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Scimitar Syndrome:

Multimodal Imaging Before and After Repair

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A 19-year-old woman was noted on an incidental chest radiograph to have dextroposition of the heart secondary to a hypoplastic right lung, with a linear opacity to the right of the right atrium extending below the diaphragm (Fig. 1). Although the patient was asymptomatic, a subcostal view via transthoracic echocardiography showed substantial dilation of the right atrium and right ventricle, a pulmonary artery systolic pressure of 27 mmHg, and an abnormal vessel and flow entering a dilated inferior vena cava (IVC) (Fig. 2). Subsequent cardiac catheterization and angiography of the right pulmonary artery revealed complete anomalous right pulmonary venous (PV) drainage from the right lung to the infradiaphragmatic IVC with a pulmonic-to-systemic flow ratio (Qp:Qs) of 2.11:1 (Fig. 3). Cardiac magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) were performed before and after gadofosveset-trisodium contrast administration. This nonin-

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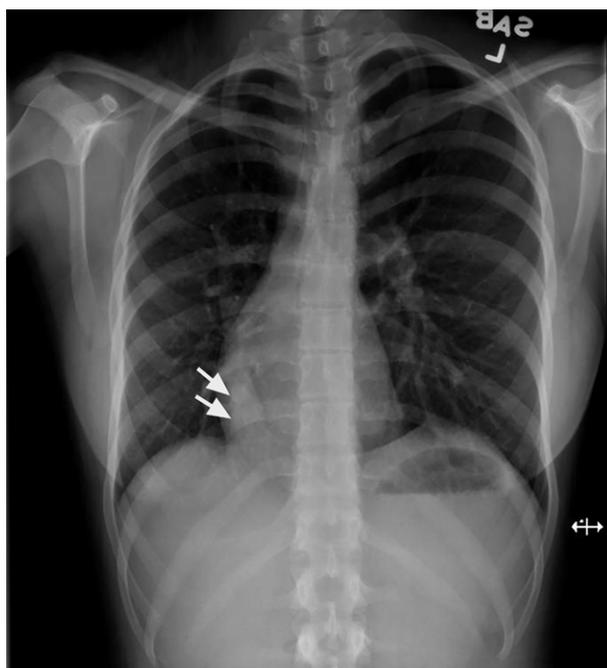


Fig. 1 Frontal chest radiograph shows a hypoplastic lung with cardiac dextroposition and a linear opacity to the right of the atrium, extending below the diaphragm (arrows).

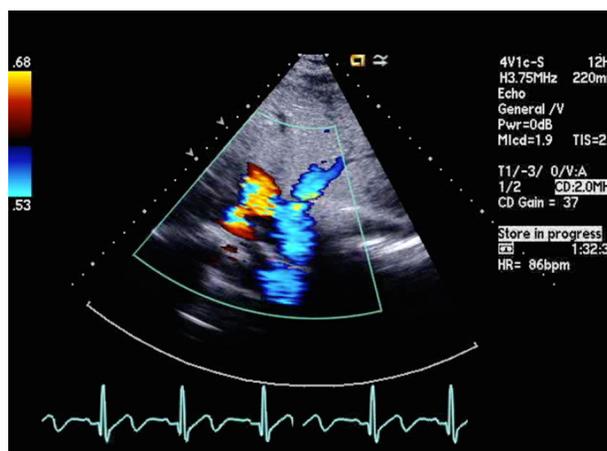


Fig. 2 Transthoracic echocardiogram in color-flow Doppler mode (subcostal view) shows Doppler inflow (red) of the anomalous right pulmonary vein as it drains into the inferior vena cava.

[Supplemental motion image is available for Figure 2.](#)

vasive study revealed anomalous drainage of the entire right lung into the infrahepatic IVC with a calculated Qp:Qs ratio of 2.1:1, confirmed the prior results, and suggested no anomalous pulmonary arterial supply, left PV drainage, horseshoe lung, or accessory diaphragm (Fig. 4). Because of substantial shunting, the scimitar right PV was repaired by means of an end-to-side anastomosis of the right PV to the left atrium adjacent to the atrial septum (Fig. 5). An outpatient postoperative MRI revealed the surgical connection, a decrease in right ventricular cavity size, and a Qp:Qs of 1:1 (Fig. 6). Postoperatively, the patient did well.



Fig. 3 Right pulmonary arteriogram (anteroposterior projection) shows anomalous pulmonary venous drainage of contrast medium into the inferior vena cava, inferior to the diaphragm (arrows).

Supplemental motion image is available for Figure 3.



Fig. 4 Cardiac magnetic resonance image with contrast medium (posterior view in 3-dimensional rendering) defines the entire right pulmonary venous drainage to the infrahepatic inferior vena cava (arrows).



Fig. 5 Postoperative frontal chest radiograph shows sternotomy wires and an unchanged heart size, with the cardiac apex lying to the right of the midline. Note the small basilar atelectasis.

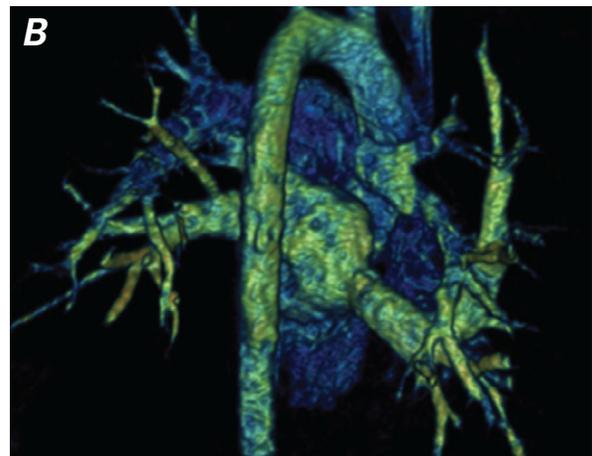


Fig. 6 Cardiac magnetic resonance angiograms with contrast medium. **A)** Anterior view shows the surgical connection between the right pulmonary vein and the left atrium. **B)** Posterior view, in 3-dimensional rendering. Note the postoperative changes associated with repair of the partial anomalous pulmonary venous return: in particular, the surgical connection of the right pulmonary vein to the left atrium.

Comment

The term “scimitar syndrome” in these patients is derived from the resemblance of the anomalous right pulmonary venous course to a curved sword of Middle Eastern origin, known as a scimitar. The tip of the “sword” is the downward, anastomotic point of the PV entry to the IVC below the diaphragm. Symptoms can arise from this abnormal anastomosis, should obstruction occur. In experienced and multidisciplinary congenital heart disease centers, the use of 3-dimensional MRI and MRA enables exquisite anatomic definition in both the pre- and postoperative conditions, distinct from that afforded by ultrasonography and conventional angiography. The calculation of shunt ratios, comparable to the calculations enabled by catheterization, allows MRI and MRA to assist clinicians in deciding between surgical and conservative management of patients who have anomalous PV drainage—perhaps sparing them the morbidity of invasive and ionizing procedures.