Infra-cardiac totally anomalous pulmonary venous drainage

V Dahya, *MB BCh, FCRad (SA)*
Tuft and Partners Radiology, and Department of Radiology, Groote Schuur Hospital, Cape Town

We report on a male neonate born at 37 weeks' gestation with severe respiratory distress immediately after birth. The child was cyanosed, requiring high oxygen pressures and continuous positive airway pressure. The pulmonary pattern on the earliest radiographs was difficult to definitively diagnose but, with serial radiographs, it became clear that the radiographic pattern was that of pulmonary venous congestion (pulmonary oedema). However, at no stage during serial radiographs over 8 days was the cardiothoracic ratio (CTR) greater than 60%. This suggested a likely specific diagnosis – infra-cardiac totally anomalous pulmonary venous drainage (TAPVD) with obstruction. An echocardiogram confirmed an atrial septal defect (ASD) and suggested anomalous pulmonary venous drainage, but the pulmonary veins were incompletely visualised owing to acoustic window limitations. Multidetector computed tomography (CT) of the thorax confirmed infra-cardiac TAPVD with common venous channel obstruction at the oesophageal hiatus. CT was used instead of MRI because of the unstable clinical condition of the patient, enabling rapid diagnosis and minimising time out of the intensive care unit. After the definitive diagnosis was made, surgical correction was attempted. Unfortunately, the patient died of respiratory failure shortly after surgery.

Discussion

The radiologist usually suspects congenital heart disease in the face of an increased CTR (cardiomegaly). Assessment of the CTR is a routine basic component of evaluation of the paediatric chest radiograph. Before the age of 2 years, the CTR should not exceed 60%; after 4 years it should not exceed 50%, and between 2 - 4 years of age it should range between 60 - 50%. The great majority of congenital cardiac lesions will manifest with cardiomegaly. To enable a differential diagnosis, further evaluation is made in assessing the state of the pulmonary vasculature (i.e. oligaemia or plethora) and determining whether the patient is clinically cyanosed or not.

The presented case demonstrates a rare but important exception to the algorithm outlined above. Recognising pulmonary oedema (severe pulmonary venous congestion) with normal cardiac size is a difficult diagnosis. In the neonatal period, pulmonary oedema is often mistaken for primary pulmonary pathology such as surfactant deficiency syndrome (hyaline membrane disease), transient tachypnea of the newborn or infection on chest radiographs. Recognition of septal lines (usually central Kerley A lines rather than Kerley B lines) is key to the diagnosis.[1] The differential diagnosis of infantile pulmonary oedema is limited (Table I). If the heart is not enlarged, the differential is further limited and the most likely diagnosis would be infra-cardiac TAPVD. Only the very rare entities of cor triatrium and congenital mitral stenosis could mimic the radiographic pattern but, in reality, these diagnoses can usually be diffentiated by recognising atrial enlargement. With infra-cardiac TAPVD, the common pulmonary venous channel is almost invariably obstructed at the diaphragmatic hiatus, as in this case (Figs 1 and 2); this leads to severe pulmonary congestion. The point of obstruction is too far removed from the heart (from a haemodynamic point of view) to result in chamber enlargement. TAPVD usually results in severe cyanosis. There is a mandatory cardiac shunt (usually ASD or patent

Table I. Causes of neonatal pulmonary oedema

- Infantile coarctation
- Congenital aortic stenosis
- Hypoplastic left heart syndrome
- Congenital mitral stenosis
- Cor triatrium
- Obstructed TAPVR

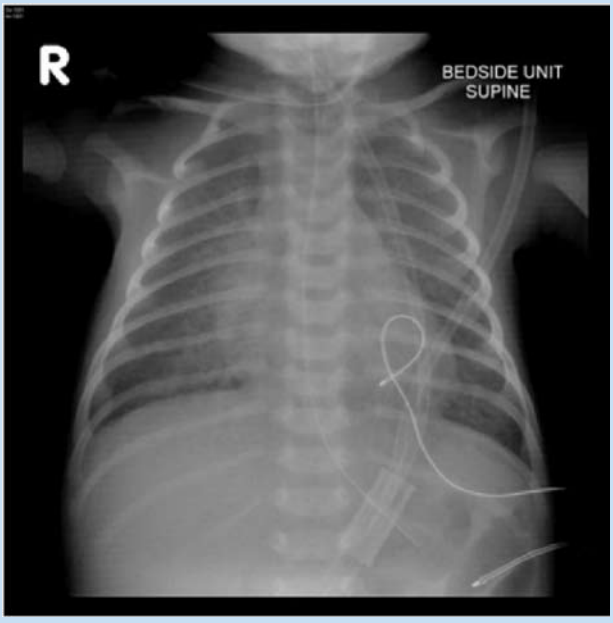

Fig. 1. Chest radiograph of the patient showing evidence of severe pulmonary venous congestion with a normal cardiothoracic ratio.

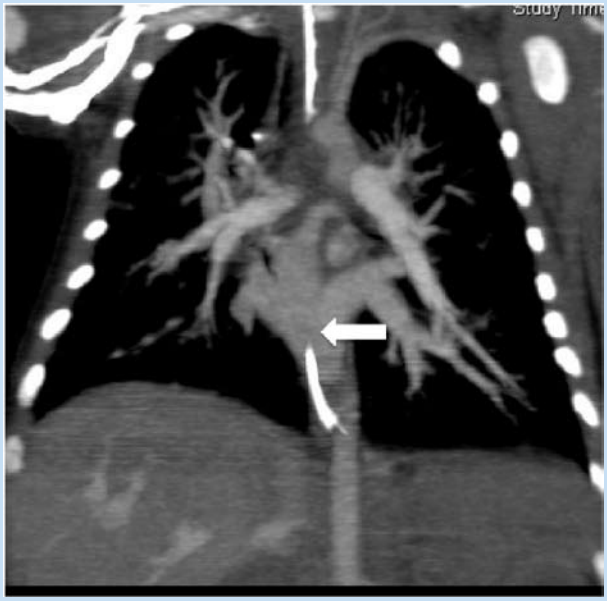

Fig. 2a. MIP reconstruction of the CT angiogram showing the four pulmonary veins converging to form a common channel (arrow) rather than entering the left atrium.

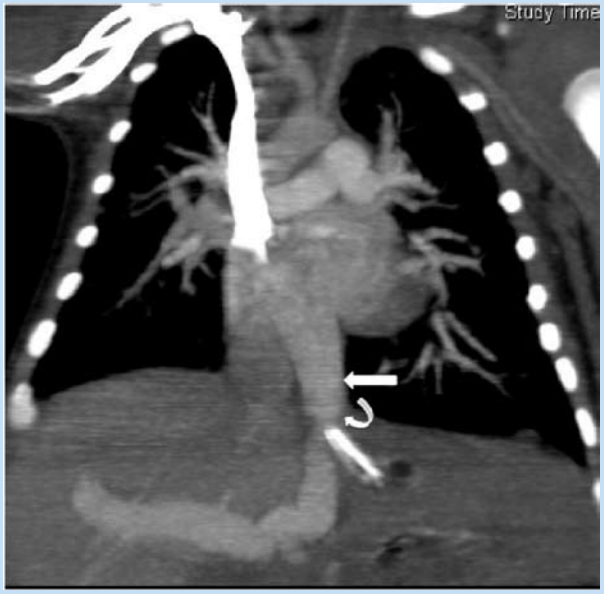

Fig. 2b. MIP reconstruction showing the common pulmonary venous channel (straight arrow) descending through the diaphragm to drain into the portal venous system. Note the narrowing of the common venous channel at the oesophageal hiatus (curved arrow), the position confirmed by the nasogastric tube.

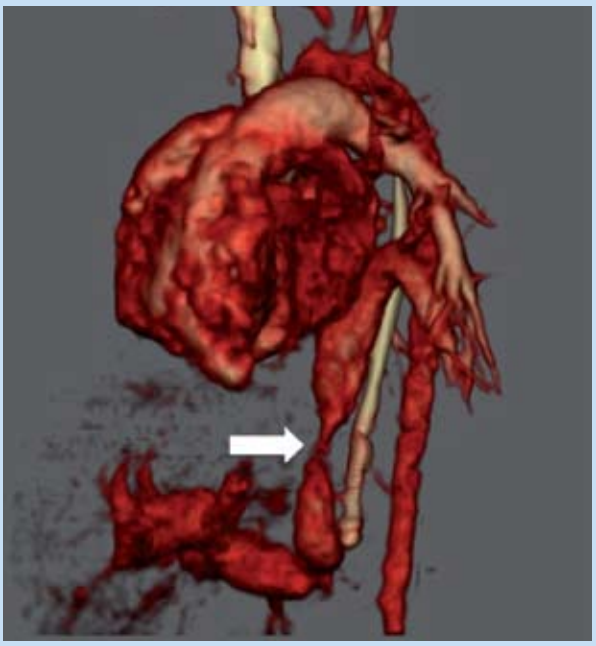

Fig. 2c. Volume-rendered image of the CT angiogram showing the narrowing of the common venous channel (arrow) at the oesophageal hiatus.

ductus arteriosis (PDA)) in patients with TAPVD, the absence of which is a fatal condition.

Infra-cardiac TAPVD represents approximately 10% of congenital TAPVD. The other variations are supracardiac, intracardiac and mixed, with the former two not usually being obstructed and radiographically classically manifesting as pulmonary plethora. Partially anomalous pulmonary venous return (PAPVD) is in contrast usually a minor congenital abnormality, often first detected in adulthood. PAPVD is often associated with an ASD or pulmonary lobar hypogenesis (e.g. Scimitar syndrome).[2]

Conclusion

The role of the general radiologist is vital in suspecting the diagnosis of infra-cardiac TAPVD based on the plain radiograph. Recognising pulmonary venous congestion without cardiac enlargement is a difficult radiological assessment but, when recognised, the diagnosis of infra-cardiac TAPVD can be strongly suggested.

1. Grainer R. Congenital Heart Disease: General Principles. In: Grainger RG, Allison D. *Diagnostic Radiology: A Textbook of Medical Imaging*. 3rd ed. New York: Churchill Livingston, 1997: 657-675.
2. Dahya V, Mayosi BM. Assessing scimitar syndrome-use of MRI and MRA. *S Afr Med J* 2007; 97(4): 248-249.