

Pulmonary Atresia with Intact Ventricular Septum (PAIVS) – A Comprehensive Review of Management Practices & Outcomes

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Background/Hypothesis: Because of the heterogeneity of cardiac anatomy in patients with pulmonary atresia with intact ventricular septum (PA/IVS), an optimal management strategy has not been established. By reviewing our institution's experience with PA/IVS, we aimed to determine which strategy improved survival rates and outcomes. We hypothesized that patients with severely hypoplastic RV and tricuspid valve and/or right ventricle dependent coronary circulation (RVDCC) who underwent heart transplants would have better survival and outcomes than those who underwent single ventricle palliation or hybrid palliation (one-and-a-half ventricle repair).

Patients/Methods: A retrospective cohort study was performed between January 1, 2000 and December 31, 2016. We collected information about procedures, complications, length of stay in the intensive care unit and in the hospital as well as outcomes and survival.

Results: Of 64 patients, 39 (69%) were male. The median age at last follow up was 1669 days. 56 (87%) patients survived to discharge. Of the 8 deaths (median age 103 days), 1 (12.5%) followed a catheterization procedure, 5 (62.5%) followed shunt procedures, and 2 (25%) followed cardiac transplantations. The initial hospital survival-to-discharge for all operations was 97%. During this period: 5 (7.8%) underwent bidirectional Glenn; 9 (14%) were transplanted; 11 (17%) underwent bi-ventricular palliation; 3 (5%) underwent hybrid palliation; 19 (30%) underwent the Fontan; 13 (20%) were palliated to a shunt; and 4 (6%) underwent an interventional catheterization procedure. All patients who underwent single ventricle palliation, hybrid procedures or biventricular repair survived (38). 78% of transplanted patients survived (7/9). 16 (25%) of the patients with PA/IVS had RVDCC; 8 (50%) underwent transplant, 5 (31%) underwent the Fontan, and 3 (19%) underwent shunt placement and subsequently died.

Conclusions: The survival was lowest in PA/IVS patients who underwent transplant. However, most of the transplanted patients had RVDCC. This subset of patients may be associated with worse outcomes.

