The Fontan Operation: Experience at Siriraj Hospital

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Abstract

Seventy five patients underwent modified Fontan operation at Siriraj Hospital from October 1987 to December 1998. Cardiology data was analyzed retrospectively. Four patients' data was unavailable. Median age at operation was 9.7 (1.8-34) years old . Tricuspid atresia accounted for 38 per cent of the patients. Ten patients (14.1%) died in the acute post operative period due to consequence of low cardiac output. Another 3 patients (4.2%) expired in the intermediate and late post operative period. Age at operation, pulmonary artery size, pre-operative oxygen saturation, and mean pre-operative pulmonary artery pressure were not different between those who survived and those who died. Abnormal pulmonary vein, atrioventricular valve regurgitation, and underlying ventricular morphology statistically affected the acute survival of modified Fontan operation. Intraoperative aortic cross clamp time, and post operative mean pulmonary artery pressure on day 0,1 and 2 post operation were found statistically shorter and lower in the survival group. Survival rate at 5 years was 83 per cent. Modified Fontan operation is the final palliative operation of choice for low risk single ventricle physiology in our institution with acceptable outcome. Thorough pre-operative hemodynamic and anatomic studies and staging modified Fontan procedure may include a higher number of candidates and improve the outcome of the operation.

Key word : Fontan Operation, Complex Univentricular Heart

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Fontan and Baudet[1] first described the palliative surgical technique for tricuspid atresia patients in 1971. This has been modified for single ventricle physiology and biventricle physiology with unfavorable anatomy for biventricular repair. Surgical techniques have been developed to atriopulmonary connection, lateral tunnel Fontan, total cavopulmonary connection, and extracardiac Fontan. These techniques may be done in one step with/without fenestration, partial exclusion of hepatic vein or by staging with bidirectional Glenn operation. The essential aim is to minimize the mortality and morbidity[2] e.g. prolonged pleural effusion, cardiac arrhythmia, thromboembolism, protein losing enteropathy, aortopulmonary collateral vessels, pulmonary arteriovenous malformation and cardiac failure. Many studies have analyzed the risk factors of the Fontan operation[3-10] and low risk criteria for modified Fontan operation have been established[11]. At Siriraj Hospital, the favorable criteria have been followed as strictly as possible. Our modified Fontan operation data has not yet been analyzed since the operation was started in 1987. To find out the risk factors and survival rate were the main objectives of this study which is the first and largest study of the Fontan operation in Thailand.

PATIENTS AND METHOD

The cardiovascular surgical database of Siriraj Hospital was reviewed. Between October 1987 and December 1995, 75 consecutive patients underwent a modified Fontan operation. Due to incomplete data, only 71 patients were subjected for the study. Cardiological files, medical records, cardiac catheterization files, cineangiocardiograms, and cardiovascular surgical files were reviewed. Demographic data, preoperative cardiovascular data, operative data, post operative data and follow-up data were collected including the most recent data which was obtained by a questionnaire and cardiological file of the last visit. The data was analyzed statistically to find out the risk factors for acute survival, and the survival rate.

Statistical Analysis

Measured variables were expressed as mean ± SD and/or median (minimum & maximum value) and were compared by using Mann-Whitney U-test. Proportions (in percentages) were computed for categorical data and were compared by Chi-square and Fisher exact test as appropriate. A p-value less than 0.05 was considered significant. Survival function was calculated by the Kaplan-Meier method.

RESULTS

Pre operative data

A total of 75 patients underwent the modified Fontan operation between October 1987 and December 1998. Four patients' data was missing. Male to female ratio was 1.2:1. Median age at Fontan operation was 9.7 years old (range from 1.8 to 34). Underlying heart diseases were tricuspid atresia 38 per cent, complex univentricular heart with unidentified ventricular morphology 19.7 per cent, complex univentricular heart with right ventricular morphology 18.3 per cent, complex univentricular heart with left ventricular morphology 16.9 per cent, complex heart with double inlet left ventricle 5.6 per cent, and double outlet right ventricle physiology 1.4 per cent. Associated cardiovascular abnormalities were bilateral superior vena cava (SVC) 22.9 per cent, atrioventricular valve regurgitation (mild & moderate regurgitation) 9.9 per cent, abnormal pulmonary veins 8.6 per cent, and abnormal systemic vein drainage 7.1 per cent. Demographic, pre operative hemodynamic, operative and post operative data are shown in Table 1. Thirty two patients (44%) had one previous cardiac surgery and another 6 patients (8.8%) underwent two cardiac operations prior to modified Fontan operation.

Operative & post operative data

There were 4 types of modified Fontan operation: 40 (56.3%) lateral tunnel, 23 (32.3%) right atrial appendage- pulmonary artery connection type, 5 (7.0%) extracardiac type, and 3 (4.2%) right atrium-right ventricular-pulmonary artery connection (Bjork). Only eleven had fenestrated modified Fontan. Median time for extubation, inotropic infusion and intensive care stay in the post operative period were 3.3 hours (range from 0 to 32), 4 days (range from 0 to 26) and 3 days (range from 0.2 to 14) respectively. Median chest tube drainage was 7 days (range from 1 to 90). Ten patients (14.1%) died in the acute post operative period (within 30 days) due to consequence of low cardiac output. No death was found in the acute post operative period in tricuspid atresia and complex univentricular heart with left ventricular morphology. There were

Table 1. Demographic, pre-operative and post-operative data.

	Number of Patients	Minimum	Maximum	Mean	Standard Deviation
Wt (kg)	70	9.5	55.0	26.4	12.5
Ht (cm)	65	82.5	173.0	126.8	23.0
BSA (m^2)	65	0.47	1.63	0.94	0.29
Hosp stay (days)	58	11.0	207	54.4	34.7
ICU stay (days)	58	0.2	14	3.5	2.4
McGoon ratio	53	1.54	3.0	2.2	0.3
Nakata Index	53	170.0	725.9	386.2	127.2
Mair Index	20	1.4	4.6	2.9	1.0
Ao cross clamp (min)	66	0	127	58.4	28.2
Bypass time (min)	68	36	326	135.0	57.4
MeanPAp cath	32	6	25	13.7	4.2
MeanPAp OR	36	8	23	15.2	4.2
PVRI (Wood.u.m2)	22	0.7	4.8	2.1	1.1
Pre-op Ao Sat	52	60	99	29.9	8.7
Qp:Qs	27	0.3	4.7	1.2	0.9
PO. PAp day0	56	10	21	14.5	2.6
PO. PAp day1	55	8	21	14.4	3.1
PO. PAp day2	47	8	23	15.1	3.1

PVRI = pulmonary vascular resistance index
McGoon ratio = (right pulmonary artery diameter + left pulmonary artery diameter) / abdominal aorta diameter.
Nakata index (11) = {right pulmonary artery cross sectional area (mm2)+ left pulmonary artery cross sectional area (mm^2)} / body surface area (m^2)
Mair index (12) = pulmonary vascular resistance index + {ventricular end diastolic pressure/ (systemic blood flow index + pulmonary blood flow index)}

1 (25%) and 4 (30%) deaths in the acute post modified Fontan operation in double inlet left ventricle and complex univentricular heart with right ventricular morphology respectively. Another 3 patients died in the intermediate and late post operative period (6 months, 8 months, and 6 years). The causes of death were related to hemodynamic status in the first two and unknown in the last one. Seven patients developed recurrent pleural effusion and required chest tube drainage. Eleven cases underwent cardiac catheterization post operation: 4 had coil embolization for multiple aorto-pulmonary collateral arteries (MAPCA), 2 had stent placement at pulmonary artery for pulmonary artery stenosis, 2 required re-operation to redirect systemic vein to pulmonary system. Two of eleven had Fontan patch leak and one had moderate to severe decrease ventricular function demonstrated by angiogram.

Abnormal pulmonary vein and atrioventricular valve regurgitation had statistical significance in the acute post operative survival. Underlying cardiac anatomy was different between the survive and the death. Aortic cross clamp time, post operative PAp on day 0 to day 2 were lower in the survive than the death with statistical significance (Table 2, 3).

Follow-up

The median follow-up period was 3.0 years (range from 15 days to 8.3 years). Sixteen patients were lost to follow-up after discharge from the hospital, post modified Fontan operation. At the most recent follow-up and information from the questionnaire, there were: sinus rhythm 96.6 per cent (atrial flutter 1.7%, heart block 1.7%), normal exercise tolerance 88.1 per cent (mild and moderate exercise intolerance 10.2% and 1.7%), New York Heart Association (NYHA) class 1 88.1 per cent (class II 10.2%, class III 1.7%), and no cyanosis 84.5 per cent (mild 13.8%, moderate 1.7%). Thirty per cent are on no medication, 27 per cent on 1 and 24 per cent on 3 kinds of medication. Five-year survival rate was 83 per cent. The survival curve is demonstrated in Fig. 1.

Discussion

Pre operative data

Choussat et al developed 10 guidelines for relatively low risk Fontan in 1978[13]. Almost

Table 2. Statistical data.

	Survive (mean ± SD)	Death (mean ± SD)	p value
Age at Fontan (years)	11.6 ± 6.9	10.9 ± 10.1	0.392
BSA (m^2)	0.94 ± 0.30	0.95 ± 0.33	0.95
PA size: McGoon ratio	2.2 ± 0.3	2.2 ± 0.4	0.915
Nakata Index	387.5 ± 128.6	374.1 ± 125.2	0.830
Mean PAp (mmHg)	13.2 ± 3.5	16.2 ± 6.2	0.167
Pre-op O2 Saturation (%)	80.7 ± 8.3	73.7 ± 9.9	0.140
Surgical bypass time (mins)	130.4 ± 50.7	161.9 ± 85.9	0.295
Aortic cross clamp time (mins)	54.8 ± 25.2	81.4 ± 36.6	0.027*
Duration of chest drain (days)	7.5 ± 4.9	22.6 ± 37.9	0.898
Duration of inotropes (days)	4.6 ± 2.4	7.1 ± 8.5	0.731
Mean PAp post-op day0	14.1 ± 2.4	17.8 ± 2.6	0.003*
Mean PAp post-op day1	13.9 ± 2.8	19.0 ± 1.4	<0.001*
Mean PAp post-op day2	14.7 ± 2.8	19.8 ± 2.9	0.007*

* statistical significant ($p < 0.05$)

Table 3. Statistic data.

Underlying cardiac anatomy	Survive	%	Death	%	p Value
Abnormal systemic vein	4	7	1	10	1.000
Bilateral superior vena cava	11	18.3	5	50	0.072
Mild decrease ventricular function	5	8.2	0		0.785
More than 1 source of pulmonary flow	19	32.8	3	42.9	0.912
Previous shunt surgery	27	44.3	3	33.3	0.797
Abnormal pulmonary veins	2	3.3	4	40	0.002*
Atrioventricular valve regurgitation	5	8.5	2	20	0.010*
Tricuspid atresia or univentricular heart with LV morphology	39	63.9	0		0.002*

* statistical significant ($p < 0.05$)

all of our patients fulfilled the criteria. However, there were some who had age < 4 years old, mildly decreased ventricular function (ejection fraction 40-50%), bilateral SVC, and azygous or hemiazygous continuation to SVC in our study and not found to affect acute post operative survival. Abnormal pulmonary vein drainage and atrioventricular valve regurgitation were the risk factors of acute survival post modified Fontan operation in our study. These were also reported in many studies[9,13,14]. Masuda M et al reported a staging Fontan procedure by bidirectional Glenn shunt for high risk patients which resulted in reducing the operative mortality rate to the same number as the low risk group[14]. This is due to decreased volume overload which results in decreasing degree of atrioventricular valve regurgitation, reducing ventricular mass and improving ventricular function. Five patients in our study had bidirectional Glenn shunt prior to modified Fontan operation and all of them survived until the latest follow-up in late 1999. Imai Y et al[4] also did a study of atrioventricular valve repair in incompetent valves by using circular annuloplasty in the same setting as modified Fontan procedure with a much improved hospital mortality rate (12% to 3%). Ventricular hypertrophy was another important risk factor for both early and late post operation mortality[6]. This compromises the diastolic function in the acute post operative period. Younger age (<2-4 years old), complex congenital anomalies other than tricuspid atresia, especially heterotaxy syndrome were claimed to be risk factors in post operative survival[3,7,9,15]. Our study also revealed all patients who had tricuspid atresia or univentricular heart left ventricular type survived the acute post Fontan procedure and ventricular morphology significantly affected acute survival.

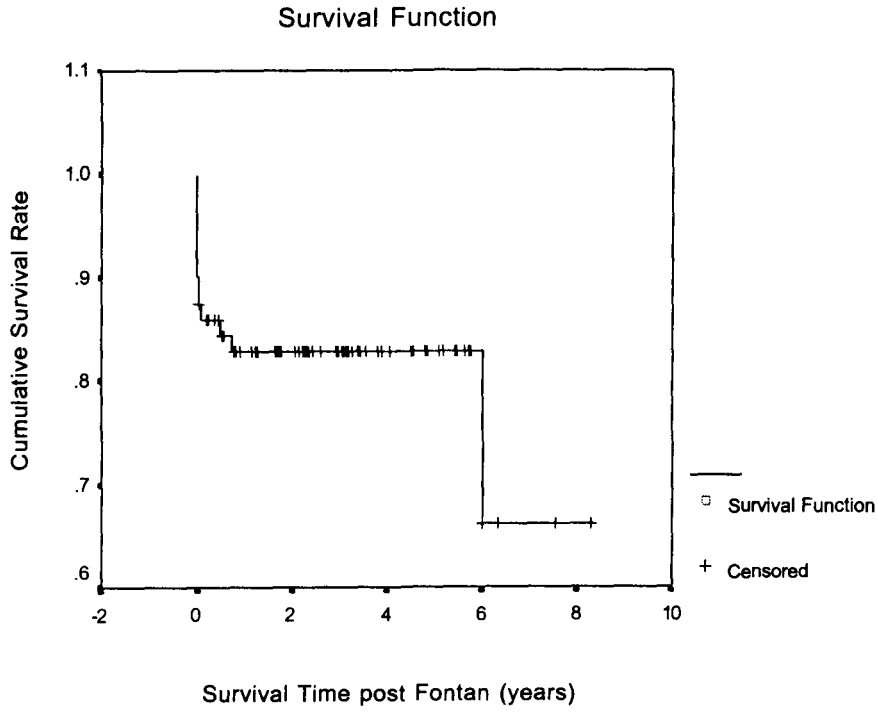

Fig. 1. Kaplan-Meier estimates survival rate post Fontan operation.

Operative and post operative period

Many technical modifications have been reported to decrease the acute post operative mortality and early and late morbidity, especially in imperfect candidates e.g. staging modified Fontan (bidirectional Glenn shunt, or hemi Fontan), lateral tunnel Fontan, fenestrated or partial hepatic vein exclusion in modified Fontan, and extracardiac Fontan etc.[2,4,16-20]. Bidirectional Glenn shunt and hemi Fontan procedure allows reduction of volume work up to 33 per cent of single ventricle and remodeling of ventricular geometry before completion of the modified Fontan procedure. At these operations, > 3.9 years old or body surface area > 0.65 m^2 were at significant risk for excessive post operative cyanosis[21]. Jacobs ML et al reported that fenestrated or partial hepatic veins exclusion in modified Fontan procedure reduced acute post operative mortality from 16 per cent to 4.5 per cent, and reduced the incidence of serous effusion from 45 per cent to 14 per cent. Pulmonary artery distortion, use of a non oxygenated glucose K+ cardioplegic solution and cardiopulmonary bypass time over 180 minutes were risk factors for early failure (death or takedown) of modified Fontan procedure[3]. Higher pre- operative PAp, operated on before 1980, asplenia syndrome, longer aortic cross clamp time, higher intraoperative (post procedure) right atrial pressure (>20 mmHg) were also reported by Knott-Craig CJ to be risk factors for early failure[7]. The two last parameters were also found to significantly affect early death in our study. Four patients were found to have significant MAPCA and two of them also had narrowing of the pulmonary artery which raised pressure in the Fontan circuit and resulted in prolonged chest tube drainage. The problem was solved by coil embolization and stent placement[22]. Most of these vessels originated from internal mammary arteries and thyrocervical trunk. Desaturation could occur post operation if there was the collateral form from the systemic vein to pulmonary vein. Triedman JK et al [23] found that collateral vessels occurred in 65 per cent and 30 per cent in post bidirectional Glenn

shunt and modified Fontan procedure respectively, especially in patients who had a history of Blalock-Taussig shunt. Step up of oxygen saturation in the distal pulmonary artery and/or an upper lobe filling defect helped to demonstrate this problem in cardiac catheterization.

Follow-up

Although modified Fontan techniques have decreased mortality and morbidity, sinus node dysfunction, intra-atrial or incisional reentrant tachycardia (IART), and sudden death are still not uncommon in post operative patients. Extensive atrial incision and stretch atrial wall are widely accepted as causing these problems. As many as 27 per cent of survivors at 20 years after operation has been reported[9,24]. Cohen MI et al found 23 per cent and 44 per cent in early and late (>4 years) with post Fontan procedure[25]. However, only 6.7 per cent of the patients in that study received a pacemaker and 4.1 per cent documented IART at mean follow-up 3.7 ± 1.7 years. These problems could be related to older age at operation, sinus node dysfunction either before or after operation, extensive atrial baffling, and longer follow-up period[24,26]. However, in our study this was documented in only 1.7 per cent who developed IART. This could be due to incomplete follow-up data in each patient and lost to follow-up. There were 15.3 per cent who had cyanosis at the most recent follow-up. This could be caused by some leakage of the intraatrial conduit, fenestrated Fontan, or veno-venous collateral vessels. Thromboembolic event was reported 3.9 per 100 patient-year post Fontan procedure and mainly in the venous circulation[27]. The occurrence was not different in all modification of the Fontan operation. The use of synthetic material in the procedure may be associated with the development of such an event. Routine long-term anticoagulant is still controversial. We had no incidence of thromboembolic events in our patients but this could be due to incomplete or lost follow-up. Progress of systemic ventricular outflow obstruction was also found in post Fontan. Finta KM. et al described this problem with the incidence of 12 per cent at the mean follow-up period of 28 months especially in the aorta arising from the hypoplastic ventricle and previous pulmonary artery banding [28]. Staging with bidirectional Glenn shunt and construction of proximal pulmonary artery-aorta connection or ventricular septal defect enlargement were reported as the options to neutralize these factors[29]. Protein losing enteropathy was one important intermediate to late morbidity post Fontan. An incidence of 3.7-10 per cent at 15 years follow-up period[8,9]. It was hypothesized that elevated systemic venous pressure leads to disturbed lymph drainage through the thoracic duct, increased intestinal congestion, lymph production and resulted in lymphocytes and proteins leaking from dilated lymphatics. These problems have not yet been clearly documented in our study. Survival rate post Fontan procedure was described as 77-88 per cent, 70-86 per cent, 60-81 per cent, and 73 per cent at 1, 5, 10, and 15 years respectively[9,10]. The number was not much different from our study, 83 per cent at 5 years post operation. Mortality was significant during the first 6 months and after 6 years post procedure[10]. This observation was also found in our study. Moreover, the longer the patients live, the higher the risk of morbidity. It is tempting to speculate that many patients may be best served by a longer period of palliation with bidirectional Glenn shunt, or other arterio-pulmonary shunts than total cavopulmonary or modified Fontan operation, especially in the imperfect candidates.

Limitation of this study

Incomplete pre operative hemodynamic and anatomic data in some patients, subjective and incomplete follow-up data and lost follow-up in many patients were the drawbacks of this study.

SUMMARY

Abnormal pulmonary vein, atrioventricular valve regurgitation, and underlying right ventricle or unidentified ventricular morphology were risk factors of the acute survival of modified Fontan operation. Intraoperative aortic cross clamp time, and post operative mean pulmonary artery pressure on day 0, 1 and 2 post operation were statistically shorter and lower in the survival group. Survival rate at 5 years was 83 per cent. Thorough preoperative hemodynamic and anatomic studies, appropriate type of modified Fontan or staging modified Fontan operation, close monitoring of post operative care might further decrease mortality and morbidity in the acute and late post operative period.

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