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Original Article

Fontan procedure with extracardiac conduit in pediatrics and children: Short-term results

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Abstract

Introduction

The concept of Fontan circulation was first clinically introduced in 1971. Technical modifications have been advocated with the concept of total cavopulmonary connection, introduced in the late 80s. Recently, an extracardiac conduit to divert inferior vena cava flow to the pulmonary arteries has been advocated, and it affords excellent palliation for patients with various forms of the anatomic or functional single ventricle.

Patients and methods

From September 2019 to August 2021, 12 patients (five males and seven females) with a mean age of 10.7 (6–15) years underwent a Fontan procedure with extracardiac conduit at the National Heart Institute. The mean weight was 34.75 (18–60) kg. The underlying diagnoses included tricuspid atresia ($n = 2$), double-inlet left ventricle ($n = 3$), transposition of great arteries ($n = 1$), and double-outlet right ventricle ($n = 6$). All patients were in sinus rhythm.

Results

There was one operative mortality. The type of conduit used was Gortex in 11 patients (91.7%) and Dacron in one patient (8.3%). The mean conduit size was 20 mm (16–26 mm). The mean cardiopulmonary bypass time was 127 (90–200) min. Aortic cross-clamping time was 25 min in two patients who needed total circulatory arrest. The mean duration of chest tube drainage was 31.8 days (15–130 days). All patients had sinus rhythm before the operation remained sinus. The mean ejection fraction was 65% (55–71%). The arterial oxygen saturation increased from a preoperative mean level of 64.2% (55–70%) to 90% (85–93%). There were nine patients (75%) who had fenestrations. The mean postoperative hospital stay was 36.7 (15–150) days, ICU stay was 6.5 (3–8) days, and mechanical ventilation time was 13.7 (4–27) h. There were three patients (25%) who needed reoperation, one who needed reintervention with chylous pleural effusion (8.3%), protein-losing enteropathy (8.3%), and infection. All patients had New York Heart Association (NYHA) functional class ≤II.

Conclusion

The extracardiac conduit is an easy way to perform the Fontan procedure, as it provides an excellent early result, maintenance of sinus rhythm, and preservation of ventricular function.

Keywords: Conduit, extracardiac, Fontan procedure

Introduction

The Fontan operation is described for univentricular physiology [1], there were modifications of techniques, like a lateral atrial tunnel, with improvement clinically [2,3]. However, systemic venous hypertension and supraventricular arrhythmias were the most disadvantages of flow dynamics in the systemic venous pathway.

The postoperative mortality rate of Fontan is high when the pulmonary artery pressure (PAP) greater than 18 mmHg, end-diastolic pressure greater than 12 mmHg, valve regurgitation, pulmonary artery (PA) distortion, pulmonary vascular resistance (PVR) greater than 2 Woods' units, ventricular outflow obstruction, and a complex anatomy,

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because of elevated systemic venous pressure and decreased cardiac output [4,5].

The Fontan procedure with extracardiac conduit has a maximum laminar blood flow, less atrial wall tension, and intra-atrial suture lines [6,7].

The extracardiac Fontan operation is an excellent palliation for patients with various forms of the anatomic or functional single ventricle. The incidence of infective endocarditis after the extracardiac Fontan is very low [8].

The advantages of the extracardiac conduit are the preservation of ventricular and pulmonary vascular function in the early postoperative period, a reduction of sinus node dysfunction or supraventricular arrhythmias, and improvement of hydrodynamics [9].

Patients and methods

From September 2019 to August 2021, 12 patients (five males and seven females) with a mean age of 10.7 years (range: 6–15 years) underwent a Fontan procedure with extracardiac conduit at the National Heart Institute. The mean weight was 34.75 kg (range: 18–60 kg) (Table 1). The underlying diagnoses included tricuspid atresia (n = 2), double-inlet left ventricle (n = 3), transposition of great arteries, ventricular septal defect, and pulmonary stenosis (n = 1), and double-outlet right ventricle (n = 6) (Table 2). All patients were in sinus rhythm. The mean ejection fraction was 64% (range: 58–68%). All patients had undergone prior operations. No patient with a bidirectional Glenn had an additional pulmonary blood flow in the form of an M-BT shunt or an antegrade flow from the right ventricular outflow tract. All patients had preoperative echocardiography and catheterization. All patients were examined for the presence of protein-losing enteropathy, which was defined as increased enteric loss of α1-antitrypsin (normal value <27 ml/24 h) or the occurrence of clinical symptoms (persistent or intermittent edema with hypoproteinemia without evidence for deficient protein production or excessive protein loss from organ systems other than the gastrointestinal tract).

Hemodynamics of Fontan pathway

While the normal systemic and pulmonary circulations are running in a parallel pattern, the Fontan pathway is a single circulation, in which the systemic and pulmonary circulations are supported by a single ventricle, and they are running in series [10].

There are many factors affecting the cardiac output of this univentricular circulation, including ventricular contractility, heart rate, preload, afterload, and transpulmonary blood flow [11].

Keeping low PVR is essential to obtain adequate ventricular preload, and the presence of systemic venous hypertension is needed to maintain the transpulmonary blood flow [10].

Table 1: Preoperative data

Number of patients	12
Age (years) [mean (range)]	10.7 (6-15)
Sex [n (%)]	
Male	5 (41.6)
Female	7 (58.3)
Weight (kg) [mean (range)]	34.75 (18-60)
Echocardiography (preoperative)	
EDD (cm) mean	4
ESD (cm) mean	2.6
EF% [mean (range)]	64 (58-68)
PAP (mmHg) [mean (range)]	12 (10-14)
O_2 Sat.% [mean (range)]	64.2 (55-70)
Sinus rhythm	12

EDD, end-diastolic diameter; EF, ejection fraction; ESD, end systolic diameter; O_2, oxygen sat; PAP, pulmonary artery pressure.

Table 2: Operative data

Etiology	n (%)
TA, ASD, VSD	2 (16.7)
DILV	3 (25)
TGA, VSD, PS	1 (8.3)
DORV	6 (50)
DORV, VSD	1 (8.3)
DORV, VSD, PS	3 (25)
DORV, VSD, PS, LSVC	1 (8.3)
DORV, VSD, PA	1 (8.3)
Previous operation	
PAB, Glenn + atrial septectomy	1 (8.3)
Glenn + Ligation of MPA	6 (50)
M-BT Shunt, Glenn	1 (8.3)
Bilateral Glenn	1 (8.3)
Glenn	3 (25)
Extracardiac conduit (n)	12
Type	
Gortex (PTFE) [n (%)]	11 (91.7)
Dacron [n (%)]	1 (8.3)
Size [mean (range)] (mm)	20 (16-26)
Fenestration [n (%)]	9 (75)
CPB time [mean (range)] (min)	127 (90-200)
ACC time (min) (n)	25 (2)
TCA time (range) (n) (min)	(15-25) (2)

ACC, aortic cross clamp; ASD, atrial septal defect; CPB, cardiopulmonary bypass; DILV, double-inlet left ventricle; DORV, double-outlet right ventricle; PA, pulmonary atresia; PAB, pulmonary artery banding; PS, pulmonary stenosis; PTFE, polytetrafluoroethylene; TA, tricuspid atresia; TCA, total circulatory arrest; TGA, transposition of great arteries; VSD, ventricular septal defect.

Pre-Fontan operation echocardiographic assessment

(1) The single-ventricular systolic and diastolic function.
(2) Regurgitation of atrioventricular valve.
(3) The outflow of the single ventricle.
(4) In patients suspected of aortic coarctation, assessment of ascending aorta, aortic arch, and isthmus.
(5) Superior vena cava (SVC) to PA communication.
(6) Flow across PA [12].

Pre-Fontan operation assessment by cardiac catheterization

Conventional cardiac catheterization should be performed for hemodynamic and anatomical assessment, including before the setting of Fontan operation; the following items should be assessed:

(1) PAP.
(2) PVR.
(3) Cavopulmonary shunt mean pressure.
(4) End-diastolic pressure of the single ventricle.
(5) Pressure gradient across the main PA and branches.
(6) Angiography for diagnosis of anatomical defects, for example, PA-branch stenosis, significant systemic-to-pulmonary-artery collaterals, abnormal flow, or anatomy of cavopulmonary shunt [12].

Calculation of indexed pulmonary blood flow (QPI) and indexed pulmonary vascular resistance (PVRI) using Fick's principle was done:

(1) Qp = VO_2/(pulmonary venous O2 content–pulmonary arterial O_2 content).
(2) PVR = mPAP–mLAP/Qp. where Qp is the pulmonary blood flow.
(3) VO2 is oxygen consumption, and the value used is indexed to body-surface area.
(4) PVR is pulmonary vascular resistance, mPAP is mean pulmonary-artery pressure, and mLAP is mean left-atrial pressure [13].

Indexed pulmonary-artery resistance was reported as woods units/meters^2.

Ten criteria were described by Choussat *et al.*[14] in 1977 for obtaining optimal results from the Fontan pathway as follows:

(1) Age greater than 4 years.
(2) Sinus rhythm.
(3) Normal systemic venous return.
(4) Normal right atrial volume.
(5) Mean pulmonary artery pressure less than 15 mmHg.
(6) Pulmonary arteriolar resistance less than 4 Woods units/m^2.
(7) Pulmonary artery aorta ratio greater than 0.75.
(8) Left ventricular ejection fraction greater than 60%.
(9) Competent mitral valve.
(10) Absence of pulmonary-artery distortion.

Currently, the criteria described by Choussat and colleagues are no more essential to obtain ideal Fontan results; many modifications were supposed by different authors.

The presence of low PVR and pulmonary artery pressure is still essential for successful Fontan operation, which can be performed in all patients younger than 4 years old. The presence of decreased left ventricular functions, mitral valve insufficiency, and anomalous systemic venous return is considered a relative contraindication for the operation. The patient's age and size should also be taken into consideration [15].

The postoperative data include ICU stay, hospital stay, duration of chest tube drainage, presence of chylous effusions, and protein-losing enteropathy as a morbidity and death.

The preoperative TTE is performed to measure the left ventricular systolic function and tricuspid regurgitation [16]. All data were presented by mean (range) like age, sex, weight, hemodynamics, and echocardiographic data. The mortality and morbidity like the presence of protein-losing enteropathy, chylous effusions, or mediastinitis were stratified as risk factors.

Exclusion criteria

(1) Conversion of a lateral atrial tunnel to an extracardiac conduit Fontan.
(2) Nonstaged Fontan procedure.

Inclusion criteria

(1) Staged Fontan procedure after Glenn.
(2) Tricuspid atresia and univentricular heart.
(3) Complex congenital heart disease that had a Glenn repair instead of biventricular repair due to lack of valved conduits.
(4) Age is greater than 3 years old.
(5) Body weight greater than 18 kg.
(6) Mean pulmonary artery pressure less than 15 mmHg.

Surgical technique

The idea of the extracardiac conduit Fontan circulation is completed by connecting the inferior vena cava to the inferior aspect of the right PA with a polytetrafluoroethylene conduit (Gore-Tex; W.L. Gore and Assoc., Elkton, MD) or a dacron vascular-tube graft. The best patient's age is greater than 3 years old and body weight greater than 18 kg. The size of the conduit is related to the body-surface area of the patient and should be at least 20 mm (range from 16 to 26 mm). Smaller sizes like16–18 mm are used in patients with polysplenia, left isomerism, and interrupted inferior vena cava (IVC) who had bilateral bidirectional Glenn to double SVC to reconnect the hepatic veins to pulmonary arteries.

The procedure is done with a double venous cannulation cardiopulmonary bypass (CPB), one with a metal-tip right-angled IVC cannula after dissection of the IVC from its pericardial attachment and the other in the right atrial appendage to avoid distortion of the SVC and the Glenn circulation. If the pulmonary arteries were small in size or distorted during the anastomosis, a patch augmentation was necessary. The pulmonary anastomosis was performed first with or without CPB by partially clamping the right PA so that the bidirectional Glenn is perfusing one or both lungs.

The aortic cross-clamping was avoided to preserve ventricular function and to decrease the postoperative pulmonary artery pressure, unless a bleeding occurred either before the procedure due to rupture of the aorta or after the procedure due to a tear in the right PA at the site of the anastomosis. Then a total circulatory arrest at 18–20°C is needed in order to visualize the tear and deal with it, so aortic cross-clamp and cardioplegia are needed to preserve the ventricular function.

After weaning from CPB, the Fontan pressure and the transpulmonary gradient were measured. If the Fontan pressure was more than 17 mmHg with a transpulmonary gradient of 10 mmHg or more, without the use of CPB, a fenestration (4–5 mm) was made between the extracardiac conduit and the right atrial free wall with side-to-side anastomosis without CPB.

After the operation, the following outcome measures were assessed: duration of mechanical ventilation, the presence of arrhythmias, ICU stay, duration of chest tube drainage, and hospital stay. Independent variables analyzed included the following: age, sex, duration of CPB, use of cardioplegic arrest or total circulatory arrest, and the presence of fenestration or not.

Results

The extracardiac conduit used at the procedure was a nonringed Gortex in 11 patients (91.7%), and one had a Dacron tube graft (8.3%) to connect the inferior vena cava with the pulmonary arterial system. The mean conduit size was 20 mm (range: 16–26 mm). Patch augmentation of the PA was needed in one patient (8.3%) with a pericardial patch due to a tear in the right PA during the anastomosis.

The mean CPB time was 127 min (range: 90–200 min) (Table 2). In the last 11 patients (91.7%), cannulation of the right atrial appendage and inferior vena cava to avoid SVC cannulation. Aortic cross-clamping with cardioplegic arrest (mean 25 min) was performed in two patients who needed total circulatory arrest. A total circulatory arrest was done in two patients, one at the beginning of the procedure due to aortic rupture for 25 min and the other for 15 min after completion of the procedure due to excessive uncontrolled bleeding from a tear at the pulmonary anastomosis.

The mean Fontan pressure was low, 12 (10–14) mmHg. A fenestration was constructed in nine patients (75%). There was one mortality (8.3%) due to bleeding during the opening of the sternum. The mean duration of chest tube drainage was 31.8 days (range: 15–130 days). Prolonged chest tube drainage (>30 days) occurred in two patients (16.7%).

All the 12 patients who were in sinus rhythm before the operation remained in sinus rhythm. During the first 48 postoperative hours, transient supraventricular tachyarrhythmia was observed in one patient (8.3%) with no hemodynamic instability and returned after medications. No patient was discharged on antiarrhythmic treatment.

The mean postoperative ventricular ejection fraction was 65% (55–71%). The arterial oxygen saturation increased from a preoperative mean level of 64.2% (range: 55–70%) to 90% (range: 85–93%). The lower saturations were because of the fenestration. There were nine patients (75%) who had fenestrations during the Fontan operation, and one patient had a fenestration in a later intervention after 5 months as the patient had ascites, infection, lower-limb edema, chylous effusion, and protein-losing enteropathy. During operation, pockets of pus in both pleurae were evacuated, and a thrombus found in the extracardiac conduit was removed.

Anticoagulation treatment consisted of acetylsalicylic acid in all patients and coumadin in all patients, except two patients, one with a congenital hemangioma in the colon and the other had shooting international normalized ratio (INR).

The mean postoperative hospital stay was 36.7 days, ranging from 15 to 150 days, and the ICU mean stay was 6.5 days that ranged from 3 to 8 days, with a mean mechanical ventilation time 13.7 h that ranged from 4 to 27 h (Table 3). There were no neurological complications postoperatively during this study.

There were three patients (25%) who needed reoperation during the early postoperative period: the first was reopened for bleeding, the second patient had mediastinitis, and she was reoperated for debridement with a pectoral flap and sternal rewiring. The third patient had complications of Fontan in the form of ascites, lower-limb edema, chylous pleural effusion (8.3%), protein-losing enteropathy (8.3%), and infection, which needed reintervention (readmission after one month of discharge and reoperation) for evacuation and fenestration.

Clinical follow-up was made at a mean time of 12 months in all survivors. All patients had NYHA functional class ≤II. An echocardiographic follow-up to all patients at a mean time of 1 year. All patients who had echocardiographic follow-up were asymptomatic. Only one patient who had echocardiographic follow-up was symptomatic.

Table 3: Postoperative data

Postoperative echocardiography [mean (range)]	
EDD (cm)	3.9 (3.2-4.5)
ESD (cm)	2.5 (2.1-2.9)
EF (%)	65 (55-71)
TR [*n* (%)]	
+ 1	4 (57.1)
+ 1-2	2 (28.6)
+4	1 (14.3)
Mortality [*n* (%)]	1 (8.3)
Reopening [*n* (%)]	3 (25)
Reintervention [*n* (%)]	1 (8.3)
Arrhythmia (SVT) [*n* (%)]	1 (8.3)
Mechanical ventilation (h) [mean (range)]	13.7 (4-27)
O_2 Sat.% [mean (range)]	90 (85-93)
ICU stay (days) [mean (range)]	6.5 (3-8)
Hospital stay (days) [mean (range)]	36.7 (15-150)
Chest tube drainage (days) [mean (range)]	31.8 (15-130)
Protein-losing enteropathy [*n* (%)]	1 (14.3)
Chylous pleural effusion [*n* (%)]	1 (14.3)

EDD, end-diastolic diameter; EF%, ejection fraction; ESD, end systolic diameter; TR, tricuspid regurgitation.

Discussion

The extracardiac conduit Fontan has an excellent hemodynamic with preservation of ventricular function and absence of major complications. In our study, preservation of sinus rhythm and ventricular function was achieved.

The potential advantages of the extracardiac conduit Fontan operation include technical ease, no manipulation in the atrium or the sinus node [7,17,18], avoiding energy dissipation by turbulence and stasis in the cavopulmonary connection [9,19].

Fontan procedure with extracardiac conduit has the advantage over the classic atriopulmonary connections in reducing of right atrial volume to avoid increased atrial wall tension and chronic elevation of coronary sinus pressure with depression of ventricular function [20]. In addition, optimum delivery of blood flow of IVC into the pulmonary arteries due to the geometry and uniform caliber of the extracardiac conduit tube, which creates a laminar flow, compared with progressive dilatation or growth of the lateral tunnel, which leads to asymmetry and distortion of the systemic venous pathway causing loss of laminar flow.

Atrial arrhythmias, particularly atrial flutter, are a major source of morbidity, particularly during late follow-up after the Fontan operation, with reported rates of up to 40% [16,18]. The extracardiac conduit Fontan reduces arrhythmias due to avoidance of all four of the factors thought to be the cause of rhythm disturbances after the Fontan procedure:

(1) Exposure of the right atrium to the elevated systemic venous pressure.
(2) Extensive atrial incisions and suture lines.
(3) Surgical manipulation in the vicinity of the sinus node.
(4) Ventricular dysfunction resulting from ischemic arrest and prolonged CPB.

Transient supraventricular tachyarrhythmias usually occurred in 13% of patients in the early postoperatively stage. In our study, there was one patient (8.3%) and returned to sinus rhythm with medication.

Patients with a single ventricle like double outlet right ventricle (DOVR) and malposed great vessels, which had a long period of pulmonary artery banding with the resultant ventricular hypertrophy, have a significant late failure [3]. When banding cannot be avoided, it is advisable to have a short duration of the banding. There was one patient (8.3%) who had pulmonary artery banding for DORV and malposed great vessels, then a bidirectional Glenn, and then nonfenestrated Fontan with a 20-mm extracardiac conduit. He was the only case with protein-losing enteropathy and prolonged chylous drainage in our study.

The noncylindrical pathway of intracardiac Fontan is often dilated, and the turbulent flow enhanced by atrial arrhythmias may lead to the stasis of blood and thrombus formation [21–23]. While the cylindrical pathway of extracardiac conduit Fontan is fixed in size with no turbulent flow, so the atrial arrhythmias are less frequent. In our study, there was one patient (8.3%) who had postoperative atrial arrhythmia in the form of SVT and returned to sinus rhythm with medications.

Thromboembolism, conduit stenosis, and lack of growth are the disadvantages of the Fontan procedure related to the extracardiac conduit. We had one patient (8.3%) who had conduit stenosis due to partial thrombosis and needed reintervention for evacuation and fenestration.

The extracardiac conduit most likely never becomes fully endothelialized. It may permit partial or full endothelialization, as in humans, the endothelialization of prosthetic material usually extends to the first 5–10 mm. There is an increased risk of thromboembolism with pulmonary emboli and systemic emboli if a fenestration is constructed.

That is why all patients were discharged on acetylsalicylic acid and coumadin therapy, except the patients with noncompliance to coumadin with high uncontrolled INR and patients with frequent attacks of rectal bleeding due to colonic hemangeoma.

An adult-sized conduit provides delay reoperation after completion of the Fontan procedure. We used adult-sized conduit greater than or equal to 20 mm in 10 patients (83.3%) and conduit sized 16 mm in one patient (8.3%) who had bilateral Glenn for the left SVC and conduit sized 18 mm for one patient (8.3%).

Amodeo and associates evaluated the patency of the extracardiac conduit by magnetic resonance imaging and reported an 18% reduction in conduit internal diameter in the first 6 postoperative months, with no progression during the next 5 years. However, Dacron conduits were used initially, which may account for an increased tendency toward peel formation as opposed to Gore-Tex grafts [24]. In our study, we had conduit partial thrombosis in one patient (8.3%) who needed evacuation and fenestration.

Ukai and colleagues reported in a study that 66 patients with various forms of anatomic or functional single ventricle had Fontan procedures. Thrombus formed in the extracardiac conduit in two patients (3.0%), infective endocarditis after the Fontan operation occurred only in this one patient (1.5%). In our study, we had one patient (8.3%) with a thrombus in the extracardiac conduit. There was no infective endocarditis [25].

Although the law of transplantation in Egypt has been made, there is a lack of valved conduits like aortic and pulmonary homografts, which are needed for patients with complex congenital heart disease for their biventricular repair. This issue forces some surgeons to go for univentricular repair and start with Glenn cavopulmonary anastomosis. After a while, the patient begins to complain and needs to go for Fontan.

At the end, which is good for these patients? To have two-stage repair with Glenn, then Fontan, and the patient will have a chance to live near-normal life (if the Fontan goes smoothly without complications) or instead, biventricular repair, and the patient will have a chance to live normally after repeated reoperations every decade to replace the valved conduit that will jeopardize the patient to hazards of reopening.

Limitations

This study has a small number of the patient population. All the procedures were done according to the surgeon's point of view. This study was dealing with different kinds of anomalies and different risk levels. All surgical outcomes mentioned earlier might be influenced by the surgeon's learning curve.

Conclusion

The extracardiac conduit is an easy way to perform the Fontan procedure, as it provides an excellent early result, maintenance of sinus rhythm, and preservation of ventricular function.

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Conflicts of interest

There are no conflicts of interest.

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