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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 neo-aortic 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], orgothic [angular]), indexed neo-aortic dimensions, and distensibility. Comparisons were made with 50 single-ventricle controls without prior arch reconstruction. Factors associated

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

Clinical trials registration: www.ClinicalTrials.gov (NCT00115934).

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

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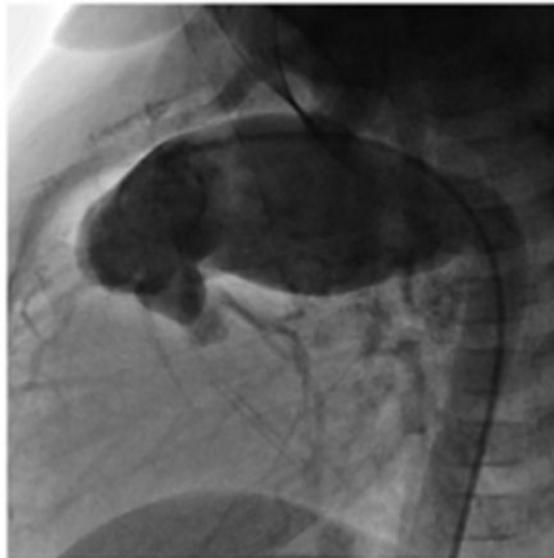
with ascending neoartical dilation, reduced distensibility, and decreased ventricular function on the 14-month echocardiogram were evaluated using univariate and multivariable logistic regression.

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 neoartical 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 neoartical 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 neoartical demonstrates abnormal anatomy and physiology. Further study is needed to evaluate the longer-term impact of these features.

Graphical abstract

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



Keywords

Norwood; aorta; coarctation

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 neoarteria where higher pressures and greater flow velocities predict greater potential for energy loss than in the total cavopulmonary connection.¹⁰ Potentially maladaptive neoarterial properties, including abnormal geometry, ascending neoarterial 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 neoarterial 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 neoarterial 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 neoarterial arch anatomy and physiology in a large multicenter cohort. The objectives of this analysis were (1) to describe neoarterial 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 neoarterial dilation and reduced distensibility; and (3) to determine the impact of ascending neoarterial 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 the neo-aorta was performed in the lateral projection by a single investigator who was blinded to both patient and center. Global aortic arch geometry was objectively quantified by measuring the arch width to height ratio and subjectively classified on the basis of previously described definitions as gothic, crenel, or normal (Romanesque).¹⁷ The gothic arch has a triangular form with a more acute angulation between the plane of the ascending and descending aorta and is typically characterized by an arch height that is greater than the arch width. The crenel arch has a more rectangular form with an arch width that is typically greater than the arch height. Normal arch geometry (Romanesque) is defined as a smooth semicircular form with an approximate 1:1 height to width ratio (Figure 1, A). The diameter of the ascending neo-aorta was measured during systole and diastole at 2 separate locations: at the largest point and just above the sinotubular junction. The descending aorta was measured during systole and diastole at the diaphragm. Additional measurements of aortic diameter were made at the proximal transverse aorta, distal transverse aorta, isthmus, and descending aorta at the area of poststenotic dilation (Figure E1). Arch measurements were indexed to body surface area. Ascending aortic Z scores were calculated using previously reported nomograms for echocardiography.¹⁸

The aortic index was defined as the ratio between the ascending aortic diameter in systole measured at the largest point and the diameter of the descending aorta in systole measured at the level of the diaphragm. The distensibility index was calculated for the ascending neo-aorta based on an average of the 2 measurements described earlier (above the sinotubular junction and at the largest point) and the descending aorta at the diaphragm as previously

described using the following calculation: $Distensibility (10^{-3} mm Hg^{-1}) = (A_{max} - A_{min}) / [A_{min} \times (P_{max} - P_{min})]$. A_{max} and A_{min} represent the maximal (systole) and minimal (diastole) cross-sectional area of the aorta measured at the same point, and P_{max} and P_{min} are the systolic and diastolic ascending aortic blood pressures obtained during catheterization hemodynamic assessment, respectively. Aortic cross-sectional area was calculated using the following calculation: $cross-sectional\ area = \pi(D/2)^2$, where D represents the measured diameter of the aorta on angiogram.

Echocardiograms were obtained after the Norwood procedure, before stage II surgery during the preoperative evaluation, and at 14 months of age. Echocardiograms were interpreted centrally at the SVR trial core echocardiography laboratory. Core laboratory procedures for image analysis (including the primary echocardiographic outcome of interest, RV fractional area change) and data management have been described.¹⁹

Statistical Methods

Standard summary statistics were used to describe study variables, including means and standard deviation or frequency counts with percentages. Wilcoxon rank-sum test or chi-square test was used for comparisons. We evaluated risk factors for 2 a priori identified binary outcomes: increased ascending neoarteria dilation (aortic index >75th percentile) and reduced distensibility (distensibility index <25th percentile) using univariate analysis. Multivariable logistic regression was performed to evaluate factors associated with neoarteria dilation while allowing for within-center correlation. Clinically relevant covariates identified as significant by univariate analysis were included in the multivariable model, and odds ratios (ORs) with 95% confidence intervals (CIs) are reported. After model assumption diagnostics and collinearity testing, the following covariates remained in the model: arch geometry (categorical variable), aortic atresia (binary variable), shunt type (binary variable), native ascending aorta diameter (continuous variable), and presence of recoarctation (binary variable). We used Spearman's rho to assess the correlation between ascending aortic dilation or reduced distensibility and change in RV function between prestage II and 14-month echocardiogram. We also assessed a discrete outcome measure, defined a priori as 5% or greater decrease in RV fractional area change between the 2 echocardiograms. We conducted all analyses using Stata 13.1 (StataCorp LP, College Station, Tex).

Results

Cohort Demographics

There were no significant differences between the SVR and control subjects with respect to age at prestage II catheterization (4.6 ± 1.8 months vs 5.0 ± 1.7 months; $P = .06$). However, the SVR subjects weighed less (5.6 ± 1.1 kg vs 6.2 ± 1.7 kg; $P < .01$) and were more frequently male (62% vs 44%; $P < .01$).

Aortic Arch Geometry

Arch geometry is summarized in Figure 1, A. Overall, 33% ($n = 108$) of the SVR cohort had Romanesque (normal) arch geometry compared with 78% ($n = 39$) of the single-ventricle controls ($P < .001$). The most common geometric subtype for the SVR cohort was the

elongated or crenel subtype (n = 186, 58%) with a width-to-height ratio of 2.0 ± 0.6 , followed by the Romanesque subtype (n = 108, 33%, ratio = 1.1 ± 0.3). Least common was the gothic subtype (n = 29, 9%, ratio = 0.8 ± 0.3). Figure 1, B demonstrates select examples of arch geometry, and the Video demonstrates still frames of all of the arch angiograms for the analytic cohort.

Arch geometry varied depending on the underlying aortic morphology. Patients born with aortic atresia or an ascending aorta 2 mm or less were significantly less likely to have normal arch geometry (28% vs 45% for aortic atresia vs stenosis [$P < .01$] and 20% vs 36% for native ascending aorta ≤ 2 vs >2 mm [$P = .01$]). There was also significant variability by center with the frequency of normal arch geometry ranging from as low as 17% to as high as 57% ($P = .003$) (Figure 2, A). Arch geometry was not significantly associated with performance of coarctectomy at the time of the Norwood or with intervention for recoarctation.

Aortic Arch Dimensions and Distensibility

Compared with the controls, SVR subjects had increased ascending neo-aortic diameters (55 ± 14 mm/m² vs 47 ± 10 mm/m²; $P < .01$) and an increased aortic index (ratio of ascending to descending aorta) (2.3 ± 0.5 vs 2.0 ± 0.3 for SVR cohort vs controls, respectively; $P < .01$). However, ascending aortic Z scores for both cohorts were dilated (Z score 3.8 ± 2.2 vs 2.6 ± 2.0 for SVR subjects vs controls, respectively, $P < .01$). Again, there was substantial variability across centers (Figure 2, B).

SVR subjects had similar transverse arch diameters as the controls (32 ± 11 mm/m vs 28 ± 6 mm/m; $P = .14$) but demonstrated narrowing at the aortic isthmus (19 ± 6 mm/m² vs 22 ± 6 mm/m² for subjects vs controls, respectively; $P < .01$). There were no differences between the SVR cohort and controls in the indexed diameter of the descending aorta or the aorta at the diaphragm.

Aortic distensibility also was abnormal in the SVR cohort. Relative to controls, reduced distensibility was seen for both the ascending (distensibility index = 2.2 ± 2.7 mm Hg vs 8.0 ± 3.8 mm Hg $\times 10^{-3}$, respectively; $P < .01$) and descending aorta (7.4 ± 5.3 vs 12.6 ± 6.4 ; $P < .01$).

Factors Associated With Aortic Dilation and Reduced Distensibility

Table 1 summarizes the factors associated by univariate analysis with an ascending aortic index 2.5 or greater (upper 25th percentile) in the SVR cohort. By multivariable analysis (Table 2), factors predicting increased ascending aortic index included receipt of an RVPAS (OR, 3.4 [95% CI, 1.8-6.8] vs mBTS, $P < .01$) and gothic arch geometry (OR, 3.2 [95% CI, 1.1-9.2] vs crenel geometry, $P < .01$). Conversely, patients with aortic atresia (OR, 0.7; 95% CI, 0.5-0.8; $P < .01$) or who received intervention for recoarctation (OR, 0.3; 95% CI, 0.1-0.9; $P = .03$) tended to have a smaller ascending aortic index. The association between recoarctation and aortic index was unchanged when excluding patients with intervention for recoarctation occurring before the prestage II catheterization (n = 2) and when excluding those who did not have recoarctation intervention until stage II surgery or later (n = 18). There were no identifiable risk factors for decreased neo-aortic distensibility (Table E1).

Outcomes

When assessed as a continuous outcome measure, there was no correlation between change in RV fractional area change and ascending aortic dimension or ascending aortic distensibility index. Overall, 34% (n = 77) of the SVR trial patients met the prespecified “cut point” for decreased RV function (5% decrease in fractional area change between prestage II echocardiogram and 14-month follow-up echocardiogram). When using this measure of decline in RV function, there was again no association with ascending aortic dilation or reduced distensibility (Table E2).

Discussion

In this analysis of aortic angiograms from the multicenter PHN SVR trial cohort, we demonstrate a high prevalence of potentially maladaptive aortic features, including abnormal arch geometry, ascending aortic dilation, and markedly reduced distensibility. The relative prevalence of these anatomic and physiologic features varied substantially between PHN centers, suggesting that physiology and postsurgical anatomy may be modifiable. However, we found no association between ascending aortic dilation or reduced distensibility and RV fractional area change measured by echocardiogram at 14-month follow-up.

An emerging body of literature indicates that abnormal geometry, morphology, and physiology in the reconstructed aorta contribute to increased wall shear stress, energy loss, and increased afterload.^{12,20,21} By using computational fluid dynamic modeling, Itatani and colleagues¹⁰ demonstrated up to 15% energy loss in the reconstructed neoorta after the Norwood surgery that was due to abnormal geometry and wall stress. They found that greater curvature of the arch resulted in the lowest amount of energy loss and that this was related to surgical technique. Similar findings have been demonstrated in patients with abnormal arch geometry after surgically repaired coarctation of the aorta, and of note, in these patients abnormal arch geometry is associated with increased left ventricular mass and increased prevalence of hypertension.²²⁻²⁴ These analyses after coarctation repair suggest that gothic (increased height to length ratio) arch geometry is the least favorable.²²⁻²⁴ Our findings are consistent with this interpretation because gothic arches demonstrated the greatest propensity for ascending aortic dilation and reduced distensibility, although the latter finding did not reach statistical significance. Relative to single left ventricle controls, few patients with single RV demonstrated the optimal curved geometry of the Romanesque aortic arch.

The typical arch morphology in our analysis also is abnormal, characterized by a markedly dilated ascending aorta with tapering across the transverse aorta to a narrowed aortic isthmus (Video 1). Voges and colleagues¹² had similar findings in their single-center analysis with an ascending aortic index of 1.9-2.1 in their single-ventricle patients (vs normal controls in whom the aortic index was 1.2-1.4). Dilation of the neoortic root and ascending neoorta has been described in other single-center analyses.^{12,25} Cohen and colleagues¹¹ found that the neoortic valve annulus, aortic root, and sinotubular junction dilate progressively over time and that the extent of dilation is associated with an increased prevalence of neoortic insufficiency. A recent analysis of the PHN cohort demonstrated a low 4% prevalence of significant (moderate or more) neoortic insufficiency.²⁶ However, follow-up in this analysis

was short with no data after Fontan surgery. It is possible that neo-aortic insufficiency associated with root dilation is a longer-term problem. Aortic root dilation also is seen in patient populations in whom the pulmonary valve functions as the neo-aortic valve, such as after the Ross procedure or arterial switch operations.²⁷⁻²⁹ The Ross procedure increasingly is being performed with reinforcement of the aortic root to prevent dilation and associated aortic insufficiency.^{30,31} The Norwood operation differs from the Ross and arterial switch operations because the entire ascending aorta is reconstructed, and, as we demonstrate, dilation extends beyond the aortic root into the ascending and transverse arch.

Patients with single RVs also demonstrated markedly abnormal physiology with approximately 4-fold reduced ascending neo-aortic distensibility when compared with single left ventricle controls. Studies using magnetic resonance imaging (MRI) data have demonstrated that reduced distensibility of the reconstructed neo-aorta contributes to suboptimal ventricular-vascular coupling, increased afterload, and energy loss.^{12,14} Voges and colleagues,¹² in an analysis of 40 patients with palliated single-ventricle lesions, found that reduced distensibility and late gadolinium enhancement (a marker of fibrosis and scar tissue) of the ascending neo-aorta correlate closely with decreased RV ejection fraction measured by cardiac MRI.¹² Although we found no correlation with RV fractional area change, there are limitations associated with echocardiographic quantification of RV function, and our analysis included a relatively short time frame of follow-up. Without definitive data, our concerns regarding the long-term implications of abnormal arch geometry, morphology, and physiology remain relatively speculative, founded on simulation models and single-center analyses, or extrapolated from other patient populations. More in-depth analysis may be feasible in the future as plans are under way to obtain MRI data at long-term follow-up in the SVR trial cohort.

Although hard evidence of a deleterious impact is lacking, it seems prudent to evaluate potentially modifiable factors that may influence neo-aortic form and function. Native aortic anatomy obviously is not a modifiable factor, and the presence of aortic atresia was associated with abnormal geometry and a smaller ascending neo-aortic diameter. However, the substantial center variability suggests that surgical technique plays a role. In support of this, Mahle and colleagues²⁵ demonstrated that long-term neo-aortic dimensions are most closely related to the immediate postsurgical dimension. Approaches to the Norwood arch reconstruction have evolved over time using variations of patch material (homografts, autografts, and extracellular matrix biomaterials) and differing techniques (eg, using coarctectomy with or without interdigitation). We did not demonstrate an association between aortic dilation and the use of coarctectomy; however, further analysis in this cohort was limited because we did not have detailed operative data (including data on use of interdigitation or the specifics of patch material).

Physiologic factors also may influence ascending aortic dimensions. We hypothesized that we would see greater ascending aortic dilation in the mBTS group due to the wider pulse pressure and greater volume load traversing the ascending neo-aorta. However, we found the opposite; patients with an mBTS had a significantly smaller ascending aortic index than those with an RVPAS. It is possible that the ascending aorta in the RVPAS group dilates more in the anteroposterior plane because of space constraints from the shunt. Because of

difficulties visualizing the ascending aorta in the anteroposterior projection, our measurements were all made in the lateral projection. Alternatively, this finding may be spurious or related to imbalance between the 2 shunt groups because of increased prestage II mortality in the mBTS group.³ In addition to shunt type, we also demonstrated that patients undergoing intervention for recoarctation were less likely to demonstrate ascending neo-aortic dilation. This likely reflects a higher risk of recoarctation in patients with smaller native aortae. However, this finding does suggest that distal obstruction is not the cause for ascending aortic dilation.

Study Limitations

Limitations of the current study include all of the limitations associated with a retrospective study, including the absence of specific data on surgical technique, native aortic arch anatomy, and other anatomic considerations that may affect postoperative anatomy. In addition, cardiac catheterization and aortic angiography were not required for SVR trial participants. Although generally accepted as standard of care, there were some patients who did not undergo prestage II cardiac catheterization or did not have interpretable aortic angiograms. Another limitation is that our distensibility index and ascending aortic *Z* scores were calculated using data from single-plane (lateral) angiograms, reducing a 3-dimensional structure to a 2-dimensional construct. In addition, these indices were developed using echocardiographic or 3-dimensional MRI data in patients with normal aortae. They have not been previously used to describe angiographic data after aortic arch reconstruction. It is possible that our absolute numbers do not compare accurately with normative echocardiographic or MRI data. However, angiographic comparisons between SVR subjects and controls should be reliable. Finally, there are several study limitations that may have affected our ability to detect a relationship between aortic properties and RV function, including the well-recognized limitations associated with echocardiographic quantification of RV function, the relatively short time frame of follow-up, and the fact that a significant percentage of our cohort had abnormal baseline RV function, suggesting that RV dysfunction is multifactorial, making it harder to isolate any single factor that might contribute to RV dysfunction. For all of these reasons, we are hesitant to reject the hypothesis that aortic morphology and physiology have a deleterious impact on long-term RV function.

Conclusions

We analyzed neo-aortic angiograms from the multicenter PHN SVR trial cohort. Neo-aortic arch geometry, morphology, and distensibility all are abnormal in these patients with single RV lesions. The implications of these variations in form and function are not yet known, and we did not find a specific association between any of these features and RV function measured by echocardiography at intermediate-term follow-up. Nonetheless, it is clear that there is substantial variability across centers, and future studies are needed to determine whether these neo-aortic abnormalities might influence patient outcomes.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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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

Central Message

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

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Perspective

We analyzed prestage II palliation aortic angiograms from the PHN Single Ventricle Reconstruction Trial subjects. Reconstructed neoortas 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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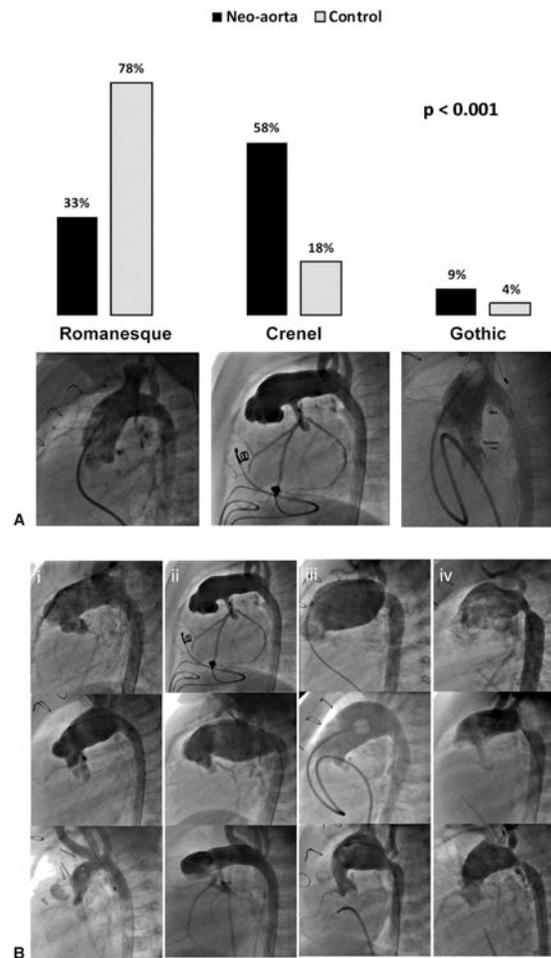


Figure 1.

A, Arch geometry in the SVR trial cohort (neoaorta) and single ventricle controls. The P value shown ($<.001$) is for individual comparisons. The Romanesque arch has a smooth semicircular form with an approximate 1:1 height to width ratio, the crenel arch has a rectangular form with width greater than height, and the gothic arch has an angular appearance with height greater than length. B, Representative examples of neo-aortic morphology and geometry. *i*, Romanesque (normal) arch geometry; *ii*, crenel arch geometry; *iii*, ascending neo-aortic dilation; *iv*, recoarctation.

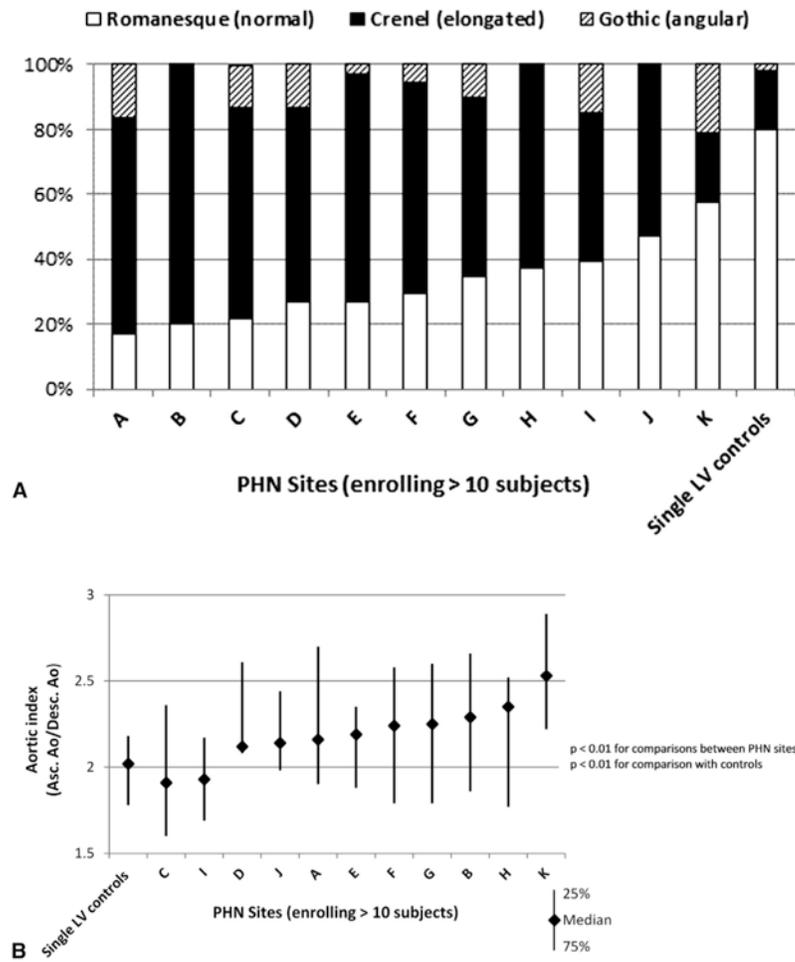


Figure 2. A, Variability was seen in neo-aortic arch geometric classification between PHN centers. B, Reconstructed neo-aortas had a greater aortic index when compared with the single-ventricle control population. Significant center variability was seen again. *PHN*, Pediatric Heart Network; *LV*, left ventricle; *Asc*, ascending; *Ao*, aorta; *Desc*, descending.

Table 1
Univariate analysis: Factors associated with ascending aortic dilation (aortic index >2.5)

	AI	2.5 (n = 82)	AI <2.5 (n = 229)	P value
Demographic features				
Gestational age, wk	311	38.1 ± 1.7	38.4 ± 1.3	.30
Birth weight, g	311	3167 ± 548	3176 ± 527	.94
Arch geometry	311			<.01
Romanesque (normal)	106	35 (33%)	71 (67%)	
Crenel (elongated)	176	35 (20%)	141 (80%)	
Gothic (angular)	29	12 (41%)	17 (59%)	
Morphologic features				
HLHS subtype	276			<.01
Aortic atresia		41 (21%)	151 (79%)	
Aortic stenosis		31 (37%)	53 (63%)	
Native AAO size	307			.19
2 mm		9 (19%)	39 (81%)	
>2 mm		72 (28%)	187 (72%)	
Recoarctation	311	12 (16%)	62 (84%)	.03
Aortic isthmus, mm/m ²	311	18.3 ± 5.3	16.7 ± 10.1	.28
Surgical variables				
Bypass time, min	311	139 ± 41	140 ± 54	.63
Coarctectomy	311	23 (23%)	76 (77%)	.39
Shunt type (intention to treat)	311			.02
mBTS	141	28 (20%)	113 (80%)	
RVPAS	170	54 (32%)	116 (68%)	
Shunt type (actual shunt received)	311			<.01
mBTS	133	22 (17%)	111 (83%)	
RVPAS	178	60 (34%)	118 (66%)	
Aortic physiology				
AAo distensibility index	226	2.4 ± 1.6	2.3 ± 3.0	.20
DAo distensibility index	226	7.64 ± 5.26	8.81 ± 5.29	.07

Data represent mean ± standard deviation or n (%) as appropriate. Percentages represent row percentages. *AI*, Aortic index; *HLHS*, hypoplastic left heart syndrome; *AAo*, ascending aorta; *mBTS*, modified Blalock-Taussig shunt; *RVPAS*, right ventricle-pulmonary artery shunt; *DAo*, descending aorta.

Table 2
Multivariate analysis: Factors associated with ascending aortic dilation (aortic index 2.5)

	OR	95% CI	P value
Arch geometries			
Crenel	Referent		
Romanesque	1.7	0.6-4.5	.32
Gothic	3.2	1.1-9.2	.03
Aortic atresia (vs stenosis)	0.7	0.5-0.8	<.01
RVPAS (vs mBTS)	3.4	1.8-6.8	<.01
Native AAO diameter (per mm increase)	1.1	0.9-1.3	.24
Recoarctation	0.3	0.1-0.9	.04

OR, Odds ratio; CI, confidence interval; RVPAS, right ventricle-pulmonary artery shunt; mBTS, modified Blalock–Taussig shunt; AAO, ascending aorta.

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