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Impact of Initial Shunt Type on Cardiac Size and Function in Children With Single Right Ventricle Anomalies Before the Fontan Procedure

The Single Ventricle Reconstruction Extension Trial

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ABSTRACT

BACKGROUND In children with single right ventricular (RV) anomalies, changes in RV size and function may be influenced by shunt type chosen at the time of the Norwood procedure.

OBJECTIVES The study sought to identify shunt-related differences in echocardiographic findings at 14 months and ≤6 months pre-Fontan in survivors of the Norwood procedure.

METHODS We compared 2-dimensional and Doppler echocardiographic indices of RV size and function, neo-aortic and tricuspid valve annulus dimensions and function, and aortic size and patency at 14.1 ± 1.2 months and 33.6 ± 9.6 months in subjects randomized to a Norwood procedure using either the modified Blalock-Taussig shunt (MBTS) or right ventricle to pulmonary artery shunt (RVPAS).

RESULTS Acceptable echocardiograms were available at both time points in 240 subjects (114 MBTS, 126 RVPAS). At 14 months, all indices were similar between shunt groups. From the 14-month to pre-Fontan echocardiogram, the MBTS group had stable indexed RV volumes and ejection fraction, while the RVPAS group had increased RV end-systolic volume ($p = 0.004$) and decreased right ventricular ejection fraction (RVEF) ($p = 0.004$). From 14 months to pre-Fontan, the treatment groups were similar with respect to decline in indexed neo-aortic valve area, >mild neo-aortic valve regurgitation (<5% at each time), indexed tricuspid valve area, and ≥moderate tricuspid valve regurgitation (<20% at each time).

CONCLUSIONS Initial Norwood shunt type influences pre-Fontan RV remodeling during the second and third years of life in survivors with single RV anomalies, with greater RVEF deterioration after RVPAS. Encouragingly, other indices of RV function remain stable before Fontan regardless of shunt type. (Comparison of Two Types of Shunts in Infants with Single Ventricle Defect Undergoing Staged Reconstruction—Pediatric Heart Network; NCT00115934) (J Am Coll Cardiol 2014;64:2026–35) © 2014 by the American College of Cardiology Foundation.

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The Pediatric Heart Network (1) SVR (Single Ventricle Reconstruction) trial compared outcomes in 549 infants undergoing a Norwood procedure for single right ventricle anomalies randomized to either a modified Blalock-Taussig shunt (MBTS) or right ventricle to pulmonary artery shunt (RVPAS) at 15 North American centers (2). The trial's primary finding was superior 1-year transplant-free survival in subjects who received a RVPAS compared with those who had an MBTS (3). However, initial shunt type for the Norwood procedure did not impact echocardiographic indices measured after shunt removal (4). Specifically, at 14 months, right ventricular (RV) systolic, diastolic, and global function; cardiac and vascular dimensions; neo-aortic and tricuspid annulus dimensions and valve function; and neo-aortic flow patterns all were similar for survivors with MBTS or RVPAS. Although significant interstage differences were noted in neo-aortic annular size and flow patterns between shunt types before stage II palliation, these were explained by the differing shunt physiologies.

SEE PAGE 2036

On the basis of these results, we hypothesized that differences in echocardiographic findings between the shunt groups would emerge with longer-term follow-up. RV dysfunction related to the creation of a right ventriculotomy in the RVPAS group was of particular concern. The Pediatric Heart Network's SVR Extension Study was undertaken to provide longitudinal surveillance of the cohort, including echocardiography within 6 months of Fontan palliation. In this analysis, we compared 2-dimensional (2D) and Doppler indices of RV size and function, neo-aortic and tricuspid valve (TV) annulus dimensions and function, and aortic size and patency to evaluate these changes for the second and third years of life for our cohort, picking up at 14 months when the original analysis left off and then conducting echocardiographic analysis again before the Fontan procedure. By investigating changes over time, we sought to identify echocardiographic features at 14 months that predict clinical outcomes.

METHODS

SUBJECTS. Entry criteria for subjects in the SVR trial have been reported previously (2). In the current study, all SVR trial survivors who underwent a transthoracic echocardiogram within 6 months of planned Fontan palliation and had that study submitted to the SVR core echocardiography lab for interpretation were eligible for inclusion. We excluded subjects who underwent biventricular repair (n = 3), had their pre-Fontan echocardiogram outside the 6-month window before Fontan surgery (n = 2), or had no Fontan procedure scheduled at the time of this data collection (n = 16).

STUDY DESIGN. The SVR study design also has previously been published (2). Briefly, SVR is a randomized clinical trial of infants with a diagnosis of single, morphologically right ventricle anomaly undergoing a Norwood procedure. The primary outcome has been published (incidence of death or cardiac transplantation at 12 months after randomization) (3), as have secondary outcomes, including hospital morbidity and rate of serious adverse events through 12 months, and risk factors for mortality and cardiac transplantation (5–7). The secondary echocardiographic markers of outcome for the SVR trial previously reported included 14-month indices of RV function, cardiac and vascular dimensions, valve annulus dimensions and function, and neo-aortic flow patterns (4).

ECHOCARDIOGRAPHIC ANALYSIS. For the SVR Extension Study, an echocardiography core laboratory at the Medical College of Wisconsin reviewed 2D/Doppler echocardiograms performed at each clinical center within 6 months of a planned Fontan procedure. Core lab procedures for image analysis and data management have been previously described (4). Echocardiographic measures obtained from the imaging are summarized in **Tables 1 and 2**.

STATISTICAL ANALYSIS. Summary descriptive statistics of echocardiographic indices are presented by intended shunt type for the Norwood procedure. Distributions of echocardiographic indices were

ABBREVIATIONS AND ACRONYMS

BSA = body surface area
MBTS = modified Blalock-Taussig shunt
MPI = myocardial performance index
RV = right ventricular
RVEF = right ventricular ejection fraction
RVPAS = right ventricle-to-pulmonary artery shunt
SVR = single ventricle reconstruction
TV = tricuspid valve

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