Interventional catheterization in pediatric patients after Fontan procedure

To the Editor,

The advances made in surgical techniques followed by the best candidate selection process possible and optimal preparation of patients eligible for surgery have contributed to reducing early postoperative mortality in patients treated with the Fontan procedure. However, the balance provided by the Fontan procedure is precarious and can fail for multiple reasons that can be resolved through interventional catheterizations (IC).

This study describes the experience of our center performing IC and focuses on the type of interventional procedures performed and predictors of these.

We conducted a retrospective study of all patients < 18 years-old who underwent a Fontan procedure from January 2000 through December 2021 and were treated with IC due to suspected anatomical injury detected by echocardiography (annually) or magnetic resonance imaging (every 10 years and then every 3 years) or complications like protein-losing enteropathy, plastic bronchitis or hepatic injury detected by echocardiography (annually) or magnetic resonance imaging. A total of 79 interventional procedures were performed 10 years after the Fontan procedure.

All patients gave their prior written informed consent, and the study was approved by the hospital ethics committee.

Descriptive statistics of the demographic, anatomical, hemodynamic, and surgical variables was used. Normally distributed variables were expressed as mean and standard deviation while those without a normal distribution were expressed as median and interquartile range [IQR]. Kaplan-Meier curves were drawn to estimate the IC-free survival rate. To identify predictors of the need for IC, univariate Cox logistics regression analysis was conducted. Variables with significance levels < .2 were included in the multivariate analysis. Also, hazard ratios (HR) with a 95% confidence interval (95%CI) were estimated.

A total of 74 patients treated with a Fontan procedure were identified. Their demographic, anatomical, and pre-catheterization characteristics are shown on table 1. After a median follow-up of 10.3 years after the Fontan procedure, the IC-free survival rate was 63% and 45% at 5 and 10 years, respectively (figure 1A). No deaths were reported associated with cardiac catheterizations. A total of 4 patients (5%) experienced complications associated with the catheterizations (pulmonary thromboembolism, brachial neuropraxia, vasoactive drug administration during the procedure, and pulmonary atelectasis).

The diagnosis of hypoplastic left heart syndrome (HR, 2.62; 95%CI, 1.18-5.78), and the values of mean pulmonary artery pressure (HR, 1.2; 95%CI, 1.02-1.41), the transpulmonary gradient (HR, 1.64; 95%CI, 1.21-2.22), and the McGoon index (HR, 0.18; 95%CI, 0.07-0.44) prior to the Fontan procedure behaved as independent predictors of the need for IC after this surgery (figure 1B).

In our patients, the rate of interventional procedures performed [47%] is similar to that reported in the series by Downing et al. with an IC-free survival rate of 53% at 15 years. Although the number of procedures performed is quite similar, in their case, the closure of fenestration was the most common procedure of all due to their high rate of fenestrated Fontan (90%) compared to ours (35%). Nonetheless, when our series was compared to others with older patients, significant differences were found. A total of 49% of the patients from the series of Van Dorn et al. (1978 through 2002) were treated with a traditional atrio pulmonary connection. Most interventional procedures were pacemaker implantation or replacement (26%) or arrhythmia ablation (20%).

Our clinical practice attempts the closure of the fenestration 6 months after the Fontan procedure if the patient’s disease progression is favorable, pressure remains < 16 mmHg during the occlusion test, and proper cardiac output is preserved (> 2 L/min/m² with a decrease of < 20% compared to baseline levels). The presence of aortopulmonary collaterals has proven to have a negative effect on Fontan circulation, thus extending the duration of pleural effusions and causing ventricular volume overload. Therefore, we delve into an aggressive search and embolization of these collaterals in the catheterizations performed before and after the Fontan procedure.

The lack of stenosis in the Fontan conduit and pulmonary branches is essential to keep proper hemodynamics in Fontan circulation. Therefore, it seems logical to treat stenoses even in asymptomatic patients.

Regarding the risk factors associated with performing IC, the diagnosis of hypoplastic left heart syndrome was seen as an independent predictor of this event in both the series of Downing...
et al. and our own. Elevated pulmonary pressures and resistances, and smaller pulmonary arteries are known factors of poor prognosis in this population.

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CONFLICTS OF INTEREST

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