Case Report

Transcatheter Treatment of Sinus Venosus Atrial Septal Defect With Pulmonary Hypertension: A Case Report

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ABSTRACT

Sinus venosus atrial septal defects (SVASDs) account for less than 10% of all atrial septal defects (ASDs). These defects are characterized by a lack of a wall separating the superior vena cava (SVC) from the right upper pulmonary vein, leading to a left-to-right shunt at a high atrial level (partial anomalous pulmonary venous drainage). Historically, all SVASDs required surgical treatment. Nonetheless, in the last decade, the SVC type of SVASD, which is amenable to device closure, can be closed via percutaneous angiography. SVC-type SVASDs result in a left-to-right shunt, progressively leading to right ventricular enlargement, tricuspid valve insufficiency, and pulmonary hypertension. Patients typically present with symptoms such as exertional dyspnea, palpitations, and atypical chest pain. Most commonly, SVASDs are associated with the SVC, where blood from the right upper and/or middle pulmonary veins drains into the SVC or the right atrium.

This report describes the successful transcatheter treatment of an SVC-type SVASD with pulmonary hypertension in a 59-year-old patient who presented with atypical chest pain and progressive dyspnea with moderate-to-severe activity. ***(Iranian Heart Journal 2025; 26(4): 89-97)***

KEYWORDS: Sinus venosus atrial septal defect; Partially abnormal pulmonary venous drainage; Device closure; Pulmonary hypertension

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Received: April 22, 2025 **Accepted:** July 9, 2025

Sinus venosus atrial septal defects (SVASDs) account for approximately 4% to 11% of all ASDs. [1] Surgical closure of SVASDs is the standard management. A safe and effective transcatheter approach would be an attractive option. From July 2011 through October 2013, four patients with large SVASDs and anomalous right upper pulmonary venous (RUPV) drainage underwent transcatheter closure of their defects at Ibn-Albitar Center for Cardiac Surgery, Baghdad, Iraq. [2]

This anomaly is characterized by a defect in the atrial septum at the junction of the superior vena cava (SVC) and the right atrium. Blood may shunt from systemic to venous circulation and is often accompanied by anomalous pulmonary venous return. [3,4] Patients with SVASD likely present with exercise intolerance, palpitations, or syncope. Owing to its rarity and the potential overlap

of symptoms with other heart diseases, diagnosis can be challenging. [1, 3, 4]

Generally, transthoracic echocardiography and transesophageal echocardiography are used to visualize the defect and assess right heart size to diagnose SVASD. Further, cardiac magnetic resonance imaging or computed tomography (CT) angiography is important to evaluate the anatomy of the SVC and partial anomalous pulmonary venous connections (PAPVCs) and to determine the suitability of the ASD for percutaneous closure. [3, 5] In general, most centers use CT angiography rather than magnetic resonance imaging.

Surgical repair is the standard treatment for SVASD worldwide. Surgical outcomes are typically favorable, with low mortality and significant symptom improvement reported in many cases. [1, 4]

Recent advances have led to the emergence of transcatheter techniques as a less invasive alternative. These approaches involve the placement of covered stents in the SVC to redirect blood flow. Preliminary studies suggest that transcatheter repair may be suitable for some patients. [5, 6]

CASE PRESENTATION

A 59-year-old woman presented with atypical chest pain and progressive dyspnea with moderate-to-severe exertion. Transthoracic echocardiography showed right atrial and right ventricular enlargement and mild-to-moderate tricuspid regurgitation, with a systolic pulmonary artery pressure of 41 mm Hg. Contrast echocardiography revealed a left-to-right shunt. Transesophageal echocardiography demonstrated an SVC-type ASD, 21 mm, with anomalous drainage of the RUPV to the SVC. Subsequently, CT angiography was performed for further evaluation of device closure feasibility and showed that the RUPV and the middle pulmonary vein drained into the junction of the SVC and the right atrium via a common trunk; the anatomy of the defect was suitable for device closure.

The procedure entailed closure of the SVASD with an Optimus CVS PTFE–covered stent (XXL, 48 mm) and an Optimus CVS PTFE–covered stent (XL, 57 mm), followed by postdilation with a balloon-in-balloon (BIB) balloon (10 × 40 mm), a VACS II balloon (30 × 40 mm), and an AltoSa-XL Gemini balloon (30 × 60 mm), without complications (Figures 4 and 10). CT angiography images obtained before and 48 hours after the procedure are presented in Figures 4 and 5. Therefore, ASD closure with two covered stents was successfully performed.

METHODS

On initial examination, the patient's ECG showed sinus rhythm with a right bundle-branch block pattern and right-axis deviation (Figure 1). A posteroanterior chest radiograph showed cardiomegaly and a uniform increase in the pulmonary vascular pattern with a prominent pulmonary hilum (Figure 2). Echocardiography showed moderate-to-severe right ventricular enlargement with normal function, right atrial enlargement, moderate mitral regurgitation, moderate aortic regurgitation, and moderate tricuspid regurgitation, with mild pulmonary hypertension and a systolic pulmonary artery pressure of 41 mm Hg (Figure 3A and 3B).

CT angiography was performed to assess the anatomy and size of the SVC and PAPVCs. The images illustrated a normal-sized, unobstructed right SVC and demonstrated that the RUPV and right middle pulmonary vein (RMPV) drained into the distal SVC through a common trunk (Figure 4A & B, Figure 4C & D, and Figure 4E & F).

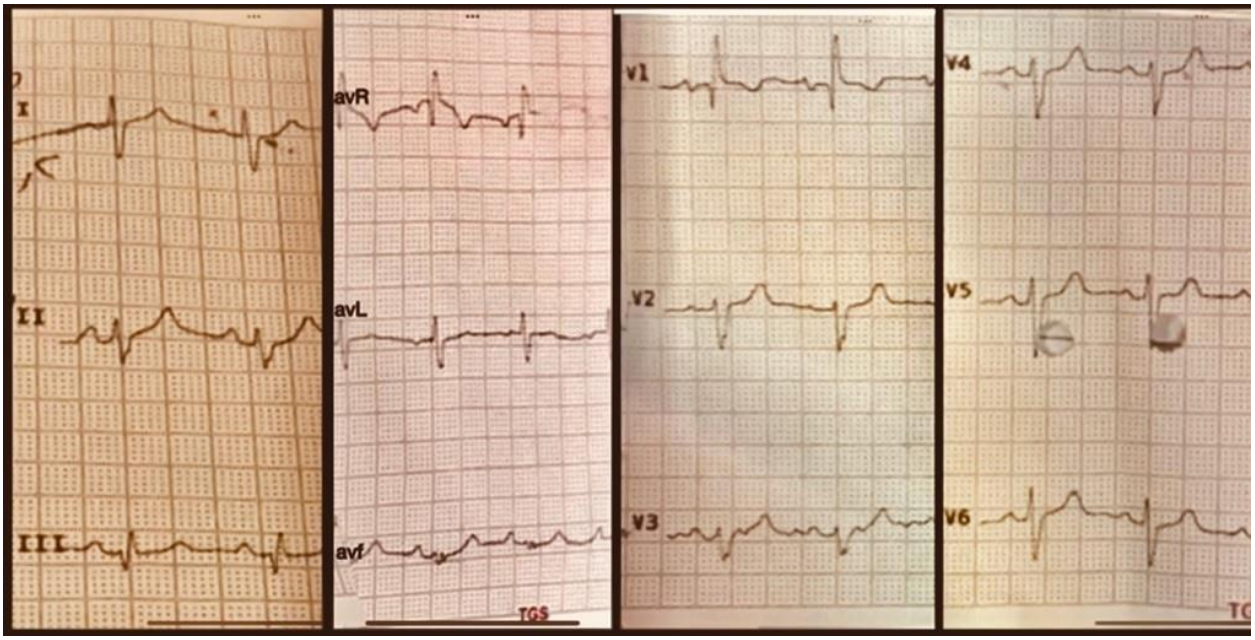

Figure 1. The patient's ECG shows sinus rhythm with a right bundle branch block pattern and right-axis deviation.

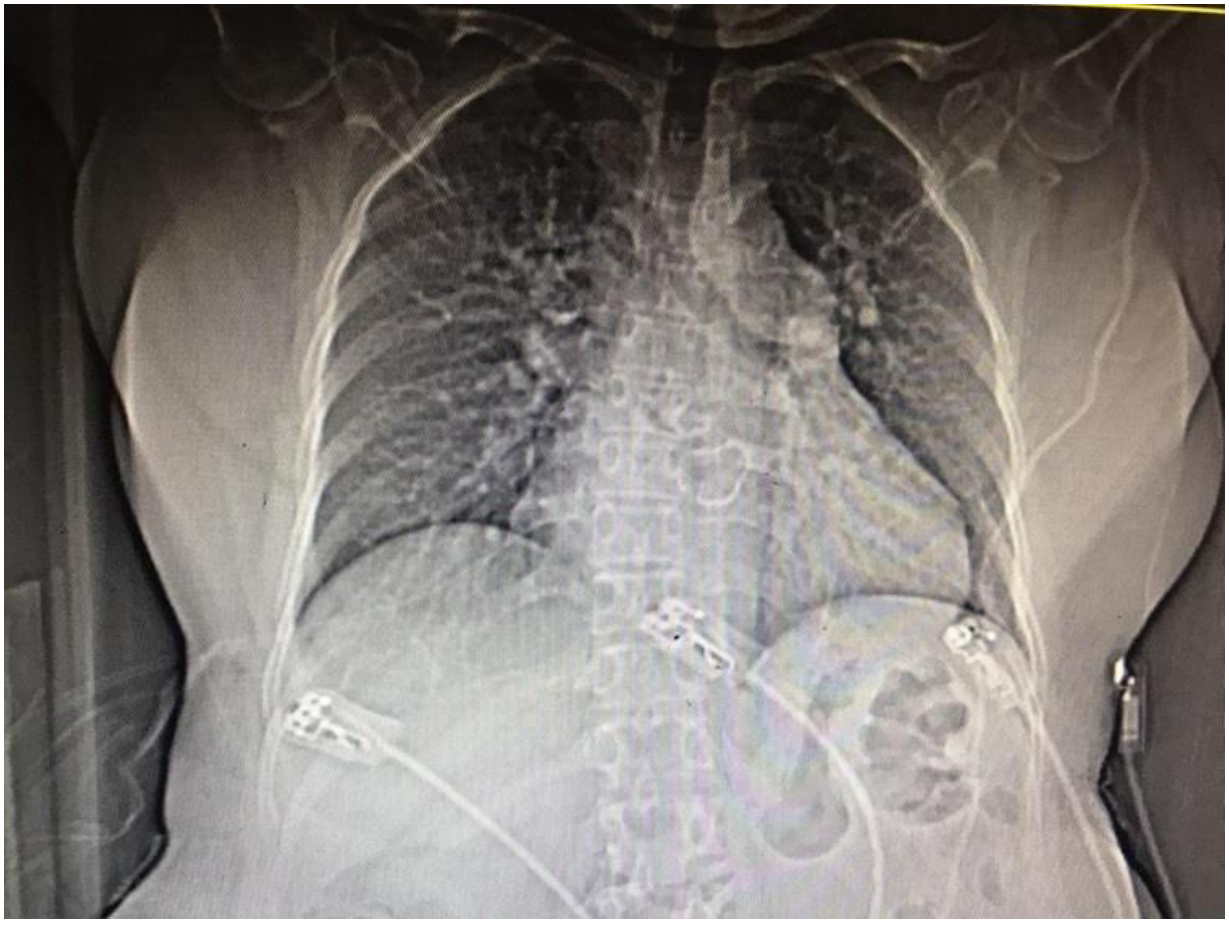

Figure 2. The patient's chest radiograph shows cardiomegaly and a uniform increase in the pulmonary vascular pattern with a prominent pulmonary hilum, consistent with shunt vascularity.

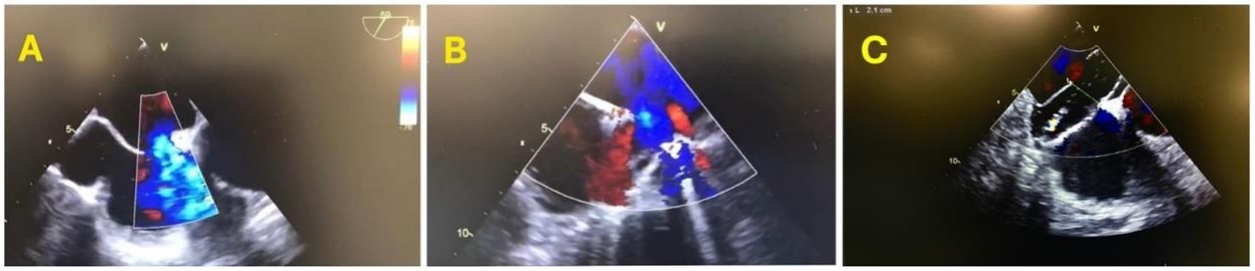

Figure 3. A and B) Transesophageal echocardiography shows an SVC-type SVASD with a left-to-right shunt and abnormal drainage of the right upper pulmonary venous to the SVC. (C) Full balloon inflation with a waist of 21 mm to stop flow from the partial anomalous pulmonary venous connections to the right atrium, without significant compression of the abnormal pulmonary veins.

SVC: superior vena cava, SVASD: sinus venosus atrial septal defect

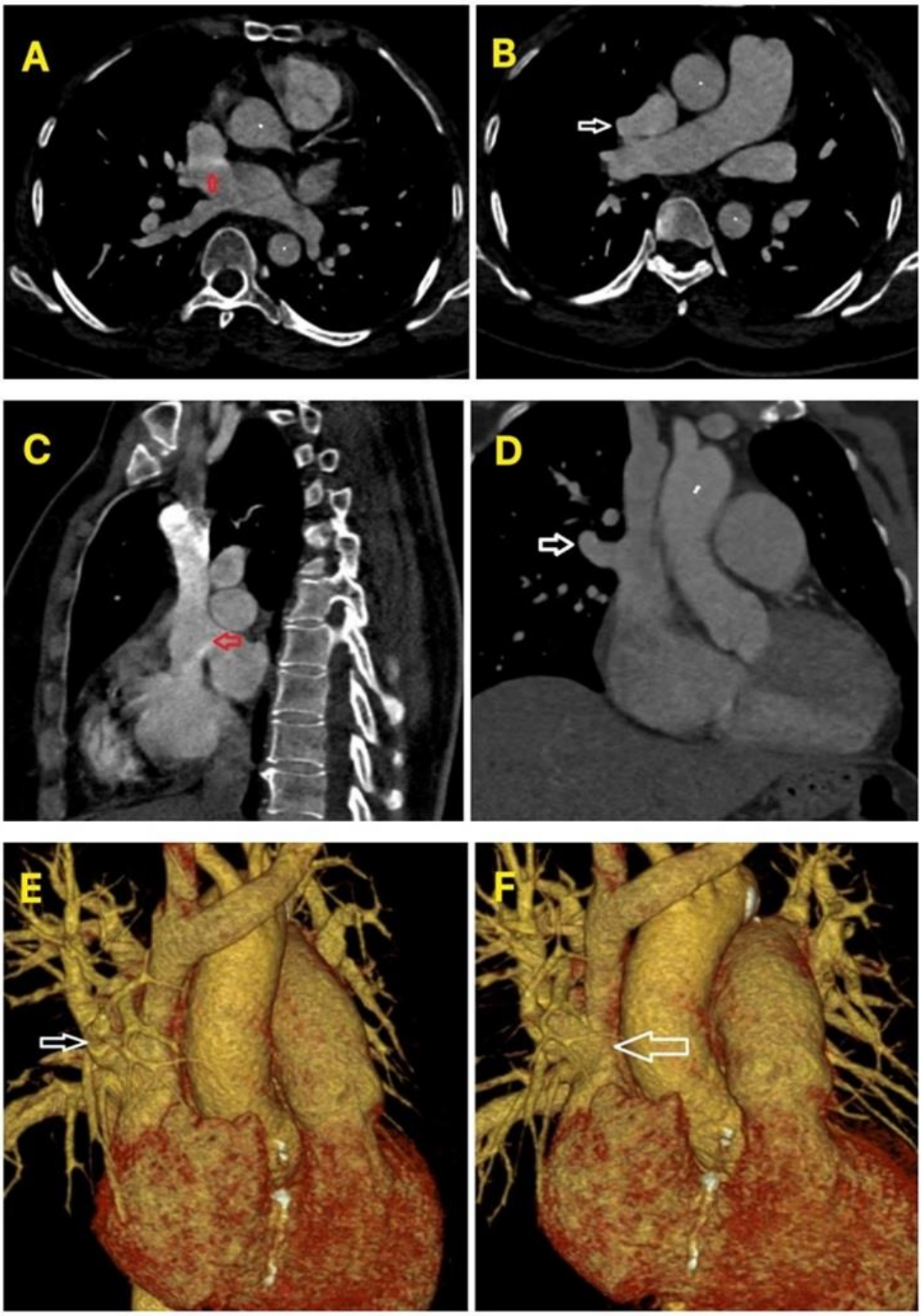

Figure 4. (A and B) Axial images reveal a sinus venosus atrial septal defect (red arrow) and a partial anomalous pulmonary venous connection (white arrow). (C and D) Multiplanar reconstruction images show the sinus venosus atrial septal defect (red arrow) and the partial anomalous pulmonary venous connection (white arrow). (E and F) Volume-rendered technique (images demonstrate the location of the stent and the partial anomalous pulmonary venous connection.

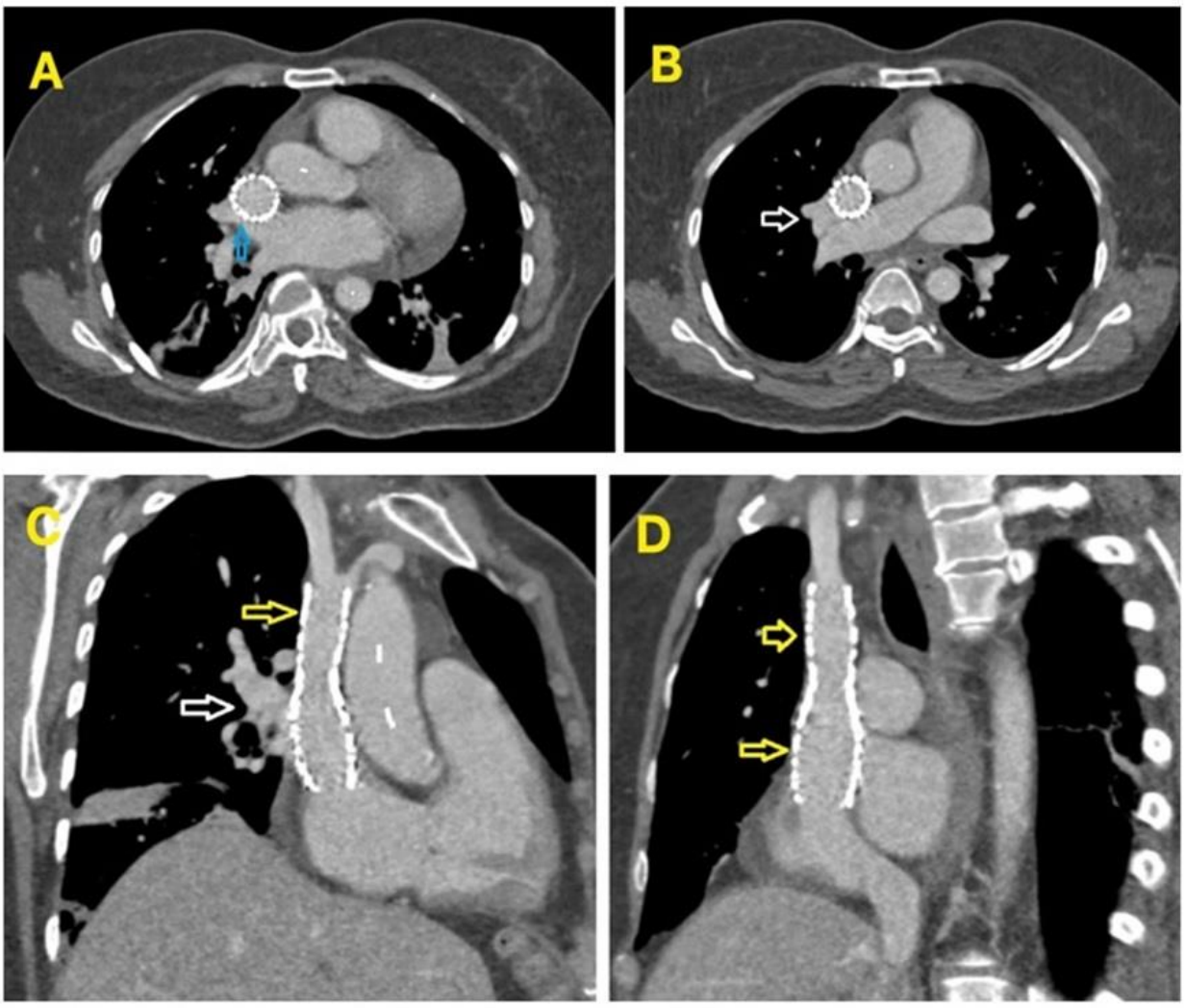

Figure 5. (A and B) The new drainage pathway of the partial anomalous pulmonary venous connection behind the stent into the left atrium is indicated (blue arrow). (C and D) Axial images show the stent and its relationship to the partial anomalous pulmonary venous connection and the sinus venosus atrial septal defect.

In addition, the findings indicated that the anatomy was suitable for device closure and that there was no evidence of pulmonary disease or chronic thromboembolic pulmonary hypertension. Given the SVASD with PAPVC and resultant right ventricular enlargement, the patient had an indication for ASD closure and repair. Nonetheless, due to pulmonary hypertension and concomitant mitral and aortic valve insufficiency, right and left heart catheterization was first performed to evaluate pulmonary artery pressure, the cause of pulmonary hypertension, pulmonary artery wedge pressure, and the coronary arteries. Coronary angiography via the right radial artery showed normal, patent coronary arteries. Right heart catheterization via the right femoral vein yielded the following values: systolic pulmonary artery pressure: 55 mm Hg; pulmonary vascular resistance: 2.7 Wood units; pulmonary artery wedge pressure: 8 mm Hg; systemic oxygen saturation: 96%; and pulmonary-to-systemic flow ratio (Qp:Qs): 2.4.

Based on the data from right heart catheterization and precapillary mild

pulmonary hypertension, the decision was made to perform transcatheter closure with careful monitoring in the postprocedural phase. Under mild sedation and simultaneous transesophageal echocardiography guidance, the patient underwent device closure of the SVASD with partial anomalous pulmonary venous drainage via percutaneous angiography.

Procedure Description

The procedure was performed using a Cheatham-Platinum-covered stent. Initial angiography of the RUPV was performed with and without balloon inflation in the SVC to confirm drainage of the common trunk of the RUPV and the RMPV into the left atrium. Angiography of the SVC in the frontal projection was employed to measure its diameter, particularly at the entrance to the right atrium. An appropriately sized balloon catheter was advanced into the SVC and inflated, with its inferior margin protruding into the right atrium, until flow through the defect ceased. This step, performed under transesophageal echocardiography guidance, confirmed separation between the SVC and the common venous trunk (Figure 3A, B). Simultaneously, a multipurpose catheter was advanced from the left femoral vein through the defect into the RUPV. Angiograms in a four-chamber view before and after balloon inflation confirmed persistent contrast drainage into the left atrium. This was further verified through the injection of agitated saline into the RUPV under transesophageal echocardiography guidance, which showed passage of bubbles into the left atrium without residual shunt. A cine-fluoroscopic image with the balloon inflated served as a landmark for subsequent stent implantation.

The covered stent was manually positioned on a BIB catheter. A long 14F delivery sheath was advanced over a 0.035-inch rigid guidewire into the SVC. The stent was deployed under combined fluoroscopic and transesophageal echocardiography guidance. Because the initial stent position was suboptimal, with a persistent shunt, an additional Cheatham-Platinum-covered stent was placed overlapping the cranial end of the first stent. Repeat angiography confirmed optimal stent placement, achieving complete separation of the common trunk of the RUPV and RMPV from the SVC.

Transthoracic echocardiography performed the following day showed no complications. A follow-up CT scan 48 hours after the procedure demonstrated no pulmonary vein thrombosis (Figure 5). The stents were correctly positioned, and the new pathway demonstrated normalized drainage without evidence of pulmonary vein stenosis or obstruction.

The patient was discharged on a regimen of aspirin (80 mg) for 6 months and clopidogrel (75 mg daily) for 3 months. During follow-up examinations at 6 months and again 3 months after aspirin discontinuation, there was no evidence of thrombosis, endocarditis, stent migration, or shunt enlargement. The patient reported a reduction in exertional dyspnea (New York Heart Association functional class I). Echocardiography confirmed improvement, showing a decrease in right ventricular size, mild-to-moderate tricuspid regurgitation, and a systolic pulmonary artery pressure of 31 mm Hg.

Sagar et al. [5] reported a procedural success rate of 97% for transcatheter SVASD closure. The use of long, premounted, covered stents has simplified the procedure. Pulmonary vein protection and the inclusion of patients with additional high-draining veins have now broadened the applicability of this technique.

In our patient, residual shunts observed during follow-up were clinically insignificant and decreased over time.

DISCUSSION

SVC-type SVASD closure, often associated with PAPVC, can be successfully performed non-surgically with percutaneous angiography if the SVC and PAPVC anatomy are suitable. The procedure is facilitated by using a suture technique with premounted long Cheatham-Platinum stents. A critical aspect during the procedure is to ensure that there is no pressure on the anomalous pulmonary vein and that it drains completely into the left atrium. The presence of a trivial residual shunt is not considered clinically significant.

Clinicians and researchers are increasingly exploring less invasive alternatives for treating congenital heart defects and diseases. For instance, covered stent correction for SVASD was initially regarded as an experimental approach for some patients and was thought to be of limited utility for most. [2, 7, 8] Nevertheless, recent years have witnessed an exponential increase in the volume of procedures and the number of centers providing covered stent correction for SVASD. [5, 9] In 2025, an international registry on the rate of percutaneous covered stent closure of SVASD was published, indicating that covered stent repair for SVASDs is an emerging alternative to surgical repair. [10]

In 2023, a study involving 58 patients (mean age: 45.4 years; range: 14.8–73.8 y) who underwent surgical or catheter-based repair of superior SVASD with anomalous pulmonary venous drainage was conducted. The findings of this study suggested that catheter-based repair of superior SVASDs was a safe and effective procedure in selected patients and could be considered a suitable alternative to surgery. [9] Our results are consistent with these findings and confirm that this therapeutic approach is appropriate and successful.

Overall, transcatheter closure is a practical and safe option for carefully selected patients with SVC-type SVASDs, even in the presence of pre-existing pulmonary hypertension.

The present report describes a 59-year-old patient with SVASD, pulmonary hypertension, and atypical chest pain who was successfully treated with catheter closure. As described previously, this procedure was highly successful and effectively reduced symptoms such as dyspnea.

This report demonstrates the successful transcatheter closure of an SVC-type SVASD with associated mild pulmonary hypertension, highlighting the feasibility and efficacy of this minimally invasive approach. The accumulating evidence for catheter-based interventions for SVC-type SVASD indicates a paradigm shift in managing these congenital defects. As illustrated, careful patient selection and advanced techniques, such as covered stent repair, can yield significant symptomatic improvement and lower procedural risks compared with traditional surgery. This case adds to the literature supporting less invasive solutions for patients with complex congenital heart disease and may help guide future research and clinical protocols.

Conflict of Interest

The authors of this article have no conflicts of interest to declare.

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