

Imaging of persistent left sided superior vena cava with echocardiography and multi-slice computed tomography: Implications for daily practice

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Abstract

Persistent left sided superior vena cava is a congenital abnormality encountered not uncommonly by the echocardiographer or cardiac radiologist. Recognition of its presence is important in the differential diagnosis of dilatation of the coronary sinus. We discuss the echocardiographic and computed tomography findings of this congenital abnormality, and the implications for clinical practice. (Cardiol J 2011; 18, 3: 332–336)

Key words: persistent left sided superior vena cava, echocardiography, multi-slice computed tomography, coronary sinus

Introduction

Persistent left sided superior vena cava (PLSVC) is a rare but clinically relevant congenital anomaly. First described by Edwards and Dushane in 1950, it is the commonest congenital venous anomaly of the thorax with a 0.5–2% incidence in the general population, and approximately 10% incidence in patients with congenital abnormalities of the heart [1]. Although echocardiographic diagnosis of PLSVC is well-established, as we discuss below, the increasing use and interpretation of multi-slice computed tomographic (MSCT) angiography of the coronary arteries by cardiologists suggests the need to be aware of this incidental finding on MSCT. This case-based review discusses the approach to systematic diagnosis of this venous anomaly by two commonly used cardiac imaging modalities: namely, echocardiography and MSCT.

Case reports

Case 1

An asymptomatic 55 year-old man was referred for echocardiographic evaluation of possible hypertensive heart disease. The patient's past medical history was significant for long-standing poorly controlled hypertension as well as diabetes. He had no known history of coronary artery disease. His physical examination and basic laboratory evaluation were within normal limits.

The patient underwent standard transthoracic echocardiography (TTE) utilizing harmonic imaging and Doppler evaluation by commercially available ultrasound equipment. Normal left ventricular systolic function and absence of significant valvular abnormalities was confirmed in multiple standard views. Dimensions of the cardiac chambers and Doppler measurements were within normal limits.

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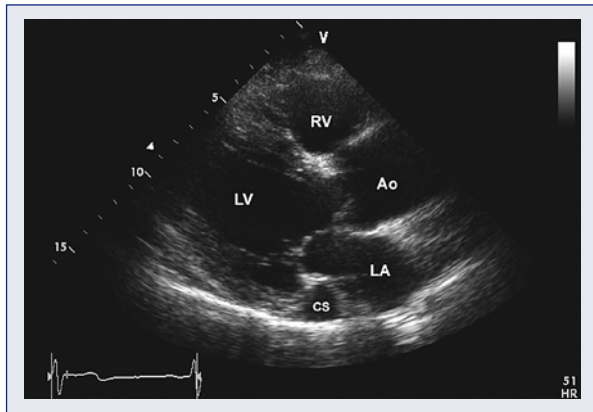


Figure 1. Parasternal long axis view demonstrating marked dilation of the coronary sinus (CS) with no other apparent echocardiographic abnormalities; LV — left ventricle; RV — right ventricle; Ao — aorta; LA — left atrium.

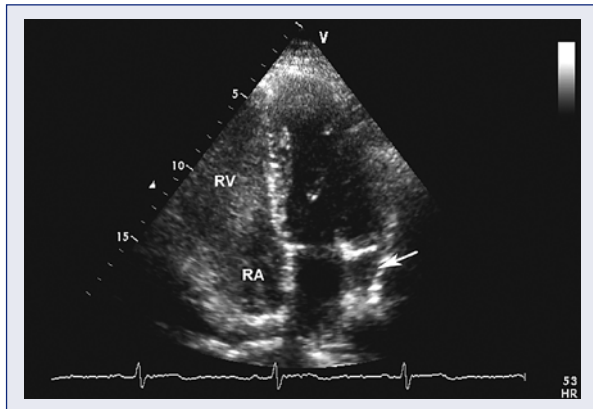


Figure 2. Apical four-chamber view after the injection of agitated saline into the right arm vein demonstrating normal opacification of the right atrium (RA) followed by opacification of the right ventricle (RV). The coronary sinus does not opacify (arrow).

However, the coronary sinus (CS) was seen to be abnormally dilated (Fig. 1). There was no significant enlargement or dysfunction of the right heart chambers or the tricuspid valve. PLSVC was suspected. A right arm peripheral vein was injected with agitated saline, and normal opacification was noted in the right atrium, followed by opacification of the right ventricle (Fig. 2). Next, the left arm peripheral vein was injected with agitated saline and abnormal opacification of the coronary sinus was noted first, followed by the right atrium and then

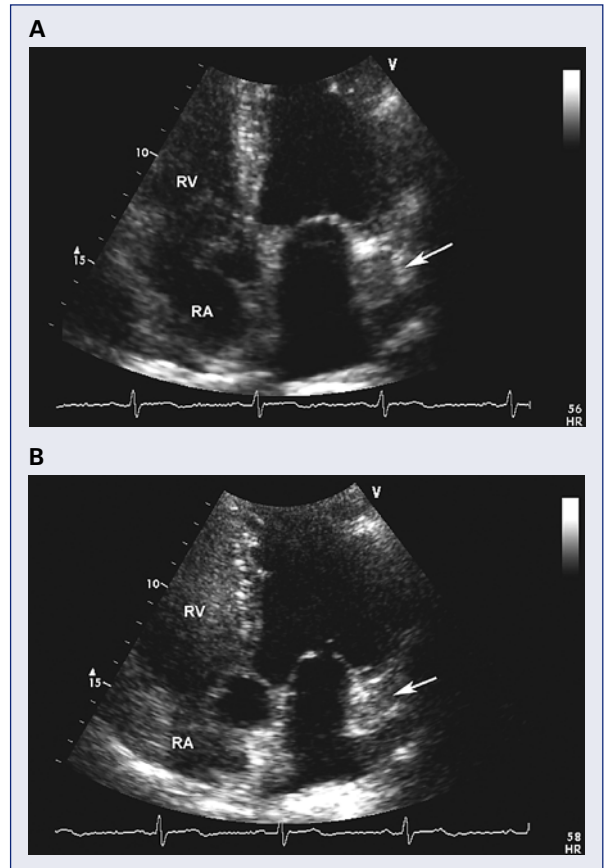


Figure 3. Apical four-chamber view after the injection of agitated saline into the left arm vein demonstrating initial opacification of the coronary sinus (arrow) (A) followed by sequential opacification of the right atrium (RA) then right ventricle (RV) (B).

the right ventricle (Fig. 3). This confirmed that the most likely etiology for CS dilatation was PLSVC.

Case 2

A 71 year-old woman with a history of chronic obstructive pulmonary disease underwent a MSCT to evaluate a hilar mass on chest X-ray. After a contrast injection through a left arm i.v., the patient was found to have an incidental finding of PLSVC. The well opacified PLSVC, located adjacent to the aortic arch, can be seen in axial and 3D volume-rendered reconstructions in Figures 4 and 5. The right SVC was not opacified during the exam because of the contrast injection through the left arm. TTE done subsequently for ejection fraction assessment prior to chemotherapy showed abnormally dilated CS without signs of right or left ventricular failure.

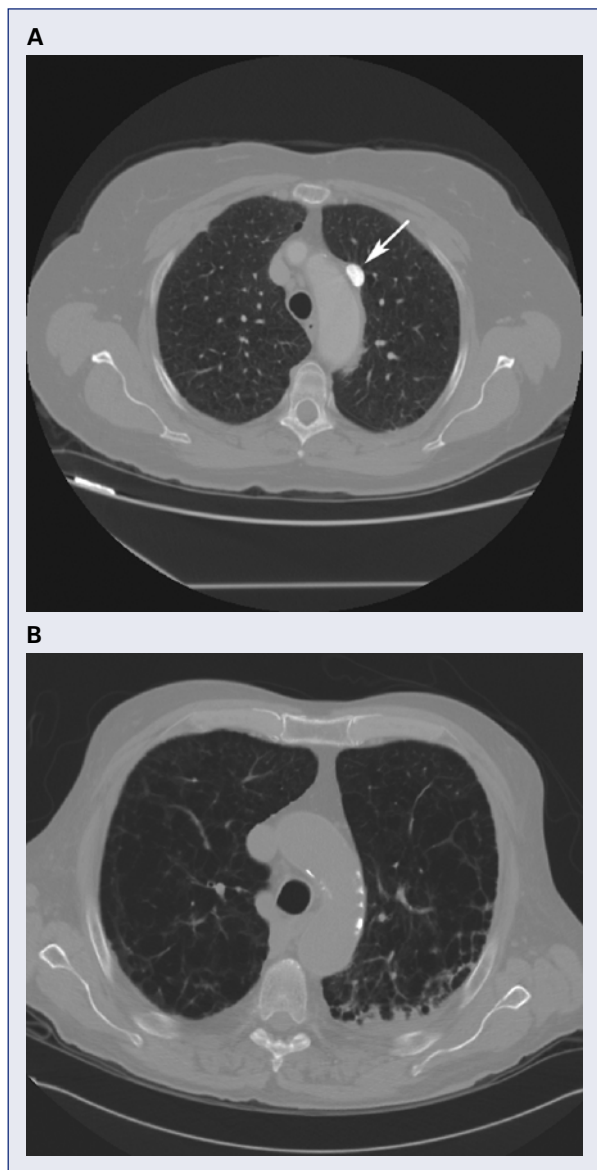


Figure 4. **A.** Axial images of a patient with persistent left sided superior vena cava (PLSVC) (arrow) opacified by a contrast injected through left arm i.v.; **B.** Corresponding axial image demonstrating absence of PLSVC in a patient without the congenital anomaly.

A ‘bubble study’ was not performed because of the previous knowledge of the PLSVC found on MSCT.

Discussion

PLSVC is a common congenital anomaly and the commonest cause of dilated CS [2]. During the eighth week of gestation, the left anterior cardinal vein, caudal to the innominate vein, obliterates to form the ligament of what Marshall described as the “vestigial fold of pericardium” [3]. PLSVC repre-

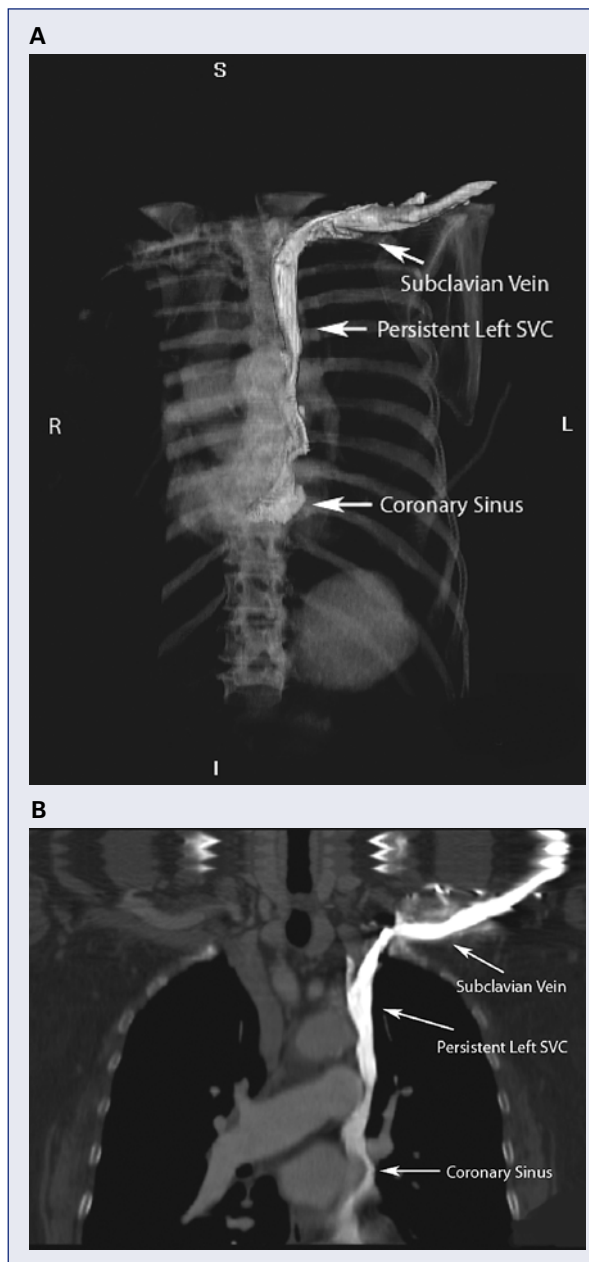


Figure 5. 3-D volume-rendered reconstruction (**A**) and coronal image (**B**) showing opacified persistent left sided superior vena cava (SVC) which drains to the coronary sinus.

sents the persistence of the left anterior cardinal vein that continues into the left CS [2]. The PLSVC opens into the right atrium via the CS and with presence of normal right SVC, the patient has a double SVC. This is commonly associated with a small or absent anastomosis that forms the left brachiocephalic vein [4].

PLSVC should be suspected whenever a dilated CS is discovered. Thus knowledge of CS anat-

Table 1. Causes of dilated coronary sinus, other than persistent left sided superior vena cava.

Etiology	Associated echocardiographic features
RV failure due to any cause	Severe RV dilatation, RV hypokinesis, RA dilation, increased RA pressures, plethoric inferior vena cava
ASD	Uncommon form of ASD due to 'unroofing' of CS. Left to right shunt detected by color Doppler or intravenous injection of agitated saline, Doppler quantification shows Qp/Qs ratio of > 1.2. Other forms of ASD can also lead to CS dilation in the setting of large shunts and RV dilation due to volume overload
Severe pulmonary hypertension	RA and/or RV dilation, severe TR, reduced mitral valve E/A ratio

RV — right ventricle; ASD — atrial septal defect; RA — right atrium; CS — coronary sinus; TR — tricuspid regurgitation

my and pathophysiology is integrally linked to a diagnosis of PLSVC and is also discussed here.

The coronary sinus is best visualized in the parasternal long-axis view as a circular structure of approximately less than 1 cm in diameter in the posterior atrioventricular groove [5]. CS dilation can stem from various forms of anomalous venous communication such as PLSVC with drainage into the CS, total anomalous pulmonary venous return with CS drainage, coronary AV fistula with drainage into the CS, and CS atrial septal defect. Other causes include right ventricular dysfunction, right atrial hypertension, and severe pulmonary hypertension.

TTE serves as an excellent modality to diagnose PLSVC. Typical findings are usually well appreciated on either the parasternal long axis or apical four-chamber view (Fig. 1). The presence of PLSVC can be confirmed by performing a bilateral 'bubble study' with the injection of agitated saline from both the left and right peripheral arm veins [6, 7]. Normally, given the presence of a single SVC, bubbles injected from either peripheral arm vein should first opacify the right atrium, with subsequent opacification of the right heart (Fig. 2). Agitated saline bubbles are destroyed during transpulmonary passage, and are therefore not seen in the CS. In the setting of PLSVC, agitated saline injected into the left arm drains through the abnormal connection between the left-sided venous return and the CS. The agitated saline bubbles can clearly be seen first in the CS, followed by right heart opacification (Fig. 3). A similar procedure to diagnose PLSVC can also be safely and accurately performed utilizing contrast echocardiography [8].

Methodical echocardiographic evaluation of patients found to have CS dilation helps the clinician correctly diagnose PLSVC or exclude it. When a dilated CS is identified, the echocardiographer should attempt to first rule out causes of right atrial volume/pressure overload such as right ventricu-

lar failure, atrial septal defect, or severe pulmonary hypertension (Table 1). Color flow and Doppler evaluation assists in ruling out coronary fistulas and other anomalous drainage patterns into the CS or right atrium. Finally, the bilateral injection of agitated saline serves as a confirmatory test for the commonest cause of CS dilation i.e. PLSVC.

Currently, MSCT is frequently applied by cardiologists to study coronary anatomy, coronary anomalies and aortic pathology. Since imaging of the thorax with MSCT also sheds light on systemic and pulmonary veins, incidental anomalies such as PLSVC merit attention. Electrophysiologists use MSCT almost routinely as a noninvasive 'road map' for pulmonary vein anatomy prior to ablation procedures for atrial fibrillation. Some institutions use computed tomography venography of the thorax as a road map for defining positions for biventricular pacemaker leads and implantable cardioverter defibrillators. The feasibility of MSCT to visualize the venous anatomy was recently demonstrated in studies with 16- and 64-slice detector row computed tomography [5, 9]. Newer generation MSCT with 64-slice technology along with 3D volume rendering provides near-isotropic data (i.e. longitudinal resolution approximates in-plane resolution). These data sets allow methodical post-processing to produce images optimized for these vessels, which are often not appreciated as planar axial images alone. Imaging of the thoracic venous system is best done in a caudal-cranial fashion, because any breathholding failures will usually occur at the end of this imaging sequence at the thoracic inlet, and contrast artifacts in the subclavian veins will be minimized [9].

Z-axis coverage is from the thoracic inlet to the celiac axis usually for imaging the thoracic venous system with MSCT, and this enables detection for even uncommon anomalies such as interrupted inferior vena cava, partial anomalous pulmonary veins etc. The presence of PLSVC and its drainage into

the CS can be well visualized on both axial slices and 3D volume-rendered reconstructions. Figure 4 shows the incidental detection of a different patient undergoing MSCT with left arm venous access. The axial images performed by 64-slice MSCT show contrast-filled persistent left SVC traveling adjacent to the aorta, as compared to the absence of the structure in a patient without the congenital anomaly. The contrast was injected through a left arm i.v. which permitted prolonged opacification of the PLSVC. The course and connection of PLSVC to the coronary sinus can be seen clearly in Figure 5.

Clinical implications

Although PLSVC is a benign condition, it may have important clinical implications. It may be associated with a variety of other congenital malformations of the heart and great vessels. It may cause technical difficulties in introducing central venous catheters [10], pulmonary artery catheters [11], or pacemaker and defibrillator leads to reach the right atrium, which may inadvertently lead to injury of the vessel wall [12]. Cardiac arrhythmias, including atrial and ventricular fibrillation, have been reported in these patients [13, 14]. Arrhythmogenesis of the PLSVC may result from dilatation of the CS opening, due to stretching of the atrioventricular node and His bundle or from focal discharges originating from muscular bundle extensions into the CS [15].

Conclusions

This case-based review illustrates echocardiographic and MSCT diagnosis of PLSVC and the evaluation of a dilated CS. It highlights the importance of a thorough evaluation of cardiac anatomy and knowledge of anatomic variations. The echocardiographer and the MSCT interpreter should be able to recognize this incidental finding and proceed with complete evaluation given its clinical implications. Once identified, PLSVC should be documented in a patient's medical history so that it is accounted for in any intravenous procedures in the thorax.

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