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RESEARCH ARTICLE

Prognosis of children with cardiomyopathy submitted to Fontan surgery: 30 years of experience at the Hospital Infantil de México Federico Gómez

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ABSTRACT

Background. At the present time, the Fontan procedure is indicated for patients with univentricular cardiac pathology. In Mexico, few tertiary centers are performing Fontan surgery in the pediatric population. The objective of this study was to analyze the results of patients with congenital heart disease with univentricular physiology who have undergone Fontan surgery since 1980 in the Hospital Infantil de México Dr. Federico Gómez.

Methods. We present a retrospective analysis of pediatric patients undergoing Fontan surgery. We included variables related to preoperative mortality and morbidity along with those related to surgery and to the postoperative period. A bivariate analysis with the comparison of averages and odds ratio was performed. Kaplan-Meier survival curves were constructed.

Results. From 1983 to 2012, 53 Fontan surgeries have been performed. Of the total, comprehensive data of 32 patients were gathered. The most common congenital heart anomaly was tricuspid atresia IB (39.4%). According to type of surgery, in 48% of patients extracardiac Fontan was performed followed by intracardiac Fontan (24%). In 87% of patients fenestration was performed. The most common complications were acute renal failure, neurological problems and pleural effusion. Overall survival was 65.6%; 90.9% of patients died within the first month of surgery. Patients have been reported with >20 years of survival (average actuarial survival 14.5 years). Variables associated with mortality were younger age at surgery, type of Fontan performed, surgical technique complications, lack of fenestration and acute renal failure.

Conclusions. Evolution of Fontan surgery during the past 30 years has been satisfactory. In patients undergoing this procedure, mortality and quality of life have shown improvements.

Key words: Fontan surgery, congenital heart disease, prognosis.

INTRODUCTION

It is estimated that approximately half the incidence of congenital heart disease (CHD) varies between 3 and 8/1000 of live newborns. This frequency represents about 10% of all birth defects. Without treatment, 25% of patients with CHD die during the neonatal period, 60% in childhood and only 15% surviving to adolescence.[1]

In general, management of these patients is surgical. Depending on the type of heart disease, surgical treatment can be done for total or definitive correction (which occurs in most cases) or palliative surgery (in the case of patients with a heart disease in which only one of the ventricles is functional). In few situations is a cardiac transplant required.[2]

The concept of palliation for treatment of congenital cardiopathies has as a goal to alleviate signs, symptoms or pathophysiological situations of the disease that are limiting in terms of probability of death or developing other irreversible disorders difficult to manage (i.e., pulmonary hypertension, cyanosis or heart failure). In general, per-

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forming these procedures is temporary while a definitive correction is carried out, directed at repair, as much as possible, to anatomic and physiological changes in the original congenital cardiopathy. In some cases, palliative procedures may have a definitive character due to the complex nature of the congenital cardiopathy.[2,3]

The concept of "repair" of univentricular type implies that patients need a sequence of palliative interventions that end, in the best case scenario, in a total cavopulmonary bypass, also called Fontan procedure. The surgical principle consists of establishing a connection between systemic venous return and the pulmonary artery branches and of the only functional ventricle with the systemic circulation, with the goal of decreasing the work of the functional ventricle, separate the pulmonary venous blood from the systemic and improve hypoxemia.[4] This technique was first applied in 1971 by Francis Fontan and, despite multiple modifications, still does not guarantee a definitive cardiovascular functionality.[2,5] Currently, Fontan surgery is indicated in patients with univentricular heart pathologies such as the following:[2,5]

- Complex congenital heart diseases that behave as a sole functional ventricle of left, right or indeterminate morphology
  1. Hypoplastic left heart syndrome
  2. Hypoplastic right heart syndrome
     a. Pulmonary atresia with complete septum, with severe hypoplasia of the right ventricle
     b. Tricuspid atresia
     c. Some severe forms of Ebstein disease
  3. Heterotaxy syndromes with left or right isomerism
  4. Criss-cross type anomalies
- Heart disease with marked ventricular disequilibrium associated with the baseline defect
  1. Atrioventricular canal
  2. Double outlet right ventricle
- Heart disease with multiple "Swiss cheese" type intraventricular communications

Initially criteria of Choussat were formulated as the ideal conditions for performing Fontan surgery.[6] However, these are not always followed. These criteria are as follows:

1. Patients age >3 years
2. Pulmonary resistances <2 Wood units
3. Pulmonary artery pressure <15 mmHg
4. Ejection fraction >45%
5. Left ventricular end-diastolic pressure <10 mmHg
6. Minimal mitral regurgitation
7. Adequate diameter pulmonary arteries (McGoon index >1.8)

From the first description of the Fontan procedure, some modifications have been made to the technique, improving survival. During the 1970s, survival ranged between 75 and 83% and is currently ~90%.[7-9] Also, risk factors for mortality have been described, such as not meeting the Choussat criteria, type of heart disease, type of surgery performed, absence of fenestration in the conduit that connects the systemic circulation with the pulmonary, pump time and anatomy of the atrioventricular (AV) valve.[10,11] Other factors are those associated with early failure.

In a study conducted at Boston Children's Hospital it was reported that the following variables were associated with failure: pulmonary artery pressure of >19 mmHg, heterotaxy syndrome, young age at the time of surgery, a tricuspid valve as the only atrioventricular system, distortion of the pulmonary artery, atriopulmonary connection originating in the right appendage or right atrium, absence of fenestration and prolonged pump time during surgery.[7,8]

There are few tertiary level care centers in Mexico that perform Fontan surgery in the pediatric population, in addition to there being scarce information on the prognosis of these patients. In a study performed at the National Institute of Cardiology Ignacio Chavez, a mortality rate of 28% was reported. It was determined that the variables associated with a greater risk of death were absence of fenestration, left atrial pressure >10 mmHg and pulmonary artery pressure >20 mmHg.[4] The objective of the present work was to analyze the results of the patients with congenital heart disease of univentricular physiology who underwent Fontan surgery at the Hospital Infantil de México Federico Gómez (HIMFG) since 1980.

SUBJECTS AND METHODS

We performed a retrospective analysis of all patients who underwent Fontan surgery from 1980 to date in the HIMFG. A search was conducted of the surgical reports in the clinical records. From all records, patient-related variables were recorded, the procedure and postopera-

tive events. Among the variables related with the patient were included the age, gender, type of heart disease, prior palliative surgery, time of the last re-operation, complications during the last surgery, distortion of the pulmonary artery (stenosis, distortion in the central pulmonary artery or marked hypoplasia in the pulmonary branches, which should have been documented with a cardiac catheterization or during surgical findings), as well as the variables related with the Choussat criteria.[6]

Among the considered variables related to the surgical procedure were type of cavopulmonary bypass, fenestration in the conduit connecting the systemic circulation to the pulmonary circulation, development of technical complications during the procedure, as well as the extracorporal circulation and aortic clamp time. Furthermore, postoperative variables considered were days of intubation, use of amines, stay in surgical therapy and hospital stay, as well as the development of neurological, renal, or infectious complications, arrhythmias, pleural effusion or death. Operative mortality was defined when it occurred within the first 30 days after surgery, late mortality when it occurred after this time period. Among the patients who survived, the period of time from the Fontan surgery to date was identified.

Statistical analysis

Qualitative variables are presented as absolute numbers and percentages, the quantitative as median and minimum and maximum values. χ^2 was used for comparison of proportions and Mann-Whitney U test to compare medians. For analyzing the possible factors associated with mortality and morbidity, we calculated odds ratios (OR) and 95% confidence intervals. For mortality, Kaplan-Meier survival curves were done; p values <0.05 were considered statistically significant. All analyses were performed using SPSS v.15.0 statistical package (Chicago, IL).

RESULTS

From 1983 to 2012, there were 53 patients admitted to the HIMFG with congenital heart disease who were scheduled for total cavopulmonary anastomosis or Fontan surgery. Of these, four procedures were done in 1983-1990, 23 in 1991-2000, and 25 from 2001-2012. The analysis included 32 patients because the complete records for 15 patients were not located. An additional six patients were excluded because it was not possible to perform Fontan surgery due to hemodynamic instability at the time of the start of the surgical procedure. It should be mentioned that some of the variables considered were not described in the records of the 32 patients included.

Of the 32 patients, 22 (68.8%) were male and 10 (31.2%) female. The age at the time of Fontan surgery ranged from 8 months to 16 years. Table 1 shows the criteria for selection of patients eligible for this type of surgery. It was observed that with the exception of age and pulmonary vascular resistance, in general the criteria for the Fontan procedure were met.

According to type of heart disease, diagnoses encountered were tricuspid atresia IB (39.4%) followed by double entry path to the left ventricle (12.1%), right ventricular hypoplastic syndrome (9.1%), polysplenia visceral heterotaxy (6.1%), tricuspid atresia IC, IIA and IIB (3% each), asplenia visceral heterotaxy (3%) and other less frequent diagnoses in 21.3% of cases. Moreover, as important antecedents it was found that 21 patients (65.6%) had undergone a prior palliative surgery. Of these, 13 (61.9%) had undergone a Glenn cavopulmonary procedure, five (23.8%) had a modified Blalock-Taussig, two (9.5%) had pulmonary artery banding, and there was one (4.7%) atrial septostomy. Of the 13 patients who underwent Glenn procedure, most (n =11) had no surgical complications. One patient had significant bleeding that required surgical re-exploration, and the other had veno-venous fistulas and pericardial effusion. In these 13 patients, the mean time between Glenn procedure and Fontan surgery was 10 years (minimum 6, maximum 15 years).

According to type of surgery, 48% underwent extracardiac fenestrated Fontan, 24% intracardiac fenestrated Fontan, 20% atriopulmonary bypass surgery, and 8% Kawashima type surgery. Of the principal technical complications that occurred when performing surgery, there were firm adhesions and bleeding (13%) and difficult anatomy of the heart itself (17.4%). The remainder (69.6%) had no technical complications. In 87%, fenestration of the conduit connecting the systemic circulation to the pulmonary artery was left. In 90%, extracorporeal circulation was used with a mean of 150 min (minimum 80, maximum 515 min). In 51.7% of patients aortic clamping was done, with a mean of 60 min (minimum 8, maximum 180 min). Only in 17.9% was it necessary to perform circulatory arrest, where the mean was 53 min (minimum 5, maximum 110 min).

With regard to the variables during the postoperative period, the average days of amine use was 4 (minimum 1, maximum 13 days). The average days of intubation was 2 (minimum 1, maximum 22 days). Of the total, 25 (78.1%) patients developed complications (Table 2). The principal complications were acute renal insufficiency (28.1%), neurological (21.9%), pneumonia (15.6%), sepsis (15.6%), and pleural effusion (15.6%). Of the nine patients with renal insufficiency 33% required peritoneal dialysis. With reference to neurological complications, two patients had ischemia or cerebral infarction, two hemiparesis, one had seizures, and two patients had brain death. Regarding infectious complications, in five patients nosocomial pneumonia was documented and in another five patients nosocomial sepsis was detected. There was mediastinitis in 3.1% and it was necessary to carry out surgical debridement. In total, 15 patients (46.8%) required antibiotics to treat infectious complications; the average time of use was 7 days (minimum 1, maximum 21 days). In total, the average intensive care stay was 7 days (minimum 1, maximum 40 days) and the average hospital stay was 17 days (minimum 1, maximum 182 days).

Of the 32 patients subjected to Fontan surgery, 11 died (34.3%). The medical record described the cause of death in seven patients. In three patients it was related to neurological complications, in one patient due to severe ventricular dysfunction, in another acute pulmonary edema, one was secondary to severe bleeding and the last due to thrombosis of the tube that bypasses the systemic circulation to the pulmonary. Analysis of mortality demonstrates that it has decreased through time according to decades: 1980-1990 (50%), 1999-2000 (26%), and 2001-2010 (12%). Patients who survived have been followed for several years (Figure 1). On average, survival was 14.5 years although there was a patient who had been followed for >22 years after surgery.

Of the patients who died, 63% occurred within the first 5 days of the postoperative period and 90.9% within the first 30 days. It is notable that only one patient died at 6 months after surgery.

Risk factors associated with morbidity and mortality

Table 3 presents the results of univariate analysis of risk factors associated with mortality in patients undergoing Fontan surgery. As seen, variables that reached statistical significance were type of surgery performed (in particular, atrioventricular shunt), presence of significant bleeding or difficult anatomy, lack of fenestration of the conduit connecting the systemic circulation with the pulmonary, and development of postoperative renal insufficiency. However, despite not reaching statistical significance for circulatory arrest and the presence of atrioventricular valve failure, the OR were ≥5.0 but with very wide intervals (95% CI). On the other hand, on analyzing the quantitative variables it was determined that the median age of patients who died was lower than those who survived (5 years *vs.* 8 years), which was statistically significant ($p = 0.012$). Averages both for oxygen saturation before surgery, pulmonary artery pressure, ejection fraction, and left ventricular end-diastolic pressure were similar between groups ($p > 0.05$).

Factors related with morbidity were also analyzed. For acute renal insufficiency, factors found to be associated

Table 1. Characteristics of patients at the time of Fontan surgery

Characteristic	*Median (minimum-maximum values)*
Age (years)	7.5 (0.6-16)
Ejection fraction (%)	68.8 (49-88)
Pulmonary vascular resistance (μW)	2.4 (0.48-4.28)
Pulmonary artery pressure (mmHg)	8.4 (3-15)
Telediastolic pressure of LV (mmHg)	5.21 (0-12)
Left atrial pressure (mmHg)	3.86 (0-10)
Nakata index	210. 5 (111-323)
McGoon index	1.9 (1.48-3.19)

LV, left ventricle.

Table 2. Principal complications in Fontan surgery patients

Complication	*n*	*%*
ARI	9	28.1
Neurological complications	7	21.9
Pneumonia	5	15.6
Nosocomial sepsis	5	15.6
Pleural effusion and chylothorax	5	15.6
Cardiogenic shock	5	15.6
Arrythmias	4	12.5
Bleeding	3	9.4
Mortality	11	34.3

ARI, acute renal insufficiency.

were the type of Fontan performed (OR 5.3, 95% CI, 0.52-54.0), technical complications during surgery (OR =2.4, 95% CI, 0.261-22.10), lack of fenestration (OR =2.5, 95% CI, 0.16-37.0), and aortic clamping (OR =5.5, 95% CI, 0.54 to 55). Other variables such as age, pulmonary artery pressure, pulmonary vascular resistance, alteration in the anatomy of the pulmonary arteries, ejection fraction or infectious complications were not statistically significant.

With regard to neurological complications, risk factors were type of Fontan performed (OR =2.16; 95% CI 0.26-17.8), technical complications (OR =6.0; 95% CI, 0.67-53.0), lack of fenestration (OR =9.3; 95% CI 0.62-139.0) and circulatory arrest (OR =4.2; 95% CI 0.45-40.0). Finally, it was determined that prior to Fontan surgery, patients who presented cardiogenic shock had lower oxygen saturation (66 *vs* 75 mmHg, p =0.043) as well as ejection fraction (65 *vs* 70%, p = 0.05); additionally, circulatory arrest was a factor related with its presentation (OR =14.6; 95% CI 0.99-215.0).

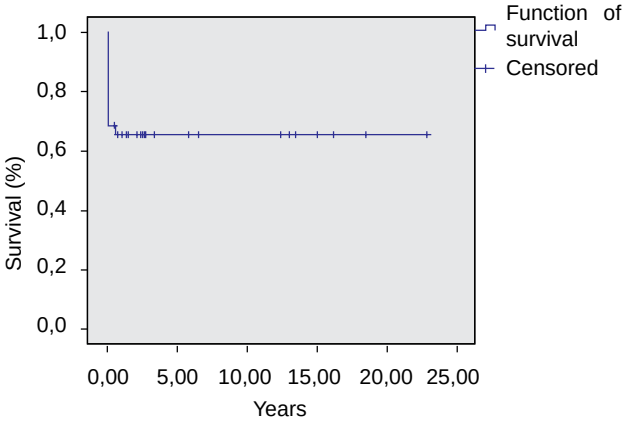

Figure 1. Survival curve of pediatric patients undergoing Fontan surgery.

DISCUSSION

Fontan surgery involves making a connection between the right atrium and the pulmonary artery to bypass the systemic venous flow to the pulmonary artery. Initially, an anastomosis was made between the roof of the right atrium and the pulmonary artery. Modifications were then made that consist of an anastomosis between the superior vena cava directly to the superior margin of the right pulmonary artery and an anastomosis of the vena cava to the inferior border of the pulmonary artery using a synthetic conduit, which can go inside the right atrium or be extracardiac, excluding the right ventricle I (modified Fontan). Generally, a perforation is made in the middle third of the graft that functions as a fenestration and has advantages during the postoperative period, especially when done in high-risk patients or in cities with altitudes high above sea level.[12-15]

This type of surgery for correction of univentricular cardiac pathology has evolved greatly. The principal problem for these patients is the pressure that the sole ventricle must support because it manages the volume of two ventricles. In patients with this type of heart disease, the initial surgery is the bidirectional Glenn procedure, and when the patient becomes symptomatic or severely cyanotic, another procedure known as total cavopulmonary bypass or Fontan surgery is required. The latter is due to the fact that during the child's growth there are changes in the blood volumes contributed by the superior and inferior

Table 3. Risk factors associated with mortality in patients undergoing Fontan surgery

Risk factor	*OR*	*95% CI*	*p*
Pleural effusion	0.42	0.04-4.35	0.82
Anatomic alteration of pulmonary artery branches	1.50	0.078-28.8	0.63
Aortic clamping	1.80	0.34-9.71	0.76
Arrythmias	3.00	0.34-26.19	0.67
Atrio-ventricular valvular insufficiency	5.00	0.21-117.8	0.89
Circulatory arrrest	5.40	0.69-41.7	0.25
Atrio-pulmonary derivation	13.50	1.34-135.9	0.06
ARI	14.80	2.19-100.6	0.009
Absence of fenestration	18.00	1.08-298.9	0.05
Significant bleeding or difficult anatomy	20.00	1.61-247.9	0.034

OR, odds ratio; 95% CI, 95% confidence interval; ARI, acute renal insufficiency.

hemibody, with the higher percentage of the inferior hemibody at ~4 years of age and between 15 and 20 kg weight, providing the hemodynamic conditions are adequate.[16-18] In the present study, patients who died were younger than those patients who survived. This may be due to the fact that at older ages the hemodynamic changes are better tolerated. In addition, the surgical procedure is easier because the pulmonary branches are larger.

In the series of patients presented, 39.4% had undergone prior surgery referred to as palliative bidirectional Glenn. Approximately half of these patients had abnormalities in the anatomy of the pulmonary artery branches, which makes performance of the Fontan procedure difficult. However, in the patient analysis, these changes did not increase mortality risk. On the contrary, it should be noted that there were six patients with a history of anterior Glenn surgery who were scheduled for Fontan surgery but it was not able to be performed due to the presence of firm adhesions that caused lesions in the large blood vessels, leading the patients to hemodynamic instability. Half the patients died during the immediate postoperative period.

Among the included Choussat criteria for selecting candidates for Fontan surgery are measurement of pulmonary resistance and ejection fraction.[6] Despite some recent publications,[19] these variables were not associated with mortality observed in the analysis in 32 patients. However, in some patients who did not meet all the criteria, these factors influenced the postoperative morbidity, particularly with the development of pleural effusion, ventricular dysfunction or cardiogenic shock. Therefore, these complications increased the postsurgical hospital stay. High pulmonary pressures before surgery seems to favor fluid leak to the third space after performing total cavopulmonary bypass.

The analysis also found that there was less risk of dying in patients who had fenestrated extracardiac Fontan surgery performed. Similar to the results of this study, different authors have described the convenience of performing this procedure.[2] Therefore, we believe that fenestration is a protective procedure and useful for improving hemodynamic condition postoperatively. Although there were no differences in mortality with respect to the time of extracorporeal pump circulation or aortic clamping, there was greater mortality due to neurological and acute renal insufficiency complications occurring with greater frequency, which is related to injury of these organs due to ischemia. Postoperative management of these patients is a very important aspect for the success of the procedure because hemodynamic and physiological changes are poorly tolerated and may require immediate intervention, such as stopping of the procedure.[15,20,21] Of the total patients, only one warranted abandoning the procedure due to presenting a transpulmonary gradient of 17 mmHg. It should be noted that in other centers around the world,[22] children who survive the postoperative period with or without complications have a satisfactory median and long-term progression. This was observed in the group of patients described in this study with the exception of one patient who died at 6 months after the surgery. The remaining patients have not required hospitalizations and are evaluated on an outpatient basis.

In conclusion, for patients with univentricular heart disease, performing Fontan surgery or total cavopulmonary bypass has successfully evolved over the years and has managed to reduce mortality and improve quality of life.

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