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Images in Congenital Heart Disease

Incomplete scimitar syndrome

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A 17-YEAR-OLD WOMAN PRESENTED IN OUR institution for evaluation of suspected arterial hypertension. She was born in 1983 at a gestational age of 26 weeks. In childhood, she suffered from recurrent pulmonary infection which was presumed to be due to bronchopulmonary dysplasia. In the last year before presentation however, there had been no pulmonary infections.

The chest radiograph showed a normal cardiac shadow and lung fields, but there was a paracardial opacity on the right side resembling a scimitar vein (arrows in Fig. 1). Further diagnostic evaluation, including monitoring of blood pressure for 24 h, ruled out arterial hypertension. Magnetic resonance angiography was performed for assessment of the paracardial opacity. Rapid T1-weighted transversal sequences revealed an atypical vessel originating from the inferior caval vein (ICV). We then proceeded

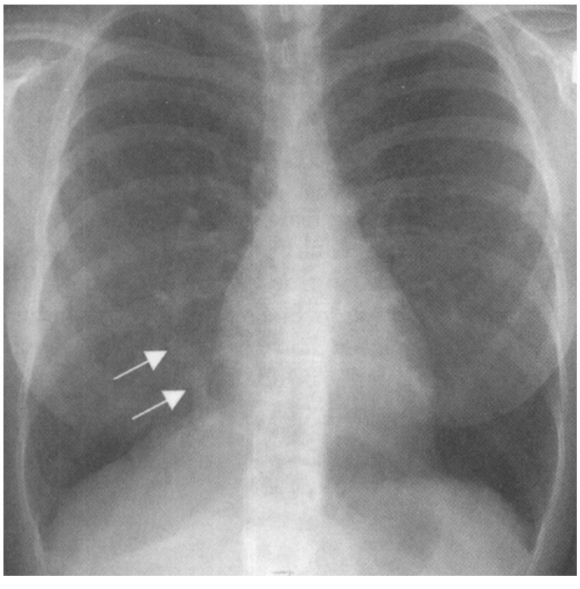

Figure 1.

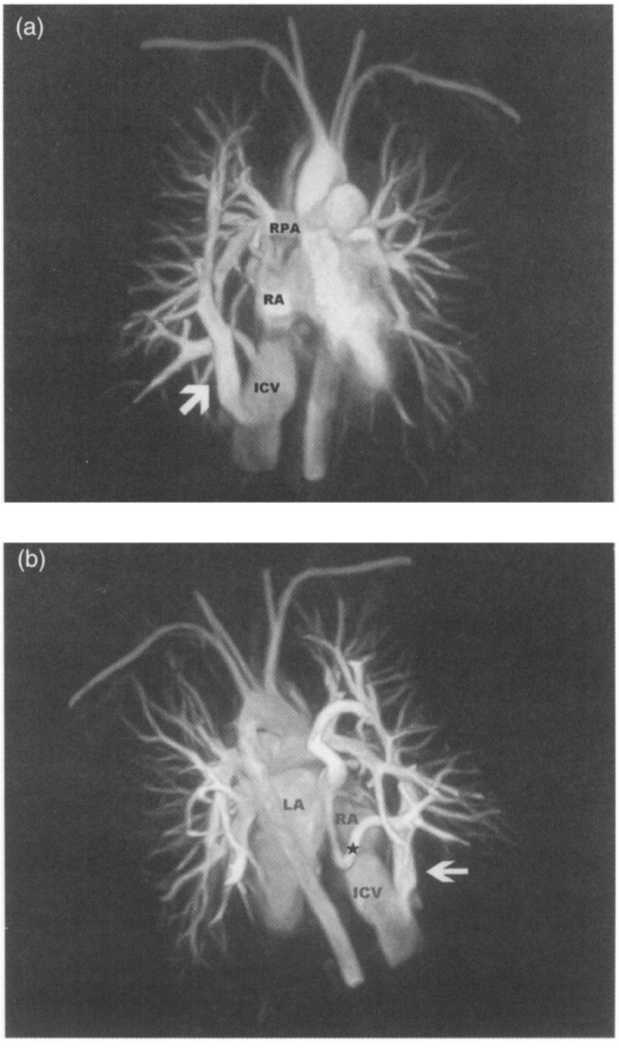

Figure 2.

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to native and contrast-enhanced sequences profiled in the coronal plane to include the lung hilums, the atriums (RA, LA) and the pulmonary and caval veins. Postprocessing was performed by shaded surface rendering. We confirmed the presence of a large vein descending from the hilum of the right lung to the inferior caval vein (arrow in Fig. 2a). The dorsal view revealed a small pulmonary vein from the lower right lobe to the left atrium (star in Fig. 2b). It also showed normal distribution of the right pulmonary artery (RPA) and excluded additional significant collateral flow of blood from the aorta to the lung.

Isolated partially anomalous pulmonary venous return to the inferior caval vein, or so-called incomplete scimitar syndrome, is a rare finding. It may be suspected on a chest radiograph when the typical scimitar sign is present. Patients may also show some unspecific pulmonary complaints, but they are usually asymptomatic. Diagnosis can be confirmed by computed tomography or magnetic resonance imaging. Cardiac catheterization can be performed to rule out any anomalous pulmonary arterial supply, and for hemodynamic assessment. Since the left-to-right shunt will not exceed 25%, no surgical intervention is justified.

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