A 56-year-old woman who underwent secundum atrial septal defect closure at the age of 25 years was referred to the Mayo Clinic for exertional dyspnea and symptomatic atrial fibrillation. On examination, she was acyanotic, with an irregularly irregular heart rate of 86 beats per minute and a blood pressure of 140/80 mm Hg. She had a fixed S₂ split with a normal apical impulse.

During the course of her work-up, transthoracic echocardiography demonstrated anomalous pulmonary venous return of the right upper and lower pulmonary veins to the inferior vena cava just above the diaphragm (Scimitar syndrome). The right-sided chambers of the heart and pulmonary arteries were dilated because of left-to-right shunting. The left and right middle pulmonary veins entered the left atrium normally. The calculated pulmonary-to-systolic flow ratio was 2.5:1 (Figures 1 through 3 and Movies I through IV). Contrast-enhanced computed tomography scan and cardiac catheterization to delineate the anatomy of the vascular and bronchial anatomy also confirmed our findings and revealed a pulmonary arterial pressure of 46/31 mm Hg and pulmonary vascular resistance of 3.26 Woods unit \( \cdot \) m² (Figure 4 and Movies V through VII). The patient was referred for surgical correction.

When anomalous pulmonary venous drainage is suspected because of the characteristic radiographic curvilinear appearance of vein shaped like a “scimitar” or Turkish sword (scimitar sign) or increased caval flow into the right atrium, the diagnosis can be made easily by transthoracic echocardiography delineating the abnormal anatomy with the use of the “crab view” coupled with subcostal imaging to demonstrate anomalous pulmonary venous flow into the inferior vena cava below or just above the diaphragm.

Acknowledgments
The authors thank Mark Zangs for his expert technical assistance.

Disclosures
None.
Figure 1. Subcostal view with 2-dimensional and color Doppler showing the scimitar vein (SV) draining into the dilated inferior vena cava (IVC) just above the diaphragm. LA indicates left atrium; RA, right atrium.

Figure 2. Pulse wave Doppler image of the anomalous vein showing the typical monophasic flow pattern of the Scimitar vein into the inferior vena and extending throughout the cardiac cycle with an abnormally high peak velocity.
Figure 4. A, Anterior view; B, posterior view. Three-dimensional computed tomography reconstructed images showing a large scimitar vein that drains most of the right lung into the inferior vena cava just above the diaphragm. IVC indicates inferior vena cava; LA, left atrium; LLPV, left lower pulmonary vein; LUPV, left upper pulmonary vein; RMPV, right middle pulmonary vein; and Scim.V, scimitar vein.

Figure 3. “Crab view” with 2-dimensional and color Doppler (obtained with the transducer in the suprasternal notch position) demonstrating the absence of the right upper and right lower pulmonary veins. The right middle (RMPV), left upper (LUPV), and left lower (LLPV) pulmonary veins drain normally into the left atrium (LA). Note the dilated right pulmonary artery (RPA) caused by left-to-right shunting. Ao indicates aorta.
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Circulation. 2006;114:e373-e375
doi: 10.1161/CIRCULATIONAHA.105.603670

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