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Images in Congenital Cardiac Disease

An accessory left pulmonary artery

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Keywords: Pulmonary arterial sling; anatomy; congenital heart malformation

We present an unusual pattern of pulmonary arterial branching in an infant with antenatal diagnosis of a muscular ventricular septal defect. He was born at 36 weeks gestation with soft dysmorphic features, including micrognathia and clinodacyly, but no signs of respiratory distress. Ultrasonic interrogation also revealed pelvicalyceal dilation of the kidneys. Genetic testing confirmed a normal male karyotype, with no 22q11 deletion. The post-natal echocardiogram showed an unrestrictive muscular ventricular septal defect opening to the inlet of the right ventricle. The left-sided aortic arch was slightly hypoplastic in its transverse segment, with mild coarctation. The arterial duct was not patent. In the parasternal short axis view (Fig. 1), the pulmonary trunk (PT) was seen to divide normally into right (RPA) and left (LPA) branches. An accessory pulmonary artery (AP) was also present, arising from the right pulmonary artery and extending to supply the left lung (AO: aorta). (To see movie clips of the echocardiogram visit website http://journals.cambridge.org/cty). Computed tomographic axial cuts (Figs 2 and 3) confirmed these findings, excluding a pulmonary arterial sling, while three-dimensional reconstruction (Fig. 4) showed the left upper lobe to be perfused by the left pulmonary artery, and the left lower lobe, by the accessory pulmonary artery. Both of these arterial branches lay anterior to the oesophagus. We believe the first branch arising from the left side of the pulmonary trunk to be the "true" left pulmonary artery, as it has a normal pattern of distal arterial supply,

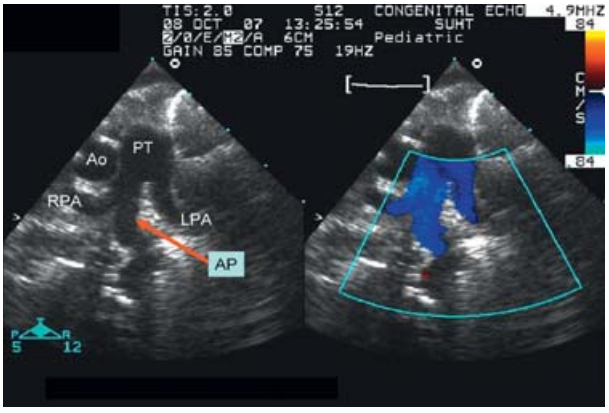

Figure 1.

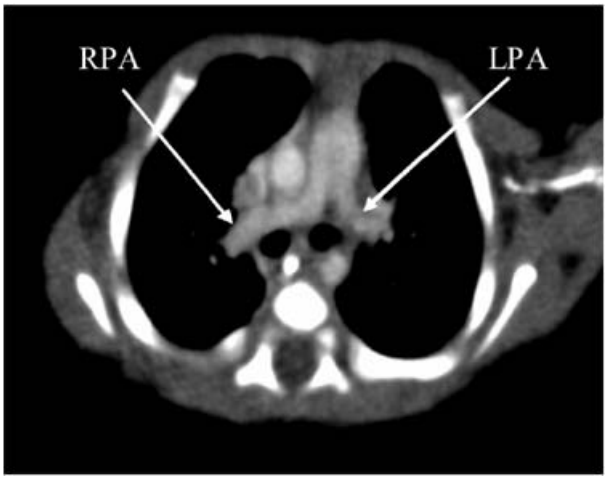

Figure 2.

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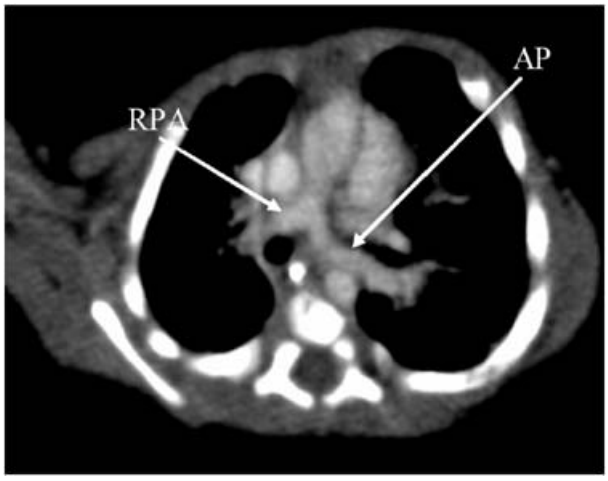

Figure 3.

confirmed on computed tomography. The patient did not undergo surgery.

In the setting of a pulmonary arterial sling, the left pulmonary artery arises from the right pulmonary artery, encircles the distal trachea, and courses between the posterior oesophagus and the anterior trachea. It is an uncommon but potentially lethal vascular anomaly that can produce obstruction of the airways.[1] In contrast, the left pulmonary artery in our patient arose in the normal place. The accessory left branch arose from the right pulmonary artery, but passed anterior to the oesophagus, without causing tracheal obstruction. To our knowledge, this pattern has not previously been described. The embryological basis for this anomaly might be similar to the explanation for pulmonary arterial sling as offered by Sade and colleagues,[1] namely persistence of the distal half of the sixth pharyngeal arch artery and left lung bud, with normal development of a "true" left pulmonary artery.

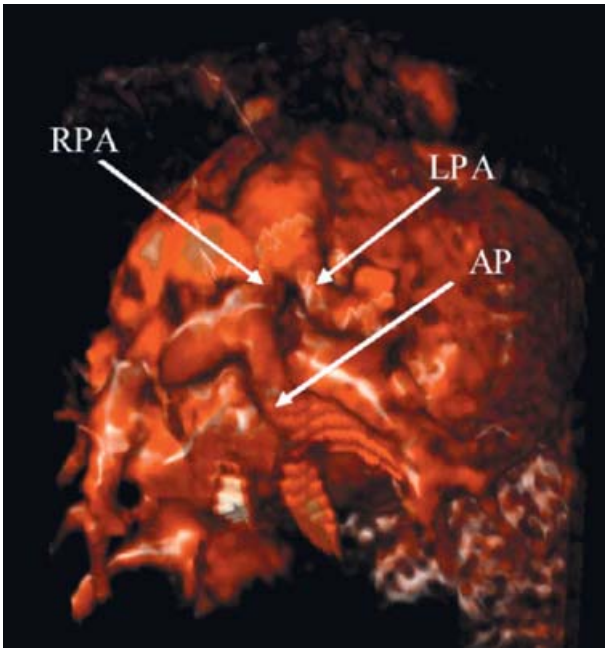

Figure 4.

Reference

1. Sade RM, Rosenthal A, Fellows K, Castaneda AR. Pulmonary artery sling. J Thorac Cardiovasc Surg 1975; 69: 333–346.

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