



# An infant with left pulmonary artery sling

Ka-lan Ip 葉嘉欣 and Theresa Ngan-Ho Leung 梁銀河

Department of Paediatrics and Adolescent Medicine, Pamela Youde Nethersole Eastern Hospital, Hong Kong

## Abstract

Pulmonary artery sling (PAS) is a rare congenital vascular anomaly with variable clinical presentations with stridor being the commonest symptom in early infancy due to airway obstruction. Here we describe a male infant with PAS presented as viral laryngotracheobronchitis and subsequently diagnosed to have left pulmonary artery sling. The clinical presentation and subsequent management were discussed followed with a literature review of this condition.

Keywords: Pulmonary artery sling, Stridor

## Case report

A male baby was noted to have intermittent mild noisy breathing since 1 month of age. He was admitted at 4 months old with hoarseness, moderate severe stridor and respiratory distress preceded by coryzal symptoms. He was treated as viral laryngotracheobronchitis with dexamethasone and nasal continuous positive airway pressure (CPAP). He recovered gradually after the acute episode but remained to have intermittent stridor. While fibre-optic bronchoscopy was planned he was re-admitted for increasing biphasic stridor and dyspnea. Review of chest radiography (CXR) showed the lower part of the trachea was inconspicuous indicative of possible narrowing. Echocardiogram revealed the left pulmonary artery (LPA) originated from the right pulmonary artery (RPA) suggestive of left pulmonary artery sling (LPAS) while other cardiac structures were normal. Contrast computed tomography (CT) thorax confirmed the diagnosis of type IB LPAS with narrowing of the trachea above carina due to extrinsic compression by the LPAS and a right tracheal bronchus. The diameter of trachea above the LPAS measured 5.1 mm while the narrowed portion measured 3.2 mm. The length of the stenotic portion measured 20.1 mm, around 30% of the total tracheal length (Figures 1a & 1d). There was an incidental finding of a tracheal diverticulum arising from the right posterior aspect of the trachea just above the level of aortic arch (Figure 1c). He had corrective surgery for the vascular and airway abnormality within one month

after diagnosis. Pulmonary arterioplasty with ligation of a patent ductus arteriosus (PDA) found intra-operatively together with sliding tracheoplasty was successfully performed under cardiopulmonary bypass. The operation was complicated by left vocal cord palsy which was not causing any significant airway obstruction. Postoperatively he had mild residual stridor when agitated but otherwise no significant respiratory distress. Rigid laryngotracheobronchoscopy showed an adequate airway admitting a size 4 endotracheal tube. Follow up CT thorax showed no significant tracheal narrowing (Figure 1b). Upon follow up his stridor and hoarseness resolved.

## Discussion

### Clinical presentation of LPAS

LPAS was first described in 1897 by Glaevecke and Doehle.<sup>1</sup> It is a rare congenital condition in which the LPA originates from the posterior aspect of the RPA and courses over the right main bronchus and then posteriorly between the trachea and the esophagus to reach the left hilum. Timing and severity of clinical manifestations can be highly variable ranging from life-threatening airway obstruction to mildly symptomatic cases, depending on the extent of airway obstruction mainly contributed by associated tracheal stenosis. Most cases present in early infancy with stridor and respiratory distress which could be exacerbated by infections as in our patient. Other symptoms include dysphagia due to compression of the esophagus, failure to thrive and recurrent chest infections. Some patients may be having mild symptoms or remained asymptomatic and present in late childhood or adulthood with non-specific

\*Author to whom correspondence should be addressed.

Email: iki481@ha.org.hk

respiratory symptoms like chest pain, cough, orthopnea and exertional symptoms. In cases with associated bronchial obstruction there could be hyperinflation of the right lung and mistaken as congenital emphysema.<sup>2</sup>

**Classification**

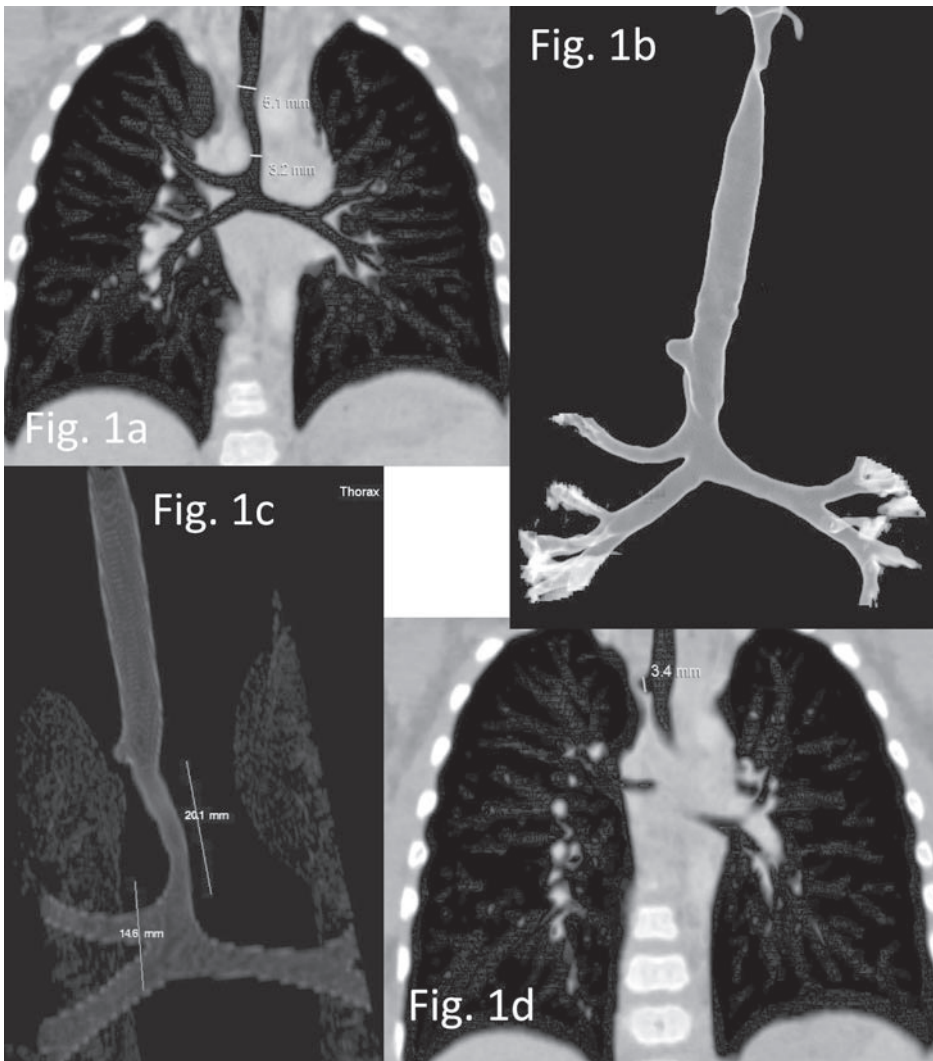
LPAS can be classified according to the level of sling and associated tracheal abnormality.

- Type I: PA sling at T4-5 level just above the normally positioned carina
  - ◆ Type IA: no right tracheal bronchus
  - ◆ Type IB: with right tracheal bronchus
- Type II: PA sling at T6-7 level just above the abnormally low and horizontal carina
  - ◆ Type IIA: with a right upper lobe "tracheal" bronchus at usual level of carina, distal airway stenosis and a bridging right bronchus

- ◆ Type IIB: same as type IIA except no separate right upper lobe bronchus

Types II LPAS have a long-segment tracheal stenosis of ring sling complex extending from the usual level of carina to the abnormally low carina and therefore it has a more severe clinical manifestation and carries a poorer prognosis as compared. Yet Type II LPAS is more common than Type I and Type IIB is the commonest.

The commonest association is tracheal stenosis (100%), followed by PDA (39%), right tracheal bronchus (22%), underdeveloped right lung (22%) and persistent left superior vena cava (22%) according to a retrospective review by the National Taiwan University College of Medicine comprising 18 children (6 girls and 12 boys aged 5 days to 17.5 years with mean age 1.5 years).<sup>3</sup>



**Figure 1.** (a & d) CT thorax, sagittal cut demonstrating the LPAS, tracheal bronchus and the tracheal stenosis; (c) 3D reconstruction of the trachea before repair, and (b) post-repair.



Other associated vascular anomalies include aberrant right subclavian artery, aortic coarctation, partial anomalous pulmonary venous return, atrial septal defect, ventricular septal defect, Tetralogy of Fallot, and more complex anomalies with single ventricle physiology. In our patient his LPAS was associated with tracheal stenosis, right tracheal bronchus, tracheal diverticulum and a PDA.

### Embryology

Development of the great vessels begins at 20-22 days and the six arches develop and regress in a craniocaudal fashion. The left post-brachial vessels connect to the left 6th brachial arch to form the LPA while the right post-brachial vessels capture a major vascular supply from the right 6th brachial arch to make an RPA. LPAS occurs when there is failure of the connection of left post-brachial vessels with the left 6th brachial arch. Instead, the connection of the left post-brachial vessels occurs with the right 6th brachial arch dorsally to the trachea forming the pulmonary artery sling.<sup>3,4</sup>

### Diagnosis

Narrowing of the trachea may be visualised on CXR and unilateral hyperinflation (usually right side of the lung) may be present due to airway obstruction. In the early newborn period, the abnormally ventilated right lung may be fluid filled and appear more solid due to prolonged retention of fetal lung fluid.<sup>5</sup> Echocardiogram can aid in diagnosis by identification of the vascular anomaly, other associated vascular and cardiac lesions. CT thorax with contrast is the ultimate diagnostic modality for diagnosis and pre-operative planning, outlining the anatomical relationship between airway and vascular anomalies and associated airway or pulmonary abnormalities.<sup>5-7</sup> MRI is an alternative imaging modality with its advantage being the lack of ionising radiation exposure.<sup>7</sup> However, CT is preferred to MRI because it provides much better visualisation of lung parenchyma and airways, higher spatial resolution, and faster scanning with lower requirement for sedation. It also allows 2D and 3D display of anatomy including visualisation of airway in the form of virtual bronchoscopy. The new low dose protocol for CT scan also minimises ionising radiation.

Fibre-optic bronchoscopy is a common investigation for stridor in children. Fibre-optic bronchoscopy is good at evaluating mucosal details, dynamic airway changes, location of extrinsic compression and the extent of airway stenosis. However, in cases of tracheal stenosis, the scope may not be passed beyond the stenotic part of the trachea<sup>8,9</sup> and it has potential risks of complete

obstruction of the airway leading to acute respiratory crisis.<sup>9</sup> Moreover, the unusual anatomy of airways seen via bronchoscope could be misinterpreted.<sup>9</sup> For a patient who is highly suspicious for the condition as in our patient, we may consider CT thorax and echocardiogram as first line investigations. Bronchoscopic evaluation (before and after operation) for direct visualisation of the airway is useful but is safer to be done in the operating room, where airway management can be carefully controlled.

### Surgical management and outcome

Untreated pulmonary sling associated with significant tracheal stenosis carries a high morbidity and mortality. The first successful surgical repair of LPAS was performed at Children's Memorial Hospital in 1953 by Potts, Holinger and Rosenblum on a 5-month old baby with recurrent attacks of dyspnea and cyanosis. Preoperative bronchoscopy showing distal airway obstruction and diagnosis was made intraoperatively. Reconnection of the LPA to the MPA at the hilum was performed. The child was well up to 24 years of follow up with normal exercise tolerance.<sup>10</sup> Over the past decades recognition of the importance of the associated tracheal anomaly and the evolution of techniques in tracheal repair from pericardial patch tracheoplasty to tracheal autograft to sliding tracheoplasty further improved surgical outcome. Sliding tracheoplasty was first reported in 1998 by Oshima and colleagues of Kobe Children's Hospital<sup>11</sup> and currently one of the commonly employed surgical treatments for tracheal stenosis.

A review by Yong et al<sup>12</sup> of the Royal Children's Hospital in Melbourne including 21 patients all diagnosed LPAS with age at surgery ranging from 2.3 to 16.4 months (median 5.9 months) undergoing repair surgery over a period of 27 years from 1984 to 2011. Twelve (57.1%) of patients suffered from tracheal stenosis and 9 (42.9%) had more than or equal to 50% involvement of the total tracheal length. Tracheal repair surgery was performed in all cases with tracheal stenosis. There were three deaths (14.3%) occurring at 19 days, 4.4 months and 5 months post-operatively with two deaths (22.2%) in the first 10 years (1984-1993) and one death (14.3%) the next 10 years (1994-2003) and none (0%) in the most recent 7 years (2004-2011). All deaths occurred in patients with tracheal stenosis who underwent tracheal repair. There was no death case since year 2004 after the introduction of the slide tracheoplasty technique.

Backer et al<sup>13</sup> analysed the database of all patients diagnosed to have LPAS and undergoing surgery at



Children's Memorial Hospital from 1985 to 2012. There were 34 patients with age ranging from 9 days to 43 years (mean  $2.1 \pm 7.5$  years) and majority were less than 1 year old. Twenty-seven (79.4%) had tracheal stenosis and 24 (70.6%) suffered from stenosis involving more than or equal to 50% of the total tracheal length. Since year 2000 all patients in the case series had CT thorax with 3D reconstruction and all patients had preoperative rigid bronchoscopy in the operation theatre immediately before the surgery. For the repair technique of tracheal stenosis 7 had pericardial tracheoplasty, 10 had tracheal autograft, 4 had tracheal resection and 5 had sliding tracheoplasty. There was no early post-operative death reported in this series. There were four late deaths (11.8%) at 2 months to 5.8 years after operation. Two patients with patch tracheoplasty and tracheal autograft done developed cardiac arrest after patch dissection and tracheal perforation respectively. Two other patients died of liver failure due to concomitant biliary atresia and pneumonia 5.8 years after the operation. Three out of the 7 patients who had pericardial patch tracheoplasty required surgical revision. One out of 10 patients receiving a tracheal autograft required placement of a stent and permanent tracheostomy.

All patients with LPAS should be repaired with correction of the vascular structural abnormalities. For tracheal stenosis, tracheal resection may be done for short segment stenosis involving less than six tracheal rings and sliding tracheoplasty is the choice of operation for longer segment of stenosis.<sup>13</sup>

## Conclusion

Pulmonary artery sling is commonly associated with tracheal stenosis which determines the severity of symptoms and prognosis. With the advance in multi-detector CT with virtual bronchoscopy, the anatomical abnormalities can be displayed for early diagnosis and pre-operative planning, hence CT thorax with contrast should be the first line investigation in highly suspicious cases. With the introduction of cardiopulmonary bypass and sliding tracheoplasty, most cases of LPAS could be repaired with good prognosis.

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