

Late Presentation of Isolated Persistent Left Superior Vena Cava with Absent Right Superior Vena Cava Manifesting as Heart Failure: A Case Report

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Background: Persistent left superior vena cava (PLSVC) with absent right superior vena cava (RSVC) is a rare congenital venous anomaly, typically asymptomatic and discovered incidentally. In rare cases, it may contribute to hemodynamic disturbances and arrhythmias, potentially leading to heart failure, especially when associated with other cardiac anomalies or conduction abnormalities.

Case Presentation: We report the case of a 45-year-old male who presented with progressive symptoms of heart failure, including dyspnea, fatigue, and reduced exercise tolerance. Initial workup revealed an unexplained dilated coronary sinus and reduced left ventricular function with dilated heart chambers. Further imaging with contrast-enhanced echocardiography and CT angiography confirmed the presence of a PLSVC draining into the coronary sinus with absence of right superior vena cava. No other congenital heart defects were identified like coronary sinus atrial septal defect (ASD). The delayed diagnosis of PLSVC likely contributed to chronic atrial stretch, arrhythmia development- atrial fibrillation in this case, and eventual heart failure symptoms.

Conclusion: This case highlights the importance of considering vascular anomalies like PLSVC in the differential diagnosis of unexplained heart failure or dilated coronary sinus. Early detection through appropriate imaging can guide management and potentially prevent progression of symptoms.

Keywords: persistent left superior vena cava, PLSVC, echocardiography, atrial fibrillation, heart failure

Introduction

Persistent left superior vena cava (PLSVC) is the most common congenital anomaly of the thoracic venous system, occurring in approximately 0.3–0.5% of the general population and up to 12% in individuals with congenital heart disease. A much rarer variant, isolated PLSVC with absent right superior vena cava (RSVC), has a reported prevalence of 0.07% to 0.13% in the general population.^{1–3} While often asymptomatic and incidentally discovered during imaging or procedures, PLSVC can present with various clinical manifestations, including arrhythmias.^{1,4} The association between PLSVC and atrial fibrillation is rare but clinically significant, as the anomalous venous return may influence the heart's

conduction system and serve as a source of ectopic electrical activity, potentially leading to rhythm disturbances.^{5,6} Understanding this relationship is crucial for accurate diagnosis and effective management of patients presenting with unexplained arrhythmias, especially when standard workups fail to identify a structural or ischemic cause. In such cases, recognizing PLSVC as a potential contributor allows for targeted imaging, better risk stratification, and tailored therapeutic strategies.⁷

Case Presentation

A 45-year-old male patient presented to the hospital with complaints of progressive symptoms of palpitation, dyspnea, fatigue, and reduced exercise tolerance. There was no history of chest pain, syncope, or cyanosis. No clear triggers were identified, but symptoms were exacerbated by exertion. The patient had no significant past medical history of hypertension, diabetes mellitus, or thyroid disorders. There was no family history of arrhythmias, sudden cardiac death, or congenital heart defects. He was a non-smoker, did not consume alcohol, and had no history of illicit drug use. On physical examination, the patient appeared mildly distressed due to dyspnea. His vital signs were as follows: blood pressure of 110/75 mmHg, heart rate of 110 beats per minute with an irregular rhythm, respiratory rate of 22 breaths per minute, and oxygen saturation of 96% on room air. Cardiovascular examination revealed no visible jugular venous distention, no parasternal heave, and an apex beat was laterally displaced. Auscultation revealed normal S1 and S2 heart sounds, with no obvious murmurs, but an irregular cardiac rhythm was noted. Respiratory examination was unremarkable with clear breath sounds and no signs of pulmonary congestion. There were no signs of peripheral edema, hepatomegaly, or neurological deficits.

An initial electrocardiogram (ECG) revealed an irregular rhythm suggestive of atrial fibrillation (Figure 1). Echocardiography showed a reduced left ventricular ejection fraction (35–40%), enlargement of the heart chambers, moderate mitral regurgitation, and a notably dilated coronary sinus—findings that raised suspicion for a persistent left superior vena cava (PLSVC). To confirm this, a bubble contrast study was conducted, which demonstrated contrast filling the coronary sinus from the left side, consistent with PLSVC drainage (Figure 2A and B). Further imaging with cardiac CT angiography revealed the presence of a persistent left superior vena cava (PLSVC) draining into a markedly dilated coronary sinus, accompanied by

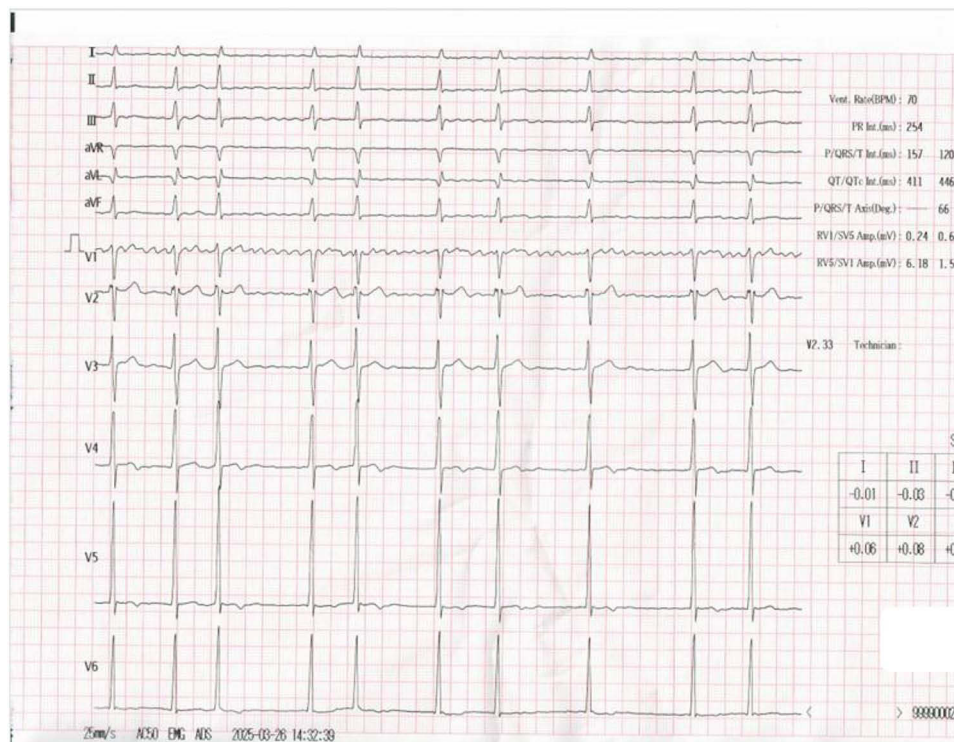


Figure 1 Electrocardiography showing atrial fibrillation with normal ventricular response.

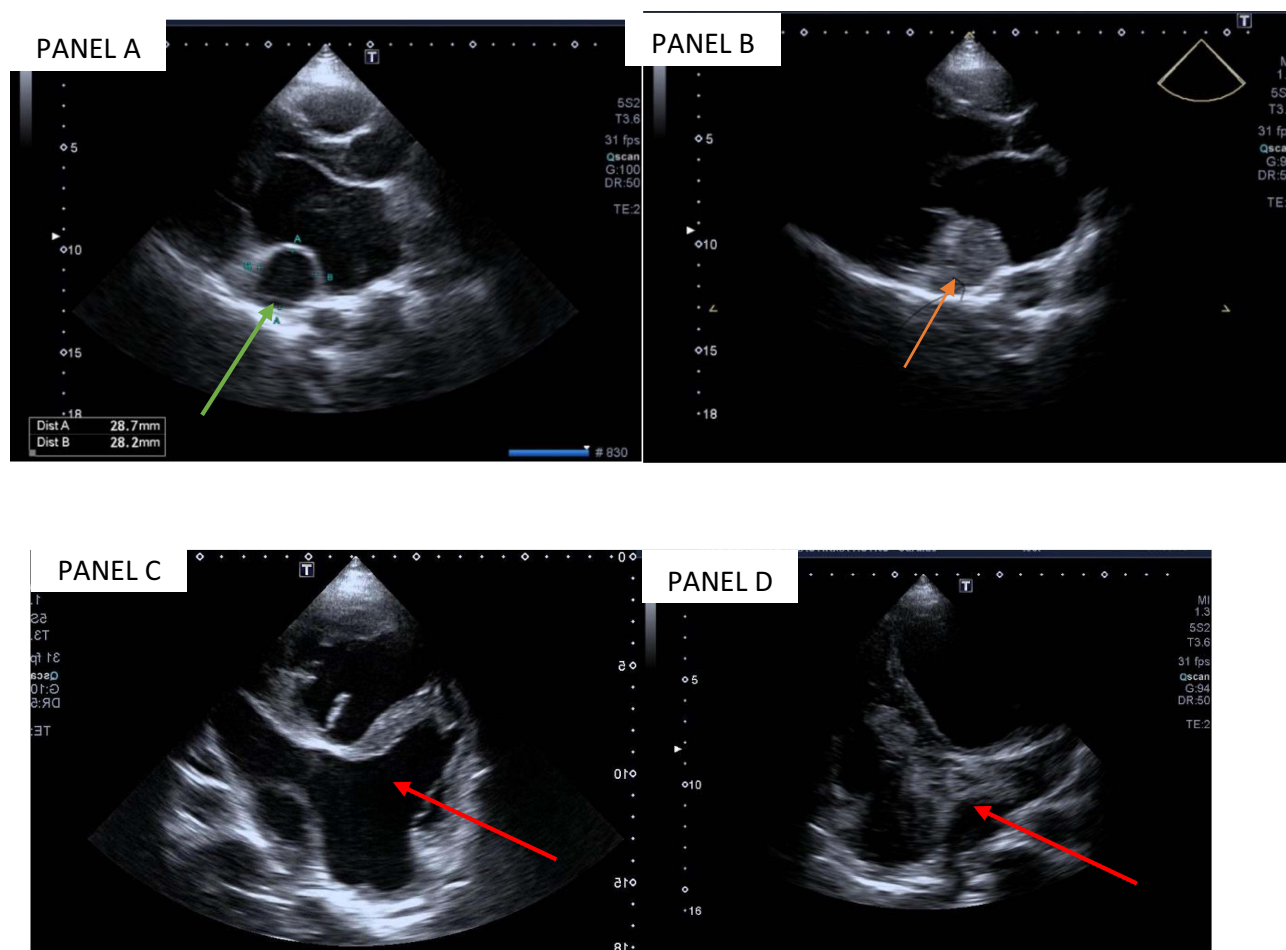


Figure 2 Echocardiography: (A) Parasternal short axis view showing a dilated Coronary sinus (28x28mm) with dilated left atrium (green arrow). (B) A bubble study showing direct filling of Coronary sinus from left system (yellow arrow). (C) Angulated apical 4 chamber view showing dilated coronary sinus (red arrow). (D) A bubble appearance in CS before right atrium (red arrow).

complete absence of the right superior vena cava. Instead, the left and right innominate veins were observed converging to form a prominent left SVC, which drained into the right atrium via the coronary sinus. These anatomical findings confirmed the venous anomaly and excluded other structural cardiac abnormalities. The axial CT view (Figure 3) demonstrates coronary sinus dilatation (red arrow) and the course of the left superior vena cava (green arrow). Coronal and sagittal reconstructions (Figure 4) illustrate the left and right innominate veins (blue arrow) merging to form the left SVC (red arrow), which ultimately drains into the right atrium via the coronary sinus (yellow arrow).



Figure 3 Axial CT image showing dilated coronary sinus (A; red arrow) and the course of the left SVC (B, C; green arrow).

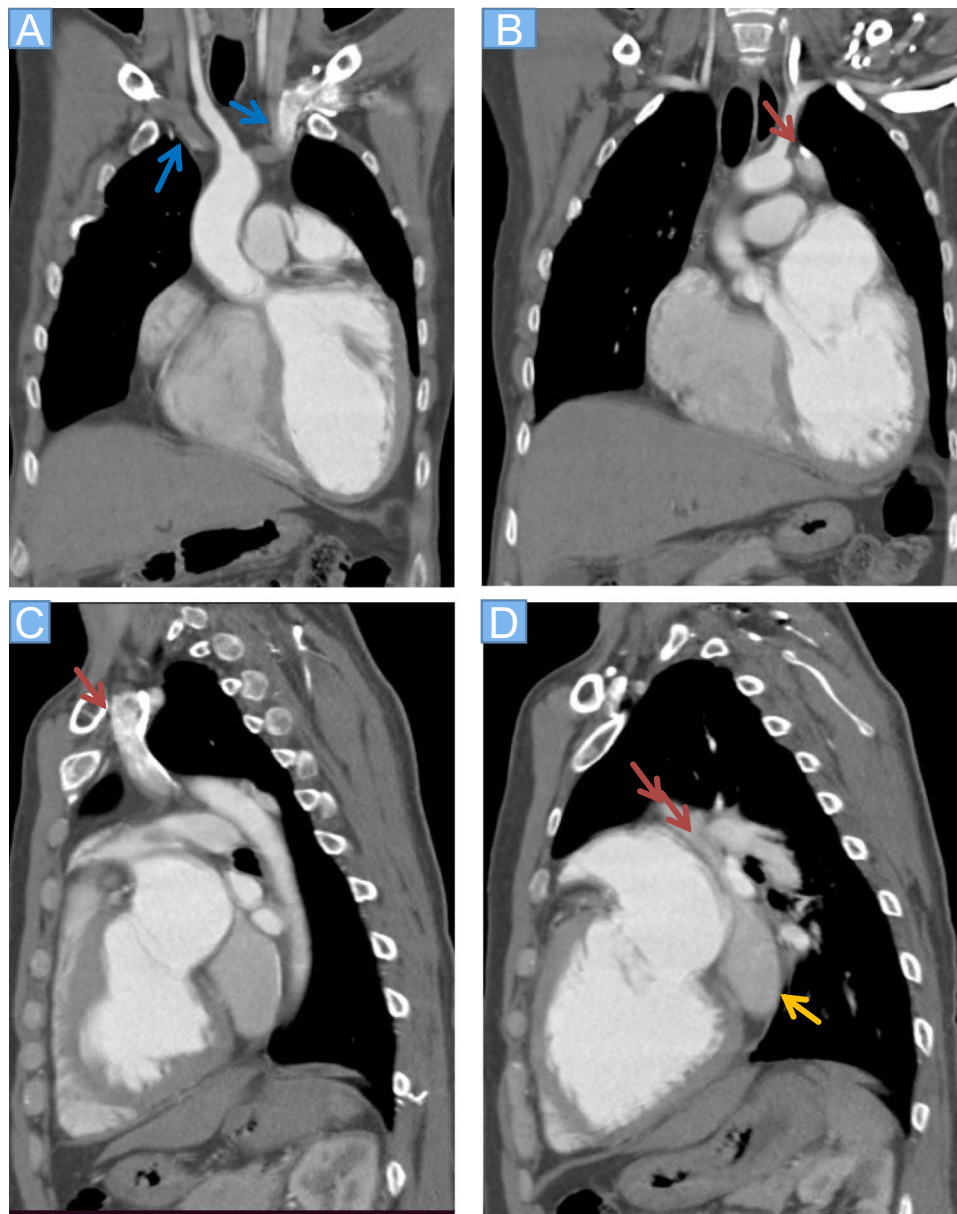


Figure 4 Coronal and sagittal CT views showing innominate veins (A; blue arrow) merging into the left SVC (B-D; red arrow) draining into the coronary sinus (D; yellow arrow).

Full laboratory tests including thyroid function tests and serum electrolytes were within normal limits. The patient was managed with rate control using beta-blockers, and anticoagulation therapy was initiated due to the presence of atrial fibrillation. After stabilization, he was referred for an electrophysiological study (EPS) to assess the conduction abnormalities associated with PLSVC.

This case highlights a rare presentation of persistent left superior vena cava with absent of right superior vena cava manifesting with arrhythmia. Although isolated persistent left superior vena cava (PLSVC) with absent right superior vena cava (RSVC) is often asymptomatic and discovered incidentally, its association with conduction abnormalities—particularly atrial arrhythmias—highlights the importance of comprehensive cardiovascular evaluation in patients presenting with unexplained rhythm disturbances. Early recognition of this rare vascular anomaly is essential for guiding appropriate management and minimizing the risk of procedural complications, thereby optimizing patient outcomes.

Discussion

PLSVC results from aberrant anterior cardinal venous system development during embryonic development. The right anterior cardinal vein forms the right SVC while the left anterior cardinal vein typically regresses by the tenth week of pregnancy. In approximately 90% of cases, a persistent left superior vena cava (PLSVC) drains into the right atrium through the coronary sinus, whereas in 10–20% of cases, it drains directly into the left atrium or pulmonary veins, causing left-to-right shunting, cyanosis, or paradoxical embolism.^{8–10} This abnormality is often linked to other congenital conditions that worsen haemodynamic effects, like unroofed coronary sinus or atrial septal defects (ASD),¹¹ ventricular septal defects, bicuspid aortic valves, coarctation of the aorta, coronary sinus atresia, abnormal pulmonary venous return, and tetralogy of Fallot.¹² The association between PLSVC and AF is well-documented. The presence of myocardial sleeves extending into the PLSVC can serve as arrhythmogenic foci, potentially initiating and sustaining AF. In the case presented, a 41-year-old patient experienced progressive shortness of breath and palpitations, leading to the diagnosis of PLSVC with associated AF.

Advanced imaging modalities play a crucial role in diagnosing PLSVC. Transthoracic echocardiography may reveal a dilated coronary sinus, prompting further investigation. Definitive diagnosis is often achieved through computed tomography (CT) or magnetic resonance imaging (MRI), which can delineate the anomalous venous anatomy. In this case, CT angiography confirmed the presence of PLSVC draining into the right atrium, facilitating appropriate management strategies.¹³

Although central venous catheterization is generally straightforward in patients with a normal right superior vena cava (RSVC), the procedure becomes technically challenging in the absence of the RSVC. In such cases, left-sided venous access is required, which increases the risk of catheter malposition. Similarly, pacemaker or implantable cardioverter-defibrillator (ICD) lead placement, typically uncomplicated via the right SVC, becomes more complex and technically demanding when advancement must occur solely through the left-sided persistent superior vena cava (PLSVC).^{14,15}

Management of AF in the context of PLSVC often involves catheter ablation, targeting the arrhythmogenic sites within the PLSVC. Recent advancements have introduced pulsed-field ablation (PFA) as a novel technique with promising outcomes. PFA utilizes non-thermal energy to achieve myocardial tissue ablation, potentially reducing collateral damage to surrounding structures. Studies have reported successful isolation of the PLSVC using PFA, resulting in effective arrhythmia control.¹⁶

Conclusion

Although PLSVC is often an incidental finding, early recognition is crucial—particularly in patients presenting with unexplained arrhythmias or a dilated coronary sinus. A thorough diagnostic workup, including multimodal imaging, is essential for accurate identification. Given its potential role in arrhythmogenesis, especially atrial fibrillation, timely detection of PLSVC enables appropriate therapeutic planning. Advances in ablation techniques, such as pulsed-field ablation (PFA), offer promising treatment options, underscoring the importance of targeted intervention in reducing the burden of recurrent AF.

Ethics Approval

Based on the regulations of the review board of Mogadishu Somali Türkiye Training and Research Hospital, institutional review board approval is not required for case reports.

Consent for Publication

Written informed consent was obtained from the patient's daughter for publication of this case report and accompanying images.

Author Contributions

All authors made a significant contribution to the work reported, whether in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas took part in drafting, revising, or critically reviewing

the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

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Disclosure

The authors report no conflicts of interest in this work.

References

1. Corici OM, Gaşpar M, Mornoş A, et al. Cardiac arrhythmias in patient with isolated persistent left superior vena cava. *Curr Health Sci J.* 2017;43(2):163.
2. Goyal SK, Punnam SR, Verma G, et al. Persistent left superior vena cava: a case report and review of literature. *Cardiovascu Ultrasound.* 2008;6(1):1–4. doi:10.1186/1476-7120-6-50
3. Sanku K, Nemalikanti S, Colna M, et al. A rare case of an isolated persistent left superior vena cava. *J Ame College Cardiol.* 2024;83(13_Supplement):3128. doi:10.1016/S0735-1097(24)05118-0
4. Bisoyi S, Jagannathan U, Dash A, et al. Isolated persistent left superior vena cava: a case report and its clinical implications. *Ann Cardiac Anaesth.* 2017;20(1):104–107. doi:10.4103/0971-9784.197847
5. Pavai J, Nayak S. A persistent left superior vena cava. *Singapore Med J.* 2007;48(3):e90–3.
6. Morgan LG, Gardner J, Calkins J. The incidental finding of a persistent left superior vena cava: implications for primary care providers—case and review. *Case Rep Medicine.* 2015;2015:198754. doi:10.1155/2015/198754
7. Mingyang GAO, Bian Y, Huang L, et al. Catheter ablation for atrial fibrillation in patients with persistent left superior vena cava: case series and systematic review. *Front Cardiovascu Med.* 2022;9:1015540. doi:10.3389/fcvm.2022.1015540
8. Kesieme EB, BUCHAN KG. Clinical anatomy of the coronary venous system and relevance to retrograde cardioplegia and cardiac electrophysiological interventions. *Clin Anat.* 2025;38(1):43–53. doi:10.1002/ca.24195
9. Azizova A, Onder O, Arslan S, et al. Persistent left superior vena cava: clinical importance and differential diagnoses. *Insights Imaging.* 2020;11(1):1–19. doi:10.1186/s13244-020-00906-2
10. Nguyen Duy T, Nguyen Van L, Pham Phuong Thao A, et al. Transvenous dual-chamber pacemaker implantation in a patient with persistent left superior vena cava undergoing maintenance hemodialysis. *Inter Med Case Rep J.* 2025;273–279.
11. Batouty NM, Sobh DM, Gadelhak B, et al. Left superior vena cava: cross-sectional imaging overview. *La radiologia medica.* 2020;125(3):237–246. doi:10.1007/s11547-019-01114-9
12. Stiver C, Ball MK, Cua CL. Pulmonary venous anomalies. In: Anderson RH, Bacha E, Webb G, editors. *Pediatric Cardiology: Fetal, Pediatric, and Adult Congenital Heart Diseases.* Cham: Springer International Publishing; 2024:1377–1419.
13. Turagam MK, Atoui M, Atkins D, et al. Persistent left superior vena cava as an arrhythmogenic source in atrial fibrillation: results from a multicenter experience. *J Interv Card Electrophysiol.* 2019;54(2):93–100. doi:10.1007/s10840-018-0444-x
14. Kaur S, Firdaus S, Solano J, et al. Incidental finding of a persistent left superior vena cava during permanent dual-chamber pacemaker implantation: a case report. *Cureus.* 2024;16(11). doi:10.7759/cureus.72865
15. Kasarla R, Jordan D, Elias M, et al. Persistent left superior vena cava: clinical and procedural challenges. *Cureus.* 2025;17(4).
16. Menè R, Sousonis V, Combes S, et al. Pulsed field ablation of a persistent left superior vena cava in recurrent paroxysmal atrial fibrillation and its effect on the mitral isthmus: a case report. *HeartRhythm Case Rep.* 2024;10(1):6–10. doi:10.1016/j.hrcr.2023.10.009

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