

Noncardiac cause of atypical chest pain: a 40-year-old female presented to the emergency department with atypical chest pain. The cardiac CT (A: axial image; B: sagittal image) showed absence of coronary stenosis and plaque but revealed a very large hiatal hernia (*), likely the cause of her symptom. RA: right atrium; LA: left atrium; LV: left ventricle; RV: right ventricle
Image courtesy of Su Min Chang, M.D.

Imaging Vignette: Scimitar Syndrome

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A 51-year-old female with a past medical history of diabetes mellitus, hypertension, and pulmonary hypertension was referred for cardiac magnetic resonance (CMR) evaluation of possible left-to-right shunt due to a suspected increase in oxygen saturation in her right atrium during right heart catheterization. CMR demonstrated partial anomalous pulmonary venous connections (PAPVC) with both right superior and inferior pulmonary veins draining into the inferior vena cava, consistent with scimitar syndrome (Figure 1). There was normal left-sided pulmonary venous return, and no atrial septal defect was detected. Phase contrast CMR demonstrated left-to-right shunt of 6.0 L/min with a Qp:Qs of 1.9:1.0.

Scimitar syndrome is a rare congenital anomaly (3–5% of all PAPVC, incidence of which is 0.4–0.7% of adults at autopsy) characterized by anomalous venous connections from the right lung into the inferior vena cava. The symptoms in the adult form are often mild initially, but chronic right-sided overload and pulmonary hypertension are late complications. The term “scimitar syndrome” derives from the shadow created by the anomalous vein on the chest radiograph that resembles a scimitar — a curved Turkish sword. Gadolinium-enhanced MR angiography is a non-invasive, radiation-free technique that exploits its high spatial resolution to provide three-dimensional anatomically detailed images. Phase-contrast CMR can add functional data sets to allow quantification of shunting and calculation of Qp:Qs ratio.

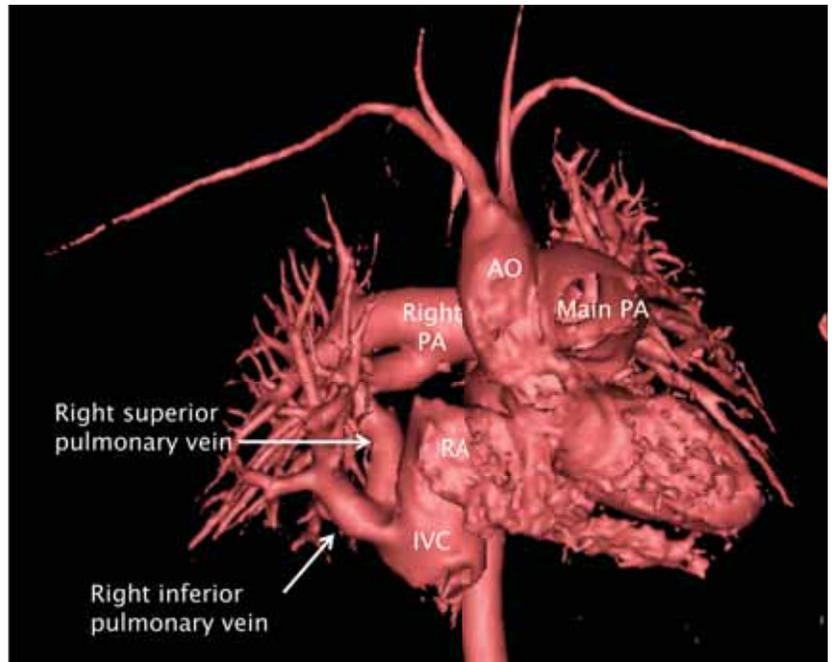


Figure 1. Anterior surface shaded display of gadolinium-enhanced chest magnetic resonance angiography; right superior and inferior pulmonary veins drain into the inferior vena cava (scimitar syndrome). AO: aorta; PA: pulmonary artery; RA: right atrium; IVC: inferior vena cava