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Congenital Cardiology Solutions

CHANGES IN CARDIAC SIZE AND FUNCTION IN CHILDREN WITH SINGLE RIGHT VENTRICLE ANOMALIES AFTER NORWOOD-RV REMODELING AFTER THE FIRST YEAR VARIES BY INITIAL SHUNT TYPE

Moderated Poster Contributions
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Background: We sought to compare changes in right ventricular (RV) size and function after the 1st year and prior to Fontan surgery in children with single RV anomalies enrolled in the Single Ventricle Reconstruction trial who underwent Norwood randomized to a modified Blalock-Taussig shunt (MBTS) vs. right-ventricle-pulmonary-artery shunt (RVPAS) .

Methods: We compared the MBTS and RVPAS shunt groups with respect to core-lab echocardiographic indices of RV size and function, tricuspid valve (TV) and neo-aortic valve (NAV) dimensions and function, and aortic arch gradient from protocol exams at 14.3±1.2 months and 33.6±9.6 months (≤ 6 months before planned Fontan).

Results: 233 subjects (111 MBTS; 122 RVPAS) had acceptable echoes at both time periods. At 14 months, there was no difference by shunt type for any measure. From the 14 mo to pre-Fontan echo, the MBTS group had stable indexed RV volumes and ejection fraction (EF), while the RVPAS group had significantly increased end-systolic volume (49.1±6.6 to 55±17.8 ml/BSA1.3, p=0.004) and decreased RVEF (44.2±3.1 to 40.9±6.7%, p=0.004). Indexed NAV area was similar in the shunt groups and became significantly smaller from 14 mo to pre-Fontan (z-score 6.8±2.9 to 5.7±2.5, p<0.001 for MBTS; 6.4±3.2 to 5.1±2.9, p<0.001 for RVPAS). Although the NAV remained markedly dilated pre-Fontan, >mild regurgitation was rare (<5%) and similar at both intervals for the 2 shunts. By the pre-Fontan study in both groups, indexed TV area also decreased, nearly normalizing in size (z-score 1.9±2.2 MBTS; 2.1±2 RVPAS). The incidence of ≥moderate TV regurgitation was stable and uncommon (18% at both intervals for both groups). Finally, aortic arch gradients were stable and usually less than 2 m/s pre-Fontan (1.7±0.5 m/s MBTS; 1.8±0.6 m/s RVPAS).

Conclusion: Initial shunt type at the Norwood procedure influences RV remodeling during the 2nd and 3rd years of life prior to Fontan surgery in survivors with single RV anomalies; longer-term systolic RV function appears to deteriorate after the RVPAS. Encouragingly, NAV and TV size decrease in both shunt groups over time, and both valves continue to function well for most survivors, without progressive regurgitation before the Fontan.