

Long term follow-up after biventricular repair of pulmonary atresia with intact ventricular septum and critical pulmonary valve stenosis.

Bonello B. (1), Sorensen C. (2), Schouvey S. (1), Aldebert P. (1), Fouilloux V. (3), Gorincour G. (2), Macé L. (3), Fraisse A. (1), Jacquier A. (2).

Congenital heart disease departement, La Timone University Hospital, Marseille, France (1)

Imaging departement, La Timone University Hospital, Marseille, France (2)

Congenital heart and cardiovascular surgery departement, La Timone University Hospital, Marseille, France (3)

Long term follow-up of repaired pulmonary atresia with intact ventricular septum (rPAIVS) and critical pulmonary valve stenosis (rCPVS) is not well described. Residual lesions such as atrial septal defect, pulmonary and tricuspid regurgitation (PR and TR) may lead to right ventricle (RV) enlargement. We sought to determine long term follow-up of patients with rPAIVS or rCPVS.

Methods: Retrospective study of patients with biventricular rPAIVS or rCPVS attending our centre. Cardiac magnetic resonance (CMR) imaging was performed using standardized protocols. Numbers are expressed as median [interquartile range]. Pearson correlation analysis and X² test were used to assess the relationships between different parameters.

Results: Eleven patients were studied at a median age of 13.2 years[10.3-16.2]. Two patients had previous palliation. Repair was a transannular patch in 4 patients and a percutaneous dilatation in 7. Four patients had atrial septal defect requiring surgery. Age at repair was 10 days[2.5-22.7] and delay between repair and CMR study was 13.1 years[9.9-16.2]. RV was dilated 9 patients 133 ml/m²[110-164] of which 6 patients had decrease RV ejection fraction. RV volume, ejection fraction and QRS duration were normal in 2 patients. More than mild TR was present in 7 patients. All but 1 patient had PR(32% [30-39]). Late gadolinium enhancement was found in 3 patients, at infundibular level in 2. All patients had normal left ventricle volume and function. QRS duration was ≥ 120 ms in 5 patients with right bundle branch. RV dilatation was associated with age at CMR($r=0.62$, $p=0.04$), decrease RV ejection fraction($r=0.78$, $p=0.006$), as a trend with TR($r=0.57$, $p=0.06$) but not with PR($r=0.38$, $p=0.2$) or late gadolinium enhancement ($r=-0.26$, $p=0.5$). QRS duration was not associated with the type of repair, the presence late gadolinium enhancement or RV dilatation ($p=0.8$, $p=0.4$ and $p=0.5$ respectively) but was associated with RV ejection fraction ($r=-0.7$, $p=0.02$).

Conclusions: RV dilatation, decrease RV ejection fraction and QRS enlargement are common in rPAIVS and rCPVS. Mechanisms of RV dilatation and decrease RV function appear to be multifactorial. T1 mapping studies should be performed. Determining the optimal timing for pulmonary valve replacement and tricuspid valve surgery is challenging.