Scimitar Syndrome
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Clinical History
A 75-year-old patient presented to MGH for non-cardiac surgery. During his pre-operative evaluation, a review of a remote chest radiograph demonstrated a slight rightward shift of the mediastinum. The patient reported no cardiac or pulmonary symptoms, and an MRI was ordered to investigate for possible congenital cardiovascular anomalies prior to surgery.

Findings
Cardiac MRI demonstrated an abnormal vessel draining the right middle lobe medial segment as well as the right lower lobe (Figure 1-4). The vessel coursed inferiorly and drained into the IVC at the level of the diaphragm, consistent with Scimitar syndrome. The left common trunk pulmonary vein and the right superior pulmonary vein drained normally into the left atrium. Mild hypoplasia of the right lung was noted.

Discussion
Partial anomalous pulmonary venous return (PAPVR) refers to a type of congenital vascular anomaly where one or more pulmonary vein drains not into the left atrium, but instead into the SVC, IVC, right atrium, coronary sinus, hepatic veins or portal veins. When the pulmonary vein drains into the IVC, it is also called “Scimitar syndrome”, named after the Turkish sword for its curvilinear opacity adjacent to the right heart border on a chest radiograph. Physiologically, Scimitar syndrome represents a left to right shunt. Most patients are either minimally symptomatic or asymptomatic, and the treatment is usually conservative.

Scimitar syndrome is also known as “hypogenetic lung syndrome”, because it is often associated with right lung hypoplasia. Imaging often reveals a mediastinal shift towards the right, as in this patient. Other documented associations include atrial septal defect, pulmonary artery hypoplasia, and sequestration.

Due to their high resolution and large field-of-view (FOV) capabilities, CT and MRI are excellent noninvasive tools that can help delineate the anatomy and course of many congenital cardiovascular anomalies. MRI has the added advantage of being free of radiation. MRI can be performed either without or with gadolinium contrast agents. In our patient, gadolinium-enhanced MRA was able to demonstrate the anomalous draining pulmonary vein with superb quality, and the patient was able to undergo surgery without any additional studies.

REFERENCES