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

Short title: AFR in IPAH

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We present a 20-year-old male with severe drug resistant idiopathic pulmonary hypertension (IPAH) diagnosed two years before. IPAH-specific triple combination therapy with intravenous epoprostenol was introduced initially. Since diagnosis progressive deterioration started with several hospitalizations due acute worsening. Since February 2019 patient’s mobility had been restricted significantly (World Health Organization [WHO] functional class IV with six-minute walk distance [6MWD] 197 meters) and severe right heart failure (RHF) with fluid retention had developed. The epoprostenol dosage reached 91.5 ng/kg/min. The patient received intravenous infusion of levosimendan three times every few months and was on an active waiting list for lung transplantation. Taking into consideration patient’s progressive worsening and no other medical therapy alternative, PAH specialist team decided to implant the atrial flow regulator (AFR) to decompress and improve right ventricle (RV) function. Approval of local ethics committee was obtained, and patient was given detailed information on the risks and benefits of the procedure and signed informed consent form.

During the first step of the procedure, detailed right heart catheterization (RHC) was performed with stepwise oximetry and hemodynamics measurements (Fick’s method) according to current recommendations [1]. The mean right atrium pressure (mRAP) was 16 mmHg, mean pulmonary artery pressure (mPAP) was 66 mmHg, pulmonary artery wedge pressure (PAWP) was 10 mmHg, pulmonary vascular resistance (PVR) was 13.9 Wood’s units, pulmonary flow (Qp) was 4.1 l/min, pulmonary-to-systemic flow ratio (Qp:Qs) was 1.0, and systemic blood pressure was 90/60 mmHg. Oximetry measurements without oxygen supplementation demonstrated mixed venous oxygen saturation from PA (SvO₂) 42.3% and arterial blood saturation from the aorta 84.7%, respectively. Under three-dimensional transesophageal echocardiography (TEE) guidance transseptal puncture was performed, followed by septostomy of atrial septum with the EverCross balloon (Medtronic, Mineapolis,
USA) (balloon size 8.0 mm x 4.0 cm) [Figure 1A]. Immediately, after septostomy patient’s oxygen saturation dropped to 72%, so the decision was made to implant Occlutech Atrial Flow Regulator (AFR) (Occlutech, Helsingborg, Sweden) device with 6 mm fenestration diameter. The device was introduced through the femoral vein using 10 F delivery sheath and implanted under angiography and TEE control without complications [Figure 1B-F]. Within several hours’ saturation improvement up to 90% within oxygen supplementation (3.0 L/minute) was observed.

Eight weeks after AFR insertion the patient’s clinical status improved (WHO functional class III, 6MWD 210 meters). The follow-up RHC (also Fick’s method) demonstrated mRAP 14 mmHg, mPAP 55 mmHg, PAWP 8 mmHg, PVR 5.7 Wood’s units, Qp 8.2 l/min, Qp:Qs ratio of 0.85, and SvO₂ of 65.6% and arterial saturation of 84.2%. Nevertheless, analyses were performed few days after levosimendan and iron intravenous infusions and oxygen supplementation. Twelve weeks after AFR implantation patient underwent successful lung transplantation.

IPAH is a severe, progressive disease leading to RHF, and ultimately death [2]. The present case shows that creation of an interatrial right-to-left shunt in end-stage IPAH patients may reduce the clinical signs of RHF and improve CO [3-4]. Increasing CO may ameliorate effective oxygen delivery to the systemic vascular bed despite arterial oxygen desaturation, especially if the desaturation is properly titrated [5].
References:


Figure 1

A. Angiogram from the balloon atrial septostomy (arrow).

B. Angiogram from the procedure showing AFR (arrow) just before the final locking.

C. Angiogram from the procedure showing AFR (arrow) after release.

D. Occlutech Atrial Flow Regulator (AFR) - self-expanding nitinol wire mesh device with fenestration (in presented case the diameter was 6 mm).

E. Final transesophageal echocardiogram (TEE) visualizing a three dimensional left atrial enface view of Occlutech AFR device.

F. Final TEE showing the shunt through Occlutech AFR device.