# Percutaneous Correction of a Large Left Superior Vena Cava to Left Atrium Fistula

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## INTRODUCTION

Among the multiple etiologies of a right-to-left shunt, an anomalous venous connection with the left atrium (LA) is a rare cause. Such defects usually occur in conjunction with other cardiac anomalies. A persistent left-sided superior vena cava (PLSVC) is a developmental anatomical defect wherein 80% to 90% of cases bear a connection with the right side of the heart.[1] These patients are usually asymptomatic. However, very rarely, the left-sided superior vena cava (SVC) empties into the LA instead. This malformation has largely been reported in young patients and has been linked to a range of potential complications. We present a rare case of an anomalous fistula between the PLSVC and the LA.

## CASE PRESENTATION

A 63-year-old woman with multiple previous small cerebrovascular accidents presented to the adult congenital cardiology clinic with shortness of breath that started 1 year prior. The patient was acyanotic with an oxygen saturation of 92% on room air at rest and 84% with exertion. Blood pressure was 160/84 mm Hg in both arms. The physical examination was unremarkable, with no S3 gallop, rales, or edema of legs. An electrocardiogram demonstrated sinus rhythm with RSR′ pattern in V1 (Figure 1). Laboratory testing revealed a hemoglobin of 14.5 g/dL, hematocrit of 46%, and N-terminal pro b-type natriuretic peptide of 89 pg/mL.

Transthoracic echocardiography (TTE) with injection of bubbles in the left antecubital fossa showed early rapid appearance of bubbles in the LA suggestive of a right-to-left shunt (Video 1, Figure 2). Remarkably, although the LA was severely dilated (>48 mL/m$^2$), the right ventricle had normal size and function without hypertrophy.

A cardiac computed tomography (CCT) scan demonstrated a PLSVC entering the posteriosuperior aspect of the LA immediately anterior to the upper left pulmonary vein (Figure 3). A bridging vein between the right SVC and PLSVC was present (Figure 4, Video 2).

A cardiovascular magnetic resonance scan showed that the coronary sinus (CS) was intact, but the PLSVC had a large fistula emptying into the LA. Given the patient's dyspnea, hypoxemia, and previous cerebrovascular accidents, we recommended they have the fistula closed. Given the high surgical risk, a shared decision was made with the patient to address the fistula via a percutaneous approach. We obtained vascular access in the left internal jugular vein with an 11-French sheath. Right heart catheterization showed normal left- and right-sided filling pressures, normal cardiac output, normal pulmonary artery pressure, and normal pulmonary vascular resistance. No significant shunt was detected. A hand injection of contrast into the PLSVC revealed a bridging vein as well as an opening between the PLSVC and the LA (Video 3). We advanced a 24 mm sizing balloon to the fistula. The balloon was inflated, and cineangiography was performed. The balloon revealed a noncircular opening that did not appear optimal for device closure (Figure 5).

Therefore, we decided to use a covered stent. We placed an expanded polytetrafluoroethylene-covered stent graft (16 mm × 14.5 mm × 7 cm) at the distal portion of the PLSVC covering the fistula into the LA. We were careful to avoid the cardiac veins. Postprocedure angiography showed that the fistula had been largely covered. However, a small opening at the distal end of the stent remained, which we initially felt was inconsequential (Video 4). Transthoracic echocardiography with a bubble study was also performed postprocedure, which noted clinical improvement with only a few bubbles entering the left ventricle at the end of 3 cardiac cycles. The patient received a loading dose of aspirin and clopidogrel after the procedure and was subsequently discharged on aspirin 81 mg orally daily and clopidogrel 75 mg orally daily with follow-up scheduled at 1 month. The patient was admitted to the hospital twice within the next month with continued dyspnea and hypoxemia. Consequently, another catheterization procedure was performed. Angiography showed abundant collaterals from the lateral cardiac vein to the middle cardiac vein indicating that covering the lateral cardiac vein would not compromise venous drainage (Video 5). We therefore placed another polymer-based stent graft (22 mm × 45 mm) from the distal edge of the previous stent into the CS. At completion of the procedure, angiography demonstrated no residual defect (Video 6). A TTE after percutaneous intervention showed minimal bubbles appearing in the LA with injection through the left arm (Video 7). The patient's symptoms and hypoxemia resolved; they have had no further complaints for 3 years of clinical follow-up.

## DISCUSSION

Persistent left-sided superior vena cava is a congenital anomaly of the thoracic venous system with an incidence of 0.3% to 0.5% in the

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## VIDEO HIGHLIGHTS

**Video 1:** Two-dimensional TTE, 4-chamber view, demonstrates rapid appearance of bubbles in the LA suggesting right-to-left shunt.

**Video 2:** Three-dimensional reconstruction of CCT scan demonstrating the PLSVC and bridging vein in *dark blue* and right-sided SVC in *light blue*.

**Video 3:** Angiography during percutaneous intervention reveals a bridging vein between the PLSVC and the RSVC.

**Video 4:** Angiography after placement of an expanded polytetrafluoroethylene-covered stent graft demonstrates a persistent small opening at the distal end of the stent.

**Video 5:** Angiography reveals abundant collaterals from the lateral cardiac vein to the middle cardiac vein.

**Video 6:** Angiography after placement of a polymer-based stent graft demonstrates no residual defect.

**Video 7:** Two-dimensional TTE, 4-chamber view after percutaneous intervention, demonstrates minimal appearance of bubbles in the LA.

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general population.[2] Many cases with PLSVC also have other associated cardiac malformations, such as atrial septal defect, endocardial cushion defect, or tetralogy of Fallot.[3] Our case demonstrates a large fistula connecting the PLSVC to the LA in an older patient, which is a rare finding. Understanding the development of the fetal cardiovascular system is central to understanding the pathophysiology of PLSVC. The primitive fetal cardiovascular system consists of 3 distinct pairs of major veins, namely, the umbilical veins, the vitelline veins, and the cardinal veins. The anterior and posterior cardinal veins on each side merge to form the right and left common cardinal veins. Anastomoses begin to appear between the right and left anterior cardinal veins, which leads to the formation of the left brachiocephalic vein. The portion of the right anterior cardinal vein caudal to the anastomosis along with the right common cardinal vein forms the normal right-sided SVC. The segment of the left anterior cardinal vein caudal to this anastomosis normally regresses. If this physiologic regression does not take place, a persistent left-sided vascular structure results, known as the PLSVC. The most common subtype of a left-sided SVC has a normal right-sided SVC present in 90% of cases. In our patient, we also found a bridging vein, which has been reported in 30% of cases with PLSVC.[4] Most cases of PLSVC drain into the right atrium via a dilated CS.[5,6] Most such patients are asymptomatic. If developmental arrest occurs at an earlier embryologic stage, the CS is absent and the PLSVC drains into the LA instead.

A PLSVC emptying into the LA is seen in about 7.5% of cases, and it leads to a right-to-left shunt and thus is often symptomatic.[7] More than half of the patients with a PLSVC are at risk of paradoxical embolism owing to a right-to-left shunt.[8] It is unclear whether our patient's previous strokes were due to paradoxical embolism. Paradoxical embolism may lead to development of a brain abscess.[9] A right-to-left shunt can also result in cyanosis[10] and heart failure.[11]

Owing to the anomalous connection, a PLSVC draining into the LA can affect procedures that involve getting access to the right side of the heart, such as right heart catheterization, Swan-Ganz catheter placement, and permanent pacemaker and implantable cardioverter-defibrillator placement.[6] In such cases, access to the right heart and CS should be performed via the right subclavian vein. There are

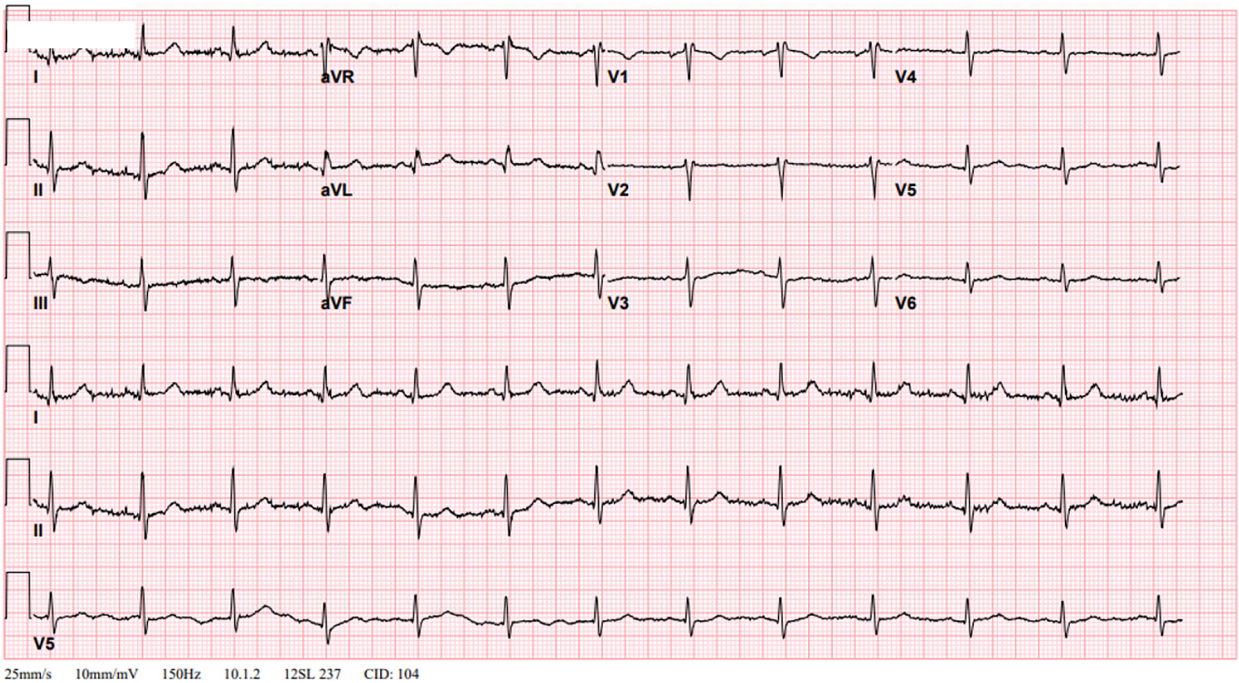


Figure 1 Electrocardiogram demonstrates sinus rhythm (ventricular rate 75 bpm) with RSR' pattern in V1.

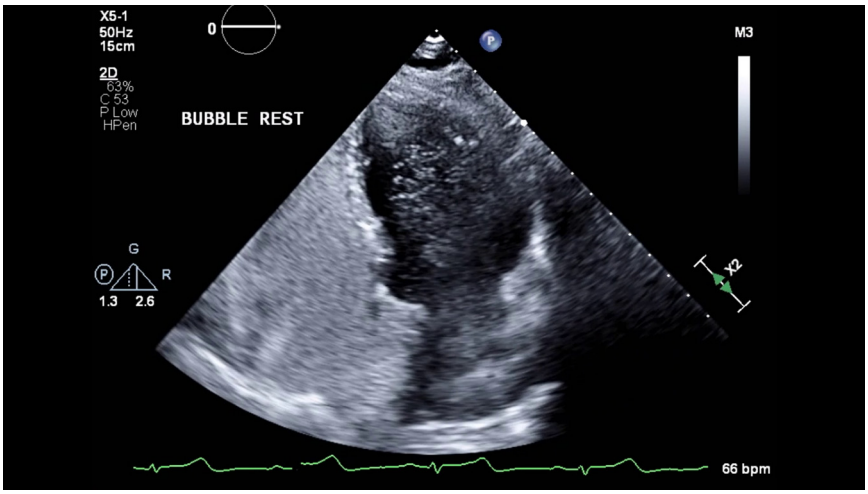


Figure 2 Two-dimensional TTE, apical 4-chamber diastolic display after injection of agitated saline via the left arm, demonstrates the early and rapid appearance of bubbles in the LA suggesting a right-to-left shunt.

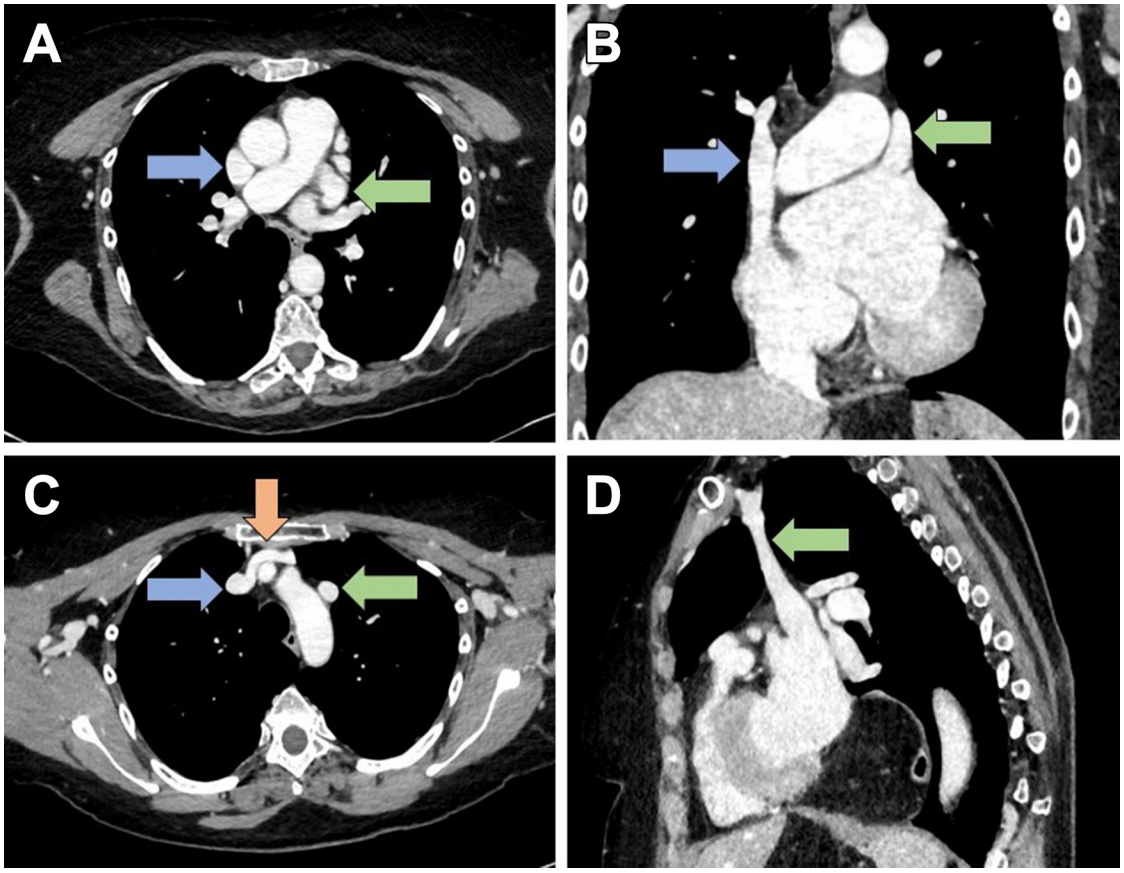


Figure 3 Cardiac computed tomography, dual-phase angiography with venous and arterial contrast, axial **(A, C)**, coronal **(B)**, and sagittal **(D)** displays, demonstrates the PLSVC (*green arrow*), right SVC (*blue arrow*), and bridging vein (*orange arrow*).

several other complications associated with the presence of a PLSVC. A PLSVC predisposes patients to potential arrhythmias including atrial fibrillation, owing to repetitive rapid discharges, and a shorter activation cycle length, due to multiple anatomical and electrical communications with the atria.[1] Additionally, catheterization of the CS through a PLSVC is much more likely to cause rhythm disturbances such as supraventricular tachycardia than catheterization through a normal right SVC.[12] The presence of a PLSVC also acts as an impediment to the administration of retrograde cardioplegia during cardiac surgery.[6]

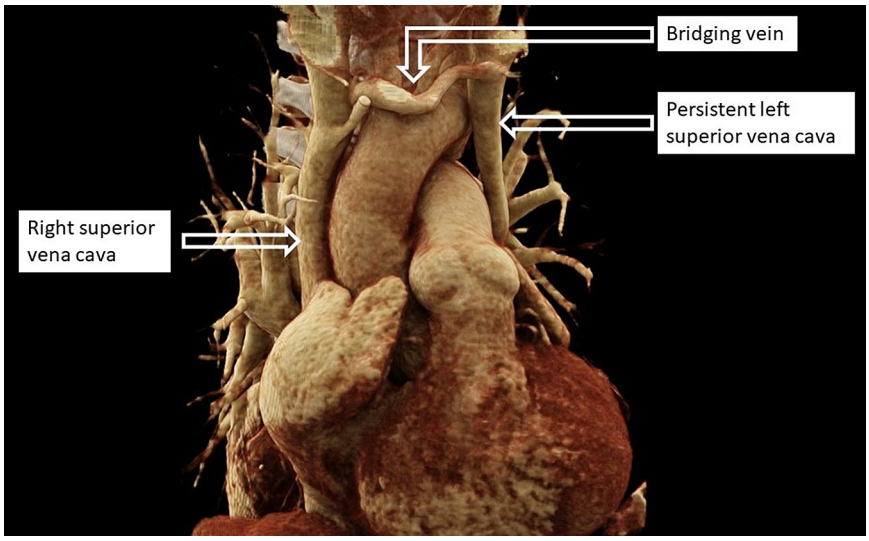


Figure 4 Three-dimensional CCT, whole-heart, volume-rendered reconstruction, demonstrates a bridging vein between the PLSVC and right SVC.

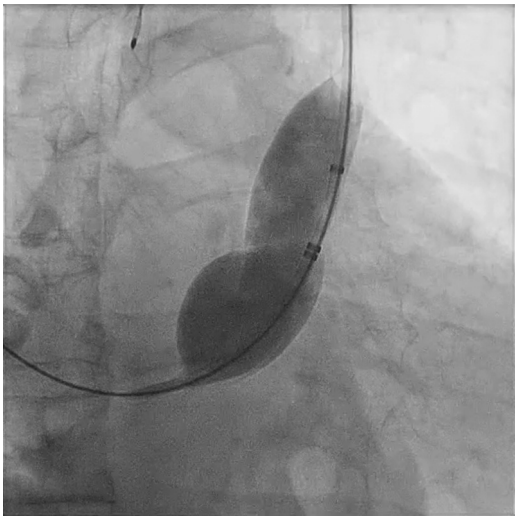

Figure 5 Angiogram demonstrates the 24 mm sizing balloon that was inflated across the defect revealing a noncircular opening.

## CONCLUSION

A PLSVC to LA fistula can present with shortness of breath and exertional hypoxemia. The use of multimodality imaging is essential for the diagnosis. Due to the serious nature of complications associated with PLSVC with a right-to-left shunt, correction or repair is indicated, which is usually surgical. Because of our patient's high surgical risk, the repair was done percutaneously using a covered stent. The patient had resolution of symptoms and continues to do well post-procedure.

## DATA AVAILABILITY

Data supporting this research article are available from the corresponding author or first author on reasonable request.

## REVIEW STATEMENT

Given his role as CASE Editor-in-Chief, Vincent L. Sorrell, MD, had no involvement in the peer review of this article. Full responsibility for the editorial process for this article was delegated to Benjamin Eidem, MD.

## CONSENT STATEMENT

Complete written informed consent was obtained from the patient (or appropriate parent, guardian, or power of attorney) for the publication of this study and accompanying images.

## ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

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## SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2023.12.023.

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