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Outcome after Univentricular Surgery in A National Center of Developing Country

Alfredo Mario Naranjo Ugalde*, Eugenio Selman-Houssein Sosa, Eutivides Aguileras Sanchez, Maria Teresa Consuegra Chouarey, Gilberto Bermúdez Gutierrez

Cardiocentro Pediátrico William Soler, Ave. 100 y Perla, Altahabana, Boyeros, CP 10800, La Habana, Cuba.
**naranjob@infomed.sld.cu*

***Corresponding Author:** Alfredo Mario Naranjo Ugalde Cardiocentro Pediátrico William Soler, Ave. 100 y Perla, Altahabana, Boyeros, CP 10800, La Habana, Cuba.*

Abstract

Goal of cavopulmonary bypass in patients with univentricular heart, is the separation of systemic and pulmonary circuit and facilitate transpulmonary circulation with pressure differences between systemic venous bed and the main ventricle. There are several controversies about the indications, techniques and timing of the cavopulmonary bypass. The application of intratrial and extracardiac bypass in the William Soler Pediatric Cardiocenter began in 1992. A prospective cohort study was carried out from 1992 to 2011 in 92 patients, 39 with the extracardiac technique and 53 with the intratrial technique in order to assess the evolution and identify risk factors for complications. There were no significant differences in early morbidity and mortality. Surgery at one time is a risk factor for hospital mortality. No risk factors were identified for persistent pleural effusions, while postoperative atrioventricular insufficiency was identified for postoperative arrhythmia. Post-surgical ventricular dysfunction is related to the decrease in the ventricular mass index, predisposes to protein-losing enteropathy and decreased functional capacity. All complications first appeared in a greater number of patients with intratrial bypass but without significant differences. The extracardiac variant exposes a lower probability of late complications.

Keywords: *Univentricular, Congenital heart diseases, Fontan*

Introduction

Surgical treatment of congenital heart disease (CHD) in Cuba, contributed to reducing the infant mortality rate for this cause up to 0.5 per 1,000 live births in 2015(1). The William Soler Pediatric Cardiac Center (WSPCC) is the national reference center for treatment of CHD.

Incidence of CHD which only one ventricle keeps all circulation is reported from 0.3 to 3 per 1000 live births. (2) Without palliative surgeries 5% reaches two years of age and a lower percentage reaches adolescence.(3)

Total cavopulmonary connections or Fontan type operations (TCPC) were described on the theoretical basis to avoid the loss of kinetic energy by the turbulence of blood flow in previous atriopulmonary techniques.(4)(5)

Total intratrial cavopulmonary shunt or lateral tunnel (LT) consists in an intracardiac conduit with prosthetic material in half its circumference and atrial tissue to the other, which joins the inferior vena cava (IVC) with the right branch of the pulmonary artery, and a bidirectional Glenn or hemifontan operation. (6)(7)

Hemodynamic implications and complications with possible association to LT, led to the description of extracardiac conduit (EC). Prosthetic conduit diverts blood from the inferior vena cava to the right branch of the pulmonary artery, outside of atrial cavity. (8)

This system allows the flow according to pressure differences between the venous return, pulmonary vessels and ventricular diastole. When not working, they displayed clinical signs of low cardiac output, systemic congestion and hypoxemia. “Fontan failure” refers to decreased exercise capacity, ventricular dysfunction, arrhythmias, thromboembolism, of protein losing enteropathy and liver failure. (9)

Arrhythmias are the clinical expression of an electro physiology circuit substrate caused by dilation, surgery or abnormal arrangement native conduction pathways. In decreasing order relate to techniques atriopulmonary, LT and EC. However, no evidence benefit of using extracardiac derivation regarding long term arrhythmias. (10) Hemodynamic response to exercise is decreased in these patients, associated with arrhythmias, decreased ventricular function or ventricular morphology. (9), (11) Protein losing enteropathy is a rare complication of poor prognosis congestion caused by gastrointestinal or some deficiency in intestinal epithelial cells. (12)

Mortality associated with the operation is comparable to biventricular correction procedures. (13) The survival operated, despite the associated morbidity in 20 years reported up to 69%. (14) This result although not optimal, is a consequence of the better patient selection, the application of surgical protocols and improved postoperative treatments.

Risk factors of univentricular circulatory system dysfunction are difficult to identify. There is no effective strategy. (15) Current therapy is based on findings from retrospective data collection and experiences of each center. Although there is a selection of patient with adherence to preoperative hemodynamic criteria, the aforementioned complications appear unrelated demonstrated with one kind or another TCPC. (16)

Cavopulmonary connections were introduced in William Soler Cardiac Center in in 1990.It was considered necessary to identify in the evolution of patients with univentricular surgery factors that predispose to major complications and thereby modify the surgical medical strategies.

Methods

An observational, analytical, prospective cohort study, was conducted from January 1992 to January 2012 at the William Soler Pediatric Cardiac Center (WSPCC). Were included patients with univentricular circulation operated with total intratrial or extra cardiac cavopulmonary connection whose guardians expressed in the informed consent process compliance with the inclusion in this study. There were excluded patients with previous atriopulmonary operated for conversion to cavopulmonary shunt and those who would be impossible to maintain follow up in the shortest time postoperative of one-year.

Groups were formed according to surgical technique, which was not manipulated by the researcher but the result of the decision of the acting surgical team in each patient.

Surgical Techniques

Preoperative evaluation was performed according to the protocols of the Cardiovascular Surgery Pediatric Cardiology William Soler, taking into account the hemodynamic risk criteria. (17) Surgical teams worked according to the WSPCC guidelines for anesthesia, surgery, cardiopulmonary bypass, myocardial protection and postoperative care on both procedures. Methods to assess the evolution

To assess the evolution with both TCPC, mean timesof cardiopulmonary bypass, mean aortic cross-clamping, the occurrence of immediate postoperative complications, mortality and the occurrence of complications were compared during the study period.

Mean change in pulmonary pressures and ventricular diastolic in follow up, in patients who were prescribed cardiac catheterization at least once after DCPT was found. Variation was associated with the occurrence of pleural effusions, arrhythmias, decreased functional capacity, protein losing enteropathy and ventricular dysfunction.

Ratio between the ventricular mass found by echo cardiogram and body surface area was obtained. To calculate the variation of ventricular mass index, the difference between the measurement on the preoperative and the last postoperative examination was found.

Ventricular function was assessed by echocardiography through the ejection fraction (EF). The diagnosis of arrhythmias was explored using electrocardiogram scheduled per year, three years, five years and one final moment coincides with the completion of the study or the last query.

Functional capacity (FC) or exercise tolerance of the patients was evaluated through an exercise stress test with Bruce protocol stress on rolling mat. It was evaluated according to the percentage achieved the expected functional capacity. (3)(18)

For the analysis of free time of complications and survival were collected dates of occurrence of the events analyzed and times between them if occurred and if not, between the end time of the study and the realization of the TCPC.

Data Collection

Data were collected prospectively during scheduled hospital admissions in the preoperative period of TCPC, during admission for operation and four times; a year after the operation, three, five years after the intervention and an end time. A database was created.

Demographic variables, morphological diagnosis, presence of heterotaxia syndrome, history of previous palliative surgery, staged or not surgery, date of partial cavopulmonary, preoperative electrocardiogram, echocardiography and cardiac catheterization were studied. Perioperative variables were age at the time of the surgery, the type of bypass, extracorporeal circulation time, aortic clamping, pulmonary artery pressure and the pressures in the superior vena cava and the ventricle at the end of the operation. In the immediate postoperative, presence of complications was collected, duration of pleural effusions and outcome at discharge.The outcome variables were the result of electrocardiogram, echocardiogram and stress test, in addition to the presence or absence of complications. Time between partial and total derivation and time lived with TCPC until the end of the study for each patient were calculated.

Processing Techniques and Data Analysis

Data collection was performed prospectively in each scheduled income. All variables were stored and processed in a database created in the statistical program SPSS 13.5 (SPSS Inc., Chicago, Illinois, United States).

Frequency distributions and contingency tables were constructed. As summary measures percentages for qualitative variables and mean and standard deviations for quantitative variables were used. For analysis of association between qualitative variables the nonparametric test of independence Pearson chi-square and Fisher's exact test was used.

For quantitative variables parametric Student t test after analysis of variance homogeneity was used. A nonparametric test (Mann - Whitney) was used, if not meet the assumptions of normality t test for comparison of means.

Binary logistic regression was used to identify risk factors for complications and mortality. The following variables were related:

- Dependents: persistent pleural effusions, arrhythmias, protein losing enteropathy, risk ventricular dysfunction and decreased functional capacity.

- Independent: Age, morphological diagnosis, heterotaxia, type of TCPC, staged surgery, interval between partial and total bypass, type main ventricle, morphological diagnosis, moderate or severe valvular insufficiency, pulmonary pressure and preoperative ventricular risk, postoperative variation of these pressures, variation ventricular mass index and time of evolution.

The exponential coefficients (βExp) model as estimates of odds ratio (OR) was also analyzed. To validate the results in terms of significance a confidence level of 95% was used and all ≤ p value of 0.05 for the statistician associated with the test was considered significant.

To analyze the clinical relevance relative risk (RR), was calculated. Absolute risk reduction (ARR) and number needed to treat to reduce an event (NNT) were calculated as measures of impact. If the NNT was found a negative value, the number needed to harm (NNH) was found.

The project received the endorsement of the Academy of Sciences of Cuba (ACC) under the Ministry of Science, Technology and Environment (CITMA). It respected the provisions of the Helsinki Declaration. (19)

In accordance with that patients older than 18 years or guardians of children under 18 years of age signed consent to participate in research.

Results

Evolution of 92 patients with TCPC were studied; 53 LT and 39 EC variant in an average of 8.85 ± 5.83 years. Their characteristics are summarized in Table 1.

The EC was performed without cardiopulmonary bypass (CPB) in four patients without use of aortic cross-clamping in nine. Aortic cross-clamping time was higher in LT group (p = 0.046, 95% CI 0.18 to 20.17) (Table 2).

Table 1. *Patient characteristics*

	EC % n=39	LT %n=53	p
Age (years) ∞	8 ± 6,01	7,7 ± 2,8	0.77 τ
Male	53.8	41.5	0.34 χ
TA	30,8	35,8	0.77 χ
Non TA	69,2	64,2	
Left Mean Ventricle	48,7	54,7	0.83 χ
Right Mean Ventricle	46,2	41,5	
No determined Ventricle	5,1	3,8	
Heterotaxy	20,5	13,2	0,51 χ
Preop.AV regurgitation	38,5	56,6	0.13 χ
Previous palliative surgery	59	56,6	0.99 χ
Staged surgery	41	88,7	0,0001 χ
Follow up time ∞	10.54 ±7	7.23 ± 4.66	0,01 ů

EC: Extracardiac conduit; LT: Lateral Tunnel; TA: Tricuspid atresia; AV: Atrioventricular; preop: preoperative, X: χ^2 Pearson, T: t Student ; ů: Mann-Whiney U; ∞ Mean ± standard deviation

Table 2. *Aortic clamp and cardiopulmonary bypass time.*

Time. minutes ∞	LT	EC	p (CI 95%)τ
Aortic clamp	68,61 ± 22,49 n=51	58,43 ± 20,62 n=30	0,046 (0,18 - 20,17)
CPB	132,83 ± 47,09 n=53	128 ± 69,11 n=35	0,71 (-21,94 to 31,6)

CPB: cardiopulmonary bypass; LT: Lateral tunnel; EC: Extracardiac conduit. T: Student t; ∞ Mean ± standard deviation

Immediate postoperative complications were presented in 30 patients (76.9%) with EC and 71.7% (38) with LT. Frequency of complications found were not significantly associated with surgical technique (p = 0.57, RR = 1.07, 95% IC 0.84 to 1.37).

Survival at hospital discharge was 80.3% (Table 3). Mortality was significantly associated with surgery performing at a time (p = 0.04). The RR value showed that probability of death after surgery is doubled if the TCPC is performed in a surgical time [ARR -0.22 (95% CI -0.41 to -0.03) NNH 5 (95% CI 3-39)]. Diagnosis different to tricuspid atresia was also associated with mortality (p = 0.03) [ARR -0.2 (95% CI -0.34 to -0.06) NNH 6 (95% CI 3-18)] (Table 3). Although not significant, died 33.3% of 15 patients with heterotaxy.

Table 3. *Hospital mortality.*

Univariate analysis.	% N=18	p^x	RR (CI 95%)
LT	18,7	0,94	1,08 (0,47 - 2,5)
EC	20,5		
Non staged surgery	34,5	0,04*	2,72 (1,2 - 6,16)
Staged surgery	12,7		
TA	6,5	0,03*	0,25 (0,06 - 1)
Non TA	26,2		
Heterotaxy	33,3	0,16	1,97 (0,82 - 8,71)
Mean right ventricle	25	0,45	1,62 (0,7 - 3.7)
Mean left ventricle	14,6		
Time (year)∞	Alive	Dead	
Glenn-TCPC	4,51 ± 2,16	3,57 ± 1,84	0,25 τ (-0,66 to 0,94)
Age TCPC	8,36 ± 4,38	8,92 ± 6,25	0,72 τ (-3,77 to 2,65)

Multivariated analysis	Sig.	Exp(B)
TCPC type	0,26	2,89
Age	0,46	0,94
Non staged surgery	0,04*	6,46
Mean ventricle	0,39	1,74
Preoperative ventricle disfunction	0,21	5,31
Preoperative AV regurgitation	0,12	0,31
Arterial pulmonary pressure	0,43	2,05
Diastolic pressure	0,19	0,30

TA: Tricuspid atresia. TCPC: Total cavopulmonary connection

X: χ^2 Pearson, CI: 95 % confidence interval. $^{\tau}$: p value, Student t ; ∞ Mean ± standard deviation; *: p<0,05

Multivariate analysis (Table 3) identified performing TCPC in one time as a risk factor for postoperative mortality (p = 0.04; β Exp 6.45); which establishes the advantage of surgical strategy in two stages.

Persistent pleural effusions occurred in 18 patients with variant IA and 12 with the EC. Although 19 (63.3%) of 30 patients with persistent pleural effusions underwent the TCPC with over six years of age, no significant relationship between age and this complication was found. CPB time and aortic clamping were slightly higher in those who developed this complication.

Although univariate analysis did not reflect relationship, 38.5% of those with preoperative ventricular dysfunction, 36.4% (8 patients) with mean pulmonary pressuresand 41.7% (10 patients) with diastolic pressures of risk developed this complication.Multivariate analysis it failed to identify any risk factors among the variables analyzed for persistent pleural effusions.

Postoperative follow-up was performed in 74 patients, 43 with LT and 31 with EC. Variation of the ventricular mass was calculated in 50 patients (67.5%) and 31 (41.9%) underwent hemodynamic study in any of the income scheduled after surgery.

Arrhythmia occurred in 13 patients (Table 4). The RR was higher in patients who had moderate or severe postoperative atrioventricular regurgitation (AVR) [p <0.01; RR 7.47 (95% CI 3.15 to 17.7)]. The value of the absolute risk reduction of postoperative arrhythmia with AVR was negative [ARR -0.61 (95% CI -0.9 to -0.31)], indicating an increased risk. The number needed to harm was low [NNH 2 (95% CI 2 to 4)], which means a high frequency of occurrence of arrhythmias in patients with impaired AV postoperative more than light range.

No differences that relates changes in diastolic ventricular and mean pulmonary pressure withnew arrhythmias were found.

Multivariate analysis showed five times more likely to develop arrhythmias (p = 0.036; β Exp 5.59), when there is postoperative atrioventricular regurgitation (Table 4).

Table 4. *Arrhythmias.*

Arrhythmias Univariate analysis	Yes % n=13	p ^{X}	RR § (IC 95%)
LT	14	0,51	1,61 (0,6 - 4,34)
EC	22,6		
Preop. AVR	20,6	0,75	1,37(0,51 - 3,69)
No preop AVR	15		
Postop. AVR	70	0,0001*	7,47 (3,15 - 17,7)
No postop AVR	9,3		
Staged surgery	16,4	0,32	1,28 (0,44 - 3,69)
One time surgery	21,1		
Age TCPC> 6 y.o.	20,9	0,56	1,62 (0,55 - 4,79)
Age TCPC <6 y.o.	12,9		

Time (years)∞	Arrhythmias.	No arrhythmias.	
Follow up	6,78 ±4,46	9,01 ±6,17	0,22 τ (-5,87 to 1,37)
Glenn-TCPC	4,47 ± 2,03	4,52 ± 2,21	0,94 τ (-1,37 to 1,27)
Multivariated analysis		Sig.	Exp(B)
TCPT type		0,380	1.961
Age at TCPC		0,575	0,949
Staged surgery		0,326	0,382
Follow up time		0,076	0,873
Postoperative AVR		0,036*	5,594
Preoperative AVR		0,444	1,671

Preop. AVR: preoperative atrioventricular regurgitation; posop: posoperative; TCPC: Total cavopulmonary connection; LT: Lateral tunnel; EC : extracardiac conduit; X: χ^2 Pearson value; τ : p value, Student t; ∞ Mean ± standard deviation; § relative risk

Ventricular dysfunction occurred in 11 patients, seven of them with LT option. It was more frequent in patients with different TA diagnosis and main left ventricle. Of patients with preoperative dysfunction, 25% had postoperative dysfunction. Although the interval between partial and total connections and follow-up time in patients in whom ventricular dysfunction was detected were higher compared to those without this complication, the differences were not significant.

Association between ventricular dysfunction and decreased ventricular mass index (Tabla5) were explored. No significant differences that related changes in pulmonary arterial or diastolic ventricular pressure with ventricular dysfunction were found.In multivariate analysis, it was not identified as a risk factor for postoperative ventricular dysfunction.

Table 5. *Ventricular dysfunction and variation of ventricular mass index*

Ventricular dysfunction	Yes n=9	No n=41
Variation of ventricular mass index (g / m^2 BA) ∞	-23,38 ± 35,86	6,66 ± 29,15
p=0,01 * (IC 95% -52,5 to -7,5) T		

BA: body superficial area, T: p value, t Student. *: p<0,05; ∞ Mean ± standard deviation

Protein losing enteropathy (PLE) was diagnosed in six patients (Table 6), in an average of 4.42 ± 3.36 years. Five patients undergone LT and all had left ventricular morphology. Five were operated with more than six years old, everyone involved two stages surgery and the interval between the two operations was more than four years in four of them.

There was association between EPP and postoperative ventricular dysfunction [p = 0.004; RR 11.45 (95% CI 2.37 to 55.16)]. The absolute risk reduction EPP in the presence of ventricular dysfunction, was -0.33 (95% CI -0.62 to -0.04), which actually means an increased risk and NNH was 4 (95% CI 2-23). Variations in ventricular and pulmonary arterial pressure were not related to the EPP.

Multivariate analysis showed a greater probability of developing enteropathy if postoperative dysfunction appears (p = 0.004; βExp 24.63) (Table 6).

Table 6. *Protein-Losing Enteropathy.*

Univariate analysis	Yes % n=6	p X	RR§ (IC 95%)
LT	11,6	0,39	0,27 (0,03 - 2,25)
EC	3,2		
Postop. Vent. disfunction	36,4	0,004*	11,45 (2,37 - 55,16)
No postop disfunction	3,2		
Pleural effusions	9,7	0,32	2 (0,5 - 9,34)
No pleural effusions	4,8		

Time (years)∞	No PLE	PLE	
Glenn-TCPC	5,62 ± 2,71	6,18 ± 2,89	0,63 ᵀ (-1,75 to 2,87)
Age at TCPC	7,35 ± 3,94	9,33 ± 2,94	0,23 ᵀ (-1.31 to 5.27)
Follow up	8,71 ± 6,1	7,59 ± 3,97	0,66 ᵀ (-6,19 to 3,95)
Multivariated analyisis		Sig.	Exp(B)
TCPT type		0,169	0,13
Age at TCPC		0,167	1,22
Postop. Ventricular disfunction		0,004*	24,63
Previous pleural effusions		0,41	2,39

∞ Mean ± standard deviation

TCPC: Total cavopulmonary connection; postop: posoperative; ^{x}: p value, χ^2 Pearson ; $^{\tau}$: p value, t Student §: Relative risk. *: p<0,05

Moderate or severe decrease in functional capacity was detected in 33 patients. Table 7 shows a higher incidence in patients with LT. Association was found between decreased functional capacity and postoperative ventricular dysfunction [p = 0.04; RR 1.83 (95% CI 1.14 to 2.9)]. Decreased functional capacity during postoperative course with ventricular dysfunction, showed an increased risk [ARR of -0.33 (95% CI -0.62 to -0, 04)] and NNH 4 (95% CI 2-25).No significant relationship between changes in pulmonary artery pressures, ventricular pressures and functional capacity was found.

After hospital discharge and until the end of this study, 5 patients died, three with IA TCPC. Two EPP, one with gastrointestinal bleeding as the final event. A patient who left antiplatelet therapy died of embolism and neurological damage intervened four years early. One patient with EC TCPC presented system dysfunction caused by high systemic blood pressure and ventricular failure.

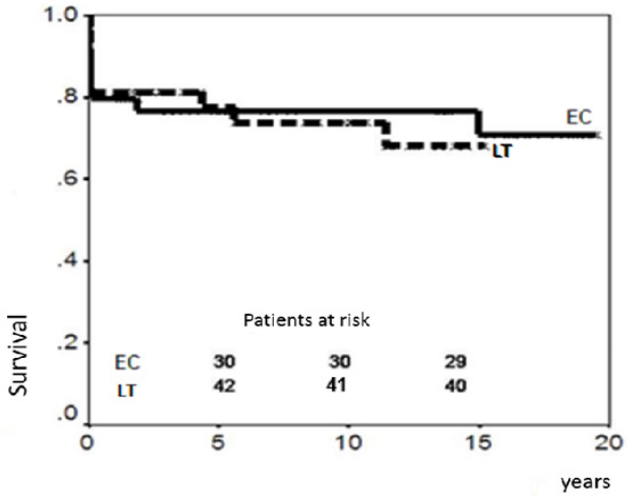
Tabla 7. *Diminished functional capacity. Univariate analysis.*

Diminished functional capacity	Yes % (n=33)	p ^{x}	RR§ (IC 95%)
LT	51,2	0,18	0,69 (0,39 - 1,21)
EC	35,5		
Mean right ventricle	43,3	0,89	0,93 (0,55 -1,58)
Mean left ventricle	46,3		
Preop. ventr. disfunction	41.7	1	0,92 (0,45 -1,9)
No preop ventr. disfunction	45,2		
Postop. ventr. disfunction	72,7	0,04*	1,83 (1,14 - 2,9)
No postop. ventr. disfunction	39,7		
Staged surgery	49,1	0,29	1,55 (0,76 - 3,71)
Time (years)∞	No diminished	Diminished	
Glenn-TCPC	5,73 ± 2,87	5,59 ±2,53	0,82 ᵀ (-1,41 to 1,13)
Age at TCPC	7,29 ± 4,58	7,79 ± 2,85	0,57 ᵀ (-1,23 a 2,33)
Follow up	8,48 ±6,03	8,79 ±5,92	0,82 ᵀ (-2,48 a 3,09)

∞ Mean ± standard deviation

TCPC: Total cavopulmonary connection; postop: posoperative; ventr.: ventricular; ^{x}: p value, χ^2 Pearson; $^{\tau}$: p value, t Student §: Relative risk. *: p<0,05

In LT survivors of immediate postoperative period the estimated survival was 97.7% at 5 years and equal to 10 years and 93.02% at 15 years. For EC was 96.7% at 5 and 10 years and 93.54% also at 15. The cumulative survival for LT was 77.4%, 73.7% and 68% at 5, 10 and 15 years. For EC was 76.6%, 76.6% and 70.7% respectively without significant differences between both techniques (log rank p = 0.85) (Figure 1).



Log Rank p=0,85 EC: Extracardiac conduit, LT: Lateral tunnel

Fig 1. *Cumulative Survival*

Discussion

Univentricular surgery is indicated in different morphological entities and their realization is not always attached to current indication criteria regarding the surgical time for reasons such as age at diagnosis, age at previous surgical palliation, the adoption of the strategy in one or two stage and hemodynamic risk assessment. (16)(20)(21)

In the early years of CPWS, patients who were the prevalence of cardiovascular malformations in Cuba were operated in advanced school age or adolescence. In two decades, the indication for surgery has dropped to internationally recommended ages moment. (22)

Anderson (15), described the average age of completion of the first referral to 1.6 ± 1.6 years and 3.4 ± 2.1 completion years old. Diller (23) studied a retrospective cohort of 321 patients from four institutions, in whom the shunt was completed between six and seven years old.

The average of eight years old at the time of completion of the TCPC in this paper, is a consequence of the extended period under study, which has reduced the age of indication from age 19 until the third year of life.Early completion of the CPC reduces the time living with hypoxemia and promotes the nutritional recovery. (24)

There is no experience WSPCC in TCPC in hypoplastic left heart syndrome, which is in many reports a higher percentage of casuistry associated with an early indication, which also justified a mean age greater indication of the TCPC in the present.(25)

As others reports, age and time between partial and total connectionswas not identified as a risk factor of morbidity.(22)However, older age at the time of completion of the bypass is associated with the presence of valvular insufficiency and arrhythmia, which relates to a longer period living with volume overload, possible cause of the annular dilatation AV and atrial dilation. (15)

It seems essential the realization of the partial CPC in a previous surgical time to avoid dilation ventricles with volume overload due to previous surgical shunts, hypertrophy in patients withpulmonary banding or native pulmonary stenosis, resulting myocardial fibrosis caused by maintained hypoxemia and abrupt ventricular unloading when performing a total derivation. (26)

Gerelli(27) stated that the delay in TCPC could not influence the evolution from the experience in 70 patients at high risk for type Fontan circulation. Age indication TCPC and the interval between the CPC was greater than the CPWS experience of 9 years (2-23) and 7 years (1 to 16) respectively.

To delay TCPC could allows a better selection of conduit for extracardiac variant, reducing the morbidity associated with the disparity of the conduit with the weight or size of the patient and the delay in the presentation of altered blood flow in Fontan circulation. (28)

The mean age of completion of CPC of the deceased and survivors in the study of WSPCC were no different, there is no significant difference in the interval between

the partial bypass and total, however in the group of the deceased the interval was lower. It is likely that more time alone with the partial bypass allows better remodeling in response to ventricular unloading and more favorable evolution.

Regarding type of TCPC could be advantageous extracardiac option to take into account the significant difference in the time of aortic clamping; however, no significant difference was found in the occurrence of postoperative complications related to the technique, so perhaps the determining factor is not the use of aortic clamping if there are strategies to treat or prevent negative effects.

Partial bypass allows ventricular remodeling, progressive normalization of the ratio of the mass with the volume of the ventricle,better diastolic function at the time of completing the derivation and lower morbidity. (29)(30) Although the TCPC survivors at one stage not face the complications of reoperation to complete a previous partial bypass, suffer the consequences of sudden ventricular unloading, which increases the risk of intolerance circulatory model(31)

In this paper, mortality of the group operated in a stage doubled the group operated in two stages it time was identified as a risk factor for mortality.This strategy was associated with the historical moment, the age of adolescents and adults attended in the early years, its hemodynamic conditions and experience of cardiovascular surgical team in WSPCC, as reported by other authors. (25)(28)

The different tricuspid atresia (AT) diagnosis was also associated with mortality. These patients may have ventricular misbalance and single valve AV or double ventricular outflow mostly ventricles right morphology and although it was not significant relationship between ventricular morphology and mortality, more than 50% of the dead were main right ventricle, described by its architecture with less chance of facing systemic vascular resistance and ventricular download.(32)

The absence of identification of a risk factor for the onset of persistent pleural effusions in this work suggests the possibility of finding relationship of this complication with some element not explored as pulmonary vascular resistance opposing the transpulmonary flow.

For some authorsarrhythmias are associated with the type of technique and frequently cited in LTvariety, although the difference between incidence rates for both variants decreases with time observation. (33)

Experience in this paper differs because a higher percentage of patients with this complication with EC option, coinciding with Kumar,(34)it could be related with use of conduit in older patients, who have lived in greater hypoxia time and possible atrial dilation changes, structural basis for electrical alterations and clinics.

Both surgical techniques include incisions on vagal receptors sites terminations, the upper right atrial wall and crista terminalis, so there is always a stimulus to the onset of reentry circuits.(35)

In a study of 520 patients from seven centers, no differences were found in the occurrence of arrhythmias relatedwith the type of TCPC.(36) Other risk factors include age at the TCPC, preoperative arrhythmias, heterotaxia and time lived with a TCPC. (37)(38)None was identified in the study of WSPCC.

Arrhythmias are related to age in the natural history of Tetralogy of Fallot, transposition of great vessels or total anomalous pulmonary venous drainage patients without surgical treatment. Probable causes are hypoxemia and increased atrial pressures as in univentricular patients.(37)(39)

Strategy in the WSPCC is to fenestrate all variants of TCPC, probably this has affected the appearance of fewer episodes of postoperative arrhythmia in this cohort.

Sinha(40) found relationship between preoperative AV failure and arrhythmias in the immediate post operative period. At follow-up after discharge only found relation with the intervention to older age and higher mean pulmonary pressures preoperatively.

Preoperativefactor for arrhythmias was not identifiedin this research. A group Heart Institute in Berlin 218 found that the risk factors for the occurrence of tachyarrhythmias are the LT variant and right ventricular morphology, which was not demonstrated in the work of WSPCC.(36)

As a risk factor for postoperative arrhythmias, it was identified the existence of more than mild postoperative atrioventricular insufficiency, which gives importance to the effort to avoid this residue or surgical sequel

and coincides with the experience of Brown, who advocates LT option as it allows repair of the AV valve. (41)(42)

The analysis of options to Fontan dysfunction, the absence of ventricular assist devices and heart transplantation indications which include severe ventricular dysfunction attributable to chronic arrhythmias and negative inotropic effect of antiarrythtmics drugs (43)focus the surgical option to treats the cause of the arrhythmia before it deteriorates general conditions.

Detection AV moderate range insufficiency in the postoperative evaluation should, in the author's opinion, justify patient reoperation in order to repair the valve lesion and avoid the appearance of limiting arrhythmias.

Univentricular surgery has beneficial effects on preoperative ventricular dysfunction.(44)However, there is the experience in patients operated between two and four years who did not reach the third decade their lives, when there was deterioration of the ventricular function, which prints importance in thetime living with total bypass. (45)

Hemodynamic studies of 48 patients for an average of 18 years identified better function in patients with main left ventricle and as a risk factor the age at indication of the derivation and time lived with it.(46) There were not found in this report matches in this regard.

Survivors TCPC with excellent performance can show ventricular dysfunction but normal ventricular volume and mass.(47)

It had been found a greatly increased ventricular mass in patients undergoing surgery in adulthood compared to that found in younger patients operated without significance, with better ventricular function and exercise tolerance in those were completed Fontan before 18 years old.(45)

Both the increase in mass as its diminution outside normal limits correspond to the significant and progressive increase in diastolic pressures that accompany ventricular dysfunction. (21)(46)

Evolutionary decreased ventricular mass index, finding of this report, should draw attention to the reduction of postoperative ventricular function and encourage the adoption of therapeutic measures that prevent or delay the consequent ventricular remodeling.

Risk factor were not identified for ventricular dysfunction, so maybe it should look in future studies, the role of the possible variation of resistance or systemic afterload in the origin of this complication.

Protein losing enteropathy (PLE) is reported from 1% to 11% of patients living with a TCPC, in this reportwas 8.1%.(48)It is associated with a mortality of 30% to 50% five years after the diagnostic.(49) No etiology has been identified.

In a study of hemodynamics before and after the onset of PLE in 7.8% of patients with TCPC , it was found that the central venous pressure and pulmonary resistance were significantly elevated compared the group of excellent evolution. Ejection fraction was also significantly lower.(50)

Unlike Rogers,(28) who states as a risk factor of the PLE history of pleural effusions, it was not found in this investigation relationship between them.

In this research, a high likelihood of this complication after detecting ventricular dysfunction of moderate to severe postoperative evolution was detected. Treatment and monitoring must then goto the search early development this indicator.The NNH was low and this makes it more likely the clinical expression of this complication if a decrease in ejection fraction appears in the echocardiographic study.

The functional capacity of patients with TCPC circulation is limited as an expression of the decreased cardiac output. In this report there was a higher percentage in patients operated with more than six years of age and intervals between CPC over four years with decreased functional capacity, however, no significant differences were observed in the age of surgical indication, intervals between derivations or time lived with TCPC. This result is perhaps result of the application of a uniform hemodynamic conditions for surgical indication as protocol.

Higher percentage of patients with two staged surgery showed decreased functional capacity, which could be explained by the impact of two postoperative and rehabilitation periods, family control over the patient's physical activity, individual perception of health, as well as the difference in ages that were operated and follow up time. (51)

Exercise intolerance is associated with increased morbidity but not related to mortality or need for heart transplantation.(23)An analysis of the evaluation

every five years of patients with Fontan operations, found a progressive reduction in the expected functional capacity between 55% and 50% in patients classified as non-excellent. The only hemodynamic changes that distinguished patients who required no income was increased ventricular diastolic pressure, which expresses dysfunction.(47)

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

Regarding late mortality, it had been cited as risk factors to be operated with more than 16 years of age, heterotaxia syndrome, right morphology ventricular and AV valve regurgitation, which highlights the importance of prevention and detection the latter modifiable factor.(44)

In a study of 261 operated at Children's Hospital Boston (14), 78% of deaths were perioperative collected. For the group of living patients, the percentage of freedom of transplantation or survival at 5, 10, 15 and 20 years was 93.7%; 89.9%; 87.3% and 82.6%. The higher percentages estimated in CPWS work are attributable to that study included atriopulmonary derivations.

WSPCC experience coincides with reports of presentation of the final event were long-term ventricular dysfunction, sudden death and arrhythmia probably associated with thrombosis duct and embolism through the fenestration. (14) Immediate morbidity showed no significant differences in relation to the bypass technique so the appearance of complications responded to other causes and not the type of procedure. The emergence of medium-term complications studied also showed no relation to the type of procedure. Early detection of identified risk factors may decrease the chances of dying or dysfunction with serious complications such as those described.

Time between follow-up studies should be related to the possible progression of geometric and functional changes described. There were established complications detected during at least one year so exploration every six-month could be vital to detect any risk factors before. Echocardiographic studies can be performed, if the training of specialized medical personnel is aimed at early detection of risk factors described, can undertake medical actions to prevent or delay associated morbidity, reducing hospital or family and personal expenses cost.

Limitations

In the design of the study the influence of medical treatments in the incidence of complications, the appearance of them in the period of follow-up was not included, which could modify the original. Another limitation is the difference in time tracking, taking into account that the longer a patient with TCPC, it is most likely that there appear some explored complications. Because of the risks associated with the procedure, not all patients agreed to study postoperative cardiac catheterization.

Conclusions

Variation in ventricular mass index is a risk factor of ventricular dysfunction and this is for protein losing enteropathy and reduced functional capacity. We recommend the study and surgical treatment of major slight residual atrioventricular valvular insufficiency which is a risk factor of new arrhythmias and promote the design of new research on the way to explore other possible actors of this pathophysiology, as transpulmonary resistance and systemic circulation, which seem to gain importance once studied are not identified as determinants of complications most limiting, with the ultimate goal to improve and prolong the lives of patients with total cavopulmonary connection.

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