in children with PAH. However, its role as predictors of outcome value has been poorly characterized. We aimed to characterize RV function in children with idiopathic PAH (iPAH) according their clinical status, and to assess the prognostic value of several echocardiographic indices of cardiac function.

Methods and results: Clinical, biological and echocardiographic variables were assessed in 39 children (28 girls) with iPAH (type 1, Dana Point classification). Patient's median age at inclusion was 5.8 years old 95%CI [2.9 – 10.4] and the median follow-up was 12.8 months. Forty-seven echocardiographic scans were performed in children at time of worsening defined by NYHA=III or IV and/ or history of syncope and/or RV failure, and 226 echocardiographic scans were performed in stable clinical status defined by NYHA≤II, without syncope and without RV failure. Outcome events were defined as changing from stable to worsening clinical status, death, transplantation, and Potts intervention. Patients in worsening status were significantly younger and had lower body mass index (median age 5.7 years old, BMI=15 kg/m²; p<0.01). TAPSE, RV peak systolic myocardial velocity, pulmonary ejection time, aortic and pulmonary velocity-time integral were significantly reduced in this group (p<0.01). LV filling was also impaired in worsening status group: mitral early (E)/late(A) ratio and LV myocardial early diastolic velocities (mitral E') were significantly reduced (respectively p=0.02 and p<0.01). TAPSE was the strongest predictor of events in multivariate Cox regression analysis (HR=0,80, 95%CI[0,75 to 0,86, p<0,01), and a TAPSE value ≤18mm was strongly predictive of clinical outcomes (area under curve 0,82).

Conclusion: Echocardiographic parameters of RV function and LV filling were impaired in children with iPAH. TAPSE can predict adverse cardiovascular events. These parameters could be used in the follow-up of children with iPAH, by comparing serially within a given patient.

0484

Outcome of coronary artery lesions after neonatal arterial switch operation

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Arterial switch operation (ASO) for transposition of the great arteries (TGA). Outcome and treatment of CL after ASO remains an unresolved question.

Objective: To study the long-term outcomes of CL after ASO.

Methods: We identified 75 (6.3%) patients with CL in our database of ASO that covers a period of 30 years. CL were either ostial or proximal, and involved the left main artery in 34 patients, the left anterior descending artery in 19, the circumflex artery in 10 and the right coronary artery in 12 patients. 35% of patients were symptomatic and diagnosed at time of an ischemic event. 65% of patients were asymptomatic and diagnosed during a systematic screening. Myocardial ischemia (MI) was demonstrated in 45% of the asymptomatic patients.

Results: First intention treatment was coronary revascularization in 32 patients (43%) (surgical angioplasty=25, graft by-pass= 3, percutaneous balloon dilatation= 5), medical treatment in 15 (20%), and surveillance in 25 (33%). Three patients died before any treatment. Mean follow up was 10.6±7.9 years. Survival was 90% at 20 years. A second intention treatment was needed because of a new anatomical lesion or new onset MI in 27% of patients who received medical treatment as first line therapy, in 20% of patients who were not treated, and in 12.5% patients who underwent revascularization. Overall, revascularization was performed in 73% of symptomatic patients, 72% of asymptomatic patients with MI, and in 22% of asymptomatic with no MI at diagnosis. At last follow-up, one patient has a residual MI.

Conclusion: Coronary lesions after ASO are not uncommon. In patients with MI, revascularization seems to be the treatment of choice. In non-ischemic patients at diagnosis, early revascularization needs to be considered in light of the severity of the lesion and MI can appear during follow-up.

0490

Long term follow-up of repaired aortic coarctation between 1978 and 2012. A single centre cohort

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Background: Repaired aortic coarctation (CoA) is associated with high long-term mortality and morbidity. Despite optimal early correction, patients with CoA remain at high risk of hypertension and death. We evaluated outcomes and follow-up of all patients who underwent CoA surgery, and alive at 30 days, between 1978 and 2012.

Methods: Retrospective cohort study in a single university teaching hospital. All data were obtained from medical records. Vital status, medical history and results of imaging were analyzed

Results: A total of 85 patients (63 men) were included. Median age (range) at surgery was 69 (2-13356) days; 19 (2-54) years at examination. Mean follow up was 15.8 years (1343 person-years). No deaths were observed. Re-intervention was common (31%) and most often due to ReCoA (13 patients, 16%). In case of ReCoA, 8 patients had thoracic surgery, and 5 percutaneous angioplasty with stenting. Half (53%) had a bicuspid aortic valve, associated with aortic valve regurgitation (21 cases). Aneurysm of the ascending aorta was diagnosed in 8 patients, without surgical indication. Prevalence of hypertension was high (18 pts, 21%); no related complications were noted; 17 patients were being treated. All patients had normal LV ejection fraction, but half (56%) had increased LV mass. In total, 28 MRI were performed. MRI showed 22 Romanesque aortic arches, 2 Crenel arches and 1 Gothic arch. Regarding follow-up, 66% were followed as recommended by ESC guidelines. Only 4 patients were lost to follow up. There were 15 pregnancies (8 patients); no complications were noted.

Conclusions: Primary repair of CoA was performed with low rates of mortality and morbidity. However, prevalence of hypertension, aortic valve abnormalities and ascending aorta aneurysms remain high, confirming CoA as an overall vascular disease. Furthermore, many patients required further reoperation. These findings emphasize that patients with CoA need lifelong follow-up.

0534

Antenatal echocardiographic parameters to predict postnatal outcome of neonates with Ebstein anomaly

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Ebstein tricuspide valve anomaly is a rare CHD with uncertain postnatal prognosis. Criteria to predict outcome are still a matter of debate. The aim of this study was to determine antenatal echocardiographic predictive parameters.

Methods: Retrospective multicentric analysis of fetus with diagnosis of Ebstein anomaly. Echocardiographic measurements of ventricles, atria, great vessels and tricuspid regurgitation were collected. Comparisons were made between group I (poor outcome= death occurred in utero or within the first 3 months of life) and group II (favourable outcome: postnatal survival >3 months).

Results: 16 fetuses were included in the study: 10 in group I (62.5%: 2 TOP, 2 fetal deaths, 6 postnatal deaths) and 6 in group II (37.5%). Mean gestationnal age at diagnosis was 29weeks (22 to 38). The mean number of echocardiographic records per patient was 2 (1 to 6). LV to RV ratio, tricuspid valve regurgitation grade and retrograde or anterograde ductal flow did not differ between the 2 groups. Significative differences were found between groups I and II regarding the presence of pulmonary flow (none or mild RV to PA flow: 8 of 9 cases died= 89%), AO to PA ratio (75% death if > 97°p vs 25% if 3-97°p), RA diameter (77.3% death if > 97°p vs 0%), PA diameter (100% death if < 3°p) and pericardial effusion (80% death vs 0%). Only 1 case had arrhythmia and died.

▶

Conclusion: this small sample size study showed that the absence of RV to PA flow and/ or pulmonary valve opening, increased AO to PA ratio, RA and decreased PA diameter and the presence of pericardial effusion might represent prognosis factors in fetus with Ebstein anomaly. These results should be confirmed by large scale prospective study.

0328

Evaluation of coronary artery wall echogenicity in Kawasaki disease acute phase

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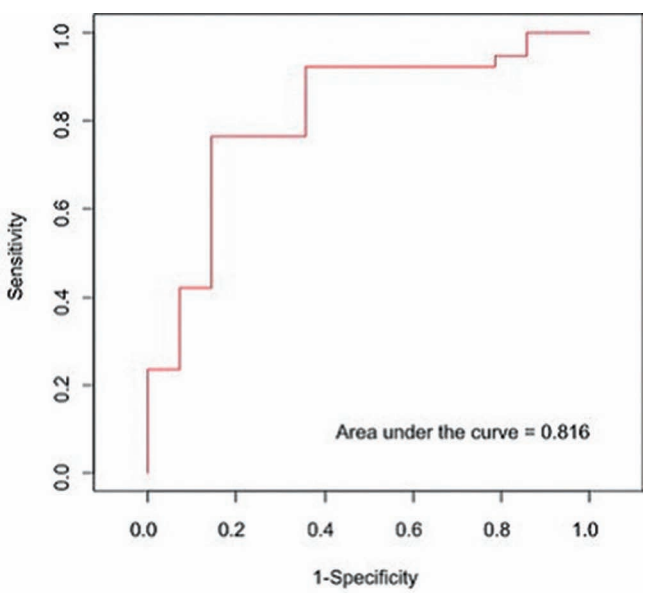
Background: Kawasaki Disease (KD) diagnostic is based on clinical, laboratory and echocardiographic criteria. Coronary artery brightness has been proposed as a criterion for diagnosis KD at the acute phase, but it is qualitative and subjective. Evaluation depends on the experience of individual echocardiographers.

Objectives: Diagnostic value of quantitative evaluation of echogenicity of the coronary artery wall (CAW) in KD at the acute phase.

Methods: A retrospective case-control study was performed in Clermont-Ferrand university hospital. Echocardiograms of 0 to 18 years old children between January 2012 to January 2014 was enrolled: 19 KD confirmed, 14 acute febrile illness and 34 apyretic children with congenital heart disease without coronary disease. The value of echogenicity was evaluated in decibel on $2mm^2$ Regions Of Interest (ROI) using Q LAB software -Philips-. For each examination, precordial short axis cross-section at level of aortic valve, 3 ROI are placed at the coronary artery environment, 3 ROI at the proximal segment of the left or right CAW, and 3 ROI at the aorta. Quantitative evaluation of echogenicity of CAW was performed with the measurement differences between ROI. Intraobserver variability was 0.8.

Results: Echogenicity of right CAW was significantly higher in the acute phase of KD than the other groups – p = 0.004, area under curve ROC = 0.8 –. Also it has been observed for left CAW – p = 0.17 –.

Debate and outcome: This first study highlights the interest of echogenicity quantitative measure of CAW for KD diagnostic. Echogenicity of CAW might be helpful in diagnostic for atypical or incomplete KD. The reproducibility has to be confirmed by a prospective study with more children suspected of KD.



Abstract 0328 – Figure: Area under curve ROC of the CAW echogenicity

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0377

Atrial septostomy in very low birth weight infants

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The interventional management of D-Transposition of great arteries D-TGA is more required with the advances in NICU and pediatric cardiology.

Objective: To describe our experience in balloon atrial septestomy (BAS) in D-TGA patients weighing < 1500 g.

Methods: we reported retrospectively all BAS in VLBW infants from January 2002 to June 2013. We collected information about the vital parameters before and after BAS. We described the atrial septestomy.

Results: We have 3 patients with a mean weight of 1333 g. D-TGA was diagnosed at a mean age of 3 days. Four atrial septestomy attempts were done at a mean age of 10.3 days. One infant had two attempts with balloon dilatation in the first, then BAS 24 h later. Procedures were done in catheterization laboratory in three cases and in NICU in one case. Patients were ventilated, sedated and transported by the NICU team. Venous access was obtained by the femoral vein puncture in two cases, femoral vein cutdown in one case and by the umbilical vein in one case. BAS were performed successfully in two cases, one using 4Fr. Swan Ganz catheter, and the other with 5 Fr. Z-Med septestomy catheter. BAS failed in two procedures, one because of atrial wall perforation, and the second because of restrictive foramen ovale which was dilated by a coronary balloon. Complications occurred in 3 cases, one minor (balloon rupture), and two major; atrial perforation in one patient, and severe bradycardia in one patient. BAS was effective with sustained clinical improvement. BAS failure in two patients was followed by heart failure in one patient and the death in the other.

Conclusion: BAS is mandatory in VLBW infants with D-TGA and restrictive atrial communication. It should be performed quickly if possible to use the valuable umbilical access, and to avoid the clinical compromise. A hole-end BAS catheter is required for successful procedure. FO balloon dilatation constitutes another life-saving option.

0397

Cardiac remodeling and factors determining occurrence of atrial arrhythmia after surgical closure of atrial septal defect in adults

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Objectives: the purpose of this study was to assess cardiac remodeling and to determine factors predicting the occurrence of atrial arrhythmia after surgical closure of atrial septal defect (ASD) in adults.

Methods: Retrospective study including 33 adult patients (>20 years old, mean age: 34±11 years, 26 women) who underwent surgical closure of secundum or sinus venosus ASD. Before operation, all patients had dyspnea and 15% were in NYHA III-IV. Sinus rythm was present in 85% of patients. The ratio of pulmonary to systemic blood flow was calculated, yielding a mean of 2,8±1 and pulmonary artery hypertension(PAH) was observed in 80% of patients with a mean value of 41,3±10 mmHg. The ASD were closed by pericardial or Dacron patch in 97% of cases.

Results: operative death was observed in 2 cases. In survival patient, with a follow-up of 97± 57 months, regression of right ventricular dilatation and PAH occurred in the first post-operative month and was maintained at late follow-up. Atrial arrhythmia occurred in 4 patients and were determined by older operative age (p=0.003) and the absence of cardiac remodeling after surgery.

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