Case Report

Single-Stage Total Cavopulmonary Venous Connection in Adults: Three Cases Report Describing the Early Results of This Procedure

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Background: The modified Fontan procedure has routinely been performed in children in the present study hospital. Recently, some adults with functionally univentricular hearts were potential candidates for a Fontan completion.

Objective: To review the early results of the single stage Fontan procedure in three adult patients in the last two years.

Case Report: There was no mortality. Re-operations were observed in two patients. The mean extubation time, ICU stay, and hospital stay were three days, five days, and ten days, respectively. The median post-operative oxygenation was 95%. The NYHA class improved significantly in all patients.

Conclusion: Single stage Fontan completion can be performed in carefully selected adult patients. The present report is the early results in three consecutive patients over the last two years. The long-term outcome needs to be followed. The authors hope to enroll more adult patients with univentricular heart and to determine the criteria for a potential adult candidate in the future.

Keywords: Single-stage total cavopulmonary venous connection; Adults; Fontan procedure; Single ventricle; Thailand

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It has been almost five decades since Fontan and Baudet introduced the first successful Fontan procedure in 1971[1]. Since then, this procedure has been modified and ubiquitously accepted for single ventricle palliation. It aims to directly divert venous return to the pulmonary artery, which can alleviate cyanosis and ventricular volume overload. One of the ten requirements that need to be strictly enforced include an age older than 4 years but younger than 15 years. However, this has been modified to pursue the Fontan operation in most selected patients. Nowadays, the ideal age to undergo the operation has been thought to be between 18 months and six years[2-6]. Patients that underwent the Fontan completion in adulthood are universally considered to be susceptible to unsatisfactory results[7-9]. The Fontan procedure, completed in early childhood, is to avoid long-term volume overload and cyanosis causing atrioventricular valve regurgitation and myocardial fibrosis, as well as to preserve systemic ventricular function in the long run[10]. The extracardiac conduit Fontan originally introduced by Marcelletti et al[11] in 1990 and adopted in centers all around the world, has become the most popular Fontan procedure in the present study institute. In Thailand, some of the single ventricle patients has become adult with either first stage palliation or without any previous interventions. The present study institution has recently reassessed and pursued single stage Fontan operation in adulthood. The purpose of the present study was to examine the clinical course and early outcome of three adult patients who underwent single-stage total cavopulmonary venous connection (TCPC) in the present study institute.

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Case Report

Between March 2018 and May 2019, three consecutive adults with single ventricle physiology underwent a single-stage Fontan operation.

Institutional Review Board approval for this retrospective clinical study was obtained [SIRB No. 493/2562(EC2)]. The hospital medical records, operative reports, cardiac catheterization data, and non-invasive studies on all patients were reviewed.

Case presentations

Their mean age was 22 years with a range of 16 to 28 years. The first patient (number 1) had known to have cyanotic heart disease since childhood, but her family had socioeconomic problems. She was referred to the present study institute when she was 24 years old and was diagnosed with heterotaxy syndrome, unbalanced atriventricular septal defect with hypoplastic left ventricle, moderate atrioventricular valve regurgitation, and severe pulmonary stenosis. The second patient (number 2) was diagnosed with double inlet left ventricle, common ventricle with left ventricle morphology, and coarctation of aorta. She had developed cyanosis at two months of age. She underwent persistent ductus arteriosus (PDA) ligation and coarctation of aorta (COA) repaired at 21 months of age and subsequently underwent pulmonary artery banding three days later at the referral center. She had also brain abscess that required craniectomy and drainage at five years old. She was initially considered to be unsuitable for the Fontan palliation and became adult. Therefore, she was referred to the present study institute for continuing of care. The third patient (number 3) was diagnosed with tricuspid atresia and severe pulmonary stenosis. She underwent right modified Blalock Taussig shunt and subsequently underwent left modified Blalock Taussig shunt owing to previous shunt thrombosis during childhood at the present study institute. However, she had lost medical attention since then.

All patients were in the New York Heart Association (NYHA) class II-III preoperatively. Preoperative arterial oxygen saturations (SpO_2) were 60%, 82%, and 82%, respectively. Demographic, echocardiographic, hemodynamic data are shown in Table 1. All patients presented with progressive deterioration of functional capacity with signs and symptoms dominated by dyspnea and cyanosis.

Table 1. Summary of pre-operative patient data

	Patient No. 1	Patient No. 2	Patient No. 3
Age	25	16	28
Prior operation	None	Coarctectomy, PAB	RMBTS, LMBTS
NYHA class	II-III	II-III	II-III
SpO_2 (%)	60	82 to 89	82 to 85
EF (%)	58	55	68
Mean PAP (mmHg)	16	17	17 to 18
Ventricular EDP (mmHg)	12	10	15
PVRi (Wood unit·m^2)	2.12	2.38	0.17
AVVR	Moderate	No	No
PA abnormalities	No	Severe	Moderate

PAB=pulmonary artery banding; RMBTS=right modified Blalock-Taussig shunt; LMBTS=left modified Blalock-Taussig shunt; NYHA class=New York Heart Association (NYHA) Functional Classification; SpO_2=oxygen saturation; EF=systemic ventricular ejection fraction; PAP=pulmonary arterial pressure; EDP=end diastolic pressure; PVRi=pulmonary vascular resistance index; AVVR=atrioventricular valve regurgitation; PA=pulmonary artery

Surgical techniques

The procedures were performed with the patient under moderate to deep hypothermic cardiopulmonary bypass. The second patient needed a brief period of circulatory arrest for a difficult pulmonary arterioplasty owing to severely bilateral proximal pulmonary artery stenosis with limited exposure. The third patient required several short periods of circulatory arrest for an extensive pulmonary arterioplasty. Aortic cross-clamping was applied with administration of intermittent antegrade blood cardioplegia in all patients.

The surgeon had performed 4-mm fenestrated extracardiac Fontan using 24-mm polytetrafluoroethylene (PTFE) conduit in all patients. A fenestration had uniformly been created by 4-mm coronary punch making a hole incorporated to a superior opening of the atrium with dunk technique. The surgeon left a superior opening after partially closed the atrium, which was separated from the inferior vena cava. However, the surgeon needed to revise a thrombosed conduit in the third patient and decided to abandon a fenestration when revisited at the time of re-operation. The surgeon felt that the patient did not need fenestration because she was able to maintain high SpO_2 with low Fontan pressure before a conduit thrombosis happened. The surgeon also reduced conduit size to 22-mm PTFE, which seemed to be more suitable for the right pulmonary artery opening after extensive pulmonary arterioplasty with bovine pericardial patch previously done.

Results

There was no mortality in the present study. Patient 1 required atrioventricular valve repair due to moderate regurgitation. Patients 2 and 3 needed pulmonary arterioplasty using bovine pericardial

Table 2. Peri-operative variables and adverse events

	Patient No. 1	Patient No. 2	Patient No. 3
Associated procedure during TCPC			
Atrioventricular valve repair	Yes	No	No
Main pulmonary division	Yes	Yes	Yes
Left superior vena cava ligation	Yes	No	No
Pulmonary arterioplasty	No	Yes	Yes
Cardiopulmonary bypass time (minute) (1st, 2nd time)	230	322, 104	243, 155
Aortic cross-clamping time (minute) (1st, 2nd time)	140	179, 51	181, 84
Circulatory arrest time (minute) (1st, 2nd, 3rd, 4th time)	-	2	8, 1, 4, 20
Duration of mechanical ventilation (days)	1	4	4
ICU length of stay (days)	1	7	7
Hospital stay (days)	7	15	15
Chest tube drainage >7 days	No	Yes (13 days)	Yes (14 days)
Reexploration for packing removal	No	Yes	No
Reexploration for circuit trombosis	No	Second re-operation for suspected conduit thrombosis	Yes

TCPC=total cavopulmonary venous connection; ICU=intensive care unit

patch. Associated procedures during TCPC and other operative variables are shown in Table 2. There were no significant arrhythmias postoperatively. All patients were treated with an oral anticoagulant therapy based on the international normalized ratio range 2 to 3.

Patient 2, who underwent pulmonary artery banding during childhood and had severe pulmonary artery distortion, developed prolonged pericardial and pleural effusion postoperatively. Early cardiac catherization four weeks postoperatively was performed and found to have 29 mmHg of pulmonary artery pressure and residual proximal left pulmonary artery stenosis. Even though there was no significant gradient between the right and left pulmonary arteries or in the Fontan pathway, a stent was placed to correct some residual stenosis. Transesophageal echocardiography demonstrated 35 to 40% ventricular systolic function. The cardiologist adjusted the dose of diuretics and increased sildenafil. Follow-up echocardiogram revealed markedly improved ventricular contraction and the effusion resolved, but the pulmonary artery pressure and resistance were still high three months after surgery (Table 3). She was prescribed endothelin receptor antagonist (bosentan), and her sildenafil was increased. At one year postoperatively, she was the NYHA functional class I-II with good clinical status. However, two years after surgery, she was hospitalized for leg edema and watery diarrhea, found to have protein losing enteropathy, which was confirmed by albumin scan. Her symptoms were improved by symptomatic treatment with diuretics, diet supplements, intermittent albumin infusion, and octreotide administration.

Table 3. Summary of post-operative patient data

	Patient No. 1	Patient No. 2	Patient No. 3
NYHA class	I	II	I
SpO_2 (%)	95	92 to 94	97
EF (%)	58	50	68
Mean PAP (mmHg)	NA	22*	NA
PVRi (Wood unit·m^2)	NA	6.78*	NA
AVVR	Moderate RAVVR, Trivial LAVVR	Mild	No

NYHA class=New York Heart Association Functional Classification; SpO_2=oxygen saturation; EF=systemic ventricular ejection fraction; PAP=pulmonary arterial pressure; EDP=end diastolic pressure; PVRi=pulmonary vascular resistance index; AVVR=atrioventricular valve regurgitation; RAVVR=right atrioventricular valve regurgitation; LAVVR=left atrioventricular valve regurgitation; NA=not available

* Cardiac catheterization was performed for the second patient at 3 months post-operatively

It should be noted that reoperation was required for patients 2 and 3. The surgeon encountered extremely difficult pulmonary arterioplasty in patient 2. Furthermore, prolonged bypass time and use of hypothermic circulatory arrest led to coagulopathy accompanied by uncontrolled bleeding. The surgeon finally applied BioGlue (CryoLife Inc., Kennesaw, GA) and packed the pulmonary artery with gauze, but she developed hypotension, increased deoxygenation, and increased pulmonary artery pressure six hours

later. The surgeon suspected that these were the result from gauze compression, so, mediastinal re-exploration was performed to remove the packing, but the patient did not sufficiently improve. Transthoracic echocardiography did not reveal any evidence of thrombosis in the Fontan pathway. The patient was returned to the operating theater and found that the BioGlue was forming like rigid material and compressed the pulmonary artery. The surgeon also revisited a Fontan conduit by disconnecting the pulmonary artery anastomosis but found no evidence of any significant thrombus that causing obstruction. However, the surgeon trimmed a conduit that seemed to be too long before suturing it back to the pulmonary artery. Her hemodynamics markedly improved after the second reoperation. Interestingly, patient 3 developed acute Fontan conduit thrombosis the morning after the operation. The surgeon hypothesized that this may have been caused by a mismatch between the 24 mm Gore-Tex graft and the pulmonary opening previously augmented with an extensive bovine pericardial patch. Another factor that could have been a contributing role was the aggressive use of blood components and prothrombin complex concentrates during and after surgery. Accordingly, the surgeon switched the conduit to 22 mm Gore-Tex graft, and enlarged the pulmonary artery opening, which was carried further medially behind the aorta. After reoperation, her postoperative course was complicated by pleural effusion, which was treated, and she was able to go home on postoperative day 15.

Discussion

Nowadays, there are still encounters with univentricular heart patients who have survived to be adults. These patients have reached adulthood without Fontan palliation or even bidirectional Glenn operation because of several reasons:

1. There was no medical attention after diagnosis since childhood or after undergoing first stage palliation.
2. The patients were initially thought to be unsuitable for any further operations.
3. Avoiding performing Fontan palliation because of high risk to benefit ratio.
4. Referring from other centers due to some reasons.

As mentioned in literatures, the authors have pursued a Fontan procedure as early as possible to alleviate cyanosis and ventricular volume overload[12,13]. The authors believe this strategy can preserve ventricular function resulting in better long-term outcome. Therefore, adults with univentricular heart who have not received to staged palliation in a timely manner were typically considered not a good candidate. Neglected adult patients would develop ventricular hypertrophy with elevated left ventricular end-diastolic pressure[4,14], and increased pulmonary vascular resistance resulting from several causes[5,15,16]. However, the authors' strategy was to reconsider the procedure for adult patients by re-evaluation. The selected patients were based on pulmonary pressure and resistance and the rigid numbers should be compromised to enroll a potential Fontan candidate. All patients had mean pulmonary artery pressure of more than 15 mmHg. The first and second patients had pulmonary vascular resistance of more than 2 Wood unit·m², but all were less than 3. The second patient had severe distorted pulmonary bifurcation because of a long-standing migrated pulmonary artery band.

Prolonged pleural effusions, defined as effusions that required chest tube drainage for more than 14 days, happened in the second and third patients. Prolonged pleural effusion could be derived from preoperative low normal ejection fraction, 17 mmHg of mean pulmonary artery pressure, pulmonary artery abnormality requiring extensive pulmonary arterioplasty, reoperation, non-functioned fenestration, and prolonged bypass time. The patient number 2 also needed stent placed in the left pulmonary artery when she was re-admitted owing to recurrent pleural effusion.

In term of anticoagulation, it needs to be justified in every patient. The cardiologist decided to give warfarin in all patients for many reasons:

1. Maintaining patency of fenestration
2. A consequence of borderline hemodynamics
3. Event of circuit thrombosis

However, the present report revealed the early result of single stage Fontal completion in only three adults. The first and third patients were in NYHA class I and the second patient was in NYHA class II. All patients have markedly improved in quality of life as well as functional status. According that increasing disability is to be anticipated in this group as time from operation extended[17], the second patient, who did not initially seem to be a perfect candidate for Fontan palliation, got protein losing enteropathy very shortly after Fontan completion. Her symptoms have been improved by symptomatic treatment comprising diuretics, diet supplements, intermittent albumin infusion, and octreotide administration. The authors needed to optimize her hemodynamics. Ultimately, she may need a cardiac transplant.

Conclusion

Single stage Fontan completion can be cautiously performed in carefully selected adult patients. Relaxing the rigid criteria, strictly followed in the past, to include patients who could potentially be a candidate for Fontan completion is possible. According to the reports from several centers[9,10,18-20], the Fontan operation can be accomplished in adult patients with good outcomes. However, mortality and complications are slightly higher than those seen in pediatric population. This is a preliminary report of current experience of the single stage Fontan completion in the authors' institute. The patient selection is the key. Therefore, carefully defining the criteria for adult patients must be done to obtain favorable outcomes.

What is already known on this topic?

Single-stage Fontan is feasible to perform in selected adult patients.

What this study adds?

The authors reported three adult cases of Fantam operation on univentricular hearts and suggest being extremely cautious to perform single-stage Fontan in some patients.

1. Delayed stage operation after pulmonary artery banding resulting in inappropriate protection of pulmonary vascular bed.

2. Severe distorted pulmonary artery resulting in prolonged operative time, bleeding, and decreased ventricular function.

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Conflicts of interest

All authors declare no personal or professional conflicts of interest, and no financial support from the companies that produce or distribute the drugs, devices, or materials described in this report.

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