

ECHO ROUNDS Section Editor: Edmund Kenneth Kerut, M.D

Scimitar Vein with Anomalous Pulmonary Venous Return in an Adult

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An asymptomatic 40-year-old female was referred for preoperative clearance for gynecological surgery. The patient's routine preoperative electrocardiogram (EKG) (Fig. 1) and chest x-ray (Fig. 2) were abnormal and for this reason the patient was referred for preoperative cardiac evaluation. Physical examination was significant for a prominent pulmonic component (P_2) of the second heart sound (S_2), with wide splitting but physiological respiratory variation. A grade 2 systolic murmur was heard along the left sternal border.

History, reported from another institution, included an evaluation for a suspected atrial septal defect (ASD) based on an abnormal EKG and finding of an enlarged right heart by echocardiography. At that time, saline contrast studies via the right antecubital vein, with normal respirations and following release of the Valsalva maneuver, failed to demonstrate any evidence of a right to left shunt. Calculation of a pulmonic-to-systemic flow ratio (Q_p/Q_s) was not performed.

Based on the above, an echocardiogram was performed. The right heart was somewhat enlarged suggesting volume overload. Pulmonary artery pressures were normal (pulmonary artery systolic pressure <30 mmHg) as calculated by Doppler of the tricuspid regurgitant jet, and Q_p/Q_s calculated as 1.4:1. Subcostal imaging demonstrated drainage of an anomalous vessel (scimitar vein) into the inferior vena cava (IVC), near the IVC-RA junction and just above the diaphragm (Fig. 3). Computed tomography (CT) also demonstrated drainage of the scimitar vein near the IVC-RA junction and also a single small right pulmonary vein was noted to drain into the

left atrium. The left pulmonary veins appeared normal (Fig. 4).

Scimitar syndrome is a form of partial anomalous pulmonary venous connection (PAPVC), defined as a partial or total anomalous pulmonary venous return of the right lung to the IVC (just below or above the right hemidiaphragm) or less commonly the azygous system. The finding of a PAPVC is found in up to 0.7% of adult autopsies, with the Scimitar syndrome comprising up to 5% of these. Hypoplasia of the right lung with a dextroposition of the heart is often noted.^{1,2} Malformation of the right pulmonary artery, bronchial vessels, and an anomalous arterial blood supply to the right lung from the thoracic or abdominal aorta may be found.^{3–5} Up to 25% of patients with scimitar syndrome have associated defects, the most common being a secundum type ASD.^{6,7} Of patients found to have a secundum type ASD, about 10 percent will have a PAPVC, but not necessarily the scimitar syndrome.⁸

The infantile form may present as early as immediately after birth, having respiratory distress or other signs of heart failure, with right lung haziness on chest x-ray. Pulmonary hypertension with an infantile presentation is associated with a poor outcome. Other associated cardiovascular abnormalities are uniformly found in this form.⁹

The adult form is often asymptomatic, with diagnosis after the first year of age. Most lead a normal life without surgical correction. If, however, the Q_p/Q_s is more than 1.5–2:1, patients may develop fatigue, exertional dyspnea, recurrent chest infection, and pulmonary hypertension.^{7,10}

Use of the term "scimitar" was first coined by Halasz et al.¹¹ describing the appearance of the anomalous right pulmonary vein by chest x-ray. The phrase "scimitar syndrome" was first used by Neill et al.¹² The scimitar syndrome is a constellation of findings which includes the scimitar vein

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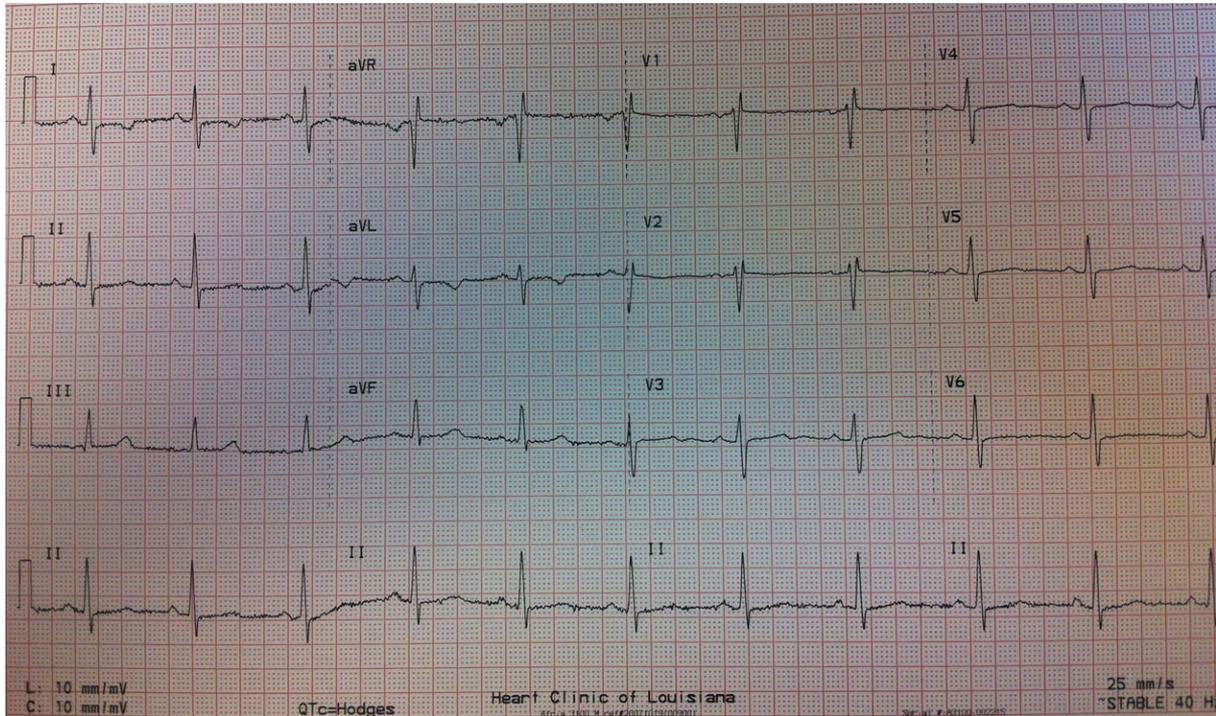


Figure 1. EKG includes evidence of right heart volume overload with a right-axis deviation and an rsr' pattern in V₁ and V₂. Mild right atrial enlargement is also suggested. These EKG findings may be found with the relatively common secundum ASD.

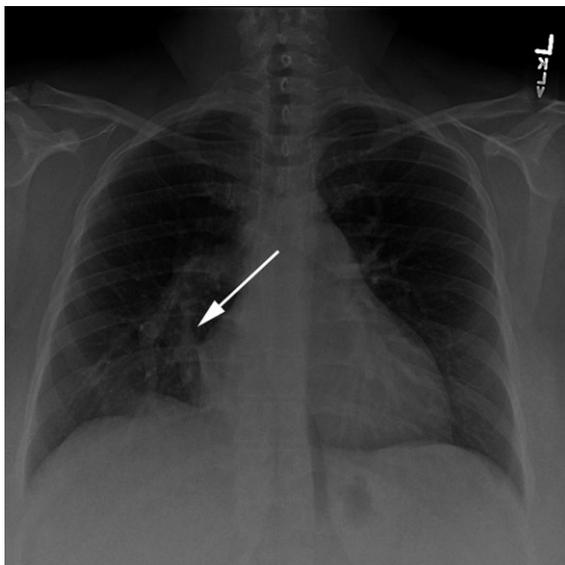


Figure 2. PA chest x-ray demonstrates a prominent size cardiac silhouette with a typical scimitar vein noted within the right lung field (arrow).

draining into the IVC, partial arterial blood supply, and hypoplasia of the affected lung with bronchial abnormalities and abnormal lung lobation.¹³ The term scimitar stems from the chest x-ray appearance of the anomalous right pulmo-

nary vein akin to a Turkish/Persian curved scimitar sword (Fig. 5). Clinically, this patient only had evidence of PAPVC with a scimitar vein.

The “scimitar sign” is seen with anomalous pulmonary vein drainage into the IVC/RA location, either just below or above the right hemidiaphragm. This chest x-ray sign has been thought to be pathognomonic for the “scimitar syndrome,” but also may be seen with a single large right pulmonary vein (ASPV) having an anomalous course, but with drainage normally into the left atrium (LA).^{14,15}

In addition to the appearance of the scimitar vein, chest x-ray findings may include hypoplasia of the right lung with a rightward mediastinal shift and an indistinct right heart border.¹⁶ CT will delineate the scimitar vein and importantly its insertion site into the IVC, RA, or the LA (ASPV).¹⁷

In summary, the scimitar syndrome describes a form of PAPVC in which a large scimitar vein drains into the IVC/RA location. In the adult form, physical examination, EKG, and echocardiography findings are suggestive of a secundum type ASD. A secundum type ASD is the most common congenital defect associated with the scimitar syndrome.

Chest x-ray will demonstrate the scimitar vein, but it should be noted is not pathognomonic of the scimitar syndrome, as ASPV may have a simi-

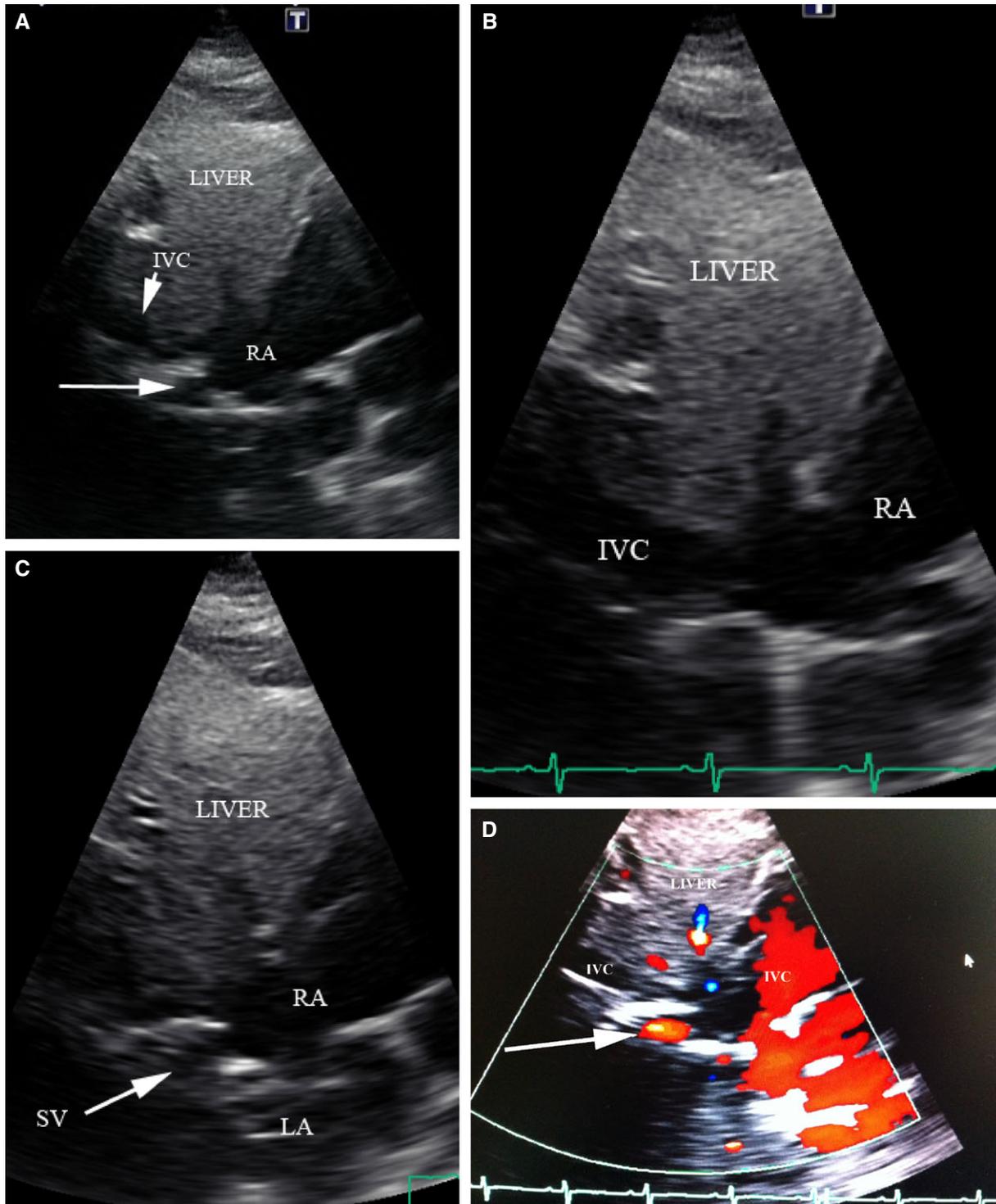


Figure 3. Subcostal imaging demonstrates the SV draining into the IVC just above the diaphragm. **A.** Visualization of the IVC, SV (horizontal arrow) and IVC-RA junction. **B.** Best visualization of the IVC, but loss of the SV. **C.** While imaging the IVC in its long axis, movement of the transducer slightly clockwise and toward the right shoulder, allowed the SV to be visualized. **D.** Color Doppler demonstrates laminar flow within the SV (red jet).

lar chest x-ray appearance. Echocardiography readily identifies drainage of the scimitar vein into the IVC/RA, and contrast enhanced CT or

magnetic resonance imaging (MRI) will identify the course and insertion site of the anomalous pulmonary vein.

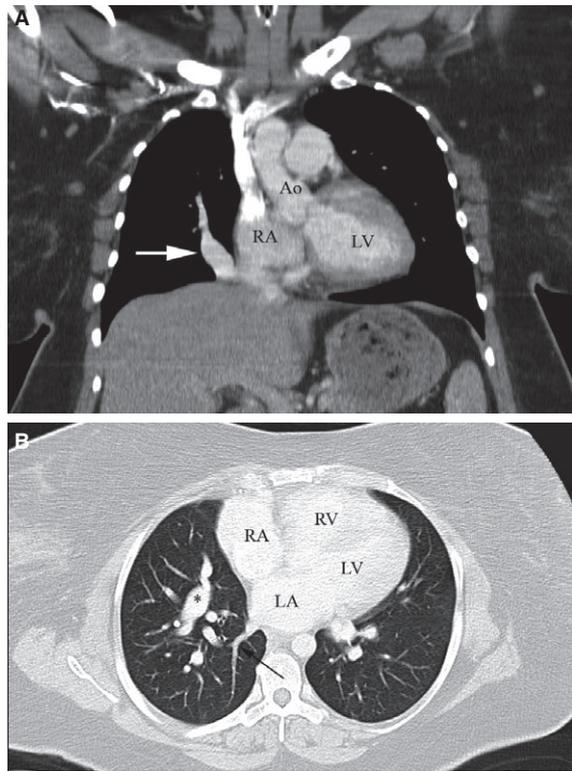


Figure 4. Contrast enhanced CT. **A.** Coronal view demonstrates scimitar vein (arrow) anomalous drainage into the RA/IVC junction. **B.** Axial view (lung window) demonstrates a single right pulmonary vein entering normally (arrow) into the left atrium (LA). The scimitar vein (*) is also visualized. Ao = ascending aorta, LA = left atrium, LV = left ventricle, RA = right atrium, RV = right ventricle.



Figure 5. Curved Turkish/Persian scimitar sword.

Summarizing points and observations include:

- 1 When an ASD is suspected by physical exam, EKG and/or echocardiography, but none found, consider PAPVC.
- 2 Calculation of Qp/Qs despite not finding an ASD may be a clue to existence of PAPVC.
- 3 Ten percent of secundum ASDs have an associated PAPVC.
- 4 The most common associated defect with scimitar syndrome is a secundum type ASD.
- 5 If a PAPVC is suspected, a chest x-ray may reveal a scimitar vein.
- 6 A SV noted by chest x-ray is not diagnostic of scimitar syndrome, as ASPV will also have this finding.
- 7 Drainage of a SV into the IVC may be above or below the diaphragm.
- 8 The SV may not be readily found, unless looking for it. In this case it was noted that while imaging the IVC in its long axis, movement of the transducer slightly clockwise and toward the right shoulder, allowed the SV to be visualized.

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