CLINICAL IMAGES

Assessing scimitar syndrome – use of MRI and MRA

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A 36-year-old woman was assessed as a live-related kidney donor. Her chest radiograph showed unsuspected dextroposition of the heart, diminished volumes of the right middle and lower lobes, and a scimitar-shaped shadow extending from the right hilum to the region of the inferior vena cava (a scimitar is a short sword with a curved blade originally used in Eastern countries) (Fig. 1).

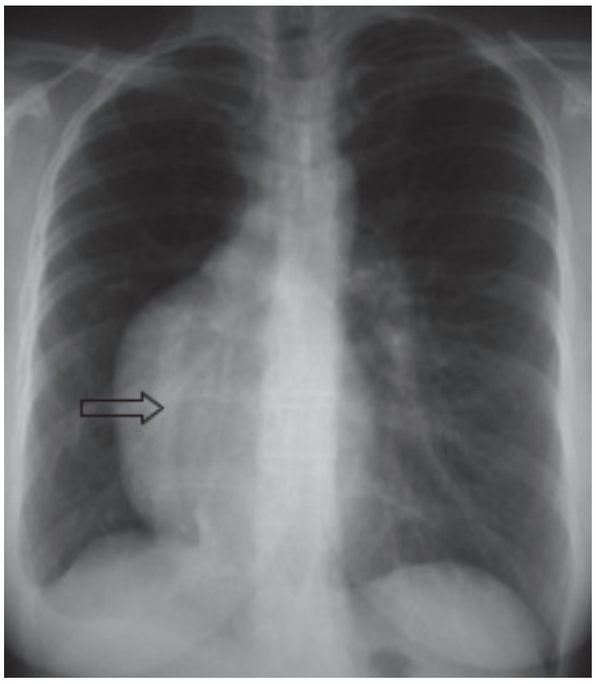

Fig. 1. Frontal chest radiograph showing dextroposition of the heart with volume loss involving the lower and middle lobes of the lung. A 'scimitar' vein (open arrow) is suspected on the right.

At the time of writing, Vijay Dahya was a radiologist in the Division of Radiology, Groote Schuur Hospital and University of Cape Town. He has an interest in cardiac magnetic resonance imaging and is a patron of the classical music and dance forms, particularly when his lovely wife, who is an accomplished classical dancer, is performing.

Bongani Mayosi, Professor and Head of the Department of Medicine, GSH and UCT, is a researcher, teacher and clinician working on the elimination of heart diseases of the poor. When not working he enjoys the company of his wife and daughters, which is about the only pastime he has.

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Non-invasive examination using computed tomography (CT) (Fig. 2), magnetic resonance imaging (MRI) (Figs 3 - 5) and magnetic resonance angiography (MRA) (Fig. 6) confirmed the diagnosis of scimitar syndrome, which is characterised by partially anomalous pulmonary venous drainage, pulmonary lobar hypogenesis and cardiac dextroposition. The associated

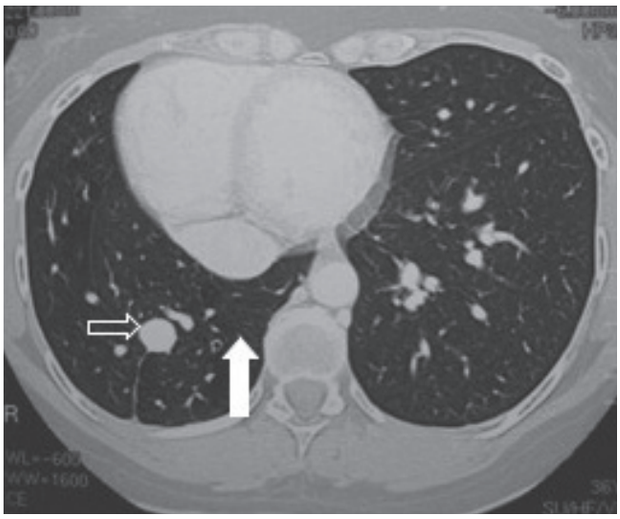

Fig. 2. Contrast-enhanced CT scan of the chest confirming the pulmonary hypogenesis involving the right middle and lower lobes (solid arrow) with cardiac dextroposition. The 'scimitar' draining vein is confirmed (open arrow).

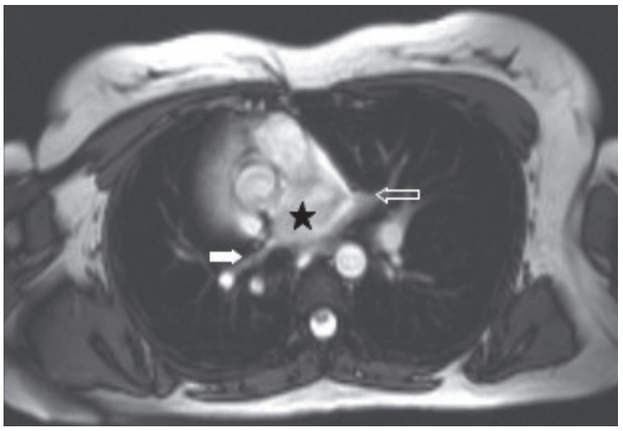

Fig. 3. Axial true-FISP (bright blood) ECG-triggered MRI showing cardiac dextroposition and normal drainage of the left (open arrow) and right (solid arrow) superior pulmonary veins into the left atrium (star) (FISP = fast imaging with steady-state precession).

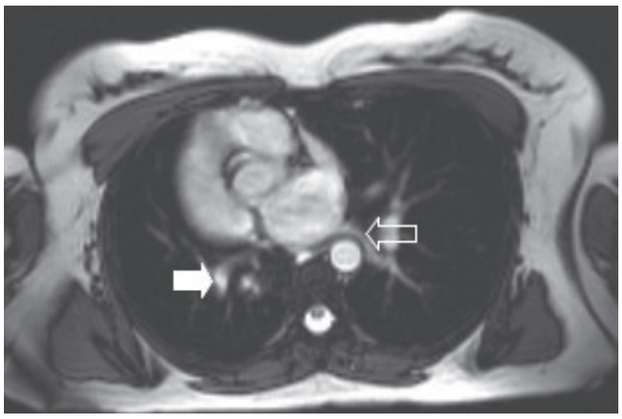

Fig. 4. Axial true-FISP (bright blood) ECG-triggered MRI showing normal drainage of the left inferior pulmonary vein (open arrow) into the left atrium. The right inferior pulmonary vein is seen draining into a 'scimitar' vein (solid arrow) representing partially anomalous pulmonary venous drainage. Cardiac dextroposition is again clearly noted.

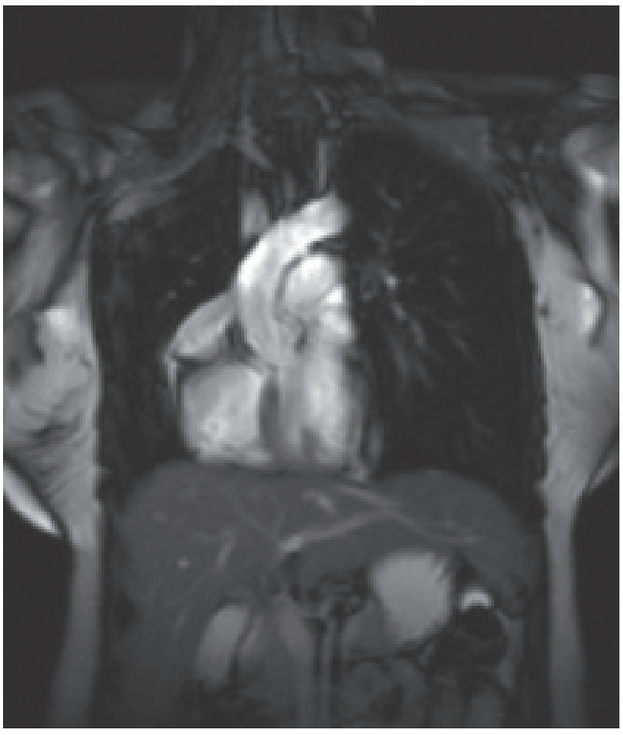

Fig. 5. Coronal true-FISP (bright blood) ECG-triggered MRI demonstrating normal cardiac anatomy besides the dextroposition.

abnormalities, which include atrial septal defect, anomalous branching of the pulmonary artery, systemic arterial supply of lung segments (pulmonary sequestration) and bony spine and/or chest wall anomalies, were not found in our patient; she proceeded with kidney donation. This case demonstrates how comprehensive, non-invasive imaging has replaced invasive techniques such as cardiac catheterisation and pulmonary angiography in the diagnosis of scimitar syndrome.

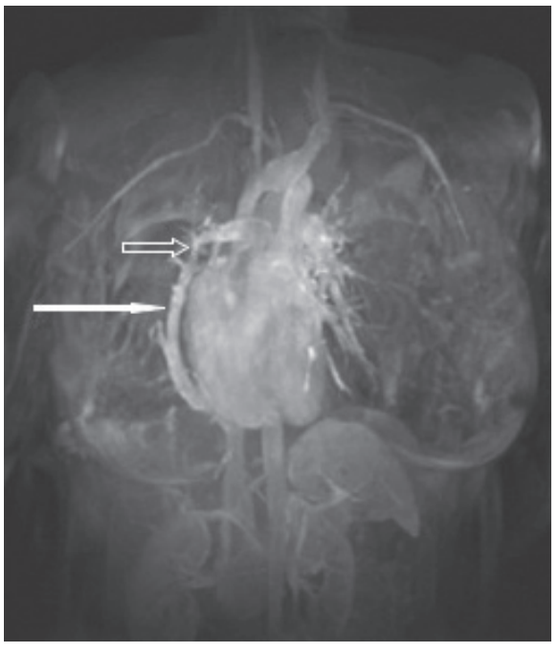

Fig. 6. Three-dimensional gadolinium-enhanced magnetic resonance angiogram (MRA) showing the 'scimitar' vein draining into the supra-hepatic inferior vena cava (solid arrow). A mild stenosis is noted in the superior portion at the point of junction between the right inferior pulmonary vein and the 'scimitar' vein (open arrow).