The Modified Fontan Procedure With Use of Extracardiac Conduit in Adults: Analysis of 32 Consecutive Patients

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Background. Uncommonly, adults with functionally univentricular hearts are becoming candidates for a Fontan procedure. The purpose of this study was to evaluate the course of patients undergoing the modified Fontan procedure with an extracardiac conduit in recent years.

Methods. Between January 2003 and December 2013, 32 adult patients (17 female and 15 male) underwent total cavopulmonary connection (TCPC) with extracardiac conduit. The median age at procedure was 24.5 years (interquartile range [IQR] 20 to 33 years). The diagnoses included double-inlet left ventricle (DILV) in 10 patients (31.2%), tricuspid atresia in 8 patients (25%), double-outlet right ventricle in 4 patients (12.5%), heterotaxia in 4 patients (12.5%), and mitral atresia in 2 patients (6.2%). Seventy-eight percent of patients had undergone at least one prior palliative procedure; the most common procedures were Blalock-Taussig shunt (16 patients), superior cavopulmonary shunt (12 patients), and pulmonary artery banding (6 patients). All patients underwent cardiac catheterization preoperatively. Aortic cross-clamping was necessary in 15 patients for intracardiac procedures. Fenestration was required in 9 patients (28%). Four concomitant intraoperative cryoablation procedures were performed.

Results. There was no hospital mortality. One patient (3.1%) died 6 months after undergoing TCPC. Morbidities included prolonged pleural effusion lasting more than 7 days in 20 patients (62.5%), atrial arrhythmias in 4 patients (12.5%), and permanent pacemakers in 3 patients (9.3%). The median follow-up time was 33 months (interquartile range [IQR], 10.5 to 50 months). Actuarial survival was 91.83% (95% confidence limits, 71.07 to 97.89) at 1 year and 5 years. Ninety-two percent of patients were in New York Heart Association class I or II at follow-up. The median postoperative oxygen saturation was 95% (IQR, 93% to 95.5%). Cardiac arrhythmia occurred in 4 patients. Systolic ventricular function improved during follow-up for all patients except 1 patient, who underwent cardiac transplantation 7 months after the TCPC.

Conclusions. The modified Fontan procedure with use of an extracardiac conduit can be performed in adults with encouraging early and midterm results. The majority of late survivors had improved quality of life. The incidence of late death, reoperation, arrhythmias, and thromboembolic events was low during follow-up.

Fontan and Baudet described the first successful Fontan procedure in 1971 [1]. Since then, this technique has become the procedure of choice for the palliation of functionally univentricular hearts (UVH) and has been performed predominantly in children. The extracardiac conduit was introduced by Marcelletti and colleagues [2] in 1990 and is now routinely performed around the world. Despite a significant number of studies assessing several thousands of patients, the ideal age for total cavopulmonary connection (TCPC) still remains under debate. Nowadays, early ventricular unloading by bidirectional superior cavopulmonary anastomosis (BSCPA) in the first year of life and TCPC with use of extracardiac polytetrafluoroethylene (PTFE) conduits between 2 and 6 years of age is commonly performed [3, 4]. Our institution has advocated an early TCPC strategy since 2001. Before that period, in our institution but also in general in our country, because of the influence of the concept of definitive palliation with BSCPA associated with additional pulmonary blood flow, it took a long time for several patients to be considered for Fontan completion [5, 6]. The present study assessed the indications for, and the results of TCPC with the use of extracardiac conduit in adults.
Patients and Methods

Between January 2003 and December 2013, 32 patients underwent TCPC in our institution with the use of an extracardiac PTFE conduit (Gore-Tex tube) between the inferior vena cava and the right pulmonary artery. Institutional review board approval for a retrospective clinical study was obtained. The hospital records, operative reports, cardiac catheterization data, and noninvasive studies on all patients were reviewed (Table 1). Late follow-up data were obtained from the patients’ cardiologists. Their ages ranged from 18 to 47 years. There were 17 female and 15 male patients. Eighty-four percent of patients had a single ventricle. All patients presented with progressive deterioration of functional capacity and with signs and symptoms often dominated by dyspnea and cyanosis.

Preoperative complex cardiac lesions are summarized in Table 2. Fifty-seven palliative operations (Table 3), including 12 BSCPA, had been performed before the TCPC. Six patients had a single-stage TCPC. All patients underwent elective preoperative heart catheterization to evaluate hemodynamic status and record pulmonary artery anatomy before TCPC. As we perform in all patients before Fontan completion, pulmonary vascular resistance was calculated as the mean pressure drop across the lung’s vascular bed divided by the flow passing through it [7]. We used the Fick principle of pulmonary blood flow that requires sampling of pulmonary arterial and pulmonary venous blood and measurement of metabolic gas exchange [8] that we perform by hood indirect calorimetry during the catheterization [9]. By use of the predictive equations, the estimated values differed from measured values by more than 20% in approximately one fifth of the cases. These data indicate that frequently oxygen consumption cannot be accurately estimated from predictive equations. Therefore, if blood flow is to be accurately determined by the Fick method in infants and children, oxygen consumption should be measured during the catheterization procedure [10].

| Table 1. Demographic, Echographic, and Hemodynamic Characteristics |
|----------------|----------------|---------------|
| Variable       | Median         | IQR           |
| Age, years     | 24.5           | (20–33)       |
| >30 years      | 10 (31.2%)     |               |
| Male sex       | 15 (46.8%)     |               |
| Weight, kg     | 57             | (49–68)       |
| Preoperative NYHA class | 1, II 18, III 8, IV 1 |
| Arterial oxygen saturation, % | 82 | (80–85) |
| Cardiac fractional shortening, % | 34 | (31–37) |
| Moderate or more atrioventricular valve regurgitation | 6 (18.7%) |
| Mean pulmonary artery pressure, mm Hg | 11 | (9–13) |
| Atrial pressure, mm Hg | 6 | (5–7) |
| Ventricular end-diastolic pressure, mm Hg | 8 | (7–10) |
| Pulmonary vascular resistance, Wood unit | 1.8 | (1.4–2.4) |

IQR = interquartile range; NYHA = New York Heart Association.

| Table 2. Preoperative Diagnosis |
| Cause of Single-Ventricle Physiology | n (%) |
| Single-ventricle morphology |         |
| Left | 28 (84.8) |
| Biventricular left dominant | 9 (28.2) |
| Right | 4 (12.5) |
| Double-inlet left ventricle | 10 (31.2) |
| Tricuspid atresia | 8 (25) |
| Double-outlet right ventricle | 4 (12.5) |
| Heterotaxy syndrome | 4 (12.5) |
| Mitral atresia | 2 (6.2) |
| Other | 4 (12.5) |

Surgical Technique

The procedures were performed with the patient under normothermic continuous cardiopulmonary bypass (CPB) and only in patients (n = 15) who required an intracardiac-associated procedure, aortic cross-clamping with the use of intermittent antegrade normothermic blood cardioplegia was used. In this series, only 1 patient with previous BSCPA and extreme cyanosis (oxygen saturation = 65% at rest) underwent off-pump TCPC. The management of hemorrhagic tendency caused by chronic cyanosis was mainly done, in addition to commercially available hemostatic agents (tranexamic acid, fibrinogen concentrate, PPSB [prothrombine II, proconvertine VII, Stuart factor X, antihemophylic factor B IX], by the extensive use of fresh-frozen plasma and also avoidance of hypothermia. The decision for fenestration (n = 9) was taken essentially intraoperatively in the presence of a surgical transpulmonary gradient of 12 mm Hg or more with use of an 8-mm PTFE conduit, as short as possible. The patient who underwent the off-pump procedure required double fenestration. No ultrafiltration was performed. Patients with preoperative documentation of atrial tachyarrhythmia (flutter) underwent a right-sided Maze procedure by means of cryoablation under aortic cross-clamping. All patients were treated by oral anticoagulant therapy (International Normalized Ratio between 2 and 3) before discharge.

| Table 3. Cardiac Procedures in 57 Patients Before TCPC |
| Procedure | n |
| Blalock-Taussig | 27 |
| Bidirectional Glenn | 12 |
| Pulmonary artery banding | 8 |
| Atrial septectomy (Blalock-Hanlon) | 3 |
| Pulmonary arterioplasty | 3 |
| Potts shunt | 2 |
| Central shunt | 1 |
| Kawashima repair | 1 |

TCPC = total cavopulmonary connection.

Some patients had more than one procedure.

Seven patients had no prior cardiac procedure.
Data Analysis

Early postoperative death was defined as that occurring before discharge or before 30 days after operation. Data were collected retrospectively and analyzed for all patients. Continuous variables were expressed as means and standard deviation, and nonparametric data were expressed as medians and interquartile range (IQR). Categoric variables were compared with the $\chi^2$ test, and continuous variables were compared with the Mann Whitney U test. Univariate and multivariate analysis of risk factors were performed with logistic regression, and $p$ values less than 0.05 were deemed statistically significant.

Results

Early Results

There was no hospital mortality. Three patients required pacemaker insertion at the same time as TCPC. One patient had a complete heart block before TCPC. Two other patients required a pacemaker after tricuspid annulus and cryoablation procedures. Three patients required atrioventricular valve repair because of severe regurgitation. The median PTFE conduit size was 20 mm (IQR, 16 to 24 mm). Associated procedures during TCPC and operative variables are shown in Table 4. The median times of ventilation and intensive care stay were 17.5 hours (IQR, 11 to 24 hours) and 3 days (IQR, 2 to 5 days) respectively. The postoperative variables and adverse events are listed in Table 5. Prolonged pleural effusions requiring extended chest tube drainage for more than 7 days developed in 20 patients (62.5%). Decreased ven- tricular function, CPB time, and older age influenced the risk for prolonged pleural effusion on multivariate analysis (Table 6).

Late Results

One late death occurred in a 20-year-old man who had previously undergone Kawashima repair and presented with left ventricular dysfunction and severe cyanosis resulting from pulmonary arteriovenous fistulas. His in-hospital course was uneventful; however, the patient died 6 months later because of massive hemoptyis attributed to coronary-bronchial fistulas. Because of the low number of deaths, no statistically proper analysis of risk factors could be performed. A second patient was 42 years old and had DILV, pulmonary stenosis, and a previous modified Blalock-Taussig shunt. He underwent TCPC with severely decreased ventricular function. The ventricular function was moderately improved at discharge with medical treatment; however, heart transplantation was performed in the seventh postoperative month for the same reason, with favorable subsequent late outcome (Table 7). Freedom from events, including death and reoperation, was 91.8% at 1 year (95% confidence limits, 81 to 100) (Fig 1).

Arrhythmias

Atrial arrhythmias were observed in 4 patients. Three patients had paroxysmal atrial tachycardia treated by antiarrhythmic medication. One patient had an episode of new-onset paroxysmal atrial fibrillation. This patient had heterotaxy syndrome and experienced moderate valvular regurgitation. The clinical course of arrhythmia is shown in Table 8.

Follow-Up

The median follow-up time was 33 months (range, 2 to 99 months). Late follow-up was obtained in all surviving patients.

Table 4. Associated Procedures During TCPC and Operative Variables

<table>
<thead>
<tr>
<th>Procedure</th>
<th>n</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intercavitary septectomy</td>
<td>7</td>
</tr>
<tr>
<td>Pulmonary arterioplasty</td>
<td>5</td>
</tr>
<tr>
<td>Arrhythmia procedure</td>
<td>4</td>
</tr>
<tr>
<td>AV valve repair</td>
<td>3</td>
</tr>
<tr>
<td>PM implantation</td>
<td>3</td>
</tr>
<tr>
<td>Off pump</td>
<td>1</td>
</tr>
<tr>
<td>CPB time, min</td>
<td>116 (IQR, 64–164)</td>
</tr>
<tr>
<td>Aortic cross-clamping time, min, 15/32 pts</td>
<td>33 (IQR, 22.5–77)</td>
</tr>
</tbody>
</table>

AV = atrioventricular; CPB = cardiopulmonary bypass; IQR = interquartile range; PM = pacemaker; pts = patients; TCPC = total cavopulmonary connection.

Table 5. Postoperative Variables and Adverse Events

<table>
<thead>
<tr>
<th>Variable</th>
<th>n</th>
</tr>
</thead>
<tbody>
<tr>
<td>Duration of mechanical ventilation, hours</td>
<td>17.5 (IQR, 11–24)</td>
</tr>
<tr>
<td>ICU length of stay, days</td>
<td>3 (IQR, 2–5)</td>
</tr>
<tr>
<td>Hospital length of stay, days</td>
<td>20.5 (IQR, 15–35.5)</td>
</tr>
<tr>
<td>Postoperative saturation, %</td>
<td>93 (75–98)</td>
</tr>
<tr>
<td>Pleural effusion, days</td>
<td>8 (IQR, 7–13)</td>
</tr>
<tr>
<td>Chest tube drainage &gt;7 days</td>
<td>20 (62.5%)</td>
</tr>
<tr>
<td>Renal failure with dialysis</td>
<td>1</td>
</tr>
<tr>
<td>Sternal wound infection</td>
<td>2</td>
</tr>
<tr>
<td>Reexploration for bleeding</td>
<td>1</td>
</tr>
<tr>
<td>Serious postoperative arrhythmias</td>
<td>1</td>
</tr>
<tr>
<td>Ascites</td>
<td>2</td>
</tr>
</tbody>
</table>

ICU = intensive care unit; IQR = interquartile range.

Table 6. Multivariate Analysis Related to Prolonged Pleural Effusion >7 days

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>Odds Ratio (95% CL)</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arterial oxygen saturation</td>
<td>0.95 (0.77–1.17)</td>
<td>0.66</td>
</tr>
<tr>
<td>Cardiac fractional shortening, %</td>
<td>1.64 (1.1–2.43)</td>
<td>0.013</td>
</tr>
<tr>
<td>Pulmonary vascular resistance, WU</td>
<td>5.06 (0.65–39.8)</td>
<td>0.12</td>
</tr>
<tr>
<td>Transpulmonary gradient</td>
<td>1.10 (0.5–2.04)</td>
<td>0.74</td>
</tr>
<tr>
<td>CPB time (min)</td>
<td>1.02 (1.003–1.05)</td>
<td>0.027</td>
</tr>
<tr>
<td>Age &gt;30 years</td>
<td>0.017 (0.0003–0.99)</td>
<td>0.049</td>
</tr>
<tr>
<td>Fenestration</td>
<td>0.08 (0.002–2.55)</td>
<td>0.15</td>
</tr>
</tbody>
</table>

CL = confidence limits; CPB = cardiopulmonary bypass; WU = Wood unit.
patients except 5 who were referred from overseas. Twenty-three surviving patients improved their New York Heart Association (NYHA) classification to I (56%) or II (36%). The median systemic oxygen saturation was 93% (IQR, 92% to 99.5%) at discharge and was 95% (IQR, 93% to 95.5%) at the last visit (p = 0.027). During follow-up, the quality of ventricular function improved slightly. The median cardiac fractional shortening was 34% (IQR, 30% to 35%) and 35% (IQR, 32.5% to 36%) before and after TCPC, respectively (p = 0.0063).

Comment

Adult survivors presenting with UVH without Fontan-type palliation are rare in countries where congenital cardiac operations are currently performed. In our series, the study group patients had reached adulthood without undergoing Fontan procedures for essentially three reasons: previous decisions about unsuitable hemodynamics or previous early Fontan failure; the “French” trend of the 1990s for avoiding, or at least delaying, Fontan completion; and finally the referral of 5 patients from overseas. The past decade saw a significant increase in adult patients with congenital heart disease undergoing the procedure. Notable progress has occurred regarding the perioperative treatment of those patients and surgical results [11]. In addition, improvements in the understanding and management of congenital heart disease justify the regular reevaluation of all kinds of patients with congenital heart disease. The indications for TCPC we applied in the past have improved in time. Some patients previously considered to be unsuitable for Fontan-type palliation have been reconsidered for the procedure despite their adult age. By contrast, we also observed during follow-up a modification of the hemodynamic status of UVH patients who had not been operated on or who had received partial palliation; in recent evaluations, their pulmonary pressures, transpulmonary pressure gradients, and pulmonary vascular resistances were significantly lower than in early investigations, and TCPC was safely performed despite their adult age. Recent changes in the management of UVH have included an attempt to reduce the adverse effects of prolonged volume load on ventricular function by performing a volume unloading operation at an earlier age [12, 13]. The advantages of early volume unloading currently correspond to the common practice of BSCPA in infancy. Although the optimal timing for TCPC still remains under debate, the recent follow-up studies more likely support the strategy for early TCPC [14]. In 1990s, in our country, the common attitude toward the management of UVH was conditioned by the trend of the “ticking clock theory,” limiting the Fontan failure essentially to the increase in central venous pressure, early completion resulting, by definition, in early failure. Beyond this strategy, as a valuable alternative to Fontan circulation, BSCPA associated with additional pulmonary blood flow was proposed and performed [5]. The study group included some of the survivors of this strategy until they were adults, when their clinical status deteriorated and they were still eligible for definitive palliation. Despite their individual clinical histories and anatomic variations, the study group was homogenous and consisted of cyanotic patients presenting with different degrees of limitation in exercise capacity, with often recent deterioration of the latter. In preoperative investigations, UVH function was found to be preserved in the majority. The indication for TCPC was essentially based on cardiac catheterization findings, the transpulmonary pressure, gradient and particularly the calculated pulmonary vascular resistance. Despite a median pulmonary artery pressure of 11 mm Hg (in 3 patients ≥15 mm Hg) and a median transpulmonary gradient of 6 mm Hg (in 2 patients ≥10 mm Hg), pulmonary vascular resistance above 3 Wood units was observed in 4 patients. This discrepancy can be explained by a particularly low pulmonary blood flow and an extremely low oxygen saturation of the pulmonary artery blood, with some degree of

### Table 7. Late Outcome Except for Arrhythmias

<table>
<thead>
<tr>
<th>Variable</th>
<th>n</th>
<th>Preoperative</th>
<th>Follow-Up</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Late mortality</td>
<td>1</td>
<td>...</td>
<td>6 months</td>
<td>...</td>
</tr>
<tr>
<td>Cardiac transplantation</td>
<td>1</td>
<td>...</td>
<td>7 months</td>
<td>...</td>
</tr>
<tr>
<td>Arterial oxygen saturation</td>
<td>25</td>
<td>83% (75% to 98%)</td>
<td>95% (82% to 98%)</td>
<td>0.027</td>
</tr>
<tr>
<td>Cardiac fractional shortening</td>
<td>25</td>
<td>34% (30% to 35%)</td>
<td>35% (32.5% to 36%)</td>
<td>0.006</td>
</tr>
<tr>
<td>NYHA class</td>
<td>25</td>
<td>I 5, II 18, III 8, IV 1</td>
<td>I 14, II 9, III 2, IV 0</td>
<td>...</td>
</tr>
</tbody>
</table>

NYHA = New York Heart Association.
vasoconstriction of the pulmonary vascular bed. Thus, the overall preoperative pressure profile was more favorable than we usually observe in children scheduled for TCPC.

In our series, only 1 patient underwent off-pump TCPC. Fifteen TCPC patients required aortic cross-clamping for associated cardiac procedures. For Petrossian and colleagues [15], an extracardiac conduit Fontan procedure coupled with minimal use of extracorporeal circulation is associated with a favorable early outcome.

The commonest postoperative morbidity was prolonged chest tube drainage for more than 7 days in 20 patients and more than 14 days in 7 patients. On multivariate analysis, the decreased preoperative ventricular function, CPB time, and older age seemed to influence the risk for prolonged pleural effusion but not the fenestration. The second cause of postoperative morbidity was atrial arrhythmias, observed in 4 patients, whereas in only 1 patient were arrhythmias documented preoperatively. Patients (n = 4) who underwent a concomitant Maze procedure with cryoablation did not have arrhythmia postoperatively. A cryoablation lesion with a right-sided Maze procedure is reserved to treat paroxysmal atrial flutter, and a left-sided Cox-Maze III procedure is used for paroxysmal atrial fibrillation [16]. Extracardiac Fontan, cryoablation, and pacemaker placement have reduced atrial arrhythmias and improved NYHA classifications [17].

Regarding the latest observations, we were surprised by the very high rate of patients (84%) presenting with sinus rhythm. Our policy is to give oral anticoagulants to every TCPC patient. In children, the referring cardiologist decides whether to pursue the treatment in follow-up. In general, adult patients known to be at risk, continue taking anticoagulant medication for life. Four patients had late cerebrovascular events: 3 with minor cerebral thromboembolism who recovered medically and 1 with hemorrhage who required an operation. Cerebrovascular accidents are a cause for concern in this particular population.

The functional benefits of the TCPC were significant: all but 2 patients were in NYHA class I or II at the last follow-up contact, continuing an active life. Increase in oxygen saturation and, eventually, a reduction of ventricular workload must principally be involved in this improvement. Our results let us expect a better and maybe a longer life for this cohort.

The present study included patients who had never been proposed for Fontan completion and also patients who were initially refused for such a procedure. The principal weakness of the present study was our inability to survey adult patients with UVH who were not found suitable for Fontan completion and also the ones who “missed the boat.” Because our center is a tertiary referral center, patients are referred for procedures from many medical (sometimes surgical) units all around the country; unfortunately, a survey of UVH patients who did not undergo Fontan completion requires a national multicenter trial, unavailable at present.

In conclusion, TCPC with the use of extracardiac conduit can be safely performed in properly selected adults, and it ameliorates exercise tolerance and subsequently quality of life. The present study shares the limitations of retrospective nonrandomized studies. In addition, unfortunately no data are available (and probably are obtainable) to identify the outcomes in UVH patients eligible for TCPC and for whom Fontan completion was intentionally postponed beyond childhood.

References


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