Persistent Shunt After Closure of Ostium Secundum Atrial Septal Defect

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A train can hide another one.

–French adage

INTRODUCTION

We describe a rare case of a patient previously diagnosed with ostium secundum atrial septal defect (ASD) and treated with transcatheter occluder device who presented 12 years later with dyspnea and persistent left-to-right shunt. Transesophageal echocardiography (TEE) and cardiac computed tomography (CCT) showed new findings of superior sinus venosus ASD associated with partial anomalous pulmonary venous return (PAPVR) and persistent left superior vena cava (SVC). Given these findings and the patient's significant symptom burden, the consensus of the heart team was to proceed with surgical intervention.

CASE PRESENTATION

A 44-year-old patient was referred to the cardiology clinic of our institution because of dyspnea on exertion and poor exercise tolerance. Medical history was significant for transcatheter ASD closure device 12 years earlier. On physical examination, the patient was afebrile, with blood pressure of 144/79 mm Hg, a heart rate of 89 beats/min, a respiratory rate of 14 breaths/min, and an O_2 saturation of 98% on room air. There was no cyanosis, finger clubbing, or signs of heart failure. On cardiac auscultation, a soft 1/6 systolic murmur was heard at the left sternal border, but a split S2 was not heard. Transthoracic echocardiography with a bubble study performed at the referring institution showed a dilated right atrium and right ventricle associated with the presence of bubbles in the left atrium after intravenous injection of agitated saline consistent with persistent atrial shunt.

Before proceeding with imaging investigations, our thoughts were focused mainly on residual shunt after transcatheter ASD closure device that could have occurred from dislodgment or improper positioning of the device against the rim of tissue surrounding the ASD. However, although a small residual shunt may be present at the completion of transcatheter ASD occluder device deployment, it usually becomes undetectable years after the procedure.[1] Alternatively, we considered the presence of other sources of left-to-right shunt at the atrial level, such as sinus venosus ASD or coronary sinus defect that may have been missed at the time of previous transcatheter closure device deployment.

Two-dimensional TEE with color flow Doppler showed a well-positioned ASD closure device. Injection of agitated saline into a right arm vein showed an early appearance of large number of bubbles in the left atrium without clear evidence of bubbles crossing through or around the device (Figure 1A, Video 1). This increased the likelihood of another shunt at the atrial level. Indeed, slight pull-back of the transesophageal probe medially revealed a defect in the superior and posterior aspect of the SVC consistent with a superior sinus venosus ASD with an orifice 1.35 cm in diameter (Figure 1B, Video 2). The next step was to identify the pulmonary veins (PVs) and their connections, as abnormal pulmonary venous connections are usually associated with sinus venosus ASD. The left PVs were identified without difficulty and showed normal drainage of left PVs into the left atrium (Figure 2, Video 3). Visualization of the right PVs was more challenging and demonstrated the right upper PV draining into the right atrium (Figure 3, Video 4). Upon further TEE, the presence of a persistent left SVC was also noted and confirmed by injection of agitated saline into a left arm vein, with appearance of bubbles in a dilated coronary sinus draining into a dilated right ventricle (Figure 4, Video 5).

Because of challenging anatomy, and to confirm the findings on TEE, the patient underwent CCT, which confirmed the superior sinus venosus ASD as well as the anomalous right and middle pulmonary venous drainage into the right SVC just before its junction with the right atrium, as well as a persistent left SVC (Figure 5). Given the presence of left-to-right shunt, right heart catheterization was performed to measure the pressures in the right heart and the shunt ratio. This revealed a right atrial pressure of 12 mm Hg, right ventricle 36/10 mm Hg, and PA 33/11 mm Hg with a mean of 18 mm Hg. Oximetry showed step-up O_2 saturation with inferior vena cava O_2 saturation of 72%, SVC O_2 saturation of 80%, and a calculated ratio of pulmonary to systemic flow of 2.1:1.0.

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Keywords: Ostium secundum ASD, Sinus venosus ASD, Partial anomalous pulmonary venous return, Multimodality imaging, Management options

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VIDEO HIGHLIGHTS

Video 1: Two-dimensional TEE, midesophageal biplane image with agitated saline contrast study (from the right arm), demonstrates the well-positioned ASD occluder device with the early appearance of bubbles in both atria.

Video 2: Two-dimensional TEE, upper esophageal long-axis (110°) view with agitated saline contrast (from the right arm), demonstrates the sinus venosus ASD.

Video 3: Two-dimensional TEE, midesophageal long-axis (102°) view with color flow Doppler, demonstrates the left normal pulmonary venous drainage into the left atrium.

Video 4: Rightward rotated two-dimensional TEE, midesophageal long-axis (102°) view with color flow Doppler, demonstrates the anomalous right upper PV draining into the right atrium.

Video 5: Two-dimensional TEE, deep esophageal view (20°) with agitated saline contrast (from the left arm), demonstrates the bubble-filled dilated coronary sinus and right ventricle.

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

Subsequently, transesophageal echocardiography and cardiac computed tomographic images were reviewed by our multidisciplinary team, including clinical and interventional cardiologists and a congenital heart surgeon, who considered both surgical repair and transcatheter closure with a covered stent. Given the patient's symptoms, dilated right ventricular (RV) cavity (4.4 cm at the RV base), as well as a shunt ratio of 2.1:1, a shared decision was made to proceed with surgical repair, which was done using the Warden procedure via median sternotomy. A transcatheter covered stent was considered as an option; however, out of concern of overlapping the covered stent with the previous device, it was decided to proceed with surgery, which consisted of closing the sinus venosus ASD and redirecting the anomalous PVs into the left atrium. The patient's postoperative course was uneventful, and they were discharged in stable condition on the fourth postoperative day. Follow-up echocardiography before the patient's discharge showed a membrane in the right atrium consistent with a surgical patch. No residual shunt by color flow Doppler was visualized.

DISCUSSION

The association of sinus venosus ASD with PAPVR is well known.[2,3] However, our case is unique in that it shows a rare occurrence of ostium secundum and sinus venosus ASD with partial anomalous pulmonary venous connections in one patient. It also illustrates the importance of a comprehensive diagnostic approach in patients with congenital heart disease in general and ASD in particular, as the risk for missing a defect is real. ASDs represent 25% to 30% of congenital heart disease in adults.[2] Ostium secundum ASDs represent 80% of all ASDs and are due to defects within the fossa ovalis.[3,4] They are usually single, although on occasion, several small defects or an atrial septum with multiple fenestrations is also observed.[5] Ostium primum ASDs are part of atrioventricular septal defects and account for 15% of all ASDs.[5] Sinus venosus defects represent 5% to 10% of all ASDs and are located outside the confines of the true septum, resulting from deficiency of the tissue that separates the right upper PV from the SVC.[6] The superior sinus venosus defect allows the orifice of the SVC to override the septum and drain into both atrial chambers. It is often associated with partial anomalous connection of the right upper PV to SVC.[6] PAPVR is present in 10% of patients with ostium secundum ASDs and in 80% of patients with sinus venosus ASDs.[7] Therefore, it is important to identify all PVs in both ostium secundum and sinus venosus ASDs. Transthoracic echocardiography has limited use in the assessment of anomalous pulmonary venous connections, but TEE plays an important role in diagnosing the type of ASD and associated anomalies of pulmonary venous connections.[6-8] For patients in whom PV connections are not well visualized on TEE, cross-sectional imaging with cardiovascular magnetic resonance and CCT provides excellent images of ASD type, location, and abnormal PV connection, particularly those associated with veins that may be difficult to image on TEE, such as the innominate vein or vertical

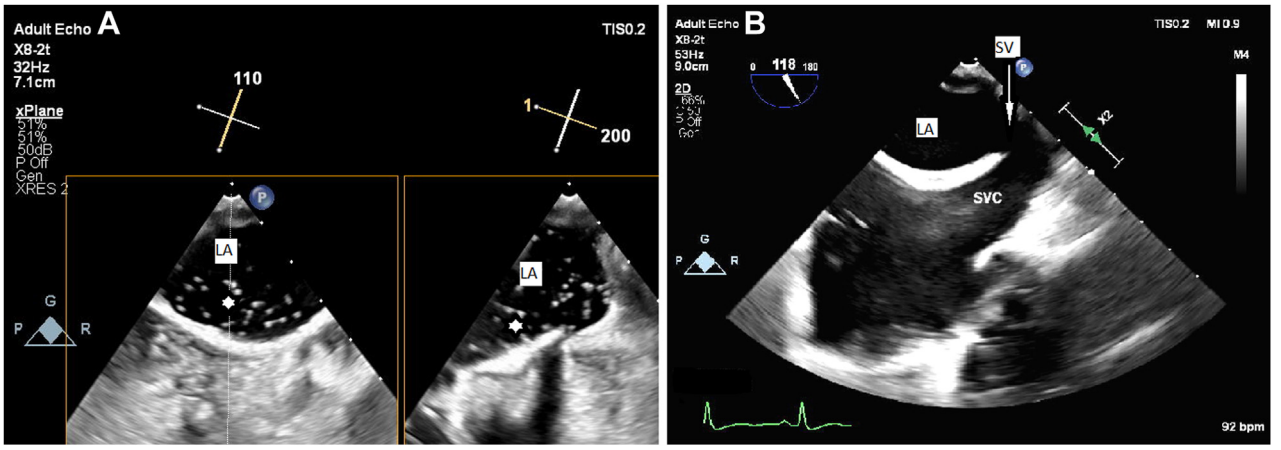

Figure 1 Two-dimensional TEE **(A)**, midesophageal biplane image with agitated saline contrast study (from the right arm) and **(B)** upper esophageal long-axis (110°) view of the atrial septum, demonstrates the well-positioned ASD occluder device with the early appearance of bubbles in both atria and the sinus venosus ASD. *LA*, Left atrium; *SV*, sinus venosus.

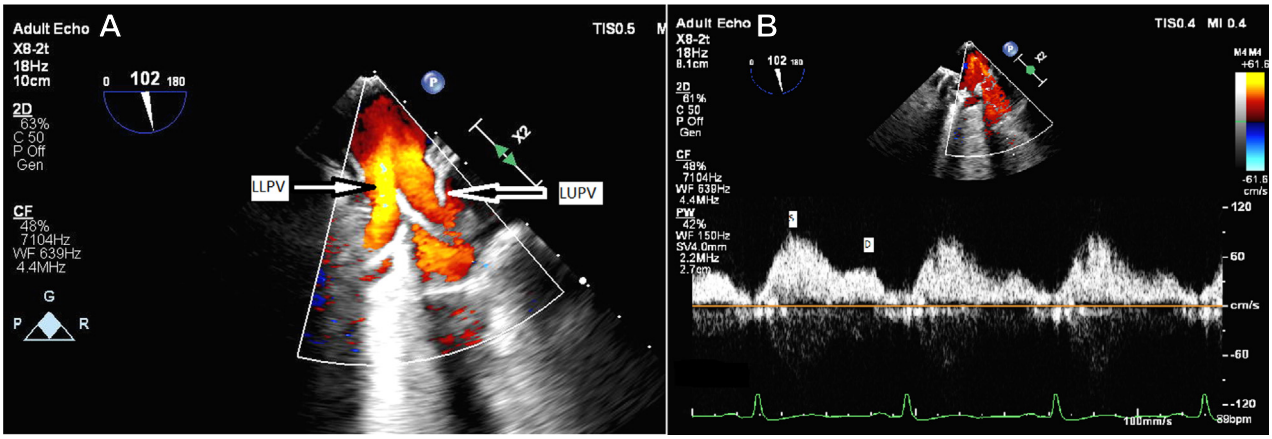

Figure 2 Two-dimensional TEE, midesophageal long-axis (102°) view with **(A)** color flow Doppler and **(B)** pulsed-wave Doppler, demonstrates the normal left pulmonic venous drainage into the left atrium. *D*, Diastolic flow; *LLPV*, left lower PV; *LUPV*, left upper PV; *S*, systolic flow.

vein.[9] At our institution, we perform CCT with retrospective electrocardiographic gating. Electrocardiographic gating minimizes motion artifacts for detailed anatomic evaluation and allows the assessment of cardiac function. For adequate opacification of cardiac chambers, iodinated contrast is power-injected at a rate of 5 mL/sec, with a region of interest placed on the ascending aorta. Image acquisition is triggered once attenuation of ascending aorta reaches 100 Hounsfield units. Images are reconstructed in thin isotropic slices ($\geq$1.0 $\times$ 1.0 mm) during diastole and systole and at 10% intervals along the cardiac cycle to improve quality of multiplanar reformatted images and to allow functional analysis. Because of its high spatial and temporal resolution and wide field of view, CCT is an excellent tool for the detection and characterization of septal defects and associated anomalies of pulmonary and systemic connections and is critical for accurate reporting and planning of surgical or transcatheter therapy.[9]

The decision to close an ASD in an adult is related to shunts large enough to cause functional impairment, evidence of right atrial and RV enlargement, atrial arrhythmias, net left-to-right shunt ratio $>$ 1.5:1 without cyanosis at rest or during exercise, systolic pulmonary artery pressure $<$50% of systolic systemic pressure, and pulmonary vascular resistance less than one-third of the systemic vascular resistance (class 1 recommendation).[10] Transcatheter closure of ASDs has become the treatment of choice for most patients with suitable anatomy and adequate surrounding rim of tissue.[11] Three-dimensional TEE provides an en face view of ostium secundum ASDs with clear visualization of the defect shape and the surrounding tissue and allows measurement of the defect size with better correlation against balloon sizing.[12] In recent years, advanced imaging with three-dimensional printing and multimodal fusion imaging process have provided simulation on printed model to ensure preprocedural guidance for transcatheter covered stent deployment for sinus venosus ASD, with excellent results in select patients.[13]

Surgery is an option for patients with sinus venosus ASDs who are not good candidate for transcatheter covered stents. The basic concept behind the surgical repair of a sinus venosus ASD is to close the interatrial defect and redirect the anomalous pulmonary venous drainage into the left atrium. Several procedures have

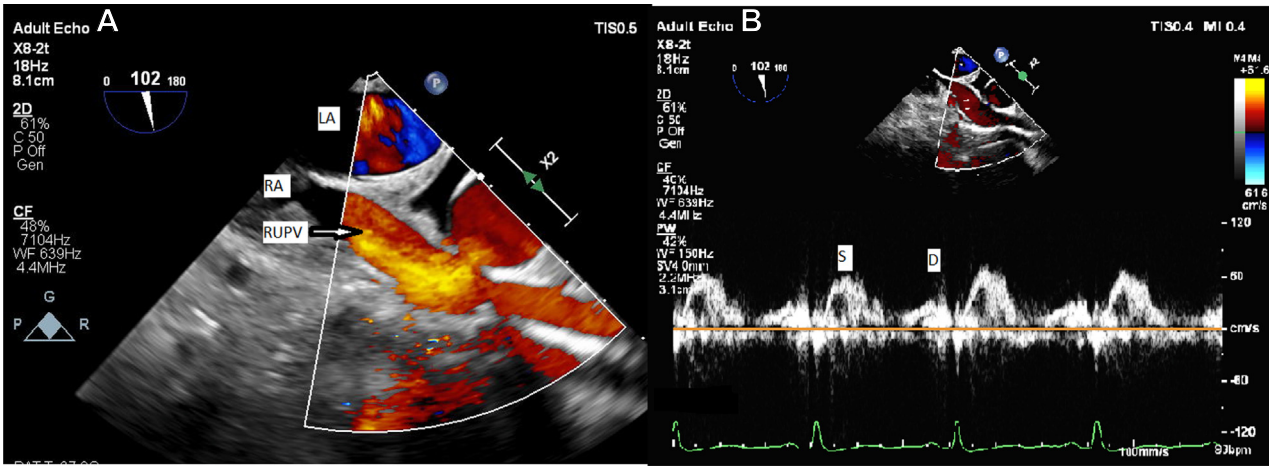

Figure 3 Rightward rotated two-dimensional TEE, midesophageal long-axis (102°) view with **(A)** color flow Doppler and **(B)** pulsed-wave Doppler, demonstrates the anomalous right upper PV draining into the right atrium (RA). *LA*, Left atrium; *RUPV*, right upper PV.

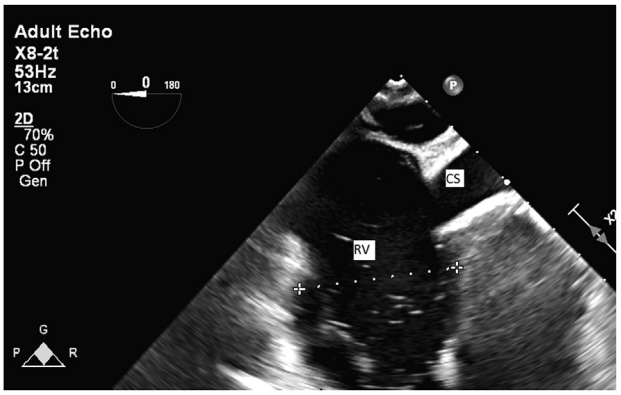

Figure 4 Two-dimensional TEE, deep transesophageal view (0°), demonstrates the dilated coronary sinus (CS) and right ventricle (RV).

been described. Warden repair, also called double patch repair, consists of transecting the right SVC above the drainage of the abnormal PV. This portion of SVC along with anomalous PV connections are rerouted into the left atrium. Then the SVC is anastomosed to the right atrial appendage.[14] Long-term results are excellent, with survival comparable with that among a normal age-matched population.[15]

CONCLUSION

Our case demonstrates the challenges in the diagnosis of sinus venosus ASD associated with PAPVR, its rare coexistence with ostium secundum ASD, as in our case, and the importance of using multimodality imaging (TEE, CCT, cardiovascular magnetic resonance) to delineate the anatomy and guide the appropriate intervention.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

The authors declare that since this was a non-interventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status.

FUNDING STATEMENT

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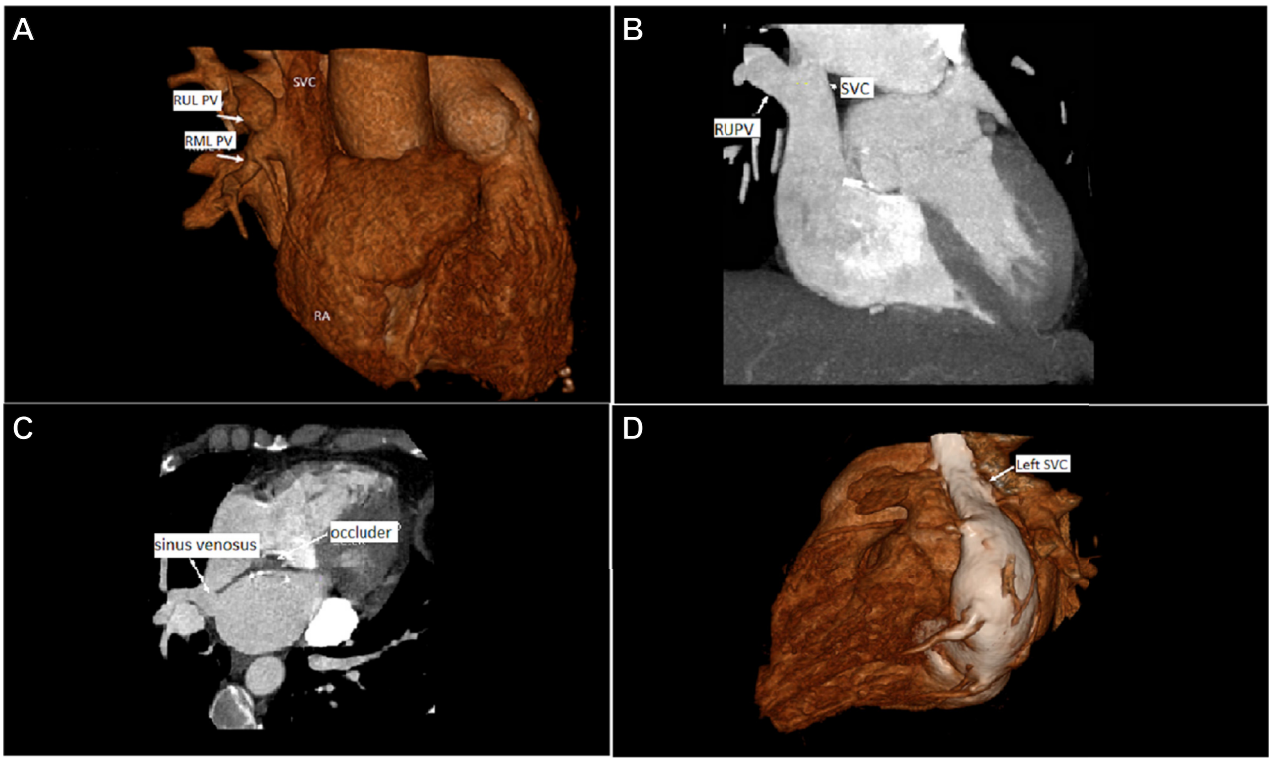

Figure 5 CCT demonstrates the anomalous right and middle PV connections to the right SVC and the sinus venosus ASD with **(A** and **D)** whole-heart, volume-rendered reconstruction displays and **(B)** oblique coronal and **(C)** axial multiplanar reformatted views.

DISCLOSURE STATEMENT

The authors report no conflict of interest.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2024.08.002.

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