Intervention for Recoarctation in the Single Ventricle Reconstruction Trial

Incidence, Risk, and Outcomes

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Background—Recoarctation after the Norwood procedure increases risk for mortality. The Single Ventricle Reconstruction (SVR) trial randomized subjects with a single right ventricle undergoing a Norwood procedure to a modified Blalock-Taussig shunt or a right ventricle–pulmonary artery shunt. We sought to determine the incidence of recoarctation, risk factors, and outcomes in the SVR trial.

Methods and Results—Recoarctation was defined by intervention, either catheter based or surgical. Univariate analysis and multivariable Cox proportional hazard models were performed with adjustment for center. Of the 549 SVR subjects, 97 (18%) underwent 131 interventions (92 balloon aortoplasty, 39 surgical) for recoarctation at a median age of 4.9 months (range, 1.1–10.5 months). Intervention typically occurred at pre–stage II catheterization (n=71, 54%) or at stage II surgery (n=38, 29%). In multivariable analysis, recoarctation was associated with the shunt type in place at the end of the Norwood procedure (hazard ratio, 2.0 for right ventricle–pulmonary artery shunt versus modified Blalock-Taussig shunt; P=0.02), and Norwood discharge peak echo-Doppler arch gradient (hazard ratio, 1.07 per 1 mm Hg; P<0.01). Subjects with recoarctation demonstrated comorbidities at pre–stage II evaluation, including higher pulmonary arterial pressures (15.4±3.0 versus 14.5±3.5 mm Hg; P=0.05), higher pulmonary vascular resistance (2.6±1.6 versus 2.0±1.0 Wood units·m^2; P=0.04), and increased echocardiographic volumes (end-diastolic volume, 126±39 versus 112±33 mL/BSA$^{1.3}$, where BSA is body surface area; P=0.02). There was no difference in 12-month postrandomization transplantation-free survival between those with and without recoarctation (P=0.14).

Conclusions—Recoarctation is common after Norwood and contributes to pre–stage II comorbidities. Although with intervention there is no associated increase in 1-year transplantation/mortality, further evaluation is warranted to evaluate the effects of associated morbidities.

Clinical Trial Registration—URL: http://www.clinicaltrials.gov. Unique identifier: NCT00115934.

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Children born with the hypoplastic left heart syndrome and other single right ventricle lesions have a high mortality risk.[1,2] Individual centers have improved outcomes by identifying modifiable risk factors, including residual or recurrent anatomic lesions.[3,4] Recoarctation is one such lesion and contributes to mortality and morbidity after the Norwood procedure.[5,6]

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Although recoarctation is recognized as an important comorbidity, its true impact remains unknown. Current understanding is based on single-center retrospective analyses that have reported an incidence of recoarctation ranging from as low as 2% to as high as 40%.[7–13] These reports have not accurately defined risk factors for recoarctation, and it is unclear whether intervention occurs sufficiently early in the post-Norwood course when the risk of mortality and morbidity is most significant. Furthermore, there are limited data on the impact of intervention on morbidities and mortality.

The National Heart, Lung, and Blood Institute–sponsored Pediatric Heart Network Single Ventricle Reconstruction (SVR) trial includes the largest multicenter, prospective cohort of infants with hypoplastic left heart syndrome or related single right ventricle anomalies with longitudinal follow-up after the Norwood procedure.[1] The purpose of this analysis is to describe the incidence and timing of intervention for recoarctation by 12 months after randomization, to assess factors that predict intervention, and to determine the impact of recoarctation with intervention on morbidity and mortality in the first year after the Norwood procedure.

Methods

Study Design and Sample

The SVR trial was a prospective trial that compared outcomes between subjects randomized to either a right ventricle–pulmonary artery shunt or a modified Blalock-Taussig shunt at the time of the Norwood procedure. Details of the trial design and main results have previously been reported.[1] We performed a retrospective cohort study using data collected from the SVR trial. Of the 555 subjects enrolled in the SVR trial, 6 were excluded from the primary end point: 5 infants did not undergo a Norwood procedure after randomization, and 1 patient's family withdrew research consent after the Norwood procedure. Thus, 549 subjects formed the analytic cohort for the trial. The Institutional Review Board at each participating center approved the trial, and written informed consent was obtained from 1 or both parents.

Study Measurements

Before the Norwood procedure, a detailed preoperative medical history was recorded, including demographics, subject characteristics, and anatomic diagnosis. Operative variables included shunt type, type of arch reconstruction (including juxtaductal coarctectomy), use of patch material, origin of polytetrafluoroethylene graft (innominate, subclavian, common carotid, aorta, or right ventricle), and additional cardiac operations. All post-Norwood surgical or percutaneous interventions were recorded, including additional cardiac surgeries such as repair of recoarctation at the time of stage II (superior cavopulmonary anastomosis) palliation. Hemodynamic data were collected for all subjects undergoing cardiac catheterization before stage II surgery.

Catheterizations were performed at the discretion of the individual centers after the Norwood procedure. A total of 389 catheterizations were performed. Hemodynamic data were submitted to the data coordinating center. Angiograms were interpreted at a core laboratory. Interpretable aortic angiograms were available for 331 of 389 study subjects (85%) with catheterization data. Angiographic measurements were performed in the lateral projection during systole. The coarctation index was defined as the ratio between the narrowest isthmic diameter and the diameter of the descending aorta at the level of the diaphragm. Cases of recoarctation were defined as those cases for which surgical or catheter-based intervention was performed by 12 months after randomization. All other variables were defined as per the SVR study.[1]

Echocardiograms were obtained after the Norwood procedure (either at the time of discharge or at ≈30 days of age if still hospitalized), before stage II surgery (during the preoperative evaluation for the stage II procedure), and at 14 months of age (end-of-study visit). There was no clinically meaningful difference in age at echocardiography for those with and without coarctation intervention (age at Norwood echocardiogram, 18±10 versus 23±14 days; age at stage II echocardiogram, 5.0±1.5 versus 4.8±1.5 months; and age at 14-month echocardiogram, 14.5±1.4 versus 14.5±1.2 months for those with and without coarctation by 12 months, respectively). Echocardiograms were interpreted centrally at a core laboratory. Echocardiographic variables included native ascending aorta diameter, distal transverse arch diameter, continuous-wave Doppler of the descending aorta, right ventricular fractional area change, body surface area–indexed end-diastolic dimension, body surface area–indexed end-systolic dimension, and degree of atrioventricular valve regurgitation (none, mild, moderate, severe).

Statistical Methods

The unit of observation for this analysis was an infant enrolled in the study. We used standard summary statistics to describe the study variables, including means with standard deviations and frequency counts with percentages. We evaluated risk factors for residual or recurrent coarctation using a Cox proportional hazards model with a shared γ frailty to account for clustering by site.[14] Individual centers included in the SVR trial defined the groups over which the frailties were shared. We treated the frailties of the final multivariable model as γ distributed with a mean of 1 and a variance estimated from the data of θ=0.394 with P=0.02 from the likelihood ratio test of H0: θ=0, suggesting a significant frailty effect. The hazard ratios (HRs) were treated conditionally on θ fixed at its optimal value that maximized the penalized log likelihood.[14] For the purpose of the model, time was defined as days from randomization to first intervention for recoarctation (transcatheter or surgical). The outcome of interest was an intervention for recoarctation defined as a dichotomous variable, and infants were censored at the time of death or at 12 months after randomization. Variables significantly associated with recoarctation in a univariate analysis and those associated with coarctation in previous studies were included in the multivariable model. We tested the proportional hazards assumption using observed (Kaplan–Meier) versus expected (Cox model) plots and goodness-of-fit tests on the basis of Schoenfeld residuals. For all variables included in the model, the observed versus predicted curves were visually inspected, and the P values from the goodness-of-fit tests were all >0.05. We compared catheterization, stage II operation, and 14-month echocardiographic variables between subjects with and without intervention using χ^2 tests of association for categorical variables and Wilcoxon rank-sum tests for continuous variables. We used a Wilcoxon signed-rank test to compare matched paired measurements of the coarctation index before and after balloon angioplasty in the same subject. For mortality outcomes, we used the Kaplan–Meier method and compared 12-month transplantation-free mortality between the groups with and without coarctation using the log-rank test. We conducted all analyses using Stata 12.0 (Stata Corp, College Station, TX) and considered a value of P<0.05 statistically significant.

Results

Of the 549 SVR subjects, 97 (18%) underwent a total of 131 interventions (92 balloon aortoplasty, 39 surgical) for recoarctation within the first 12 months after randomization. The incidence of recoarctation intervention at 1, 3, 6, and 12 months was 0%, 3%, 16%, and 23%, respectively (Figure 1). Intervention typically occurred at the time of pre–stage II cardiac catheterization (n=71, 54%) or concomitant with stage II surgery (n=38, 29%). Median age at initial intervention was 4.9 months (range, 1.1–10.5 months). There was wide center variation, with intervention incidence rates ranging from 0 to 50 interventions per 100 patient-years for the 14 centers enrolling ≥10 patients. Center variation did not appear to

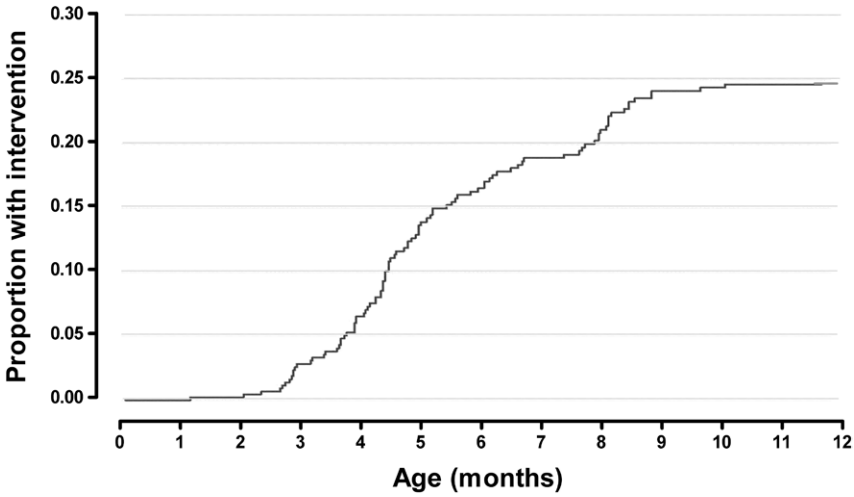

Figure 1. Kaplan–Meier curve for recoarctation intervention by 12 months after randomization.

reflect differing indications for intervention because centers with higher intervention incidence rates did not use lower catheterization peak recoarctation gradients as an indication for intervention (Figure 2).

By univariate Cox proportional hazards analysis (Table 1), intervention for recoarctation by 12 months was not associated with any demographic or surgical variables but was associated with a smaller native ascending aorta (P=0.03) and echocardiographic indexes assessed at Norwood hospital discharge, including smaller echocardiographic distal transverse arch diameter (P=0.04) and increased peak echo-Doppler arch gradient (P<0.01). Overall, 46 of 355 subjects (13%) demonstrated a pre-Norwood discharge peak echo-Doppler gradient >20 mm Hg. Of those, 22 ultimately received an intervention, 4 died in the interstage period, and 20 survived without any intervention (sensitivity, 31%; specificity, 92%; positive predictive value, 57%; negative predictive value, 83%).

By multivariable Cox proportional hazard modeling adjusted for site, predictors of recoarctation intervention included the peak echocardiographic arch gradient at the time of Norwood discharge (HR, 1.07 per 1 mm Hg; P<0.01) and receipt of a right ventricle–pulmonary artery shunt (HR, 2.0 for right ventricle–pulmonary artery shunt; P=0.02). However, when the model was repeated with the assigned shunt type (intention to treat), there was no association with recoarctation (HR, 1.4; P=0.24; Table 2). The multivariable results were unchanged when all Norwood hospital deaths and transplantations were excluded.

Recoarctation Intervention and Pre–Stage II Comorbidities

By univariate analysis (Table 3), intervention for recoarctation before stage II surgery was associated with hemodynamic variables measured at pre–stage II catheterization, including a higher peak systolic catheterization gradient (P<0.01), higher mean pulmonary arterial pressures (P=0.05), and higher pulmonary vascular resistance (P=0.04). Subjects undergoing intervention also demonstrated increased echocardiographic body surface area–indexed right ventricular end-systolic (P<0.04) and end-diastolic (P<0.02) volumes at the time of stage II surgery. However, right ventricular volumes were not different for those with and without recoarctation at the 14-month follow-up echocardiogram. This relationship remained when only the 182 subjects who had both stage II and 14-month echocardiographic data were compared. Importantly, subjects who underwent intervention for recoarctation did not demonstrate decreased right ventricular fractional area change or increased atrioventricular valve regurgitation at the time of stage II surgery or at the 14-month follow-up compared with those who did not have a recoarctation intervention by 12 months.

Intervention Outcomes

Overall, 33 of 97 subjects (34%) received a second (n=31) or third (n=2) intervention for recoarctation within 12 months after randomization. Reintervention rate was 39% (n=33 of 83) for subjects undergoing balloon angioplasty as their initial intervention. Although balloon angioplasty significantly improved the coarctation index (0.6±0.1 versus 0.8±0.2 for preangioplasty versus postangioplasty index; P<0.01), there was no association between degree of improvement and need for subsequent reintervention. For those subjects undergoing surgical recoarctation intervention, 5% (2 of 39) required subsequent reintervention for recoarctation.

Outcomes

At 12 months after randomization, there were 7 deaths and 2 heart transplantations in the 97 subjects with prior intervention for recoarctation. Comparison of all patients undergoing pre–stage II cardiac catheterization (n=380) shows that 12-month postrandomization transplantation-free survival did not differ for those with intervention (86% versus 91% for intervention versus no intervention, respectively; P=0.14). There was also no difference in 1-year transplantation-free survival in the subset of subjects who survived to stage II surgery (88% versus 93% for prior intervention versus no intervention, respectively; P=0.07).

Although mortality in the SVR cohort was more likely during the Norwood hospitalization or interstage period, very few subjects with early mortality had prior intervention for recoarctation (1 of 97 and 1 of 51 for Norwood hospital and

Table 1. Univariate Predictors of Intervention for Recoarctation by 12 Months

	n	Overall (n=449)	Recoarctation (n=97)	No Recoarctation (n=452)	HR	*P*
Demographic features						
Gestational age, wk	549	38.2±1.6	38.1±1.5	38.2±1.6	0.96	0.56
Birth weight, g	549	3103±541	3058±478	3113±554	1.0	0.14
Age at Norwood, d	549	5.8±4.1	4.8±3.3	6.0±4.2	0.94	0.08
Race, n (%)	544		95	439		0.16
White		436 (80.2)	80 (84.2)	356 (81.1)	0.95	
Black		86 (15.8)	13 (13.7)	73 (16.6)	0.88	
Asian		10 (1.8)	2 (2.1)	8 (2.0)	Referent	
Hispanic, n (%)	539		95	444	0.94	0.84
Yes		101 (18.7)	20 (21.3)	81 (18.2)		
Morphological features, n (%)						
HLHS subtype	480		87	393		0.47
Mitral atresia/aortic atresia		209 (43.5)	42 (48.3)	167 (42.5)	1.43	
Mitral stenosis/aortic atresia		138 (28.8)	19 (21.8)	119 (30.3)	1.16	
Mitral atresia/aortic stenosis		23 (4.8)	6 (6.9)	17 (4.3)	1.92	
Mitral stenosis/aortic stenosis		110 (22.9)	20 (23.0)	90 (22.9)	Referent	
Aortic atresia	480	347 (72.3)	61 (70.1)	286 (72.8)	1.02	0.47
Native ascending aorta diameter	527		97	430	0.59	0.03*
≤2 mm		98 (18.6)	24 (24.7)	74 (17.2)		
Surgical variables						
Bypass time, min	549	144±54	137±50	145±55	1.0	0.65
Coarctectomy, n (%)	548		96	452	1.1	0.63
Yes		184 (33.6)	30 (31.2)	154 (34.1)		
Shunt type (intention to treat), n (%)	549		97	452		
RVPAS		274 (49.9)	56 (42.2)	218 (48.3)	1.2	0.32
MBTS		275 (50.1)	41 (57.8)	234 (51.7)	Referent	
Shunt type (actual shunt received), n (%)	549		97	452		
RVPAS		281 (51.2)	60 (61.9)	221 (48.9)	1.3	0.19
MBTS		268 (48.8)	37 (38.1)	231 (51.1)	Referent	
Echocardiography variables (stage I discharge)						
Peak arch gradient, mm Hg	355	11.2±7.9	14.9±1.1	10.2±6.5	1.06	<0.01*
Distal arch diameter, mm	442	6.3±1.5	5.9±1.6	6.4±1.5	0.18	0.04*
RV end-diastolic volume, mL/BSA$^{1.3}$	309	93±50	93±32	93±24	1.0	0.61
RV end-systolic volume, mL/BSA$^{1.3}$	309	50±17	49±21	50±16	1.0	0.89
RV fractional area change, %	376	37±7	37±8	37±7	1.5	0.61

HRs are from univariate Cox proportional hazards models. Data represent n±SD or n (%) as appropriate. BSA indicates body surface area; HLHS, hypoplastic left heart syndrome; HR, hazard ratio; MBTS, vs modified Blalock-Taussig shunt; RVPAS, right ventricle–pulmonary artery shunt; and RV, right ventricle.

*Significant.

interstage mortality, respectively). However, 29% of subjects (7 of 24) who died after stage II surgery through 12 months had prior recoarctation intervention. To assess whether this disproportionate contribution to late mortality might reflect a higher incidence of unrecognized recoarctation in those with interstage mortality, pre-Norwood discharge echo-Doppler arch gradients were assessed. Although peak gradients were significantly higher in those with interstage mortality, quantifiable differences were small (11±8 versus 13±6 mm Hg for survival versus interstage deaths, respectively; *P*=0.03), and only 4 of 51 subjects with interstage mortality demonstrated a pre-Norwood discharge gradient >20 mm Hg.

Discussion

The major findings of the present multicenter analysis include an 18% cumulative incidence of intervention for recoarctation, with incidence by center ranging widely from 0% to 50%. Recoarctation appears difficult to predict early in the clinical

Table 2. Multivariable Predictors of Intervention for Recoarctation by 12 Months

Predictor	Hazard Ratio (95% CI)	Model *P* Value
Birth weight	0.99 (0.99–1.0)	0.87
Bypass time	1.0 (0.99–1.0)	0.44
Age at Norwood	0.9 (0.9–1.0)	0.18
RVPAS (actual shunt received)*	2.0 (1.1–3.6)†	0.02†
Coarctectomy	0.99 (0.5–2.0)	0.98
Native ascending aorta >2 mm	0.66 (0.3–1.3)	0.22
Norwood discharge echo gradient	1.07 (1.0–1.1) †	<0.01†
Aortic atresia	0.65 (0.4–1.2)	0.19

CI indicates confidence interval; and RVPAS, right ventricle–pulmonary artery shunt.

*Actual shunt received represents the shunt in place at the end of the Norwood procedure. When multivariable modeling was repeated with the intention-to-treat shunt type, use of an RVPAS was no longer associated with recoarctation (hazard ratio, 1.4; 95% confidence interval, 0.8–2.4; *P*=0.24).

†Significant.

course, and intervention typically occurred later in the post-Norwood course, most commonly at the time of pre–stage II catheterization. Subjects with intervention demonstrated pre–stage II comorbidities including higher pulmonary vascular resistance and right ventricular dilation; however, echocardiographic dimensions demonstrate relative normalization after intervention. Initial intervention for recoarctation was most often balloon aortoplasty, which was acutely effective at increasing the coarctation index, but the result was either inadequate or short-lived, requiring subsequent reintervention in 39%. There was no difference in overall survival between those with and without recoarctation intervention; however, recoarctation may have been unrecognized in those with pre–stage II mortality who did not undergo cardiac catheterization.

Recoarctation after single-ventricle surgery has been shown to exacerbate atrioventricular valve regurgitation and ventricular dysfunction and to increase mortality risk.[5,7,15] Despite these recognized consequences, the true scope of the problem has been difficult to define, with reported incidence ranging from 2% to 40%.[7–13] These prior reports have been retrospective, single-center analyses. The present report is the first multicenter analysis with prospectively collected data. These data confirm that recoarctation is a significant concern, with intervention in nearly 1 of 5 Norwood recipients.

In this analysis, there was no difference in 12-month post-randomization transplantation-free survival or post–stage II survival for subjects with intervention. Although it is encouraging that with intervention late mortality outcomes are unaffected, it is less clear whether earlier mortality in the predischarge or interstage period might be affected by

Table 3. Hemodynamic and Echocardiographic Consequences

	n	Recoarctation	No Recoarctation	*P*
Pre–stage II catheterization				
End-diastolic pressure, mm Hg	69/311	8.6±3.3	8.2±3.3	0.30
Wedge pressure, mm Hg	45/205	10.7±3.4	10.8±3.9	0.80
PA pressure, mm Hg	48/204	15.4±3.0	14.5±3.5	0.05
PVRI, WU·m^2	40/230	2.6±1.6	2.0±1.0	0.04*
AVO_2 difference	64/306	25.4±7.6	24.8±7.4	0.80
Coarctation index	68/257	0.6±0.2	0.9±0.1	<0.01*
Peak systolic gradient, mm Hg	79/280	19.8±14.3	3.3±7.1	<0.01*
Stage II echocardiography				
RV end-diastolic volume, mL/BSA$^{1.3}$	57/210	126±39	112±33	0.02*
RV end-systolic volume, mL/BSA$^{1.3}$	57/210	74±32	63±23	0.04*
RV fractional area change, %	84/276	32±8	34±8	0.24
Tricuspid insufficiency	68/329			0.95
None/mild		50 (73.5)	242 (73.6)	
Moderate/severe		18 (26.5)	87 (26.4)	
Echocardiography at 14 mo				
RV end-diastolic volume, mL/BSA$^{1.3}$	48/181	94±32	89±30	0.51
RV end-systolic volume, mL/BSA$^{1.3}$	48/181	54±24	52±23	0.71
RV fractional area change, %	66/225	33±7	32±7	0.56
Tricuspid insufficiency	73/247			0.48
None/mild		56 (76.7)	191 (77.3)	
Moderate/severe		17 (23.3)	56 (22.7)	

Data represent mean±SD when appropriate. *P* values are from the Pearson χ^2 or Wilcoxon rank-sum test when appropriate. AVO_2 difference indicates the difference between systemic and mixed venous saturation, BSA, body surface area; n, number of subjects with/without recoarctation; PA, pulmonary artery; PVRI, pulmonary vascular resistance index; RV, right ventricle; and WU, Wood units.

*Significant.

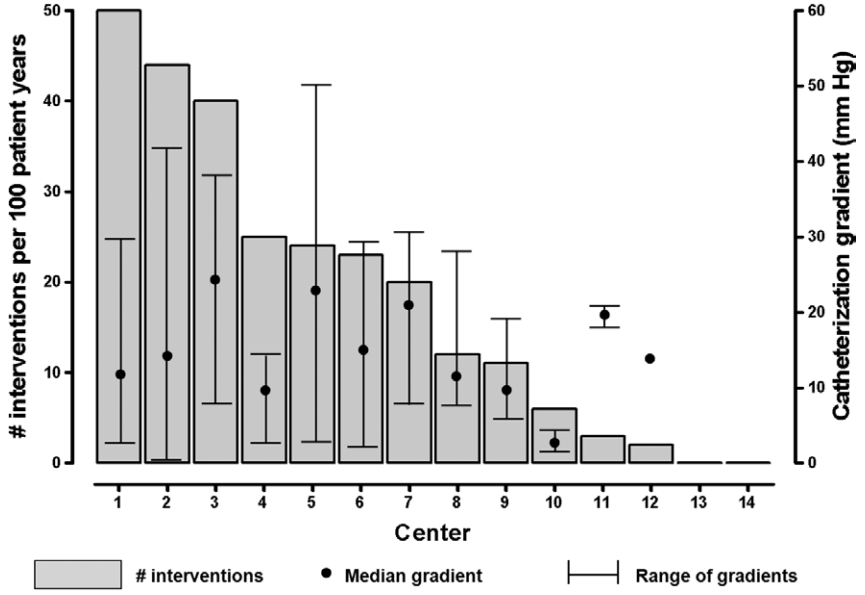

Figure 2. Intervention rates for recoarctation and catheterization recoarctation gradients by center. There was wide center variability in intervention rates (0%–50%) for the 14 Single Ventricle Reconstruction (SVR) trial centers that enrolled ≥10 subjects during the trial. Median and range of catheterization recoarctation gradients are also presented by center.

recoarctation. Mortality risk is significantly higher during these earlier stages. However, in this cohort, intervention for recoarctation was performed rarely in subjects during Norwood hospitalization or in those children with interstage mortality. This suggests that recoarctation either does not develop until late in the course or is underdiagnosed in those with earlier mortality. Underdiagnosis is a concern; Fraisse et al[6] demonstrated 40% mortality in patients with a missed diagnosis of recoarctation that was subsequently identified by autopsy. To assess the likelihood of missed diagnoses, pre-Norwood discharge echocardiographic arch gradients were evaluated. Although gradients were significantly higher in those with interstage death, the measured differences were small and the sensitivity of gradients commonly used to predict recoarctation (ie, >20 mm Hg) was poor. Previous studies have evaluated similar pre–stage I discharge echocardiographic indexes, including arch gradients, atrioventricular valve regurgitation, and ventricular dysfunction, and have demonstrated variable sensitivity with differences typically seen only with more severe recoarctation.[6,16] There were also no specific anatomic or demographic risk factors that predicted intervention. Our impression is that recoarctation is not easy to predict early in the postsurgical course with echocardiographic or clinical variables. This conclusion is supported by the timing of intervention, which occurred relatively late in the stage I course, typically close to stage II surgery. Some centers are now moving toward catheterization or alternative imaging before Norwood discharge. It remains to be seen if this will allow earlier identification and intervention for recoarctation or if earlier intervention would change outcomes.

Beyond mortality, several morbidity measures were associated with recoarctation intervention, including increasing echocardiographic ventricular dimensions, mean pulmonary arterial pressures, and pulmonary vascular resistance. Although echocardiographic ventricular end-systolic and end-diastolic dimensions were increased at the time of intervention, these dimensions demonstrated relative normalization at the 1-year follow-up. This is encouraging and suggests that some of the deleterious effects of recoarctation can be reversed with intervention.

Surgical Risk Factors for Recoarctation

Several reports have suggested that recoarctation is dynamic and may result from contraction of residual ductal tissue. In autopsy specimens, ductal tissue has been demonstrated at the recoarctation site, and surgeons have reported decreased incidence when using techniques that eliminate ductal tissue.[8,9,17,18] These techniques include complete resection using coarctectomy or coarctectomy with an interdigitating technique to further prevent ductal constriction.[9,17] In this present analysis, interdigitation was not reported, but complete coarctectomy was used in one third of subjects and did not decrease the incidence of recoarctation intervention. Despite the absence of a clear-cut surgical risk factor, the wide center variability suggests that perhaps surgical technique may be an important factor.

An interesting finding in this analysis was the association of assigned shunt type but not intention-to-treat shunt type with reintervention. One possibility is that recoarctation is a greater risk for early mortality in those with a modified Blalock-Taussig shunt than in those with an right ventricle–pulmonary artery shunt. Another SVR trial analysis has demonstrated increased interstage mortality with a modified Blalock-Taussig shunt, and it is plausible that this circulation is more tenuous and incapable of tolerating the hemodynamic burden of recoarctation.[19] Another explanation is that factors leading to crossover from 1 shunt type to the other might have affected the risk of recoarctation. The intraoperative crossover rate in the SVR trial was 9%, with an approximately equal number in each direction.[1] The most frequent reason for shunt crossover at the time of Norwood was cardiac or arch anatomy that prevented the use of the assigned shunt. Those subjects with complex arch anatomy that prohibited the use of the modified Blalock-Taussig shunt were potentially at increased risk for recoarctation.

Interventions for Recoarctation

Balloon aortoplasty has become the intervention of choice for recoarctation at most centers. Our data confirm the findings of prior reports indicating that balloon aortoplasty is effective at acutely increasing isthmic diameter.[7,11–13,20–22] However, 39% of subjects required subsequent reintervention, which is higher than in previous reports. Although surgical intervention is associated with a lower recurrence rate, many caretakers find it reasonable to use aortoplasty as the first-line intervention because surgical intervention is more invasive. However, aortoplasty is not risk free, and several prior reports have demonstrated intraprocedural arrest or bradyarrhythmias with the need for cardiopulmonary resuscitation in 20% to 30% of those undergoing aortoplasty.[12,13,20,22]

Limitations of the present study include the noted use of intervention to define recoarctation. The advantage of this definition is that it is a well-defined end point. However, indications for intervention and the timing of intervention likely vary by center, which introduces some heterogeneity. This definition also limits the ability to detect recoarctation in subjects who did not undergo cardiac catheterization. No invasive hemodynamic or angiographic data were available for the majority of subjects with early mortality. Other limitations include the absence of specific data on surgical technique or procedural details during catheterization, including specific details on balloon size, postintervention hemodynamics, and procedural complications.

Conclusions

Intervention for recoarctation was performed in almost 1 in 5 patients after single-ventricle surgical reconstruction. Intervention was typically performed at the time of pre–stage II catheterization, and in this analysis, very few patients with early mortality (before stage II) had intervention for recoarctation. It is counterintuitive that recoarctation is protective against early mortality; more likely, recoarctation is underdiagnosed. It is enticing to speculate that earlier diagnosis may decrease interstage mortality. There was wide center variability, and this finding suggests either that some centers have managed to significantly decrease recoarctation incidence or alternatively that some centers are simply more aggressive with intervention. The former explanation seems more likely because the gradients at the time of intervention did not vary substantially by center. Regardless of the explanation, recoarctation remains a common problem. Although associated intermediate-term morbidities such as ventricular dilation appear to be reversible with intervention, it remains unclear whether long-term outcome is improved.

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See the online-only Data Supplement for a complete list of the Pediatric Heart Network Investigators.

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Disclosures

None.

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CLINICAL PERSPECTIVE

Children born with the hypoplastic left heart syndrome and related single right ventricle anomalies suffer from significant morbidities and high mortality rates. Residual or recurrent recoarctation of the aorta is an important contributor to morbidity and may increase mortality. We performed a retrospective cohort study using data collected from the Single Ventricle Reconstruction trial to describe the incidence and timing of intervention for recoarctation, factors that predict intervention, and the impact of recoarctation with intervention on morbidity and mortality. We found that recoarctation intervention is common, with an incidence at 1, 3, 6, and 12 months of 0%, 3%, 16%, and 23%, respectively. This varies widely by center (from 0–50 interventions per 100 patient-years) and often occurs relatively late in the interstage course. By multivariable analysis, factors associated with recoarctation intervention included the peak echocardiographic arch gradient at Norwood discharge and receipt of a right ventricle–to–pulmonary artery shunt. However, when modeling was repeated using the assigned shunt type (intention to treat), there was no longer a significant association, perhaps implicating factors associated with shunt crossover or a potential survivor bias. Recoarctation with intervention was associated with comorbidities assessed before stage II surgery, including higher pulmonary vascular resistance and increased echocardiographic right ventricular chamber dimensions. Recoarctation with intervention was not associated with decreased 12-month transplantation-free survival. Thus, intervention for recoarctation is relatively common but may be difficult to predict early in the clinical course. Although recoarctation contributes to morbidities, with intervention, 12-month outcomes were not affected.

Intervention for Recoarctation in the Single Ventricle Reconstruction Trial: Incidence, Risk, and Outcomes

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