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Results of 100 Consecutive Extracardiac Conduit Fontan Operations

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Abstract Throughout the years, the experience with Fontan's operation has increased and has opened the way to a much wider application of this principle. A number of major risk factors have been identified and managed by several modifications of the original Fontan procedure. In the past 15 years, operative and postoperative risks have been controlled to a major extent by the application of intermediate surgical procedures. Modifications to the original technique have been designed to simplify surgery and better protect the myocardium by avoiding, as much as possible, prolonged ischemic time. In 1988, we developed a new form of total extracardiac right heart bypass by means of associating an extracardiac conduit placed between the inferior vena cava (IVC) and the pulmonary artery with a bidirectional cavopulmonary anastomosis (BCPA)—the so-called total extracardiac cavopulmonary connection (TECC).Between November 1997 and October 2003, 100 patients with complex cardiac anomalies underwent a modified Fontan operation by TECC. In 88 patients, the repair was staged by preliminary BCPA that was bilateral in 15 patients and associated with a modified Damus–Kaye–Stansel procedure to bypass a subaortic obstruction in 24 patients. Early (in hospital) deaths occurred in 6% of patients and the extracardiac conduit was taken down in 2 additional patients for a total early failure rate of 8%. The cause of death was myocardial failure in 5 patients. Pulmonary artery distortion or hypoplasia appeared to be the cause of death in 1 patient and the cause of failure in 1 patient. Atrioventricular valve regurgitation was the cause of failure in 1 patient. Follow-up to December 2003 was available in all surviving patients. There were no late deaths. At follow-up, 87 patients (89%) were in New York Heart Association (NYHA) functional class I, 4 in class II, and 3 in class III due to moderate to severe atrioventricular valve regurgitation. Use of the following guidelines can result in the achievement of orthoterminal repair with complete separation of pulmonary and systemic circulation, with negligible early mortality and a long-term NYHA class I: (1) Not all patients with univentricular heart (UVH) should undergo the extracardiac Fontan procedure; (2) in UVH with favorable streaming, a Q_p/Q_s ratio of approximately 1 to 1, and mild cyanosis, natural history might be preferable to surgical history; (3) any form of fenestration is contradictory to orthodox application of Fontan's principle. Children in whom a planned fenestration seems necessary because of suboptimal conditions should instead undergo a combination of BCPA and associated forward pulmonary blood flow; and (4) negligible mortality should be considered mandatory in UVH, normal pulmonary arteries, and absent cardiomegaly after appropriated and correct staging.

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Throughout the years, experience with the Fontan operation has increased [2, 3, 7], allowing for a much wider application of this principle [8]. A number of major risk factors have been identified and managed by several modifications of the original Fontan procedure. During the past 15 years, the impact of operative and postoperative risks has been controlled to a major extent by the application of intermediate surgical procedures—the so-called

staging toward the Fontan operation [5]. Modifications to the original technique have been designed to simplify surgery and better protect the myocardium by avoiding, as much as possible, prolonged ischemic time.

In 1988, while treating a child with one ventricle and anomalies of pulmonary and systemic venous return, we developed a new form of total extracardiac right heart bypass by means of an extracardiac conduit placed between the inferior vena cava (IVC) and the pulmonary artery with a bidirectional cavopulmonary anastomosis—the total extracardiac cavopulmonary connection (TECC). This technique was applied to patients presenting with other anomalies, such as auricular juxtaposition, hypoplasia, or atresia of the left atrioventricular valve or common atrioventricular valve [6].

Because of the successful outcome of these patients, this preliminary experience was extended to all patients with a functional single ventricle. In our early experience, we were concerned about this procedure because of the need for an artificial conduit in the systemic venous pathway and the possible risk of late stenosis and thrombus formation. Based on careful follow-up, along with long-term clinical results, TECC is our technique of choice, and it has been renamed extracardiac Fontan [1].

Rationale for a Staged Fontan Procedure

In general, two major categories of univentricular heart can be identified according to the pattern of pulmonary blood flow (PBF): those with restrictive PBF and those with unrestricted PBF. Patients with restrictive PBF, depending on the severity of pulmonary stenosis, might require surgical palliation at a younger age. We believe that a systemic-to-pulmonary artery shunt is preferred in patients younger than 4–6 months. In older patients, a bidirectional cavopulmonary anastomosis (BCPA) should be the palliation of choice, allowing growth and protection of the pulmonary vascular bed. Patients with unrestricted PBF have a typical clinical presentation with congestive heart failure and failure to thrive during the first months of life. These patients often have a Q_p/Q_s of more than 2 to 3:1, a volume overload that may impair long-term ventricular function. All these findings indicate the need for surgical intervention early in life. In the absence of systemic outflow tract obstruction, pulmonary artery banding (PAB) may represent a good surgical option.

After 6–8 months, the patient undergoes a second procedure by Damus–Kaye–Stansel anastomosis, if systemic outflow tract obstruction is present or likely, and BCPA. When the pulmonary artery pressure distal to the band is sufficiently low and PBF is well controlled, an isolated BCPA may be added to the PAB.

Potential Advantages of the Extracardiac Conduit Fontan Procedure

TECC has several theoretical and practical advantages that may lead to improved postoperative recovery and long-term outcomes:

1. It optimizes laminar flow patterns within the cavopulmonary connections, thereby minimizing energy dissipation.
2. It avoids extensive suture lines in the right atrium, thereby decreasing the substrate for postoperative atrial tachyarrhythmias.
3. It prevents right atrial exposure to the elevated pressures of the systemic venous circuit, thereby decreasing the propensity for atrial dilatation and secondary supraventricular tachyarrhythmias, thrombus formation, and right pulmonary venous obstruction.
4. The risk of obstruction to the pulmonary venous return is reduced.
5. Intraatrial placement of prosthetic materials is avoided.
6. It preserves the option of creating a fenestration without returning to cardiopulmonary bypass in the event that an elevated transpulmonary gradient exists following repair.
7. In case of late conduit obstruction, the conduit can be replaced in a totally extracardiac procedure.

Candidates for an Extracardiac Fontan Procedure

Hemodynamic criteria are already well established. However, one condition that must be upheld is that the patient should weigh at least 15 kg. This allows insertion of a 20- to 22-mm conduit, which is almost the same size as that used in adults. The patients who are properly staged are those who have undergone BCPA at the correct time. The only contraindication is poor ventricular function.

Surgical Technique

The Fontan procedure is almost always performed in patients who have undergone at least one median sternotomy and therefore have mediastinal adhesions. The procedure can be performed during hypothermic circulatory arrest or during continuous perfusion if the femoral vein is cannulated for separate drainage of the IVC. The SVC is cannulated at the junction with the innominate vein.

While cooling the patient, the anteroinferior border of the right pulmonary artery is incised over its entire length. The incision must reach the confluence of the pulmonary

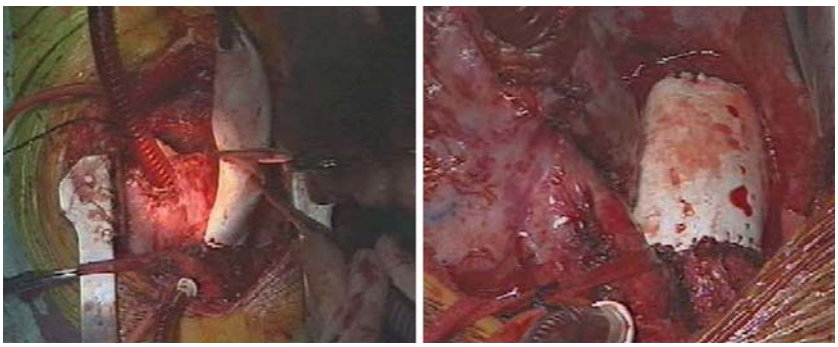

Fig. 1 The extracardiac Fontan procedure. After performing the distal conduit-to-right pulmonary artery anastomosis, gentle traction is applied to the conduit and it is divided. *Inf*, inferior; *L*, left; *R*, right; *Sup*, superior

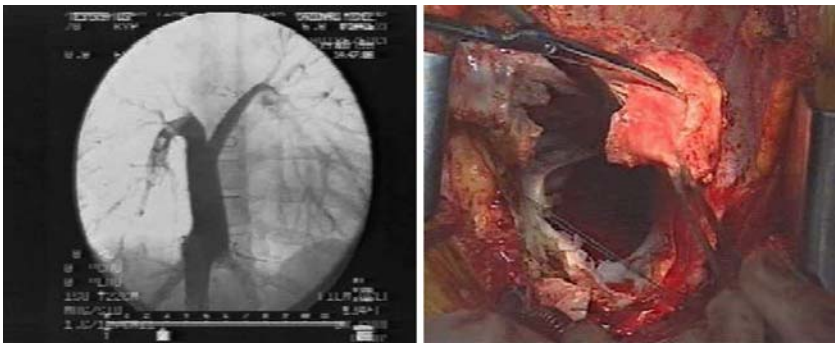

Fig. 2 The completed extracardiac Fontan procedure. *Inf*, inferior; *L*, left; *R*, right; *Sup*, superior

arteries and the left pulmonary artery origin must be seen. The polytetrafluoroethylene (PTFE) stretch conduit (we have never used one with a diameter <18 or >22 mm) is cut slightly oblique to direct the IVC blood flow toward the confluence of the pulmonary arteries and sutured in place using a 5–0 monofilament running suture.

From the atrial opening of the IVC, the atrial septum is inspected for adequate interatrial communication. This opening is then closed with a running 5–0 suture. The PTFE stretch conduit is pulled down toward the IVC and cut slightly short to place the IVC and right pulmonary artery under gentle traction (Fig. 1). The correct length of the conduit varies between 3 and 5 cm according to the patient's anatomy (Fig. 2). The anastomosis with the IVC is performed with a running 5–0 suture. If the IVC is not present, as is the case in patients with left isomerism and azygos continuation, surgery is the same because the hepatic veins usually join together before reaching the atrium. A separate vein is sometimes present. In such cases, the vein should be "unifocalized" with the others to create a common single vein.

Cardiopulmonary Bypass Technique

When we started using this technique, deep hypothermic circulatory arrest was used in all patients. The systemic venous return was drained by means of only one single cannula in the right atrium to facilitate the dissection of the IVC and the termino-terminal anastomosis between the latter structure and the conduit.

The negative effects of the deep hypothermic circulatory arrest and the ischemic cardioplegic arrest in patients with ventricular hypertrophy prompted us to begin using continuous perfusion. Single-stage cannulation is utilized at the SVC–innominate vein junction and at the femoral vein for IVC flow.

In the later patients in our series, we used continuous perfusion without aortic cross-clamping. The surgical procedure was totally extracardiac since the atrioseptectomy had already been performed at the BCPA stage.

Conduit Selection

In our early experience, we used homografts, which we thought represented the ideal long-term conduit, over synthetic materials. However, two of the three implants underwent early calcification, suggesting a nonoptimal performance in the venous position and prompting us to abandon the use of this conduit.

Later, we used woven Dacron. It is important to note that the neo-intimal (peel) formation inside the woven Dacron tubes does not develop uniformly and does not adhere firmly to the Dacron. Gradual thickening of the peel or dissection of the peel may result in complete obstruction. Due to these concerns, we abandoned the use of Dacron and used PTFE in the later TECC. PTFE is a form

Table 1 Preoperative diagnoses

Diagnostic group	No. of patients	Deaths	Takedown	Global failure	%
SV with LAVV hypoplasia or atresia	43	3	0	3	7
Heterotaxy syndrome	23	3	1	4	17
Criss-cross hearts	5	0	0	0	0
CAVC	11	0	0	0	0
Unbalanced CAVC	4				
DORV, CAVC	7				
SV with left atrial juxtaposition	6	0	0	0	0
CCA with situs solitus and dextrocardia	12	0	1	1	8
Total	100	6	2	8	8

CAVC, common atrioventricular canal; CCA, complex cardiac anomalies; DORV, double-outlet right ventricle; LAVV, left atrioventricular valve; SV, single ventricle

of Teflon in which the polymer is arranged as a lattice of nodes interconnected by filaments. A disadvantage of PTFE is persistent needle hole bleeding in the heparinized patient, which is only partially overcome by the use of PTFE suture and by the fact that suture lines are subjected to low venous pressure. We use only a 3- to 5-cm maximum conduit length between the IVC and the pulmonary artery, which promotes laminar flow from the IVC.

Fenestration

We believe that use the systemic venous pressure decompression, creating a right-to-left communication at the atrial level, is justified only when there are transient and reversible physiologic increases in pulmonary vascular resistance.

We generally place a fenestration by performing a 4- or 5-mm side-to-side anastomosis between the conduit and the right atrial free wall. Another option is to place a synthetic tube graft (6–8 mm) from the conduit to the free wall.

One Hundred Consecutive Extracardiac Conduit Fontan Operations

Patient Population

Between November 1997 and October 2003, 100 patients with complex cardiac anomalies underwent a modified Fontan operation by TECC. Preoperative diagnoses are listed in Table 1.

Ninety-three patients (93%) had previously undergone a total of 169 palliative operations before TECC. Anomalies of systemic or pulmonary venous return (or both) were present in 44 patients, and 9 patients had total anomalous pulmonary venous connection (1 infradiaphragmatic, 2 supracardiac, and 6 intracardiac). In 88 patients, the repair was staged by preliminary BCPA that was bilateral in 15 patients and associated with a modified Damus–Kaye–Stansel procedure to bypass a subaortic obstruction in 24 patients. Age and weight at operation were 67 ± 34 months (range, 19–198) and 19.8 ± 9.1 kg (range, 12–67), respectively. Forty-three patients were younger than 4 years at operation. Mean pulmonary artery pressure was 11 ± 5 mmHg (range, 6–20), mean arterial oxygen saturation was 83 ± 8% (range, 54–93), and mean end diastolic ventricular pressure was 7 ± 3 mmHg (range, 2–14). Mean hematocrit value was 50.5 ± 7% (range, 40–66) and hemoglobin content 15.9 ± 2.1 g/dl (range, 12.2–19.4).

Early Results

Early (in-hospital) deaths occurred in 6% of patients and the extracardiac conduit was taken down in two additional patients for a total early failure rate of 8%. The cause of death was myocardial failure in five patients. Pulmonary artery distortion or hypoplasia appeared to be the cause of death in one patient and the cause of failure in one patient. Atrioventricular valve regurgitation was the cause of failure in one patient. Most of the patients received inotropic agents in the immediate postoperative period. The mean duration of mechanical ventilation was 28.9 hours (range, 8.7–78.5), excluding seven patients in whom ventilatory support was prolonged (>100 hours). Mean arterial oxygen saturation on room air at discharge was 95% (range, 89–100).

Prolonged pleural and peritoneal effusions, defined as drainage for more than 10 days or the need for multiple drainage procedures, have decreased in frequency from 33% (our first experience) to 10% in this group, even without the use of fenestration.

Perioperative arrhythmias (less than 30 days after Fontan operation) were observed in 11 patients. Eight patients had transient junctional rhythm with no hemodynamic consequences. One patient had atrial flutter and underwent electrical cardioversion. Two patients had junctional ectopic tachycardia that responded to antiarrhythmic drugs. None of these patients had arrhythmias at the time of hospital discharge.

No patient underwent early surgical revision of either anastomosis. Three patients underwent plication of one hemidiaphragm because of phrenic nerve injury. Mediastinitis developed in two patients.

Late Results

Follow-up to December 2003 was available in all surviving patients. There were no late deaths. At a mean follow-up of

393 ± 20.7 months (range, 3–75), 87 patients (92%) were in New York Heart Association functional class I, 4 in class II, and 3 in class III due to moderate to severe atrioventricular valve regurgitation. Two patients had a stent placed in the left pulmonary artery. Four patients underwent drainage of a pleural and/or pericardial effusion in the late postoperative period. No neurologic complications occurred in the follow-up period.

Sixty magnetic resonance imaging studies were performed in 30 patients of our early experience to assess the prosthetic tube. The studies showed a mean reduction of the internal conduit diameter of 17.8 ± 7.6% during the first 6 months in 20 patients. There was no progression over the following 5 years (17.2 ± 6.17%, p = not significant). Even those 10 patients with the maximal internal diameter reduction did not progress over time (32.0 ± 7.9 vs 32.1 ± 7.9%).

Arrhythmias

Major late postoperative arrhythmias (more than 30 days after Fontan procedure) were present in six patients, all of whom had recurrent atrial flutter. Four of these patients were receiving antiarrhythmic drugs. In the other two patients, the flutter was associated with symptomatic bradycardia (sick sinus syndrome). These patients were treated by pacemaker implantation.

Conclusions

This 25 year long search through the best possible treatments for children with single ventricle physiology has resulted in both disillusions and several gratifying experiences.

Currently, we strongly suggest the following synthetic guidelines to the ideal candidate on orthoterminal repair with complete separation of pulmonary and systemic circulation, with negligible early mortality and a long term New York Heart Association (NYHA) class I:

1. Not all patients with univentricular heart (UVH) should undergo the extracardiac Fontan procedure;
2. In UVH with favorable streaming, a Q_p/Q_s ratio of approximately 1 to 1, and mild cyanosis, natural history might be preferable to surgical history;
3. Any form of fenestration is contradictory to orthodox application of Fontan's principle. Children in whom a planned fenestration seems necessary because of suboptimal conditions should instead undergo a combination of BCPA and associated forward pulmonary blood flow;
4. Negligible mortality should be considered mandatory in UVH, normal pulmonary arteries, and absent cardiomegaly after appropriated and correct staging

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