctation and extended end-to-end anastomosis was performed (ischaemic clamp time 24±4 min). PA banding was performed in 4 patients (See Image). Patch material used included treated autologous pericardium (4), Xenopericardium (5) and autologous arterial tissue (2). Duration of ventilation was 1.7±1.3days; hospital stay was 36±12days. 6 patients underwent corrective operation mostly for VSD or AVSD repair in course of their infancy. Follow-up was 100% complete with a mean duration of 9±4years.
Results: Early and late mortality was 0%. No patient developed any neurological complication. 2 reimplanted LSA underwent anastomotic revision/thrombectomy early postop; 1 patient needed balloon angioplasty of isthmus at 6 months. Only 1 patient with a preoperative proximal arch hypoplasia had a significant gradient (18 mmHg) at follow-up. All children enjoy good general condition with subjective normal exercise tolerance. No patient is on antihypertensive medication. Clinical gradient across the distal arch is 0±13 mmHg. Echocardiography shows a gradient of 6±4 mmHg and normal LVEF. Z value of the ascending aorta, arch and isthmus were 0.8 ±1.6, -1.5±1.3 and -0.5±0.9 respectively.
Conclusions: This technique of arch reconstruction without CPB is a less invasive, reproducible and transferable option for selective neonatal hypoplastic arch repair. It renders a more harmonic shape to the arch, allows consequential growth without development of relevant gradients and leaves children free of hypertension.

P240

Older age at repair and arm-leg blood pressure gradient at follow-up predict systolic hypertension after treatment for coarctation of the aorta

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Introduction: Long-term morbidity and mortality following Coarctation of the Aorta (CoA) repair has been reported to be linked with hypertension. We evaluated prevalence and perioperative predictors of hypertension 10 years after primary native CoA repair in a population based national cohort.
Methods: We collected the most recent blood pressure and growth data of 304 patients with isolated native CoA treated either by surgery or cath intervention in Finland 2000-2012. Systolic blood pressure (SBP) z-scores were generated based on US fourth report data (<18 years) and FinHealth2017 data (≥18 years). Systolic hypertension was defined as SBP ≥ +2SD or as use of blood pressure medication at latest follow-up. Arm-leg-SBP-gradients was calculated as the difference between right arm and ankle oscillometric cuff measurements.
Results: In all, we collected follow-up data of 284/304 (93%) patients originally treated by surgery (n=235) or balloon angioplasty/stent (n=37/12). The median follow-up time from primary procedure was 9,7 years (range 3,3-18,1) and median age at follow-up was 11,8 years (range 3,3-29,5). Blood pressure medication was prescribed in 28/284 (9,8%) patients. Mean SBP was +0,9 SD and 34/280 (12,1%) had SBP-hypertension. Age at primary procedure was higher in patients with SBP-hypertension (median 0,6 years, range 0,0-17,4) compared with non-hypertension (0,1 years, range 0-16,8; p<0,01). Further, at follow-up patients with SBP-hypertension were older (median 13,3 years, range 4,0-27,0) compared with non-hypertensive patients (11,3 years, range 3,3-29,5; p<0,01). However, follow-up time was not associated with SBP-hypertension. Arm-leg-SBP-gradient at follow-up was higher in patients with SBP-hypertension (mmHg, mean±SD, -0,3±14,6) compared with non-hypertensive patients (-6,4±11,6; p=0,048). In logistic regression, older age at

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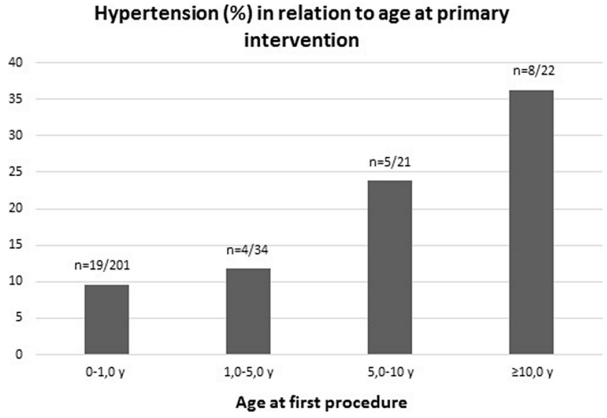
primary procedure and higher arm-leg-SBP-gradient at follow-up were independent predictors of SBP-hypertension.
Conclusions: Development of SBP-hypertension is common after CoA treatment, and related with older age at primary treatment. In the present cohort, SBP-hypertension was also associated with an arm-leg-SBP-gradient.



P241

Outcome of univentricular heart palliation in the context of associated truncus arteriosus

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Introduction: The association of a univentruicular heart defect with truncus arteriosus (UVH-TA) is extremely rare. There is a lack of population based outcome studies reported in the literature
Methods: Single centre retrospective outcome review of cases presenting with UVH-TA over a 30 year period (1990-2020). The hospital records including case notes, echocardiographic and other imaging modality data, out patients records, operation notes and other electronic data were reviewed. Patient characteristics, primary and associated cardiac lesions as well as the need for surgical intervention and the final outcome of surgery were observed.
Results: Six cases (2 male) with UVH-TA presented over a 30 year period. Five had complete unbalanced atrioventricular septal defect (uAVSD) (83%) and one (17%) had tricuspid atresia (TA) associated with truncus arteriosus. All had antenatal diagnosis. Two cases (33%) were excluded from initial surgical palliation due to Trisomy 21 in 1, and severe truncal valve regurgitation in 1. Initial surgical palliation was performed in 4 cases (67%) at median of 31 days (2-60) and consisted of disconnection and reconstruction of the pulmonary arteries and establishing controlled pulmonary blood flow. There were no early deaths. Conversion to cavopulmonary shunt was not possible in 2 due to PA anatomy in 1 and severe airway problems in 1. They died at 14 and 17 months respectively. 2 patients (33%) underwent CP shunt with 1 being alive (17%) at 6 months – 1 months after CP shunt. The second patient proceeded to Fontan completion at 2.3 years but required catheter takedown 3 months later. He died at 5.3 years of age.
Conclusions: Univentricular heart defects associated with truncus arteriosus carry extremely poor short to medium-term outcome. This should be considered during antenatal and postnatal counselling and decision making.

P242

Problems of elevated pulmonary pressure in the NICU patient- differentiation in the most critical patients

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Introduction: The neonate presents with a vast range of problems in the Neonatal Intensive Care Unit and the Pediatric Cardiologist is asked to consult for various reasons. Differential diagnosis begins when the neonate presents with clinical features of pulmonary hypertension, different degrees of cardiac failure and echocardiographic signs of pulmonary hypertension. The aim of the study was to look into the problems of neonates with echocardiographic signs of elevated pulmonary pressure as a major reason for cardiological consultations.
Methods: A retrospective analysis of the cardiological consultations reported by the Department of Intensive Care and Congenital Malformations of Newborns and Infants (NICU) between 2018-2019 to the Department of Pediatric Cardiology of the Polish Mother's Memorial Hospital Lodz, Poland regarding patients with signs of elevated pulmonary pressure.
Results: Between 2018-2019 a total number of 1245 patients were hospitalized in the NICU, during this time a total number of 1108 cardiological consultations were performed for different reasons. The total number of consultations (planned and urgent) due to elevated pulmonary pressure amounted to 244 (22 %), with certain patients requiring repeated evaluations. Each patient upon echocardiographic assessment presented with signs of elevated pulmonary pressure and different degrees of compromised systemic output. We observed dilated right heart chambers with flattening or bowing of the intraventricular septum and right ventricular hypertrophy. Different degrees of tricuspid regurgitation were observed but all high velocity. Patients with patent arterial ducts shunted from right to left. Depending on underlying cause each patient presented with different degree of compromised systemic output, the most severe cases showed underfillment of left atrium and left ventricle with insufficient systemic flow and impaired abdominal aortic pulsation. A summary of the patient population presenting with systemic/suprasystemic pressure noted upon echocardiographic examination and implemented treatment is presented in table 1.
Conclusions: The differential diagnosis of the neonate in severe clinical condition with echocardiographic signs of increased pulmonary vascular resistance remains a challenge. Signs of persistent pulmonary hypertension in the newborn, whether primary or secondary, besides congenital heart defects are a major cause for planned or urgent cardiological consultations in the neonate and remain a major problem in NICU patients.

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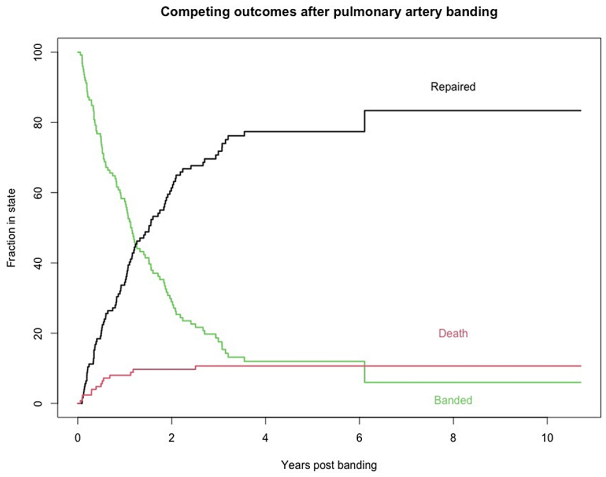
Irregularity	No of cases	Main treatment	Need for Sildenafil	Need for iNO	Need for ECMO	Additional ECHO findings
Pulmonary sequestration	2	Surgery / interventional treatment	(-)	(-)	(-)	(-)
Combined arteriovenous and lymphatic malformation of the neck	1	Propranolol, interventional treatment	(-)	(-)	(-)	Retrograde flow aortic arch
Cerebral arteriovenous fistula	3	Ventilation, inotropic support, qualification for interventional treatment	(-)	(-)	(-)	Retrograde flow aortic arch
Congenital Cystic Adenomatoid Malformation	2	Surgical treatment	(-)	(-)	(-)	(-)
Patent arterial duct with bidirectional shunting	6	iNO followed by Surgical PDA ligation if needed	(+) after ligation	(+)	(-)	Doubts regarding aortic isthmus
Bronchopulmonary dysplasia	9	Oxygen therapy + assisted ventilation	(+)	(+)	(-)	Hyperechogennic lungs impeding assessment
Congenital Diaphragmatic Hernia	20	Surgery	6 cases required after surgery	14 cases required	8	May be accompanied by various degrees of aortic arch narrowing
Pneumonia	2	Targeted treatment, ventilation, inotropic support	(-)	(+)	2	Hyperechogennic lungs impeding assessment
Right pulmonary artery embolism	2	Alteplaza i.v	(-)	(-)	(-)	Embolus located in RPA, almost absent flow through MPA
Omphalocele with signs of PH	3	Clinical compromise after surgery. Ventilation	(-)	(+)	(+) 1 case	Dextroposition of heart
Sepsis	4	Targeted treatment, ventilation, inotropic support	(-)	(-)	(-)	Frequent - regurgitation of other valves
Meconium aspiration syndrome	5	Surfactant lavage, ventilation	(-)	(-)	(-)	Hyperechogennic lungs impeding assessment
Intrauterine premature constriction of ductus arteriosus	5	Ventilation (invasive/ noninvasive)	(-)	(+)	(-)	No duct first hour after delivery
Intrauterine premature constriction of foramen ovale	4	Ventilation (invasive/ noninvasive)	(-)	(-)	(-)	Restrictive FO with thickened septum
Pulmonary vein stenosis with normal 4-chamber view	1	Surgical treatment	(-)	(-)	(-)	Turbulent flow pulmonary veins,

Table 1: Patient population summary with implemented treatment. Number of patients with systemic/suprasystemic pulmonary pressure noted upon echocardiographic examination requiring the most intensive treatment.

Conclusions: In a cohort with a high incidence of comorbidity, PAB carries significant risks of reintervention and of mortality. In the absence of a comparator group or randomization, comparison of the relative risk of this approach to that of primary repair in high-risk patients is difficult. PAB is successful in achieving weight gain prior to definitive repair in the majority but does not lessen the incidence of heart block following repair.



P243
pulmonary artery banding – still a role for staged biventricular repair of intracardiac shunts?

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Introduction: Although primary intracardiac repair has become increasingly safe in small infants and neonates, pulmonary artery banding (PAB) remains a useful palliation in patients with additional complexity. We reviewed outcomes in this group over a 10-year period.
Methods: Retrospective cohort study including all patients who underwent PAB between January 2010 and December 2019, excluding single ventricle physiology and left ventricular training.
Results: During this period, 125 patients underwent PAB (2.5% of all surgical procedures) and were followed up for a median of 4 years. The median age at banding was 41 (2-294 days) and weight 3.4 (1.8- 7.32 kg). Multidisciplinary team consensus was for PAB due to significant comorbidity in 81 (64.8%) and for complexity of anatomy in 44 (35.2%). The median hospital stay was 14 (4-303 days) and 14 patients (11.2%) required complete intra-cardiac repair during the same admission. After discharge, 19/105 patients (18.1%) needed interstage readmission due to respiratory infection and 18 (14.4%) required unplanned re-intervention. Six patients (4.8 %) died during the initial inpatient admission and 5 following discharge (4%) for a total interstage mortality of 8.8%. Full repair was completed in 93 (74.4%) at a median age of 13 (3.1-49.9) months and weight of 8.5 (3.08-16.8) kg. Prior to PAB, 54% were below the 0.4th weight centile, while at the time of repair, only 28% remained so. Following repair, 5 (5.5%) developed complete heart block requiring permanent pacemaker, and 11 (11.8%) needed other re-interventions after repair. The post-repair mortality was 6/93 (6.5%), bringing the overall mortality of the staged approach to 17/125 (13.6% - See Figure 1). The great majority of deaths were on review considered to be related to non-cardiac comorbidity.

P244
Realignment of the ventricular septum in Tetralogy of Fallot using direct partial closure – long-term follow-up

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Introduction: Aortic dilatation and insufficiency after surgical repair of Tetralogy of Fallot (TOF) have been described, and beside an intrinsic aortopathy with wall structure abnormalities, unfortunate alignement of the left ventriculare outflow tract (LVOT) after VSD patch closure is a risk factor for these complications. In 2011, the influence of realingement of the left ventricular outflow tract (LVOT) by (partial) direct closure of the ventricular septal defect (VSD) in Tetralogy of Fallot (TOF) was assessed in our institution. We aimed to evaluate the follow-up of this cohort and to compare the results to a matched group of TOF patients with classical VSD patch closure.
Methods: In total, 40 patients with TOF treated between 2004 and 2010 are included in the study, with 20 patients each in the (A) VSD direct closure and (B) patch closure group. Follow-up time after surgery was 12.8 years (± 1.6).
Results: Baseline characteristics including age, weight, height, and echocardiographic estimations of VSD size, size of the aortic annulus and ascending aorta, angle between interventricular septum (IVS) and anterior aortic annulus (aAoA), gradient across the right ventricular outflow tract (RVOT) and surgical and ICU parameters (bypass time, cross-clamp time, duration of ventilation, ICU stay) were not significantly different between groups A und B. After surgery and during long term follow-up, realignement of the LVOT shown by the IVS/aAoA-angle was better in group A (34 vs 45°, $p < 0.0001$). No differences in LVOT size, aortic

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annulus size, aortic regurgitation or dilation of the ascending aorta and RVOT gradients were found. Postoperative, rhythm disturbances were found in 3 patients in each group, with only one complete AV-block remaining in group B.
Conclusions: (Partial) direct closure of the VSD in TOF showed comparable long-term results without higher risk for rhythm disturbances after surgery but with a better realignement of the LVOT. As it is known, that dilations of aortic annulus and ascending aorta with consecutive aortic insufficiencies are described after classical patch repair of VSD in TOF, this new surgical approach can be beneficial concerning this unfavourable outcome.

P245
Retrospective evaluation of the effects of arginine vasopressin in the postoperative management of patients with Hypoplastic Left Heart Syndrome
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Introduction: The postoperative course of Hypoplastic Left Heart Syndrome (HLHS) patients remain very challenging. Low Cardiac Output Syndrome (LCOS) is common, requiring meticulous management with inotropic and vasoactive drugs. Arginine vasopressin (AVP) represents an interesting alternative to other drugs used to modulate vascular tone, because of its distinctive effect of increasing systemic blood pressure and tissue perfusion without increasing heart rate or myocardial oxygen demand. Our hypothesis is that the use of vasopressin, when initiated within 24 hours of Cardiopulmonary Bypass (CPB), leads to improvements in the postoperative outcomes, length of stay and complication rate.
Methods: All patients with HLHS having undergone surgery between March 2014 and March 2017, at Children's Hospital Colorado, were included in this retrospective cohort study. Charts were reviewed from study subjects identified from existing databases. Patients were stratified into those who received AVP (AVP group) and age, diagnosis and surgical procedure-matched control patients who did not receive AVP (Control).
Results: Of the 226 records reviewed, 69 patients who received AVP postoperatively were included. The distribution of the patients according to the surgical palliation stage was as follows, for AVP and non AVP receiving subjects respectively: Stage 1, 56.2% vs 34.0%, Stage 2, 7.5% vs 46.1% and Stage 3, 36.2% vs 19.9%. On average, AVP group had significantly lower pre AVP administration mean blood pressure (-13.51 mm Hg) than controls, but was 1.97 mm Hg higher 2 hours post administration ($p<0.0001$), following an increase over 24 hours for both groups, with a slightly higher mean blood pressure in non the AVP group (-0.12 mmHg, $p>0.273$). Average pre AVP administration lactate in the AVP group was 1.53 mg/dL significantly higher than control group ($p<0.002$), trended lower 2 hours post administration (0.142 mg/dL, $p<0.26$), and remained significantly lower over the 24-hour period (-0.066, $p<0.01$). No significant difference was observed on the VIS score or total fluid given over 24 hours between the groups prior to the AVP administration ($p>0.05$).
Conclusions: Considering lactate and VIS scores for both groups, the use of vasopressin seems to provide good hemodynamic results. Further studies are recommended to better characterize AVP in this population.

P246
Risk factors for prolonged mechanical ventilation after congenital heart surgery in pediatric patients.
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Introduction: Prolonged mechanical ventilation (PMV) is a well-recognized factor as a quality metric for pediatric cardiac surgical programs. It is associated with complications and financial recourse utilization. In recent years studies, a lot of risk factors for PMV are described and analyzed. Even more, some authors had established predictive models to detect proactively patients in risk for PMV. This study aims to analyze risk factors for PMV after congenital heart surgery in pediatric patients in our center.
Methods: Medical files of patients 0-18y who underwent heart surgery in 2016 and 2017 were reviewed. Patients that met the inclusion criteria were studied. PMV was defined as invasive mechanical ventilation ≥96h. The patients were divided in two groups according to duration of mechanical ventilation: group 1- patients with PMV, group 2 - patients without PMV. The risk factors were divided in preoperative, operative and postoperative. Data were presented as medians with IQR or as means ± standard deviation. A non-parametric Mann-Whitney U test, binomial logistic regression test and ROC curve analysis integrated in the statistical software SPSS 24.0 were used. A value of $P< 0.005$ was considered significant.
Results: 438 patients were operated in 2016 and 2017 and 384 of them were included in the study. 80 patients (20.8%) needed PMV (group 1) and 304 (79.2%) did not need PMV (group 2). The following risk factors for PMV were established: younger age with cut-off value of ≤ 270 days with AUC 0,820 (95% CI 0,777-0,857) with sensitivity of 90% and specificity of 60.86%, preoperative procedures with OR 9.97, prematurity with OR 2.76, malnourishment with OR 2.07, delayed sternal closure (DSC) with OR 21.29, RACHS-1 with OR 6,96, sepsis with OR 39.86 and failed extubation with OR 9.98.
Conclusions: In our center the strongest predictors for PMV are younger age, postoperative systemic infection, DSC, need for preoperative procedures and higher RACHS-1 scores. Focusing on these factors, makes it possible to detect patients in risk for PMV.

Table 1: Odd's ratios or AUC for the probability for prolonged mechanical ventilation after congenital heart surgery in pediatric patients

Risk factors for prolonged mechanical ventilation	OR/AUC	95% Confidence interval		p
		Lower limit	Upper limit	
Preoperative				
○ Age ≤ 270days	**0,820**	0.777	0,857	0.001***
○ Prematurity	**2.76**	1.42	5.38	0.002***
○ Malnourishment	**2.07**	1.25	3.44	0.004***
○ Preoperative procedures	**9.97**	4.89	20.32	0.001***
Operative				
○ Delayed sternal closure	**21.29**	10.49	43.21	0.001***
○ RACHS-1	**6.96**	4.14	12.42	0.001***
Postoperative				
○ Sepsis	**39.86**	14.80	107.35	0.000***
○ Failed extubation	**9.98**	4.12	24.14	0.000***

*** - Statistical significance for $p \leq 0.005$

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P247

Single center experience of enhanced training of morphologically left ventricle in congenitally corrected transposition of great arteries

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Introduction: Method of enhanced training of (eLVT), proposed by Peter Zartner (Germany) in 2015 - atrioseptostomy plus pulmonary artery banding (PAB) heightened strength of morphologically left ventricle (mLV) in ccTGA by increasing both preload and afterload. We want to demonstrate that it is enough for double switch (DS) preparing without supplementary surgeries.
Methods: Between April 2015 and November 2019 eLVT was performed on 11 patients. There were 8 patients with isolate ccTGA, 2 patients - after conventional surgeries, one has ccTGA, VSD. Decreased contractility of morphologically right ventricle (mRV) and moderate-to-severe tricuspid regurgitation (TR) became the indications for procedure. Median age was 37 months (3 weeks - 9 years). Intraatrial septum was intact in 7 patients, 4 had patent foramen ovale.
Results: Postoperative course was uneventful in all patients. The median time of ICU stay was 2 days. Echocardiography follow-up was fulfilled every 3 months. The median time before MRI and cardiac catheterization studies was 14 months. At that moment echo data revealed an increase of the LVi mass from a mean 28 g/m2 to 51 g/m2 ($p < 0,01$), mean LVi volume increased from 25,5 ml/m2 to 54 ml/m2 ($p < 0,001$). The mean echo gradient across PAB was 77 mm Hg. MRI confirmed all echo data. The mean invasive pressure in mLV was 96 % of mRV. Shift of intraventricular septum to the mRV, good ventricular function with concentric contraction of mLV, reduction of TR degree were observed in vast majority of patients. 8 patients underwent DS procedures at mean time 22,5 months after the first surgery in 3 different centers. 2 are still waiting for it. There was one eLVT fail: 7 y.o. girl after conventional surgery didn't show any difference in mLV mass, volume and pressure in it. Closing ASD with fenestration and loosing of PAB was done 3 years after. There was one early death (12,5 % mortality) after DS due to myocardial infarction. The median follow-up after the DS is currently 20 months. Stable LV function was observed in all patients.
Conclusions: ELVT is an effective method of mLV preparation for DS in short period of time without additional procedures.

P248

Successful single ventricle palliation of common arterial trunck, unblanaced atrioventricular septal defect and interrupted aortic arch

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Introduction: Common arterial trunk (CAT) is a relatively rare cardiac anomaly accounting for less than 1% of all congenital heart lesions. Its association with unbalanced atrioventricular septal defect (uAVSD) is very rare. Interrupted aortic arch (IAA) is present in approximately 15% of patients with CAT, but successful management of a patient with the combination of CAT with uAVSD and IAA has not been reported in the literature. We describe our management of such a case from fetal life through to second stage univentricular palliation.
Methods: A baby boy was delivered by elective Caesarean section at 39 weeks' gestation weighing 3.83 kg with antenatal diagnosis of uAVSD on the hypoplastic left heart syndrome spectrum. Postnatal echocardiogram demonstrated unbalanced AVSD with significant hypoplasia of the left ventricle. A cardiac computed tomography angiogram confirmed presence of short common truncal root arose from the dominant right ventricle. The aortic arch was interrupted between the left common carotid artery and left subclavian artery (Type B). There was a large ductal arch supplying the descending aorta which arose from the superior aspect of the pulmonary trunk (Figure 1).
Results: First stage univentricular palliation was undertaken at 7 days of age. The pulmonary arteries were detached in continuity from their common truncal origin, uncrossing the origins, and the bifurcation augmented using a patch of pulmonary artery homograft. A 5mm Polytetrafluoroethylene (PTFE) conduit was anastomosed to the RPA and a right ventriculotomy. Echocardiographic concern of evolving RV-PA conduit narrowing prompted balloon dilatation of the proximal RV-PA conduit at 3 months of age. The cavo-pulmonary (CP) shunt and PA plasty was done at around 5 months of age and 7.6 Kg weight. The child was discharged home on day 7 and doing well.
Conclusions: Our case represents a rare and equally complex constellation of congenital cardiac defects, which are individually challenging in their own right. The combination of these three lesions has a guarded short-term prognosis but the child was able to undergo first and second stage of univentricular palliation with no significant complications. Although a smooth journey thus far, the possibility and outcome of future interventions, remains guarded.

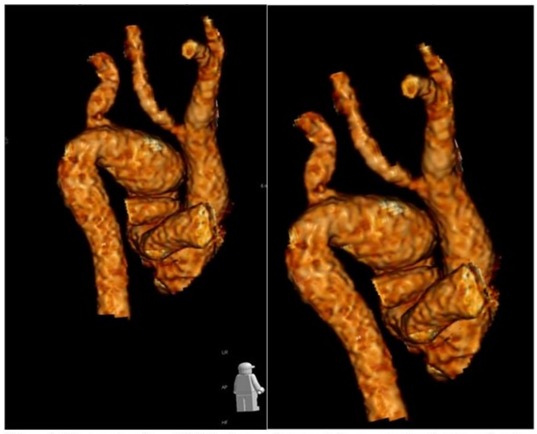

Figure 2: 3D reconstruction of CT Angiogram from posterior aspect, showing truncal root, criss-crossing of PA's. Type B interrupted aortic arch with ascending aorta giving rise to Right Brachio-Cephalic Artery, Left Common Carotid Artery, and ductal arch connecting to descending aorta which gives rise to Left Subclavian artery.

P249

Surgical options in patients with different anatomical types of congenitally corrected transposition of the great arteries

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Introduction: Surgical treatment of CC-TGA includes a vast majority of interventions depending on different anatomical types of cc-TGA

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Methods: Were analyzed surgical options in consequent young patients with different types of CC-TGA.
Results: Between January 2007 and December 2018 we observed 101 patients at the age of 0-18 y.o. with CC-TGA. Isolated cc-TGA was diagnosed in 23 (22,7%) patients and 78 patients (77,3%) had associated defects. Situs solitus was found in 78 patients (77,2%), dextrocardia – in 21 patients (20,8 %),VSD – in 64 (63 %), TV insufficiency severe – in 12 (12 %), PS – 21 (21 %), PA I-II – 14 (14%), MAPCAs – 2 (1,9%), CoA – 6, TAPVC – 1 (0,9%). Sixty patients (59,4%) underwent surgical repair, 109 operations were performed. Four patients were operated with isolated cc-TGA and 56 patients – with associated defects. Double switch operation was performed in 16 patients at the age 65,9 mo ± 50,1 mo. Intraoperative mortality was 6,25% (n=1). Pulmonary artery banding before DSO was performed in 9 patients and 4 patients underwent pulmonary banding with coarctation repair. Three patients had reoperations during follow-up – two patients due to pulmonary veins collector obstruction and one due to residual VSD. Aorto-pulmonary anastomosis was performed in 19 patients, central shunt - in two patients. Glenn shunt was performed in 20 patients ant 15 patients underwent TCPC with intraoperative mortality 0%. One patient underwent Kawashima operation. One patient died 5 months after TCPC probably due to arrhythmia. Pulmonary artery banding was performed in 25 patients at the age of 0,1 month - 94 mo (mediana 6 mo), including 6 patients with pulmonary artery banding and aorta coarctation repair. Total mortality was 15,7 % (3 patients died, including one intraoperative death). TV plasty was performed in 2 patients and one patient underwent TAPVC repair with mixed type (intracardiac + supracardiac). VSD closure with PA obstruction repair was performed in one patient
Conclusions: CC-TGA is a complex congenital heart disease associated with other congenital heart defects. Presence of associated congenital heart defects needs specified surgical correction - palliative operation, physiologic or anatomic repair, with good early and long-term follow-up results

P250
Surgical treatment of cardiac fibromas in different ventricle localization in children.

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Introduction: Primary cardiac tumors are rare in children, with a prevalence of 0.0017–0.28% in autopsy series. Cardiac fibromas (CF) represent the second most common benign cardiac tumor in the pediatric population following rhabdomyomas. Fibromas are usually single and intramural and are mainly located in the septum and the free-wall of the left ventricle. Even though these tumors show a slow rate of growth, sudden deaths from malignant ventricular arrhythmias have been described in approximately 10% of patients with this pathology.
Methods: Between February 2004 and September 2019 nine consecutive children (5 males and 4 females) were diagnosed with intramural fibromas in different ventricular localizations. One patient had a right ventricle CF, five of them had free-wall left ventricle localization, two patients had septal localization of CF. They underwent complete (N=7) or partial (N=1) primary preventive surgical resection. Surgery was performed on cardiopulmonary bypass (CBP) using aortic and bi-caval cannulation with aortic cross-clamping and antegrade blood cardioplegia. The remaining patient received an implantable cardioverter-defibrillator (AICD) in secondary prevention after being resuscitated from cardiac arrest. Tumor resection was not performed due to parents' disagreement.
Results: Median age at surgical intervention was 2,57 (Q1 0.8 - Q3 3,5) years. Median follow up was 6,3 (Q1 1,3 - Q3 10) years. There were no early or late deaths. There were three surgical re-interventions. Two patients had the revision of the tumor cavity due to bleeding at the time of primary surgery. The patient with AICD had AICD revision for subcutaneous array displacement and a high defibrillation threshold. All patients are in NYHA functional class I with normal ventricular function. No ventricular arrhythmias have been detected after excision of CF in the follow-up.
Conclusions: In this small series primary preventive surgical CF resection was an effective strategy to prevent life-threatening ventricular arrhythmias and was associated with excellent survival and well-preserved ventricular function.Supported by grant MH CZ - DRO, 00064203

P251
The borderline left ventricle in neonates – help in decision making for treatment

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Introduction: There exists no golden standard for decision making to either strive for uni- or biventricular repair of borderline left ventricles, although several echocardiographic and morphologic scores have been developed. The variety of the disease complex does not allow to set one single approach, but requires additive diagnostics.
Methods: Failing of established definitions, we follow international consensus and define the borderline LV with 3 basic measurements: mitral valve annulus ≤8mm, aortic valve annulus ≤4.5mm and a non-apex forming LV in the echocardiographic 4 chamber view. We show different treatment pathways in our series of 12 neonates (weight 2.2 to 3.1 kg) with borderline LV. We performed interventional and surgical treatment, also in combination. Procedures included balloon aortic valvuloplasty (BAV) and arch stenting (n=3), fetal BAV followed by a neonatal Ross (n=1), a Giessen procedure (n=2), and a primary Norwood type univentricular palliation (n=6).
Results: Of 3 patients with initial transcatheter palliation, 2 have continued to do well with biventricular status (including staged removal of the arch stent and surgical repair of the arch), while one infant developed pulmonary hypertension (PA pressure at 80% of systemic, but surviving). The neonate with the Ross procedure could not be weaned from ECMO and parents refused the univentricular approach; cardiac catheterization 4 weeks post-surgery demonstrated an LVEDP of 23mm Hg; the patient subsequently died. Both infants with a primary Giessen type palliation were converted to a Norwood type palliation at 4 and 6 weeks respectively, due to absence of LV growth. All 6 patients with primary univentricular palliation survived; 4 of them have proceeded to second stage Glenn palliation and 2 are awaiting surgery. In two of these 6 patients, preoperative cardiac catheterization demonstrated LVEDP of 18 and 20mm Hg respectively, which enabled the choice of single ventricle palliation.
Conclusions: Choosing appropriate candidates for biventricular repair is challenging. In case of failing clear treatment pathway,

invasive preoperative measurement of LVEDP might help. Further studies are necessary to determine critical LVEDP limits.

P252

The long term fate of subaortic stenosis in childhood

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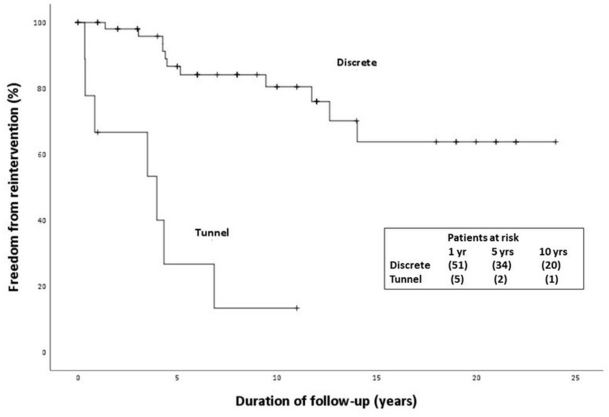
Introduction: Subaortic stenosis (SAS) is a common cause of left ventricular outflow tract (LVOT) obstruction. It can be divided into a discrete form consisting of a circular membrane or fibromuscular ridge and a long tunnel-type. The ideal timing of intervention remains controversial. While immediate surgical results are satisfying, reoperation rate amounts up to 20 % and surgery.
Methods: We conducted a retrospective study of 84 children (0-18y) diagnosed with SAS between 1992 and 2017. General characteristics, hemodynamic and surgical data were reviewed.
Results: Seventy-one patients underwent surgery. Median age at diagnosis was 2.3 (IQR 5.3) years and peak gradient 31.8 (± 24.6) mmHg. Discrete SAS was present in 89 % (63/71) and tunnel-shaped SAS in 11 % (8/71). Median age was 3.4 (IQR 7.3) years at intervention and peak gradient was 53.0 ± 31.3 mmHg. Reoperation rate was 20 % in case of discrete SAS and even 75% in patients with tunnel-shaped SAS ($p = 0.003$). Reoperation was associated with higher residual peak gradient immediately after surgery ($p = 0.003$), irrespective of the type of surgery (resection +/- myectomy). Tunnel-shaped SAS, small aortic annulus, age < 2 years at initial intervention and absence of VSD were other predictors of reoperation. Aortic insufficiency (AI) was present in 41 % of the patients before intervention and in 37% after. Prevalence of AI at latest follow-up was 61%. Thirteen patients had only conservative followup. Their median age at diagnosis was 3.8 (IQR 8.0) years and peak gradient at diagnosis was 11.6 ± 12.3 mmHg. Median follow-up time was 9.3 (IQR 6.2) years with a peak gradient of 18.5 ± 12.5 mmHg at latest follow-up. Peak gradients were significantly lower ($p = 0.001$) and median age was significantly higher at diagnosis in this group ($p = 0.04$). AI was present in 15% of patients at diagnosis and in 54% at latest follow-up.



Conclusions: SAS remains a complex disease. Reoperation rate remains high, especially in tunnel-shaped SAS and depends on the early postoperative gradient, small annulus, and young age, but not on the type of operation. Low initial gradient and late diagnosis are predictors for a milder disease. AI is frequently associated and is often progressive, despite surgery.

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The results of complete valve-sparing repair of tetralogy of Fallot

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Introduction: Tetralogy of Fallot (TF) is the most common congenital heart defect with cyanosis. Preserving pulmonary valve during surgery can provide long-term benefits for quality of life, heart function, and risks of rhythm disturbances. Special protocol with the aim of maximum preservation of pulmonary valves during early primary complete elective repair was created in our center. The aim of this study was to analyze the immediate results after the valve-sparing complete repair of tetralogy of Fallot.
Methods: Patients age 1-7 months with TF who underwent complete repair between January 2017 and October 2020 were included in our study. All patients were divided into two groups: valve-sparing repair (VSR) and transannular patch (TAP). Valve-sparing repair performed with annular dilation with Balloon or Hegar if necessary, and combined with leaflets mobilization and delamination techniques. Data were obtained from medical records and include echocardiography findings and surgical protocols. The dimensions of the pulmonary valve were assessed using Z-scores.
Results: Eighty-five patients were included in this study. The VSR group includes 54 (64%) patients. The VSR group had lower cardiopulmonary bypass time (<0.01), epinephrine support duration (<0.01), but higher pulmonary valve Z-scores than the TAP group (-2,5 vs -3,3 $p<0.01$).
Conclusions: Valve-sparing complete repair of tetralogy of Fallot is possible in more than 60% of cases in elective patients. Pulmonary valve preservation reduces cardiopulmonary bypass time and provides a more stable early postoperative period.

P254

Transesophageal echocardiographic predictor of significant right ventricular outflow tract obstruction after tetralogy of fallot repair

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Introduction: The optimal method for reconstruction of the right ventricular outflow tract in tetralogy of Fallot repair is yet to be established. Transatrial repair with pulmonary valve preservation avoids the detrimental effects of ventriculotomy and transannular patch, however because of moderately hypoplastic pulmonary valve these techniques are associated with undesirable residual right ventricular outflow tract gradients resulting in reoperations. The study sought to define intraoperative echocardiographic predictors

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of significant right ventricular outflow tract obstruction after tetralogy of Fallot repair.
Methods: From July 2016 to December 2017 58 patients with tetralogy of Fallot underwent repair with direct intraoperative pressures measuring and intraoperative transesophageal echocardiography performing after tetralogy of Fallot repair completion. Multivariate analysis was performed using logistic regression analysis to identify independent predictors of right ventricular outflow tract obstruction.
Results: One year after tetralogy of Fallot repair significant right ventricular outflow tract obstruction was present in 12 patients (20.6%): 10 patients (26.2%) after pulmonary valve sparing procedure and 2 patients (12.5%) after transannular patch (p=0.35).The AUC of the RVOT z-score value was significantly greater than that of Prv/Plv ratio, RV-PA gradient by direct measurements, RV-PA gradient by ITEE and TEE and its superiority was significant (p<0.0001, p=0.0013, p=0.0021, p=0.0016, respectively). A cutoff value of RVOT z-score for significant RVOTO development was -3.2 (AUC=0.979, p<0.001) with 95.9% sensitivity and 96.8% specificity. Multivariate analysis found that the right ventricular outflow tract z-score reached a statistical significance as a predictor of significant right ventricular outflow tract obstruction (p<0.0001).
Conclusions: Right ventricular outflow tract z-score < -3.2 measured by intraoperative transesophageal echocardiography is an indicator of postoperative right ventricular outflow tract obstruction right after surgery and a predictor of obstruction development following tetralogy of Fallot repair.

P255
Unexpected death in children with severe congenital heart defects in Norway 2004–2016
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Introduction: Despite major improvements in diagnostic and treatment modalities, congenital heart defects (CHDs) still represent an important cause of early mortality in children. Updated knowledge on the rates and causes of death among children with severe CHDs is needed to further improve treatment and survival. This study investigated nationwide mortality rates and clinical details in children with severe CHDs with emphasis on unexpected mortality unrelated to cardiac intervention.
Methods: Data on all pregnancies in Norway from 2004 to 2016 were extracted from mandatory national registries. Oslo University Hospital is the national centre for invasive treatment of children with CHDs, and information on live-born children with severe CHDs was retrieved from the Oslo University Hospital's Clinical Registry for CHDs, medical records and autopsy reports.
Results: Among 2359 live-born children with severe CHDs, 234 (10%) died before 2 years of age. Of these, 109 (46%) died in palliative care; 58 (25%) died unexpectedly per (n=3)-/postoperative (n=55); and 67 (29%) died unexpectedly and unrelated to a cardiac intervention, either before (n=26) or following (n=41) discharge after a cardiac intervention. We found no significant time trends during the study period. Comorbidity (38/67, 57%), persistent low oxygen saturation (SaO_2, <95%; 41/67, 61%), staged surgery (21/41, 51%), residual cardiac defects (22/41, 54%), and infection (36/67, 54%) were frequent in children who died unexpectedly unrelated to an intervention. Two or more of these risk factors were present in 62 of 67 children (93%). The main causes of death in children discharged from hospital after cardiac intervention were cardiac (15/41, 37%) and infection (23/41, 56%), and atrioventricular canal defect was the most frequent CHD (9/41, 22%).
Conclusions: The numbers of unexpected deaths unrelated to cardiac intervention in children <2 years of age without comorbidity were low in Norway. However, close follow-up is recommended for infants with comorbidities, persistent low oxygen saturation, staged surgery or residual cardiac defects, particularly when an infection occurs.

9. Heart failure

P256
A case of erythrokeratodermia-cardiomyopathy syndrome caused by dominant de novo DSP mutation
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Introduction: Erythrokeratodermia-cardiomyopathy syndrome (EKC) is caused by missense mutations which cluster tightly within a single spectrin repeat of DSP. The cardiomyopathy is characterised by left ventricular dilatation and appears to be malignant in nature; of three previously reported cases, 1 patient died from heart failure aged 3, the other 2 cases had progressive and moderately dilated left ventricles (LV) with low-normal left ventricular systolic function at most recent follow-up.
Methods: Case report
Results: A Caucasian child presented at 6 months of age with erythroderma, ichthyosis and hypotrichosis. The child had recurrent sepsis episodes in infancy and significant co-morbidities, including mild global developmental delay, failure to thrive requiring PEG feeding, egg and peanut allergies, conductive deafness and oligodontia. The patient was subsequently diagnosed with a de novo heterozygous missense mutation in theDSP gene (p.His586Pro). Similarly to the previously reported cases 1, this encodes for a proline substitution within the alpha-helix of spectrin repeat 6 which likely disrupts the alpha-helix. Our patient was screened with echocardiography from 9 months of age. At 3 years of age, the patient developed a mild gradient across the aortic valve. At 8 years of age, dilatation of the LV was first noted. The left ventricular outflow tract obstruction (LVOTO) gradually progressed (peak 115mmHg) with associated moderate aortic stenosis, mild aortic incompetence and concentric left and right ventricular hypertrophy. LVOTO was secondary to valvar and subvalvar

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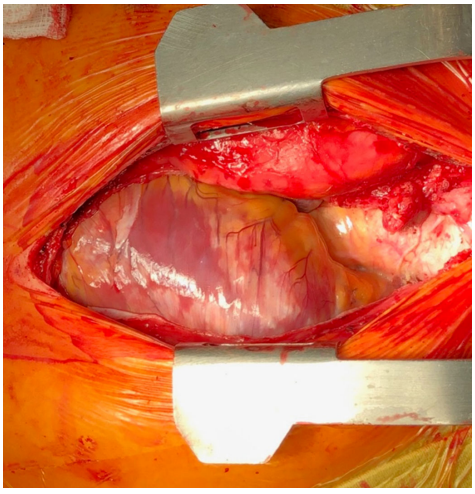
obstruction. At 10 years of age, the patient underwent a subaortic membrane resection with aortic valve repair. Intra-operatively, diffuse fibrosis was macroscopically visible (Figure 1). Three months later, at most recent follow-up, there is no residual LVOTO although both ventricles remain persistently dilated (LVIDd 5cm, z score +4) with mildly impaired left ventricular systolic function. Cardioprotective medications (spironolactone and bisoprolol) were started post-operatively. We plan to monitor our patient closely due to the potentially malignant nature of this disease with interval cardiac MRIs and Holter recordings.
Conclusions: To the best of our knowledge, there are just 3 reported cases of EKC in the literature. We aim to contribute to the knowledge base of this condition in order to better prognosticate and treat future patients.



P257
A severe case of myocarditis
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Introduction: Giant Cell Myocarditis (GCM), unlike most forms of viral myocarditis, is a rapidly progressive disease, and fatal if untreated. Even though GCM is rare its diagnosis is crucial, since initiation of immunosuppressive therapy can be very effective.
Methods:.
Results: A previously healthy 15-year-old male was admitted to our department with a history of two weeks of chest pain, fatigue, dyspnoea, and dry cough. He had acute otitis one week before the onset of symptoms. On presentation he was haemodynamically stable, had good peripheral perfusion, and basal rales on lung auscultation. ECG showed sinus tachycardia and diffuse T wave inversion; troponin I and BNP levels were elevated (see figure 1). An echocardiogram was performed, demonstrating a structurally normal heart and left ventricular (LV) systolic dysfunction, with an ejection fraction (EF) of 47%. Diagnostic workup for myocarditis aetiology was initiated, and the patient was started on enalapril and carvedilol. In the next days, symptoms and LV function worsened (EF 25%). He was then admitted to the Intensive Care Unit (ICU) and started on dopamine and milrinone, and EF increased to 43%. In spite of inotrope support and one levosimendan cycle there was clinical worsening with decreasing LV function (EF 31%, global longitudinal strain -5.5%) and increasing troponin and BNP levels (maximum 18287 and 3929 pg/mL respectively). An endomyocardial biopsy (EMB) was performed, which revealed an inflammatory infiltrate with eosinophil predominance and with presence of giant multinucleated cells. Triple immunosuppression was started with prednisolone, azathioprine and cyclosporine. There was a significant improvement in the following days, with progressive improvement of LV function and biomarkers and subsiding of symptoms in two weeks, allowing weaning of inotropic support. The patient was discharged home one month later, in NYHA class 2, with troponin I of 55pg/mL, BNP 305pg/mL, normalization of ECG abnormalities, an EF of 53%, with fibrosis but without segmental contractility abnormalities on MRI. He has been stable since (17 months follow up time).
Conclusions: This case illustrates the need for EMB, although sensitivity is not perfect. Even though this patient is now well, GCM has a poor prognosis, with average time to death/transplant less than one year.

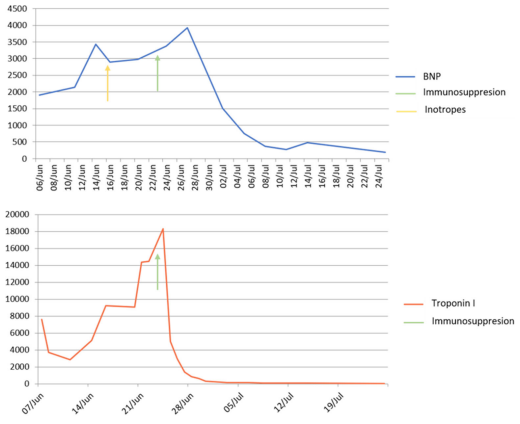

Figure 1: biomarker evolution

P258
Absence of cardiac involvement in children with uncomplicated SARS-CoV-2 infection
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Introduction: The pandemic of SARS-CoV-2 is a major health issue, and involvement of the cardiovascular system is common amongst adult with acute coronavirus disease 2019 (COVID-19). Since the beginning of the epidemic, children seem relatively spared with a low morbidity and mortality. However, multisystem inflammatory syndrome in children (MIS-C) is a rare but severe complication following SARS-CoV-2 infection. Cardiovascular involvement is reported in about 80% of MIS-C cases, with elevated cardiac enzymes, left ventricular dysfunction, shock, coronary artery dilatation, mitral regurgitation and arrhythmias. Although MIS-C seems to be a post-infectious complication, its pathogenesis has not yet been clearly elucidated. It is unknown

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whether children with uncomplicated SARS-CoV-2 infection can develop subclinical cardiac implication and coronary artery dilatation.
Methods: Children with an acute infection of SARS-CoV-2 confirmed by positive RT-PCR test on nasopharyngeal swab between March and May 2020, who didn't meet MIS-C diagnostic criteria, were proposed an outpatient cardiology appointment. Electrocardiogram and echocardiography were performed in all participants.
Results: 35 children (17 female) aged 2 months to 16 years (mean: 9.2 years) were enrolled after informed consent. Cardiology assessment took place 66 days (range 44 to 100 days) after the test. Shortening fraction of the left ventricle was normal in all subjects (mean shortening fraction 35.25%, range 30-43%). Coronary arteries were normal without dilatation in all 35 children. Moreover, there was no valvar abnormalities and no pericardial effusion. ECGs were normal without conduction abnormalities.
Conclusions: We didn't observe any subclinical cardiac involvement in our cohort of pediatric patients with uncomplicated SARS-CoV-2 infection. Cardiac dysfunction and coronary artery dilatations reported in MIS-C, but never or rarely reported in acute pediatric COVID-19 cases corroborate the hypothesis of a post-infectious syndrome. Further researches are necessary to better understand the underlying mechanisms of cardiovascular involvement after SARS-CoV-2 infection.

P259
Ambulatory blood pressure monitoring in Duchenne muscular dystrophy

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Introduction: Duchenne muscular dystrophy (DMD) affects not only skeletal, respiratory and the heart muscle but also impacts the circulatory system. Authors' clinical experience (unpublished), in concordance with Cheeran et al. and Masood et al., suggested significant hypotension by casual BP measurements. This appeared to be an unaddressed concern of possibly clinical significance as some patients receiving angiotensin-converting enzyme (ACEi) report dizziness and lightheadedness leading to noncompliance. Therefore, we conducted a cross-sectional observational study using 24-hour ambulatory blood pressure monitoring (ABPM) as a reference method.
Methods: Of 76 DMD patients being under care of our Department, 58 patients (aged 10.7±3.5) were successfully examined using ABPM. The inclusion criteria were age ≥5years, body height ≥120 cm and willingness to cooperate. We analyzed the whole group as well as ACEi treatment subgroups. The protocol included nighttime measurements between 10pm and 6am. All values were normalized for sex and height (z-score) with reference data by Wühl et al. Statistical analysis was performed using Wizard Pro 1.9.38 (Evan Miller, Chicago, IL). All data are reported as mean ± standard deviation of the calculated z-scores.
Results: The outcomes in all studied patients for 24-hour period were SBP 0.3±1.3, DBP 0.3±1.1 and MAP 0.6±1.2, daytime SBP -0.1±1.3, DBP -0.2±1.2 and MAP 0.2±1.2, nighttime SBP 0.9±1.3, DBP 0.9±1.3 and MAP 1.3±1.2. There were 14.9% of patients having hypertension (>97 percentile) and 4.3% of patients with hypotension (<3 percentile). Seventeen patients were treated with ACEi drugs and 33 did not receive such treatment. Most frequently applied were perindopril (19.6%) and enalpril (7.8%), cilazapril, ramipril and lisinopril were prescribed for single patients. Table 1 presents results for ABPM subgroup analysis, showing significantly lower BP in the ACEi treated group.
Conclusions: ACEi exert a significant effect by lowering BP, having the cardioprotective, beneficial effect on DMD patient's survival as previously reported. True hypotension in DMD patients (even with ACEi treatment), is infrequent, in contrast to what was previously suggested by casual BP measurements, yet the treated patients reporting symptoms should be carefully evaluated. Conversely hypertension is prevalent, and this requires further elucidation.

Parameter	On-ACEi	No-ACEi	p-value
Age	12.0 ± 3.1	10.2 ± 3.7	0.095 ns
SBP 24-hours [z]	-0.1 ± 1.0	0.8 ± 1.4	0.041 *
DBP 24-hours [z]	-0.2 ± 0.9	0.7 ± 1.0	0.003 **
MAP 24-hours [z]	0.2 ± 0.8	1.1 ± 1.1	0.009 **
SBP daytime [z]	-0.5 ± 1.0	0.3 ± 1.3	0.041 *
DBP daytime [z]	-0.7 ± 0.9	0.3 ± 1.2	0.005 **
MAP daytime [z]	-0.3 ± 0.8	0.7 ± 1.2	0.005 **
SBP nighttime [z]	0.5 ± 1.0	1.3 ± 1.4	0.062 ns
DBP nighttime [z]	0.5 ± 1.0	1.3 ± 1.4	0.051 ns
MAP nighttime [z]	0.9 ± 1.1	1.5 ± 1.3	0.121 ns

Table 1. Comparison of ambulatory blood pressure monitoring between a subgroup taking ACEi and not. The differences are significant during the 24-hour period and the day and not the night.

P260
Bisoprolol in paediatric cardiomyopathy: a single centre experience.

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Introduction: The efficacy of beta-blockers is well established in the treatment of adult heart failure, providing mortality benefit over placebo.Bisoprolol is also of proven benefit in adult hypertrophic cardiomyopathy (HCM). We conducted a retrospective audit to investigate the safety and efficacy of bisoprolol in children with cardiomyopathy (CM).
Methods: We identified children prescribed bisoprolol, from 2014 to 2019 using pharmacy records. We reviewed dose at commencement and on titration, blood pressure before and on treatment, heart rate before and on treatment, side effects, and noted if they received another beta-blocker before commencement.
Results: 42 children were prescribed bisoprolol: 19 HCM, 21 dilated CM, 1 restrictive CM and 1 arrhythmogenic ventricular CM. Median age at commencement was 130 months (range 4-225 months). 24 (57%) were previously on another beta-blocker (carvedilol 8, atenolol 6, propranolol 2, propranolol and then carvedilol 2). In 20 (47%) the dose was up-titrated. Mean dose at commencement was 0.07mg/kg/day (SD 0.04mg/kg/day) and after titration 0.13mg/kg/day (SD 0.06mg/kg/day). In 6 (14.3%), bisoprolol was stopped : 2 due to deterioration, 1 improvement, 1 mechanical support and then heart transplant, 1 bisoprolol was changed to nebivolol, 1 due to side effects (fatigue

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despite reducing the dose). 4 children (9.5%) reported side-effects: 3 fatigue and 1 fatigue and nausea. However, documentation of side effects was limited and relied on documentation in clinic letters. Blood pressure (BP) and heart rate (HR) did not drop significantly at baseline (BP p=0.43, HR p=0.65), and up-titration (BP p=0.11, HR p= 0.38).
Conclusions: From our limited preliminary experience, bisoprolol is safe in children with cardiomyopathy. It was well tolerated. Further studies are needed to assess its efficacy, optimal dose and safety.

Patients (total number)	42
Diagnosis	Dilated CM 21 HCM 19 Restrictive CM 1 Arrhythmogenic Ventricular CM 1
Weight at commencement mean (SD) (kg)	37.0 (25.2)
Age at commencement median (range) (months)	130 (4-225)
Patients on another beta-blocker (patients)	24: Propranolol 2 Carvedilol 8 Atenolol 6 Propranolol and then Carvedilol 2
Dose at commencement mean (SD) (mg/kg/day)	0.07 (0.04)
Dose at up-titration mean (SD) (mg/kg/day)	0.13 (0.06)
Side-effects	4 Fatigue 3 Fatigue and nausea 1
Discontinuation	6 Improvement 1 Deterioration 2 Mechanical support and heart transplant 1 Side-effect 1 Change agent 1
Follow up median (range) (months)	21 (0, 1428)

P261
Cardiac troponin I is a useful biomarker for early detection of anthracycline-induced cardiac dysfunction during chemotherapy in pediatric cases.

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Introduction: Cardiac troponin (cTn) is expected for early detection of cardiac damage before decline of LVEF or symptomatic heart failure among pediatric cancer patients who receive chemotherapy including anthracycline. Now, we evaluated the clinical picture of those with elevated serum level of cTnI.
Methods: We enrolled 19 children (male: 10, age at onset: 64 (1-206) months old) who received chemotherapy including anthracycline and underwent routine cardiac examinations more than once from April 2019 to December 2020 in our institute. Twelve subjects were diagnosed as hematological malignancies and 7 were solid tumors, the cumulative dose of anthracycline equivalent to doxorubicin was 120 (30-520) mg/m (2). They were divided into high-cTnI (H) group (n=9) and normal-cTnI (N) group (n=10) by normal limit of cTnI (<26.3 pg/mL). Baseline characteristics, other blood biomarkers and echocardiographic parameters such as LVEF and global strain were retrospectively compared. Data were described as median with range and statistical significance was determined when p value was <.05 by Mann-Whitney U test.
Results: The median of the peak of serum levels of cTnI were 62.0 (26.4-7,947.1) pg/mL in H group and 8.9 (4.0-23.1) pg/mL in N group (p=.002). Four cases in H group developed cardiac dysfunction in the course of treatment, which 3 had subclinical LV systolic dysfunction but recovered within a month, and the other had overt heart failure and required medication. There was no significant difference between 2 groups in age at onset, plasma levels of natriuretic peptide and LVEF. However, although not significant, there was likely to be higher cumulative anthracycline dose (180 vs 66 mg/m (2), p=.070) and lower global longitudinal strain (-9.6 vs -14.3 %, p=.065) in H group.
Conclusions: It is possible that those with higher levels of cTnI might be related to subclinical decline of LVEF or symptomatic heart failure, more cumulative administration of anthracycline, and lower global longitudinal strain in pediatric population. Routine cardiac screening combining echocardiography and cTnI could be useful for early detection of cardiac complication in the treatment.

P262
Changes in the blood pressure in Duchenne and Becker muscular dystrophies suggest subclinical adaptation to heart failure in disease early stages

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Introduction: Duchenne muscular dystrophy (DMD) and Becker muscular dystrophy (BMD) lead to disability, respiratory and heart failure. Many pathophysiological aspects of cardiovascular system involvement remain unknown. The aim of the study was to assess changes in the blood pressure (BP), pulse pressure (PP) and mean arterial blood pressure (MAP) in dystrophic patients.
Methods: We studied 85 patients aged 10.4 years (3.0 to 17.9) with a genetically confirmed mutation (88% DMD, 12% BMD). During the 3-year period and 218 ambulatory follow-up visits BP was measured using oscillometric method. The measurements were normalized (z-score) based on the current reference for Polish population. PP and MAP were calculated by standard formulas. For lack of reference centile charts for PP and MAP we dichotomized the data around mean normal values for PP and MAP for respective age and assessed the distribution. Data is presented as mean ± standard deviation or median (range) dependently on the distribution. Chi-square test and Pearson correlation were employed.
Results: SBP was 105mmHg (68–140; z-score -0.2±1.2) and DBP 68±109mmHg (z-score 0.9±1.1), normal in 82% and 71% of measurements respectively. SBP more commonly indicated hypotension (12.6%) than hypertension (5.4%). DBP was high in 27.0% of measurements and low in 1.8%. Both SBP and DBP z-scores dropped with age (r=-0.50, p<0.001 and r=-0.26, p=0.006 respectively). Calculated PP was 37±10mmHg, and MAP 80mmHg (52–105). PP was below the expected mean value in 76% of cases, more commonly in the older patients (p<0.001). Conversely MAP was above the expected mean value in 66% of cases, predominantly in younger patients (p<0.001). There

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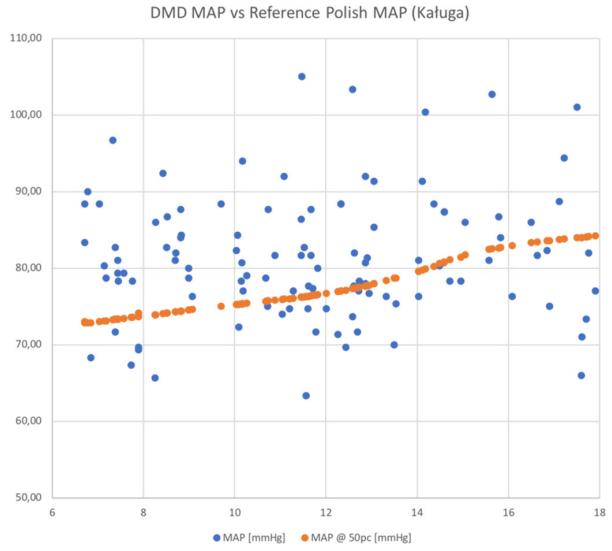
were statistically significant differences in both PP and MAP in comparison to normal population (p=0.001, p=0.014 respectively).
Conclusions: Based on the windkessel model of arterial circulation the results suggest early rise of MAP as proxy for peripheral vessel constriction (a compensatory mechanism). That is followed by a progressive decrease of SBP and PP dependent on the (initially subclinical) reduction of left ventricular stroke volume. These novel findings call for further studies.



P263
Characterization of children and adolescents with pulmonary arterial hypertension. Data from the Polish Registry of Pulmonary Hypertension (BNP-PL)

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Introduction: In Poland the care of children suspected of pulmonary hypertension (PH) is centralized in 8 medical pediatric centers and all pulmonary arterial hypertension (PAH) children are treated based on the National Health Fund criteria. The Working Group on Pulmonary Circulation of the Polish Cardiac Society established a Data Base of pulmonary hypertension in the Polish population (BNP-PL) in 2018. We present the results of the pediatric arm of the registry including epidemiology, clinical characteristics and treatments of Polish PAH children
Methods: We prospectively enrolled all PAH patients at the age of 3 months to 18 years who had been under care between March 1st 2018 and September 30th 2018. Patients diagnosed before March 1st 2018 were defined as prevalent cases while patients diagnosed on 1st March 2018 and later were defined as incident cases. To be enrolled to the study all patients had to have their diagnostic right heart catheterization which confirmed the PAH diagnosis. Functional class was assessed based on the World Health Organization (WHO-FC). Anthropometric measurements were evaluated on the basis of percentile charts of height and BMI. Data is presented as mean ± standard deviation or median (range) dependently on the distribution.
Results: 80 PAH children (males, n=40, 50%) were enrolled in the present analysis. The median percentile of height was 27.2 and median BMI percentile was 31.5. In Table 1 we present demographic data of the patients including clinical characteristics and treatments. Most frequent cause of PAH was congenital heart disease (n=54; 67.5%), followed by idiopathic PAH (n=25; 31.25%) and portopulmonary PAH (n=1; 1.25). At enrollment 31% of patients were still in WHO-FC III. The majority of children (95%) were treated with PAH specific medications but only a half of them with double combination therapy.
Conclusions: Almost all PAH cases were associated with CHD or IPAH without significant representation of other PAH subclasses which may indicate that some children are underdiagnosed. Most of children had a growth retardation which, suggests the need for active dietary interventions. The low use of a combination PAH therapies calls for significant improvement in therapy of this rare group of severely diseased patients.

Table 1. Demographic and Baseline Characteristics of Children with idiopathic pulmonary arterial hypertension (IPAH) and pulmonary arterial hypertension associated with congenital heart disease (CHD-PAH).

Characteristics	All, n=80	IPAH, n=25	CHD-PAH, n-54	p
Age at diagnosis	5.1 (2.1-8.1)	5.9 (2.5-8.3)	4.6(2.1-7.5)	0.5
Age at enrollment	10.4 (7.9-15.2)	9.8 (7.6-11.5)	11.5(7.9-15.4)	0.15
Female, n(%)	40 (50%)	12 (48%)	27(50%)	0.29
Incident cases, n(%)	10 (12.5%)	9 (36%)	1 (1.9%)	<0.001
mPAP (at diagnosis)	48.0 (37.0-57.0)	48.0(39-67)	48 (36-55.5)	0.54
Functional class at enrollment, n (%)				
I	9	2	9	0.5
II	45	13	45	0.5
III	25	10	25	0.3
IV	0	0	0	---
6MWD[m]	429(360-500) n=50	450(383-524) N=18	420 (353-469) N=32	0.46
NT-proBNP [ng/l]	272 (104.6-628.0)	268.7 (69.2-502.7)	290.0 (115.8-676.0)	0.38
Sildenafil [n,%]	57 (71%)	18 (72%)	38 (70.4%)	0.88
Tadalafil [n,%]	1 (1.3%)	0	1 (1.9%)	0.50
Bosentan [n,%]	61 (76.3%)	17 (68%)	44 (81.5%)	0.29
Treprostinil [n,%]	3 (3.75%)	3 (12%)	0	0.009
Riociguat [n,%]	1 (1.3%)	1 (4.0%)	0	0.14
PAH monotherapy [n,%]	35 (44%)	10 (40%)	24 (44.4%)	0.71
PAH double combination therapy [n,%]	39 (49%)	10 (40%)	29 (53.7%)	0.7
PAH triple combination therapy [n,%]	3 (3.8%)	3 (12%)	0	0.01

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P264

Complete resolution of a left ventricular thrombus following continuous enoxaparin therapy in three pediatric patients

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Introduction: Patients with a dilated and impaired left ventricle (LV) are at an increased risk of developing LV thrombus. The associated risks are embolic events. Vitamin K antagonists are often used as first line therapy in adults. Because of its lytic effect and the short half-life period we used enoxaparin. In order to achieve therapeutic levels without fluctuations, we used enoxaparin as continuous intravenous infusion (CII). However, there are almost no reports regarding the continuous intravenous use in children. We report the complete resolution of LV thrombus in 3 pediatric patients.
Methods: Three patients with apical LV thrombus were treated with enoxaparin as CII in our unit between 2013 and 2018.The first patient was a 16-year-old boy, the second patient a 26-year-old woman with trisomy 21, both had tetralogy of Fallot, corrected in infancy. Both presented with severe heart failure. The third patient was a 3-year-old boy, presenting with dilative cardiomyopathy following Parvovirus B19 myocarditis. He also presented with a thrombus formation in the left apex. In all patients enoxaparin was administered as CII with anti-Xa levels between 0.7 - 1 IU/ml, the doses required were between 1.5mg/kg/d (patient with renal insufficiency) and 3.5mg/kg/d.
Results: In all patients, complete dissolution of the thrombus occurred within 7-10 days. During this therapy there was no evidence for thrombus embolization. After complete resolution of the thrombus the CII was temporarily switched to a subcutaneous administration with similar anti-Xa levels. After 2 weeks enoxaparin was switched to acetylsalicylic acid because of expected poor compliance in a difficult family situation (patient 1). Because of the poor left ventricular function in patient 2 and 3 enoxaparin was switched to a Phenprocoumon therapy with INR levels of 2-3.
Conclusions: CII of enoxaparin was safe without secondary bleeding and resulted in complete dissolution of the thrombi without secondary embolization. Therefore, CII of enoxaparin, which maintains stable high drug levels compared to the subcutaneous application, is an attractive alternative for the treatment of LV thrombus formations, especially in children with contraindications for conventional thrombolytic therapy. Large population studies are needed to confirm these encouraging preliminary results.

P265

heart transplantation in children with congenital heart disease: a tertiary center experience

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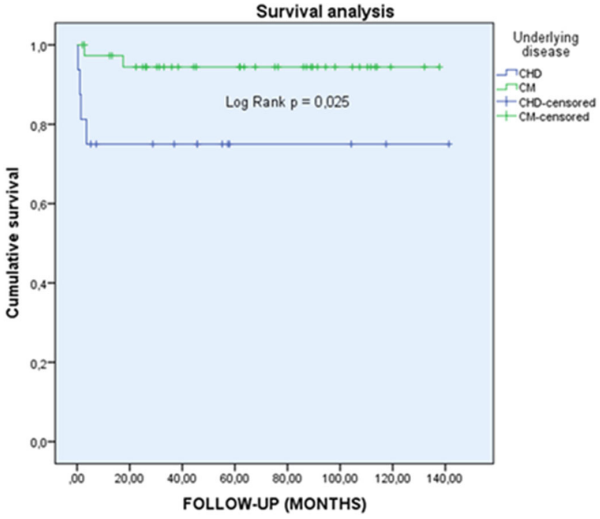
Introduction: Paediatric heart transplantation (PHTx) in patients with congenital heart disease (CHD) represents a challenge given the presence of unique factors such as complex anatomies, sternotomies, or sensitization due to previous surgeries.
Methods: We retrospectively registered PHTx performed in our hospital from December 2008 to November 2020.
Results: 56 transplants were performed in 23 women (41%) with a median age of 73 months (p25-p75 17-163 months). The indication was for cardiomyopathy (CM) in 70% (N = 39) and for CHD in 30% (N = 17). Of the CHD group, 53% (N = 9) had biventricular physiology. Prior to transplantation, 12% (N = 2) in the CHD group and 44% (N = 17) in the CM group required mechanical circulatory support, p= 0.021. There was significant difference in the cardiopulmonary bypass time (CBT), being greater in patients with CHD (248 ± 16 vs 198 ± 14 minutes, p= 0.029). After the transplant, the CHD group required more frequently mechanical circulatory support (41% vs 13%, p= 0.017), longer mechanical ventilation (312 [240-744] hours vs 48 [36-127] hours, p= 0.006), had more incidence of renal failure (82% vs 51% p= 0.029), severe right ventricular dysfunction (29% vs 8%, p= 0.033) and prolonged Intensive Care Unit (ICU) stay (23 [13-45] vs 12 [9-23] days, p= 0.023). There were no significant differences regarding the incidence of acute rejection (cellular 2R or greater: 35% in CHD vs 33% in CM, ns; humoral: 35% in CHD vs 26% in CM, ns). With a median follow-up of 52 months (p25-p75 25-95 months), survival was 95% (N= 37) in CM vs 76% (N= 13) in CHD (p= 0.025). There was a significant difference in mortality in the first three months after transplant (deaths: N = 4 in CHD vs N= 1 in MC, p= 0.011), while in the subsequent follow-up no differences were found (1 death in each group).
Conclusions: Patients with CHD have longer CBT time, prolonged ICU admission and invasive support, with higher post-operative mortality, but despite his initial outcome is slightly worse, the evolution is similar in both groups after the initial postoperative period.



P266

Impairment of left ventricular function is one year earlier than introduction of respiratory aid in Fukuyama muscular dystrophy

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Introduction: Fukuyama congenital muscular dystrophy (FCMD) is a rare entity that affects 2.9/100000 in Japanese population, and characterized by progressive skeletal muscle weakness, severe intellectual deficit and brain malformation. Cardia dysfunction is often

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observed, but its clinical manifestation is unclear.Purpose of this study is to clarify the cardiac involvement in FCMD.
Methods: Retrospective chart review was carried out at our institute on patients with genetically confirmed FCMD. (n=15) Age at diagnosis of FCMD, first cardiology clinic visit, declining mortor function, introduction of respiratory aid, echocardiographic data, medical treatiment for heart failure, and its prognosis were reviewd.
Results: FCMD was confirmed at 1.1±0.6 years old (y), and first cardiology clinic visit was at 6.9±4.1 y. Observation period was 12.3±8.3 years. Declining of motor function was observed at 6.1±2.3 y and respiratory aid was required at 14.1±3.2 y in 11 patients (73%). Decreased fraction shortening (FS) of the left ventricle on echocardiography (<0.25) was observed at 13±4.7 y in 13 patients (87%). Carvedilol was used in 12 patients, ACE-I or ARB was used in 12 patients, and 11patients were taking both. FS was 0.35±0.04 at the first clinic visit, and 0.24±0.09 at the last clinic visit. There were two death, and two lost of follow up duing the study period.
Conclusions: Cardiac involvement is frequent and progressive in FCMD. Impairment of left ventircular function was one year earlier than introduction of respiratory aid in FCMD.

P267
Long-term outcome of LVAD in Duchenne population with end stage cardiomyopathy

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Introduction: The aim of this study was to report the effect of long-term use of VAD in patients affected by Duchenne Muscular Distrophy and end stage cardiomyopathy.
Methods: We collected data of DMD patients with end-stage cardiomyopathy implanted with LVAD in our center (group 1) and the ones in regular follow-up because not eligible for LVAD (group 2). Each patient of group 1 underwent extensive pre-operative assessment, in-hospital treatment and was enrolled in a dedicated DMD HF (heart failure) clinic during the post-discharge period. Patients of group 2 were followed in a dedicated DMD HF clinic or as inpatient in case of need of inotropic support. All data during follow-up were recorded including echocardiography, ECG examination, laboratory blood tests, BNP dosing and assessment of nutritional status and respiratory function.Survival of groups was represented by Kaplan Meier analysis and compared by log rank.
Results: Eight DMD patients (group 1) with end-stage cardiomyopathy were implanted with LVAD and treated at our center from 2011 until 2018 (mean age at implantation was 16.9 ± 2.9 years), whilst 4 patients (group 2) were not eligible for LVAD implantation. At the last follow-up, 4 out of 8 patients were alive and 2 did not have post-operative complications. The main complications were bleeding and infections with one episode of acute heart failure successfully treated. The principal causes of 3 deaths at the median follow-up of 22 months were sepsis, tracheal bleeding, and cerebral hemorrhage. After 1 year from implantation we noticed a significant decrease in heart rate (p=0.002), in LV volumes and diameters (LVEDD p=0.03, LVESD p=0.02, EDV p=0.01 and ESV p=0.02) and a significant increase in EF (p=0.0036). However, relative wall thickness did not change over time. A significant improvement of survival in DMD patients treated with LVAD compared to the group that wasn't implanted (log rank<0.001) was noticed, with a 5-year increase in life expectancy.
Conclusions: Our data showed that LVAD improved outcomes in patients with LVAD when compared to those without. An integration of outpatient DMD HF clinic and patient-centered palliative care will most likely increase the quality of life and delivery of care for this population.

P268
Midterm outcomes after the modified Fontan Operation are not related to ventricular morphology

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Introduction: Outcomes of single ventricle patients undergoing the Fontan operation improved over the past decades. Nowadays, the majority of patients survives into adulthood. We aimed to evaluate the impact of ventricular morphology on survival and morbidity after the modified Fontan operation in a single centre cohort.
Methods: Medical records of patients with single ventricle physiology who underwent a modified Fontan operation between 1996 and 2018 utilizing the interatrial lateral tunnel technique or an extracardiac conduit were reviewed. The underlying diagnoses, ventricular morphology, types of all preceding operations and time and causes of death, heart transplantation or Fontan take-down were recorded (primary endpoint). In addition, freedom from Fontan related complications (Protein losing enteropathy, bronchitis fibroplastica, arrhythmias, thromboembolic events) was assessed. Event-free survival was compared between patients with left and right ventricular morphology and between patients with Hypoplastic left heart syndrome and other types of single ventricle heart defects.
Results: 398 patients were included. Median age at surgery was 2.7 (IQR 2.3-3.2) years. Right ventricular morphology was present in 252 (63.3%) patients. The interatrial lateral tunnel technique was utilized in the majority of the cases (n=346, 86.9%). Median follow-up was 7.0 (2.4-12.1) years. During follow-up, 20 patients died, 1 underwent heart transplantation and 3 patients had Fontan take-down. Freedom from death, transplantation or take-down was 95% and 90% after 10 and 15 years, respectively. There was no difference between right or left ventricular morphology (p=0.563) or between patients with Hypoplastic left heart

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syndrome and other underlying diagnoses (p=0.359). Complication free survival was 81% and 71% after 10 and 15 years with no difference between right and left ventricular morphology (p=0.248). Hypoplastic left heart syndrome was not a risk factor for increased morbidity (p=0.382). The occurrence of arrhythmias during follow-up after Fontan completion was associated with impaired survival (p=0.001). Patients with the lateral tunnel technique or the extracardiac conduit showed no difference in transplant-free survival (p=0.343) and freedom from morbidity (p=0.602).
Conclusions: Midterm survival rates and morbidity after the modified Fontan operation are not related to ventricular morphology. The occurrence of arrhythmias during follow-up after Fontan completion is a risk factor for impaired outcome.

P269
Pediatric myocarditis: a 14-year experience in a tertiary center
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Introduction: Myocarditis is a life-threatening disease in the pediatric population. Its presentation can range from chest pain to heart failure or sudden cardiac death. 20-30% progress to dilated cardiomyopathy, with poor prognosis.The diagnostic gold standard is endomyocardial biopsy (EMB), rarely used in pediatrics. Usually myocarditis is indirectly diagnosed with biomarkers and imaging techniques among which cardiac magnetic resonance (CMR) stands out. The aim of this study is to describe demographic, virological and clinical variables, therapy and outcome, focusing on the role of EMB in establishment of specific treatments.
Methods: Pediatric patients with myocarditis were included prospectively for 14 years (2007 to 2020). Diagnosis was made either by EMB or by CMR (Lake Louise criteria), electrocardiogram and biomarkers. 7 patients with imaging criteria and inflammatory infiltrates within the myocardium but genetic study compatible with genetic cardiomyopathy were excluded.
Results: 51 patients were included, median age 3 (0-17) years. 57.8% were male. The main clinical onset was cardiogenic shock (35.3%) or heart failure (29.4%) but 21.6% were diagnosed after ischemic-like chest pain; 5 had a fulminant presentation and 2 presented with rhythm disturbances. A mild viral episode was reported in 42 patients (82.3%), a median of 4.5 days before the diagnosis. 82.4% were admitted to the ICU, 49% required mechanical ventilation, 76.4% inotropic drugs and 29.4% mechanical circulatory support with ECMO (median 15 days) or Berlin-Heart (8 days). The diagnosis was confirmed by EMB in 21 patients (41.2%) a median of 8 days after the onset; CMR was performed in 38 patients (74.5%), 60.5% before 2 weeks. As part of an intern protocol, targeted therapy with immunosuppressive and antiviral treatment was administered depending on EMB analysis from 2015; until then, treatment was empirical. During follow-up, 5 patients died within a median of 8 days and 4 required heart transplantation. The rest recovered a median of 22 days after the onset.
Conclusions: Despite the good results with targeted therapy, large studies are needed to assess long-term evolution. The creation of a Spanish pediatric registry of myocarditis to collect data about onset, examinations, treatment and outcomes, could detect strong endpoints and contribute to consolidate a national protocol.

P270
Persistent Myocardial Atrophy despite Left Ventricular Reverse Remodeling in Duchenne Cardiomyopathy treated by Left Ventricular Assist Device
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Introduction: Dilated cardiomyopathy (DCM) is the leading cause of death in Duchenne patients. Left ventricular assist devices (LVADs) are considered therapeutic options as a destination therapy in advanced heart failure. The aim of our study was to evaluate left ventricle (LV) remodeling of Duchenne patients after LVADs and chronic therapy.
Methods: Demographic and echocardiographic data were collected and analyzed for 8 LVADs implanted Duchenne patients. All parameters were measured in three phases: pre-LVAD, 1 month and 1 year post- LVAD implant.
Results: All patients were affected by end-stage DCM, mean age at implantation was 16.9 ± 2.9 years. Patients were treated with maximal medical therapy. One year post-implantation, heart rate decreased from a mean of 110 ± 19 bpm to 82 ± 2 bpm (p=0.002), a significant decrease in LV volumes and diameters (LVEDD p=0.03, LVESD p=0.02, EDV p=0.01 and ESV p=0.02) was noticed together with a significant increase in ejection fraction (p=0.0036). However, relative wall thickness did not change over time, showing an eccentric remodeling pattern before and after LVADs.
Conclusions: Our data showed that cardiac atrophy is persistent in Duchenne cardiomyopathy despite the improvement in LV function secondary to a significant ventricular unloading through prolonged use of LVADs coupled with chronic therapy. Further multicentric studies are needed to derive solid conclusions.

P271
Prevalence and course of cardiac complications in children with long chain fatty acid oxidation disorders before and after newborn screening
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Introduction: Long chain fatty acid oxidation disorders lead to deficient energy production and accumulation of potentially toxic 3-hydroxy fatty acids thereby affecting especially organs that use long chain fatty acids as the primary energy source such as skeletal muscles and the heart. Cardiac involvement typically presents as severe cardiomyopathy during the first days of life. However, various rhythm disorders and later onset cardiomyopathy have also been described.
Methods: We performed a retrospective analysis of 32 patients aged 3 days to 26.5 years (median 6 years) diagnosed with long chain fatty acid oxidation disorders from 1990 until 2020 with special regard to their cardiac complications. 19/32 patients were diagnosed by newborn screening and the remaining 13 were diagnosed due to their symptoms. Median follow up period was 6 years.
Results: 6/13 (46%) patients diagnosed before newborn screening had a documented cardiac involvement including hypertrophic,

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restrictive and undulating cardiomyopathy. Their age at diagnosis of the metabolic disorder ranged from 5 months to 7 years (median 1 year), 2 were diagnosed post mortem. The median age at first documented cardiac complications was 1 year. 6/13 (46%) patients died (median 6.5 months). 9/19 (47%) patients diagnosed after newborn screening had cardiomyopathy, with myocardial hypertrophy being present in 5, dilated cardiomyopathy in 2 and mild hypertrophy that evolved into dilated phenotype during metabolic crisis in 2. The median age at first documented cardiac complications was 0.5 years. 3/18 (17%) patients died (median 3.5 years). Ventricular fibrillation was documented in 2 patients. No other clinically relevant arrhythmias were observed. Only two of the surviving patients with cardiomyopathies have moderate or severe phenotype, the rest manifest only mild heart muscle involvement. However, all deceased patients diagnosed by newborn screening had suffered from mild cardiomyopathy that deteriorated rapidly during a metabolic crisis.
Conclusions: This study provides an overview of cardiac findings in patients with long chain fatty acid oxidation disorders with special focus on one patient with rare restrictive phenotype. Heart failure and/or fatal arrhythmia were the major cause of death in most deceased patients. Newborn screening has reduced mortality in our cohort.

P272
Recovery of Ventricular function in children Dilated Cardiomyopathy Phenotypes: Role of long-term Milrinone Therapy

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Introduction: Management of children with severe heart failure (HF) due to dilated cardiomyopathy (DCM) phenotypes is challenging. Limited donor pool and long transplant waiting list (TWL) are barriers to orthotopic heart transplantation (OHT) in smaller infants. Milrinone, a phosphodiesterase 3 inhibitor has unique inotropic and vasodilator properties, however data regarding its long-term use in paediatric HF is lacking. This report evaluates the role of long-term milrinone therapy and potential for recovery of cardiac function in children with severe HF.
Methods: Single-centre observational study (between 2014-20) involving 28 children with severe HF and DCM treated with long-term (> 7 days) milrinone infusion. Severity of HF defined by vasoactive-inotrope score (VIS). Serial monitoring of haemodynamic, biochemical, echocardiographic, ECG and cardiac biomarker parameters to assess recovery. Initiation of mechanical cardiac support (MCS) and transplant referral in the event of deterioration. Data analysed at initiation and discontinuation of milrinone infusion and last clinic visit.
Results: Demographics: 15 female: 13 male. Median age (months): 3.5 (range 0.5 to 77). Median weight (kg): 5.85 (range 2.0 to 18.9). DCM phenotypes: viral myocarditis (15), idiopathic (11), genetic (2). PICU course: All patients in decompensated HF (mean lactate 9.14) requiring ventilatory and inotropic support (mean VIS :17). 4 patients required CVVH renal support. Median PICU length of stay was 6.5 days (range 1 to 189 days). Inotrope therapy: All patients received milrinone infusion at 0.5 to 0.75mcg/kg/min. 19 needed additional inotropic support in the acute stage. The median time on milrinone was 42 days (range 7 to 315 days). Outcome: No early deaths. Two patients had successful OHT. Long- term follow up: median 799 days (range 58 to 2202 days) 3 late non cardiac deaths. 3 patients on TWL (2 on milrinone/1 MCS) Overall, 20 (71%) patients recovered cardiac function and discharged home on medications. 4 (14%) patients needed MCS support in addition milrinone infusion. (See table for details)
Conclusions: Long-term milrinone therapy provides safe and effective pharmacological bridge to recovery of cardiac function in small infants and children with decompensated HF. It can be a bridge to advanced MCS and OHT in refractory cases.

Table of results.
20 children with recovery of cardiac function on long term Milrinone (9 patients were < 5.0 kg at presentation)

	At presentation (or at nadir where specified)	At time of stopping Milrinone	At most recent follow up
Milrinone alone N=16 Median time on milrinone 37 days Interquartile range (IQR) 42.5.			
Echocardiographic parameters			
Median fractional shortening (IQR)	15% (5)	18% (6.75)	27% (8.5)
Median ejection fraction (IQR)	32% (13.2)	37% (15)	54% (15.5)
Median LVIDd (IQR)	32.4mm (12.5)	32.5mm (9)	36mm (5.75)
Median LVIDd z score (IQR)	+4.7 (4.03)	+4.7 (3.42)	+3.1 (1.81)
ECG Parameters			
Evidence of strain /abnormality number of patients (percentage of patients)	16/16(100%)	12/14 (85.7%)	5/14 (35.7%)
Biochemical parameters			
Median BNP (IQR)	20,326 ng/L (52,978)	3055 ng/L (5,847.5)	275 ng/L (184.5)
Median eGFR nadir(IQR)	51.5 mL/min/1.73m^2	>75 mL/min/1.73m^2	
Severe liver impairment (nadir) number of patients (percentage of patients)	13/16 (81.2%)	1/16 (6.3%)	
MCS with Milrinone N=4 Median time on milrinone 45 days. Interquartile range 12.			
Echocardiographic parameters			
Median fractional shortening (IQR)	6% (3)	20% (4.5)	23% (7)
Median ejection fraction (IQR)	14% (7.5)	42% (10)	55% (13.25)
Median LVIDd (IQR)	40mm (14.25)	34mm (12.5)	35.5mm (11.5)
Median LVIDd z score (IQR)	+1.54 (4.17)	+3.65 (4.53)	+1.7 (1.72)
ECG Parameters			
Evidence of strain /abnormality number of patients (percentage of patients)	4/4 (100%)	3/3 (100%)	2/4 (50%)
Biochemical parameters			
Median BNP (IQR)	52273 ng/L (32,336)	5663ng/L (2608)	124ng/L (-)
Median eGFR nadir(IQR)	33mL/min/1.73m^2 (2.4)	>75mL/min/1.73m^2 (0)	
Severe liver impairment (nadir) number of patients (percentage of patients)	0%)	0 (0%)	

P273
Treatment of pulmonary arterial hypertension associated with congenital heart diseases of left-right shunt - Bosnian and Herzegovinian experience

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Introduction: Pulmonary arterial hypertension (PAH), especially associated with congenital heart disease (CHD) of left-to-right shunt, is a big problem in medicine in terms of diagnostics and treatment. It evolves through increase of vasculare resistance and/or increase of blood flow through lungs, where the mean pulmonary artery pressure is above 25 mmHg and during catheterization above 30 mmHg. Due to volume and pressure load of blood from the left heart cavities in right, hypertrophy and insufficiency with increased production of PAH mediators, necrotizing arteritis of pulmonary capillaries occurs. PAH is nonspecific, and shortened 6 MWD occurs later. On ECG we see right heart hypertrophy, specific x-ray image, and on ultrasound we have significant tricuspidal regurgitation, right ventricular hypertrophy, small left heart

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cavity, pulmonary valve fluttering, paradoxical interventricular septal motion. The diagnosis is completed by CT and pulmonary angiography, ventilation/perfusion lung scintigraphy, golden standard of PAH diagnosis is catheterization of the right heart.
Methods: Paper represents retrospective analytical study over a period of 23 years.
Results: In the last 23 year we treated 62 patients with PAH related CHD of left-to-right shunt. In the last 8 years on therapy with endothelin receptor antagonists we have 12 patients who have continuosly received therapy everyday in the doses of 1 – 2.14 mg/kg with evaluation every 1-3 months. In the recent period pediatric heart catheterization is done in 48 children due to evaluation of possible surgical intervention of CHD with L-D shunt. At 18 patients we proved fixed pulmonary hypertension, at 9 of those patients it was associated with Down syndrom. In 30 patients who underwent catheterization with unfixed reversible PAH, 4 children were not surgically treated. During the follow-up of patients with PAH and CHD of the L-D shunt so far 11 patients (2-38 years of age) have died.
Conclusions: Continuous administration of endothelin 1 receptor antagonists indicates satisfactory health status of patients with PAH and CHD of the L-D shunt, combined with phosphodiesterase 5 inhibitors and prostanoids as well as other supportive therapy for comorbidities. However, new randomized clinical trials are neccessary to find an optimal therapy for this condition and to evaluate other possible treatment modalities.

P274
Vein of Galen aneurysm as an extracardiac cause of severe heart failure in the neonatal period
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Introduction: Extracardiac arteriovenous malformations are considered to be an extremely rare cause of severe heart failure in the neonatal period. Intracranial arteriovenous malformation associated with the large-vein aneurysm of Galen (VGA) is the most common pathology among these malformations, due to impaired development of the embryonic precursor - Markovskiy's medial prosenecephalic vein. Presence of this congenital pathology significantly complicates the diagnostic search, since clinical manifestations are characteristic only for heart failure.
Methods: A clinical case of a newborn girl with VGA, who was admitted to the neonatal intensive care unit of the Regional Children's Hospital with signs of severe heart failure in January 2019.
Results: On the third day after birth, the child developed cyanosis (O2 saturation was 50%). Therefore, congenital heart disease was suspected.

On examination in the neonatal intensive care unit: cyanosis (50% saturation), dyspnea (respiratory rate 50-60/min), tachycardia (heart rate 140-160 bpm) and significant swelling and pulsation of the neck veins, large fontanelle 3x3 cm in size, at the level of the bones of the skull, auscultation of the fontanel was not performed. Pulsation was determined on the femoral arteries, and the liver was enlarged (+ 4 cm) on palpation. During auscultation - pansystolic murmur across left lower sternal border 4 \ 6 by Levine scale. Cardiomegaly (CTI> 80%) with enhanced pulmonary pattern and hepatomegaly where revealed on the roentgenogram. During an echocardiographic examination, the child was diagnosed with dilated right heart chambers with tricuspid insufficiency, patent foramen ovale and a dilated innominate vein. After excluding the diagnosis of critical heart disease, neurosonography was carried out and a diagnosis was made — arterio-venous malformation — Vein of Galen aneurysm, which was confirmed on computed tomography.
Conclusions: Aneurysm of the vein of Galen is an extremely rare pathology, however, the swelling and pulsation of the cervical veins during the examination and the presence of severe cardiovascular insufficiency in newborns in the absence of congenital heart disease requires children's cardiologists to suspect and exclude arterio-venous malformation of the cerebral vessels. Endovascular treatment of VGA is considered the most effective, despite the high postoperative mortality.

10. Psychosocial

P275
Cardiac health status and quality of life in children and teenagers after arterial switch operation for transposition of the great arteries 10. Psychosocial
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Introduction: As the number of patients after arterial switch operation (ASO) for transposition of the great arteries (TGA) is still growing, it is essential to investigate the relationship between the subjective quality of life (QoL) and the objective cardiac health status in this population.
Methods: A total of 65 consecutive patients aged 8-18 with TGA, who underwent ASO at our institution, were included in the study. Detailed analysis was conducted on the clinical data: routine 12-lead electrocardiography (ECG), Holter ECG, transthoracic echocardiography and cardio-pulmonary exercise test. Evaluation of the QoL was performed with the KIDSCREEN-52 questionnaire.
Results: The analysed population (46 boys, 19 girls, mean age 13,49 ± 3,23 years) showed good overall cardiac health status. Exercise performance decreased significantly with age ($p<0.001$). A significant negative correlation between VO2 peak % predicted and OUES % predicted with the Body Mass Index (BMI) percentile was found ($p<0,001$ and $p=0,025$, respectively). VO2 peak % predicted was positively related to the QoL assessment: physical well-being ($p=0,006$), psychological well-being ($p=0,004$) and self-perception ($p<0,001$), whereas OUES % predicted was positively associated with the QoL in the domain of self-perception ($p=0,025$). Children and teenagers after ASO for TGA assessed their QoL higher in all domains when compared to the reference population, though their subjective QoL decreases with age in the majority of the domains, with significant decrease found for physical and psychological well-being, self-perception, school environment and general QoL index. The QoL assessment (physical and psychological well-being, self-perception, school environment and general QoL index) was negatively related to the BMI percentile ($p=0,012$, $p=0,049$, $p=0,06$, $p=0,03$ and $p=0,08$, respectively).
Conclusions: Although ASO for TGA provides good long-term outcomes, exercise capacity of these patients deteriorates with age, which may be related to the increasing tendency for overweight and obesity, as well as a reduced level of physical activity.The QoL of children and teenagers after ASO for TGA is high in all dimensions, nevertheless it deteriorates with age. Due to significant correlation found between exercise capacity and subjective assessment of some QoL domains, therapeutic

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approaches in TGA patients with an impact on exercise capacity may have a positive influence on their QoL.

P276
Caring for children with congenital heart diseases: economic burden of pre-surgical treatment on nigerian families

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Introduction: Managing children with Congenital Heart Disease (CHD) attracts enormous cost especially in resource-poor settings like Nigeria. This study sought to determine the healthcare costs of pre-surgical management of CHD and describe its catastrophic effects on households
Methods: Using a semi-structured interviewer-administered questionnaire, caregivers of children with CHD were interviewed. Family income, type of CHD, co-morbidity, healthcare payment mechanism and healthcare cost were explored over 3 months prior to the study. Healthcare costs were then averaged to obtain monthly estimates. Catastrophic health expenditure (CHE) was defined as healthcare spending above 10% family monthly income. Factors associated with increased healthcare spending in CHD management were explored using the Kruskal Wallis test of significance.
Results: Of the 108 parents interviewed, 81.0% paid for healthcare using out-of-pocket payment mechanism. The median direct medical and non-medical costs were ₦3,625 (Range:₦200–₦59,350) [$10.07;Range:$0.56–$164.86] and ₦420 (Range: ₦150–₦11,000) [$1.17;Range $0.42–$30.56] monthly, respectively. Hospitalization and transportation accounted for majority of the direct medical and non-medical costs, respectively. About 36.1% of families suffered financial catastrophe. Catastrophic overshoot and Mean positive overshoot were 5.6% and 30.8% above the 10% income threshold, respectively. The healthcare spending was significantly higher in families of CHD children with heart failure(p=0.001), pulmonary hypertension(p=0.038) and those who suffered financial catastrophe(p=0.001). Health insurance did not significantly reduce healthcare spending among the insured(p=0.630).

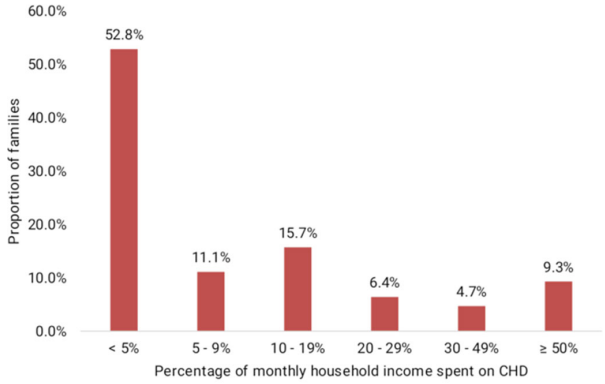

Figure 1: Proportion of families and percentage of household income expended on care of children with Congenital heart diseases

Conclusions: The economic burden of pre-surgical management of children with CHD is high in Nigeria. Appropriate government interventions are needed to cushion the burden of healthcare cost on affected families.

P277
COVID-19 related anxiety among parents of children suffering from Duchenne muscular dystrophy

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Introduction: Duchenne (DMD) and Becker (BMD) muscular dystrophies are genetic disorders affecting skeletal, respiratory, and the cardiac muscles leading to respiratory and/or circulatory failure and death in early adulthood. Therefore, the clinical course of the disease potentially puts these patients in a high-risk group for severe and complicated course of coronavirus disease 2019 (COVID-19).
Methods: We present a cross-sectional, observational study based on survey research conducted from 04th July to 6th August 2020. Parents of children with DMD or BMD were invited via online groups and forums to fill in an online questionnaire regarding demographics, medical and social information and assessment of level of anxiety. Only complete surveys were considered eligible. Statistical analysis was performed using Wizard 2beta239 (Evan Miller, Chicago, IL). All data are reported as percentage (n) or mean ± standard deviation or median (range) dependently on the data type and distribution. Appropriate tests were used and $p<0.05$ was considered statistically significant.
Results: Sixty-three respondents, primarily female (82%), aged 39.5 ±7.8 years took part in the study. Majority of responders (51%, n=32) had two children and 35% (22) had one child, 25% (8) of responders had more than one child with BMD/DMD. Mean age of the children with dystrophy was 9.6±5.2 years. Eighty-nine percent of respondents were moderately-to-highly afraid of COVID-19-related complications in their children, 70% feared for their children lives and 72% reported concern due to healthcare system reorganisation (Figure 1). Guardians of younger children (<12 y.o.) were more concerned than the parents of older children (p=0.044). Information about protecting DMD/BMD patients against SARS-CoV-2 infection (94%) and resuming of planned hospitalizations and out-patient visits (95,3%) were indicated as factors much-to-greatly increasing guardians' sense of security. Moderate or severe level of anxiety (as assessed by General Anxiety Disorder - 7 Questionnaire) was present in 20% (12) guardians. There was no difference in concern or anxiety level between DMD and BMD groups.
Conclusions: COVID-19 pandemic as a new health hazard and simultaneously culprit of reorganisation in health system both resulting in increased anxiety and concern in parents, including fear of offsprings' life. Yielding adequate information by healthcare providers may ease parents' fear.

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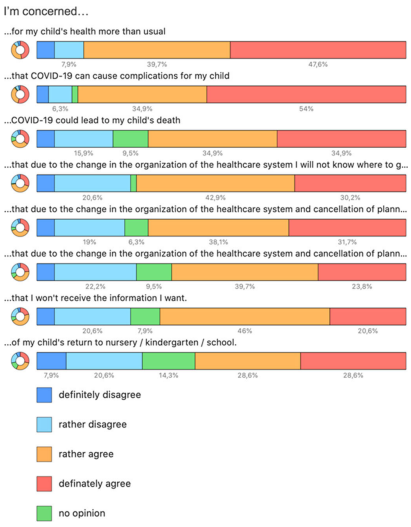

P278

Do parents want to be updated during their child's cardiac surgery? If so, how when and what?

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Introduction: During cardiac surgery parents face a long anxious wait. There can be critical complications as well as logistical delays. Current standards are that parents should be updated as soon as possible after surgery, but in other international centres parents receive regular updates, or focussed updates if there are complications or delays. We sought to investigate what parents of children who have had cardiac surgery would have liked in terms of contact and information while their child is in theatre.
Methods: Semi-structured interviews were carried out with nineteen families covering the spectrum of congenital cardiac surgery. Families were divided into three groups representing higher risk (5), medium risk (6) and lower risk (8) surgery (average RACHS score 5, 3 and 2 respectively).
Results: All participants regardless of procedural complexity concurred they would like to be updated in the event of further unexpected waiting times. 74% specified that they were happy to be contacted via telephone call (not text). In the instance of complications, we received a mixture of responses as to whether the parents would prefer to be informed during the surgery or after. Results suggested a correlation between the level of risk involved with 80% of the high-risk group wanting updates during the surgery but the medium/lower risk groups all preferring to be told after. Of the eleven families that specified how they wished to be contacted in the event of a complication, 82% preferred face to face contact and preferably a 'familiar face'. A number of parents expressed high levels of anxiety and uncertainty when waiting over the expected time which was heightened with a lack of a defined point of contact, with some describing feeling "lost" and as if they were "free-falling" when no updates were provided.
Conclusions: Our results emphasised the need for improved communication between family members and staff during cardiac surgery. Telephone updates seem appropriate if surgery is taking longer than anticipated, but in the event of complications face to face contact with a member of staff known to the family should be considered. We are now working to implement this within our institution.

P279

Health-related quality of life, parenting stress and illness cognitions in children and adolescents with Ebstein anomaly and their families

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Introduction: Due to the improved survival rates of children and adolescents with congenital heart disease, more attention is now being directed towards the impact of disease on the whole family. The aim of the current study was to evaluate both self- and proxy-reported health-related quality of life (HRQoL), and to assess levels of parenting stress and parental illness cognition.
Methods: In this cross-sectional, multicentre study, we included EA patients (aged 8-17 years) and their parents. Patients underwent routine clinical assessments in one of the Dutch university hospitals between May 2017 and March 2019. The Generic Paediatric Quality of Life Inventory 4.0 (for ages 8-12/13-17 years) was used to assess HRQoL. Parenting stress was evaluated using the Dutch short version of the Parenting Stress Index (NOSIK). Results were compared to available normative data. The Illness Cognition Questionnaire-parent version was used to assess parental illness cognition. Results were compared to available data of parents of children with childhood cancer.
Results: Questionnaires of thirty-eight patients (median age: 13 years; 48% male) and their parents were completed. Self-reported HRQoL was not significantly impaired, except for social functioning in children aged 8-12 years. Gender, severity of tricuspid valve regurgitation or previous cardiac surgery did not predict self-reported HRQoL ($p=0.606$, 0.846 and 0.426, resp.). There was a poor agreement of HRQoL Total Scores between patients and their mothers (ICC 0.514, $F(27) = 3.104$, $p=0.002$) as well as their fathers (ICC 0.567, $F(27) = 3.552$, $p=0.01$). Agreement between both parents was good (ICC 0.858, $F(27) = 15.948$, $p=0.000$). Normal levels of parenting stress were observed: the median NOSIK score was borderline average (median 43.0, range 25-107). Nevertheless, four parents (10%) reported high to extremely high scores (for mothers ≥ 74 and for fathers ≥ 64),

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indicating worrisome levels of stress. Parents reported good acceptance of their children's disease and few cognitions of helplessness. *Conclusions:* We recommend routine assessment of HRQoL and timely screening for psychosocial problems not only in children with EA, but also in their parents, as their perspectives might differ substantially. Early identification and intervention may positively influence the whole family's wellbeing.

P280

Intellectual functioning in children with congenital heart defects: a follow up study

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Introduction: **Background**: Studies report risk for children with congenital heart defects (CHD) to develop lower intellectual functioning compared to healthy children. However, few studies look at intellectual functioning in these children over time and describe factors related to differences on intellectual functioning. This study compares intellectual functioning in children with congenital heart defects in relation to severity of the heart defect, the child´s age and the socioeconomic status of the family (SES) in children with CHD over time.
Methods: **Methods**: 227 children with CHD were tested using the Wechsler intelligence scales to determine Full Scale IQ (FSIQ). FSIQ was then analyzed in relation to age (3-, 5-, 9-, and 15-year-olds), severity of the diagnosis (mild, moderate, and severe), and SES (low, medium, and high). 109 children (48 %) were tested one time and 118 children (52 %) tested 2 or more times. A mixed model repeated-measures is used for statistical analysis. The analysis was performed using JMP statistical software (SAS institute).
Results: **Results**: Children with severe CHD had significantly lower FSIQ than children with mild CHD ($p = 0.0079$). The 9 and 15-year-olds had significantly lower FSIQ compared to the 3-year-olds ($p = 0.0009$ and $p = 0.0050$ respectively), the 9-year-olds had also significantly lower FSIQ compared to the 5-year-olds ($p = 0.0310$). Children from families with low SES had significantly lower FSIQ than children from high SES-families ($p = .0008$) and medium SES- families ($p = 0.0001$). All these tests were performed controlling for the other factors.
Conclusions: **Conclusions**: Severity of the heart defect, age of the child and SES of the family had significant main effects on FSIQ over time. Late effects of the disease were noteworthy. These factors should be considered when planning interventions and follow-up programs for children with CHD.

P281

Life satisfaction and Quality of Life in children and adolescents treated for valvular aortic stenosis: Self-report and proxy-ratings

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Introduction: Congenital valvular aortic stenosis is a life-long disease which occurs in 3-5% of children with CHD. The severity of the disease varies from a life-threatening condition at birth to a condition which may only result in interventions later in life. Due to the seriousness of the defect and the possible restrictions it may impose on the child or adolescent, it is important to investigate the life satisfaction and quality of life for the children diagnosed with the condition. Life satisfaction is the evaluation of one's life as a whole and have been found to be correlated with a number of different factors such as general health as well as happiness.
Methods: 48 patients 8-18 years old treated for valvular aortic stenosis were recruited. Among the patients 14 (30%) have had two or more aortic valve interventions including 9 having had an aortic valve replacement. The patients were matched with 43 healthy controls by age, gender, geography and measurement period. The participants filled out the age-appropriate Satisfaction with Life Scale as well as Kidscreen-52, which measures quality of life. The parents of the children and adolescents answered the proxy-version of the Kidscreen- 52.
Results: Life satisfaction and quality of life scores did not differ significantly between children and adolescents treated for aortic stenosis compared to children without the condition. Neither was any difference found between parents´ rating of their children's life satisfaction and quality of life. Parents of adolescents treated with aortic stenosis did however experience that their adolescents had less autonomy compared to view of the parents of the healthy controls.
Conclusions: Overall children and adolescents with aortic stenosis report a similar degree of life satisfaction and quality of life as children without the diagnosis according to self-reports and proxy-reports.

P282

Reduction in Health-related Quality of Life during Covid-19 pandemic in Children with Congenital Heart Disease – Findings from self and proxy report

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Introduction: Social distancing, extensive bans on contacts, curfews, and required wearing of masks in public places have - while unavoidable for disease containment purposes - caused major disruptions to everyday life in face of the Coronavirus Disease 2019 (COVID-19) pandemic. The aim of this study was to figure out how the COVID-19 pandemic affects the Health-related Quality of Life (HRQoL) of children and adolescents with CHD, as well as how the parents perceive the HRQoL of their children.
Methods: HRQoL was assessed by KINDL® questionnaire during the COVID-19 pandemic and compared to the children's most recent completion of the questionnaire out of the FOOTLOOSE study setting (German-Clinical-Trial-Register-ID: DRKS00018853) at the outpatient department of the German Heart Center Munich (DHM). From May 27th to June 29th 2020, 160 German children with various CHD (15.2 ± 2.5 years, 62 girls) completed this re-assessment of HRQoL. Mean follow-up period was 2.1 ± 1.7 years. Difference between children's self-reported HRQoL and parents' proxy report was calculated with a paired student T-Test, and agreement of the respective ratings with intraclass correlation coefficient (ICC) and their 95% confidence intervals.

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Results: HRQoL in children with CHD was significantly lower during COVID-19 pandemic compared to before in total KINDL® score (by -2.1 ± 12.3, p=.030), and the subcategories emotional well-being (by -5.4 ± 1.2, p<.001) and friends (by -4.5 ± 1.7, p=.009). Parents of children with CHD rate the HRQoL in total KINDL® score (mean difference: 3.9 ± 1.2, p=.002), and the subcategories family (mean difference: 8.8 ± 1.7 SEE, p<.001) and friends (mean difference: 7.6 ± 2.2 SEE, p<.001) even worse than their children. Only moderate degree of agreement was found between most of the sub-categorical HRQoL assessment of children with CHD and their parents.
Conclusions: The COVID-19 pandemic had a negative impact on HRQoL in children and adolescents with CHD and their families. Furthermore, parents rate the HRQoL of their chronically ill children even worse than the children themselves. Specifically, psychological concerns of children with CHD and their families should also be considered by health care providers during the COVID-19 pandemic.

P283

Results reported by patients with CHD and their parents regarding the knowledge of their disease valued graphically and descriptively

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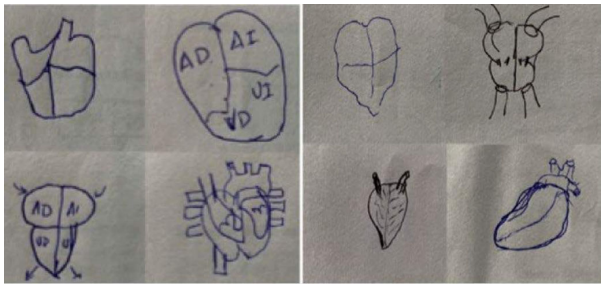
Introduction: Patients with congenital heart disease (CHD) are a chronically ill population. The knowledge of the disease is a key factor in promoting an appropriate attitude towards health. For this, it is necessary a correct understanding of the disease, both on the part of the children and of the parents.The objective of this study is to assess the compression the heart condition in children with CHD and their parents, by locating the heart, drawing a picture and describing the heart defect, and assess whether their understanding correlates with age.
Methods: Descriptive cross-sectional study over a period of 6 months in a Tertiary Center of Cardiology Paediatric, involving 157 parents, 60 patients up to age 8 (41% girls) and 54 with 9-18-year-old (46% girls). It is carried out using a questionnaire, which includes a silhouette of a person where must locate the heart, a blank box where children over 8 years old and their parents must draw a picture of the heart disease, and a question from which they must describe the disease
Results: 76.4% of parents know the name of their child's illness. 53.7% of children over 8 years old know it and 35% of children under 8 know it. 82.6% of parents correctly locate the heart in a person's silhouette as the 70.4% of children between the ages of 9 and 18. However only 61% of children up to the age of 8 place it. Only 3.4% of parents correctly draw their children's CHD. Of children over 8 years of age, 3.8% correctly draw their heart disease. 18.5% of children over 8 years old adequately describe their illness. Of children under this age, 100% do not know how to describe it. Of the parents, only 46.5% describe it correctly.
Conclusions: The compression of CHD reflected in graphic and descriptive form is inappropriate in both parents and children. An effort should be made on the part of the professionals, in trying to explain the CHD to achieve a better understanding of the disease and thus to optimize the behaviour in terms of health.



P284

The Impact of COVID-19 on paediatric cardiology trainees in the UK.

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Introduction: In March 2020 the UK reported its first coronavirus related death. The weeks following saw rapid and frequent changes to the delivery of healthcare; face-to-face appointments were limited, elective procedures cancelled, and concerns about the availability of PPE began to surface. The potential impact of changing personal and professional circumstances on junior doctor well-being has been alluded to in statements from all UK Royal Colleges. **We present results from the first part of a planned longitudinal prospective cohort study tracking trainee experiences during the pandemic.**
Methods: In May 2020, two months after than start of the pandemic, all UK paediatric cardiology trainees were invited to participate in an electronic survey. Questions addressed the work environment, training experiences and personal circumstances. Individual interviews were also conducted with 4 trainees.
Results: 30/45 (67%) paediatric cardiology trainees from 7/10 U.K training regions (Liverpool, Cardiff and Belfast were not represented) completed the survey. Respondents were mostly aged 30-40 (77%), 48% female and 6 (20%) from ethnic minority backgrounds. The majority were working full time 27 (90%) and in higher/sub-speciality training 23 (77%). Most felt safe at work (89%) with access to appropriate PPE. Perceptions of workload intensity varied by region. 79% received formal teaching at 50%-100% of pre-pandemic levels. However, 93% reported reduced opportunities for sub-speciality training; 61% characterised this as very significant. Well-being was almost unanimously negatively affected.
Conclusions: Even during the first wave of the pandemic, the majority of U.K paediatric cardiology trainees felt safe at work. Workload intensity varied, reflecting changes in the configuration of cardiac services; in 1 London Centre and the East-Midlands, surgical/interventional activity was paused. Higher/sub-speciality training was most impacted; where elective activity provides most experience. Well-being was virtually unanimously negatively impacted. COVID-19 has been the defining global healthcare

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crisis of the modern era. There has been a measurable impact on paediatric cardiology trainees; fewer cardiac catheterisations', restricted fetal screening, and a more emergency-driven case mix. Consideration of adjustment to training duration may be required pre-certification and our follow-up survey will aim to evaluate the longer-term implications of the pandemic on training.

P285

What about mental health in children with CHD compared to their healthy twin? - First preliminary results of the "Same Same, but different?" project

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Introduction: Mental health is a construct strongly influenced by epigenetics, socioeconomic aspects as well as family structure. Congenital heart defect (CHD) accompanies patients all their life and is assumed to have greater influence on mental health. This study aims to figure out the impact of CHD without the confounding factors epigenetics, socioeconomic aspects and family structure in twin siblings.
Methods: Out of the "Same Same, but different?" project the parents of 25 pairs of twin siblings (one with CHD and the other without, 30 girls (60%), 10.2±4.2 years) filled in the strength and difficulties questionnaire (SDQ) for mental health for both of their children. The SDQ consists of five dimensions and a total difficulties score for mental health. The distribution of the values for interpreting and defining a disorder in the reference is defined that about 80% of the children are classified as normal, 10% as borderline and 10% as abnormal.
Results: In all 50 children, the total difficulties score showed 82% normal scores, 12% borderline and 6% abnormal. In terms of sub dimensions they showed 68% normal, 18% borderline and 14% abnormal in the dimension of emotional symptoms, 78% normal, 12% borderline and 10% abnormal in conduct problems dimension, 80% normal, 2% borderline and 18% abnormal in hyperactivity, 76% normal, 6% borderline and 18% abnormal in peer relationship problems an in prosocial behavior 90% normal, 4% borderline and 6% abnormal. For 27 (54%) children, parents reported conspicuous behavior, which in 7 (14%) children led to clearly family burdens, in 11 (22%) to slight family burdens, and in 9 (18%) to no burdens. The direct twin sibling comparison using the paired-sample t-test showed significant differences in total difficulties score (p=.008), in peer relationship problems (p=.018) and hyperactivity (p=.008). The other dimension showed no significant difference between the twins.
Conclusions: Children with CHD suffer from mental health problems in a direct comparison with their twin siblings in terms of total difficulties score, peer relationship problems, and hyperactivity, and the parents reported on family burdens that cannot be ignored. These results underline the importance of psychological guidance for the whole family.

11. Morphology

P286

3D Printing in Congenital Heart Disease: A Single Tertiary Cardiac Centre Experience 11. Morphology

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Introduction: 3D printing of congenital cardiac anatomy is still in its infancy with only a handful of units utilizing this technology on a regular basis. More user friendly and widely available 3D segmenting software, better 3D printing technology and materials as well as reducing costs, will enable more widespread use of this. 3D printed cardiac models can be used for surgical planning, teaching and hands-on training and education and counselling for patients.
Methods: We reviewed indications and outcomes for 3D printed cardiac models at our institution since 2017.
Results: There were 40 requests made over 4 years (6 in 2017, 10 in 2018; 8 in 2019 and 16 in 2020). The most common indications for printing were to assist in surgical or interventional planning and parental counselling in cases of complex cardiac anatomy, to demonstrate interesting cardiac anatomy for teaching purposes, to aide surgical training and parental education in common cardiac abnormalities. Models comprised a mix of blood volume and hollow-heart models printed depending on the indication. There is evidence of service development with time.
Conclusions: We have enhanced the quality of our 3D segmenting and printing processes using more pliable materials where necessary, providing more accurate representation of life-size cardiac anatomy and improving turnaround time. We have also increased selectivity in the cases printed. We envisage that 3D printed heart models will be increasingly used in teaching and training of the future generation of doctors and technicians. 3D printing can complement conventional imaging modalities, can assist anatomy teaching and can enhance interventional and surgical skills. Cost continues to be the limiting factor for more widespread use of this modality.

P287

A long-term survival case with neonatal Marfan syndrome

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Introduction: Neonatal Marfan syndrome(nMFS) is a rare and life-threatening disease due to refractory heart failure from severe mitral valve regurgitation (MR). We experienced a successful followed-up case with nMFS after surgical repair for mitral valve regurgitation and aortic root dilation, both were rapidly progressed with age. Optimum surgical timing and medical treatment is crucial in management of nMFS.
Methods: case report
Results: She was born in 38 weeks gestation without asphyxia, and diagnosed as nMFS in neonatal period because of characteristic phenotype and large deletion in FBN1 gene.From the age of 6

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months, severe MR due to mitral valve prolapse (MVP) and despite early angiotensin-converting-enzyme inhibitor administration, LV dilation progressed and the end-diastolic left ventricular diameter (LVDd) in echocardiography exceeded the normal ratio by 150%. Thus, mitral valve surgery was undergone. Mitral valve was repaired with edge to edge technique for MVP and ring annuloplasty for annular dilatation using Physio 26 mm ring (Edwards Lifesciences, Irvine, Calif). (Figure1) Mitral valve regurgitation remains mild postoperatively with no progression, and the LVDd remains normalized (Figure2). Mitral stenosis has not progressed. During subsequent follow-up, aortic root dilatation progressed. Beta blocker was started when Valsalva size exceed 25mm, and then angiotensin II receptor blocker was added-on. At the age of 9 years, valve-sparing aortic root replacement using the David technique was employed with normalization of her aortic root dimension as Valsalva size reached to 35mm. She has been doing well postoperatively without progression of MR and AR. Changes in left ventricular diameter and aortic root size over time on echocardiogram are shown in the figure2.

Conclusions: This case, to our knowledge, is the first report on neonatal Marfan syndrome after mitral valve repair with favorable late outcome. Early medical management and an optimal surgical intervention would prevent from refractory heart failure and sudden death.

Figure1

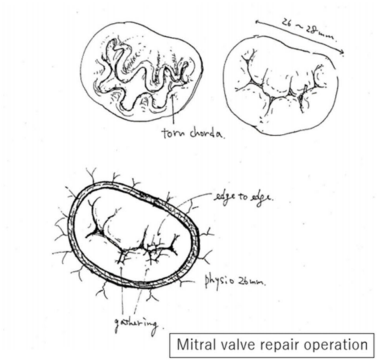
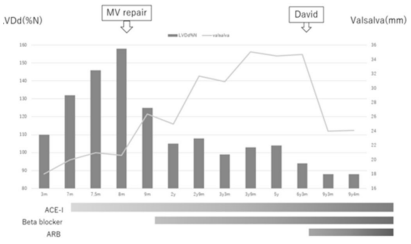


Figure2



P288

A single centre case series of atrial isomerism in the context of congenital heart disease

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Introduction: This case series describes the progress of children following a diagnosis of atrial isomerism in the context of congenital heart disease. Atrial isomerism is widely regarded to have a poor prognosis especially in children whose anatomy leads to a single ventricle circulation. The aim of this series is to follow the child from antenatal or postnatal diagnosis; through corrective and palliative surgery; assessing the outcome in terms of morbidity and mortality at each stage.

Methods: This was a single centre retrospective case notes review of all patients diagnosed with atrial isomerism (antenatally or postnatally) between 1/1/2011 and 31/12/2016. The electronic record was used to identify patients at diagnosis and then to follow the progress of the fetus or child.

Results: 40 fetuses were suspected to have atrial isomerism antenatally and a further 12 babies were diagnosed postnatally. The clinical course is illustrated in figure one. Cardiac diagnosis included atrioventricular septal defect in 72.4% (n=21) which was unbalanced in 8 (38%). Extracardiac co-morbidities included primary ciliary dyskinesia (2), biliary atresia (3) and oesophageal atresia (2). In total just 36.5% of babies that received a diagnosis of isomerism were alive at one year of age (n=19) compared to 70.8% of babies who survived to first intervention. 5 babies died pre cardiac intervention (17.2%) and 2 died within 7 days of their first intervention. There were 5 further deaths in the first year of life. 4 children died over the age of 1 of whom 2 died in the post-operative period (one post redo relief of obstructed pulmonary venous drainage aged 2 years, one post Fontan completion aged 9 years). The other 2 deaths were not related to cardiac intervention. The outcome was equally poor regardless of antenatal diagnosis, right or left sided isomerism and single ventricle status.

Conclusions: This case series has highlighted the poor prognosis associated with a diagnosis of atrial isomerism. The majority of children required multiple interventions with mortality associated with each stage. This clearly illustrates the importance of careful counselling at diagnosis to give the family a realistic expectation of the clinical course that is to be expected for their child.

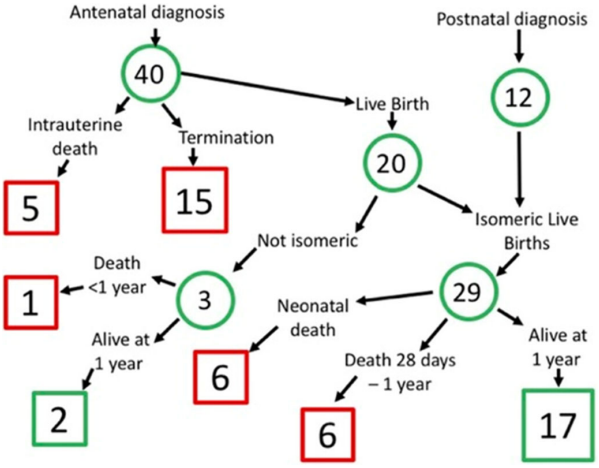

Figure 1: A study diagram depicting the outcome of the patients with atrial isomerism and congenital heart disease.

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P289

Case report: Aortic interruption in a 5-year old male presenting with hypertension

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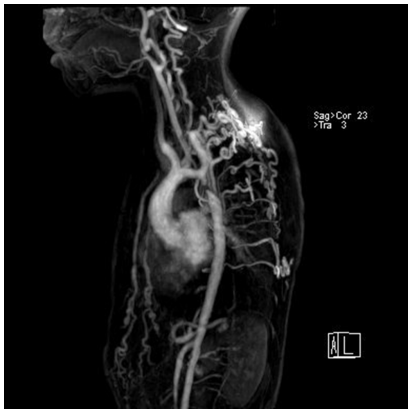
Introduction: An interrupted aortic arch is rare congenital malformation occurring in 3 per million newborns. It is commonly detected in the neonatal period and characterized by a complete loss of anatomical continuity between the ascending and descending portions of the aorta. If left without surgical repair, it is associated with a high mortality.
Methods: We present a 5-year old male who was transferred to our clinic due to arterial hypertension.
Results: The patient was found with abnormal blood pressure with a mean level of 130/74 mm Hg and significant difference between the upper and lower limbs. Transthoracic echocardiography was suspicious for an interrupted aortic arch. During cardiac catheterization, the arterial catheter could not advanced above the fifth thoracic vertebrae. With a transseptal approach, an aortic angiography showed a discontinuity between the aortic arch and the aorta descendens with a blood pressure difference of 55 mm Hg. Cardiac MRI revealed an aortic interruption with a length of 3.5 mm and numerous collaterals, including intercostal and mammal arteries, mainly from the left side (Figure 1). Surgical repair was successfully performed with an end-to-end anastomosis and a patch angioplasty. Postoperative controls showed a good result with a Vmax of 2.6 m/s in transthoracic echocardiography, no postoperative difference of blood pressure in the upper and lower limb and no hypertension.
Conclusions: If left untreated, an interrupted aortic arch is associated with a high mortality. The only chance of surviving is by developing a collateral circulation. In this case symptoms can be mild or absent. In many patients the only sign is arterial hypertension, which is often diagnosed late. Our patient showed no other symptom than hypertension, which was diagnosed in the routine examination in children. This circumstance underlines the importance of the examination of the pulses and blood pressure in the upper and lower extremities. According to the literature, diagnosis of an interrupted aortic arch at a later stage in life is associated with a higher complication rate and can lead to a different treatment regimen.Our situation made it possible to repair the interrupted aortic arch with an end-to-end anastomosis, which is the preferred surgical repair procedure.



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Complications Derived from Bicuspid Aortic Valve (bav) in a Pediatric Population

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VALL D'HEBRON HOSPITAL (1)

Introduction: To determinate complications derived from bicuspid aortic valve (BAV) in a pediatric population.
Methods: Review of patients with diagnosis and follow-up of BAV from 1997 to 2019 in a tertiary hospital.
Results: 281 patients were included, 68.5% male, median age at follow-up 8.24±2.4y. Family history of BAV was known for 7.5% of patients. At diagnosis, aortic stenosis (AS) was described in 117/281(42.6%) patients: mild 21.9%, moderate 7.9% and severe 16.1%. The majority was male (72%). In AS greater than mild, valve morphology was vertical in 37/66 (56%) and more often with raphe 55/66 (83%, P=0.014). A total of 66 aortic valve procedures were performed in 47 patients (22%). First procedure at median age of 2.4±3.8y: 36/47(76%) percutaneous valvuloplasty and 11/47(23%) open heart surgery (10/47 valvulotomy, 1/47 mechanical aortic valve replacement). Severe AS was the indication in 46 patients, and severe aortic root dilatation in 1. During follow-up, 12/47 patients (25%) underwent a second procedure at a median age of 6.48±5.5y: 5/12 percutaneous valvuloplasty, 4/12 open-heart aortic valve repair, 2/12 mechanical aortic valve replacement and 1 Ross-Konno. Indication was severe AS in 8 patients, severe aortic regurgitation (AR) in 3 patients and severe double aortic lesion in 1. A third procedure was performed in 6 patients at a median age of 8.7±5.3y: 3/6 Ross-Konno, 1/6 mechanical aortic valve replacement, 1/6 open-heart valvulotomy and 1/6 percutaneous valvuloplasty. Procedure was secondary to a severe AS in 3 patients and severe AR in 3. A fourth procedure was performed in 1 patient at 14 years of age secondary to an aortic aneurysm. Residual AR was described in 38/47 patients (80%): 23/38 mild (60%), 5/38 moderate (13%), 10/38 severe (26%). A total of 6/38 patients (15%) underwent a procedure as consequence of severe residual AR. A case of endocarditis was described in a patient with three procedures over BAV.
Conclusions: There is a statistically significant relationship between vertical valve morphology with raphe and procedures over BAV. Most of severe AR in the pediatric population are secondary to procedures performed on AS.

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Fontan procedure: 35-years single centre experience

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Introduction: Fontan procedure is surgical treatment for about 6% of the patients with congenital heart defects when biventricular circulation cannot be achieved. Contributing factors to long-term survival and freedom of complication are still researched.
Methods: Patients with Fontan circulation, treated at Mother and Child Health Institute of Serbia from December 1983 to September 2018, were retrospectively analysed regarding heart defect, age, type of surgery, complications and outcome.
Results: The study included 57 patients, with following diagnoses: tricuspid atresia (40.4%), double inlet left ventricle or

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univentricular heart (26.3%), pulmonary atresia with intact ventricular septum (10.5%), hypoplastic left heart (15.8%) and unbalanced complete atrioventricular (AV) canal (7%). There were 33/57 males. The first stage procedure included modified Blalock-Taussig shunt in 45.5% and pulmonary artery banding in 30.9% of the patients. Glenn anastomosis was performed in 79.6% of the patients, at median age 11.8 months. Fontan procedure was completed at median age 4.3 years and considered atriopulmonary connection (10.5%), lateral tunnel (7%) and extracardiac conduit (82.5%), with fenestration in 26% of the patients. Cardiopulmonary bypass (CPB) duration was 111.4 ± 50.7 min. Follow-up of the whole group and survivors was 6.7 ± 7.4 and 9.5 ± 7.3 years, respectively. Thrombosis occurred in 18.5% of the patients averagely 9.2 years after surgery, with elevating chance for thrombosis by every additional year of follow-up (OR = 1.1, CI: 1.012-1.217). Other complications were early and late arrhythmias (21.8%), along with protein losing enteropathy (10.9%) on average 1.6 years postoperatively. Mortality rate was 22.8%, entirely in the first three postoperative months. Mean predicted survival age was 20 years. Favourable factors for survival showed on the Cox regression model were dominant left ventricle (OR = 3.55, CI: 1.04-12.11) and shorter CPB measured in hours (OR = 3.23, CI: 1.42-7.41). Mean predicted survival age among patients with dominant left and right ventricle was 21.9 and 8.8 years, respectively.

Conclusions: After critical first trimester, long-term outcome in our group of patients with Fontan circulation is optimal, in accordance with the findings of other authors. During time the risk of thrombosis increases. Independent predictors of survival are dominant left ventricle and shorter CPB time.

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Mitral stenosis as primary anomaly of multilevel left heart obstruction in Shone complex with insignificant hypoplasia of the left ventricle

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Introduction: Objective: Patients with multilevel obstructions of left heart structures have a high risk for neonatal and infant morbidity and mortality. We analyzed and demonstrate the morphological spectrum of 7 neonatal autopsy cases of the Munich Heart Collection (1974-1992), with the clinical diagnosis of coarctation of the aorta (CoA)and ventricular septal defect (VSD), and 10 infant autopsy specimens from Boston Children's Hospital (1964-1994) with Shone complex, considering diagnosis and surgery.

Methods: Examination of all 17 heart specimens included measurements of the length from apex to AV-valves and semilunar valves, heart valve diameters, and thickness of right and left ventricular myocardium.

Results: Mean age at death of the Munich collection was 14 days (5 to 33 days) and that from Boston 99 days (1 day to 19 months). All hearts presented: Mitral stenosis with proximal orifice hypoplasia and distal complete or partial type parachute mitral valve, muscular subaortic stenosis with prominent "bulge" of superior-anterior LV-outlet septum (mean thickness 12mm), CoA with aortic arch hypoplasia, significant hypoplasia of both mitral and aortic valves (mean diameter ratio of left-to-right AV-valves 0.5 and 0.6 for semilunar valves, insignificant LV-hypoplasia (mean LV/RV length ratio of inlet and outlet equally 0.9), normal segmental connection, patent foramen ovale (PFO) (in one half restrictive). Additional findings: VSD (11 perimembraneous-inlet, 4 muscular-trabecular, none in 2), supramitral ring in one, endocardial fibroelastosis (EFE) in three. Surgery in 11 cases: CoA repair with duct-closure in 8, additional pulmonary artery banding (PAB) in 5, Norwood in 2, and incision of muscular subaortic stenosis in 1, VSD-closure in 4.

Conclusions: Our results imply that early multilevel intervention, possibly including temporary aorto-pulmonary amalgamation, may be crucial for survival of infants with Shone complex and insignificant LV– hypoplasia and VSD. Efforts to relieve mitral stenosis must be considered besides CoA-repair and myectomy of muscular subaortic stenosis ("bulge").

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Pitfalls in echocardiography: Anomalous origin of the right vertebral artery from the right carotid artery simulating normal brachiocephalic branching

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Introduction: Determination of brachiocephalic branching is an integral part of echocardiographic studies in children with CHD. The normal left aortic arch (LAA) is characterized by origin of the brachiocephalic artery as the first vessel to the right, branching into right subclavian and carotid artery. We report three patients with left aortic arch and origin of the right vertebral artery (RVA) from the right common carotid artery (RCCA) associated with an aberrant right subclavian artery (ARSA) originating from the descending aorta.

Methods: Our first patient presented in the neonatal period with tetralogy of Fallot. Echocardiographic imaging of the aorta revealed LAA with seemingly normal branching of the first vessel to the right. Later cardiac catheterization following corrective surgery showed ARSA from the descending aorta. The first vessel from the aorta was the RCCA giving origin to the RVA and thereby imitating normal brachiocephalic branching. Our second patient presented with aortic coarctation. The first vessel from the aortic arch showed seemingly normal brachiocephalic branching. Further evaluation however revealed ARSA from the descending aorta. CT-scan confirmed ARSA and anomalous RVA origin from the RCCA. A third patient presented with AVSD and trisomy 21. Again echocardiographic evaluation of the aortic arch showed seemingly normal branching of the first vessel to the right while it was possible to demonstrate ARSA from the descending aorta.

Results: Prevalence of the RVA originating from the RCCA is given with 0.18% in the literature. However this anomaly is not uncommon in patients with ARSA, which frequently occurs in children with trisomy 21 or 22q11.2 deletion syndromes. While almost all vertebral artery anomalies have no clinical impact in childhood, anomalous origin of the RVA from the RCCA can imitate normal brachiocephalic branching in LAA despite the presence of ARSA. This is of significance in patients requiring surgical treatment of coarctation or interrupted aortic arch.

Conclusions: Echocardiographic visualization of branching of the first aortic arch vessel does not exclude aberrant origin of the right subclavian artery. Especially in infants requiring surgery for obstructive aortic arch lesions echocardiography should also focus on direct visualization of possible ARSA from the descending aorta.

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Unilateral Pulmonary Artery Agenesis and Vascular Ring, a Rare Association

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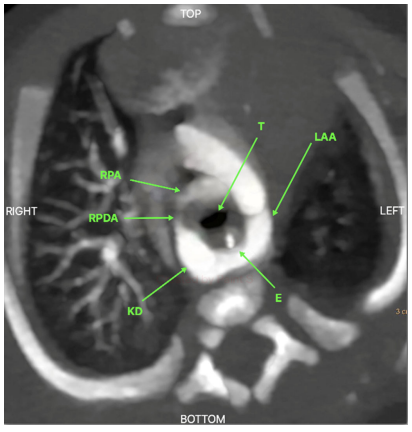
Introduction: Congenital unilateral pulmonary artery agenesis (UPAA) and the aortic arch anomalies forming vascular ring are uncommon anomalies. We describe for the first time the association of both rare malformations.
Methods: We report a patient with left-sided congenital pulmonary artery agenesis with a vascular ring, since the fetal life evolution to the postnatal management.
Results: The patient was diagnosed at the 20 gestational weeks on a left retroesophageal circumflex aortic arch with a right ductus arteriosus. After delivery, clinical examination detected mild polypnea and inspiratory stridor. Chest radiography was normal. A transthoracic echocardiography showed a structurally normal heart with a restrictive patent ductus arteriosus and a vascular ring. A distal absence of the left pulmonary artery was suspect, so a pulmonary computerized tomographic (CT) angiogram and a thoracic magnetic resonance imaging (MRI) were performed; both detected an agenesis of the proximal left pulmonary artery and a vascular ring composed by a left aortic arch (AA), a right ductus arteriosus (DA) that emerges from a Kommerell diverticulum (KD) and reaches the right pulmonary artery, closing the ring. There was an aberrant right subclavian artery (ARSA) near the DA (Figure1). Surgical repair was performed. Further evaluation by angiography showed mild hypoplasia in the transverse AA and mild segmental coarctation proximally to the KD and the ARSA. The absence of the left pulmonary artery was confirmed.
Conclusions: Due to the low incidence and the usual lack of symptoms, vascular rings and UPAA are frequently underdiagnosed malformations. Prenatally, it could be suspected with an echocardiography, but in the postnatal life, echocardiography usually need a complemetary image technique, as in the case we described. MRI is noninvasive and has a superior diagnostic power, so in some cases it could replaces CT and angiography. Other diagnosis images that could be useful are the chest radiography, ventilation-perfusion scanning, and esophagography. The election treatment in vascular rings is the early surgical repair, especially in symptomatic patients; in the UPAA the surgical options are limited and have no very successful long-term outcomes.



12. Preventive

P295

An intracardiac foreign body: The case of a missing suture in an operated infant with congenital heart disease

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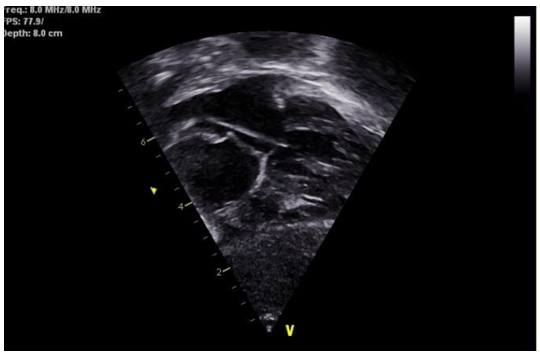
Introduction: Intra-cardiac foreign bodies, without a history of penetrating trauma, are a rare occurrence and there is minimal literature on retained sutures post cardiac surgery. Due to the fine structure of the sutures they are difficult to identify on many traditional imaging techniques. Foreign bodies can reach the heart in different ways including intra-operatively, the most common route is via migration of a medical device. Techniques for removing post traumatic or iatrogenic foreign bodies include percutaneous, endovascular and an open surgical approach. The need for removal of foreign bodies should be individualised depending on the nature of the structure, and the actual or potential side effects.
Methods: A 6 month old infant with a background of VACTERL was accepted for routine tetralogy of fallot repair. The patient had a background of a previous repair of long gap oesophageal atresia and therefore an intraoperative transosophageal echocardiogram was contraindicated and an epicardial echo was performed. The patient recovered well from surgery but routine postoperative echocardiograms demonstrated a linear structure originating in the right upper pulmonary vein and extending into the left atrium. This was seen to cross the mitral and aortic valve and discontinue at the origin of the innominate artery.
Results: This case was discussed in the joint cardiology and surgical conference and the decision made to perform a surgical re-exploration and removal of the presumed foreign body due to the uncertainty around the nature of the foreign body. The previous sternotomy site was re-opened and the patient placed onto bypass. The aorta was opened transversely and a length of prolene thread was identified lying within the left atrium originating from the purse-string suture that had been used for the LV vent.
Conclusions: Detecting this suture using various imaging modalities was a challenge due to its radiolucent nature. Our unit practice is to perform a transoesophageal echocardiogram in the operating theatre but in this particular case, this was contraindicated and therefore an epicardial echocardiogram was performed. This case highlighted the importance of unit familiarity with both tranoesophageal and epicardial echocardiography intra-operatively and the importance of echocardiography by multiple operators.



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P296

Arterial wave reflection in patients with congenital cardiac anomalies - comparison of three methods

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Introduction: Central augmentation index (cAIx) is a well-established marker for arterial stiffness. Alternative methods that assess arterial stiffness based on the peripheral wave form are less well established. It is the aim of this study to compare different methods that analyze arterial wave reflection in congenital aortopathy patients and healthy controls.
Methods: Demographic parameters were recorded. CAIx, defined as the difference between the reflected (P2) and forward (P1) wave heights divided by the pulse pressure, and central mean pressure were measured (SphygmoCor XCEL, AtCor). Arterial wave reflection was evaluated peripherally by 1. peripheral AIx (pAIx) which is defined as (P2-P1)/P1 (EndoPAT, Itamar), and 2. aging index (AI) which is based on the acceleration curve of the digital pulse wave (DPA, Meridian). For statistical analyses, linear regression analysis correcting for covariates was performed. Correlations between different methodologies were assessed using Spearman correlation coefficient.
Results: 113 patients with congenital aortopathies (53 with repaired aortic coarctation without restenosis, 44 bicuspid aortic valve with at most moderate stenosis or insufficiency, 16 with Marfan syndrome), 27 Fontan-palliated patients and 78 controls were included in the study. Age range 8-65 years. Following correction for the effects of age, sex, height, heart rate and central blood pressure, all three parameters of wave reflection were significantly abnormal in the Aortopathy and Fontan groups compared to Controls. In the Control and Aortopathy groups, all parameters of arterial wave reflection correlated strongly $r>=0.7$, $p<0.001$), while there was no significant correlation between any of the above parameters in the Fontan group ($r<0.13$, p NS).
Conclusions: Using multiple modalities we show that arterial wave reflection is increased in patients with congenital aortopathies and Fontan patients. While the three methods of assessing arterial wave reflection correlate strongly with each other in the Aortopathy and Control groups, this is not seen in Fontan patients. Thus, parameters of arterial wave reflection should be interpreted with caution in Fontan patients.

P297

Assessment of a Palivizumab prophylaxis program in children with congenital heart disease

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Introduction: Respiratory syncytial virus (RSV) causes potentially severe respiratory infections in children. Infants with congenital heart diseases (CHD) are at higher risk. Prophylaxis with Palivizumab, a humanized IgG monoclonal antibody, is part of the most effective prevention method nowadays, reducing severity of infections. The aim of this study is to evaluate a prophylaxis program with Palivizumab in infants with CHD, regarding safety, follow-up and family impact.
Methods: An observational prospective study is presented, including patients 0-24 months of age with CHD and indication for prophylaxis with Palivizumab based on current European guidelines, along October 2018 to February 2019 vaccination program. All of them were followed-up in a national reference hospital for CHD. Palivizumab was administrated intramuscularly monthly (15 mg/kg dose). Families also received health education. Data were collected from medical records and forms completed by parents at the beginning and end of the program. Statistical study was carried out by Stata/IC 15.1.
Results: 69 patients with CHD were included in the program. 53 completed the study, 51% females, 17% with Down syndrome. Median age at the time of first administration was 8.5 months. Along vaccination period, 35% patients presented more than 2 respiratory affections, compared to 17% who did not present any. 50% did not need medical assistance, versus 12% who did so more than twice. 85% of children were not hospitalized, and only 2 required admission to PICU. All infants admitted required oxygen therapy, only 1 invasive MV. RSV was isolated in less than 25%. 4 patients had their cardiac surgery delayed due to bronchiolitis. These results show similar admission and oxygen therapy rates comparing to intervention groups in international scientific researches with control group. 0-12 months group presented more respiratory affections and medical assistance than 12-24 months group. 50% parents had to miss work any time. 4 children presented mild side effects after Palivizumab administration (3 agitation, 1 fever).
Conclusions: Children with CHD are a higher risk group for RSV infection, with significant clinical and family impact. Prophylaxis with Palivizumab reduces disease severity and admission needs, without relevant side effects. Identification and inclusion of children who may benefit from prophylaxis is essential.

P298

Better lung function after catheterization therapy vs surgical closure in patients with congenital cardiac shunts

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Introduction: In left-to-right cardiac shunts (atrial septal defect, ventricular septal defect, patent foramen ovale or persistent ductus arteriosus) surgical closure used to be the only treatment. Unlike catheterization, iatrogenic injuries of the rib cage, lungs or phrenic nerves may occur. Since catheter closure of these defects is available, lower co-morbidities are noticed. This study assesses differences of lung functions between the two procedures.
Methods: Spirometry data from October 2001 to March 2019 from 293 patients (31.97 ± 15.77 years, 174 females) were retrospectively analyzed. Forced expiratory volume within the 1st second (FEV_1), forced vital capacity (FVC), and Tiffeneau-Pinelli index (FEV_1/FVC) were compared in patients who had surgical closure (n=188) vs catheter therapy (n=105). Restrictive (FVC) and obstructive (FEV_1/FVC) were defined with a z-score <-1.64 (GLI 2012 references).
Results: FVC ($z=-1.18 \pm 1.18$, $p<0.001$), FEV_1 ($z=1.28 \pm 1.23$, $p<0.001$), and FEV_1/FVC ($z=-0.23 \pm 1.15$, $p<0.001$) were markedly reduced in patients with congenital shunt defects on long term after closure. Of all subjects, 97 patients (33%) were classified restrictive and 34 (12%) obstructive. The lung volume reduction was more pronounced after surgical closure compared to catheter

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closure for FVC (z=-0.92 ± 0.99 vs. z=-1.28 ± 1.21, p=0.010) and in FEV_1 (z=-0.96 ± 1.08 vs. z=-1.42 ± 1.24, p=0.001). No differences in obstructive patterns (FEV_1/FVC) were seen. While overall the older the patient at surgery was, the better the patients' FVC (ρ=0.207, p=0.004) results, no such correlation was seen in the catheterization group.
Conclusions: Patients with surgically closed repaired left-to-right cardiac shunts show smaller lung volumes resembling restrictive lung patterns compared to catheterization. This effect is directly related to the younger age at surgery in the childhood.

P299
Cardiopulmonary Evaluation of Adolescent Elite Sport Climbers

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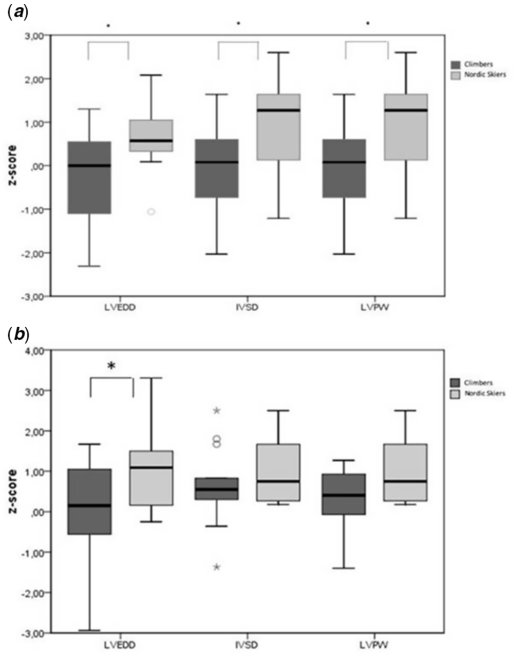
Introduction: Sport climbing has evolved into a highly professional sporting discipline and will debut at the 2020 Olympic Games in Tokyo. The IOC advocates protection and promotion of athlete health during training and competition. This requires a specific understanding of the physiologic adaptations inherent within the sport. In 2019 the first sudden cardiac death in a climber was reported during a climbing competition in Canada. So far, all the research investigating the cardiopulmonary capacity in climbers focused on climbing performance. The effects of elite climbing on the athlete's heart are unknown. This study focuses on the elite level climber's heart.
Methods: Over 5 years, all the members of the German Junior National Team Climbing (GJNTC) received an electrocardiogram, an echocardiogram, and a cardiopulmonary exercise test. 14 athletes were evaluated twice in a longitudinal approach over a timeframe of 2 years and compared to 14 athletes from the GJNT in Nordic skiing (GJNTN).
Results: 47 young climbers (20 girls, 27 boys) were examined at least once and 14 climbers were reevaluated after 27.5 months. In the echocardiogram the recorded values fell between the measurements for athletes and those for non-athletes. All left ventricular measurements increased over time but only interventricular septal thickness (IVSD) reached significance. At the second evaluation these measurements were comparable to those of the nordic skiers except for the end-diastolic diameter. The cardiopulmonary exercise test revealed a good physical fitness with limitation due to muscular fatigue in the legs. When compared to the Nordic skiers the climbers showed inferior results but high lactate tolerance.
Conclusions: Some cardiac adaptations in the sense of an athlete's heart can be observed in young elite climbers but still within normal limits. However, the significant increase of IVSD over time and the fact that the left ventricular (LV) dimensions are comparable to high-level Nordic skiers after being in the GJNTC for two years implies structural changes over time. Even though the cardiopulmonary exercise capacity is inferior to endurance athletes the lactate tolerance seems to be high in climbers.



Z-scores of LVEDD, IVSD, and LVPW of climbers and nordic skiers at the first visit (A) and after two years (B) (*signifies a significant difference between the two groups

P300
Cardiovascular effects after Assisted reproductive techniques - A pilot study.

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Introduction: More than five million children are born after assisted reproductive techniques worldwide. Long-term outcome of these children is still unclear. Several studies indicate cardiovascular changes in children, including significantly changes in arterial blood pressure and vascular dysfunction. The question raises if there are also influences on the cardiovascular health of the mother due to the necessary hormone intake.The aim of our study is to evaluate systematically cardiovascular changes in pregnancy from second trimester onwards
Methods: This prospective pilot study evaluates cardiovascular changes in the mother and her child possibly influenced by assisted reproductive techniques. Pregnant women are recruited from a center of reproductive medicine and are evaluated in a cardiovascular center in Germany. Examinations include vascular examinations of both, mother and fetus. Specific questionnaires,

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elaborate the social status, previous illnesses and living habits of the parents including physical activity. Long- term follow-up examinations of both, mother and her child, are scheduled.
Results: Since the start of the study, 13 pregnant women were examined after assisted reproductive techniques. These include fresh embryo transfer following controlled ovarian hyperstimulation and frozen-thawed embryo transfer. Healthy pregnant women serve as controls (n=107). The data are also compared to other high risk pregnancies (n=104 women with gestational diabetes) The mean age in the study group was 35.08 years (SD ±3.96), which was significantly higher than in the control group (mean age: 32.38 years SD±4.24). The average weight of the study group was 80.46 kg (SD ±11.22) which was also significantly higher than in the control group (61.95 kg SD±9.58; p=0.00). The mean thickness of the intima media (A. carotis) from the mother was 0.46 mm (SD±0.03mm) and similar to the high-risk pregnancy group with gestational diabetes (0.47mm (SD±0.004), which was also significantly higher than in the control group. The average blood pressure was 111/69 mmHg (SD±11.35), equal in all groups.
Conclusions: The intima media thickness and the weight of the mother in the study group are already comparable to other high-risk pregnancies and might indicate cardiovascular risks for the mother. However these preliminary results have to be interpreted with caution as further investigations are needed.

P301
Cardiovascular risk clustering and vascular phenotype associations in mothers and six-year-old children

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Introduction: Cardiovascular risk factors tend to cluster and their combination seems to be more predictive for atherosclerosis development and progression. Parental cardiovascular disease increases the risk of cardiovascular disease in the offspring long term. Genetics and family-shared lifestyle contribute, but the magnitude of their influence on child vascular structure and function in early childhood is unknown.
Methods: In this cross-sectional analysis of 201 mother-child pairs originating from the Finnish Gestational Diabetes Prevention Study (RADIEL) longitudinal cohort, we studied associations of child and maternal cardiovascular risk clustering, and maternal subclinical atherosclerosis with child arterial phenotype. Dyads were evaluated at child age 6.1 ± 0.5 years with ideal cardiovascular health (ICVH – set of seven health metrics: BMI, blood pressure, fasting glucose, total cholesterol, diet quality, physical activity, smoking), body composition, very-high frequency ultrasound of carotid arteries (25 and 35 MHz), carotid-femoral and carotid-radial Pulse Wave Velocity.
Results: We found no association between child and maternal ICVH, but report evidence of particular metrics correlations: total cholesterol (r=0.24, P=0.003), BMI Z-score (r=0.17, P=0.02), diastolic blood pressure Z-score (r=0.15, P=0.03), and diet quality (r=0.22, P=0.002). Child vascular phenotype was not associated with child or maternal ICVH. In the multivariable regression explanatory model adjusted for child sex, age, systolic BP, lean body mass, and body fat percentage, maternal carotid intima-media thickness (IMT) was the only independent predictor of child carotid IMT (adjusted R (2)=0.08). Child carotid IMT increased by 0.1 mm (95% CI 0.05, 0.21, P=0.001) for each 1 mm increase in maternal carotid IMT. Children of mothers with subclinical atherosclerosis had decreased carotid artery distensibility (1.1 ± 0.2 vs 1.2 ± 0.2 %/10 mmHg, P=0.01) and trend toward increased carotid IMT (0.37 ± 0.04 vs 0.35 ± 0.04 mm, P=0.06).
Conclusions: ICVH metrics are heterogeneously associated in mother-child pairs in early childhood. Maternal carotid IMT predicts child carotid IMT, but underlying mechanisms remain unclear. Maternal subclinical atherosclerosis is associated with local carotid arterial stiffness in early childhood.

P302
CHiC-D- Cardiovascular Health in Children with Type 1 Diabetes- early detection, cardiovascular prevention and treatment monitoring

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Introduction: CVD mortality in adulthood is still more than 4 times higher among people with T1D than in the general population. The CHIC-D study is an ongoing project that aims to determine the time course of vascular changes in children with T1D and the impact of the degree of metabolic control and blood pressure on changes in the different layers of the arterial wall. By using Ultra-high frequency ultrasound (UHFUS) that enables visualization of the different layers of the arterial wall combined with measurements of vascular elasticity, 24h blood pressure and assessment of metabolic and inflammatory profile our objective is to establish a novel, highly sensitive, clinically applicable method of cardiovascular risk evaluation and treatment monitoring for paediatric patients with T1D.
Methods: Children (6-15,99yr) with T1D duration of ≥5 years living in the Gothenburg region have been randomly selected from the national pediatric diabetes registry SWEDIABKIDS, and asked to participate in the study. UHFUS on the radial, dorsal pedal and carotid arteries combined with measurements of vascular elasticity, endothelial function and 24h ambulatory blood pressure (ABP) as well as metabolic and inflammatory profile have been collected in 27 patients and 16 controls so far.
Results: The preliminary results show an increased media thickness and intima- media thickness (IMT) in the carotid artery among the children with T1D as compared to healthy controls. The children

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with T1D also show increased intima thickness, media thickness and IMT in the dorsal pedal artery. Differences in blood pressure are already present between our study groups with a tendency towards higher total and daytime diastolic blood pressure and a significantly higher nighttime diastolic blood pressure in children with T1D.

Conclusions: Increase of the wall thickness that predominantly involve the medial layers of the peripheral arteries in children with T1D already 8 years after diagnosis may be important markers for early vascular changes. Interestingly, as a measure of vascular function, only nighttime diastolic ABP is affected, and not PWV. Modifiable factors associated with these changes are yet to be discovered in this study. Using our sensitive methods, different treatment strategies may be tested in future studies.

Table 1: Demographic data and results from vascular measurements.

	T1D patients n=27	Healthy controls n=16	p- value
Age (years)	12.3 ± 2.0	10.8 ± 2.8	0.05
Weight (kg)	49.2 ± 13.7	42.7 ± 15.2	0.138
Height (cm)	157± 14	150 ± 20	0.137
T1D duration (years)	8.02 ± 1.56	-	
HbA1c (IFCC mmol/mol)	48 ± 6.7	30 ± 2.0	<0.01
HbA1c (DCCT %)	6.6 ± 0.6	4.9 ± 0.2	<0.01
SBP (mmHg)	106 ±7	104 ± 8	0.35
DBP (mmHg)	66 ± 4	63 ± 5	0.04
Total 24h SBP	112 ± 9	109 ± 7	0.19
Total 24h DBP	67 ± 6	64 ± 3	0.08
Daytime SBP	116± 9	114 ± 7	0.52
Daytime DBP	71± 6	69 ± 4	0.27
Night time SBP	103 ± 10	97 ± 7	0.08
Night time DBP	57 ± 5	54 ± 2	0.02
Brachial PWV (m/s)	7.3 ± 0.7	7.3± 0.9	0.94
Aortic pulse wave velocity (m/s)	5.2 ± 0.7	5.1 ± 0.7	0.57
UHFUS measurements	n=28	n=15	
Carotid diameter (mm)	5.84 ± 0.43	5.87 ± 0.47	0.87
Carotid intima (mm)	0.11 ± 0.02	0.11 ± 0.02	0.75
Carotid media (mm)	0.21 ± 0.05	0.17 ± 0.05	0.02
cIMT (mm)	0.32 ± 0.06	0.28 ± 0.06	0.03
Radial diameter (mm)	1.75 ± 0.29	1.70 ± 0.30	0.60
Radial intima (mm)	0.06 ± 0.01	0.06 ± 0.01	0.47
Radial media (mm)	0.07 ± 0.03	0.06 ± 0.02	0.38
rIMT (mm)	0.13 ± 0.03	0.12 ± 0.02	0.28
Dorsal pedal diameter (mm)	1.44 ± 0.43	1.23 ± 0.41	0.12
Dorsal pedal intima (mm)	0.07 ± 0.01	0.06 ± 0.01	0.04
Dorsal pedal media (mm)	0.09 ± 0.03	0.07 ± 0.02	0.01
dIMT (mm)	0.16 ± 0.030	0.13 ± 0.02	0.001

Values presented as mean ± SD. Abbreviations: SBP – Systolic blood pressure, DBP – Diastolic blood pressure, UHFUS - Ultra high frequency ultrasound, IMT –intima-media thickness, PVW- pulse wave velocity.

P303

Children with congenital heart defects or innocent murmurs are less active from infancy to school age, regardless of defect severity or treatment

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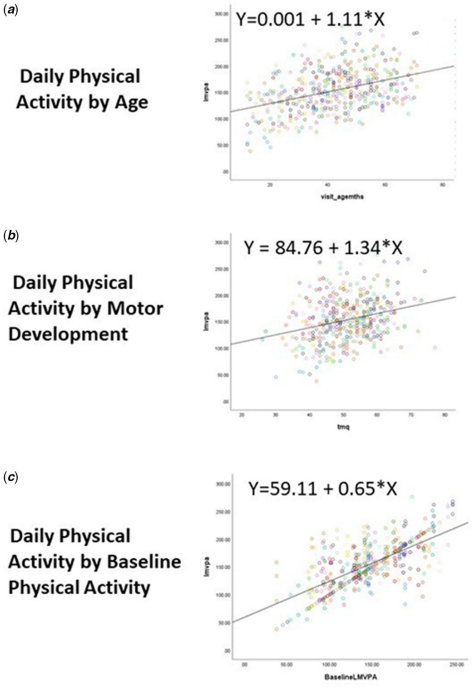
Introduction: Sedentary lifestyles and motor skill delays are common among children with congenital heart defects. Identifying the early childhood physical activity trajectory would enhance intervention timing and effectiveness.

Methods: Participants (n=158, 87 male) were recruited at 12-53 months of age. They had an innocent heart murmur (n=30), CHD with insignificant hemodynamics not requiring treatment (n=49), CHD treated without cardiopulmonary bypass (n=31), or CHD treated surgically with bypass (n=48). Physical activity (7-day accelerometry, n=103) and motor development (Peabody Developmental Motor Scales-2, n=148) measures were completed every 8 months until 72 months of age. Length of follow up was 3 (mean) to 4 (median) visits. Mixed effect linear regression models adjusted for repeated measures described physical activity trajectories.

Results: Motor development was age appropriate (Peabody 50.3 ±8.0) throughout study participation but children had primarily inactive lifestyles (mean daily minutes of physical activity=150 ±36). Only 5/133 participants achieved activity levels comparable to healthy peers. Motor development was significantly related to daily physical activity ($F=14.61$, $p<0.001$, 95%CI of estimate: 0.46, 1.45) in a simple model adjusted for study site and season of assessment (see Figure). Age significantly improved (chi-square=52.5) the model, with both age (95%CI of estimate: 0.70-1.18) and motor development (95%CI of estimate: 0.08-1.01) increasing with daily activity. Model fit again improved (chi-square=314.0) when both baseline physical activity and age were added to the model, but only baseline physical activity (95%CI of estimate: 0.91-0.97) remained significantly related to daily physical activity. CHD treatment group (chi-square=1.39) and gender (Chi-square=0.66) did not improve model fit.

Conclusions: Children with simple or complex CHD or innocent heart murmurs had low physical activity levels despite age-appropriate motor skills. Motor development, gender, age and cardiac treatment group did not impact physical activity trajectory. Results suggest that daily physical activity measured as early as 12 months of age is the strongest predictor of children's activity throughout early childhood. Effective interventions to enhance physical activity among young children with innocent heart murmurs and simple or complex CHD must be a high priority.

Figure 1: Association of Daily Physical Activity with Age, Motor Development and Baseline Physical Activity



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P304

Competitive sport in the young. Epidemiological report from the Sports Medicine Center of a major pediatric reference hospital in Italy

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Bambino Gesù Children Hospital IRCCS (1)

Introduction: Healthy young people as well as young with medical problems, refer to the Sports Medicine Center of our Hospital for a pre-participation screening as mandated by law for competitive sport in Italy. Aim of this study is to describe the cases evaluated in a 6-month period of time
Methods: We retrospectively analyzed the medical reports of athletes who were deemed fit for competitive sports in our department in the first half of 2019.
Results: 448 athletes (32.6% female, age 7-18 years), were deemed fit for competitive sport in the study period. In 192 (42.9%) medical problems emerged requiring further evaluations: 51 (26.6%, 11.4% of the total) had non-cardiovascular problems, 141 (73.4%, 31.5% of the total) showed problems of cardiovascular origin. In the non-cardiovascular group, the following conditions were found: 7(13.7%) previous hematologic malignancies; 6(11.8%) cases of type 1 diabetes; 6(11.8%) genetics syndromes; 5(9.8%) neuropsychiatric disorders; 5 cases (9.8%) of obesity; 4(7.8%) of asthma; 4(7.8%) coagulative disorders; 4(7.8%) kidney problems; 3(5.9%) cases of dyslipidemia; 3(5.9%) orthopedic problems; (3.9%) portal cavernomas; 2(3.9%) cases of cystic fibrosis. In the cardiovascular group, the following problems were found: 22(15.6%) tachy-arrhythmias; 19(13.5%) interatrial septum defects, surgically (5) or device (14) corrected; 14(9.9%) interventricular septum defects, surgically corrected (7) or in natural history (7%); 12(8.5%) cases of patent foramen ovale; 11(7.8%) mitral (mainly prolapse) and 11(7.8%) aortic (mainly bicuspid) valve abnormalities; 9(6.4%) pre-excitations; 8(5.7%) corrected abnormal pulmonary venous returns; 7(5.0%) left ventricular hypertrabeculations; 6(4.3%) pulmonary valve abnormalities; 4(2.8%) cases of hypertension; 3(2.1%) brady-arrhythmias; 3(2.1%) tricuspid valve dysplasias; 2(1.4%) cases of corrected patent ductus arteriosus; 2(1.4%) coronary fistulas; 2(1.4%) corrected pulmonary artery hypoplasias. All the remaining accounted for 1(0.7%) complex condition, surgically corrected: atrioventricular canal, aortic coarctation, great arteries transposition, tetralogy of Fallot. None of the subjects with cardiovascular abnormalities referred symptoms at rest and during exercise in the year before evaluation.
Conclusions: A large number of young subjects mainly with cardiovascular but also other medical problems have currently access to competitive sports.

P305

Enabling clinicians to translate healthy lifestyle knowledge to children with complex heart problems and their parents

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Introduction: Children with complex heart problems (CHP) are at increased risk of secondary morbidities making education about healthy lifestyles an important part of care. Children, parents and clinicians have identified the need for additional healthy, active lifestyle counseling resources.
Methods: Patients, parents, clinicians and researchers collaboratively developed 9 new knowledge-to-action tools to counsel healthy active lifestyles among children with CHP (see content information in Table). Initially, 7 paediatric cardiology professionals (2 physicians; 1 mental health professional; 3 nurses, 1 kinesiologist), 5 patient/family representatives (1 teen, 1 adult, 3 parents) and 5 recreation professionals met to review current practices and resources. Through facilitated discussions, solutions for healthy lifestyle barriers, desired resources and supports, and how needs may differ for children with different types of heart problems were identified.
Results: The new knowledge tools were distributed to 39 families of children with CHP, 3 to 17 years of age, during their clinic visit. A researcher, blind to resource distribution, interviewed each family by telephone to explore perceptions of resource benefits. Inductive thematic analysis was used to summarize feedback from 39 parents and 8 children. Families found the new resources helpful as they clarified information specific to them/their child. Most families reporting relying on the internet, family/friends, schools, and clinicians as sources of healthy lifestyle information. The cardiac clinic as an information hub for each child was important to families, who thought that general information may not apply. Participants provided suggestions to improve the content and delivery of the resources, and the need to share the information in different settings – schools, community, sports.
Conclusions: A collaborative approach developed new healthy lifestyle resources for paediatric cardiology clinicians, children with CHP and their families. Access to a variety of resources helped families to clarify information specific to their child. Families felt disseminating information through the cardiac clinic would be a simple, low-cost intervention to improve their understanding, awareness, and ability to participate in healthy active lifestyles. Future research is required to evaluate the effectiveness of these resources when used in a variety of clinical settings as a cost-effective method of increasing healthy active lifestyle awareness/knowledge among children with CHP.

Table: Description of Healthy Lifestyle Resources

Type of Resource	Description
Handout	*Title: "Be strong, Be active, Be healthy"* Information about the importance of physical activity. Different types of activity (dynamic vs static) and activity intensities that are appropriate can be specified for each child's heart condition. Distinguishing recreational activities from competitive sport and tips for participating in physical activity were also provided.
Handout	*Title: "Body Contact Restrictions"* How to be active when body contact restrictions are required and tips for helping the child cope with the required body contact limitations. The types of activities that are recommended can be specified for each child.
Handout	*Title: "Understanding Your Child's Exercise Test Results"* General information about what the child does during a cardiac exercise test, why exercise testing is used, how exercise strengthens the heart, and space to provide the specific exercise test results for the child.
Web page	*Title: "Emotions and Learning among Children with Heart Problems"* Web page on the Canadian Congenital Heart Alliance website (https://www.cchaforlife.org/emotions-learning-physical-activity-among-children-seen-heart-clinics) summarizing information regarding learning, emotional, and physical activity concerns among children with CHP.
Web page	*Title: "Network Enabling Options for a Website"* List of websites that provide support group resources. *"Abilities Online"* (http://www.abilityonline.org/public/ability-online) – online support community that welcomes kids, teens, and young adults. Provides games, homework help, other resource links, and age specific forums for conversation. To register, applicants are screened with a phone call for safety and confidentiality reasons. *"PD Heart"* (http://tchin.org/support/pdheart.htm) – an online support group for parents, family members, and adults with congenital heart disease. Registration is required to access the forums. *"Baby Center"* (https://community.babycenter.com/groups/a5055/babies_and_children_with_heart_problems) – forum about babies and children with congenital heart problems. Provides a forum for parents to ask and answer questions regarding CHD. The forum is publicly accessible. *"Yahoo Support Group"* (http://health.groups.yahoo.com/group/congenitalheartdefectsupport/) – traditional online support group sponsored by Kids with Heart National Association for Children's Heart Disorders. Members can join after being approved by a moderator.
Survey	*Title: "Healthy Lifestyle Questionnaire"* Brief questionnaire to assess the child's healthy active lifestyle need for support during the cardiac clinic visit. Multiple choice responses to each question were coded to enable healthcare professionals to quickly determine the need for additional counselling or resources.
Brochure	*Title: "Fun Physical Activities for Cardiac Families"* Brochure about the "Fun Physical Activities for Cardiac Families" event. It included an introduction to the event, the event schedule, and physical activity and discussion choices. The event focused on introducing families to a variety of physical activities suitable to their children's complex heart problems, education about physical activity, and an opportunity to connect with other parents/children with complex heart problems.
In clinic posters	Three posters suitable for display in cardiac clinic waiting and examination rooms. The posters were designed to encourage parents and children to ask their physician about what level and types of physical activity are appropriate.
Registration sheet	*Title: "Cardiac Clinic Appointments"* Cardiac clinic appointment reminder page for parents. The reminder page included a section where parents could record any questions that they would like to address during their next appointment.

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P306

Endocarditis prophylaxis in the "real life" of the general pediatrician and/or dentist

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Introduction: Endocarditis is a potentially life-threatening disease in children with CHD and correct prophylaxis (EP) is of utmost importance. EP guidelines are well known by pediatric cardiologists. However, the knowledge and correct application of these guidelines by other stakeholders of children's health is less certain.
Methods: We conducted a survey among pediatricians and dentists in Flanders, Belgium. The survey consisted of a part assessing the knowledge about EP guidelines and included a few test cases. The survey was completed by 910 dentists (16.2% response rate for 5604 dentists), 100 pediatricians (17.5%) and 16 congenital cardiologists (59%).
Results: 65% of the dentists did not know any guideline. The majority of dentists would look for information on the internet or from the child's physician. In the pediatrician's group 47% did not know any of the guidelines and the majority of pediatricians would contact the child's pediatric cardiologist. In the dentist group we focused primarily on the knowledge of high and low risk treatments and the identification of patients at risk. 87% identified correctly low risk treatments, but only 64% identified correctly all high risk procedures. Of the dentists knowing EP guidelines, 83% asked for the presence of CHD and allergy to antibiotics. Pediatricians correctly defined most defects at high risk, but scored lower for the correct identification of lower risk CHD. Most pediatricians were able to identify high risk procedures, but failed to identify many procedures correctly as low risk. Pediatricians were well aware of dental hygiene measures. The practical cases were different for dentists and pediatricians. Dentists asked more advice of the patient's physician, but would wrongly withhold treatments in high risk patients (29%). Pediatricians had difficulties with the identification of low risk procedures and the correct classification of valvar diseases.
Conclusions: The knowledge of Flemish dentists and pediatricians of the EP guidelines is low. Too many children receive unnecessary antibiotics, and some children are unduly deprived of necessary dental procedures. EP guidelines should not only be spread by the cardiology scientific publications, but also by the dentistry and pediatric scientific societies and journals in order to improve their knowledge of the EP guidelines.

P307

Evaluation of MIS-C Patients By Means of Laboratory, Electrocardiography, and Transtorasic Echocardiography

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Introduction: MIS-C is a rare complication of COVID-19 in children. The estimated incidence of laboratory-confirmed SARS-CoV-2 infection in individuals <21 years old was 322 per 100,000 and the incidence of MIS-C was 2 per 100,000. The initial reports of MIS-C emerged from the United Kingdom in April 2020. Since then, there have been reports of similarly affected children in other parts of the world, including Europe, Canada, the United States, and South Africa. While many children with MIS-C meet criteria for complete or incomplete Kawasaki disease (KD), the epidemiology differs from that of classic KD. This report is about the MIS-C children seen at a tertiary center in İstanbul-Turkey. We aimed to determine prognosis by means of cardiovascular and arrhythmia assessment.
Methods: We evaluated thirty-five MIS-C children who were hospitalized with COVID-19 infections through laboratory findings, electrocardiography (ECG), and transthoracic echocardiography (TTE) (March-December 2020). Patients were evaluated by the diagnostic criterias of both CDC and WHO. The differences were noted. On ECG, the risk of arrhythmia was observed by the evaluation of depolarization and repolarization parameters (such as Tp-Te interval, Tp-Te/QTc, Tp-Te/QT ratio) and by TTE, cardiac ventricle systolic and diastolic functions employing ejection fraction, fractional shortening, and Doppler parameters were studied. All coronary arteries were evaluated with TTE.
Results: Mean age of the patients were 15.5±5.7 years (23 boys, 12 girls). We observed significantly increased levels fibrinogen, D-Dimer, LDH, ferritin, and IL-6 levels. There was significant changes by means of TpTe, Tp-Te/QT, and Tp-Te/QTc on ECG.19 patients had no coronary artery involvement, while 16 had, and three of them also had thrombosis at the time of prognosis. Patients were treated with IVIG, Metilprednisolon, Pulse steroid, asetylsalicylic acid(anti-inflammatory and anti-thrombotic doses), clexane, and heparinization that depends on the course of the disease. Ten patients had inotropic support.
Conclusions: The thought that children are less affected by COVID-19 may be a misconception after the diagnosis of MIS-C. There are a lot of unknowns about COVID-19 and MIS-C. Therefore, one should be alert while evaluating a child with COVID-19 infection, because of sudden clinical detoriation of MIS-C patients.

P308

Health and activity status of children with congenital heart diseases and other chronic diseases after taking part in a special sports program.

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Introduction: The current WHO-led study shows that there is insufficient physical activity among adolescents worldwide. Physical activity is known as an important part of daily routine in healthy youth population as well as in children with chronic diseases. An active lifestyle supports the motor, psycho-social and individual development of children. Previous investigation describe that frequent physical exercise can improve cardiovascular health parameters. Therefore, KidsTUMove was developed as a holistic inclusive program, which counteracts and prevents the general increase of lack of movement plus giving impulses for healthy way of living. The aim of this study was to analyse the health and activity status of children with chronic diseases taking part in a special sports program.
Methods: To evaluate the effects of the KidsTUMove program regarding health and activity status, all former and current participants (2007-2018) were contacted to complete a self-reported online survey. Items were physical activity behavior (KIGGS), use of media (KIGGS), nutrition behavior (KIGGS), health related quality of life (KINDL®), self-efficacy (SWK) and individual

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lifestyle changes due to the program. Results were compared with data of KIGGS study.
Results: 49 children (age 12.9 ± 4.5 years, response rate 64.47%) attended the survey. 80.9% stated that KidsTUMove has a positive influence in daily life. Reasons for participation: meeting friends (49%), new impulses for exercise (28.6%), joint sports activities (8.2%). Pursued aims: motivation for healthy nutrition (28.6%), healthy everyday life (20.4%), physical exercise (34.7%). 70.2% children were active at least 1 -2 times per week (KIGGS: 31.9%); 85.1% were members in a sportsclub (KIGGS: 60%). Quality of life (total score): 72.57% (KIGGS: 75.1%). Nutrition behaviour: 42.6% KidsTUMove participants ate daily fruits (KIGGS: 29.4%♂/30.4%♀); 48.9% daily vegetables (KIGGS: 17.7%♂/22.6%♀). Sweets were eaten by 42.6% 1-2 times per week (KIGGS: 57.6%♂/58 %♀). Fast food was not consumed by 85.1% (KIGGS: 9.2%♂ /15.4 %♀). Sweet drinks were never consumed by 70.2% (KIGGS: 8.4%♂ /10.8 %♀).
Conclusions: Children taking part in specially designed health programs accordingly to the needs of their target group seem to benefit in their daily life. A planned study will concentrate on the topic: Daily life of children with chronic diseases all over Europe.

P309

How good is the athletic Fontan? Cardiopulmonary exercise capacity in Fontan patients with athletic background in comparison to those with none

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Introduction: In univentricular patients after Fontan procedure the exercise tolerance is poorer compared to healthy peers. Insufficient preload and incomplete ventricular relaxation during exercise result in a limited increase of stroke volume and impaired heart rate response. Thus, the peak VO_2 in Fontans is significantly lower compared to normal values. Higher peak VO_2 represents a good predictor for mortality and morbidity in Fontan patients. Peak VO_2declines with age, representing a stronger negative predictor with a lower starting value. Cardiac rehabilitation programs promote exercise tolerance in Fontans. A higher physical activity level beginning at a young age would lead to a higher exercise tolerance.
Methods: Cardiopulmonary exercise testing as an incremental step test on a treadmill was performed by 14 Fontan patients with a history of sports activity in their youth or childhood with >/= 3h/week and 9 Fontan patients with no physical activity. For the incremental running test we used a protocol starting at 4 km/h with (boys), 3 km/h (girls), and 1% inclination, with an increase of 2 km/h every 2 min near exertion 1 km/h. A general questionnaire included medical history and sports participation since childhood.
Results: We included 23 Fontans (mean age 21,1 years, 16 males, 7 females, 14 left and 9 right systemic ventricle). Questionnaires revealed sport activities until maturity in 14 patients (sport during childhood 4,9h/week, during adolescence 5,4h/week, or continuously <3h/week). All subjects achieved maximal exertion with comparable RER of 1,08 in both groups and comparable lactate values. The mean parameters measured at peak exercisediffered significantly between both groups (mean Peak VO_2= 36,5 vs 26,2 ml/min/kg; mean Peak HR 176/min vs 144/min).
Conclusions: PeakVO_2, HR-variability and O_2-pulse as indicator of cardiac output were significantly higher in Fontan patients who have been physically active while growing up than among those who have been continuously inactive. In comparison to CPET values in Fontan patients in other studies, the patients in this study achieved much higher values. As a higher peakVO_2 is associated with a better long-term survival, this study suggests that physical activity at an early age could improve peakVO_2 and in a consequence in the long-term outcome of Fontan patients.

P310

Identifying criteria for a physical literacy screening task: an expert delphi process

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Introduction: REACH (Recreation, Education, Allied-health, Coaching, Healthcare) leaders provide critical support for children's physical literacy. This project established screening criteria to identify children needing significant support for a healthy, active lifestyle.
Methods: A 3-round expert Delphi process sought consensus regarding physical literacy screening. Group discussions (Round 1) identified screening issues. Qualitative analyses of meeting notes by two researchers represented the issues as statements. Experts rated each statement (5-point Likert scale) in Rounds 2 and 3. Experts were aware of the mean Round 2 rating for each statement during Round 3. A priori consensus was 75% of participants stating agree/strongly agree.
Results: 53 experts were invited to participate with 37 (63% female, mean career length = 16 years) providing consent. Each round comprised at least 7 experts with primary/secondary expertise for each REACH sector. Round 1 identified 60 criteria and 27 potential screening tasks, which were represented in 90 statements. Consensus was achieved for 44/90 statements in Round 2 and 51/90 statements in Round 3. The tasks should be suitable for research and practice, with individuals or groups of children, and including those with disabilities. The assessment of physical activity and sedentary behavior, motor skill, cardiorespiratory fitness and activity motivation are important. Providing results useful to REACH leaders and a decision tree for further follow up are recommended.
Conclusions: A physical literacy screening tool would enable leaders in recreation, education, allied health, coaching, and healthcare to identify children with low physical literacy. Expert consensus suggests the screening should use objectively measured tasks and questionnaires encompassing multiple facets of physical literacy, including motor competence, motivation, strength, endurance, and daily behavior. Research is required to identify potential tasks that meet these criteria and are suitable for each REACH sector.

P311

Impact of COVID-19 disease on clinical research in paediatric and congenital cardiology

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Introduction: COVID-19 triggered an unprecedented crisis affecting our society at every level. Research in paediatric and congenital cardiology is currently in full development and may have been disrupted. The aim of this study is to determine the impact of COVID-19 in paediatric and congenital cardiology clinical research and to analyse decision-making and adaptation processes, from a panel of on-going academic and industry-sponsored research at the time of the pandemic.
Methods: This observational study was carried out in April (2020), from a CHD clinical research network involving five tertiary care paediatric and congenital cardiology centres. Investigators and clinical research assistants from each participating research centre filled in an online survey questionnaire, and each principal investigator underwent a one-hour web-based videoconference interview.
Results: A total of 34 study questionnaires were collected, reporting that 18 studies were totally suspended. Upon investigator's decision, after discussion on ethical issues and with facilitating support from health authorities, 16 studies were resumed. The rate of study suspension in interventional research (53%) was similar to non-interventional research (56%). Logistical problems were predominantly reported in both continued and suspended trials. Research protocols were adapted, largely thanks to telemedicine, which in some cases even improved the course of the study.
Conclusions: The impact of the Covid-19 pandemic on clinical research in paediatric and congenital cardiology has been limited by a rapid adaptation of all research structures and an extensive use of telemedicine at all stages of the studies.

P312

Impairments in cardiovascular health and reduced physical activity early after treatment for pediatric cancer

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Introduction: Cardiovascular diseases are well known late sequelae in pediatric cancer survivors. Cardiovascular health is usually related to physical activity. Increasing evidence suggests the beneficial effect of regular physical activity on overall health in pediatric cancer survivors. However, a large proportion of this group is physically inactive. Data regarding cardiovascular aspects in combination with physical activity levels is rare in pediatric cancer patients shortly after treatment. Therefore, this study aimed at investigating non-invasive cardiovascular parameters and physical activity levels early after cessation of treatment.
Methods: Participants between 6 and 18 years of age with mixed types of cancer were recruited during follow-up care from April to June 2019. Peripheral and central systolic/diastolic blood pressure (pSBP/cSBP/pDBP/) and pulse wave velocity (PWV) were assessed using the Mobil-O-Graph® to evaluate aspects of cardiovascular health. Physical activity levels were quantified with the questionnaire of the KiGGS study (German Health Interview and Examination Survey for Children and Adolescents). All data was compared to age- and gender-matched reference values of healthy children and adolescents.
Results: In total, 40 children and adolescents (11.3±3.8 years of age, 50% ♀) were recruited within 1.6±1.8 years post-treatment. PSBP (z-score 0.9±1.7, p=0.003), pDBP (z-score 0.8±1.9, p=0.033) as well as cSBP (≥8 years: z-score 0.6±1.3, p=0.011) were significantly increased compared to reference values. PWV was elevated (<8 years: z-score 1.2±2.9, p=0.374; ≥8 years: z-score 0.6±1.9, p=0.127). Less than 20% of the participants reported 60 minutes of daily moderate-to-vigorous physical activity as recommended for children and adolescents by the World Health Organization. Half of the participants were active sports club members before diagnosis, but one third did not resume their former membership after cessation of treatment.
Conclusions: Early after treatment, children and adolescents show increased blood pressure and an elevated PWV, both risk factors for cardiovascular diseases. These findings combined with low levels of self-reported physical activity in the studied group underline the support needed regarding engagement in physical activity to potentially counteract risk factors and improve cardiovascular health. Implementation of sports offers already during and following treatment should be considered as a meaningful, cost-effective preventive approach in terms of late effects associated with physical inactivity.

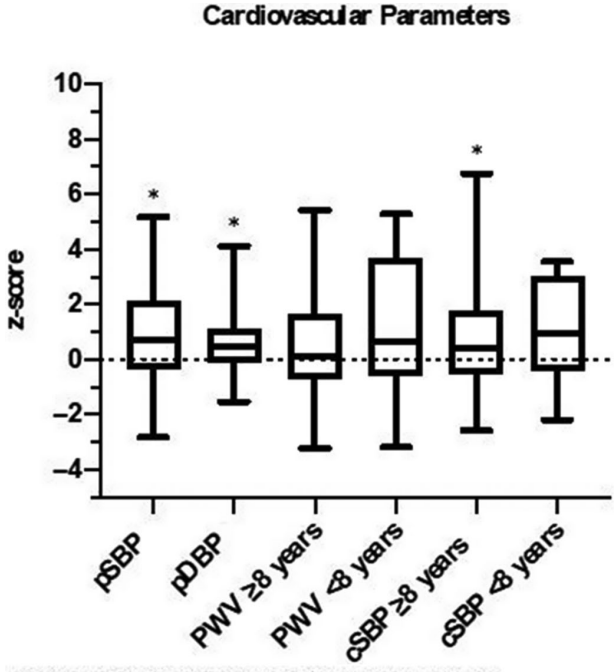

Figure 1. Cardiovascular parameters shown in z-scores. pSBP (peripheral systolic blood pressure), pDBP (peripheral diastolic blood pressure), PWV (pulse wave velocity), cSBP (central systolic blood pressure); *significant values ($p \leq 0.05$)

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P313

Incidence of Metabolic Syndrome: Accelerating atherosclerosis index, underlining late cardiotoxicity in pediatric survivors of childhood malignancies

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Introduction: Childhood cancer survivors have increased risk of dyslipidaemia, insulin resistance (IR), obesity and hypertension. American Heart Association and the Council for atheromatic Cardiovascular Disease (ACVD) in the Young, have declared the post cancer treatment survivors as a population with increased risk for ACVD in early adult life. The aim of the study is to measure the incidence of metabolic syndrome (MS) among survivors of different common childhood malignancies proving that they are in a mode of accelerating atherosclerosis.
Methods: A double blinded study examined 105 survivors from childhood malignancies. Males:57 Females:48, age range: 08-36 years, mean age 17.7 years old. Three age groups were studied: Children: 24, Adolescents: 47, adults 35. They suffered initially from leukaemia's (44), CNS tumours: (25), lymphomas (16), neuroblastomas (12), miscellaneous (8). Duration from completion of treatment: 5,5 years to 25 years. Mean years of completion treatment: 7 years and 8 months. An independent team calculated their MS incidence based on criteria by the 2007 consensus group of the International Diabetes Foundation. Then blinded to existing findings intima medial thickness (IMT) of both carotid arteries followed the proposed by the Association of European Pediatric and Congenital Cardiology (AEPC) guidelines, was calculated as a surrogate index of atherosclerosis.
Results: We calculated in all categories an incidence of MS 17.1%. in leukaemia 14.3%, in CNS 12.5%, in lymphomas 12.5%, neuroblastomas:9%, miscellaneous 8.3% survivors. By category of survivors pathological IMT found was proportional 85.6%, 83.7%, 82.5%, 81.1%,80.4%. These numbers explain the strong correlation of MS and accelerating atherosclerosis.
Conclusions: An increase incidence of MS that serves as a clustering risk factor for early atherosclerosis can be found among children, adolescents and young adults' survivors for childhood malignancies. These worrying findings, even in the childhood age group, must draw the attention to early preventive measures to avoid clinical events of ACVD.

P314

Inspiratory ventilatory training improves patients exercise capacity with repaired tetralogy of Fallot: results of a randomized and controlled trial

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Introduction: Patients with tetralogy of Fallot (ToF), the most common cyanotic congenital heart disease, have limited blood flow to the lungs before corrective surgery and ongoing dysfunction of the pulmonary valve and right ventricle leading to lower exercise capacity and impaired lung volumes in many patients throughout life. Training of deep inhalation may increase lung volumes, improve pulmonary blood flow, and consequently exercise capacity. This study examined the effects of a volume-oriented inspiratory breathing training in patients with repaired ToF on exercise capacity and lung volumes.
Methods: From February 2017 to November 2018 sixty patients (age at inclusion: 14.7 ± 4.8, 39% female) underwent spirometry (forced vital capacity, FVC; forced expiratory volume within the first second, FEV1), a cardiopulmonary exercise test (CPET: peak oxygen uptake, peak $V'O_2$) and breathing excursion measurement. They were randomized into immediate breathing exercise or a control group with training after a delay of six months. Patients were re-examined six months after randomization as well as after their training.
Results: In the first six months (intention to treat analysis) the training group increased in exercise capacity (Δpeak $V'O_2$ 0.5 ± 3.4 vs. -2.1 ± 4.7 ml/min/kg, P=0.017), FVC (Δ0.17 ± 0.17 vs 0.06 ± 0.15 L, P=0.010), and FEV1 (Δ 0.14 ± 0.17 vs -0.02 ± 0.16 L, P<0.001). Including the delayed training data from the control group (n=54), this increase in peak $V'O_2$ (Δpeak $V'O_2$ 1.2 ± 3.6ml/min/kg, P=0.017) correlated with self-reported training days per week (r=0.282, P=0.039).
Conclusions: Young patients with repaired ToF benefit from a daily inspiratory breathing training regarding exercise capacity and lung function.

P315

Is there an increase in carotid intima-media thickness in children with inflammatory bowel disease?

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Introduction: Inflammatory bowel disease (IBD) patients are suitable as a model for investigating the effects of inflammation on the vascular system for they do not carry the burden of additional cardiovascular risks (obesity, diabetes or dyslipidemia). Carotid intima-media thickness (cIMT) reflects early arterial structural changes and independently predicts cardiovascular events in asymptomatic individuals. Some of the previous studies on adult IBD patients showed increased cIMT, but not all have confirmed these results. The only pediatric study by Aloi et al. failed to demonstrate difference comparing 52 IBD patients with healthy controls.
Methods: The study included 161 children and adolescents at the age of 14.08 ±2.88 (6-18 years) - 55 with newly discovered active disease, 53 in clinical remission defined by pediatric activity indices for Crohn disease and ulcerative colitis (PCDAI/ PUCAI), and 53 healthy controls. CIMT was measured by the automatic edge tracking system (Vivid E9, General Electrics, software 112 (1.7); linear probe 11L-D). The measurements were performed 1 cm from common carotid artery bifurcation at the posterior wall in longitudinal view in the length of 1 cm. The mean of three end-diastolic measurements at each side was calculated. The

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groups were compared using ANOVA (SPSS 20.0), with the statistical significance threshold at $p<0.05$
Results: The mean cIMT in active IBD was 0.49±0,05 mm (0,38-0,66 mm), in patients in remission 0,48±0,04 (0,36-0,56 mm), and in controls 0,47±0,04 mm (0,42-0,57 mm). Although a slight increase in cIMT depending on disease activity can be observed, the difference between the groups did not reach statistical significance (p=0,373). In patients with Crohn disease cIMT was 0,48 ±0,04 (0,36-0,56 mm), and in ulcerative colitis 0,49±0,05 mm (p=0,336).
Conclusions: Increase in cIMT reflects structural changes, which require time. It seems that such changes in blood vessel walls induced by inflammation remain inapparent in the pediatric population. This emphasizes the possible protective role of timely and adequate treatment of IBD.

P316
Objectively Assessed Physical Activity Behavior is Associated with Health-Related Quality of Life in Pediatric Patients with Congenital Heart Disease

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Introduction: With increasing survival rates, a multidimensional approach to health becomes more important to improve long-term outcome in patients with congenital heart disease (CHD). Recent studies using objective measures have children with CHD shown to be almost as active as healthy peers, and the positive impact of physical activity (PA) on optimal physical and mental well-being is widely established. Furthermore, findings on whether Health-related Quality of Life (HRQoL) is reduced in children with CHD are mixed. This study aimed to determine whether there is an association between objectively assessed PA and HRQoL in children with CHD.
Methods: Within the ongoing cardiovascular screening study FOOTLOOSE (DRKS00018853) at the outpatient clinic of the German Heart Center Munich, 339 children with CHD (12.1 ± 3.4 years, 138 girls) provided PA data after a 7-day objective physical activity assessment from September 2017 to November 2020. PA was evaluated as average daily steps and moderate-to-vigorous physical activity (MVPA) minutes assessed via wearable bracelet Garmin vivofit® Jr. These children also completed the KINDL® - a 24 Likert-scaled item questionnaire assessing HRQoL in the six dimensions physical well-being, emotional well-being, self-esteem, family, friends and everyday functioning. Answers were transferred to a score ranging from 0-100.
Results: Daily Steps (r=.138, p=.010) and daily MVPA minutes (r=.124, p=.022) were both correlated to total KINDL® score. Furthermore, both steps and average daily MVPA minutes were also associated with the subcategories physical well-being (steps: r=.150 p=.005; MVPA: r=.128, p=.018), friends (steps: r=.157, p=.003; MVPA: r=.141, p=.009).
Conclusions: PA was positively associated with HRQoL in children with CHD, but further investigations need to explore the magnitude PA really has on quality of everyday life in this pediatric population. Especially CHD subgroup analysis should receive focused attention in future research.

P317
Oral changes are important for diagnostic processes in patients with Kawasaki disease presenting with faint and limited principal clinical features.

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Introduction: Timely treatment of Kawasaki disease (KD) is important for avoiding coronary artery abnormalities (CAAs). It is, however, sometimes difficult to establish a diagnosis of KD due to faint and/or limited principal clinical features. We examined the clinical features in such patients.
Methods: We retrospectively reviewed the clinical records of patients who presented with limited and faint clinical features at admission. A clinical feature that was recognizable by even junior doctors was defined as a definite feature (d-Feature) and a feature that was faint and recognizable by only experienced doctors was defined as a faint feature (f-Feature). Total features (t-Features) consisted of d- and f-Features.
Results: Among 383 KD patients, 62 patients (16%) presented with fever and ≤3 clinical features at admission and treatment of antibiotics was initiated. Of these, 82% of patients presented with fever and ≤1 d-Feature. Most patients (89%) presented with ≤2 t-Features. Among the d-Features at admission, cervical lymphadenopathy was the most common (53%), followed by oral changes (19%) and conjunctivitis (15%). Rash or extremity changes were rarely observed (3% and 2%, respectively). Most f-Features were rarely observed (2-6%), except for oral changes (18%). When the number of features increased and treatment of KD were initiated, the proportions of all d-Features were around 70%. For f-Features, oral changes were most frequently observed (23%) and the prevalence of the other features was 15-16%. Thirty-two patients presented with fever and ≥4 d-Features, whereas 30 patients presented with ≤3 d-Features. Among these 30 patients, d-Features of cervical lymphadenopathy was the most common (60%) and remaining features' d-Features were 40-43%. Among the f-Features, proportion of oral changes was the highest (43%), and the others were 23-33%. The experienced doctors decided to commence KD treatment by considering the patients' clinical courses and echocardiographic findings, and delayed treatment (> seven day of illness) were only three patients and CAA was observed in one patient.
Conclusions: During diagnostic process of KD, evaluating f-Features of oral changes are important for avoiding delayed treatments and CAAs.

P318
Parents of Very Young Children with congenital heart defects report good quality of life for their children and families regardless of defect severity

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Introduction: School-age children with congenital heart defects have been reported to have lower quality of life, particularly

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among those with complex heart defects. Little is known about the quality of life of very young children (less than 5 years of age) with heart defects or innocent murmurs, and the relationship between quality of life and defect severity or treatment. The purpose of this project was to investigate parent reports of quality of life for their very young children with congenital heart defects (CHD), and to compare their scores to published data for healthy children.
Methods: Parents of children 1-3 years old with CHD or innocent heart murmurs completed the Pediatric Quality of Life Inventory (PedsQL) core, cardiac and family impact modules. Multivariable regression analyses assessed the impact of age, sex, family income, and CHD treatment history (study group) on PedsQL scores. Correlations between family impact and core/cardiac modules were examined. PedsQL scores were compared to healthy norms.
Results: 140 parents of young children participated within four study groups: CHD no treatment (n=44), CHD treatment without bypass (n=26), CHD treatment with bypass (n=42) or innocent heart murmurs (n=28). Male sex was associated with higher core (F=4.16, p=0.04, σ2=.03) and cardiac quality of life (F=4.41, p=.04, σ2=0.04). Higher family income was associated with higher family quality of life (F=8.89, p<.01, σ2=0.13). Parents of children with innocent heart murmurs and children with CHD not requiring treatment had higher core QoL compared to young healthy children. Cardiac related quality of life scores were associated with family impact (r=0.68) and core module (r=0.63) quality of life scores.
Conclusions: Parents of very young children with CHD report good quality of life for their children and families. Quality of life exceeds that of children with innocent murmurs or CHD not requiring repair. Parents report a lower quality of life among girls, and lower family quality of life is associated with lower family income.

P319
Physical activity around the world for health prevention

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Introduction: Since obesity in children based on the lack of physical activity and unhealthy lifestyle is rapidly increasing, the AEPC working group "Sports Cardiology and Prevention" decided to create a poster with all important aspects and current recommendations at a glance. The idea was to implement the information as short as possible and as early as possible, that means information for young ages (and parents) to lay the foundation for a healthy active life.
Methods: After intense research on literature and recommendations, a first draft was shown at the AEPC 2018 in Athen during the working group session. The second overworked draft was shown during the working group meeting in Styria in November 2018. After a working period in close cooperation with a graphical designer, the final versions of the KIDS (up to 11 respectively 13 years) and the TEENS poster version could be shown at the AEPC working group session in Sevilla 2019 and has been confirmed by the whole working group as an expert committee.
Results: Different kinds of design as well as content-related issues were in discussion and ended up in the decision to include recommendations on sleep time, screen time, physical activity and suggestions on how to implement them in everyday life. The result was, due to extensive recommendations on all the included aspects and to the desire of getting all information at a glance, to split the poster into two different age groups- KIDS and TEENS. Up to date we already have English, German, Hungarian, Croatian, Swedish, Spanish, Italian, Bulgarian, Rumanian, Portugese and Finnish languages translated versions.
Conclusions: There are high demand and interest on these posters and on getting the information spread by putting them on the walls of the waiting rooms of resident doctors and at all hospitals. There are still some languages missing and there is a need to get founded to have the chance to spread the posters all over the world for a healthier lifestyle for children, teenagers and in the end even adults. It should now be spread out by international pediatric congresses.

P320
Physical activity in children and adolescents with congenital heart disease: Review from a measurement methodological perspective

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Introduction: the aim was to compile a literature overview of physical activity in children with congenital heart disease and to critically evaluate the methodology used for physical activity measurement.
Methods: A review of the literature was performed using PubMed to identify studies 2009-2020 examining accelerometer and subjectively assessed physical activity in children and adolescents with congenital heart disease.
Results: A total of 15 studies were included (six studies using subjective measures and nine articled using accelerometers for the assessment of physical activity). The patients generally failed to meet the recommendations of physical activity. When compared to healthy controls, the results were widely divergent in the subjectively assessed measures and the accelerometer-based studies showed a tendency of no difference in physical activity. Neither subjective methods nor accelerometer-based studies reported any difference in physical activity in general, due to the severity of the heart disease.
Conclusions: No consensus could be found regarding physical activity in children with congenital heart disease compared to healthy controls or due to the severity of the heart disease. These results may largely be explained by methodological differences and limitations in the assessment of physical activity.

P321
Physical Activity in Children with Congenital Heart Defects: results of a nationwide survey

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Introduction: In children with congenital heart defects (CHD), a sedentary lifestyle should be avoided and usually WHO recommendations on physical activity (PA) are supposed to be followed. Whether these recommendations are considered and implemented remained unanswered so far. In order to obtain representative data of the actual amount of PA (and potential influencing factors) in children with CHD we performed a nationwide online survey.
Methods: All patients aged 6 to 17 years registered in the German National Register for CHD were contacted by email and asked to participate in the survey using the comprehensive questionnaire of the "Motorik-Modul" from the German Health Interview and Examination Survey for Children and Adolescents (KiGGS), thus allowing the comparison with a representative age-matched subset of 3.385 participants of the KiGGs study. The questionnaire for CHD-patients was amended by specific questions regarding medical care, sports recommendations and PA restrictions.
Results: 1.198 CHD patients with available complete datasets were included for evaluation. Compared to the reference group, CHD patients significantly less frequently reached the WHO recommended level of 60 minutes of daily PA (8.8% versus 12%; $p < .001$). Enjoyment in sports was almost equally distributed across CHD and reference groups, and strongly correlated with the level of PA ($r=0.41$; $p < .001$). Remarkably, 49.2% of children with complex CHD, 31.7% with moderate, and even 13.1% with simple CHD were advised by their physician to restrict PA. Regression analysis demonstrated a significant impact of age ($p < .001$) and gender ($p =0.05$) on PA, whereas girls with CHD demonstrated lower levels of PA. Also, number of interventions ($p=0.005$) and enjoyment in sports ($p < .001$) play a relevant role on the level of sports activity. Multiple regression analysis revealed physician-recommended PA restrictions having significant impact on the amount of PA ($p=0.01$), followed by sports activity of the father ($P=0.05$).
Conclusions: According to this nationwide survey, PA is markedly reduced in children with CHD. Our study provides information on possible contributing factors that may influence the amount of PA beyond the burden of the heart defect itself, including an unexpected high rate of physician-recommended restrictions on levels of PA.

P322

Quantitative analysis of NT-pro-BNP in neonates using dried blood samples: validation in healthy term babies compared to infants with cardiac disease.

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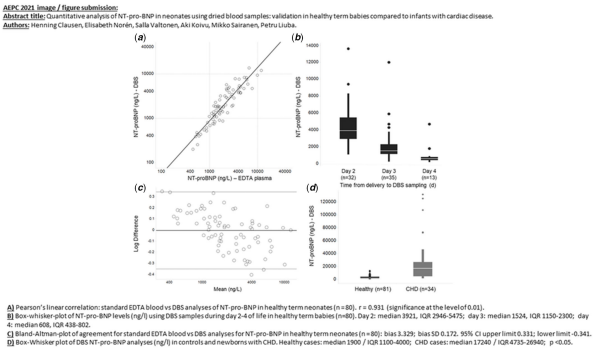
Introduction: N-terminal prohormone of brain natriuretic peptide (NT-pro-BNP) is an established biomarker in adults with heart failure and there is emerging evidence about its role in children with various types of cardiac disease. It has been used to identify adults and children with deteriorating cardiac function and we hypothesized that NT-pro-BNP may be an early marker of cardiovascular changes after birth, which may result in improved identification of neonates at risk of cardiovascular compromise or collapse.
Methods: We established a new quantitative NT-pro-BNP analysis on dried blood samples (DBS) as those universally used for neonatal screening programs. Results were compared to standard NT-pro-BNP assay analyses and predefined neonatal patient criteria reviewed. Healthy participants were prospectively followed-up for a minimal of 12 months. Controls were compared to a retrospectively identified paediatric patients with established cardiac disease (CHD) in whom stored DBS were available for analysis.
Results: Newly developed DBS assay was used to analyze NT-pro-BNP in 115 babies (healthy controls: 81, CHD cases: 34; males: 54%) on day 2-4 of life using 3.5 microliters of blood with no loss to follow-up in this single centre pilot study (2018-2020). None of the prospectively included healthy babies born 2018-2019 showed signs of cardiovascular disease postnatally. 31 CHD cases born 2003-2019 were used for comparison. We found a significant difference between diseased and healthy babies using DBS analysis of NT-pro-BNP ($p <0.05$). 13/19 (68%) of critical congenital heart disease could be identified by DBS NT-pro-BNP results alone. Detection of any CHD type improved to 81% when combining pulse oximetry and NT-pro-BNP results and to 89% for critical CHD. DBS assay performed well based on receiver operating characteristics with an area under the curve of 0.86, which improved to 0.96 when matching controls and cases born between 2018-2019.
Conclusions: NT-pro-BNP analysis using DBS is feasible in newborns using minimal blood amounts. Comparison of healthy neonates with children suffering from various types of heart disease showed significant differences of NT-pro-BNP levels soon after birth suggesting cardiovascular maladaptation may be detectable early on and future studies should explore the role of NT-pro-BNP as a potential neonatal screening marker of cardiovascular disease.



P323

Rapidly increasing left ventricular wall thickening is a risk factor for ventricular arrhythmias in pediatric hypertrophic cardiomyopathy

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Introduction: Primary prevention of sudden cardiac death (SCD) is an important issue in pediatric patients with hypertrophic

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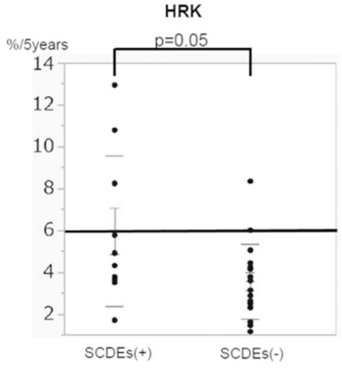
cardiomyopathy (pHCM). However, risk stratification has not yet been determined. This study focused on verifying the current risk model for pHCM and determining additional risk factors.
Methods: The subjects were 30 patients with pHCM who were under 17 years of age when they first visited our center, with a median age of 13 years (range: 1-16 years). Risk scores (HRK) were calculated using the formula published in JAMA cardiology 2019. SCD related events (SCDEs) included ventricular tachycardia (VT) and ventricular fibrillation (Vf) (n=10). The following parameters were compared between the SCDEs-positive and SCDEs-negative groups: HRK values, maximum wall thickness (mWT), maximum annual change (mWT per year), left atrial diameter index by echocardiography, left ventricular mass index (LVMI), extent of late gadolinium enhancement (LGE%) by magnetic resonance imaging, and maximum annual changes (mLVMI per year and mLGE% per year). Patient follow-up was terminated at 22 years of age. Values are expressed as mean ± standard deviation. The Wilcoxon test was used for comparison. To evaluate the prognostic ability of the risk factors, receiver operating characteristic curve analysis was performed. P <0.05 was considered statistically significant.
Results: The results of the comparison are shown in the table. As seen in the figure, HRK values did not significantly differ between SCDEs-positive and SCDEs-negative groups. However, if the number of cases is sufficient, a significant difference is likely to be observed. There was a significant difference in mWT per year between both groups, and the cut-off value for predicting VT/Vf was an increase in the wall thickening of 2.6 mm per year (sensitivity 70% and specificity 100%).
Conclusions: Perhaps HRK could be applied to patients with pHCM in the Japanese. However, additional factors are required to identify those who need implantable cardioverter-defibrillator insertion. This study suggested that a rapidly increasing left ventricular wall thickening (>2.6 mm per year) can be an additional risk factor for VT/Vf in pHCM.

Table.
Comparison between SCDEs(+) group and SCDEs(-) group in various parameters

	SCDEs(+)	SCDEs(-)	p value
HRK values (% / 5 years)	6.0±3.6	3.6±1.8	0.05
mWT (mm)	18.7±9.7	12.7±4.0	0.17
LADI (mm / m^2)	26.3±12.9	25.6±9.2	0.88
LVMI (g / m^2)	91.4±33.3	78.4±23.2	0.27
LGE% (% of LVM)	9.8±6.0	5.9±3.0	0.11
mWT / year (mm / year)	**3.9±2.9**	**0.5±0.7**	**0.0004**
mLVMI / year (g / m^2 / year)	18.8±5.6	-1.2±6.2	N/A
mLGE% / year (% of LVM / year)	2.8±1.1	0.7±0.5	N/A

Abbreviation: HRK, HCM Risk-Kids; mWT, maximum wall thickness; LADI, left atrial diameter index; LVMI, left ventricular mass index; LGE, late gadolinium enhancement; LVM, left ventricular mass; SCDEs, sudden cardiac death related events
N/A indicates that analysis is not possible due to insufficient number of cases.

Figure.
Comparison between SCDEs(+) group and SCDEs(-) group in HRK values



Abbreviation: HRK, HCM Risk-Kids; SCDEs, sudden cardiac death related events

P324

Reduced spirometric volumes in children with a right heart defect – are they all restrictive?

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Introduction: Children and adolescents with a right heart defect (RHD) often have a reduced forced vital capacity (FVC) and forced expiratory volume within the first second (FEV1). Total lung capacity (TLC) is not routinely tested, despite this testing may detect reasons for a supposed restrictiction – or even an over-inflation of the patients. This study evaluated lung volumes in children and adolescents with Tetralogy of Fallot, Ebstein's anomaly, pulmonary regurgitation, or pulmonary stenosis by body plethysmography.
Methods: In total, 42 children with a RHD and a mean age of 12.9 ± 2.7 years (31.0% female) were tested with a spirometry and body plethysmography (31.0% female, April 2018 to August 2020). Reduced patterns were defined as z-scores < -1.64 in FEV1/FVC, FVC and TLC as referred to Global Lung Initiative 2012 and Stocks et al., 1995.
Results: Of all patients, 57.2% had normal results in spirometry and body plethysmography. A sole restrictive pattern (↓FVC, ↓FEV1, ~FEV1/FVC) in spirometry was seen in 15 patients (35.7%), a sole obstructive (~FVC, ↓FEV1, ↓FEV1/FVC) in three (7.1%), while none had combined patterns. Body plethysmography identified most of the restrictive pattern as true (11 of 17, additional ↓TLC) and only four as originated as obstruction with hyperinflation (~TLC).
Conclusions: Almost half of the children with RHD in our cohort show abnormalities in lung volumes. Most of these abnormalities (64.7%) express themselves in a sense of restrictive pattern, probably a small lung.

P325

Screening for end-organ damage in Fontan survivors should include routine thyroid function testing: a report from the Fontan Care Network

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Introduction: A recent guideline recommends screening for end-organ damage in Fontan patients. However, screening for thyroid dysfunction was not recommended, although thyroid disorders are common in patients with congenital heart disease. The aim of this

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study was to assess the prevalence of subclinical hypothyroidism (SHT) and to identify potential factors associated with SHT in Fontan patients.
Methods: A retrospective chart review was performed of all pediatric Fontan patients routinely followed at one of the participating institutions of the Fontan Care Network in whom thyroid function was tested between 2013 - 2019. SHT was defined as TSH levels above 4.5 mU/L, with free T4 being in the normal range. Associations between thyroid function, clinical data and laboratory results were explored. NTproBNP measures were log transferred. The relationship between the presence of SHT and covariates was analysed using binary logistic regression.
Results: Seventy-four Fontan patients (36.5% female; age 12.4 ± 7.3 years) were included. SHT was present in 16 (21.6%) patients. Patients with SHT had significantly lower systolic blood pressure (103.1 ± 11.0 vs 112.1 ± 12.5 mmHg, p = 0.011), lower weight z-score (-1.5 ± 1.9 vs -0.1 ± 1.5, p = 0.004), more frequently NYHA functional class ≥ II (62.5% vs 31.0%, p = 0.022), and protein-losing enteropathy (50% vs 22.4%, p = 0.035). Significant inverse correlations were observed between TSH levels and systolic blood pressure (r = -0.284, p = 0.014), weight z-score (r = -0.312, p = 0.008) and height z-score (r = -0.281, p = 0.016). Higher TSH (p = 0.020) and logNTproBNP (p = 0.013) levels were demonstrated in patients with moderate and severe AV valve regurgitation. Using binary logistic regression analysis, we found that NYHA functional class was a predictor of SHT (OR 8.95; 95% CI 1.18 – 67.82; p = 0.034).
Conclusions: SHT is common in Fontan patients, particularly in patients with impaired NYHA functional class. Routine screening for thyroid dysfunction in the Fontan population is therefore recommended.

P326
Spirometric patterns in children with congenital heart disease

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Introduction: Children with congenital heart disease (CHD) often have decreased lung capacity. Previous studies have demonstrated he relationship between some CHD and abnormal spirometry patterns. The aim of this study was to evaluate lung function in children with CHD and if there is any correlation between them.
Methods: Observational, descriptive and prospective study in children with CHD in a Tertiary Center of Cardiology Paediatric. Spirometry was indicated in all these patients as a part of test exercise. Only the first spirometry of each patient was tested. The study included 70 patients aged between 6 to 17 years old who have done this test from January till November 2019. Spirometry was corrected for age, gender, weight, size and race; and analyzed using percent predicted z-scores.
Results: Eighty-five spirometries were analyzed, only two were excluded because they were not valid. 52.6 % of patients were male. Only 10.14% were overweight. 53.6% had cyanosant CHD. 92.6% were complex CHD. The most common was Tetralogy of Fallot (21.7%), followed by single ventricle circulation (Fontan, 13.3%). 72.6% had undergone surgical intervention. Only 22.4% had an abnormal spirometry pattern. 20.5% patients with cyanosant CHD had a restrictive pattern versus 5.3% patients with non-cyanosant CHD (p 0.04). Highlights, 60% of patients with pulmonary atresia with ventricular septal defect (PAVSD) had a restrictive pattern (p> 0.05). The mean z-score of forced vital capacity (FVC) was reduced in 31.4% patients with cyanosant CHD (p 0.05). The analysis revealed statistically significant difference between surgeries' number and restrictive pattern (p 0.02). In addition, obstructive disease as studied. The mean z-score of FEV1/FVC in patients with cyanosant and non cyanosant CHD were not significantly different and there was also not different between obstructive pattern and bronchospasm.
Conclusions: Lung function is decreased in one third of patients wih CHD. Restrictive pattern is related to cyanosant CHD and with surgeries´ number. Respiratory physioterapy programs could be promoted both before and after cardiac surgery to minimise the impact on lung function in patients with CHD.

P327
The Congenital Cardiology Cloud – optimizing long-term care by connecting ambulatory and hospital medical attendance via telemedicine

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Introduction: Patients with complex congenital heart disease frequently undergo a life-long ambulatory therapy with the need for repeated hospital interventions. To optimize the interplay between patients, ambulatory care and hospital care we designed and implemented a telemedicine service, the Congenital Cardiology Cloud (CCC) in a regional network consisting of a University pediatric heart centre, 2 University pediatric hospitals, 14 peripheral pediatric clinics and 25 pediatric cardiology practices. This study aims at exploring the CCC´s evolution, through a comprehensive assessment of the CCC´s development and installation and impact on patient´s care and decision making.
Methods: CCC development comprises analysis of historically risen admission and discharge management, definition of technical and organizational requirements with respect to feasibility, usability and data protection, processing soft- and hardware and supervision of implementation and usage.
Results: In the CCC a large variety of different file formats from different diagnostic and archiving in clinic-programs and outpatient clinics can be easily exchanged. Automated dispatch of medical reports and workflow optimized, data-secure bidirectional exchange of diagnostical data facilitates contacts between ambulatory and hospital sector and lead to a decrease of time-consuming interdisciplinary congenital heart conferences. Implementation of

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videoconference for interdisciplinary and intersectoral case discussion as well as empowerment of patient-physician relationship due to provisioning of medical data and real-time discussion of treatment options have been initiated. Telemedicine helps to change the interplay of ambulatory and hospital care from hierarchic coexistence to cooperation and mutual patient advice. CCC implementation required willingness, perseverance and professional engagement. The need for human resources and technical investment and the lack of reimbursement are critical barriers.
Conclusions: The CCC enables sharing of complex clinical information, overcoming sectoral barriers of ambulatory and hospital care and improving mutual patient advice. Daily life penetration and mutual acceptance will have to be improved. Future application analysis and possible introduction of refinancing concepts will show its long-term feasibility.

P328

The efficacy and side effect of ibuprofen in acute rheumatic fever: The comparative results of experience with acetylsalicylic acid and ibuprofen

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Introduction: The arthritis of rheumatic fever is very responsive to treatment with acetylsalicylic acid (ASA), but there are many side effects, especially hepatotoxicity, due to ASA therapy. The efficacy of the other non-steroidal anti-inflammatory drugs, especially naproxen and tolmetin were studied before; hovewer enough reports are not available for the ibuprofen. The aim of this study was to compare the efficacy and safety of ibuprofen to ASA in the treatment of acute rheumatic fever (ARF).
Methods: Our study was performed retrospectively. We studied 38 patients with ARF who were admitted to our hospital between 2015 and 2018. All of patients had arthritis and/or mild carditis. Seventeen patients were treated with ibuprofen (30 mg/kg per day; group I) and 21 patients were treated with ASA (75-100 mg/kg per day, group II).
Results: There was no significantly difference between the groups for the sex, age and duration of the treatment ($p>0.05$). The erythrocyte sedimentation rates (ESR) and C-reactive protein levels (CRP) were high in all of the patients before the treatment, and the differences were not significant between the groups ($p>0.05$). Clinical symptoms had disappeared at the same time in both the ibuprofen and ASA groups ($p>0.05$). ESR was decreased to normal limits on 15±7 and 12±4 days after the treatment in the ibuprofen and ASA groups, respectively ($p>0.05$). At end of treatment, ESR was normal in all patients, however it was found to be lower in the ASA group than the ibuprofen group ($p<0.05$). No gastrointestinal, dermatologic or renal side effects were observed in the two groups. Liver enzyme elevations were detected in 3 and 13 patients in the ibuprofen and ASA groups, respectively ($p<0.01$). Liver enzyme elevations were higher in the ASA group than the ibuprofen group ($p<0.01$). None of the patients in the two groups developed carditis over the following 1 year period.
Conclusions: Ibuprofen appears to be effective for the treatment of rheumatic fever. It can be used particularly in patients who cannot tolerate aspirin.

P329

The impact of COVID-19 on Children and Adolescents with Pulmonary Arterial Hypertension. Data from the Polish Registry of Pulmonary Hypertension

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Introduction: COVID-19 has had a significant impact on all aspects of pulmonary arterial hypertension (PAH), from diagnosis and management to an increased risk of death in patients with PAH. Pandemic made it necessary to revisit the manner in which patients receive care to decrease risk of contracting the virus.
Methods: Eight Tertiary Paediatric Centres participating in the paediatric arm of the registry "Database of Pulmonary Hypertension in the Polish Population" (BNP-PL) took part in this multi-centre cross-sectional observational study. Eligible children were diagnosed with PAH before March 20, 2020, the day the pandemic was officially announced by the Polish government. All parents of children with PAH in the BNP-PL were invited to take part in an anonymous electronic questionnaire. It comprised of several question panels: demographic data, fear of COVID-19 and other pandemic related problems, standard anxiety scale (GAD-7), presence of alarming symptoms, which had appeared or exacerbated since March 20, 2020. Data collection was seized on the September 09, 2020.
Results: We have received 62 responses (response rate 77%), 4 were incomplete. The remaining 58 were further analysed. Ninety-five percent of the responders were women (n=55) at a median age of 41 years. Ninety-five percent of the parents were afraid of possible negative consequences of the SARS-CoV-2 on their children's health including death in 77%. Sixty to 71% of the parents were also afraid of various potential problems resulting from re-organising health-care system. As a result, 72% of parents had symptoms of anxiety as assessed by GAD-7, noticeably 34% in moderate-to-severe degree. No cases of COVID-19 were reported in the studied group of children.
Conclusions: The impact of COVID-19 on health care delivery and on society at large is going to be felt for years to come. This information is vital to provide prognostic information on cardiac health after COVID-19, to inform long-term clinical care pathways, and to guide lifestyle recommendations. COVID-19 pandemic had an overall negative impact on children with PAH and their families. Several risk factors for poor outcome were identified. Long-term

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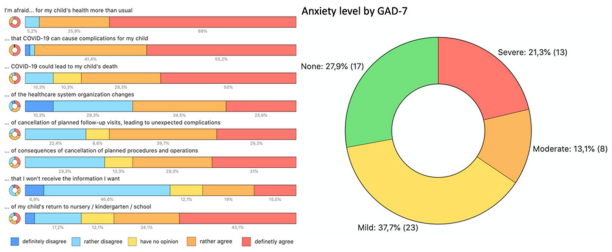
strategies should be validated and implemented to deliver quality care for children with PAH, with emphasis on psychosocial well-being



P330

Utilization of clinical criteria and laboratory investigations to identify high risk children for coronary involvement in Kawasaki disease

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Introduction: Kawasaki disease is an acute medium and small vessel vasculitis, which has a predilection for coronary arteries. Coronary artery involvement can vary from mild ectasia occurring in nearly 40% to giant coronary artery aneurysms. Hence, we sought to determine a correlation between selected clinical and laboratory criteria in order to predict high risk children with Kawasaki disease for coronary involvement in order to optimise the initial management and follow up.
Methods: Serial analysis of Clinical, biochemical and echocardiographic parameters of 101 patients with diagnosed Kawasaki disease within the first 10 days of the illness and followed them up paying special attention to coronary artery dilatation according to coronary artery standard deviation charts and visual assessment until 12 weeks of the disease for 15 months at the National Paediatric Cardiology unit, Colombo Sri Lanka. Maximum recorded internal diameter of the coronaries was considered for the analysis during the follow up. Thereafter, analysis had been performed to identify coronary artery risk predictors with Pearson chi square test and odds ratio.
Results: Mean age of the sample was 44months (±36.8 months), 62(61.4%) were males and 42(41.6%) patients had coronary involvement (Coronary artery diameter >2 SD adjusted to the Body Surface Area/ visual dilatation to Coronary artery aneurysm formation). Age between 7 months to 60 months, fever more than 102^0F, CRP more than 100 mg/L and increased coronary echogenicity within first 10 days of the illness showed significant association with coronary involvement. However, duration of fever, platelet count, AST (Aspartate aminotransferase), ALT (Alanine aminotransferase), Serum bilirubin and Serum sodium or ESR did not demonstrate statistically significant association with coronary dilatation or aneurysm formation in Sri Lankan sample, although utilised in standard risk stratification scores. Also, diagnosis of complete Kawasaki disease, hypoalbuminemia and hyper echogenic coronaries found to be important predictors of coronary aneurysms.
Conclusions: In a Sri Lankan centre, Age between 7 months to 60 months, fever more than 102^0 F, CRP more than 100mg/dl, increased coronary echogenicity, hypoalbuminemia and diagnosis of complete Kawasaki disease during early stage of Kawasaki disease demonstrated high predictive utility for the development of coronary pathology.

P331

Vitamin D, liver-related biomarkers and distribution of fat and lean mass in young patients with Fontan circulation

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Introduction: Young patients with Fontan circulation may have low serum 25-hydroxyvitamin D levels, an affected liver, and altered body compositions. This study aimed to explore vitamin D intake and serum levels, liver biomarkers, and lean and fat masses in children with Fontan circulation.
Methods: Prospective data was collected in 2017 to 2018, obtained with food-frequency questionnaires, biochemical analyses of liver biomarkers, serum 25-hydroxyvitamin D, and dual-energy X-ray absorptiometry scans in children and adolescents with Fontan circulation (n=44). Body compositions were compared to matched controls (n=38). Linear regression analyses were used to investigate associations of biomarkers, leg pain and lean mass on serum levels of 25-hydroxyvitamin D. Biomarkers were converted to z-scores and compared to reference values.
Results: Although the daily vitamin D mean intake (9.9 μg/d) was in accordance with recommendations, 42% of the Fontan population had <50 nmol/L serum 25-hydroxyvitamin D. This was not associated with fat or lean mass, leg pain, or biomarkers of liver status. The Fontan group was significantly shorter but no differences were seen between groups regarding weight, body mass index (BMI) or BMI for age z-score. Analysis of body composition showed that lean mass index in the Fontan population was lower in both arms and legs and fat mass index significant higher in arms, abdomen and in total body. Younger Fontan children within the age 6-12 years, had a significant higher abdominal fat mass index, but no difference in other body fat mass index or lean mass index, compared to controls. At the age of 13 to 18, the Fontan group showed statistically significant lower lean mass index of arm, legs and total body and higher fat mass of abdomen and total body. Male adolescents with Fontan circulation had a greater mean abdominal fat mass than male controls and higher cholesterol levels than females with Fontan circulation.
Conclusions: Vitamin D intake or serum levels were not associated with body composition or liver biomarkers in the Fontan group, but they had lower lean mass and higher fat mass than controls. The more pronounced abdominal fat mass in male adolescents with Fontan circulation might increase metabolic risks later in life.

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Characteristic	Fontan N=38	Control N=38	P value
Female: Male ratio	16:22	16:22	
Age, years	12.3 (3.9)	12.4 (4.0)	0.23
Weight, kg	46.3 (19)	47.4 (20)	0.53
Height, cm	150.1 (23)	155.7 (24)	0.003
Body Mass Index for age z-score	0.22 (1.2)	-0.13 (1.0)	0.15
Lean Mass Index, arms	1.26 (0.4)	1.36 (0.4)	0.02
Lean Mass Index, legs	4.01 (0.9)	4.4 (1.1)	0.001
Lean Mass Index, abdomen	6.30 (1.0)	6.29 (1.1)	0.91
Lean Mass Index, total body	12.90 (2.0)	13.28 (2.4)	0.15
Fat Mass Index, arms	0.65 (0.2)	0.53 (0.2)	0.03
Fat Mass Index, legs	2.05 (0.6)	1.85 (0.7)	0.18
Fat Mass Index, abdomen	2.70 (1.4)	1.68 (0.9)	<0.001
Fat Mass Index, total body	5.75 (2.2)	4.4 (1.8)	0.005

P332
What do Physical Education teachers know about their students' heart disease?

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Introduction: The advances in recent years in the diagnosis and treatment of heart disease have meant that many children with heart disease can lead an almost normal life including physical exercise. It is common that for fear of adverse events these children are restricted from practising physical activity at school. Physical Education teachers play a fundamental role in integrating children with cardiopathy into society. It is necessary that they have adequate knowledge about the state of health of their students, to know the limits for the practice of physical exercise if they had and adequate knowledge about the techniques of cardiopulmonary resuscitation. The objective of the study was to evaluate the knowledge of Physical Education teachers about the limitations or necessary adaptations in the group of patients with congenital heart disease (CHD).
Methods: Cross-sectional descriptive study carried out over a period of 2 months in which 77 Physical Education teachers participated (44.2% with a diploma in teaching, 44.2% with a degree in sports sciences, 11% with other degree), using an anonymous questionnaire with 9 questions.
Results: 76.6% claim to have specific training in cardiopulmonary resuscitation. 27 (35.1%) teachers had a student with CHD. Of these, only 53.1% had a recommendation report on physical exercise. 44.4% of these needed curricular adaptation. On a scale of 1 to 5 to evaluate the knowledge about the limitations and/or risks that these students may have in the area of physical education, 45.5% were placed in the centre, scoring 3; and on the knowledge of action protocols with these students, more than half of the responses were placed in position 2 (39%) and 3 (27.3%).
Conclusions: Basic knowledge of some clinical aspects of children with CHD is vital for their optimal Physical Education. It is therefore appropriate that teachers have both the necessary clinical information and know the characteristics of these pathologies, in order to improve their teaching practice. There is a need for strategies that favor communication and the transmission of information between health staff and teachers.

13. Nursing

P333
Between Heaven and Hell–a qualitative study on the experiences of parents with a critically ill child with extracorporeal membrane oxygenation (ECMO)

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Introduction: It is inevitably a traumatic experience for parents when their child's severe illness necessitates a period of intensive care. This situation becomes even more challenging for parents if a highly technical therapy, such as extracorporeal membrane oxygenation (ECMO) in the Pediatric Intensive Care Unit, is required.
Methods: The study examines the experiences of parents of critically ill children undergoing ECMO therapy, with the aim of better understanding their needs and identifying courses of action for healthcare professionals. The qualitative study was based on interviews with couples whose critically ill child is or has been receiving ECMO therapy during the last year, which were used to examine their experiences and perceptions. The analysis of six such interviews was performed using intensive paraphrasing and inductive coding.
Results: The parents' situation can be best characterised as 'ambivalent'. This was most clearly demonstrated by simultaneous feelings of hope and fear. The parents were unexpectedly torn from their existing family life, leading to intense feelings of helplessness with strong emotional reactions.
Conclusions: The parents' ambivalence is a central aspect, and healthcare professionals are an important point of contact who have a major influence on them. The study contributes to raising healthcare professionals' awareness of the importance of perceiving the parents' ambivalence and of adjusting their behaviour accordingly.

P334
Family feedback in relation to adapting the parent and toddler cardiomyopathy coffee mornings to a virtual platform in response to COVID-19

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Introduction: The Inherited Cardiac Conditions (ICC) clinical nurse specialist (CNS) team host coffee mornings for parents and their children who are affected by cardiomyopathy. These were held in response to parents' requests to meet other families affected by the condition. Support groups for families of children with heart disease can identify positive coping strategies and the need for psychological support (Jackson et al., 2018). Since the COVID-19 outbreak in the UK, incoming calls to the nurse specialists from families were greatly increased, due to heightened anxieties surrounding COVID-19 and its potential effects on children with cardiomyopathy. Therefore, the CNS team responded by adapting the coffee morning forum to a virtual platform with questions and answers surrounding COVID-19 and cardiomyopathy.

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Methods: The first virtual cardiomyopathy coffee morning was held on the 23rd June 2020 via Microsoft teams, a secure and accessible platform. Parents of children with cardiomyopathy under the age of 5 years were invited. Parents were asked to submit anonymous questions prior to the morning if they wished. It aimed to be more accessible for families, rebuild a support network and help ease anxieties surrounding COVID-19 in the presence of the consultant and the clinical nurse specialists.
Results: 6 families attended. Feedback from parents was collated in relation to their experience of the first virtual coffee morning via email and feedback forms. This assisted in identifying the benefits of the event and how this can be adapted for future practice i.e. more regular sessions with guest speakers. Results identified that all parents found it very reassuring, informative and it eased their anxiety in relation to COVID-19 and cardiomyopathy.
Conclusions: Overall, the adaptation of the coffee morning to a virtual platform was deemed valuable as evidenced by feedback from staff and families that attended. Although the coffee morning was focused around COVID-19 for the event that took place during the pandemic, the team envisage that the focus can change in accordance with participants wishes. The virtual coffee morning is an initiative that will be incorporated into the future practice of the CNS team to further support families with children affected by cardiomyopathy.

P335
Needs and experiences of adolescents with congenital heart disease and parents in the transition process: a qualitative study

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Introduction: Most patients with congenital heart disease (CHD) need lifelong cardiac follow-up. Transitioning to adulthood and transferring to adult-focused care is often challenging. This study explored experiences and needs of adolescents with CHD and parents during the entire transitional process, including the post transfer period.
Methods: We performed a qualitative study according to the phenomenological approach, focusing on adolescents with CHD (n=9) and parents (n=12). Semi-structured interviews were conducted with patients and parents after being transferred to the adult department. Data were analyzed with inductive thematic analysis. Firstly, data collection and analysis of both samples were done separately. Finally, results were merged in a second data analysis to discover common themes.
Results: Five common themes between adolescents and parents were identified: 1) Having mixed feelings about leaving the pediatric care; 2) Being prepared and informed; 3) Shifting responsibilities and roles; 4) Being accompanied during consultations; and 5) Gaining trust in new healthcare providers.
Conclusions: Adolescents with CHD and parents would like to be offered adequate preparation and personalized guidance to reduce their anxiety and uncertainty. Transition might be better smoothed by focusing on improving adolescents´ disease knowledge and transitional skills. This might facilitate parents in handing over responsibilities and adapting to their new roles. Adolescents appreciate the presence of parents during consultation, but with reduced input. Finally, the participation of a transition coordinator and a joint transfer consultation involving the pediatric cardiologist seems to be of great importance for a fluent transitional process and especially in establishing a new treatment relationship.

P336
The evolution of a transition service for inherited cardiac conditions

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Introduction: The Inherited Cardiac Conditions (ICC) team at the Royal Brompton and Harefield Hospital (RBHT) offer a dedicated transition service for patients with ICC's. In 2015, the ICC service had no formalised transitional care provisions for adolescent patients. This is a unique patient group due to the sudden nature of development of the condition, risk of sudden cardiac death mode of inheritance. Therefore, an individualised service considering these factors was essential. This abstract aims to demonstrate the evolution from no pre-existing service to an integrated transition pathway utilised throughout the ICC team, meeting national standards and featuring patient co-design in its development.
Methods: In 2015, a focus group was held with seven ICC patients aged between 15-19 years to obtain feedback in relation to the implementation of a transition service. This was followed by analysis of results discussed with the wider ICC team to formulate a formal transition care pathway for patients. The transitional care pathway was further reviewed in 2018, utilising patient feedback collated via quick feedback sheets and emotional touchpoints. Further evaluation was obtained following the introduction of patient support days in 2018.
Results: The ICC transition service now consists of two dedicated adolescent clinics to provide MDT care and individualised transition plans. Additional nurse led transition consultations occur across ICC services providing education and support. Recently, the ICC team launched patient support days offering an opportunity for patients to meet other young people with ICC's and engage with appropriate charities. This has been expanded to include multiple specialist centres. Links are established with charities with social media forums supervised by trained staff, this is promoted within the ICC service.
Conclusions: Through service development and innovation, the team have successfully expanded the care that is provided utilising patient input to create a bespoke service for adolescents with ICC's. In addition to this the service has been successful in a grant application through Kings Health Partnership (KHP) for a transition research nurse. This will enable further re-evaluation of services, the implementation of nurse-led video clinics and the streamling of ICC transition services across three London trust sites.

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