

# Catheter-based closure of a large atrial septal defect with inferior rim deficiency using pulmonary vein slide-out assisted implantation technique: a case report

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<b>Background</b>	Transcatheter approach for large and complex atrial septal defects may represent a therapeutic challenge, particularly when the postero-inferior rim is deficient and floppy.
<b>Case summary</b>	Here, we describe a successful catheter-based closure of a large (>30 mm) secundum atrial septal defect associated with postero-inferior rim deficiency in a 35-year-old female with congestive heart failure using pulmonary vein slide-out assisted implantation technique.
<b>Discussion</b>	Inferior–posterior rim deficiency is a well-known risk factor for device instability or embolization. Transcatheter closure may represent a safe and effective alternative to the traditional surgical approach provided that modified implantation techniques are employed.
<b>Keywords</b>	Atrial septal defect • Transcatheter closure • Transoesophageal echocardiography • Atrial septal rims deficiency • Case report

## Learning points

- Despite the transcatheter closure of atrial septal defect has become the most frequent treatment modality worldwide, the anatomical complexity of the interatrial septum makes the procedure particularly demanding in some cases.
- For these reasons, a comprehensive echocardiographic imaging and monitoring are recommended in order to select alternative strategies in case of improper positioning or incomplete anchoring of the occluder.
- The choice of a highly flexible more conformable device like Occlutech FSO and the modified implantation technique (pulmonary vein slide-out assisted technique) have been key to success in this complex catheter-based closure procedure.

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## Introduction

Transcatheter closure of secundum atrial septal defect (ASD) is currently accepted as the treatment of choice in most patients, showing excellent efficacy as well as lower complication rate compared to surgery.<sup>1-3</sup>

However, the feasibility of a transcatheter approach for large defects is much less clear particularly when the rim of septal tissue at the aortic root is absent, when the postero-inferior rim is deficient (<5 mm), and when the ASD is associated with excessively bulging atrial septal aneurysms (ASA) particularly if multifenestrated. The treatment decision for these complex defects (approximately 20% of ASD patients)<sup>4,5</sup> is unclear. Traditional management includes surgical closure, but morbidity of the open-heart procedure, presence of scar, and longer hospital stay makes surgical closure unattractive to many patients.

## Timeline

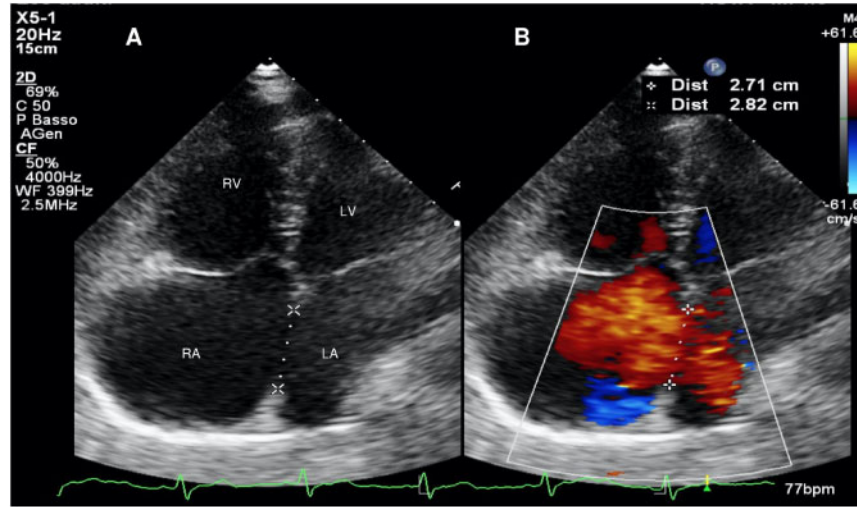
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- Day 1. Patient admitted for palpitations, chest pain, and long-term worsening dyspnoea (New York Heart Association class III)
  - Day 2. Chest X-ray revealed signs of increased pulmonary flow, right chambers dilatation as well as pulmonary artery and its branches
  - Day 3. Transthoracic and transoesophageal echocardiography detected a large secundum atrial septal defect with severe left-to-right shunt and significant right heart enlargement due to volume overload. Deficient floppy postero-inferior rim (<5 mm) was shown.
  - Day 5. The patient underwent a technically demanding catheter-based closure of her complex atrial septal defect anatomy with the help of 'pulmonary vein slide-out' assisted implantation technique
  - Day 7. Transthoracic echocardiography at discharge confirmed the stable position of the device without residual left-to-right shunt
  - Day 180. The patient remained symptom free. Transthoracic echocardiography confirmed the abolition of the left-to-right shunt, showing in addition a marked reduction in right cardiac chambers volume overload
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## Case presentation

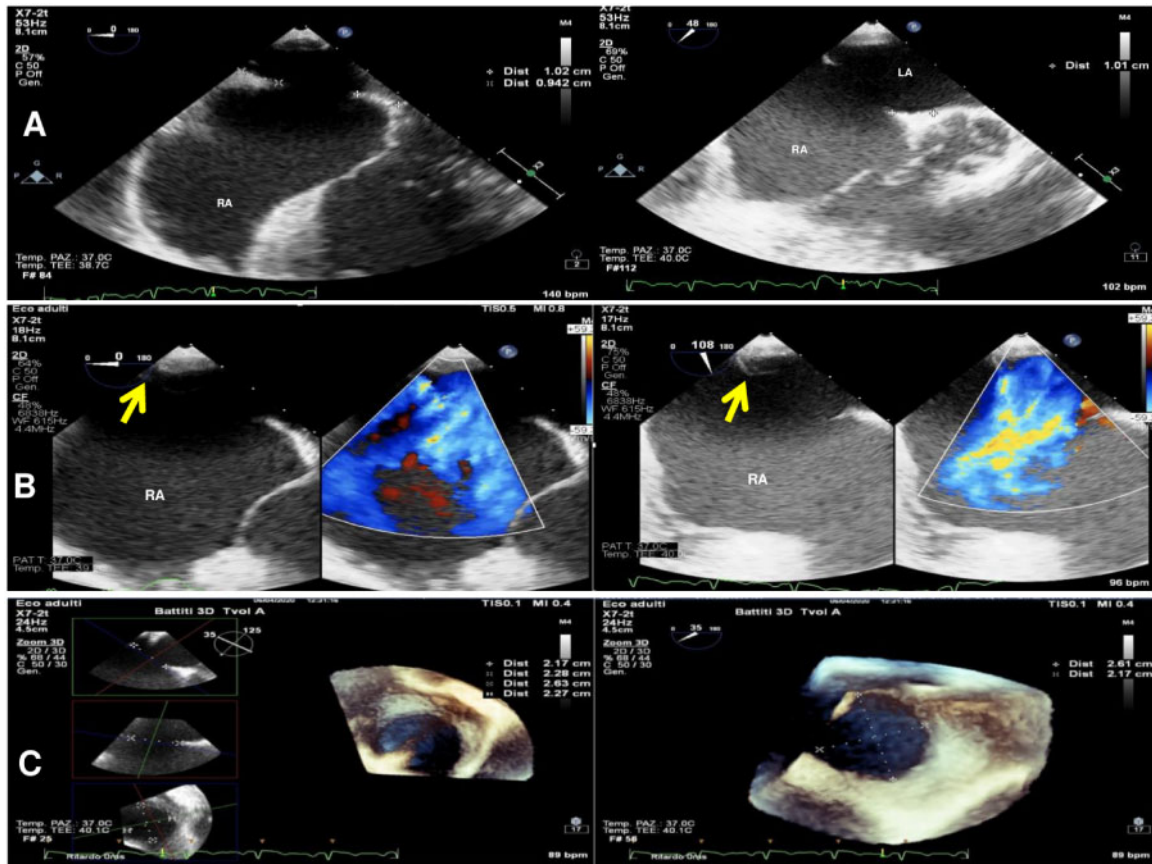
A 35-year-old female patient from Bangladesh without cardiovascular risk factors was admitted for chest pain, palpitations, and long-term exertional dyspnoea. Past medical history was unremarkable except for a heart murmur heard since childhood. Right ventricular enlargement was indicated by a palpable right ventricular lift; a systolic ejection murmur in the second and third intercostal spaces and a fixed splitting of the second heart sound throughout the respiratory cycle were audible. Electrocardiogram showed sinus rhythm with complete right bundle branch block and right atrium (RA) enlargement. Chest X-ray revealed signs of increased pulmonary flow, right cham-

bers dilatation as well as the pulmonary artery and its branches. After the initial diagnostic work-up and medical treatment optimization, transthoracic echocardiography showed a large secundum ASD (ranging from 27 to 28 mm in diameter) with severe left-to-right shunt with pulmonary to systemic flow ratio (Qp:Qs) ratio of 2.6, mildly increased mean pulmonary artery pressure (40/15 mmHg, mean 25 mmHg) and significant right heart enlargement due to volume overload (Figure 1). Transoesophageal echocardiography (TOE) colour Doppler (Figure 2A,B) imaging of the septal defect was done with 0–120° sweeping in addition to the standard imaging angles at 0°, 45°, and 90° and three-dimensional (3D) TOE (Figure 2C) in order to better define the secundum ASD (21.7 mm × 26 mm) showing a deficient floppy postero-inferior rim (<5 mm), adequate remaining rims (>10 mm), total septal length of 50 mm, normal drainage of pulmonary veins into the left atrium, and a thrombi-free left atrial appendage.

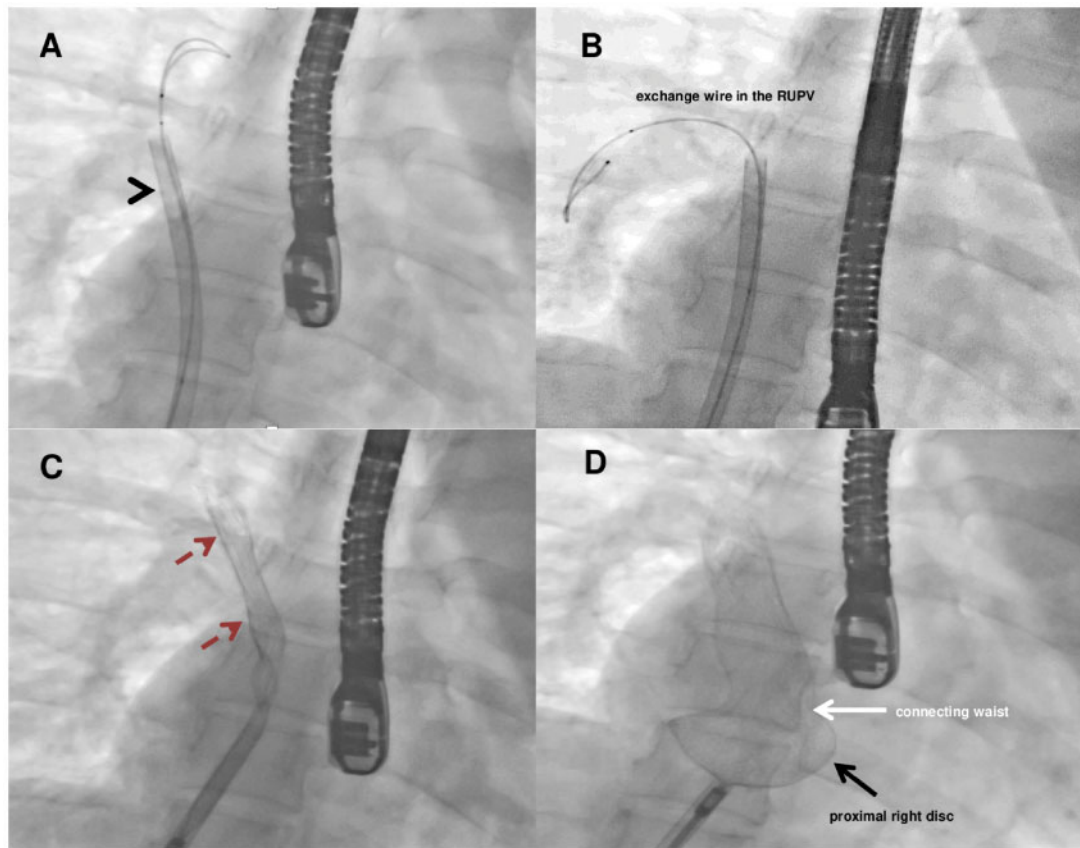
Considering that the particular ASD anatomy was likely to increase the technical difficulties of the percutaneous closure procedure, the patient was offered a surgical option, but she firmly rejected. In view of her clinical conditions (New York Heart Association class III), it was decided to address her congenital heart disease using a catheter-based technique. An informed consent was signed by the patient. The procedure was performed in the catheterization laboratory under deep sedation and continuous real-time 2D/3D TOE colour flow Doppler and fluoroscopic guidance. Balloon-stretched defect size was 25 mm in diameter and this measurement was used as a guide for device size selection. A third-generation Figulla Flex II ASD occluder (FSO, Occlutech GmbH, Jena, Germany) with a flexible titanium-oxide coated nitinol-mesh and double-disk design was selected. In consideration of the significant floppiness of the postero-inferior rim, an oversized by 8 mm occluder (33-mm FSO) and a modified implantation technique (pulmonary vein-assisted technique) was adopted. To delay the deployment of the left distal disc of the device, we started its placement in the right upper pulmonary vein (RUPV). When the left distal disc was uncovered in the RUPV, we held it stationary in an elongated form, allowing unsheathing of the occluder device so that the proximal disc would deploy in the RA, engaging the right aspect of the interatrial septum (IAS). A short wiggle of the delivery system released the left atrial disc from the RUPV position, engaging the IAS from the left aspect with a perfect configuration for ASD closure (Figures 3 and 4, Video 1). Being postero-inferior rim deficiency a well-known risk factor for device migration, a vigorous pull and push manoeuvre was therefore done to check the final device stability. Transoesophageal echocardiography investigation showed a perfect device alignment, with no mitral valve regurgitation or aortic impingement of the disc tips to the aortic root and the device was finally released successfully. Transthoracic echocardiography at discharge confirmed the stable position of the device without residual left-to-right shunt (Video 2). The patient was treated with aspirin 100 mg and clopidogrel 75 mg once daily for 6 months. At 6-month follow-up the patient remained symptom free. Transthoracic echocardiography confirmed the stable position of the device with no left-to-right shunt and marked reduction in right cardiac chambers volume overload (Figure 5).



**Figure 1** Two-dimensional transthoracic echocardiography (A) colour Doppler (B) apical four-chamber views showing a large atrial septal defect ranging from 27.1 to 28.2 mm in diameter with right heart chambers dilatation. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.



**Figure 2** (A,B) Two-dimensional transoesophageal echocardiography (TOE) colour Doppler images showing a large secundum atrial septal defect with severe left-to-right shunt, significant right atrial enlargement due to volume overload and adequate septal rims except for a deficient and very floppy postero-inferior one (yellow arrows); (C) three-dimensional (3D) transoesophageal echocardiography images demonstrating the circular index of the atrial septal defect defined as the ratio of the maximal diameter (26.1 mm) to the minimal diameter (21.7 mm) and the relationship with the surrounding structures. LA, left atrium; RA, right atrium.



**Figure 3** Fluoro-angiographic procedural steps. (A,B): after crossing the atrial septal defect, the delivery system (DS, arrowhead) with the exchange 0.035 inch–260 cm long wire are positioned in the right upper pulmonary vein (RUPV); (C,D) deployment of the uncovered distal disc in the RUPV holding it stationary in an elongated form (red dotted lines), allowing then unsheathing of the connecting waist (white arrow) and proximal right disc (black arrow) of the device in the right atrium, engaging the right aspect of the interatrial septum.

## Discussion

There is no universally accepted definition for large ASDs. Studies have variously described the lower cut-off levels for large ASDs in the diameter range 20–30 mm and an upper cut-off level of between 36 and 40 mm.<sup>6–8</sup> Indeed, main anatomical limitations to percutaneous ASD closure may be insufficient surrounding rims, multiple defects and excessively bulging ASAs.

Romanelli et al. demonstrated that the vast majority of large ASDs, defined as maximal ASD diameter in the range 20–39 mm, can be closed with transcatheter techniques with a very low complication rate. However, attempts at transcatheter closure of extremely large defects ( $\geq 40$  mm) are associated with a low success rate and the potential for complications.<sup>9</sup>

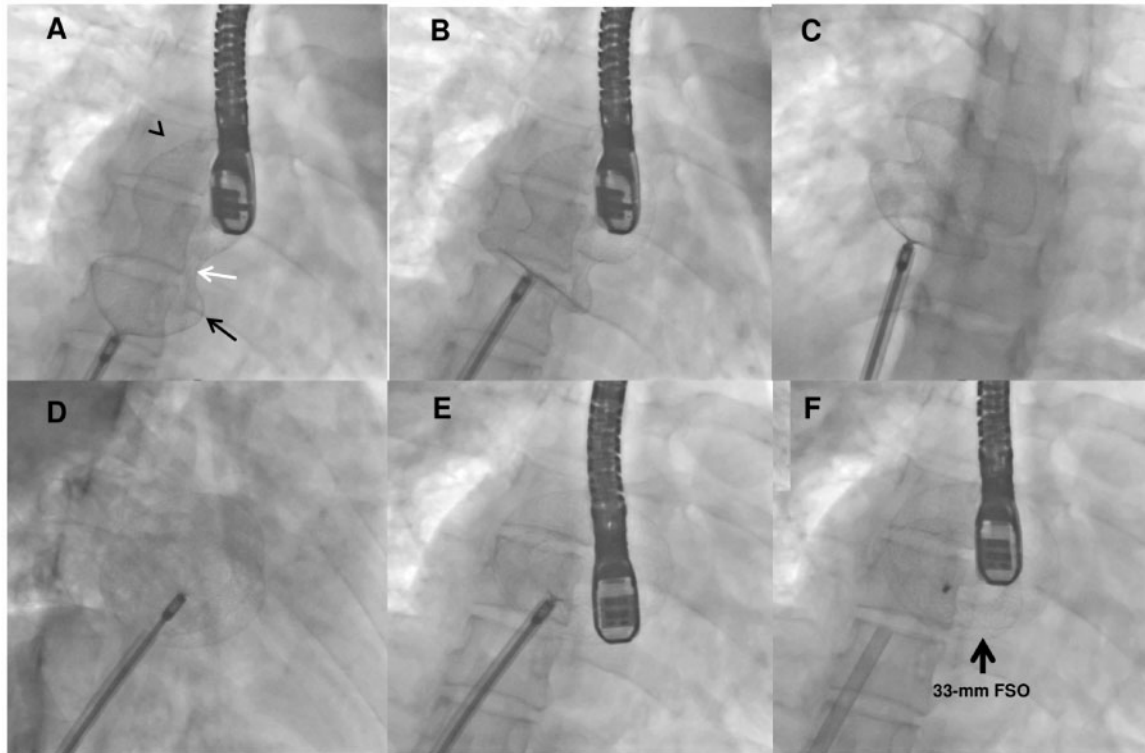
Recently, several studies have reported the feasibility of transcatheter closure in complex cases with a variety of modified implantation methods including the balloon-assisted technique.<sup>10</sup>

Lack of adequate aortic rim is often granted for success since it is well demonstrated that deficiency in the rim toward the aorta does not influence the success rate for transcatheter closure.<sup>11</sup> On the

contrary, deficiency of the postero-inferior rim was associated with failure or device embolization, because almost always the bigger distal left disc flips tangentially across the defect.<sup>12</sup>

The success of complex ASD closure mainly lies in the proper imaging techniques.<sup>13,14</sup> Secundum ASD is often oval or even crescentic in shape and measurements vary in different planes. A single dimension does not reflect the true ASD size. An oval ASD measuring  $>40$  mm in its long axis can be suitable for transcatheter closure as the diameter in the short axis is much smaller. Certainly, 3D echocardiography describing measurements such as the circular index of the ASD (defined as the ratio of the maximal diameter to the minimal diameter on a 3D-image) better clarify the indications for transcatheter closure.<sup>15</sup>

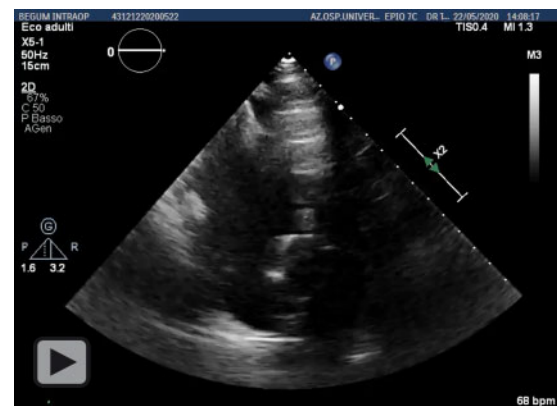
The Occlutech FSO has several innovative features compared to previous technologies: minimized metal contents especially in the distal left disc with no-clamping hub on it, which may provide more flexible and less traumatic feature. In addition, it has a distinct release mechanism resembling a biptome which enables flexible movement between the device and delivery cable, allowing device self-alignment resulting in a smooth conformability to the atrial septal contours.



**Figure 4** Fluoro-angiographic procedural steps. (A) a short wiggle of the delivery system released the left atrial disc (arrowhead) from right upper pulmonary vein position allowing proper left disc deployment; white arrow is pointing at the connecting waist and black arrow at the proximal right disc of the device; (C–E) engagement of the interatrial septum from the left and right atrial aspect with a perfect configuration for atrial septal defect closure; (F) 33-mm FSO (black arrow) finally deployed in a stable and correct position.

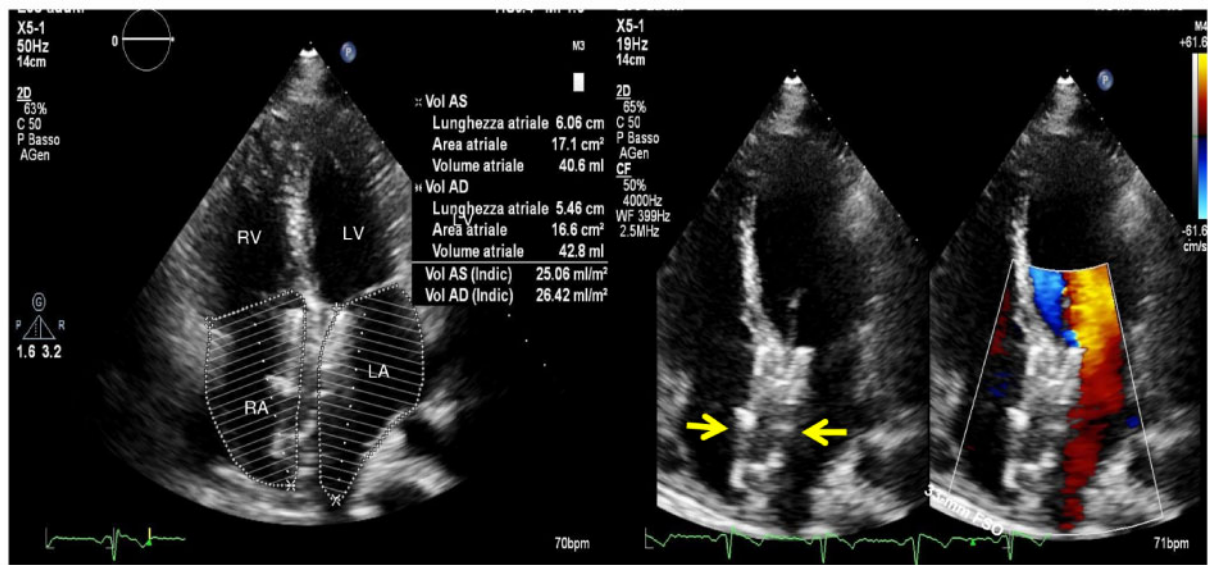


**Video 1** Fluoro-angiographic procedural steps under transoesophageal echocardiography monitoring. The uncovered distal left disc has been deployed in the right upper pulmonary vein (RUPV) holding it stationary in an elongated form allowing then unsheathing of the connecting waist and proximal disc of the occluder device in order to deploy the proximal right disc in the right atrium, engaging the right aspect of the interatrial septum. After a short wiggle of the delivery system the left atrial disc from the RUPV position engaged correctly the interatrial septum from the left aspect with a perfect configuration for ASD closure.



**Video 2** 2D transthoracic echocardiography apical four-chamber views transthoracic echocardiography at discharge confirmed the 33-mm FSO device in place.

• The pulmonary vein slide-out assisted technique offered substantial chances of success in this complex catheter-based closure procedure.



**Figure 5** Six-month follow-up 2D transthoracic echocardiography colour Doppler apical four-chamber views showing the 33-mm FSO *in situ* (yellow arrows) without residual left-to-right shunt and significant reduction in right heart chambers volume overload. LA, left atrium; LV, left ventricle; RA: right atrium; RV, right ventricle.

## Lead author biography



Dr Eustaquio Maria Onorato is an interventional adult and pediatric cardiologist particularly active in structural and complex congenital heart disease interventions. Senior Consultant, Congenital and Structural Heart Diseases Interventions at Centro Cardiologico Monzino, University School of Medicine, Milan, Italy. SCAI and ESC fellow. Honorary Membership of the Venezuelan and Russian Scientific Society of Interventional Cardiology.

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## Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

**Slide sets:** A fully edited slide set detailing these cases and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** E.M.O. is a consultant for Occlutech, manufacturer of the device. The remaining authors declare no conflict of interest to disclose.

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