Profile of Surgically Modified History of Heterotaxy Syndrome

Objective: Surgical treatment for patients with heterotaxy syndrome is still a challenge. We sought to analyze the risk factor for worse clinical outcome in patients undergoing single-ventricle palliation.

Methods: 160 patients who were treated as single ventricle were retrospectively reviewed. Co-existent anomalies include TAPVC in 59 (37%), CAVV as valve morphology in 113 (71%), pulmonary atresia in 52 (33%). Results: 1st palliation includes BTS in 87 (54%) patients, PAB in 24 (15%) patients, and RV-PA in 6 (4%) patients. TAPVC repair was performed in 45 (69% of total TAPVC) patients. Kaplan-Meier survival curve showed a survival of 87% at 1-year, 80% at 5-year, and 77% at 10-year, respectively. Mortality was observed in 39 (24%) patients, mostly seen before BDG (before BDG in 24 (62%), after BDG in 7 (18%), and after Fontan in 8 (20%). Kaplan-Meier survival curve showed that patients with TAPVC had a significantly lower survival compared to those without TAPVC (TAPVC: 74% vs. non TAPVC: 95% at 1-year, p<0.001). Patients with pulmonary atresia had a tendency towards higher mortality but did not reach to a statisticsal significance (PA: 22% vs. non PA: 9% at 1-year, p=0.111).

Need for atrioventricular valve repair was not associated with mortality. Logistic risk analysis revealed presence of TAPVC as a risk factor for mortality (p<0.001). Conclusions: Presence of TAPVC is a significant risk factor associated with mortality in the first year of life. Pulmonary atresia and significant atrioventricular valve regurgitation was not related to the mortality.