

## Abstract Session: Congenital and Pediatric Cardiology

### O23–O27

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### O23

#### Long-term outcome and blood pressure profile in adult patients with repaired coarctation

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**Introduction:** Aortic coarctation (CoA) is a congenital narrowing of the proximal descending aorta. Despite improvement in interventional techniques, prevalence of cardiovascular events and aortic re-interventions is high with an increased prevalence of arterial hypertension (aHT). The aim of this study is to assess the incidence of adverse cardiovascular events, defining outcome predictors, and to assess blood pressure (BP) profile in patients (Pts) with repaired CoA.

**Methods:** We retrospectively evaluated adult Pts with repaired CoA who underwent clinical and imaging follow-up. Nocturnal BP was assessed by ambulatory blood pressure monitoring (n = 141 Pts). The Primary endpoint was defined as the composite of death, heart failure, presence of atrial fibrillation, cerebrovascular events, coronary revascularization, aortic dissection, re-CoA requiring intervention and pseudoaneurysm formation. Residual aHT

was defined as the presence of antihypertensive therapy, or a BP  $\geq$  140/90 mmHg.

**Results:** Overall, 280 Pts (112 females, 40%), age  $33,6 \pm 13,2$  years were included in the study. After a mean follow up period of  $25,5 \pm 12,7$  years after first intervention for CoA repair, primary endpoint occurred in 133 patients (48%): re-CoA requiring intervention (102; 36%), pseudoaneurysm formation (27; 10%), atrial fibrillation (20; 7%), cerebrovascular events (9; 3%), heart failure (5; 1,8%), coronary revascularization (4; 1,4%), aortic dissection (3; 1,1%). Residual aHT was present in 137 Pts (49%), and 134 Pts were under anti-hypertensive treatment (48%). Impaired nocturnal BP response was found in 45 of 141 Pts (32%) after ambulatory BP monitoring, and in 14 Pts (10%) with normotensive BP in the 24-hour-BP monitoring. By multivariate Cox regression analysis, primary stenting procedure (HR 28.877, 95% CI 13.343-62.493, p <0.001) was predictor of the composite endpoint. Pts with age >8 years at first CoA repair (HR 3.381, 95% CI 2.052-5.571, p <0.001) and pts with hypoplastic aortic arch (HR 1.530, 95% CI 1.069-2.190) were at higher risk for residual aHT.

**Conclusions:** Adult Pts with repaired CoA present a high prevalence of residual aHT, which is a driving risk factor for the development of cardiovascular disease in older age. Careful clinical and imaging follow-up is needed to address the occurrence of re-CoA or pseudoaneurysm formation and tailor antihypertensive treatment. CoA-Pts treated with stenting procedure are at greatest risk for cardiovascular events.

**Table:** O23-1. Prevalence of outcomes related to type of intervention.

Type of primary intervention	Age at first CoA repair, years	Re-coarctation n (%)	Pseudoaneurysm formation, n (%)	Residual arterial hypertension, n (%)
End-to-end anastomosis (n = 181)	1,2 IQR 0,1-5,4	71 (39)	11 (6)	87 (48)
Stenting (n = 39)	22,7 $\pm$ 12,6	11 (28)	5 (13)	20 (51)
Aortic patch plasty (n = 22)	9,2 $\pm$ 7,6	11 (50)	7 (32)	16 (73)
Subclavian flap-repair (n = 22)	0,1 IQR 0,1-0,3	7 (32)	1 (5)	5 (23)
Extra-anatomic aortic ascends-to-descends bypass (n = 11)	26 $\pm$ 14,6	1 (9)	1 (9)	5 (45)
Interposition graft (n = 5)	23,9 $\pm$ 18,6	1 (20)	2 (40)	4 (80)

## O24

**The role of exercise testing in congenital heart disease patients after coronary artery reimplantation**

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**Introduction:** The Ross procedure is an appealing alternative to treat aortic valvular diseases in children and young adults. As this procedure requires reimplantation of the coronary arteries, one could expect coronary artery stenosis as complication, similar to what has been found in patients after an arterial switch procedure for transposition of great arteries (TGA). Exercise testing is usually performed to detect myocardial ischemia due to coronary artery stenosis. Our study aimed therefore to 1) assess the prevalence of pathological exercise tests in patients after Ross and arterial switch procedure for TGA, and 2) evaluate whether coronary artery stenosis was confirmed by supplementary non- and/or invasive testing. Patients with tetralogy of Fallot (TOF) who do not undergo coronary reimplantation during surgical repair, served as the control group.

**Methods:** Clinical data, results of exercise tests and supplementary exams like coronary computer tomography (CCT), cardiac magnetic resonance imaging (CMR) or coronary angiography were retrospectively reviewed.

**Results:** Thirty Ross, 30 TOF and 14 TGA patients were included. Table 1 shows that duration of follow-up since surgical repair was significantly longer in TGA and TOF patients and that exercise capacity and the prevalence of pathological exercise tests were similar between groups. In 5 patients, a doubtful test was described. Nine patients received supplementary investigations because of pathological exercise tests (table 1). CCT was performed in a total of 30% of Ross, 21.4% of TGA and 3.3% of TOF patients ( $p = 0.023$ ). Cardiac catheterization with coronary angiography was performed in 20% of Ross, 21.4% of TGA and

13.3% TOF patients ( $p = 0.727$ ). None of these investigations revealed a coronary artery stenosis.

Patients with pathological exercise tests presented significantly more often a severe pulmonary stenosis and a trend to higher prevalence of pulmonary regurgitation than patients with normal exercise test (table 2).

**Conclusions:** Pathological exercise tests in patients after Ross procedure, arterial switch procedure for TGA and TOF repair, appear to be related to pathologies of the RV outflow tract rather than to coronary artery stenosis. The role of exercise testing in detecting coronary artery complications in these patients is probably limited and should be evaluated prospectively. At the present, additional non-invasive testing is mandatory to confirm or exclude coronary artery stenosis.

## O25

**ECG screening in paediatric athletes: a multi-centre retrospective study in 891 Swiss athletes**

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**Introduction:** Controversy persists regarding the utility of ECG in the screening of young athletes. Evaluation criteria have been revised and now achieve high sensitivity and specificity in adults. However, the lack of information on ECG screening in paediatric athletes illustrates the critical need to evaluate the risk factors in children with growth-dependent cardiac changes.

**Methods:** We conducted a retrospective study at six medical centres in Switzerland from 2011 to 2017. We regis-

**Table:** O24-1. Patient characteristics and result of exercise tests at last follow-up.

	Ross	TGA	TOF	p
Duration of follow-up since surgical repair (years)	8 (0-20)	18 (6-30)	20 (8-41)	<0.001
Age at the last consultation (years)	22 (17-47)	20 (14-32)	23 (17-48)	0.463
Reintervention rate, N (%)	7 (23.3)	6 (42.9)	17 (56.7)	0.031
%METs/Watt of predicted	93 ± 22	105 ± 21	94 ± 29	0.653
%Maximal heart rate	91 (68-103)	92 (80-100)	88 (59-98)	0.088
Results of exercise test				0.349
• Pathological tests, N (%)	6 (20)	1 (7.1)	2 (6.7)	0.234
• Doubtful tests, N (%)	2 (6.7)	0 (0)	3 (10)	0.469
Further investigation in case of pathological exercise test, N (%)	6 (75)	1 (100)	2 (40)	0.326

**Table:** O24-2. Comparison of groups with pathological and negative exercise tests.

	Pathological exercise tests (N = 14)	Negative exercise tests (N = 60)	p
Pulmonary insufficiency/RV-PA conduct insufficiency, N(%)	12 (86)	36 (60)	0.118
Degree of pulmonary stenosis/RV-PA stenosis, N(%)			0.004
• Absent (no stenosis), N (%)	4 (29)	24 (40)	0.381
• Mild, N (%)	3 (21)	24 (40)	0.161
• Moderate, N (%)	4 (29)	11 (18)	0.301
• Severe, N (%)	3 (21)	0 (0)	0.006

tered the demographic, anthropometric, sport-related and clinical data in athletes younger than 18 years, practising sports with different dynamic/static composition, on regional, national or international level, with a frequency of at least 6 hours per week consulting for a pre-participation screening. The pre-recorded 12-lead ECG was analysed by the most recent International Criteria of 2017.

**Results:** Of the 891 athletes enrolled in our study, 672 (75.4%) presented one or more training-related findings. There were 38 (4.3%) athletes with borderline criteria, thereof 2 (0.2%) with two borderline criteria, requiring further investigations. 17 (1.9%) athletes bared abnormal findings with one athlete presenting two of them. Of the 19 athletes with pathologic criteria, 10 (53%) presented a T wave inversion. Further investigations included echocardiography, Holter-ECG, exercise stress test and cardiac MRI. No underlying pathology was detected. Therefore, none of the athletes had to be excluded from sports. Total costs of this screening program were 91,595 CHF (103 CHF per athlete) for basic screening with history, clinical examination and ECG. When adding the costs for further examinations of the 19 athletes with pathologic findings, the total amount increased to 99,919 CHF (112 CHF per athlete).

**Discussion:** In our study, few ECG were abnormal, underlining the high specificity of the International Criteria in our cohort. Though, lack of longitudinal follow-up did not allow us to analyse their sensitivity. Most of inherited cardiac diseases develop with age and it is not yet clear if they can be detected with screening in early age. These findings question the utility of ECG screening in paediatric athletes. Based on our first results, we initiated the national register Swiss PAED (Swiss Paediatric Athletes ECG Database) in order to gather more data and allow follow-up studies.

## O26

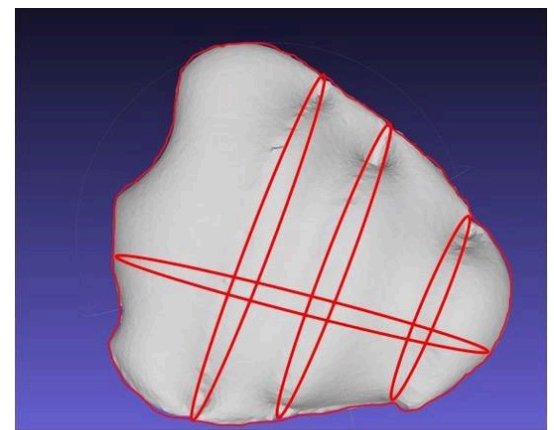
### Compressed sensing single-breath-hold cardiac magnetic resonance for quantification of right ventricular volumes in congenital heart disease

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**Background:** Cardiac magnetic resonance (CMR) has become the gold standard for the evaluation of the right ventricle (RV) in patients with congenital heart disease (CHD). Recently, novel compressed sensing (CS)-based single-breath-hold multislice CMR cine techniques have emerged as a mean to accelerate data acquisition. The aim of this study was to compare a CS-CMR cine technique with the standard multi-breath-hold technique cine images

**Figure:** O26-1. 3D reconstruction of the right ventricle.



3D reconstruction of the right ventricle. The five 2D planes which are acquired with the compressed sensing based single-breath-hold multislice cine technique, are shown in red.

for the assessment of right ventricular (RV) volumes and function.

**Methods:** The CS cine sequence acquires 2 long-axis and 3 short-axis cine loops in 1 single breath-hold. To measure RV ejection fraction (RVEF(CS)) as well as RV volumes, a non-model-based 3D reconstruction software was used (figure). For comparison, a conventional stack of multi-breath-hold short axis cine images was acquired. As a reference for the RV stroke volume (RVS), pulmonary antegrade flow was measured by phase-contrast CMR in the main and both pulmonary branch arteries.

**Results:** Seventeen patients with different CHD lesions were included (age 32±14 years; 7 (41%) female; CHD lesions: 3 tetralogy of Fallot, 5 Ross-operation, 4 pulmonary stenosis, 2 ventricular and 1 atrial septal defect, 1 Marfan, 1 partial anomalous pulmonary venous connection). None of the patients had more than a trace of tricuspid regurgitation as determined by echocardiography. CS derived RV enddiastolic volume, RVS and RVEF were significantly lower than those determined by the standard short axis cines (table). The agreement of RVS(CS) with total antegrade pulmonary flow determined either in the main pulmonary artery (MPA) or by the sum of antegrade flow in both pulmonary branch arteries (PPA) was superior to that of RVS(standard).

**Conclusions:** The results demonstrate the feasibility of applying the CS strategy to evaluate the RV function and volumes in CHD patients. The preliminary data show that the single-breath-hold CS better determines RV stroke volume as it avoids misalignment of the different long and short axis cine loops. The CS strategy has the potential to replace the multi-breath-hold standard CMR technique.

## O27

### Evolving hypoplastic left heart syndrome: assessment of single ventricle physiology prediction score and rate of potential candidates for fetal aortic valvuloplasty

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**Introduction:** The 3-stage surgical treatment has significantly improved the prognosis of patients with hypoplastic left heart syndrome (HLHS), but there is still a significant number of comorbidities associated with the single ventricle physiology. Due to the progressive nature of HLHS and the importance of abnormal flow through the left ventricle during fetal life, fetal aortic valvuloplasty (FAV) has been proposed to restore a biventricular physiology. Candidates for FAV should have 1) a high likelihood for evolving toward HLHS (eHLHS) without FAV (assess with eHLHS score) and 2) a high likelihood for maintaining a biventricular physiology after FAV (assess with Boston score). We performed a retrospective study to: 1) determine the performance of the eHLHS score for predicting evolution toward HLHS in our population, and 2) determine the number of fetuses seen at our clinic with underdevelopment of left heart aorta-complex that could have been candidate for FAV according to Boston score.

**Methods:** Based on surgical, catheterization and echocardiographic databases, we identified all patient with some underdevelopment of the left heart-aorta complex seen at the fetal cardiology clinic of our tertiary centre between 2010 and 2018. Morphologic and hemodynamic measurements were extracted from echocardiographic exams, birth and pregnancy data were extracted from the computerized medical reports.

**Results:** Twenty-six fetuses were identified, with 17 satisfying the criteria for eHLHS: 5 fetuses evolved to HLHS, 1 to biventricular physiology at birth, and 11 pregnancies

**Table:** O27-1. eHLHS outcome.

	eHLHS score predicting univentricular physiology	eHLHS score predicting biventricular physiology
Univentricular physiology at birth	5	1
Biventricular physiology at birth	1	5
Termination of pregnancy	11	3
Boston score $\geq 4$ (successful FAV)	0	0
Boston score $< 4$ (unsuccessful FAV)	17	9

were terminated. On the other hand, 9 fetuses didn't satisfy the criteria for eHLHS: 5 fetuses evolved to a biventricular physiology at birth, 1 to HLHS and 3 pregnancies were terminated. Sensitivity, specificity, positive and negative predictive values of the eHLHS score in our population are 83%. Based on the Boston score, none of the fetuses included in our study would have been a good candidate for FAV (Boston score  $\geq 4$ ), even among those for which eHLHS score predicted a biventricular physiology.

**Conclusion:** The performance of the eHLHS score in our population is decent but not excellent. This should be taken into account when counseling families. The lack of good candidates for FAV in our cohort illustrates the challenge of prenatal diagnosis of initially subtle lesions that may lead to HLHS and the rarity of the condition due to the high selectivity of the Boston score.