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Morbidity risk factors after total cavopulmonary shunt

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ARTICLE INFORMATION

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Acronyms
ARR: absolute risk reduction
AV: atrio-ventricular
CPWS: William Soler Pediatric Cardiology Hospital, for its acronym in Spanish
NNH: needed number to harm
NNT: number of patients needed to treat
TCPC: total cavopulmonary connection

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ABSTRACT

Introduction: Arrhythmia, ventricular dysfunction and decreased functional capacity appear in the surgical outcome of patients with total cavopulmonary shunt. Detection of risk factors for this morbidity is essential to improve outcomes.

Objective: To identify these risk factors in patients with intraatrial and extracardiac total cavopulmonary shunt.

Method: A prospective cohort study was conducted from January 1992 to January 2012 at William Soler Cardiology Hospital. Frequency distributions and contingency tables were built. The association between qualitative variables was analyzed by Ji-square test of independence, for quantitative, with the Student t test. The binary logistic regression was used to identify risk factors.

Results: 74 patients were studied at an average follow-up time of 8 years. Thirteen patients had arrhythmias and it was identified as a risk factor for postoperative occurrence of moderate or severe atrioventricular failure. Eleven patients developed moderate or severe ventricular dysfunction. Association with ventricular mass variation was detected. Decreased functional capacity was detected in 33 patients with associated ventricular dysfunction.

Conclusions: Atrioventricular failure that appears after total cavopulmonary shunt should be treated since it is a risk factor for new arrhythmias. The decrease in ventricular mass in evolutionary studies should guide the diagnosis and treatment of ventricular dysfunction and decreased functional capacity in patients operated with total cavopulmonary shunt.

Key words: Total cavopulmonary shunt, Univentricular heart, Risk factors, Morbidity

Factores de riesgo de morbilidad luego de la derivación cavopulmonar total

RESUMEN

Introducción: La arritmia, la disfunción ventricular y la disminución de la capacidad funcional aparecen en la evolución posquirúrgica de los pacientes con derivación cavopulmonar total. La detección de factores de riesgo de esta morbilidad es esencial para mejorar la evolución.

Objetivo: Identificar estos factores de riesgo en pacientes con derivación cavopulmo-

nar total intraatrial y extracardíaca.

Método: Se realizó un estudio de cohorte prospectivo desde enero de 1992 hasta enero de 2012 en el Cardiocentro Pediátrico "William Soler". Se construyeron distribuciones de frecuencias y tablas de contingencia. La asociación entre variables cualitativas se analizó mediante la prueba de independencia Ji cuadrado; para las cuantitativas, con la prueba *t de Student*. Para la identificación de factores de riesgo se utilizó la regresión logística binaria.

Resultados: Se estudiaron 74 pacientes en un tiempo promedio de seguimiento de 8 años. Trece pacientes presentaron arritmias y se identificó como factor de riesgo a la aparición posoperatoria de insuficiencia aurículo-ventricular moderada o grave. Once pacientes desarrollaron disfunción ventricular moderada o grave. Se detectó asociación con la variación de la masa ventricular. La disminución de la capacidad funcional se detectó en 33 pacientes con asociación a la disfunción ventricular.

Conclusiones: La insuficiencia aurículo-ventricular que aparece después de la derivación cavopulmonar total debe tratarse por ser un factor de riesgo de nuevas arritmias. La disminución de la masa ventricular en estudios evolutivos debe orientar el diagnóstico y tratamiento de disfunción ventricular y disminución de la capacidad funcional en pacientes operados con derivación cavopulmonar total.

Palabras clave: Derivación cavopulmonar total, Corazón univentricular, Factores de riesgo, Morbilidad

INTRODUCTION

The goal of surgical treatment of patients with univentricular heart is to create a hemodynamic system to divert the systemic venous return to the pulmonary arteries without the contractile impulse of a ventricular cavity. Total cavopulmonary connection (TCPC) or Fontan procedure[1] is the current palliative surgical option. It is done in one-stage or two-stage surgery and the most used techniques are the intraatrial[2] and the extracardiac[3].

After TCPC, the advantage of prolonging the life expectancy of these patients is accompanied by the appearance of morbidity associated with the new form of blood circulation. The single ventricle functions as a pump which passively receives the pulmonary circulation and actively propels the systemic circulation. Blood flow depends on the pressure differences between the venous return, pulmonary vessels and ventricular diastole. The increase in pulsatile impedance (aortic) and non-pulsatile (pulmonary) in series increases vascular resistance and afterload[4]. When it does not work, clinical signs of low cardiac output, systemic congestion and hypoxemia appear.

The sudden discharge of the return of both territories of cava veins produces imbalance between ventricular and blood volumes, increased venous pressure and slowing of transpulmonary flow. Treatment in two stages, first the partial bypass of the superior vena cava and then the inferior, allows changes to be progressive[5].

Although there is a selection of patients, according to preoperative hemodynamic criteria of good bypass function, there is a decreased exercise capacity, as well as ventricular dysfunction, arrhythmias, thromboembolism , protein-losing enteropathy and medium and long term renal dysfunction[6].

Arrhythmias are the clinical expression of an electrophysiological substrate of multiple potential circuits by expansion, surgery or anomalous arrangement of conduction paths. The relationship between the occurrence of postoperative arrhythmias with age when TCPC is performed seems logic; and for some, with the type of technique[7], with the presence of preoperative arrhythmias, with the loss of sinus rhythm, valvular insufficiency, heterotaxy syndrome, the dominant right ventricle and follow-up time[8].

In these patients there is a decrease in functional capacity and a decrease in oxygen consumption of 50 to 60 % for the age[9]. During exercise there is an increase in pulmonary pressure, abnormal decrease in end-diastolic volume and heart rate increases little[10].

Risk factors for this outcome are difficult to determine. The following risk factors are mentioned: patient selection, technical options, variability in follow-up time, age at surgical indication, heterotaxy

syndrome, right ventricular morphology, moderate or severe atrio-ventricular (AV) failure, and elevated pulmonary pressure and resistance[11].

Cavopulmonary connections were introduced in William Soler Pediatric Cardiology Hospital in 1990 along with the two-stage surgical approach. In the national medical literature there are approaches to the experience in the surgical treatment of univentricular hearts[12], however there is no study on postoperative morbidity in the middle and long terms. Based on the hypothesis that during the postoperative course new risk factors for arrhythmias, ventricular dysfunction and decreased functional capacity can be identified, a prospective study in order to identify patients with extracardiac and intraatrial TCPC was designed.

METHOD

A prospective cohort study was conducted from January 1992 to January 2012 in William Soler Pediatric Cardiology Hospital (CPWS, for its acronym in Spanish).

The sample consisted of patients who underwent extracardiac or intraatrial TCPC and who survived at one year of surgery, whose legal guardians expressed their consent for the inclusion in this research. Patients with previous atriopulmonary bypass with conversion to cavopulmonary connection were excluded as well as those to whom the evolutionary follow-up in the shortest time of postoperatively study of one year would be impossible to maintain. Those patients whose guardians decided to revoke the consent granted and those who did not undergo the evolutionary follow-up in the shortest time of one year for causes other than postoperative mortality left the study.

The sample was composed of 74 patients: 43 operated with intraatrial TCPC and 31 with extracardiac TCPC.

The performing of one or another surgical technique resulted from the decision of the attending surgical team in each patient. The guides to good practices for TCPC were followed for anesthesia, surgery on both technical modifications, and the use of cardiopulmonary bypass, myocardial protection and postsurgical care[13].

The diagnosis of arrhythmias was tested by electrocardiogram and new arrhythmia was defined as the postoperative occurrence of any pathological alteration different from sinus rhythm.

Ventricular function was assessed by echocardiogram analyzing the ejection fraction. Ventricular dysfunction was defined as ventricular ejection fraction lower than 50 %[14].

The functional capacity or exercise tolerance of patients was assessed through an ergometric test, with The Bruce Protocol Treadmill test[15]. The expected functional capacity equal or less than 65 % was defined as diminished[16].

Pulmonary arterial and diastolic ventricular pressures were measured at the end of surgery. The mean variation of these pressures was found in the follow-up time of patients to whom a hemodynamic study was indicated at least once after TCPC. This variation was associated with morbidity.

The ratio between the ventricular mass found by echocardiography and the body surface area was obtained. To calculate the change in ventricular mass index, the difference between the measurement made in the preoperative of the total connection and the last postoperative examination was found.

The times between total and partial bypass in addition to the time lived with total bypass until the end of the study for each patient were calculated. Data were collected during hospitalization scheduled in the preoperative period to the TCPC and in four subsequent times, a year after surgery, at three and five years from the procedure and in a final moment. Data were stored and processed in a database created in the SSPS 13.5 statistical software (SPSS Inc., Chicago, Illinois, United States).

Frequency distributions and contingency tables were built. As summary measures percentages for qualitative variables and means and standard deviations for the quantitative variables were used. For analysis of association between qualitative variables the Pearson's chi-square test for independence and Fisher's exact test were used. For quantitative variables the parametric Student t test was used with the previous analysis of homogeneity of variance.

To identify risk factors binary logistic regression was used. The following variables were related:

- Dependent variables: arrhythmias, ventricular dysfunction of risk and decreased functional capacity.
- Independent variables: Age, morphologic diagnosis, heterotaxy, type of total cavopulmonary connec-

tion, one-stage or two-stage surgery, interval between partial and total pulmonary connection, type of primary ventricle, morphologic diagnosis, moderate or severe valvular insufficiency, pulmonary and ventricular-arterial pressures of preoperative risk, postoperative variation of these pressures, variation of ventricular mass index and time of evolution.

The coefficients exponential (Expβ) of the model as estimators of the odds ratio (OR) was analyzed. To assess the quality of adjustment the Hosmer and Lemeshow statistic was used.

To validate the results in terms of significance all p value was considered P ≤ 0.05 for the statistic associated with the test and a confidence level of 95 % was used. To analyze the clinical relevance, the relative risk (RR) was calculated. As measures of impact, the absolute risk reduction (ARR) and number of patients needed to treat (NNT) to reduce an event or the needed number to harm (NNH) with their respective confidence intervals of 95 %. The factors that in the univariate analysis showed a significant relationship with the observed effect were identified as intervention.

The research was approved by the management, the Scientific Council and the Ethics Committee of William Soler Pediatric Car-diology Hospital. The project received the support of the Academy of Sciences of Cuba, from the Ministry of Science, Technology and Environment. The provisions of the basic principles of the Helsinki Declaration containing recommendations to follow in biomedical research were observed[17].

Table 1. Arrhythmias. Univariate analysis.

Arrhythmias	Yes (n=13) %	No (n=61) %	p x	RR § (CI 95%)
Intraatrial	14,0	86,0	0,51	1,61 (0,6 a 4,34)
Extracardiac	22,6	77,4		
Preop. AV Failure.	20,6	79,4	0,75	1,37 (0,51 a 3,69)
No preop. AV Failure.	15,0	85,0		
Postop. AV Failure	70,0	30,0	0,0001*	7,47 (3,15 a 17,7)
No postop. AV Failure	9,3	90,7		
One-stage surgery	21,1	78,9	0,73	1,28 (0,44 a 3,69)
Two-stage surgery	16,4	83,6		
Tricuspid atresia	10,3	89,7	0,32	2,14 (0,64 a 7,15)
No tricuspid atresia	22,2	77,8		
Age > 6 years old	20,9	79,1	0,56	1,62 (0,55 a 4,79)
< 6 years old	12,9	87,1		
Time in years (Mean ± SD)				
Evolution	6,78 (±4,46)	9,01 (±6,17)	0,22 τ	(-5,87 a 1,37)
P-TCPC Interval	4,47(± 2,03)	4,52 (± 2,21)	0,94 τ	(-1,37 a 1,27)

Caption. Preop.: preoperative; Posop.: posoperatoria; P-TCPC: Partial-Total cavopulmonary connection; x: p value, Pearson χ^2 test; τ: valor de p, Student t test; CI: confidence interval; §: relative risk; *: p < 0,05

RESULTS

Postoperative follow-up was performed in 74 patients, 43 with intraatrial TCPC and 31 with extracardiac technique. The variation of the ventricular mass was calculated in 50 patients (67.5%) of the 74 assessed at least once a year after surgery and 31 patients (41.9%) underwent hemodynamic study in one of the scheduled admissions a year after the surgery. The rest of the variables analyzed a year after the intervention was collected in 100 % of survivors of

Table 2. Arrhythmias. Multivariate analysis.

Variables	Wald	Sig.	Exp(B)
Type of TCPC	0,77	0,38	1,96
Age at TCPC	0,31	0,57	0,94
One-stage surgery	0,96	0,32	0,38
Evolution time	3,15	0,07	0,87
Postoperative AV failure	4,38	0,03*	5,59
Preoperative AV failure	0,58	0,44	1,67

Hosmer and Lemeshow test p=0,630
TCPC: Total cavopulmonary connection; AV: atrioventricular.
*p<0,05

the immediate postoperative period.

Arrhythmia occurred in 13 patients (**Table 1**). The RR was higher in patients with a diagnosis other than tricuspid atresia and with a significant relationship in those who had moderate or severe postoperative AV impairment [p < 0.01, RR 7.47 (95% CI 3.15 to 17.7)]. The value of the absolute risk reduction of arrhythmia with postoperative AV impairment was negative [ARR -0.61 (95% CI -0.9 to -0.31)], which indicates an increased risk. The number needed to harm was low [NNH 2 (95% CI 2-4)], which means a high frequency of occurrence of arrhythmias in patients with moderate or severe AV impairment.

Multivariate analysis showed that when there is postoperative AV impairment the probability of developing arrhythmias is five times higher (p = 0.03, Exp β 5.59), (**Table 2**).

Ventricular dysfunction occurred in 11 patients, seven of them with intraatrial therapeutic option (**Table 3**). It was more frequent in patients with a diagnosis other than tricuspid atresia with left main ventricle. Of patients with preoperative dysfunction, 25 % had postoperative dysfunction. Although the interval between partial and total shunts and follow-up time in patients in whom ventricular dysfunction was detected were higher than in those without this complication, the differences were not significant.

An association between ventricular dysfunction and decreased ventricular mass index was found, without significant changes that related changes in ventricular and pulmonary arterial pressures with ventricular dysfunction (**Table 4**). Multivariate analysis did not identify any risk factor of postoperative ventricular dysfunction.

Moderate or severe decrease in functional capacity was detected in 33 patients. **Table 5** shows a higher incidence in patients with intraatrial TCPC, in those who underwent surgery with more than six years old, and in those operated in two stages. An association between the decrease in functional capacity and postoperative ventricular dysfunction was found [p = 0.04, RR 1.83 (95% CI 1.14 to 2.9)]. The decrease in functional capacity during the postoperative course with ventricular dysfunction, showed an absolute risk reduction with a negative value, that is, an increased risk [ARR of -0.33 (95% CI -0.62 to -0, 04)] and NNH 4 (95% CI 2 to 25).

The multivariate analysis (**Table 6**) showed a higher probability of occurrence of this complication related to surgical stages and postoperative ventricular dysfunction, but with no significant relationship.

Table 3. Ventricular dysfunction. Univariate analysis.

Ventricular dysfunction	Yes (n=11) %	No (n=63) %	p [x]	RR [§] (CI 95%)
Intraatrial	16,3	83,7	0,75	0,79 (0,25 a 2,47)
Extracardiac	12,9	87,1		
Tricuspid atresia	10,3	89,7	0,51	1,71 (0,49 a 5,75)
No tricuspid atresia	17,8	82,2		
Right ventricle	13,3	86,7	0,69	0,78 (0,25 a 2,42)
Left ventricle	17,1	82,9		
No preoperative dysfunction	12,9	87,1	0,37	1,93 (0,59 a 6,29)
Preoperative dysfunction	25,0	75,0		
Risk VEDP	16,7	83,3	0,72	1,16 (0,34 a 3,93)
No Risk VEDP	14,3	85,7		
Two-stage surgery	14,5	85,5	1	1,1 (0,32 a 3,67)
One-stage surgery	15,8	84,2		
Time in years (Mean ± SD)				
Glenn-Fontan interval	6,06 ± 2,82	5,6 ± 2,7		0,61 τ (-1,31 a 2,23)
Evolution time	10,5 ± 6,15	8,28 ± 5,9		0,25 τ (-1,64 a 6,08)
Age	7,73 ± 4,02	7,48 ± 3,9		0,84 τ (-2,3 a 2,8)

Caption. VEDP, ventricular end-diastolic pressure; x: p value, Pearson χ^2 test; τ: p value, Student t test, CI: confidence interval.

DISCUSSION

Arrhythmias are presented related to age in the natural history of patients who have not undergone univentricular surgery or with tetralogy of Fallot, transposition of great vessels or total anomalous drainage of pul-

monary venous without surgical treatment[8]. Probable causes are hypoxemia and increased atrial pressures, which makes complication not exclusive of TCPC but of atrial hemodynamic characteristics and its consequences over time. TCPC patients show node dysfunction over time as a result of decreased response of nodal cells to neural modulation[18]. The cause of tachyarrhythmias could be found in the areas of slow conduction between the inferior vena cava, the tricuspid valve and the coronary sinus that produce electrophysiological delay after incisions, atrial patches, increased atrial wall stress or congenital lesion[8].

Table 4. Disfunción ventricular y variación del índice de masa, presión pulmonar y ventricular.

Variation Mean ± standard deviation	Ventricular dysfunction Yes	 No	p (CI 95%) [T]
Ventricular mass index (g/m2 body surface area)	-23,38 ± 35,86 n=9	6,66 ± 29,15 n=41	0,01 * (-52,5 a -7,5)
Mean pulmonary artery pressure (mmHg)	-1 ± 4,27	1,69 ± 3,78	0,07 (-5,66 a 0,28)
Ventricular diastolic pressure (mmHg)	-1,37 ± 1,4 n=8	0,13 ± 3,34 n=23	0,14 (-6,43 a 1,05)

T: P value, Student t test; CI: confidence interval; *: p<0,05

Table 5. Decreased functional capacity. Univariate analysis.

Decreased functional capacity	Yes (n=33) %	No (n=41) %	p [x]	RR [§] (CI 95%)
Intraatrial	51,2	48,8	0,18	0,69 (0,39 a 1,21)
Extracardiac	35,5	64,5		
Left ventricle	46,3	53,7	0,89	0,93 (0,55 a 1,58)
Right ventricle	43,3	56,7		
No preoperative VD	45,2	54,8	1	0,92 (0,45 a 1,9)
Preoperative VD	41,7	58,3		
Postoperative VD	72,7	27,3	0,04*	1,83 (1,14 a 2,9)
No postoperative VD	39,7	60,3		
One-stage surgery	31,6	68,4	0,29	1,55 (0,76 a 3,71)
Two-stage surgery	49,1	50,9		
Age > 6 years old	38,7	61,3	0,53	1,26 (0,73 a 2,16)
< 6 years old	48,8	51,2		
Time in years (Mean ± SD)				
P-TCPC Interval	5,59 ± 2,53	5,73 ± 2,87		0,82 [τ] (-1,41 a 1,13)
Follow-up time	8,79 ±5,92	8,48 ±6,03		0,82 [τ] (-2,48 a 3,09)

Caption. VD: ventricular dysfunction; P-TCPC: Partial-Total cavopulmonary connection; x: p value, Pearson χ^2 test; τ: P value, Student t test; CI: confidence interval; §: relative risk; *: p<0,05

Postoperative arrhythmias are cited more frequently in the intraatrial variety although the difference between the incidence rates for both variants decreases with the observation time[19,20]. The experience in this work differs because a higher percentage of patients with the extracardiac option had this complication, but coincides with the results of Kumar et al.[21], which could be explained by the performing of surgery to older patients, who have lived longer in hypoxia and with possible atrial changes due to dilation. Both surgical techniques include incisions in sites with receptors or vagal terminations, the upper right atrium and the terminal ridge[22], therefore the stimulus for the development of reentry circuits exists in both.

The extracardiac variant does not prevent arrhythmias and makes difficult its treatment with ablation techniques for the inability to map the atrial territory through deep venous access[23].

In a study of 520 patients from seven centers[24], no differences were found in the occurrence of arrhythmias in relation to the type of TCPC. Other cited risk factors for arrhythmias include age for TCPC, existence of preoperative arrhythmias, moderate or severe AV insufficiency in the preoperative phase, heterotaxy syndrome and time lived with a TCPC[8,25]. None were identified in the study of CPWS.

Ono *et al.*[26] showed that patients with ventricular dysfunction and arrhythmias improved after perform-

Table 6. Decreased functional capacity. Multivariate analysis.

Variables	Wald	Sig.	Exp(B)
Kind of TCPC	0,53	0,46	0,61
Interval between connections	1,08	0,29	1,19
TCPC Age	0,00	0,98	0,99
Evolution time	1,05	0,30	1,06
Preoperative ventricular dysfunction	0,39	0,53	0,63
Preoperative pulmonary artery pressure	0,36	0,54	1,47
Preoperative ventricular diastolic pressure	0,00	0,97	1,02
Main ventricle	0,08	0,77	0,86
Two-stage surgery	2,07	0,15	5,27
Postoperative ventricular dysfunction	2,47	0,12	3,43

Hosmer and Lemeshow test p=0,421
TCPC: Total cavopulmonary connection

ing shunt fenestration. The working strategy in CPWS is to perform fenestration in all TCPC variants, probably this has influenced in the occurrence of fewer episodes of postoperative arrhythmia in this cohort.

Robbers-Visser et al.[27] relate the occurrence of arrhythmias in six years of follow-up, with the right ventricular morphology, without finding either a relationship with this technique. A group from the Heart Institute in Berlin[28] detected the intraatrial variant and right ventricular morphology as risk factors, which was not demonstrated in the work of the CPWS.

A study of six-year average follow-up of 65 patients with extracardiac TCPC found the occurrence of arrhythmias in 4.7 % compared with the previous performing of partial cavopulmonary connection[29]. Due to the manipulation of the sinus node area, the two-stage surgery exposes the risk of inflammation and healing process with increased arrhythmogenic potential with respect to those who had no prior surgery. Perhaps we should accept the existence of an alteration of the conduction system in these patients, which despite the strategies used, produces loss of sinusal rate[29].

Sinha et al.[30] found a relationship between preoperative AV failure and the occurrence of arrhythmias in the immediate postoperative period. Valve surgery associated with completion of the cavopulmonary connection increases surgical times but does not significantly increase perioperative morbidity and mortality[31].

In CPWS research no preoperative factor for the occurrence of arrhythmias was identified, perhaps due to the selection that favored good performance criteria or low dysfunction risk[13,32].

As a risk factor of postoperative arrhythmias, the existence of postoperative AV failure was identified, which gives importance to the effort to avoid this waste or surgical sequel and matches the experience of Brown et al.[33], who advocate the intraatrial option since it allows repair of the valve through the atrial access to the organic lesion detected preoperatively.

The analysis of options for TCPC dysfunction, the absence of ventricular assistance devices and indications for heart transplantation includes severe ventricular dysfunction attributable to chronic arrhythmias and to the negative inotropic effect of antiarrhythmic drugs[34,35]. This experience suggests the surgical option that deals with the cause of the arrhythmia before it deteriorates the general condition of the patient.

Detection of moderate AV impairment in postoperative assessment should justify reoperation of the patient in order to repair the valve lesion and prevent the occurrence of arrhythmias limiting the TCPC function. The reference to valve repair after TCPC is very rare[36,37]. Menon et al.[36] present, in a retrospective study of 61 patients, the reoperation of 61 patients, 72 % with arrhythmias, at an average time of 4.7 years from the Fontan procedure. Those who also had protein losing enteropathy or severe ventricular dysfunction showed poorer outcome, so the surgical indication should precede the appearance of some other complication associated with univentricular circulation.

Prospective randomized trials with both techniques are unlikely due to ethical implications, so perhaps with systematized Holter studies the incidence (as close as possible to the real one) of the type of arrhythmia and its time of onset could be detected, data not explored in this study.

The largest interval between partial cavopulmonary connection and its completion favors, for some authors, the time free of complications, so they re-

commend delaying the time of TCPC and considering the use of the extracardiac variant because of the possibility of using conduits according to body weight [38]. This study showed no relationship of the interval between the two cavopulmonary connections with the morbidity studied.

The completion of partial cavopulmonary connection prevents dilation in ventricles with volume overload due to previous surgical fistulas, hypertrophy in patients in whom pulmonary banding was performed or in patients with native pulmonary stenosis, myocardial fibrosis resulting from sustained hypoxemia and abrupt ventricular discharge when performing TCPC in one-stage surgery[39]. Probably a longer time with partial connection allows a better remodeling in response to ventricular discharge and a more favorable outcome in the completion of the bypass.

The reduction in ventricular preload enhances contractility and decreases the effect of the sustained hypoxemia on the myocardium with fibrosis and subendocardial ischemia[5]. However, the experience in patients operated between two and four years of age who did not reach the third decade of life due to impairment of ventricular function, gives importance to time lived with TCPC[40].

Nakamura et al.[41] looked for risk factors for ventricular dysfunction in hemodynamic studies with 48 patients for an average of 18 years and identified better function in the left ventricles, and the age as a risk factor when the bypass was indicated and time lived with it.

In the CPWS study no coincidences in this regard were found. The risk for postoperative ventricular dysfunction could be higher if there was preoperative dysfunction, perhaps because the full recovery of the previous ventricular damage is not guaranteed with the bypass, but rather its clinical manifestation or aggravation is delayed.

In a study with hemodynamic variables of TCPC patients with excellent outcome by an average of 18 years[42] a mean ejection fraction of 49.3 % (range 20-63 %) was found showing significant differences with the control group and it was concluded that survivors with excellent outcome may show ventricular dysfunction but normal volume and ventricular mass.

Both the increase in mass and its decrease outside normal limits correspond to a significant and progressive increase in diastolic pressures that accompany ventricular and bypass dysfunctions[41,43].

Evolutionary decrease of ventricular mass index, found in this CPWS study, should draw attention to the reduction of postoperative ventricular function and encourage the adoption of therapeutic measures to prevent or delay the subsequent ventricular remodeling.

Among potential risk factors analyzed, none of them was identified for ventricular dysfunction, so the role of changes in systemic resistance or afterload in the origin of this complication should be analyzed in future studies.

The increase in cardiac output during exercise is minimal in Fontan-type circulation due to the inability to increase preload and to the dependence on the pulmonary vascular resistance[44]. In exercise tolerance studies with an average interval of five years, parameters in functional capacity were better in children than in adults and a significant difference between the first and the last study with decrease in both ages. Exercise tolerance decreases as time spent with the total connection increases[45,46].

Ohuchi *et al.*[42] in the analysis of the evaluation every five years in patients with Fontan procedures, found a progressive reduction in functional capacity between 55 and 50 % in patients classified as not excellent. The only hemodynamic changes that distinguished patients who required hospitalization from those who did not, was increased ventricular diastolic pressure, which expresses dysfunction.

In this CPWS study, the presence of postoperative ventricular dysfunction doubled the likelihood of moderate or severe decrease in functional ability (RR 1.83, 95 % 1.14 to 2.29). The NNH was low and this makes the clinical expression of this complication more likely if a decrease in ejection fraction appears in the echocardiogram.

There was a higher percentage of patients with decreased functional capacity that underwent surgery with more than six years of age, with no significant differences in the age when total connection was indicated, time intervals between connections or time lived with TCPC. This result may be due to the application of a uniform protocol in the surgical indication regarding hemodynamic conditions at all times of the period studied.

Although there was no significant relationship, the percentage of patients with intraatrial TCPC who presented decreased functional capacity was greater than that of the extracardiac group, which is consistent with

the observations made by Anderson et al.[47] who found no relationship either with the primary ventricular morphology.

It was noticeable that a higher percentage of patients with two-stage surgery showed decreased functional capacity, which could be explained by the impact of two postoperative periods and the rehabilitation periods of each of them, the family control over the patient's physical activity, rehabilitation, individual perception of health as well as the difference in the ages at which they underwent surgery and the follow-up time[48].

In the experience of CPWS, the connection technique or time lived with the TCPC were not identified as risk factors for morbidity.

The periodic studies were conducted one or two years apart. Interval reduction of the future postoperative follow-up will allow early detection of identified risk factors and reduce the chances of developing signs of dysfunction with severe complications as those described.

In design of the study, the influence of medical treatments in the incidence of complications was not included, only their appearance in the follow-up period, which could modify the original data by not counting patients who did not present any of the complications derived from previous treatments. Another limitation is the difference in follow-up time, taking into account that the longer a TCPC patient is followed, the more likely some of the complications explored will be evident. Because of the risks associated with the procedure, not all patients agreed to the study by cardiac catheterization postoperatively.

CONCLUSIONS

The age at which total connection is performed, the interval between partial and total connection or type of TCPC are not risk factors for arrhythmias, ventricular dysfunction or decreased functional capacity. Moderate or severe AV valvular insufficiency of postoperative appearance is a risk factor for arrhythmias and its detection should produce therapeutic changes to avoid one of the most limiting complications in patients with TCPC. Detecting a decreased mass index during the postoperative follow-up is an indicator of the likely occurrence of ventricular dysfunction in patients with TCPC, which should generate therapeutic actions as it is a risk factor for reduced functional capacity. Postoperative regular and systematic assessment allows early detection of indicators of potential complications of univentricular circulation and their prevention.

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