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# A Rare Association of Sinus Venosus-Type Atrial Septal Defect and Persistent Left Superior Vena Cava Detected by Transthoracic Echocardiography and Cardiac Magnetic Resonance Imaging

Authors' Contribution:  
Study Design A  
Data Collection B  
Statistical Analysis C  
Data Interpretation D  
Manuscript Preparation E  
Literature Search F  
Funds Collection G

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**Conflict of interest:** None declared

**Patient:** Male, 60  
**Final Diagnosis:** Persistent left superior vena cava  
**Symptoms:** —  
**Medication:** —  
**Clinical Procedure:** Transthoracic echocardiogram and cardiac magnetic resonance imaging  
**Specialty:** Cardiology

**Objective:** Congenital defects/diseases  
**Background:** Association of persistent left superior vena cava (PLSVC) and sinus venosus-type atrial septal defect (SVASD) is rare. We describe a patient with dilated coronary sinus (CS) found to have PLSVC and SVASD.  
**Case Report:** The patient is a 60-year-old man with history of stroke who underwent a transthoracic echocardiogram (TTE) for evaluation of shortness of breath. TTE demonstrated a markedly dilated CS. Agitated saline was injected into the left antecubital vein to further assess CS. The parasternal long axis view demonstrated immediate filling of the CS and confirmed the presence of a PLSVC. Apical 4-chamber view with injection of agitated saline into the right antecubital vein demonstrated immediate contrast opacification of both atria, consistent with a right to left cardiac shunt. Cardiac magnetic resonance (CMR) was performed, which confirmed the TTE findings of PLSVC and defined the cardiac shunt as SVASD.  
**Conclusions:** PLSVC should be suspected in a patient with an abnormally dilated CS. In this case we identified a rare association of PLSVC with a SVASD. TTE with agitated saline contrast injection and CMR are useful diagnostic tools for PLSVC and associated cardiac congenital anomalies, respectively.

**MeSH Keywords:** Cardiac Imaging Techniques • Echocardiography • Heart Defects, Congenital

**Full-text PDF:** <http://www.amjcaserep.com/abstract/index/idArt/894394>

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## Background

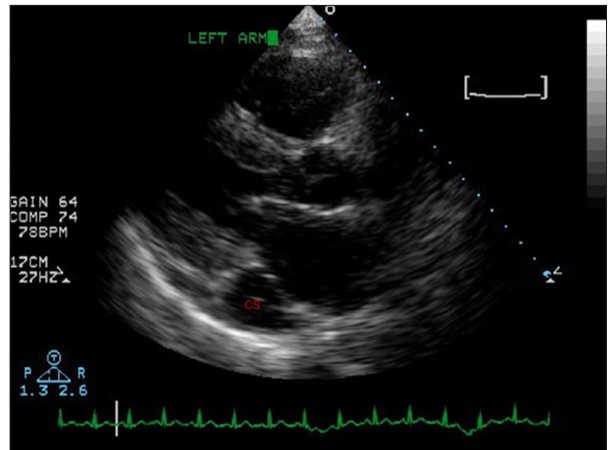
Persistent left superior vena cava (PLSVC) is the most commonly described thoracic venous anomaly in the medical literature [1]; however, the association with sinus venosus-type atrial septal defect (SVASD) is very rare [2]. SVASD, if left unrepaired, may eventually lead to right-heart volume overload and subsequent hypertensive pulmonary vascular disease, highlighting the need for heightened awareness of this association when PLSVC is diagnosed. We describe a case of PLSVC identified in a patient with markedly dilated coronary sinus (CS) and a rare association with SVASD.

## Case Report

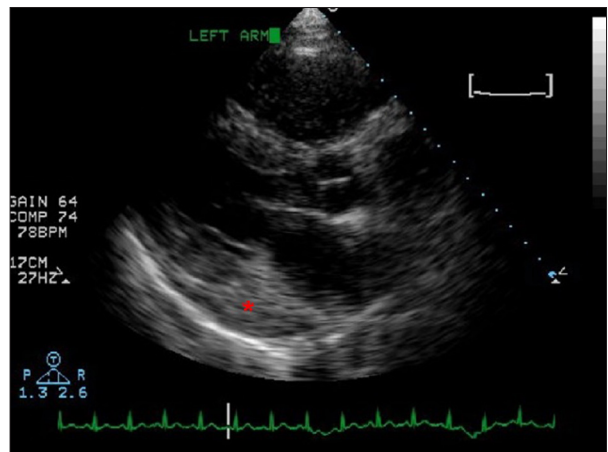
The patient was a 60-year-old male with history of stroke who underwent a transthoracic echocardiogram (TTE) for evaluation of shortness of breath. Initial TTE demonstrated a markedly dilated CS (Figure 1) and elevated pulmonary artery systolic pressure. To define the cause of the dilated CS, agitated saline was injected into the left antecubital vein. The parasternal long axis view demonstrated immediate filling of the CS and confirmed the presence of a PLSVC (Figures 1, 2). Imaging from the apical 4-chamber view with injection of agitated saline into the right antecubital vein demonstrated immediate contrast opacification of both atria, consistent with a right-to-left cardiac shunt (Figures 3, 4). Cardiac computed tomography (CT) was considered for better delineation of the venous anomaly and to define the location of the shunt; however, the patient had impaired renal function. Therefore, cardiac magnetic resonance (CMR) was performed on a 1.5T cardiac magnetic resonance imaging scanner, which confirmed the TTE findings of PLSVC (Figure 5, (arrow)), and defined the cardiac shunt as SVASD (Figure 6). Moreover, CMR helped to rule out some cardiac anomalies more commonly reported in association with PLSVC such as bicuspid aortic valve, coarctation of the aorta, coronary sinus ostial atresia, and cor triatriatum [1].

## Discussion

PLSVC is the most common thoracic vein anomaly, occurring in approximately 0.3–0.5% of the normal population [1,3] and about 3–10% in those with congenital heart disease (CHD) [1]. There are variable insertions of the PLSVC. It drains into the right atrium via the coronary sinus in 80–90% of individuals, resulting in no hemodynamic consequence, but can cause a right-to-left-sided shunt when it drains into the left atrium [4]. During the diagnostic procedure for our patient, SVASD, another congenital anomaly, was found. SVASD accounts for 5–10% of all atrial septal defects [2]. It is located along the superior aspect of the atrial septum in most cases, near the entry of



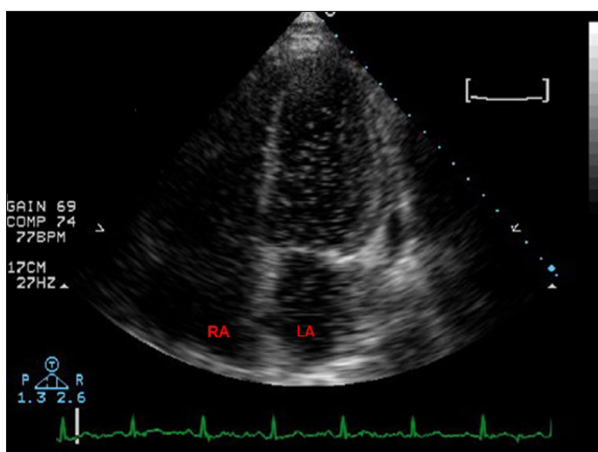
**Figure 1.** Parasternal long axis view of the CS without saline contrast injection into the left antecubital vein. CS – coronary sinus.



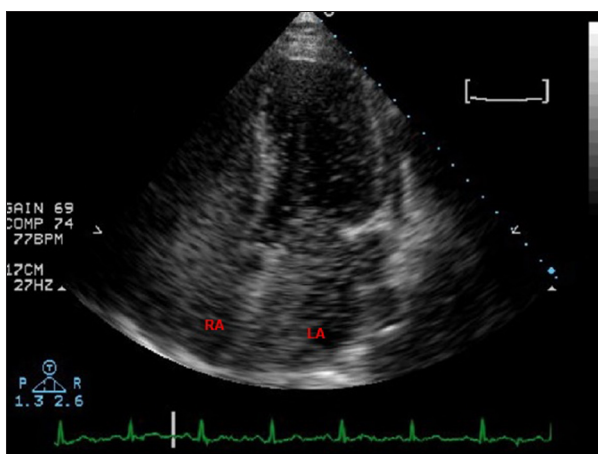
**Figure 2.** Parasternal long axis view with saline contrast injected into the left antecubital vein demonstrating immediate filling of the coronary sinus consistent with the presence of a persistent left superior vena cava. \* Filling of coronary sinus.

the superior vena cava. SVASD is caused by abnormal fusion between the embryonic sinus venosus and the atrium [5]. It may be asymptomatic in childhood but may become symptomatic with age. Unrepaired SVASD eventually leads to right-heart volume overload and can ultimately lead to hypertensive pulmonary vascular disease as was present in our patient.

PLSVC have practical implications when performing procedures such as right-heart catheterization or new therapeutic techniques that make use of the cardiac veins overlying the left ventricle, such as left ventricular pacing or percutaneous *in situ* coronary venous arterialization [4,6]. Swan-Ganz catheter placement can also be challenging as it is performed at the bedside. PLSVC can complicate permanent pacemaker and implantable cardioverter defibrillator placement. Serious



**Figure 3.** Apical 4-chamber view without saline contrast injection into the right antecubital vein. RA – right atrium, LA – left atrium.

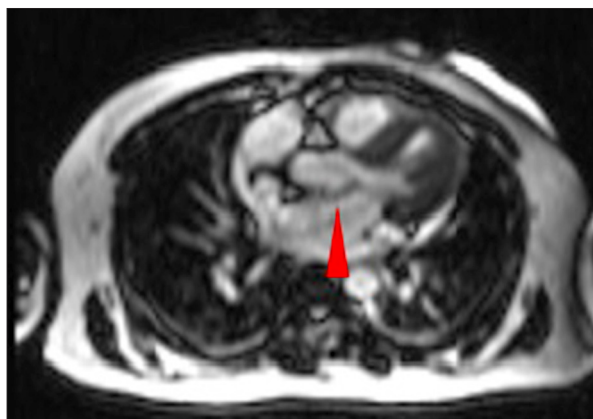


**Figure 4.** Apical 4-chamber view with saline contrast injected into the right antecubital vein demonstrating immediate contrast opacification of both atria, consistent with a right-to-left cardiac shunt. RA – right atrium, LA – left atrium.

complications such as arrhythmia, cardiogenic shock, cardiac tamponade, and coronary sinus thrombosis have been reported when pacemaker leads or catheters have been inserted via PLSVC [4]. However, the association of PLSVC and SVASD is very rare [2], and few cases have been reported in the literature. The presence of dilated CS in the absence of elevated right-sided filling pressure, enhancement of dilated CS before the right atrium (RA) following the injection of contrast into the left arm vein, and normal transit of contrast with RA opacification before the CS after injection of contrast in right arm are echocardiographic criteria for PLSVC diagnosis [4]. Transesophageal echocardiogram (TEE) improves diagnostic accuracy of SVASD [7] and it has also been shown to be superior to TTE in this regard [8].



**Figure 5.** Cardiac magnetic resonance confirming the findings of persistent left superior vena cava (arrow).



**Figure 6.** Cardiac magnetic resonance confirming the cardiac shunt as sinus venosus-type atrial septal defect (arrow head).

Cardiac CT was not performed due to our patient's poor renal function. Cardiac CT has been used by numerous studies to assess congenital cardiac malformations [9,10] and PLSVC [6]. It is a non-invasive test that allows for accurate and rapid evaluation of cardiac and vascular structures [11] and shows a clear spatial relationship of the heart with its surrounding structures [12]. A recent study pointed out the usefulness of cardiac CT, especially 3D helical CT angiography, in the evaluation of extracardiac anomalies in infants with complex CHD [13].

CMR can achieve a detailed cardiac morphologic imaging, assess vascular flow and cardiac function, and can also be applied in the work-up of CHD [14]. It is capable of providing qualitative and quantitative information regarding the anatomic layout and function of the heart due to recent technical advances in data acquisition and reconstruction, spatial and temporal resolution, and radiation dose reduction [15]. In a recent study comparing the use of CMR in place of diagnostic cardiac catheterization in the management of pediatric patients with CHD, CMR was associated with shorter hospital stay and

lower rate of complications, and, importantly, the use of CMR-acquired images in guiding surgical intervention did not result in a statistically significant decline in 30-day postsurgery survival when compared to catheterization [16].

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## Conclusions

PLSVC should be suspected in a patient with an abnormally dilated CS. In this case we identified a rare association of PLSVC with a SVASD. TTE is a useful diagnostic tool and agitated saline contrast injected into the right and left antecubital veins can establish the diagnosis of PLSVC. CMR is an alternative to cardiac CT in identifying venous and cardiac congenital anomalies.