Incidental finding of a left persistent subclavian vein during a routine insertion of an indwelling venous catheter

R Arrangoiz, T Niglaizzo, B Mosher, R Kareti

Citation

Abstract
The embryological development of the systemic venous system is a complex process during which developmental abnormalities may occur. One variation is the persistence of a left superior vena cava (LSVC). When present, it usually drains into the right atrium via the coronary sinus. In 8% of the cases, however, the anomalous vein directly drains into the left atrium. We describe a patient with a history of polycythemia vera who received multiple infusions of blood products each month and who required the placement of an indwelling venous catheter. A persistent left superior vena cava draining into the left atrium was diagnosed during the insertion of the catheter.

CASE REPORT
A 51-year-old woman with a history of polycythemia vera and poor intravenous access was referred by her primary care physician for the insertion of an indwelling venous catheter. Her past medical history was significant for asthma, GERD, Barrett’s esophagus, esophageal cancer and bipolar disorder. Her past surgical history included esophageal resection for esophageal cancer with postoperative radiation therapy, right-sided nephrectomy, as well as hysterectomy and bilateral salpingoophorectomy, left wrist fracture with hardware insertion and removal and uvalectomy. Placement of the indwelling catheter was initiated uneventfully by cannulation of the left subclavian vein using standard anatomic landmarks.

Initial venipuncture returned dark, non-pulsating blood. The guidewire threaded easily but appeared to go not into the superior vena cava but into the left side of the heart (Figure 1). Blood drawn from the venipuncture site in the subclavian vein had oxygen saturation and partial pressure of oxygen compatible with systemic venous blood.

DISCUSSION
Persistent superior vena cava is a rare disorder that was first described in the 18th century (1). This congenital malformation may occur without any other associated anomalies or may be associated with total anomalous pulmonary venous return, pulmonary atresia, tricuspid atresia, hypoplastic left ventricle and endocardial cushion defects, and tetralogy of Fallot (2).

Figure 1
Figure 1: Chest X-ray following the insertion of a left subclavian central line
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A persistent left superior vena cava is the most common form of anomalous venous drainage involving the superior vena cava (Figure 2) and represents persistence of the left horn of the embryonic sinus venosus, which normally involutes during normal development to become the coronary sinus. This is an uncommon disorder with an incidence of 0.3% to 0.5% in the general population and 1.5 to 11% in patients with congenital heart disease.

**Figure 2**
Figure 2: Anomalous venous drainage involving the superior vena cava

In approximately 90% of cases a persistent left superior vena cava enters the right atrium through the orifice of an enlarged coronary sinus, and this lesion is considered to be an anomaly of the coronary sinus. The persistent LSVC characteristically reaches the heart in the angle between the left atrial appendage and the left pulmonary veins. The left superior vena cava then travels down the back of the left atrium to enter the left atrioventricular groove and channel draining blood from the head and both arms. In the normal heart, this is the site of the oblique ligament and vein of Marshall. Blood returning from the left upper extremity and head through the left subclavian and left internal jugular veins drains into the persistent left superior vena cava. From there, it drains into the coronary sinus and thence into the right atrium. The coronary sinus dilates secondary to this large volume of blood. The left innominate vein is either absent or small.

A persistent left superior vena cava is usually of no clinical significance since the systemic venous blood continues to return to the right atrium but in rare occasions may produce cardiac dysrhythmias due to stretching of the atrioventricular node and bundle of His, it may also cause left ventricular inflow obstruction because of partial occlusion of the mitral valve. The dilated coronary sinus can protrude into the left atrium and can be mistaken for a left atrial mass.

The persistent left superior vena cava has important clinical implications in certain situations. It may complicate placement of cardiac catheters or pacemaker leads. Awareness of this anomaly may reduce confusion about the position of a catheter that may appear as to be inserted in an abnormal location.

<table>
<thead>
<tr>
<th>Table 1</th>
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<td>Classification of Persistent Left Superior Vena Cava</td>
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<tr>
<td>A. 90% of the cases:</td>
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<tr>
<td>1. Persistent left superior vena cava connecting to the right atrium via coronary sinus.</td>
</tr>
<tr>
<td>B. 10% of the cases:</td>
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<tr>
<td>1. Persistent superior vena cava that connects to left atrium:</td>
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<tr>
<td>i. Often associated with:</td>
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<td>1. Atrial septal defect.</td>
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<td>2. Heterotaxy syndromes.</td>
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There are two types of persistent left superior vena cava (Table 1).
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In 10% of the cases a persistent LSVC drains into the roof of the left atrium between the left atrial appendage and pulmonary veins rather than to the coronary sinus (Figure 3). This congenital malformation is called complete unroofing of the coronary sinus. The orifice of the coronary sinus then persists as an interatrial communication.

Figure 4
Figure 3: Persistent LSVC drains into the roof of the left atrium

CONCLUSION
Persistent superior vena cava is a rare disorder with an incidence of 0.3% to 0.5% in the general population that should always be part of the differential diagnosis when a surgeon is presented a difficult central line insertion where the guidewire is going to the left side of the heart.

References
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