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Scimitar syndrome in infancy

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MeSH: Heart Catheterization, Scimitar Syndrome, Heart defects, congenital, Infant

Scimitar syndrome, if presenting in infancy, is associated with signs of heart failure and pulmonary hypertension.[1] The typical pathological features are sequestration of a segment of the lung, usually the right lower lobe, with arterial supply arising from the abdominal aorta, and partial anomalous pulmonary venous connection, with the sequestered segment draining to the inferior caval vein. Additional features include an atrial septal defect and hypoplasia of the right pulmonary artery and lung. Standard therapy consists of occlusion of the anomalous arterial supply to abolish the left to right shunt, and surgical repair, if required, of associated cardiac defects.[2]

We present the typical angiographic features of an infant with Scimitar syndrome. In addition to occlusion of the aortopulmonary collateral vessel, the infant underwent successful surgical closure of an atrial septal defect with rerouting of the anomalous pulmonary venous connection.

Fig. 1 Abdominal aortogram demonstrating anomalous arterial supply to the right lower lobe of the lung.

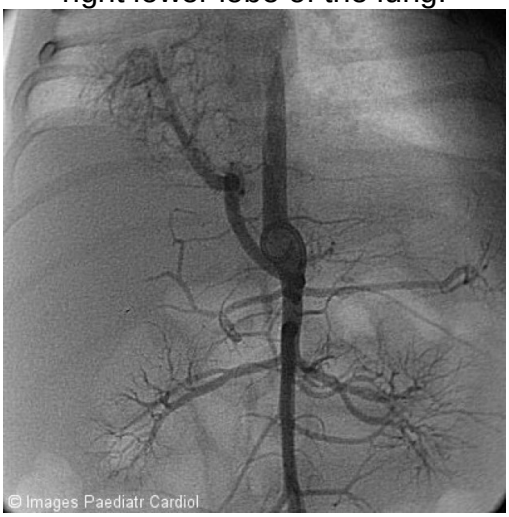


Fig. 2 Follow through demonstrating the extent of the sequestered segment.

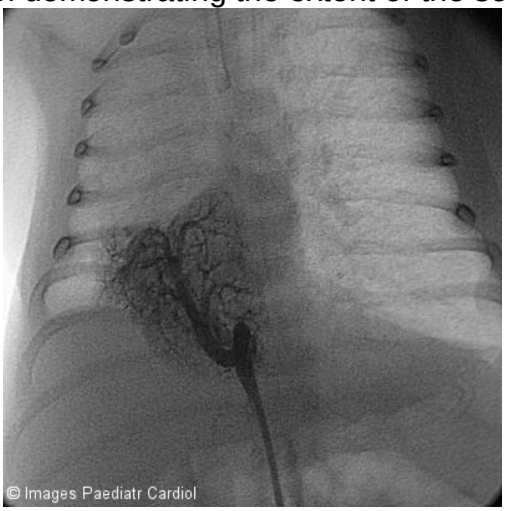


Fig. 3 Aortogram following single coil occlusion of the aortopulmonary collateral vessel.

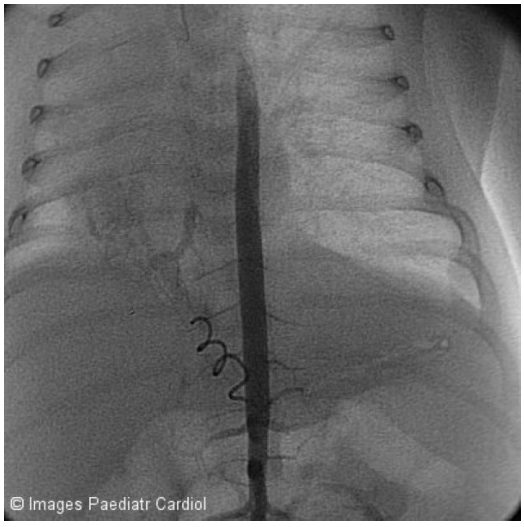


Fig. 4 Pulmonary artery angiogram demonstrating hypoplasia of the right pulmonary artery.

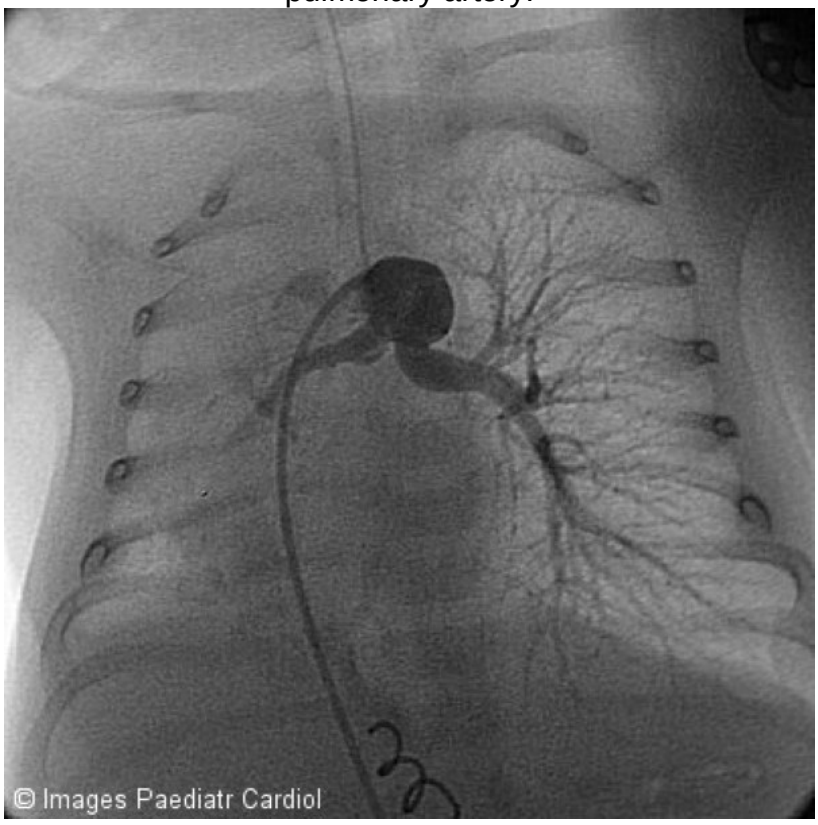


Fig. 5 Follow-through showing a small right lung supplied by the native pulmonary artery.

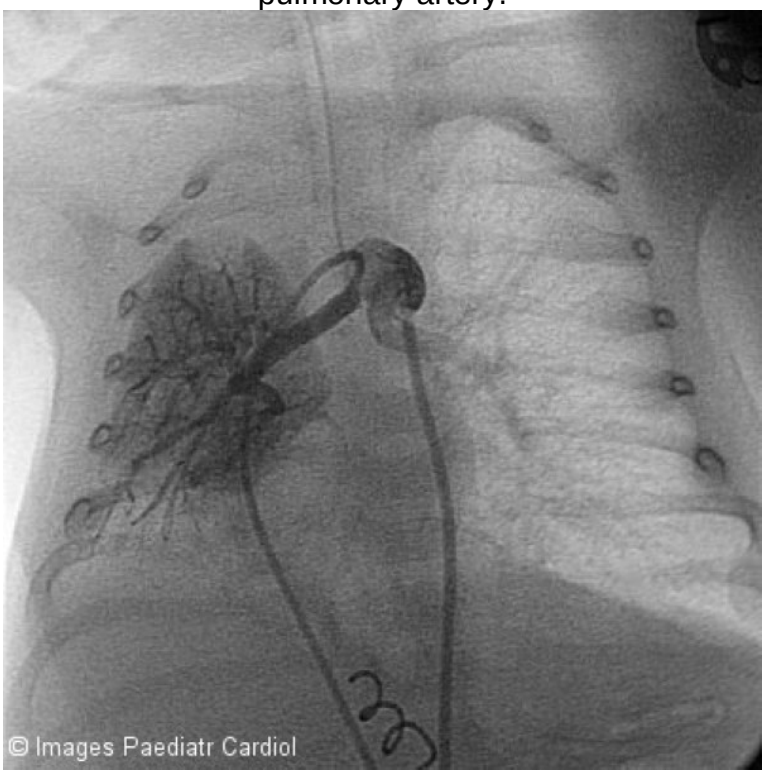
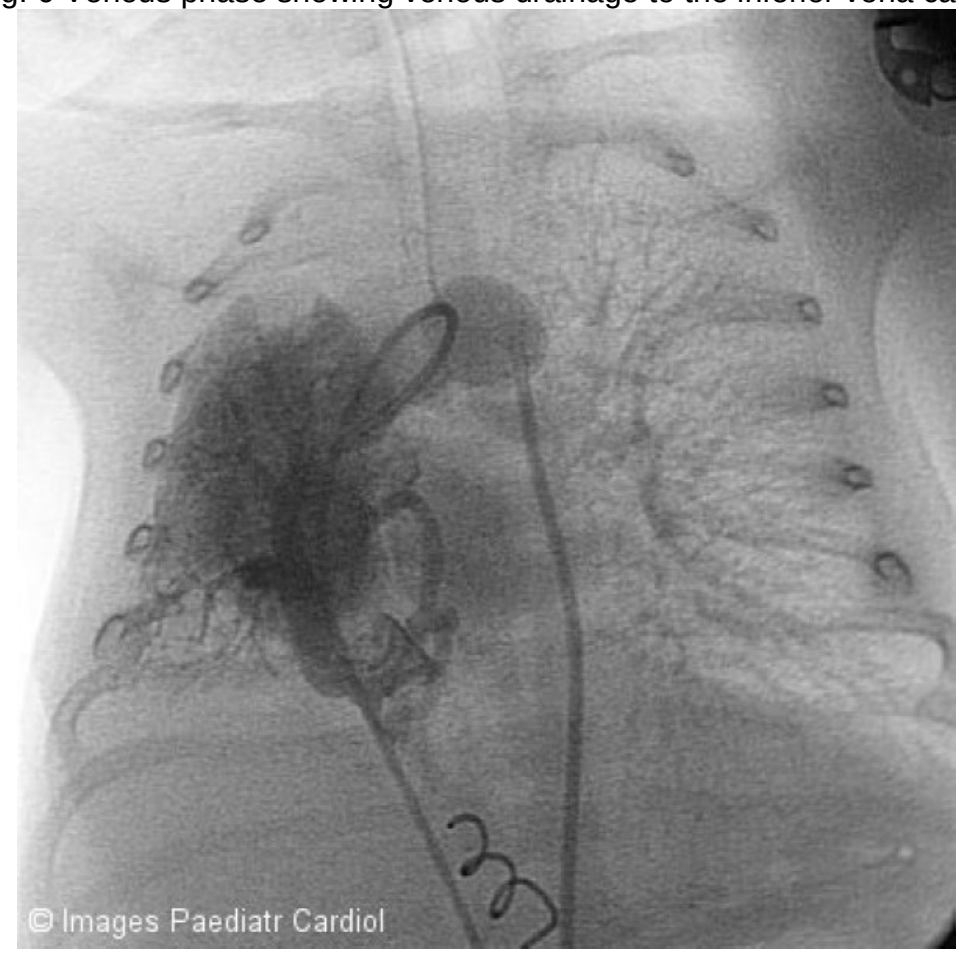


Fig. 6 Venous phase showing venous drainage to the inferior vena cava.



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