

Isolated Persistent Left Superior Vena Cava, Role of Echocardiography Screening and CT angiography

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Abstract

Background: An isolated persistent left SVC with concomitant agenesis of right SVC in adult patients is a very rare abnormality. Physician should consider it particularly in patients, in which venous access will be performed. Our rare case deals with the importance of detailed echocardiographic examination with screening of coronary sinus dilatation before the electrophysiology study.

Case: A 65-year-old woman came to outpatient clinic for a chief complaint of palpitations. Her ECG showed paroxysmal SVT with WPW syndrome. She underwent echocardiography examination before electrophysiology study and it was found that she had a dilated coronary sinus. Therefore we performed cardiac CT. It was found that she had a persistent left superior vena cava (SVC) and an absence of a right SVC with no other congenital anomaly.

Conclusion: A comprehensive echocardiography examination to look for a dilation of coronary sinus is a first suggestion to screen this anomaly, eventually followed by echocardiography with agitated saline injection and/or computed tomography can help physician to anticipate the anomaly before the invasive procedure involving the thoracic vein.

Keywords: echocardiography, cardiac CT angiography, vascular malformation, superior vena cava

BACKGROUND

Persistent left superior vena cava (PLSVC) is the most common congenital malformation of the thoracic venous system and it affects about 0.3 to 0.5% of the general population.¹ This incidence increases 10-fold in patients with cardiac malformations.² PLSVC with an absent right superior vena cava (RSVC), which is also referred to as isolated PLSVC, is very uncommon, occurring in 0.07 to 0.13% of patients who have congenital heart defects with viscerotransposition. Nearly half of the patients with isolated PLSVC have other cardiac malformations, such as atrial septal defect, endocardial cushion defect or tetralogy of Fallot.³ In this case report we present a patient with isolated PLSVC with no other cardiac structural abnormalities. The PLSVC was diagnosed by means of echocardiography. The diagnosis was supported by cardiac CT angiography.

CASE

A 65-year old woman went to outpatient clinic with a chief complaint of palpitations. Her ECG showed paroxysmal supraventricular tachycardia (SVT) with Wolf-Parkinson-White (WPW) syndrome. It was decided that she should undergo electrophysiology study and ablation. The echocardiography examination revealed a dilated coronary sinus. A CT angiography was performed because of suspicion of a persistent left superior vena cava (PLSVC). The examination showed a bridging vein draining the right jugular and right subclavian veins; it joined the left brachiocephalic vein and formed the PLSVC, which descended at the left side of the mediastinum, leftward of the pulmonary artery and left atrium (LA) before draining into the right atrium (RA) via a dilated coronary sinus (CS) (Figs 1–3). The RSVC was absent and the PLSVC carried all venous blood from

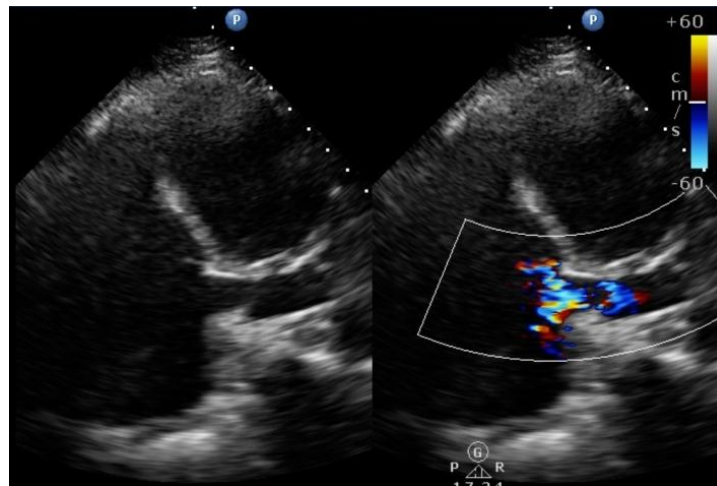


Figure 1. Transthoracic echocardiography showed a dilatation of coronary sinus draining to RA.

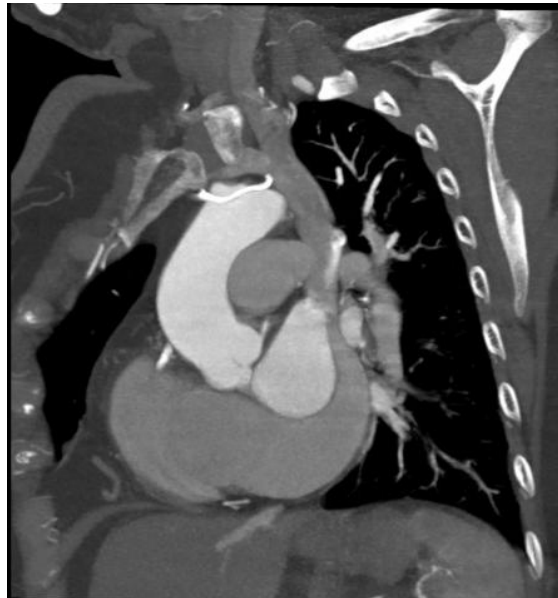


Figure 2. Multiplanar reformatted image demonstrates that the right superior vena cava is absent and there is persistent left superior vena cava (PLSVC).

the head, neck and upper extremities. There was no other intra- or extracardiac pathological structural finding.

DISCUSSION

PLSVC with absent RSVC (isolated PLSVC) is a very rare venous malformation. During normal

fetal development, the left-sided anterior venous cardinal system regresses, leaving the CS and the ligament of Marshall. Failure of the closure of the left anterior cardinal vein results in PLSVC.⁴ In general, PLSVC is associated with RSVC and drains into the RA via a dilated CS. When developmental arrest occurs at an earlier stage, the CS is absent and the PLSVC drains

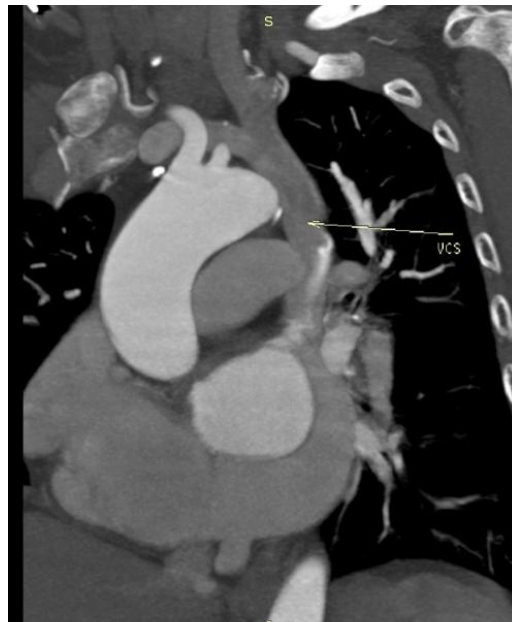


Figure 3. Multiplanar reformatted image reveals the persistent left superior vena cava (PLSVC) draining into a dilated coronary sinus (CS)

into the LA. Either isolated or associated with RSVC, this venous malformation itself causes no haemodynamic disturbance and is usually diagnosed incidentally.⁵⁻⁷

However, it has several clinical implications. A PLSVC can cause problems during central venous catheterisation (access to the CS can cause hypotension, angina, perforation of the heart, tamponade and arrest),⁸ pacemaker implantation (due to the circuitous path taken by the electrode, it can be difficult to obtain a stable electrode position and sustained capture),⁹ or cardiopulmonary bypass (isolated PLSVC impairs the use of retrograde cardioplegia).

In addition, a higher incidence of arrhythmias and conduction system abnormalities has been described in patients with PLSVC. There are two proposed mechanisms for this association: a dilated CS stretches the atrioventricular nodal tissue, which prepares a substrate for re-entrant tachycardias; or, the early conduction tissue has close

proximity to the cardinal venous tissue and this leads to sinus node dysfunction. Lenox *et al.* found sino-atrial node abnormalities in some patients with absent RSVC and this condition may predispose to sick sinus syndrome.¹⁰⁻¹²

In 10% of patients, a PLSVC may drain into the LA either directly or via an unroofed CS. This creates a right-to-left shunt and the risk of paradoxical embolism is markedly increased. In addition, drugs directly enter the systemic circulation when they are applied from the left brachiocephalic vein. A final clinical implication of PLSVC (especially when isolated) is a high incidence of accompanying congenital heart defects, for example ventricular septal defect, atrial septal defect, endocardial cushion defect or tetralogy of Fallot.^{3,13} Therefore associated congenital heart disease should be meticulously searched for.

When PLSVC is present, the ECG often shows an abnormal P-wave axis and a normal or shortened PR interval. A geometric change in the LA may be a possible mechanism for the

left-axis deviation of the P wave.¹⁴ On chest X-ray, a crescent-shaped shadow of the PLSVC can be seen at the aortic knob or left upper mediastinum. After insertion of a pulmonary artery catheter into the left subclavian or jugular vein, a control chest X-ray gives the false appearance that the catheter has passed through the vessel. The diagnosis can be confirmed by TTE, transoesophageal echocardiography (TEE), venous angiography, computed tomography (CT) or magnetic resonance imaging (MRI).

On two-dimensional B-mode TTE, the characteristic finding is a dilated CS on parasternal long-axis view. The normal diameter of the CS is smaller than 1 cm and in the case of isolated PLSVC, severely increased flow can cause a truly giant CS.^{15,16} Other causes of dilated CS are: increased RA pressure, an anomalous systemic or pulmonary venous connection or a fistulous connection with the coronary arteries.¹⁷

The next step in the echocardiographic evaluation should be contrast application with agitated saline. In normal individuals, agitated saline injection from the left or right antecubital vein results in opacification of the RA. In isolated PLSVC, as in our case, contrast given from the left or right arm opacifies the CS. When PLSVC is associated with an unroofed CS, contrast injection from either arm results in opacification of the LA. If RSVC accompanies the PLSVC, contrast given from the left arm first appears in the CS, whereas contrast given from the right arm first appears in the RA. On TEE, the anomalous PLSVC and absence of RSVC can be well visualised. In mid-oesophageal views, the PLSVC can be seen near to the left atrial appendage and left upper pulmonary vein. In the bicaval view, the absence of RSVC can be demonstrated.

Other techniques (venous angiography, CT, MRI) directly visualise the venous anatomy and confirm the diagnosis. In the absence of an RSVC, central venous access should be made from the

femoral vein in patients with PLSVC. During right-sided open-heart surgical procedures, a PLSVC has to be drained by inserting a separate cannula into it. If the PLSVC drains into the LA and creates a large right-to-left shunt, surgical correction should be made. Again, central venous access via the femoral vein is a safer choice in this variation. When implanting permanent pacemakers, the left subclavian vein is preferred, as lead manipulation is easier. There is an acute angle between the CS ostium and the tricuspid valve, therefore the lead should be looped in the RA in order to enter the right ventricle.¹⁸ Hand-shaped stylets and active fixation leads are also helpful to overcome technical difficulties.¹⁹

Finally, a wide spectrum of clinicians (radiologists, sonographers, interventionalists, intensivists, anaesthesiologists, cardiothoracic surgeons) should be aware of PLSVC and its variations in order to avoid possible complications.

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