

Interventions and strategies in patients with pulmonary atresia and multifocal lung perfusion

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Introduction: Patients with pulmonary atresia (PAtr) and ventricular septal defect (VSD) occur in 7 of 100000 births and account for 1% of the patients with a congenital heart disease (CHD). They usually present with multifocal lung perfusion in form of multiple aorto-pulmonary collaterals (MAPCA).

Methods: Between 2003 and 2017 we collected 60 patients with PAtr or critical pulmonary valve stenosis (cPS) and VSD, age ranged from 0.17. to 42.5 years (mean 5.1 years). All had MAPCA and the first aim was to differentiate who had native pulmonary arteries (nPA). These were selected as the later main branch pulmonary arteries and all strategies applied targeted to promote growth of these vessels, by closing collaterals doubly supplying lung areas and stenting of stenotic arteries. As a next step these vessels were connected to the right ventricle either by intervention or surgery and unifocalisation of all available MAPCA was performed. Patients without recognizable nPA were set on a palliative approach with optimisation of the lung perfusion and later VSD closure only if the right ventricular (RV) pressure was to expect lower than the systemic pressure.

Results: Follow-up time is 1.1 to 11.8 years (mean 4.7 years). Patients underwent 0 to 4 surgical operations (mean 2.1) and 0 to 14 catheterisations (mean 6). Two patients died during follow up. 15 patients (25%) had no native pulmonary artery (PA). 45 patients had a native pulmonary artery and followed our biventricular approach, while 11 of these patients still await VSD closure with or without fenestration. The RV pressure compared to the left ventricular (LV) pressure was significantly higher in the patients without nPA, in mean 94% (standard deviation 31%), than in mean 73% (standard deviation 21%) in the group with nPA, after the VSD was closed.

Conclusions: The use of MAPCA as pulmonary arteries increases the afterload of the RV after "corrective" surgery like uni-focalisation and VSD closure, so the recruitment of nPA, even if hypoplastic, improves the prognosis in many patients. They should be recognised and their growth pursued consequently by increasing forward blood flow through these vessels.