"Intracardiac Tumors in Neonatal Period: Report of 2 Cases of Myxoma"

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Document type : *Communication à un colloque (Conference Paper)*

C 05 • Emerging MRI techniques in cardiovascular research

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INTRODUCTION In cardiovascular research, imaging of small animals with high-frequency cardiac ultrasound is considered as a standard approach. However, its limited accuracy and reproducibility requires large sample sizes. The recent development of high-field MRI offers the potential to better define structural and functional changes in murine models of cardiovascular diseases.

AIM The purpose of our work was to evaluate left ventricular systolic function in mice using a 11.7 Tesla MRI scanner.

METHOD A total of eight C57Bl/6 wild-type mice were studied, including 3 animals having undergone a surgical transverse aortic constriction (TAC). During the entire MRI scanning, mice were anaesthetized with Isoflurane 1-3%, in a temperature-controlled setting. Scans were prospectively gated for electrocardiogram (ECG) and respiration. Imaging was performed on a 11.7 Tesla Bruker MR scanner. Cardiac scout images were obtained in the conventional planes (2-chambers and 4-chambers) with a tripilot sequence. A FLASH-cine sequence was applied to produce a stack of short-axis images covering the entire ventricles, perpendicular to the LV long-axis. The left ventricular (LV) systolic function was assessed from the full stack of short-axis cine images using the Segment software (Medviso, Sweden) by tracing epicardial and endocardial borders, including papillary muscles. The following volumes (µl) were determined: end-diastolic (LVEDV), end-systolic (LVESV) and stroke volume (LVSV). LV ejection fraction (LVEF, in %) and LV mass (mcg) were subsequently deduced.

RESULTS Accurate quantitative data were obtained in all animals. The mean LVEDV, masses and EF obtained in wild-type mice were 29.5±4.5 µl, 43.5±5.5mg and 68±12 % respectively. As expected, the mice with TAC presented significant LV hypertrophy, with a LV mass of 80.5mg±7.5mg ($p<0.05$), but unchanged LVEDV and EF. When compared to high-frequency ultrasound, preliminary data showed that inter- and intra-experiment reproducibility was likely better in MRI (e.g. almost identical LVEDV, LV masses and LV EFs (n=2-3)). Using light anesthesia with isoflurane, we also showed that mice survive the scanning procedure, and can be re-studied later on when appropriate.

CONCLUSION High-field cardiac MRI is a very promising tool to assess LV systolic function and hypertrophy in mouse models of cardiovascular diseases. Although the development of other sequences already used in the clinical field (i.e. tagging, velocity mapping or contrast-enhanced sequences) is still under investigation, we are confident that this technique will soon allow a non-invasive and reproducible approach of diastolic function and myocardial viability, as needed in translational cardiovascular research.

C 06 • Intracardiac Tumors in Neonatal Period: Report of 2 Cases of Myxoma

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INTRODUCTION Cardiac tumors are uncommon in children. Primary benign tumors are more frequent than malignant, and during perinatal period, rhabdomyomas are the most frequent tumors. Myxomas are exceedingly rare.

METHOD We report 2 cases of myxoma, diagnosed during fetal life and early neonatal period.

RESULTS A 29-year-old G1P0 woman was referred for evaluation of a heart mass detected at 21 weeks gestation. At fetal echocardiogram, the tumor appeared unique (7x5mm) and was localized at the apex of the right ventricle with a broad basis. The echogenicity was homogenous without calcifications and slightly hyperechogeneous compared to the neighbouring myocardium. According to power-Doppler, the tumor wasn't highly vascularized. The rest of the cardiac anatomy was normal, with normal biventricular function and sinus rhythm. At 27.4 weeks, the tumor was lightly increased with as only repercussion a mild tricuspid valve regurgitation. A possible multi-organic damage was excluded by ultrasound and fetal MRI. Fetal karyotype was normal and there was no deletion or duplication in both genes for tuberous sclerosis. A 4.4kgs full-term boy was born uneventfully. Clinical examination and ECG were normal. Postnatal echocardiogram defined the tumor as a sessile multilobed mass in the apex of the right ventricle. At 1-month age, MRI sequences led to the diagnosis of an isolated myxoma (17x11x12mm) without hemodynamic repercussion. Our second case was diagnosed in a 2-week-old girl, born full term with 3.5kgs, without any personal or familial history. During a check-up for cyanotic breath-holding-spells, an hyperechogeneous, homogeneous, sessile cardiac mass was shown in the apex of the left ventricle. The biventricular function, cardiac anatomy and electrical activity were normal. MRI suggested the etiology of a myxoma of oval form, 8x9x3mm. In both cases, conservative approach was adopted because of the tumor stability and the absence of symptoms, arrhythmia and hemodynamic compromise. Our two patients are currently well at respectively 5-month and 3-year follow-up.

CONCLUSION Myxomas are very rare tumors in the pediatric age. They may however already be present antenatally and should be part of the differential diagnosis of cardiac mass. MRI may confirm the diagnosis. Surgical abstention is conceivable, especially when the patient is asymptomatic.

C 07 • Self-expanding stent Sinus superflex Visual to create an unrestrictive atrial communicatio

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INTRODUCTION In infants with complex CHD an unrestrictive atrial septal communication may be necessary. Balloon septostomy and dilation is however sometimes inefficient in creating an adequate atrial shunt; blade septostomy requires a big sheath and balloon expandable stents may slide and embolise.

METHOD Procedure done in 6 infants requiring a stable atrial shunt after failed Rashkind or thick atrial septum predicting poor result. Access through a 5 Fr sheath in the femoral vein. A 0.014" stiff coronary wire is preferably curled in left atrium; balloon interrogation of atrial septum with a soft balloon (10 mm Tyshak, Numed); optimizing the beamer angulation (LAO-cranial) perpendicular to the atrial septum; positioning of Optimed Sinus superflex Visual® 5F 10/20 mm; opening the distal part in the LA, hand injection through sidearm of the sheath to delineate relationship with the septum; pull back if required; opening the stent completely; post dilation if required. Low dose acetylcalicylic acid is given at 2 mg/d.

RESULTS 6 infants age median 3.7 months (range 1.0 – 7.8); CHD: complex TGA (1), tricuspid atresia (1), small left heart (4). In all patients the stent was successfully deployed; the anti-jump technique and visual markers allowed perfect placement; the open cell design allows the stent to hook at the septum, preventing sliding or embolisation. In all patients the stent provided an adequate unrestrictive atrial communication until the next surgical step [switch (1) or Glenn (5)]. The timing of the next operation was elective and not imposed by inadequate atrial shunt. All stents could easily be removed by the surgeon. No evidence of any thombo-embolic event during a follow-up of 4.7 months (range 1.4-7.0). The stent was firmly attached to the atrial wall.

CONCLUSION Sinus superflex Visual® stent delivered safely through a 5F sheath allows to obtain an adequate atrial connection for several months in infants with complex congenital heart disease.

C 08 • Sternotomy As a Safe Alternative 'Access' for Interventions In Low Birth Weight infants

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INTRODUCTION Low birth weight infants with congenital heart defect remain a therapeutic challenge. Prolonged administration of prostaglandin has many disadvantages; the surgical options yield high morbidity and mortality; vascular access for percutaneous interventions may seriously damage vessels required for later procedures. We used sternotomy for hybrid direct cardiovascular access in low birth weight infants to perform a bail out stenting procedure.

METHOD 4 patients: hybrid suite; sternotomy; A/ 3 patients (weight 1620, 2190 & 2630 g) extreme Fallot – pulmonary atresia with hypoplasia pulmonary trunk (2-3 mm); purse string on the right ventricle; 2 vascular clips were placed as radio-opaque markers: one at the pulmonary valve annulus, one at the puncture site within the purse string. Double needle technique: 2 identical 21G needles: 1 needle as a reference adjacent on the surface of the RVOT to determine precise puncture depth and direction; the other needle was used to perform the punction. The punction was performed under direct vision in two motions: first access perpendicular to the surface for 10-15 mm into the right ventricular cavity with free blood, second angulation of the needle towards and advancement through the atretic outflow tract into the pulmonary trunk. A 0,014" coronary wire into the pulmonary arterial branch; needle exchanged for a 4 Fr short sheath; angio by mini 1cc injections through side-arm; a premounted coronary stent was deployed into the RVOT 5/16 mm to obtain an "intracardiac Sano shunt"; sheath and clips removed. B: 1 patient (900g) with critical aortic coarctation and open duct under PG; purse-string on the ascending aorta with radiomarker clip; puncture with 21 gauge needle; 0,014" wire into the descending aorta; needle exchanged for a 4 Fr short introducer sheath with the tip at the aortic cross; angios through side-arm. A 3/8 mm coronary stent deployed into the aortic isthmus (from LSA until beyond coarctation); sheath and clip removed, arterial duct clipped.

RESULTS Fallot: adequate palliation with good anterograde flow to the pulmonary arteries (sat> 92%). After a median of 3 months additional transvenous stenting required in all patients because of progressive muscular infundibular stenosis. Two patients evolved to full repair at 5 months; one patient with multiple hilar stenoses requires additional percutaneous procedures through the stented RV outflow tract. Coarctation: good aortic flow, stent resected at 4 months. No associated morbidity as frequently seen in premature infants with CHD.

CONCLUSION Medial sternotomy can be a safe alternative access for bail out transluminal cardiac interventions in low birth weight infants, allowing conventional repair at bigger weight. The technique with 2 identical needles and radio-opaque markers markedly simplifies the hybrid procedures.