

# IMAGES

## in PAEDIATRIC CARDIOLOGY

**Morrison ML,<sup>1</sup> Sands AJ,<sup>1</sup> Paterson A.<sup>2</sup> Primitive hepatic venous plexus in a child with scimitar syndrome and pulmonary sequestration. *Images Paediatr Cardiol* 2013;15(3):1-5**

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### **Key Words**

Primitive hepatic venous plexus & Scimitar syndrome

### **Abstract**

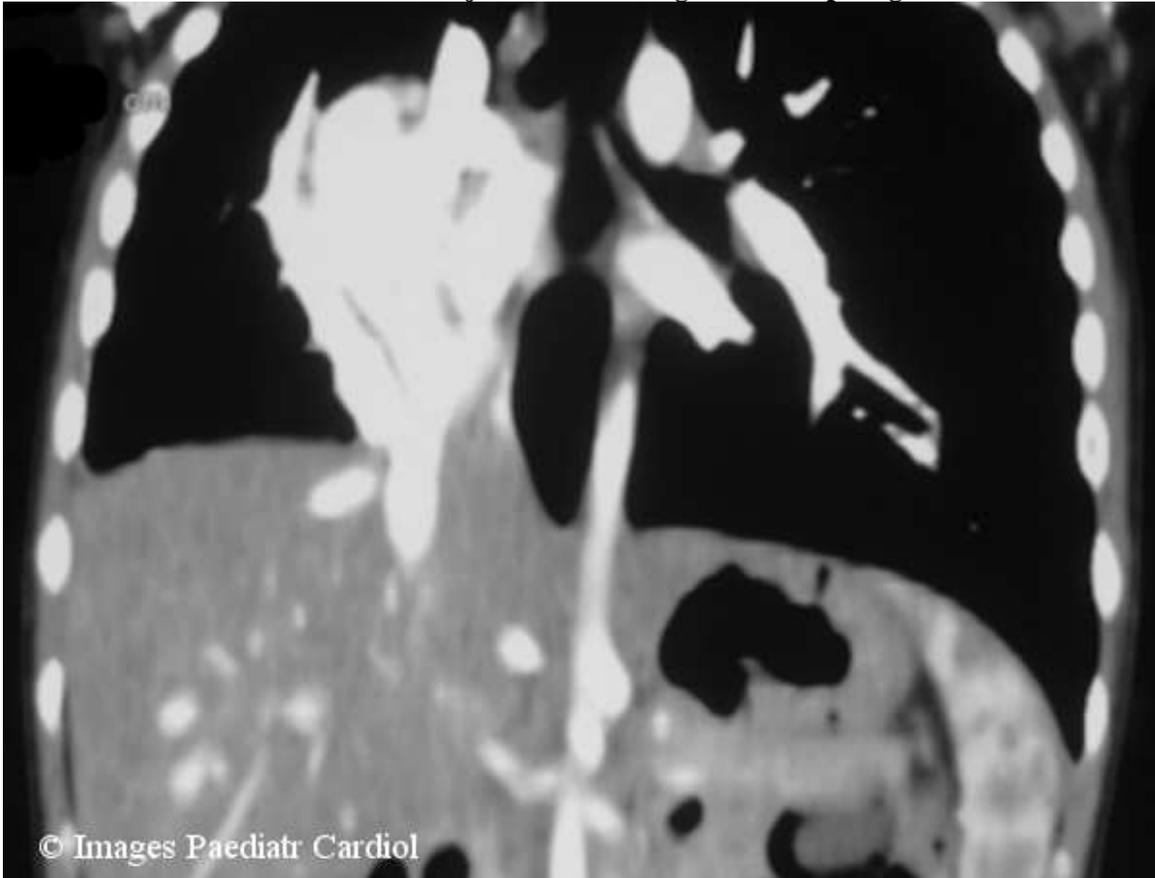
We report a case of scimitar syndrome with pulmonary sequestration, persistent primitive hepatic venous plexus and stenosis of the inferior vena cava in a child presenting with failure to thrive. Such associations are rare but may have implications when planning interventions for patients with complex congenital heart disease

### **Case Report**

A 2 month old girl was referred for assessment of respiratory distress, murmur and failure to thrive. Echocardiography confirmed dextroposition of the heart with dilated right atrium, right ventricle and pulmonary artery. There was anomalous pulmonary venous drainage with the right-sided pulmonary veins appearing to drain to coronary sinus. Bilateral superior vena cavae were noted. At cardiac surgery partial anomalous drainage of the right-sided pulmonary veins to the upper inferior vena cava was identified. The right-sided veins were successfully baffled to left atrium through the atrial septal defect.

Despite surgery she remained unwell with frequent respiratory infections and did not thrive. A CT angiogram was performed which demonstrated scimitar syndrome with an area of pulmonary sequestration in the lower lobe of the right lung (figure 1).

**Figure 1: Coronal multiplanar reconstruction demonstrating the scimitar vein draining to the inferior vena cava just below the right hemidiaphragm.**



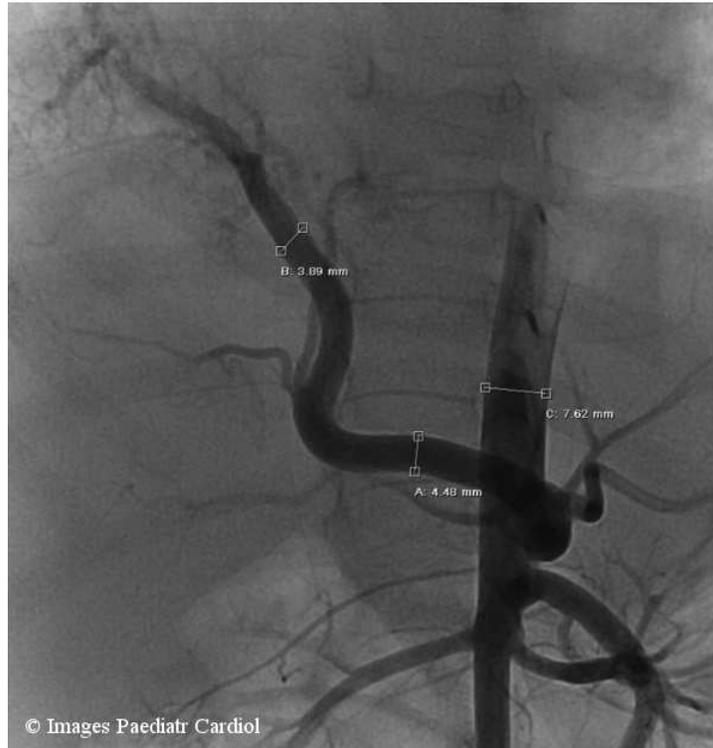
The right pulmonary artery was underdeveloped and there was a large collateral artery from the descending aorta supplying the right lower lobe. Interesting, a complex racemose network of venous channels was noted connecting the inferior vena cava to the hepatic veins and the right atrium. There was mild stenosis of the inferior vena cava (figure 2).

**Figure 2: 3D-volume rendering image from the CT study. The primitive hepatic venous plexus is seen draining to the infra-diaphragmatic inferior vena cava; at the point of entry of these veins, the inferior vena cava is stenotic (white arrow). The sequestered segment lies posteriorly, immediately adjacent to the inferior vena cava (grey accented arrow). The systemic arterial supply vessel arises from the descending aorta just above the origin of the coeliac trunk (black accented arrow).**



This appearance represents persistence of the primitive hepatic venous plexus. The large collateral artery was successfully occluded during cardiac catheterisation with 14 micro coils, which greatly improved her symptoms. The patient made a good recovery from this procedure and continues to do well at follow-up. To date she has required no further intervention for the sequestered lobe of right lung (figures 3 and 4).

**Figure 3: Angiogram demonstrating the large collateral artery arising from descending aorta near the coeliac trunk.**



**Figure 4: Selective angiogram in the collateral artery following successful occlusion with micro coils.**



## Discussion

Scimitar syndrome, a rare congenital abnormality consisting of anomalous drainage of one or all of the right-sided pulmonary veins to the inferior vena cava producing a classic scimitar-like shadow on the chest x-ray, was first described in 1960.<sup>1</sup> Scimitar syndrome has a number of associations including absence or abnormalities of the right lung, dextroposition and congenital heart defects such as atrial septal defect. It has been described in association with anomalies of the systemic venous connections including, very rarely, underdevelopment of the inferior vena cava and persistence of the primitive hepatic venous plexus as seen here.<sup>2-4</sup> Surgical correction of scimitar syndrome is considered high risk. The use of vascular plugs and coils has emerged as a successful, alternative method to occlude vascular connections.<sup>5</sup> Such large aorto-pulmonary collaterals carry high flows and often require multiple devices to achieve complete occlusion (as described here).

The primitive hepatic venous plexus may represent the embryological precursors to the hepatic veins and the hepatic segment of the inferior vena cava.<sup>6</sup> It is usually benign, although its presence can have implications for patients undergoing palliation for complex congenital heart disease who may remain desaturated post operatively.<sup>6</sup> There are no reports in the literature to date describing scimitar syndrome +/-pulmonary sequestration in conjunction with persistent hepatic venous plexus. The recognition of such associations, although rare, can prevent unnecessary investigations of the abdominal and hepatic vasculature and aid decision-making when planning surgical or other interventions for patients with complex congenital heart disease.

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