Early and medium-term results after modified Fontan operation in adults


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Received 28 August 2002; received in revised form 28 November 2002; accepted 9 December 2002

Abstract

Objective: Single ventricle palliation is rarely performed in adults and the results are less optimal than in children. In this article we analyze our experience with the modified Fontan operation in this age group.

Methods: Data of 15 consecutive patients with single ventricle with a mean age of 26 (range 16–38) years, who underwent Fontan operation between 3/92 and 1/2000 were retrospectively analyzed. Five patients had previously had an aortopulmonary shunt in childhood and two patients had previously received a bi-directional cavopulmonary shunt as adults. Eleven patients were preoperatively in NYHA class III and four in class II. The main factors for the selection of the patients before surgery were well-developed pulmonary arteries with lower lobe index $120 - 30 \text{mm}^2/\text{m}^2$, pulmonary artery pressure $< 18 \text{mmHg}$, good cardiac function and enddiastolic systemic ventricular pressure $12 \text{mmHg}$. The lateral tunnel Fontan operation (LTFO) was performed in ten patients and extracardiac Fontan operation (ECFO) in five. A fenestration 4–5 mm in size was constructed in all patients with LTFO and in three of five patients with ECFO.

Results: There was one intraoperative and one late death (total mortality 13%). The mean extubation time and hospital stay were 24 h and 21 days, respectively. Severe postoperative complications were observed in three patients (20%). Two LTFO patients out of a total of eight patients (53%) with perioperative arrhythmias received a permanent pacemaker due to bradyarrhythmia. During the median follow-up of 5.0 (range 2.3–10.1) years, four patients developed arrhythmias; one of them had new onset bradyarrhythmia after LTFO and required permanent pacemaker implantation. The median postoperative oxygen saturation was 93% (range 90–98%). NYHA class improved significantly in 12 survivors. Cardiac catheterization (0.5–4 years postoperatively, $n = 12$) showed excellent Fontan hemodynamics in all patients.

Conclusions: The modified Fontan operation can be performed in adults with acceptable early and midterm mortality and morbidity and leads to either complete or marked relief of cyanosis and enhanced exercise tolerance in all survivors. Postoperative arrhythmias are one of the main drawbacks but the incidence of arrhythmias after ECFO seems to be lower. The long-term follow-up has yet to be established.

Keywords: Fontan operation in adult; Extracardiac; Lateral tunnel

1. Introduction

In patients with single ventricle, the Fontan operation relieves cyanosis and ventricular volume overload. It improves exercise tolerance and preserves ventricular function. The ideal time for the operation is in early childhood to avoid long-term volume overload and cyanosis, which may lead to valve regurgitation and myocardial fibrosis. In adult patients the Fontan operation bears these special risk factors for an adverse outcome [1–3]. To establish whether late surgical Fontan palliation in adults is beneficial, we analyzed our results of the last 8 years.

2. Patients and methods

Between March 1992 and January 2000, 15 consecutive adults (23%) of the total of 64 patients with single ventricle physiology underwent a Fontan operation at our institution. Their mean age was 26 years (range 16–38 years) and mean body weight 60 kg (range 42–90 kg). Eight patients were older than 25 years. Previous surgical procedures during childhood were establishment of an aortopulmonary shunt in five patients and of pulmonary artery banding in two. Two patients had received a bi-directional cavopulmonary shunt prior to the definitive Fontan operation. Preoperatively four patients were in NYHA class II and 11 in class III (Fig. 1). Preoperative arterial oxygen saturation was 82% (range 64–85%, Fig. 2). The anatomical diagnoses are presented in Table 1. All patients were seen for the first time in our outpatient department as adults.
2.1. Selection criteria

Preoperatively, all patients underwent echocardiography (ECG) and heart catheterization and pulmonary, venous and arterial angiography was performed to exclude significant venous and arterial collateral vessels. Patients were considered to be candidates for the Fontan procedure if the mean pulmonary artery pressure did not exceed 18 mmHg and the enddiastolic systemic ventricular pressure (SVEDP) was less than 12 mmHg. None of the patients had more than mild atrioventricular or aortic valve regurgitation. The size of the pulmonary arteries was estimated using the main pulmonary artery index (Nakata index \( \text{[4]} \)) and the lower lobe index \( \text{[5–8]} \). Although three patients with local stenosis of the central pulmonary arteries had a Nakata index of less than 330 \( \text{mm}^2/\text{m}^2 \) (lowest 180 \( \text{mm}^2/\text{m}^2 \)), all patients had a lower lobe index of 120 \( \text{mm}^2/\text{m}^2 \) (normal range, \( \text{[6]} \)) or more. The preoperative data are summarized in Table 2.

Preoperatively all patients had normal sinus rhythm.

Two other adult patients underwent successfully conversion of atrio-pulmonary connection to ECFO, but were not included in this report.

2.2. Surgical techniques

All operations were performed through median sternotomy on cardiopulmonary bypass. In ten patients a lateral tunnel Fontan operation (LTFO) and in five patients an extracardiac Fontan operation (ECFO) was performed. In the group with LTFO, moderate hypothermic cardiopulmonary bypass (CPB) and cardioplegia were used. The intraatrial tunnel was created by a polytetrafluoroethylene (PTFE, Gore-Tex \( ^{\text{\textregistered}} \) ) patch. In four of ten patients, an incision of the right atrial roof was made to enlarge the atrio-pulmonary anastomosis. A fenestration of 4–5 mm diameter was punched into the intraatrial patch in all patients.

In the group with ECFO, only one patient was operated on with mild hypothermic CPB and cardioplegic aortic cross-clamp of 7 min. The remaining four patients were operated on under normothermic perfusion and with beating heart. The inferior vena cava-pulmonary artery anastomosis was created with a PTFE vascular graft (diameter 24 mm) between the inferior caval vein and the right pulmonary artery. In three patients a fenestration was created between the conduit and right atrium with 3–5 mm PTFE tube graft.

All previous aortopulmonary shunts were closed. Additional surgical procedures included closure of the pulmonary trunk in 14 patients, enlargement of a stenotic pulmonary artery in five patients and atrial septectomy in four patients. One patient received a left bidirectional cavo-pulmonary anastomosis. Intraoperative transoesophageal echocardiography was performed in all patients.

<table>
<thead>
<tr>
<th>Table 1</th>
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<tbody>
<tr>
<td>Cardiac anatomical diagnosis</td>
</tr>
<tr>
<td>Diagnosis</td>
</tr>
<tr>
<td>Double inlet left ventricle</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
</tr>
<tr>
<td>Double outlet right ventricle</td>
</tr>
<tr>
<td>Mitral atresia</td>
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<tr>
<td>Total</td>
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<th>Table 2</th>
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<tr>
<td>Preoperative patients data ( ^{a} )</td>
</tr>
<tr>
<td>Data</td>
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</tr>
<tr>
<td>Age (years)</td>
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<tr>
<td>Weight (kg)</td>
</tr>
<tr>
<td>Arterial oxygen saturation (%)</td>
</tr>
<tr>
<td>Hemoglobin level (mg/dl)</td>
</tr>
<tr>
<td>EDP (mmHg)</td>
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<tr>
<td>MPAP (mmHg)</td>
</tr>
<tr>
<td>PAI (mm(^2/\text{m}^2))</td>
</tr>
<tr>
<td>LLI (mm(^2/\text{m}^2))</td>
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\( ^{a} \) EDP, systemic ventricular enddiastolic pressure; MPAP, mean pulmonary artery pressure; PAI, pulmonary artery (Nakata) index; and LLI, lower lobe artery index.
Table 3
Early postoperative complications (n = 6)

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>Diagnosis</th>
<th>Op. method</th>
<th>Respiratory support (days)</th>
<th>Requirement of prolonged inotropic support</th>
<th>Pulmonary vasodilation (PAP &gt;20 mmHg)</th>
<th>Requirement of dialysis</th>
<th>Prolonged pleural effusions (&gt;10 days)</th>
<th>Dysrhythmias</th>
</tr>
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<tbody>
<tr>
<td>18</td>
<td>DILV, TGA</td>
<td>LTFO</td>
<td>4</td>
<td>Yes</td>
<td>Nitric oxide</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>18</td>
<td>MA, TGA</td>
<td>LTFO</td>
<td>4^</td>
<td>No</td>
<td>Prostacyclin</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>29</td>
<td>DORV, TGA</td>
<td>LTFO</td>
<td>5</td>
<td>No</td>
<td>Nitric oxide</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>27</td>
<td>DORV, TGA</td>
<td>ECFO</td>
<td>8</td>
<td>Yes</td>
<td>Nitric oxide</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>37</td>
<td>DORV, TGA</td>
<td>ECFO</td>
<td>24</td>
<td>Yes</td>
<td>Nitric oxide</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>37</td>
<td>TA</td>
<td>LTFO</td>
<td>1</td>
<td>No</td>
<td></td>
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</table>

^a Open chest for 24 h.
^b Because of septic shock on the 12 p.o. day.
^c Renal failure in septic shock; requirement of continuous diuretics perfusion for 2 days.

2.3. Follow-up

The median follow-up period in the LTFO group and the ECFO group was 6.2 (4.2–10.1 years) and 3.5 years (2.3–6.1 years), respectively. All patients underwent echocardiographic examination at least twice a year. Twelve patients underwent catheterization 6–48 months (median 27 months) after the operation.

In ten patients, exercise capacity was tested by spiroergometry 11–84 months (median 36 months) after surgery. Six of them were tested twice, the second time being 24–84 months (median 80 months) postoperatively.

Perioperatively, all patients had at least one pre- and several postoperative 12-lead standard ECG recordings. During the immediate postoperative period continuous ECG monitoring with the possibility of on- and offline analysis as well as optional printout was utilized for the detection and documentation of rhythm disturbances. After the hospital stay, ECGs were registered three times a year. A Holter ECG was performed at least once a year. Atrial flutter, atrial fibrillation or junctional ectopic tachycardia were defined as supraventricular tachyarrhythmias (SVT). Sinus bradycardia and slow junctional rhythm were defined as bradyarrhythmias.

2.4. Statistical analysis

To compare the different surgical techniques (intraatrial tunnel and extracardiac conduit) with regard to the length of ventilatory support and hospital stay, postoperative oxygen saturation and aerobic capacity, the Mann–Whitney test was used. To detect differences between the two techniques with regard to the incidence of arrhythmias, the chi-square test was employed.

Four patients developed early postoperative SVT (Table 5).

3. Results

Median duration of cardiopulmonary bypass was 157 min (range: 107–474 min) for the LTFO group and 131 min (range 78–210 min) for the ECFO group (\(P = \text{n.s.}\)). Median aortic cross-clamp time for LTFO was 63 min (range 45–79 min) versus only 7 min in one patient for ECFO (\(P < 0.001\)).

3.1. Early results

There was one (6.25%) intraoperative death due to severe tracheal bleeding in a 38-year-old patient with LTFO. Despite intraoperative bronchoscopy the source of bleeding was not found. No patients died during early postoperative hospital stay.

Six out of 14 patients developed complications (Table 3). They needed mechanical ventilation for a mean of 93 h (range 14–576 h) and had a mean hospital stay of 27 days (range 20–44 days).

In eight out of 14 patients the postoperative course was uneventful. They were extubated after a mean of 11 h (range 6–48 h) after operation and were discharged after 13 days (range 7–20 days). Ten of 11 patients (91%) with preoperative SVEDP lower than 10 mmHg were extubated within 72 h, compared to one of three patients (33%) with preoperative SVEDP >10 mmHg (\(P = 0.04, r = 0.650\)).

There was no statistically significant difference in the incidence of postoperative complications or in the duration of mechanical ventilation or hospital stay between the two different surgical groups. However, the median arterial oxygen saturation on discharge was higher after ECFO (see Table 4).

The mean pulmonary artery pressure and the left atrial pressure on the first postoperative day were 14 (range 10–16 mmHg) and 7 mmHg (range 4–11 mmHg), respectively. The median oxygen saturation on discharge was 93% (range 87–98%) in patients with fenestration and 98–99% in three patients without fenestration.
Three of them were discharged in sinus rhythm under antiarrhythmic medication with either amiodaron, sotalol or isoptin. Four patients, all with LTFO, developed bradyarrhythmia. Two of them required the implantation of a permanent pacemaker.

In summary, four patients with LTFO (44%) but no patient with ECFO lost sinus rhythm after surgery; however, this difference did not reach statistical significance ($P = 0.09$).

### 3.2. Midterm mortality

One patient with double inlet left ventricle died suddenly 5 years after LTFO, probably due to ventricular arrhythmia. The last control catheterization in this patient 3 years after surgery showed favorable Fontan hemodynamics with good ventricular function, left atrial pressure of 6 mmHg and mean pulmonary artery pressure of 8 mmHg.

### 3.3. Midterm follow-up

During a median follow-up period of 5.0 years (range 2.3–10.1 years) no patient required reoperation and none has so far developed protein-losing enteropathy.

Cardiopulmonary exercise-testing by spiroergometry showed an exercise capacity of $1.6 \pm 0.49$ w/kg (54.2 ± 16.1% of the norm for this age group) and an oxygen consumption capacity of $23.6 \pm 6.3$ ml/min per kg (58.4 ± 13.5% of the norm for this age group). Furthermore, patients, who were tested twice showed decreased exercise and oxygen consumption capacity of $1.4 + 0.28$ w/kg (48.8 + 8.8%) and $19.9 + 4.6$ ml/kg per min (48.5 + 7.7%), respectively, compared to earlier postoperative testing ($P = 0.028$). However, NYHA class improved significantly in 12 patients (86%). Postoperatively four patients were in NYHA class I and eight in NYHA class II; two patients remained in class III at the end of the follow up (Fig. 1, $P = 0.001$).

Echocardiography documented stable ventricular dimensions and performance, and no significant increase in valve incompetence, in all patients.

In 12 patients, including one who died, the diagnostic catheterization 0.5–4 years after the Fontan operation showed mean pulmonary artery pressure of 11 mmHg (range 8–14 mmHg). No patient had systemic venous obstruction or pulmonary artery stenosis. In one patient, who was reported previously [5,9] with vena azygos continuation, a parietal thrombosis of the extracardiac conduit was observed 1.5 years postoperatively without hemodynamic relevance. No further extracardiac conduit or lateral tunnel thrombosis and no symptomatic thromboembolic events were observed during the follow up in other patients.

One patient with double outlet right ventricle and transposition of the great vessels developed mild subaortic outflow tract obstruction of 40 mmHg 9 months after surgery. Antiarrhythmic treatment with digoxin and amiodaron was replaced by atenolol, which resulted in a reduction of the gradient to 15 mmHg. Thus, no surgical intervention was necessary.

The fenestrations were closed by intervention in five patients, were insignificant in five, and had closed spontaneously in three patients. At the control cardiac catheterization and after closure of fenestrations the median arterial oxygen saturation was 95% (range 91–97%) (Fig. 2).

Four patients developed arrhythmias 2–4.5 years after operation. They required antiarrhythmic medication ($n = 4$), repeated cardioversion ($n = 3$), or a permanent pacemaker.
(n = 1). All these four patients have an intratrial tunnel, but no patient with an extracardiac conduit has developed new onset arrhythmias during the short follow-up time of 2 years after the last extracardiac procedure in the group (P = 0.04).

4. Discussion

The Fontan procedure can improve ventricular function and enhance aerobic capacity. In adult patients, however, long-standing cyanosis with myocardial fibrosis, chronic volume overload and valve incompetence have to be taken into account as risk factors for the procedure. Strict selection criteria should improve postoperative results; however, no generally accepted guidelines exist. In our series we considered enddiastolic systemic ventricular pressure (SVEDP) greater than 10 mmHg and mean pulmonary artery pressure (mPAP) higher than 15 mmHg as risk factors, as did Gates et al. [3] and others. However, we accepted patients with SVEDP of 12 mmHg or mPAP of 17 mmHg for the Fontan operation if the ventricular function was adequate. We waived the measurement of the ventricular ejection function because of the absence of standardized data for single ventricle physiology and the great variety in cardiac anatomy in the group, but we measured the contractility and kinetics of the ventricular wall using two-dimensional echocardiography and contrast angiography.

In the last 12 patients we replaced the Nakata index by the lower-lobe index, as a better predictor for a well-developed pulmonary vascular bed [6,8,10]. Two patients who otherwise would have been judged as poor candidates for the Fontan procedure because of significant pulmonary artery distortion were able to be included in this series. In these patients the central pulmonary arteries were enlarged with patches during the Fontan operation, resulting in obstruction-free flow because of normal lower lobe artery index (LLI).

It is noteworthy that the technique with extracardiac conduit seems to be superior to the intratrial lateral tunnel with respect to postoperative complications and hospital stay, although this did not reach statistical significance in this series, compared to our data on a group of children, because the numbers are small.

In the absence of preoperative hemodynamic risk factors such as high pulmonary artery pressure or high SVEDP, a routinely performed fenestration is not necessary and should be avoided, because it is a possible source of cyanosis, thrombembolic events [11–14] or, in rare cases, complications during transcatheter closure [15]. Thus, in our opinion it should be reserved for patients with suboptimal hemodynamics, such as slightly elevated pulmonary artery pressure and/or pulmonary vascular resistance or decreased lower-lobe pulmonary artery index [16].

Arrhythmias are a common complication after the Fontan procedure [2,12,17,18]. In our series 28% of the patients lost sinus rhythm and 26% developed arrhythmias during the follow-up period. However, these patients with univentricular heart frequently develop sinus node dysfunction in adult life whether they remain without surgery or are operated on palliatively with aortopulmonary shunt or undergo definitive cavopulmonary connection [19–21]. On the other hand, despite the relatively small number of adult patients in this series, there is a significant difference in postoperative arrhythmias between the patients with extracardiac conduit and with intratrial tunnel. We speculate that this difference may be even more significant as time goes by, due to the ongoing onset of arrhythmias in the LTFO group, in which extensive intratrial suture lines remain a permanent risk factor [18,22].

Adult patients have a high risk of postoperative arrhythmias, which with increased postoperative thrombogenicity may lead to pulmonary emboli and, if a fenestration is placed, to cerebrovascular events [12–14,21]. We prefer lifelong anticoagulation for all adult patients with phenprocoumon.

Iserin et al. have shown that patients with Fontan circulation do not necessarily have a better exercise tolerance compared with those without right heart bypass [23]. Thus, it is not surprising that the patients in this study showed reduced exercise tolerance compared to healthy people [24]. It is known that, at least in patients with original Fontan operation after initial improvement of exercise tolerance, continuous reduction of the physical capacity due to the progressive heart failure is to be expected, especially after late Fontan palliation, such as in some patients in this series [25]. However, in daily life most of the patients showed a performance of NYHA class II and a clear improvement in comparison to their preoperative status. Furthermore, relief from cyanosis and polycythemia and the reduction in stroke risk are clear advantages of the successful Fontan procedure.

5. Conclusion

The Fontan procedure in adults can be performed with low early and midterm mortality and morbidity. It leads to improved exercise tolerance and marked relief of cyanosis. Compared to the intratrial tunnel procedure, the technique using an extracardiac conduit with normothermic cardiopulmonary bypass and without cardioplegia seems to improve the early postoperative course and leads to a significant reduction in the development of arrhythmias both in the early postoperative period and in the midterm follow-up.

Acknowledgements

We are grateful Anne Gale for editorial advice.

References


