

Long-term outcome of pulmonary atresia with ventricular septal defect

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Introduction: We reviewed data of all patients with pulmonary atresia and ventricular septal defect examined and treated at our institution between January 1970 and December 2006. The aim was to evaluate anatomical substrates and outcome in this patient group.

Methods: Patient records of 106 patients (48 females) with median follow-up of 9.4 years (range 0-36.5 years) were studied. Angiographic data was available and analyzed in 76 cases.

Results: Confluent true pulmonary arteries (CTPA) were present in 93 (88%) patients. 44 (42%) patients had on average 2.6 major aortopulmonary collateral arteries (MAPCAs). 19 patients (18%) had completely MAPCA dependent pulmonary blood circulation (cMDPC), and of them 8 (42%) had CTPA. 104 patients had altogether 258 operations. Mean age at first operation decreased from 19.6 months (95% CI 8.0; 31.1) between 1970-1997 to 1.4 months (95% CI 0.4; 2.4) between 1998-2007. Full repair was possible in 73 patients (69%) at median age of 3.1 years, of whom 66 (90%) patients had CTPA and 13 (18%) patients had cMDPC. Reintervention rate after full repair was 49%. Overall freedom from reintervention at 2 and 5 years postoperatively was 79% and 74 %, respectively. Native pulmonary artery size was significantly bigger in patients who survived (n=60) compared to those who died (n=46). Median McGoon index was 1.4 vs. 1.0 (p<0.02) and median Nakata index was 110 vs. 62 (p<0.01). In patients surviving full repair, CTPA were significantly bigger compared to those who died after repair (Nakata 114 vs. 74, p<0.04). The origin of the pulmonary blood circulation whether from native pulmonary arteries or from MAPCAs, didn't significantly affect the survival. Extremely small or absent intrapericardial pulmonary arteries negatively affected 10-year survival. Estimated 10-year survival was 60%, and 78% for patients who achieved full repair. Additional extracardiac anomalies were present in 36% patients.

Conclusions: In our material of 106 patients with PA+VSD, complete repair has been possible in 73 patients (67%) of which 56 (76%) are alive. Extracardiac anomalies were present in 36% underscoring importance of their recognition. These observations are important for pre- and postnatal counseling.