Persistent Left Superior Vena Cava with Absent Right Superior Vena Cava

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ABSTRACT: Persistent left superior vena cava (PLSVC) is the most common congenital abnormality of the thoracic systemic venous drainage; in fact, cardiology fellows in training are often quizzed on this when a dilated coronary sinus is noted on an echocardiogram. However, its association with an absent right superior vena cava (RSVC) and how to diagnose this combined condition is less commonly known. We present two cases of PLSVC with an absent RSVC, describe how to recognize and confirm this diagnosis, and discuss its clinical relevance.

PATIENT 1

A 62-year-old male with a history of coronary artery disease, hypertension, and hyperlipidemia was admitted with chest pain. An electrocardiogram showed ectopic atrial rhythm, RSR' pattern in V1, and nonspecific ST- and T-wave abnormalities. Chest x-ray revealed fine reticular opacities in the mid to lower lungs, possibly from early edema or atypical infection.

The patient's echocardiogram showed an enlarged coronary sinus (Figure 1 A, B). Saline contrast study through a left peripheral intravenous (IV) catheter revealed saline bubbles in the coronary sinus before the right atrium and right ventricle, confirming PLSVC (Figure 2). However, an injection of saline contrast through peripheral access in the right upper extremity also generated saline bubbles in the coronary sinus before the right atrium (Figure 3). This finding suggested that the right subclavian vein drained into the left superior vena cava.

The patient was referred for a coronary angiogram for chest pain, but results were unremarkable. A venogram was subsequently performed. An injection of iodinated contrast through an IV catheter in the left upper extremity vein confirmed the diagnosis of PLSVC draining into the coronary sinus. Next, iodinated contrast was injected through an IV catheter in the right upper extremity vein. This confirmed that the right subclavian vein emptied into the left superior vena cava, affirming the absence of RSVC. This was noted in the patient's record for future reference.

PATIENT 2

A 69-year-old male with a history of nonischemic cardiomyopathy and severe left ventricular (LV) systolic dysfunction (despite optimal medical therapy), chronic atrial fibrillation, chronic obstructive pulmonary disease,

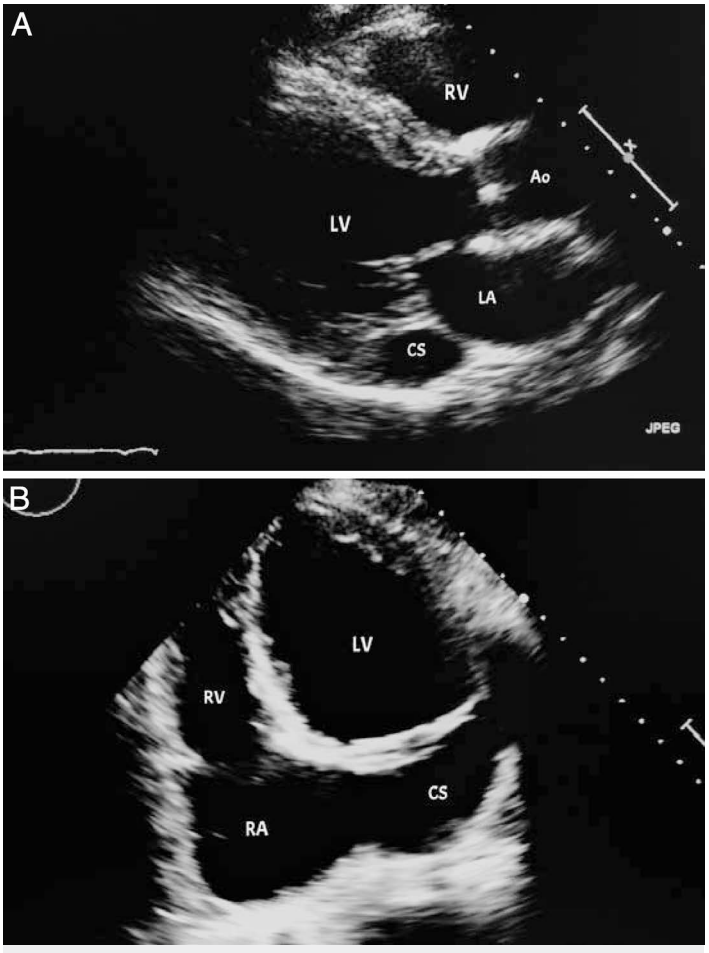

Figure 1.
(A) Two-dimensional transthoracic echocardiogram with a parasternal long view showing dilated coronary sinus, raising the suspicion for persistent left superior vena cava (PLSVC). (B) Apical view showing dilated coronary sinus, also raising the suspicion for PLSVC. LV: left ventricle; LA: left atrium; RA: right atrium; CS: coronary sinus; RV: right ventricle; Ao: aorta

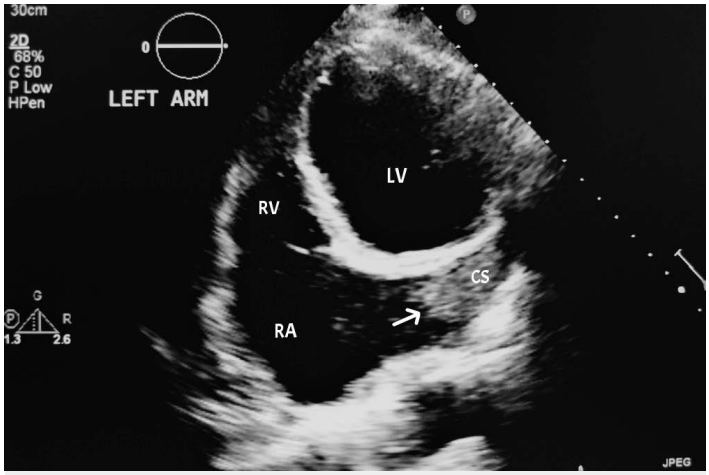

Figure 2.
Two-dimensional transthoracic echocardiogram with an apical view and injection of saline contrast through an IV catheter in the left upper extremity demonstrates saline contrast entering the coronary sinus (arrow) prior to the right atrium and right ventricle, confirming persistent left superior vena cava. RA: right atrium; RV: right ventricle; LV: left ventricle; CS: coronary sinus

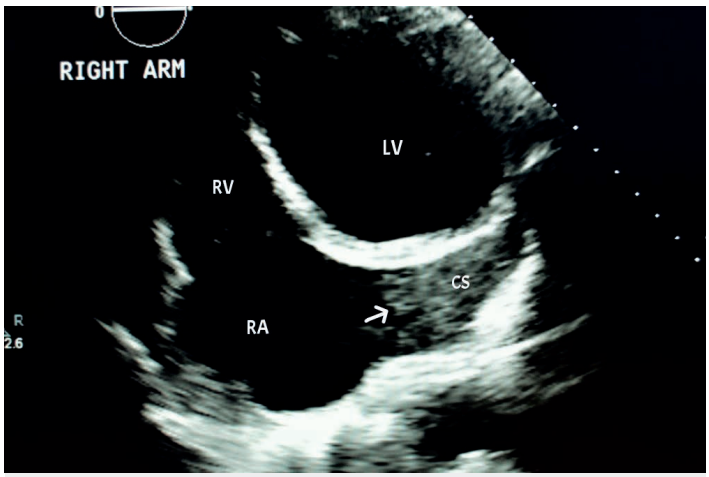

Figure 3.
Two-dimensional transthoracic echocardiogram with an apical view and injection of saline contrast through an IV catheter in the right arm demonstrates opacification of the coronary sinus (arrow) prior to the right atrium and right ventricle, confirming the absence of right superior vena cava. RA: right atrium; RV: right ventricle; LV: left ventricle; CS: coronary sinus

hypertension, obstructive sleep apnea, and anemia was referred for an implantable cardioverter defibrillator (ICD) placement for primary prevention. His electrocardiogram showed atrial fibrillation with occasional premature ventricular contractions and a ventricular rate of 86 beats per minute. His echocardiogram prior to the procedure confirmed severe LV systolic dysfunction with a visually estimated ejection fraction of 30%. The right atrium was mildly dilated, but the right ventricle was normal in size and systolic function. A markedly dilated coronary sinus was noted. Although a dilated coronary sinus may be found in patients with heart failure, PLSVC was suspected due to the severity of the dilatation. An injection of saline contrast in a left upper extremity IV catheter revealed the presence of contrast in the coronary sinus prior to right atrium, confirming PLSVC. Next, injection of saline contrast through a right upper extremity IV catheter also showed contrast in the coronary sinus before the right atrium, consistent with an absent RSVC.

Figure 4.
Venogram with injections from left and right subclavian veins confirms persistent left superior vena cava and absent right superior vena cava (RSVC) with the right subclavian vein draining into the left SVC.

The patient was taken for an ICD knowing that the procedure would be challenging due to the congenital anatomical anomaly. A bilateral upper-extremity venogram performed prior to the procedure demonstrated the presence of PLSVC with absent RSVC (Figure 4). Due to the absent RSVC, ICD access was attempted through the left subclavian vein rather than from the right. Unfortunately, an ICD lead could not be placed into a suitable position in the right ventricular apex. After much difficulty, a lead was positioned in the septal aspect of the right ventricular outflow tract but could not demonstrate acceptable sensing and pacing thresholds. Thus, the procedure was aborted and the patient was referred for subcutaneous ICD placement, which was placed without complications.

DISCUSSION

PLSVC is a rare congenital abnormality that is found in about 0.3% of the general population. Its incidence increases by 10- to 30-fold in patients with congenital heart disease.[1-3]

The fetal development of systemic and pulmonary veins is complex and variable. In normal cardiac development, the anterior cardinal veins draining the cephalic portion of the embryo combine with the posterior cardinal veins to form common cardinal veins that drain into the sinus venosus. During the eighth week, an anastomosis develops between the right and left anterior cardinal veins, creating the left brachiocephalic vein. Portions of the right anterior cardinal vein and the right common cardinal vein develop into the RSVC. Part of the left anterior cardinal vein caudal to the brachiocephalic vein regresses, leaving the ligament of Marshall and coronary sinus. In rare cases in which the left anterior cardinal vein fails to regress, a PLSVC forms and usually drains into the right atrium via the coronary sinus.[4-6] The majority of patients with PLSVC have an intact RSVC. In rare circumstances, the right anterior cardinal vein regresses during fetal development, resulting in absent RSVC associated with PLSVC.[7-9]

PLSVC should be suspected when a dilated coronary sinus is noted on echocardiography. This condition can be diagnosed by injecting saline contrast through an intravenous catheter in the left arm. Under normal circumstances, injection of saline contrast through a vein in the left arm will opacify the right atrium before draining into the right ventricle. In the presence of PLSVC, opacification of the coronary sinus will occur before the right atrium.[10,11] Absence of RSVC associated with PLSVC can be further demonstrated by injecting saline contrast though an intravenous catheter in the right arm. In these cases, the coronary sinus will still opacify before the right atrium. Diagnosis of PLSVC and absent RSVC can be confirmed by venography, CT, or magnetic resonance imaging.[9,12,13]

PLSVC draining into the right atrium via the coronary sinus is not hemodynamically significant, and patients are generally asymptomatic. Therefore, surgical correction or other treatment is not needed.[14] However, it is still important to recognize this condition, particularly if a surgical procedure is planned. PLSVC may make it difficult to insert central venous catheters and pacemaker leads or affect the performance of coronary artery bypass grafting and other cardiac surgeries.[7,15] Patients with PLSVC are more likely to have other congenital defects and may also be associated with rhythm abnormalities, such as sinus node dysfunction and atrioventricular block.[2,16-18]

In patients with PLSVC with absent RSVC, all the blood from the cranial aspect of the body drains via the left superior vena cava, but this does not have any significant hemodynamic consequences.[5,18] Bartram et al. reviewed 121 patients with PLSVC associated with absent RSVC and found that 46% had other cardiac malformations. The most common defects included atrial septal defects (16%), endocardial cushion defects (11%), and tetralogy of Fallot (9%). Notably, 54% of the patients did not have other cardiac malformations, suggesting that PLSVC associated with absent RSVC is often seen in patients without other congenital heart disease.[18]

In patients with PLSVC who require a pacemaker or defibrillator, the device may be inserted through the right subclavian vein to avoid technical difficulties associated with the abnormal anatomy of the left venous system. However, in patients with both PLSVC and absent RSVC, venous access should be limited to the left side. Alternatively, use of epicardial leads or more recent advances such as subcutaneous ICD implantation may be considered.[19,20]

CONCLUSION

PLSVC should be considered when a dilated coronary sinus is noted on echocardiogram. It may be confirmed by intravenous injection of saline contrast in the left upper extremity with filling of the coronary sinus and then the right atrium. PLSVC may be associated with an absent RSVC, with the right subclavian vein draining into the left superior vena cava. This can be established by intravenous injection of saline contrast in the right upper extremity with similar filling of the coronary sinus before the right atrium. Although the combination of PLSVC with absent RSVC is reportedly uncommon, it is very important to recognize this combination in patients undergoing placement of central catheters, insertion of pacemaker leads, or surgical procedures. Therefore, we recommend that all patients with a diagnosis of PLSVC by echocardiography should also undergo saline contrast injection through the right arm to ascertain the presence or absence of RSVC.

Conflict of Interest Disclosure:
The authors have completed and submitted the *Methodist DeBakey Cardiovascular Journal* Conflict of Interest Statement and none were reported.

Keywords:
persistent left superior vena cava, congenital, venous drainage, right superior vena cava

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