Mid-Term Outcomes of Transcatheter Intervention for Pulmonary Atresia with Intact Ventricular Septum

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Objective: We report a single-institution experience and mid-term results for pulmonary atresia with intact ventricular septum (PA-IVS).

Method: Enrolled in the study were 34 PA-IVS patients who underwent transcatheter procedures in the neonatal period between April 2010 and October 2013. Six of the patients were lost in follow-up before the second stage. We retrospectively evaluated the mid-term results of the remaining 28.

Results: Out of 28 patients, 6 had a monopartite right ventricle (RV), 19 - bipartite and 3 - tripartite. Three patients dropped out of follow-up (2 bipartite, 1 monopartite). The remaining 25 had a mean age of 10.8±6.1 months and mean weight of 8.8±4.1 kg. Mean post-procedural follow-up period was 414±362 days. Out of 3 tripartite patients, 2 had RF and balloon valvuloplasty performed and required no further intervention, while 1 underwent 1.5-ventricular repair. In the bipartite group, 6 patients underwent the Glenn procedure, 1 had biventricular repair and 4 had surgery and biventricular repair. Of these 4, 3 also had right ventricular outflow tract (RVOT) reconstruction, infundibular muscle resection, pulmonary valve commissurotomy and a combination of the two, respectively; only 1 patient had RVOT reconstruction due to stent obstruction. Out of the 6 patients that had the Glenn procedure, 2 were lost in the early post-operative period, and 2 also underwent RVOT reconstruction with pulmonary valve commissurotomy and pulmonary valve reconstruction, respectively. Shunt surgery was only performed in 1 patient. Two monopartite patients underwent the Glenn procedure. Three monopartite and 3 bipartite patients have very recently undergone procedures and are continuing follow-up.

Conclusion: In evaluation of PA-IVS patients, many factors influence the choice of univentricular, biventricular or 1.5-ventricular repair. We had patients with a tripartite RV that needed 1.5-ventricular repair as well as those with a bipartite RV that progressed to biventricular repair. For this reason, repair options must be considered based on RV development. RVOT reconstruction and pulmonary valve surgery may be needed for biventricular repair in bipartite patients. The survival rate in monopartite patients appears to be low.