

Pulmonary atresia with intact ventricular septum: medium term follow-up

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Introduction: Pulmonary atresia with intact ventricular septum (PAIVS) is a rare and complex congenital heart disease characterized by imperforate pulmonary valve and intact ventricular septum. Treatments have been improved over time: cardiosurgical techniques have become safer during the neonatal period and percutaneous intervention has been developed as an alternative. This resulted in a reduction mortality over time, but clinical trend, final cardiac physiology (biventricular, one-and-a-half, univentricular), long term complications and therapeutic history can be very complex and currently not well known. Purpose of this study is to clarify this topic by analyzing a surviving population affected by PAIVS in retrospect.

Methods: 59 PAIVS patients and older than 6-year were selected in our Institution. Neonatal heart anatomy (tricuspid valve z-score, right ventricular morphology), therapeutic history, final cardiac physiology, clinical outcome and complications of these patients were evaluated in a median follow-up of 16 years.

Results: Patients with more unfavorable neonatal characteristics were immediately directed to univentricular palliation (9). In the other patients percutaneous valve perforation (PVP) was performed to less diminutive right ventricle and was effective in 64%.

Almost all patients (90%) underwent further surgery/percutaneous intervention during follow-up and each patient underwent an average of 3,1±1,2 interventions.

Final biventricular physiology was reached by 78% of candidates of biventricular repair. They had higher tricuspid z-score (-1,04 vs -2,89, p<0,05), severe tricuspid regurgitation (97% vs 64%, p<0,05), a more represented right ventricle (bi/tri-parted 100% vs 55%, p<0,05), no difference in surgical/percutaneous approach .

Half patients with biventricular repair has severe pulmonary regurgitation and 12 patients underwent pulmonary valve replacement for right ventricle enlargement (141±19 ml/mq vs non-valve replacement 99±38 ml/mq, p<0,05). Those who developed dilatation of the right ventricle enlargement had higher neonatal tricuspid z-score (-0,37 vs -1,29, p<0,05) but there wasn't difference about surgical/percutaneous treatment.

Most patients have NYHA I-II regardless final physiology. Only 10% has clinical arrhythmias (all supraventricular).

Conclusions:

Most of APSI patients reached biventricular physiology, they had higher tricuspid z-score, more tricuspid regurgitation, more represented right ventricle. A significant number of these patients underwent pulmonary valve replacement. There were few clinical arrhythmias.