Journal of Pediatric Surgery (2011) **46**, 2190–2194

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Journal of Pediatric Surgery

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Pediatric surgical image

Left pulmonary artery sling with right lung aplasia

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Received 31 May 2011; revised 26 July 2011; accepted 27 July 2011

Key words:
Pulmonary artery sling;
Lung aplasia;
CT scan;
Congenital

Abstract We report 2 cases of right pulmonary aplasia with left pulmonary artery sling responsible for severe respiratory symptoms. Repositioning of the left pulmonary artery without tracheal surgery was successful in both patients. The postoperative course was simple, and the outcome was favorable at last follow-up (after 2 years and 3 months, respectively). Computed tomography provided an accurate diagnostic evaluation that helped to choose the best surgical technique.

Unilateral absence of a lung is a rare congenital abnormality with a birth prevalence of about 1 in 15,000. In pulmonary aplasia, the lung is absent, but there is a rudimentary bronchus, whereas both the lung and the airway are missing in pulmonary agenesis [1]. Unilateral absence of a lung affects the 2 sides equally. Right-sided pulmonary aplasia or agenesis carries a worse prognosis because the greater displacement of the heart and mediastinum results in substantial distortion of the great vessels and airway [2]. Aberrant left pulmonary artery (LPA) is also a rare congenital abnormality in which the LPA arises from the right pulmonary artery and courses between the trachea and esophagus to the left hilum. An aberrant LPA responsible for tracheal compression is known as *LPA sling* and is accompanied in 50% of patients with complete tracheal rings manifesting as congenital tracheal hypoplasia [3].

Here, we report 2 cases of right-sided pulmonary aplasia with LPA sling. We emphasize the role for computed tomography (CT) in clarifying the anatomic features of this rare birth defect.

1. Case reports

1.1. Clinical history and physical examination

The first patient was a girl born at full term to nonconsanguineous parents. The birth and perinatal course were uncomplicated. At 3 weeks of age, she required admission for respiratory distress from bronchiolitis caused by the respiratory syncytial virus. At the time, absence of the right lung was diagnosed together with several other birth defects (left kidney agenesis, second left rib agenesis, and supernumerary T9/T10 hemivertebrae). She was referred to our institution at the age of 4 months with severe respiratory distress requiring emergency intubation. Intubation proved extremely challenging, and mechanical ventilation was difficult to manage. Her body weight at the time was 5 kg.

The second patient was diagnosed with absence of the right lung during the second trimester of pregnancy. After birth, she experienced recurrent respiratory tract infections that were treated with physical therapy and continuous

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0022-3468/$ – see front matter © 2011 Elsevier Inc. All rights reserved.
doi:10.1016/j.jpedsurg.2011.07.026

antibiotic therapy. She was 2 years old and weighed 12 kg when she was referred to our institution because of respiratory status deterioration. The only abnormal physical finding was asymmetry of the chest wall with prominence of the left side.

1.2. Initial investigations

In both patients, the chest x-ray showed hyperinflation of the left lung with displacement of the mediastinum and heart into the right hemithorax. By echocardiography, dextroposition of the heart was the only abnormality. There were no structural cardiac anomalies or patent ductus arteriosus. No right pulmonary artery was seen. Flexible bronchoscopy was performed in both patients. Neither patient had complete tracheal rings. The right mainstem bronchus was present but was very short and completely obstructed before its division. In the first patient, severe compression of the origin of left main bronchus was noted. The second patient also had compression of the left bronchus, but the most striking abnormality was marked deviation of the distal trachea.

1.3. Preoperative CT

Preoperative CT was performed using a dual source CT scanner (Somatom Definition; Siemens AG, Forchheim, Germany) with a single prospective electrocardiogram-gated axial thoracic acquisition at the systolic phase (40% of the R-R interval), without padding, from the upper thoracic inlet to the diaphragm. A low-dose protocol with imaging at 80 kV and weight-based adjustment of the tube current was used. Iodinated contrast medium (Iopromide, 300 mg/mL; Ultravist 300, Bayer Schering, Pharma, Berlin, Germany) was injected using a single-head power injector (Stellant; Medrad, Indianola, PA). The contrast dose was 2 mL/kg body weight, and the injection rate was also adapted to body weight. All images were transferred to an external workstation (Leonardo, Siemens Medical Solution, Forchheim, Germany). Three-dimensional techniques such as multiplanar reformation, maximum intensity projection, and volume rendering were used.

Herniation of the left lung and complete aplasia of the right lung were seen in both patients. The right lung parenchyma and right pulmonary artery and veins were completely absent. A rudimentary right bronchus was visible, but there was no right bronchial tree. The aortic arch, which was rotated and displaced to the right, crossed the anterior aspect of the trachea, causing tracheal compression. An aberrant LPA located between the trachea and esophagus arose directly from the main pulmonary trunk and caused compression of the posterior aspect of the trachea (Fig. 1A-B). The left pulmonary veins emptied normally into the left atrium.

The tracheal reconstruction images showed differences in the type of tracheobronchial compression between the 2 patients. In the first patient, the main site of compression was the left bronchus. In the other patient, airway compression was chiefly related to an S-shaped deformation of the trachea (Fig. 2), with anterior compression by the aortic arch and posterior compression by the LPA (Fig. 3A-B).

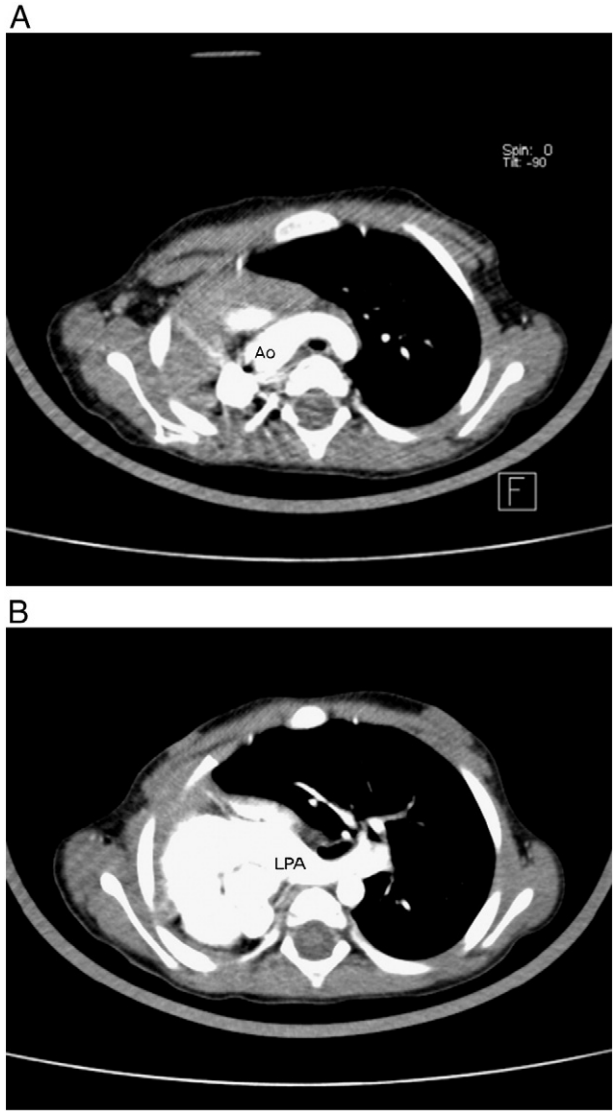

Fig. 1 Preoperative axial multidetector CT view in the second patient. A, The aortic arch, which is rotated and displaced to the right, can be seen to cross the trachea anteriorly. B, The aberrant LPA runs behind the trachea. Ao indicates aorta.

1.4. Operative technique

Translocation of the LPA was performed in both patients. A midline sternotomy was performed, and the pericardium was opened. A normothermic cardiopulmonary bypass without cross-clamping was established between the right atrium located deep in the right side of the chest and the ascending aorta. The LPA was dissected, separated from the trachea, and divided between 2 clamps. The retrotracheal portion of the artery was translocated anterior to the trachea.