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CASE REPORT

A different approach to treatment of failing Fontan: Transcatheter covered stent implantation

Fontan yetersizliği tedavisinde farklı bir yaklaşım: Transkateter kaplı stent yerleştirilmesi

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Summary– A 5-year-old male with a double outlet right ventricle with noncommitted ventricular septal defect and pulmonary stenosis underwent a bidirectional Glenn operation at 2 years and a Fontan operation with ligation of the pulmonary trunk at 5 years. He presented with pleural effusion 3 months after the Fontan operation. Physical examination revealed a grade 3/6 systolic murmur in the pulmonary area. Echocardiographic evaluation revealed an antegrade pulmonary flow (APF) of gradient 80 mmHg across the ventriculopulmonary communication. Cardiac catheterization and angiography demonstrated the presence of residual antegrade pulmonary flow and stenosis at the pulmonary artery bifurcation. Both pathologies were treated using a single covered stent. Relief of the pulmonary artery stenosis and total occlusion of the residual APF was demonstrated on a control angiogram.

Özet– İki yaşında iki yönlü Glenn ameliyatı geçirmiş ve beş yaşında pulmoner trunkus bağlanması ve kalp dışı Fontan ameliyatı geçirmiş çift çıkımlı sağ ventrikül, ventriküler septal defekt ve pulmoner stenozlu bir hasta sunduk. Hasta Fontan ameliyatından üç ay sonra plevra sıvısıyla başvurdu. Fizik muayenesinde pulmoner odakta 3/6 sistolik üfürüm duyuldu. Ekokardiyografik değerlendirimede pulmoner öne doğru akım ve burada 80 mmHg basınç farkı tespit edildi. Kalp kateterizasyonu ve anjiyografi çatallanma düzeyinde darlık ve pulmoner öne doğru akımı gösterdi. Her iki patolojiyi de transkateter yolla kaplı stent yerleştirerek giderdik. Kontrol anjiyogramında çatallanma düzeyinde darlığın giderildiği ve öne doğru akımın kaybolduğu izlendi.

Fontan surgery is the final stage operation of a series of palliative operations. It is performed on patients with complex congenital heart disease who are not suitable for biventricular repair. The procedure diverts systemic venous blood returning to the right atrium via the inferior vena cava to the pulmonary arteries. Since blood is transmitted passively to the lungs in Fontan circulation, pulmonary artery size, preoperative pulmonary arterial pressure, pulmonary vascular resistance, and the presence of branched pulmonary artery stenosis are of great importance, and determine the outcome of the procedure in these patients. Even a minor increase in pulmonary arterial pressure or an alteration in pulmonary blood flow may affect the hemodynamics of circulation and result in complications such as pleural effusion, ascites, or protein losing enteropathy.[1,2]

Abbreviation:

APF *Antegrade pulmonary flow*

This report describes successful transcatheter treatment using a single covered stent of a pleural effusion from pulmonary artery stenosis and residual pulmonary antegrade flow in a 5-year-old male with Fontan circulation.

CASE REPORT

A 5-year-old male presented with pleural effusion 3 months after an extracardiac conduit Fontan operation. His initial diagnosis was double outlet right ventricle with noncommited ventricular septal defect and pulmonary stenosis. The patient had undergone a bidirectional Glenn operation at 2 years and an extracardiac conduit Fontan operation and ligation of the pulmonary trunk 3 months prior to presentation. On

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physical examination, a systolic murmur was heard in the pulmonary area. Doppler echocardiography revealed an antegrade pulmonary flow (APF) with a gradient of 80 mmHg across the ventriculopulmonary communication. The patient was subjected to cardiac catheterization and angiography with the aim of APF occlusion. A 6 Fr sheath was introduced into the right femoral vein and right jugular vein. A right ventricular angiogram revealed the presence of residual APF (Figure 1a). The initial hemodynamic study demonstrated identical pressure levels in the systemic veins and pulmonary arteries. However, a subsequent hemodynamic study performed after placement of a 6 × 8 mm duct occluder across the ventriculopulmonary communication revealed a pressure gradient of 7 mmHg between the distal left pulmonary artery and right pulmonary artery, and the superior vena cava. Distal left pulmonary arterial pressure was found to be 11 mmHg, while the superior vena cava, inferior vena cava, and right pulmonary arterial pressures were 18 mmHg. On a simultaneous right ventricle and superior vena cava angiogram, stenosis at the pulmonary artery bifurcation was observed, and the device was found to be causing no obstruction to the pulmo-

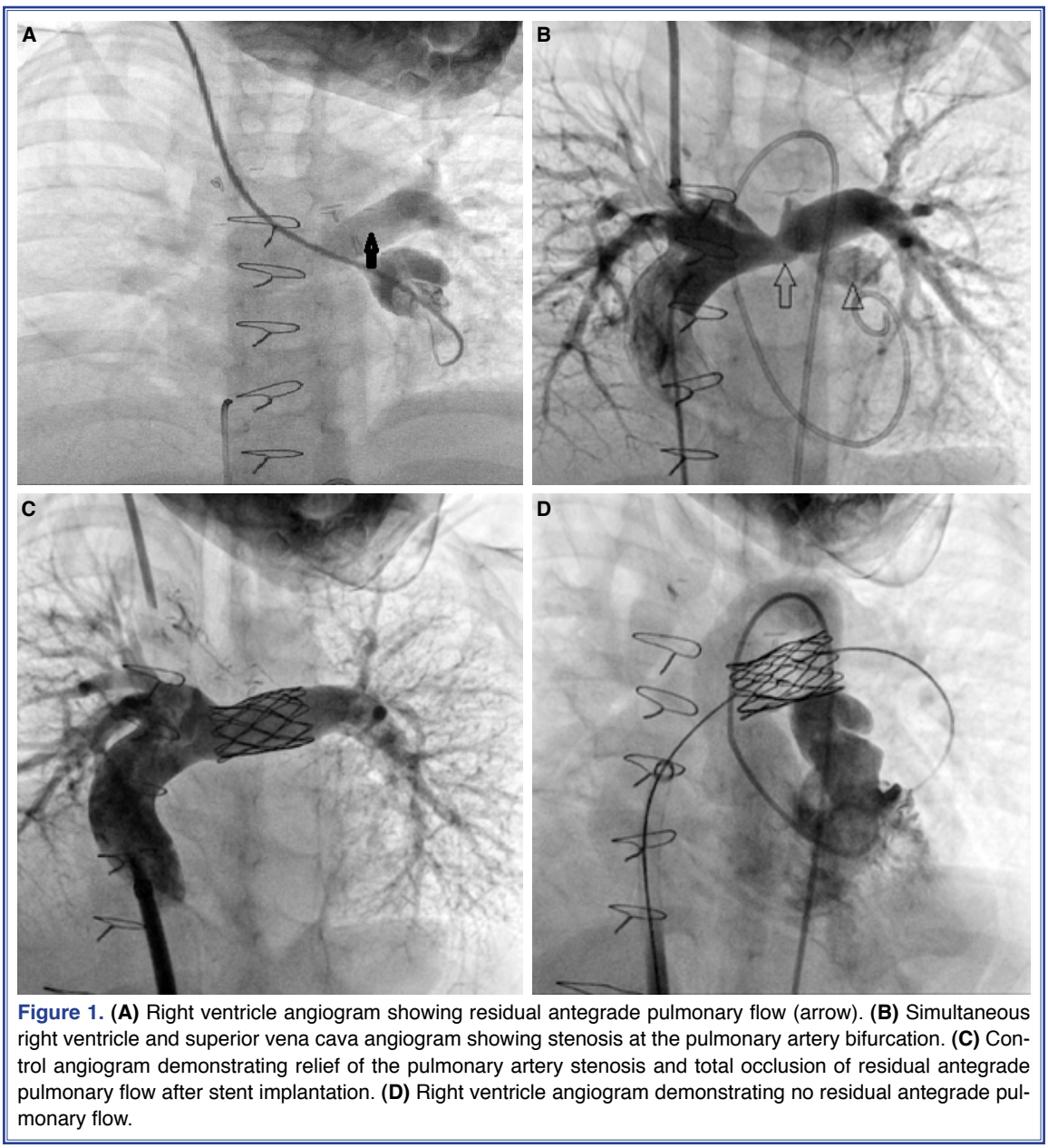

Figure 1. **(A)** Right ventricle angiogram showing residual antegrade pulmonary flow (arrow). **(B)** Simultaneous right ventricle and superior vena cava angiogram showing stenosis at the pulmonary artery bifurcation. **(C)** Control angiogram demonstrating relief of the pulmonary artery stenosis and total occlusion of residual antegrade pulmonary flow after stent implantation. **(D)** Right ventricle angiogram demonstrating no residual antegrade pulmonary flow.

nary arteries (Figure 1b). The duct occluder was then removed, and a decision made to treat both pathologies with implantation of a covered stent. The smallest diameters of stenosis at the pulmonary artery bifurcation and distal left pulmonary artery were 6 mm and 14 mm respectively. A 28-mm covered Cheatham platinum stent (Numed, Hopkinton, New York, USA), mounted on a 15 mm × 3.5 cm balloon-in balloon catheter (Numed, Hopkinton, New York, USA) was advanced via the femoral vein and deployed at the pulmonary artery bifurcation. The site of the implanted stent was at a safe distance from the Glenn anastomosis. A control angiogram demonstrated relief of the pulmonary artery stenosis and total occlusion of the residual APF (Figures 1c, d). Following implantation, final pressure in the left pulmonary artery, right pulmonary artery and superior vena cava was 14 mmHg. Pulmonary effusion improved dramatically in a few days and had not recurred at 1-year follow-up after stent implantation.

DISCUSSION

Fontan circulation diverts systemic venous return directly to the pulmonary arteries. The presence of aortapulmonary collaterals, branch pulmonary artery stenosis, atrioventricular valve regurgitation, elevated ventricular end diastolic pressures, APF, and stenosis of venous–pulmonary artery anastomosis can lead to a dysfunction in the Fontan circulation and thus to persistent pleural effusion, ascites, or protein-losing enteropathy.[3]

Ligation of the pulmonary trunk in the total cavopulmonary connection has previously been described.[4] Residual pulmonary flow after this procedure causes a rise in pulmonary arterial pressure and hence dysfunction in the Fontan circulation. After simple ligation of the pulmonary trunk, recanalization of APF can occur at any time postoperatively, as occurred in the present patient.[5] As simple ligation of the pulmonary artery is associated with recanalization, the main pulmonary artery should be divided and sutured.[5,6] Surgery has historically been the treatment of choice in elimination of APF. However, in recent years, transcatheter elimination of ventriculopulmonary communication with a duct occluder has been reported in patients with bidirectional cavopulmonary anastomosis, and in those who developed ascites or pleural effusions after a Fontan procedure.[1,2] Torres et al. reported left pulmonary stenosis in a patient after a duct occluder device implantation for occlusion of APF. The stenosis was treated with a stent implanted in the left pulmonary artery in the same procedure.[7] In the present case, no pressure gradient was obtained on the hemodynamic study prior to the occlusion of APF with a duct occluder. Following occlusion of the APF, a pressure gradient of 7 mmHg was found due to a stenosis at the pulmonary artery bifurcation demonstrated on a simultaneous right ventricle and superior vena cava angiogram. This gradient persisted after removal of the device, which led us to consider the initial hemodynamic evaluation as inaccurate.

Pulmonary artery stenosis is frequently observed in patients with univentricular circulation. During staged palliative operations, reconstruction of pulmonary arteries should be performed. Stenosis in the pulmonary arteries or systemic veins, if noticed, should be corrected preoperatively.[3] Stenosis may also develop postoperatively mainly at the anastomosis area. In Fontan circuits, pulmonary artery stenosis causes an increase in blood pressure of the contralateral lung and can result in persistent pleural effusion and ascites. A relationship between the development of ascites or pleural effusions and pulmonary arterial pressure greater than 15 mmHg has been reported.[3] Stent implantation into the stenotic segment in Fontan circuits is reportedly effective.

Blind pouch formation at the ligated segment of the pulmonary artery is a serious risk factor for thromboembolic events. Although thrombosis is a known and serious complication following a Fontan circuit, there is no consensus on the prophylaxis protocol. Presentation of the thromboembolic event may be silent or alarming, as in an embolic stroke.[7] What is known is that the rate of thromboembolism is higher in a group that takes no prophylaxis compared to a group treated with acetylsalicylic acid or warfarin.[8] As occlusion of the APF resulted in the formation of a pulmonary stump in our patient, we began coumadin prophylaxis. Although thrombus formation in Fontan is multifactorial, attention must be paid in order to avoid creating areas, such as a pulmonary stump, which are prone to thrombus formation.[9]

If Fontan circulation fails due to APF and pulmonary artery stenosis, both pathologies can be treated with a single intervention by implanting a covered stent. Transcatheter intervention is considered safe

and effective. To the best of the authors' knowledge, this is the first reported case of residual APF and pulmonary artery stenosis after a Fontan operation treated with a single covered stent.

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