Late Clinical Outcomes of the Fontan Operation in Patients with Tricuspid Atresia

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Objective *- Evaluation of the long-term clinical results of the Fontan operation in patients with tricuspid atresia.*

Methods *- A retrospective analysis was made at the Instituto de Cardiologia do Rio Grande do Sul (Institute of Cardiology of Rio Grande do Sul), from August 1980 through January 2000, of 25 patients with a long-term follow-up, out of a series of 36 patients who underwent the Fontan operation or one of its variants due to tricuspid atresia. Their mean age at surgery was 5.4±3.1 years, and their mean weight was 15.8±6.1 kg, the majority of them (63.9%) being males. Four patients underwent the classical Fontan operation, 12 the Kreutzer variant, 6 the Björk variant, 9 total cavopulmonary shunt with a fenestrated tube, and 5 total cavopulmonary shunt with a nonfenestrated tube.*

Results *- The patients were followed-up on an outpatient basis, with a mean long-term survival time of 5.5±4.2 years (50 days to 17.8 years) and a late mortality rate of 8%. Arterial saturation increased from 77.2±18.8% in the preoperative period to 91±6.7% upon the last outpatient visit (p>0.05). At the final check, most (67%) patients were asymptomatic and 87% could tolerate exercise. Ten (40%) patients experienced some kind of complication during the long-term follow-up, such as cardiac arrhythmia, cyanosis, protein-losing enteropathy, neurological events, right heart failure, intolerance to exercise and reoperation.*

Conclusion *- The results indicate that, once the immediate postoperative period is over, during which the adaptations to the new circulatory physiology occur, the evolution of patients with tricuspid atresia who underwent the Fontan operation is satisfactory, in spite of a low, yet significant, morbidity.*

Keywords: Fontan, tricuspid atresia, cavopulmonary shunt

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Tricuspid atresia is a rare cardiac malformation, in which no communication exists between the right atrium and the right ventricle, bringing about complex physiological adaptations and culminating with high morbidity and mortality, thus requiring the utilization of an adequate surgical technique for its correction [1]. So, in 1971, Fontan and Baudet [2] introduced the basis for an operation that became the final palliative surgery for patients with tricuspid atresia. Basically, a communication is established between the systemic venous return and the pulmonary arterial circulation, bypassing the right ventricle flow [2,3]. As years went by, however, a number of changes were proposed to the classical Fontan operation, such as the Kreutzer technique [4], the Björk technique [5], and the total cavopulmonary shunt (TCPC) [6-9], and very few studies deal with the long-term follow-up of patients who undergo these procedures [10-18].

Considering that Fontan surgery is a relatively recent therapeutic procedure and that, in addition to this, some important changes were added to the original surgical technique, this study aims at evaluating late survival, as well as the complications that occurred during the long-term follow-up of patients with tricuspid atresia who underwent Fontan surgery or its variants.

Methods

From August 1980 through January 2000, at the Instituto de Cardiologia do Rio Grande do Sul, 36 patients underwent the Fontan operation or its variants, to treat tricuspid atresia. This being a descriptive, anterograde, retrospective, noncontrolled cohort study, the patient data were collected from patients' medical records. For this purpose, we considered as valid for the study only patients with tricuspid atresia who survived the immediate postoperative period and whose test results were expressed by reports written and signed by the doctor or technician in charge, and where the assisting doctor's own notes and the preoperative evaluation chart were available.

All patients had transthoracic echocardiography

performed at our institution to determine the preoperative anatomical diagnosis, as well as a hemodynamic study for the evaluation of the systemic and pulmonary arterial pressures, in addition to an electrocardiogram, chest X-ray, hemogram, arterial gasometry, glycemia, creatinine and e electrolytes.

The data concerning surgery were obtained from the report made by the surgeon, and the surgery date, the times of extracorporeal circulation and of myocardial ischemia, and the surgical technique used were evaluated.

The mean age at surgery was 5.4±3.1 years, and the mean weight was 15.8±6.1 kg, most (63.9%) of the patients being males. As divided by decades, 11 patients underwent surgery in the 1980s, at a mean age of 6.1±4.4 years, and 25 in the 1990s and in the year 2000, at a mean age of 4.9±2.2 years. The echocardiography and hemodynamics data are presented in tables I and II, respectively. Most (93.1%) of the diseased had a sinusal rhythm on the electrocardiogram. Thirty-four patients had alterations on the thoracic X-rays, 31 due to cardiomegaly, 17 due to pulmonary arterial hyperflow, and 27 to hypoflow. As for the palliative surgery, 28 patients had undergone at least 1 previous procedure, which included Blalock-Taussig anastomosis (n=21), pulmonary artery bandage (n=9), Glenn surgery (n=4), and aorto-pulmonary anastomosis (Cooley) (n=2).

With regard to the surgical technique used, 4 patients underwent the classical Fontan operation (2 in the 1980s and 2 in the 1990s), 12 the Kreutzer variant (8 in the 1980s and 4 in the 1990s), 6 the Björk variant (1 in the 1980s and 5 in the 1990s), 9 TCPC with a fenestrated tube (all in the 1990s) and 5 TCPC with a nonfenestrated tube (3 in the 1990s and 2 in 2000). The mean time of extracorporeal circulation was 105.4±34.2 minutes and of myocardial ischemia was 64.5±25.5 minutes; no significant difference occurred with regard to the technique used (p>0.05). The immediate in-hospital mortality was 30.5%, where 6 deaths were due to cardiogenic shock (1 Fontan, 1 TCPC with a fenestrated tube, 2 Kreutzer, 2 Björk), 2 due to multiple organ failure (both TCPC with a fenestrated tube), 1 due to cardiac arrhythmia (Kreutzer), and 2 for undefined causes (both Kreutzer). No difference existed in mortality in the immediate postoperative period between the techniques used (p>0.05). As for the surviving patients, 100%, 66.6%, 66.6%, 75%, and 58.3% underwent TCPC with a nonfenestrated tube, classical Fontan, TCPC with a fenestrated tube, Björk, and Kreutzer techniques, respectively. When the decade in which surgery was performed was evaluated, independently from the operating technique, we found that the immediate survival was 54.6% in the 1980s and 76% in the 1990s and in the year 2000. With regard to the systolic pressure of the pulmonary artery, the patients who died in the immediate postoperative period had a mean of 30.8±18.8 mmHg, and those who survived had 20.3±8.1 mmHg (p>0.05).

Table I - Anatomical variables of patients with tricuspid atresia who underwent the Fontan operation or its variants, established by transthoracic echocardiography

Anatomical variable	Patients (n=36)	%
Situs		
Solitus	35	97.2
Right atrial isomerism	1	2.8
Dextrocardia	2	5.6
Left atrioventricular valve		
Normal	32	88.9
Valve failure	4	11.1
Ventricle-arterial connection		
Concordant	22	61.1
Discordant	7	19.4
One-way exit	6	16.7
Double-way exit of right ventricle	1	2.8
Associated anomalies		
Pulmonary stenosis	8	22.2
Pulmonary atresia	3	8.3
Persistence of arterial channel	1	2.8
Coarctation of the aorta	1	2.8

Table II – Preoperative pulmonary artery and left ventricle pressures of patients with tricuspid atresia who underwent the Fontan operation or its variants

Variable	Mean
Pulmonary artery	
Systolic pressure (n=23)	23.2 + 12.9mmHg
Diastolic pressure (n=23)	10.7 + 5mmHg
Mean pressure (n=24)	15.8 + 6.3mmHg
Left ventricle	
Systolic pressure (n=29)	81.7 + 14.7mmHg
Final diastolic pressure (n=26)	11.5 + 6mmHg

The follow-up of these patients was done with regular check-ups in the pediatric cardiology outpatient service of our institution, from where if necessary they were referred for transthoracic echocardiography or a new hemodynamics study. An evaluation was made of the incidence of complications and reoperations, arterial gasometry, hematocrit, and long-term survival. The time of survival after the Fontan operation was determined by subtracting the date of surgery from the date of the last outpatient check-up (or the date of death).

Statistical analysis of the data was performed with the EPI-INFO 6.0 and SPSS by frequency calculation programs, the Student *t*, Fischer exact, and chi-square tests, and the Kaplan-Meier actuarial curve; the established significance level was 5%.

This study was evaluated and approved by the ethics committee at our institution.

Results

Twenty-five patients were followed-up ever since they underwent the Fontan operation or one of its variants, with a mean survival time of 5.5±4.2 years (from 50 days to 17.8 years) (Fig. 1).

The mean hematocrit decreased from 52.6±8% in the preoperative period to 44.1±10% in the late postoperative period (p>0.05); the mean arterial saturation, available for 10 patients, increased from the preoperative 77.2±18.8% to 91±6.7% (n=5) at the last outpatient visit (p>0.05).

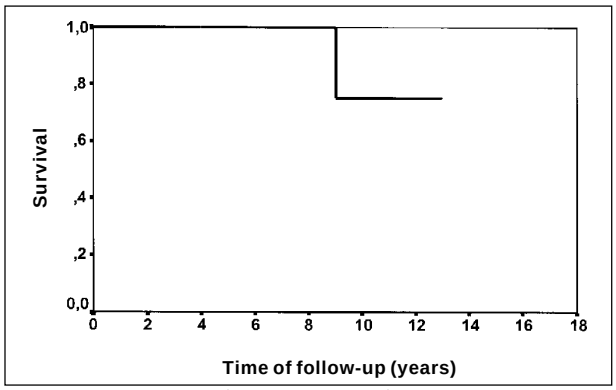

Fig. 1 - Kaplan-Meier curve of long-term survival of patients with tricuspid atresia w**ho underwent** the Font**an operation** or its variants, with exclusion of immediate mortality. (Ordenada): Survival; (Abscissa): Time of follow-up (years)

Seventeen patients underwent a control echocardiography, on which a pericardial spill with no hemodynamic repercussion was seen in 1 patient; a residual interventricular communication was found in 2 patients, and in another a minimal mitral regurgitation. Cardiac catheterization was performed in only 4 patients, 2 of which were reoperated on later.

Ten patients experienced some complications during the long-term follow-up (tab. III). Of the patients with neurological events, 1 had mental retardation and another had a seizure, both having undergone surgery in the 1990s. Three patients experienced cardiac arrhythmia, 2 having been operated on in the 1990s and 1 in the 1980s; 1 patient had an atrial flutter, 1 had paroxysmal atrial tachycardia, and another had atrial fibrillation. Two patients who had been operated on in the 1980s were reoperated on during the follow-up period: 1 patient 7 years and 5 months after a classical Fontan due to anasarca resulting from intratubular fibrosis that was resected, and a Dacron graft used; another patient 8 years after a Kreutzer procedure due to venous congestion resulting from stenosis of the valvate tube, replaced by a heterologous pericardium graft at the atriopulmonary connection.

On the last check-up, most (67%) of the patients were asymptomatic, and 87% tolerated exercise, based on direct patient questioning (a method resembling that used by Gentles et al [16]).

Two (8%) patients died during the long-term follow-up, 1 due to cardiogenic shock, 17 and 1/2 years after a classical Fontan performed in 1981, and another due to multiple organ failure, 8 years and 7 months after Kreutzer surgery performed in 1989.

Discussion

Tricuspid atresia, anatomopathologically described ever since the XIXth century [1], has, until our time, undergone a continuous evolution regarding its surgical handling. Having been attributed a high mortality rate until the early 1970s, this complex heart disease had its natural history changed by the surgical technique described by Fontan and Baudet [2], giving rise to the so-called Fontan Era [14]. An important aspect to be taken into consideration in the new circulatory physiology resulting from this surgery is that, after this procedure, the systemic and pulmonary vascular resistances are serial, and therefore the energy responsible for the blood flow in these 2 systems comes from a single ventricle. As it is a recent procedure, we have no conclusive answers yet whether this remodeling is beneficial to the good performance of ventricular function in the long run [1].

In this study, we evaluated 25 patients with tricuspid atresia and an average follow-up of about 5 years after Fontan surgery or its variants, similar to that reported by other authors [15,16,19]. Our patients were operated on, on the average, at about 5 years of age, as with other groups [16,19,20]; they had a tendency toward being younger during the last decade of our experience, like in the Toronto [15] and Boston [17] groups. Older age at surgery is a risk factor of late mortality after the Fontan operation, because the prolonged volume overload in the working ventricle and persistent cyanosis contribute to the progressive deterioration of the cardiac muscle [1,19], although Yeh et al [15] were unable to prove a significant increase in survival by anticipating the moment of surgery.

Although the immediate mortality in this series was higher than that reported in the literature [1,10-13,16,17], the late mortality in our group of patients was similar to that found by other authors [15,19], reaching a mean survival of 5.5±4.2 years. Yeh et al [15] found that 65.8% of their patients were alive 20 years after Fontan surgery. Weipert et al [18] evaluated long-term survival in patients with tricuspid atresia who

Table III - Complications experienced during long-term follow-up of patients with tricuspid atresia who underwent the Fontan operation or its variants, according to the technique used (n=25)

Complication	Fontan (n=3)	Kreutzer (n=7)	Björk (n=4)	Total cavopulmonary shunt with fenestrated tube (n=6)	Total cavopulmonary shunt with nonfenestrated tube (n=5)
Cardiac arrhythmia	2	-	-	1	-
Cyanosis	-	2	-	-	1
Protein-losing enteropathy	2	1	-	-	-
Neurological events	-	1	-	-	1
Right heart failure	1	-	-	1	-
Intolerance to exercise	1	1	-	-	1
Reoperation	1	1	-	-	-

underwent the right atrium-pulmonary artery or Björk techniques, and found a survival rate of 15 years in about 70%, for both techniques.

In our group of patients, a high mortality occurred in the immediate postoperative period. However, if we compare mortality by decades, it decreased by 53% from the 1980s to the 1990s and the year 2000. This decrease was also observed by other authors [10,11,20,21], proving the technical improvement inherent in to the accumulated experience and to the incorporation of new techniques. In the Boston group [11], immediate mortality in their initial experience was 27%, rather similar to that in our group.

We must also consider that the patients who died in the immediate postoperative period had a tendency toward higher pulmonary blood pressure than those who survived, and this correlation was reported as being significant by Mayer et al. [17]. The presence of pulmonary arterial hypertension can be a considerable determinant of survival, because the systemic venous return will have to overcome the high pulmonary arterial resistance for a long time. Thus, with the increase in resistance in the pulmonary arterial bed, an early failure of this serial circulatory system can occur, which may explain, at least in part, the high immediate mortality.

By comparing the surgical technique used and the immediate survival, we observe that all patients with TCPC and the great majority of those who underwent classical Fontan, Björk and TCPC with a fenestrated tube survived the immediate postoperative period. As for the late postoperative period, likewise no patient with TCPC, fenestrated or nonfenestrated, and Björk died, whereby most of these techniques began to be used only in the decade of the 1990s and in the year 2000.

When we evaluated the functional capacity of our patients, even though based on a subjective answer, we found that most of them could tolerate exercise on their last outpatient visit. Similar to our results and measuring methodology, Gentles et al [16], from Boston, observed that 91.3% of their patients were in excellent or good physical shape, 5.4 years on the average after a Fontan operation. The same authors showed that, the longer lasting the follow-up period of patients who had symptoms, the worse the patient's functional class, suggesting that the new circulatory physiology brings about a progressive decrease in cardiac performance. Driscoll et al [22] found improvement in physical tolerance and cardio-respiratory response in patients with tricuspid atresia after a Fontan operation, although they nevertheless still remained below normal, a fact observed as well by other authors [18], who, in addition to that, did not find any difference between Björk and the right atrium-pulmonary artery regarding the ventilatory response. On the other hand, Driscoll et al [22] reported a worse physical tolerance with increasing age, most probably due to the chronic volume and pressure overload in the single ventricular cavity. This physiopathological outcome will often culminate with the inevitable indication of a heart transplant for these patients [11]. In this sense, Fontan et al [22] also consider this operation a palliative procedure, because their patients also showed a decreased functional capacity in the long run.

Although the arterial saturation in the late postoperative period of only 5 patients was available to us, its mean value was 91±6.7%, rather similar to that found by Gentles et al [16]. It is important to consider that the other patients did not have this measurement made in their long-term follow-up because, as they were seen on an outpatient basis and were in stable clinical conditions, no need existed to request such a test. Thus, we can presume that this mean reflects the approximate levels of the patients without this test. Besides that, a tendency existed towards higher arterial saturation during the late follow-up, as compared with that in the preoperative period. Likewise, the hematocrit showed a tendency toward being lower in the late postoperative period. These 2 facts together indicate a better state of late oxygenation after Fontan surgery or its variants.

When we analyzed the complications that occurred according to the surgical technique used, we found that they tended to be less prevalent in Björk and fenestrated or nonfenestrated TCPC patients, although Gentles et al [11] concluded in their study that the long-term follow-up results are not directly related to the performed procedure. Cyanosis, present in 3 patients, may arise out of a deficient pulmonary flow, deriving from high pulmonary arterial resistance or decreased pulmonary perfusion pressure [1]. The development of cardiac arrhythmia, which appears to occur less with TCPC, is another severe complication of difficult clinical control that is more prevalent the worse the functional class is and the longer the follow-up time [1]. Bando et al [13] observed arrhythmia in 6 (4.7%) of their patients, in greater association with large fenestrations or right atrium-pulmonary artery anastomosis. In our series, 3 patients had arrhythmia, 2 having had a classical Fontan and 1 TCPC with a fenestrated tube. Protein-losing enteropathy, present in 3 (12%) patients during the long-term follow-up, had a higher prevalence than that (2.5%) reported by Gentles et al [16], and by Bando et al [13], in whose patients this complication did not occur. Two (8%) of our patients had neurological complications. In the Boston [23] and Indianapolis [13] groups, a cerebrovascular accident occurred in approximately 2% of the patients. The Boston [23] authors found that the risk of this neurological complication lasts from the immediate postoperative period until 2 and 1/2 years of follow-up. The prevalence of reoperations during long-term follow-up was 8% in our study, whereas the literature reports a variety of data, such as 20.1% [16] and 8.6% [15].

Despite the fact that this series consists of a small number of patients, our results are comparable to those in the literature. This and other studies prove that the Fontan operation or its variants give satisfactory results in the long run, contributing to considerable survival. In addition to that, the improvement in quality of life brought about by the

operation has as a result, among the benefits discussed above, an improvement in the patient's social development conditions [16]. In this series, Björk and TCPC patients had fewer complications and no late deaths. In addition to that, a clinically important improvement occurred in arterial oxygenation, and a great number of patients had satisfactory physical capacity. Thus, almost 30 years after its first description, the Fontan operation proves to be an important therapeutic step for patients with tricuspid atresia. The evolution of the technique used, with the current preference for the total cavopulmonary shunt with intra- or even extracardiac morphology, associated with better preoperative selection and larger surgical experience, has contributed to better immediate and long-term results.

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