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 dell22q11.2 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: intraventricular 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 8st 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.