Persistent Left Superior Vena Cava Draining Directly into Left Atrium and Normal Coronary Sinus

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Abstract

A persistent left superior vena cava is a frequent variation of the thoracic venous system and is explained by the persistence of the left superior cardinal vein. However, in most cases, it drains into the coronary sinus. It sometimes terminates directly into the left atrium but this anomaly is usually associated with the absence of the coronary sinus. To our knowledge, only rare cases of a persistent left superior vena cava terminating into the left atrium with a normal coronary sinus have been described. In this article, we present a case with a direct connection between the left superior vena cava and the left atrium, diagnosed by saline contrast echocardiography, and describe the contribution of echocardiography in the diagnosis of a persistent left superior vena cava. Echocardiography is a reliable and easy diagnostic tool that allows a bedside approach in a patient in whom there is suspicion of a persistent left superior vena cava, without the administration of radiographic contrast. ***(Iranian Heart Journal 2012; 13(2):49-53).***

Keywords: Persistent left superior vena cava■ Echocardiography

Case report

24-year-old woman with atypical chest pain was referred for echocardiographic evaluation. She had a history of a large ventricular septal defect (VSD) and patent ductus arteriosus (PDA) at childhood and surgical repair of the PDA and pulmonary artery banding nineteen years previously. That surgical operation was followed by VSD surgical repair three years afterward. She had remained symptom free until she referred to us.

Physical examination was unremarkable except for a systolic murmur (grade II/VI) at the lower left sternal border. No cyanosis was detected in her routine physical examination. Her ECG showed sinus rhythm with right bundle branch block.

Transthoracic echocardiography showed a mild left ventricular (LV) enlargement with a normal systolic function (LV ejection fraction =55%), moderate right ventricular (RV) enlargement with mild systolic dysfunction, and moderate tricuspid regurgitation (peak gradient =25 mm Hg). There was no residual VSD or PDA, and the other parameters, including the coronary sinus diameter, were normal.

The injection of agitated saline (creating air-filled micro bubbles by shaking the saline solution in a syringe) via the left antecubital vein showed immediate opacification of the left atrium (LA) and subsequently the LV, suggesting the diagnosis of a persistent left superior vena cava (PLSVC) connection to the LA (Fig.1).

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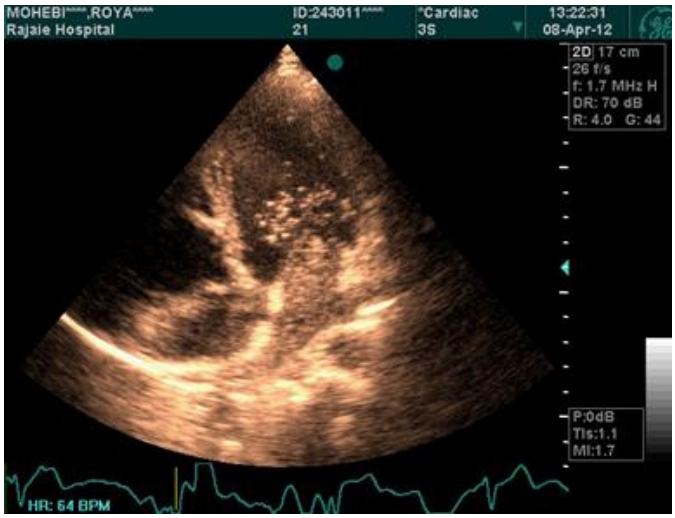
A left supraclavicular echocardiographic examination also confirmed the diagnosis of a PLSVC to the LA (Fig. 2 and Fig. 3).



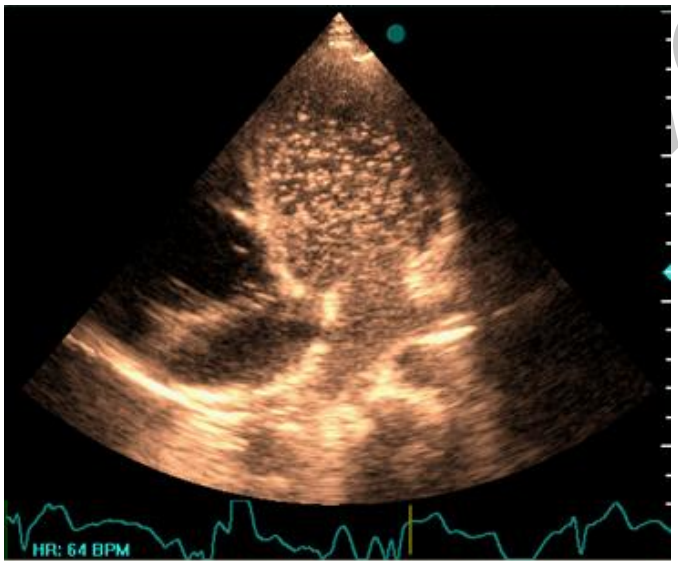


Fig.1. Agitated saline contrast injection via the left antecubital vein shows the immediate appearance of the contrast in the left atrium and then in the left ventricle, suggesting a diagnosis of a direct connection between a persistent left superior vena cava and the left atrium.

Fig. 2. Left supraclavicular echocardiographic view demonstrates an abnormal venous structure adjacent to the upper descending aorta and left atrium.

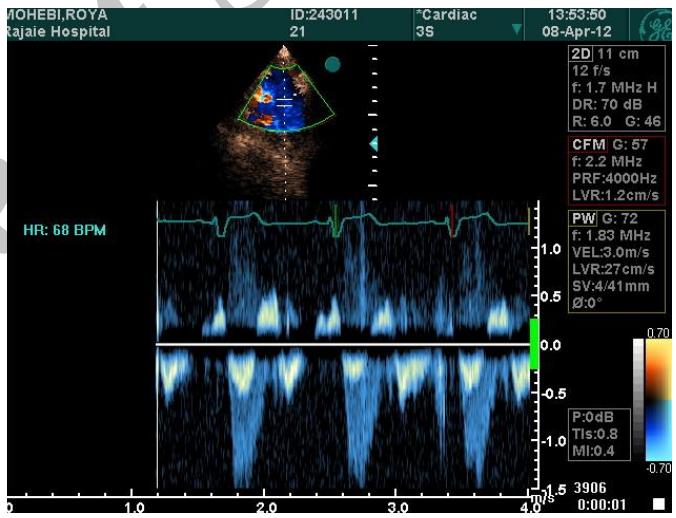


Fig. 3. Pulsed wave Doppler recording of the persistent left superior vena cava

Epidemiology

The PLSVC is a congenital abnormality with an estimated incidence of 0.3–0.5% in the normal population (1,2). The PLSVC is most frequently seen in patients with other congenital heart abnormalities, with a reported incidence of 1.5–10% in these patients (2,3). Despite its low incidence, the PLSVC is the most common congenital abnormality of the venous system. A more uncommon abnormality is the persistence of an LSVC connecting directly to the LA. This anomaly was found in about 7.5% of the cases of PLSVC (4).

Embryology

Venous system development is a complex process, during which several malformations may occur such as the absence or duplication of some sections and malposition. In the embryo, the venous sinus consists of a central section, called transversal portion, and the right and left horns. The coronary sinus originates from the fusion of the transversal portion of the venous sinus with the proximal part of the left horn (5). A PLSVC results from the permanence of the left horn of the venous sinus and the cardinal common vein; this phenomenon affects the development of the sinus node, atrioventricular (AV) node, and His bundle (5, 6). A PLSVC, patent during fetal life, normally closes before birth. In some cases, however, it remains patent after birth and its venous blood drains into the coronary sinus in 50% of cases and into the right superior vena cava through a venous trunk in 50% (6).

Various researchers believe that the embryological anomaly of the LSVC connecting to the LA cannot exist with a normal coronary sinus. Be that as it may, there is a theory to explain this association of the LSVC with the LA and a normal coronary sinus. There is no obvious reason for the coronary sinus to become involved when the left common cardinal vein retains a connection with the LA (7).

PLSVC Subtypes

Ninety-two percent of PLSVC cases drain into the right atrium via the coronary sinus or sinus venosus. These patients are asymptomatic. Eight percent of PLSVC cases drain into the LA, causing right-to-left shunting. These latter patients may have unexplained cyanosis and are at risk for systemic embolization (8). The PLSVC has been classified as follows: 1) isolated, a very uncommon pattern (9) characterized by an extremely dilated coronary sinus and 2) complete unroofed coronary sinus (10), in which the LSVC is connected to the roof of the LA in a site between the LA appendage and the pulmonary veins. In the latter situation, the right superior vena cava is small or absent (11), associated with the right superior vena cava: in this case the PLSVC can be connected to the atria through the right superior vena cava or separately. Nevertheless, there are some reports that show the association between the rare anomaly of a PLSVC to the LA and a normal coronary sinus. We do not know whether this rare association ever occurs without other intracardiac malformations. The hemodynamic results of a small right-to-left shunt in such a situation are unlikely to be noticeable (7).

Associated anomaly

In 10% of cases, the PLSVC is associated with other congenital abnormalities, particularly AV channel, ASD, VSD, Tetralogy of Fallot, and situs inversus (11 , 12).

Diagnosis

The PLSVC is a congenital anomaly that is not uncommon. Its discovery often follows the finding of an abnormally positioned

catheter, pacemaker, or internal defibrillator lead placed into a left subclavian or left jugular vein. Most physicians, confronted with a central venous catheter with its tip in the left paramediastinal region, will think of the diagnosis of a PLSVC, but probably will desire confirmation of the presence of a PLSVC to rule out catheter perforation or migration. Multi-slice cardiac computed tomography scan with or without three-dimensional reconstruction, cardiac magnetic resonance imaging, venography with left-sided injection of the iodinated contrast, and echocardiography are described as diagnostic tools to confirm the diagnosis of a PLSCV. The advantages of transthoracic echocardiography compared with the other techniques are: 1) no need for nephrotoxic contrast agents in a patient with pre-renal failure; 2) no radiation exposure; 3) no hemodynamic effects after the injection of agitated saline (micro bubbles) compared with iodinated contrast; and 4) no possible anaphylaxis to the injected saline (8). The diagnostic echocardiographic procedure includes three important steps: 1) measurement of the coronary sinus by two-dimensional echocardiogram, using the off-axis four-chamber apical view for superior-inferior diameter measurement

(normal value 10–12 mm), and the parasternal long-axis view for area evaluation (normal value 0.9 ± 0.3 cm^2); 2) injection of contrast in the left antecubital vein in order to confirm the passage of the medium first into the coronary sinus and then in the right atrium; and 3) distinction of the isolated PLSVC from the form associated with a right superior vena cava by means of an injection of a contrast medium in the right antecubital vein: the isolated form results in the visualization of the coronary sinus first and the right atrium later on. This finding demonstrates the absence of a right superior vena cava, whereas in case of an unroofed coronary sinus, the LA is visualized first, independently of either the right or left antecubital vein used (Table 1) (13).

Echocardiographic findings	Normal	Isolated PLSVC	Associated PLSVC	Unroofed CS
CS diameter	10-12mm	>12mm	>12mm	10-12mm
Contrast injection in left arm vein	RA	CS>>RA	CS>>RA	LA
Contrast injection in right arm vein	RA	CS>>RA	RA	LA

Table 1. Echocardiographic findings to diagnose persistent left superior vena cava

PLSVC, persistent left superior vena cava; LA, left atrium; RA, right atrium; CS, coronary sinus

Conclusion

Most instances of a PLSVC drain into the coronary sinus, leading to characteristic coronary sinus dilation with confirmation by saline contrast echocardiography. However, approximately 10% of PLSVC cases drain into the LA but this anomaly is usually associated with the absence of the coronary sinus. To our knowledge, only rare cases of a PLSVC terminating into the LA with a normal coronary sinus have been described. Contrast echocardiography is safe and highly informative for the definite diagnosis of a PLSVC and its drainage site.

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