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 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 (27.8%). 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.

**Surrogate echocardiographic parameters to assess right ventricular global systolic function in children with congenital heart disease or pulmonary hypertension**

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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 years. 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 Anular Peak Systole Excursion, RV 2 dimensional longitudinal strain and Tissue Doppler derived parameters: tricuspid systolic excursion velocity (Sa), myocardial acceleration during isovolumic contraction (IV A), 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.