

Topic 38 – Congenital

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0141

Spectrum of congenital heart disease in Moroccan patients with Down's syndrome

Sanaa Benhaourech (1), Ayoub El Hammiri (2), Abdenasser Drighil (2), Rachida Habbal (2)
(1) *CHU Ibn Rochd, Cardiologie, Casablanca, Maroc* – (2) *CHU Ibn Rochd, Casablanca, Maroc*

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 atrio-ventricular 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 (7,8%), VSD+PDA (7,8%), ASVD+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

Olivier Guillard (1), Diala Khraiche (1), Magalie Ladouceur (1), Francesca Raimondi (1), Laurence Iserin (2), Damien Bonnet (1)
(1) *APHP-Hôpital Necker-Enfants Malades, M3C, Paris, France* – (2) *APHP-Hôpital Européen Georges Pompidou (HEGP), Paris, France*

In congenital heart diseases and pulmonary hypertension, right ventricle dysfunction is associated with prognosis. Surrogate parameters of RV systolic function are used in routine. However, their dependence to RV loading conditions has not been tested. Moreover, studies have suggested that these parameters are altered by cardiac surgery. The aim of our study was to assess the feasibility, reproducibility and relevance of surrogate echocardiographic parameters of RV systolic function in children with CHD involving the RV or pulmonary hypertension. We recruited 294 consecutive children with RV pathological loading conditions: 159 have barometric overload, 102 have volumetric overload, 33 have mixed overload. The median age was 9.2+/- 2.3. We assessed RV global systolic function by measuring the Fractional area change. We analyzed the feasibility, reproducibility and relevance of parameters of RV longitudinal systolic function: the Tricuspid Annular Peak Systole Excursion, RV 2 dimensional longitudinal strain and Tissue Doppler derived parameters: tricuspid systolic excursion velocity (Sa), myocardial acceleration during isovolumic contraction (IVA), and RV Tei index. TAPSE and Sa are the surrogate parameters with the best feasibility (>98%) and reproducibility (inter and intra observer variability<6%). 2D strain, IVA and Tei index have

worse feasibility and reproducibility. TAPSE and Sa were correlated to RV loading conditions. In patients with history of cardiac surgery, no parameter of longitudinal function was correlated to FAC or RVEF. Only in PH, TAPSE and Sa were correlated to RV global systolic function assessed by FAC (respectively r 0.84 and r 0.54, p<0.001). Parameters of longitudinal RV systolic function correlate with loading conditions. They cannot be used to predict RVEF in children with CHD or in children who had undergone cardiac surgery. They can only be used to predict RVEF in PAH.

0106

Knowledge-based 3D reconstruction compared to MRI for evaluation of right ventricular volumes and function in congenital heart diseases affecting the right ventricle

Olivier Guillard, Diala Khraiche, Francesca Raimondi, Magalie Ladouceur, Damien Bonnet
APHP-Hôpital Necker-Enfants Malades, M3C, Paris, France

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 Ventripoint Medical Systems unit. Parameters analysed 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, CC = 0.90; ESV, ICC = 0.87; RVEF, ICC = 0.64). For inter-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: sensibility = 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

Abdelaziz Hadadi, Hafdi Karim, Youssef Jarmoune, Rachida Habbal
CHU Ibn Rochd, Cardiologie, Casablanca, Maroc

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 ducts 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 Gynecologic 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