Staged Ventricular Septation in Double-Inlet Ventricle - a strategy to avoid Fontan?

Objective: Single stage ventricular septation for double-inlet left or right ventricle (DILV or DIRV) has historically been associated with poor early outcomes. We hypothesize that staged ventricular septation may demonstrate favorable clinical outcomes and avoid the need for Fontan palliation.

Methods: This single-center retrospective study reviewed patients with DILV or DIRV who underwent staged ventricular septation between 2015 to 2021. The strategy involves pulmonary artery (PA) banding or Norwood procedure during infancy (stage 1), followed by fenestrated ventricular and atrial septation to anchor the ventricular septum and procedures to provide favorable streaming, while maintaining systemic RV pressure to avoid septal shift (stage 2). Residual septal defects are closed with pulmonary artery band removal at stage 3. Patients are considered candidates if they have two adequate atrioventricular valves and an aortic valve. Results are reported as median (interquartile range).

Results: Staged ventricular septation strategy was applied in 12 patients (DILV = 10, DIRV = 2). Patient characteristics are outlined in Figure 1. Median combined indexed ventricle end-diastolic volume by MRI prior to stage 2 was 132 ml/m2 (117 – 148). Immediate post-operative median right and left atrial pressures following stage 2 were 7 mmHg (6 – 9) and 8 mmHg (5 – 9), respectively. Median intubation time was 2 days (1 – 2), ICU length of stay (LOS) was 4 days (2 – 6), and hospital LOS was 11 days (9 – 14). At discharge, median oxygen saturation was 90% (84 – 95). At a median follow-up time of 33 months (12 – 55) after stage 1, 11 months (0.6 – 18) after stage 2, and 17 months (9 – 23) after stage 3, there were no interstage deaths or cardiac transplants. No patients developed complete heart block. Neo-LV dysfunction (moderate or greater) was observed in 1 patient (1/12, 8%) after stage 2, necessitating placement of a resynchronization pacemaker and medical management on oral heart failure regimen. Hemodynamic evaluation with cardiac catheterization and MRI after stage 2 demonstrated median left atrial pressure of 9 mmHg (9 – 10), cardiac index of 3.4 L/min/m2 (2.9 – 3.5), and indexed end diastolic volumes of the RV and LV of 52 ml/m2 (39 – 66) and 105 ml/m2 (100 – 109), respectively. Unplanned reintervention at any stage was required in 5 patients (5/12, 42%), for transcatheter branch PA balloon dilation in 3, resynchronization pacemaker in 1, and atrioventricular valve repair in 1 patient. Five patients (5/12, 42%) have progressed to stage 3, and the remaining patients are awaiting complete septation. No patients have required Fontan palliation.

Conclusions: Staged ventricular septation is an alternative to single-ventricle palliation in a subset of double-inlet ventricle patients and is associated with acceptable early outcomes. Further studies are necessary to determine long-term outcomes of this strategy.

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Figure 1. Staged Ventricular Septal Defect Closure and Arterial Switch Operation (ASO = arterial switch operation) for transposition of great arteries.