

Transcatheter closure of extracardiac Fontan fenestration in a cyanotic patient

Fikri Demir¹, Tevfik Karagöz², Ebru Aypar¹, Semra Atalay¹, Ercan Tutar¹

Pediatric Cardiology Units of ¹Department of Pediatrics, Ankara University Faculty of Medicine, and ²Department of Pediatrics, Hacettepe University Faculty of Medicine, Ankara, Turkey

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Fontan operation is the connection of the systemic venous return to the pulmonary arteries, performed in patients with single ventricle physiology. The Fontan circuit is commonly fenestrated because of early postoperative risks such as high systemic venous pressure and low cardiac output. As it causes progressive cyanosis and increased risk of paradoxical embolism in the follow-up period, occlusion of these fenestrations is generally suggested. Successful closure of extracardiac Fontan fenestration with atrial septal occluder in a 10-year-old girl is reported herein. To our knowledge, this is the first such report from Turkey.

Key words: extracardiac Fontan, fenestration, transcatheter closure.

Fontan operation is a palliative surgical procedure, connecting systemic venous return to the pulmonary arteries, and it is used in patients with a single ventricle physiology. Because of its advantages, such as lower risk of ventricular dysfunction, arrhythmias and baffle leaks and shorter cardiopulmonary bypass duration, a modified form, extracardiac Fontan, involving placement of an extracardiac conduit to connect the inferior vena cava with the pulmonary artery, is frequently preferred^{1,2}. In order to avoid early postoperative risks such as high systemic venous pressure and low cardiac output, a fenestration between the Fontan circuit and pulmonary venous side is commonly placed, especially in high-risk patients³.

Although fenestration is beneficial in early postoperative states, it causes progressive cyanosis, polycythemia and increased risk of paradoxical embolism because of right-to-left shunt in the follow-up period^{3,4}. Thus, transcatheter occlusion of these fenestrations should then be performed, in the event it does not close spontaneously^{5,6}. Herein, we report a successful closure of extracardiac Fontan fenestration with an atrial septal occluder in a cyanotic girl. To the best of our knowledge, it is the first report from Turkey.

Case Report

A 10-year-old girl had been diagnosed with tricuspid atresia, atrial septal defect, ventricular septal defect, hypoplastic right ventricle, and pulmonary stenosis in the newborn period. A bidirectional Glenn shunt had been constructed between the superior vena cava and right pulmonary artery at the age of nine months. She had undergone an extracardiac Fontan procedure with a fenestration (at 5 years of age).

During the follow-up visits in the last five years, she had moderate cyanosis (peripheral oxygen saturation of 75-80% on room air), polycythemia, clubbing, and mild effort intolerance. Transthoracic echocardiography revealed considerable right-to-left shunt across the fenestration. Accordingly, to eliminate cyanosis and increased risk of paradoxical embolism, it was decided to close the fenestration.

After an informed consent was obtained from the parents, two right femoral venous and one right femoral arterial sheaths were placed under complete midazolam and ketamine sedation. Heparin was given in a dose of 100 IU/kg. Three doses of cefazolin (50

mg/kg/dose) were administered at 8-hour intervals for antimicrobial prophylaxis. The pressures and oxygen content in systemic veins, pulmonary artery, common atrium and aorta were measured. An extracardiac conduit angiography was performed, and a significant shunt across the fenestration into the systemic circulation was observed (Fig. 1). No other cause of cyanosis was determined. The mean pressure in Fontan circuit was 9 mmHg, and peripheral oxygen saturation was 92%. The fenestration was temporarily occluded with a 12 mm balloon catheter passed over an exchange wire (Fig. 2) and the diameter of the fenestration was measured as 6 mm. Hemodynamic measurements were repeated 15 minutes after balloon occlusion. Peripheral oxygen saturation increased from 92% to 99%. As no significant increase in mean systemic venous pressure (12 mmHg) and no decrease in cardiac output were observed, it was regarded as suitable for permanent occlusion.

A long 8 Fr device delivery sheath with its dilator was conducted in the right atrium from the fenestration. The dilator was withdrawn and the position of the sheath was confirmed by transthoracic echocardiography. A 7.5 mm Occlutech™ atrial septal occluder was introduced through the sheath with its delivery wire and the left atrial disc was opened in the atrial side. After it was slightly pulled back

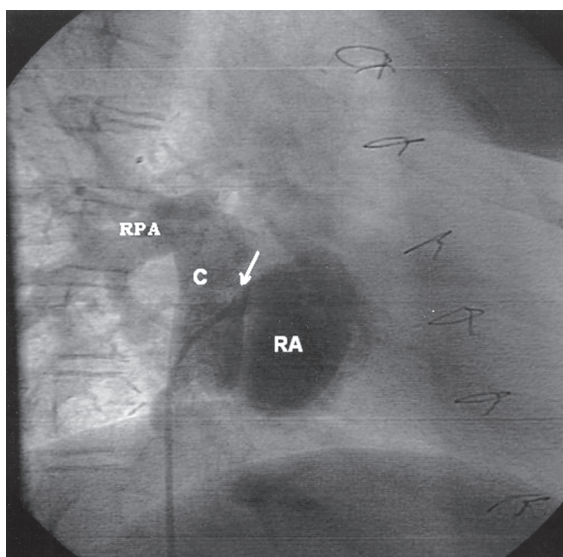


Figure 1. Angiography showing the fenestration (arrow) between Fontan conduit (C) and right atrium (RA). RPA: Right pulmonary artery

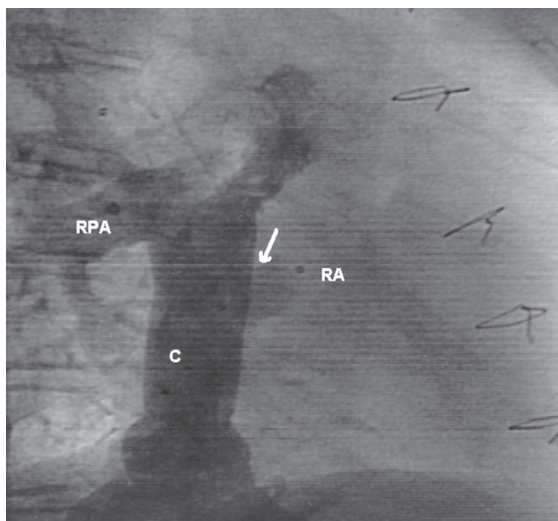


Figure 2. Temporary balloon (arrow) occlusion of the fenestration. C: Conduit, RA: Right atrium, RPA: Right pulmonary artery

and stabilized, the counterpart was opened on the Fontan conduit side. After disappearance of the shunt on a second angiogram, absence of any hemodynamic deterioration and confirmation of the position of the occluder by echocardiography, the device was released from the delivery wire (Fig. 3). The peripheral oxygen saturation during oxygen administration of 5 L/min with mask immediately increased from 90% to 99%. The device did not obstruct blood flow in the conduit, and no shunt from the fenestration or intracardiac thrombus was observed in the follow-up echocardiography. The follow-up electrocardiogram did not change. We have continued the treatment of acetylsalicylic acid in a dose of 3 mg/kg once daily and warfarin 5 mg once daily, which she had taken over the previous five years.

During follow-up, echocardiography showed that the position of the device was suitable, and there was no thrombus or any flow abnormality in Fontan circulation. No arrhythmia, ascites, edema or protein-losing enteropathy was observed. The oxygen saturation on room air was 95%.

Discussion

Because of early postoperative risks of increased systemic venous pressure and low cardiac output, the Fontan circuit is frequently fenestrated. Although fenestration was initially applied

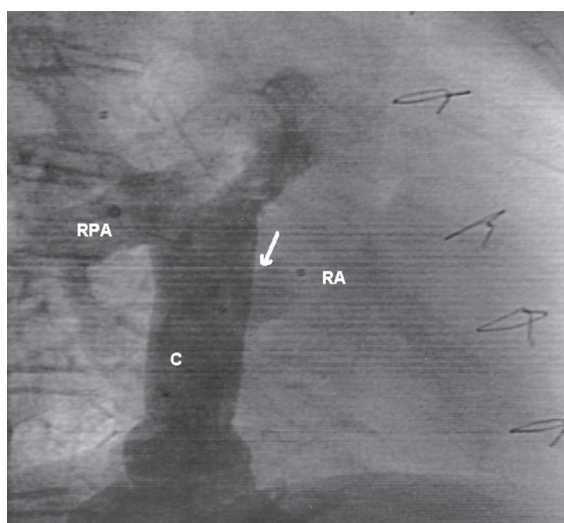


Figure 3. Permanent closure of the fenestration with atrial septal occluder (arrow) and disappearance of the right-to-left shunt. C: Conduit, RA: Right atrium, RPA: Right pulmonary artery

only in high-risk patients with ventricular dysfunction, distortion of pulmonary arteries or increased pulmonary vascular resistance, it has become a routine practice in many centers^{3,7}. On the other hand, some researchers have argued that the fenestration was not essential at all especially in patients with extracardiac conduit⁸. In our institution, construction of a fenestration has been preferred only in high-risk patients.

Although fenestration may be a life-saving intervention in some early postoperative conditions, it generally loses its importance after cardiovascular adaptation in the postoperative period. In fact, it causes progressive cyanosis and polycythemia and increased risk of paradoxical embolism that necessitate occlusion of the fenestration^{3,4}. Our patient's oxygen saturation was 75-80%. Likewise, clubbing, polycythemia and mild effort intolerance had also been present for five years. She had no thromboembolic event.

It is important to fully evaluate the patients in order to determine the suitability of the fenestration for closure and the presence of other possible causes of cyanosis, such as pulmonary arteriovenous fistulae, systemic-pulmonary venous collaterals, or connection of a left superior caval vein to the left atrium⁹. Improper decision for closure may result in heart failure, ascites, protein-losing enteropathy,

the need for Fontan takedown, or death associated with high central venous pressure and/or low cardiac output. Furthermore, early closure (≤ 6 months after Fontan operation) may predispose to cardiac decompensation and may be unnecessary in cases of spontaneous closure¹⁰.

Balloon occlusion test is a good tool for observing immediate consequences of the closure. Different criteria were proposed for closure. Some investigators recommend not closing the fenestration if cardiac output drops more than 30% with greater than 4 mmHg increase in mean systemic venous pressure^{6,11}. Similarly, other authors suggest that the fenestration should not be occluded in patients with a high venous pressure greater than 16 or 20 mmHg^{12,13}. Cardiac catheterization is also important for evaluation of any pathology that can be the cause of decreased cardiac output during temporary occlusion, such as distorted pulmonary arteries or aortopulmonary collaterals³. In our catheterization laboratory, we did not observe any other pathology that would cause cyanosis, or any significant increase in the mean systemic venous pressure or decrease in cardiac output after balloon occlusion. As the occlusion test was well tolerated, it was decided to close the fenestration permanently.

The other matter worthy of discussion is the type of device to be chosen. Different alternatives have been proposed, such as umbrella devices, septal or duct occluder devices, coils or stents^{3,13-16}. We preferred atrial septal occluder because the device consisted of two overlapping retention discs. The smaller right-sided disc could be properly placed in the semicircular wall of the conduit and would not deteriorate the flow in the Fontan circulation. In addition, the polyester patches inside the device promote thrombosis and prevent shunting. It can also be easily delivered and released through a long sheath. The diameter was measured as 6 mm and a 7.5 mm OcclutechTM atrial septal occluder was placed. The angiogram of the extracardiac conduit was repeated, and no shunt was observed through the fenestration. The peripheral saturation increased from 92% to 99% while breathing oxygen with mask. Position of the device and flow pattern in Fontan circulation were normal, cyanosis disappeared, and no adverse event was observed during the follow-up period.

In conclusion, prompt management of Fontan patients is quite important. The decision of fenestration, its closure and the type of device used seem to be the crucial parameters in the follow-up. Lastly, if closure is essential, atrial septal occluder device is a simple, safe and effective alternative.

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