The extracardiac conduit Fontan operation using minimal approach extracorporeal circulation: Early and midterm outcomes

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Objective: Our approach to the extracardiac conduit Fontan operation has evolved over time from full-pump, to partial-pump, to completely off-pump. This study is designed to report our overall experience with the extracardiac conduit Fontan operation and to evaluate the evolution in bypass technique on postoperative outcomes.

Methods: From September 1992 to April 2005, 285 patients, median age 4.5 years (1.4-44 years), median weight 16 kg (9.4-94 kg), underwent a primary extracardiac conduit Fontan procedure. Early and late outcomes were analyzed for the entire cohort and for 2 patient groups depending on whether an oxygenator was used in the bypass circuit (166 patients; 58%) or not (119 patients; 42%).

Results: Early failure (including death and takedown) occurred in 7 patients (2.5%). Prevalence of new early postoperative sinus node dysfunction necessitating a permanent pacemaker was 0.4%, and that of new tachyarrhythmias necessitating discharge home on a regimen of antiarrhythmia medications was 2.5%. Ten-year actuarial freedom from Fontan failure, new sinus node dysfunction necessitating a permanent pacemaker, and reoperation for conduit thrombosis or stenosis was 90%, 96%, and 98%, respectively. Fenestration rate was lower ($P = .001$) in the no-oxygenator group (8%) than in the oxygenator group (25%). Patients in the no-oxygenator group had lower intraoperative Fontan pressure (12.0 ± 2.3 vs 13.5 ± 2.4 mm Hg, $P < .001$), common atrial pressure (4.6 ± 1.8 vs 5.3 ± 1.8 mm Hg, $P = .003$), and transpulmonary gradient (7.5 ± 2.1 vs 8.3 ± 2.2 mm Hg, $P = .013$) than did the oxygenator group.

Conclusions: The extracardiac conduit Fontan operation coupled with minimal use of extracorporeal circulation is associated with favorable intraoperative hemodynamics, low fenestration rate, minimal risk of thrombosis or stenosis, and minimal early and late rhythm disturbance.

The optimal Fontan connection requires a combination of factors that, when viewed in light of the dichotomy of the currently available options between the lateral tunnel (LT) and extracardiac conduit (EC) Fontan operations, are frequently conflicting. Among these factors are the provisions for growth potential,

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Abbreviations and Acronyms

ASA	= acetylsalicylic acid
CPB	= cardiopulmonary bypass
EC	= extracardiac conduit
IVC	= inferior vena cava
LT	= lateral tunnel
PA	= pulmonary artery
PTFE	= polytetrafluoroethylene
SVC	= superior vena cava

avoidance of atrial surgery, and avoidance of atrial distention. We believe that the rhythm-stabilizing advantages of the EC Fontan, related to avoidance of atrial surgery and atrial distention, outweigh its disadvantage related to lack of growth potential, which can be controlled for by doing the operation at an older age. This study in part is a report of our overall early and midterm experience with the EC Fontan operation and examines the evidence for and against this technique.

Given the inherently marginal nature of the post-Fontan hemodynamics related to lack of a pulmonary ventricle, improved early postoperative outcomes require optimization of perioperative cardiac output and reduction of systemic venous/Fontan pressure. This can be achieved by reducing the inflammatory and ischemic stresses of the operation and by fenestrating the Fontan circuit. Fontan fenestration, however, has both short- and long-term consequences, and with the EC technique, it requires limited additional atrial surgery. Instead, we have chosen to focus on minimizing the perioperative inflammatory and ischemic stresses by continuing to modify the extracorporeal circulatory support associated with the operation. This study is therefore also a report of our evolving technique and results with the minimal approach extracorporeal circulation.

Patients and Methods

This is a retrospective analysis of 302 consecutive patients who underwent an EC Fontan operation between September 1992 and April 2005. Of these, 285 patients underwent a primary Fontan, and 17 had revision of an atriopulmonary or LT Fontan to an EC procedure. The 285 patients who underwent a primary Fontan are the focus of this study. All operations were performed by a single group of pediatric cardiac surgeons with an internally consistent strategy. All patients underwent an extracardiac connection. The patients' preoperative management was designed to avoid concomitant intracardiac procedures by performing them at the time of the bidirectional Glenn or at a separate procedure. To reduce the need for reoperation for Fontan pathway obstruction related to lack of conduit growth potential, the operation was not performed until the patient reached a weight of about 15 kg (about 3-5 years of age), so that a large, at least 18-mm and more commonly a 20-mm conduit could be inserted. Intraoperative management centered on performing a large pulmonary artery (PA) anastomosis using a

beveled conduit and incorporating it into aggressive pulmonary arterioplasties. Care was taken to avoid PA, conduit, and inferior vena cava (IVC) distortion and pulmonary venous compression. During the mediastinal dissection, the sinus node area was carefully identified and direct trauma and traction on the node were avoided. The circulatory support technique evolved over the course of our experience. Four different circulatory techniques were used.

Full Cardiopulmonary Bypass (CPB): 1992 to 1998

Early in our experience, the entire operation, including the conduit-PA and conduit-IVC anastomoses, was performed on full CPB. The aorta, superior vena cava (SVC), and IVC were cannulated, and an oxygenator was used in the bypass circuit. From the outset, induced cardiac arrest was avoided and the operation was performed on a warm beating heart unless a concomitant intracardiac operation was necessary.

Partial CPB: 1998 to 2000

Midway in our experience, we realized that the PA anastomosis could be safely performed without the need for circulatory support. The PA anastomosis site was first isolated with vascular clamps placed toward the contralateral lung and immediately adjacent to the Glenn anastomotic site, such that adequate Glenn flow was maintained to the ipsilateral lung. Care was taken to ensure adequate oxygenation and ventilation by monitoring the systemic oxygen saturation and end-tidal carbon dioxide level and by taking care to avoid obstruction of SVC blood flow. In patients who had a jugular central venous line in place, with the PA clamps temporarily in place, the Glenn pressure was monitored to ensure that it did not go above 15 to 20 mm Hg. The clamps were accordingly adjusted before committing to the incision and anastomosis. After completion of the PA anastomosis, the conduit was clamp occluded and the PA clamps were removed, re-establishing Glenn flow to both lungs. The aorta and IVC were then cannulated, and with an oxygenator in the circuit, the IVC anastomosis was performed on partial warm CPB. With this technique, CPB times were reduced dramatically to about 10 to 20 minutes.

Active IVC Decompression: 2000 to 2002

With further experience it became evident that safe performance of the IVC anastomosis did not require the use of an oxygenator. Since oxygenation and ventilation were maintained by Glenn flow to the lungs, only a pump was necessary to decompress the IVC and support systemic blood flow. After completion of the PA anastomosis, the aorta and IVC were cannulated and, without an oxygenator in the circuit, the IVC blood was drained and pumped to the aorta with either a roller pump or, more commonly, a centrifugal pump during performance of the IVC anastomosis. With this technique, the oxygenator was completely avoided and pump times remained low in the 10- to 20-minute range.

Passive IVC Decompression (Off-pump): 2002 to 2005

In the most recent phase of our experience with the EC Fontan operation, the pump was also eliminated, allowing the operation to be performed completely off-pump. After completion of the PA anastomosis, the IVC and right atrium were cannulated and connected to each other directly, allowing passive lower body venous

TABLE 1. Distribution of patients among the four different circulatory strategies and in the oxygenator and no-oxygenator groups

Circulatory Strategy	No. (%)
Oxygenator group	166 (58)
Full CPB	115 (40)
Partial CPB	51 (18)
No-oxygenator group	119 (42)
Active IVC decompression	66 (23)
Passive IVC decompression	53 (19)

CPB, Cardiopulmonary bypass; IVC, inferior vena cava.

decompression during the IVC anastomosis. In patients with a femoral central venous line in place, with the IVC clamps temporarily in place, the pressure was monitored to ensure that it did not go above 15 to 20 mm Hg. If necessary, the patient was placed in a mild Trendelenburg position to aid lower body drainage. Mild systemic hypotension was treated with volume administration.

Fenestrations were never routinely performed, but they were added to the procedure when the surgeon deemed them important to patient outcome. After completion of the operation, hemodynamic status was evaluated intraoperatively. If Fontan pressure was consistently above 18 mm Hg, with a transpulmonary gradient of 12 mm or more, a fenestration was placed between the conduit and the right atrial free wall. This was accomplished without CPB, either with a direct 4- to 6-mm side-to-side anastomosis or with a 4- to 8-mm polytetrafluoroethylene (PTFE) tube graft.

Data Collection and Follow-up

Institutional review board approval for a retrospective clinical study was obtained. Perioperative data were obtained by review of hospital medical records. Late follow-up data were obtained from the patients' cardiologists or pediatricians. Follow-up information collected included copies of physician clinic notes, resting electrocardiogram, ambulatory rhythm monitor, echocardiogram, and other diagnostic information as indicated. The early postoperative period was defined as the time from surgery to hospital discharge. The late postoperative period was defined as the time from initial hospital discharge to latest follow-up. Late arrhythmia data were based on the patients' histories of symptoms and follow-up electrocardiograms. There was no protocol for systematic arrhythmia detection such as regularly scheduled Holter monitoring. Early and late outcome variables were compared among the four different circulatory support categories. Data analysis in these four categories revealed that the full CPB subgroup and the partial CPB subgroup showed no major differences in outcome. Thus these two subgroups were combined into one group, the oxygenator group. Similarly, the active IVC decompression (pump only) subgroup and the passive IVC decompression (off-pump) subgroup showed no major differences; thus they were combined into another group, the no-oxygenator group. Early and late outcome variables were compared between the oxygenator and no-oxygenator groups. There were no significant differences in age, sex, weight, dominant ventricle, prevalence of heterotaxy, or conduit size between the oxygenator and no-oxygenator groups. Mean follow-up time was 3.7 years (0.1-11.7 years). At least one postdischarge follow-up was

available in 96% of the patients. A total of 12,206 patient-months of actual follow-up were available. Ratio of actual to potential follow-up months yielded an 85% completeness of follow-up. Continuous variables between the two groups were compared with independent samples *t* test. Correlation between two continuous variables was assessed with linear regression analysis. Freedom from event was calculated by Kaplan-Meier survival estimates.

Results

Median age was 4.5 years (1.4-44 years) and median weight was 16 kg (9.4-94 kg). In 45 patients (16%), the operative weight was less than 14 kg, and in 20 patients (7%), it was less than 13 kg. The morphologic characteristic of the dominant ventricle was that of right or indeterminate ventricle in 102 of 282 patients (36%) and that of a left ventricle or combined unseptatable left and right ventricles in 180 patients (64%). A PTFE conduit (Gore-Tex Stretch Vascular Graft; W. L. Gore & Associates, Inc, Flagstaff, Ariz) was used in 270 of 285 patients (95%). Early in our experience, we used homograft tissue in 12 patients. Three patients underwent autologous tissue extracardiac Fontan by anastomosing the main PA directly to the IVC. Median conduit size was 20 mm (16-24 mm). Conduit size was 18 mm or larger in 97% of the patients and 20 mm or larger in 69%. By definition, no patients in the no-oxygenator group had a concomitant intracardiac operation. By contrast, 10 patients in the oxygenator group (6%) had a concomitant intracardiac operation. Crossclamping of the aorta was avoided in 271 patients (95%). The PA-conduit anastomosis was performed off-pump, before the IVC anastomosis, in 174 patients (65%). Distribution of patients among the different bypass strategy groups is depicted in Table 1. There was a significant difference in the rate of oxygenator use in the first 143 cases (87%) in our series compared with the last 142 cases (30%) ($P < .001$). In the oxygenator group, a significant correlation was found between duration of CPB and intraoperative Fontan pressure; longer CPB duration was associated with higher Fontan pressure (Figure 1). Intraoperative hemodynamic data including Fontan and common atrial pressures and transpulmonary gradients were all lower in the no-oxygenator group (Table 2). There were no differences in these parameters within the subgroups of either the oxygenator or no-oxygenator groups. Early mortality occurred in 3 patients (1.1%). The circulatory support technique used in these patients was full CPB. There have been no deaths in our last 186 patients. Four patients (1.4%) required early Fontan takedown on postoperative days 0 to 4, and survived. Three of these patients underwent a second successful Fontan operation 1 to 4 years after their initial operation. Early Fontan failure rate (including death and takedown) was therefore 2.5%. All of the Fontan failures occurred in the oxygenator group. There were no perioperative cerebral events, hepatic injury, cardiac ischemia, or arrhythmias attributed to the limited extracorporeal tech-

Figure 1. Correlation between CPB time and intraoperative Fontan pressure (regression coefficient = 5.5, $P = .01$). OR, Operating room; CPB, cardiopulmonary bypass.

nique. There were no patients in the partial CPB, active IVC decompression, or passive IVC decompression groups who required conversion to full CPB because of difficulty with completion of the anastomoses. One patient in the passive IVC decompression group was subsequently found to have bleeding from the PA anastomosis and was placed on full CPB for repair. Four patients (1.4%) had early Fontan thrombosis 3 to 5 days postoperatively, necessitating reoperation and thrombectomy. Two of these events occurred in the oxygenator group and 2 in the no-oxygenator group. The circulatory support techniques used included full CPB in 2 patients, active IVC decompression in 1 patient, and passive IVC decompression in 1 patient. One of these patients underwent an autologous tissue direct PA-IVC anastomosis without use of a conduit. Anastomotic suture-line tension was suspected as the cause of the thrombosis. At reoperation, a PTFE graft was inserted. The second patient was 2.3 years old and weighed 12.5 kg. A 20-mm conduit was initially inserted. After thrombosis on postoperative day 4, presumably caused by patient-conduit size mismatch, the conduit was removed and was replaced with an 18-mm conduit. The third patient was a 13.2-kg patient who received a 20-mm conduit. Fontan thrombosis occurred on postoperative day 3

TABLE 2. Intraoperative hemodynamic data in the oxygenator versus no-oxygenator groups

Hemodynamic data	Oxygenator	No-oxygenator	P value
Fontan pressure	13.5 ± 2.4	12.0 ± 2.3	$P < .001$
Common atrial pressure	5.3 ± 1.8	4.6 ± 1.8	$P = .003$
Transpulmonary gradient	8.3 ± 2.2	7.5 ± 2.1	$P = .013$

All pressures are expressed in millimeters of mercury $\pm$ standard deviation.

TABLE 3. Hospital milestones

Milestone	Value
Ventilator duration less than 24 h	77%
Inotropic support less than 48 h	70%
Median chest tube duration (d)	8 (range 2-91)
Median hospital stay (d)	11 (range 4-106)

after administration of blood products at the time of reoperation for evacuation of a substernal hematoma. The fourth patient had marginal preoperative ventricular diastolic function with an end-diastolic pressure of 14 mm Hg and had low cardiac output postoperatively. There was no mortality or Fontan takedown in these 4 patients. A nonobstructive thrombus was detected on echocardiogram in 1 other patient before hospital discharge. It was treated with anticoagulation and resolved.

Forty-nine patients underwent intraoperative fenestrations. Of these procedures, 14 were side-to-side anastomoses and 31 were PTFE tube grafts (preferred technique). In 4 patients the fenestration type was not indicated in the operative report. Most intraoperative fenestrations were performed early in our experience between 1992 and 1995. Since 1995, intraoperative fenestration was used in only 24 (9.7%) of 247 patients undergoing an EC Fontan procedure. Fenestration rate was lower ($P = .001$) in the no-oxygenator group (9 patients, 8%) than in the oxygenator group (40 patients, 25%). At the extremes of circulatory management, the full CPB subgroup had the highest fenestration rate of 34%, whereas the fenestration rate in the latest subgroup of passive IVC decompression was 8% ($P < .001$).

Ninety-six percent of patients had preoperative sinus or nonsinus atrial rhythm. Among 278 patients who survived to hospital discharge with a functional Fontan (excluding early deaths and takedowns), 95% were in sinus or nonsinus atrial rhythm. Fixed (nontransient) sinus node dysfunction necessitating early postoperative implantation of a permanent pacemaker occurred in 1 patient (0.4%). Two other patients required a pacemaker for postoperative complete heart block (1 after ventricular septal defect enlargement). Five other patients underwent a planned intraoperative pacemaker insertion for preoperatively diagnosed dysrhythmias. Seven patients (2.5%) had recurrent (nontransient) newly diagnosed early postoperative supraventricular tachyarrhythmias necessitating discharge home on a regimen of antiarrhythmia medications: 3 with junctional tachycardia, 1 with atrioventricular tachycardia, 1 with intra-atrial reentrant tachycardia, and 1 with atrial fibrillation.

Of 259 patients in whom information was available, 200 (77%) were extubated within 24 hours after the operation. Other hospital milestones are listed in Table 3. Duration of chest tube drainage was longer ($P = .003$) in the full CPB group (11.8 ± 12.0 days) than in the passive IVC decom-

TABLE 4. PredischARGE and follow-up anticoagulant and antiplatelet management

Regimen	PredischARGE (%)	Follow-up (%)
Warfarin only	50	10
ASA only	17	74
Warfarin and ASA	30	8
No warfarin or ASA	3	8

ASA, Acetylsalicylic acid.

TABLE 5. Late outcomes

Outcome	No.
Mortality	8
Fontan takedown	1
Cardiac transplantation	1
Intracardiac thrombi	5
Cerebrovascular accident	3
Protein-losing enteropathy	3
Plastic bronchitis	3
Late pacemaker for sinus node dysfunction	5
Late-onset supraventricular tachyarrhythmia	4

pression (off-pump) group (7.8 ± 3.5 days). There were no other differences in early outcome variables between the oxygenator and no-oxygenator groups or their subgroups. Chest tube drainage lasting more than 14 days occurred in 15% of the patients. Although somewhat variable, the general criteria for chest tube discontinuation was drainage of less than 1 to 2 mL $\cdot$ kg⁻¹ $\cdot$ d⁻¹ for 2 consecutive days. Among survivors to hospital discharge, 2 patients (1.1%) had an early postoperative cerebrovascular accident. Two of these patients underwent surgery on full CPB, and 1 patient

had active IVC decompression. Neither of these patients had a fenestration. PredischARGE room-air median oxygen saturation (including patients with patent fenestrations) was 96% (75%-99%). Ninety-seven percent of the patients were discharged home on a regimen of acetylsalicylic acid (ASA) or warfarin or both (Table 4).

Late Postoperative Data

Late outcomes are summarized in Table 5. Three patients with early mortality and 1 patient with early Fontan failure necessitating permanent takedown are excluded from the denominator in the late data analysis. Three other patients who had a second successful Fontan after initial takedown were counted only once (since the first operation) in the late data analysis. A total of 278 patients were therefore available for late analysis. Late mortality occurred in 8 of 278 patients (2.9%), from 3 months to 8 years after the initial operation (mean 2.4 years). Late Fontan takedown was required 3.4 years after the initial operation in 1 patient with protein-losing enteropathy. One heterotaxy patient underwent successful cardiac transplantation for worsening ventricular failure 4 months after the initial operation. Ten-year actuarial freedom from Fontan mortality was 91%, and freedom from Fontan failure (including mortality, takedown, and cardiac transplantation) was 90% (Figure 2).

There were no cases of late Fontan conduit thrombosis or stenosis necessitating reoperation. One patient in the passive IVC decompression group underwent repair of right PA stenosis during reoperation for atrioventricular valve regurgitation 23 months postoperatively. Ten-year actuarial freedom from reoperation for conduit thrombosis or stenosis was 98%. Late intracardiac thrombi were detected in 5 patients (1.8%) 2 months to 5 years postoperatively. Of these, 3 were in

Figure 2. Actuarial freedom from Fontan failure including mortality, takedown, and cardiac transplantation. Vertical bars indicate 70% confidence limits (CL). Numbers of patients at risk are shown in parentheses.

the systemic venous side, 1 was in the pulmonary venous side, and 1 was in the neo-aortic root. Late cerebrovascular accident occurred in 3 patients 2 months to 9 years after the operation. Protein-losing enteropathy occurred in 3 patients and plastic bronchitis developed in 3 other patients (Table 5).

At follow-up, 91% of the patients were in sinus or nonsinus atrial rhythm. Excluding 11 nonsurvivors, 3 survivors with early or late failure, 5 patients with pre-existing pacemakers, and 5 patients with planned intraoperatively implanted pacemakers, there were 262 patients available for rhythm analysis. Among these 262 patients, late rhythm information was available in 247 patients (95%). Among these 247 patients, the incidence of late sinus node dysfunction requiring insertion of a pacemaker was 2% (5 patients). Ten-year actuarial freedom from needing a pacemaker for sinus node dysfunction was 96%. Two patients were asymptomatic with chronic stable junctional rhythm and did not require pacemaker insertion. Three patients required a pacemaker for late-onset heart block. Excluding patients with pre-existing tachyarrhythmias, 4 patients (1.6%) had identified late-onset supraventricular tachyarrhythmias. At follow-up, 6 patients (2.3%) were receiving long-term antiarrhythmia medications for new or pre-existing supraventricular tachyarrhythmias.

There were no significant differences between the oxygenator and the no-oxygenator groups for any of the above late outcome measures. At a mean follow-up time of 3.7 years, 90% of the patients were in New York Heart Association functional class I, 9% were in class II, 0.4% were in class III, and none were in class IV. Late follow-up echocardiogram was available in 248 of 268 late survivors with a functional Fontan (excluding 3 with early Fontan failure who underwent successful reoperation). Among these 248 patients, ventricular function was graded qualitatively as normal or adequate in 237 patients (96%), whereas 3% had mild dysfunction and 1% had moderate or severe dysfunction. Late oxygen saturation was available in 185 patients. Median room-air oxygen saturation was 96% (73%-100%). This included patients with a patent fenestration. Breakdown of patients on the basis of type of anticoagulation regimen at follow-up is provided in Table 4. Ninety-two percent of the patients were receiving ASA or warfarin or both. The referring cardiologist managed the patients' anticoagulation after hospital discharge.

Discussion

The current options for long-term palliation of patients with functional single ventricle hearts remain primarily limited to the LT and the EC Fontan operations. Both procedures have distinct advantages and disadvantages that, given today's technology, cannot yet be consistently incorporated into a single operation. Since all of the patients in the current study had EC Fontan operations, direct comparison of the two

procedures is not possible from the information provided in this analysis. The study, however, does provide useful information addressing the various unproved claims, both in praise and in criticism, that have been made in relation to the EC Fontan operation.

The early and midterm Fontan failure rate, intraoperative hemodynamics, early and late outcomes, and midterm quality of life data all support the conclusion that the EC Fontan operation is a safe and reliable procedure that provides a durable and functional conduit. Favorable results with the EC Fontan have also been reported by several other centers.¹⁻⁴ A potential disadvantage of the EC Fontan operation is related to the lack of growth potential incorporated into the circuit.⁵ In our experience, this issue has been effectively dealt with by waiting to perform the operation until the patient reaches a weight of about 15 kg (about 3-5 years of age), so that a near adult IVC-sized conduit (≥ 18 mm in 97% of our cases) can be inserted. This strategy is designed to minimize the potential for radial outgrowth of the conduit and to accommodate the patient's future exercise demands. The potential for axial distortion with patient growth seems at least as likely with the PTFE baffle used in the LT as it is with an EC Fontan operation. Although long-term data are not available, to our knowledge none of our patients has required conduit replacement for growth-related pathway obstruction. Delaying the Fontan operation until 3 to 5 years of age incurs minimal additional risk. Worsening cyanosis does not generally develop until later, when patients start using more extensive lower body exercise and the head/lower body size ratio decreases further. For patients operated on before about 5 years of age, preoperative complications related to cyanosis and right-to-left shunt have been extremely uncommon in our series and have not justified earlier operation. Excluding patients who were referred for surgery at a much later age, development of clinically significant pulmonary arteriovenous malformations has been rare.

Another potential disadvantage of the EC Fontan operation, in the view of some, is the possibility of an unacceptable incidence of thrombosis. It is acknowledged by all that multiple factors including obligatory foreign material, systemic coagulation profile, nonpulsatile flow, and stasis related to lower postoperative cardiac output that may exist in all patients with a functional single ventricle after a Fontan operation, whether EC or LT, put them at higher risk for clotting problems; however, there seems to be no basis for the position that the EC Fontan is a greater risk. The area of foreign body in contact with the bloodstream might seem at first to be higher with the EC; however, it is important to note that the LT baffle is in contact with the blood twice, on both the left side and the right, so that the total area of contact is roughly similar between the EC and the LT. An important corollary to this is that the LT exposes both the

systemic and pulmonary circulations to the risk of foreign body–related thromboembolic complications, whereas with the EC the risk is confined to the pulmonary circulation.

Our experience supports the position that the risk of both early and midterm thrombosis is low for the EC Fontan, with freedom from reoperation of 98% at 10 years. Of the 4 cases of thrombosis necessitating reoperation, all occurred in the early postoperative period, with no late occurrence. One of the cases was technical and did not involve a conduit; it is not relevant to our current technique. This patient received a primary IVC-PA anastomosis, which was under tension. Of the 3 patients with PTFE grafts, 1 was considered a very high-risk candidate because of ventricular diastolic dysfunction, suggesting a hemodynamic cause resulting in low cardiac output and stasis. The other 2 patients received 20-mm conduits, despite having body weights of 12 and 13 kg. This suggests that patient weight–conduit diameter mismatching (oversizing) may be causative.⁶ Careful attention to patient selection and appropriate graft sizing should reduce even further the already low incidence of early postoperative graft thrombosis. Beyond the early postoperative period, only 5 patients in our series were found to have intracardiac thrombi. Of these thrombi, 3 were in the systemic venous side, 1 in the pulmonary venous side, and 1 in the neo-aortic root. These results compare favorably with the incidence of post-Fontan thromboembolic events reported in the literature.⁷⁻¹⁰ Our prophylactic strategy has been warfarin alone or in combination with ASA for the vulnerable first 3 months postoperatively to allow for endothelialization of the suture lines. This is then followed by lifelong ASA therapy with yearly transthoracic echocardiographic monitoring.

Institutions that perform primarily an LT have reported favorable overall results with the Fontan operation.¹¹⁻¹³ Atrial arrhythmias, however, continue to be a concern with the LT technique. In one large series of exclusively LT Fontan operations, freedom from new bradyarrhythmias was 79% at 10 years.¹¹ Our data show a high incidence of either sinus rhythm or nonsinus atrial rhythm, both early and midterm, and low incidences of new-onset pacemaker placement and supraventricular tachyarrhythmias necessitating antiarrhythmia medications. Although the outcome variables examined are different between studies, these results compare favorably with reports in the literature on rhythm disturbance after the LT Fontan^{11,14} and are supported by other centers performing the EC Fontan.^{2,4,15} There are important rhythm-related differences between the two techniques. The LT Fontan involves extensive atrial surgery, including in the vicinity of the sinus node and crista terminalis, whereas the EC Fontan involves minimal atrial surgery, confined to the IVC transection site, an area that is remote from the sinus node. Furthermore, after the LT Fontan, a portion of the right atrium, including the sinus

node and crista, is suddenly exposed to the higher-pressure Fontan circuit, whereas after the EC Fontan, the entire atrium is in contact with the low-pressure pulmonary venous circuit. Avoidance of pressure-related atrial stretch on the sinus node and crista, along with the minimal atrial incisions and suture lines associated with the EC Fontan, may possibly avoid the long-term electrophysiologic substrates for development of atrial arrhythmias.

An important principle of the Fontan operation is to achieve a nonobstructive Fontan pathway. Pathway obstruction is more frequently an issue at the superior (PA) and at the Fontan connection. The main objectives are to create a large PA anastomosis and to avoid branch PA stenosis. An LT Fontan performed after, or incorporating, a hemi-Fontan type operation (which creates a large superior connection) is frequently nonobstructive. The same cannot be said with certainty regarding an LT connection through the cardiac end of the SVC or the roof of the atrium, after a previous bidirectional Glenn. The price to be paid for a hemi-Fontan, however, is the need for significant atrial surgery in the vicinity of the sinus node and potential long-term arrhythmogenicity.¹⁴ The EC Fontan, on the other hand, can be done after, or incorporating, the technically simple and less invasive bidirectional Glenn, and it can be performed with a large PA anastomosis incorporated into PA plasty, thereby achieving a nonobstructive superior connection, without the need for atrial surgery. In our series, only 1 patient required additional surgery for branch PA stenosis, at the time of planned surgery for atrioventricular valve regurgitation.

Given the inherently marginal nature of the post-Fontan hemodynamics related to lack of a pulmonary ventricle, improved early postoperative outcomes require optimization of perioperative cardiac output and reduction of systemic venous/Fontan pressure. This can be achieved by fenestrating the Fontan circuit and by reducing the inflammatory and ischemic stresses of the operation. We have chosen to focus on the latter by continuing to modify the extracorporeal circulatory support associated with the operation. From the outset, we avoided ischemic arrest of the heart, unless occasionally necessitated by a concomitant intracardiac procedure. Our circulatory support management gradually evolved into the four support techniques described. We initially used full CPB, then transitioned to partial CPB with IVC cannulation only, then eliminated the oxygenator but kept the pump (active IVC decompression), and finally eliminated the pump as well (passive IVC decompression, or “off-pump”). Thus, the opportunity exists to examine the outcomes of an operation performed quite uniformly in essentially all respects, with the exception of a wide spectrum of circulatory support techniques. The four groups were then consolidated into two—the oxygenator and no-oxygenator groups. Further comparisons between

the oxygenator and the no-oxygenator groups showed a number of important differences, and thus this categorization of patients became the basis for further analysis. It should be emphasized from the outset, however, that the groups are not fully matched for comparison for several reasons. The groups are not contemporaneous, and the patients were not prospectively randomized to the various circulatory support techniques, but rather the technique was gradually modified over time and experience, based on perceived overall benefit to the patient. In addition, in the oxygenator group, 10 patients (6%) underwent a concomitant intracardiac operation, whereas in the no-oxygenator group, by definition, no patients had a concomitant intracardiac procedure. These issues create distinct limitations for interpreting the data and introduce potential bias; nevertheless, a number of interesting and potentially important insights can be gained by the analysis of the experience.

The transition from full to partial CPB dramatically reduced the bypass times and afforded a wide spectrum of CPB times with which to assess Fontan hemodynamics. There was a direct correlation between CPB time and higher Fontan pressure (Figure 1). Comparison of the oxygenator and no-oxygenator groups revealed that the Fontan pressure, common atrial pressure, and transpulmonary gradient were all higher in the oxygenator group (Table 2). All of the early Fontan failures occurred in the oxygenator group. Fenestration rate was considerably higher in the oxygenator group (25%) than in the no-oxygenator group (8%), with the highest rate being in the full CPB subgroup (34%). These results agree with those of several studies that show the length of CPB to be associated with worse early outcomes.^{2,3,13,16,17} Although these observed differences should be viewed with caution given the inherent differences between the groups, it can be argued that elimination of the oxygenator certainly did not result in worse outcomes, but it may not have been the only factor that contributed to the improved outcomes. The observation that important differences are present in a variety of early outcome measures between the oxygenator group and the no-oxygenator group, but that there are no differences in these same outcome measures between the two subgroups that made up the no-oxygenator group (the active IVC decompression subgroup and the passive IVC decompression subgroup), suggests that elimination of the oxygenator is important but elimination of the pump is not.

The feasibility of performing the EC Fontan operation with limited extracorporeal support should be evaluated individually in each patient depending on the anatomy. Regarding the conduit-PA anastomosis, important factors to consider are the relative position of the Glenn connection (central versus eccentric) with respect to the branch PAs, the size of the branches, and the presence or absence of important branch PA stenosis. At the most favorable extreme, an eccentrically positioned Glenn with large nonstenotic

branch PAs is best suited for an off-pump technique, whereas a centrally placed Glenn with bilateral branch PA stenoses is not. The final determinant of suitability of an off-pump approach is the avoidance of intraoperative oxygen desaturation, low end-tidal carbon dioxide readings, or cerebral venous hypertension during temporary clamp occlusion of the PAs. Since the conduit-PA anastomosis is performed toward the lung contralateral to the Glenn connection (while the Glenn perfuses the ipsilateral lung), offsetting of the Glenn and Fontan anastomoses is a natural consequence of the off-pump technique. The IVC anastomosis does not require special maneuvers, as it is performed in a similar manner whether accomplished on-pump or off-pump. The determinant of suitability of the off-pump technique is the avoidance of important hypotension during temporary clamp occlusion of the IVC while the IVC–right atrial shunt is open. Mild degrees of hypotension can be dealt with by placing the patient in a Trendelenburg position to aid lower body drainage and administration of volume.

It has been argued that routine Fontan fenestration improves early postoperative outcomes.^{12,18,19}

At institutions that have primarily performed an LT Fontan, a large majority of the patients are discharged home with a fenestration.¹¹⁻¹³ In contrast, fenestration rate in our last 247 patients has been less than 10%. Fenestration rate as low as 15% has also been reported with the EC Fontan operation performed with a short mean duration of full CPB.³ We would agree that if the Fontan completion were to be done with prolonged full extracorporeal support, hypothermia, ischemic cardiac arrest, and total circulatory arrest, a fenestration would be a more frequent necessity, with either the LT or the EC Fontan. This has also been the case early in our experience when the operation was performed on full CPB with more prolonged bypass times associated with the early learning curve and more frequent intracardiac procedures.²⁰ The extracardiac nature of the EC Fontan operation, however, allows for the operation to be performed with a variety of bypass techniques and avoidance of ischemic cardiac arrest. Our approach has been to take full advantage of this flexibility and to minimize extracorporeal support to the extent possible. This approach has helped in achieving favorable early postoperative outcomes without the need for a fenestration. Short- and long-term postoperative issues related to a patent fenestration, including cyanosis, potential complications related to a right-to-left shunt, and need for an additional procedure to close the fenestration, are therefore avoided. It should also be mentioned that with the EC Fontan technique, there is a distinct incentive to avoid a fenestration. Fenestrating an EC requires a limited amount of additional atrial surgery (with either a side-to-side or a tube graft fenestration). Since avoiding atrial surgery is one of the important advantages of the extracardiac technique, optimizing the intraoperative

hemodynamics in an attempt to avoid the need for a fenestration has been a natural consequence of our commitment to the EC Fontan operation.

In the current study, there were no important differences between the oxygenator group and the no-oxygenator group for any late outcome measures, implying that the influence of circulatory support technique is much greater in the perioperative period and this influence fades as time after surgery increases.

Conclusions

The EC Fontan procedure is a safe, reliable, and effective modification of the Fontan operation. This study supports the view that this procedure provides a unique benefit by minimizing early and possibly also midterm atrial rhythm disturbances, as well as the view that concerns over a high rate of growth-related conduit obstruction and conduit thrombosis are unwarranted. The EC Fontan operation also provides the unique opportunity for maximal flexibility with respect to the circulatory support methods used in the operation. Avoidance of an oxygenator seems to confer a number of early benefits, including improved hemodynamics and a reduced need for fenestration. In contrast, presence or absence of a pump appears to have no effect on early outcome. The operation can therefore be performed either off-pump or with active IVC decompression using a pump, at the discretion of the operating surgeon, with equal benefit.

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Discussion

Dr Marshall L. Jacobs (Philadelphia, Pa). With this superb series of EC Fontan procedures, Dr Hanley's group has set the bar high with respect to operative outcomes. Emphasis has been placed on increasing independence from CPB or partial circulatory support over the course of the series. In the manuscript, the authors acknowledge that the influence of circulatory support strategy is entirely in the early perioperative period. The principal hemodynamic benefit of avoiding bypass appears to be a decrement of approximately 1 mm Hg in the atrial pressure, the transpulmonary gradient, and the venous pathway pressure at the end of the operation.

There is no question as to the quality of these outcomes; they are excellent. One could stand here and present a series of comparable outcomes with Fontan operations using different means of cardiopulmonary support or different means of constructing the total cavopulmonary connection, but that would not make for an interesting discussion. What it would do is to raise an interesting question. There is clearly no question that a no-bypass strategy limits the surgeon's choice to that of EC Fontan only, eliminating the possibility of an intra-atrial tunnel reconstruction. One must decide, then, whether the EC Fontan is the optimal palliation for all functional univentricular hearts. If not, does avoidance of bypass justify limiting the scope of surgery to this one type of reconstruction?

In 1999, Fogel at the University of Pennsylvania used magnetic resonance pulse labeling to study the distribution of venous flow to the right and left PAs in patients on whom we had performed

hemi-Fontan procedures followed by LT completion Fontans. The distribution of flow to the right and left lungs was essentially equal. Of the blood in the right PA and the left PA, 48% and 31%, respectively, came from the IVC.

More recently, using computational fluid dynamics, Ed Bove and Mark de Leval not only found that the LT after the hemi-Fontan had considerably lower power losses than the EC Fontan, but they corroborated our earlier finding with IVC flow to the right lung being 52% for the LT, just 19% for the EC, and only 15% for the EC with a bevel to the left lung, as illustrated by Dr Hanley. Clearly, the type of surgical reconstruction affects where the venous blood goes, and Dr Hanley's own group has been among those taking the lead in emphasizing the important influence of hepatic venous blood on the health of the pulmonary vasculature.

My first question is this. Recognizing the potential for late-phase deterioration of the Fontan circulation and notwithstanding the excellent short-term results that you have achieved, do you have any concern that the effort to minimize the use of bypass has relegated your patients to a type of cavopulmonary connection that might have less than optimal properties with respect to energy conservation or with respect to the long-term health and function of the pulmonary vascular bed?

Dr Hanley. Thank you, Marshall, for your pertinent and thoughtful questions and analysis. Yes, we do have these concerns, and, of course, we are monitoring these patients in an ongoing fashion. I think you can pick your study from those studies available in the literature, in terms of computational fluid dynamics, and certainly there are others that state very definitively that an EC provides the least amount of overall energy loss.

In terms of the distribution of flow to each lung, we position the EC so that it goes off to the left side. I see that as a very good arrangement in terms of energy loss. It may bias the IVC flow to the left side, but certainly not exclusively to the left. We have magnetic resonance images that back this up. However, more importantly, from a practical standpoint, our mean oxygen saturation is 95% at late follow-up up to 12 years, indicating that pulmonary arteriovenous malformations are not developing, which is what I think you are concerned about here.

Dr Jacobs. My second and only other question pertains to potential consequences of cannulation, either for bypass or for minimal circulatory support. In 2001 at this meeting, Dr Hanley's group read a paper emphasizing the important adverse consequence of hemidiaphragm paralysis in Fontan patients, with a reported incidence of 2.7%. This complication was associated with significant morbidity, early from effusions and late from protein-losing enteropathy.

For this and other reasons, in our practice we completely avoid caval cannulation for the Fontan operation and instead have used a single venous cannula in the atrial appendage, and, a very different strategy from Dr Hanley's, a very brief period of hypo-

thermic circulatory arrest, to construct the cavopulmonary connection. Avoidance of redissection of the area around the cavae has resulted in complete freedom from phrenic nerve injury, and in 93 consecutive patients there have been no deaths and no Fontan failures. The only neurologic morbidity has been transient seizures in 1 patient with previous cerebral disease, which is a low incidence comparable to your series.

With the dissection and presumably tourniquet placement around caval cannulation sites, has there continued to be an incidence of postoperative diaphragm paralysis in your Fontan patients?

Dr Hanley. The earlier study that you referred to evaluated diaphragm palsy in patients late after Fontan, and it was looking at the impact of the inefficient diaphragm in the context of the limited Fontan physiology. Many of those diaphragm palsies had occurred before the Fontan operation, and the study was also multicentered, including several different Fontan techniques. Thus only a small number of diaphragm palsies have occurred in our hands at the time of the EC Fontan operation. However, your point is very well taken. A very precise surgical dissection is required here.

I think Ed Petrossian is in the audience here, but I do not think in the past 5 years we have had any diaphragm palsies in the series.

Dr Pedro J. del Nido (*Boston, Mass*). This is an excellent series, and I would echo what Dr Jacobs has said, that in other series using other types of cavopulmonary connections, other centers, including ours, have reported 10-year follow-up with similarly low mortality and supraventricular tachycardia rates.

My question relates to the issue of the oxygenator. You have identified this as a risk factor, but is that a fair comparison given the fact that your study is not contemporaneous and that it was a univariate analysis?

How many of the patients who were on CPB also had aortic crossclamping and, therefore, ischemic time, and was this a factor? In other words, if you had taken into account these other variables, would this risk factor of an oxygenator simply fall out?

Dr Hanley. That is a very good question. This was not a randomized prospective study of oxygenator versus no oxygenator. There has been an evolution of the technique, and I do not think we can tease the factor that you are referring to out of the data. Our standard policy right from the beginning was warm normothermic beating bypass with no crossclamp. Even the typical full bypass runs early in the series were performed with no aortic crossclamp except for a very small number of patients.

In general, I agree with your mortality comment, but I have not seen numbers in the literature from series performed predominantly using techniques other than the EC that show supraventricular tachycardia rates, and certainly not sinus node dysfunction rates, as low as we have seen.