Conclusion: surgical correction of ASD in adults is safe and efficacious. Cardiac remodeling after ASD closure in the adult is a common and an early event and prevents late morbidity which is in most cases due to arrhythmias. The mode of closure does not seem to significantly impact remodeling.

0459

MRI evaluation of coronary anatomy and myocardial perfusion after arterial switch for transposition of great arteries

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Background: Coronary arteries obstruction is the main cause of mortality and morbidity in patients who underwent arterial switch for transposition of great arteries. Long-term outcome of coronary transfer and its consequences on myocardial perfusion is scarcely known. Objective: To evaluate feasibility of cardiac MRI to describe coronary anatomy, myocardial perfusion and fibrosis after arterial switch operation.

Methods: 110 patients (mean age 13.5 y) were included. Twenty-five/110 had had previously documented coronary artery obstruction. cMRI protocol included cine SSFP in short axis, two-chamber, three and four chamber view, and perfusion analysis before and after diprydiamole infusion. Anatomy was evaluated by 3D heart sequence in 78 patients. Finally, late enhancement was evaluated ten minutes after injection of contrast agent in 71 patients.

Results: Perfusion could be evaluated in all patients. One perfusion defect was identified. All patients with prior negative myocardial ischemia test had normal perfusion on cMRI even those with mild to moderate coronary stenosis. Anatomical evaluation of proximal coronary arteries was possible in 71/78 patients. Finally, we found limited myocardial fibrosis in only two/78 patients who had left coronary artery obstruction that had been repaired.

Conclusion: cMRI is feasible and gives comprehensive information on coronary artery anatomy and physiology after the arterial switch operation. Use of cMRI as a screening tool for late coronary artery obstruction should be evaluated in larger series of patients to estimate its sensitivity and specificity.

0462

Chest CT findings in idiopathic and heritable pediatric pulmonary arterial hypertension

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Objectives: To describe CT findings in paediatric idiopathic pulmonary arterial hypertension (PAH).

Methods: Lung and cardiac CT of 30 children with idiopathic PAH or heritable PAH (median age 7.5 years (range 11 months – 15.5 years)) were compared to 30 children without cardiac or lung disease matched for age. PAH diagnosis was always confirmed by a right heart catheterization. All patients were considered as having idiopathic or heritable PAH after a complete diagnostic work-up according to the Nice international recommendations. Seven/30 patients had mutations in one of the known PAH genes (BMPR2, Alk1, Tbx4, Eif2ak4). 

Results: CT findings were significant increase of the ratio of main pulmonary artery/ascending aorta (median 1.5 [1.5-1] versus 0.98 [0.7-1.1]) in controls and right ventricle/left ventricle (median 1.4 [0.8-2] versus 0.8 [0.6-1]). Mediastinal adenopathies were observed in 9 cases. Parenchymal anomalies were present in 25/30 patients with mosaic pattern of attenuation in 9, ground glass nodules in 17, micronodules in 12, interlobular septal thickening in 13, consolidation in 3, and distal pulmonary arteries anomalies in 16. No correlation was found between CT findings and WHO functional class or hemodynamic characteristics of patients. Veno-occlusive disease was diagnosed at CT in 3 patients and confirmed subsequently either genetically or histologically. Two/4 diagnosis of Rendu-Osler (ALK1 mutations) were diagnosed at CT before genetic confirmation.

Conclusion: CT is a valuable tool in the diagnostic work-up of PAH in children. Specific parenchymal anomalies are highly suggestive of EIF2AK4 or ALK1 mutations and may orientate genetic confirmation of the disease.

0528

Long term follow-up and prognosis of anomalous origin of left coronary from pulmonary artery

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The aim of the study was to assess the long-term outcome of patients with ALCAPA

Methods: Retrospective single-center analysis of patients who underwent surgical correction of ALCAPA from 1980 to 2012. Surgical techniques, demographics, echocardiographic parameters and outcomes were collected. Patients were divided into group I <2 years at diagnosis, group II: >2 years.

Results: 48 patients (28 females) were included, median age 6 months (min 4mos, max 65y), median weight 6kg (min 1.9kg); 36 in group I and 12 in II. HF was patent in 39 patients (mean age 4y), 9 were asymptomatic (mean age 1y). Qwave on ECG was present in 87% of cases. Mean LVSF=24.2% (group I= 26.6% vs II= 37.7%, and 16.6% in patients <6mos of age), LVEDD and LVEF Z-scores were respectively +2 and +5 in groups I and II; 73% had MR: severe in 6%, moderate in 46%, mild in 21%. Left coronary artery ostium located in the left posterior sinus in 31cases, right posterior sinus in 12 and in right pulmonary branch in 4. Direct coronary artery reimplantation was performed in 71%, Takeuchi technique in 6%, Meyer technique in 20% and LCA ligation in 3%. Mean age at surgery was 29mos, mean weight 9kg. Post-operative mechanical circulatory support was required in 3 cases, who had more severe HF, lower LVSF and longer bypass duration. Mean FU was 8mos (6 to 31mos). Freedom from reoperation was 100% at 1y, 91% at 10y and 88% at 20y. LVSF increased by 20% in the early postoperative course and 36% at late FU. MR improved significantly in most of the cases. Overall mortality was 33% (15 in group I died before Day-30, none in group II), decreasing over time from 55% to 11%, and was lower in patients who had direct reimplantation. Q wave disappeared in 82% of the cases; 95% of survivors were asymptomatic at latest evaluation.

Conclusion: ALCAPA patients have good long-term survival and outcome. Age >2y at diagnosis and direct implantation are factors of favourable prognosis.

0535

Percutaneous valvuloplasty versus surgical valvulotomy in neonates with critical aortic stenosis

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This study was to compare outcomes and prognosis of newborns with critical aortic valve stenosis (AVS) undergoing either percutaneous or surgical intervention as first therapeutic option.

Methods: Neonates with diagnosis of critical AVS (defined as aortic flow dependent on ductus arteriosus patency) were included in the study, and divided in group I (percutaneous valvuloplasty) and group II (surgical valvulotomy).

Results: From 2000 to 2013, 23 cases (19 males) were analyzed: 9 in group I and 14 in II, aged 0 to 28 days at diagnosis (< 7 days of age: 78% of group I vs 50% in II, p< NS). Weight was > 2.5kg in 100% of group II and 78% of group I. Symptoms of heart failure were more frequent in group I.