

AP Target Symposium

AP Target Symposium 2 (I-APT2)

Dealing with the borderline Right Ventricle - Fontan vs One-and-a-Half Ventricle Repair vs Biventricular Repair: what are the criteria and how to get there –

Chair: Munetaka Masuda (Department of Cardiovascular Surgery, Yokohama City University Hospital, Japan)

Chair: Hiroyuki Yamagishi (Department of Pediatrics, Keio University School of Medicine, Japan)

Fri. Jul 7, 2017 4:15 PM - 5:45 PM ROOM 3 (Exhibition and Event Hall Room 3)

4:15 PM - 5:45 PM

[I-APT2-03] Decision-making process in theater: PAIVS

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Pulmonary atresia with an intact ventricular septum (PAIVS) is an uncommon congenital heart disease with a variable degree of right ventricular (RV) hypoplasia and coronary artery anomalies. Patients with RV-dependent coronary circulation should be better managed with single ventricular circulation, however achieving biventricular repair with good functional status and low systemic venous pressure is the ideal goal even to the patients with PAIVS. For biventricular circulation to be achieved, the tricuspid valve and RV must have adequate size to support pulmonary blood flow. Previous studies of PAIVS showed that RV and tricuspid valve diameters had poorer growth when RV–pulmonary artery continuity was not achieved. To aim a growth of right-sided heart structures, our repair strategy of the first palliation for patients with PAIVS includes modified BTS with pulmonary valvotomy.

We reviewed the data at our institution and analyzed the impact of the first palliation on the growth of right-sided heart and factors associated with a choice of definitive surgical procedure.

Methods and Results: Fifty patients with PAIVS underwent a staged surgical approach in Okayama university hospital since 1991. Six (12%) patients died after 1st palliation or inter-stage. Thirty patients could achieve a biventricular repair (BVR group), 6 patients had a 1+1/2 ventricular repair (1+1/2V group), and 5 patients had Fontan completion (Fontan group). After modified BTS with pulmonary valvotomy, normalized tricuspid valve (TV) diameter did not increase in any of group (BVR: pre 80% vs. post 83%, 1+1/2V: pre 63% vs. post 51%, Fontan: pre 57% vs. post 49%). Normalized RVEDV increased in only BVR group (BVR: pre 32% vs. post 64%, 1+1/2V: pre 43% vs. post 42%, Fontan: pre 29% vs. post 32%). Major coronary artery fistula was a strong factor with proceeding single-ventricle palliation (BVR 4/30 (13%) patients, 1+1/2V 1/6 (17%), and Fontan 4/5 (80%)).

Conclusions: TV growth was not obtained by modified BTS with pulmonary valvotomy, therefore TV size at birth appeared to be a predictor for achieving BVR. Proportionate RV growth was seen only in patients achieved BVR. However, RV growth was not seen in patients having 1+1/2 ventricular repair. Therefore, indication of biventricular repair is TV size of more than 80% of normal and Fontan is TV size of less than 50% of normal. Major coronary artery fistula was a strong predictor for proceeding single-ventricle palliation.