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ORIGINAL ARTICLE

Early and intermediate-term results of the extracardiac conduit total cavopulmonary connection for functional single-ventricle hearts

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KEYWORDS
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Background/Purpose: Fontan operation has evolved from atriopulmonary connection to total cavopulmonary connection (TCPC) due to its advantages in terms of hemodynamics and reduction of atrium-related complications. We analyzed the early and intermediate-term results of extracardiac conduit TCPC (EC-TCPC) procedure in patients with functional single ventricle to investigate the risk factors of surgical mortality and intermediate failure.
Methods: Retrospective review of the medical records of 88 consecutive patients with functional single ventricle who underwent EC-TCPC from 2000 to 2013 was conducted.
Results: The follow-up was 100% complete, ranging from 3 months to 13 years (mean 7.0 ± 3.8 years). There were two (2.3%) hospital and 18 (20.4%) late deaths. The estimated event-free survival rates at 1 year, 5 years, and 10 years were 90.6%, 89.3%, and 77.2%, respectively. On univariate analysis, fenestration was the only risk factor for surgical mortality ($p = 0.027$). On multivariate analysis, the significant atrioventricular valve regurgitation was the only risk factor for intermediate failure ($p = 0.017$).
Conclusion: The clinical results of EC-TCPC in patients with functional single ventricle were satisfactory. The patients who needed fenestration during operation had higher risk of surgical

Conflicts of interest: The authors have no conflicts of interest relevant to this article.

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mortality. Significant atrioventricular valve regurgitation had negative impact on intermediate survival.
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Introduction

Fontan-type operation has been performed in a wide variety of functional single-ventricle hearts.[1–3] This procedure has evolved from atriopulmonary connection to total cavopulmonary connection (TCPC) with either an intracardiac lateral tunnel (LT) or the extracardiac conduit (EC) technique due to its advantages in terms of hemodynamics and reduction of atrium-related complications.[4–8]

In recent times, the EC type of reconstruction has become more popular than the LT type due to the following reasons: easier technique; suitable for all kinds of single-ventricle patients, especially those with heterotaxy syndrome with anomalous systemic or pulmonary venous return; avoidance of systemic cooling and cardioplegic arrest; and extensive atrial manipulation, all of which may have a negative effect on survival.[9] However, there are still some unsolved issues regarding the use of EC-TCPC, including absence of growth potential, the most optimal size, risk of thromboembolism, and the significance of fenestration. In this study, we retrospectively analyzed our early and intermediate-term results during the 13-year period in 88 consecutive patients with functional single-ventricle pathology to investigate the risk factors of surgical mortality and intermediate failure following EC-TCPC.

Methods

Patient selection

A single-center retrospective analysis of 88 consecutive patients who underwent EC-TCPC for various functional single-ventricle hearts from 2000 to 2013 was conducted (Table 1). This study was approved by the Institutional Review Board of Kaohsiung Veterans General Hospital and the need for written informed consent from the patients was waived.

The study cohort included 48 (54.5%) male and 40 (45.5%) female patients. The mean age at operation was 6.3 ± 5.3 years (range 1.5–33 years) and the mean body weight at operation was 20.9 ± 13.3 kg (range 9.2–70 kg).

Exclusion of patients with failed palliation

Three patients with right atrial isomerism, total anomalous pulmonary venous connection, and pulmonary venous obstruction who failed to survive the initial palliation were not included in this study.

Operative characteristics

Although the use of noninvasive computed tomography might reduce the use of diagnostic cardiac catheterization, two-dimensional echocardiography and complete cardiac catheterization were performed in all patients before the operation.[10] The indications for EC-TCPC in our institute are as follows: aged over 1 year, body weight of more than 9 kg, mean pulmonary artery pressure of less than 20 mmHg, ejection fraction of more than 30%, and Nakata index of more than 150 mm^2/m^2. Primary EC-TCPC was performed in only three patients and 85 patients had received various kinds of palliative procedures before the completion of EC-TCPC. On average, 2.2 palliative procedures were performed for each patient before EC-TCPC completion (Table 2). The bidirectional Glenn shunt was

Table 1 Diagnosis of the 88 patients.

Diagnosis	*N* (%)
Heterotaxy, double-outlet right ventricle, CAVSD	37 (42.0)
Tricuspid atresia	18 (20.5)
Double-inlet single ventricle	11 (12.5)
Pulmonary atresia with intact ventricular septum	7 (8.0)
Mitral atresia	4 (4.5)
Ebstein anomaly	4 (4.5)
CAVSD	3 (3.4)
Hypoplastic left heart syndrome	2 (2.3)
L-Transposition of great arteries	2 (2.3)
Total	88 (100.0)

CAVSD = complete atrioventricular septal defect.

Table 2 Previous procedures.

Procedure	*n*
Bidirectional Glenn shunt	85
Systemic pulmonary arterial shunt	47
Pulmonary artery banding	16
Pulmonary artery angioplasty	12
Atrial septectomy	10
Damus–Kaye–Stansel procedure	3
Repair coarctation	3
Atrioventricular valve repair	2
Bulboventricular resection	1
Atrioventricular valve replacement	1
Pulmonary vein angioplasty	1
Atriopulmonary anastomosis	1
Total (average procedure/patient)	182 (2.2)

performed at the mean age of 2.9 ± 3.1 years, and the EC-TCPC was completed at the mean age of 6.0 ± 4.5 years. The mean procedural interval between the bidirectional Glenn shunt and EC-TCPC completion was 3.1 ± 5.2 years. More-than-moderate degree (3+) of atrioventricular valve regurgitation was identified in 12 patients and valve repair, whenever feasible, was performed on five of these 12 patients.

Surgical technique

The operation was performed through the median sternotomy using standard cardiopulmonary bypass. The superior vena cava was cannulated just proximal to the innominate vein and the inferior vena cava was cannulated just above the cavophrenic junction. Normothermic or mild hypothermic cardiopulmonary bypass was instituted initially. If an intracardiac procedure such as atrioventricular valve repair, atrial septectomy, or resection of subaortic conus was required, cold crystalloid cardioplegic arrest was used for myocardial protection. The inferior vena cava was transected at the atriocaval junction and a polytetrafluoroethylene (PTFE) straight tube graft (Gore-Tex Stretch Vascular Graft, W.L. Gore & Associates, Flagstaff, AZ, USA) was interposed between the inferior vena cava and the right pulmonary artery in all patients except in a 33-year-old patient with a 26-mm Dacron graft. The size of the graft was 16 mm in six patients, 18 mm in 24 patients, 20 mm in 35 patients, and 22 mm in 22 patients (Figure 1).

A fenestration was created by interposition of the PTFE graft (diameter ranged from 4 mm to 7 mm) between EC and atrium in 15 (17.1%) patients with low cardiac output after TCPC completion. The size of the fenestration was further adjusted by stepwise clipping of the graft with hemoclip to achieve the most balanced hemodynamic and systemic oxygen saturation.

Follow-up

Follow-up was 100% complete and the period ranged from 3 months to 13 years (mean 7.0 ± 3.8 years). *Surgical mortality* was defined as any death within 30 days after surgery or after 30 days but during the same hospitalization period subsequent to the operation.

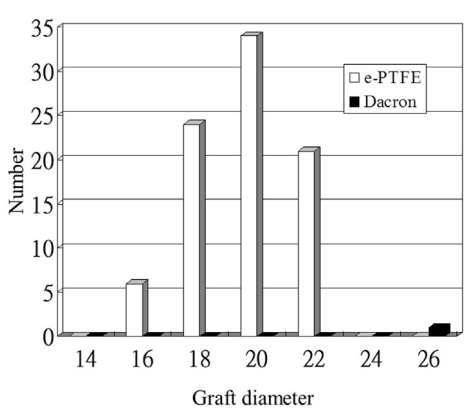


Figure 1 Distribution of the size of the extracardiac conduit. e-PTFE = expanded polytetrafluoroethylene.

Two kinds of antiplatelet drugs (aspirin and dipyridamole) were prescribed for all patients to circumvent thromboembolic complication postoperatively. Furthermore, warfarin was added initially in eight patients with atrial fibrillation ($N = 7$) or after a mechanical valve replacement ($N = 1$). The warfarin dose was titrated to the international normalization ratio of 2.0–3.0.

Chest roentgenogram, standard 12-lead electrocardiogram, and two-dimensional echocardiography were performed every 6 months. Cardiac catheterization had been performed in 51 patients 1 year postoperatively for evaluating the postoperative pulmonary arterial pressure and pulmonary vascular resistance change.

Statistical analysis

Statistical analysis was performed using the Statistical Package for the Social Sciences for Windows software system, version 17.0 (SPSS Inc., Chicago, IL, USA). Continuous variables acquired from the clinical characteristics were expressed as mean ± standard deviation and were compared using the independent *t* test. The Chi-square test or Fisher exact test according to the number of events for variables appraised was used for comparisons made between categorical variables. Actuarial survival and cardiac-related event-free survival were estimated by the Kaplan–Meier method with the log-rank test. Cox proportional hazard analysis was used for identifying the significant risk factor for the intermediate failure. A two-tailed *p* value of less than 0.05 was considered statistically significant.

Results

Operative characteristics

Forty-five patients (51.1%) underwent EC-TCPC at the age of more than 4 years. Twenty-one patients (23.9%) underwent 26 concomitant procedures during EC-TCPC completion (Table 3). The cardiopulmonary bypass time ranged from 48 minutes to 442 minutes (mean 125.6 ± 89.7 minutes). The cardioplegic cardiac arrest was used in 15 patients in whom the cardiac ischemic time

Table 3 Concomitant procedures.

Procedure	*n*
Pulmonary artery angioplasty	8
Atrioventricular valve repair	5
Atrial septectomy	4
Right ventricular exclusion	4
Permanent pacer implantation	2
Damus–Kaye–Stansel procedure	1
Resection of subaortic conus	1
Takedown of previous atriopulmonary anastomosis	1
Total procedure number (patient)	26 (21)

ranged from 13 minutes to 84 minutes (mean 49.4 ± 27.5 minutes). Atrioventricular valve repair for regurgitation (>3+) was performed in five of the 12 patients.

Outcomes

The endotracheal tube was extubated within 24 hours in 77 patients (87.5%). Prolonged pleural effusion for more than 14 days occurred in 40 patients (45.5%). The mean pulmonary artery pressure was 12.7 ± 4.0 mmHg before EC-TCPC and 15.9 ± 2.5 mmHg after EC-TCPC; the preoperative pulmonary vascular resistance was 2.11 ± 0.79 WU·m^2; the arterial oxygen saturation was 84.2% ± 6.4% before EC-TCPC and 96.9% ± 2.6% after EC-TCPC; the ventricular end-diastolic pressure was 8.7 ± 4.2 mmHg before EC-TCPC and 7.5 ± 2.8 mmHg after EC-TCPC; the pulmonary-to-systemic flow ratio was 0.9 ± 0.2 before EC-TCPC and 1.0 ± 0.2 after EC-TCPC. There were two surgical mortalities. A 28-month-old boy with dextrocardia, tricuspid atresia, and transposition of great arteries developed refractory supraventricular tachyarrhythmia. He died of multiple organ failure 4 days after the operation. The other one was a 6-year-old girl with Ebstein anomaly, who developed severe low cardiac output after the operation. After 7-day extracorporeal membranous oxygenation, she died of multiple organ failure.

Late death occurred in 13 patients. The causes of death were ventricular dysfunction in four patients, pneumonia with sepsis in three patients, protein-losing enteropathy associated with congestive heart failure in two patients, conduit thrombosis due to high pulmonary vascular resistance in two patients, pulmonary arteriovenous malformation in one patient, and nephrotic syndrome in one patient. One patient underwent heart transplantation for intractable heart failure 1 year after EC-TCPC. On Cox proportional hazard analysis, more-than-moderate atrioventricular valve regurgitation was the only risk factor for intermediate death ($p = 0.017$).

Fenestration

In total, 15 patients (17.1%) required fenestration and four of them (4.6%) were fenestrated under the subsequent rescue condition in the intensive care unit due to low cardiac output. Eventually, two of these 15 patients (1 out of the 4 rescued patients) died of multiple organ failure. All the residual fenestrations ($n = 13$) were noted to be occluded spontaneously during the follow-up cardiac catheterization 1 year after the operation. On univariate analysis, fenestration was the only risk factor for surgical mortality ($p = 0.027$; Table 4).

Table 4 Analyzed predictors for surgical mortality (N = 2).

	No *n/N*, %	Yes *n/N*, %	*p*
Demographics			
Age ≥ 4 y	1/43, 2.3	1/45, 2.2	>0.99
Male	1/48, 2.1	1/40, 2.5	>0.99
Pre-PAP ≥ 15 mmHg	1/64, 1.6	1/24, 4.2	0.473
Pre-PVR ≥ 2.5 WU·m^2	1/73, 1.4	1/15, 6.7	0.313
Pathology			
Asplenia	2/80, 2.5	0/8, 0	>0.99
AVVR ≥ 3+	2/76, 2.6	0/12, 0	>0.99
CAVV	2/52, 3.8	0/37, 0	0.511
Dextrocardia	1/66, 1.5	1/22, 4.5	0.440
Heterotaxy	2/51, 3.9	0/37, 0	0.507
RV morphology	2/40, 5.0	0/48, 0	0.204
None-situs solitus	2/62, 3.2	0/26, 0	>0.99
SVDA	2/58, 3.4	0/30, 0	0.545
TAPVC	2/86, 2.3	0/2, 0	>0.99
Previous palliation			
Atrial septectomy	2/78, 2.6	0/10, 0	>0.99
BDG	0/3, 0	2/85, 2.4	>0.99
PA angioplasty	2/76, 2.6	0/12, 0	>0.99
PA banding	2/72, 2.8	0/16, 0	>0.99
Shunt	2/41, 4.9	0/47, 0	0.214
Operative variables			
Aortic clamp	1/73, 1.4	1/15, 6.7	0.313
CPB ≥ 2 h	1/59, 1.7	1/29, 3.4	>0.99
Fenestration	0/73, 0	2/15, 13.3	0.027
Postoperative course			
PLE ≥ 2 wk	2/48, 4.2	0/40, 0	>0.99
Post-PAP ≥ 15 mmHg	0/39, 0	2/49, 4.1	0.501
SV arrhythmia	2/81, 2.5	0/7, 0	>0.99

AVVR = atrioventricular valve regurgitation; BDG = bidirectional Glenn; CAVV = common atrioventricular valve; CPB = cardiopulmonary bypass; PA = pulmonary artery; PLE = pleural effusion; post-PAP = postoperative pulmonary arterial pressure; pre-PAP = preoperative pulmonary arterial pressure; pre-PVR = preoperative pulmonary vascular resistance; RV = right ventricle; SV = supraventricular; SVDA = systemic venous drainage anomaly; TAPVC = total anomalous pulmonary venous connection; WU = Wood units.

Arrhythmia

Two patients developed complete atrioventricular block before operation and a permanent pacemaker was implanted during EC-TCPC. Seven patients developed atrial fibrillation before operation due to high atrial pressure and this was resolved in four patients after EC-TCPC. Medical rate control with anticoagulation therapy was appropriate in the three residual patients.

Reoperation

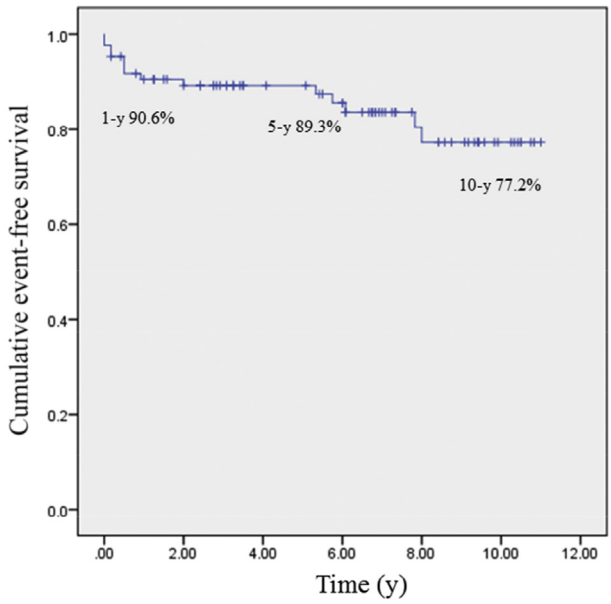
Seven patients (8.0%) were reoperated and five of them died during the follow-up period. Atrioventricular valve operations were performed in four patients (3 replacements and 1 repair) because of the deteriorated atrioventricular valve regurgitation with ventricular failure. Three of them died and one patient received heart transplantation at last. The Damus—Kaye—Stansel procedure was performed in one patient for the progressive subaortic stenosis, but this patient eventually died of protein-losing enteropathy. An EC-TCPC was taken down in one patient due to ventricular failure and conduit thrombosis, and this

patient finally died. The hemiazygos to left pulmonary arterial anastomosis was performed in one patient for progressive cyanosis and satisfactory result was achieved. The estimated event-free survival rates at 1 year, 5 years, and 10 years were 90.6%, 89.3%, and 77.2%, respectively (Figure 2).

Discussion

EC-TCPC

In Taiwan, severe congenital heart diseases account for 11.5% of all cases. Among them, the single-ventricle heart accounts for 0.6% of all cases. The estimated survival at 5 years of age is 66.0% in this population.[11] Significant improvements of outcomes after Fontan operation for this group of patients have evolved since the original description of the procedure by Fontan and Baudet in 1971.[1] The factors improving early survival include better understanding of Fontan physiology, careful selection and preparation of candidate, earlier and staging operation, and refinement in procedure. LT-type TCPC and EC-TCPC are the two most common modifications of the Fontan operation with comparable operative survival and midterm results. However, EC-TCPC has the advantages of applicability to almost all kinds of complex cardiac anomalies, especially for patients with heterotaxy and abnormal systemic or pulmonary venous return; less cardiac arrest and cardiopulmonary bypass times; avoidance of atriotomy and suture line loading in atrium; more efficient laminar flow with less energy loss; and fewer postoperative dysrhythmias.[7,9,12,13]



Year	1	2	3	4	5	6	7	8	9	10
Patient at risk	82	77	68	60	59	55	44	38	32	22

Figure 2 Kaplan—Meier curve demonstrated the estimated event-free survival rates at 1 year, 5 years, and 10 years to be 90.6%, 89.3%, and 77.2%, respectively.

Although the artificial conduit used for the EC-TCPC technique has been reported to limit growth potential, recent reports have demonstrated that longitudinal growth of the autologous vessels above and below the conduit can compensate this limitation.[14,15]

Conduit size

In our early series, one of six patients with 16-mm EC experienced conduit thrombosis 6 months after EC-TCPC despite dual-antiplatelet therapy and eventually, revision for the EC replacement was mandatory. Theoretically, the smaller size of EC might be less relevant to the thrombotic episode and the elevated pulmonary vascular resistance in this particular patient might play an important role in flow stasis, and then in subsequent EC thrombosis.[16] Therefore, considering the general adult inferior vena cava diameter of around 20 mm, consistent with the previous report, the PTFE with diameter of 18—22 mm was used as the EC size of choice for patients with body weight of more than 10 kg.[8,16]

Fenestration

The effect of fenestration is still controversial. In most contemporary literatures, the fenestration was used routinely; additionally, it was advocated that fenestration would not affect early survival, resource use, or pleural drainage amount.[7,17—19] In this study, however, we performed fenestration selectively in high-risk patients with elevated pulmonary vascular resistance ($>$2.5 WU$\cdot$m^2).[20] This strategy might bias the fenestration as the only risk factor for surgical mortality on univariate analysis. From our limited experiences, we felt that fenestration was a valuable adjunctive procedure without significant drawback and might be beneficial for the high-risk patients during the perioperative period.

Antithrombotic issue

Routine and lifelong anticoagulation therapy was advocated to prevent the thrombosis and late stenosis of EC.[8,21] Without anticoagulation therapy, 20—23% of thrombus formation in the EC was also reported.[22,23] However, the optimal anticoagulation level for a patient after EC-TCPC is still unknown.[24,25] We reserved the anticoagulant therapy for those with mechanical valve or supraventricular arrhythmias to avoid major bleeding complication. Instead, dual-antiplatelet therapy with aspirin and dipyridamole was used for all other patients. During the mean follow-up of 7.0 $\pm$ 3.8 years, EC thrombosis was noted in two patients (2.3%). Further study is necessary to confirm the role of anticoagulation and antiplatelet therapies in ameliorating the EC thrombosis.

Outcomes and reoperation

The surgical mortality regarding Fontan-type operations declined dramatically to less than 5% during the past two decades and this rate in our series was 2.3%, which was

fairly comparable to the published results.[7,13,19] The reoperation rate of Fontan population after 5–15 years' follow-up was reported to be 29%.[2] In this study, the reoperation rate was 8% (7/88). Four of the seven reoperations were mandatory for the intractable ventricular failure associated with significant atrioventricular valve regurgitation and the results were unsatisfactory. This fact made the significant atrioventricular valve regurgitation the only risk factor for intermediate failure on multivariate analysis. Heart transplantation was advocated as another therapeutic option for this difficult population.[26] However, donor heart shortage, especially in this age group, is an unsolved global issue. We only had a single transplantation experience for failing Fontan.

Risk analysis

On univariate analysis, fenestration was the only risk factor for surgical mortality ($p = 0.027$). Significant atrioventricular valve regurgitation was the only risk factor for intermediate failure. Driscoll and colleagues[2] also demonstrated the same result. Early operation and more aggressive management of atrioventricular valve regurgitation might be mandatory for salvaging this critical population.

Limitation of the study

The natures of retrospective, nonrandomized, and small population are the inevitable weakness of this study. A larger, prospective, and randomized multicenter cooperative study will certainly circumvent this predicament.

Conclusion

In conclusion, we demonstrated 88 consecutive patients with functional single ventricle who underwent EC-TCPC with satisfactory long-term clinical outcomes. The patients who needed fenestration during operation had higher risk of surgical mortality. Significant preoperative atrioventricular valve regurgitation had negative impact on intermediate survival.

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