Risk Factor Analysis for Second-Stage Palliation of Single Ventricle Anatomy

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Background. **Single ventricle hearts can be surgically palliated by a series of operations culminating in the Fontan procedure, which establishes a total cavopulmonary connection. The second-stage procedure creates a physiologic connection between the superior vena cava and the pulmonary artery.**

Methods. **From 1998 to 2010, 557 patients with single ventricle heart disease underwent second-stage surgical palliation. This cohort was retrospectively analyzed to assess patient outcome by a number of anatomic, physiologic, and procedural factors. The analysis excluded patients undergoing hybrid first-stage procedures.**

Results. **The median age at operation was 165 days (range, 59 days to 49 years). The most common anatomic subtypes were hypoplastic left heart syndrome (52%), tricuspid atresia (12%), unbalanced atrioventricular septal defect (10%), double inlet left ventricle (9%), or other (17%). Left ventricular hypoplasia was present in 70%. A hemi-Fontan procedure was done in 89%, and 11% received a bidirectional Glenn. Concomitant atrioventricular valve repair was necessary in 9%. Early mortality was 4.7%, and 5.9% died after discharge but before Fontan. No early or late deaths occurred in patients with tricuspid atresia and double inlet left ventricle. Multivariate analysis demonstrated ventricular dysfunction, atrioventricular valve regurgitation, and unbalanced atrioventricular septal defect were significant adverse risk factors for survival to Fontan.**

Conclusions. **Second-stage palliation can be performed at low risk for patients with left ventricular dominance, but significant risk remains for patients with left ventricular hypoplasia and unbalanced atrioventricular septal defect. Atrioventricular valve insufficiency is a persistent problem that has not been neutralized by repair strategies.**

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Surgical creation of a total cavopulmonary connection has been established as the goal for the management of patients with a functionally univentricular heart [1, 2]. This is achieved by a series of operations that culminate in the Fontan procedure. The second-stage procedure involves construction of a physiologic connection between the superior vena cava and the pulmonary artery. In practice, this connection is accomplished by the bidirectional Glenn (BDG) or hemi-Fontan procedure (HFP). Second-stage palliation has been shown to relieve the volume load on the single ventricle and allow for favorable ventricular remodeling, which has the result of improving outcomes for the eventual Fontan [3–5].

Many reports have documented improvements in outcomes after staged palliation of single ventricle heart disease [6–8]. This study was designed to evaluate a large group of patients undergoing second-stage palliation to identify factors that influence outcome.

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Patients and Methods

All patients undergoing second-stage reconstruction for single ventricle heart disease from 1998 to 2010 were identified through the University of Michigan Congenital Heart Surgery Database. Patient demographics, clinical characteristics, imaging, operative reports, hospital records, and clinical reports were collected, and a retrospective analysis of the data was performed. Patient follow-up was complete in 98%. Institutional Review Board approval was obtained before the initiation of the study.

We defined the primary end point as survival to Fontan (we had recently reported our results with the Fontan operation and we did not want to duplicate the subsequent follow-up) and failure as a composite variable of death, transplant, or unsuitability for Fontan. Early mortality was defined as a death occurring within 30 days or before discharge. Patients undergoing hybrid first stage procedures were excluded from this analysis.

Univariate analysis was performed to identify factors significantly linked to outcome. Normally distributed continuous variables were analyzed using the Student *t* test. The Fisher exact test was used for binary variables, and the Wilcoxon rank sum test was used for nonparametric variables. Multivariable models were tested with

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logistic regression using variables found to be significant by univariate analysis. Data analysis was performed using SPSS 18 software (SPSS, Inc, Chicago, IL). A value of $p < 0.05$ was used to determine significance.

Results

A total of 557 patients (38% female) underwent second-stage palliation. Patient characteristics are summarized in Table 1. The median age at time of repair was 165 days (range, 48 days to 49 years), and the median weight was 6.8 kg (range, 3 to 57 kg). The most common anatomic subtypes were classic hypoplastic left heart syndrome (HLHS) in 52%, tricuspid atresia in 12.2%, unbalanced atrioventricular septal defect (UAVSD) in 10.2%, double inlet left ventricle in 9%, or other in 17%. Preoperative echocardiography was used to establish ventricular dominance. Left ventricular hypoplasia was present in 70% and right ventricular hypoplasia in 30%.

Preoperative assessment of atrioventricular valve (AVV) function and ventricular function was determined by echocardiography performed immediately before op-

Table 1. Univariate Analysis of Preoperative Patient Characteristics in Outcome Groups

Variable[a]	All Patients n = 557	Survival to Fontan n = 408	Failure n = 70	*p* Value
Age, months	5.5 (1.6–601)	5.5	5.3	0.256
Weight, kg	6.00 ± 4.87 (3–57)	6.10 ± 4.77(3–57)	5.25 ± 5.58 (3–37)	<0.0005
BSA	0.31 ± 0.13 (0.16–1.57)	0.32 ± 0.13 (0.16–1.57)	0.29 ± 0.23 (0.19–1.57)	<0.0005
Sex, % male	61.8	62.4	70	0.431
Anatomic diagnosis				
LV dominance	159 (29.4)	148 (31.6)	11 (16.4)	0.011
HLHS	290 (52.0)	252 (52.0)	38 (52.1)	0.988
Tricuspid atresia	66 (12.2)	63 (13.2)	3 (4.3)	0.032
DILV	49 (8.8)	44 (9.2)	3 (4.3)	0.167
UAVSD	56 (10.2)	38 (8.0)	18 (25.7)	<0.0005
TGA				
Dextro	64 (11.5)	55 (11.6)	6 (8.6)	0.459
Levo	58 (10.4)	50 (10.5)	6 (8.6)	0.619
DORV	84 (15.1)	66 (13.9)	16 (22.9)	0.071
PA/IVS	17 (3.0)	16 (3.4)	1 (1.4)	0.385
Dextrocardia	29 (5.2)	24 (5.0)	3 (4.3)	0.785
Ebstein anomaly	8 (1.6)	6 (1.3)	2 (2.9)	0.299
Heterotaxy	47 (8.6)	37 (7.8)	10 (14.3)	0.035
Interrupted IVC	14 (2.5)	13 (2.6)	1 (1.4)	0.632
Echocardiography				
Dysfunction > mild	40 (7.5)	25 (5.3)	15 (21.4)	<0.0005
AVVR > mild	84 (17.7)	66 (13.9)	28 (40.0)	<0.0005
Catheterization data				
EDP, mm Hg	8.00 ± 3.42	8.00 ± 3.50	8.50 ± 2.73	0.043
PAP, mm Hg	14.0 ± 4.77	13.0 ± 4.72	14.0 ± 5.11	0.325
Qp/Qs	1.72 ± .68	1.7 ± .60	1.8 ± .58	0.387
SVC saturation, %	50 ± 8.53	50 ± 8.13	45 ± 10.7	0.014
McGoon ratio	1.51 ± .44	1.53 ± .44	1.38 ± .40	0.037
Nakata index	225.7 ± 139.94	228.06 ± 139.28	200.46 ± 114.83	0.180
Diameter, mm				
RPA	6.50 ± 2.21	6.58 ± 2.171	5.84 ± 1.94	0.017
LPA	6.13 ± 2.13	6.19 ± 2.10	5.58 ± 1.84	0.043
SVC	5.38 ± 1.06	5.38 ± 1.05	5.30 ± 1.14	0.824
SVC/BSA	78.39 ± 34.37	77.45 ± 33.68	81.43 ± 35.44	0.515
SVC/aorta	0.65 ± .16	0.65 ± 0.15	0.67 ± 0.20	0.635

[a] Continuous data are presented as median (range) or median ± standard deviation (range), and categoric data are presented as number (%), or as indicated.

AVVR = atrioventricular valve regurgitation; BSA = body surface area; DILV = double inlet left ventricle; DORV = double outlet right ventricle; dysfunction = ventricular dysfunction; EDP = end diastolic pressure; HLHS = hypoplastic left heart syndrome; IVC = inferior vena cava; LPA = left pulmonary artery; LV = left ventricle; PA/IVS = pulmonary atresia/intact ventricular septum; PAP = pulmonary artery pressure; Qp/Qs = ratio of pulmonary blood to systemic blood flow; RPA = right pulmonary artery; SVC = superior vena cava; TGA = transposition of the great arteries; UAVSD = unbalanced atrioventricular septal defect.

eration. AVV function was normal in 58.2%, whereas insufficiency was identified as mild in 24.1%, moderate in 13.9%, and severe in 3.8%. Dominant ventricular function was normal in 77.1%, and dysfunction was mild in 15.4%, moderate in 6.9%, and severe in 2 patients. Pressures, saturations, and vascular measurements were recorded from cardiac catheterizations performed within 3 weeks of the second-stage palliation. The findings at catheterization are reported in Table 1.

On the basis of institutional preference, isolated HFP was performed in 81% of patients. Isolated BDG was constructed in 7% and was reserved for patients with unfavorable venous anatomy, heterotaxy syndrome, or other unusual complexity. The 12% of patients with bilateral superior vena cavae underwent HFP with contralateral BDG (8%) or bilateral BDGs (4%). Concurrent procedures were performed in 12%, most commonly an AVV repair (9%). Valve repair techniques were individualized to the patient's valve pathology. Patients with HLHS most frequently underwent a partial annuloplasty, whereas other patients underwent a variety of techniques selected by the operating surgeon. Other procedures included AVV replacement in 1 patient and repair of aortic arch obstruction in 3%.

Patient outcomes are illustrated in Figure 1. To summarize, survival to Fontan was 87%. Median follow-up to a primary end point was 18.5 months. Failure occurred due to early deaths (4.7%), late deaths (5.9%), transplantation (1.5%), and poor candidacy for Fontan (1.3%). Most patients who were not candidates for Fontan palliation were listed for transplantation. There were no early or late deaths in patients with tricuspid atresia or double inlet left ventricle. Patients with UAVSD were at greatest risk of early death (12.3%), whereas early death occurred in only 5% of HLHS patients. Early mortality was 5.7% in the context of left ventricular hypoplasia and 1.3% for right ventricular hypoplasia. Complications occurred in 15.8% of patients and are delineated in Table 2. At discharge, normal sinus rhythm was present in 98.2%, and the remainder ultimately required pacemaker implantation.

Univariate analysis was performed to identify risk factors for poor outcome by patient characteristics, anatomy, echocardiographic data, and catheterization data. The results are summarized in Table 1. Significant categoric variables with associated hazard ratios are highlighted in Table 3. Left ventricular dominance and diagnosis of tricuspid atresia were protective factors, whereas age younger than 3 months, heterotaxy syndrome, diagnosis of UAVSD, clinically significant AVV insufficiency (worse than mild), and clinically significant ventricular dysfunction (worse than mild) were associated with poor outcomes. Several continuous variables, including weight, body surface area, end-diastolic pressure, superior vena cava saturation, McGoon ratio, and pulmonary artery diameter showed significant differences in the two outcome groups, but no critical values or cutoffs could be identified to guide patient selection. During the first-stage palliation, 85% of patients underwent shunt placement, of which 91% received a systemic-to-pulmonary artery shunt and 9% underwent placement of a right ventricle-to-pulmonary artery shunt. The difference in outcomes by shunt type was not statistically significant ($p = 0.08$).

Multivariate analysis was performed using logistic regression. A model was constructed using preoperative variables that were statistically significant by univariate analysis. This model determined that three factors were statistically independent prognostic risk factors: clinically significant (more than mild) AVV regurgitation (odds ratio, 2.33; $p = 0.008$), clinically significant (more than mild) ventricular dysfunction (odds ratio, 3.77; $p = 0.001$),

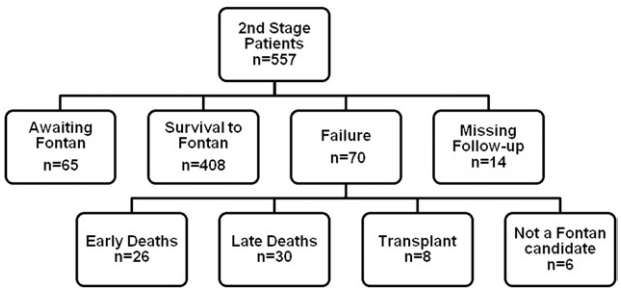

Fig 1. Flowchart demonstrates outcomes for all patients undergoing second stage reconstruction.

Table 2. Complications

Complication	No. (%)
Total complications	88 (15.8)
Chylothorax	11 (2)
Diaphragm paralysis	37 (6.6)
Extracorporeal membrane oxygenation	8 (1.4)
Hypoxemia requiring reintervention	36 (6.4)
Mediastinitis	4 (0.7)
Pathway thrombosis	15 (2.7)
Renal failure	15 (2.7)
Seizure	20 (3.6)
Sepsis	12 (2.2)
Stroke	6 (1.1)
Takedown	10 (1.8)

Table 3. Univariate Analysis of Significant Preoperative Categoric Variables

Variable	HR	95% CI	*p* Value
Age <3 months	4.91	2.13–11.30	0.001
Dominant left ventricle	0.47	0.24–0.90	0.011
Tricuspid atresia	0.29	0.08–0.95	0.032
UAVSD	3.58	1.90–6.76	<0.0005
Heterotaxy	2.27	1.10–4.7	0.035
Ventricular dysfunction > mild	5.48	2.75–10.89	<0.0005
AVVR > mild	3.48	2.00–6.05	<0.0005

AVVR = atrioventricular valve regurgitation; CI = confidence interval; HR = hazard ratio; UAVSD = unbalanced atrioventricular septal defect.

and the anatomic diagnosis of UAVSD (odds ratio, 2.79; $p = 0.016$).

Concomitant AVV repair was performed in 50 patients and was successful (defined as mild or less insufficiency at the time of discharge) in 27 (54%). The repair was durable in 74% of these patients as assessed at the time of Fontan or latest follow-up. An additional 5 patients in the early repair failure group crossed over to the late successful repair group due to subsequent AVV repair (n = 1) or improvement in ventricular function (n = 4). Overall, 25 patients (50%) had a successful late valve repair outcome, and among these, survival to Fontan was 88%. In contrast, only 32% of patients with an unsuccessful late valve repair outcome survived to Fontan ($p < 0.0005$). No correlation between valve morphology and repair outcome was identified.

Early reintervention for hypoxemia was required in 36 patients (6%). Cardiac catheterization was performed in all of these patients to delineate the cause of hypoxemia. A clear anatomic defect (pulmonary artery stenosis or baffle leak) could be identified in 16 patients. Correction of the defect resulted in a survival of 75%. In contrast, no anatomic defect could be identified in 20 patients. These patients underwent addition of a small shunt to the hemi-Fontan pathway (n = 10) or takedown of the HFP to a shunted circulation (n = 10). Mortality was 90% for the former group and 80% for the latter group. Absence of an anatomic defect in patients requiring early reintervention for hypoxemia represented a significant risk ($p = 0.001$).

Comment

Staged palliation of the functionally univentricular heart, culminating in a total cavopulmonary connection, has dramatically improved patient outcomes. The goal of second-stage reconstruction includes early reduction of the volume load imposed by shunt physiology, allowing for favorable ventricular remodeling. This reduction in volume load results in a smoother transition to Fontan physiology achieved in the final stage [6].

Our center adopted the HFP in 1993, and we remain strong supporters of this technique, which is used for most patients. One advantage of the HFP, as we perform it, includes the routine augmentation of the proximal branch pulmonary arteries [6]. Many patients, especially those with HLHS, will have some degree of narrowing of the left pulmonary artery near the ductal insertion or narrowing of the right (or left) pulmonary artery at the site of shunt insertion. These lesions are routinely managed at the time of HFP, and late intervention on the pulmonary arteries is exceedingly rare in our patient population. This ensures an optimal anatomic substrate for the eventual Fontan. Our center also prefers the lateral tunnel Fontan procedure for ultimate palliation. The HFP provides the optimal intermediate stage for progression to the lateral tunnel Fontan [1].

A final advantage of the HFP is the theoretic benefit of providing a more energetically favorable total cavopulmonary connection. Computer flow-modeling demonstrates that the anteroposterior caval offset created by the HFP and subsequent lateral tunnel Fontan results in much less energy loss than the BDG and extracardiac conduit Fontan or other anatomic arrangements [9].

The greatest recognizable disadvantage of the HFP involves its technical difficulty and requirement for the use of cardiopulmonary bypass with brief cardioplegic arrest. We believe that the advantages of the HFP outweigh its difficulty. The HFP and subsequent lateral tunnel Fontan techniques have also been questioned due to the potential risk of atrial dysrhythmias, including sick sinus syndrome and late tachyarrhythmias. We and others have seen a low late incidence of atrial dysrhythmias and no significant impact of the HFP on sinus node function [10–12]. Much longer-term follow-up (on the order of decades) will be necessary to determine ultimate outcomes with respect to energetics and arrhythmia development.

Our center previously reported the results of the HFP for patients with HLHS in 1998 [6]. Early mortality in this group of 114 patients was 2%, as was late mortality. The mortality in this current study is much higher, including a second interstage mortality rate of nearly 6%. This increase in death appears to be secondary to an increase in patient complexity. As we have gained confidence with first-stage and second-stage palliation, we have been more aggressive about accepting higher-risk patients. In addition, as our management of first-stage patients has improved, a greater number of patients are considered candidates for second-stage palliation, many of whom would not have survived to be considered in the past.

The primary goal of this study was to identify preoperative risk factors for poor outcome after second-stage palliation. Univariate analysis revealed a series of significant factors, and subsequent multivariate logistic regression determined that three of these were statistically independent prognostic factors: significant AVV regurgitation, significant ventricular dysfunction, and the anatomic diagnosis of UAVSD. Clearly, these factors are anatomically and physiologically linked.

The importance of significant AVV regurgitation cannot be overemphasized, especially because it is the only one of these factors that we can potentially modify [13]. Our subgroup analysis was performed to determine the effect of AVV repair on patient outcome. Removal of the volume load during second-stage reconstruction is recognized to result in some improvement in AVV function [14]. Currently at our center, AVV repair is performed at the time of second-stage palliation routinely in patients with more than moderate insufficiency and selectively in patients with moderate insufficiency [15].

Valve repair comprises some form of annuloplasty (most frequently in HLHS patients) and more complex forms of valve repair, including commissuroplasty, chordal shortening, edge-to-edge repair, and other variations of leaflet reapproximation [16, 17]. Repair of the dysfunctional common AVV typically required multiple techniques. In the current study, 50 patients underwent concomitant AVV repair, but a successful early repair was achieved in only 54%. Late successful valve repair

was found in 50%. Successful valve repair was accompanied by a much more favorable outcome. No correlation between valve morphology and repair outcome was identified. These data suggest that a more aggressive approach to valve repair, including reintervention, if necessary, is indicated in patients with significant AVV insufficiency. Dominant ventricular dysfunction certainly represents a confounding variable, and normal AVV function may be impossible to achieve in patients with underlying dysfunction.

The diagnosis of UAVSD represents a very high-risk group of patients, likely due to the common association with other complex anatomic defects and the frequent occurrence of AVV insufficiency, which can be very difficult to repair surgically [18]. Interestingly, our analysis showed that even in the absence of associated defects and physiologic derangements, the diagnosis of UAVSD appears to confer risk. We sought to identify additional prognostic factors based on pulmonary artery size, superior vena cava diameter, end-diastolic pressure, and pulmonary artery pressure, and although some trends were observed, no critical values could be determined that could be used as criteria for optimal patient selection.

An interesting subgroup of patients required early reintervention for hypoxemia (6% of cases). Catheterization revealed an anatomic defect (pulmonary artery stenosis or baffle leak) in 44%. Not surprisingly, correction of the defect resulted in favorable outcomes. The remaining 56% of patients had no recognizable anatomic defect, and hypoxemia was secondary to inadequate pulmonary blood flow. The identification of "pop-off" veins was common, but coiling or other embolization of these proved to be nearly universally futile. Management of these patients involved one of two strategies: addition of a small central aortopulmonary shunt to the hemi-Fontan pathway or takedown of the second-stage reconstruction to a systemic-to-pulmonary shunt. Neither strategy was advantageous, because the outcomes were uniformly poor. Owing to the extremely high-risk nature of this complication, the data suggest that early listing for cardiac transplantation should be considered.

In summary, second-stage palliation can be achieved at low risk, especially for patients with left ventricular dominance. Despite improvements in technique and patient management, significant risk remains for certain subgroups. Clinically significant AVV regurgitation and ventricular dysfunction, as well as the anatomic diagnosis of UAVSD, were statistically independent risk factors for poor outcomes. Successful late AVV repair mitigates risks and improves candidacy for Fontan. Patients with early hypoxemia without anatomic defect are at particularly high risk and should be considered for transplantation.

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DISCUSSION

DR TARA KARAMLOU (Seattle, WA): Great presentation, really nice job. You showed your results with atrioventricular (AV) valve repair. One of the things that I think some of us struggle with is the timing of the AV valve repair, and whether, when you unload the ventricle and do your second-stage repair, if some of the AV valve insufficiency may be mitigated by doing that second stage. You mention an aggressive approach, and I wonder if you could educate us all as to what your current criteria for intervening on the AV valve are.

MR LEE: What we currently do is choose to operate on patients who have moderate or severe regurgitation or insufficiency. We either do an annuloplasty for those patients who have hypoplastic left heart syndrome or, alternatively, we will do a more complex repair of those who have complex anatomy, such as unbalanced AVSD. I think that the next presentation will probably go over a little better of those patients who do require valve repair among single ventricle patients.

DR KIRK KANTER (Atlanta, GA): Congratulations, as a medical student, that is a very good presentation for a resident or an attending. In your univariate analysis, was one of the variables you looked at whether or not they had a right ventricle-to-pulmonary artery conduit or a Blalock-Taussig shunt at the original palliation?

MR LEE: Yes, we also looked at that, but I didn't find any significant association with failure.

DR STEPHEN LANGLEY (Portland, OR): I'd like to congratulate you, that was a tremendous presentation. Patients with Down syndrome and single ventricle physiology seem a particularly high-risk group. Did you look at the influence of Down syndrome on the outcomes, particularly the patients with an unbalanced atrioventricular septal defect (AVSD)?

MR LEE: We don't have that data currently. I did look at some genetic abnormalities, but I didn't divide it between those with Down syndrome vs some other genetic abnormality. I do believe that is a good question. We'd like to look into that in the future.

DR JOHN MAYER, JR (Boston, MA): At some risk of getting over my statistical head, which is pretty easy, the concern I would have is about how the variables segregate since you have the possibility of highly linked variables, namely, AV valve regurgitation and heterotaxy. Is that an issue? And if it was, how did you deal with it?

MR LEE: Yes, we also looked at that using our multivariate analysis. Actually in my model I included heterotaxy and unbalanced AVSD. And what we found was that there was no confounding effect between the two.