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Intracardiac Cavopulmonary Connection in Patients with Univentricular Heart Using Intra-atrial Lateral Tunnel and Intra-atrial Conduit Techniques

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ABSTRACT

Background: In this study, we analyzed the time course of hemodynamic efficiency and follow-up in Fontan candidates who underwent the bidirectional Glenn procedure for staged intracardiac cavopulmonary connection (ICPC).

Methods: Between 1991 and 2008, 52 patients with univentricular heart (mean age, 3.3 years; range, 2-8 years; 27 female patients [51.9%]) underwent ICPC. The cardiac malformations were as follows: tricuspid atresia, 25 cases (48.0%); common ventricle, 16 cases (30.7%); and pulmonary atresia with intact ventricular septum, 11 cases (21.1%). The intracardiac cavopulmonary procedure was indicated for all 52 cases. In 42 patients (80.7%), an intra-atrial lateral tunnel was constructed with a bovine pericardium patch. In the last 10 consecutive cases (19.3%), we performed a modified surgical technique in which we implanted an intra-atrial corrugated bovine pericardium tube sutured around the superior and inferior vena cava ostium. In all cases, a 4-mm fenestration was made to reduce the intratunnel pressure. All 52 patients had previously undergone a Glenn operation.

Results: There were 2 hospital deaths (3.8%) and no recorded late deaths. During the follow-up, all patients were medicated with antiplatelet drugs. To evaluate the hemodynamic performance, we used Doppler echocardiography, computed tomography, and magnetic nuclear resonance studies. There were no prosthesis thromboses during this follow-up period. To evaluate cardiac arrhythmias, we conducted a Holter study. The last 10 patients with an intra-atrial conduit (IAC) presented with sinus rhythm and no arrhythmias during the last 4 years. The 50 surviving patients (96.1%) have been followed up for 6 to 204 months; all these patients are free of reoperation.

Conclusion: The Glenn operation, which is performed at an early age, prepares the pulmonary bed to receive the ICPC. The midterm results of the intracardiac Fontan procedure seem to be good. The modified surgical procedure (IAC) can be a good alternative technique to the Fontan procedure in suitable patients.

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INTRODUCTION

The Fontan repair [Fontan 1971] and its modifications can be performed in selected groups of patients, and relatively low rates of mortality have been reported. Among the later modifications of the Fontan-type procedure, de Leval and associates introduced a significant alteration when they described the intracardiac cavopulmonary connection (ICPC) [de Leval 1988]. The modified Fontan procedure represents the final stage for the palliation of hearts with a single-ventricle physiology.

The introduction of the bidirectional Glenn (BDG) procedure preceding an ICPC extended the indications for the Fontan procedure [Bridges 1990; Trusler 1990]. High-risk Fontan candidates who have undergone BDG and staged ICPC (the staged strategy) have exhibited excellent clinical results [Maluf 1994; Alejos 1995; Masuda 1998]. The exact mechanism responsible for the superiority of the BDG procedure is still poorly understood, however.

In addition, several management strategies have been incorporated to reduce the mortality rate, including the use of universal risk factors that improve patient selection [Alejos 1995], the use of fenestration [Prigjian 1993], and the use of modified ultrafiltration after cardiopulmonary bypass (CPB) [Fogel 1995; Maluf 1999].

Many reports have discussed the importance of the pulmonary artery size [Sievers 1983] and systemic ventricular function in Fontan candidates. A few studies have investigated in detail the hemodynamic conditions in the Fontan circulation by focusing on ventricular efficiency. According to an evaluation with a theoretical model, Nogaki et al [2000] reported that the contractility of the Fontan circulation was lower than that of the normal circulation and that the afterload Fontan circulation was higher than that of the normal circulation.

When the ICPC is performed, the lower resting heart rate or the higher peak heart rate is frequently part of the operation history; each is weakly associated with an improved physical function. Cardiac arrhythmias are very

common in patients who undergo intracardiac lateral tunneling with a long suture line along the right atrial wall [Blaufox 2008].

The objective in the present study was to focus on the surgical results and ICPC follow-up after staged Fontan operation with 2 techniques: (1) intra-atrial lateral tunnel (IALT) and (2) intra-atrial conduit (IAC) Fontan, in which we sutured around the superior and inferior vena cava (IVC) ostium. In the latter technique, our intention was to reduce or avoid cardiac arrhythmias.

MATERIALS AND METHODS

Patient Information

Between March 1990 and December 2008, 105 patients underwent BDG procedures at our institution. We included 52 patients in this series, which consisted of 27 female and 25 male patients (mean patient age ± SD, 3.3 ± 1.0 years; range, 1-8 years). These patients underwent ICPC procedures in the Cardiovascular Division of Universidade Federal de São Paulo between March 1991 and December 2008. These patients constituted a consecutive series (Table 1). This study was approved by the Ethical Commission of Hospital São Paulo, UNIFESP.

Hemodynamic Variables

The preoperative hemodynamic variables of heart rate, mean pulmonary artery pressure, ejection fraction, and Nakata pulmonary arterial index [Nakata 1984] were evaluated in all patients in accordance with the protocol established by our institution.

The operative procedures carried out before the BDG procedure were a modified Blalock-Taussig shunt performed in 39 of the patients and pulmonary artery banding in 9 patients. In 52 patients (49.5%), the ICPC was indicated after the first stage of the BDG procedure (Figure 1).

Patient Inclusion Criteria

Indications for intra-atrial cavopulmonary anastomosis surgery included the following: diagnosis of univentricular heart, previous BDG procedure, presentation with cyanosis and decreased exercise tolerance, preserved ventricular function (ejection fraction >50% and an end-diastolic pressure of the systemic ventricle <11 mm Hg), a pulmonary pressure <18 mm Hg, a pulmonary resistance of <2.0 Wood units, a Nakata index >250 $mm^2 \times m^2$, a staged period (BDG to ICPC) of <5 years, and a New York Heart Association functional class of I to II.

Table 1. Anatomic Diagnosis

Diagnosis	Patients, n (%)
Tricuspid atresia	25 (48.2)
Single ventricle	16 (30.7)
Pulmonary atresia w/ IVS*	11 (21.1)

*IVS indicates intact ventricular septum.

Patient Exclusion Criteria

Exclusion criteria included the following: presence of pulmonary fistulae, low ventricle function (an ejection fraction <50% and an end-diastolic pressure of the systemic ventricle ≥11 mm Hg), a pulmonary pressure >18 mm Hg, a pulmonary resistance of >2.0 Wood units, a Nakata index <250 $mm^2 \times m^2$, a staged period (BDG to ICPC) >5 years, and a New York Heart Association functional class ≥III.

Fifty-two patients had a previous bidirectional cavopulmonary shunt performed by direct end-to-side anastomosis between the superior vena cava (SVC) and the right pulmonary artery (RPA) (BDG). This group was referred for an ICPC 1 to 5 years later. An IALT was indicated in 42 cases (80.7%), and an IAC technique was performed in the last 10 consecutive patients (19.2%).

Operative Technique

Anesthesia was performed via standard intravenous access with fentanyl, midazolam, and pancuronium for muscle relaxation. Although the ICPC technique was performed via aortic, SVC, and IVC cannulation to initiate CPB and to perform the surgical approach with a beating heart, myocardial preservation was achieved with a cold blood cardioplegic solution when heart arrest was necessary. We have described our surgical technique in previous publications [Maluf 1994].

IALT Technique. Forty-two patients (80.7%) had undergone a previous bidirectional cavopulmonary shunt with a direct end-to-side anastomosis between the SVC and the RPA (BDG). These patients underwent IALT operations, which were performed with a bovine pericardium patch (Figure 2).

The details of the surgical procedure used are as follows: After dissection of the BDG, the operation was performed with CPB, moderate hypothermia, and a beating heart. After

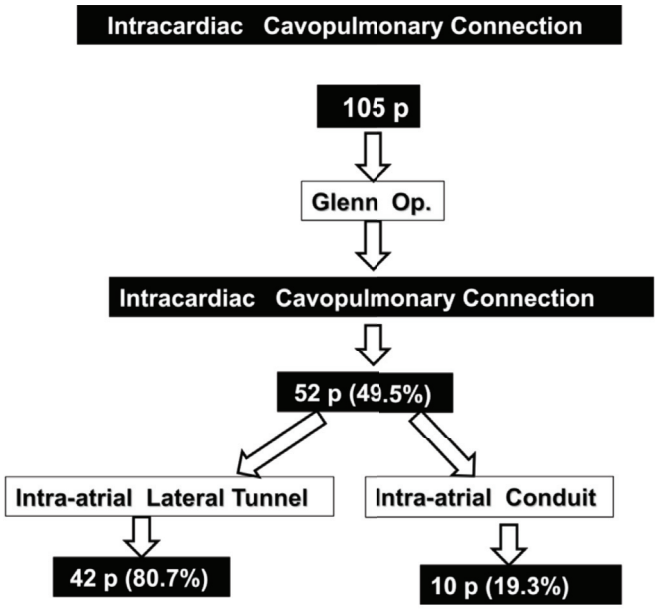

Figure 1. Staged Intracardiac cavopulmonary connection. p indicates patients; Op, operation.

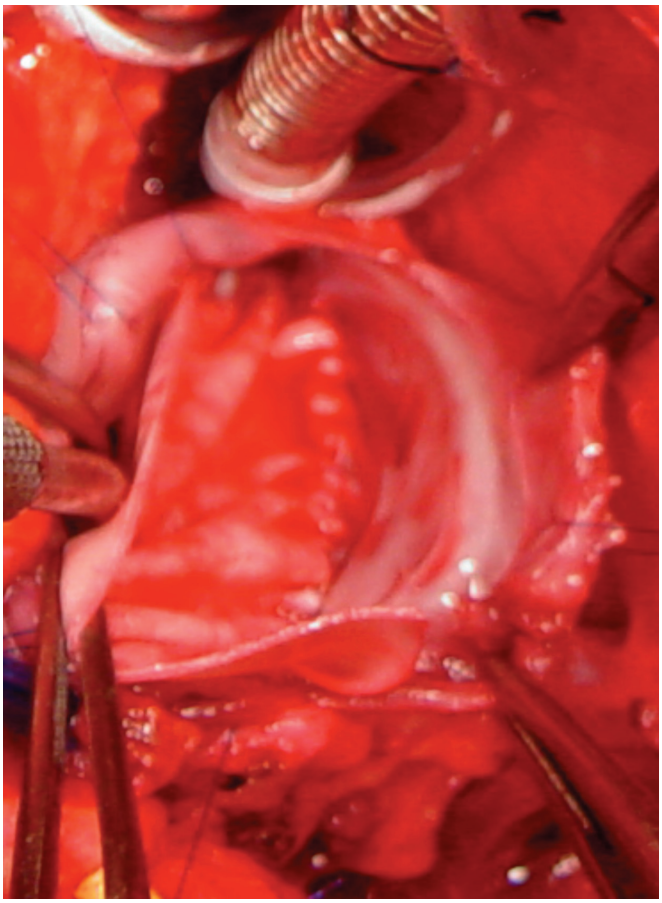

Figure 2. An intra-atrial lateral tunnel operation was performed by suturing a bovine pericardium patch to the lateral right atrium wall.

the right atrium approach, the lateral tunnel was made with a bovine pericardium patch sutured around the IVC ostium and to the lateral right atrium wall. The upper tip of the tunnel was connected to the SVC ostium. The RPA was open and stitched to the lateral tunnel by the proximal tip of the SVC. Finally, the anastomosis was enlarged with a bovine pericardium patch.

IAC Technique. In the last 10 consecutive patients (19.2%), we introduced a technique modification to avoid the lateral suture in the free wall of the right atrium, which increases the likelihood of cardiac arrhythmias (Figure 3). After a longitudinal right atrial approach, a corrugated bovine pericardium tube measuring 18 to 20 mm in diameter was sutured around the ostium of the IVC and SVC. Finally, the connection with an RPA was performed with the same technique as the lateral tunnel procedure.

A 4-mm trapdoor-type fenestration was performed in both techniques to promote its spontaneous occlusion during the follow-up period.

Statistical Analysis

The incidences of cardiac arrhythmias in the 2 groups were evaluated with the Fisher exact test. A P value of <.05 was considered statistically significant.

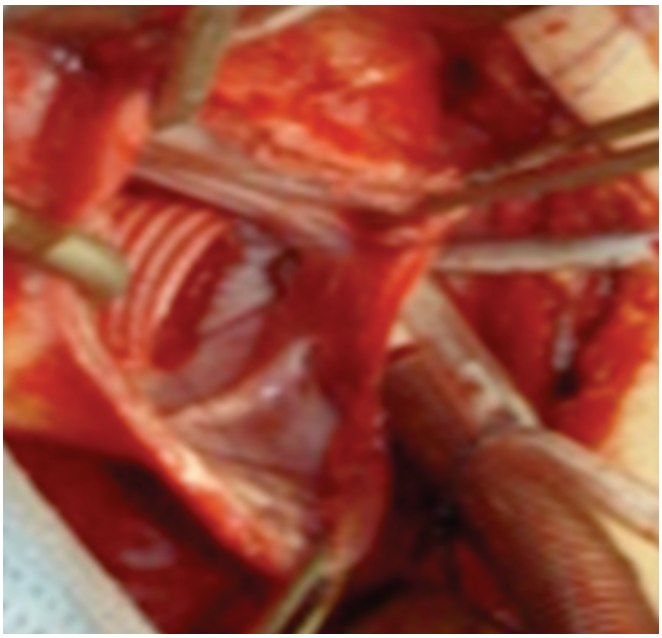

Figure 3. The intra-atrial conduit operation is performed by implanting a corrugated bovine pericardium tube around the ostium of the superior vena cava and inferior vena cava (IVC).

RESULTS

There were 2 hospital deaths (mortality, 2.8%) in the IALT group. One patient presented with a compromised ventricular function and acute renal insufficiency that was not reversed with peritoneal dialysis. The patient died 30 days after the operation. The other patient presented with multiple organ failure, with death occurring 20 days after the operation.

No mortality was recorded among the 10 patients (19.3%) who received an IAC implant.

The mean (±SD) CPB time was 114 ± 27 minutes (range, 50-152 minutes). Thirty patients underwent aortic cross-clamping, and the mean aortic cross-clamping time was 42 ± 33 minutes (range, 20-80 minutes). The mean duration of mechanical ventilation was 12 hours (range, 0-204 hours), the mean intensive care unit stay was 5 days (range, 3-29 days), the mean duration of chest tube drainage was 6 days (range, 2-38 days), the mean postoperative hospital stay was 16 days (range, 7-60 days), and the mean follow-up period was 87 ± 45 months (range, 6-204 months). All patients received antiplatelet drug therapy during follow-up (Table 2).

Postoperative problems were minimal. Medications applied during the hospital stay for symptoms of fluid retention, congestive heart failure, or both included digoxin, diuretics, and captopril. Five patients had occasional early-morning facial edema; all of these patients remained on a regimen of diuretics and captopril.

There was a significant improvement in mean O_2 saturation after surgery in both groups: IALT group, 78% preoperatively and 92% postoperatively; IAC group, 79% preoperatively and 93% postoperatively. Table 3 summarizes the postoperative data.

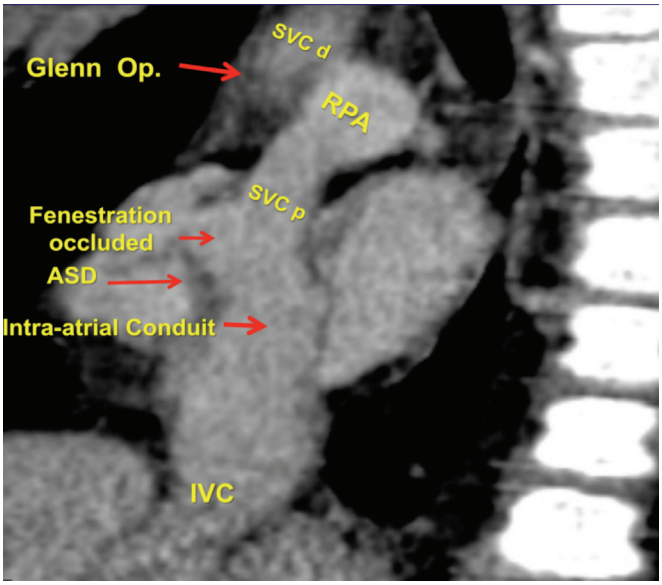

Figure 4. Image of the magnetic nuclear resonance performed in a patient 3 years after intra-atrial conduit implantation (a variant procedure). SVC indicates superior vena cava; Op, operation; d, distal; RPA, right pulmonary artery; p, proximal; ASD, atrial septal defect; IVC, inferior vena cava.

In the IALT group, which consisted of 40 surviving patients (80%), cardiac arrhythmias were observed in 13 (30.9%) of the cases. No arrhythmias were observed in the IAC group. This difference was statistically significant (P = .048). Ten patients (23.8%) received antiarrhythmic drugs daily. One patient developed late postoperative paroxysmal supraventricular tachycardia and was treated with a selective β-blocker.

All patients underwent periodic clinical and cardiac-imaging examinations. Doppler echocardiography was performed at 6-month intervals; magnetic nuclear resonance (MNR) and computed tomography evaluations were performed at 2 years postoperatively. The details of the IAC operation were evaluated in the MNR study. According to the MNR examinations, there were no cases of open fenestration by 2 years postoperatively in this group (Figure 4).

Table 2. Operative Data*

Variable	Time
CPB, min	114 ± 27 (50-152)
Aortic clamping time, min	42 ± 33 (20-80)
Mechanical ventilation, h	12 ± 7 (0-204)
ICU stay, d	5 ± 3 (3-29)
Chest tube drainage, d	6 ± 4 (2-38)
Hospital stay, d	16 ± 8 (7-60)

*Data are presented as the mean ± SD (range). CPB indicates cardiopulmonary bypass; ICU, intensive care unit.

The Kaplan-Meier actuarial curve shows that 96.1% of the patients with an intra-atrial tunnel survived. During follow-up, there were no detected thromboembolism events according to Doppler echocardiographic criteria. No reoperations were performed during that time (Figures 5 and 6).

DISCUSSION

The experience with the Fontan operation has been increasing over the years and has opened the way to a much wider application of this principle. The subsequent development of the cavopulmonary connection approach thus avoids the stasis of blood in the right atrium, as seen in the atrial-pulmonary anastomosis [de Leval 2010; Kreutzer 2010], or turbulence, as seen in cases of atrial-ventricular-arterial connections. The hemodynamic analysis of Jonas and Castaneda [1988] demonstrated the advantages of laminar flows with the introduction of new surgical techniques, with less outflow and lower incidences of arrhythmias and thrombosis. A number of major risk factors have been identified and managed with several modifications of the original Fontan procedure, and the late outcomes of the Fontan circulation are encouraging. Ventricular morphology, surgical techniques, and fenestration do not appear to influence the early or late outcome. In the preoperative preparation of the patient, it is important to detect the impaired ventricular function and the elevated pulmonary artery pressure, because they have an adverse influence on both early and late outcomes.

Table 3. Postoperative Data*

Variables	IALT Group	IAC Group	P
Group, n	42	10	—
BDG, n	42	10	—
Fenestration: tunnel/conduit, n	42	10 (trapdoor)	—
Cardiac arrhythmia, n	13 (30.9%)	0 (0%)	.048
Takedown operation, n	0	0	—
Death, n	2	0	.649
Mortality rate	4.76%	0 (0%)	—

*IALT indicates intra-atrial lateral tunnel; IAC, intra-atrial conduit; BDG, bidirectional Glenn.

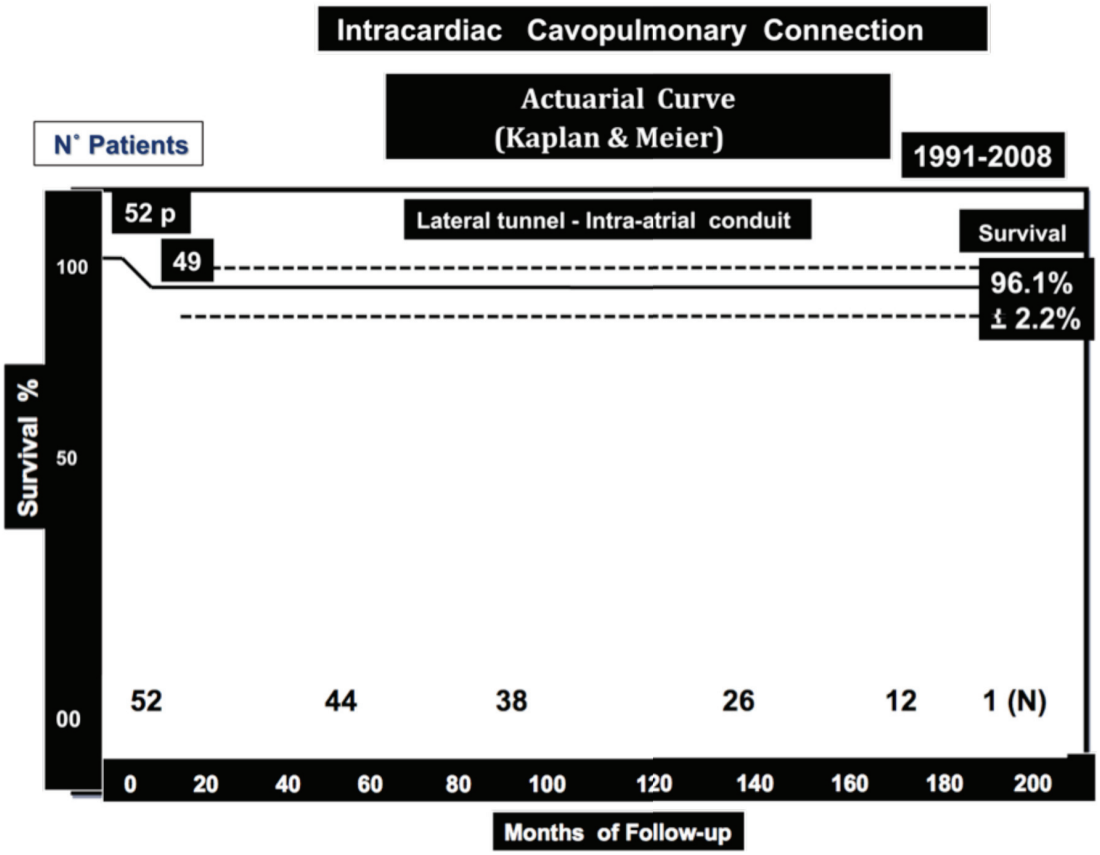

Figure 5. Actuarial curve obtained with the Kaplan-Meier method to analyze the surgical and follow-up results of intracardiac cavopulmonary connection. p indicates patients.

Reoperations are common, with a small preoperative pulmonary artery size being an additional risk factor.

This report has focused on the only 2 different options of intracardiac connection of the SVC and the IVC with the RPA for reducing cardiac arrhythmias and tunnel thrombosis (ie, IALT and IAC implantation).

In our institution, the total cavopulmonary connection is performed in a staged strategy. The first stage, the BDG procedure, was performed beginning in March 1990, and the second stage, the ICPC procedure, was performed beginning in April 1991. The BDG procedure was indicated preferentially for young patients (0.8-2 years of age), and the ICPC procedure was indicated 1 to 3 years later [Maluf 1994].

In our series, the 40 surviving patients who underwent the lateral tunnel Fontan procedure presented with 13 cases (30.9%) of cardiac arrhythmias. To prevent frequent cardiac arrhythmias, we changed the lateral tunnel technique to IAC implantation. A corrugated bovine pericardium tube is sutured around the ostium of the IVC and SVC to avoid the stitched line in the right atrium wall. None of the last 10 patients who underwent the IAC technique (and received Holter monitoring during the follow-up) presented with cardiac arrhythmias at any of 3 different times: at rest, during exercise, or asleep. None of these patients were referred for pacemaker implantation.

Gewilling et al [1992] reported that early and late arrhythmias in the postoperative period after a Fontan operation seemed to be a consequence of adverse hemodynamic function. The early follow-up results are therefore poor even when the patient has been restored to sinus rhythm. Medical and surgical modifications to improve the hemodynamic disturbances associated with arrhythmias are therefore indicated.

Other authors have reported that Fontan conversion with concomitant arrhythmia surgery and permanent pacemaker placement is safe, improves the New York Heart Association functional class, and has a low incidence of recurrent arrhythmias. In most patients, implantation of a concomitant permanent pacemaker is needed [Kim 2005; Fujii 2009].

The IALT operation produces excellent midterm outcomes, even when it is used in patients with a complex anatomy. The incidence of postoperative atrial tachyarrhythmias is low and depends largely on the underlying cardiac morphology and incidence of preoperative arrhythmia. The good midterm outcome after an IALT operation should serve as a basis for comparison with other surgical alternatives to completing the Fontan circulation [Brown 2010].

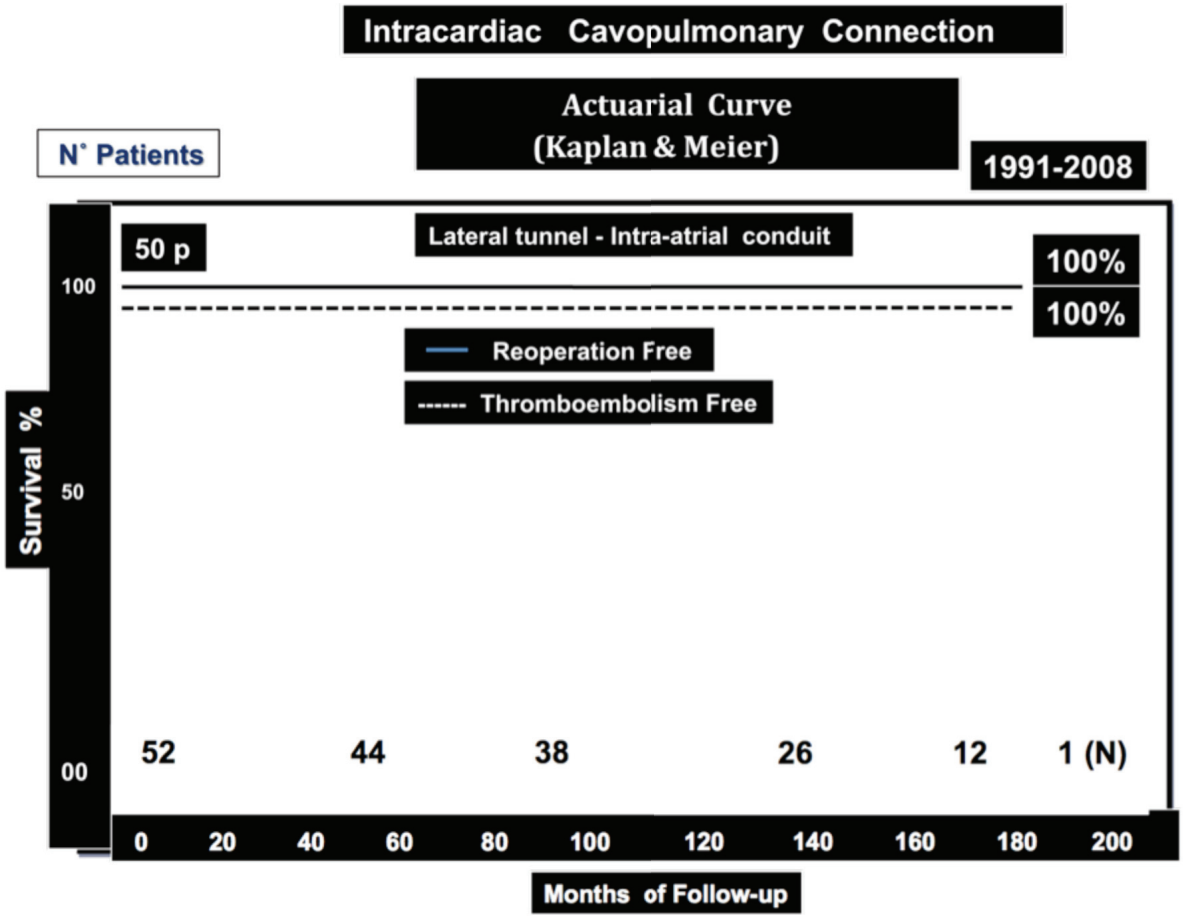

Figure 6. Probability of overall reoperation and thromboembolism over the last 9 years after lateral tunnel intracardiac conduit surgery. p indicates patients.

The outcomes after staged IALT operations are good, with comparable freedom from late reoperations and freedom from Fontan failure by the 6-year follow-up. The right ventricular morphology has been identified as a risk factor for arrhythmias [Robbers-Visser 2010].

A staged operation is considered one of the most important factors for obtaining excellent clinical results with low mortality in the treatment of high-risk Fontan candidates. Tanoue et al [2001] reported that the reduction in volume load achieved with the BDG procedure preceding ICPC allowed any afterload mismatch to be corrected, thereby improving ventricular efficiency after staged ICPC in patients.

The most significant findings in patients who undergo the Fontan procedure with a staged operation are: (1) a reduction in the volume load of the systemic ventricle, which improves contractility both after BDG and after staged ICPC; (2) afterload increases both after BDG and after staged ICPC; and (3) these changes improve ventricular efficiency during the interval between the BDG procedure and ICPC.

Improved early morbidity and mortality after the Fontan operation have been described for the clinical experience at the Mayo Clinic from 1987 to 1992 [Cetta 1996]. In our institution, the pulmonary angiographic examination results were available for the first 18 surviving patients who underwent the BDG procedure. These results showed a significant reduction in the total index of the left pulmonary artery after a mean follow-up of 23.6 months [Westphal 2001].

An alternative approach to the homogeneous distribution of SVC blood flow into the RPA and left pulmonary artery is the hemi-Fontan technique, which includes an atriopulmonary anastomosis that uses an atriopulmonary patch that directs the SVC flow into both pulmonary arteries and the IVC flow into the ventricle, thus maintaining cardiac output (modified Glenn physiology) [Douville 1991].

The extracardiac cavopulmonary connection using a nonvalved conduit described by Marcelletti et al [1990] is gradually becoming accepted as a new alternative for the surgical treatment of certain complex congenital heart diseases. Among the advantages of the use of an extracardiac conduit is the possibility of using it under normothermic CPB with a beating heart, which is important for preserving left ventricular function. The extracardiac cavopulmonary connection, however, is not always advantageous. Among the complications that can arise are thromboembolism, cellular hyperplasia, fibrosis and calcification, and the absence of growth. All are severe complications that can occur in the early follow-up

or even in the months or years after the intervention and can shadow the late evolution [Giannico 1992; Burke 1997; McElhinney 1998].

The recent national experience of Paulista et al [2003] involved 18 consecutive patients with univentricular heart who underwent total cavopulmonary connection. These patients underwent their operations with the extracardiac conduit procedure to make the connection between the IVC and the pulmonary circulation. Three patients died in the immediate postoperative period (hospital mortality rate, 16.6%). There were 4 cases of thrombosis, with 1 case evolving to death. The patients in all 4 cases underwent reintervention. Apart from the case described above, the patient in a fifth case experienced thrombosis and progressive calcification that led to total occlusion of the conduit. Therefore, the patients in this study were routinely prescribed warfarin sodium, with acetylsalicylic acid used only when use of the former was impossible or after the 12th postoperative month, when it was used for the rest of the patient's life. This precaution, however, did not prevent the occurrence of thrombosis in the conduit, which began within 24 hours in one of the patients and totally occluded the tube. This occurrence forced replacement, but it did not impede the lethal result of this complication.

The various ways of performing the Fontan operation have always been associated with a thromboembolism incidence in the postoperative period [Rosenthal 1995].

The Kaplan-Meier actuarial curve in our series of 50 patients who survived the intra-atrial connection technique and underwent 12 to 204 months of follow-up by Doppler echocardiography, MNR, and computed tomography indicated a survival rate of 96.1%, free of reoperation. We have not recorded thromboembolism events during this period (Figures 4-6). All of our patients received antiplatelet drugs (acetylsalicylic acid) only.

CONCLUSION

The bidirectional cavopulmonary anastomosis with intracardiac repair is a feasible alternative for patients with right ventricular hypoplasia. Early unloading of the functional univentricular heart by means of the BDG technique produced good conditions for completing the intra-atrial cavopulmonary connection.

Technical modifications of the intra-atrial operation (IAC implanted) have produced results with no cardiac arrhythmias and no thromboses in the prosthesis during follow-up. The modified surgical procedure can be a good alternative to the Fontan procedure in suitable patients.

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