Predictors of outcome after the Fontan operation: Is hypoplastic left heart syndrome still a risk factor?

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Objective: This study was undertaken to evaluate factors contributing to a decrease in early mortality and morbidity after the Fontan procedure between January 1, 1992, and December 31, 1999.

Methods: Outcomes evaluated were early survival, duration of pleural effusions, and duration of hospitalization. Potential predictors evaluated included anatomic diagnosis, presence of a common atrioventricular valve, preoperative pulmonary artery pressure, type of Fontan operation, type of intentional right-to-left shunt or baffle fenestration, and use of modified ultrafiltration.

Results: The modified Fontan procedure was performed in 332 patients at a median age of 22 months (range, 11-380 months) and a median weight of 11 kg (range, 5.8-120 kg). Prior stage I reconstructive surgery for classic or variant hypoplastic left heart syndrome had been performed in 205 (53%) of 332 patients, and 318 (96%) had undergone an interim superior cavopulmonary connection. A lateral-tunnel Fontan operation was performed in 281 patients, and an extracardiac conduit Fontan operation was performed in 51 patients. An intentional right-to-left shunt was created in 298 (90%) patients. Between 1992 and 1999, the outcome after the modified Fontan operation improved significantly. Overall mortality was 6.6% (22/332), with only 2 deaths since 1994. Morbidity was also reduced, with a decreased duration of pleural effusions and decreased hospital stay. In a multivariable analysis of the entire cohort, only the presence of a common atrioventricular valve (odds ratio, 7.64; 95% confidence limits, 2.07-28.14; $P = .0002$) and increased preoperative pulmonary artery pressure (odds ratio, 1.46/1 mm Hg increase; 95% confidence limits, 1.2-1.78; $P < .001$) increased the risk of early death, whereas use of a single-punch fenestration in a lateral-tunnel Fontan (odds ratio, 0.06; 95% confidence limits, 0.01-0.65; $P = .02$) and use of modified ultrafiltration (odds ratio, 0.14; 95% confidence limits, 0.03-0.72; $P = .019$) decreased the risk of death. The risk of prolonged pleural effusions (>3 days) was increased in patients with hypoplastic left heart syndrome (odds ratio, 1.73; 95% confidence limits, 1.07-2.81; $P = .03$) and was decreased by use of a single-punch fenestration in a lateral-tunnel Fontan operation (odds ratio, 0.17; 95% confidence limits, 0.07-0.4; $P < .001$), as well as by the use of modified ultrafiltration (odds ratio, 0.25; 95% confidence limits, 0.15-0.40; $P < .01$).

Conclusions: In a contemporary series of Fontan operations performed largely in patients with hypoplastic left heart syndrome or variants, systemic ventricle morphology had no effect on mortality. Some patient characteristics, however, continue to influence outcome. The decrease in mortality and morbidity in the current era is attributed to changes in management strategies, specifically the use of modified ultrafiltration and baffle fenestration.

Management strategies for neonates and infants with a functional single ventricle have evolved significantly in recent years, with most undergoing a series of staged palliative procedures culminating in a modified Fontan operation. Mortality and morbidity after the Fontan operation have been dramatically reduced in recent years. A variety of modifications of the Fontan operation have been introduced, including interim superior cavopulmonary connection, baffle fenestration, the lateral-tunnel Fontan procedure, and the extracardiac conduit Fontan procedure. Improved outcome after stage I reconstruction for patients with hypoplastic left heart syndrome (HLHS) has led to an increasing number of these patients undergoing the Fontan operation. Previous reports have suggested that HLHS was a risk factor for mortality after the Fontan operation. The current study was undertaken to review our institutional experience with the modified Fontan operation in a cohort of patients composed largely of subjects with HLHS or variants to identify patient and management characteristics that might influence early mortality and morbidity.

Methods and Patients

Patient Population

The study was approved by the Institutional Review Board of The Children's Hospital of Philadelphia. Review of the Cardiology and Cardiac Surgery Databases at The Children's Hospital of Philadelphia identified 332 patients who underwent a modified Fontan operation between January 1, 1992, and December 31, 1999.

Perioperative Data Acquisition

Patient-related variables. Demographic and anatomic variables were recorded from the medical record (Table 1). Patients were assigned to one of 6 diagnostic groups on the basis of cardiac anatomy (Table 2). The atrioventricular (AV) valve anatomy was classified as a common AV valve or "other" on the basis of the preoperative echocardiogram. AV valve regurgitation was classified as none, mild, moderate, or severe on the basis of the preoperative echocardiogram. The mean pulmonary arterial pressure and ventricular end-diastolic pressure were recorded from the preoperative cardiac catheterization.

Procedure-related variables. During the study period, 2 techniques for the creation of the systemic venous pathway were used exclusively: either a lateral tunnel with an intra-atrial baffle or an extracardiac conduit connection. No patient underwent an atriopulmonary connection. Use and type of intentional right-to-left shunt were recorded, as were details of intraoperative management (Table 1).

Postoperative variables. The duration of hospital stay (in days) was recorded. The duration of pleural drainage (in days) was defined as the period from the date of the operation until the last thoracentesis or final removal of pleural catheters. Prolonged pleural effusions were considered as pleural effusions requiring drainage for greater than 3 days. Very prolonged effusions were defined as those necessitating drainage for greater than 14 days. Prolonged hospital stay was defined as 14 days or longer.

TABLE 1. Preoperative variables

Demographics	Age at operation Year of operation Weight
Diagnosis	Single left ventricle with normally related great arteries Single left ventricle with transposed great arteries Single right ventricle HLHS Heterotaxy syndrome Other
AV valve anatomy	Common AV valve Other
AV valve regurgitation	None Mild Moderate Severe
Initial procedure	None Balloon atrial septostomy Pulmonary artery band Pulmonary artery band with coarctation repair Aortopulmonary shunt Stage I with interrupted aortic arch repair Stage I
Superior cavopulmonary connection	None Bidirectional Glenn shunt Hemi-Fontan Bilateral cavopulmonary connection
Preoperative hemodynamics	Mean pulmonary arterial pressure Ventricular end-diastolic pressure
Procedure-related variables	
Type of Fontan	Lateral tunnel Extracardiac conduit
Creation of intentional right-to-left shunt	Yes/No
Type of intentional right-to-left shunt	None Adjustable atrial septal defect Multiple small holes Hepatic vein exclusion Side-to-side anastomosis with extracardiac conduit Single-punch fenestration in lateral-tunnel baffle
CPB time	
Aortic crossclamp time	
Deep circulatory arrest time	
MUF	Yes/No
Postoperative outcomes	
Hospital mortality (operative or <30 d)	
Duration of pleural drainage	
Duration of hospital stay	

TABLE 2. Diagnostic categories

Diagnoses	No. (%)
Single left ventricle with normally related great arteries	42 (13)
Single left ventricle with transposed great arteries	46 (14)
Heterotaxy syndrome	26 (8)
Single right ventricle (not HLHS)	27 (8)
HLHS	176 (53)
Other diagnoses	15 (4)

TABLE 3. Initial procedures

Procedure	No. (%)
None	28 (8)
Balloon atrial septostomy	3 (1)
Pulmonary artery banding	23 (7)
Pulmonary artery banding with coarctation repair	4 (1)
Aortopulmonary shunt	64 (19)
Stage I procedure	205 (62)
Stage I procedure with interrupted aortic arch repair	5 (2)

Statistical Analysis

Data are presented as medians and ranges where appropriate. Specific outcomes evaluated were hospital mortality (before hospital discharge or less than 30 days postoperatively), duration of postoperative pleural effusions, and duration of postoperative hospital stay. Patient and management characteristics that were evaluated as possible effectors of outcome are listed in Table 1. The effect of patient and surgical characteristics was evaluated by means of univariate and multivariable analysis. For the univariate analysis, the χ^2 test, the Fisher exact test, and logistic regression were used as appropriate. A *P* value of less than or equal to .1 was the criterion for inclusion in the multivariable model. Multivariable analysis was performed by means of a logistic regression model. Results are expressed as odds ratios (ORs) with 95% confidence limits (CLs). Patient characteristics were adjusted for year of operation in the multivariable analysis to control for possible changes in the patient population over time. Risk factors for prolonged effusion and hospital stay were evaluated in the hospital survivors.

The effect of year of operation and operating surgeon could not be separated from each other (Figure 1) or from certain surgical characteristics (eg, modified ultrafiltration [MUF]). In addition, it is expected that all surgeons' results improve over time, and therefore comparing the results of one surgeon in 1992 with those of another in 1999 would potentially introduce bias. For these reasons, the effect of year of operation and operating surgeon are presented descriptively and not incorporated into the multivariable analysis. There were no deaths among the 96 patients undergoing a lateral-tunnel Fontan operation in whom a single-punch fenestration was created; as a result, these patients would be eliminated from a multivariable logistic regression model where death is the outcome of interest. Therefore, a fictitious patient coded as having died after a lateral-tunnel Fontan procedure with a single-punch

Figure 1. Distribution of cases by surgeon and by year. **Figure 2. Type of Fontan procedure by year.**

fenestration was added to the model. This fictitious patient is not included in the descriptive data. The use of a single-punch fenestration in a lateral tunnel-type Fontan operation was highly correlated with the use of MUF. Their effects were therefore analyzed in separate models. The OR and significance of other characteristics in the model were not substantially different when combined with the single-punch fenestration versus with MUF.

Results

A total of 332 patients underwent a modified Fontan procedure at The Children's Hospital of Philadelphia between January 1, 1992, and December 31, 1999. Anatomic diagnoses are shown in Table 2. HLHS was present in 156 (53%) of 332 patients. A common AV valve was present in 34 patients. The majority of patients underwent at least one palliative procedure before cavopulmonary connection, with the stage I reconstruction performed most frequently (*n* = 205, 62%; Table 3). An interim superior cavopulmonary connection was performed in 318 (96%) patients. A hemi-Fontan procedure was performed in 257 patients, a bidirectional Glenn shunt in 36 patients, and bilateral cavopulmonary connections in 25 patients. At the time of the Fontan procedure, AV valve regurgitation was absent in 211 patients, mild in 100 patients, moderate in 17 patients, and severe in 2 patients. Preoperative echocardiograms were not available for 2 patients.

Figure 3. Type of intentional right-to-left shunt by year. LT, Lateral tunnel; EC, extracardiac conduit; ASD, atrial septal defect.

Figure 4. Fontan mortality by year of surgery. There was a significant decrease in mortality between 1994 and 1995, from 17% to 3% ($P = .026$). In addition, when the years 1992 through 1994 are compared with the later era of 1995 through 1999, there is a statistically significant decrease in overall mortality ($P < .001$).

Figure 5. Duration of effusions (in days) by year of operation. There was a dramatic decrease in the duration of effusions between 1994 and 1995. The median duration of pleural drainage for each year is shown at the bottom of the figure.

The median age at the time of the operation was 22 months (range, 11-380 months), and the median weight was 11 kg (range, 5.8-120 kg). A lateral tunnel-type Fontan connection was created in 281 patients, and extracardiac

TABLE 4. Right-to-left shunts

Type of intentional right-to-left shunt	No. (%)
None	34 (10)
Adjustable atrial septal defect	6 (2)
Multiple small holes	85 (26)
Partial hepatic vein exclusion	65 (20)
Side-to-side anastomosis between extracardiac conduit and atrial wall	40 (12)
Single-punch fenestration in lateral-tunnel baffle	96 (29)
Type of fenestration not determined	6 (2)

conduits were used in 51 patients (Figure 2). An intentional right-to-left shunt was created in 298 (90%) patients. The type of shunt varied according to the year of the operation. Early in the series, hepatic vein exclusion or multiple, small-baffle fenestrations were used, whereas in the most recent era, use of a single-punch fenestration in a lateral-tunnel baffle or a side-to-side anastomosis between the extracardiac conduit and atrial wall were used exclusively (Table 4 and Figure 3). Whenever possible, associated lesions, such as AV valve regurgitation and pulmonary artery stenosis, were addressed at the time of the superior cavopulmonary connection to avoid the need for additional procedures at the time of Fontan completion. Additional procedures were performed at the time of the Fontan procedure in 16 patients, including atrial septectomy in 8, tricuspid valve repair in 7, and arch augmentation in 1. The median duration of cardiopulmonary bypass (CPB) was 60 minutes (range, 32-236 minutes). A period of deep hypothermic circulatory arrest (DHCA) was used in 319 patients, with a median duration of 24 minutes (range, 12-93 minutes). Continuous CPB with bicaval cannulation was used in the remaining 13 patients (all extracardiac conduits). The median duration of aortic crossclamping was 26 minutes (range, 12-99 minutes). For patients undergoing a lateral-tunnel Fontan procedure with a single-punch fenestration, the median duration of CPB was 56 minutes (range, 40-147 minutes). The median duration of aortic crossclamping was 17 minutes (range, 12-60 minutes), with a median duration of DHCA of 17 minutes (range, 12-59 minutes). MUF was introduced in December 1995 and was used in all subsequent patients.

Overall, there were 22 deaths among the 332 patients in the study, for a mortality of 6.6%. The yearly mortality ranged from 0% to 17% (Figure 4) and decreased significantly after 1994 ($P < .001$). Since 1994, mortality after the Fontan procedure has been 1% (2/174). During the study period, Fontan failure during the early postoperative period, requiring takedown to a cavopulmonary connection or cardiac transplantation, occurred in 4 patients. Of these, 3 patients operated on in 1993 ($n = 2$) and 1994 ($n = 1$) died

TABLE 5. Variables with no demonstrated effect on mortality

Age
Weight
Anatomic diagnosis
AV valve insufficiency
Ventricular end-diastolic pressure
Heterotaxy syndrome
Initial procedure
Type of staging operation
Type of Fontan operation
Crossclamp time
CPB time
DHCA time

TABLE 6. Variables with no demonstrated effect on duration of pleural drainage

Age
Weight
Presence of common AV valve
AV valve insufficiency
Preoperative pulmonary arterial pressure
End-diastolic ventricular pressure
Initial procedure
Type of staging operation
Type of Fontan operation
Crossclamp time
CPB time
DHCA time

TABLE 7. Duration of pleural effusions by type of Fontan baffle fenestration

Type of baffle fenestration	No. of patients	Days of pleural drainage (median)	Days of pleural drainage (range)
None	33	7	1-258
Adjustable ASD	6	8	1-53
Hepatic vein exclusion	58	7	1-253
Multiple small holes	77	2	1-213
Single-punch hole in lateral tunnel	93	1	1-29
Single hole in extracardiac conduit	38	1	1-24

TABLE 8. Effect of type of intentional right-to-left shunt on duration of pleural drainage

Type of fenestration	OR (95% CL) for drainage >3 d	P value	OR (95% CL) for drainage >14 d	P value
Adjustable ASD	1.58 (0.25-9.82)	.49	0.71 (0.11-4.45)	.72
Multiple small holes	1.38 (0.61-3.09)	.44	1.00 (0.45-2.24)	1.00
Hepatic vein exclusion	0.72 (0.31-1.66)	.44	0.78 (0.33-1.83)	.57
Extracardiac conduit with side-by-side hole	0.053 (0.21-1.33)	.18	0.25 (0.08-0.76)	.01
Lateral tunnel with single-hole punch	0.18 (0.08-0.42)	<.001	0.13 (0.05-0.34)	<.001

ASD, Atrial septal defect.

after takedown of the Fontan procedure to a superior cavopulmonary connection, and 1 patient (1997) underwent orthotopic cardiac transplantation and survived. In the multivariable analysis characteristics that increased the risk of death included the presence of a common AV valve (versus all other AV valve types; OR, 7.64; 95% CL, 2.07-28.14; $P = .002$) and increased mean pulmonary artery pressure at the preoperative cardiac catheterization (OR, 1.46/1 mm Hg increase; 95% CL, 1.2-1.78; $P < .001$; Table 5). Characteristics that decreased the risk of death in the multivariable analysis were use of a single-punch fenestration in the lateral-tunnel Fontan operation (OR, 0.06; 95% CL, 0.01-0.65; $P = .02$) and the use of MUF (OR, 0.14; 95% CL, 0.03-0.72; $P = .019$).

The median duration of postoperative pleural drainage among survivors was 2 days (range, 1-258 days), and this

decreased significantly after 1994 ($P < .001$, Figure 5). A diagnosis of HLHS, however, was associated with an increased risk of prolonged effusion (OR, 1.73; 95% CL, 1.07-2.81; $P = .03$; Table 6). The duration of pleural drainage was related to the type of fenestration (Tables 7 and 8). Use of single-punch fenestration in a lateral-tunnel Fontan operation in patients with HLHS reduced the OR for prolonged effusions to 0.47. Use of MUF in patients with HLHS reduced the OR for prolonged pleural drainage to 0.91. Creation of an extracardiac conduit Fontan with a side-by-side anastomosis and a lateral-tunnel Fontan with a single-punch fenestration were both found to significantly reduce the incidence of very prolonged pleural effusions (>14 days, Table 7). Other types of intentional right-to-left shunts (hepatic vein exclusion and multiple small-baffle fenestrations) did not reduce the incidence of prolonged effusions.

Figure 6. Hospital stay (in days) by year of operation. There was a significant decrease in the duration of hospital stay between 1994 and 1995 ($P = .04$). In addition, when the early era of 1992 through 1994 is compared with the later era of 1995 through 1999, there is a significant decrease in the duration of hospital stay ($P < .001$). The median duration of hospital stay for each year is shown at the bottom of the graph.

The median length of hospital stay among survivors was 9 days (range, 3-142 days). As with mortality and prolonged effusions, the incidence of prolonged hospitalization decreased significantly after 1994 ($P < .001$, Figure 6). Patients with HLHS had a significantly increased risk of prolonged hospitalization (OR, 2.24; 95% CL, 1.23-2.07; $P = .008$). The risk of prolonged hospitalization was decreased by use of a single-punch fenestration in a lateral-tunnel Fontan operation (OR, 0.09; 95% CL, 0.03-0.4; $P < .001$), as well as by use of MUF (OR, 0.07; 95% CL, 0.03-0.16; $P < .002$).

Discussion

Management of neonates with a functional single ventricle has evolved considerably in the 3 decades since the introduction of the Fontan operation. Most of these infants undergo a series of staged reconstructive operations beginning in the neonatal period and culminating in an eventual Fontan procedure. The goals of neonatal management include creation of unobstructed systemic outflow and pulmonary venous pathways, limitation of pulmonary blood flow to prevent development of pulmonary vascular disease, and avoidance of pulmonary artery distortion. A variety of technical modifications have been introduced since the first report of the Fontan operation,¹⁻³ including staging with an interim superior cavopulmonary connection,^{4,5} use of an intra-atrial baffle to create a lateral-tunnel Fontan,⁶ creation of an intentional right-to-left shunt (baffle fenestration),⁷⁻⁹ and introduction of the extracardiac conduit Fontan.¹⁰ Because of improved management in the neonatal period and early infancy, an increasing number of patients previously thought to be at high risk, specifically those with HLHS, are being referred for the Fontan operation.¹¹

Despite the change in patient population, there has been a dramatic reduction in mortality and morbidity after the Fontan operation. Multiple factors, some difficult to measure, probably have contributed to the decline in mortality and morbidity, including modifications of the procedure and improved pre-Fontan management of infants with single ventricle, as well as improvements in CPB, anesthesia management, and intensive care unit care.

Our study reports a single-institution experience with a large number of patients undergoing the modified Fontan procedure in the current era. Despite a patient population composed largely of patients with HLHS or variants, mortality has decreased dramatically in recent years, as has morbidity, especially prolonged pleural effusions. In contrast to previous reports, HLHS was not a risk factor for mortality. However, a diagnosis of HLHS does remain a risk factor for morbidity, specifically for prolonged pleural effusions and prolonged hospitalization. Interestingly, there was no difference in mortality or morbidity between patients undergoing a lateral-tunnel Fontan or an extracardiac conduit Fontan. However, creation of a single-punch fenestration in a lateral-tunnel Fontan operation was associated both with the lowest mortality and the lowest morbidity, suggesting that a single-punch fenestration provides a more reliable and consistent method of baffle fenestration than other techniques. Modifications of CPB, specifically the introduction of MUF, were also associated with a decrease in mortality and morbidity in this study.

Gentles and colleagues,¹² from Boston Children's Hospital, reviewed a large (500 patients) single-center experience of outcomes after the Fontan operation between 1973 and 1991. This early time period encompassed many of the technical modifications and changes in management strategies since the introduction of the Fontan operation. In their study a higher preoperative mean pulmonary arterial pressure, younger age at operation, presence of heterotaxy syndrome, and presence of a tricuspid valve as the systemic AV valve were identified as risk factors for early failure. Use of an atriopulmonary connection-type Fontan procedure and absence of baffle fenestration were also associated with an increased risk of early failure. HLHS was present in fewer than 10% of the patients but was noted to be a risk factor for early death. In addition, age at the time of the Fontan operation of less than 4 years remained a risk factor for failure. It should be noted that the last patient in their series was operated on in 1991, and thus their report represents the evolution of the Fontan procedure rather than results in the current era.

Hsu and associates¹³ reported outcomes after a single-stage, nonfenestrated Fontan procedure in 61 consecutive patients between 1990 and 1996. The mean age at the time of the operation was 3.3 years, and early mortality was 3.9%. The median duration of pleural drainage was 5.5

days. However, these 61 patients represented only two thirds of the patients undergoing the Fontan procedure at their institution. Use of the single-stage, nonfenestrated Fontan procedure was restricted to patients thought to be at low risk, and there were only 2 patients with HLHS among the 61 patients studied. Their experience demonstrated that a single-stage, nonfenestrated Fontan procedure can be performed with reasonably low morbidity and mortality in selected patients.

Thompson and colleagues¹⁴ reported results in 81 patients undergoing an extracardiac Fontan operation between 1992 and 1997 after a previous bidirectional Glenn shunt. They used a technique of selective fenestration after separation from CPB, for a pressure in the Fontan circuit of greater than 18 mm Hg or in the transpulmonary gradient of greater than 10 mm Hg. There were 2 operative deaths, and prolonged chest tube drainage (>14 days) occurred in 13 patients, 8 with and 5 without fenestration. HLHS was present in fewer than 10% of the patients. Their experience demonstrates that an extracardiac conduit Fontan operation without fenestration is associated with acceptable morbidity and mortality in selected low-risk patients.

Van Arsdel and coworkers¹⁵ evaluated risk factors for mortality and morbidity in 100 patients undergoing the Fontan procedure between 1992 and 1995. HLHS was present in fewer than 10% of the patients. Mortality in the first half of the series was 16%, and it was 0% in the second 50 patients. Duration of pleural effusions and hospital stay were also shorter in the more recent era. Patient characteristics and risk factors were comparable throughout the study; however, patients in the recent era of lower mortality were more likely to have routine staging, an extracardiac Fontan completion, and use of MUF. The authors attribute the improvement in outcome to increasing use of the extracardiac Fontan procedure, as well as introduction of MUF after CPB.

Mosca and associates¹⁶ recently reported the outcome of the Fontan procedure in 100 consecutive patients with HLHS between 1992 and 1998. Two different surgical techniques were used. In the earlier era (1992-December 1995), patients underwent a bidirectional Glenn procedure and then a lateral-tunnel completion Fontan procedure with the right atrium reconnected to the pulmonary artery at the time of the Fontan operation (technique 1). In December of 1995, a new technique was introduced, whereby patients underwent a hemi-Fontan procedure as the staging procedure. The Fontan procedure (lateral tunnel) was performed with single right atrial venous cannulation, profound hypothermia, and a brief period of DHCA (technique 2). Baffle fenestration with a 4-mm single-punch fenestration was routinely used. MUF was introduced in 1996 and thus was used only in patients undergoing technique 2. Hospital survival for patients treated with technique 1 was 79% com-

pared with 98% for those treated with technique 2. The findings of this study are consistent with those of the current study in that use of a single-punch fenestration in the lateral-tunnel conduit, as well as routine use of staging and MUF, resulted in low mortality for the Fontan procedure in patients with HLHS.

We previously evaluated the effects of MUF in a small group of patients undergoing the Fontan operation.¹⁷ The study was a retrospective review, yet it showed that use of MUF was associated with a decrease in the duration of pleural effusions and length of hospital stay. The patients in that study are included in the current study. In the current study the use of MUF was also shown to decrease mortality and to reduce the incidence of prolonged pleural effusions and prolonged hospitalization. Studies at other institutions have also demonstrated beneficial effects of MUF after the Fontan operation.^{15,16} The mechanisms by which MUF results in beneficial effects have not been fully determined but likely include removal of excess body water and inflammatory mediators. MUF has been shown to improve ventricular function, lower pulmonary vascular resistance, improve pulmonary compliance, and decrease postoperative bleeding, all of which may contribute to improved outcome after the Fontan operation.^{18,19}

Several centers have reported improved hemodynamic results and improved patient outcomes after the modified Fontan operation when an intentional right-to-left shunt (baffle fenestration or adjustable atrial septal defect) is used.²⁰⁻²² Benefits of baffle fenestration include improved ventricular preload, increased cardiac output, improved oxygen delivery, reduced systemic venous pressure, and decreased severity of pleural effusion. The findings of the current study suggest that the specific type of fenestration is an important determinant of outcome.

The current study demonstrates that a management strategy, including routine staging, effective baffle fenestration, and use of MUF, is associated with very low mortality and morbidity after the Fontan procedure. Because almost all patients underwent an interim superior cavopulmonary connection, the effect of staging cannot be evaluated. A variety of techniques were used for baffle fenestration, and only creation of a single-punch fenestration in a lateral-tunnel baffle was associated with a decrease in both mortality and morbidity. Neither hepatic vein exclusion nor the creation of multiple small holes in the baffle resulted in a decrease in the incidence of prolonged effusions. Other studies have suggested that routine baffle fenestration is not necessary; however, these studies included few patients with HLHS, and the reported duration of pleural drainage has consistently been longer than in the current study. The type of fenestration is important because techniques other than single-punch fenestration in a lateral-tunnel baffle appear to be less effective and less consistent.

This study has several limitations. The study is retrospective. Many of the management strategies evolved over time and are highly related to both the surgeon and the year of the operation. Decisions, such as time of the Fontan operation, type of fenestration, and use of MUF, were made at the surgeon's discretion and not according to protocol. In addition, certain practices are highly linked, such as the side-to-side anastomosis type of fenestration and the extracardiac conduit, and thus the effects cannot be separated. Because of the nature of the study, it is not possible to determine whether similar results could have been achieved with alternate management strategies, such as a single-stage, nonfenestrated Fontan procedure without MUF. However, there are no reports of equally successful applications of such a strategy to a large group of patients with HLHS. This study demonstrates that a strategy of interim superior cavopulmonary connection, effective baffle fenestration, and use of MUF results in excellent outcome after the Fontan operation, with very low morbidity and mortality, even in high-risk patients, such as those with HLHS.

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Discussion

Dr Ralph Mosca (New York, NY). I congratulate the authors on a well-written review of a large series of patients undergoing the Fontan procedure at the Children's Hospital of Philadelphia. The overall results were excellent, a tribute to the expertise of all involved in these patients' care. Those who care for large numbers of these patients will attest to the fact that 15 years ago the appearance of a Fontan procedure on the operating room schedule was met with some trepidation. This often portended a difficult postoperative time for patients and caregivers alike. The procedures were reserved for a highly selected subgroup of patients with single ventricle, and those with HLHS were few and far between. Today, HLHS has become the leading diagnosis in many centers, the procedures are routine, and the patients are fast tracked, extubated early, and often discharged in a week.

I have a few questions. A common AV valve was a risk factor for early death, yet AV valve insufficiency was not. What is it about the presence of a common AV valve, aside from its competency, that increases the risk?

In the article, you described 19 patients with either moderate or severe AV valve insufficiency, yet only 7 underwent attempted AV valve repair. Despite its lack of predictive value concerning early morbidity and mortality, do you believe that AV valve competency is important in the long-term, and what is your policy regarding selection of patients for valve repair?

Elevated preoperative pulmonary artery pressure was found to be a significant risk factor for early death, with an OR of 1.46 for each millimeter increase in pressure, yet there are no references to absolute values. At what point did this become clinically significant?

The punch-style fenestration decreased the risk of early death in the lateral-tunnel group. Interestingly, there was no difference in mortality when compared with that in the extracardiac Fontan group. Were all these fenestrations as reliably patent? Did selection bias favor your extracardiac group? Finally, should we, as some groups are saying, do extracardiac Fontans if this is the result?

The median duration of pleural drainage was 2 days, the shortest I have seen in this patient population. Many institutions keep the tubes in place until the patients are reliably taking enteral nutrition to check for increases in chylous drainage. What is your opinion on these policies, and have you noticed any increase in late postoperative effusions with early removal of the pleural tubes?

Finally, it strikes me that when viewed from the neonatal period, survival after a Fontan procedure for single-ventricle lesions can be likened to the challenge of a game of pool. Now that we understand the important issues and angles involved, the morphologic surface on which we are playing seems less important. What is paramount is that each shot or stage is well made and, just as important, sets up the next. You have certainly shown us that this is true for HLHS.

Dr Spray. Thank you for your comments. The issue of the common AV valve is a significant one. In the small number of patients with a common AV valve, we could not identify the presence of AV valve regurgitation as a risk factor. The common AV valve may therefore be a surrogate marker for some other factor. In single-ventricle malformations, especially those with right ventricular morphology, AV valve regurgitation is not uncommon when these patients come to the Fontan operation. We have not actually found that repair of the AV valve at that stage, unless the regurgitation is severe, has changed the overall survival, hospital stay, or morbidity in these patients. However, if significant AV valve regurgitation is present at the time of the Fontan operation, at least an attempt to limit the amount of regurgitation is warranted. Unfortunately, common AV valve may be the most difficult anomaly to repair surgically, and these patients may ultimately come to valve replacement.

There was no absolute value of pulmonary artery pressure that was associated with mortality. This variable was assessed continuously. I would say, however, that patients who have pulmonary artery pressures after the hemi-Fontan in the 16- to 18-mm Hg

Discussion Figure 1. Relationship between pulsatile pressure at the pre-Fontan catheterization and probability of operative mortality.

range are relatively poor candidates but not contraindicated for completion operations, especially if a fenestration is done. As noted in Discussion Figure 1, pre-Fontan pulmonary artery pressures of greater than 12 to 14 begin to be associated with increased risk. Fenestration has been shown to take high-risk patients and make them into low-risk patients, which I think our data tend to support.

We could not identify any difference in mortality, hospital stay, or hemodynamics in patients with extracardiac versus lateral-tunnel Fontan operations. There were no deaths at all in patients with single-punch fenestration lateral-tunnel operations. In the patients with extracardiac Fontan operations, the only thing that was slightly different was that their oxygen saturation levels were slightly higher postoperatively, and their duration of effusion was slightly higher than that in the lateral-tunnel group, probably reflecting the fact that the fenestration was not as reliably performed with a side-by-side fenestration in an extracardiac conduit.

We now generally remove the chest tubes on the first postoperative day if there is no drainage, and we have not found a significant increase in chylous effusions later with that policy. This also limits, clearly, the hospital stay in these patients.