

## Beware of a duplicate superior vena cava

Christina Zhu BS, BA, Ferris Zeitouni MS, M Logan Warren DO, Ariel P Santos MD, MPH

### ABSTRACT

A duplicate superior vena cava (SVC) with a persistent left SVC (PLSVC) is present in 0.3% of the population with an incidence as high as 10–11% in patients with congenital heart disease. While a PLSVC is rare and usually asymptomatic, clinicians should be aware of this anomaly during central line placement due to potential complications. Our case involves a 76-year-old man with end-stage renal disease, on peritoneal dialysis (PD), who was admitted to our hospital for treatment of generalized peritonitis secondary to a descending colon perforation. He underwent left hemicolectomy and removal of an infected PD catheter. During preparation for discharge, he underwent hemodialysis catheter replacement. During the procedure, it was noted that the wire did not traverse the midline. A subsequent venogram with contrast showed a duplicate SVC draining into the coronary sinus. The catheter was inserted into the proximal left SVC and verified with fluoroscopy. One hour later, the patient went into atrial fibrillation and death ensued. Persistent left superior vena cava, if known or suspected, requires additional caution in central venous line placement to avoid potentially fatal complications, such as arrhythmias. Immediate removal and reassessment of an alternative access site should be pursued to avoid secondary complications.

**Keywords:** Persistent left superior vena cava, vascular malformations, catheterization, central venous catheter

### INTRODUCTION

While uncommon in the general population, congenital anomalies of the great vessels can lead to significant adverse consequences if unnoticed during catheter placement. A duplicate superior vena cava (SVC) with persistent left superior vena cava (PLSVC) is the most common central venous anomaly with an incidence of 0.3% in the general population. The incidence increases up to 10% in patients who have congenital heart disease.<sup>1–5</sup> A persistent left SVC occurs when the left common cardinal and precardinal veins fail to obliterate, resulting in a patent left SVC and duplicate SVC. Most PLSVCs drain into the coronary sinus and right atrium and are asymptomatic.<sup>1–5</sup> A duplicate SVC is

usually unrecognized and can cause confusion during catheter placement if the guidewire or catheter fails to cross the midline on imaging. Irritation of the coronary sinus from catheter placement can cause hypotension, arrhythmia, and myocardial ischemia. Duplicate SVCs that drain into the left atrium can introduce emboli into the systemic circulation. Due to these possible serious complications, clinicians should be aware of this anomaly during central venous line placement. We present a case of an incidentally discovered duplicate SVC with PLSVC during placement of a central venous catheter that led to atrial fibrillation and sudden death.

### CASE REPORT

A 76-year-old man with known end-stage renal disease on peritoneal dialysis (PD) presented to the emergency room for persistent abdominal pain with signs of peritonitis on physical examination; computed tomography of the abdomen showed free fluid

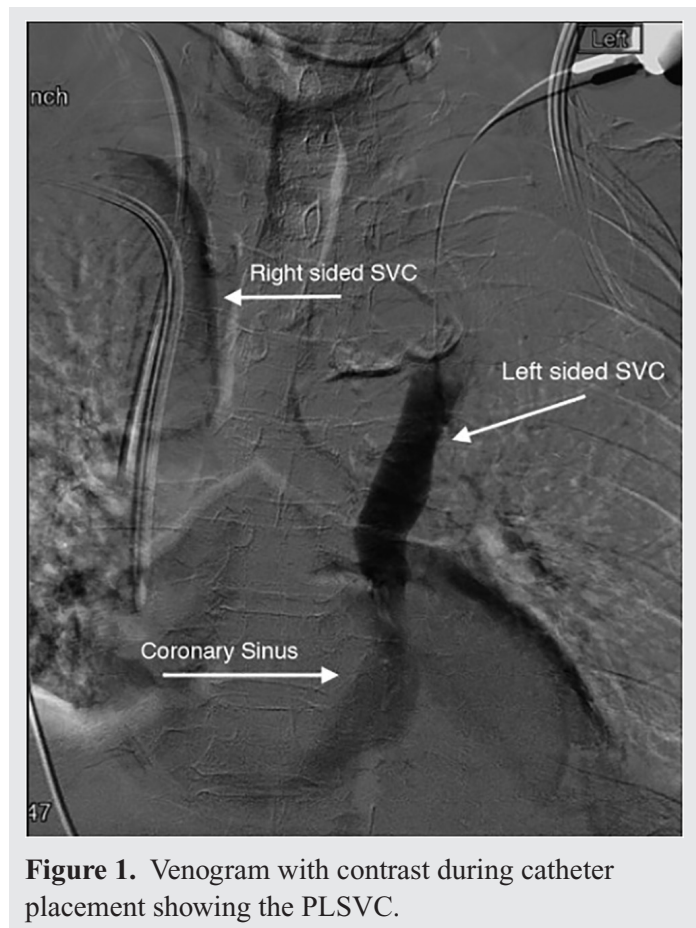
**Corresponding author:** Christina Zhu  
**Contact Information:** Christina.Zhu@ttuhsc.edu  
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and descending colon thickening. Findings on emergent exploratory laparotomy were consistent with generalized peritonitis secondary to descending colon perforation. He underwent infected PD catheter removal, left hemicolectomy with end colostomy, and splenectomy due to a capsular tear secondary to adhesions in the left upper quadrant. Cultures were taken from the PD catheter and confirmed infection. Pathology reported focal perforation of the colon and perisplenitis. Immediately post-operation, a temporary hemodialysis catheter was placed in the right internal jugular vein. Physical therapy was started when the patient finished a course of IV antibiotics. Seventeen days after initial presentation, during preparation for potential discharge, it was discovered that his temporary right internal jugular hemodialysis catheter had ceased to function.

A new tunneled hemodialysis catheter was then placed in the left internal jugular vein (IJV) using ultrasound guidance and fluoroscopy by the interventional radiologist in the radiology suite. Catheters in the right IJV and subclavian were already present. During placement, it was noted that the wire failed to traverse the midline. A venogram with contrast showed a duplicate SVC draining into the coronary sinus (Figures 1 and 2). Fluoroscopy verified the presence of the hemodialysis catheter within the proximal left SVC. Approximately one hour following catheter placement, the patient went into atrial fibrillation with rapid ventricular rate, which progressed to pulseless electrical activity without return of spontaneous circulation. A code blue was called, but the patient died.

## DISCUSSION

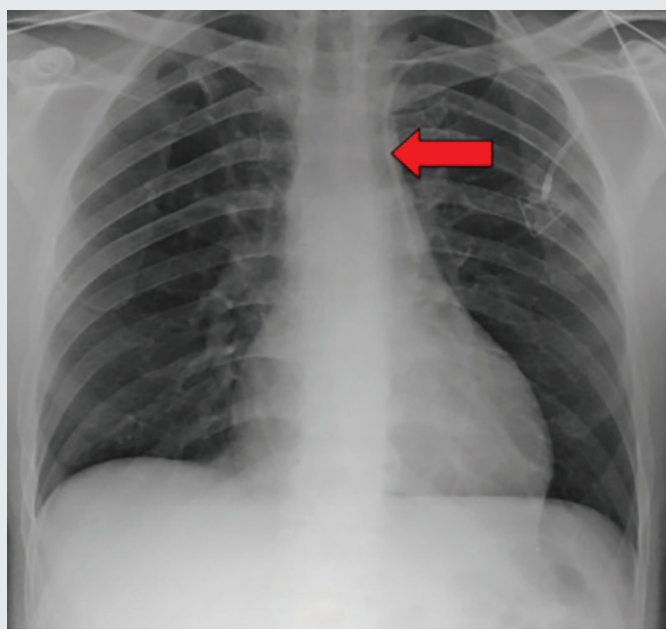
A duplicate SVC usually presents as a PLSVC and is rarely symptomatic in patients without congenital heart disease. Its discovery is typically incidental and commonly occurs following catheter placement with subsequent imaging showing an abnormal pathway for the catheter.<sup>2</sup> Surgery and cardiac imaging are also common procedures that lead to the discovery of this anomaly. In normal embryological development, about four weeks into gestation, the right and left precardinal veins drain the upper portion of the embryo before draining into their respective common cardinal veins and entering the fetal heart.<sup>3</sup> The right SVC forms from



**Figure 1.** Venogram with contrast during catheter placement showing the PLSVC.

the junction of the right precardinal and right common cardinal vein. After this occurs, an oblique anastomosis develops, forming a connection between the left precardinal vein and the right SVC. This anastomosis will then become the left brachiocephalic vein. In normal development, the left precardinal and common cardinal veins undergo atrophy leaving only the right precardinal and common cardinal veins which become the SVC. In rare cases, a PLSVC develops from lack of obliteration of the left common cardinal vein and a portion of the left precardinal vein.<sup>1-4</sup>

A persistent left SVC typically is asymptomatic, since approximately 90% of these anomalies drain to the coronary sinus, which feeds into the right atrium of the heart.<sup>6</sup> During central line, catheter, or pacemaker insertion with a PLSVC, the coronary sinus can become irritated which could lead to complications, including arrhythmias, hypotension, and myocardial ischemia.<sup>4</sup> The other 8–10% of cases involve the PLSVC draining



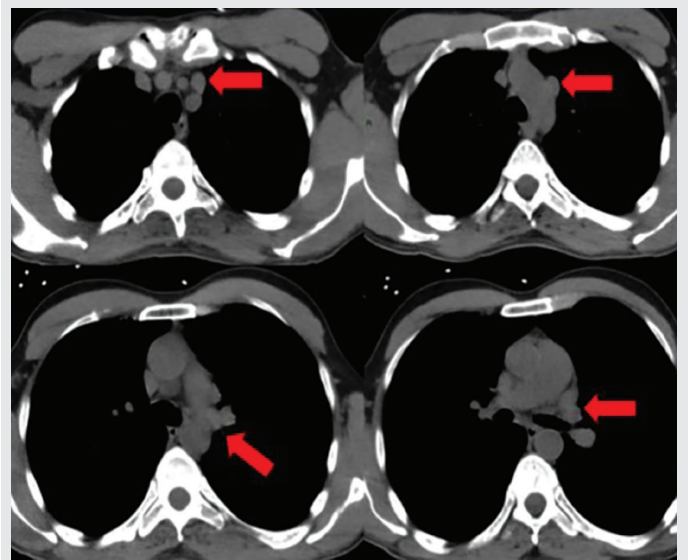
**Figure 2.** Chest x-ray of catheter placement with the wire not traversing the midline.

into the left atrium, which requires additional caution. In this variant, a right-to-left shunt is formed. This allows right sided emboli to be transferred into systemic circulation which can have dire consequences, such as ischemia or stroke and even death.<sup>1,2,4,5</sup>

It is important for physicians to be aware of these congenital cardiac abnormalities and to be able to identify them prior to surgery and catheter placement. Identification of a dilated coronary sinus on imaging should raise suspicion for PLSVC and confirmatory tests should be performed.<sup>7,8</sup> Confirmatory diagnostic studies include chest radiography with contrast, transthoracic and transesophageal echocardiography, venography, magnetic resonance imaging, and computed tomography scans (Figure 3).<sup>9</sup> Recognition of congenital cardiac abnormalities, such as PLSVC, should prompt catheter or central line removal and identification of an alternative access site to avoid complications.

## CONCLUSION

A duplicate SVC with PLSVC is a rare cardiac anomaly often discovered incidentally. Identification



**Figure 3.** Diagnosis of PLSVC by tracking the vessel from the left internal jugular vein and subclavian veins to coronary sinus.

and recognition of cardiac abnormalities are important prior to central venous line, catheter, and pacemaker placement. If PLSVC is unnoticed, coronary sinus irritation, atrial fibrillation, and technical difficulties during catheter and central line placement can develop.<sup>2,3</sup> Subsequent removal and reassessment of an alternate access point should be pursued to avoid these serious and possibly lethal outcomes.

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**From:** School of Medicine (CZ), Graduate School of Biomedical Sciences (FZ), Department of Surgery (LW, APS), Texas Tech University Health Sciences Center, Lubbock, Texas

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