Maladaptive aortic properties after the Norwood procedure: An angiographic analysis of the Pediatric Heart Network Single Ventricle Reconstruction Trial

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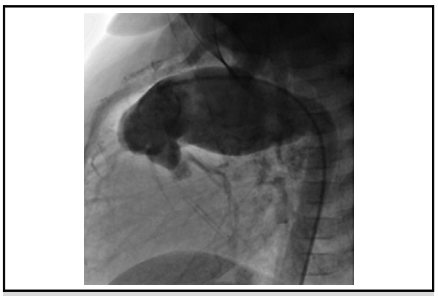
ABSTRACT

Objectives: Aortic arch reconstruction in children with single ventricle lesions may predispose to circulatory inefficiency and maladaptive physiology leading to increased myocardial workload. We sought to describe neoaortic anatomy and physiology, risk factors for abnormalities, and impact on right ventricular function in patients with single right ventricle lesions after arch reconstruction.

Methods: Prestage II aortic angiograms from the Pediatric Heart Network Single Ventricle Reconstruction trial were analyzed to define arch geometry (Romanesque [normal], crenel [elongated], or gothic [angular]), indexed neoaortic dimensions, and distensibility. Comparisons were made with 50 single-ventricle controls without prior arch reconstruction. Factors associated with ascending neoaortic dilation, reduced distensibility, and decreased ventricular function on the 14-month echocardiogram were evaluated using univariate and multivariable logistic regression.

Results: Interpretable angiograms were available for 326 of 389 subjects (84%). Compared with controls, study subjects more often demonstrated abnormal arch geometry (67% vs 22%, $P < .01$) and had increased ascending neoaortic dilation (Z score 3.8 ± 2.2 vs 2.6 ± 2.0, $P < .01$) and reduced distensibility index (2.2 ± 1.9 vs 8.0 ± 3.8, $P < .01$). Adjusted odds of neoaortic dilation were increased in subjects with gothic arch geometry (odds ratio [OR], 3.2 vs crenel geometry, $P < .01$) and a right ventricle-pulmonary artery shunt (OR, 3.4 vs Blalock–Taussig shunt, $P < .01$) but were decreased in subjects with aortic atresia (OR, 0.7 vs stenosis, $P < .01$) and those with recoarctation (OR, 0.3 vs no recoarctation, $P = .04$). No demographic, anatomic, or surgical factors predicted reduced distensibility. Neither dilation nor distensibility predicted reduced right ventricular function.

Conclusions: After Norwood surgery, the reconstructed neoaorta demonstrates abnormal anatomy and physiology. Further study is needed to evaluate the longer-term impact of these features. (J Thorac Cardiovasc Surg 2016;■:1-9)



After the Norwood, the neoaorta shows abnormal geometry and ascending aortic dilation.

Central Message

After Norwood surgery, the reconstructed neoaorta demonstrates abnormal geometry, ascending aortic dilation, and reduced distensibility, with variation seen between centers.

Perspective

We analyzed prestage II palliation aortic angiograms from the PHN Single Ventricle Reconstruction Trial subjects. Reconstructed neoaortas demonstrated abnormal arch geometry, ascending dilation, and reduced distensibility. These factors were not associated with reduced RV function on the 14-month echocardiogram. The longer-term impact of these abnormalities warrants further study.

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See Appendix for a complete list of the Pediatric Heart Network Investigators.

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Abbreviations and Acronyms

CI = confidence interval
mBTS = modified Blalock–Taussig shunt
MRI = magnetic resonance imaging
OR = odds ratio
PHN = Pediatric Heart Network
RV = right ventricle
RVPAS = right ventricle-pulmonary artery shunt
SVR = Single Ventricle Reconstruction

Scanning this QR code will take you to supplemental figure, tables, and imaging video for this article.

Despite advances in care, children born with hypoplastic left heart syndrome and related single right ventricle (RV) lesions continue to demonstrate high mortality rates.[1,2] Mortality risk is greatest in neonates and infants. However, there is continued attrition beyond infancy, and survivors have significant morbidities and reduced functional and exercise capacity.[3-5]

To reduce long-term morbidity and mortality, surgeons have sought to optimize efficiency of the palliated single-ventricle circulation. Several studies have identified alternative surgical approaches to improve flow dynamics of the total cavopulmonary connection.[6-9] In contrast, a relatively understudied source of circulatory inefficiency is the reconstructed neoaorta where higher pressures and greater flow velocities predict greater potential for energy loss than in the total cavopulmonary connection.[10] Potentially maladaptive neoaortic properties, including abnormal geometry, ascending neoaortic dilation, and reduced wall distensibility, have been described in single-center analyses and have been associated with increased power loss and ventricular dysfunction.[10-14] However, in the absence of multicenter data, it remains unclear whether these maladaptive neoaortic properties are universally present after aortic arch reconstruction or they may be related to individual surgeon technique or other center-specific factors. Moreover, risk factors that contribute to abnormal arch geometry, morphology, and physiology have not been evaluated, and no multicenter evaluation has assessed whether maladaptive neoaortic properties affect outcomes.

The National Heart, Lung, and Blood Institute–sponsored Pediatric Heart Network (PHN) Single Ventricle Reconstruction (SVR) trial enrolled the largest multicenter prospective cohort of infants with hypoplastic left heart syndrome or related single RV congenital heart disease with longitudinal follow-up after the Norwood procedure.[3,15] As part of the SVR trial, prestage II angiograms were evaluated at an angiographic core laboratory, providing a unique opportunity to study neoaortic arch anatomy and physiology in a large multicenter cohort. The objectives of this analysis were (1) to describe neoaortic arch morphology and physiology in this cohort in comparison with single-ventricle controls who did not require aortic reconstruction; (2) to assess the frequency of and risk factors for ascending neoaortic dilation and reduced distensibility; and (3) to determine the impact of ascending neoaortic dilation and reduced distensibility on RV function after arch reconstruction.

MATERIALS AND METHODS

Study Design and Sample

The SVR trial was a prospective study comparing outcomes in patients with single RV between subjects randomized to a modified Blalock–Taussig shunt (mBTS) or a right ventricle-pulmonary artery shunt (RVPAS) at the time of the Norwood procedure. Details of the trial design and primary results have been reported.[3,15] We performed a retrospective cohort study using data from the SVR trial and supplemented the analysis by making comparisons with a concurrent single-ventricle control population with no history of surgical arch intervention. For the SVR trial population, 549 subjects comprised the original analytic cohort. Of these, 389 survived and underwent cardiac catheterization before stage II palliation (superior cavopulmonary anastomosis), and 326 (84%) had aortic arch angiographic images of adequate quality for the required measurements. The control population was identified from the angiographic database at Duke University and consisted of 50 patients with single left ventricles with no prior arch reconstruction who were undergoing prestage II catheterization during the concurrent time period as the SVR trial. Diagnoses in the control population included pulmonary atresia/intact ventricular septum (n = 19), tricuspid atresia (n = 11), double inlet left ventricle (n = 7), right dominant unbalanced atrioventricular canal (n = 7), and a subset of complex single left ventricle lesions (n = 7); 5 patients (10%) had no surgical intervention before undergoing superior cavopulmonary anastomosis, 9 patients (18%) underwent pulmonary artery banding, and the majority (n = 36, 72%) received a systemic to pulmonary artery shunt. Arch measurements (descriptions follow) were all similar between control subjects receiving a systemic to pulmonary arterial shunt and those who did not; therefore, the control subjects were compared collectively with the SVR cohort. For the SVR cohort, the Institutional Review Board at each participating center approved the trial. Written informed consent was obtained from 1 or both parents. For controls, the Duke University Medical Center Institutional Review Board approved the data collection with waiver of informed consent.

Study Measurements

In the population with SVR, detailed preoperative medical history was recorded before the Norwood procedure, including demographics, subject characteristics, and anatomic diagnosis. Operative variables collected included shunt type, use of coarctectomy and patch material (yes/no), origin of shunt, bypass time, and additional cardiac operations. All post-Norwood interventions were recorded, and cases of recoarctation were defined as those for which surgical or catheter-based intervention was performed by 12 months after randomization as previously described.[16] Cardiac catheterizations before stage II palliation were performed at the discretion of the individual centers. Hemodynamic data were collected for all subjects who underwent catheterization and submitted to the data coordinating center. Angiographic analysis of both the SVR study cohort and the control population was performed at Duke University, which served as the SVR trial angiographic core laboratory. Angiographic assessment of