A therapeutic Fontan fenestration closure

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Introduction

The Fontan procedure directs systemic venous blood to the lungs without a ventricle and is used for various single-ventricle heart defects. Fenestration improves early postoperative outcomes, but may lead to long-term issues such as persistent cyanosis, thromboembolism, and possible need for later closure.

Case report

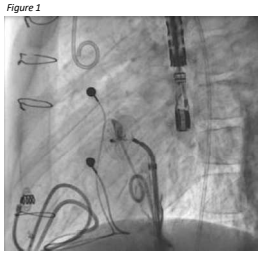
The case involves a 9-year-old girl with an unbalanced atrioventricular septal defect and left ventricular hypoplasia, treated with staged palliation including Glenn and fenestrated extracardiac Fontan, plus an epicardial pacemaker. She has recurrent fainting and exertional cyanosis. After a recent episode with desaturation and tachycardia, cardiac catheterization was performed.

Catheterization procedure

A mean pressure of 18 mmHg was observed in both the pulmonary circulation and the Fontan circuit. The left ventricular end-diastolic pressure and the left atrial pressure were normal (8 mmHg). A fenestration occlusion test was carried out using an 18 mm AGA balloon, with no increase in the mean pulmonary arterial pressure (PAPm), which remained stable at 18 mmHg. No changes were observed in systemic aortic pressure. A marked increase in systemic oxygen saturation was noted, rising from 84-85% at baseline to 98-100%. Subsequently, the closure of the Fontan conduit fenestration was performed using an 18 mm Amplatzer Talisman PFO Occluder device. (*Fig. 1*)

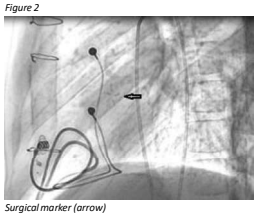
Following the closure of the fenestration, an increase in oxygen saturation levels was observed (SpO2 93-96%), and no arrhythmic events were noted during the hospitalization.

Figure 1



Amplatzer Talisman PFO Occluder device 18 mm

Figure 2



Surgical marker (arrow)

Discussion

Thanks to the presence of a surgical marker (Figure 2) placed by the surgeons during the Fontan procedure, we were able to easily locate the fenestration and use this marker to correctly place the closure device. This is an interesting case because, typically, surgeons place surgical markers in non-fenestrated Fontan conduits to provide interventionists with a reference for a future percutaneous fenestration and to give electrophysiologists a marker for perforation, should electrophysiological studies be necessary. In this case, however, the marker was placed during the surgical Fontan fenestration specifically to give interventionists a marker to close the fenestration if needed. We deemed the closure of the fenestration appropriate based on the immediate improvement in saturation during the fenestration occlusion test: the episodes of fainting and desaturation were frequent and debilitating. We are currently evaluating the possible need to administer sildenafil to the patient during follow-up.

Conclusions

Surgical Fontan fenestration can improve early outcomes, but in many cases, a subsequent percutaneous fenestration closure may be necessary to improve long-term outcomes. In this context, collaboration between surgeons and interventional cardiologists is essential to ensure optimal outcomes for patients following surgery or percutaneous procedures.