Familial Scimitar Syndrome
Three-Dimensional Visualization of Anomalous Pulmonary Vein in Young Sisters

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Scimitar syndrome is a rare congenital disorder characterized by an anomalous connection of the pulmonary vein with the inferior vena cava. The anomalous vein appears as a “scimitar”-like shadow on a chest x-ray. We recently encountered Scimitar syndrome in 2 sisters and demonstrated the 3D structure of the anomalous vein by computed tomography (CT).

Case 1
A 27-year-old woman was referred to our hospital for further evaluation of an abnormal shadow in the right lower lung field on chest x-ray (Figure 1). She was asymptomatic. The 3D CT clearly demonstrated an abnormal pulmonary vein that drained into the suprahepatic inferior vena cava (Figure 2). Cardiac catheterization showed a mean pulmonary artery pressure of 15 mm Hg, a pulmonary/systemic flow ratio of 1.86, and a left-to-right shunt ratio of 46%. Corrective surgery was not performed because of her lack of symptoms and relatively low shunt ratio.

Case 2
This patient was the 31-year-old sister of the patient described in case 1. She also had an abnormal shadow in the right lower lung field on chest x-ray (Figure 3). She had mild dyspnea on exertion, which began at ≈20 years of age. The 3D CT demonstrated an abnormal pulmonary vein, which drained into the suprahepatic inferior vena cava (Figure 4). On cardiac catheterization, the mean pulmonary artery pressure was 16 mm Hg, the pulmonary/systemic flow ratio was 2.05, and the left-to-right shunt ratio was 53%. The patient subsequently underwent surgery to switch the anomalous pulmonary vein to the left atrium.
Figure 3. Case 2. Chest x-ray showing an abnormal shadow in the right lower lung field (arrows).

Figure 4. Case 2. Left, 3D CT in right anterior oblique view. Arrows indicate the anomalous vein. Center, 3D CT in posteranterior view. Arrows indicate the anomalous vein. IVC indicates inferior vena cava; RV, right ventricle; RA, right atrium; LV, left ventricle; and Ao, descending aorta. Right, Late phase of pulmonary arteriogram showing the anomalous pulmonary vein (arrows).
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