

IMAGING VIGNETTE

INTERMEDIATE

CLINICAL VIGNETTE

Persistent Left Superior Vena Cava Draining Directly Into the Left Atrium With Occluded Coronary Sinus



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ABSTRACT

Combined persistent left superior vena cava entering the left atrium with a congenitally atretic coronary sinus is a rare imaging finding. In the absence of a significant right-to-left shunt, it is generally asymptomatic and can be an incidental discovery. Assessing the anatomy of the cardiac vasculature is crucial before transcatheter cardiac procedures.

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A 43-year-old woman presented with exertional shortness of breath and palpitations. Her past medical history included complete heart block with dual-chamber pacemaker (PM) placement at the age of 40 years, paroxysmal atrial fibrillation, supraventricular tachycardia, and ventricular septal defect repair at 2 years of age. Transthoracic echocardiography performed revealed newly reduced left ventricular ejection fraction from 60% to 40% over the past year with interventricular septum dyskinesia consistent with a paced rhythm. Device interrogation revealed a normally functioning dual-chamber PM with >99% right ventricular pacing. A decision was made to proceed with revision to a biventricular PM to allow ventricular synchrony by implantation of a coronary sinus (CS) lead. During the procedure, after revision of the generator, numerous failed attempts were made to access the right atrium (RA) using a deflectable catheter through the left subclavian vein. Left upper extremity venography exhibited a persistent left superior vena cava (PLSVC) suspected to be entering the RA through the CS. However, navigation of the CS was unsuccessful. As a result, the previously implanted leads were plugged into respective ports of the new generator, and a third left ventricular epicardial lead was plugged in anticipating future surgical correction and fixation.

With these concerns, the patient was admitted for further cardiac imaging evaluation. Transesophageal echocardiography demonstrated an abnormal blood vessel in the left atrium (LA) between the left atrial appendage and the left upper pulmonary vein. Agitated saline injection into left arm confirmed the presence of an anomalous systemic venous return into the LA (**Figures 1A and 1B**). This resounded a concern for an unroofed coronary sinus. For further clarification, cardiac computed tomography angiography (CTA) was performed with a selected left superior vena cava injection (**Figures 1C to 1F, Videos 1 and 2**), which unexpectedly revealed a PLSVC from the left brachiocephalic vein into the LA posteriorly (**Supplemental Figure 1A to 1D**). Normal

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**ABBREVIATIONS
AND ACRONYMS****CS** = coronary sinus**CTA** = computed tomography
angiography**LA** = left atrium**PLSVC** = persistent left
superior vena cava**PM** = pacemaker**RA** = right atrium

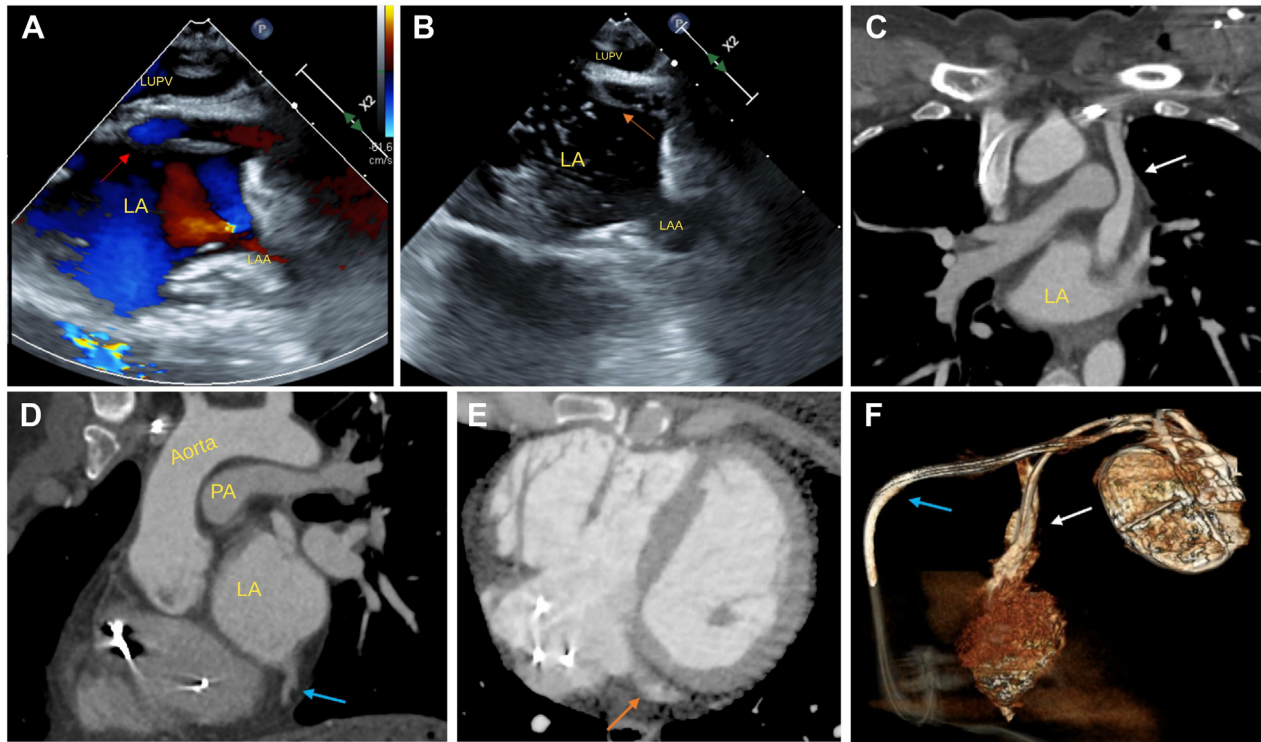
anatomy of the left innominate vein and the right-sided superior vena cava was observed. An atretic CS was noted in the posterior atrioventricular groove, and cardiac venous drainage through small veins was noted in the LA.

A PLSVC is a rare cardiac anomaly secondary to failure of regression of the left common cardinal and left superior cardinal veins embryonically. The estimated prevalence among the general population is 0.2% to 3%.¹ Commonly, the PLSVC drains into the RA through the expanded CS with different types identified on CTA.² Rarely, the PLSVC drains into the LA via an unroofed CS.³ Uniquely in our patient, the PLSVC drained into the LA with an atretic coronary sinus and no LA-to-RA communication.

A PLSVC is usually asymptomatic and is detected incidentally, particularly when it drains into the RA and without a significant right-to-left shunt. Our patient underwent a treadmill stress test after encountering challenges with the upgrade and maintained O₂ saturation >95% despite 7 minutes on the treadmill. Cardiac arrhythmias and paradoxical embolization are linked with a PLSVC.¹

Crowdedness of the right superior vena cava with previously implanted leads and a relatively small-sized right superior vena cava in the presence of double vena cava drainage were most likely the reasons for failed passage of the catheter into the RA. A transthoracic echocardiography short suprasternal axis view can identify a PLSVC. When in doubt, diagnosis can be confirmed through transesophageal echocardiography with agitated saline contrast injection into the left arm and cardiac CTA.

In conclusion, a PLSVC is usually an incidental and rare finding. It can complicate invasive procedures such as cardiac resynchronization procedures, particularly when associated with CS atresia.

FIGURE 1 Diagnostic Imaging Modalities Showing the Persistent Left Superior Vena Cava

(A) A midesophageal transesophageal echocardiography view showing an anomalous persistent left superior vena cava (PLSVC) (red arrow) between the left atrial appendage and the left upper pulmonary vein. (B) Agitated saline injection into left arm was detectable first in the left atrium (orange arrow) through the PLSVC. (C and D) Cardiac computed tomography angiograms demonstrated a PLSVC (white arrow) originating from the left brachiocephalic vein into the left atrium, coronary sinus atresia, and coronary venous drainage to the left atrium (blue arrow). (E) Cardiac computed tomography angiograms exhibiting the coronary sinus atresia (orange arrow). (F) A 3-dimensional reconstructed image from left upper extremity venography showing the PLSVC (white arrow) and PM leads passing through the right superior vena cava to the right ventricle (blue arrow).

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KEY WORDS cardiac resynchronization therapy, congenital heart disease, persistent left superior vena cava, right-to-left shunt, unroofed coronary sinus

 **APPENDIX** For supplemental videos and a figure, please see the online version of this paper.