CLINICAL VIGNETTE

Closure of secundum atrial septal defect with a fenestrated occluder in a patient with severe pulmonary hypertension

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A 53-year-old woman with several years' history of exertional dyspnea (New York Heart Association class III) and recently diagnosed secundum atrial septal defect (ASD II) was admitted to our department for further evaluation and treatment.

Transthoracic echocardiography showed a markedly dilated right ventricle (61 mm) with moderate tricuspid regurgitation (peak velocity, 3.1 m/s), normal function of both ventricles, and enlargement of both atria (right atrial area, 31 cm²; left atrial area, 27 cm²) with the presence of ASD II. Transesophageal echocardiography (TEE) confirmed moderately large ASD II sized 27 mm with sufficient rims and left-to-right shunt on color flow Doppler analysis. Blood tests revealed an increased level of N-terminal pro-B--type natriuretic peptide (1064 pg/ml; reference range, <125 pg/ml). On cardiopulmonary exercise testing, the patient's peak oxygen uptake was 16.8 ml/kg/min and the minute ventilation/carbon dioxide production slope was 28.9.

Right heart catheterization (RHC) with stepwise oximetry and measurement of hemodynamic parameters (using the Fick's method') indicated precapillary pulmonary hypertension (PPH), with a mean pulmonary arterial pressure (mPAP) of 39 mm Hg, pulmonary artery wedge pressure (PAWP) of 7 mm Hg, pulmonary vascular resistance (PVR) of 3.7 Wood units (Wu), systemic vascular resistance index (SVR) of 21.3 Wu, and pulmonary-to-systemic flow ratio (Qp:Qs) of 2.7.

Due to the severe pulmonary hypertension, it was decided to close the defect with a fenestrated device. Approval of the local Bioethics

Committee was obtained, and the patient was given detailed information on the risks and benefits of the proposed procedure and signed an informed consent form. During the first step of the procedure RHC was performed again, confirming the presence of severe PPH (mPAP, 53 mm Hg; PAWP, 7 mm Hg; PVR, 3.3 Wu; SVR, 17.5 Wu) and a significant left-to-right shunt (Qp:Qs, 2.7). The Swan-Ganz catheter was positioned in the pulmonary artery throughout the procedure and PAP and PAWP were monitored continuously. Using the femoral venous access and a 14-F delivery sheath, a custom-made fenestrated ASD occluder (Occlutech, Helsingborg, Sweden) with waist and fenestration diameters of 27 mm and 6 mm, respectively, was successfully implanted with the standard ASD closure technique under fluoroscopic and TEE guidance, without complications (FIGURE 1A-1F).

The patient's symptoms improved significantly during the 3-month follow-up and control TEE evaluation demonstrated proper position of the fenestrated ASD occluder with a residual left-to-right shunt across the fenestration.

A therapeutic approach in patients with ASD II and severe PPH remains controversial due to limited data. The precise cutoff value of PVR which precludes ASD II closure in the presence of PPH does not exist. ^{2,3} Current guidelines recommend closure if the defect is significant and PVR is lower than 5 Wu, but it may also be correctable with a fenestrated device when PVR drops below 5 Wu after targeted PPH treatment. ^{4,5} However, the extent of improvement decreases with the increase of mPAP. In such patients, the presence

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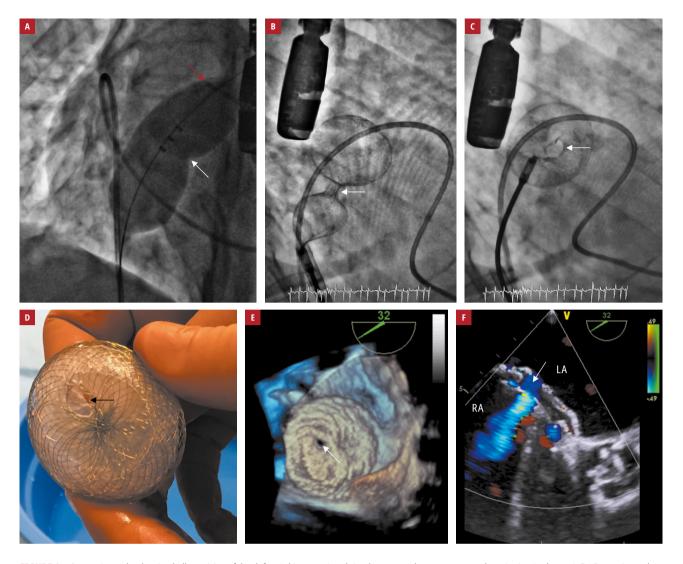


FIGURE 1 A – angiography showing balloon sizing of the defect (white arrow) and simultaneous pulmonary artery catheterization (red arrow); **B**, **C** – angiography performed during the implantation procedure showing the fenestrated ASD occluder (FASD; arrows) just before the final locking (**B**) and after release (**C**); **D** – the FASD, a self-expanding nitinol wire mesh device with fenestration (fenestration diameter, 6 mm; arrow); **E**, **F** – final transesophageal echocardiography visualizing a 3-dimensional left atrial en face view of the FASD (**E**) and the shunt through the FASD (**F**); fenestration indicated by the arrows

of a fenestration within the occluder may allow a blood overflow in the right-to-left direction as a reduction of right heart volume overload. To our best knowledge, this is the first report describing the usage of a custom-made fenestrated ASD occluder in Poland. Our findings indicated that percutaneous closure of the defect with a fenestrated device in carefully selected adults with ASD II and developed PPH may be feasible and safe.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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