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Use of Occlutech® Fenestrated Atrial Septal Defect Occluder in ASD-Associated Pulmonary Arterial Hypertension

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Introduction

Pulmonary Arterial Hypertension (PAH) is a chronic disease with progressively increasing right ventricular (RV) pressure, Right-Heart Failure (HF), and death.[1] An association between PAH and secundrum-type Atrial Septal Defect (ASD) is observed in 9 to 35% of patients, especially in females. It is speculated that ASD-Associated PAH resolves after intervention and rarely progresses especially with early intervention.[2-4] However, as most of these patients are left with untreated ASDs, there is a dearth of information. Fenestrated ASD closure is preferable in patients with moderate to severe PAH. A restricted interatrial shunt in these patients can enhance systemic ventricular output at the expense of desaturation if shunt reversal occurs when progressive PAH ensues. Maintaining a sustainable restricted interatrial communication is challenging without the use of a dedicated device such as the Occlutech® Fenestrated Atrial Septal Defect (FASD) occluder (Figure 1). We describe compassionate use of the FASD Occluder with optimal outcomes in a 56-year-old female with ASD-Associated PAH.

Case Report

A 56-year-old female with progressive PAH was referred to the Congenital Heart Center for evaluation and management. She was receiving combination medical therapy with

Figure 1: Occlutech® Fenestrated Atrial Septal Defect Occluder.

macitentan and tadalafil. However, she continued to experience dyspnea when attempting to climb a flight of stairs and had bilateral lower extremity edema.

At the age of 48 years, she did not fully recover from bronchitis with symptoms including orthopnea, paroxysmal nocturnal dyspnea, dyspnea on exertion, fatigue, wheezing, cough, near-syncope, and nausea, which was later diagnosed as severe PAH with moderately elevated Pulmonary Vascular Resistance (PVR). Right ventricular (RV) systolic pressure was 95 mmHg, pulmonary artery (PA) pressure was 95/30 (53) mmHg with systemic blood pressure of 112/66 (81) mmHg. Cardiac output was 3.87 L/min (Fick method).

She had a 27 mm ostium secundum ASD. A three-dimensional transesophageal echocardiogram (3DTEE) showed predominantly left-to-right shunt with some flow reversal. Her six-minute walk test distance dropped from 1580 to 1400 feet in a span of three months. As a result of worsening PAH on maximal medical therapy, a decision was made by a multidisciplinary team including a pulmonologist and lung transplant director, and congenital heart specialists to close the defect with a fenestrated device. The Occlutech® FASD Occluder was selected for use under the U.S. Food and Drug Administration's compassionate use guidance. The patient was given comprehensive education on the risks and benefits of the procedure, including complications of general anesthesia, TEE, cardiac catheterization and the occluder itself. The potential intraprocedural and postprocedural risks including air embolus, allergic reaction to nickel, arrhythmia, bleeding, injury to blood vessels, device embolization and migration, and thromboembolic events were also discussed. The patient signed the compassionate use informed consent form after all questions were addressed to her satisfaction prior to the procedure.

Procedure

Under general anesthesia, 3DTEE confirmed the presence of a significant atrial communication with persistent left-to-right shunt. A detailed right-heart catheterization was performed under aseptic precautions with stepwise oximetry and hemodynamics. The patient's RA pressure was 12/7 (9) mmHg, PA pressure was 80/29 (47) mmHg with systemic blood pressure of 100/50 (67) mmHg. Oximetry in 30% oxygen included superior vena cava (SVC) 78%, inferior vena cava (IVC) 75%, RA 80%, RV 81%, PA 80%, and LA 92%. Although the pre-procedure calculated Qp:Qs was 0.91 suggesting a net right-to-left shunt through the ASD, the pulmonary reactivity testing showed Qp:Qs of 1.5: 1 on 100% oxygen and nitric oxide. A balloon occlusion test showed no hemodynamic instability on complete occlusion. Cardiac output was 3.81 L/min (Fick method).

A 0.035-inch extra-stiff guidewire was placed into the pulmonary vein and a 14F Mullins sheath was advanced over the guidewire and positioned into the uppper-left pulmonary vein. A 27 mm FASD occluder with 6 mm fenestration was loaded on the delivery cable system and advanced in through the sheath. The LA disk was deployed followed by deployment of the RA disk under fluoroscopic and 3DTEE guidance. After confirming secure deployment of the FASD occluder on 3D TEE (Figures 2 and 3), hemodynamic

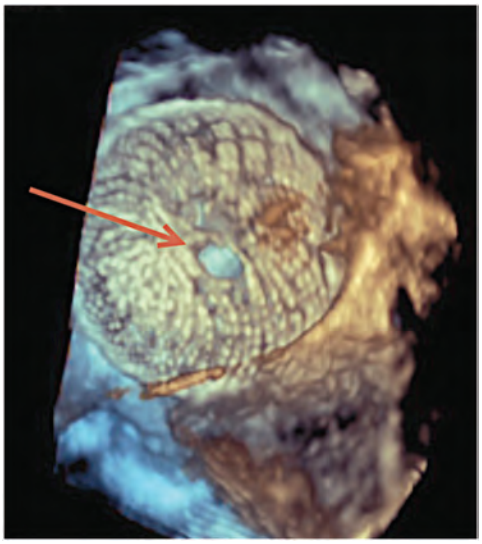

Figure 2: Post-deployment 3DTEE shows a well-seated FASD. The red arrow shows the 6 mm fenestration in the occluder.

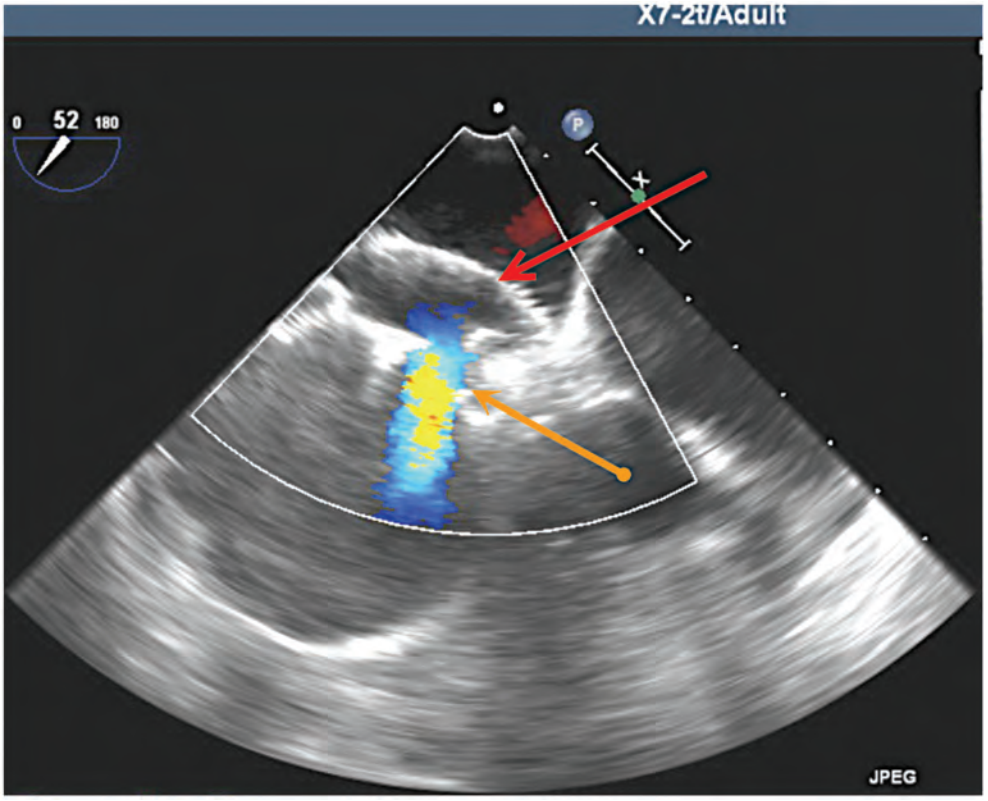

Figure 3: The FASD Ocludder (red arrow) with left-to-right shunting through the fenestration (orange arrow) on two-dimensional TEE post deployment.

measurements were repeated. The patient's RA pressure was 10/8 (8) mmHg, PA pressure was 72/26 (43) mmHg with systemic blood pressure of 139/68 (94) mmHg. Cardiac output improved to 5.97 L/m (Fick Method) with no significant demonstration of left-to-right shunt post device deployment. The delivery cable was then released from the device and withdrawn.

The patient's hemodynamics were stable throughout the procedure and there were no arrhythmias or other complications. She was also extubated without any complications and discharged home the next day. As PAH warrants anticoagulation therapy; the patient's anticoagulation regimen (clopidogrel and aspirin) was continued after the deployment of the device.

"In patients with ASD-Associated PAH, the unrestricted shunting can lead to severe symptoms and progressive PAH. A fenestrated device must be considered in these patients to restrict significant left-to-right shunting, but simultaneously allow for any necessary overflow if and when right HF develops in the future....
The significant symptomatic improvements and the sustained atrial communication four months after implantation of the FASD Occluder in our patient shows that it may be a useful closure device in patients with ASD-Associated PAH."

Following discharge, the patient developed a right pseudoaneurysm with an arteriovenous fistula at the catheterization site requiring two thrombin injections. The complication resolved over time and was determined to be unrelated to the FASD occluder.

At her one-month follow-up visit, the patient reported significant improvement in her exercise tolerance and she had more energy with recorded resting saturation of 99%. Her six-minute walk test distance was relativity unchanged at 1430 feet. She remained without pedal edema on the same diuretic therapy. An echocardiogram demonstrated good device placement, improved RV pressures, and continuous left-to-right shunt across the FASD Occluder at rest.

Four months after the procedure, the patient reported feeling very well with significant improvement in her stamina and exercise tolerance. An echocardiogram demonstrated improved RV

pressures with continuous left-to-right shunt across the FASD Occluder at rest. Her six-minute walk test distance significantly improved to 1520 feet.

Discussion

The association between PAH and ASD in young adults, especially in females is well - recognized. It is typically independent of the degree of shunting and increased pulmonary blood flow through the ASD.[2-4] Although it can be slow in becoming symptomatic, these patients can develop progressive PAH. It is critical to appreciate the difference between this group of patients from those who do not develop mild PAH with large ASDs. In the latter group, it can resolve after intervention, especially when performed early and rarely do patients develop progressive PAH. When considering our patient's clinical presentation and cardiac catheterization findings at the time of ASD closure, she is likely to have had ASD-Associated PAH.

Patients who develop PAH immediately or several months or years after ASD closure have poorer prognosis when compared to Congenial Heart Disease (CHD) patients with PAH.[5-7] As such, a fenestrated ASD closure is preferred in patients with ASD and moderate-to-severe PAH to decrease significant left-to-right shunting, but allow possible overflow for right HF in the future.[7] Creating a restricted and sustainable atrial communication can be challenging and compelled us to use the FASD Occluder that can maintain a fenestration. However, spontaneous closure can occur in fenestrated devices.[7-9]

Conclusion

In patients with ASD-Associated PAH, the unrestricted shunting can lead to severe symptoms and progressive PAH. A fenestrated device must be considered in these patients to restrict significant left-toright shunting, but simultaneously allow for any necessary overflow, if and when right HF develops in the future. It is desirable to achieve higher systemic ventricular output with marginal increase in cyanosis in these patients with an optimal saturation range of 87-90% at rest. The significant symptomatic improvements and the sustained atrial communi cat ion four months after implantation of the FASD occluder in our patient shows that it may be a useful closure device in patients with ASD associated PAH.

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