0141
Spectrum of congenital heart disease in Moroccan patients with Down’s syndrome
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Background: Down’s syndrome is the most common chromosomal abnormality caused by trisomy of chromosome 21; this anomaly is frequently associated with congenital heart defects (CHDs).

Objectives: The aim of the study was to identify types, frequency and outcomes of CHD associated with Down syndrome.

Methods: This retrospective descriptive study was conducted in pediatric cardiology department of the University Hospital IBN ROCHD of Casablanca in Morocco, based on the CHD registry identified between 2008 and 2014.

Results: 2156 patients with CHD were identified including 128 patients with Trisomy 21(6%). The median age of diagnosis was 9.5 months (Min: 2d-Max: 16 years), the sex ratio was 1. The most common CHD was the atrioventricular septal defect (AVSD) (42.2%), followed by ventricular septal defect VSD (31.3%) then atrial septal defect ASD (28.9%) and patent ductus arteriosus PDA(24.2%). The most common associations of CHD were: AVSD+ASD (10%), VSD+ASD (7.6%), VSD+PDA (7.6%), AVSD+PDA (7%) and ASD+PDA (6%). The indication for surgery was the most common treatment’s modality (54.3%) and the total mortality was 20%.

Conclusion: In our study, the spectrum of congenital heart disease in Down syndrome was similar to the one described in the literature, and also in our Moroccan patients the AVSD was the most described abnormality, the mortality rate in this population remain high therefore we should insist on the early screening of congenital heart disease in this specific population.

0104
Surrogate echocardiographic parameters to assess right ventricular global systolic function in children with congenital heart disease or pulmonary hypertension
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Cardiac Magnetic resonance imaging (CMR) is considered the reference method for RV volumes and RVEF measurements. Three-dimensional knowledge-based reconstruction (3D-KR) derived from two-dimensional echocardiographic imaging is a novel technique. The aim of this study was to assess the feasibility and reliability of this novel echocardiographic technique in children with diverse CHD involving the right ventricle.

114 children (mean age 12.3 years, range 0.5 to 18) referred for cardiac MRI were included. Among them, 27 patients had barometric overload, 63 patients had volumetric overload, and 24 patients had mixed overload. Echocardiographic image acquisition was performed using a standard ultrasound scanner linked to a Ventrimed Medical Systems unit. Parameters analyzed were RV end-diastolic volume (EDV), RV end-systolic volume (ESV), and RVEF. The method of disks was used for CMR RV volumes. Feasibility of 3D-KR was 100%. Echocardiographic RV volumes correlated well with CMR (EDV, $r = 0.90$; ESV, $r = 0.87$; RVEF, ICC = 0.64). For intra-observer analyses, COV were 8% for EDV, 15% for ESV, and 17% for EF. For intra-observer analyses, COV were 4% for EDV, 7% for ESV, and 9% for EF. Characteristics of 3DKR as screening tool in detecting RV-EDV < 150mL were: sensitivity = 83%, specificity = 93%, positive predictive value = 90%, and negative predictive value = 88%. 3D-KR volumes are feasible and correlated well with CMR volumes in children. This new technique can be used as an accurate routine tool to assess RV function in CHD with barometric or volumetric overload and as a good screening tool in detecting patients with RV volumes < 150mL/m2 or RVEF >40% in repaired conotruncal heart defects.

0390
Vascular emergencies revealing a coarctation of the aorta in pregnant women
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Coarctation of the aorta is defined as a circular narrowing at the junction of the aortic arch and descending aorta, opposite the start of the ductus arteriosus. It represents 6-8% of congenital heart disease. It represents the first etiology of congenital heart disease in pregnant women. We report in this paper, two cases of coarctation of the aorta complicated in pregnant women, collected within the maternal resuscitation Lalla Meryem in Ibn Rochd UH center of Casablanca. The complications occurring during this condition makes the poor prognosis. Thus, the incidence of hypertension and pre-eclampsia is higher, putting them at risk of aortic dissection and rupture, rupture of cerebral aneurysm and heart failure. Infective endocarditis is rare but it is a serious complication. Preterm delivery is not common in coarctation of the aorta as well as Fetal growth retardation. While the incidence of miscarriages and abortions is much higher. The management of these patients is based on a good understanding of the specific risks to the coarctation of the aorta, pregnancy planning and a multidisciplinary approach involving the cardiologist, the anesthesiologist, the intensivist, the gynecologist obstétrien, the neonatologist and pediatrician. Research factor for cardiovascular risk is an essential part of monitoring of pregnancies. A preconception assessment and appropriate care throughout pregnancy are necessary to avoid a late diagnosis of this
Cerebral tissue oxygen saturation monitoring during balloon atrial septostomy in neonates with transposition of the great arteries. Preliminary data

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Balloon atrial septostomy (BAS) increases peripheral oxygen saturation in neonates with transposition of the great vessels (TGV). Effect of BAS on cerebral oxygenation remains little known. We aimed to describe the modification of regional cerebral tissue oxygen saturation (rcSaO²) during the catheterization.

Methods: we prospectively included 6 neonates with TGV and restrictive inter-atrial shunt who required BAS. BAS was performed in catheterization laboratory by an interventional pediatric cardiologist. rcSaO² was measured using near-infrared spectroscopy (NIRS) during the whole procedure.

Results: Median rcSaO² at the beginning of the procedure was 52.5% ranging from 21% to 78%. Median rcSaO² after the BAS was 69.5% ranging from 64% to 94%. The rcSaO² increased significantly immediately after the BAS (p=0.0273 by Wilcoxon signed rank test). Median rcSaO² delta between before and after BAS was 19% ranging from 11 to 43%. The rcSaO² delta was higher although not significantly when rcSaO² before the BAS was less than 50% (31% vs 16%, p=0.14). Linear regression analysis revealed that the delta of rcSaO² was significantly inversely related to the rcSaO² at the beginning of the procedure (Delta=-0.45 x rcSaO²av + 45.8, p=0.37, R²=0.70).

Conclusion: BAS improves cerebral oxygen saturation during the catheterization in neonates with TGV and restrictive inter-atrial shunt. The increase is proportional to the degree of alteration before the procedure.

Dissecting progenitor cell contributions to the developing heart

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Cardiac progenitor cells of the second heart field (SHF) contribute to the poles of the elongating embryonic heart. Perturbation of SHF development leads to a spectrum of congenital heart defects. Recent evidence suggests that distinct regions of the heart are pre-patterned in the SHF. For example the del22q11.2 or DiGeorge syndrome gene Tbx1 is required in the SHF for development of the inferior wall of the embryonic outflow tract, giving rise to subpulmonary myocardium. Characterization of the expression of an enhancer trap transgene at the Hes1 locus, encoding a transcriptional repressor, has identified a complementary Notch-dependent Hes1+ TBX1+ subpopulation of SHF cells giving rise to future subaortic myocardium. Using transcriptomic analysis we have characterized the genetic signatures of future subaortic and subpulmonary myocardium and identified Pparg among the genes enriched in future subpulmonary myocardium. Genetic and explant analyses have shown that Hes1 controls the molecular signature of future subaortic myocardium through direct transcriptional repression of Pparg. Our results reveal that distinct genetic regulatory networks control different progenitor cell contributions to the developing heart. We also investigated the potential role of Hes1 in the maintenance of residual SHF progenitors in the fetal heart. Our initial results have identified Hes1+ cells in the fetal heart and suggest that Hes1 deletion impacts negatively on residual progenitor cell numbers. Together, our study identifies a role for Hes1 in the regulation of cardiac progenitor cell fate and maintenance in the definitive heart of clinical importance for heart repair.

Supravalvular mitral ring: an underestimated form of congenital mitral stenosis

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Introduction: The congenital supravalvular mitral ring is a rare subtype of congenital mitral stenosis. This form is poorly understood and thus underestimated. It has a good prognosis with appropriate management. The identification of this entity remains unrevealed in the literature, only less than 100 cases were reported. The aim of this study was to share the diagnostic and therapeutic experiences of the Casablanca pediatric cardiology unit.

Methods: Between December 2008 and January 2014, 1927 patients with congenital heart disease were collected retrospectively in the Ibn Rochd cardiology department of Casablanca university hospital.

Results: 19 patients had congenital mitral stenosis (0.01% of the entire series), 8 of them were related to a supravalvular mitral ring. The sex ratio M / F was 1 : 1. The median age was 10 months [5-168]. For all patients, the diagnosis was made by echocardiography. The average mean mitral valve gradient was 14±8mmHg. Associated anomalies were: interventricular communication (n=3), coarctation of the aorta alone (n=1), coarctation of the aorta associated with a bicuspid aortic valve (n=2), Double Outlet Right Ventricle (DORV) (n=1) and a complex congenital cardiopathy (n=1). The surgery was indicated to 7 patients; the 8th patient had DORV with Eisenmenger. The supra-mitral ring was surgically resected to 4 patients with a good surgical outcome, 2 patients are awaiting for surgery and one patient was lost to view. No deaths per or postoperative were noted.

Conclusion: The supravalvular mitral ring is a rare variety of congenital mitral stenosis. It is often underestimated and potentially serious. The echocardiography is the diagnosis key. Surgical treatment is usually indicated. After resection, the prognosis is good without risk of recurrence and that improves long-term survival.