ORIGINAL ARTICLE

Total Right Heart Bypass: Long-Term Complications and Survival

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ABSTRACT

Introduction
Total right heart bypass (RHB) is a palliative procedure with high incidence of complications. Early recognition and individualization of these complications is crucial to define adequate therapeutic strategies.

Objective
The aim of this study was to evaluate the incidence of events and mortality in total RHB during long-term follow-up and compare atriopulmonary (AP) with extracardiac conduit (EC) techniques.

Methods
Between 1987 and 2010, 191 patients submitted to total RHB with a mean follow-up (X) of 6.5 ± 5 years (1-20 years) after surgery were analyzed.
Patients were divided, according to the surgical approach, in group I: AP, 39 patients, X=14 years and group II: EC, 152 patients, X=4 years.

Results
The following complications were present in 57% (n = 116) of patients:

Complications	Group I: AP	Group II: EC	p
Long-term events (n = 111)	77%	53%	0.0076
Arrhythmias (n = 49)	56%	17%	0.0000
Atrial flutter (n = 13)	30%	0.7%	0.0000
Thrombosis (n = 31)	31%	12.5%	0.0058
Protein-losing enteropathy (n = 9)	10.3%	3.3%	0.06
Subaortic stenosis (n = 7)	10.3%	2%	0.01
Interventional procedures (n = 43)	7.7%	24.7%	0.05
Reoperations (n = 20); conversions (n = 6)	25.6%	6.6%	0.0005
Mortality (n = 9)	15.4%	2%	0.0004

Overall long-term mortality was 4.6% (n = 9).
At univariate analysis, mortality was associated with ventricular dysfunction (p=0.0000), protein-losing enteropathy (p=0.0000), atrial flutter (p=0.0012), reoperations (p=0.0006), subaortic stenosis (p=0.0024), thrombosis (p =0.01) and AP technique (p=0.0004).
Multivariate analysis revealed that mortality was associated with ventricular dysfunction [OR 27.7 (4.64-165.24); p = 0.0003], AP technique [OR 2.5 (16.2-105.9); p = 0.0036] and protein-losing enteropathy [OR 9.31 (1.53-56.66); p = 0.01].

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Conclusion

- Adverse events were present in 57% of patients submitted to total RHB during long-term follow-up.
- Long-term mortality was associated with ventricular dysfunction, atrial flutter, protein-losing enteropathy, subaortic stenosis, reoperations, thrombosis and atriopulmonary technique.
- Ventricular dysfunction, protein-losing enteropathy and atriopulmonary technique were predictors of mortality.

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Key words > Congenital Heart Defects - Single Ventricle - Cardiovascular Surgical Procedures - Fontan-Kreutzer Procedure - Postoperative Follow-up.

Abbreviations >

CVA	Stroke	**EC**	Extracardiac conduit
AP	Atriopulmonary	**PLE**	Protein-losing enteropathy
AV	Atrioventricular	**X**	Mean postoperative follow-up
RHB	Right heart bypass		

INTRODUCTION

During the seventies, Dr. Francis Fontan in France and Dr. Guillermo Kreutzer in Argentina initiated the development of total right heart bypass (RHB) using the atriopulmonary (AP) technique. (1) Several years later (1988) M. de Leval (2, 3) and Castañeda described the lateral tunnel technique (4) and in 1990 Marceletti introduced the extracardiac conduit (EC) procedure based on Puga's research made during the previous year. (6, 7) These procedures have increased the survival and improved the outcome and quality of life of patients with single ventricle; (1, 8) however, they are palliative interventions with high incidence of complications. (9)

The evolution of total RHB shows a significant reduction in operative mortality due to several reasons: the better understanding of the physiological and anatomic cornerstones, (10) surgical advances and postoperative care.

The current challenge is to optimize the functional outcome. For this reason, it is necessary to recognize, understand and know how to manage the complications of total RHB. The goal of this study is to evaluate the incidence of late events in total RHB and to analyze long-term mortality comparing the traditional technique with the extracardiac conduit.

METHODS

During the period 1987-2010, 257 patients were operated with the RHB technique at the Hospital Prof. Dr. Juan P. Garrahan. A cohort of 191 patients operated on with AP and EC variants was included in this retrospective study. Median follow-up was 5 years (25-75% interquartile range: 2-10 years). Median age at the moment of surgery was 5 years (25-75% interquartile range: 4-7 years)

Patients undergoing vena cava lateral tunnel (19 patients), those who died in the immediate postoperative period (30 patients) and those lost during follow-up (17 patients) were excluded from the study.

The following types of single ventricle were identified: left ventricular type (137 patients), right ventricular type (28 patients), biventricular type (24 patients) and indeterminate type (2 patients).

The patients were divided, according to the surgical approach, in group I: AP, 39 patients, with X of 12 years (± 4.7 years) and group II: EC, 152 patients, with X of 4.6 year (± 3.3 years).

All the patients were evaluated with physical examination, electrocardiogram, pulse oximetry, chest X-ray, transthoracic color-Doppler echocardiography, exercise stress test and 24-hour Holter monitoring. Transesophageal color-Doppler echocardiography, contrast echocardiography, cardiac catheterization or multislice computed tomography were indicated according to the clinical findings.

Statistical Analysis

Microsoft Office Excel 2003 was used to store data. All calculations were performed using Statistix 8.0 software package.

Frequency and/or percentage distributions were established for all the variables in relation with the total number of cases; accordingly, values were expressed as proportions, mean and standard deviation or median and interquartile range.

Fisher's exact test or the chi square test was used to compare proportions. A p value < 0.05 was considered statistically significant.

Odds ratio (OR) and the corresponding 95% confidence interval were determined.

Long-term survival was estimated using the Kaplan-Meier method.

RESULTS

Long-term events

Fifty-eight percent of patients (n = 111) presented events: 30 patients (77%) in group I and 81 patients (53%) in group II (p = 0.0075) (Table 1).

Arrhythmias

Arrhythmias occurred in 25.6% of patients with X = 10.4 years (± 4.9 years) in group I and X = 3.7 years (± 3 years) in group II, and were more common in group I (n = 22) (p = 0.0000). In this group, atrial flutter (n = 12) was the most common arrhythmia (p = 0.0000), followed by non-sustained atrial arrhythmia (n = 7), loss of sinus rhythm (n = 2) and ventricular tachycardia (n = 1). Conversely, in group II (n = 27), junctional rhythm was more frequent (n = 13), followed by supraventricular tachycardia (n = 4), ventricular tachycardia (n = 4), ventricular premature

Table 1. Long-term complications in patients undergoing total RHB

	Group I: AP	Group II: EC	p
Long-term events (n = 111)	77%	53%	0.0076
Arrhythmias (n = 49)	56%	17%	0.0000
Atrial flutter (n = 13)	30%	0.7%	0.0000
Thrombosis (n = 31)	31%	12.5%	0.0058
Stroke (n = 4)	5.1%	1.3%	0.13
Protein-losing enteropathy (n = 9)	10.3%	3.3%	0.06
Subaortic stenosis (n = 7)	10.3%	2%	0.01
Interventional procedures (n = 43)	7.7%	24.7%	0.05
Reoperations (n = 20); conversions (n = 6)	25.6%	6.6%	0.0005
Mortality (n = 9)	15.4%	2%	0.0004
Ventricular dysfunction (n = 21)	15.4%	10%	0.32
Plastic bronchitis (n = 1)	0	0.7%	0.6

RHB: Right heart bypass. AP: Atriopulmonary. EC: Extracardiac conduit.

beats (n = 3), sinus node dysfunction (n = 2) and atrioventricular (AV) block (n = 1) (Figure 1).

Atrial flutter occurred in 30% of patients in the AP group after X = 10 years (6 to 16 years), while 9.8% of those in the EC group presented loss of sinus rhythm or sinus node dysfunction after X = 5 years (1 to 10 years).

A definite pacemaker was implanted to 10 patients (5.2%) during follow-up: 6 in group I (4 associated with conversion to sinus rhythm with the Maze procedure and 2 with sinus node dysfunction) and 4 in group II (3 with junctional rhythm associated with ventricular dysfunction and 1 with sinus node dysfunction).

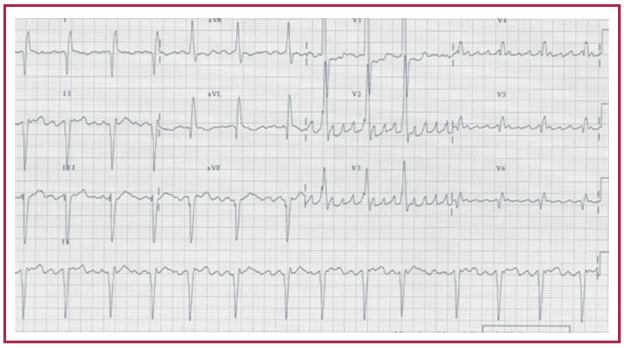

Fig. 1. Electrocardiogram from a patient with atrial flutter 7 years after total RHB with the atriopulmonary technique.

Thrombosis and stroke

Thrombosis was detected in 31 patients (16.2%): 12 (31%) in group I and 19 (12.5%) in group II (p = 0.0058) after X = 12 years (± 3.9 years) and 3.8 years (± 3.8 years), respectively. Five patients were receiving anticoagulation therapy, 22 were taking aspirin and 4 were not taking any medication.

Thrombi were most common in the venous system in both groups while systemic thrombi occurred in only 3 patients.

Five episodes of stroke (CVA) were detected in 4 patients, 2 in each group.

In group I, one female patient presented 2 episodes of CVA while taking antiplatelet agents at 4 and 6 months after AP with fenestration. The fenestration was closed and no further episodes occurred. The other patient, who presented CVA 17 years after surgery, was not taking antiplatelet or anticoagulation agents. None of these two patients presented arrhythmias, thrombosis or ventricular dysfunction.

In group II, two patients developed CVA 10 and 17 years after total RHB without fenestrations. Thrombi were present in both patients. Junctional rhythm and ventricular dysfunction were found in one patient who was receiving anticoagulation agents. The other patient had normal ventricular function, did not present arrhythmias and was receiving antiplatelet agents.

Protein-losing enteropathy

Protein-losing enteropathy (PLE) affected 4.7% of the series (n = 9): 4 patients (10.3%) in group I with X = 11 years (11-13 years) and 5 patients (3.3%) in group II with X = 4 years (9 months-10 years) (p = 0.06).

In group I, PLE was associated with arrhythmias in 75% (n = 3) of patients, while in group II 80% (n = 4) had ventricular dysfunction.

Subaortic stenosis

The diagnosis of subaortic stenosis was made in 3.6% of patients (n = 7): 4 patients in group I and 3 patients in group II (p = 0.01).

Recurrence of subaortic stenosis occurred in 3 patients (1 with previous banding), 2 and 3 months after AP and 6 months after EC. Four patients (2 with previous banding) developed subaortic stenosis 3 and 15 years after AP and 3 and 8 years after EC.

The association between subaortic stenosis and banding was seen in 43% of cases.

Ventricular dysfunction

Ventricular dysfunction was observed in 15.4% (n = 6)

of patients in group I and in 10% (n = 15) of those in group II (p = 0.32) and was not isolated but associated with subaortic stenosis, arrhythmias, thrombi and collateral vessels in the AP technique, and with junctional rhythm, collateral vessels, thrombi, cyanosis, subaortic stenosis and PLE in the CE procedure.

The resolution of these factors was associated with recovery of ventricular function in 52% of cases. The following procedures were performed: embolization of collateral vessels, definite pacemaker implant in cases of junctional rhythm, use of antiarrhythmic agents, anticoagulation and reoperations (surgery for subaortic stenosis and conversion).

Plastic bronchitis

Only 1 patient in group II presented bronchial cast expectoration 6 months after RHB, requiring angioplasty of the right pulmonary artery, embolization of collateral vessels, repair of pulmonary artery branches and fenestration.

Interventional procedures

Fifty-eight interventional procedures were performed in 43 patients with X = 2.7 years (± 2.4 years): 3 patients (7.7%) in group I and 40 (24.7%) in group II (p = 0.05) (Figure 2).

In group I, these procedures were: embolization of collateral vessels (n = 4), fenestration closure (n = 1) and closure of AV valve (n = 1). In group II the procedures included fenestration closure (n = 23), embolization of collateral vessels (n = 22), stent implant (n = 1), enlargement of extracardiac conduit (n = 1), closure of the hepatic veins (n = 1), pulmonary artery closure with Amplatzer device (n = 1), pulmonary artery balloon angioplasty (n = 1) and stent implant (n =1) and coil exclusion of the right ventricle (n = 1).

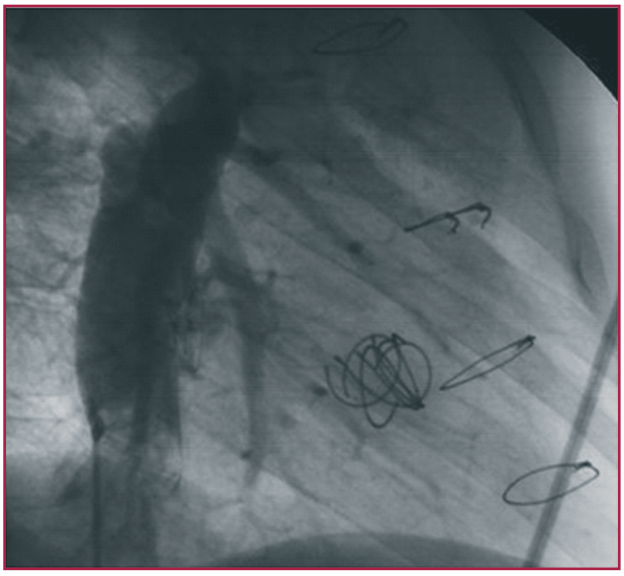

Fig. 2. Patient with total RHB with extracardiac conduit who underwent an interventional cardiology procedure one year after surgery, with stent implant in the conduit and coil exclusion of the right ventricle.

Fenestration closure was indicated at a median of 1.5 years (0.5-6 years). Five fenestrations were performed in the group operated with the AP surgical variant: 2 closed spontaneously, 1 was occluded by an interventional procedure and the remaining fenestration was not occluded due to the results of the occlusion test. Fenestrations were performed to 63/152 patients in the group operated with the EC technique: 23 underwent interventional occlusion and 18 closed spontaneously. The remaining 22 patients are under clinical surveillance.

Reoperations

Twenty patients underwent 22 reoperations: 10 (25.6%) in group I and 10 (6.6%) in group II (p = 0.0005). The indications, procedures and years after total RHB are indicated in Table 2.

Six patients underwent conversion to EC: 3 due to atrial flutter, 2 due PLE and 1 due to both events. The Maze procedure was performed in 5 of them and antitachycardia pacemaker was implanted in 4. One patient died in the immediate postoperative period. All the patients received antiarrhythmic agents and anticoagulant therapy. Only one patient presented recurrent atrial arrhythmia (20%).

Mortality

Long-term mortality was 4.6% for the total group (n = 9): 15.4% in group I (n = 6) and 2% (n = 3) in group II (p = 0.0004).

Two patients died in the AP group 1 and 2 years after surgery, associated with subaortic stenosis and ventricular dysfunction. Later deaths were due to PLE (n = 1) 16 years after surgery and arrhythmias (n = 3) at X = 12.3 years. Of the 3 patients with arrhythmias, 1 also had thrombosis and 2 had ventricular dysfunction.

In patients with EC, deaths were due to: subaortic stenosis at X = 3 months, sudden death at X = 1 year and ventricular dysfunction associated with PLE at X = 5 years.

At univariate analysis, mortality was associated with ventricular dysfunction (p = 0.0000), PLE (p=0.0000), atrial flutter (p=0.0012), reoperations (p=0.0006), subaortic stenosis (p = 0.0024), thrombosis (p = 0.01) and AP technique (p = 0.0004).

Multivariate analysis revealed that mortality was associated with ventricular dysfunction [OR 27.7 (4.64-165.24); p= 0.0003], AP technique [OR 2.5 (16.2-105.9); p = 0.0036] and PLE [OR 9.31 (1.53-56.66); p = 0.01].

The Kaplan-Meier curve showed that in the group operated with the AP technique survival at 5 years was 95% (CI 83-98%), 92% at 10 years (CI 78-97%) and 77% at 20 years (CI 52-91%), while in the group operated with the CE procedure, survival at 5 years was 97% (CI 89-99%) and 97% at 10 years (CI 89-99%) (p = 0.29) (Figure 3).

Table. 2. Reoperations in patients with total RHB.

Patient	Group	Indication	Type of surgery (number)	Time interval after RHB	Maze	Conversion
1	EC	AV regurgitation	(1) Closure of AV valve	5 years	no	no
2	EC	Diaphragmatic paralysis Cyanosis-thrombosis	(1) Diaphragmatic plication	3 years	no	no
3	EC	Sinus node dysfunction	(1) Pacemaker	6.8 years	yes	no
4	EC	Junctional rhythm + thrombosis + AV regurgitation	(1) Pacemaker + conduit replacement + AV valve repair	13.5 years	no	no
5	EC	Aortic regurgitation	(1) Aortic valve repair	2 years	no	no
6	EC	Subaortic stenosis	(1) enlargement of the bulboventricular foramen	4 years	no	no
7	EC	Enteropathy + junctional rhythm	(1) Fenestration + anastomosis release + pacemaker	5 years	no	no
8	EC	AV regurgitation Subaortic stenosis + AV regurgitation + junctional rhythm	(1) AV closure (2) Enlargement of the bulboventricular foramen + AV closure + pacemaker	6 years 8 years	yes	no no
9	EC	AV regurgitation	(1) AV repair	4 years	no	
10	EC	Plastic bronchitis	(1) Fenestration + repair of the pulmonary artery branches	3 years		
11	AP	Subaortic stenosis	(1) Stansel	8 years	no	no
12	AP	Enteropathy + atrial flutter + pauses	(1) Conversion + fenestration (2) Maze + pacemaker	13 years 14 years	yes	yes
13	AP	Thrombosis + enteropathy	(1) Conversion + Maze + pacemaker + fenestration	13 years	yes	yes
14	AP	Atrial flutter + thrombosis	(1) Pacemaker	13 years	no	no
15	AP	Atrial flutter	(1) Conversion + Maze + pacemaker	13 years	yes	yes
16	AP	Pulmonary artery antegrade flow	(1) Pulmonary artery ligation	12 years	no	no
17	AP	Atrial flutter	(1) Conversion + Maze + pacemaker	12 years	yes	yes
18	AP	Atrial flutter	(1) Conversion + Maze + pacemaker	10 years	yes	yes
19	AP	Enteropathy	(1) Conversion + fenestration	13 years	no	yes
20	AP	Subaortic stenosis	(1) Stansel	3 years	no	no

RHB: Right heart bypass. EC: Extracardiac conduit. AP: Atriopulmonary. AV: Atrioventricular.

DISCUSSION

Elevated leukocytes at admission in patients with Total RHB is the best palliative option for patients with single ventricle physiology. Current publications have reported that mortality of total RHB has decreased from 30% to less than 5% in the last years. (11) Staged surgical approach and EC with fenestrations have contributed to this progress. Despite these promising results, the risk of failure and late complications after the Fontan circulation, (12, 13) as arrhythmias, thrombosis and CVA, PLE, outflow tract obstruction, ventricular dysfunction and valve regurgitation still occur and deteriorate total RHB.

The incidence of supraventricular arrhythmias in our series was 30%. Some authors have reported up to 50% incidence at 20 years. (12-15) Loss of sinus rhythm in EC increases during follow-up and can affect cardiac output, predisposing to atrial flutter (10% of our series). (16-18)

Thromboembolic events represent another factor of morbidity and mortality, with an incidence of 16.2% in our series that is similar to the one reported by other publications. (19) This complication was more common in RHB with the AP technique, in which the giant right atrium generates a predisposing hemodynamic factor in the long-term follow-up period and consequently, when patients are older. The incidence of CVA (13%) was not related with fenestrations as described in previous series (19, 20) and increased at long-term follow-up. Silent pulmonary thromboembolism is another complication. There is no agreement on the indications of anticoagulation therapy versus antiplatelet treatment. (11, 21-25) Although most publications recommend antiplatelet agents over anticoagulants, they all agree on the need of prospective studies to establish definite recommendations.

The incidence of PLE in our patients (4.7%) was similar to the one reported by other series (26). The pathophysiology of this condition is still unknown; (26) it can occur early or several years after surgery and an effective treatment is still unavailable. (27)

Subaortic stenosis developed at the beginning of our experience and was associated with previous banding in 43% of patients. Later, the surgical strategy was

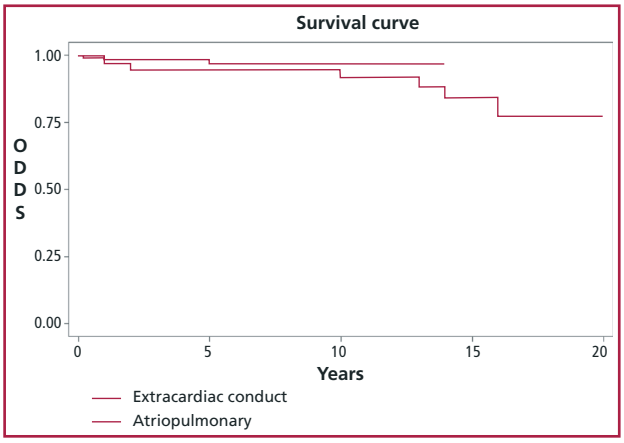

Fig 3. Patient survival with total RHB.

replaced by the Damus-Kaye-Stansel procedure or the palliative arterial switch operation in order to protect ventricular function. (28, 29)

In agreement with other authors, (30) ventricular dysfunction (11%) was not an isolated event but was related to volume and pressure overload, myocardial fibrosis secondary to ischemia, arrhythmias, valvular regurgitation and chronic low cardiac output.

In our series, 15.7% of patients had to be reoperated due to several causes: arrhythmias, valvular regurgitation, subaortic obstruction, definite pacemaker implant and PLE. (31, 32) In our experience, conversion to EC has been successful in agreement with the report by Mavroudis et al. (33-35) and with 20% incidence of recurrent arrhythmias, as reported by other authors. (36)

Cardiac catheterization is useful to detect complications and eventually treat them. In our series, 22.5% of patients underwent an interventional cardiology procedure. A significant number of our patients submitted to cardiac catheterization presented collateral circulation as an additional finding which generated chronic volume overload. The development, prevalence and pathogenesis (cyanosis, Glenn stage) of collateral circulation are not clearly understood yet. (37)

Many effects of total RHB are related to the endothelial dysfunction induced by this circulatory system. Our future challenge is to understand the molecular basis of these conditions and differentiate between those developing as compensatory mechanisms and those emerging as abnormal responses, with significant therapeutic implications. (38)

The balance between pulmonary resistance, systemic resistance and ventricular function is essential for the proper functioning of total RHB circulation. (39) Long-term mortality (4.6%) is related to events that modify this labile circulation. In our population, the survival curve was similar to that described in previous series: (40) 95% and 92% at 5 and 10 years, respectively, in the AP group and 97% at 5 and 10 years in the EC group, with a pronounced fall (77%) after 20 years for the AP group.

Study Limitations

The main study limitations include: the retrospective nature of the analysis, treatment strategies have been modified throughout the years, the surgical strategies compared are not contemporary, the sample is heterogeneous (number of patients, anatomical variants) and the follow-up period is different in both groups, though this variable was adjusted in the statistical analysis. Although the difference in the follow-up period is a determinant factor for the development of complications, as they can occur in any patient and at any moment independently of the surgical technique and the anatomical variant, this preliminary study encourages us to continue with the analysis of this complex group of patients.

CONCLUSIONS

Total RHB is the treatment of choice for patients with single ventricle physiology with high incidence of adverse events that in our series was 57%.

Mortality was associated with ventricular dysfunction, atrial flutter, PLE, subaortic stenosis, reoperations, thrombosis and with the AP technique which showed greater incidence in long-term events compared to the EC procedure. Ventricular dysfunction, PLE and AP technique were identified as predictors of mortality.

Total RHB is a palliative procedure. The early detection and treatment of complications is the current challenge.

RESUMEN

Bypass total del ventrículo pulmonar: complicaciones y sobrevida en el seguimiento alejado

Introduction

La cirugía de bypass total del ventrículo pulmonar (BPTVP) es un procedimiento paliativo con una incidencia elevada de complicaciones. El reconocimiento e individualización precoz de estas complicaciones es esencial para definir estrategias terapéuticas adecuadas.

Objetivo

Evaluar los eventos alejados del BPTVP, analizar la mortalidad alejada y comparar la técnica auriculopulmonar (AP) con el conducto extracardíaco (CE).

Material y métodos

Entre 1987 y 2010 se analizaron 191 pacientes sometidos a BPTVP con un tiempo medio de seguimiento posquirúrgico (X) de 6,5 ± 5 años (1-20 años).
Los pacientes se dividieron, de acuerdo con la variante quirúrgica, en grupo I: AP, 39 pacientes, X = 14 años y grupo II: CE, 152 pacientes, X = 4 años.

Resultados

El 57% de los pacientes (n = 116) presentaron las siguientes complicaciones:
La mortalidad global alejada fue del 4,6% (n = 9).
En el análisis univariado, la mortalidad estuvo asociada con disfunción ventricular (p =0,0000), enteropatía

Complications	Group I: AP	Group II: EC	p
Long-term events (n = 111)	77%	53%	0.0076
Arrhythmias (n = 49)	56%	17%	0.0000
Atrial flutter (n = 13)	30%	0.7%	0.0000
Thrombosis (n = 31)	31%	12.5%	0.0058
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Subaortic stenosis (n = 7)	10.3%	2%	0.01
Interventional procedures (n = 43)	7.7%	24.7%	0.05
Reoperations (n = 20); conversions (n = 6)	25.6%	6.6%	0.0005
Mortality (n = 9)	15.4%	2%	0.0004

perdedora de proteínas (p = 0,0000), aleteo auricular (p = 0,0012),reoperaciones (p = 0,0006), estenosis subaórtica (p = 0,0024), trombos (p = 0,01) y la técnicaquirúrgica AP (p = 0,0004).
En el estudio multivariado, la mortalidad estuvo relacionada con disfunción ventricular [OR 27,7 (4,64-165,24); p = 0,0003], técnica AP [OR 2,5 (16,2-105,9); p = 0,0036] y enteropatía perdedora de proteínas [OR 9,31 (1,53-56,66); p = 0,01].

Conclusiones

– El BPTVP presentó eventos adversos en el 57% de los pacientes durante el seguimiento alejado.
– La mortalidad alejada estuvo asociada con disfunción ventricular, aleteo auricular, enteropatía perdedora de proteínas, estenosis subaórtica, reoperaciones, trombos y técnica quirúrgica auriculopulmonar.
– Los predictores de mortalidad fueron la disfunción ventricular, la enteropatía perdedora de proteínas y la técnica auriculopulmonar.

Palabras clave > Defectos cardíacos congénitos - Ventrículo único - Cirugía cardiovascular - Cirugía de Fontan-Kreutzer -Evolución posquirúrgica.

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