Outcomes of Straightforward Extracardiac Fontan Operation in Advanced-Age Group

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Introduction: *Fontan operation is a palliative procedure used in patients with univentricular heart (UVH). There have been numerous modifications to the technique. The appropriate strategy for treating patients with UVH in Thailand is still a major concern.*
Objective: *To describe the outcomes of patients after straightforward Fontan operation at advanced-age and evaluate their post-surgical cardiac performance.*
Material and Method: *A cross sectional study was performed in post-Fontan operation patients at Rajvithi Hospital between 2000 and 2009. Post-operative cardiac performance was analyzed using the echocardiogram and treadmill exercise stress test (EST).*
Results: *Thirty-nine patients were enrolled in the present study. The mean age was 11.7 years and the mean age at operation was 7.4 years. The predominant systemic chamber was morphologic left ventricle. Straightforward Fontan operation was performed in 60% of cases. Mean of Fontan index, mean of McGoon ratio, and mean of Nakata index were 2.63, 2.32, and 414.15 sqmm/sqm, respectively. Mean of the PVR and PAP were 1.98 U/m^2 and 11.05 mmHg. Eighteen percent of patients died in the early post-operative period. Most of the patients died from septicemia. The mean EF was 58.43 %. Systolic dysfunction in 17.9% of the cases was abnormal, whereas diastolic dysfunction was present in half of the cases. Thirty-nine percent had MPI abnormality. During EST, three cases developed arrhythmias and 3 cases had hypotension. Patients were categorized into 3 groups (Group 1: normal systolic and diastolic functions, Group 2: diastolic dysfunction, Group 3: impairment of both systole and diastole). Pre-operative cardiac catheterization parameters, surgical data and cardiac performance showed no statistical significance. However, there was a significant correlation between those with abnormal cardiac performance and arrhythmias or hypotension during EST (p = 0.003).*
Conclusion: *The selection of suitable cases and good pre-operative evaluation could decrease the morbidity and mortality in patients undergoing the Fontan procedure. This study also found a correlation between abnormal cardiac performance and transient cardiac arrhythmia during exercise. The evaluation of cardiac performance and EST remains to be performed for following-up of patients who have undergone the Fontan operation, even for the asymptomatic cases.*

Keywords: *Post, Fontan operation, Outcome*

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Univentricular heart (UVH) is an anomaly in which all blood flows through one ventricle or the atrioventricular (AV) valves empty blood into only one chamber in the ventricular mass. Of the children affected with cardiac problems in Thailand, 80 percent were congenital, with 30 percent of these being cyanotic. Of the cyanotic cases, 30 percent suffer from UVH[1].

In 1971, Francis Fontan reported a new approach to the surgical palliation of tricuspid atresia[2]. Since its original description, the Fontan operation has undergone numerous modifications. The modified Fontan operation such as the extracardiac conduit has become the treatment of choice for patients born with

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UVH, with favorable results[3-9].

A bidirectional Glenn shunt has been used as an intermediate palliative step prior to the Fontan operation. The advantage of this palliative surgery is decreased volume load on the ventricle. Some surgeons prefer the straightforward Fontan procedure in order to prevent complications later, after the procedure.

The patient's age for undergoing the Fontan operation has been decreasing steadily. Several centers tend to operate on patients with favorable anatomy and physiology at the age of 2 years, or even younger. In contrast to our center, the patients' age was relatively older, because many patients receive late medical care and there is usually a long waiting list for complex lesion surgeries.

Since there is currently limited information on the outcomes of patients after the Fontan operation for advanced-age patients, further investigation on these particular cases may lead to appropriate strategies for treating UVH in Thailand.

Objective

The objectives of the present study are to describe the outcomes of a group of patients with UVH who had straightforward Fontan operation at an advanced-age and to evaluate their post-surgical cardiac performance.

Material and Method

A cross sectional study was performed on post-Fontan operation patients at Rajvithi Hospital by a single surgeon between 2000 and 2009. Post-operative cardiac performance was evaluated by a single investigator using two different methods: echocardiogram and treadmill exercise stress test (EST). Patients with a pacemaker were excluded from the study.

The standard protocol for echocardiographic assessment were as follows: 1) Evaluation of ventricular systolic function by Simpson's method, 2) Assessment of diastolic function by conventional pulse wave (PW) Doppler measurement at pulmonary vein, mitral valve and/or tricuspid valve, 3) Assessment of the tissue using Doppler Imaging (TDI) by S', E', A', isovolemic contraction time (IVCT), isovolemic relaxation time (IVRT), and E/E', and 4) Evaluation of myocardial performance index (MPI) by tracking tissue Doppler signal at medial and lateral aspects of the AV-valve[10-14]. The staging of diastolic dysfunction was determined by the characteristics of wave forms according to previous studies[15-18].

EST was performed using the modified Bruce protocol. Maximum heart rate, blood pressure, oxygen saturation, rhythm disturbance, and patient status were continuously monitored throughout the test. The patients' well-being was evaluated by a questionnaire to determine the New York Heart Association (NYHA) functional class and events after surgery, including syncope, chest pain, palpitation and leg/face edema.

According to the characteristics of their cardiac performance, patients were then categorized into four groups. Group 1 comprised of patients with normal systolic and diastolic functions. Group 2 comprised of patients with exclusive systolic dysfunction. Group 3 comprised of those who solely had diastolic dysfunction, and Group 4 comprised of cases with combined systolic and diastolic dysfunctions.

Statistical analysis

Statistical analysis was performed by SPSS version 16.0. For continuous variables, data was presented as mean $\pm$ standard deviation (SD). For categorical variables, data were presented as numbers and percentages. One way ANOVA/Kruskal-Wallis test was used for the comparison of mean between each group. Differences in frequency of events among patient groups were analyzed by Chi-square test. The *p*-value <0.05 is considered to be statistically significant.

Results

Thirty-nine patients were enrolled in the present study (Table 1). The mean age was 11.7 ± 5.2 years and the mean age at operation was 7.4 ± 4.1 years. The youngest age at operation was 2 years and the oldest was 18 years. Male gender predominated the study population (66.7%). Complex double outlet of the right ventricle (DORV) was the most common diagnosis (30.8%). The predominant systemic chamber was morphologic left ventricle (82.1%). Sixty percent of the cases underwent straightforward total cavopulmonary anastomosis, while 24% received a Blalock-Taussig shunt as palliative treatment prior to receiving the Fontan procedure. Extracardiac tunnel with fenestration was the most common type of Fontan modification (71.8%). The mean duration of follow-up after Fontan procedure was 3.4 ± 2.9 years. Pre-operative cardiac catheterization parameters are demonstrated in Table 2.

The overall mortality rate was 20.5% (8 out of 39 cases). Early post-operative morbidity and mortality

Table 1. The demographic data

	n (%)
Gender	
Male	26 (66.7)
Female	13 (33.3)
Diagnosis	
TA	9 (23.1)
Unbalanced AVSD	8 (20.5)
Complex DORV	12 (30.8)
DILV	4 (10.3)
Others	6 (15.4)
Predominant chamber	
Left ventricle	32 (82.1)
Right ventricle	7 (17.9)
Previous surgical operation	
None	24 (61.5)
RMBTS	6 (15.4)
LMBTS	3 (7.7)
PAB	3 (7.7)
BDGS	3 (7.7)
Type of fontan operation	
Extra-cardiac without fenestration	4 (10.3)
Extra-cardiac with fenestration	28 (71.8)
Others**	7 (17.9)

TA = tricuspid atresia; Unbalanced AVSD = unbalanced atrio ventricular canal defect; Complex DORV = complex double outlet of right ventricle; DILV = double inlet of left ventricle; RMBTS = right modified Blalock-Taussig shunt; LMBTS = left modified Blalock-Taussig shunt; PAB = pulmonary artery banding; BDGS = bidirectional Glenn shunt

* hypoplastic right ventricle, straddling atrioventricular valve, transposition of great vessel with ventricular septal defect with pulmonary stenosis, severe Ebstein's anormaly

** intra-cardiac Fontan, lateral tunnel Fontan, Kawashima operation

was defined within 30 days after surgery. Eighteen percent of patients died in the early post-operative period and 2.6% died in the late post-operative period (more than 30 days). Most of the patients died from septicemia. None of the deaths was sudden and unexpected death immediately post-operation was due to cardiac arrhythmias. The one remaining late death was a car accident. Two patients were lost during follow-up. One case was excluded from the study due to the patient being on a pacemaker.

In terms of morbidity, re-operation was performed in three cases (7.9%). The first case had residual hepatic sinusoid, the second case suffered from deterioration of ventricular diastolic functions after the surgery, and the last case had progressive ascites, requiring re-operation to take down the Fontan circuit. One case (2.6%) developed protein-losing enteropathy (PLE) 1 year and 6 months after the operation. No thrombosis of the pathway or post-operative pancreatitis was observed in our patients.

The mean ejection fraction (EF) was 58.43$\pm$7.43%. Patients with PLE and significant post-operation AV-valve regurgitation had the minimum EF of 28%. Measurements of conventional PW and TDI echocardiography are demonstrated in Table 3 and 4.

Systolic dysfunction was abnormal in five cases (17.9%), whereas diastolic dysfunction was present in half of the cases (50%). Regarding diastolic abnormalities, 57.1 percent had abnormal relaxation (Stage 1) and 42.9 percent had a pseudo-normalization pattern (Stage 2). Thirty-nine percent had MPI abnormality.

The association between cardiac performance and the type of surgery showed that straightforward Fontan and staged Fontan had 16.66% and 20% systolic dysfunction, respectively.

For EST, the modified Bruce protocol was used. Instead of using conventional exercise testing protocol, Naughton protocol was used for patients with poor left ventricular function and the six-minute walk was used for patients with suspected thrombus formation in the Fontan circuit. One patient with mental retardation was excluded. During the exercise, three cases developed arrhythmias (paroxysmal junctional escape beats, transient atrial fibrillation, and transient third degree atrioventricular block) and three cases had hypotension at peak exercise.

For the post-operative symptoms, NYHA functional class 1 was found in 17 cases (63%) and functional class II was found in 11 cases (37%). None of the patients experienced syncope attacks or chest pain after the surgery. Two cases exhibited palpitations related to exercise. One patient with very poor cardiac function and PLE had frequent leg edema and puffy eyelids.

Patients were categorized into four groups according to cardiac performance. Half of the cases (14/28) had normal systolic and diastolic functions (Group 1), 32 percent had exclusive diastolic dysfunction (Group 2), and 18 percent had impairment of both systolic and diastolic function (Group 3). None of the cases had only systolic dysfunction (Group 4). Among each group, there was no statistical significance between systolic or diastolic dysfunction and the pre-operative cardiac catheterization parameters. In addition, the correlation between the surgical data and metabolic equivalent tasks (METs) with cardiac

Table 2. Pre-operative catheterization data

	Mean ± SD	Minimum	Maximum
Fontan index	2.63±1.30	0.63	8.30
McGoon ratio	2.32±0.47	1.10	3.30
Nakata index (sqmm/sqm)	414.15±177.37	103	915
PVR (U/sqm)	1.98±1.30	0.40	6.90
MPAP (mmHg)	11.05±4.98	3	21
RPA size (mm)	13.6±4.33	2.90	26.60
LPA size (mm)	13.00±4.24	4.40	25.70

PVR = pulmonary vascular resistance; MPAP = mean pulmonary artery pressure; RPA = right pulmonary artery; LPA = left pulmonary artery

Table 3. Measurements of conventional pulse wave Doppler

	Mean ± SD	Minimum	Maximum
Mitral valve inflow velocity			
E (cm/s)	62.26±21.09	23	110
A (cm/s)	52.36±21.20	26	133
E/A ratio	1.40±0.72	0.22	2.89
E wave deceleration time (msec)	165.13±57.77	66.85	308.80
Pulmonary vein flow			
Systolic (cm/s)	52.25±18.48	18	102
Diastolic (cm/s)	66.54±20.93	31	104
S/D ratio	0.82±0.31	0.25	1.48
Atrial reversal (cm/s)	24.28±14.79	11	87
A-duration (msec)	132.53±45.18	59.15	266.17

Table 4. Measurements of TDI

	Lateral wall (n = 28)	Septal wall (n = 12)
S' (cm/s)	6.25±2.00	4.83±2.08
E' (cm/s)	8.07±3.44	6.25±2.66
A' (cm/s)	7.46±2.56	6.83±2.12
IVCT (msec)	79.31±17.80	81.23±34.32
IVRT (msec)	73.47±16.18	79.61±7.09
E/E'	8.72±3.50	10.38±3.68

performance also showed no statistical significance. However, there was a significant correlation between those with abnormal cardiac performance and cardiac arrhythmias or hypotensive episode during EST (p = 0.003).

Discussion

Pre-Fontan operation data

In the present study, the mean operation age was 7.4 years, which is considerably older than the usual 2 to 4 years, due to the constraints of surgical service and late presentation of the patients. Some disadvantages have been reported for late Fontan operation. However, several cardiac surgeons still elect to perform the straightforward Fontan at the advanced-age since they believe in 'the ticking clock theory' that Fontan operation has a limited lifetime and should be postponed to as late as possible[19,20].

For risk stratification, pulmonary vascular resistance (PVR) above 4U/sqm remains the absolute contraindication for completion of the Fontan procedure. Additionally, the Fontan index of less than 4 is a favorable indicator of good outcomes[21]. Some reports showed that McGoon ratio of more than 1.8, mean pulmonary artery pressure (MPAP) of less than 15 mmHg and Nakata index of more than 250 sqmm/sqm are considered to be positive factors[21,22]. For the present study, 8 patients presented with MPAP over 15 mmHg, which may have resulted from high pulmonary blood flow, and one patient presented with high PVR (6.9U/sqm). All of them had immediate post-operative complications.

Table 5. Comparison between cardiac performance and type of surgery

	Straight-forward Fontan n = 18	Staged Fontan n = 10	*p*-value
Systolic function			
Normal	15 (83.3)	8 (80)	0.60
Abnormal	3 (16.6)	2 (20)	
Diastolic function			
Normal	9 (50)	5 (50)	0.65
Abnormal	9 (50)	5 (50)	

Table 6. Comparison of pre-operative catheterization data between groups according to cardiac performance

Parameters	Group 1	Group 2	Group 3	*p*-value
Fontan index	2.29±1.05	2.39±0.73	3.20±1.38	0.22
McGoon ratio	2.45±0.47	1.93±0.45	2.35±0.44	0.05
Nakata index (sqmm/sqm)	407.14±174.03	373.90±107.24	444.20±180.94	0.08
PVR (U/sqm)	1.47±0.90	2.49±1.35	1.55±0.57	0.77
MPAP (mmHg)	9.93±3.66	10.00±5.45	10.40±6.35	0.98
RPA size (mm)	14.00±3.50	12.40±5.19	15.70±6.31	0.44
LPA size (mm)	13.50±3.86	11.5±2.45	14.70±6.76	0.35

PVR = pulmonary vascular resistance; MPAP = mean pulmonary artery pressure; RPA = right pulmonary artery; LPA = left pulmonary artery

Table 7. Correlation between group of the patients and surgical information

Surgical data	Group 1 (n)	Group 2 (n)	Group 3 (n)	*p*-value
Previous surgery				
None	9	6	3	0.80
RMBT	1	2	1	
LMBT	1	1	1	
PAB	1	0	0	
BDG	2	0	0	
Type of Fontan operation				
Extra cardiac without fenestration	0	2	0	0.32
Extra cardiac with fenestration	11	6	4	
Others*	3	1	1	

RMBT = right modified Blalock-Taussig shunt; LMBT = left modified Blalock-Taussig shunt; PAB = pulmonary artery banding, BDG = bidirectional Glenn shunt
*intracardiac Fontan, lateral tunnel Fontan, Kawashima operation

Table 8. Comparison between group of the patients and METs

Parameters	Group 1 (Mean ± SD)	Group 2 (Mean ± SD)	Group 3 (Mean ± SD)	*p*-value
METs	9.48±3.44	8.10±2.50	7.82±4.06	0.42

METs = metabolic equivalent tasks; 1MET = 3.5 mL/kg/min aerobic capacity

Morbidity and mortality

The overall morbidity and mortality in this study was acceptable. A robust pre-operative assessment of ventricular volume, function, and compliance, as well as AV valve function, plays an important role in reducing the morbidity rate. Moreover, prevention and control of hospital-acquired infections was also a major contributor in improving the surgical outcomes.

Cardiac performance post-Fontan operation

Cheung et al found that diastolic function in 23% post-Fontan operation patients was 23%. However, after follow-up, it increased to 77%. Approximately 30-35% of the patients had systolic dysfunction after the operation[23]. In the present study, cardiac dysfunction was lower than previous studies and results showed restrictive physiology, which may be due to the nature of the cross-sectional study and shorter follow-up time.

MPI measurement in the present study correlated well with abnormal ventricular systolic or diastolic function, which is in contrast to previous studies, which found that the MPI did not correlate with the EF[24]. Nonetheless, in the future, evaluation of cardiac function before and after surgery may predict the outcomes of UVH in short- and long-term periods.

Exercise stress test post-Fontan operation

According to previous literatures, cardiac arrhythmia is a common problem during exercise[25]. Interestingly, the results showed significant correlation between abnormal cardiac performance and the presence of cardiac arrhythmias during exercise (p = 0.003). As a result, patients with systolic and/or diastolic dysfunction need close monitoring. Moreover, exercise testing should be performed to unmask silent arrhythmias.

Symptoms post-Fontan operation

Though our patients had surgical repair at an advanced-age, most of the patients were symptom-free in the long-term follow-up, except for two patients who experienced palpitations related to exercise. Interestingly, these two cases also developed transient atrial fibrillation and transient complete heart block during EST.

Conclusion

The outcomes of the Fontan operation studies are different among various institutions and have different issues of concern. However, the patients still require follow-ups in the long term and need to have regular hemodynamic evaluation. Selection of suitable cases and good pre-operative evaluation could decrease the morbidity and mortality in patients who have undergone the Fontan procedure. Results from the present study showed the correlation between abnormal cardiac performance and the transient cardiac arrhythmia during exercise; therefore, the evaluation of cardiac performance and the EST plays an important role in following-up of patients who have undergone the Fontan operation, even for the asymptomatic cases.

Potential conflicts of interest

None.

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การศึกษาผลการรักษาผู้ป่วยหลังการผ่าตัด Straight-forward Extracardiac Fontan ในเด็กโต

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ภูมิหลัง: การผ่าตัด Fontan เป็นการผ่าตัดรักษาโรคหัวใจแต่กำเนิดชนิดเขียวที่มีหัวใจห้องล่างที่สามารถใช้ได้เพียงห้องเดียว การผ่าตัดชนิดนี้ปัจจุบันมีการพัฒนาวิธีการผ่าตัดและแนวทางการรักษาอย่างมาก การศึกษานี้จึงจัดทำขึ้นเพื่อพัฒนาแนวทางการรักษาในประเทศไทยและสร้างแนวทางในการติดตามผู้ป่วยหลังการรักษาอย่างมีระบบ

วัตถุประสงค์: เพื่อศึกษาผลการผ่าตัด straight-forward Fontan ในเด็กโตและ cardiac performance หลังการผ่าตัด

วัสดุและวิธีการ: ทำการศึกษาโดยศึกษาแบบ crossectional ในผู้ป่วยโรคหัวใจชนิดมีอาการเขียวที่ได้รับการผ่าตัด Fontan ที่โรงพยาบาลราชวิถีในช่วง พ.ศ. 2543 ถึง พ.ศ. 2552 โดยศึกษาข้อมูลทั่วไป การทำงานของหัวใจโดยคลื่นเสียงสะท้อนหัวใจ (echocardiogram) และการวิ่งสายพาน (treadmill exercise stress test) หลังการผ่าตัด Fontan

ผลการศึกษา: ผู้ป่วยในการศึกษานี้มีทั้งหมด 39 ราย อายุโดยเฉลี่ยในการศึกษานี้เท่ากับ 11.7 ปี และอายุเฉลี่ยในการผ่าตัดเท่ากับ 7.4 ปี Complex DORV เป็นโรคที่พบบ่อยที่สุดในการศึกษานี้คิดเป็น 30% หัวใจห้องล่างที่ใช้เป็นห้องหลักในการศึกษานี้คือห้องซ้าย การผ่าตัดชนิด Straight-forward Fontan พบเป็น 60% ของการศึกษา และระยะเวลาหลังการผ่าตัดกับการศึกษานี้มีค่าเฉลี่ยเท่ากับ 3.4 ปี ผลการสวนหัวใจก่อนการผ่าตัดผลว่ามีค่าเฉลี่ยของ Fontan index, McGoon ratio และ Nakata index เท่ากับ 2.63, 2.32 และ 414.15 sqmm/sqm ตามลำดับ ค่าเฉลี่ยของ PVR เท่ากับ 1.98 U/sqm และ MPAP เท่ากับ 11.05 mmHg. ผู้ป่วยที่เสียชีวิตหลังการผ่าตัด ไม่เกิน 30 วัน พบเป็น 18% และสาเหตุที่พบมากที่สุดคือการติดเชื้อในกระแสเลือดค่าเฉลี่ยของ ejection fraction เท่ากับ 58.43% ผู้ป่วยที่มี systolic dysfunction พบเป็น 17.9% ส่วนผู้ป่วยที่ diastolic dysfunction พบครึ่งหนึ่งของผู้ป่วยที่ศึกษาและผู้ป่วยที่มี MPI abnormality พบเป็น 39% การวิ่งสายพานพบว่ามีผู้ป่วย 3 ราย มีภาวะหัวใจเต้นผิดจังหวะและผู้ป่วย 3 รายมีภาวะความดันโลหิตต่ำ

การศึกษานี้ได้แบ่งผู้ป่วยออกเป็น 3 กลุ่มโดยการทำงานของหัวใจ (กลุ่มที่ 1: normal systolic and diastolic functions, กลุ่มที่ 2: diastolic dysfunction, กลุ่มที่ 3: impairment of both systole and diastole) ผลการสวนหัวใจก่อนการผ่าตัด และข้อมูลเกี่ยวกับการผ่าตัดก่อนการผ่าตัด Fontan รวมทั้งชนิดการผ่าตัด ไม่มีความสัมพันธ์ทางสถิติกับการทำงานของหัวใจ แต่มีความสัมพันธ์กับการเต้นผิดจังหวะของหัวใจและภาวะความดันโลหิตต่ำขณะออกกำลังกายอย่างมีนัยสำคัญทางสถิติ ($p = 0.003$)

สรุป: การเลือกผู้ป่วยที่เหมาะสมก่อนการผ่าตัดจะช่วยลดอัตราการตายและภาวะแทรกซ้อนหลังการผ่าตัดได้ และการศึกษานี้ยังพบว่ามีความสัมพันธ์ระหว่างหัวใจเต้นผิดปกติและความดันโลหิตต่ำขณะออกกำลังกาย การติดตามผู้ป่วยหลังการผ่าตัด Fontan โดยการประเมินการทำงานของหัวใจโดยการตรวจคลื่นเสียงสะท้อนหัวใจและการวิ่งสายพานจึงมีความสำคัญมาก