Transcatheter closure of a fistula between the right pulmonary artery and left atrium using the Amplatzer septal occluder

Sağ pulmoner arter-sol atriyum fistülünün Amplatzer septal tıkayıcı cihaz ile transkateter yoldan kapatılması

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Summary – A congenital fistula between the right pulmonary artery (RPA) and left atrium (LA) is a rare condition that results in central cyanosis. An 11-year-old boy was admitted with exertional dyspnea and easy fatigability. He had severe cyanosis of the lips and limbs with clubbing of the fingers. Systemic oxygen saturation was 70%. There was no abnormal finding on electrocardiography, chest radiography, and echocardiography. Agitated saline injection showed early appearance of contrast bubbles in the LA. A pulmonary arteriovenous fistula was suspected and diagnostic cardiac catheterization was performed. Angiography demonstrated a large fistula between the proximal RPA and LA. The narrowest part of the fistula was 13.8 mm in balloon sizing. A 14-mm Amplatzer septal occluder was deployed at the narrowest site; however, the device migrated to the LA and then to the aortic arch. The device was removed and was successfully reimplanted to the fistula. After the procedure, arterial oxygen saturation increased from 70% to 96% and control angiography demonstrated complete occlusion of the fistula. The patient was symptom-free on follow-up evaluations at 6, 12, and 18 months, with a mean oxygen saturation of 96%. This case represents the first pediatric patient in whom a septal occluder was used.

Özet – Doğuştan sağ pulmoner arter (SPA) ile sol atriyum (SA) arasında fistül oluşumu, santral siyanozla karşımıza çıkan nadir bir durumdur. On bir yaşında bir erkek çocuk efor dispnesi ve çabuk yorulma yakınmalarıyla yatırıldı. Hastanın dudak ve ekstremitelerinde ciddi siyanoz ve parmaklarında çomaklaşma vardı. Sistemik oksijen satürasyonu %70 bulundu. Elektrokardiyografi, göğüs radyografisi ve ekokardiyografide bir anormalliğe rastlanmadı. Ajite salin enjeksiyonuyla SA'da erken kontrast baloncukları görüldü. Pulmoner arteriyovenöz fistülden şüphelenilerek, hastaya tanısal kardiyak kateterizasyon yapıldı. Sağ pulmoner arter anjiyografisinde proksimal SPA ile SA arasında büyük bir fistül görüldü. Balonla ölçümde fistülün en dar yeri 13.8 mm idi. Fistülün en dar yerine 14 mm'lik Amplatzer septal tıkayıcı yerleştirildi; ancak, cihaz SA'ya ve sonra da arkus aorta göç etti. Cihaz çıkarılarak ikinci girişimde fistüle başarıyla yerleştirildi. İşlem sonrasında arteryel oksijen satürasyonu %70'ten %96'ya yükseldi ve kontrol anjiyografisinde fistülün tamamen kapandığı görüldü. Hasta 6, 12 ve 18. aylardaki takiplerinde semptomsuzdu ve arteryel oksijen satürasyonu ortalama %96 idi. Olgumuz, fistül kapatılmasında septal tıkayıcının kullanıldığı ilk çocuk olgudur.

A direct communication between the right pulmonary artery and left atrium is a rare anomaly. Central cyanosis with clubbing of fingers and toes, exertional dyspnea, and decreased arterial oxygen saturation usually accompanies the lesion.[1,2] Surgical repair was the only treatment available till the introduction of interventional procedures.[1,3] We report on a case in which a congenital RPA-to-LA fistula was closed in the cardiac catheterization laboratory using the Amplatzer septal occluder.

Abbreviations:

LA *Left atrium*
RPA *Right pulmonary artery*

CASE REPORT

An 11-year-old boy was admitted with exertional dyspnea and easy fatigability. He had severe cyanosis.

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His mother stated that he had had a cyanotic color for many years. His height and weight were measured as 165 cm and 33 kg, respectively, and his blood pressure was 110/70 mmHg. He had severe cyanosis of the lips and limbs with clubbing of the fingers. No tachypnea, dyspnea, or murmurs were noted. The liver and spleen were not palpable. Laboratory findings were as follows: hemoglobin 20.5 g/dl, hematocrit 62.2%, and platelet count 256000/m^3. Systemic oxygen saturation was 70%.

Electrocardiography showed a normal sinus rhythm. There was no abnormality on the chest X-ray, nor on two-dimensional and color flow imaging. Agitated saline injection showed early appearance of contrast bubbles in the LA (within two beats). A pulmonary arteriovenous fistula was suspected and, after obtaining informed written consent of the parents, diagnostic cardiac catheterization was performed. Right heart catheterization was done under local and general anesthesia to measure hemodynamics and oxygen saturation. Selective right and left pulmonary arteriograms were obtained. Angiography of the RPA demonstrated a large fistula between the proximal RPA and LA (Fig. 1a). Mean RPA pressure was measured about 20 mmHg. Then, the catheter was sent through the right atrium to the right ventricle and to the main pulmonary artery in order to reach the fistula. The guide wire was left in the left ventricle and balloon sizing was performed. The narrowest part of the fistula was measured as 13.8 mm (Fig. 1b). The fistula was crossed from the right heart with an 8-F long sheath and a 14-mm Amplatzer septal occluder was deployed at the narrowest site. However, the device migrated to the LA and then to the aortic arch. Through an arterial route, the device was removed using a 12-F long sheath and was successfully reimplanted to the fistula (Fig. 2). After the procedure, arterial oxygen saturation rose from 70% to 96% and control angiography demonstrated complete occlusion of the fistula. The device caused no obstruction in the RPA and remained stable on release. The patient was symptom-free on follow-up evaluations at 6, 12, and 18 months with a mean oxygen saturation of 96%. Echocardiograms with agitated saline injection at 6 and 18 months showed no contrast bubbles on the left side of the heart.

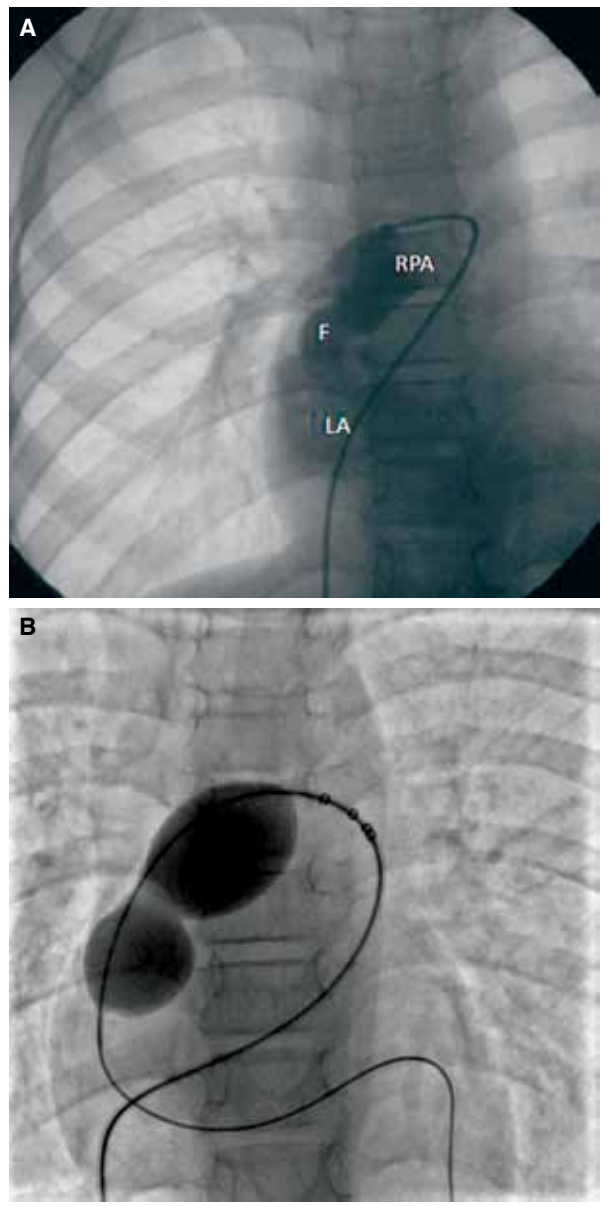

Figure 1. **(A)** Pulmonary arteriogram in the posteroanterior projection shows a communication between the right pulmonary artery (RPA) and the left atrium (LA) directly draining to the LA. **(B)** Balloon sizing of the fistula in the posteroanterior view. The waist is seen at the fistulous communication.

DISCUSSION

A direct communication between the RPA and LA is a rare congenital cardiac malformation.[1,2] It may present in the neonatal period as intense cyanosis requiring urgent intervention. If desaturation is less severe, presentation may be delayed till childhood or even adult life.[4] De Souza e Silva et al.[5] categorized communication between the RPA and LA into three types. In type I, the RPA branches normally, pulmonary venous return is normal, and an additional fistulous channel connects the RPA to the LA. In type II, the right lower branch of the pulmonary artery connects directly with the LA, forming an aneurysmal sac. In type III, all pulmonary veins drain into the abnormal

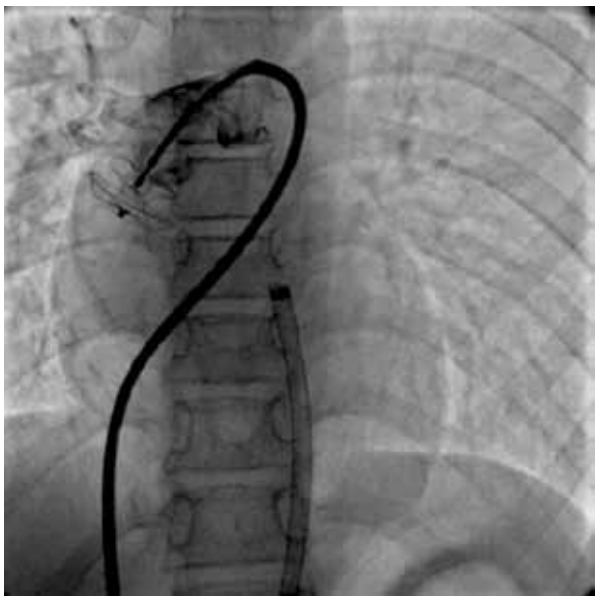

Figure 2. Pulmonary arteriogram in the posteroanterior projection shows complete occlusion of the fistula by the Amplatzer septal occluder.

channel that connects the RPA to the LA. According to this classification, our patient had type I anomaly.

Symptoms may vary depending on the size and location of the right-left shunt. The characteristic signs and symptoms of the anomaly are cyanosis of the lips, cheeks and extremities, digital clubbing, and dyspnea on exertion. The condition is difficult to diagnose because patients may present without a heart murmur.[1-4] Laboratory examination usually shows arterial hypoxemia and polycythemia. Electrocardiography may show left atrial dilatation and left ventricle hypertrophy with left axis deviation, but the fistula causes a huge volume load of the left heart only if there is a very large shunt. Cardiomegaly may be apparent on the chest X-ray in only extreme cases. However, if the hemodynamic burden is modest, both electrocardiography and chest X-ray may be completely normal.[1,6] Echocardiography and cardiac catheterization are necessary to demarcate the lesion clearly. Selective angiography of the involved pulmonary artery reveals the anomaly with rapid opacification of the LA and poor opacification of the lungs, corresponding to the involved pulmonary artery.[1]

Early repair should be performed to avoid complications, particularly systemic embolism, infective endarteritis, cerebral abscess, and rupture of aneurysmal communications.[1,4] Surgical ligation of the fistula (with or without cardiopulmonary bypass) had been the preferred treatment of this condition until recently.[1,3] Transcatheter coil occlusion of an RPA-to-LA fistula was successfully performed in a neonate.[7] The Amplatzer device offers an alternative occlusion for larger communications.[4] Francis et al.[8] occluded an RPA-LA fistula, the narrowest part of which was 8 mm by balloon sizing, using a 14-12 mm Amplatzer duct occluder in a 12-year-old girl. Uthaman et al.[9] occluded an RPA-LA fistula using the Amplatzer septal occluder in a 25-year-old male with cerebral abscess. We occluded the RPA-LA fistula, the narrowest part being 13.8 mm by balloon sizing, using a 14-mm Amplatzer septal occluder through the venous system. However, protrusion of the inferior part of the distal disc to the LA caused embolization to the LA and aorta after the first deployment. The same device was advanced second time to the fistula area and occlusion was achieved by fixing both discs in the fistula tract. As there was no atrial septal defect in our case, we implanted the device from the pulmonary arterial site. Fistulas involving the RPA and LA can also be closed by a muscular ventricular septal defect occluder or vascular plugs of proper diameter and size. During the diagnostic assessment before closure, angiographic evaluation of the length and diameter of the fistula and determination of the narrowest part by balloon sizing seem to be very important in device selection.

This case report describes the first pediatric patient in whom a septal occluder was used for the treatment of an RPA-to-LA fistula. Catheter closure of the RPA-to-LA fistula with the Amplatzer septal occluder is feasible and appears to be a safe and effective alternative to surgical treatment.

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Key words: Arteriovenous fistula/therapy; embolization, therapeutic; heart atria/abnormalities; heart catheterization; heart defects, congenital/therapy; pulmonary artery/abnormalities.

Anahtar sözcükler: Arteriyovenöz fistül/tedavi; embolizasyon, terapötik; kalp atriyumu/anormallik; kalp kateterizasyonu; kalp defekti, doğuştan/tedavi; pulmoner arter/anormallik.