Transcatheter Closure of Secundum Atrial Septal Defect with Large Multifenestrated Septum Primum Aneurysm and Double Atrial Septum: A Challenging Transesophageal Echocardiography–Guided Procedure

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INTRODUCTION

The feasibility of catheter-based closure for large atrial septal defects (ASDs) depends on anatomic criteria including the adequate length of the defect rims, the ASD size in relation to the total septal length, and the mobility of the multifenestrated atrial septal aneurysm (ASA). Double atrial septum is a rare cardiac anomaly caused by malposition of the septum primum, which fails to fuse with the septum secundum while the inferior vena cava is posteriorly placed and somehow overriding the interatrial septa, leaving a persistent space as a passage between the two atria and the inferior vena cava.[1-7] This anomaly represents a technical challenge for ASD device closure. We report the echocardiographic findings and outcome of a patient undergoing successful transcatheter correction of a peculiar atrial septal anomaly combining large multifenestrated ASA associated with an accessory interatrial chamber.

CASE REPORT

A 65-year-old male admitted for dyspnea and congestive heart failure underwent transthoracic echocardiography (TTE), which demonstrated enlarged right atrium, right ventricle, and a large multifenestrated ASA with severe left-to-right shunting (Figure 1, Video 1). More detailed anatomic definition of the defect was provided by two-dimensional/three dimensional (2D/3D) transesophageal echocardiography (TEE): a large multifenestrated secundum ASD (30 × 35 mm, area 9.7 cm^2) with highly mobile fibrous bands across the defect (Figure 2, Video 2) with adequate but partially redundant rims (aortic 14 mm, posterior 14 mm, superior 16 mm) and a very floppy inferior rim (13 mm). Furthermore, a double-membrane structure with an interatrial accessory chamber was seen in the bicaval view in the inferior portion of the atrial septum (Figures 3 and 4, Video 3).

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The patient underwent cardiac catheterization demonstrating a large left-to-right shunting (QP/QS ratio of 3.6) with increased mean pulmonary artery pressure (28 mm Hg) and normal pulmonary vascular resistance (1.86 Wood unit · m^2). Coronary arteries were normal.

Catheter-based closure of this peculiar ASD was deemed challenging. The patient was offered surgical closure but firmly rejected this option. Percutaneous closure was then performed under deep sedation, 2D/3D TEE color-flow Doppler monitoring, and fluoroscopic guidance. The device size was determined by the waist diameter of the inflated sizing balloon (30 mm; Figure 5). A third-generation 33 mm Figulla Flex II ASD occluder (FSO; Occlutech, Jena, Germany) with a flexible titanium oxide coated nitinol mesh and double-disc design was selected.

Inferior rim deficiency/floppiness has been described as a risk factor for device migration. Therefore, a modified balloon-assisted technique (BAT) to facilitate device closure of this large ASD was utilized. The BAT consists of using a balloon catheter to support the left atrial disc of the device during its deployment. The balloon support prevents prolapse of the left atrial disc into the right atrium (Video 4). A 33 mm FSO was advanced through the introducer sheath, and the left distal disc was opened into the left atrium after gently rotating the sheath clockwise. During deployment, the balloon was partially inflated into the defect to prevent left atrial disc prolapse into the right atrium. The introducer sheath and the device were pulled against the atrial septum until the complete apposition of the left disc on the septum was obtained. During the deployment of the connecting waist and the right atrial disc, the balloon was gradually deflated and gradually retrieved until the device was considered properly positioned. Moderate residual left-to-right shunting due to failed alignment of the device to the atrial septum was confirmed by color Doppler. A larger 36 mm FSO was used, but this did not prevent unstable inferoposterior anchoring of the device (Figure 6). Finally, the largest available 39 mm FSO was used, accomplishing correct device alignment to the defect without mitral interference or impingement of the discs to the aortic root. Color Doppler identified a trivial eccentric left-to-right shunt (Figure 7). A pull-and-push maneuver was performed to check occluder stability, and the device was finally released. Predischarge TTE confirmed stable position of the device with trivial residual left-to-right atrial shunting. One month later the patient was asymptomatic, and TTE did not demonstrate any residual shunt flow. Right atrium and right ventricle dimensions were markedly reduced (Figure 8).

DISCUSSION

Double atrial septum is a very rare atrial septal anomaly, with about 20 cases reported in the literature. An interatrial chamber is delineated by

VIDEO HIGHLIGHTS

Video 1: Transthoracic echocardiogram showing a multifenestrated ostium secundum ASD associated with a highly mobile aneurysmatic septum primum bulging toward the left atrium and the right atrium.

Video 2: Three-dimensional TEE of the multifenestrated interatrial septal defect (en face view from the left atrial perspective). The large ASD with a slightly elliptical shape is partially covered by aneurysmatic highly mobile fibrous bands.

Video 3: Two-dimensional TEE in the bicaval view showing a very large aneurysmatic multifenestrated fossa ovalis and a double-membrane structure, with an interatrial accessory chamber in the inferior portion of the atrial septum. A small Eustachian valve is also seen.

Video 4: Balloon-assisted technique under fluoroscopic and TEE guidance. With the inflated balloon in situ across the defect, the left atrial disc is deployed and the whole assembly is pulled back against the septum. The balloon supports and prevents prolapse of the left atrial disc into the right atrium and then allows the delivery of the right atrial disc outside of the sheath. Then after checking the discs on their respective sides of the septum by TEE, the operator deflates the balloon slowly, allowing the discs to expand and stent the septum followed by careful slow and steady withdrawal of the balloon.

View the video content online at www.cvcasejournal.com.

a parallel double-layer septal membrane.[8,9] This chamber is separated from the left atrium by the septum primum and from the right atrium by an additional septal structure, which is presumably the remnant of a persistent left venous valve of the embryologic sinus venosus.

This anomaly poses a significant challenge for ASD device closure, which needs stable attachment of left and right atrial discs on either side of the atrial septum. In our case, the device had to appose both the primum and secundum atrial septal structures to achieve a stable occluder device position.

Initial attempts at device deployment were unsatisfactory because the right atrial disc gripped only the septum primum, leading to rocking of the device, which was deemed unsatisfactory. Large oversizing of the third device plus a modified BAT allowed the right atrial disc to be deployed against the firm margins of the septum secundum while obliterating the space between the septum primum and septum secundum, without impingement on the surrounding structures.

In the FSO occluder device, structural metal components have been minimized (no clamping hub on the distal disc), making the device more flexible and less traumatic. In addition, the FSO has a distinct release mechanism resembling a bioptome, which enables flexible movement between the device and the delivery cable. This allows for device self-alignment and safe conformability to the atrial septal anatomy.

The procedure would not have been successful without precise definition of the unusual atrial septal anatomy by 2D/3D TEE, TEE procedural guidance, modified BAT technique to assist in holding the device in place, and the use of an adequate device.

CONCLUSION

The combination of secundum ASD with a large multifenestrated ASA and double atrial septum is a very rare anomaly and represents a technical challenge during catheter-based interventions. Keys for

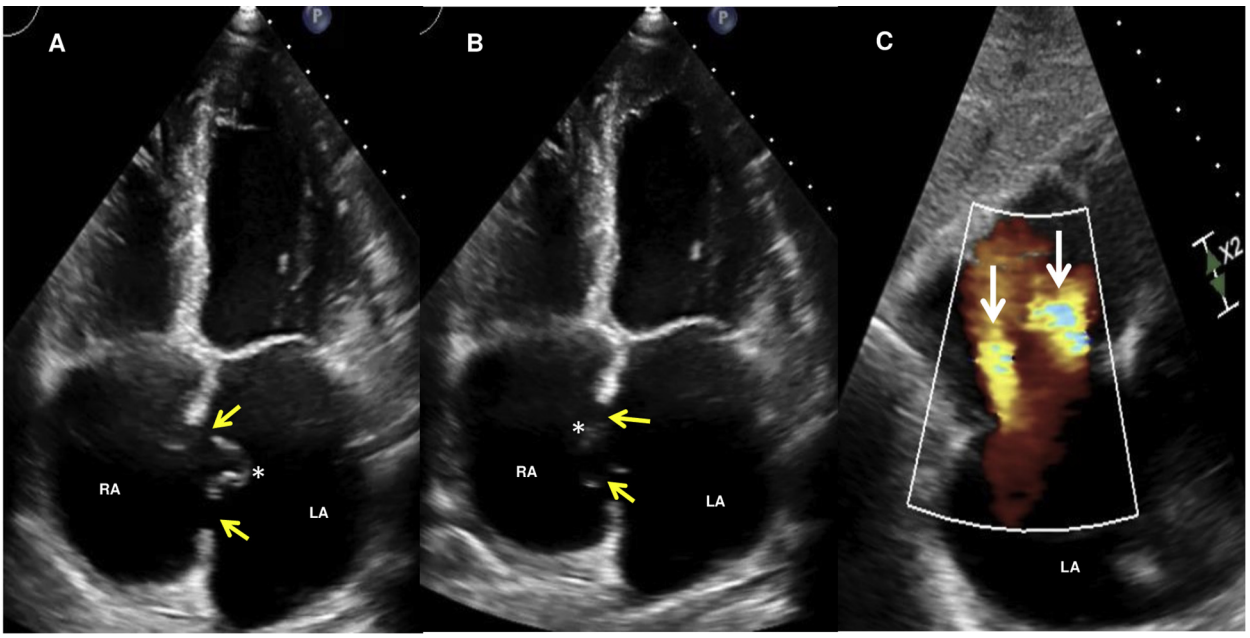

Figure 1 Transthoracic echocardiogram showing a multifenestrated (*yellow arrows*) ostium secundum ASD associated with a highly mobile aneurysmatic septum primum (*white star*) bulging toward the left atrium **(A)** and the right atrium **(B)**; TTE color Doppler in subcostal view **(C)** confirms the presence of the two defect fenestrations (*white arrows*). *LA*, Left atrium; *RA*, right atrium. See also Video 1.

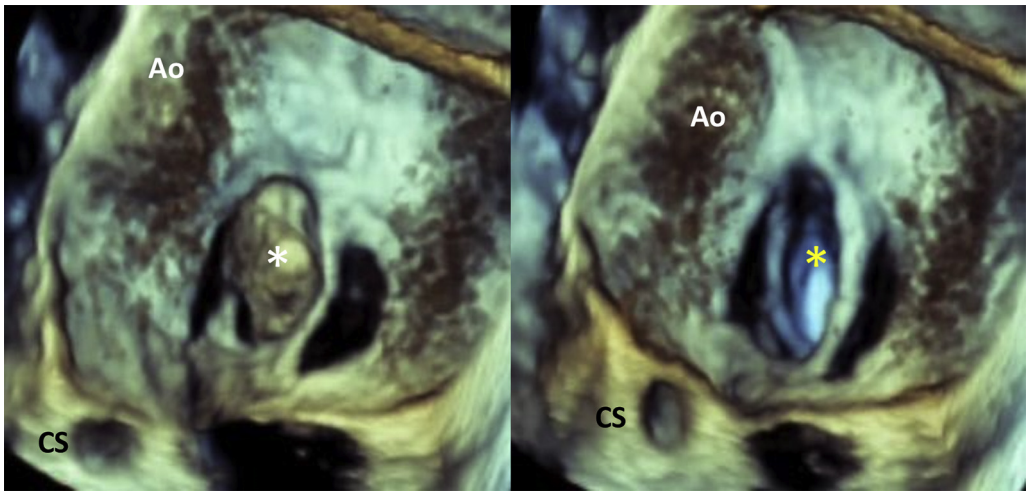

Figure 2 Three-dimensional TEE (en face view from the left atrial perspective) displays the multifenestrated interatrial septal defect. The large ASD with a slightly elliptical shape is partially covered by aneurysmatic highly mobile fibrous bands (*white and yellow asterisks* in two subsequent frames). *Ao*, Aortic root bulging on the left atrium; *CS*, coronary sinus. See also Video 2.

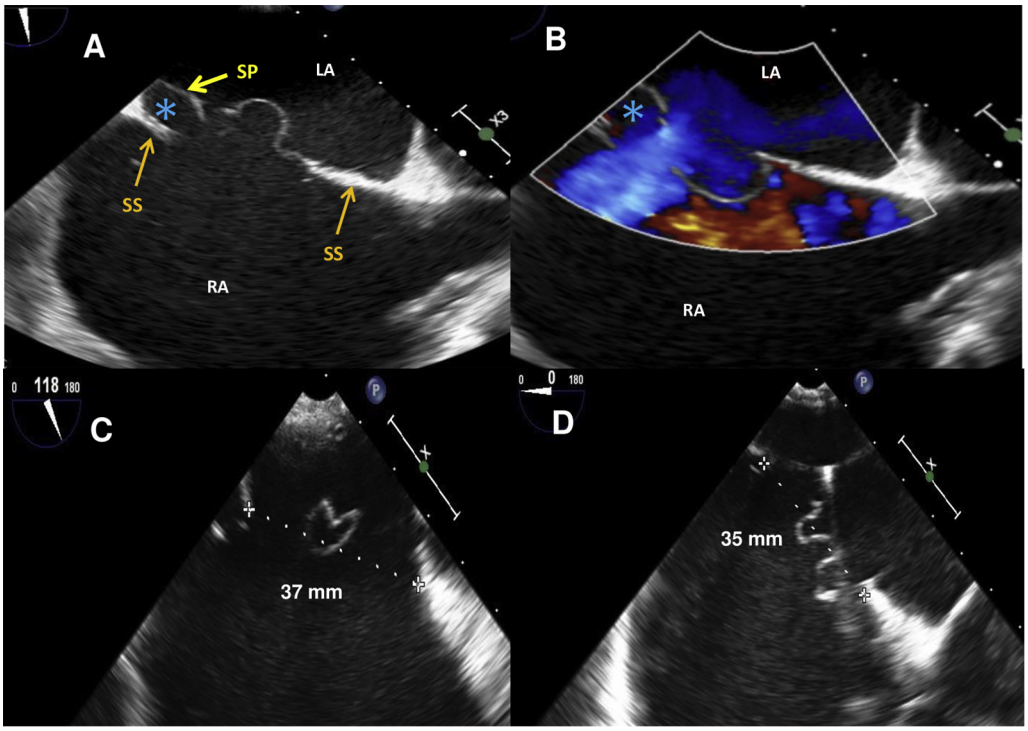

Figure 3 Two-dimensional TEE in the bicaval view showing the anomalous attachment of the septum primum, which fails to fuse with the septum secundum, leaving a persistent space (interatrial space) as a passage between the two atria **(A, B)**; the total length of the aneurysmatic fossa ovalis floor at 118° and 0° TEE views is also accurately measured **(C, D)**. *LA*, Left atrium; *RA*, right atrium; *SP*, septum primum; *SS*, septum secundum. See also Video 3.

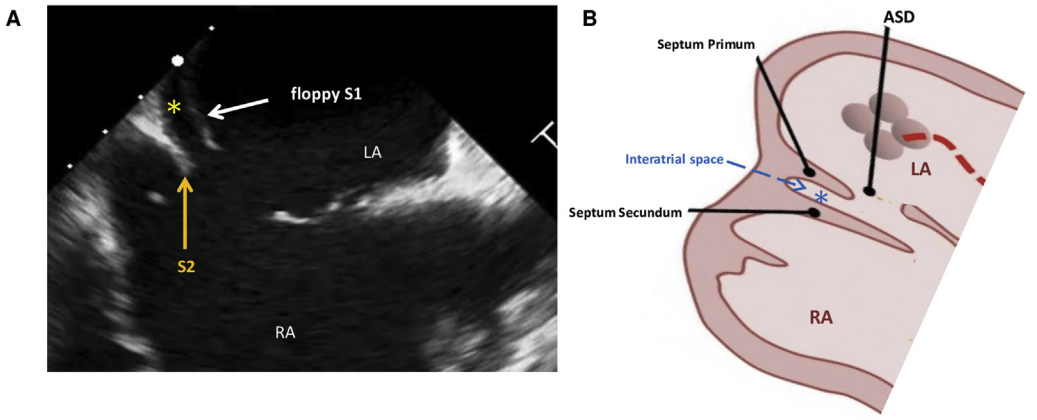

Figure 4 Two-dimensional TEE in the bicaval view image **(A)** and the corresponding drawing **(B)** showing a double-membrane structure with an interatrial accessory chamber (*yellow asterisk*) in the inferior portion of the atrial septum. This rare cardiac anomaly is caused by malposition of the septum primum, which fails to fuse with the septum secundum, leaving a persistent space as a passage between the two atria. *LA*, Left atrium; *RA*, right atrium; *S1*, septum primum; *S2*, septum secundum.

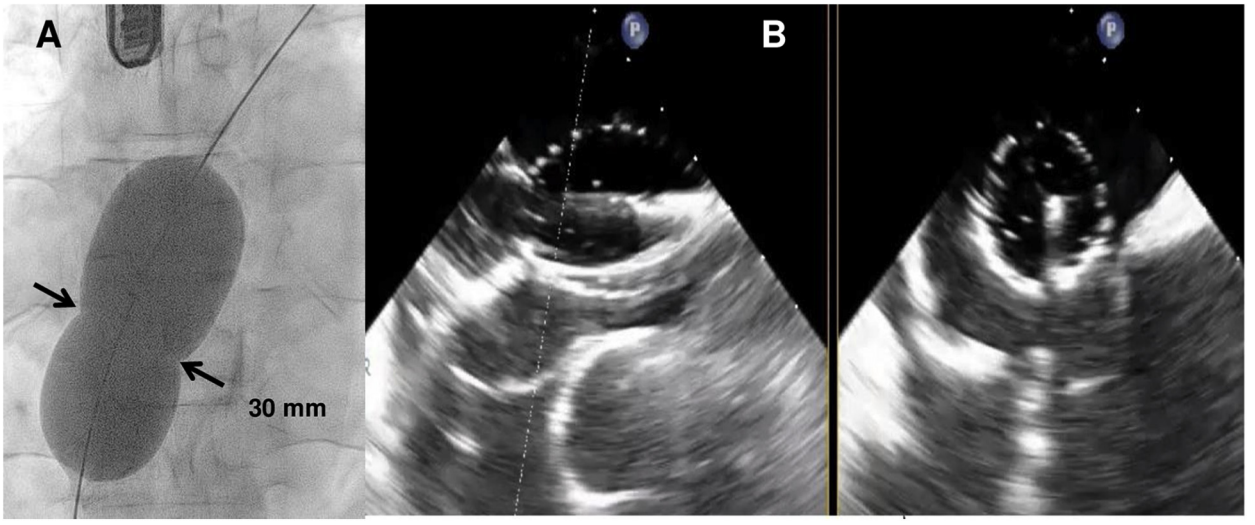

Figure 5 The balloon-stretched diameter (*black arrows*) shown in cardiac fluoroscopy in the straight anteroposterior projection **(A)** and in two-dimensional TEE multiplanar views **(B)**.

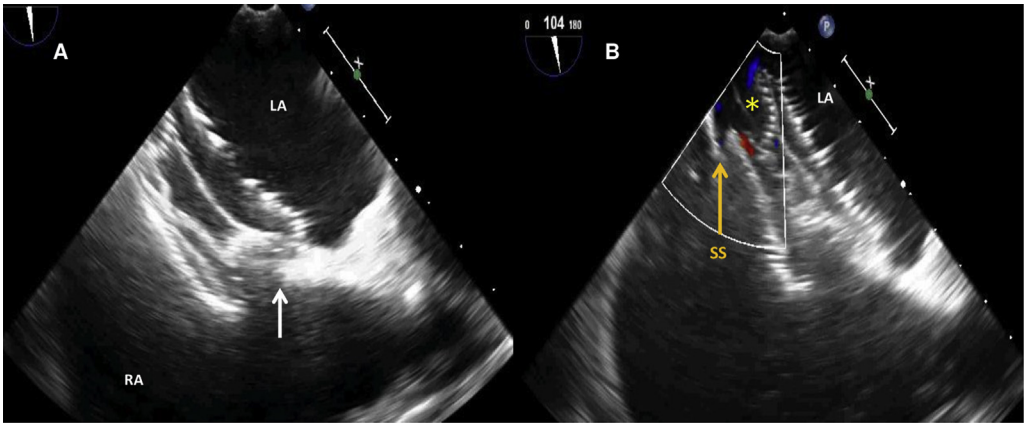

Figure 6 Two-dimensional TEE color Doppler **(A, B)** in the bicaval view showing good superior rim and unsafe inferoposterior rim anchoring of the 36 mm FSO (*white arrows*). Interatrial space, *yellow star*; *LA*, Left atrium; *RA*, right atrium; *SS*, septum secundum (*orange arrow*).

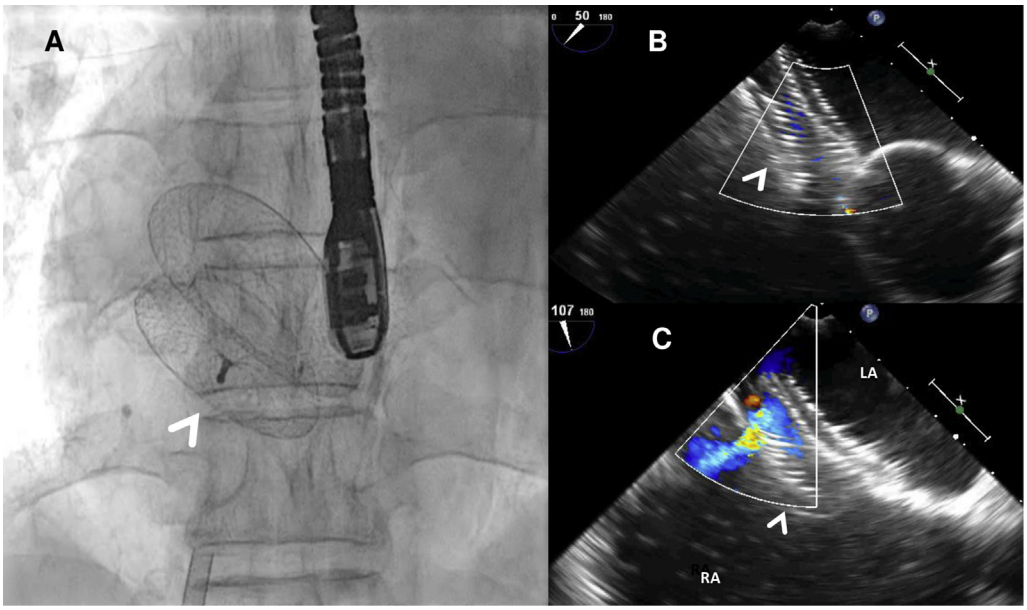

Figure 7 Cardiac fluoroscopy in anteroposterior projection **(A)** and two-dimensional TEE color Doppler images at 50° **(B)** and at 107° **(C)** showing the 39 mm FSO (*white arrowhead*) with better device alignment. The final result was successful superior rim anchoring without aortic impingement of the disc tips to the aortic root, and a trivial eccentric residual left-to-right shunt was located inferoposteriorly with color Doppler. *LA*, Left atrium; *RA*, right atrium.

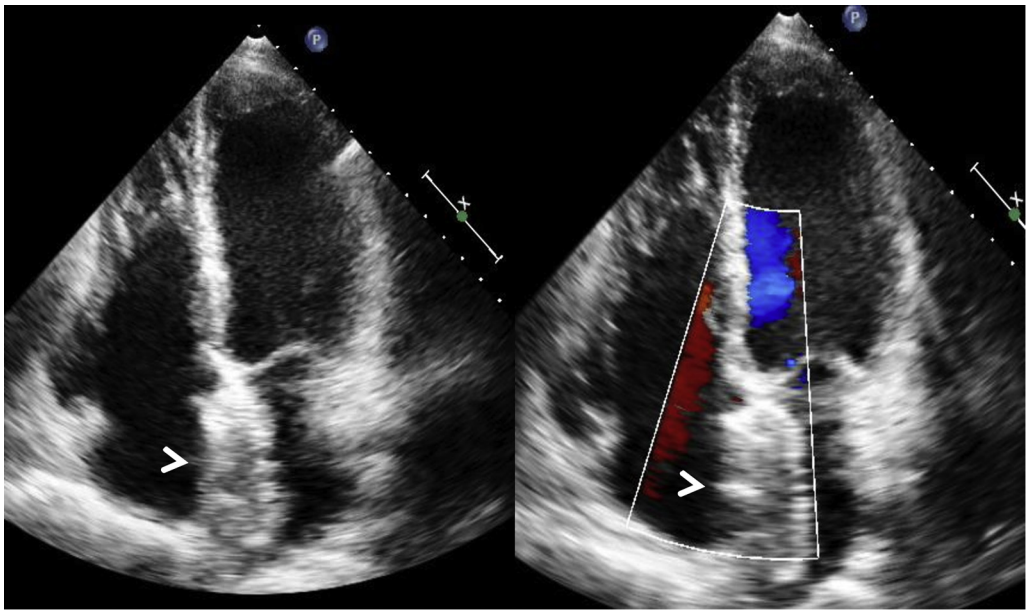

Figure 8 Thirty-day follow-up TTE in the apical four-chamber view showing a well-positioned occluder device (*white arrowhead*) without residual shunt and significant decrease of right heart overload.

success are comprehensive 2D/3D TEE assessment along with intraprocedural imaging guidance, modified BAT, and availability of an innovative cutting-edge device technology.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2021.01.005.

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