Successful transcatheter closure of a Fontan fenestration with a bioabsorbable Biostar occluder

Fontan fenestrasyonunun biyoemilimi olan Biostar tıkayıcı cihazı ile başarıyla kapatılması

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Summary– We report the successful closure of an extracardiac Fontan fenestration with a bio-absorbable device, which may be refenestrated by a transcatheter route when needed, in a 10-year-old boy. The patient presented with cyanosis two years after an extracardiac Fontan operation. Echocardiography revealed a moderate shunt from the Fontan circulation into the systemic circulation with a mean pressure gradient of 3-4 mmHg. Treadmill testing revealed a significant decrease in oxygen saturation (down to the low 50's from a baseline level of 80-85%). Cardiac catheterization revealed normal pressure in the Fontan circuit. A temporary balloon occlusion test showed that the defect was suitable for permanent occlusion. The fenestration was then occluded by a bio-absorbable Biostar (NMT medical, Boston, USA) atrial septal occluder device. The oxygen saturation on room air increased up to 95% after closure.

Özet– Bu yazıda, ekstrakardiyak Fontan fenestrasyonu biyoemilimi olan ve gerektiğinde perkütan yolla tekrar fenestre edilebilecek bir cihazla başarı ile kapatılan 10 yaşında bir erkek çocuk sunuldu. Hasta Fontan operasyonundan 2 yıl sonra siyanoz yakınmasıyla başvurdu. Ekokardiyografik incelemede Fontan dolaşımından sistemik dolaşıma ortalama 3-4 mmHg basınç farkıyla orta düzeyde şant saptandı. Egzersiz testiyle hastanın oksijen doygunluğu %85 olan bazal seviyeden %50'ye kadar düştü. Kalp kateterizasyonunda Fontan dolaşımının basıncı normal bulundu. Balonla geçici tıkama testi açıklığın perkütan kapatma için uygun olduğunu gösterdi. Bunun üzerine fenestrasyon biyoemilimi olan Biostar (NMT medical, Boston, ABD) atriyal septum tıkayıcı cihazı ile kapatıldı. İşlem sonrası herhangi bir komplikasyon görülmedi. Hastanın oksijen doygunluğu oda havasında %95'e yükseldi.

Fenestration of the Fontan circulation, which results in a residual right-to-left shunt, has improved the operative survival rates among high-risk patients. However, progressive cyanosis, decreased exercise capacity, eritrocytosis, and increased risk of paradoxical embolism in the follow-up period are common late complications of fenestration. Because of these complications, most centers recommend occlusion of these fenestrations.[1] Use of bio-absorbable devices in these patients may provide the possibility to re-fenestrate in cases of rare complications such as protein-losing enteropathy and ascites, both of which may be seen after fenestration closure.[2]

We report the successful closure of an extracardiac Fontan fenestration with a bio-absorbable atrial septal occluder in a 10-year-old boy two years after his initial operation. To our knowledge, this is the first use of such a device for Fontan fenestration closure in Turkey.

CASE REPORT

A 10-year-old boy with a known diagnosis of unbalanced complete atrioventricular septal defect and morphological left ventricular hypoplasia with pulmonary stenosis presented to our outpatient clinic with complaints of cyanosis, shortness of breath, and

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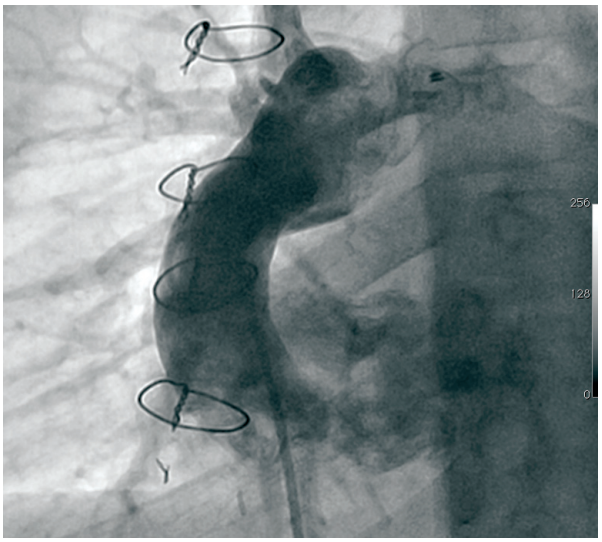

Figure 1. Extracardiac conduit angiography revealed a significant shunt across the fenestration into the systemic circulation.

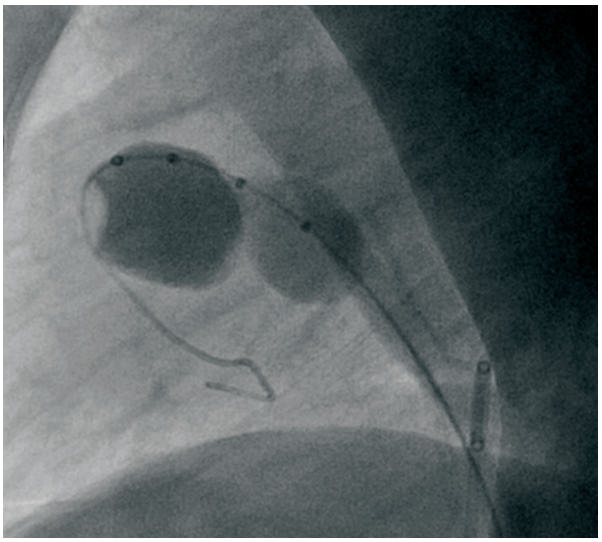

Figure 2. Temporary balloon occlusion test with a 12 mm balloon catheter passed over an exchange wire. The diameter of the fenestration was measured to be 6 mm.

fatigue. His past medical history was significant for multiple surgical interventions including a pulmonary artery banding procedure in infancy followed by a bilateral cava-pulmonary anastomosis at 3 years of age and a fenestrated Fontan procedure about two years prior to this presentation. The latter procedure was performed because he presented with borderline high pulmonary artery pressure and high pulmonary vascular resistance during his preoperative evaluation. His physical examination revealed clubbed fingers and mild cyanosis at rest. Cardiac oscultation revealed single first and second heart sounds and a soft 1-2/6 systolic murmur along the left sternal border. There was no organomegaly and his peripheric pulses were equally palpable. A complete blood count revealed moderate erytrocytosis. Echocardiographic evaluation showed normal ventricular function with mild atrioventricular valve regurgitation. There was no left ventricular outflow obstruction. Flow in the vena cava superior and pulmonary arteries was laminar with no evidence of obstruction. There was a moderate shunt from the extra cardiac conduit into the right atrium with a mean pressure gradient of 3-4 mmHg, which suggested low pressure in the Fontan circulation. Treadmill testing revealed a significant decrease in oxygen saturation (down to the low 50's from a baseline level of 80-85%). Because of this, a hemodynamic study to evaluate the possibility of transcatheter closure of the fenestration was planned, and the patient was taken to the catheterization laboratory.

A left and right heart catheterization was performed under deep sedation with ketamine and midazolam. Arterial and venous sheaths were placed into the right groin. Intravenous (IV) heparin (100 U/kg) for anticoagulation and cefazolin IV (50 mg/kg/dose) for antimicrobial prophylaxis was administered. The pressures and oxygen content in the systemic veins, pulmonary artery, common atrium, and aorta were measured. 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 the Fontan circuit was 9 mmHg and the mean right atrial pressure was 6 mmHg. His peripheral oxygen saturation was 85%. The fenestration was temporarily occluded with a 12 mm balloon catheter, which was passed over an exchange wire (Fig. 2), and the diameter of the fenestration was measured to be 6 mm. Hemodynamic measurements were repeated 15 minutes after balloon occlusion. Peripheral oxygen saturation increased from 85% to 95%. The mean systemic venous pressure increased from 6 mmHg to 7 mmHg, and the cardiac index decreased from 2.9 L/min/m^2 to 2.5 L/min/m^2. This hemodynamic condition was regarded as suitable for permanent occlusion. A long 11 Fr device delivery sheath was conducted in the right atrium from the fenestration. The

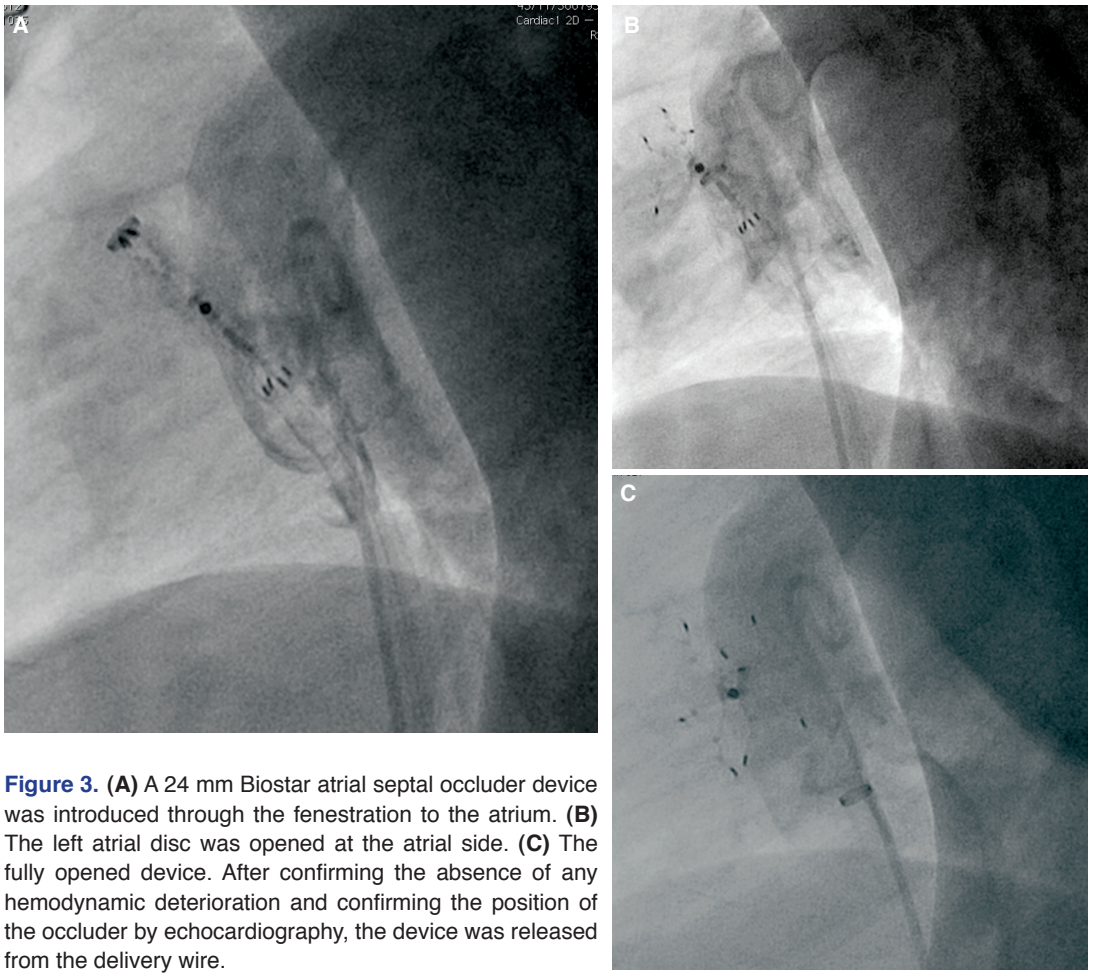

Figure 3. **(A)** A 24 mm Biostar atrial septal occluder device was introduced through the fenestration to the atrium. **(B)** The left atrial disc was opened at the atrial side. **(C)** The fully opened device. After confirming the absence of any hemodynamic deterioration and confirming the position of the occluder by echocardiography, the device was released from the delivery wire.

position of the sheath was confirmed by transthoracic echocardiography. A 24 mm Biostar (NMT medical, Boston, USA) atrial septal occluder device was introduced through the sheath and the left atrial disc was opened in the atrial side. Then, the device was slightly pulled back and stabilized, and then the right atrial disc was opened on the other side. A control angiogram revealed no residual shunt. After confirming the absence of any hemodynamic deterioration and the position of the occluder by echocardiography, the device was released from the delivery wire (Fig. 3A-C). Then, the peripheral oxygen saturation increased from 85% to 95%. The device did not obstruct blood flow in the conduit. No flow through the fenestration and no intracardiac thrombus were observed during follow-up echocardiography.

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

DISCUSSION

Surgical procedures based on the Fontan principle separate pulmonary and systemic blood flow in patients with single ventricular physiology. Risk factors including high pulmonary vascular resistance, poor systolic or diastolic ventricular function, hypoplastic or distorted pulmonary arteries, and systemic outflow obstruction may increase the prevalence of death and poor outcomes after Fontan-like procedures. Since fenestration of the Fontan pathway is effective for reducing perioperative morbidity and mortality rates in high-risk patients,[3] our center and others have elected to leave a surgical fenestration in high-risk Fontan patients. The patients who have persistent patent fenestration are mildly cyanotic but clinically well. The

"correct" management of this group is controversial. Several centers have demonstrated a significant incidence of thrombus formation within the Fontan pathway,[4] which places involved patients at an increased but undefined risk of systemic thromboembolic events as long as the residual right-to-left shunt is present. These children may be treated prophylactically with warfarin, but the benefit of such an approach is not known. Achieving a therapeutic international normalized ratio is a difficult task in young patients. The long-term bleeding risks of warfarin in young, active children are unknown.[5] It has also been reported that these children can increase the right-to-left shunt with exertion, which may cause them to become more cyanotic and thus exercise intolerant. Last of all, the chronic effects of persistent mild cyanosis on the developing child are poorly defined.

On the other hand, closure of the defect in the catheterization laboratory has been consistently shown to cause an acute reduction in cardiac index and tissue oxygen delivery, which has led some authors to speculate that there may be a hemodynamic benefit to maintaining fenestration patency.[6]

Based on his experience with transcatheter techniques to close various intracardiac defects,[10-12] Bridges et al. described a technique for the late postoperative closure of Fontan baffle fenestrations with the use of test occlusion and subsequent permanent closure with an intracardiac device.[7]

It is important to fully evaluate these patients in order to determine the suitability of the fenestration for closure. They must also be evaluated for 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 atrium. The 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 the patient to cardiac decompensation and may be unnecessary in cases of spontaneous closure.[6]

We have found that test occlusion and the subsequent transcatheter closure of Fontan fenestrations constitute a successful clinical strategy in the management of patients with single-ventricle physiology. Different criteria have been proposed for closure. Some investigators do not recommend closing the fenestration if cardiac output drops more than 30% with greater than a 4 mmHg increase in mean systemic venous pressure.[8] Some authors suggest that the fenestration should not be occluded in patients with a high venous pressure greater than 16 or 20 mmHg. Cardiac catheterization is also important for the evaluation of any pathology that can cause decreased cardiac output during temporary occlusion, such as distorted pulmonary arteries or aortopulmonary collaterals.[7] 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. If the occlusion test was well tolerated, we decided to close the fenestration permanently.

The other matter worthy of discussion is the type of device to be used. Different alternatives have been proposed, such as umbrella devices, septal or duct occluder devices, and coils or stents.[9] Although successful Fontan fenestration closure has been previously reported from our country,[10] this is the first time where it has been performed with a bio-absorbable device. We used the Biostar atrial septal occluder. Biostar is a bioabsorbable device consisting of a matrix of acellular collagen isolated from porcine intestine, which is mounted on the metal framework of a Starflex occluder in a double umbrella configuration and coated with heparin. After the device is positioned, it is rapidly incorporated into the septum and cellular remodeling ensues, with the collagen being absorbed and replaced by host tissue. This process can take approximately 24 months, after which only the metal struts remain. It is well known that the results of fenestration closure are improved oxygenation and the reduced use of anticongestive medications. However, in rare cases it may predispose the patient to cardiac decompensation, increased systemic venous pressure, and the development of ascites or protein losing enteropathy. We believe that bioabsorbable devices are most suitable for fenestration closure in these patients since it is possible to re-fenestrate them in the case of a rare situation such as protein-losing enteropathy and ascites after fenestration closure.

Conflict-of-interest issues regarding the authorship or article: None declared

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Key words: Child; cyanosis; Fontan procedure/adverse effects; heart catheterization/instrumentation; heart defects, congenital.

Anahtar sözcükler: Çocuk; siyanoz; Fontan işlemi/yan etki; kalp kateterizasyonu/enstrümantasyon; kalp defektleri, doğumsal.