EARLY RESULTS OF THE FONTAN PROCEDURE IN ONE HUNDRED CONSECUTIVE PATIENTS WITH HYPOPLASTIC LEFT HEART SYNDROME

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Objectives: The purpose of this study was to review a large, single institutional experience with the Fontan procedure for patients with hypoplastic left heart syndrome.

Methods: One hundred consecutive patients with "classic" hypoplastic left heart syndrome underwent Fontan palliation between February 1992 and April 1998. Patient demographic, morphologic, and procedural variables were examined and analyzed. In particular, two different surgical techniques were used: technique I (February 1992 to December 1995) employed cardiopulmonary bypass and moderate systemic hypothermia, and technique II (December 1995 to April 1998), profound hypothermia and circulatory arrest. A retrospective review of medical records was performed and variables were examined and analyzed.

Results: Hospital survival for the entire cohort was 89% (95% CI 83%-95%). The technique of operation, cardiopulmonary bypass time, and aortic crossclamp time were each strongly associated with survival. Survival for patients treated by technique I was 79% (95% CI 68-91%; n = 48) and for those treated by technique II, 98% (95% CI 94%-100%; n = 52). Cardiopulmonary bypass and crossclamp times were also highly correlated with time to extubation and length of intensive care unit stay. Preoperative pulmonary artery pressure was correlated with survival; preoperative oxygen saturation, right atrial pressure, pulmonary vascular resistance, pulmonary artery size, extent of aortopulmonary artery collaterals, and echocardiographic estimates of ventricular function and tricuspid regurgitation were not correlated with survival.

Conclusions: Our recent experience with Fontan palliation for patients with hypoplastic left heart syndrome suggests that it is attended by low perioperative mortality. The precise operative technique used appears to be an important determinant of outcome, with the duration of cardiopulmonary bypass and crossclamping being particularly significant. (J Thorac Cardiovasc Surg 2000;119:1110-8)

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The Fontan procedure and its modifications have been applied to nearly all forms of single ventricle cardiac lesions. A more thorough understanding of post-Fontan physiology, modifications in surgical technique, and advances in perioperative care have led to improved outcomes. The average early survival after the Fontan procedure has improved from 75% to greater than 90% over the past 20 years.[1-5] Despite improved overall survival, many authors still consider specific cardiac lesions to carry an elevated risk of early postoperative mortality. Usually included in this higher risk category are patients with hypoplastic left heart syndrome (HLHS).[6-9]

Certain preoperative and intraoperative variables, aside from the basic cardiac morphology, have been identified as important risk factors for early postoperative mortality after Fontan palliation. These include elevated pulmonary artery pressure and resistance, ventricular dysfunction, systemic atrioventricular valve regurgitation, longer cardiopulmonary bypass (CPB) time, and significant pulmonary artery distortion.[6,7] In addition, there is a sense that systemic–pulmonary artery collateral blood vessels, by providing pulmonary arterial flow that competes with systemic venous return, may also increase morbidity and mortality.[10,11] The purpose of this study was to review our experience with the Fontan procedure performed for patients with HLHS in an effort to ascertain whether these and other perioperative variables influenced patient outcome (hospital survival, duration of intubation, length of time in the intensive care unit, and length of total hospital stay). Although these techniques are used in all of our patients with various forms of univentricular heart, the cohort of patients with HLHS was chosen since it represents the largest and most homogeneous group at our institution. A significant change in surgical technique for the Fontan operation occurred approximately halfway through this series, and patient outcomes were therefore also related to the type of surgical management used.

Methods

Patients. This is a retrospective study of 100 consecutive patients with HLHS who underwent a Fontan procedure between February 1992 and April 1998. Only patients with "classic" HLHS (aortic valve atresia or hypoplasia and a right ventricle–dependent circulation) were included.[12] Patients with other single right ventricle lesions, such as an unbalanced atrioventricular septal defect and left ventricular hypoplasia or double-outlet right ventricle with mitral atresia, were excluded. All patients had previously undergone neonatal Norwood palliation followed by stage 2 palliation consisting of a bidirectional Glenn (BDG) or hemi-Fontan procedure (HFP).

Operative technique. The final anatomy of the Fontan palliation was essentially the same in all the patients in this report: a total cavopulmonary connection with a polytetrafluoroethylene* intra-atrial lateral tunnel, with a 4-mm fenestration within this baffle. However, two significantly different surgical techniques were used to effect this palliation:

Technique I. Forty-eight patients operated on between February 1992 and December 1995 underwent the Fontan procedure with CPB, bicaval venous cannulation, and moderate systemic hypothermia (28°C). All patients who had undergone a BDG procedure[13,14] for stage 2 palliation (n = 36) were included in this group, as well as 12 patients whose stage 2 palliation was an HFP.[15] The right pulmonary artery and the cephalad aspect of the right atrium were mobilized. Incisions in the inferior aspect of the right pulmonary artery and in the superior portion of the right atrium were fashioned. The caudal aspect of the right pulmonary artery and right atrium were connected, creating the floor of the new atriopulmonary connection. A lateral tunnel (polytetrafluoroethylene) was then constructed incorporating the orifice of the inferior vena cava, as well as the new atriopulmonary connection. A 4-mm fenestration (punch or adjustable snare) was placed within the baffle.

Technique II. Fifty-two patients operated on between December 1995 and April 1998 underwent the Fontan procedure with single right atrial venous cannulation, profound hypothermia, and deep hypothermic circulatory arrest as previously described.[16] Minimal dissection of the cardiac structures was performed, and the pulmonary arteries and venae cavae were not exposed. All patients in this group had previously undergone an HFP as their stage 2 palliation. A right atriotomy anterior to the crista terminalis was fashioned, the atrial septal defect was enlarged as needed, and the patch separating the atria from the pulmonary arteries was excised. The branch pulmonary arteries were sounded and calibrated with dilators to confirm their size. A lateral tunnel incorporating the orifices of both venae cavae was fashioned with polytetrafluoroethylene. A 4-mm diameter punch fenestration was placed in the baffle. The anterior suture line was sewn to the edge of the atriotomy, avoiding the crista terminalis, completing the baffle, and closing the atria. The remainder of the operative procedure was identical to technique I.

Concurrent procedures included tricuspid valvuloplasty (n = 14) and placement of a pulmonary artery stent (n = 13). Tricuspid valvuloplasty was performed with standard techniques of suture commissuroplasty and/or annuloplasty.[17] Artificial annuloplasty rings were not used. A Palmaz stent (generally sizes P-188 or P-308; Johnson & Johnson Interventional Systems, Warren, NJ) was inserted into the left pulmonary artery when diffusely hypoplastic or when exposure of the branch pulmonary artery would have required extensive dissection.

Modified ultrafiltration (MUF) was introduced in April of 1996 and thus was used only in patients undergoing technique II (all but 2 patients undergoing technique II were treated with MUF at the Fontan procedure). This technique has been described elsewhere[18,19] and was routinely performed for 10 minutes after weaning from CPB.

Data acquisition. Demographic and other patient-related data (eg, the occurrence of postoperative complications) were obtained from hospital records. For patients operated on with technique II, the CPB time does not include the period of circulatory arrest. Survival is defined as survival to hospital discharge.[1] Lengths of stay in the intensive care unit and in the hospital were chosen as end points that encompass other potentially complicating factors, that is, the use of inotropic agents and persistent pleural effusions.

*W. L. Gore & Associates, Inc, Flagstaff, Ariz.

Table I. *Patient demographic data, preoperative characteristics, intraoperative variables, and outcomes shown for the entire group and a comparison between subgroups (technique I and technique II)*

	Total group (n = 100)		*Technique I (n = 48)*		*Technique II (n = 52)*		
Preop and intraop parametric data	*Mean ± 95% CI (range, n)*		*Mean ± 95% CI (range, n)*		*Mean ± 95% CIP value (range, n)*		P *value (technique I vs II)*
Age (mo)†	23.5 ± 1.2	(14-50, 100)	24.8 ± 2.1	(14-50, 48)	22.8 ± 1.2	(16-36, 52)	.05
Weight (kg)†	10.7 ± 0.33	(7-17, 100)	11.0 ± 0.58	(7-17, 48)	10.4 ± 0.33	(8.5-13, 52)	.09
SAS (%)†	83 ± 1.0	(68-89, 96)	83 ± 1.0	(68-89, 46)	83 ± 1.0	(76-89, 50)	>.2
RAP (mm Hg)†	6.4 ± 0.52	(1-16, 90)	6.6 ± 0.79	(1-14, 45)	6.1 ± 0.8	(2-16, 43)	>.2
PAP (mm Hg)†	11.5 ± 0.53	(7-18, 94)	11.7 ± 0.72	(7-17, 46)	11.9 ± 0.7	(7-18, 48)	>.2
PVR (WU × m^2)†	2.1 ± 0.23	(0.4-6.2, 80)	1.9 ± 0.35	(0.4-6.2, 41)	1.9 ± 0.4	(0.66-4.5, 39)	.13
McGoon ratio†	1.49 ± 0.07	(0.86-2.11, 76)	1.42 ± 0.10	(0.86-2.11, 33)	1.53 ± 0.09	(0.93-2.1, 43)	.11
CPB time (min)	84.8 ± 7.1	(43-196, 100)	115.4 ± 8.2	(67-196, 48)	56.6 ± 2.6	(43-90, 52)	<.0001
Crossclamp time (min)	48.1 ± 3.4	(24-105, 100)	58.9 ± 5.2	(30-105, 48)	38.2 ± 2.2	(24-64, 52)	<.0001
Circulatory arrest time (min)	NA		NA		31.1 ± 2.3	(14-57, 51)	NA
Preop nonparametric data	*n (%)*		*n (%)*		*n (%)*		
Systolic function†	n = 98		n = 47		n = 51		>.2*
Normal ventricular function	77 (79%)		37 (79%)		40 (78%)		
Mild to moderate ventricular dysfunction	17 (17%)		8 (17%)		9 (18%)		
Severe ventricular dysfunction	4 (4%)		2 (4%)		2 (4%)		
Tricuspid regurgitation†	n = 98		n = 47		n = 51		>.2*
No regurgitation	60 (61%)		30 (64%)		30 (59%)		
Mild regurgitation	27 (28%)		12 (26%)		15 (29%)		
Moderate regurgitation	6 (6%)		4 (9%)		2 (4%)		
Severe regurgitation	5 (5%)		1 (2.1%)		4 (8%)		
Collateral blood flow† (qualitative scale)	n = 76		n = 34		n = 42		.0004*
0	10 (13%)		7 (21%)		3 (7%)		
1	20 (26%)		15 (44%)		5 (12%)		
2	25 (33%)		7 (21%)		18 (43%)		
3	12 (16%)		2 (6%)		10 (24%)		
4	6 (8%)		3 (9%)		3 (7%)		
5	2 (3%)		0 (0%)		2 (5%)		
6	1 (1%)		0 (0%)		1 (2%)		
Outcome data							
Survival	89 ± 6.1*** (100)		79 ± 11.5*** (48)		98 ± 3.8*** (52)		.003*
Intubation	2** (1-31, 97)		3** (1-31, 47)		1** (0-6, 50)		<.0001*
ICU stay	3** (1-144, 98)		5.5** (2-144, 48)		2** (1-8, 50)		<.0001*
Hospital stay	13.5** (2-146, 96)		16** (2-146, 48)		10.5** (6-40, 48)		<.0021*

CI, Confidence interval; NA, not applicable; *SAS*, systemic arterial saturation; *RAP*, right atrial pressure; *PAP*, pulmonary artery pressure; *PVR*, pulmonary vascular resistance; *WU*, Wood units; *CPB*, cardiopulmonary bypass; *ICU*, intensive care unit.
*Statistical testing with Wilcoxon rank sum test. ** Median value (range, n). *** Percentage ± 95% CI (n).
†Preoperative values.

Echocardiographic data. A single examiner who was blinded to the remainder of the patient data reviewed preoperative 2-dimensional and Doppler echocardiograms, which had been obtained within 10 days of the Fontan procedure. Right ventricular systolic function was qualitatively estimated to be normal, mild to moderately depressed, or severely depressed. The magnitude of tricuspid regurgitation was qualitatively estimated to be none or trace, mild to moderate, or severe.

Cardiac catheterization hemodynamic data. The following data were extracted from the pre-Fontan catheterization record: mean right atrial (RAP) and pulmonary arterial (PAP) pressures, pulmonary blood flow (PBF, calculated by the Fick principle with the use of either measured or assumed oxygen consumption), and systemic arterial oxygen saturation. Pulmonary vascular resistance (PVR) was calculated by the following formula: PVR (indexed) = (Mean PAP – Mean RAP)/PBF (indexed).

Angiographic data. A single observer inspected cineangiograms of the pulmonary artery and ascending aorta. Branch pulmonary artery diameters just proximal to the origins of the right and left upper lobe branches and the diameter of the descending thoracic aorta at the diaphragm were measured and used to calculate the McGoon ratio.[20,21] The magnitude of blood flow into the lungs from aortopulmonary collaterals was judged by a semiquantitative estimate of the amount of contrast material present in the pulmonary arteries

Table II. *Patient preoperative and intraoperative data as shown in survivors and nonsurvivors*

	Survivors	*Nonsurvivors*	
Parametric data	*Mean ± CI (range, n)*	*Mean ± CI (range, n)*	P *value*
Age (mo)†	23.8 ± 1.3 (16-50, 89)	21.3 ± 2.5 (14-26, 11)	>.2
Weight (kg)†	10.76 ± 0.30 (7-17, 89)	9.86 ± 0.85 (8.4-12, 11)	.09
SAS (%)†	83 ± 1.0 (74-89, 85)	83 ± 4.0 (68-89, 11)	>.2
RAP (mm Hg)†	6.2 ± 0.60 (1-16, 77)	7.6 ± 1.6 (2-10, 10)	.12
PAP (mm Hg)†	11.3 ± 0.54 (7-18, 83)	13.3 ± 1.24 (10-17, 11)	.02
PVR (WU × m^2)†	2.1 ± 0.25 (0.4-6.2, 71)	2.2 ± 0.72 (0.72-3.5, 9)	>.2
CPB time (min)	79.8 ± 7.0 (43-172, 89)	124.9 ± 19.5 (82-197, 11)	<.0001
Crossclamp time (min)	46.5 ± 3.5 (24-105, 89)	61.2 ± 10.83 (35-87, 11)	.008
McGoon ratio†	1.49 ± 0.07 (0.86-2.11, 69)	1.46 ± 0.20 (1.25-1.92, 7)	>.2
Nonparametric data			
Systolic function†	n = 87	n = 11	>.2*
Normal ventricular function	67 (77%)	10 (91%)	
Mild-moderate ventricular function	16 (18%)	1 (9%)	
Severe ventricular dysfunction	4 (5%)	0 (0%)	
Tricuspid regurgitation†	n = 87	n = 11	>.2*
No regurgitation	54 (62%)	6 (54%)	
Mild regurgitation	24 (28%)	3 (27%)	
Moderate regurgitation	5 (6%)	1 (9%)	
Severe regurgitation	4 (5%)	1 (9%)	
Collateral blood flow†	n = 67	n = 9	>.2*
0	9 (13%)	1 (11%)	
1	16 (24%)	4 (45%)	
2	23 (34%)	2 (22%)	
3	11 (16%)	1 (11%)	
4	5 (7.5%)	1 (11%)	
5	2 (3%)	0 (0%)	
6	1 (1.5%)	0 (0%)	

Survival was significantly associated with shorter cardiopulmonary bypass and aortic crossclamp times. In addition, pulmonary artery pressure was also significantly associated with survival, however the differences in pressures are likely clinically unimportant. *CI*, Confidence interval; *SAS*, systemic arterial saturation; *RAP*, right atrial pressure; *PAP*, pulmonary artery pressure; *PVR*, pulmonary vascular resistance; *WU*, Wood units; *CPB*, cardiopulmonary bypass.

*Statistical testing with Wilcoxon rank sum test.

†Preoperative values.

and veins after injection of contrast medium into the ascending aorta. An ordinal scale was developed that is roughly proportional to the magnitude of collateral flow. For each lung, 0 = no contrast material seen in the pulmonary vessels, 1 = barely detectable contrast material in the pulmonary vessels, and 3 = relatively dense contrast material easily seen in the large branch pulmonary arteries and veins. A score of 2 was assigned when the amount of contrast material entering the pulmonary vessels was intermediate in magnitude between 1 and 3. Each lung was scored and the scores were added together (the maximum score was therefore 6).

Statistical analysis. Four outcome measures were evaluated in the postoperative period: survival, number of postoperative days intubated, length of stay in the intensive care unit, and length of hospital stay. For survival, bivariate analyses of possible predictors were performed with χ^2 for all categoric variables (tricuspid regurgitation, right ventricular function, and aortopulmonary collateral blood flow) and the Student *t* test for all continuous variables. The number of days intubated postoperatively, the length of stay in the intensive care unit, and the hospital length of stay were not normally distributed. Therefore, bivariate comparisons for each of these variables were performed by means of the Wilcoxon rank sum test. Additionally, these outcomes were each categorized into binomial variables, either less than or equal to the 75th percentile or greater than the 75th percentile. These binomial variables were assessed by means of χ^2 for all categoric predictor variables and the Student *t* test for all continuous predictor variables. Nonparametric, ordinal data were tested as a bivariate model; 75% were chosen to be tested for each of these variables. Multivariable models with the use of logistic analyses were then composed for each outcome to investigate independent outcome predictors. All data were stored in Microsoft Excel and then analyzed with the SAS statistical package (SAS Institute, Inc, Cary, NC).

Results

Outcomes. Table I summarizes patient demographics, preoperative characteristics, intraoperative variables, and outcomes for all patients considered together and when segregated according to the technique used for the Fontan procedure. Patients operated on by means of technique II had better outcomes (better sur-

vival, a shorter length of time intubated, and a shorter length of stay in the intensive care unit and the hospital) than those operated on with technique I. Although the preoperative characteristics of the patients in the 2 groups were very similar, there was a statistically significant difference in ages between the 2 groups of patients, and the aortopulmonary collateral blood flow "score" was significantly higher for patients undergoing technique II (Table I). CPB and aortic crossclamp times were both significantly shorter for the patients operated on with technique II, suggesting that these variables might account for the difference in outcomes between the 2 groups.

Predictors of survival. Survival was also associated with lower preoperative pulmonary artery pressure and shorter CPB and aortic crossclamp times (Table II). Patients treated by technique I had an increased risk of death by logistic modeling (odds ratio 13.3; 95% CI 1.6-108.6). The surgical method used for stage 2 palliation was also a predictor of survival; 78% for BDG patients (95% CI 65%-91%) and 95% for HFP patients (95% CI 90%-100%; $P = .017$), with an increased risk of mortality associated with the use of a BDG procedure as stage 2 palliation (odds ratio 3.6; 95% CI 1-13.4). However, because all patients with a BDG shunt had been operated on with technique I, it is difficult to ascertain whether the reduced mortality in technique II is related to the type of stage 2 palliation or to the Fontan operative technique (or both) due to the high correlation of these 2 variables. Subgroup analysis reveals that although most patients undergoing technique I had previously undergone a BDG shunt (n = 36), 12 patients in this group had undergone an HFP as stage 2 palliation. There was no significant difference in mortality between patients treated by technique I who had undergone a BDG and those who had an HFP (22% vs 17% mortality, respectively; $P = .682$). In contrast, patients treated by technique I who underwent an HFP tended to have a higher mortality than patients treated by technique II who underwent an HFP (17% vs 2% mortality; $P = .09$), Therefore, the type of technique used at the Fontan operation appears to be a more important predictor of mortality than the type of stage 2 palliation used.

Longer CPB time, longer aortic crossclamp time, and surgical technique I were significantly associated with longer intubation time and longer stay in the intensive care unit (Tables III and IV). The total length of hospital stay was significantly longer in patients undergoing technique I and in those with longer CPB times (Table I and Table V).

Discussion

The Fontan procedure originally described for the treatment of tricuspid atresia is now being applied for the palliation of virtually all forms of univentricular lesions. A few factors appear to influence survival, including the diagnosis of HLHS, elevated pulmonary artery pressure and/or resistance, pulmonary arterial distortion, impaired ventricular function, and prolonged CPB times.[1-9] In addition, aortopulmonary collateral blood flow, by competing with venous return, also seems to increase the chance of an unfavorable outcome.[10,11] Relatively little data pertaining to the results of the Fontan procedure in patients with HLHS are available regarding these possible risk factors. This analysis was undertaken to determine whether these and other variables affected short-term morbidity and mortality in a relatively homogeneous patient population.

The care of patients with univentricular heart at our institution has evolved over the past decade. Initially, patients underwent neonatal palliation followed by a Fontan procedure. As we realized the advantages of the BDG procedure in increasing the effective pulmonary blood flow and reducing the volume load on the single ventricle, this became a routine part of our staged palliation. The Fontan procedure then required construction of an atriopulmonary connection and often augmentation of narrowed branch pulmonary arteries. These operations were performed with bicaval venous cannulation and moderate hypothermia, which required significant CPB and aortic crossclamp times and occasionally were complicated by phrenic nerve injury, postoperative bleeding, and reduced pulmonary compliance. These factors appeared to negatively affect the outcome of our patients with single ventricle lesions. The HFP was then used as a routine method to augment the branch pulmonary arteries and provide a large potential communication within the right atrial chamber for the eventual lateral tunnel Fontan procedure. When performed with a single atrial cannula and profound hypothermia, use of an HFP as an intermediate-stage procedure greatly facilitated the Fontan procedure by reducing the time required for dissection of mediastinal structures, markedly reduced phrenic nerve injury as a complication, and made reconstructive work on the branch pulmonary arteries rare. For this reason the latter technique has been adopted as our preferred method of palliation of patients with single ventricle lesions. Recognizing the potential deleterious effects of circulatory arrest, we have continued to develop our technique. We currently use brief periods of complete circulatory arrest (5-10 minutes) in association with very low flow

Table III. *Preoperative and intraoperative variables associated with intubation time*

	Intubation time ≤3 days	*Intubation time >3 days*	
Parametric data	*Mean ± CI (range, n)*	*Mean ± CI (range, n)*	P *value*
Age (mo)	23.0 ± 1.3 (14-45, 74)	25 ± 3.1 (12-50, 24)	>.2
Weight (kg)	10.6 ± 0.37 (7-17, 77)	11.1 ± 0.8 (8.6-15, 23)	>.2
SAS (%)	83 ± 1.0 (74-89, 73)	83 ± 2.1 (68-89, 22)	>.2
RAP (mm Hg)	63 ± 0.6 (2-16, 69)	6.6 ± 1.3 (0.4-6.2, 20)	>.2
PAP (mm Hg)	11.3 ± 0.6 (7-18, 71)	12.0 ± 1.8 (7-17, 22)	>.2
PVR (WU × m^2)	2.05 ± 0.24 (0.66-4.5, 61)	2.18 ± 0.7 (0.4-6.2, 18)	>.2
McGoon ratio	1.51 ± 0.08 (0.86-211, 61)	1.38 ± 0.13 (1.05-1.68, 14)	.13
CPB time (min)	76.5 ± 8.2 (43-196, 74)	108.0 ± 9.8 (69-155, 23)	.0001
Crossclamp time (min)	44.5 ± 3.7 (24-105, 77)	57.6 ± 6.1 (34-98, 23)	.0026
Nonparametric data			
Systolic function	n = 75	n = 23	>.2*
Normal ventricular function	60 (80%)	17 (74%)	
Mild-moderate ventricular dysfunction	11 (15%)	6 (26%)	
Severe ventricular dysfunction	4 (5%)	0 (0%)	
Tricuspid regurgitation	n = 73	n = 24	>.2*
No regurgitation	45 (62%)	15 (63%)	
Mild regurgitation	20 (27%)	7 (29%)	
Moderate regurgitation	4 (5.5%)	1 (4%)	
Severe regurgitation	4 (5.5%)	1 (4%)	
Collateral blood flow	n = 62	n = 15	>.2*
0	8 (13%)	2 (13%)	
1	14 (23%)	6 (40%)	
2	22 (35%)	4 (27%)	
3	10 (16%)	2 (13%)	
4	5 (9%)	1 (7%)	
5	2 (3%)	0 (0%)	
6	1 (2%)	0 (0%)	

Degree of association of preoperative and intraoperative variables with intubation time. Prolonged intubation time was highly associated with longer cardiopulmonary bypass and aortic crossclamp time. *CI*, Confidence interval; *SAS*, systemic arterial saturation; *RAP*, right atrial pressure; *PAP*, pulmonary artery pressure; *PVR*, pulmonary vascular resistance; *WU*, Wood units; *CPB*, cardiopulmonary bypass.
*Statistical testing with Wilcoxon rank sum test.
†Preoperative values.

bypass and venous return provided by nonocclusive suckers to the CPB circuit.

Although the short-term survival of the patients in this report (79%) is not as good as published reports of all patients undergoing the Fontan procedure when viewed as a whole, the survival of patients treated by technique II (98%) compares favorably with the best reported results.[4] Thus, the diagnosis of HLHS in itself does not militate against low operative morbidity and mortality.

A marked improvement in morbidity and mortality was observed between patients undergoing the Fontan procedure with technique II as compared with technique I. The preoperative characteristics of the patients in the 2 groups were very similar. The difference in age between these groups was statistically significant, although a mean age difference of 2 months is likely clinically irrelevant. These differences could be the result of one or more factors, including technique of stage 2 palliation (BDG vs HFP), differences in patient selection, improved postoperative patient care, reduction in CPB time and/or aortic crossclamp time, the use of MUF, or yet other unknown factors. Because of the nearly concurrent nature of these factors, discerning those responsible for this improvement in outcome may not be possible. We favor the notion that the reduction in CPB and aortic crossclamp time largely accounts for the improved mortality for several reasons. As previously noted, the trend toward lower mortality in HFP patients operated on by technique II rather than technique I would suggest that the type of stage 2 palliation was less important than the type of surgical technique used for the Fontan operation. Although patient selection may have tended to eliminate higher-risk patients during the time that technique

Table IV. *Preoperative and intraoperative variables associated with length of ICU stay*

	ICU stay ≤ 7 days	*ICU stay > 7 days*	*ICU stay*
Parametric data	*Mean ± CI (range, n)*	*Mean ± CI (range, n)*	P *value*
Age (mo)†	23 ± 1.3 (14-45, 73)	24.8 ± 2.9 (18-50, 26)	>.2
Weight (kg)†	10.6 ± 0.4 (8.4-17, 73)	10.9 ± 0.8 (7-15, 25)	>.2
SAS (%)†	83 ± 1.0 (74-93, 72)	82 ± 3 (68-89, 24)	>.2
RAP (mm Hg)†	6.4 ± 0.6 (2-16, 68)	6.5 ± 1.3 (1-14, 22)	>.2
PAP (mm Hg)†	11.5 ± 0.6 (7-18, 71)	11.5 ± 1.2 (7-17, 24)	>.2
PVR (WU × m^2)†	2.0 ± 0.2 (0.66-4.5, 61)	2.3 ± 0.7 (0.40-6.2, 19)	>.2
McGoon ratio†	1.52 ± 0.08 (0.86-2.11, 61)	1.33 ± 0.13 (1.05-1.97, 15)	.09
CPB time (min)	76.0 ± 8.0 (43-196, 73)	109.0 ± 9.8 (69-157, 25)	<.0001
Crossclamp time (min)	44.0 ± 3.7 (24-105, 73)	57.6 ± 6.9 (34-98, 25)	.0009
Nonparametric data			
Systolic function†	n = 73	n = 25	>.2*
Normal ventricular function	59 (81%)	18 (72%)	
Mild-moderate ventricular dysfunction	10 (14%)	7 (28%)	
Severe ventricular dysfunction	4 (5%)	0 (0%)	
Tricuspid regurgitation†	n = 73	n = 25	>.2*
No regurgitation	45 (62%)	15 (60%)	
Mild regurgitation	19 (26%)	8 (32%)	
Moderate regurgitation	4 (5%)	2 (8%)	
Severe regurgitation	5 (7%)	0 (0%)	
Collateral blood flow†	n = 58	n = 16	>.2*
0	8 (14%)	1 (6%)	
1	14 (24%)	6 (38%)	
2	19 (33%)	5 (31%)	
3	10 (17%)	2 (13%)	
4	4 (7%)	2 (13%)	
5	2 (3%)	0 (0%)	
6	1 (2%)	0 (0%)	

Degree of association of preoperative and intraoperative variables with length of stay in the intensive care unit. Prolonged intensive care unit stay was associated with longer cardiopulmonary bypass and aortic crossclamp times. *ICU,* Intensive care unit; *CI,* confidence interval; *SAS,* Systemic arterial saturation; *RAP,* right atrial pressure; *PAP,* pulmonary artery pressure; *PVR,* pulmonary vascular resistance; *WU,* Wood units; *CPB,* cardiopulmonary bypass.

*Statistical testing with Wilcoxon rank sum test.

†Preoperative values.

II was being used, we have not consciously modified our selection criteria for the Fontan operation, and the preoperative characteristics of the patients in the technique I and II groups were nearly identical (Table I). Similarly, we have not made any changes in postoperative management that likely account for the difference in mortality. Other investigators have found longer CPB and crossclamp times to correlate with less favorable outcomes in the patient requiring the Fontan operation.[3,4,22] Given the marked sensitivity of the Fontan patient to reduced ventricular compliance and increased pulmonary vascular resistance, both of which can be caused by CPB and myocardial ischemia related to a period aortic crossclamping, it is not surprising that these factors may adversely affect outcome. Of note, despite a period of circulatory arrest, technique II resulted in a significantly shorter duration of CPB. Recently, hypothermic low-flow CPB has been reported to produce greater pulmonary dysfunction than periods of circulatory arrest.[23] Perhaps reducing this period of CPB increases lung compliance in patients undergoing technique II, thereby improving their postoperative course. Importantly, although our technique involves a period of circulatory arrest, we do not want to suggest that circulatory arrest imparts any survival advantage. The use of a single atrial cannula and circulatory arrest has significantly simplified the procedure, but given the potential risks associated with its use, we have tried to minimize the periods of complete circulatory arrest. Currently, we use brief periods of circulatory arrest and perform the majority of the procedure with reduced-flow sucker bypass.

Right ventricular function, as assessed by preoperative right atrial pressure and a subjective echocardiographic estimate of systolic function, did not prove to be a predictor of survival. It is possible that this is a reflec-

Table V. *Preoperative and intraoperative variables associated with length of hospital stay*

	Hospital stay ≤ 16 days	*Hospital stay > 16 days*	
Parametric data	*Mean ± CI (range, n)*	*Mean ± CI (range, n)*	P *value*
Age (mo)	22.9 ± 1.4 (14-45, 68)	24.8 ± 2.4 (17-50, 32)	.15
Weight (kg)	10.3 ± 0.33 (7-14.5, 70)	11.5 ± 0.7 (8.6-17, 30)	.007
SAS (%)	83 ± 1.0 (74-93, 67)	82 ± 2.0 (68-88, 29)	>.2
RAP (mm Hg)	6.3 ± 0.7 (2-16.0, 63)	6.6 ± 1.0 (1-14, 27)	>.2
PAP (mm Hg)	11.5 ± 0.6 (7-18, 65)	11.6 ± 1.0 (7-17.0, 29)	>.2
PVR (WU × m^2)	2.1 ± 0.3 (0.4-4.2, 57)	2.2 ± 0.5 (1.1-6.2, 23)	>.2
McGoon ratio	1.5 ± 0.09 (0.86-2.11, 54)	1.5 ± 0.10 (1.1-1.87, 22)	>.2
CPB time (min)	79.0 ± 8.2 (45-196, 68)	95 ± 12.9 (43-172, 31)	.05
Crossclamp time (min)	46.2 ± 3.8 (24-98, 68)	51.5 ± 6.5 (29-105, 31)	.16
Nonparametric data			
Systolic function	n = 67	n = 31	>.2*
Normal ventricular function	51 (76%)	26 (84%)	
Mild-moderate ventricular dysfunction	12 (18%)	5 (16%)	
Severe ventricular dysfunction	4 (6%)	0 (0%)	
Tricuspid regurgitation	n = 67	n = 30	>.2*
No regurgitation	41 (61%)	19 (63%)	
Mild regurgitation	17 (25%)	9 (30%)	
Moderate regurgitation	4 (6%)	2 (7%)	
Severe regurgitation	5 (7%)	0 (0%)	
Collateral blood flow	n = 54	n = 21	>.2*
0	7 (13%)	2 (10%)	
1	16 (29%)	4 (19%)	
2	15 (28%)	10 (48%)	
3	10 (19%)	2 (10%)	
4	3 (6%)	3 (14%)	
5	2 (4%)	0 (0%)	
6	1 (2%)	0 (0%)	

Analysis of variables associated with total hospital stay. Total hospital stay was associated with longer cardiopulmonary bypass time. In addition, a statistically significant but likely clinically unimportant association of preoperative weight with hospital stay was found. *CI*, Confidence interval; *SAS*, systemic arterial saturation; *RAP*, right atrial pressure; *PAP*, pulmonary artery pressure; *PVR*, pulmonary vascular resistance; *WU*, Wood units; *CPB*, cardiopulmonary bypass.
*Statistical testing with Wilcoxon rank sum test.

tion of selection bias (that patients with poor function did not undergo Fontan palliation). However, in this series 21 patients estimated to have moderately to severely depressed function were operated on, and 20 survived. Thus, moderate degrees of right ventricular dysfunction may be well tolerated at least in the short to intermediate term. During the time period of the study only 2 patients underwent cardiac transplantation after an HFP: one with moderate right ventricular dysfunction, moderate to severe tricuspid regurgitation, and elevated pulmonary vascular resistance, and the second with moderate right ventricular dysfunction and bilateral pulmonary artery stenoses after bilateral endoluminal stent placement. In both cases it was the combination of right ventricular dysfunction with another perceived risk factor that fostered the decision to proceed with transplantation. Both of these patients are alive and doing well after their transplant operations.

The degree of preoperative tricuspid regurgitation also did not have a significant effect on survival. This may be due to selection bias (only 11 patients had moderate to severe tricuspid regurgitation) and perhaps an aggressive policy of performing tricuspid valvuloplasty at the time of the Fontan procedure for all but minimal degrees of regurgitation.[24] Pulmonary artery size appears to influence survival in some but not all previous reports[25-27] and did not predict survival in our series. This may be related in part to the routine bilateral augmentation of the pulmonary arteries at the HFP, which resulted in adequate-sized pulmonary arteries for the vast majority of patients at the Fontan procedure. In addition, the use of intravascular pulmonary artery stents at the Fontan operation may also have reduced preoperative pulmonary artery size as a risk factor. Our data also suggest that the magnitude of aortopulmonary collateral blood flow is not a significant

short-term risk factor for Fontan patients. However, the method used to estimate this flow was crude and this question requires more quantitative exploration.

Unfortunately, we do not know what effect MUF may have had on outcomes in these patients. It is possible that the use of MUF improved outcomes, perhaps by reducing extravascular fluid or reducing circulating inflammatory mediators.[28-30] We began using MUF in April 1996; hence, almost all of the technique II group and none of the technique I group had been treated with this modality. We cannot therefore separate the possible confounding effect of MUF from differences related to surgical technique.

In conclusion, survival after the Fontan procedure for patients with HLHS has improved markedly and is equivalent to the results for other univentricular lesions.

The potential negative effects of pulmonary artery size can be ameliorated, at least in the case of the surgically accessible pulmonary arteries, by the routine augmentation of the pulmonary arteries at the time of the HFP or with the use of endoluminal stents. Tricuspid regurgitation should be aggressively treated and, if surgically remediable, does not unduly increase the risk of the Fontan procedure.

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