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Percutaneous interventions in Fontan circulation

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ABSTRACT

Introduction and objectives: Different percutaneous interventional procedures are needed to reach and maintain adequate anatomical and physiological conditions for the Fontan circulation. We aim to describe the experience gained at a children's hospital in such interventions, and to analyze the clinical outcomes.
Methods: Retrospective study of all patients with Fontan circulation completed between 1995 and 2013. We analyzed the clinical characteristics and the different types of percutaneous interventions performed, considering three different periods of time: before Glenn surgery, between Glenn and Fontan surgeries, and after Fontan was completed. Survival and time to indication of percutaneous interventions in each period were analyzed, as well as the clinical situation at last follow-up.
Results: Of the 91 patients analyzed, 46 (50.5%) required percutaneous interventions. The most frequent procedures were pulmonary artery angioplasty and angioplasty of the Fontan conduit. Estimated survival at 10, 20 and 30 years of age was 96.2%, 94.7% and 89.4%, respectively. There were no significant differences in survival of patients undergoing percutaneous interventions or not. Overall survival and time to indication of percutaneous interventions were significantly lower in the group of patients with right morphology systemic ventricle. Patients with fenestrated Fontan required interventions more frequently. At the end of follow-up, 66 patients (72.5%) were asymptomatic, without significant differences between patients who underwent or did not undergo percutaneous interventions.
Conclusions: Interventional catheterization procedures are often necessary to reach and maintain the fragile Fontan circulation, mainly in patients with right morphology systemic ventricles and fenestrated Fontan conduits.

1. Introduction

Total cavopulmonary derivation (TCD) surgery consists, regardless of the changes emerged since the first description of the technique by Fontan in 1971 [1], in the connection of the systemic venous flow to the pulmonary arteries passively, bypassing the ventricular impulse. This is performed through the direct connection of the superior vena cava (SVC) to the pulmonary branches in a first staged surgery (Glenn operation), and of the inferior vena cava (IVC) in a second staged procedure (Fontan operation), achieving so a complete venous return derivation into the pulmonary circuit.

For such univentricular physiology, favorable anatomic and hemodynamic parameters are needed. These are frequently achieved only with previous percutaneous or surgical procedures when necessary. In general, these determinants include a sufficient but not excessive pulmonary flow in early stages, optimal ventricular function, absence of significant stenosis or regurgitation of the systemic atrioventricular valve, sinus rhythm, low pulmonary resistance and pulmonary arteries of adequate size [2].

In recent years, given the increased survival of patients with Fontan circulation, percutaneous techniques have emerged for new indications or to solve complications, which aim to keep this fragile physiology in optimal circumstances. We present in this paper the cumulative work experience in structural percutaneous procedures performed at our institution over the past 18 years in patients with univentricular physiology.

2. Materials and methods

2.1. Study population and analyzed variables

All patients who had a TCD completed at La Paz Children's University Hospital in Madrid from 1995 to October 2013, and at least one follow-up visit, were included. No patients with cavopulomonary bypass pending

Abbreviations: CHD, congenital heart disease; IVC, inferior vena cava; NYHA, New York Heart Association; PI, percutaneous intervention; SVC, superior vena cava; TCD, total cavopulmonary derivation.

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completion or who were considered candidates for biventricular physiology were included. The variables were collected retrospectively from medical reports and paper or computerized clinical history of patients. In addition to demographic data, information on the type of congenital heart defect (CHD), the date of Glenn surgery and Fontan surgery, the date of each interventional catheterization, and the type of procedure performed were obtained. Moreover, information on the need of other than Glenn or Fontan surgery during follow-up was collected. Three periods of time were considered for presentation of the results regarding the performance of percutaneous interventions (PIs): prior to Glenn surgery (pre-Glenn), between Glenn and Fontan surgery, and after Fontan surgery (post-Fontan). The last date of follow-up of each patient, and the date of death in patients who died, were recorded. Also, the overall clinical status of each patient at follow-up, according to their functional New York Heart Association (NYHA) class, and the presence of relevant issues related to the failure of Fontan circulation, such as protein-losing enteropathy, plastic bronchitis, desaturation (arterial oxygen saturation below 90%) or inclusion on heart transplant list, were recorded. To assess the clinical outcome and mortality of the patients with follow-up in other centers, a telephone contact was made in October 2013, by the end date of the overall monitoring of the cohort.

All data were recorded and used in accordance with the applicable data protection laws and ethical principles of the Declaration of Helsinki.

2.2. Indication of percutaneous interventional procedures

Indications of the different types of PI were mainly proposed by the clinical cardiologist responsible for each patient's follow-up in our high-resolution (clinical evaluation + electrocardiogram + echocardiography) outpatient clinics. Other times, it is the interventional cardiologist who proposed a specific PI based on the hemodynamic or anatomic findings in pre-Glenn and pre-Fontan diagnostic catheterizations that, most of the times, all patients on the way to univentricular physiology undergo. In any case, all PI proposed were discussed and finally indicated during Children Heart Team meetings where clinicians, interventional cardiologists and surgeons responsible for the care of each patient participated.

2.3. Cardiac catheterization

All catheterizations were performed under general anesthesia and mechanical ventilation with endotracheal intubation. The route of venous access was femoral or jugular and arterial access was femoral.

2.4. Statistical analysis

Qualitative variables were expressed as percentages and the relationships between them were analyzed using the χ^2 test or Fisher's exact test. Quantitative variables were expressed as mean ± standard deviation for normally-distributed variables, or as median [interquartile range] for variables which were not normally distributed. To test the hypothesis of normal distribution of quantitative variables, the Kolmogorov–Smirnov test was used. Relationship between continuous variables of normal distribution was analyzed according to the number of existing categories, using the Student t test or ANOVA with post-analysis according to the Bonferroni method; continuous variables not normally distributed were analyzed using Mann–Whitney or Kruskal–Wallis tests, depending on the number of categories. Significant value of $p < 0.05$ (two-tailed) was considered statistically significant. For the overall survival analysis and time to the indication of PI, the Kaplan–Meier method was used. Comparisons were performed using the log-rank test. The statistical analysis was performed using SPSS PASW statistics 15.0 package (SPSS Inc., Chicago, Illinois, United States).

3. Results

From 1995 to October 2013, a total of 91 patients with CHD underwent a TCD in two stages: first the superior cavopulmonary connection (Glenn surgery), and in a second time the inferior cavopulmonary connection (Fontan). In 15 patients (16.5%) our team opted for a fenestration of the Fontan conduit into the right atrium

Table 1
Basal characteristics of the cohort of patients.

	Total cohort (n = 91)	Patients with percutaneous interventions (n = 46)	Patients without percutaneous interventions (n = 45)	p
Demographic characteristics				
Age at last follow-up or death (years)	15.5 ± 5.4	14.6 ± 5.8	16.4 ± 4.9	0.107
Age at Glenn surgery (years)	1.6 [0.9–3.1]	1.5 [0.8–3.0]	2.0 [1.3–3.2]	0.249
Age at Fontan surgery (years)	7.0 ± 2.8	6.8 ± 2.7	7.3 ± 3.0	0.409
Males (%)	57 (62.6)	30 (65.2)	27 (60.0)	0.607
Cardiopathy				
Hypoplasia/atresia of left ventricle/valves (%)	37 (40.7)	26 (56.5)	11 (24.4)	**0.014**
– Hypoplastic left ventricle*	13	11	2	
– Double outlet RV with ventricular disbalance	12	5	7	
– Mitral atresia	5	4	1	
– Shone syndrome	4	4		
– Mitral-aortic atresia	2	2		
– Criss-cross heart	1		1	
Atresia or stenosis of right valves (%)	37 (40.7)	15 (32.6)	22 (48.9)	0.250
– Tricuspid atresia	20	6	14	
– Pulmonary atresia with intact septum	6	2	4	
– Hypoplastic RV*	5	3	2	
– L-TGA with pulmonary atresia	3	1	2	
– Tricuspid and pulmonary atresia	2	2		
– D-TGA with tricuspid stenosis	1	1		
Left morphology single ventricle (%)	9 (9.9)	3 (6.5)	6 (13.3)	0.317
Right morphology single ventricle (%)	8 (8.8)	2 (4.3)	6 (13.3)	0.157
Systemic ventricle				
Left morphology (%)	42 (46.2)	16 (34.8)	26 (57.8)	**0.028**
Right morphology (%)	49 (53.8)	30 (64.2)	19 (42.2)	

Continuous variables are presented as mean ± standard deviation (normal distribution) or as median [interquartile range] (non-normal distribution). Right column shows the p value for comparisons between patients with and without percutaneous interventions. RV: right ventricle; L-TGA: L-transposition of the great arteries; D-TGA: D-transposition of the great arteries.
* Cases of ventricular hypoplasia without a primary valvular cause.