

Therapeutic strategies in patients with pulmonary atresia

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Introduction: Newborns with pulmonary atresia (PA) with or without ventricular septal defects have a severe prognosis. Their future outcome depends on the growth of the right ventricle (RV) and the pulmonary arteries (PAs). Recent observations and new interventional and hybrid procedures may improve their perspective.

Methods: We report on 12 newborns with PA and ventricular septal defect (VSD) as well as on 12 patients with PA and intact ventricular septum (IVS). Mean age at first intervention was 158 days (1d – 6.7y), bodyweight 4.3 kg (2.5-19kg). In patients with PAVSD lung perfusion was secured by either surgery (3 Sano shunts, 3 transanular patches) or RVOT stent implantation in 5 and stenting of an atypical arterial duct (PDA) from the ascending aorta in one patient. In all patients with PAIVS the PDA was stented. High frequency perforation with balloon dilation of the pulmonary valve (PV) was necessary in 4, PV dilation only in 5 patients to increase the RV volume load. Interventional closure of RV to coronary sinusoids (RVCS) was possible in one patient.

Results: Follow up period ranges from 3 to 90 months (mean 20m). All patients with PAVSD have forward flow through the RV to the native PAs and await Fallot-repair, which has been already performed in two patients. Seven patients with PAIVS after PA opening and stenting of the PDA already achieved biventricular circulation. One of these 7 patients had additional RVCS which were suited for interventional closure as was done. Three patients with RVCS have univentricular palliation. Two patients died after their initial procedure, one on a pulmonary infection followed by ECMO and the other one on acute heart failure.

Conclusions: In patients with PAVSD staged interventional and/or surgical procedures can promote the growth of hypoplastic pulmonary arteries. Collaterals can therefore be closed and surgery will completely change from unifocalisation to Fallot repair. In patients with PAIVS staged interventional procedures can promote the growth of a hypoplastic RV, converting a single ventricle physiology to a biventricular scenario. The presence of RVCS remains critical, but interventional closure may become an option in some patients.