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Survival and Clinical Course at Fontan After Stage One Palliation With Either a Modified Blalock-Taussig Shunt or a Right Ventricle to Pulmonary Artery Conduit

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Objectives We sought to determine whether the type of shunt used at stage one palliation (S1P) affected the survival and the perioperative course through Fontan completion.

Background Although improved surgical and interstage survival have been demonstrated with the use of the right ventricle to pulmonary artery (RV-PA) conduit compared with a modified Blalock-Taussig shunt (BTS) at S1P, it is unknown whether this effect will be observed in long-term follow-up.

Methods All patients who underwent a S1P during 2002 and 2003 (n = 80) at our institution were included for analysis. Patients were followed until death or June 1, 2007. Perioperative variables at Fontan completion were recorded.

Results For the entire cohort, cumulative survival for those who underwent a RV-PA conduit (n = 34) was 79.4% at 3 years compared with 65.8% in the modified BTS group (n = 46) (log-rank = 0.31). At Fontan (n = 44), when compared with those who had received a modified BTS, those who had a RV-PA conduit placed at S1P had no difference in the median duration of ventilation (21 h [range 10 to 96 h] vs. 26.5 h [range 7 to 204 h], p = 0.09) or hospital stay (9 days [range 5 to 29 days] vs. 10 days [range 6 to 48 days], p = 0.89), although length of stay in the intensive care unit was shorter (2 days [range 0 to 6 days] vs. 4 days [range 1 to 25 days], p = 0.01). Sixty-seven percent of the RV-PA conduit group had at least one PA intervention 3 years after S1P compared with 42.8% in the modified BTS group (log-rank = 0.11).

Conclusions Nonstatistically significant trends toward improved cumulative survival and increased PA interventions were demonstrated in patients who had a RV-PA conduit placed at S1P. Longitudinal follow-up of larger groups of randomized patients is required to determine the influence of the RV-PA conduit on long-term outcomes. (J Am Coll Cardiol 2008;52:52–9) © 2008 by the American College of Cardiology Foundation

After the initial description of the technique by Norwood et al. (1,2), the use of a right ventricle to pulmonary artery (RV-PA) conduit as the source of pulmonary blood flow at stage one palliation (S1P) for hypoplastic left heart syndrome (HLHS) has recently been pioneered by Sano et al. (3,4). In some series, the use of the RV-PA conduit at S1P has been associated with improved surgical and interstage mortality when compared with the use of a modified Blalock-Taussig shunt (BTS) (5–7). Improved coronary and splanchnic flow resulting from increased post-operative diastolic pressure in the RV-PA conduit group could be an explanation for this observed improved short-term survival. Studies evaluating survival through and after bidirectional cavopulmonary anastomosis (BCPA) after an RV-PA conduit are limited. A requirement for earlier palliation with a BCPA, largely attributed to an earlier onset of significant cyanosis, has been reported (8). In at least one report, the type of shunt used for pulmonary blood flow had no demonstrated survival benefit through stage two palliation (9,10).

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Because of the necessity of a ventriculotomy with a RV-PA conduit and the ensuing potential for late development of ventricular dysfunction, aneurysms and/or arrhythmias, it is unknown whether the early survival benefit will continue through mid- and long-term follow-up (11–14). Clearly, at BCPA completion, the immediate impacts of the RV-PA conduit to diastolic blood flow to distal organ beds, growth of pulmonary arteries, and alterations in ventricular volume loading have been removed. Reports of impaired growth and increased need for intervention on the pulmonary arteries with RV-PA conduit through follow-up have raised further concerns (15).

To date, there is no published comparison of perioperative course or survival through Fontan completion of 2 cohorts of patients undergoing each technique, during the same era at the same institution. In this study, we retrospectively analyzed a cohort of 80 patients who contemporaneously underwent either the RV-PA conduit or modified BTS during S1P for single ventricle defects at our institution and who are now of the age for consideration for Fontan completion. We under took this study 1) to examine whether the clinical course and perioperative outcomes differ between the 2 groups at Fontan completion; 2) to define the rate of pulmonary artery interventions for the 2 groups beginning at stage one palliation through Fontan completion; and, finally, 3) to determine the impact of the RV-PA conduit on survival on the entire cohort through the usual period of Fontan completion.

Methods

Study design. The study design is a retrospective case series including all patients who underwent a S1P at our institution from January 1, 2002, through December 31, 2003. The Committee for Clinical Research at Children's Hospital Boston, Boston, Massachusetts, approved the review of patient records for this study. During the study period, 2 approaches to surgical S1P were used at our institution: one that used a modified BTS and one that used a RV-PA conduit. The choice of operative approach at S1P was at the discretion of the patient's surgeon and cardiologist (16).

Patients were grouped by the type of shunt used at S1P (a modified BTS or a RV-PA conduit). The early S1P perioperative, interstage and perioperative courses, and outcomes after BCPA surgery from this cohort of patients have been previously described (9,16). Our institutional practice for final palliation of patients with single ventricle lesions typically involves a complete echocardiogram and hemodynamic catheterization at 18 to 24 months of age followed by a Fontan operation at 24 to 30 months. In this study, only patients who underwent Fontan completion at our institution were included in the description of perioperative Fontan course.

All patients were followed until death, loss to follow-up, or June 1, 2007. It was recorded whether patients had died, underwent Fontan completion, were converted to, or were in the process of being converted to a biventricular circulation. Demographic and anatomical data were reviewed for all patients. For the purpose of multivariable hazard testing, relevant S1P perioperative variables were also reviewed for all patients.

Abbreviations and Acronyms

BCPA = bidirectional cavopulmonary anastomosis

BTS = Blalock-Taussig shunt

HLHS = hypoplastic left heart syndrome

RV-PA = right ventricle to pulmonary artery

S1P = stage one palliation

For those who underwent a Fontan procedure at our institution, the last complete echocardiogram and catheterization before the procedure were included for analysis. All catheterization and operative reports from the time of stage one palliation (S1P) through June 1, 2007, were reviewed to determine the number and type of interventions on the pulmonary arteries. Both surgical patch plasty and/or catheter-based balloon dilation, with or without stent placement, were recorded for all patients undergoing Fontan through the follow-up period.

The type of Fontan procedure performed, the length of cardiopulmonary bypass time, and the need for additional operative procedures, including atrial septectomy, pulmonary artery plasty, arch augmentation or coarctation repair, or tricuspid valvuloplasty, were recorded. Indicators of clinical course at the time of Fontan, including the length of mechanical ventilation, duration of indwelling chest tubes and amount of chest tube output in the first 48 h after surgery, and the lengths of intensive care unit and hospital stay, were recorded. By reviewing the first 48 h after surgery, we were able to calculate, by the formula modified from Wernovsky et al. (17), a peak inotrope score.

Statistical analysis. Measures of central tendency and dispersion are expressed in terms of median and range as a result of the observed skewness in the distributions of predictor variables. Bivariable associations between demographic, anatomic, and clinical variables and operative group were assessed with the Wilcoxon rank-sum test and chi-square or Fisher exact tests for continuous and categorical variables, respectively. Cumulative survival functions were constructed with Kaplan-Meier estimates and event times compared between subgroups with the use of log-rank tests. Cox proportional hazards survival modeling was used to identify independent risk factors for death for the entire cohort.

The number of hazard regression model covariates is limited by the total number of events (deaths) (18,19). Given the number of events in the present analysis ($n = 22$), we considered for inclusion in the model only those perioperative variables found to be significant in univariate analyses ($p < 0.05$), the principle variable of interest (S1P operative group), and a previously reported strong risk factor for surgical death, the presence of an intact atrial septum at birth (20,21). Both the proportional hazards assumption

and the functional form of continuous model variables were assessed based on cumulative sums of martingale-based residuals (22).

All statistical tests were 2-sided, and type I error was controlled at level 0.05. Analyses were performed using SAS (Version 9.1, SAS Institute, Cary, North Carolina), SPSS (Version 14.0, SPSS Inc., Chicago, Illinois), and Sigmaplot (Version 10.0, Systat Software, Inc., Richmond, California).

Results

Patient outcomes. During the follow-up period, 22 patients died. In the modified BTS group (n = 46), 5 died before discharge after S1P and 6 died during the interstage period before BCPA as has been previously described (Fig. 1) (9,16). In the RV-PA conduit group (n = 34), 5 patients died before discharge after S1P and no patients died in the interstage period before BCPA (9,16). Among those 6 patients with a BTS who died in the interstage period before BCPA, 5 were followed after S1P by the referral institution. Compared with those patients who resided in the U.S., underwent a BTS, and returned to have the BCPA at our institution (n = 27), those who suffered interstage death before BCPA demonstrated trends toward increased distance between primary residence and site of S1P (47.5 miles [3.45 to 1,225.9 miles] for interstage survivors vs. 724 miles [28.7 to 1,102.1 miles] for interstage nonsurvivors, p = 0.07) and increased distance between primary residence and site of interstage follow-up (18.7 miles [1.7 to 207.5 miles] for interstage survivors vs. 72.1 miles [3.4 to 178.9 miles] for interstage nonsurvivors, p = 0.12). No patient in either operative group died before discharge after BCPA. Two in each group died in the interstage period before Fontan. No patient in the RV-PA conduit group died after Fontan, whereas in the modified BTS group, 1 died before discharge after Fontan and one died in the follow-up period 12.5 months after Fontan completion. For all patients during the entire follow-up period, there was no association between distance between place of primary residence and site of S1P and survival (391.3 miles [1.43 to 2,585.25 miles] for survivors vs. 266.8 miles [0 to 1,102.1 miles] for nonsurvivors, p = 0.66).

Four patients in the RV-PA group were cannulated for extracorporeal membrane oxygenation in the post-operative S1P period versus 4 in the modified BTS group (p = 0.54). Forty-nine patients underwent Fontan completion at our institution (23 in the RV-PA conduit group vs. 26 in the modified BTS group), whereas 6 underwent Fontan completion at another institution (1 in the RV-PA conduit group vs. 5 in the modified BTS group). Two patients in the RV-PA conduit group underwent conversion to a 2-ventricle circulation after BCPA, whereas in the modified BTS group 3 patients underwent a 2-ventricle repair after S1P and 1 patient after the Fontan procedure. In the RV-PA conduit group, 1 patient is awaiting Fontan completion (Fig. 1).

Pre-Fontan characteristics. Twenty-six patients of the initial 46 (56.5%) who had a modified BTS during their

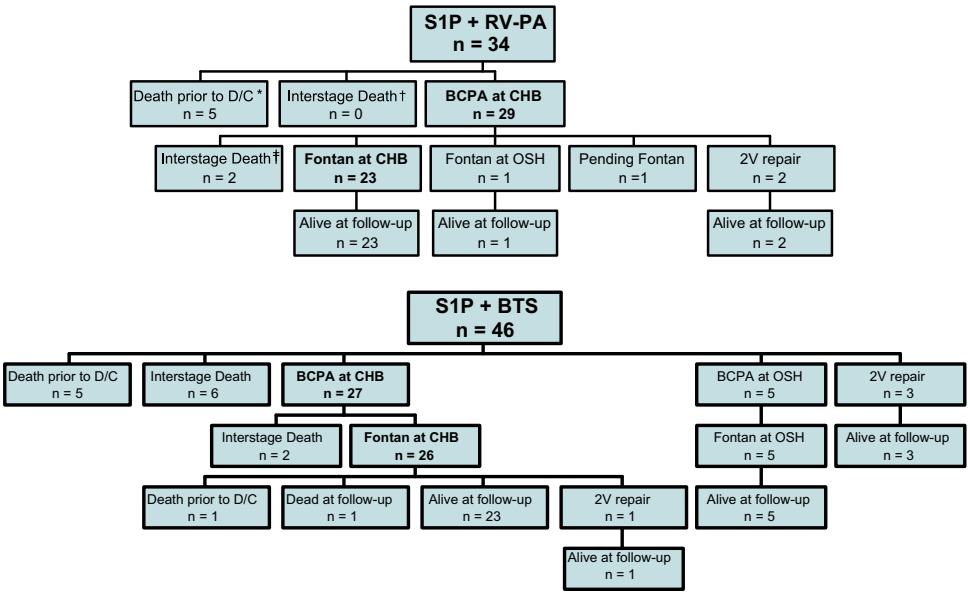

Figure 1 Outcome of 80 Patients Who Underwent S1P From 2002 Through 2003

A tree of outcomes is presented for both RV-PA and BTS. *p = 0.74 between 2 groups (Fisher exact test), †p = 0.04 between 2 groups (Fisher exact test), and ‡p = 1.0 between 2 groups (Fisher exact test). BCPA = bidirectional cavopulmonary anastomosis; BTS = modified Blalock-Taussig shunt; CHB = Children's Hospital Boston; D/C = discharge; OSH = outside hospital; RV-PA = right ventricle to pulmonary artery shunt; S1P = stage one palliation; 2V repair = biventricular repair.

Table 1 Demographics and Cardiac Morphology of Patients Undergoing Fontan

	BTS (n = 26)	RV-PA Conduit (n = 23)	p Value
Demographics			
Age at Fontan (yrs)	2.3 (1.4–5.3)	2.6 (1.6–4.3)	0.11
Weight (kg)	11.6 (8.4–16.3)	11.3 (8.8–16.3)	0.89
BSA (kg/m^2)	0.52 (0.42–0.62)	0.51 (0.42–0.66)	0.84
Gender, male	14/26 (54%)	12/23 (52%)	0.91
Morphology			
HLHS type			
MA/AA	4/26 (15%)	7/23 (30%)	0.31*
MS/AA	7/26 (27%)	3/23 (13%)	0.30*
MA/AS	0/26 (0%)	1/23 (4%)	0.47*
MS/AS	11/26 (42%)	10/23 (43%)	0.93
Non-HLHS			
DORV-MA	3/26 (12%)	1/23 (4%)	0.62*
Shone's syndrome	1/26 (4%)	0/23 (0%)	1.0*
DILV	0/26 (0%)	1/23 (4%)	1.0*

Values are median (range) or frequency (column percent) for continuous and categorical variables, respectively. p values were obtained from Wilcoxon rank-sum test (continuous variables) and chi-square test (categorical variables), except where noted. *Fisher exact test.

AA = aortic atresia; AS = aortic stenosis; BSA = body surface area; BTS = Blalock-Taussig shunt; DILV = double inlet left ventricle; DORV = double outlet right ventricle; HLHS = hypoplastic left heart syndrome; MA = mitral atresia; MS = mitral stenosis; RV-PA = right ventricle to pulmonary artery.

initial S1P have subsequently undergone Fontan completion at our institution, compared with 23 of 34 (67.6%) of those who underwent a RV-PA (p = 0.44). There were no differences between the 2 groups in the age or weight at Fontan completion, gender, the presence of morphology other than HLHS, or subtype of HLHS morphology (Table 1). There were no differences between the groups in pre-operative grades of echocardiographically assessed systemic atrioventricular valve regurgitation, neoaortic regurgitation, or ventricular dysfunction. Similarly, no differences were observed in cardiac index, systemic ventricular end-diastolic pressure, mean pulmonary artery pressure, pulmonary vascular resistance, common atrial pressure, or ratio of pulmonary to systemic blood flow the at pre-operative catheterization (Table 2). There were no differences between the 2 groups during these catheterizations in the numbers of aortopulmonary collaterals coiled (0 [0 to 3] in the RV-PA shunt group vs. 0 [0 to 7] in the modified BTS group, p = 0.69) or in the number of patients who underwent balloon dilations of residual aortic coarctation (3 [13%] in the RV-PA shunt group vs. 7 [26.9%] in the modified BTS group, p = 0.24).

Pulmonary artery and other interventions since S1P. Although there was a trend toward increasing number of patients in the RV-PA shunt group who had Fontan completion with at least one intervention on the pulmonary arteries since S1P at 5 years of follow-up, either surgical patch plasty or catheter balloon dilation with or without a stent, this did not reach statistical significance (15 [65.2%] in the RV-PA shunt group vs. 12 [46.2%] in the modified BTS group, log-rank = 0.09) (Fig. 2).

Fontan operative course. Forty-seven patients underwent a lateral tunnel Fontan procedure, and 2 patients had an extracardiac conduit Fontan. A fenestration allowing shunting of blood from the venous baffle to the left atrium was created for all patients. All previous BCPA palliations were bidirectional Glenn procedures.

Total bypass time for Fontan completion was similar between the 2 groups. Although Fontan completion in patients in the RV-PA conduit group involved an increased use of aortic-cross clamps (20/23 vs. 11/24; p < 0.01), the duration of cross-clamp use was similar between groups of those exposed to it (53.5 min [13 to 77 min] for the RV-PA group vs. 46 min [33 to 107 min] p = 0.89) (Table 3). In cases in which no aortic cross-clamp was used, induced fibrillation of the heart during hypothermic cardiopulmonary bypass was used. The use of circulatory

Table 2 Echocardiographic and Catheterization Variables Before Fontan

	BTS		RV-PA Conduit		
	Total No.	Value	Total No.	Value	p Value
Echocardiography					
AVV regurgitation, 0 (none) to 4 (severe)	26	2 (0–4)	23	2 (0–3)	0.09
Neoaortic regurgitation, 0 (none) to 4 (severe)	26	0.5 (0–4)	23	0 (0–2)	0.63
SV dysfunction, 0 (none) to 4 (severe)	26	0 (0–4)	23	1 (0–3.5)	0.18
Aortic arch obstruction, 0 (none) to 4 (severe)	26	0 (0–2)	23	0 (0–2)	0.39
Catheterization					
Cardiac index (l/min/m^2)	25	3.2 (2–4.4)	18	3.4 (2–6.1)	0.30
SVedp (mm Hg)	25	10 (4–19)	22	8.5 (3–18)	0.26
Mean PAp	26	12.5 (9–24)	22	12.5 (9–18)	0.27
Rp (U/m^2)	24	1.7 (0.9–4)	18	1.8 (0.5–4)	0.69
Atrial pressure (mm Hg)	25	10 (6–16)	22	8.5 (4–12)	0.13
Qp:Qs	22	0.68 (0.5–1.8)	15	0.65 (0.42–1.0)	0.45

Values are median (range) or frequency (column percent) for continuous and categorical variables, respectively. p values were obtained from Wilcoxon rank-sum test (continuous variables), Armitage test for trend (ordinal variables).

AVV = atrioventricular valve; BTS = Blalock-Taussig shunt; PAp = pulmonary artery pressure; Qp:Qs = ratio of pulmonary to systemic flow; Rp = pulmonary vascular resistance; RV-PA = right ventricle to pulmonary artery; SV = single ventricle; SVedp = single ventricle end-diastolic pressure.

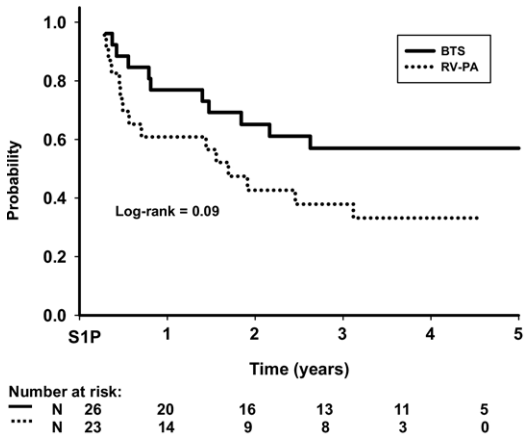

Figure 2 **Kaplan-Meier Plot of Survival After S1P by Type of Pulmonary Arterial Shunt**

Freedom from pulmonary artery intervention after S1P is shown. Abbreviations as in Figure 1.

arrest was limited to one patient in the modified BTS group.

The number of patients who received at least 1 additional operative procedure at the time of Fontan was similar as well between the groups (12 [52.2%] in the RV-PA group vs. 12 [46.2%], p = 0.67). Seven patients underwent tricuspid valvuloplasty at the time of Fontan, 3 in the RV-PA conduit group and 4 in the modified BTS group. One patient in the RV-PA conduit group underwent transverse arch augmentation, whereas one patient in each group underwent enlargement of the atrial septal defect. One patient underwent the placement of a dual-chamber pacing system in the modified BTS group, whereas one patient had epicardial leads placed in the RV-PA conduit group. A similar number underwent pulmonary artery patch augmentation at the time of Fontan (6 [26.1%] in the RV-PA conduit group vs. 8 [30.7%], p = 0.72) (Table 3).

Fontan post-operative course. The post-operative course for patients in both groups was similar. There were no differences observed in the peak inotrope score in the first 48 post-operative hours. The amount of chest tube drainage was similar between the 2 groups in the first 48 post-operative hours, as was the total duration of indwelling chest tubes. The difference in total duration of mechanical ventilation between the groups approached statistical significance (21 h [10 to 96 h] in the RV-PA group vs. 26.5 h [7 to 204 h], p = 0.09). There was a shorter length of intensive care stay in the RV-PA conduit group (2 days [0 to 6 days] vs. 4 days [1 to 25 days], p = 0.01); however, the total hospital length of stay was similar between the groups (9 days [5 to 29 days] vs. 10 days [6 to 48 days], p = 0.89) (Table 3).

Survival. Cumulative survival for the entire cohort of 80 patients was 78.7% at 1 year and 71.4% at 3 years. For the patients who underwent a RV-PA conduit during S1P (n = 34), survival at 1 year was 82.4% and 79.4% at 3 years compared with 75.9% at 1 year and 65.8% at 3 years for those who underwent a BTS (n = 46) (log-rank = 0.31) (Fig. 3). Conditional survival analysis of those who patients who underwent a RV-PA conduit, survived the perioperative period, and subsequently were discharged after S1P (n = 29) was 96.6% at 1 year and 93.1% at 3 years after discharge compared with 85.2% and 74% for those who underwent a BTS (n = 41) (log-rank = 0.06) (Fig. 4).

Table 3 **Operative and Post-Operative Courses at Fontan**

	BTS (n = 26)	RV-PA conduit (n = 23)	p Value
Operative variables			
TBT (min)	107.5 (63–189)	94 (64–159)	0.04
Use of aortic cross-clamp	11 (42%)	20 (87%)	<0.01
Aortic cross-clamp time (min)	46 (33–107)	53.5 (13–77)	0.89
Use of circulatory arrest	1 (4%)	0 (0%)	1.0*
Additional operative procedures			
Tricuspid valvuloplasty	4 (15.3%)	3 (13%)	1.0*
Atrial septum resection	1 (4%)	1 (4%)	1.0*
Aortic arch augmentation	0 (0%)	1 (5%)	0.45*
PA patch augmentation	8 (30.7%)	6 (26.1%)	0.72
Postoperative course			
Peak inotrope score	10 (5–22.5)	10 (0–25)	0.80
CT output first 48 h (ml/kg/day)	35.5 (14–117.5)	35.5 (8.2–109.4)	0.98
Duration of indwelling CT (days)	6.5 (2–31)	6 (2–21)	0.78
Duration of ventilation (h)	26.5 (7–204)	21 (10–96)	0.09
Length of ICU stay (days)	4 (1–25)	2 (0–6)	0.01
Length of hospital stay (days)	10 (6–48)	9 (5–29)	0.89

Values are median (range) and frequency (column percent) for continuous and categorical variables, respectively. p values were obtained from Wilcoxon rank-sum test (continuous variables) and chi-square or Fisher exact test (categorical variables). *Fisher exact test.

BTS = Blalock-Taussig shunt; CPB = cardiopulmonary bypass; CT = chest tube; ICU = intensive care unit; PA = pulmonary artery; RV-PA = right ventricle to pulmonary artery; TBT = total bypass time;

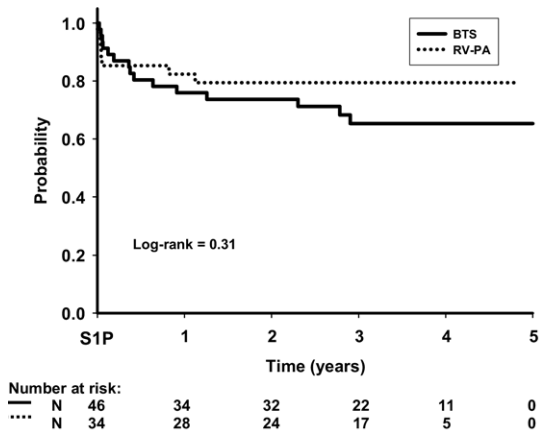

Figure 3 **Kaplan-Meier Plot of Freedom From Pulmonary Artery Intervention After S1P by Type of Shunt**

Cumulative survival after S1P by shunt. Abbreviations as in Figure 1.

A multivariable model was constructed to test the independent impact of shunt type at S1P on survival through Fontan completion using perioperative S1P variables found to be statistically significant in univariate testing. The perioperative S1P variables that did not reach significance in univariate survival analysis were gender, fetal diagnosis, ascending aorta size, and birth weight. Controlling for the use of peritoneal dialysis and extracorporeal membrane oxygenation after S1P, the presence of hypoplastic left heart syndrome, and an intact atrial septum before S1P (all variables found to be significant in univariate testing), the use of a RV-PA conduit was not associated with improved survival (hazard ratio: 1.2, 95% confidence interval: 0.4 to 3.1, p = 0.75) (Table 4).

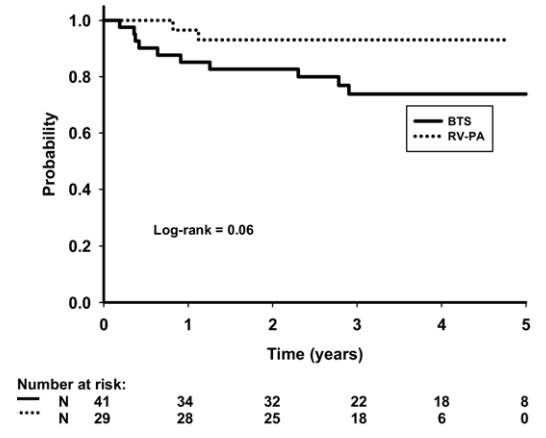

Figure 4 **Kaplan-Meier Plot of Conditional Survival After Discharge After S1P by Type of Shunt**

Survival after S1P discharge by shunt type. Time zero indicates discharge after S1P. Abbreviations as in Figure 1.

Table 4 **Independent Risk Factors at S1P for Death Through Fontan Completion***

Stage One Palliation Variables	Hazard Ratio†	95% Confidence Interval	p Value
RV-PA conduit	1.2	0.4–3.1	0.75
Intact atrial septum	1.6	0.6–4.2	0.38
Non-HLHS morphology	3.6	1.3–10.2	0.02
Post-operative peritoneal dialysis	5.6	1.5–21.1	<0.01
Post-operative ECMO	5.7	2.0–15.9	0.01

*n = 80; total number of deaths = 22. †Hazard ratios based on a 1 unit increase in the value of the corresponding covariate.

ECMO = extracorporeal membrane oxygenation; HLHS = hypoplastic left heart syndrome; RV-PA = right ventricle to pulmonary artery.

Discussion

We report a single institution's experience with Fontan completion and overall survival in a cohort of patients who underwent S1P with either a modified BTS or RV-PA conduit concurrently. We show no statistical differences between the 2 groups in pre-operative variables or perioperative outcomes for those undergoing Fontan completion, with the exception of decreased intensive care unit length of stay for those undergoing a RV-PA conduit. When the entire cohort of patients was examined, a trend toward improved cumulative survival up to 3 years after S1P for those who underwent a RV-PA conduit was demonstrated, although this difference did not reach statistical significance. Additionally, our survival rate of approximately 75% at 5 years for the RV-PA conduit is similar to that described by Sano et al. (23) in a recent noncomparative experience. Similarly, conditional survival analysis of those who patients who survived and subsequently were discharged after S1P, suggests a strong, albeit statistically nonsignificant, trend toward improved survival for the RV-PA conduit group through the period of Fontan completion. These data suggest that potential longitudinal survival advantage for the RV-PA conduit might exist after the perioperative S1P period, through the interstage period before BCPA and without decrement through Fontan completion. However, given the lack of statistically significant differences in survival in univariate or multivariable testing of our data, either cumulative or conditional, we must conclude that if actual differences exist between the 2 groups, their statistical demonstration will require greater numbers of patients with extended longitudinal follow-up.

These data should encourage the long-term longitudinal follow-up of those patients currently enrolled in the Single Ventricle Reconstruction Trial sponsored by the Pediatric Heart Network, a randomized prospective study evaluating mortality in patients undergoing a modified BTS or RV-PA conduit during S1P for single ventricle physiology.

We examined the influence on survival of the distance between home residence and site of S1P and the distance between home residence site of interstage follow-up. We found a nonstatistically significant trend towards decreased interstage survival before BCPA, but not in overall survival

through follow-up, in those patients remote in home residence from the institution of S1P and site of interstage follow-up.

Previous investigations by Ghanayem et al. (24) have found significant improvement in interstage survival before BCPA in patients enrolled in an aggressive interstage monitoring and follow-up program. Our institution, the site of S1P for these patients, does not use interstage daily home saturation monitoring and weight checks as described by the Wisconsin group. Given the findings, we are reexamining interstage follow-up for all patients, to ensure consistency and optimal outcomes.

The hemodynamics of the 2 groups before Fontan completion at our institution were remarkably similar. Although they were subjectively graded by echocardiography and retrospectively reviewed, the degrees of ventricular dysfunction and atrioventricular valve regurgitation were essentially identical. Similarly, Sano et al. (25) found no difference between groups in pre-Fontan ventricular function by echocardiography in a total of 14 patients undergoing Fontan. Although at least 1 group has reported decreased ventricular contractility at the time of pre-operative Fontan catheterization associated with the RV-PA conduit, our data revealed no differences in cardiac index or systemic ventricular end-diastolic pressures (26). Thus, ventricular dysfunction was not a factor found to alter Fontan course or survival between the 2 groups.

We found that mean pulmonary artery pressures and pulmonary vascular resistances between the groups were similar at pre-operative catheterization. Although indices of pulmonary artery size were not serially measured in our patients over time, other groups have found that, despite a significant incidence of central pulmonary artery hypoplasia at the insertion of the conduit, distal pulmonary artery growth as assessed by the Nakata or McGoon technique was increased with RV-PA conduit (10,27). Similar to other centers that have reported an increase in the need for pulmonary artery interventions in the RV-PA conduit group before or at the time of BCPA, we found a trend toward increased need for pulmonary artery intervention in this group through Fontan completion (5,28).

Interestingly however, the rate of chest tube output, as well as the duration of indwelling chest tubes was nearly identical between the groups at Fontan completion. These data would suggest to us that residual pulmonary artery stenosis had adequately been addressed either operatively or by balloon dilation before and at the time of Fontan completion and also suggest that there were no important clinical difference in post-operative Fontan baffle pressures that might result from residual pulmonary arterial stenosis or hypoplasia. Thus, our experience suggests that although patients who receive a RV-PA conduit at S1P might require more total interventions to the pulmonary arteries through Fontan completion, this reintervention rate has not negatively altered perioperative Fontan course. Future sources of study should include evaluations of the need for pulmonary artery interventions in the RV-PA conduit group after Fontan and the incidence of late pulmonary arterial hypoplasia.

The operative courses were similar between groups despite the use of aortic cross-clamp in a greater number of patients in the RV-PA conduit group. The post-operative courses were different in several respects. There was a trend toward increased duration of ventilation in the modified BTS group; however, it did not reach statistical significance in this study. The intensive care unit length of stay was increased in the modified BTS group; however, the total length of hospital stay was similar for both groups. The disparate nature of these findings can not be readily explained in our data without further examination.

Study limitations. This study is limited by its retrospective nature and the biases inherent in this type of study design. The patients were not randomized, and there is an underrepresentation of patients in the RV-PA conduit group. On the basis of this nonrandom approach, unidentified risk factors between the groups could significantly impact outcomes without detection. The cohort is small; thus, only large differences in comparisons will be able to be statistically identified. Multivariable modeling with these relatively small numbers of outcomes is subject to overfitting, although an attempt was made to limit the number of variables included for adjustment, including only those variables reaching statistical significance ($p < 0.05$).

Given the small difference in survival through Fontan completion that we found between the 2 groups, one would need to enroll approximately 125 patients in each group in a randomized prospective trial of comparing the two techniques for this difference to reach statistical significance. Because not all patients had all subsequent palliations and follow-up at our institution and because one patient has yet to complete palliation with a Fontan operation, important differences between the groups could fail to be identified.

Conclusions

Our data suggest that the pre-operative characteristics at Fontan completion are similar between the 2 groups. Despite similar pre-operative characteristics, including similar echocardiographic measurements of ventricular function, the use of a RV-PA conduit at S1P was associated with decreased intensive care unit length of stay but not hospital length of stay. Follow-up through and shortly after Fontan completion in this cohort suggest nonstatistically significant improved survival at the expense of increased need for pulmonary artery interventions. However, multivariable modeling did not reveal the use of a RV-PA to be an independent predictor of survival at this follow-up.

This work provides the first description of Fontan completion in 2 groups of patients who have undergone concurrently either a modified BTS or RV-PA conduit during S1P. These findings should be verified in a larger cohort that has been randomized to either technique at stage one

palliation. Further, prospective longitudinal follow-up of these patients will be required to evaluate pulmonary artery growth and the incidences of late ventricular dysfunction and arrhythmias.

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