Extracardiac conduit Fontan procedure: early and intermediate results

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Abstract

Objective: The extracardiac Fontan procedure, as compared with classic atriopulmonary connections, may have the potential for optimizing ventricular and pulmonary vascular function by maximizing the laminar flow principle, by the avoidance of intra-atrial suture lines and cardiac manipulation, and by minimizing cardiopulmonary bypass time. In this study the clinical results of this procedure are assessed.

Methods: From January 1990 until January 1997, 45 patients (33 males and 12 females) with a median age of 4.0 years (range 2.7–38 years) underwent an extracardiac Fontan procedure for univentricular physiology. The underlying diagnoses included tricuspid atresia (n = 19), double-inlet left ventricle (n = 11), and complex anomalies (n = 15). Forty patients (89%) were in sinus rhythm. The median ventricular ejection fraction was 60%. In 37 patients (82%) the procedure was staged.

Results: Median cardiopulmonary bypass time was 72 min, with a decrease to a median time of 24 min in the last ten patients. Aortic cross-clamping was avoided in 33 patients (73%). The intraoperative Fontan pressure and transpulmonary gradient were low: 13.6 ± 3.2 and 8.5 ± 3.9 mmHg, respectively. Transient supraventricular tachyarhythmias were observed in six patients (13%).

Conclusions: In the majority of patients, the extracardiac Fontan procedure, when performed as a staged procedure, provides excellent early and midterm results in terms of quality of life, maintenance of sinus rhythm, and preservation of ventricular function.

Keywords: Univentricular heart; Fontan operation; Bidirectional cavopulmonary anastomosis

1. Introduction

Since the original description of the Fontan operation for univentricular physiology [1], numerous technical modifications, notably the lateral atrial tunnel total cavopulmonary connection, have led to a continuous improvement in clinical outcome [2–6]. However, because of concern about the potential for systemic venous hypertension and long-term arrhythmogenic effects, since almost a decade, we have strived for an extracardiac conduit approach with the objective to reduce supraventricular arrhythmias and improve flow dynamics in the systemic venous pathway. The extracardiac conduit Fontan procedure maximizes laminar blood flow, reduces atrial wall tension, and avoids intra-atrial suture lines [7,8]. In this report the early to intermediate clinical results of this procedure are presented.

2. Patients and methods

From January 1990 until January 1997, 45 patients (33 males and 12 females) with a median age of 4.0 years (range 2.7–38 years) underwent an extracardiac Fontan procedure for univentricular physiology. The three youngest patients (2.7, 2.9 and 3.0 years) had heterotaxy (polysplenia) syndrome with azygos continuation. The median body weight was 18.8 kg (range 14–78 kg). Three patients, two with asplenia syndrome and one with hypoplastic left heart syndrome, died before the extracardiac Fontan operation could be undertaken; the causes of death were heart failure (n = 2) and sepsis (n = 1). An exclusion criterion for the present study was conversion of a previous atriopulmonary or lateral atrial tunnel Fontan connection to an extracardiac conduit Fontan procedure. The underlying diagnoses...
included tricuspid atresia ($n = 19$), double-inlet left ventricle ($n = 11$), and complex anomalies ($n = 15$) (Table 1). Forty patients (89%) were in sinus rhythm. Five patients (none of whom were staged) with complete heart block ($n = 3$) and with chronic atrial flutter ($n = 2$) had a permanent pacemaker. The median ventricular ejection fraction was 60% (range 40–65%). All but three patients had undergone prior operations, which are summarized in Table 2. In 37 patients (82%) the procedure was staged; the median age at construction of the bidirectional cavopulmonary anastomosis was 6 months (range 7 weeks–11 years). None of the patients with a bidirectional cavopulmonary anastomosis had an additional source of pulmonary blood flow in the form of a systemic-pulmonary shunt or a patent right ventricular outflow tract. All patients were subjected to preoperative echocardiography and catheterization, the results of which are summarized in Table 3. Four patients (all with heterotaxy syndrome and a common atrioventricular valve) had moderate ($n = 2$) or severe ($n = 2$) regurgitation of the atrioventricular valve. Three patients had subvalvular systemic outflow obstruction. Aortopulmonary collateral vessels were present in six patients; in four patients these were successfully obliterated by coil embolization. Postoperatively all patients were examined for the presence of protein-losing enteropathy, which was defined as increased enteric loss of $\alpha_1$-antitrypsin (normal value $< 27$ ml/24 h) or the occurrence of clinical symptoms (persistent or intermittent edema with hypoproteinemia) without evidence for deficient protein production or excessive protein loss from organ systems other than the gastrointestinal tract).

### Table 1: Diagnoses of patients undergoing extracardiac Fontan procedure

<table>
<thead>
<tr>
<th>Primary diagnosis</th>
<th>No. of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tricuspid atresia</td>
<td>19</td>
</tr>
<tr>
<td>Double-inlet left ventricle</td>
<td>11</td>
</tr>
<tr>
<td>Heterotaxy syndrome</td>
<td>6</td>
</tr>
<tr>
<td>Pulmonary atresia with intact ventricular septum</td>
<td>5</td>
</tr>
<tr>
<td>Hypoplastic left heart syndrome</td>
<td>3</td>
</tr>
<tr>
<td>Double-outlet right ventricle</td>
<td>1</td>
</tr>
</tbody>
</table>

### Table 2: Previous procedures

<table>
<thead>
<tr>
<th>Previous procedure</th>
<th>No. of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bidirectional superior cavopulmonary anastomosis</td>
<td>37</td>
</tr>
<tr>
<td>Systemic-pulmonary arterial shunt</td>
<td>26</td>
</tr>
<tr>
<td>Transection of pulmonary trunk</td>
<td>27</td>
</tr>
<tr>
<td>Atrial septectomy</td>
<td>18</td>
</tr>
<tr>
<td>Pulmonary artery banding</td>
<td>12</td>
</tr>
<tr>
<td>Relief of subaortic obstruction</td>
<td>5</td>
</tr>
<tr>
<td>Norwood operation</td>
<td>3</td>
</tr>
<tr>
<td>Repair of atrioventricular valve</td>
<td>2</td>
</tr>
</tbody>
</table>

2.1. Operative technique

The operative technique has been reported previously [8]. The extracardiac conduit Fontan circulation is completed by connecting the inferior vena cava to the pulmonary arteries with a polytetrafluoroethylene conduit (Gore-Tex, W.L. Gore & Assoc., Elkton, MD). We prefer to perform the extracardiac Fontan procedure when the patient is at least 3 years old or has a body weight of at least 18 kg. The size of the conduit is related to the body surface area of the patient and preferably should be at least 20 mm. Especially in our recent experience an effort was made to reduce cardio-pulmonary bypass (CPB) time. In case the pulmonary arteries were of adequate size and patch augmentation was not necessary, the pulmonary anastomosis was performed without CPB by partially clamping the pulmonary artery so that the bidirectional cavopulmonary anastomosis perfused one or both lungs. When this was possible, the duration of CPB was limited to the time required to perform the inferior cavopulmonary anastomosis only. Whenever possible, CPB was established with inferior vena cava cannulation only; in older patients the femoral vein was cannulated with the objective to improve exposure in the crucial area of the inferior vena cava. In case double venous cannulation was necessary, the left brachiocephalic vein was preferably cannulated to avoid distortion of the superior vena cava. In an effort to preserve ventricular function aortic cross-clamping was avoided unless a concomitant intracardiac procedure had to be performed.

After weaning from CPB, the Fontan pressure (i.e. the pressure in the extracardiac conduit) and the transpulmonary gradient were measured. If the Fontan pressure was more than 17 mmHg with a transpulmonary gradient of 10 mmHg or more, without use of CPB, a fenestration was made between the extracardiac conduit and the right atrial free wall. This was accomplished either with a direct side-to-side anastomosis (Fig. 1) or with a 4–6-mm Gore-Tex tube graft with an adjustable snare device (Fig. 2).

2.2. Data analysis

Data were retrospectively collected from patient records. The median follow-up time was 64 months (range 26–105 months). Continuous variables before and after the Fontan operation or between two groups of patients were compared with the use of the paired or independent samples $t$-test, respectively. Dichotomous variables were compared by
means of Fisher's exact test or $\chi^2$ analysis. The following outcome measures were assessed: duration of mechanical ventilatory support, occurrence of arrhythmias, duration of stay in the intensive care unit, and duration of chest tube drainage.

Independent variables analyzed included the following: age, date of operation, ventricular morphology, staged versus non-staged Fontan procedure, duration of CPB, use of cardioplegic arrest, Fontan fenestration, and additional operative procedure at the time of Fontan completion.

3. Results

3.1. Perioperative data

In all 45 patients a non-ringed Gore-Tex conduit was used to connect the inferior vena cava with the pulmonary arterial system. The median conduit size was 20 mm (range 18–25 mm). Patch augmentation of the pulmonary artery was performed in 18 patients (40%); in 12 patients the Gore-Tex conduit was fashioned in such a way that it acted as an onlay patch and in six patients a separate pericardial patch was used. In eight older patients without a prior bidirectional superior cavopulmonary anastomosis and some form of previous palliative procedure ($n = 5$), the construction of the extracardiac conduit was combined with a bidirectional cavopulmonary anastomosis. In the earlier part of the series, intracardiac procedures were performed in six patients for repair of atrioventricular valve regurgitation ($n = 3$), resection of subaortic stenosis ($n = 2$), and enlargement of a bulboventricular foramen ($n = 1$).

Median CPB time was 72 min (range 56–210 min) and decreased significantly over the course of our experience with a decrease to a median CPB time of 24 min in the last ten patients ($P \leq 0.001$). In 14 patients (31%) cannulation of the left brachiocephalic vein or superior vena cava could be avoided. Aortic cross-clamping with cardioplegic arrest (median 45 min) was performed in 12 patients, all of whom were operated on before 1996.

The Fontan pressure and transpulmonary gradient were low: $13.6 \pm 3.2$ and $8.5 \pm 3.9$ mmHg, respectively. A fenestration was constructed in 18 patients (40%), with a marked decrease in the incidence of fenestration over time, with only five of the last 25 patients (20%) undergoing a fenestration compared with 13 in the first 20 patients (65%) ($P = 0.03$). Early fenestration closure in the intensive care unit was performed in five patients.

There was no early mortality. Early postoperative outcome variables are summarized in Table 4. Median duration of chest tube drainage was 5 days (range 3–28 days). Prolonged chest tube drainage (>14 days) occurred in six
patients, all of whom were non-staged ($P \approx 0.04$). Prolonged mechanical ventilatory support ($\geq 24$ h) was associated with longer duration of CPB ($112 \pm 57$ vs. $83 \pm 37$ min, $P = 0.005$) and a non-staged Fontan procedure ($P = 0.05$). Factors associated with prolonged postoperative stay in the intensive care unit ($> 7$ days) included higher preoperative pulmonary artery pressure ($16.3 \pm 7.2$ vs. $10.8 \pm 6.8$ mmHg, $P = 0.01$), surgery with aortic cross-clamping ($P = 0.03$), and higher postoperative Fontan pressure ($15.9 \pm 2.5$ vs. $12.8 \pm 3.0$ mmHg, $P = 0.01$).

All 40 patients who were in sinus rhythm before the operation remained in sinus rhythm. During the first 48 postoperative hours, transient supraventricular tachyarrhythmias were observed in six patients (13%) and in all cases consisted of junctional ectopic tachycardia that in all cases lasted less than 20 min. None of the patients were discharged on antiarrhythmia medication. Holter monitoring in the early postoperative stage was performed in 24 patients, and demonstrated accelerated junctional rhythm in six patients and nonsustained ventricular arrhythmia in one patient. In none of the patients did the arrhythmias produce hemodynamic instability. The patients with accelerated junctional rhythm were treated with digitalization.

Other transient early postoperative complications included renal failure in five patients and elevated liver enzyme levels in four. Two patients underwent plication of the diaphragm for diaphragmatic paralysis.

### 3.2. Late follow-up

There was no late mortality during a median follow-up of 64 months (range 26–105 months). Thirty nine patients (87%) were in NYHA class I, four (9%) were in NYHA class II, and two (4%) were in class III. All of the patients who were in NYHA classes II and III had undergone a non-staged Fontan procedure ($P = 0.02$) (Table 5). In addition to functional class, maintenance of sinus rhythm ($P = 0.04$), and preservation of ventricular function ($P = 0.05$) were superior in patients who were appropriately staged. Forty patients (89%), 37 of whom were staged, remained in sinus rhythm. Holter monitoring in 15 patients demonstrated the absence of cardiac arrhythmias. The median ventricular ejection fraction was 59%. The arterial oxygen saturation increased from a preoperative median level of 82% (range 75–91%) to 97% (range 87–99%). Patients with lower saturations were those in whom the fenestration remained open. Only two patients underwent elective fenestration closure at 6 and 11 months after the Fontan operation. In nine patients, echocardiography documented spontaneous closure of the fenestration. Echocardiographically, none of the patients had evidence of atrial thrombus or thrombus in the extracardiac conduit. Echocardiographically, none of the patients had evidence of atrial thrombus or thrombus in the extracardiac conduit. None of the patients developed chronic pleural effusions or protein-losing enteropathy. We documented regression of pulmonary arteriovenous fistulae in three patients with heterotaxy (polysplenia) syndrome who had pulmonary arteriovenous fistulae after a previous bidirectional cavopulmonary anastomosis (Kawashima operation). Anticoagulation treatment consisted of acetylsalicylic acid in 38 patients (all with sinus rhythm) and coumadin in the remaining seven patients without sinus rhythm or suboptimal hemodynamics.
4. Discussion

The key to a successful short- and long-term outcome in Fontan patients is appropriate and timely surgical intervention in the systemic venous conduit combined with a tendency toward minimization of the number and duration of palliative procedures. Management of these patients must begin in the newborn period and should be directed toward reducing the likelihood of development of risk factors for a Fontan procedure. It is emphasized that the good results in the present series are mainly secondary to appropriate staging in the majority of the patients. This policy involves the use of small systemic-pulmonary shunts to provide limited pulmonary blood flow, construction of an early bidirectional cavopulmonary anastomosis, early identification and correction of systemic ventricular outflow tract obstruction, and avoidance of prolonged periods of pulmonary artery banding. If there is no pulmonary vascular obstructive disease, atrioventricular valve regurgitation, or subaortic stenosis (which in a historic sense are mainly passively or actively iatrogenic in nature), and ventricular function remains well preserved, the outcome of the Fontan operation is primarily related to the absence of obstruction in the systemic venous conduit and absence of atrial arrhythmias. For palliation of ductus-dependent univentricular physiology, it is generally our policy to construct a 3.5-mm modified Blalock–Taussig shunt. In non-ductus-dependent univentricular physiology, we attempt to manage the neonate medically until primary construction of a bidirectional cavopulmonary anastomosis becomes feasible; this is possible in the majority of cases because the physiologically elevated pulmonary vascular resistance prevents pulmonary overcirculation during the neonatal period. Subsequent construction of a bidirectional cavopulmonary anastomosis as early as 6 weeks of life avoids recirculation of both pulmonary and systemic venous return with resultant volume overload of the single ventricle and attendant sequelae [9]. Thus far we have observed adequate growth of the pulmonary arteries and the cavopulmonary anastomoses. Associated lesions, such as subaortic stenosis, atrioventricular valve regurgitation, or branch pulmonary artery stenosis, if present, are usually corrected at the time of construction of the bidirectional cavopulmonary anastomosis. The Fontan circulation is preferentially completed as an extracardiac conduit procedure when the patient is at least 3 years old or has a body weight of at least 18 kg. These criteria for Fontan completion are primarily based on physiologic and practical issues. Further delay of completion, secondary to a maturational decrease in apportionment of systemic blood flow to the upper versus the lower body segment, is often accompanied by exercise incapacity and excessive cyanosis with attendant sequelae [10]. A concomitant practical advantage of completing the Fontan operation using the aforementioned criteria is the fact that, empirically, at 3–4 years of age an adult-sized conduit (20–24 mm) can usually be inserted. An exception to these criteria may have to be made in patients with polysplenia syndrome andazygous continuation in whom there is an increased tendency toward the development of pulmonary arteriovenous fistulae after a Kawashima operation [11]. In these patients, connection of the hepatic veins to the pulmonary arterial system by means of an extracardiac or intracardiac conduit may have to be performed at an earlier age. Adhering to these principles we have been able to achieve excellent hemodynamic results with preservation of ventricular function and absence of major complications. In our experience the outcome in terms of functional class and preservation of sinus rhythm and ventricular function was superior in patients who had been appropriately staged and who had not been subjected to multiple and long-standing palliative procedures.

The potential advantages of the extracardiac conduit Fontan operation include technical ease, absence of manipulation in the atrium or in the vicinity of the sinus node, and satisfactory hemodynamic results [8,12–15]. Recent hemodynamic and computational modeling studies have convincingly confirmed the importance of avoiding energy dissipation by turbulence and stasis in the cavopulmonary connection [16,17]. As compared with classic atriopulmonary connections, the extracardiac Fontan procedure has the advantage of reduction of right atrial volume and hence avoidance of the effects of increased atrial wall tension and chronic elevation of coronary sinus pressure with its potential deleterious effects on ventricular function [18]. In addition, the circular geometry and uniform caliber of the valveless extracardiac conduit results in the optimization of blood flow into the pulmonary arterial circulation. Although a perfectly constructed lateral atrial tunnel total cavopulmonary connection, albeit a major improvement as compared with the atriopulmonary connection, may create a laminar flow pattern, at least in the early stage, we have concerns that in the long term dilatation or growth of the atrial wall segment that is incorporated in the tunnel (versus non-growth of the baffle) may lead to asymmetry and distortion of the systemic venous pathway with loss of laminar flow.

Atrial arrhythmias, particularly atrial flutter, are a major source of morbidity particularly during late follow-up after the Fontan operation, with reported rates of up to 40% [19,20]. A major advantage of the extracardiac conduit Fontan operation consists of the potential to reduce arrhythmias because it avoids all four of the factors thought to be important potential contributors to rhythm disturbances after the Fontan procedure: (1) exposure of the right atrium to the elevated systemic venous pressure; (2) extensive atrial incisions and suture lines; (3) surgical manipulation in the vicinity of the sinus node; and (4) ventricular dysfunction resulting from ischemic arrest and prolonged cardiopulmonary bypass. All 40 patients who were in sinus rhythm preoperatively remained in sinus rhythm. Transient supraventricular tachyarrhythmias occurred in 13% of our patients in the early postoperatively stage and responded
well to temporary pacing. None of the patients required a permanent pacemaker at the time of discharge. At a median follow-up of 64 months Holter monitoring in 15 patients demonstrated a normal sinus rhythm without any rhythm disturbances.

Early reduction of volume loading of the single ventricle preserves ventricular function and avoids dilatation of the atrioventricular valve(s). Severe regurgitation of a common atrioventricular valve has been reported to be a significant predictor of poor early outcome [6]. Volume overloading of the single ventricle and annular dilatation was avoided in the majority of our patients by early construction of a bidirectional cavopulmonary anastomosis. Three patients with heterotaxy syndrome and regurgitation of the common atrioventricular valve (who all had been exposed to a chronic volume overload by the construction of generous systemic-pulmonary shunts at other institutions) were successfully treated by a valvuloplasty procedure [21]. Patients with a single ventricle and transposed great arteries who are subjected to prolonged periods of pulmonary artery banding with resultant ventricular hypertrophy have been reported to have a significantly higher likelihood of late failure [6]. When pulmonary artery banding cannot be avoided, it is therefore our policy to reduce as much as possible the duration of the banding.

The extracardiac Fontan operation has potential disadvantages that are related to the use of a conduit, such as thromboembolism, conduit stenosis, and lack of growth. In classic atroipulmonary Fontan connections and, to a lesser extent, the lateral atrial tunnel total cavopulmonary connection, the turbulent flow in the noncylindrical (and often dilated) pathway, enhanced by the presence of atrial arrhythmias, may lead to the stasis of blood and thrombus formation [4,13,15]. Prosthetic material in human beings rarely becomes endothelialized beyond the first 5–10 mm. Although this may permit full or partial endothelialization of a baffle as used in the lateral atrial tunnel technique, an extracardiac conduit will most likely never become fully endothelialized. In the setting of a prothrombotic state, this may increase the risk of pulmonary emboli and, if a fenestration is constructed, systemic emboli. On the other hand, the lateral atrial tunnel technique places a large amount of prosthetic material in the pulmonary venous atrium with therefore an increased risk of systemic emboli. In view of the reported risk of postoperative thromboembolism of up to 20%, mainly in intra-atrial Fontan connections [22–24], all of our patients were discharged from the hospital on a regimen of acetylsalicylic acid. Seven patients who are not in sinus rhythm or in a suboptimal hemodynamic state are kept on chronic coumadin therapy. Thus far, no patient has had echocardiographic evidence of thrombus formation in the conduit. Anticoagulation treatment may be avoided in the rare patient with good hemodynamics in whom only autologous tissue is used for the extracardiac Fontan conduit [25].

By delay of completion of the Fontan procedure until the patient weighs approximately 20 kg, we expect to avoid reoperation by using an adult-sized conduit. Thus far, this has accommodated the patient’s growth and exercise demands. Right pulmonary vein obstruction by conduit compression has not been observed thus far. Amodeo and colleagues [14] evaluated extracardiac conduit patency by serial magnetic resonance imaging studies and reported a reduction in conduit internal diameter of 18% during the first 6 postoperative months, with no progression during the next 5 years. However, they used Dacron conduits in their initial experience, which may account for an increased tendency toward peel formation as opposed to Gore-Tex grafts.

In conclusion, in the majority of patients, the extracardiac Fontan procedure, performed as a staged procedure, provides excellent early and midterm results in terms of quality of life, maintenance of sinus rhythm, and preservation of ventricular function.

References


