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Fontan operation: a technique in evolution

Operação de Fontan: uma técnica em evolução

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Abstract

Objectives: **Recent experimental flow studies based on angiography and magnetic resonance have shown that total cavopulmonary anastomosis (TCPA) is a valid concept for surgical treatment of many congenital heart defects, but there is not agreement of the best surgical arrangement. The aim of this study is to assess the immediate results with three different techniques of TCPA.**

Methods: **Clinical study of all TCPA performed from January 2005 to July 2008; there were 40 patients, all with previous Glenn anastomosis, with mean age of 6.4 ± 3.2 years. Three different techniques were employed: Group 1 (G1) lateral tunnel, Group 2 (G2) extracardiac conduits, Group 3 (G3) intracardiac conduit directed to the left pulmonary branch. All patients had a fenestration performed and pre- and postoperative variables were assessed.**

Results: **G1 had 11 patients, G2 10 patients and G3 19 patients. Preoperative variables were similar in the 3 groups (*P*>0.05). Surgical mortality was higher in Groups 1 and 2 (9.1% and 10%) compared to Group 3 (0%) but there was no statistical significance (*P*=0.3841). Pleural effusion was absent in Group 3 (0%), which was statistically significant in relation to the other groups (*P*=0.0128). The length of hospital stay was also significantly lower in G3 (8 days) in relation to G1 (18 days) and G2 (13 days) (*P*=0.0164).**

Conclusion: **Intracardiac TCPA was associated with lower postoperative morbidity and is currently our preferred technique on total cavopulmonary anastomosis.**

Descriptors: **Fontan procedure. Heart bypass, right. Pleural effusion.**

Resumo

Objetivo: **Estudos recentes de fluxo com modelos experimentais de anastomoses cavopulmonares totais (ACPTs) baseados em ressonância magnética e angiografia demonstram que este é um procedimento bem estabelecido para o tratamento de várias cardiopatias, mas o melhor arranjo espacial continua controverso. Nosso intuito é apresentar os resultados imediatos com três diferentes técnicas de ACPTs.**

Métodos: **Ensaio clínico de ACPTs realizadas no período de janeiro de 2005 a julho de 2008 com 40 pacientes, com idade média de 6,4 ± 3,2 anos, com Glenn prévio. Os pacientes foram divididos em três grupos, dependendo da técnica cirúrgica empregada: Grupo 1 (G1) - túnel lateral; Grupo 2 (G2) - conduto extracardíaco; Grupo 3 (G3) - conduto intracardíaco dirigido para o ramo esquerdo de artéria pulmonar, todos com fenestração. Foram avaliadas variáveis pré e pós-operatórias.**

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Resultados: **Foram incluídos 11 pacientes no G1, 10 no G2 e 19 no G3. As variáveis pré-operatórias foram semelhantes nos três grupos (*P*>0,05). A mortalidade foi maior nos Grupos 1 e 2 (9,1% e 10,0%, respectivamente), comparadas ao Grupo 3 (zero), porém sem significância estatística (*P*=0,3841). Efusão pleural foi ausente no Grupo 3, diferença significativa (*P*=0,0128) em relação aos outros grupos (40,0% e 33,3%). A mediana do tempo de hospitalização pós-operatória foi menor no Grupo 3 (8 dias), em relação aos grupos 1 e 2 (18 e 13 dias, respectivamente) (*P*=0,0164).**

Conclusão: **A técnica de conduto intracardíaco foi associada a menor morbidade pós-operatória, sendo a opção atual do nosso serviço na anastomose cavopulmonar total.**

Descritores: **Técnica de Fontan. Derivação cardíaca direita. Derrame pleural.**

INTRODUCTION

Since the publication of the original technique of atrial-pulmonary shunt by Francis Fontan in 1971 [1], a series of surgical techniques was developed for the treatment of univentricular heart diseases [2-5]. However, hydrodynamic studies performed by de Leval et al. [5] in 1988, have shown that the presence of the right atrium produced considerable kinetic loss in the system and thus launched the concept of total cavopulmonary anastomosis. The procedure proposed by the authors, posteriorly named lateral tunnel, which consisted of to create a tunnel between the holes in the inferior and superior vena cava with a concuit prosthesis sutured on the lateral side of the right atrium provided a more laminar flow system, but it did not resulte in a significant reduction in the incidence of complications, particularly atrial arrhythmias and sinus node dysfunction [6,7].

In 1990, Marceletti et al. [8] presented a new technique of total cavopulmonary anastomosis, that consisted of a prosthetic conduit interposition between the inferior vena cava and the underside of the right pulmonary artery branch outside the heart. The purpose of this procedure was to keep the right atrium as low-pressure chamber and free of potentially arrhythmogenic sutures [9].

The techniques of cavopulmonary anastomoses have dominated the scenario of treatment of univentricular heart diseases since then, with the two techniques often used in the same institution [10]. However, pleural effusions remain a major source of morbidity in the immediate postoperative period, with an incidence in the literature of 13% to 39% [11]. In our institution, the two techniques were used concurrently until 2006 when, due to the high incidence of pleural complications, we developed a variant technique of total cavopulmonary anastomosis.

Flow studies with phantom models built in blown-glass based on magnetic resonance and angiography imaging performed by Amodeo et al. [12] demonstrated that the flow of inferior vena cava directed toward the left branch of the artery creates a vortex effect, which helps in the distribution of flow between the pulmonary branches and provides a more favorable pattern of energy saving.

Based on these observations, we use a surgical technique in which a olytetrafluoroethylene (PTFE) conduit is placed between the inferior vena cava and pulmonary artery, through the right atrium via the foramen ovale into the left atrium roof, where it is exteriorized and sutured to the trunk or left pulmonary artery branch. This technique allowed to reach the left pulmonary artery branch with a prosthesis smaller, with fewer curves and angles, thus minimizing the presence of inflexible material in the system.

The aim of this study was to review the immediate results of patients undergoing the Fontan operation at a single institution with three models of total cavopulmonary anastomosis.

METHODS

Study design

We performed clinical trial with 40 patients with univentricular physiology undergoing the Fontan operation in Biocor Institute, from January 2005 to July 2008. The data related to pre- and postoperative periods were obtained by review of electronic medical records of patients identified in the system established by the institution in January 2005. The diagnoses are shown in Table 1. This study was approved by the Ethics Committee of the hospital.

Table 1. Anatomical diagnosis in 40 patients

Diagnosis	No. of patients	Percentage [%]
Tricuspid atresia	20	50
Left ventricular double outflow tract	6	15
Pulmonary atresia with intact septum	5	12.5
Heterotaxy syndrome	4	10
Hypoplastic left ventricle syndrome	3	7.5
Others	2	5
Total	40	100

Surgical technique

All patients had previously undergone surgery of bidirectional cavopulmonary anastomosis (Glenn procedure) with or without maintaining accessory flow, as previously published [13,14], and in four of them this stage was supplemented with a Hemifontan procedure.

Until October 2006, technics of lateral tunnel and external conduit were performed, depending on the choice of surgeon. In all cases it was used cardiopulmonary bypass (CPB), normothermic and without aortic clamping in cases of external conduit, and slightly hypothermic with crystalloid cardioplegia in the lateral tunnel.

In the technique of lateral tunnel, the area corresponding to the proximal stump of the superior vena cava was anastomosed to the underside of the right pulmonary artery branch. After right atriotomy, a bovine pericardial patch was sutured around the internal opening of the inferior vena cava and continued superiorly in front of the right pulmonary veins bypassing the internal opening of the superior vena cava. A 4-mm fenestration was created directly in the patch implanted, followed by right atrial suturing. These patients constituted the Group 1.

For the external conduit it was used Dacron graft of 18 to 24 mm, interposed between the inferior vena cava and the underside of the right pulmonary artery branch. The inferior vena cava was trans-sectioned at the cavoatrial junction and anastomosed to the conduit, that passed beside the right atrium up to reach the pulmonary artery. The proximal stump of the inferior vena cava was closed with direct suture. In the face of the conduit in contact with the right atrium it was created a 4-mm fenestration. Part of the right atrial wall was excluded with side clamping, where a small atriotomy was performed, sutured around the fenestration. These patients were included in Group 2.

Group 3 consisted of all patients undergone surgery from October 2006, when the technique described below was adopted as the first option in the service. We used cardiopulmonary bypass with mild hypothermia and isothermal blood cardioplegia. After aortic clamping, it was performed conventional right atriotomy. The oval fossa, as well as all the resected interatrial septum was removed, creating a large interatrial communication. Small segment with 4 to 5 cm of the PTFE conduit of 18 to 22 mm was sutured around the internal opening of the inferior vena cava and passed through the interatrial septum into the left atrium. It was performed extensive pulmonary arteriotomy from the most proximal segment of the right pulmonary artery branch, extending to the right side of the pulmonary artery trunk, avoiding, at all costs convergence with the Glenn's flow. When necessary, the pulmonary artery was dissected and trans-sectioned at the level of the bifurcation, and the distal orifice was used as the aforementioned pulmonary arteriotomy.

The conduit was exteriorized and adjusted through a small atriotomy made in the roof of the left atrium. It was then performed running suture containing the pulmonary arterial and atrial walls, and the conduit. A 4-mm fenestration was made directly into the conduit implanted in all cases, after closure of the right atrium and removal of the aortic clamping (Figure 1).

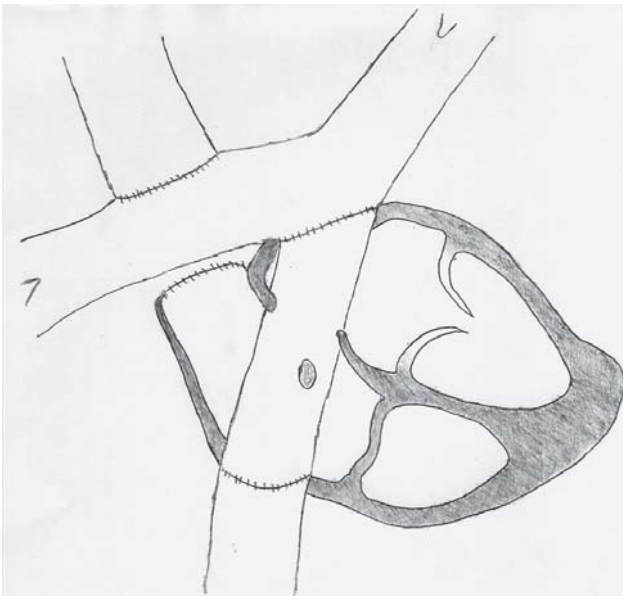

Fig. 1 – Modified Fontan operation. Notice the straight path between the inferior vena cava, the conduit and the left pulmonary artery branch. The presence of rigid material is minimized because the path is shorter

Variables studied

The following variables were collected and compared among three groups: age, weight, gender, disease, ventricular dominance, prior surgical or interventional procedures, mean arterial pressure obtained from the last cardiac catheterization performed, the oxygen saturation measured by digital oximetry on the eve of surgery, evidence of accessory pulmonary flow by cardiac catheterization or echocardiography and the presence of bicaval Glenn. As surgical variables, surgical technique used and the CPB time were recorded. The time of aortic clamping was not collected, because the cases of external conduit were performed without cardiac arrest. Durin the postoperative it was obtained the mean pulmonary artery pressure inferred by the central venous pressure measured in the catheter routinely placed in the internal jugular vein of all patients immediately after admission on the pediatric intensive care unit (PICU) and with the bed head at 30 degrees, the oxygen saturation obtained from the last gasometric laboratory analysis performed in the PICU, the presence of infection in the postoperative, the length of stay in PICU and postoperative hospit stay, the need for reoperation and the occurrence of complications and/or mortality.

It is important to emphasize that the therapeutic treatment of pulmonary vasodilation in the postoperative period was constant throughout the study, with routine infusion of milrinone in all cases and the administration of sildenafil in patients with increased pulmonary arterial pressure and low peripheral saturation.

Pleural effusion was defined as the presence of pleural drainage for more than 2 weeks, or drainage exceeding 20 ml/kg/day for more than 48 hours, or the need for repeat thoracentesis or the need of surgical or chemical pleurodesis. There was a distinction between chylous or non-chylous effusions. The criteria for persistent pleural effusion were established after review of the literature [11.15] and based on clinical observations of the service.

Statistical analysis

The hypothesis tested was that the incidence of early complications was lower with the modified technique of total cavopulmonary anastomosis surgery developed by us when compared to classical techniques of lateral tunnel and external conduit in a homogeneous group of patients who had undergone surgery in the same institution in relatively short time and with the same surgical and postoperative conditions. The data were statistically analyzed in EpiInfo software (version 6.04b, Epidemiology Program Office, Centers for Disease Control, Atlanta, United States) and are presented as mean "plus or minus" one standard deviation. The chi square test, using the Yates correction was used to compare differences between discrete variables. The Fisher exact test was used in cases of small samples in each category, when the expected value was less than five. The Student t test was used to compare differences between continuous variables with gaussian distribution and the Kruskal Wallis test when these variables did not show the same distribution. P value less than or equal to 0.05 was considered statistically significant.

RESULTS

Between January 2005 and July 2008, 40 patients underwent total cavopulmonary anastomosis at our institution, and in 11 (27.5%) the technique used was lateral tunnel; in 10 (25%) the technique of external conduit and in 19 (47.5%) the internal conduit technique. No patient required a take-down in the immediate postoperative period, due to hemodynamic failure of the cavopulmonary circuit implanted.

Table 2 shows the demographic data and preoperative variables compared between the three groups. Patients in Groups 1 and 2 were aged slightly higher when compared to Group 3, although this finding did not reach statistical significance (8.0 ± 3.7 years and 7.0 ± 4.0 years versus 5.2 ± 1 8 years). Other variables collected were similar in the three groups.

Table 2. Preoperative variables comparison in the three groups

Variables	Group 1 [n=11]	Group 2[n=10]	Group 3[n=19]	*P*
Age [mean ± sd]	8.0 ± 3.7	7.0 ± 4.0	5.2 ± 1.8	0.0819
Weight [median]	21	17	18	0.3682
Female gender	7 [63.6%]	6 [60.0%]	7 [36.8%]	0.2817
Left ventricular dominance	10 [90.9%]	6 [60.0%]	16 [84.2%]	0.1713
Previous Blalock	4 [36.4%]	6 [60.0%]	7 [36.8%]	0.4335
Previous PA cerclage	3 [27.3%]	2 [20.0%]	6 [31.6%]	0.8021
Preoperative O_2 saturation [mean ± sd]	76 ± 3	78 ± 4	78 ± 6	0.1434
Mean pulmonary artery pressure [mean ± sd]	11 ± 3	12 ± 4	11 ± 3	0.6206
Bicaval Glenn	1 [9.1%]	1 [10.0%]	2 [10.5%]	0.9921
Accesory pulmonary flow	6 [54.5%]	4 [40.0%]	6 [31.6%]	0.4651

PA= Pulmonary artery; sd = Standard deviation

Table 3. Postoperative variables comparison in the three groups

Variables	Group 1 [n=11]	Group 2 [n=10]	Group 3 [n=19]	*P*
CPB time	85 ± 28	82 ± 24	89 ± 21	0.7261
Reoperation*	1 [10.0%]	4 [44.4%]	0 [0.0%]	0.0048
Post mean PA pressure	14 ± 4	17 ± 5	14 ± 3	0.1127
Post O_2 saturation*	89 ± 3	92 ± 3	91 ± 4	0.1085
Infection*	1 [10.0%]	4 [44.4%]	2 [10.5%]	0.0701
ICU time [median]	3	3	3	0.9229
Pleural effusion*	4 [40.0%]	3 [33.3%]	0 [0.0%]	0.0128
Time of hospital stay [median]*	18	13	8	0.0164
Mortality	1 [9.1%]	1 [10.0%]	0 [0.0%]	0.3841

** Two patients were excluded due to death [one patient from Group 1 and another from Group 2]. PA = pulmonary artery; CPB = cardiopulmonary bypass; ICU = Intensive care unit*

Regarding the pre- and postoperative variables (Table 3), CPB time was similar in all groups, demonstrating that the new technique is comparable in complexity to the classical techniques. The three groups were similar also in terms of hemodynamic performance and incidence of infections. However, the occurrence of persistent pleural effusion, with high prevalence in Group 1 (40%) and 2 (33%) was not observed in Group 3 ($P = 0.0128$). Direct consequence of pleural effusion was the need for maintenance of chest tubes for longer, which reflected significantly in the time of hospital stay ($P = 0.0164$).

The hospital mortality of the whole group was 5%. There was one death in Group 1 and another in Group 2, and in group internal conduit there was no occurrence of mortality. The result, however, did not reach statistical significance.

DISCUSSION

The Fontan operation has been the surgical technique that has most changed in the pediatric cardiac surgery. Currently, it is well established the superiority of the cavopulmonary anastomosis on the atriopulmonary one [16-18], but the discussion continues about the best technique of total cavopulmonary anastomosis: lateral tunnel or external conduit [19-22]. Computer modeling of fluid dynamics of the effects of exercise on hemodynamics of two techniques recently developed have demonstrated the superiority of the external conduit when compared to the lateral tunnel [23]. For this reason, the vast majority of the techniques presented recently in the literature is based on the concept of external conduit [24-28].

In a recent article, Bove et al. [29], working on advanced computing models, analyzed the principles of the search for the ideal Fontan hemodynamics: lower power dissipation through anastomoses without sharp angles, stenosis or dilatation, reduced of the possible presence of a rigid material, fixed and impermeable; and distribution of the homogeneous pulmonary flow. Another classic study, performed by Amodeo et al. [12] showed that the direction of the flow of the inferior vena cava to the left pulmonary artery branch creates a vortex effect in the pulmonary artery, which regulates the distribution of blood between the pulmonary branches and promotes a pattern of savings more favorable compared to the situation in which the anastomoses of superior and inferior vena cava are in opposition. Based on these principles, we developed the technique of internal conduit directing the inferior vena cava to the left atrial roof, which allows to meet the majority of these premises. The path of the conduit is the most straight when compared to the existing techniques. The use of prosthetic material, that is, non-pulsatile, fixed and not permeable, is minimized, since the prostheses do not exceed 5 to 6 cm. The prosthesis is easily fenestratable and is conveyed directly to the left pulmonary artery branch. Furthermore, the intra-atrial arrangement reached with the resection of the interatrial septum allowed the use of prostheses of at least 18 mm in diameter, even in children under 3 years of life. This is consistent with the protocol established by Kirklin et al. [30] and had a direct impact on long-term outcome, with a very low incidence of reoperation for revision or replacement of conduit within 16 years of follow-up.

Stamm et al. [31] described a similar technique in patients with heterotaxy syndrome, but they conveyed the intra-atrial conduit to the orifice of the superior vena cava or to the roof of the right atrium, an arrangement very similar to the technique of lateral tunnel. In our series, only four patients presented this syndrome, and one patient in Groups 1 and 3 and two patients in Group 2. Most patients presented, therefore, the usual intra-atrial anatomy. The advantage of conveying the conduit to the roof of the left atrium through the atrial septum is the alignment that is obtained between the inferior vena cava, the conduit and left pulmonary artery branch.

Several types of material have been used in the manufacture of the shunt between the inferior vena cava and pulmonary artery [32-34]. In cases of external conduit of this experiment, Dacron tubes treated with bovine collagen were used. However, the observation of some cases of late thrombosis of the tube found in our study (no objective of this study) and study of others authors [35] led us to opt for polytetrafluoroethylene (PTFE) as the conduit of choice. This material has been widely used within the cardiac chambers, as well as a patch or conduit, with excellent results and low incidence of thromboembolic complications, even in therapeutic regimen with exclusive use of aspirin [30].

The most significant point in the present study was the low incidence of pleural effusions found in the technic of internal conduit that contributed directly to reducing morbidity and hospital stay. In the long-term period, it is well established that the pleural effusion is correlated with the incidence of protein wasting syndrome and decreased survival [17]. The mechanisms that contribute to the development of pleural effusion include hormonal, hydrostatic and inflammatory factors. The hormonal mechanisms involve the activation of the renin-angiotensin system, with involvement of the natriuretic peptide and vasopressin [11].

It is known that cardiopulmonary bypass (CPB) causes inflammatory reaction, which results in capillary leak and fluid retention. Shikata et al. [36], studying two groups of patients undergone on-pump and off-pump surgery, noticed a significant reduction in the incidence of effusion in the group without CPB. Gupta et al. [11] considered the

prolonged cardiopulmonary bypass time as independent risk factor for excessive pleural drainage volume. In this series, CPB was used in all cases and the time was similar in all groups and was not therefore decisive in the outcome. However, if the CPB has proinflammatory effects, on the other hand off-pump surgery requires the surgeon to move the anastomosis of the pulmonary artery toward the left branch due to the presence of Glenn, by creating more favorable hydrodynamic conditions. The presence of infection in the postoperative period [11] and even surgery in viral season [15] have been considered as risk factors for the occurrence of pleural effusion.

It seems, however, that the hemodynamic and hydrodynamic factors are the main responsible for early and late results of the Fontan operation, high central venous pressure, absence of fenestration, significant aortopulmonary collateral circulation [15], pulmonary vascular compliance [37], low preoperative saturation and small size of the conduit [11]. Several of these variables were tested in this study, but were similar among the three groups. Thus, we considered the good results related to the hydrodynamic performance of the technique of internal conduit.

CONCLUSION

The immediate results obtained with the total cavopulmonary anastomosis using the technique of intracardiac conduit were higher when compared to classical techniques of lateral tunnel and external conduit. The lowest postoperative morbidity expressed by the occurrence of pleural effusion, with consequent reduction in hospital stay, makes the technique of internal conduit the current option of our service in total cavopulmonary anastomosis. However, late results should be assessed, so that this preference is actually validated.

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