Hybrid Atroventricular Valve Replacement in Patients with Hypoplastic Left Heart Syndrome

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Objectives: Severe atroventricular (AV) valve regurgitation in patients with hypoplastic left heart syndrome is challenging given the lack of dedicated valve prosthesis for small infants. The Melody valve is a transcatheter pulmonic valve that can be modified and implanted in AV position in a hybrid fashion. We report our experience with hybrid AV valve replacement in patients with hypoplastic left heart syndrome (HLHS).

Methods: The Melody valve was used for hybrid AV valve replacement in 3 infants (mean age 10.2 months, range 7-14; mean weight 6.8 kg, range 6.6-6.9) with HLHS and severe AV (tricuspid) valve regurgitation. All patients were in refractory heart failure and unable to wean from the respirator. In one patient concomitant Berlin Heart implantation was performed. For hybrid AV valve replacement the following modifications of the Melody valve were performed. The Melody valves were shortened in order to suit the small ventricles and avoid pulmonary vein obstruction by inverting and crimping the last row of struts on both sides of the valve. Additionally, a skirt of pericardium or Cormatrix was sewn to the middle of the stent to anchor the valve in the native annulus and to prevent paravalvular leakage. The modified valves were then crimped on a balloon-in-balloon–catheter. After proper positioning, valves were balloon-inflated to appropriate diameters (mean 21mm, range 18-22 mm) and secured to the valve annulus with several interrupted sutures.

Results: Implantation of the Melody valve in AV-position was successful in all patients with no procedural complications. The Melody valves functioned properly in the high-pressure environment without any relevant valve regurgitation, with unrestrictive antegrade ventricular inflow and no paravalvular leaks during the study period. Thirty-day mortality in this complex patient population was 0%. One patient later on died from multi organ failure, one patient is awaiting cardiac transplantation and one patient is stable at home.

Conclusion: The Melody transcatheter pulmonic valve can be modified for hybrid AV-valve replacement in patients with HLHS. Percutaneous redilation is possible to adapt the valve to patient’s growth.