Type 2 left pulmonary artery sling –
Association with decreased right lung volume
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Introduction

Left pulmonary artery sling (LPAS) is a rare congenital anomaly in which the LPA originates from the posterior aspect of the right pulmonary artery and courses between the trachea and esophagus to reach the left lung. Approximately 50% of LPAS cases are associated with intrinsic tracheal stenosis.

There are two major types of left pulmonary artery sling. Type 1 LPAS usually compresses the distal trachea and right main stem bronchus resulting in hyperinflation of the right lung. A tracheal bronchus may be present. Type 2 LPAS is characterized by a more inferiorly located left pulmonary artery sling and abnormal bronchial branching with a low T-shaped carina. Long segment tracheal stenosis with complete cartilaginous rings is a common accompaniment often with bilateral hyperinflation. Other lung abnormalities may also occur, including right-side pulmonary hypoplasia and agenesis as well as scimitar syndrome and other foregut lesions.

We present 5 cases of Type 2 LPAS with diminished right lung volume in order to emphasize the association of this appearance with right lung vascular abnormalities.

Anatomic type of LPAS

![Anatomic types of PA sling (Wellis, et al.)](image)

Figure 1. Anatomic types of PA sling (Wellis, et al.) – solid circle denotes left pulmonary artery origin. RUL Right upper lobe bronchus, ILB intermediate left bronchus, BRB bridging right bronchus, LMB left main bronchus

Materials and Methods

5 Children (3 males and 2 females) aged 1 day to 6months presented with respiratory distress and decreased right lung volume with ipsilateral cardiome diastinal shift and poorly visualized airways on chest radiographs. All had plain chest radiographs, echocardiography and bronchoscopy; some had angiography (3), barium swallow (3), chest CT/CTA (3), and MRI(4).

Results

Case 1. Type II A LPAS in a 4-day-old boy

Fig.2 A&B. Poor tracheal visualization on both views with a low, T-shaped carina (arrow) and right upper lobe atelectasis.

Fig.3 C-F.Sup to Inf Axial MR images demonstrate right upper lobe tracheal bronchus (arrow) and adjacent atelectasis(C). D. The low LPAS (arrowhead) courses between the esophagus (star) and stenotic intermediate left bronchus (arrow). E. Just below the sling is the T-carina with a right bridging bronchus (double arrow). Posterior 3D reconstruction of the airway and vascular anatomy(F). Note also aberrant right subclavian artery.

Case 2. Type II A in a 6-month-old girl

Fig.4 A. Poor tracheal visualization; right lung decreased volume? patchy atelectasis/pneumonia.

Fig.4 B&C. Oblique coronal MR images demonstrate left pulmonary artery (arrow) arising from right pulmonary artery (arrowhead) just behind the airway. Note low T-shaped carina (arrow) with moderate long segment lower tracheal narrowing and bridging right bronchus (arrowhead).

Case 3. Type II B LPAS in a 4-month-old girl

Fig.5 A-B. Poorly visualized trachea; probable absent hypoplastic right lung with ipsilateral mediastinal shift and herniation of left lung to right. Lateral airway high KV fluoroscopic image shows narrowed lower trachea bowed posteriorly.

Fig.5 C-G. Noncontrast CT(C) and Sup to Inf axial MR images (D-G) confirm absent right lung. The lower trachea (arrow) is displaced posteriorly by the crossing aortic arch and is also stenotic. The LPA (arrowhead) passes between the airway (arrow) and the esophagus (dashed arrow). This represents left pulmonary artery sling anatomy but with absence of the right bronchial origin. Type G - 3D frontal and lateral reconstructions of the anatomy. Note common origin of carotid artery.

Case 5. Type II B with Scimitar syndrome in a newborn boy

Fig.6 A. Plain chest AP-hypoplastic right lung with probable right scimitar vein (arrow). B-C. Scimitar vein (arrow) and horseshoe lung (arrowhead) shown on chest CT scan.

Fig.6 D-E. Coronal and axial MR images demonstrate scimitar vein (arrow) draining to IVC and low T-carina with lower tracheal stenosis. The upper LPA is normal (arrowhead), the lower LPA arises from the right side (dashed arrow) as a LPS. Note the seler lower LPA (dashed arrow) crossing the aorta. Subcarinal atelectatic sequestered horsehose lung had systemic arterial supply (not shown).

Conclusions

Decreased right lung volume and poorly visualized/narrow trachea/low T-carina should raise suspicion for Type 2 LPAS. Right lung abnormalities are common ranging from atelectasis to hypoplasia/agenesis. Associated airway, lung and vascular anomalies may change the prognosis and management of these children. High quality MR and CT imaging display the complete spectrum of extracardiac pathologic anatomy.

References