

MINI-FOCUS ISSUE: TRANSCATHETER INTERVENTIONS

ADVANCED

CASE REPORT: CLINICAL CASE

Transcatheter Closure of Persistent Left Superior Vena Cava Draining to Pulmonary Vein



A Rare Entity

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ABSTRACT

We describe a patient with recurrent embolic strokes who was found to have a persistent left superior vena cava draining into a pulmonary vein. Transcatheter placement of a vascular plug resulted in successful occlusion of the superior vena cava. Repeat bubble study on follow-up imaging was negative for a right-to-left shunt. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2020;2:1120-3) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

HISTORY OF PRESENTATION

A 67-year-old female patient with a recent prior right basal ganglia and corona radiata stroke 3 weeks earlier presented with new onset dysarthria to Delray Medical Center, Delray Beach, Florida. Her initial vital signs revealed a temperature pf 97.8°F,

heart rate 97 beats/min, RR of 18/min, and a blood pressure of 159/97 mm Hg. Magnetic resonance imaging confirmed an acute infarct in the left posterior frontal lobe. Intracranial and extracranial cerebral vascular imaging was nonrevealing. She was not a candidate for tissue plasminogen activator because of the timeline of the presentation. A transesophageal echocardiogram (TEE) at another facility with an agitated saline study had confirmed an intracardiac right-to-left shunt that was believed to be a patent foramen ovale, and a closure procedure was planned. She was receiving dual antiplatelet therapy. TEE with agitated saline, injected through the left arm, at our institution showed positive results, with nearly simultaneous bubbles in the right and left sides of the heart. Prolonged cardiac monitoring during previous hospitalization and in our facility was unremarkable.

LEARNING OBJECTIVES

- To review of a rare entity, the LSVC, as a differential diagnosis of embolic stroke.
- To understand the unique features of bubble study in LSVC as compared with a patent foramen ovale.
- To establish an overview of the treatment planning and feasibility of the percutaneous closure of LSVC.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the *JACC: Case Reports* [author instructions page](#).

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PAST MEDICAL HISTORY

The patient's past medical history included hypertension, dyslipidemia, Graves disease, and former smoking.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included a large atrial septal defect, a sinus venosus defect, and a persistent left superior vena cava (LSVC) draining either into the left atrium or into an unroofed coronary sinus.

INVESTIGATIONS

TEE failed to demonstrate an atrial septal defect or a sinus venosus defect. All pulmonary veins had normal drainage into the left atrium. Right-sided heart catheterization was performed, and saturation data were obtained. There was no saturation step-up to suggest a hemodynamically significant shunt. The superior cavoatrial junction was probed for an anomalous venous connection, but none was identified. Selective angiography of the left innominate vein revealed a persistent LSVC. Selective engagement and angiography of the LSVC demonstrated drainage into the left upper pulmonary vein (LUPV) and the left atrium (Figure 1A). Subsequently, a computed tomography angiogram with immediate and delayed images was obtained. This study confirmed an LSVC traveling anterior to the aortic arch and left pulmonary artery and ruled out other abnormalities (Figure 2A). Ultrasound Doppler imaging of the left arm, along with the bilateral lower extremities, did not show evidence of venous thrombosis.

MANAGEMENT

Because this patient had multiple embolic events despite medical therapy, she was scheduled for transcatheter closure of the LSVC (Figure 1B). The largest diameter of the LSVC measured 6.8 mm on angiography, a 10-mm Amplatzer Vascular Plug II (St. Jude Medical Inc., St. Paul, Minnesota) was deployed in the midsegment successfully. Subsequent angiography revealed no flow through the device, with excellent flow through the innominate vein to the right superior vena cava. The patient was discharged to rehabilitation on dual antiplatelet therapy.

FOLLOW-UP

The patient had an uncomplicated hospital course post-procedure and was subsequently transferred

to an acute rehabilitation facility. At 4-week follow-up, the patient was in stable condition, and a transthoracic echocardiogram showed normal cardiac structures and a negative bubble study result.

DISCUSSION

Embryonically, LSVC results from failure of left anterior cardinal vein to close. Congenital anomalies of the superior and inferior vena cava are rare and are usually incidentally recognized, with a prevalence of 0.07% to 8.7% (1). A persistent LSVC is estimated to be present in <0.5% of the general population, but it is present in approximately 4% in patients with congenital heart disease (2). In the absence of other congenital heart disease, LSVC typically drains into the coronary sinus, a less common variant of LSVC drains directly into the left atrium, and much rarer variant drains into the left atrium through left upper pulmonary vein (3). This form of LSVC is associated with embolic events through the left atrium (4). The anatomic course of LSVC is anterior to the left pulmonary artery (Figures 2A to 2C); this helps to distinguish it from the "levoatriocardinal vein" (vertical vein). The levoatriocardinal vein travels between the left pulmonary artery and left bronchus. It is a remnant of the splanchnic vein connecting the pulmonary venous plexus and the left atrium and is commonly recognized in association with left-sided obstructive heart lesions.

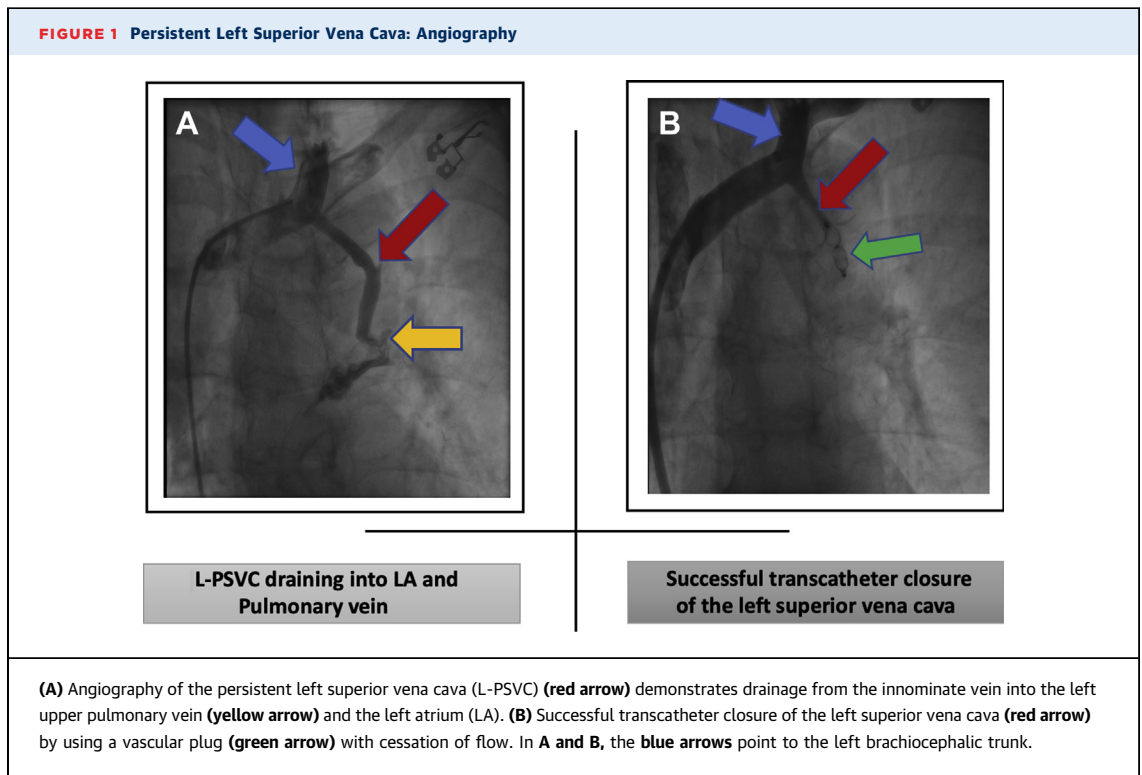
An LSVC may be occasionally detectable on a chest radiography by a focal widening of the mediastinum superior to the left side of the aortic knob. It could be revealed incidentally when it is traversed by an intravenous catheter (5) or with challenges when introducing pacemaker leads (6). An LSVC draining into the left atrium can be detected by injection of agitated saline into the patient's left arm and focusing on the timing of left atrial bubbles (7). A right arm agitated saline injection would fail to show the defect; and without attention to the timing of bubbles in the left atrium after a left arm saline injection, a false diagnosis of patent foramen ovale could be made, as was initially considered in our case. This could lead to either an unnecessary procedure or ineffective treatment planning to avoid future events. As with the general expert consensus that led to prior guidelines for comparable shunt lesions such as unroofed coronary sinus (8), closure of an LSVC with a left-sided

ABBREVIATIONS AND ACRONYMS

LSVC = left superior vena cava

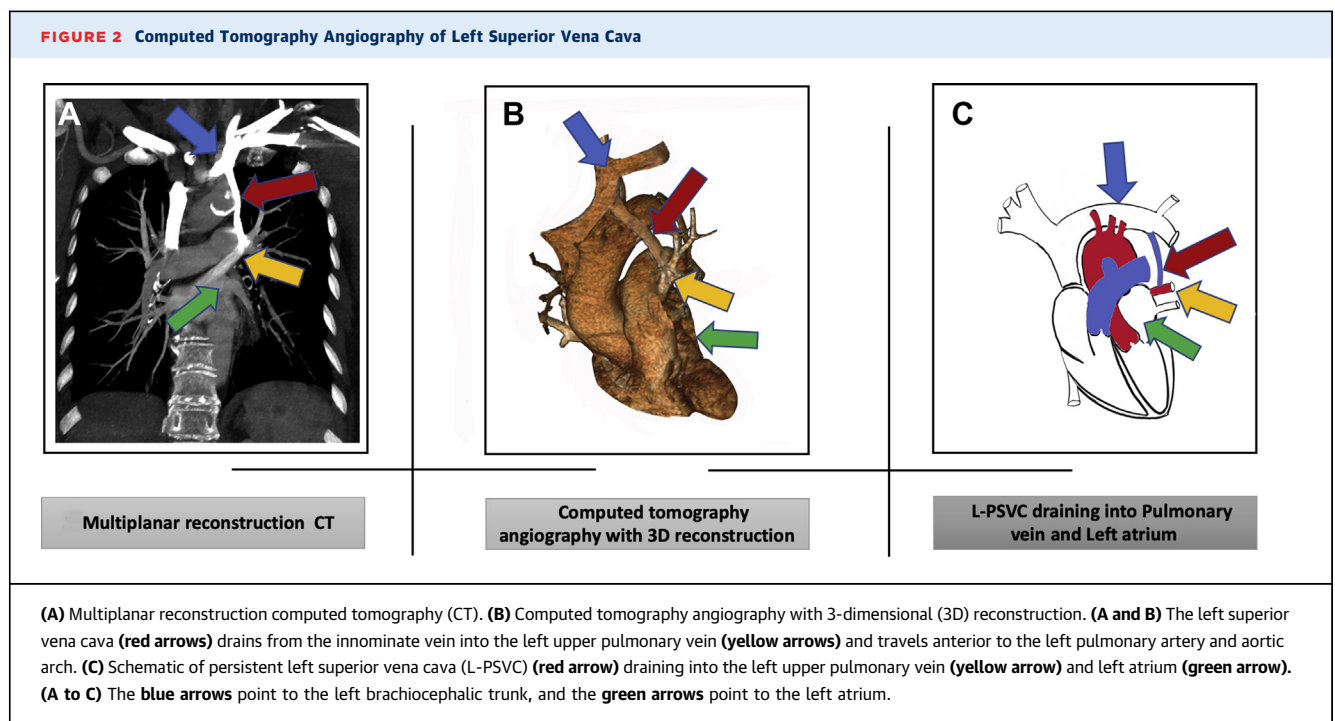
LUPV = left upper pulmonary vein

TEE = transesophageal echocardiogram



heart connection is reasonable: significant hemodynamic shunt (pulmonary-to-systemic flow ratio >1.5), exertional hypoxia, or embolic events. The transcatheter closure technique of the LSCV with

the use of vascular plug is similar to what is described in published reports (7,9). However, the more proximal location of the LSCV and LUPV confluence in this patient did require careful



deployment of the plug, to avoid jeopardizing LUPV drainage.

Transcatheter closure is a safe and feasible option in these patients.

CONCLUSIONS

This case represents a rare presentation of a persistent LSVC with drainage into the LUPV that was complicated by recurrent embolic events.

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KEY WORDS congenital anomalies, embolic stroke, persistent left superior vena cava, transcatheter closure