Predictors and outcomes of right ventricular outflow tract conduit rupture during percutaneous pulmonary valve implantation: a multicentric study

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**Aims:** Conduit rupture is a serious complication encountered during percutaneous pulmonary valve implantation (PPVI). We sought to evaluate the incidence and predictors of conduit rupture during right ventricular outflow tract (RVOT) transcatheter treatment.

**Methods and Results:** All consecutive patients who underwent transcatheter RVOT treatment from May 2008 to December 2011 were prospectively studied. Baseline demographics along with incidence, predictors and outcomes of conduit rupture with various transcatheter therapies were reviewed. Conduit rupture occurred in 9 out of 99 patients (9.09%). All conduit ruptures occurred universally during balloon dilatation. Significant risk factors included heavy calcification (p<0.05, OR=16 [1.87-357]), conduit type (homograft/others; p<0.05, OR=5.37 [1.1-27.39]) and conduit stenosis as the primary lesion (p<0.05). Other factors such as prolonged time interval between prior surgical RVOT repair and interventions, use of high-pressure balloons, balloon diameter, and overexpansion of conduit statistically failed to show any association. All patients were managed in the cardiac catheterization laboratory. There were no delayed complications during a mean follow up period of 2.3±0.95 years.

**Conclusion:** Conduit rupture is a serious complication. Heavy calcification, homograft conduit and stenosis as primary lesion were significant predictors. Immediate diagnosis with use of targeted interventional therapies should be attempted before proceeding with PPVI.
PPVI procedural success, residual gradients and in-situ duration of Melody valve were comparable in both groups with and without IE. Microbial diagnosis was achieved in all and probable source of infection and its portal of entry were identified in 4/5 cases. A significant number of patients with IE however had abruptly discontinued antiplatelet therapy (3/5, p=0.026, RR=20 [3.34-120.8]). Three patients presented with severe obstruction, of whom 2 died within 24 hours while on medical therapy and one required urgent balloon angioplasty to relief obstruction.

Conclusion: Early and late onset, de novo prosthetic valve endocarditis of the Melody valve is emerging as a catastrophic complication of PPVI. Abrupt discontinuation turns out to be a significant predictor of Melody valve endocarditis. Due to its rapidly progressive nature, aggressive invasive management should not be delayed.

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Atrial septal defect area assessed by 3D echo is relevant for calibration during percutaneous closure.

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Balloon Sizing during percutaneous atrial septal defect (ASD) closure extends radiation dose and device size but remains the gold standard. We assessed the relationship between the occlusive balloon diameter (BD), area and diameters measured using real time Transesophageal Echocardiography (RT3DTEE) and the effect of ASD shape.

Methods: From 2011 to 2013, we prospectively enrolled 30 children (mean weight 30.9kgs max64 min18) who underwent transcatheter closure of an isolated ASD under 3DTEE (3D-matrix array 2-7 MHz TEE probe and iE33 US machine Philips®). ASD diameters were measured by 2D-TEE and off-line by 3D Multplanar reconstruction (Qlab software®). ASD area was estimated by delineating the outlines of the defect on the reconstruction software. The shape of the ASD was assessed visually on the RT3DTEE “en face” view and was defined as circular (n=16) or oval (n=14). An asymmetry index was calculated by dividing the maximal divided by the minimal 3D diameter (mean 1.4±0.2 mm max1.84). A cut-off of 1.25 was set to distinguish oval (n=8) and circular shape (n=22).

Results: The Amplatzer® device number was equal to BD±1mm in 23 cases (76.7%) and higher in the remaining cases. Difference between 3D maximal diameter and BD (2.8±2.2mm min-9.6 max12.2, p=0.0051) was higher in round ASD than in oval shape (4.8±3.5 vs 1.4±4.1, p=0.04). ASD area was well correlated with BD (r=0.82, p<0.0001). After multivariate linear regression analysis, ASD area by 3D delineation was the only significant variable for the prediction of BD: BD (mm)= 4.5*ASD area (cm 2)+11. This formula allow a prediction with <1mm difference with the observed BD in ½ and <2mm in 2/3 of procedures.

Conclusion: The relationship between BD and echo parameters is influenced by the ASD shape. ASD area estimated by delineation on a 3D view is the most relevant parameter to estimate the BD. It may be sufficient to guide percutaneous ASD closure without balloon sizing in children.

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Percutaneous closure of coronary artery fistulas in paediatrics

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Background: prevalence of coronary fistulas is poorly known in the pediatric population. However complications are potentially serious with: heart failure, sudden death, arrhythmias, endocarditis. In adults, transcatheter closure is an alternative to surgical treatment. In pediatric population, few publications exist on the feasibility and effectiveness of this treatment.

Aims: To evaluate the feasibility of percutaneous closure of coronary fistulae in paediatrics, the evolution and the occurrence of complications.

Methods: Retrospective observational multicentric national study concerning all patients under 16 who underwent transcatheter closure of a congenital coronary fistula (complex cardiopathies excluded).

Results:
- ORIGIN Left coronary (28) Right coronary (28) Double (5)
- DRAINAGE SITE Right atrium (12)
- Right ventricle (12)
- Left atrium (2)
- Left and right ventricle (1)
- Pulmonary artery (1)
- Right atrium (12)
- Left atrium (1) Right ventricle (4)
- Pulmonary artery (1)
- Population: 61 patients (36 girls, 25 boys), median age at diagnosis 0.6 years [0-15.4], 3.9 years at procedure [0-16].
- Initial signs: precordial murmur (90%), congestive heart failure (11%), non specific ECG abnormalities (4,9%), left or right ventricular dilatation (31%).
- Efficiency: complete occlusion at hospital discharge in 82%, 5 procedures failures.
- Complications: no death, 3 transient STEMI, 3 Coil embolization, 1 ventricular fibrillation recovered, 1 Leg ischemia.

Combination therapy: antiplatelet (46%), mean duration 4.1 months. 1 with AVK treatment
- Evolution: data are available for 43 children (70%), median follow-up 91 days [min=0, max=4824]. At 2 years, complete occlusion rate was 72.7%±27.6%
- 5 reoperations (3 transcatheter, 2 surgical procedures). 1 late recanalization at 17 months.
- No cardiovascular complication.

Conclusion: The percutaneous closure of coronary fistulas in the pediatric population appears to be effective and relatively safe.

Figure – Right coronary to right atria fistula
Management and transcatheter therapy in partial abnormal pulmonary venous return with additional drainage to the left atrium: a multicenter study and world experience

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Background: A dormant pulmonary vein connecting a partial abnormal pulmonary venous return to the left atrium has been occasionally reported. Indications for transcatheter therapy are not clarified and data on mid-term results is lacking. We aim to report characteristics and transcatheter therapy in partial abnormal pulmonary venous return with additional drainage to the left atrium.

Methods: We retrospectively studied such patients in 5 institutions.

Results: Ten patients (6 girls) presented at a median age of 8 (0.1 to 54) year-old with 2 anatomic types: 8 vertical vein types (drainage of the left upper lobe to the innominate vein via a large vertical vein and to the left atrium via the left upper pulmonary vein) and 2 scimitar vein types (drainage of the right middle and lower pulmonary veins to the inferior vena cava and to the left atrium via a connecting pulmonary vein). Associated malformations were aortic coarctation (n=2) and secundum atrial septal defect (n=3). Two patients with vertical vein type were operated. Transcatheter occlusion of the abnormal pulmonary venous return was performed with Amplatzer devices in 7 cases, associated with coarcl-occlusion of systemic arterial supply (n=2), secundum atrial septal closure (n=2), left upper pulmonary vein stenosis stenting (n=1), and coarctation stenting (n=1). Including previously published cases, 18 patients (13 vertical vein, 5 scimitar vein types) underwent transcatheter repair. Patients over 40 year-old tend to be symptomatic at presentation (p=0.056).

Conclusion: In partial abnormal pulmonary venous return with dual drainage, transcatheter therapy can be offered in the majority of the patients.


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Introduction: very few comparative studies have yet reported health related quality of life (HRQoL) among children with congenital heart diseases (CHD).

Method: we prospectively recruited children aged 8 to 18 with CHD (group 1, n=282) and same age randomized control population selected among schools (group 2, n=164). CHD were recruited in 2 tertiary care centers in France and Belgium. 30 families refused to participate in group 1 and 150 in group 2. Primary outcome was the scoring for each dimension with Kidscreen-28 and Children-27 for parents (French version of the generic validated pediatric HRQoL questionnaire). Secondary outcomes were: severity class of CHD from 1 to 4 (from Uzark), cardiopulmonary exercise test (VO2max, SVI), and PedsQL scoring (non validated HRQoL generic questionnaire, French and Belgian versions).

Results: 282 children with CHD (sex ratio 1,9 – mean age 12.3±3) and 164 children in schools (sex ratio 1,1 – mean age 12.8±2,4) were recruited. Both centers were comparable for most demographic and clinical data. In most dimensions self reported QoL scores among children with CHD were not different from control group, except for physical well-being (mean 46.5±10.2 vs 49.9±8.6, p<0.05). QoL in lower severity classes is not significantly different from controls. Children with severe CHD (class 4) have lower QoL in physical well-being (43.3±8.64, p<0.05). In several dimensions (physical, social, school) parents’ reported QoL is lower than their children’s evaluation in group 1 but not in group 2 where scorings are identical. Teenagers have higher QoL scores than younger children.

Conclusion: QoL among children with CHD is close to that of same age healthy children except for physical well-being. HRQoL for low severity CHD patients is similar to controls. QoL for severe CHD is the most impacted. Further studies should evaluate QoL evolution in time per patient.

Quantitative assessment of right ventricular function by 2D speckle tracking imaging in Eisenmenger syndrome

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Background: 2D strain imaging (2DS) is a reliable non invasive technique for assessing global and regional right ventricle (RV) function in pulmonary hypertension (PAH). Eisenmenger syndrome (ES) differs significantly from other types of PAH (OTPAH) in its physiology and prognosis. Aim: To assess the RV function by 2DS in ES and determine whether there is a difference with OTPAH.

Methods and Results: Clinical and echocardiographic variables were assessed in 14 consecutive patients with ES (mean age 39±12), and 15 patients with other types of PAH (mean age 61±12). 17 Patients were in functional class (FC) II (10 ES) and 1 patients were in FC III (4 ES). No difference was found between the 2 groups regarding 6MWT (p=0.93), mPAP and PVR were higher in the ES group (71.7±13±mmHg vs 41.6±9±mmHg, p<0.01) and 92± vs 8.3±3 UW/ooes (p<0.05).

Correlation between 2DS and conventional parameters was good (r=0.91, FAC r=0.89, RA/LA surface ratio r=0.88, peak systolic velocity r=0.84, eccentricity index r=0.81, Tei index r=0.81). No statistical difference was found between ES and OTPAH according conventional parameters except for TAPSE (p=0.02, FAC p=0.13, S p=0.77), whereas 2DS showed a markedly less impaired RV function in the ES group (-18.8±16.6% vs -13.8±2.89%, p<0.001), even when analysis was adjusted to FC in each group (p<0.01). 2DS segmental analysis of the RV free wall showed more pronounced difference in the apical segments (p<0.001) in comparison with basal segments (p=0.57) between the 2 groups, emphasizing the importance of RV apical function in PAH.

Conclusion: 2DS provides a new tool to quantify global and regional longitudinal RV function in ES. Despite additional cyanosis-related comorbidities, the specific pathophysiology and hemodynamic conditions of ES are associated with relatively preserved RV systolic function which may be part of the better survival outcome in comparison with OTPAH.

Aortic root and ascending aortic dilatation in patients with repaired tetralogy of Fallot: determinants, rates of progression and outcomes.

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Background: We examined the features, progression and relations with outcomes of aortic dilatation in adults with repaired tetralogy of Fallot (rtToF).

Methods and Results: Retrospective study of 110 rtToF adults with native aorta, median age 30.9±10.0 years were studied by cardiovascular magnetic resonance (CMR) at baseline, and at follow-up (median 6.3 [IQR 5.1-7.6] years). Aortic diameter was measured at sinus, sino-tubular junction and mid-dilating aorta level. Aorta dilatation was defined as diameter > 2
standard deviations larger than normal. Three different phenotypes were described: 36% had normal aortic dimension at any level (Figure 1A), 42% had aortic root dilatation (Figure 1B) and 22% had aortic root and ascending aorta dilatation (Figure 1C). There was no predictor of sinus dilatation, but age at repair (OR1.1, p=0.004), male gender (OR3.6, p=0.01), pulmonary atresia (OR17.3, p=0.0007) and previous palliation were predictors of ascending aorta dilatation. On multivariate analysis, age at repair (p=0.006), male gender (p=0.008) and pulmonary atresia (p=0.0004) were independent predictors for ascending aorta dilatation. No patient experienced any aorta related event. Diameter increase was low: mean rate 0.05 ±0.1 mm/year at sinus and 0.12 ±0.26 mm/year ascending aorta level and occurred in 25% of patients at sinus, and in 35% at ascending aorta level. No predictors of aortic diameter progression were found.

Conclusions: There are different phenotypes in aorta in patients with rOoT and aortic root dilatation is the most common. Rates of diameter increase in mid-term follow-up are low. This should be taken into account when planning for imaging in follow-up and/or discussing the merits of prophylactic aortic replacement.

Figure 1 - Different aorta phenotypes.

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The medical past of adults with complex congenital heart disease impacts their social development and professional activity.

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Advances in surgery and therapeutic catheterization in recent decades increased steadily life expectancy and prevalence of adult patients with congenital heart disease (ACHD). We assessed medical and psychosocial parameters of ACHD according to the complexity of the disease.

Methods: We included from a single-center prospective observational cohort study which started in January 2013, 68 ACHD patients (40.7±29.3 ±17.8 years) followed in cardiology from January to April 2013 who answer a questionnaire assessing daily activity and psychosocial being.

Results: Underlying cardiac disease was: AVSD in 30%, VSD in 35%, ASD in 9%, PDA in 5%, associated shunts in 5%, complex CHD in 10%, left heart obstruction in 25%, pulmonary veins anomaly in 25% and TGA in 1%; CHD was native in 122 cases (77%), 7 had palliation (4%) and 30 complete repair (19%). Pulmonary oxygen saturation was 84±12% (range 44 to 98%), lower in non-operated or palliated cases (81%) than in repaired cases (92%, p=0.002). Patients were in NYHA class I (18%), class II (42%), class III (37%) or IV (3%), not different with previous repair or not. Target therapy agents were given in 35% of the cases (1 agent in 20%, 2 associated in 13%, intravenous epoprostenol in 1.5%). Death occurred in 26 patients (16%) at the age of 29.3±17.8 years. Complications occurred in most of the cases (64%) including: hemorrhages events, syncope, thrombo-embolia, cerebral abscess, infective endocarditis, heart failure or arrhythmias. NYHA class did not differ between patients with or without target therapy. SpO2 was 82% in untreated cases compared to 86% in treated cases (NS). Survival rates were: 98% at 10-years, 93% at 20-years, 87% at 30-years, 83% at 40-years, 73% at 50-years and 53% at 60-years of follow-up. Survival was lower in Down patients (p=0.0023), in males (p=0.04) and higher better up to 50-years in patients under target therapy (p=0.05).

Conclusion: Survival rates of adult patients with Eisenmenger Syndrome seem to improve up to 50-years of age with target therapy agents. These results have to be confirmed by larger scale multicentre studies.

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Is surgical repair of partial atrioventricular septal defect safe and efficient in adulthood?

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Background: partial atrioventricular septal defect (PAVSD) is usually operated in childhood with excellent long-term results. However, some patients may present during adulthood and their management as well as their outcome is still unclear. We sought to analyze the clinical characteristics at presentation and the outcome of patients with PAVSD, non operated or operated during adulthood.
Methods: Between January 2000 and March 2013, 31 adult patients with PAVSD presented and were followed in our care network. The mean age at study entry was 34 (+16.6) years.

Results: Dyspnea was the most frequent symptom at presentation in 19 cases (61%). Left atrioventricular valve regurgitation (LAVVR) and right ventricular overload were higher in patients undergoing surgical repair (p = 0.01). Twenty-two patients (71%) had their surgical repair at a mean age of 39.4 (±15.3) years old with ostium primum closure and partial or complete suture of the left atrioventricular cleft without postoperative death or major complication. There was one late reoperation for a residual shunt. Nine patients (29%) were not operated. After a mean follow-up of 7.4 (±7.1) years, 26 patients (84%) are in NYHA class I or 2 whereas 5 (16%) are in NYHA class III or IV. Operated patients have a lower NYHA class (p = 0.01), a lower grade of LAVVR (p = 0.03) and a lower systolic pulmonary artery pressure (p = 0.01) than unoperated patients at last follow-up. The onset or persistence of supraventricular arrhythmias (SAV) after surgery was associated with an older (≥40 year-old) age at repair (p = 0.02), a higher systolic pulmonary artery pressure (p = 0.04) and a higher grade of LAVVR (p = 0.03).

Conclusion: PAVSD repair is indicated in the adult patient because it carries an excellent prognosis and operated patients have better long-term clinical and hemodynamic status than unoperated patients. Moreover, SAV incidence decreases when surgery is performed before 40 year-old.

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Relationship between fluoroscopic time, morphological parameters and irradiation during catheterization in children with congenital heart disease.

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Cardiac catheterization procedures are being increasingly performed in children with congenital heart disease for diagnostic and treatment purposes. Given children’s greater sensitivity to radiation and the longer life span during which radiation health effects can develop, knowledge of factors associated with irradiation is necessary. We report the radiation doses and related factors for children who underwent cardiac catheterization in Toulouse children Hospital from January to April 2013.

Methods and results: We prospectively included 60 consecutive children (mean age 4 years old, weight 2.350-59 kg) undergoing a therapeutic (n=55, 91.7%) or diagnostic (n=5, 8.3%) procedure. We investigated the relationship between Dose Area Product (DAP), Fluoroscopic Time (FT), pulsed Fluoroscopic DAP, image acquisition DAP, age, morphological parameters and double products combining FT and morphological parameters. Body Surface Area (BSA) was calculated with the Mosteller formula.

The mean DAP was 20697±29342mgycm². The mean total fluoroscopic time was 24.6±19.7 min. DAP was not significantly different between diagnostic and therapeutic catheterization (p=0.98). Although Image acquisition DAP accounted for only 4.4±2.4% of FT, it represented 42.5±19.6% of DAP. DAP was moderately but significantly correlated with FT (r=0.73, p<0.0001), BSA (r=0.44, p=0.0011), age (r=0.37, p=0.0082), weight (r=0.43, p=0.002) and size (r=0.38, p=0.0052). DAP was strongly associated with FTXWeight (r=0.92, p<0.0001), FTXBSA (r=0.93, p<0.0001) and FTXSize (r=0.91, p<0.0001). Linear regression analysis model involving FTxBSA to predict DAP was significant (p<0.0001). Approximately 90% of the variance of DAP was accounted for by this model.

Conclusion: FT and morphological features (BSA, weight, size) are the key parameters associated with DAP. Peculiar attention to reduce FT and avoid unnecessary image acquisition may decrease irradiation during catheterization in children with congenital heart disease.
patients grew up. Neutropenia was found in nineteen patients. It was responsi-
ble for eight episodes of severe infection including three septic shocks
causing death in two cases. Survival correlates with two prognostic factors:
the severe neutropenia at diagnosis (<0.5 G/L) and the year of birth, as
patients born after 2000 had a 70% survival rate vs 22% for those birth before
2000.

Conclusion: BTHS outcome in this survey is driven by cardiac events and
with the infectious risk related to neutropenia. Intensive treatment of heart
failure and prevention of infection in infancy may improve survival of patients
with BTHS.

Incidence, diagnosis and outcomes of coronary artery compression
during percutaneous pulmonary valve implantation

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Background: Coronary compression (CC) may occur during percutaneous
pulmonary valve implantation (PPVI) and is potentially life threatening. We
sought to evaluate its incidence, diagnosis and outcome.

Methods: All consecutive patients who underwent transcatheter RVOT
treatment from May 2008 to December 2011 in 4 institutions were studied.

Results: CC occurred in 6 out of 100 patients (6%) at a median age of
24 (13 to 49) years, with RVOT conduit stenosis as the primary lesion in all cases.
The initial congenital heart disease was pulmonary atresia-ventricular
septal defect (n=3), complex transposition of the great arteries (n=2) and
critical aortic stenosis status-post Ross operation (n=1). The RVOT initial
median diameter at surgical implantation was 23 (17 to 24) mm and conduit
types were homograft (n=3), bioprosthesis (n=2) and a pericardial
patch(n=1). CC was diagnosed by coronary angiogram during balloon dila-
tion of the RVOT in all cases whereas it was suspected on pre-procedure
computed tomography (CT-scan) in only 2 cases. Compression occurred on
the left anterior descending coronary artery in 4 cases and on a right coro-
nary artery that arose from the proximal left anterior descending coronary
artery in 2 patients (single coronary artery). No risk factor was found but
there was a significantly higher incidence of CC in one of the 4th institu-
tions (p=0.04). CC was well-tolerated and resolved after the balloon was
deflated in all the cases. No patients underwent RVOT stenting or PPVI.
Surgical conduit replacement was electively performed in 3 patients. Two
patients with moderate residual RVOT stenosis are followed. One patient
with encephalopathy and respiratory insufficiency died 9 months after
catheterization.

Conclusions: CC is efficiently diagnosed by coronary angiogram during
balloon dilation in a small proportion of patients undergoing transcatheter
interventions on RVOT. Diagnosis by pre procedure CT-scan is not accurate.
No specific risk factors exist. Surgical conduit replacement is indicated when
balloon dilation fails to improve the RVOT obstruction.

Cardiac transplantation in adults patients with congenital heart
disease: long-term outcomes and influence of initial defect and repair:
a collaborative study on 97 patients

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Purpose: Due to increasing success with repair or palliation in childhood,
there is a rapidly growing population of adult patients with complex conge-
tinal heart disease who may require transplantation. But ACHD is a heteroge-
neous population. We compared outcomes in different defects and indifferent
physiopathological models of repair i.e. univentricular (1V) or biventricular
(2V) status.

Methods: Out of a multi-institutional (3 centers) series of 2257 HTx from
1988 and 2012, 100 (4.4%) were performed in 97 ACHD aged ≥16 years at
the time of operation (65 men). We evaluated clinical data, etiology, surgical
history, perioperative issues and outcomes.

Results: The mean age at the time of HTx was 29.8±10.5 years (range 16–
58). Forty-three patients (44%) had 1V physiology including 16.5% Fontan-
type circulation and 54 (56%) had 2V physiology including 26% with a sys-
temic right ventricle. Primary diagnosis included single ventricle (27%), D-
transposition of the great arteries (20%), tricuspid atresia (13%), tetralogy
of Fallot (12%), congenital valvular heart diseases (11%), corrected transposition
of the great arteries (7%). Twenty-four patients required reintervention for
hemorrhage. In-hospital mortality was high (34%) and multivariable factors
included female gender of the donor and tricuspid atresia. The 1V patients
were younger (26.9 vs 32.1 years, p<0.014), had undergone more surgeries
(2.9 vs 1.9 prior surgeries, p=0.002), had a lower requirement for ventricular
assist device implantation before HTx (4.7% vs 20.4%, p=0.02), and had
longer ischemic (229 vs 182min, p<0.001) and cardiopulmonary bypass times
(240 vs 163min, p<0.001). Long-term survival was significantly higher in 2V
patients (p=0.023).

Conclusion: HTx of ACHD poses several surgical challenges due to
complex anatomy, specific physiology and prior surgical interventions. 1V
physiology is associated with higher in-hospital and long-term mortality, in
comparison to 2V physiology.

TAPSE is correlated with right ventricle ejection fraction in children
with congenital heart disease affecting right ventricular function

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Background: In congenital heart disease, assessment of right ventricular
(RV) function is determinant for patient’s follow-up and therapeutic strategy.
However, only few data of RV systolic functioning parameters measured by
echocardiography in children are available, and their usual change in different
types of RV loading condition. We investigated the feasibility, reproducibility
and correlation to 3D RV ejection fraction (RVEF) of each parameter of RV
systolic function in children with congenital heart defect affecting the right
ventricle.

Methods and Results: We performed echocardiography in 143 consecu-
tive children aged 0 to 18 years with different pathological RV loading condi-
tion. We classified them in three groups: dominant volume overload, dominant
barometric overload, and mixed overload. We measured RV systolic function
parameters described in the pediatric and adult literature: Tricuspid Annulus
Posterior Systolic Excursion (TAPSE), Tricuspid Annulus Maximal Systolic
Velocity in Doppler Tissue Imaging (Sa), Fractional Area Change (FAC), 2D
RVEF, 3D RVEF, IsoVolumic Acceleration (IVA) and Tei index. 3D RVEF
was measured using a new system of 2D echography with Knowledge-Based
3D reconstruction allowing to reconstruct volumes from sections of right ventricle
with standard sonographer (Ventrpoint, USA). This system has been
already validated by MRI volumes measurements.

We found that TAPSE was the most feasible (n=137/143; 97.08%), reproduc-
ible (ICC=0.986; p=0.001) (Coefficient of Variation=2.1%) parameter and was sig-
nificantly correlated to 2D RVEF (R=0.301; p=0.0014) and 3D RVEF (R=0.337;
p=0.0014). Z-score TAPSE age-adjusted is significantly different in each type of
loading condition (Pressure/Volume p=0.0089; MixedPressure p=0.365; Mixed/
Volume p=0.0004). Mixed overload had the worse TAPSE Z-score value (−5) and
volume overload had the best TAPSE Z-score value (−1.5).

Conclusion: We recommend using TAPSE Z-score to assess RV func-
tion in children. However TAPSE is significantly dependent on loading
condition and further studies need to determine range values in each loading
condition (figure next page).
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Atrioventricular discordance with ventriculo-arterial concordance: diagnostic challenge, surgical management and long-term outcome

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Abstract 296 – Figure – Correlation between TAPSE Z-score and REVF

Background: Atrioventricular discordance with ventriculo-arterial concordance is a rare cardiac defect whose pathophysiology resembles transposition of the great arteries.

Objective: To report a series of ten patients with atrioventricular discordance with ventriculo-arterial concordance focusing on segmental analysis, diagnostic difficulties, surgical management and follow-up.

Methods: Monocentric retrospective review of medical files of all patients with this diagnosis seen from 01.01.1983 to 01.01.2013.

Results: Seven patients had [I,D,S] segmental arrangement, two had anatomically corrected malposition (S,L,D) and one had [S,L,S] arrangement. Only five patients were correctly diagnosed preoperatively while diagnosis had to be completed during or after surgery in the other five. Mean age at repair surgery or at last surgical intervention was 3.4 years (range: 5 months – 12.8 years). Repair surgery finally performed was atrial switch procedure of Senning or Mustard type in eight of ten patients. Repair included ventricular septal defect closure in three cases and right ventricle to pulmonary artery conduit or patch in four cases. Main postoperative complications were two cases of baffle obstruction requiring reintervention and one sick sinus syndrome needing pacemaker implantation. There was no postoperative heart block. Two early postoperative deaths and eight late survivors. Mean follow-up after repair was 6.8 years (range: 5 months-25.4 years) with fair functional status in all but one patient who will be listed for heart transplantation.

Discussion: Preoperative diagnosis of atrioventricular discordance with ventriculo-arterial concordance remains challenging. Atrial switch procedure is the surgical method of choice. If correctly diagnosed, long-term follow-up is encouraging. Rhythm disturbances and baffle obstruction are the main postoperative problems in this series.

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Premature neonates with severe congenital heart diseases: post-operative course and outcome after early cardiac surgery.

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Background: Prematurity is a recognized risk factor for morbidity and mortality following cardiac surgery. This study examined the post-operative course and the midterm outcome after early cardiac surgery.

Methods: Since 2000, 29 consecutive premature neonates (median GA: 34.4w) were admitted for severe congenital heart disease (at least 2 defects and/or defect resulting in death within the 1st month of life). All underwent cardiac surgery with cardiopulmonary bypass in the preterm period; records were retrospectively reviewed. At surgery, median age was 9d, median corrected GA 36.1w, median weight 2340g; 44% were preoperatively, ventilated. Pathologies were transposition of the great arteries (n=9), aortic arch obstruction associated with intra-cardiac defect (n=9), total abnormal pulmonary venous return (n=5), critical aortic valve stenosis (n=3), univentricular hearts (n=3); all pts except the latter had a one-stage biventricular repair.

Results: Thirty-day survival was 86% with 7 early post-operative deaths. Mortality rate raised 66% for pts with univentricular hearts. A low weight at birth and at surgery and a long delay before sternal closure were risk factors of mortality. Early outcome was independent of the gestational age, corrected gestational age at surgery, preoperative status, and duration of both cardiopulmonary bypass and aortic clamping. Median follow-up of hospital survivors was 15.7 months (including 11 pts>5 y); 3 pts died after discharge within 5 months post-surgery, 6pts required 8 reoperations for residual cardiac sequelae. At last follow-up, 13pts were in class I or II of NYHA, 2 had pulmonary artery hypertension.

Conclusion: Cardiac surgery with one-stage biventricular repair for severe congenital heart diseases is feasible in preterm neonates; mortality is high but acceptable; the degree of prematurity is not a risk factor of death, which argues for early surgery. At midterm, there are no late death, a good clinical status but frequent reoperations.

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Management of anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) syndrome during adulthood: a multi-center retrospective study in a serie of twelve patients.

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ALCAPA syndrome is a rare congenital heart disease, mainly diagnosed during the first months of life, with a high early mortality in the absence of surgery in symptomatic children. Adult form is extremely rare and no recommendation for its management is available. We undertook this study in order to analyze symptoms, management and long term outcome of adult patient with ALCAPA.

Methods: we analyzed retrospectively the data of patients who have been hospitalized in Cardiology Departments of 7 French Hospitals.

Results: 12 adults (30.2±21.4 years) were included. Main symptoms consisted of chest pain (58.3%), supra-ventricular arrhythmia (42.9%) and heart failure (33.3%). ECG was abnormal in 60% (negative T waves or ST segment depression). Echocardiogram showed a mean LVEF of 51.5±12.8% and mitral regurgitation was present in 83.3% of the cases (mainly moderate). Coronary angiography was performed in 83.3% of patients and showed ALCAPA in all cases. CT-scan, MRI and myocardial scintigraphy were also performed (respectively in 45.5%, 27.3% and 41.7%), as well for diagnosis as for follow-up...
up. 11 patients underwent reconstructive surgery, with mitral valve reconstruction in one case. 1 patient refused surgery and died 2 years later. Immediate post-operative complications were cardiogenic shock (2 patients), one non-significant narrowing of the transferred coronary artery ostium that was treated medically and in 1 case, a significant stenosis of the pulmonary artery trunk and of the LCA required surgery. A prophylactic ICD was implanted in 1 patient. All operated patients are alive with a follow-up of 16.9 ± 22 years.

Conclusion: diagnosis of ALCAPA in adult is done mainly by coronary angiography in the presence of atypical symptoms. The long term results of surgery are excellent. Based on the complicated evolution of this syndrome and the good results of surgery, it should probably be proposed systematically even in case of accidental discovery in an asymptomatic adult.

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Prognosis of severe congenital heart diseases: do we overestimate the impact of prenatal diagnosis?

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Introduction: congenital heart diseases (CHD) are the most common and serious among birth defects (incidence 0.8%). Prenatal screening is time-consuming and costly in terms of organization. It is subject to controversy because of unproven benefits on morbidity and mortality associated with a significant impact on the continuation of the pregnancy.

Method: from 2003 to 2009 we retrospectively analyzed all pregnancies and children aged less than 1 with a diagnosis of CHD in our tertiary center database. Our study population was then limited to serious or complex CHD: lethal cases, leading to medical termination of pregnancy (MTP), and CHD requiring surgery, interventional catheterization or hospitalization during the first year of life. Primary endpoint was 1-year mortality among alive neonates.

Results: 322 severe CHD were included. 62.1% had a prenatal diagnosis with an excellent screening predictability of the heart defect severity. We observed significant differences between prenatal (group 1) and postnatal (group 2) CHD diagnoses comparing: type of heart disease (hypoplastic left heart syndrome 7.8% vs. 0.8%), frequency of ducal-dependent heart defect (34.3% vs. 28.7%) and association with chromosomal abnormality or malformation syndrome (31.1% vs. 28.8%). We counted 96 MTP of 200 prenatal diagnosis (48%). Among the 224 alive neonates 15.2% died before the age of 1. Mortality at 1 year and major prognosis morbidity variables were not significantly different in both groups.

Conclusion: in our center prenatal diagnosis of severe CHD has an impact on the decision of MTP but not on prognosis in terms of 1-year mortality and morbidity. We recommend prospective multicenter studies with assessment of neurological prognosis and quality of life of patients. These studies will be facilitated by the use of standardized CHD registries.

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Impact of antenatal diagnosis on outcomes of neonates with hypoplastic left heart syndrome

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The objective of this study was to assess the influence of antenatal diagnosis of HLHS on postnatal outcomes.

Material and methods: This is a retrospective review of French single-centre records of neonates with diagnosis of HLHS. Clinical data, therapeutic management, outcomes were assessed. Comparison were made between groups with and without antenatal diagnosis, and between periods before and after 2005 (date of initiation of Norwood Program).

Results: Among 95 neonates (56 males= 59%) with HLHS, 44 (46%) were diagnosed prenatally (PreND group) and 51 (53%) after birth (PostND group). Birth weight was 3.0±0.5 kg, Age at diagnosis in PostND group was 4±6 days. All patients were free from symptom in the PreND group, while PostND group cases presented with heart failure (42%), cardiogenic shock (24%), cyanosis (12%), heart murmur (8%) or associated symptoms (14%). Age at Norwood procedure was 6±2 days in PreND and 9±4 days in PostND (p< 0.01). Time from diagnosis to surgery did not differ between groups (mean 6 days). Survival was similar between groups. Because of less termination of pregnancy, 71% (22 of 31 cases) of neonates with HLHS were diagnosed prenatally since 2005, compared with only 34% (22 of 64) before 2005 (p< 0.01).

Conclusion: Antenatal diagnosis may prevent acute postnatal cardiac failure in neonates with HLHS and allow early Norwood procedure, but it does not impact on long-term postoperative outcome.

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Echocardiographic right ventricular function variables: external validity study in a normal pediatric population

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Introduction: regular echocardiographic assessment of right ventricular (RV) function is required in patients with congenital heart disease (CHD). Although normal values are validated in the adult population, few studies are published in pediatrics. The aim of this study is to define in real clinical situ-ation normal values for RV variables in children and to compare the results to published studies.

Methods: prospective multicenter study from 2010 to 2012. 314 normal chil-dren aged 2 days to 18 years included (46% female, 88 infants under 1 year old, 26 neonates, 226 children). Following RV variables were collected by 4 pediatric cardiologists, using 2 ultrasound systems (Philips IE33, Aloka Alpha 10): tissue Doppler imaging (TDI) E’, A’ and S waves at tricuspid valve, TDI Tei Index and TAPSE. The impossibility to measure a variable was informed. A univariate poly-nomial linear regression was used to assess the relationship between these indexes and the anthropometric factors via the coefficient of determination.

Results: more than 90% variables could be easily collected. Mean values for E’, A’ and S waves were: 13.7 ± 3.8 cm/s, 10.1 ± 3.7 cm/s, and 12 ± 2.2 cm/s; mean values for TAPSE were 18.7 ± 4.9 mm and 0.41 ± 0.11 for DTI Tei index. In infant, E’, A’ and S waves were best correlated with BSA, and TAPSE with height. In children S wave and TAPSE were best correlated with weight, and A’ with age. No significant correlation was found for E’ wave in children, and for DTI Tei index. No significant differences were found between genders, ultrasound systems. Values were mostly well correlated to published studies.

Conclusion: we established echocardiographic RV function reference values in children with good feasibility and correlation to published reference studies. Routine application to children with CHD must be defined.

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Feasibility and reproducibility of tissue motion annular displacement of mitral valve in children with and without heart disease

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Objectives: Little is known about parameters of systolic left ventricular (LV) function using speckle tracking (ST) in children. The aim of this study...
was to define the feasibility (F) and reproducibility (R) of tissue motion annular displacement of mitral valve (TMAD) in healthy children (HC), in children with heart disease (HD) and to assess the correlations between TMAD and usual LV systolic function parameters.

Methods: We prospectively included 22 children with HD (13 boys, mean age 104 months) and 22 HC (12 boys, mean age 116 months). In an apical 4 chambers view 1 region of interest (ROI) was placed at the septal and lateral parts of the mitral annulus and 1 at the apex. The displacement of the midpoint between the 2 annular ROIs toward the apex was calculated by QLAB 9® software.

Results: TMAD indexed F was 100% (CI95% 94,8-100%) in children with and without HD and 86% (CI95% 71,5-100%) in HD. F of TMAD was not significantly different between HC and children with HD. Intra observer variability (V) of TMAD was respectively 19% and 11% in HC and in children with HD. Inter observer V of TMAD was respectively 19% and 16% in HC and in children with HD. TMAD was not correlated to age nor to body surface area (BSA) nor to LV ejection fraction in HC whereas indexed TMAD was correlated to stroke volume (r=0,591, p=0,0122), cardiac index (r=0,532, p=0,0241), indexed TAPSE (r=0,691, p=0,0034) and conversely correlated to end-diastolic (ED) LV diameter (r=0,677, p=0,0041), to EDLV volume (r=0,629, p=0,0076) and to end-systolic LV volume (r=0,616, p=0,0090) in HC.

Conclusion: TMAD seems an easy measurable marker with an excellent F and R to assess the mitral annular displacement. It seems independent of BSA and well correlated with stroke volume. The advantage of TMAD over Tissue Doppler imaging relies on the independence on angle. TMAD is an interesting tool in children. Its accuracy to estimate systolic function needs to be further investigated in children.

Folded melody valve technique for complex right ventricular outflow tract
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Introduction: Percutaneous Melody valve implantation has achieved standard of care for the management of patients with dysfunctional right ventricle (RV) to pulmonary artery (PA) conduits. The landing zone right ventricular outflow tract (RVOT) dimensions may vary significantly making it unfavorable for Melody valve implantation. We report a novel technique in patients with complex RVOT anatomies.

Methods: All patients were candidates for Melody insertion were evaluated with angiography. The landing zone right ventricular outflow tract (RVOT) dimensions may vary significantly making it unfavorable for Melody valve implantation. We report a novel technique in patients with complex RVOT anatomies.

Results: From 2008 to 2012, four patients received a shortened Melody. The uncovering, crimping and loading of the Melody valve was technically simple in all. The folded and crimped valve moved easily in the delivery system and the deployment was done using standard technique. All valves were positioned where intended. Patients were discharged the day after the procedure. Evaluation showed excellent performance of the Melody valve with no paraprosthetic leak, no erosion, no perforation, no stent fracture, no residual stenosis, no valvular or para valvular regurgitation immediately after implantation or at follow up.

Conclusion: The “Folded valve technique” is a safe addition to the interventional armamentarium allowing the implanting physician to modify the valve in patients with complex RVOTs. In the future, this technique may also be an option for patients with vulnerable RVOT neighborhood that may preclude conventional technique.

Surveillance of pediatric heart transplant patients in France: a national survey

Introduction: Rejection following cardiac transplant remains a important cause of morbidity and mortality as well as the complications of immunosuppressive therapy. Thus, surveillance of pediatric heart transplant (PHT) patients is crucial to prevent these risks or, at least, to allow an early treatment. However, very few international guidelines have been established concerning the modalities of this monitoring.

Aims: To collect and to compare the different practices of PHT surveillance in France.

Patients and Methods: Descriptive multicenter study. Each french cardio-logic center involved in PHT was contacted by e-mail in order to complete an electronic questionnaire.

Results: 8 centers were involved in the surveillance (including 7 centers performing PHT) of these patients. The average number of followed PHT
patients was 16.1 (2-50) per center. The average number of involved physicians was 3.7 (1-8) including 38% of pediatric cardiologists. Only two centers had a devoted nurse for this activity. Echocardiography was considered to be reliable for the early detection of transplant rejection by only 57% of centers. IVRT was always collected. Whereas TM measurements were frequently analyzed, other Doppler measurements were inconsistently reported. Myocardial strain analysis (using speckle tracking method) was almost never performed (14%). Coronarography was systematically performed in 43%; coronary CT angiogram in 29% and cardiac MRI in 14%. For patients aged >1 year, cardiac biopsies were systematically performed in 86%. The prevention of the transplant coronary artery disease was conducted using pravastatin in 86%, aspirin in 28% and clopidogrel in 14%.

Conclusion: The French practices for the monitoring of PHT patients are highly heterogeneous due to the absence of national recommendations. This study highlights the need of a national register in order to establish consensus for the management of these patients.

Intraoperative use of a new micro multplane transoesophageal probe in neonates with congenital cardiac disease: preliminary results

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Background: Transoesophageal echocardiography (TEE) has become a critical diagnostic and perioperative management tool for patients with congenital heart disease. The use of dedicated paediatric TEE probes is however not recommended in children below 5 kg which make it not available for neonatal cardiac surgery.

Aim: To assess the feasibility and the quality of imaging modalities of a new micro multiplane transoesophageal echocardiogram probe during repair of congenital heart defects in neonates.

Method: Prospective study of micro transoesophageal echocardiogram S8-3t probe used at a single institution.

Results: A total of 15 perioperative studies were performed on 15 patients, with a median weight of 3.52 kilograms (2.7-4.5 kg) and a median age of 34 days (2-150 days). We experienced none of the usual complications related to probe insertion including difficulty with probe insertion, airway compression, displacement of an endotracheal tube or damage to surrounding structures. We obtained good two-dimensional imaging quality in all patients. Pulse wave and continuous wave Doppler was consistently good across all examinations allowing complete pre and post-operative assessment of cardiac anatomy and function.

Conclusion: The use of micro multiplane transoesophageal echocardiogram probe is safe and feasible, and provides imaging of good quality in neonates. It may become an additional tool in routine for the surgical management of neonates with complex congenital heart defects.

Factors determining the nature of progression of discrete fixed subaortic stenosis

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Introduction: In discrete fixed subaortic stenosis, surgery is indicated when the systolic gradient (Gmax) between the left ventricle (LV) and the aorta exceed 50 mmHg or in the presence of significant aortic regurgitation (AR).

The aim of this study is to determine the factors that influence the progression of the obstruction and the appearance of AR.

Methods: retrospective serial echocardiographic review of 19 patients, mean age 16 years (2 years-38 years), with fixed discrete subaortic stenosis that don’t require surgery ( initial Gmax at inclusion <50 mmHg and without any symptom). The mean follow up was 5,42 years. The progression of gradient is defined by the formula (Gmax at follow up – initial Gmax).

Results: The mean velocity of increasing of Gmax was 2 mmHg/year. This progression was correlated to the patient’s age (cut off=15 years, r=0.5, p=0.02), and the initial value of the Gmax (cut off=40 mmHg, r=0.43; p=0.04).

The appearance or the aggravation of aortic regurgitation was determined by: the initial grade of AR (r=0.64 ; p=0.003), initial Gmax (r=0.65; p=0.002), progression’s velocity of Gmax (r=0.47; p=0.04), and distance between the membrane and the aortic cusps (cut off=5 mm, r=0.49 ; p=0.03).LV hypertrophy was influenced by the velocity of progression of obstruction (>2 mmHg/year).

Conclusion: the identification of factors determining the evolution of discrete subaortic stenosis (age < 15 years, initial Gmax > 40 mmHg, distance membrane- cusps > 5 mm) allow an adequate screening of patients that will require early operation.