Mean number of isthmuses was 1.9±0.6. The isthmus between the pulmonary annulus (PA) and the septal patch was found in 100% patients, the isthmus between the septal patch and tricuspid annulus (TA) was found in 62% patients, the remaining isthmuses (between the TA and the RVOT patch and between the PA and RVOT patch) were present in 12% patients.

There was no correlation between the number of isthmuses and inducibility but, interestingly, all 5 VT patients had conduction velocity in the responsible isthmus during sinus rhythm <0,5 m/s.

Conclusion: VTs isthmuses after TOF repair are located in anatomically defined isthmuses, with conduction velocity in the responsible isthmus <0,5 m/s during sinus rhythm.

**335**

Characteristics of PAH associated with pretricuspid shunts in the registry of the French PAH network

Damien Bonnet [Orateur] (1), Xavier Jais (2), Laurence Rottat (2), Isabelle Szepeanski (1), Marilyne Lévy (1), Virginie Gressin (3), Olivier Sibon (2), Gérald Simonneau (2)

AP-HP, CHU Necker-Enfants Malades, Cardiologie Pédiatrique, Paris, France – (2) Centre de Référence de l’HTAP Sévère, Université Paris Sud, Clamart, France – (3) Actelion Pharmaceuticals, Paris, France

Pulmonary arterial hypertension (PAH) associated with pretricuspid shunts is commonly delayed until adulthood but PAH may reveal the underlying unknown congenital heart defect (CHD). Characteristics of this subgroup of CHD-PAH is scarcely reported.

Objective: To review and analyze patients with PAH and pretricuspid shunts enrolled in the prospective PAH registry of the French PAH network.

Patients and results: Among the 255 pts with CHD-PAH, 105 patients had a pretricuspid shunt. 72 were female and 28 male. The age at time of CHD diagnosis ranged from birth to 81 years (mean 38.1 years): 17 pts were diagnosed before the age of 18 years and 28 after 60 years. Fifteen pts had had their ASD closed after the diagnosis of PAH. In pts with open shunts, the diagnosis of PAH ranged from the postoperative period (3 cases) to 54 years; 7 pts were repaired before the diagnosis of PAH and the delay between repair and the diagnosis of PAH was done simultaneously with the diagnosis of CHD in 56% of the cases whereas in 16% the diagnosis of CHD followed the diagnosis of PAH (from 2 months to 16 years) and in 28% PAH appeared during the follow-up of the CHD (from 2 to 47 years). At inclusion, 61% were in functional class (FC) III, 32% in FC II, 5% in FC I and 2% in FC IV. Resting oxygen saturation was below 92% in 30/92 patients. 6 minutes walking distance (n=81) was 360±107 m (range 120-700). Mean pulmonary artery pressure at RHC (n=95) was 53±16 mmHg, cardiac output 5.02±1.7 L/min and pulmonary vascular resistances 11±8.3 WU. With regards to PAH specific treatment, only 49.6% of pts in class II-III received PAH therapy. During follow-up, 9 pts died and 3 underwent heart-lung transplantation.

Conclusion: In this PAH-registry, the proportion of CHD-PAH due to pretricuspid shunts compared to post-tricuspid shunts and other CHD is higher than in previously reported series. It shows that the natural history of pretricuspid (open and closed shunts) is far from being clearly understood as PAH can be diagnosed throughout life. PAH at diagnosis was only moderately less severe than in idiopathic PAH whereas PAH specific therapies are not widely used in this setting.

**336**

Tetralogy of Fallot: impact of pulmonary valve replacement on the left ventricular function

Aurélie Chalard [Orateur] (1), Marielle Gouton (2), Roland Henaine (3), Jean Ninet (3), Philippe Douek (1), Sylvie Di Filippo (2), Loïc Bousset (1)

(1) Hôpital Louis Pradel, CREATIS-LRMN, UMR CNRS 5520, Inserm U1044, Radiologie, Bron, Lyon, France – (2) Hôpital Louis Pradel, Cardiologie Pédiatrique, Lyon, France – (3) Hôpital Louis Pradel, Chirurgie Pédiatrique, Lyon, France

Pulmonary valve regurgitation is the most common mid- to long-term complication after complete repair of Tetralogy of Fallot (ToF). The benefit of Pulmonary Valve Replacement (PVR) on right ventricular volumes is well established. However, impact on the left ventricular function is still debated. We sought to determine the evolution of the left ventricular function and septal motion after PVR.

Methods: Patients with a history of complete repair of ToF and requiring a PVR were prospectively enrolled. They all underwent a cardiac MRI (CMR) performed before and after PVR at 1.5 Tesla (Intera®, Philips Medical Systems, Best, The Netherlands). The CMR protocol included cine steady-state free precession sequences in short axis planes (slice thickness=5mm, space between slices=5mm) to assess the ventricular volumes. End-diastolic (EDV), end-systolic volumes (ESV) and ejection fraction (EF) were calculated from manual contouring of the left and right ventricular chamber. All volumes were indexed to the body surface area. In order to quantify abnormal septum motion, maximal excursion of the interventricular septum (IVS) was calculated. A Wilcoxon test was performed to compare these parameters before and after PVR.

Results: 19 patients (mean age 29.6+/–14.5 years) has been included between March 2009 and May 2011. LV-EDV and LV-ESV were respectively 76.7±26.3 and 38.5±19.6 mL/m² before the PVR and 78.6±22.2 and 34.0±13.9 mL/m² after, leading to a significant increase in LV-EF of 5.7±1.7% (51.3±8.2% before PVR and 57.1±7.2% after PVR, p=0.0001). There was also a significant reduction of RV-EDV (p=0.0001) and RV-ESV (p=0.0001) but without improvement of RV-EF. Maximal IVS excursion decreased after PVR (9.2±2.9 mm before and 6.7±3.2 mm after; p=0.002).

Conclusion: This study demonstrates a significant improvement in left ventricular function and a decrease of the abnormal IVS excursion after PVR.