

Midterm surgical results of total cavopulmonary connection: Clinical advantages of the extracardiac conduit method

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Objective: We evaluated the midterm surgical outcomes of intra-atrial lateral tunnel and extracardiac conduit total cavopulmonary connection to clarify the clinical superiority.

Methods: Patients (n = 167) underwent total cavopulmonary connection (88 with lateral tunnel and 79 with extracardiac conduit) from November 1991 to March 1999. Survival, incidence of reoperation and late complications, exercise tolerance, hemodynamic variables, and plasma concentration of natriuretic peptide type A were compared. In the lateral tunnel group, time-related change in lateral tunnel size was investigated for its relationship to postoperative arrhythmias.

Results: The 8-year survival was 93.2% in the lateral tunnel group and 94.9% in the extracardiac conduit group. Seven reoperations were performed in the lateral tunnel group but none in the extracardiac conduit group. Supraventricular arrhythmias developed in 14 patients (15.9%) in the lateral tunnel group and in 4 patients (5.1%) in the extracardiac conduit group ($P = .003$). Freedom from cardiac-related events was 72.5% in the lateral tunnel group and 89.8% in the extracardiac conduit group at 8 years ($P = .0098$). Hemodynamic variables and exercise tolerance were similar in both groups but plasma natriuretic peptide type A concentration, a parameter of atrial wall tension, was higher in the lateral tunnel group. In the lateral tunnel group, intra-atrial tunnel size increased by 19.4% during the 44.2-month interval and the percent increase in tunnel size was an independent predictor of supraventricular arrhythmias.

Conclusions: The midterm survival, hemodynamic variables, and exercise tolerance were similar and satisfactory in both lateral tunnel and extracardiac conduit groups; however, the incidence of cardiac-related events was significantly less frequent in the extracardiac conduit group. In the lateral tunnel group, careful observation is required to monitor the relationship of the dilating tendency of the intra-atrial tunnel and the development of late complications.

Fontan-type operation has been performed in a wide variety of functional single ventricle hearts, and it has evolved from atriopulmonary connection to total cavopulmonary connection (TCPC) on the basis of the theoretical advantages in terms of hydrodynamics and reduction of atrium-related complications.¹⁻⁴ As more patients survive the operation, their long-term functional status has become a major concern.

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Currently, the 2 major modifications of TCPC are the intra-atrial lateral tunnel (LT) method and the extracardiac conduit (EC) method; however, controversy has remained in terms of long-term superiority.⁵⁻⁸ In this study, we evaluated our midterm surgical results with these 2 procedures to provide some data and recommendations on this issue.

Methods

Study Group

Patients (n = 167) who underwent TCPC at Fukuoka Children's Hospital from November 1991 to March 1999 were enrolled in this study. Eighty-eight patients underwent LT-TCPC and 79 patients had EC-TCPC. Until 1996, we performed both modifications, but after 1997, we performed EC-TCPC in all patients. The age and weight at operation was 6.2 ± 3.9 years (1.5-18.4) and 17.8 ± 9.5 kg (7.3-56.5) in the LT group and 6.2 ± 4.1 years (1.6-19) and 17.9 ± 9.7 kg (8.0-55.8) in the EC group. Follow-up was achieved for all patients, ranging from 39 months to 127 months (median 78, mean 75 months). Medical records, operative records, all available electrocardiograms (ECGs), echocardiograms, and pre- and post-operative cardiac catheterization records were reviewed. The characteristics of the study population are summarized in Table 1, and preoperative hemodynamic variables are shown in Table 2. The pulmonary artery index (Nakata index) was smaller in the EC group (330.2 ± 117.7 vs 271.2 ± 171.5 mm²/m², $P = .01$); however, there was no difference between the 2 groups in the other hemodynamic variables measured. Most of the patients enrolled in this study were taking a low dose of warfarin and aspirin against hypercoagulability state and angiotensin-converting enzyme inhibitor for cardiovascular protection postoperatively.

Operative Techniques

All patients underwent TCPC using cardiopulmonary bypass. In LT-TCPC, after cardioplegic arrest, a longitudinal atrial incision was made. Intra-atrial baffling was completed to route the blood flow from the inferior vena cava to the superior vena cava orifice, which was connected to the inferior surface of the pulmonary artery (intra-atrial baffling method). The baffle material was an autologous pericardial patch in 30 patients, a xenopericardial patch in 24 patients, and a polytetrafluoroethylene (PTFE) patch in 14 patients. In the other 20 patients, the intra-atrial tunnel was constructed by use of atrial wall and intra-atrial septal tissue (autologous tunnel method).⁹ In determining the suture line, careful attention was paid to avoid the crista terminalis and the area close to the sinus node. In EC-TCPC, a connection between the inferior vena cava and the pulmonary artery was established by a PTFE straight tube graft in all patients. The size of the graft was 16 mm in 32 patients, 18 mm in 21 patients, and 20 mm in 26 patients. The heart was arrested only when intracardiac procedures such as atrioventricular valvuloplasty, atrial septectomy, and others were required in this group. Fenestration was created in 1 patient in each group. All patients had previously or simultaneously undergone bidirectional cavopulmonary anastomosis.

Follow-up Assessments

All patients had follow-up assessments with a median duration of 78 months, with ECGs, chest radiographs, echocardiograms, and

TABLE 1. Characteristics of the study population

	LT	EC
n	88	79
Age (y)	6.2 ± 3.9	6.2 ± 4.1
Sex (M/F)	51/37	40/39
Diagnosis		
SV	63 (71.6%)	53 (67.1%)
TA	12 (13.6%)	11 (13.9%)
PA-IVS	3 (3.4%)	10 (12.7%)
HLHS	0	4 (5.1%)
Others	10	1
Heterotaxy syndrome	31(35.2%)	19 (24.1%)
Prior procedure		
SP shunt	42 (47.7%)	36 (45.6%)
PAB	15 (17.0%)	19 (24.1%)
CoA repair	1 (1.1%)	4 (5.1%)
BDG	19 (21.6%)	55 (69.6%)*
Concomitant procedure		
PA plasty	23 (26.1%)	32 (40.5%)
AVV plasty	18 (20.5%)	14 (17.7%)
Atrial septectomy	12 (13.6%)	10 (12.7%)
SAS releif	5 (5.7%)	5 (6.3%)
CPB time (min)	153.3 ± 50.3	$132.3 \pm 46.9^*$
ACC time (min)	57.5 ± 22.7	$15.4 \pm 20.7^*$
Follow-up (mo)	82.0 ± 25.7	$57.7 \pm 19.7^*$

SV, Single ventricle; TA, tricuspid atresia; PA-IVS, pulmonary atresia with intact ventricular septum; HLHS, hypoplastic left heart syndrome; SP shunt, systemic to pulmonary artery shunt; PAB, pulmonary artery banding; CoA, coactation of aorta; BDG, bidirectional Glenn; PA, pulmonary artery; AVV, atrioventricular valve; SAS, subaortic stenosis; CPB, cardiopulmonary bypass; ACC, aortic crossclamp. Values are mean \pm SD or n (%).

* $P < .01$.

blood sampling every 3 to 6 months regularly. The incidence of early and late deaths, reoperations, and late complications (protein-losing enteropathy, systemic thromboembolism, and supraventricular arrhythmias) was compared between the 2 groups, and the overall incidence of these cardiac-related events was also calculated and compared. In addition, hemodynamic variables obtained from the latest postoperative catheterization and plasma natriuretic peptide type A (ANP) levels were compared. In patients over 6 years of age, treadmill exercise test with the standard Bruce protocol was performed to determine cardiopulmonary response to exercise.

Definition and Evaluation of Supraventricular Arrhythmias

Supraventricular arrhythmias included basal rhythm or any episodes of atrial fibrillation, atrial flutter, junctional rhythm, paroxysmal atrial tachycardia, junctional ectopic tachycardia, sinus node dysfunction including sick sinus syndrome and wandering pacemaker, 2° and 3° atrioventricular block, and frequent atrial ectopic beats (more than 1000 beats per day) on any available ECG and ambulatory Holter monitors. We planned Holter monitoring whenever any supraventricular arrhythmia was detected or suspected with multiple rest and exercise ECGs as well as patient's complaints. In this study, 67 patients (40.1%) had ambulatory Holter monitoring during the follow-up.

TABLE 2. Hemodynamic variables

	LT	EC
Preoperative data		
mPAP (mm Hg)	11.4 ± 3.5	10.8 ± 2.7
v-EDP (mm Hg)	4.9 ± 2.4	5.4 ± 2.8
CI (L/min/m ²)	4.6 ± 1.5	4.1 ± 1.1
Rpl (U/m ²)	1.6 ± 0.8	1.8 ± 0.7
PAI (mm ² /m ²)	330 ± 118	271 ± 172*
Qp/Qs	1.11 ± 0.54	0.92 ± 0.38
Sao ₂ (%)	83.0 ± 4.9	84.7 ± 3.8
Postoperative data		
CVP (mm Hg)	10.7 ± 3.0	10.8 ± 2.6
v-EDP (mm Hg)	4.3 ± 2.4	4.5 ± 2.8
CI (L/min/m ²)	3.4 ± 0.6	3.5 ± 0.6
Qp/Qs	0.97 ± 0.14	0.98 ± 0.08
Sao ₂ (%)	93.1 ± 4.5	94.7 ± 1.6

mPAP, Mean pulmonary artery pressure; v-EDP, ventricular end-diastolic pressure; CI, cardiac index; Rpl, pulmonary vascular resistance index; PAI, pulmonary artery index; Qp/Qs, pulmonary/systemic flow ratio; Sao₂, systemic oxygen saturation; CVP, central venous pressure. Values are mean ± SD.

*P = .01.

Time-Related Changes in the Lateral Tunnel Size

In patients in the LT group who underwent postoperative catheterization more than twice, the time-related change in the lateral tunnel size was investigated. From the lateral tunnel angiogram, the corrected maximum cross-sectional area of the lateral tunnel (max.CSA index) was calculated by the following formula: max.CSA index = πab /body surface area, where a = radius of the widest portion of the lateral tunnel in the anterior-posterior image of the angiogram, and b = radius of the tunnel in the same level as a in the lateral view of the angiogram. The time-related change of the max.CSA index was represented as the percent change in the max.CSA index obtained from the first and the last postoperative cardiac catheterizations.

Statistical Analysis

Data are expressed by mean ± standard deviation. The differences in hemodynamic variables and plasma ANP levels were compared by an unpaired t test. Actuarial survival and freedom from cardiac-related events were estimated by the Kaplan-Meier method with the log-rank test. Univariate and multivariate evaluations for predictors for postoperative supraventricular arrhythmias and cardiac-related events were performed using a multiple logistic regression model. Variables tested in the univariate and multivariate analysis are listed in the appendix. Statistical analysis was performed with SPSS software (version 9.0, SPSS, Inc, Chicago, Ill).

Results

Surgical Outcome

There was 1 hospital death in each group. A 2.6-year-old girl with LT-TCPC developed refractory paroxysmal atrial tachycardia immediately after the operation and died of severe heart failure after 5-day extracorporeal membrane oxygenator support. A 2.9-year-old boy with EC-TCPC died of low cardiac output 32 days after the operation due to

high pulmonary vascular resistance. Late deaths occurred in 5 patients in the LT group and 3 patients in the EC group. In patients with LT-TCPC, the causes of death were upper respiratory infection in 1 patient, protein-losing enteropathy associated with congestive heart failure in 1 patient, hypoxia in 1 patient, and deteriorated ventricular performance with thrombus formation in the intra-atrial tunnel in 2 patients. In patients with EC-TCPC, all 3 patients died of ventricular dysfunction. The overall incidence of death was 6.8% in the LT group and 5.1% in the EC group. The estimated survival at 8 years after surgery was 93.2% in the LT group and 94.9% in the EC group.

Reoperations

There was no reoperation in the EC group, but 7 were performed in the LT group. Two patients who showed severe ventricular failure with thrombus formation in the lateral tunnel underwent take down of TCPC and finally died. Two patients underwent conversion to EC-TCPC because of lateral tunnel dilatation accompanied by supraventricular arrhythmias in 1 patient and pulmonary venous obstruction by the dilated lateral tunnel in 1 patient. After the conversion operation, both patients were relieved from symptoms. For the other 2 patients with moderate right-to-left shunt through the anastomosis of tunnel patches, 1 received direct closure of the leak and the other received conversion to an intra-atrial conduit with a PTFE tube graft. The remaining patient underwent ligation of an abnormal hepatic vein, which connected to the atrium and caused marked systemic desaturation after TCPC. Freedom from reoperation was 91.6% at 8 years in the LT group.

Hemodynamic Assessments

Postoperative cardiac catheterization was performed in 81 of 88 patients (92.0%) in the LT group and 73 of 79 patients (92.4%) in the EC group. There were no differences in the hemodynamic variables at the latest follow-up catheterization between the 2 groups (Table 2). Angiogram of the systemic venous pathway indicated no thrombus formation among survivors in either group and no graft stenosis in the EC group. The latest echocardiogram (LT and EC groups) detected no/trivial atrioventricular valve regurgitation (AVVR) in 64.1% and 53.4%, mild AVVR in 29.5% and 42.4%, and moderate AVVR in 6.4% and 4.1%, respectively. There was no difference in the degree of AVVR between the 2 groups.

Exercise Tolerance Test

A treadmill exercise test with the standard Bruce protocol was performed in 67 patients (76.1%) in the LT group and 50 patients (63.3%) in the EC group. Mean age (years) at the test was 12.8 ± 3.6 in the LT group and 11.3 ± 4.2 in the EC group. A summary of the results is shown in Table 3. When compared with age- and sex-matched control sub-

TABLE 3. Results of exercise tolerance test

	n	EET (minutes)	Rest HR (bpm)	Peak HR (bpm)	Δ HR (bpm)	Peak VO ₂ (mL/kg/min)
LT	67	12.8 ± 2.3	85.1 ± 14.1	166.1 ± 20.2	81.1 ± 22.8	33.4 ± 6.8
EC	50	11.6 ± 1.8	89.7 ± 15.4	174.5 ± 19.1	85.6 ± 23.6	33.8 ± 6.9
<i>P</i> value		.002	.1	.024	.29	.76
Percentage of the age- and sex-matched predicted value						
LT		86.9 ± 13.5	109.1 ± 18.0	90.3 ± 11.0	76.6 ± 21.0	86.6 ± 17.5
EC		83.5 ± 11.0	111.7 ± 20.0	95.5 ± 10.1	83.7 ± 24.0	89.2 ± 19.6
<i>P</i> value		.15	.46	.01	.09	.46

EET, Exercise endurance time; HR, heart rate; Δ HR, heart rate increase; VO₂, oxygen consumption; LT, lateral tunnel; EC, extracardiac conduit.

jects (n = 653), rest heart rate (HR) was higher whereas the remaining indices were significantly lower in both groups. Between the groups, exercise endurance time and peak oxygen consumption were similar, but HR response to exercise was more blunted in the LT group than the EC group. Supraventricular arrhythmias during exercise were recognized in 17.9% in the LT group and 12.0% in the EC group. The number of patients who were treated with beta-blocker was not different (8.9% in the LT group and 8.0% in the EC group).

Cardiac-Related Events

The latest ECG and Holter monitor showed sinus rhythm in 163 of 167 patients and junctional rhythm in 1 patient in each group. Two patients in the LT group had permanent pace makers implanted due to third-degree atrioventricular block. New-onset supraventricular arrhythmias were recognized in 14 patients in the LT group and 4 in the EC group. Multivariate analysis revealed that the type of procedure (LT method, *P* = .003, odds ratio: 16.7) and the preoperative mean pulmonary artery pressure (*P* = .014, odds ratio: 0.61) were significant predictors of postoperative development of supraventricular arrhythmias. Three patients in the LT group developed protein-losing enteropathy with 1 death and 1 was observed in the EC group. Systemic thromboembolism occurred in 1 patient in each group. The overall incidence of cardiac-related events (death, reoperations, supraventricular arrhythmias, protein-losing enteropathy, and thromboembolism) was significantly higher in the LT group (Figure 1), and the estimated predictors for cardiac-related events were the type of procedure (LT method, *P* = .013, odds ratio: 16.6) and postoperative ventricular end-diastolic pressure (*P* = .032, odds ratio: 1.29). Freedom from cardiac-related events was 72.5% in the LT group and 89.8% in the EC group at 8 years after TCPC (*P* = .0098).

Plasma ANP Level

Plasma ANP level was measured in 79 patients (89.7%) in the LT group and 75 patients (94.9%) in the EC group. The ANP level (pg/mL) was 50.5 ± 37.5 (range 10-250) in the LT group and 33.8 ± 21.1 (range 10-110) in the EC group

(*P* < .001, Figure 2). The percentage of patients with values above the normal range (>40 pg/mL) was greater in the LT group (49.4% vs 24.0%).

Time-Related Changes in the Lateral Tunnel Size

In the LT group, serial angiographic data were available from 62 patients (70.5%) to investigate time-related changes in the intra-atrial lateral tunnel size. The interval between the first and the last postoperative cardiac catheterization was 44.2 ± 25.2 months (range 7.9 to 107.8 months). The max.CSA index of the lateral tunnel changed from 7.2 ± 3.6 to 8.4 ± 5.0 (cm²/m²) in 62 patients (*P* = .0005). For patients with intra-atrial baffling method, the index changed from 6.2 ± 2.6 to 6.8 ± 2.6 (*P* = .027), and for patients with autologous tunnel method, the index changed from 10.2 ± 4.7 to 12.1 ± 6.1 (*P* < .0001), respectively (Figure 3). The percent increase in the max.CSA index was 19.4% ± 38.6% (−50.5%–156.2%) during the follow-up. Thirty patients (48.4%) experienced more than 20% increase in size. There was no difference between intra-atrial baffling method and autologous tunnel method.

The Relationship Between the Change in the Lateral Tunnel Size, Plasma ANP Level, and Supraventricular Arrhythmias

Figure 4 shows the relationship between the percent increase in the max.CSA index and plasma ANP level in 62 patients. Open circles indicate patients with new-onset postoperative supraventricular arrhythmias. There was a positive linear relationship between the percent increase in max.CSA index and the plasma ANP level (*P* = .002, *r* = .4). In the LT group, multivariate analysis revealed that significant predictors for the postoperative supraventricular arrhythmias were percent increase in max.CSA index (*P* = .013) and preoperative pulmonary artery pressure (*P* = .014). Length of time after TCPC operation had a marginal correlation (*P* = .06). The incidence of supraventricular arrhythmias was not different between intra-atrial baffling method and autologous tunnel method (13.3% vs 25.0%; *P* = .36).

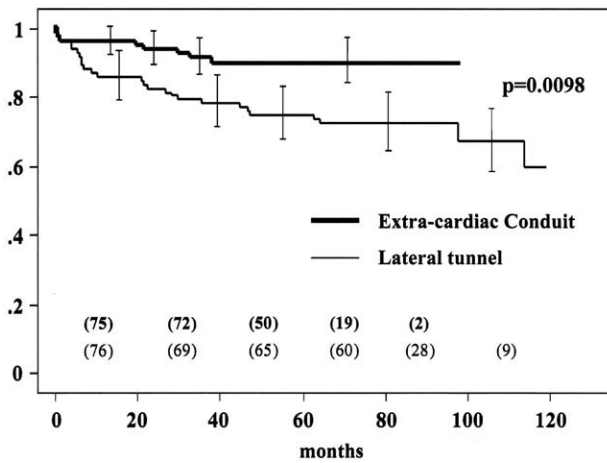


Figure 1. Kaplan-Meier estimated overall freedom from cardiac-related events. "Cardiac-related events" includes death, reoperations, and postoperative complications (protein-losing enteropathy, systemic thromboembolism, and supraventricular arrhythmias). Error bars indicate 95% confidence interval. Numbers of patients at risk are shown in parentheses. Overall freedom from cardiac-related events was lower in LT-TCPC ($P = .0098$).

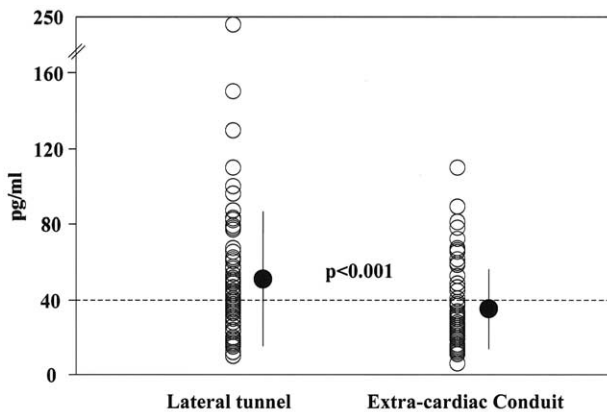


Figure 2. Comparison of the plasma ANP concentration. The values were significantly higher in the LT-TCPC group ($P < .001$). Error bars indicate 1 standard deviation.

Discussion

Since de Leval proposed the TCPC as an alternative procedure to the Fontan-type operation,¹⁰ several modifications have developed to obtain more efficient Fontan circulation.^{9,11,12} However, clinical information regarding the long-term outcomes after these procedures is still limited. Stamm and associates⁵ reported long-term results of the LT-TCPC, showing an estimated survival of 91% at 10 years with good functional status. Our previous report⁷ and reports by others^{6,13,14} showed excellent early and midterm outcome of EC-TCPC with low mortality and morbidity. A

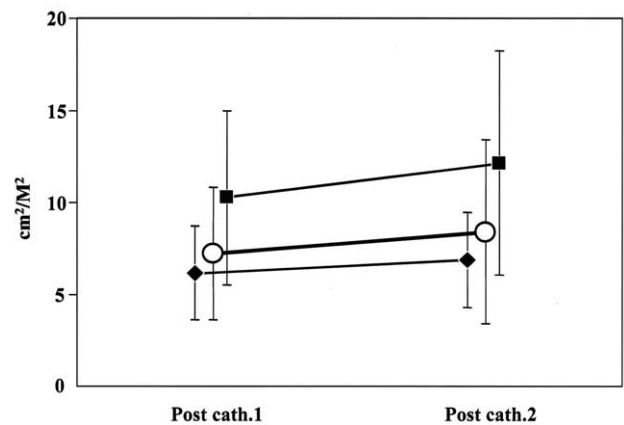


Figure 3. Time-related changes in the maximum cross-sectional area/body surface area of the lateral tunnel (max. CSA index) in 62 patients with LT-TCPC. The index increased from 7.2 ± 3.6 to 8.4 ± 5.0 (cm^2/m^2) in all the 62 patients (open circles), from 6.2 ± 2.6 to 6.8 ± 2.6 in patients with "intra-atrial baffling" (closed diamonds), and from 10.2 ± 4.7 to 12.1 ± 6.1 in the patients with "autologous tunnel" (closed squares). The mean interval was 44.2 months. Post cath.1, The first catheterization after TCPC; Post cath. 2, the last catheterization after TCPC.

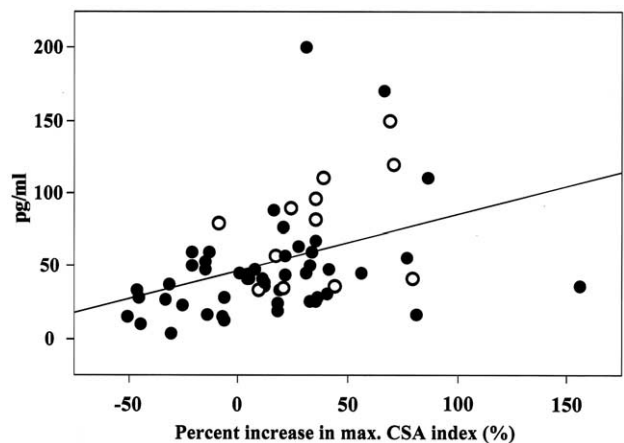


Figure 4. The relationship between the percent increase in maximum cross-sectional area/body surface area of the lateral tunnel (max. CSA index), plasma ANP level, and postoperative supraventricular arrhythmias. There was a positive linear relationship between the percent increase in max. CSA index and the plasma ANP level ($P = .002$, $r = .4$). Open circles indicate patients with supraventricular arrhythmias.

recent report by Azakie and associates⁸ compared these 2 TCPC procedures performed at a single center in terms of early and midterm outcome. However, more clinical information is necessary to discuss the long-term benefits and problems of these modifications.

In this study, midterm survival, postoperative hemodynamic variables, and exercise tolerance were equally satis-

factory in the both groups; however, the incidence of cardiac related events was significantly higher in the LT-TCPC group as compared with the EC-TCPC group. The incidence of thromboembolism and protein-losing enteropathy was low in the both groups; we cannot comment on the influence of the type of procedure on these late complications. However, the incidence of supraventricular arrhythmias was higher in patients with LT-TCPC, and multivariate analysis revealed that the LT procedure itself was a predictor of postoperative supraventricular arrhythmias. This result concurs with the recent reports from Azakie and colleagues⁸ and Ovroutski and colleagues¹⁵ that LT-TCPC is an independent predictor of early and intermediate postoperative atrial arrhythmias and that EC-TCPC decreases these arrhythmias. With regard to the cases of reoperation in the LT-TCPC group, 4 reoperations (2 cyanotic patients with baffle leak and 2 symptomatic patients with dilated lateral tunnel who underwent EC-TCPC conversion) could theoretically have been avoided if EC procedure was employed at the TCPC completion. Thus, midterm cardiac-related morbidities were more frequent in the LT-TCPC group, with a higher incidence of supraventricular arrhythmias and reoperations.

Cardiopulmonary responses to exercise were subnormal in our patients with TCPC, consistent with former reports.^{16,17} However, we could comment that over 80% normal of exercise responses in our patients with TCPC are not pessimistic results. The exercise endurance time and peak oxygen uptake did not differ between the patients with LT-TCPC and patients with EC-TCPC; however, HR response to exercise was lower in the LT-TCPC group. Butera and associates¹⁸ reported that patients with TCPC have significantly reduced HR variability and a low vagal drive. Ohuchi and associates¹⁷ reported that HR response to exercise was closely related to cardiac autonomic nervous activities and these were severely impaired in Fontan population. Thus abnormalities in the neural response seem to exist in patients with TCPC; however, it is possible that more blunted HR response to exercise in patients with LT-TCPC is attributable to increased surgical damage to intra- and extracardiac autonomic nervous system as well as damage to sinus node and sinus node artery. In addition, Ishikawa and associates¹⁹ reported abnormal neurohumoral responses, including ANP, to exercise in patients after Fontan operation. It is known that ANP is a major depressor with inhibitory actions to the pressors like norepinephrine and others²⁰ and, in this study, plasma ANP level was higher in the LT-TCPC group than in the EC-TCPC group. Therefore, it is also possible that lower HR response in the LT-TCPC group is partly due to an elevation of ANP production.

A number of studies have been done to investigate the unique feature of the Fontan circulation, and it is well known that the efficiency of systemic venous blood flow

largely affects the Fontan circulation. Fogel and associates²¹ showed that the driving force for the systemic venous blood to move into the pulmonary circulation was largely cardiac-dependent. Ascutto and associates²² implied that pressure loss from energy dissipation caused by flow turbulence within the systemic venous pathway could impair cardiac performance when a single ventricle must support both systemic and pulmonary circulations. This kind of flow turbulence may occur when the intra-atrial lateral tunnel dilates markedly. According to the energetics, EC-TCPC can provide more energetically efficient Fontan circulation because of the lack of disparity in the cross-sectional area of the systemic venous pathway. Indeed, Lardo and coworkers²³ showed that fluid power loss in the systemic venous pathway was significantly lower with EC-TCPC compared with LT-TCPC.

Our study revealed that 48% of patients with LT-TCPC showed more than 20% increase in the lateral tunnel size over time. In addition, there was a positive linear relationship between the lateral tunnel dilatation ratio and the plasma ANP level. This represents that patients with dilating lateral tunnel have a greater stretch of the atrial wall. This finding should be applied to Laplace's Law, which states that the larger the vessel radius, the larger the wall tension required to withstand a given internal pressure. Moreover, postoperative supraventricular arrhythmias in the LT-TCPC group were more frequent in patients with higher dilatation ratio and plasma ANP level, and multivariate analysis revealed the dilatation ratio of the lateral tunnel as a predictor of development of postoperative supraventricular arrhythmias. Thus our results imply that chronic stretch of the atrial wall could be another arrhythmogenic stimulus in LT-TCPC, although the lateral tunnel suture line alone could provide an electrophysiologic substrate for atrial flutter.^{24,25}

In addition to the advantages of EC-TCPC mentioned above, our experience and that of others of successful conversion of failed LT-TCPC to EC-TCPC^{26,27} can provide an additional rationale for the use of the EC-TCPC as a procedure of choice. However, EC-TCPC contains several unresolved potential disadvantages. Thrombus formation, related to both the use of foreign material and a hypercoagulability state after the Fontan-type operation,²⁸ is a major concern because entire endothelialization could never be expected for a PTFE tube graft.²⁹ We should also carefully monitor the possible risk of distortion of the artificial graft associated with the patient's growth.

Study Limitation

Accurate volumetry is crucial to discuss the time-related change in the intra-atrial lateral tunnel; however, we had no method to measure the actual volume of this complexly shaped venous pathway. We did not demonstrate that the

maximum cross-sectional area of the lateral tunnel accurately reflects the entire volume of the tunnel; however, in most cases, the shape of the lateral tunnel was almost unchanged, even when there was a time-related tendency of dilatation. Therefore, we propose adoption of this parameter to monitor the time-related changes in the overall volume of this pathway. Another limitation is the retrospective nature of this study. In evaluating postoperative arrhythmias, we did not perform ambulatory Holter monitoring in all the patients but limited our study to those who were suspected to have significant arrhythmias according to multiple rest and exercise ECGs and patient complaints. However, we believe that the fact that only 15.8% of the patients receiving the Holter test had significant arrhythmias supports the validity of our findings.

Conclusion

In conclusion, our data indicate the clinical advantages of EC-TCPC in terms of incidence of cardiac-related events, although midterm survival, hemodynamic variables, and exercise tolerance were similar and satisfactory in both patients with LT-TCPC and patients with EC-TCPC. In patients with LT-TCPC, careful observation will be required to monitor the relationship of the tunnel dilatation with late complications, especially supraventricular arrhythmias.

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Appendix**Variables Tested in Univariate and Multivariate Analysis**

Demographics

Age at TCPC
Sex
Time since TCPC

Diagnosis

Single ventricle
Tricuspid atresia
Pulmonary atresia with intact ventricular septum
Hypoplastic left heart syndrome

Heterotaxy syndrome

Morphology of ventricle

Right
Left

Prior procedure

Systemic-pulmonary artery shunt
Pulmonary artery banding
Coarctation repair
Atrial septectomy
Pulmonary artery reconstruction
Atrioventricular valve repair
Bidirectional Glenn procedure
Anomalous pulmonary venous return repair
Subaortic stenosis relief

Preoperative hemodynamics

Arterial oxygen saturation
Mean pulmonary artery pressure
Mean systemic venous pressure
Ventricular end-diastolic pressure
Cardiac index
Pulmonary vascular resistance
Pulmonary artery index (Nakata index)
Pulmonary flow/systemic flow ratio
Atrioventricular valve regurgitation

Type of TCPC

Intra-atrial lateral tunnel
Extracardiac conduit

Cardiopulmonary bypass time

Aortic crossclamp time

Concomitant procedure

Atrial septectomy
Pulmonary artery reconstruction
Atrioventricular valve repair
Anomalous pulmonary venous return repair
Subaortic stenosis relief

Postoperative hemodynamics

Arterial oxygen saturation
Mean systemic venous pressure
Ventricular end-diastolic pressure
Cardiac index
Atrioventricular valve regurgitation

Plasma atrionatriuretic peptide level

Percent increase in maximum cross-sectional area index (only for the LT group)