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LETTER TO THE EDITOR
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Partial closure with a self-made fenestrated device of secundum atrial septal defect with severe pulmonary artery hypertension in adults

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Secundum atrial septal defect (ASD) represents one of the most common congenital heart diseases, accounting for up to 10% of all congenital heart defects. It is often diagnosed in adulthood due to the absence of clinical symptoms.[1] By the age of 40 years, 90% of untreated patients have symptoms of exertional dyspnea, fatigue, palpitation, sustained arrhythmia, or even evidence of heart failure. Although ASD is less commonly associated with pulmonary artery hypertension (PAH), a subset of patients may develop moderate-to-severe PAH, often occurring in the fourth decade of life and defined as mean pulmonary artery pressure (PAP) > 20 mmHg with pulmonary vascular resistance (PVR) ≥ 3 Wood units.[2] Pulmonary hypertension has been also classified as moderate (50–59 mmHg) or severe (≥ 60 mmHg) according with the right ventricular systolic pressure calculated by echocardiography. It is noteworthy that adult ASD patients with severe PAH have a poorer prognosis than those without.[3,4] In this setting, complete closure of the septal defect by transcatheter or surgical intervention is still a matter of debate and may result in complications associated with sudden increase in PVR,[5,6] or serious acute left ventricular dysfunction leading to pulmonary edema immediately after closure.[7] We report a case series of moderate-to-large ASD with severe PAH in whom a modified incomplete closure using a self-fenestrated (SF)-ASD device has been performed in order to reduce gradually the left-to-right shunt (LRS) and promote further decrease of pulmonary arterial pressure in the mid-term and long-term follow-up.

Case 1. A 52-year-old obese male patient was admitted for progressive dyspnea on exertion and fatigue. He has a past history of long-standing arterial hypertension, dyslipidemia and coronary artery disease (previous anterior myocardial infarction) for which he underwent percutaneous coronary intervention with implantation of a drug-eluting stent in the left descending coronary artery. He was in New York Heart Association (NYHA) class II under beta-blockers (bisoprolol: 2.5 mg/day), angiotensin converting enzyme inhibitors (20 mg/day), aspirin (100 mg/day), statin (20 mg/day) and furosemide (25 mg/day). Electrocardiogram showed incomplete right bundle branch block and first-degree atrioventricular block. Chest X-ray revealed signs of increased pulmonary flow, right chambers dilatation as well as the pulmonary artery and its branches. Baseline two-dimensional (2D) transthoracic/transesophageal echocardiography (TTE/TEE) color Doppler showed significant enlarged right ventricle with a tricuspid annular plane systolic excursion (TAPSE) of 13 mm, a large ASD with adequate rims except for a diminutive antero-superior aortic one (Figure 1A & 1B), LRS with the pulmonary to systemic flow (Qp/Qs) ratio of 1.3:1, an estimated systolic PAP of 85 mmHg, mildly hypertrophied left ventricle with an ejection fraction of 50% with

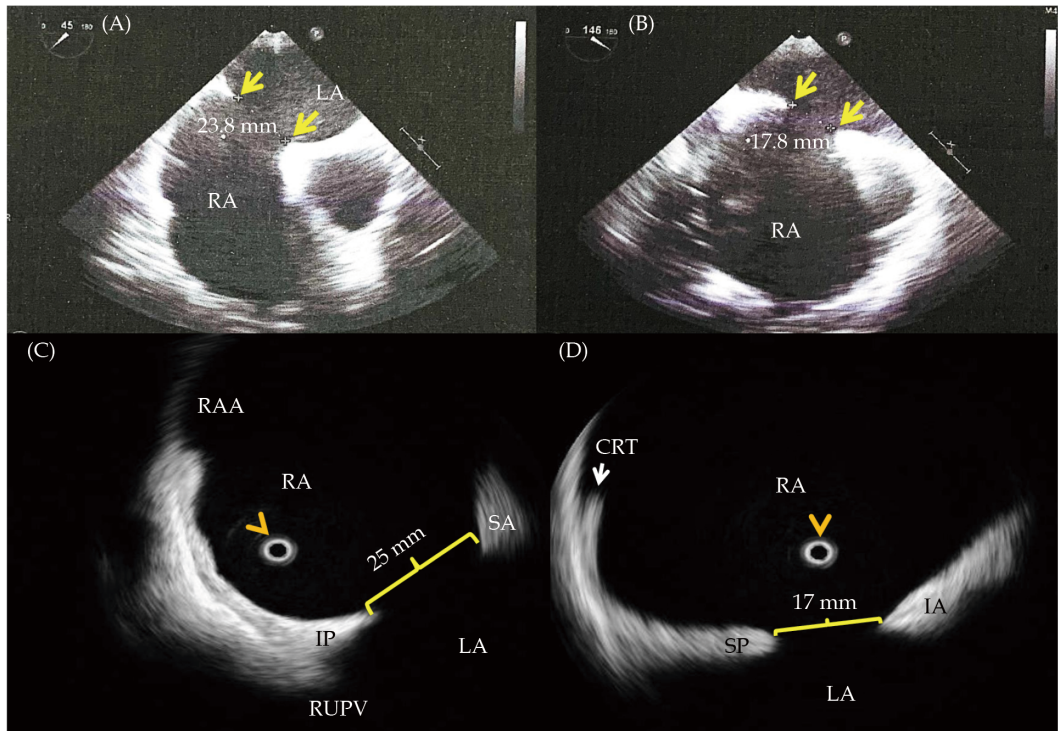

Figure 1 Baseline two-dimensional transoesophageal echocardiogram at 45 degree (A) and 146 degree (B) views showing a large secundum ASD (2.38 mm × 17.8 mm in diameter, yellow arrows) with a diminutive aortic rim. Intraprocedural rotational intracardiac echocardiography by Ultra-ICE (mechanical 9F/9 MHz 360° scan probe) views showing ASD, 25-mm diameter in the axial plane (C) and 17-mm diameter in the long-axis four-chamber (D) planes. Crista terminalis (white arrow) (panel D) appears as a linear structure at the junction between the posterior smooth wall and the anterolateral trabeculated portion of the right atrium. ICE catheter (orange arrowhead) is located at the center of the images. ASD: atrial septal defect; CRT: crista terminalis; IA: infero-anterior rim; ICE: intracardiac echocardiography; IP: infero-posterior rim; LA: left atrium; RA: right atrium; RUPV: right upper pulmonary vein; SA: supero-anterior rim; SP: supero-posterior rim.

impaired relaxation (type I diastolic dysfunction). The patient underwent right heart catheterization demonstrating LRS with QP/QS ratio of 1.4:1, a mean PAP of 40 mmHg (systolic: 75 mmHg, diastolic: 28 mmHg) and a moderately increased PVR (3.5 Woods units).

After heart team discussion, the decision to proceed with a catheter-based treatment was confirmed. Written informed consent, after explanation, was obtained from the patient. Percutaneous closure was performed under local anesthesia, fluoroscopic guidance and rotational intracardiac echocardiography (rICE) continuously obtained by the same interventional team (a single-operator use) using a commercially available 9F-9MHz Ultra ICE™ catheter-based ultra-sound transducer (EP Technologies, Boston Scientific Corporation, San Jose, CA, USA), as previously described.[8–10] Intraprocedural rICE showed secundum ASD, 25-mm in diameter on the axial plane (Figure 1C) and 17-mm in diameter on the long-axis four-chamber view (Figure 1D). A 24-mm ASD Occluder (ASDO; MemoPart, Lepu Medical Technology) with a flexible titanium-oxide coated nitinol-mesh was selected for implantation. After pushing a 7-Fr dilator through both discs and the waist of the ASDO close to the center hub of the device, a low-profile high-pressure 8 mm × 20 mm balloon (Cordis Saber™, USA) was then positioned across the device and fully inflated to its maximum rated pressure of 10 atmospheres to create a 5-mm fenestration (Figure 2). The 24-mm ASDO with the self-made fenestration of 5 mm was successfully implanted under rICE (Figure 3A) and fluoroscopic guidance (Figure 4). The patient was discharged home the day after the procedure on dual antiplatelet therapy in better clinical conditions and 2D TTE color Doppler showed a well aligned fenestrated device with a residual LRS corresponding to the fenestration's patency (Figure 3B). At three-month follow-up, he had a sig-

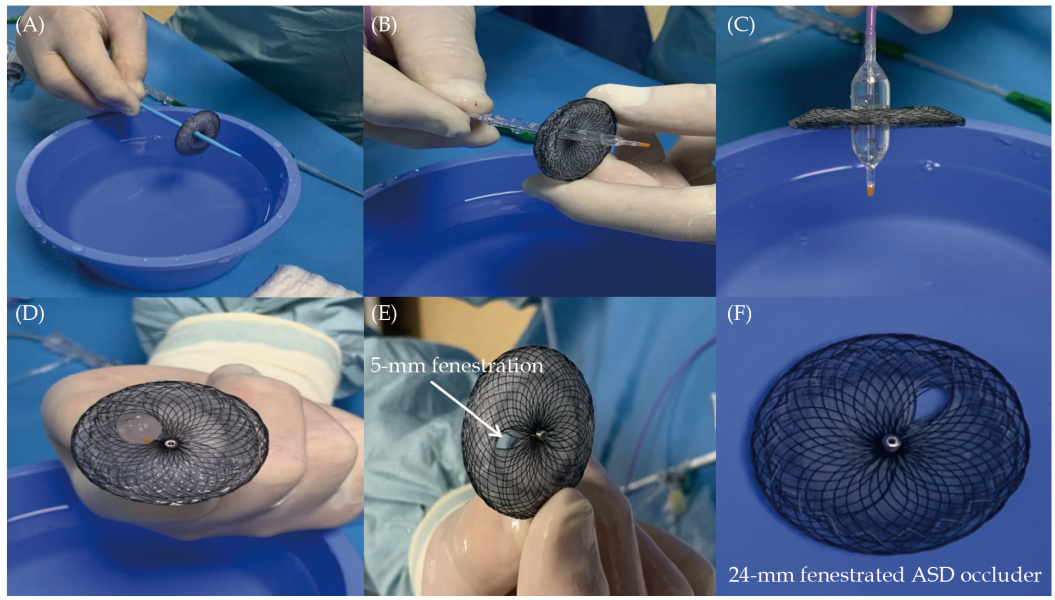

Figure 2 In vitro steps of the self-made fenestration of the 24-mm ASD device. The fenestration was created by pushing a 7-Fr dilator through both discs and the waist of the ASD device halfway between the edge of the connecting stent part and the center hub of the device (A). A low-profile, high-pressure 8 mm × 20 mm balloon (Cordis Saber™, USA) was positioned across the center of the device (B) and dilated to its maximum rated pressure of 10 atmospheres until fully inflated (C & D) to create a 5-mm fenestration (E). After removal of the balloon, the fenestration was evaluated for its diameter and correct position within the fabric ensuring the final fenestration would be centered within the fabric of the connecting stent part of the device (F). ASD: atrial septal defect.

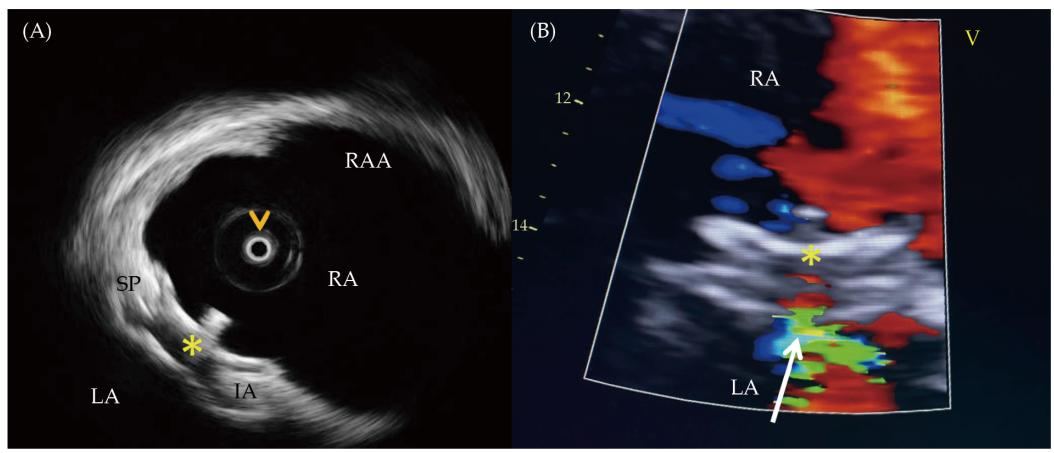

Figure 3 Post-procedure rICE by Ultra-ICE (mechanical 9F/9 MHz 360°) and two-dimensional transthoracic echocardiography color Doppler. rICE in the long-axis four-chamber view (A) showing the correctly implanted SF 24-mm ASDO (yellow asterisk). Post-procedure two-dimensional transesophageal echocardiography color Doppler (B) in the subcostal view showing the well aligned SF 24-mm ASDO (yellow asterisk) with a residual left-to-right shunt (white arrow) corresponding to the fenestration's patency. ICE catheter (orange arrowhead) located at the center of the images. ASDO: atrial septal defect occluder; IA: infero-anterior rim; ICE: intracardiac echocardiography; LA: left atrium; RA: right atrium; rICE: rotational intracardiac echocardiography; SP: supero-posterior rim.

nificant clinical improvement and 2D TTE color Doppler confirmed the stable position of the device with an estimated PAPs of 30 mmHg with no right ventricular overload. No device malfunction, thrombus formation or other complications like paradoxical embolism, endocarditis, or right-to-left shunt occurred so far.

Case 2. A 55-year-old female patient was admitted for progressive dyspnea. Her medical history included a previous diagnosis of deep venous thrombosis treated with oral anticoagulant. No cardiovascular risk factors were reported. Electrocardiogram showed atrial fibrillation and a computed tomography scan confirmed pulmonary artery

trunk dilatation. Chest X-ray revealed signs of increased pulmonary flow, right chambers dilatation as well as the pulmonary artery and its branches. She was under pantoprazole (20 mg/die) and rivaroxaban (20 mg/die). The 2D TTE/TEE color Doppler (Figure 5) showed an enlarged right ventricle with TAPSE of 16.5 mm, a large ASD (26 mm) with LRS with Qp/Qs ratio of 1.9:1, estimated systolic PAP of

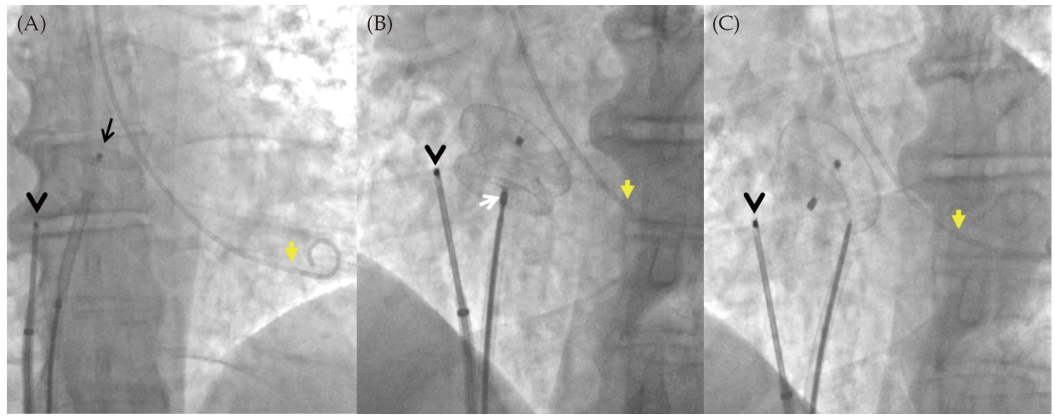

Figure 4 Fluoro-angiographic procedural steps under rotational intracardiac echocardiography (Case 1). (A): Distal left disc (black arrow) of the SF 24-mm ASDO opened in the left atrium in close proximity of the interatrial septum with the tip of the Ultra-ICE probe (black arrowhead) positioned in the right atrium; note that a 6-Fr pig-tail catheter (yellow arrow) has been placed in the apex of left ventricle as a continuous left ventricle pressure monitoring tool; (B): the proximal right disc of the SF 24-mm ASDO engaging the right aspect of the interatrial septum; the device is still anchored to the delivery catheter (white arrow); and (C): the SF 24-mm ASDO device successfully deployed. ASDO: atrial septal defect occluder; ICE: intracardiac echocardiography.

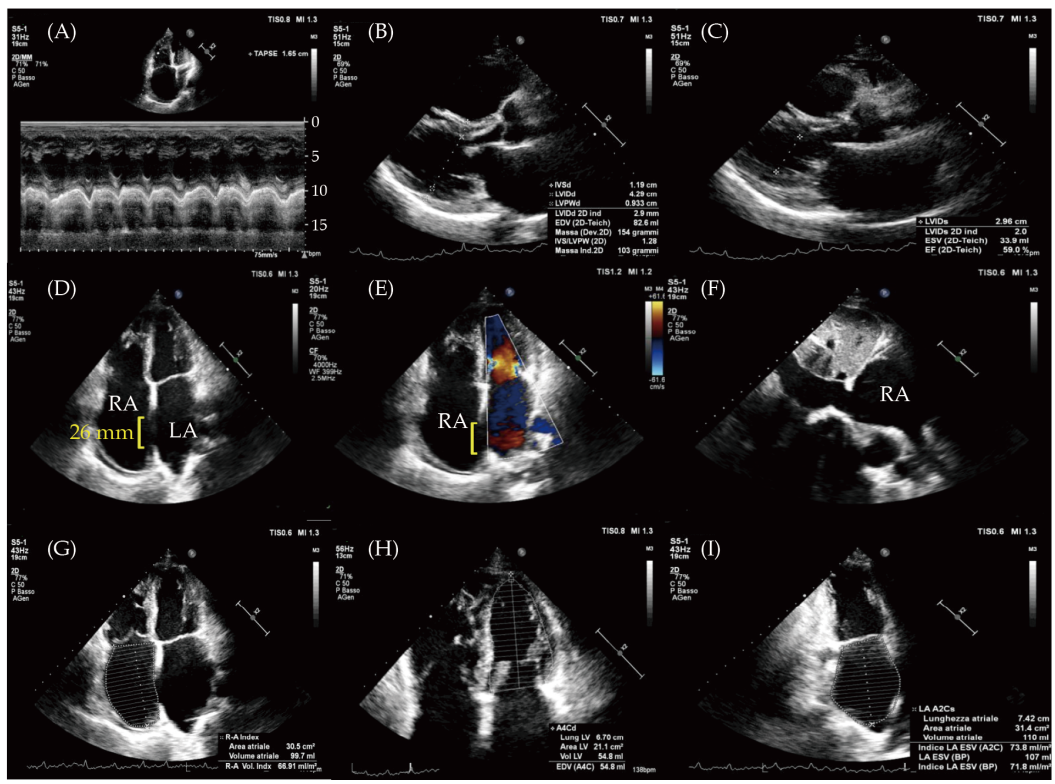

Figure 5 Baseline two-dimensional transthoracic echocardiogram color Doppler image. (A): An enlarged right atrial cavity with tricuspid annular plane systolic excursion of 16.5 mm (B-mode); (B & C): left ventricle end-diastolic and end-systolic dimensions, ventricular septum and posterior left ventricle wall diameters in parasternal long-axis views; (D–F): a large secundum ASD (26 mm in diameter) in four chamber views with a markedly dilated RA in subcostal view; and (G–I): volumes and areas of RA, left ventricle and LA in four- and two- chamber views. LA: left atrium; RA: right atrium.

80 mmHg, normal left ventricle function with an ejection fraction of 59%.

The patient underwent right heart catheterization confirming a QP/QS ratio of 2.5:1, a mean PAP of 40 mmHg (systolic: 80 mmHg, diastolic: 29 mmHg) and increased PVR (4.2 Woods units).

The case was evaluated by the heart team and a catheter-based treatment was scheduled. Following adequate explanation, written informed consent was obtained from the patient. Percutaneous ASD closure was then performed under local anesthesia, fluoroscopic and rICE guidance, without balloon-sizing maneuver, as per our routine. Preprocedural rICE showed a secundum ASD, with a diameter of 27 mm on the axial plane and 30 mm on the long-axis four-chamber view. A 28-mm ASDO was selected for implantation and a 5-mm fenestration was created across the device as previously described. The SF 28-mm ASDO was then successfully implanted under rICE and fluoroscopic guidance (Figure 6). The patient was discharged home the day after the procedure in good clinical conditions on antiplatelet therapy in addition to rivaroxaban. Three-month 2D TTE color Doppler and right heart catheterization follow-up confirmed the stable position of the SF-ASDO with no residual shunt as detected by color flow Doppler and fluoro-angiography (Figure 7). Right ventricle end-diastolic diameters decreased. Hemodynamic parameters showed PAPs of 25 mmHg and QP/QS ratio of 1:1. No device malfunction or thrombus formation occurred so far. The patient showed a remarkable clinical improvement in terms of NYHA class and quality of life.

As previously mentioned, in adult population complete closure of ASD associated with severe PAH may be risky due to the possible abrupt increase of PVR with pulmonary hypertensive crisis or acute pulmonary edema in those elderly patients with concomitant decreased left ventricle compliance or overt diastolic dysfunction.

The concept of fenestrated closure to allow for a minor shunt in both directions depending on the compliance failure of the ventricles is not new and has been already tested in the past. Several case reports on the use of SF-made or custom-made (AGA Medical Corporation) ASD devices have been reported in the literature.[11-14] Indeed, fenestrated closure allows for controlled residual shunt providing adequate cardiac output with a mechanism for decompression in the event of critical increase in PVR as in our two cases.

After reviewing all previous case reports published, it is worthy to note that self-made fenestrations closed as early as three months while custom-

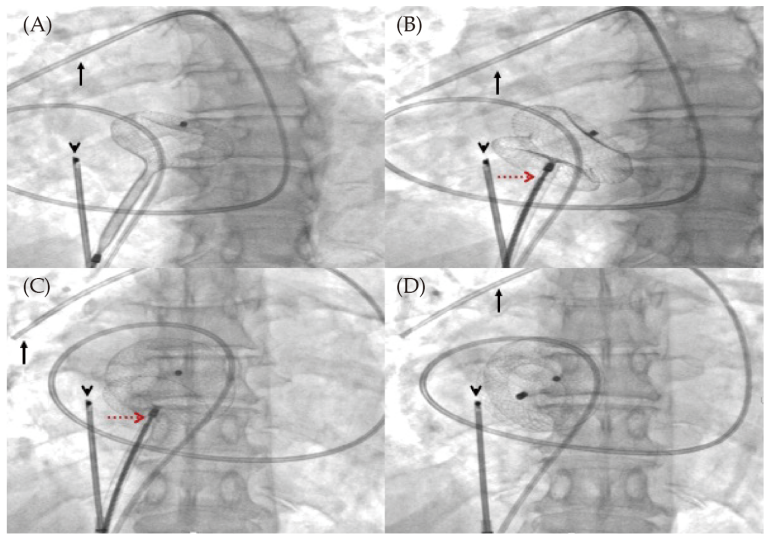

Figure 6 Fluoro-angiographic procedural steps under rotational intracardiac echocardiogram with Swan-Ganz catheter in the distal right pulmonary artery (Case 2). (A): Distal left disc of the SF 28-mm ASDO opened in the left atrium in close proximity to the interatrial septum; the tip of the Ultra-ICE probe (black arrowhead) in the right atrium and Swan-Ganz catheter (black arrow) in the distal right pulmonary artery; (B & C): the proximal right disc of the SF 28-mm ASDO engaging the right side of the interatrial septum and still anchored to the delivery catheter (red dotted arrow); and (D): the SF 28-mm ASDO finally deployed. ASDO: atrial septal defect occluder; ICE: intracardiac echocardiography.

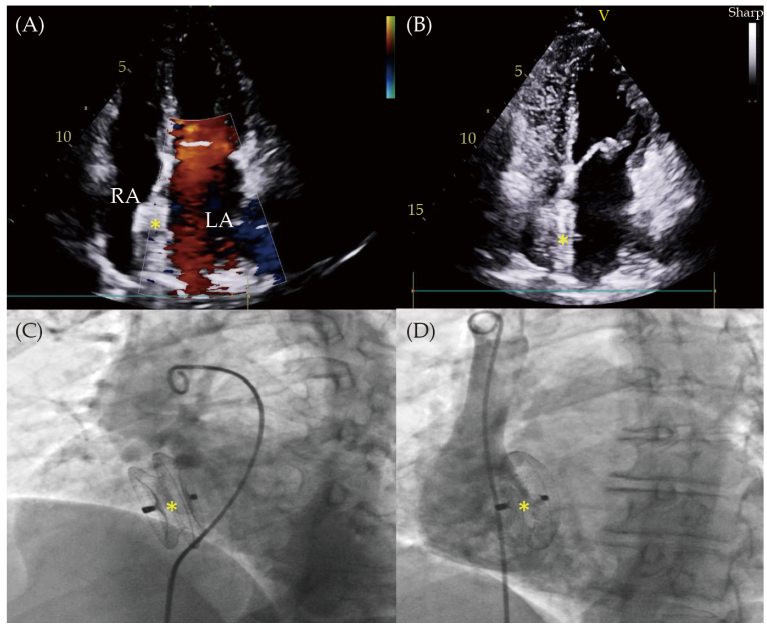

Figure 7 Three-month follow-up two-dimensional TTE color Doppler and fluoro-angiographic images (Case 2). Two-dimensional TTE color Doppler and contrast-enhanced TTE in apical four-chamber view showing the SF 28-mm ASDO (yellow asterisk) correctly aligned to the interatrial septum without any residual left-to-right or right-to-left shunt and markedly reduced right atrial cavity (A & B). Right pulmonary artery angiography in hepato-clavicular projection (C) and superior vena cava angiogram (D) showing the perfect position of the SF-28 mm ASDO without any residual left-to-right or right-to-left shunt, confirming that the self-made fenestration is definitively closed. ASDO: atrial septal defect occluder; LA: left atrium; RA: right atrium; TTE: transthoracic echocardiography.

made fenestrated devices tend to remain patent for longer duration. Differently from the previous reports, in our two patients, we did not placed sutures at the edges of the nitinol wires mesh for a long-lasting patent fenestration, allowing the memory properties of the nitinol mesh to reshape the device to its original configuration thus closing the fenestration gradually in the mid run.

Recently, a novel dedicated fenestrated device (FASD, Occlutech GmbH, Jena, Germany), available on an order-made basis, has proven to be beneficial without any major device-related complications[15] and may represent a cutting-edge technology in the cath lab.

In summary, partial occlusion of moderate-to-large ASD associated with severe but still reversible PAH using a SF-ASD device turned out to be a feasible alternative allowing gradual decrease in the magnitude of the shunt and right-sided chamber overload without any major device-related complications. Fenestrated ASD devices are an important therapeutic armamentarium for interventional ASD occlusion, especially for elderly patients in whom concomitant comorbidities may influence outcome and prognosis.

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