Aortic root dilatation and stiffness assessed by magnetic resonance imaging in adults with repaired tetralogy of Fallot

Florence Pontnau*1, Magalie Ladoueucur2,1,2,3, Antonio Ferreira4, Laurence Isnard5, Elie Mousseaux5,3
1 Adult congenital heart diseases unit, Department of cardiology, APHP-Hôpital Européen Georges-Pompidou, Paris, France.
2 Pediatric cardiology department, Centre de référence des malformations cardiaques congénitales complexes, M3C, APHP, Hôpital Necker, Paris, France.
3 INSERM, U970, PARCC, Paris, France.
4 Department of radiology, Centro hospitalar de Lisboa Ocidental, Lisboa, Portugal.
5 Department of radiology, APHP, Hôpital Européen Georges-Pompidou, Paris, France.

* Corresponding author: florence.pontnau@egp.aphp.fr

Purpose We aimed to assess dimensions and biomechanics of the thoracic aorta in patients with repaired tetralogy of Fallot (TOF), using Cardiac Magnetic Resonance (CMR).

Background Aortic root dilatation frequently occurs in TOF and can lead to aortic regurgitation (AR), aortic aneurysms and its complications. Historically, studies in TOF have shown abnormalities of the aortic media that can predispose to aortic root dilatation.

Methods 50 patients (aged 29±12 years) with repaired TOF and 50 control subjects (aged 29±11 years) matched for age and sex underwent CMR imaging, with standard cine and velocity sequences. The aortic root dimensions were assessed at end-diastole at the following levels: aortic annulus, sinus of Valsalva, sinotubular junction (STJ), ascending and descending aorta. Aortic elastic properties were evaluated by aortic distensibility and pulse wave velocity (PWV). CMR included conventional left ventricle (LV) and right ventricle (RV) systolic function and volume study. AR fraction measure.

Results Diameters of the aorta indexed to the body surface area were significantly increased in TOF compared to controls at level of sinus of Valsalva (22.6±3.8 vs 17.0±2.0mm/m²; p<0.001), STJ (18.2±4.1 vs 13.3±1.7mm/m²; p<0.001) and ascending aorta (20.0±4.3 vs 14.3±2.0mm/m²; p<0.001). In contrast, the diameter at annulus and descending aorta was not significantly different. Ascending aorta has a reduced distensibility in patients compared to controls (3.4±2.2 vs 6.6±2.4 x10⁻⁶/mmHg⁻¹; p<0.001) and PWV in the aortic arch is significantly increased (8.1±6.6 vs 4.3±1.3 m/s; p<0.001). Unlike controls, PWV and aortic velocity were not correlated with age in TOF. AR occurs more often in TOF than in controls. Aortic dilatation and stiffness were not related to age at surgical repair. Both LV and RV systolic functions were moderately impaired in TOF.

Conclusions Aortic root dilatation is associated with increase in aortic stiffness. These parameters can be easily measured by CMR.

Conflict of interest The authors have not transmitted any conflicts of interest.

Cardiac phenotype and prognosis of patients with mutations in NKX2.5 gene

Philippe Maury†1, Alban Baruteau2, Estelle Gandjbakhch4, Francis Bes- sière1, Patrice Bouvagnet4, Florence Kyndt3, Sébastien Hascoët1, Sylvie Di Filippo2, Francoise Hidden-Lucet1, Philippe Chevalier3, Damien Bonnet1, Vincent Probst2, Alice Maltret5.
1 University Hospital Louis Pradel, Lyon, France.
2 University Hospital Necker, Paris, France.

* Corresponding author: mauryjphil@hotmail.com

Introduction Mutations in NKX2.5 gene explain familial forms of atrial septal defect (ASD) associated with atroventricular conduction disturbances and unexplained sudden death (SD) but cardiac phenotype has not been described in a large population of patients with NKX2.5 mutations.

Methods All successive patients with mutations in NKX2.5 gene were included, representing the whole population of French NKx2.5 mutated patients.

Results 47 pts carried NKX2.5 gene mutations (24 men, median 25 yo, 0 to 69) (20 unrelated families, 2.5±1.5 mutated subject/family). There was an history of SD in 9 and of pace-maker implantation in 5 families. ASD was present in 70% (surgically corrected in 67% and percutaneously in 2 pts) and ventricular septal defect in 15%. Conduction disturbances were observed in 82%. 13 pts (27%) developed complete or high degree AV block. Available ECGs showed PR interval of 219±43 ms, QRS duration of 86±15 ms and a QTC of 408±27 ms. Electrophysiological study was performed in 15 pts (3 had infra hisian and 5 suprhisian block).

A pace-maker was implanted in 20 pts (with ICD in 5) and a loop recorder in one. Sustained or nonsustained ventricular tachycardia were observed in 6 pts. Mean ventricular pacing % was 77±37. Six pts were dependent of the pace-maker. Three patients deceased over the follow-up (2 SD and one endocarditis). 13 pts developed paroxysmal or permanent supraventricular arrhythmias (mainly atrial fibrillation). Five pts displayed dilated cardiomyopathy, 3 had left ventricular (LV) hypertrophy and 4 with features of noncompacted LV. LV ejection fraction was normal except in 2 cases (35%).

Conclusion Carriers of NKX2.5 gene mutations harbor a rich phenotype associating most of the time ASD and/or VSD together with evolutive AV block leading to pace-maker/ICD implantation in a significant part of them. Associated LV cardiomyopathy is less frequent but ventricular arrhythmias appear common and SD may happen.

Conflict of interest The authors have not transmitted any conflicts of interest.

3D Transthoracic echocardiography assessment of the pulmonary valve in patients with TOF

Khaled Hadeed1, Sébastien Hascoët1, Romain Amadieu1, Yves Dulac1, Sophie Breining1, Philippe Aca1
1 Pediatric cardiology unit, Children’s Hospital, Toulouse University Hos- pital, France.

* Corresponding author: hadeed.k@chu-toulouse.fr

Background Accurate evaluation of pulmonary valve (PV) morphology and pulmonary annulus (PA) diameter is crucial before surgical correction of tetralogy of Fallot (TOF). Our aim was to assess PV morphology using three-dimensional transthoracic echocardiography (3D-TTE) in infants with TOF before surgical correction. And to compare PA diameter obtained by different imaging modalities.

Methods 30 patients with TOF were prospectively included. All patients underwent 2D and 3D-TTE, 23 patients underwent CT-Scan and 7 cardiac catheterization. PA diameter was measured using 2D-TTE in parasternal short axis view as recommended. 3D dataset was acquired using zoom mode at PV. Both vertical (Dv) and horizontal (Dh) diameters of PA were measured. Mean 3D diameter (3DD) was calculated as (Dv+Dh)/2. Eccentricity index (EI) of PA was calculated (Dv – Dh/Dv). These measurements were compared to CT-Scan and angiography when available and to perioperative measurements.

Results Mean age was 7.4 months (3-24 months), mean weight was 6.6 kg (4.5-13.5 kg). PV was described as bicuspid in 15/30 patients by 3D-TTE from en face view, with 75% agreement between 3D-TTE and perioperative finding (20/30 patients). PA geometry was slightly asymmetric by 3D-TTE. Dv was significantly larger than Dh (8.4 mm vs 7.4 mm, p = 0.001), and mean
P4

Right ventricular activation mapping to determine electrical activation pattern in patients with repaired tetralogy of Fallot

Emmanuelle Fournier1,*, Zakaria Jalal2, Frédéric Sacher1, Pierre Bordonar1, Hubert Cochet1, Michel Haissaguerre1, Jean-Benoît Thambo1

1 University Hospital of Bordeaux Haut-Lévêque, Service des pathologies cardiaques congénitales de l’enfant et de l’adulte, Pessac, France.

* Corresponding author: emmanuelle.fournier01@gmail.com

Background Patients after repaired tetralogy of Fallot (TOF) frequently have right ventricular (RV) dysfunction and prolonged QRS duration (QRSd), the latter is considered as a sudden death risk factor. It has been suggested that QRSd mainly reflects abnormalities of the RV outflow tract (RVOT) rather than the RV body itself. We aimed to better understand the RV electrical activation pattern in these patients using activation mapping.

Methods 51 Adults (33±13 yo, mean QRS duration 153±21ms) referred for either catheter ablation or pulmonary valve replacement late after TOF repair underwent a MRI, with fibrosis analysis, and an invasive RV activation mapping (201±32 sites per patient; Carto 3 – Biosense Webster). RV total activation time (RVTAT) was defined as the duration between the first and the last RV EGM.

Results The delay between QRS onset and earliest RV EGM was 28±23 ms translating the absence of RV parkinje activation and the left to right ventricle activation. We observed in all patients a single RV septal breakthrough (mid-septal in 79%, septo-basal in 14% and apico-septal in 7%) followed by 2±spreads of activation: a first wave from the septum to the RV anterior wall either catheter ablation or pulmonary valve replacement late after TOF repair underwent a MRI, with fibrosis analysis, and an invasive RV activation mapping (201±32 sites per patient; Carto 3 – Biosense Webster). RV total activation time (RVTAT) was defined as the duration between the first and the last RV EGM.

Conclusion RV delayed activation in patients with repaired TOF translates an homogeneous activation pattern that is not only the consequence of an infundibular disease but also reflects a slow conduction in the RV free wall.

Conflict of interest The authors have not transmitted any conflicts of interest.

P5

Children with tetralogy of Fallot exhibit accelerated maturation of the cardiac tissue into adult phenotype

Clément Karsenty1,*, Céline Guilbeau-Frugier2, Philippe Maury2, Atul Pathak2, Philippe Aecar1, Jean-Michel Senard2, Céline Gales2, Sébastien Hascoët2

1 Inserm U1048, Institut des maladies métaboliques et cardiovasculaires, I2MC, Toulouse, France.
2 Paediatric cardiology unit, Children Hospital, Toulouse University Hospital, Toulouse, France.

* Corresponding author: clement.karsenty@hotmail.fr

Background Pressure overload continuous from the disease represented by the degree of desaturation (r=0,623; p=0,017). In all children analysed, CMs were hypertrophied but unlike healthy myocardium, CM size was heterogeneous (CV=40,4%), with alternating immature and mature area. In mature area, as expected, CMs proliferation stopped as indicated by the loss of Ki67 staining and exhibit a mature rod-shape. Ultrastructurally, CMs had structured intercalated disk and elongated contractile apparatus with an alignment of Z-strikes and apparent I-band. The lateral membrane between two CMs was compacted with periodic crests and holes.

Conclusion Our data highly suggest that the increase of pressure during childhood may act as a maturation factor. Myocardium in ToF is heterogeneous, with mature and immature zone associated with fibrosis, which are a potential substrate for arrhythmias.

Conflict of interest The authors have not transmitted any conflicts of interest.