Midterm Outcomes after Conversion to Total Cavo-pulmonary Connection

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Objective: Some investigators reported the efficacy of the conversion to total cavo-pulmonary connection in the patients having the long term issue associated with the systemic venous connection. However, it’s indication is still controversial and surgical outcomes are uncertain. The purpose of this retrospective observational study was to investigate the optimal timing and midterm results. Patients and Methods: Twenty-six patients, average age was 26.3 ± 7.6 years and years after Fontan completion was 14.6 ± 5.1 years, underwent TCPC conversion between March 1992 and January 2008. There was atrial tachyarrhythmia in 16 patients (61%) and thrombus formation in 13 patients (50%). The medical records from those patients were observed and reviewed. Results: There were 4 hospital deaths and 4 late deaths. Operative factor and timing were implied, although the exact reason was not detected statistically. In midterm survivors, New York Heart Association classification was improved from 4 (22%) to 12 (67%) in class I, from 14 (78%) to 6 (33%) in class II and from 2 (8%) to none in class III, respectively. Post operative cardiothoracic ratio and SpO2 were 51 ± 6% and was 94 ± 1%. Conclusion: Clinical status in the patients enduring TCPC conversion was improved, although total cavo-pulmonary connection conversion was still high risk operation. Further investigation was needed.

Key words: total cavo-pulmonary connection conversion, Fontan operation, tachyarrhythmia, arrhythmia surgery, thromboembolism

Introduction

The Fontan operation was originally introduced as functional repair for patients with tricuspid atresia\(^1\). Despite several improvements in surgical techniques and postoperative treatment, late deterioration in functional status could be observed with longer duration of follow-up. Late postoperative complications include obstruction of venous pathways, atrial tachyarrhythmia, cyanosis secondary to systemic venous collateralization, thrombus formation and protein-losing enteropathy (PLE). These complications are associated with high central venous pressure (CVP) and systemic ventricular dysfunction\(^2,3\). Francis Fontan himself stated that this operation had an inherent palliative nature\(^4\). In our institute, 574 patients underwent the Fontan type operation and actual survival at 10, 15 and 20 years after the procedure was 94, 91 and 81% respectively. Freedom from arrhythmia at 10, 15 and 20 years after Fontan operation was 81, 60 and 41%. Freedom from major thromboembolism at 20 years was 92%. TCPC conversion, MAZE procedure and thrombus removal were performed for the patients who had dilated right atrium, large thrombus in the right atrium and atrial tachyarrhythmia at 14.6 ± 5.1 years after Fontan completion. Total cavo-pulmonary connection (TCPC) conversion has been performed to rescue the failed Fontan circulation as a palliative strategy\(^5\). Indications for TCPC conversion are still controversial and surgical outcomes are uncertain. In this study, we observed and reviewed medical records from the patients who underwent TCPC conversion, and investigated early and midterm outcome after the TCPC conversion.

Patients and Methods

Between 1992 and 2008, 26 patients underwent TCPC conversion at Tokyo Women’s Medical University (TWMU). Preoperative characteristics were
Table 1 Preoperative patient’s characteristics

<table>
<thead>
<tr>
<th>Congenital cardiac anomalies</th>
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<tbody>
<tr>
<td>TA</td>
<td>14</td>
</tr>
<tr>
<td>SRV</td>
<td>7</td>
</tr>
<tr>
<td>SLV</td>
<td>2</td>
</tr>
<tr>
<td>AVSD</td>
<td>2</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
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</tr>
<tr>
<td>Age at conversion (year)</td>
<td>26.3±7.6</td>
</tr>
<tr>
<td>Age at Fontan (year)</td>
<td>12.0±6.5</td>
</tr>
<tr>
<td>Years after Fontan (year)</td>
<td>14.6±5.1</td>
</tr>
<tr>
<td>Weight (kp)</td>
<td>47.2±9.0</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>male</td>
<td>9</td>
</tr>
<tr>
<td>female</td>
<td>17</td>
</tr>
<tr>
<td>NYHA class, n (%)</td>
<td></td>
</tr>
<tr>
<td>I</td>
<td>4 (15%)</td>
</tr>
<tr>
<td>II</td>
<td>20 (77%)</td>
</tr>
<tr>
<td>III</td>
<td>2 (8%)</td>
</tr>
<tr>
<td>CTR (%)</td>
<td>60±9</td>
</tr>
<tr>
<td>SpO2 (%)</td>
<td>91±6</td>
</tr>
<tr>
<td>CVP (mmHg)</td>
<td>14±4</td>
</tr>
<tr>
<td>EF (%)</td>
<td>55±8</td>
</tr>
<tr>
<td>Rhythm</td>
<td></td>
</tr>
<tr>
<td>SR</td>
<td>10 (38%)</td>
</tr>
<tr>
<td>af</td>
<td>11 (42%)</td>
</tr>
<tr>
<td>AF</td>
<td>3 (12%)</td>
</tr>
<tr>
<td>PSVT</td>
<td>2 (8%)</td>
</tr>
<tr>
<td>Thrombus formation</td>
<td>13 (50%)</td>
</tr>
</tbody>
</table>


shown in Table 1. The congenital cardiac anomalies included tricuspid atresia (n=14), single right ventricle (n=7), single left ventricle (n=2), unbalanced complete atrioventricular septal defect (n=2), Ebstein’s anomaly with pulmonary atresia (n=1). These patients had undergone RA-PA conduit (n=1), Björk operation (n=6), and RA-PA anastomosis (n=19) at Fontan completion. In our institute, the inclusion criteria for TCPC conversion were supra ventricular tachyarrhythmia, dilated right atrium, thromboembolism and progressive cyanosis. There was supra ventricular tachyarrhythmia in 16 patients (64%) (atrial fibrillation in 11, atrial flutter in 3, PSVT in 2). Thrombus formation in venous channel was observed in 13 (50%). Nine (38%) patients had severe cyanosis due to residual right to left shunt resulting from collateral formation. Atrioventricular valve regurgitation (over grade III) was observed in 5. Average age and weight at TCPC conversion, age at Fontan completion and years after Fontan completion were 26.3 ± 7.6 years old, 47.2 ± 9.0 kg, 12.0 ± 6.5 years old and 14.6 ± 5.1 years respectively. Preoperative clinical status evaluated by New York Heart Association (NYHA) classification was 4 patients (15%) in class I, 20 patients (77%) in class II and 2 patients (8%) in class III. Preoperative cardiothoracic ratio (CTR) and SpO2 were 60 ± 9% and 91 ± 6% respectively. Preoperative catheterization revealed mean CVP of 14 ± 4 mmHg and systemic ventricle ejection fraction of 55 ± 8%. Clinical course of these patients was reviewed.

Result

Fundamentally, the TCPC conversion consisted of extra-cardiac TCPC using artificial conduits in all patients, of which conduits were woven Dacron graft 20 mm in 1 patient, coated kitted graft 28, 30 mm in 2 patients and expanded polytetrafluoroethylene (e-PTFE) graft 16-30 mm in 23 patients. Twelve patients (46%) underwent MAZE procedure, although MAZE procedure was applied to all patients who required TCPC conversion recently. Recent 7 patients (27%) underwent DDD pacemaker implantation simultaneously regardless of heart block. Thrombectomy was performed in 13 patients having thrombus formation in right systemic venous channel. Atrioventricular valve plasty was performed in 5 patients who had atrioventricular valve regurgitation over grade III. In 2 patients, fenestration using e-PTFE conduit measured 5 mm in diameter was placed. The mean operative time, cardiopulmonary bypass (CPB) time and CVP at the time of weaning from CPB were 633 ± 165 minutes, 215 ± 165 minutes and 17.8 ± 3.8 mmHg respectively. In 11 patients operative time was over 600 minutes and in 12 patients CPB time was over 200 minutes due to concomitant procedures or unexpected hemorrhage. All data are expressed as means ± standard deviation.

1. Early mortality

Actual survival rate following TCPC conversion was 80 and 60% at 1 and 5 years (Fig. 1). There were 4 hospital deaths due to multi organ failure
caused by persistent low output syndrome. There was no significant difference in all variables between hospital death group and late death group and midterm survivors due to small number of patients (Table 2, 3). In these patients, mean age at Fontan completion, age at TCPC conversion and years to TCPC conversion were $12.9 \pm 8.3$, $27.3 \pm 8.8$, and $14.7 \pm 3.3$ respectively. All 4 patients were in NYHA class II and 3 patients had atrial fibrillation and thrombus in right atrium before surgery. Preoperative cardiac catheterization showed mean CVP and systemic ventricle ejection fraction (EF) of $13 \pm 5$ mmHg and $51 \pm 11\%$ respectively. All 4 patients underwent TCPC conversion using artificial conduit and 3 patients who had arrhythmia and thrombus underwent MAZE procedure and thrombectomy. Fenestration was placed for 1 patient. Operation time, CPB time and CVP at weaning from CPB were $828 \pm 193$ min, $296 \pm 93$ min and $21.5 \pm 5.7$ mmHg respectively. Three patients had unexpected cardiac laceration during the revision of median sternotomy and dissection. Average blood loss during the operation was $2,625 \pm 2,652$ ml. Two patients needed intra aortic balloon pumping at weaning from CPB. All 4 patients were not able to be weaned from respirator.

2. Late mortality

There were 4 late deaths. Also there was no significant difference in all variables due to small number of patients (Table 2, 3). Their mean age at Fontan completion, age at TCPC conversion and years to TCPC conversion were $10.4 \pm 1.4$ years old, $24.0 \pm 8.7$ years old and $13.9 \pm 8.1$ years respectively. Preoperatively, 2 patients were in NYHA class II, 2 patients who had severe TR and cardiac dysfunction (preoperative ejection fraction 45%) and congestive liver cirrhosis before TCPC conversion were in NYHA class III. Three patients had something ar-

![Fig. 1 Actual survival rate after TCPC conversion](image)

**Fig. 1** Actual survival rate after TCPC conversion
Actual survival rate following TCPC conversion was 80 and 60% at 1 and 5 years.

<table>
<thead>
<tr>
<th>Table 2</th>
<th>Preoperative variables</th>
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<tbody>
<tr>
<td></td>
<td>HD (n = 4)</td>
</tr>
<tr>
<td>Age at Fontan completion (year)</td>
<td>$12.9 \pm 8.3$</td>
</tr>
<tr>
<td>Age at conversion (year)</td>
<td>$27.3 \pm 8.8$</td>
</tr>
<tr>
<td>Years to TCPC conversion (year)</td>
<td>$14.7 \pm 3.3$</td>
</tr>
<tr>
<td>NYHA</td>
<td>N.S.</td>
</tr>
<tr>
<td>I</td>
<td>5</td>
</tr>
<tr>
<td>II</td>
<td>4</td>
</tr>
<tr>
<td>III</td>
<td>2</td>
</tr>
<tr>
<td>Rhythm</td>
<td>N.S.</td>
</tr>
<tr>
<td>SR</td>
<td>1</td>
</tr>
<tr>
<td>af</td>
<td>3</td>
</tr>
<tr>
<td>AF</td>
<td>3</td>
</tr>
<tr>
<td>PSVT</td>
<td>2</td>
</tr>
<tr>
<td>CTR (%)</td>
<td>$65 \pm 9$</td>
</tr>
<tr>
<td>CVP (mmHg)</td>
<td>$13 \pm 5$</td>
</tr>
<tr>
<td>EF (%)</td>
<td>$51 \pm 11$</td>
</tr>
<tr>
<td>Spo2 (%)</td>
<td>$93 \pm 4$</td>
</tr>
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</table>

Table 3  Operative and post operative variables

<table>
<thead>
<tr>
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<th>HD (n = 4)</th>
<th>LD (n = 4)</th>
<th>SU (n = 18)</th>
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<tr>
<td>OPE time (min)</td>
<td>828 ± 193</td>
<td>627.3 ± 86.3</td>
<td>592.8 ± 158.7</td>
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<tr>
<td>CPB time (min)</td>
<td>296 ± 93</td>
<td>217.0 ± 24.4</td>
<td>196.9 ± 64.0</td>
</tr>
<tr>
<td>CVP (mmHg)</td>
<td>21.5 ± 5.7</td>
<td>17.7 ± 3.2</td>
<td>15.7 ± 2.3</td>
</tr>
<tr>
<td>Blood loss (ml)</td>
<td>2,625 ± 2,632</td>
<td>2,429 ± 1,887</td>
<td>1,927 ± 1,566</td>
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<tr>
<td>Hospital stay (days)</td>
<td>—</td>
<td>70 ± 33</td>
<td>52 ± 21</td>
</tr>
<tr>
<td>CTR (%)</td>
<td>—</td>
<td>59 ± 8</td>
<td>51 ± 6</td>
</tr>
<tr>
<td>SpO2 (%)</td>
<td>86 ± 8</td>
<td>94 ± 1</td>
<td>N.S.</td>
</tr>
<tr>
<td>NYHA</td>
<td>—</td>
<td>—</td>
<td>N.S.</td>
</tr>
<tr>
<td>I</td>
<td>—</td>
<td>0 ( 0%)</td>
<td>7 (40%)</td>
</tr>
<tr>
<td>II</td>
<td>—</td>
<td>0 ( 0%)</td>
<td>11 (60%)</td>
</tr>
<tr>
<td>III</td>
<td>—</td>
<td>4 (100%)</td>
<td>0 ( 0%)</td>
</tr>
<tr>
<td>Rhythm</td>
<td>—</td>
<td>3 ( 75%)</td>
<td>8 (44%)</td>
</tr>
<tr>
<td>SR</td>
<td>—</td>
<td>0 ( 0%)</td>
<td>6 (33%)</td>
</tr>
<tr>
<td>PMR</td>
<td>—</td>
<td>1 ( 25%)</td>
<td>4 (23%)</td>
</tr>
</tbody>
</table>


![Fig. 2 Pre and post operative NYHA classification in late death group](image)

Two patients were classified in class I, 2 patients were in class II before TCPC conversion but all patients were classified in class III after TCPC conversion.

Typhus. Mean preoperative CTR and SpO2 were 62 ± 13 and 91 ± 2% respectively. Preoperative cardiac catheterization showed that mean CVP and systemic ventricle EF were 18 ± 2 mmHg and 48 ± 4% respectively. Mean operative time, CPB time, CVP at the weaning of CPB were 627.3 ± 86.3 min, 217.0 ± 24.4 min and 17.7 ± 3.2 mmHg respectively. And mean bleeding during the operation was 2,429 ± 1,887 ml. One patient who had severe heart failure with TR preoperatively died at 3 months after TCPC conversion. One patient who complicated with liver cirrhosis died due to PLE at 3 years after TCPC conversion. And the patient who complicated with SLE died due to pneumonia at 3 years after TCPC conversion. The patient who had hepatic failure died due to rupture of esophageal varices at 7 years after TCPC conversion. Three patients restored sinus rhythm after TCPC conversion and postoperative mean CTR was 59 ± 8%. Postoperative mean SpO2 was 86% ± 8. Nevertheless after TCPC conversion, all patients started at NYHA class III (Fig. 2).

3. Midterm outcome of survivors

In mid-term follow-up, 12 (67%) of 18 survivors were in NYHA class I, 6 (33%) were in NYHA class II (Fig. 3). Postoperative CTR was 51 ± 6% and SpO2 was 94 ± 1%. 8 patients (44%) had sinus rhythm, 6 patients (33%) had a paced rhythm while 4 patients (23%) had atrial fibrillation (Fig. 4). Ratio of patients in NYHA class I and regular heart rhythm (including sinus rhythm and regular paced rhythm) was likely to increase although there was no significant difference statistically between the groups.

Discussion

The strategy of the conversion from failed Fontan to TCPC was first reported by Laks et al. Although there are some indications for TCPC conversion (including a giant atrium with thrombus, arrhythmias refractory to medical or simple electro-
Fig. 3 Pre and post operative rhythm of the midterm survivors
Fourteen patients (77%) had regular rhythm (8 patients (44%) had sinus rhythm, 6 patients (33%) had a paced rhythm) after TCPC conversion, while 6 patients (34%) had sinus rhythm before TCPC conversion.

physiologic treatment, pulmonary venous obstruction by the enlarged right atrium and ventricular dysfunction), optimal timing for this approach is not clearly defined. Since arrhythmia surgery was not performed at the TCPC conversion initially, the reduction of the energy loss was reported to improve the disadvantage in hemodynamics. Marcelletti CF et al reported that the TCPC conversion reduced the risk for arrhythmia and thrombo-embolism. Especially when anti-arrhythmic surgical intervention was performed at the TCPC conversion simultaneously, this procedure decreased arrhythmia resulting in overall improvement in quality of life and functional class. Sheikh et al reported that TCPC conversion can be achieved with low mortality and improvement in exercise ability.

These procedures sometime required difficulties because of severe adhesion resulting from the multiple previous operations. Mavroudis C et al reported that long CPB time was considered to be a risk factor for death or heart transplantation.

In our study, there was a trend to affect the hemodynamics including elevated CVP at the weaning from CPB by longer duration of CPB and operative time, which were associated with operative bleeding caused by unexpected cardiac laceration during the revision of median sternotomy and dissections, although there was no statistical significance. Thus operative technical factors including longer duration of CPB or large volume of blood transfusion might deteriorate lung condition which influences postoperative hemodynamics. Consequently, it might affect early mortality.

Preoperative condition such as cardiac function or hepatic function may affect late mortality since preoperative cardiac dysfunction and liver cirrhosis were observed in 2 patients and 3 of 4 patients died after discharge due to heart failure or complication of hepatic failure. Although it was said that severe ventricular dysfunction attributed to chronic persistent arrhythmia or anatomic pathway obstruction may be improved with surgical intervention, severe cardiac dysfunction and hepatic dysfunction are deemed as high risk factors for late death. McElhinney et al reported that the most questionable indication for TCPC conversion involves the patient with deteriorating functional status who has no specific target conditions; for example, the patient who is functionally very limited and has moderately elevated pulmonary vascular resistance, moderately depressed ventricular function, a large right atrium without specific complications and no obstructive lesions. In these cases, the improvement gained by TCPC conversion might well be outweighed by the risk of the procedure itself. Thus optimal timing of the TCPC conversion is important and the TCPC conversion should be undertaken at the early phase when patients are asymptomatic and their occult organ failure is reversible. Preop-
ervative careful assessment is mandatory for the TCPC conversion and careful follow-up is also necessary to avoid overlooking the timing of the TCPC conversion.

In midterm outcome of survivors, CTR was decreased and SpO2 was increased. Regular rhythm (including paced rhythms) restored after the TCPC conversion concomitant with arrhythmia surgery in 14 patients and NYHA classification was improved from 4 (22%) to 12 (67%) in class I and from 14 (78 %) to 6 (33%) in class II (Fig. 4). Restoration of regular rhythm might contribute to the improvement of the activity of the daily life in those patients (Fig. 3).

Conclusion

TCPC conversion is still high risk operation. Potential complications related to the operative technique and timing for this operation might affect postoperative course. Restoration of regular rhythm might contribute to the activity of the daily life. Further investigation was required.

References

Total cavo-pulmonary connection conversion 後の中遠隔期成績の検討

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上松 耕太・石原 和明・岩田 祐輔・黒澤 博身

【背景】Fontan 手術は当初三尖弁閉鎖術に対する根治術として行われていたが、次第に適応拡大され単心室性の房室結節を示す症例やさまざまな複雑心室形により二心室修復が不可能な症例に対しても行われるようになっている。

【目的】Fontan 手術遠隔期には上室性不整脈、心房細動性歯歯症、チアノーゼの進行や心不全等の合併症が認められる。こうした症例に対し Total cavo-pulmonary connection (TCPC) conversion が施行されるようになってきているが手術適応は明確でなく、術後成績も明らかでない。今回われわれは TCPC conversion 後の中遠隔期成績より手術適応を検討した。

【症例と方法】対象症例は 1992 年 3 月から 2008 年 1 月までに当科にて TCPC conversion を行った 26 例とし、手術時平均年齢、体幹 Fontan 手術年齢と初回 Fontan 手術からの期間はそれぞれ 26.3±7.6 歳、47.2±9.0 kg、12.0±6.5 歳と 14.6±5.1 年であった。上室性不整脈を 16 例 (64%) に、右房内血栓を 13 例 (50%) に認めた。術前 NYHA 心機能分類は 4 例 (15%) が class I、20 例 (77%) が class II、2 例 (8%) が class III であった。これらの症例を後方視的に検討した。

【結果】全例に TCPC conversion を施行し、同時手術として 12 例 (46%) に MAZE 手術を、右房内血栓を認め る症例には血栓除去術を施行した。病院死亡を 4 例に認め、いずれも低拍出量症候群の多臓器不全で失った。 遠隔期死亡を 4 例に認め、心不全、肺炎、肝機能障害に伴う合併症で失った。遠隔期死亡群では 2 例 (50%) が 衛前 NYHA class I、2 例 (50%) が class II で術後は全例 class III で改善が認められず手術前後時期に問題があると考えられた。生存症例の術後 rhythm は頑調例 8 例 (44%)、ベースメーカー調律 6 例 (33%) と 14 例 (77%) に整調律を認め、術前 12 例 (66%) に認めている不整脈は 4 人 (23%) に減少し、NYHA 心機能分類では術前 4 人が class I、14 人が class II であったが、術後 12 人が class I、6 人が class II と改善が認められた。手術時期および術中因子が遠隔死亡および病院死亡に関与し、整脈化が日常生活を改善していることが示唆された。

【結論】TCPC conversion は risk の高い術式で手術手技に関連する因子と手術介入時期が術後経過に関与する と考えられた。整脈化が日常生活を改善することが示唆されたが、今後更なる検討が必要である。