

Percutaneous closure of residual shunting in a patient with a fenestrated atrial septal defect occluder

A case report

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Abstract

Rationale: Fenestrated atrial septal defect (ASD) occlusion has been performed in patients complicated with severe pulmonary hypertension (PH). Nevertheless, the persistent interatrial residual shunting in the fenestration might increase the risk of paradoxical embolism. Percutaneous closure of fenestrated ASD occluder (ASO) has not yet been reported.

Patient concerns: A 26-year-old patient with a 25-mm ASD and severe PH underwent ASD closure using a Memory ASO with a waist of 32 and 6-mm custom-made fenestration. Echocardiography revealed the fenestration remained 6 mm and the pulmonary artery pressure decreased to the normal range at the 6-month follow-up.

Diagnoses: Persistent interatrial residual shunting in ASO.

Interventions: Percutaneous closure of residual interatrial shunting was performed using a waist of 8-mm ASO under guidance of TEE.

Outcomes: The fenestration was successfully closed. Neither thromboembolism nor infection events were noted during the 12-month follow-up after the procedure.

Lessons: This case illustrates that percutaneous closure of the residual shunting in fenestrated ASO was feasible and safe for short- and long-term.

Abbreviations: ASD = atrial septal defect, ASO = atrial septal defect occluder, PH = pulmonary hypertension.

Keywords: adult congenital heart disease, fenestrated device, occluder overlap

1. Introduction

Secundum-type atrial septal defect (ASD) is one of the most common types of congenital heart diseases in adulthood. Compared with the conventional surgical closure, percutaneous closure of ASD provides an effective, minimally invasive, and safe alternative for patients with ASD.^[1] Patients with ASDs and pulmonary hypertension (PH) might require atrial communication

as decompression in the event of PH crisis or left ventricular diastolic dysfunction.^[2,3] Partial occlusion with fenestrated ASD occluder (ASO) has been suggested to be beneficial in patients with severe PH, and it might be superior than complete ASD closure.^[2,4] However, the fenestration might remain patent in the long-term follow-up.^[5] The persistent residual shunting may increase the risk of paradoxical embolism. Herein, we report a case of a patient with successful deployment of the second ASO for closure of the residual shunting in the fenestrated ASO implanted previously.

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2. Case report

A 26-year-old female, recently diagnosed with secundum-type ASD accompanied by severe PH, was referred to our hospital for therapy. She presented a 1-year history of symptoms, including exertional chest distress and dyspnea. The second heart sound was prominent, and a grade 2/6 systolic murmur was heard at the left midsternal border during physical examination. Her functional class was assessed to be NYHA II. Any marked abnormality was not detected in the laboratory tests. Transthoracic echocardiography (TTE) revealed the presence of a moderately large secundum-type ASD, sized 25 mm. Both aortic and posterior left atrium rims were small (2 mm), while the other rims were sufficient. Also, a bidirectional interatrial shunt was observed, predominantly from left to right. In addition, the right chambers and pulmonary artery were dilated. The diameters of the right atrium and right ventricle end-diastolic were 45 and 51 mm, respectively. The right and left ventricular functions were normal. Tricuspid regurgitation was moderate-to-severe. The

pulmonary artery systolic pressure (PASP) gradient was 110 mm Hg as estimated by the Doppler method. Diagnostic catheterization revealed a significantly elevated mean pulmonary artery pressure (mPAP) of 55 mm Hg, and pulmonary vascular resistance index (PVRI) of 4 Wood/U/m².

The patient provided written consent and then underwent sufficient preoperative preparation, including the administration of Remodulin for reducing the pulmonary arterial pressure and oxygen inhalation. Considering the severe PH, Memory ASD (Shape Memory Alloy Co., Ltd, Shanghai, China) with a waist of 32 and 6-mm custom-made fenestration (Fig. 1A) was subsequently inserted by standard ASD closure technique under the guidance of intraoperative transesophageal echocardiography (TEE). Consequently, the interatrial shunt was successfully diminished. A mild bidirectional shunt through the fenestration could be detected by TEE; the right and left ventricular functions remained stable. After the procedure, the patient was treated with Remodulin and aspirin, and then, discharged on day 3, as the recovery was uneventful. However, Aspirin was prescribed for 3 months.

The patient's symptoms improved significantly as noted at the 3-month follow-up. TTE evaluation demonstrated excellent ASD position and a residual left-to-right shunt across the fenestration. PASP decreased to 71 mm Hg. Tricuspid regurgitation improved from mild-to-moderate degree. The enlarged right atrium and ventricle also decreased in size (from 45 to 40 mm and from 51 to 44 mm, respectively).

Half a year after the procedure, TTE revealed normal size of the right heart chambers. A further decrease in PASP was also observed, up to 34 mm Hg. The residual shunt did not exert a significant influence according to hemodynamics. Considering the possibility of paradoxical embolism and patient's concern, complete closure of residual atrial shunting was performed under general anesthesia and guidance of TEE, which estimated the diameter of fenestration in the device as 6 mm. The guidewire moved from inferior vena cava to right atrium, traversing the ASD fenestration, and lodged into the left atrium. Venous access was obtained with 8F sheath via the wire. After the withdrawal of the wire, a standard ASD (Yachuang Biotech Co., Ltd, Xuzhou, China) with 8 mm waist was delivered from the sheath. The left and right discs were successively deployed, and the small ASD overlapped the previously large discs on both sides (Fig. 2). The position relationships among the sheath, fenestration, and the center hub of the device could be visually displayed by 3-dimensional (3D) TEE (Fig. 1B). The patient was treated by Aspirin after the second procedure. Pre-discharge 24-hour TTE revealed the stationary position of the 2 devices. No interatrial residual shunt was observed. In addition, no thrombi or vegetation was detected on the devices.

Aspirin was prescribed for 3 months after discharge. No thromboembolism or infection events occurred during the 12-month follow-up after the second closure.

The patient consented to publish this case report, which was also approved by the Institutional Review Board of Shanghai Jiao Tong University Affiliated Sixth People's Hospital, Shanghai, China.

3. Discussion

Complete closure of the ASD might aggravate the right heart overload in patients with significant PH. A patent foramen ovale (PFO) allowing a pop-off to the left heart may ameliorate the effect of increased overload on the right ventricular function.^[6]

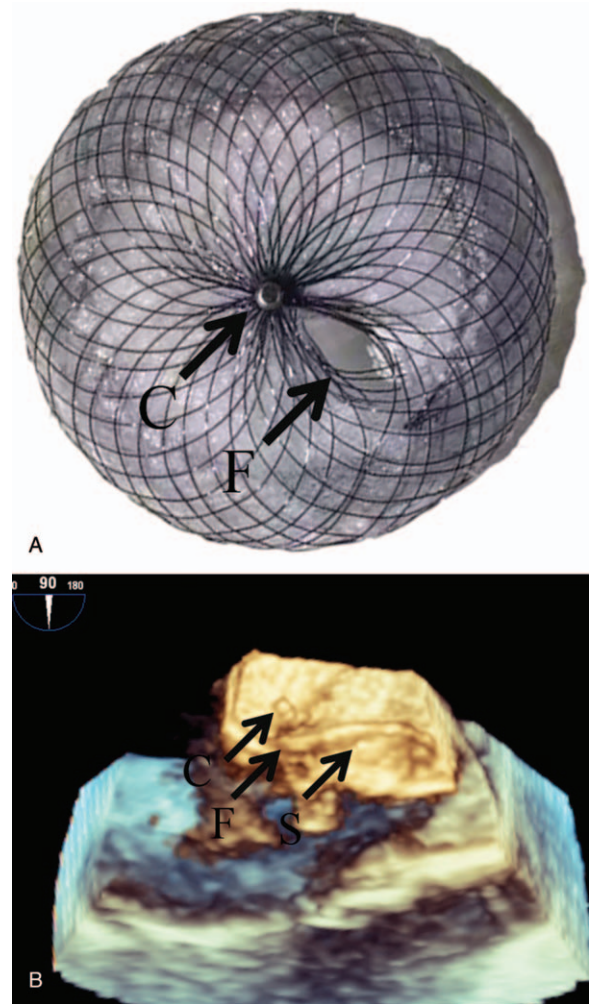


Figure 1. Actual Memory ASD fenestrated device and 3-dimensional transesophageal echocardiography (TEE) imaging: (A) Fenestration next to the center hub (arrows), (B) Position relationship among center hub fenestration and the sheath (arrows). C=center hub, F=fenestration, S=sheath.

Similarly, the fenestration in ASD could allow a blood overflow in the right-to-left direction as a reduction of right heart overextension in such patients. Therefore, the transcatheter-fenestrated ASD closure is considered to be advantageous to patients with severe PH. In order to maintain the interatrial communication patent, fenestrated ASDs were manufactured in various ways,^[2,4,5,7-9] which have been shown to be safe and associated with improvements in functional status and pulmonary arterial pressure.^[5] Despite that a few ASD fenestrations may close spontaneously after implantation during follow-up, most of the interatrial communications remained open over a prolonged duration in previous cases.^[2,4,7-10]

Nevertheless, the longstanding fenestration may not be necessary later when the severe PH and right heart overload is improved. Moreover, as the residual shunting across the fenestration may cause a paradoxical embolism. Thus, complete closure of the ASD fenestration should be beneficial to the patients, especially to those with a high risk of peripheral venous thrombosis, such as pregnant women. Venous thrombosis can be secondary to a prothrombotic state in the hemostatic system associated with pregnancy. Compared with the nonpregnant

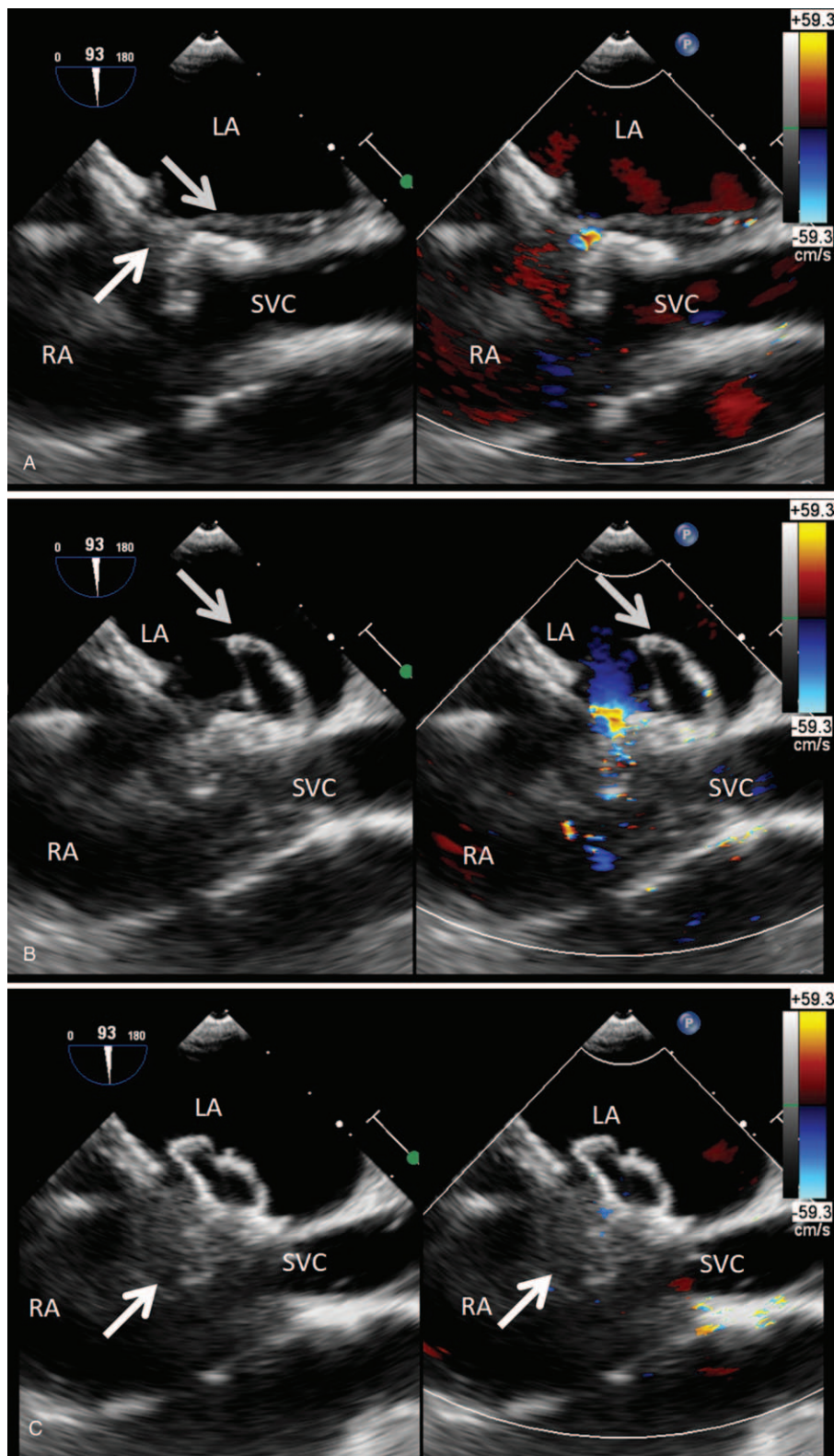


Figure 2. Color compared transesophageal echocardiography (TEE) guidance showing the procedural steps of the fenestration closure: (A) Sheath across the fenestration (arrows), (B) Deployment of the left disc (arrow), (C) Deployment of the right disc (arrow). LA=left atrium, RA=right atrium, SVC=superior vena cava.

state, the risk of venous thromboembolism was 5-fold higher in pregnant cohort,^[11] which significantly increased the risk of paradoxical embolism through atrial communication. A paradoxical embolism can lead to stroke, which is a catastrophic event

in young patients. Considering that the current patient was of childbearing age, facing an increased risk of paradoxical embolism if pregnant, the fenestration was completely closed in the absence of the need for decompression in the right heart.

To the best of our knowledge, this is the first report describing the percutaneous closure of the residual shunting in the fenestrated ASO. The complete closure was performed 6 months after the fenestrated ASO implantation. Both Doppler-estimated pulmonary arterial pressure and size of the right heart chambers were within the normal range according to TTE. Nevertheless, the improvement of PH was observed during >6-month follow-up in a majority of the previous cases.^[4,5,7] Thus, we speculated that the selection of complete closure time might be varied according to the follow-up results.

Different from the atrial septum tissue, the fenestrated device is stiff and thick. The reduced operation space due to the large device in the right atrial as well as the small fenestration would increase the difficulty while inserting the guidewire and sheath from the inferior vena cava to the fenestration. The wire was inserted across the fenestration after several attempts. As a result, the second occluder was implanted successfully under TEE guidance instead of radiation exposure. Two-dimensional (2D) TEE combined with real-time 3D TEE imaging can demonstrate the structures of fenestration and occluder or sheath in an enhanced manner.

Although the size of the second ASO was much smaller than the first, the second prescription of postoperative oral Aspirin was similar to that of the first ASD closure. This phenomenon might be attributed to the difficulty of the tissue encroachment to the second ASO, as the small ASO directly contacts with the previous device instead of the cardiac structures. Some authors proposed that incomplete endothelialization was a mechanism of endocarditis after ASO implantation.^[12] However, statistical data on endothelialization of 2 devices in such an overlapping state, as well as the effects on platelet aggregation are yet lacking. In this case, the patient did not display any events during the 12-month follow-up after the second operation; no thromboembolism or infections occurred. Furthermore, the safety of this procedure needs to be further validated in an additional number of cases and studies.

4. Conclusion

This case report illustrated that minimal invasive percutaneous complete closure of the fenestration in ASO under TEE guidance rather than radiation exposure is feasible and can serve as a therapeutic option in patients with severe PH, who were treated with fenestrated ASOs. Complete closure of the residual atrial shunting in the fenestrated ASO should have the advantage of

reversing the heart hemodynamics to the normal condition and avoiding potential paradoxical embolism.

Author contributions

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