Atrial flow regulator as a bridge to lung transplant in a young patient with drug-resistant idiopathic pulmonary arterial hypertension

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A 20-year-old man presented to our department with severe drug-resistant idiopathic pulmonary arterial hypertension (IPAH) diagnosed 2 years before his admission. Initially, IPAH-specific triple-combination therapy with intravenous epoprostenol was introduced. After the diagnosis had been established, the patient's condition started to deteriorate progressively and several hospitalizations were reported due to acute worsening. Since February 2019, his mobility has been significantly limited (World Health Organization [WHO] functional class IV; 6-minute walk distance, 197 m) and severe right heart failure (RHF) and fluid retention have developed. The dose of epoprostenol reached 91.5 ng/kg/min. The patient was receiving an intravenous infusion of levosimendan 3 times every few months and was on the waiting list for lung transplant. Due to the patient's progressive deterioration and lack of alternative medical therapy, the PAH specialist team decided to implant the atrial flow regulator (AFR) to decompress the right ventricle and improve its function. The local ethics committee approved the procedure. The patient was given detailed information on the risks and benefits of the surgery and provided written informed consent.

At the first stage of the procedure, full right heart catheterization was performed with stepwise oximetry and hemodynamic measurements (using the Fick method) according to current guidelines.[1] The mean right atrial pressure (mRAP) was 16 mm Hg, mean pulmonary artery pressure (mPAP) 66 mm Hg, pulmonary artery wedge pressure (PAWP) 10 mm Hg, pulmonary vascular resistance (PVR) 13.9 Wood units, pulmonary flow (Qp) 4.1 l/min, pulmonary to systemic flow ratio (Qp:Qs) 1, and systemic blood pressure 90/60 mm Hg. Oximetry measurements without oxygen supplementation demonstrated mixed venous oxygen saturation (SvO_2) in the pulmonary artery of 42.3% and arterial blood saturation in the aorta of 84.7%. Transseptal puncture was performed under 3-dimensional transesophageal echocardiographic guidance and was followed by atrial septostomy with the EverCross balloon (Medtronic, Minneapolis, Minnesota, United States) of 8 mm × 4 cm in size (FIGURE 1A). Immediately after septostomy, oxygen saturation dropped to 72%, so the decision was made to implant an AFR (Occlutech, Helsingborg, Sweden) with the fenestration of 6 mm in diameter. The device was introduced through the femoral vein using a 10F delivery sheath and implanted under angiographic and transesophageal echocardiographic guidance (FIGURE 1B–1F). The procedure was uneventful. Within several hours, saturation improved up to 90% with oxygen supplementation (3 l/min).

Eight weeks after the AFR insertion, the patient's clinical condition improved (WHO functional class III; 6-minute walk distance, 210 m). Follow-up right heart catheterization (performed with the Fick method) demonstrated mRAP of 14 mm Hg, mPAP of 55 mm Hg, PAWP of 8 mm Hg, PVR of 5.7 Wood units, Qp of 8.2 l/min, Qp:Qs ratio of 0.85, SvO_2 of 65.6%, and arterial oxygen saturation of 84.2%.

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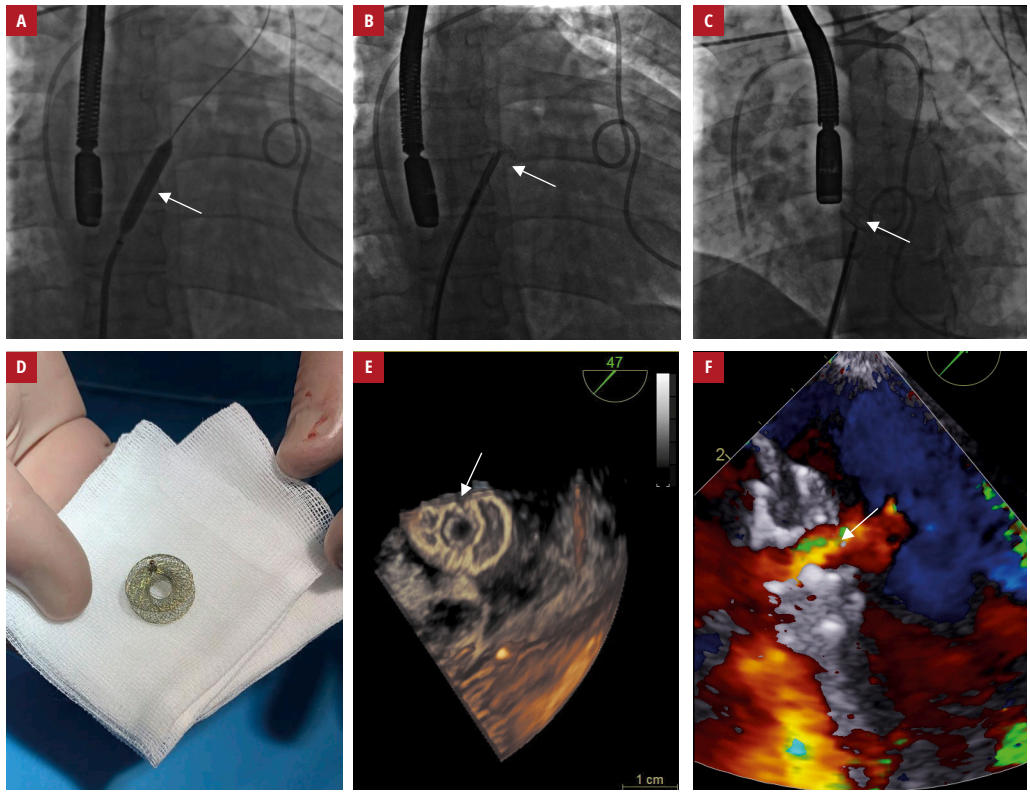

FIGURE 1 **A** – balloon atrial septostomy (arrow) seen on angiography; **B** – intraprocedural angiography showing an atrial flow regulator (AFR; arrow) just before the final locking; **C** – intraprocedural angiography showing an AFR (arrow) after release; **D** – Occlutech AFR, a self-expanding nitinol wire mesh device with fenestration (in this case, of 6 mm in diameter); **E** – final transesophageal echocardiography: 3-dimensional left atrial anterior view of the Occlutech AFR (arrow); **F** – final transesophageal echocardiography demonstrating the shunt achieved using the Occlutech AFR (arrow)

Nevertheless, analyses were performed a few days after an intravenous infusion of levosimendan and iron and oxygen supplementation. Twelve weeks after the AFR implantation, the patient underwent successful lung transplant.

Idiopathic pulmonary arterial hypertension is a severe, progressive disease leading to RHF and, ultimately, death.[2] The presented case shows that establishing an interatrial right-to-left shunt in patients with end-stage IPAH may relieve the signs of RHF and improve cardiac output.[3-4] Increasing cardiac output may ameliorate effective oxygen delivery to the systemic vascular bed despite arterial oxygen desaturation, particularly if the desaturation is properly titrated.[5]

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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