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Outcomes of the Extracardiac Fontan Procedure Using Cardıopulmonary Bypass: Early Results

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ABSTRACT

Background: Although the avoidance of cardiopulmonary bypass during the Fontan procedure has potential advantages, using cardiopulmonary bypass during this procedure has no adverse effects in terms of morbidity and mortality rates. In this study, we assessed the postoperative outcomes of our first 9 patients who have undergone extracardiac Fontan operation by the same surgeon using cardiopulmonary bypass.

Methods: Between September 2011 and April 2013, 9 consecutive patients (3 males and 6 females) underwent extracardiac Fontan operation. All operations were performed under cardiopulmonary bypass at normothermia by the same surgeon. The age of patients ranged between 4 and 17 (9.8 ± 4.2) years. Previous operations performed on these patients were modified Blalock-Taussig shunt procedure in 2 patients, bidirectional cavopulmonary shunt operation in 6 patients, and pulmonary arterial banding in 1 patient. Except 2 patients who required intracardiac intervention, cross-clamping was not applied. In all patients, the extracardiac Fontan procedure was carried out by interposing an appropriately sized tube graft between the inferior vena cava and right pulmonary artery.

Results: The mean intraoperative Fontan pressure and transpulmonary gradient were 12.3 ± 2.5 and 6.9 ± 2.2 mm Hg, respectively. Intraoperative fenestration was not required. There was no mortality and 7 patients were discharged without complications. Complications included persistent pleural effusion in 1 patient and a transient neurological event in 1 patient. All patients were weaned off mechanical ventilation within 24 hours. The mean arterial oxygen saturation increased from 76.1% ± 5.3% to 93.5% ± 2.2%. All patients were in sinus rhythm postoperatively. Five patients required blood and blood-product transfusions. The mean intensive care unit and hospital stay periods were 2.9 ± 1.7 and 8.2 ± 1.9 days, respectively.

Conclusions: The extracardiac Fontan operation performed using cardiopulmonary bypass provides satisfactory results in short-term follow-up and is associated with favorable postoperative hemodynamics and morbidity rates.

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INTRODUCTION

Since the introduction of the Fontan procedure in 1971 [Fontan 1971], numerous procedures have been advocated for the establishment of total cavopulmonary connection in patients with univentricular physiology [Marcelletti 2000; Azakie 2001; Yetman 2002]. Previous approaches generally included cavopulmonary connections by either intra-atrial rerouting using a baffle or intra-atrial grafting with a Gore-Tex tube. Although these procedures seem to be useful in patients requiring an intracardiac intervention, the use of an extracardiac conduit to achieve a total cavopulmonary connection has become the strategy of many surgeons [Alexi-Meskishvili 2001] because of the postoperative tendency for systemic venous hypertension and long-term arrhythmogenic effects associated with intraatrial approaches. The theoretical advantages of an extracardiac type connection include the avoidance of aortic cross-clamping and myocardial ischemia, shortened duration of cardiopulmonary bypass, improved short- and long-term hemodynamics, lower incidence of atrial arrhythmias, establishment of laminar flow, and reduced energy loss [Amodeo 1997; Lardo 1999; Petrossian 2000; Tireli 2006].

Another controversial issue regarding this procedure is the use of cardiopulmonary bypass during the performance of an extracardiac connection. Some authors have claimed that in patients with single-ventricle physiology, the use of cardiopulmonary bypass may be associated with persistent pleural effusion, increased risk of lung injury, and consequent prolonged mechanical ventilation requirements [Kogon 2008; LaPar 2012]. Previous studies also found that prolonged cardiopulmonary bypass is an independent predictor of adverse outcomes after an extracardiac Fontan procedure [Petrossian 2006]. On the other hand, there are also many studies that have demonstrated that the extracardiac Fontan procedure can be performed with no significant differences in terms of operative mortality and morbidity either with or without the use of cardiopulmonary bypass [Meyer 2006]. Therefore, the optimal approach for performance of an extracardiac Fontan procedure with respect to the use of cardiopulmonary bypass remains unclear; thus far, no single approach has been shown

Table 1. Demographic Data of the Patients*

Patients	Diagnosis	Sex	Age, years	Previous Operation
1	Tricuspid atresia, PS	M	17	Central shunt
2	Tricuspid atresia, PS	F	4	—
3	Unbalanced CAVSD	M	9	BD Glenn shunt
4	Unbalanced CAVSD	F	4	DOLV, PS
5	DORV, RV hypoplasia, PS	F	17	BD Glenn shunt
6	Dextrocarida, LV hypoplasia, PS	M	8	BD Glenn shunt
7	Tricuspid atresia, PS	F	10	BL Glenn shunt
8	Tricuspid atresia, PS	F	10	BT shunt + BD Glenn
9	Tricuspid atresia	F	10	PA banding +BD Glenn

*BD indicates bidirectional; BL, bilateral; CAVSD, complete atrioventricular septal defect; DOLV, double outlet left ventricle; DORV, double-outlet right ventricle; LV, left ventricle; PA, pulmonary artery; PS, pulmonary stenosis.

to be superior. We believe that using cardiopulmonary bypass during this procedure has no adverse effects in terms of morbidity and mortality rates. In this study, we assessed the postoperative outcomes of our first 9 patients who have undergone extracardiac Fontan operation using cardiopulmonary bypass.

MATERIALS AND METHODS

Between September 2011 and April 2013, 9 consecutive patients underwent an extracardiac Fontan operation using cardiopulmonary bypass at our institution, all performed by the same surgeon (M.B.). There were 3 male and 6 female patients. The age of patients ranged between 4 and 17 years (9.8 ± 4.2 years). The demographic data of the patients is summarized in Table 1. All patients were in sinus rhythm preoperatively. Previous operations performed on these patients were modified Blalock-Taussig shunt procedure in 2 patients, bidirectional cavopulmonary shunt operation in 6 patients, and pulmonary arterial banding in 1 patient. All patients were evaluated with echocardiography and angiography preoperatively. The mean pulmonary vascular resistance was 2.1 ± 0.2 Wood Units.

Surgical Technique

All operations were performed through a standard or repeat median sternotomy. The ascending aorta, pulmonary arteries, superior and inferior vena cavae, and (if present) previous Blalock-Taussig shunt were dissected free and controlled with tapes. Systemic anticoagulation was achieved with full-dose intravenous standard heparin (300 units per kilogram) with the aim of an activated clotting time in excess of 450 seconds. In cases without a previous Glenn operation, the superior vena cava was dissected up to the innominate vein level and the azygos vein was doubly ligated and divided. The inferior vena cava–atrial junction was completely liberated and dissected below the diaphragm if necessary. The lateral pericardial recess was also freed in order to avoid compression of the underlying pulmonary veins by the tube graft. Superior and inferior vena cava cannulae were directly placed on the innominate vein and inferior vena cava, respectively. The cannula was put as low as possible onto the inferior vena cava. The ascending aorta was cannulated as a standard fashion. All operations were performed at normothermia under cardiopulmonary bypass without applying cross-clamping. In 2 patients additional intracardiac intervention was carried out. One patient with a diagnosis of a double outlet right ventricle, rudimentary left ventricle, common atrioventricular valve, and severe atrioventricular valve regurgitation required valve repair. For the remaining patients, the restrictive atrial-septal defect was enlarged with a short period of cardiac arrest.

During the operation, the superior vena cava was clamped at the cavoatrial junction and transected. The atrial end was oversewn. An arteriotomy was performed on the anterosuperior aspect of the right pulmonary artery. Bidirectional Glenn anastomosis was performed to the most suitable region of the right pulmonary artery using a continuous suture technique. The inferior vena cava was clamped and transected from the cavoatrial junction, taking care to avoid injury to the coronary sinus and the right coronary artery. The cardiac end of the inferior vena cava was oversewn. The caudate end of the graft was anastomosed to the inferior vena cava in an end-to-end fashion with a running polypropylene suture. According to the diameter of the inferior vena cava, an appropriately sized Gore-Tex tube graft was selected. The cranial end of the tube graft was beveled and anastomosed end-to-side to the most suitable region at the inferior aspect of the right pulmonary artery with a continuous polypropylene suture. Fenestration was not routinely used.

Hospital review board approval for a retrospective clinical trial was obtained. Demographic and preoperative data, including anatomical diagnoses, ventricular morphology, age, weight, and prior surgical procedures were analyzed. Perioperative data including central venous pressure, blood pressure, and arterial oxygen saturation levels were obtained by review of hospital medical records. The early postoperative period was defined as the time from surgery to hospital discharge. The postoperative data during this time period, including length of stay in the intensive care unit, length of mechanical ventilation, need for blood and/or blood-product transfusion, postoperative O_2 saturation, pulmonary arterial pressure, length of intensive care unit and hospital stay periods, and surgical complications were noted. The late postoperative period was defined as the time from initial hospital discharge to latest follow-up. Follow-up data, including copies of electrocardiograms, echocardiograms, and other diagnostic information after discharge were obtained from the patients' cardiologists or family. Late arrhythmia data were based on the patients' histories of symptoms and follow-up electrocardiograms. All values are expressed as ± standard error of the mean, and categorical data are reported as a percentage.

RESULTS

Anatomical diagnoses included tricuspid atresia in 5 and single ventricle with pulmonary stenosis in the remaining 4 patients; of these, 1 patient had dextrocardia, single ventricle with left atrial isomerism, severe pulmonary stenosis, and azygos continuation of an interrupted inferior vena cava, 2 patients had unbalanced complete atrioventricular septal defect and pulmonary stenosis, and 1 patient had double-outlet right ventricle, rudimentary left ventricle, and common atrioventricular valve. Five patients had a bidirectional cava-pulmonary connection before completion of the Fontan procedure, and the mean interval from the bidirectional cava-pulmonary connection to the Fontan was 39.1 ± 6.2 months. The polytetrafluoroethylene conduit sizes ranged from 18 to 24 mm (24 mm in 2 patients, 22 mm in 2 patients, 20 mm in 4 patients, and 18 mm in 1 patient).

There was no postoperative mortality. Pressure measurements after the completion of all anastomoses revealed no pressure gradients between the pulmonary arteries and either the superior or the inferior caval veins. The mean intraoperative Fontan pressure and transpulmonary gradients were 12.3 ± 2.5 and 6.9 ± 2.2 mm Hg, respectively. Fenestration between the graft and right atrium was not required in any patient. All patients were transferred to the intensive care unit with our standard protocol of inotropic support (dobutamine 5 μg/kg per minute and afterload reduction with nitroglycerin 2.5 μg/kg per minute for 24 hours postoperatively), after which time inotropic support is stopped. All patients were extubated within 24 hours after the operation. The mean mechanical ventilation time was 10.3 ± 4.8 hours (range 3 to 24 hours) The peri- and postoperative data are presented in Table 2.

The mean arterial oxygen saturation raised from 76.1% ± 5.3% to 93.5% ± 2.2%. All patients were in sinus rhythm, and no arrhythmia developed in any patient in the early postoperative period. Five patients required blood and blood-product transfusions (2 units of packed red blood cells in 2 patients and 1 unit in 3 patients). The mean intensive care unit and hospital stay periods were 2.9 ± 1.7 and 8.2 ± 1.9 days, respectively. Mean postoperative central venous pressure was 13.7 ± 1.9 mm Hg. Mean chest tube drainage continued 3.5 ± 1.2 days.

Table 2. Postoperative Outcome Variables*

Patients	Pre-op SPO_2, %	FP, mm Hg	CT Drainage, mL	ICU/Hospital Stays, days	VS, hours
1	81	14	400	4/12	24
2	70	11	250	2/7	13
3	72	10	150	4/8	3
4	78	12	250	4/8	18
5	77	11	300	2/6	6
6	70	12	250	2/6	3
7	80	13	350	5/9	18
8	77	12	250	2/9	4
9	80	12	150	1/6	4

*CT indicates chest tube; ICU, intensive care unit; FP, Fontan pressure; Pre-op, preoperative; SPO_2, partial oxygen saturation; VS, ventilatory support.

Complications included persistent pleural effusion in 1 patient and a transient neurological event in 1 patient. There was only 1 patient with persistent pleural effusion requiring chest tube drainage (more than 7 days). This patient recovered uneventfully and was discharged at postoperative day 6. However, 3 weeks later, she was readmitted to our hospital with progressive dyspnea. Her physical and radiological evaluation revealed massive pleural effusion at the right hemithorax. A chest tube was placed and the patient was managed conservatively. At the end of 8 days, the tube drainage was stopped and the patient was redischarged without trouble. The other patient was a 17-year-old patient with a history of a previous cerebral infract. During the early postoperative period, a weakness was noticed in his left arm and leg. Only the chronic infarct area was visible on his tomographic evaluation. This patient was also managed conservatively and he recovered dramatically, without permanent neurological damage.

After discharge, no major complications were observed. All patients were anticoagulated with Coumadin and there were no incidents of graft thrombosis. Although the follow-up period is short, no patient undergoing extracardiac Fontan procedure with cardiopulmonary bypass has developed protein-losing enteropathy. One patient required 24-hour ambulatory Holter monitoring because of transient episodes of supraventricular tachyarrhythmia. This patient was treated medically and she was noted to be in sinus rhythm on her most recent evaluation. No patient had ventricular tachycardia. All patients were followed up biannually with echocardiography for assessment of ventricular function, atrioventricular valve regurgitation, and assessment of obstruction of the Fontan circuit. Two patients were noted to have mild ventricular dysfunction, and one patient mild-to-moderate atrioventricular valve regurgitation. Echocardiography revealed no evidence of pulmonary artery distortion or obstruction of the Fontan circuit on late follow-up.

DISCUSSION

With the recent refinements in cardiac anesthesia, myocardial protection, and postoperative care, many complex congenital cardiac defects may be surgically managed today with acceptable morbidity and mortality rates. In patients with univentricular physiology, the aim of physiologic correction should be the prevention of short- and long-term morbidity and mortality. Although total cavopulmonary connection is the universally accepted palliative procedure in patients with a functional single ventricle [d'Udekem 2007; Kim 2008; Ohuchi 2011], the optimal surgical strategy is a subject of considerable debate.

Nowadays, the lateral tunnel and extracardiac approaches are the current options which are widely used by many

surgeons [Kumar 2003; Meyer 2006; Kim 2008]. Each technique has its own advantages, and previous studies have demonstrated excellent outcomes for both approaches [Azakie 2001; Nakano 2004; Alphonso 2005; Van Doorn 2005; Meyer 2006]. Proponents of the lateral tunnel approach advocates this technique due to its growth potential. This technique is also relatively simple to fenestrate. Potential disadvantages for the lateral tunnel approach include the presence of multiple atrial suture lines and the exposure of a portion of the atrium to increased venous pressure. On the other hand, the extracardiac reconstruction has fewer atrial suture lines, which theoretically reduces the possibility of postoperative arrhythmias [Azakie 2001; Meyer 2006]. Avoidance of intracardiac prosthetic material and exposure of the atria to increased systemic venous pressure are the other reasons why this procedure has become the preferred technique by many surgeons. Its limited growth potential and the relative difficulty in performing fenestration are the main disadvantages of this approach.

We believe that the use of prosthetic materials at a young age increases risk of prosthesis–patient mismatch in the future. It has previously been reported that at 2 to 4 years of age, and 12 to 15 kg of body weight, the inferior vena cava to–pulmonary artery distance is up to 80% that of adult size [Yetman 2002; Alexi-Meskishvili 2001]. Other studies also revealed [Ocello 2007; van Slooten 2012] that a body weight of at least 15 kg allowed the insertion of a 20- to 22-mm conduit, which is the same size often used in adults. Our current strategy in total cavapulmonary connection is to allow patients to grow in anticipation of the final Fontan; in this way, we aimed to insert a larger tube graft during the operation. In patients weighing than 15 kg, we generally delay the surgical intervention until the patient reaches the ideal size. As may be observed in our patient cohort, the mean weight of our cases was 26.4 ± 8.2 kg, and they were generally older than those reported in the literature. Even in these patients, we generally tried to use the tube grafts that oversize the diameter of the inferior caval vein by less than 20%. In our series, we constructed the inferior cavopulmonary pathway with 24 mm in 2 patients, 22 mm in 2 patients, 20 mm in 4 patients, and 18 mm in 1 patient. By using a slightly larger extracardiac conduit, we aimed to overcome the limitation of the extracardiac technique in terms of growth potential. Indeed, although the follow-up period is very short, we have not confronted a patient requiring conduit replacement because of prosthesis–patient mismatch.

Currently, some surgeons propose the avoidance of cardiopulmonary bypass during extracardiac Fontan to prevent the harmful effects of extracoporeal circulation on single-ventricle and pulmonary functions [McElhinney 1998; Tam 1999; Tireli 2006]. Some studies demonstrated that prolonged cardiopulmonary bypass and cross-clamp times are significant predictors of mortality and risk of complications in patients undergoing Fontan procedures [McElhinney 1998]. Kawahira and colleagues [Kawahira 2006] demonstrated the attenuation of inflammatory reactions in patients undergoing an off-pump procedure. However, they also stated that there are many factors, including drugs, protamine, duration of surgical procedure, and blood transfusion, which may potentially affect the levels of cytokines in these patients. On the other hand, Petrossian [Petrossian 2006], in his series of 285 patients, demonstrated that the presence or absence of a pump seems to have no effect on early outcome and added that the operation can be performed either off pump or under cardiopulmonary bypass with equal benefit. Our series is very small and it is not possible to draw definitive conclusions from this small number. However, in our series, we did not notice any adverse effects of using cardiopulmonary bypass in terms of morbidity and mortality.

It is accepted that multiple factors, including obligatory foreign material, systemic coagulation profile, nonpulsatile flow, and stasis put Fontan patients at increased risk of thromboembolic complications [Jahangiri 1994]. Since the area of foreign body in contact with the bloodstream is higher with the extracardiac procedure, it has been hypothesized that extracardiac patients are more prone to thromboembolism. Some authors stated that the total area of bloodstream contact is roughly similar between the extracardiac and the lateral tunnel. Therefore, despite initial concerns of an increased risk of thromboembolic complications with an extracardiac procedure, we and others have noticed no thrombosis in these patients [Marcelletti 1999; Yetman 2002]. We currently follow our patients with life-long anticoagulation because of abnormal coagulation patterns of patients undergoing Fontan procedure.

In conclusion, the extracardiac Fontan procedure is a safe, reliable, and effective technique in patients with single-ventricle physiology. In light of the literature, we can also state that the use of cardiopulmonary bypass during this procedure has no adverse effects on early outcome.

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