

## CASE REPORT

## Percutaneous treatment of high-output right heart failure with pulmonary hypertension due to a large iliac arteriovenous fistula using a patent ductus arteriosus occluder

Pulmoner hipertansiyon ile birlikte yüksek debili sağ kalp yetersizliğine sebep olan bir olguda, geniş iliyak arteriyovenöz fistülün patent duktus arteriyosus kapatma cihazı kullanılarak perkütan yoldan kapatılması

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**Summary**– Presently described is the case of a 41-year-old male patient with signs of high-output right heart failure (HF) due to a possibly iatrogenic arteriovenous fistula (AVF) between the left common iliac artery and the left common iliac vein. Invasively evaluated hemodynamic measurements were consistent with severe shunting, which resulted in precapillary pulmonary hypertension (PH). The AVF was successfully closed with a non-dedicated, 16-mm Amplatzer patent ductus arteriosus occluder (St. Jude Medical, Inc., St. Paul, MN, USA). Although his signs and symptoms were documented to be dramatically improved after closure, because mild PH persisted, adjuvant pulmonary arterial hypertension-targeted treatment with bosentan was initiated to prevent late pulmonary vascular disease.

**Özet**– Bu yazıda, sol ana iliyak arter ile sol ana iliyak ven arasında, muhtemelen iyatrojenik sebeple oluşmuş bir arteriyovenöz fistüle (AVF) sahip olan ve yüksek debili sağ kalp yetersizliği bulguları ile kendini gösteren 41 yaşındaki bir erkek hasta sunuldu. Hastanın invaziv kalp kateterizasyonu ile ölçülen hemodinamik verileri, ciddi şant akımının sebep olduğu prekapiller pulmoner hipertansiyon (PH) ile uyumlu saptandı ve sonrasında bu AVF 16 mm Amplatzer™ patent duktus arteriyozus cihazı ile perkütan yolla başarılı bir şekilde kapatıldı. Kapatma işleminden sonra hastanın şikayetlerinin ve klinik bulgularının dramatik bir şekilde düzelmesine rağmen, hafif PH varlığının devam etmesi sebebiyle, uzun dönem pulmoner vasküler hastalık gelişimini önlemesi adına bosentan tedavisi başlandı.

Described is the case of a young male patient with high-output heart failure (HF) and pulmonary hypertension (PH) due to a large peripheral arteriovenous fistula (AVF) that was percutaneously treated with the implant of a non-dedicated device.

### CASE REPORT

A 41-year-old male was referred to our tertiary cardiovascular center for advanced evaluation of HF. He had a history of progressive dyspnea, orthopnea, bilateral leg swelling, and palpitations for 6 months. A coronary angiography procedure had been performed 12 months earlier via the left transfemoral

access. The initial vital signs recorded included a heart rate of 130 beats per minute, blood pressure of 135/78 mm Hg, respiratory rate of 36 breaths per minute, and oxygen saturation of 88% at room air. Bilateral, severe, pretibial pitting edema and inspiratory rales auscultable at the base of the lungs were the main findings of his physical examination.

#### Abbreviations:

AVF	Arteriovenous fistula
CT	Computed tomography
HF	Heart failure
IVC	Inferior vena cava
LCIA	Left common iliac artery
LCIV	Left common iliac vein
PA	Pulmonary arterial
PH	Pulmonary hypertension
PVR	Pulmonary vascular resistance
RV	Right ventricle
SVR	Systemic vascular resistance
VA	Veno-arterial

Received: November 06, 2017 Accepted: February 09, 2018

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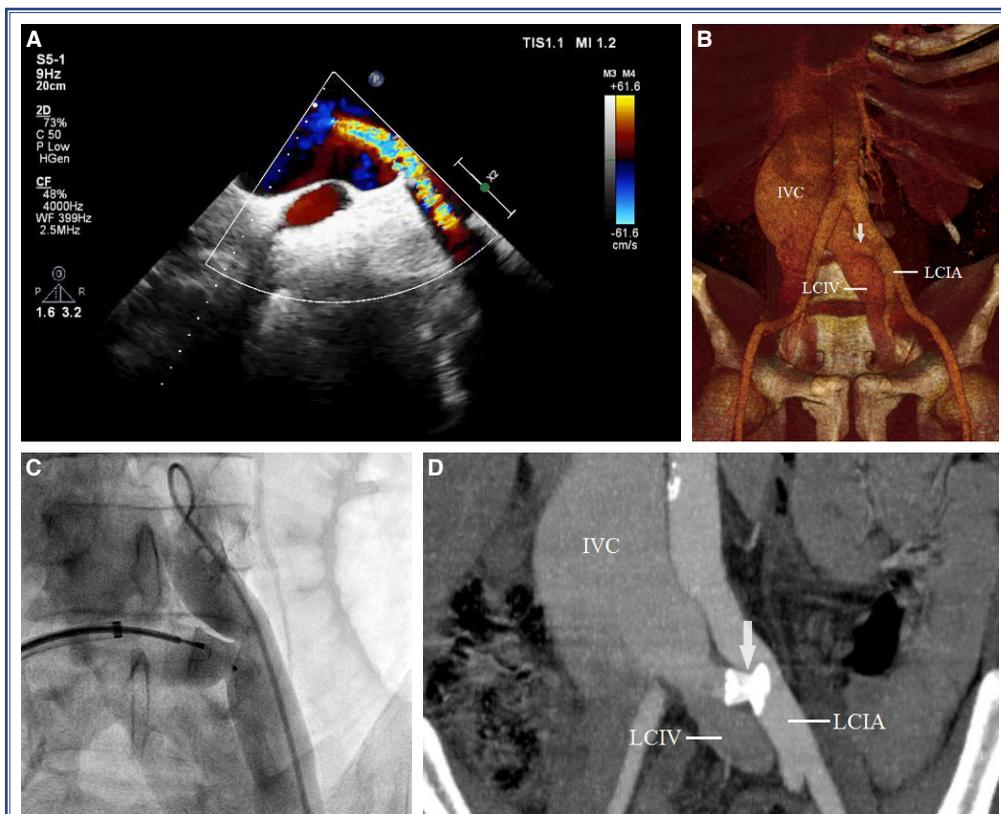
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Additionally, a palpable, continuous thrill at the right lower quadrant of the abdomen was confirmed by auscultation as a grade-5, systolodiastolic murmur over the left iliac artery. An electrocardiogram revealed nothing unusual other than sinus tachycardia. A chest X-ray revealed cardiomegaly, pulmonary arterial (PA) enlargement, and pulmonary congestion consistent with high-output HF. Echocardiography also revealed enlargement in the PAs, right atrium, right ventricle (RV); a D-shaped septum at diastole; severe tricuspid regurgitation; and moderate mitral regurgitation with a normal left ventricle ejection fraction. The estimated PA systolic pressure from the tricuspid regurgitant jet was 55 mm Hg. In addition, a continuous shunt flow from the left common iliac artery (LCIA) to the left common iliac vein (LCIV) was detected on a Doppler examination (Fig. 1a). Computed tomography (CT) angiography disclosed an enlarged infe-

rior vena cava (IVC), and an AVF between the LCIA and the LCIV (Fig. 1b). Right and left heart catheterization and coronary angiography was performed in order to both exclude coronary artery disease and to perform further evaluation of the PH. Systolic, diastolic, mean, and wedge PA pressure was measured as 63, 33, 46, and 14 mm Hg, respectively. The Qp/Qs ratio of 2.5 was consistent with hemodynamically significant systemic to pulmonary shunting via a large AVF. The pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR) were 2.7 and 15 Wood U, respectively. The diagnosis was high-output HF with PH due to a peripheral AVF, and it was decided that the shunt would be closed percutaneously. Using the left femoral arterial access, a 6-F right Judkins catheter was engaged at the site of the AVF, and a 0.035-mm hydrophilic guidewire was crossed over the defect and advanced to the IVC. A snare was ad-

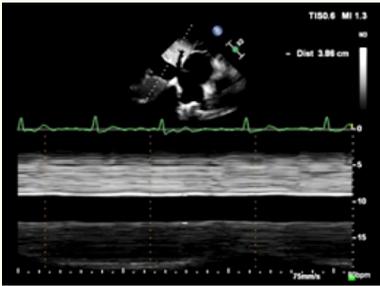
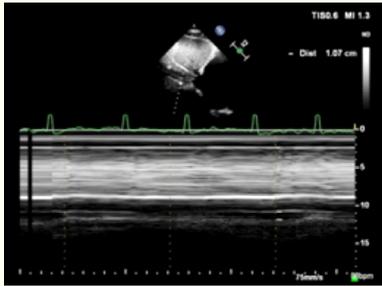
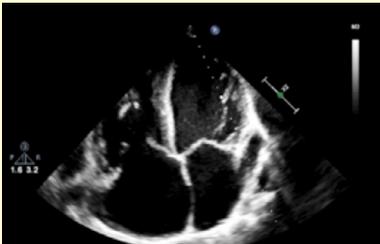
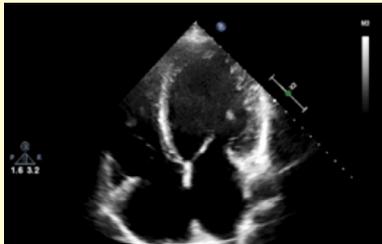


**Figure 1.** (A) A high-grade, continuous shunt flow was seen from the left common iliac artery to the left common iliac vein in a Doppler examination. (B) Three-dimensional reconstructed computed tomography demonstrated an enlarged inferior vena cava and a large arteriovenous fistula (arrow). IVC: Inferior vena cava; LCIA: Left common iliac artery; LCIV: Left common iliac vein. (C) Placement of a 16-mm Amplatzer patent ductus arteriosus occluder device (St. Jude Medical, Inc., St. Paul, MN, USA) into the left common iliac artery with a delivery sheath. (D) Postprocedural control computed tomography confirmed the stability of device (arrow). IVC: Inferior vena cava; LCIA: Left common iliac artery; LCIV: Left common iliac vein.

vanced from the right femoral vein to the IVC, and following the capture and retrieval of the guidewire, a veno-arterial (VA) loop was created between the right femoral vein and the left femoral artery systems. The delivery sheath of the occluder was pushed forward through this VA loop to the LCIA. Initial attempts to use a 16-mm Amplatzer patent ductus arteriosus (PDA) occluder device (St. Jude Medical, Inc., St.

Paul, MN, USA) to close the AVF failed to close the shunt. The use of an Amplatzer atrial septal defect (ASD) occluder (St. Jude Medical, Inc., St. Paul, MN, USA) also failed as a result of malorientation of the device within the LCIA. Finally, shunt closure was achieved with the original 16-mm Amplatzer PDA occluder device in another effort (Fig. 1c). The signs and symptoms of AVF and high-output HF were markedly

**Table 1. A comparison of the patient’s initial and postprocedural clinical, echocardiographic, hemodynamic, and neurohormonal parameters**

Measure	Initial	Postprocedure, 90 <sup>th</sup> Day
ECHO LVEDD (cm) / LVEDV (mL)	6.9 / 247	5.8 / 167
LVEF (%)	55	55
Mitral E / A / e' / E:e' (cm/s)	80 / 60 / 9 / 8.9	90 / 60 / 9 / 10
Pulmonary vein inflow	Systolic dominance	Systolic dominance
ECHO MPA diameter	4.56	3.8
ECHO PAPs (mm Hg)	56	30
TAPSE (cm)	2.0	2.5
St (cm/s)	13.2	14
IVC diameter (cm)	3.86	1.1
		
6 MWD (meters)	250	480
BNP (pg/mL)	3203	453.4
CATH PAP (mm Hg)	63 / 33 / 46	47 / 20 / 30
PVR (Wood U)	3.8	3.1
Mean RA pressure (mm Hg)	20	6
CATH Qp/Qs ratio	2.5	1.0
Apical 4-chamber ECHO view		

6MWD: Six minute walking distance; BNP: Brain natriuretic peptide; CATH: Catheterization; ECHO: Echocardiography; IVC: Inferior vena cava; LVEDD: Left ventricle end-diastolic diameter; LVEF: Left ventricle ejection fraction; LVEDV: Left ventricle end-diastolic volume, MPA: Main pulmonary artery; PAP: Pulmonary artery pressure; PAPs: Systolic pulmonary artery pressure; PVR: Pulmonary vascular resistance; RA: Right atrium; St: Tissue velocity of tricuspid annular longitudinal systolic motion; TAPSE: Tricuspid annular plane systolic excursion.

relieved immediately after device implantation. A 90<sup>th</sup>-day control CT and conventional angiography confirmed the stability of device without any significant leakage or luminal narrowing in the LCIA (Fig. 1d). The invasively evaluated mean PA pressure was determined to be 30 mm Hg, the PVR was 3.1 Wood U, and the Qp/Qs ratio was almost 1.0. Due to the borderline PVR measures, pulmonary arterial hypertension-targeted treatment with bosentan 62.5 mg twice daily followed by 125 mg twice daily was initiated to be accompanied by periodic follow-up. His initial clinical, echocardiographic, hemodynamic, and neurohormonal measurements showed marked and progressive improvement after the procedure (Table 1).

## DISCUSSION

Systemic AVF is one of the most common etiologies of high-output HF. Other etiologies include chronic anemia, sepsis, hypercapnia, and hyperthyroidism.<sup>[1]</sup> An AVF typically occurs secondary to incidental trauma, or iatrogenically during a catheterization procedure for renal dialysis or vascular intervention.<sup>[1-3]</sup> Congenital AVF may develop due to fetal-maternal viral infection or some still unclear genetic factors, and are most often seen in hepatic vascular, cerebral vascular, and pulmonary vascular areas.<sup>[3]</sup> Pulmonary AVF may also be part of the Osler-Weber-Rendu syndrome (hereditary hemorrhagic telangiectasia), and abdominal AVF may be detected in patients with tuberous sclerosis or Down syndrome.<sup>[4,5]</sup>

Clinically important high cardiac-output HF may develop in patients with large, peripheral, systemic AVF due to hemodynamically significant arteriovenous (AV) shunting.<sup>[6]</sup> Reduced SVR due to AV shunting and peripheral vasodilatation contribute to this pathophysiological change.<sup>[1]</sup> However, the pathophysiological mechanisms of the development of PH in this setting remain unclear. It is hypothesized that increased PA blood flow leads to shear stress-induced endothelial dysfunction, triggering the obstructive remodeling and vasospasm mediated by endothelin-1 in the presence of impaired nitric oxide-mediated PA relaxation.<sup>[7]</sup> End-stage renal disease provides a model for the long-term hemodynamic and cardiovascular effects of AVF.<sup>[2]</sup> Shunting reduces arterial blood pressure, body weight, and plasma volume, initially, whereas there is no statistically significant change in left ventricle end-diastolic diameter, mass index, or

ejection fraction after the creation of an AVF. In contrast, because of increased venous return, significant dilatation was observed in the left atrium and RV with reduced RV systolic function. Both RV dysfunction and wasted systemic perfusion through AVF increase myocardial demand, which initiates HF.<sup>[2]</sup>

AVF may occur in various vascular locations, including abdominal, pulmonary, intracranial, and coronary sites, as well as the upper or lower extremities.<sup>[1,3,4,5,7,9]</sup> An integrated approach that includes a physical examination disclosing the continuous shunt flow; Doppler, CT, and sometimes magnetic resonance angiography; and an invasive assessment of anatomopathological and hemodynamic characteristics of AVF, the Qp/Qs, PA pressure, and PVR are essential to the decision-making in patients with complicated AVF.<sup>[1,6,8]</sup> A classic triad, i.e., high-output HF with a precipitous onset, a pulsatile abdominal mass accompanied by a thrill and a bruit, and unilateral lower extremity ischemia or venous engorgement, is seen in only 50% to 80% of AVF patients.<sup>[5]</sup>

Although there is a lack of randomized controlled trials, a variety of treatment options, including open surgical repair or interrupting the connection between the arterial and the venous side via endovascular graft stent insertion, have been used for the treatment of AVF.<sup>[1,7]</sup> Surgical and endovascular treatments are preferable when there is the co-existence of arterial aneurysmatic dilatation. However, the long-term results of endovascular repair are unclear, and the risk of a type II endoleak with expansion of the aneurysm cannot be totally eliminated.<sup>[5]</sup> Moreover, the currently available transcatheter closure techniques, such as vascular plagues, occluder devices, covered stents, and coils for embolization may be alternative solutions that eliminate the risks of general anesthesia and perioperative problems due to large incisions, trauma, or groin infection.<sup>[3,4,9,10]</sup>

Bosentan is an orally active, dual endothelin receptor type A and B antagonist that is widely used in various types of PH, especially in group 1 PH. Its efficacy on 6-minute walking distance, time to clinical worsening episodes, functional class, hemodynamics, echocardiographic and Doppler variables, and neurohormonal parameters has been demonstrated in large, randomized control trials and meta-analyses.<sup>[11-16]</sup> According to this evidence, we decided to start temporary bosentan treatment for a couple of months due to

persistent, mild PH and borderline PVR observed in a control right-heart catheterization.

In our typical case of high-output HF and PH due to a large AVF between the LCIA and the LCIV, possibly secondary to iatrogenic injury during previous catheter procedures, we were able to close the AVF percutaneously with the placement of a non-dedicated device, a 16-mm Amplatzer PDA occluder, after several attempts. Although the signs and symptoms of the patient improved dramatically after closure, mild PH persisted and the initiation of pulmonary arterial hypertension-targeted treatment with bosentan to prevent late pulmonary vascular disease was also considered to be necessary.

**Peer-review:** Externally peer-reviewed.

**Conflict-of-interest:** None.

**Informed Consent:** Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

**Authorship contributions:** Concept: C.K.; Design: O.Y.A.; Supervision: F.Y.; Materials: A.H.; Data collection: A.H.; Literature search: S.T.; Writing: C.K., O.Y.A.

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**Keywords:** Arteriovenous fistula; duct occluder; high output heart failure; percutaneous peripheral intervention; pulmonary hypertension.

**Anahtar sözcükler:** Arteriovenöz fistül; ductus kapatma cihazı; yüksek debili kalp yetersizliği; perkütan periferik girişim; pulmoner hipertansiyon.