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CASE REPORT

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Anomalous left pulmonary artery: case reports exploring anatomic variants

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Abstract

Background Pulmonary artery sling (PAS) is a rare congenital anomaly where the left pulmonary artery (LPA) branches from the right pulmonary artery, compressing the trachea and esophagus and frequently leading to respiratory distress in infants. Surgical intervention, such as LPA reimplantation or translocation, is crucial to relieve airway compression and restore normal pulmonary function.

Case presentation This report highlights varied LPA anatomies, including a unique case of an anomalous LPA without true sling formation but causing tracheal compression, alongside two typical PAS cases. Respiratory symptoms were successfully mitigated in all three cases without concomitant tracheal reconstruction.

Conclusions This report underscores the necessity of thorough preoperative assessment of airway anatomy and highlight the importance of individualized surgical planning for anomalous LPA.

Keywords Vascular ring, Anatomy, Trachea, Pulmonary arteries, Congenital heart disease

Background

Pulmonary artery sling (PAS) is a rare congenital anomaly in which the left pulmonary artery (LPA) branches from the right pulmonary artery (RPA) [1]. The LPA then travels posterior to the trachea and anterior to the esophagus, often causing tracheal compression. Infants with PAS typically present within the first month of life with nonspecific signs of respiratory distress, such as wheezing, stridor, cyanosis, recurrent pneumonia, emphysema, and atelectasis [2]. In approximately 50–65% of the cases, PAS presents with complete tracheal rings, a condition known as the "ring-sling" complex [3]. This can further exacerbate respiratory symptoms and contribute to tracheal stenosis [4, 5]. In the absence of surgical intervention, PAS can result in fatal respiratory insufficiency.

Embryologically, the "space available" theory has been proposed to explain the development of an LPA sling, which states that all structures in the embryonic foregut mesoderm, such as the lung buds, bronchial buds, and the sixth branchial arch, compete for space [2]. Early branching of the right bronchus may create more space around the developing trachea, allowing the left pulmonary vessel to approach the right ventral sixth branchial arch and form a PAS. This theory may also explain the associated congenital anomalies, such as a patent ductus arteriosus (PDA), and the various presentations of anomalous LPA [1, 2].

This report includes a unique LPA course in which the LPA arises from the RPA and passes anteriorly to the trachea, avoiding formation of a true sling but distally

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compressing the left mainstem bronchus. To our best knowledge, three other cases of anomalous LPA without sling formation have been described in literature [6–8]. One case described an anomalous LPA that coursed anterior to the trachea, which was associated with proximal tracheal stenosis but did not cause airway compression [7]. Another case was a partial anomalous LPA that arose from the RPA, coursing anterior to the trachea without causing airway obstruction [6]. The third case had a similar anatomy of LPA coursing anterior to the trachea but was associated with a right aortic arch and an aberrant left subclavian artery [8].

In addition to a case of anomalous LPA without sling formation, our report also presents two cases of typical PAS as a comparison, highlighting the diverse anatomical variations of anomalous LPA. Together, our case reports demonstrate the importance of thorough preoperative assessment for LPA anatomy and associated airway compression to inform surgical decision-making.

Case presentations

Case 1

A 5-year-old, former 24-week premature female weighing 14 kg presented with wheezing, reactive airway disease, and frequent respiratory infections. A chest computed tomography (CT) was performed, which demonstrated a tortuous LPA originating from the RPA at the level of the left mainstem bronchus (Fig. 1). The LPA coursed anterior to the trachea, avoiding tracheal compression. The anomalous pulmonary artery traveled anterior to the left mainstem bronchus, compressing it against the descending aorta. Proximal to the site of compression, the left bronchus was 3 mm in diameter, which narrowed to less than 1 mm in diameter at the site of maximal compression. An obstructive hyperinflated pattern was seen in the left upper lung lobe, which was consistent with airway compression leading to chronic air trapping. There were no tracheal rings identified on CT. This anatomic configuration was subsequently confirmed with rigid bronchoscopy.

Surgical repair began with a median sternotomy, dissection of the pulmonary trunk and its branches, and ligation of the ligamentum arteriosum. Cardiopulmonary bypass (CPB) was initiated. LPA was ligated at its origin from the RPA and divided. The distal LPA was dissected completely. The LPA was then mobilized and separated from the left bronchus, with an anastomosis established between the LPA and the anterolateral main pulmonary artery (MPA) using 6–0 Prolene sutures. The anastomosis was widened and augmented at its anterior edge utilizing an autologous pericardial patch attached by 6–0 Prolene sutures (Fig. 2). Bronchoplasty was not performed due to significant relief of airway obstruction

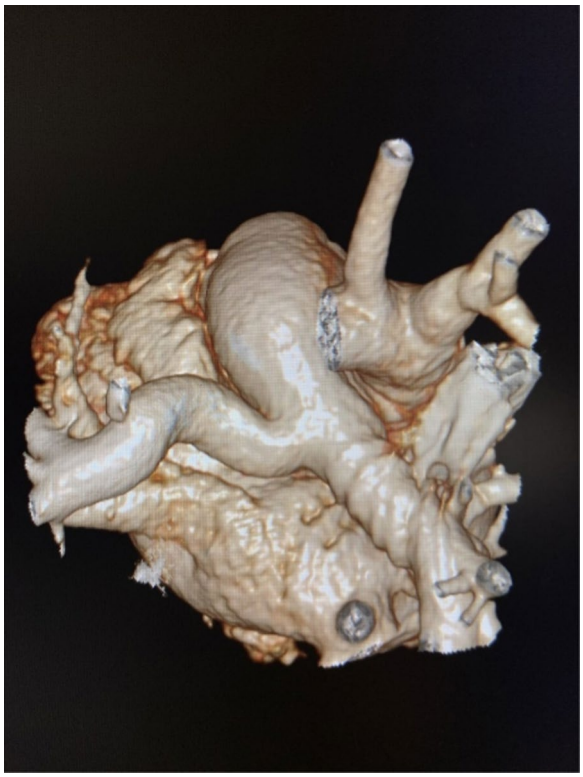

Fig. 1 Preoperative CT reconstruction of the first patient demonstrating anomalous origin of the LPA from the RPA

after reimplantation (Fig. 3). The patient was extubated at the conclusion of the case.

The patient was discharged 2 days after the operation in good condition. At 6-month follow-up, the patient had some residual hypoplasia of the LPA with a widely patent anastomosis and no respiratory symptoms. At 18-month follow-up, a Doppler ultrasound showed normal LPA velocity. A perfusion scan demonstrated 40% and 60% perfusion of the left and right lungs, respectively. The patient reported excellent health with no residual respiratory symptoms.

Case 2

A 4-year-old asymptomatic female weighing 26 kg was diagnosed with a type IA PAS. As described by Rahmath et al., a type 1A LPA sling has a normal carinal anatomy with bifurcation at T4/5 into the left and the right mainstem bronchi and is typically associated with mild airway compression [2]. During workup for an auscultated murmur, the patient was found to have subclinical tracheal compression and a small PDA. A CT scan and a rigid bronchoscopy confirmed the diagnosis. No complete tracheal rings were identified. The LPA originated from the RPA at the level of tracheal bifurcation. The LPA then traveled superior to the right

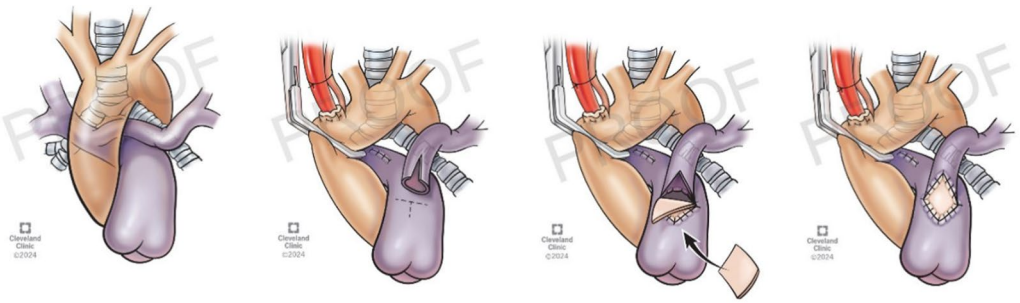

Fig. 2 Preoperative anatomy of the first patient. Mobilization of the LPA and anastomosis between LPA and MPA utilizing autologous pericardial patch

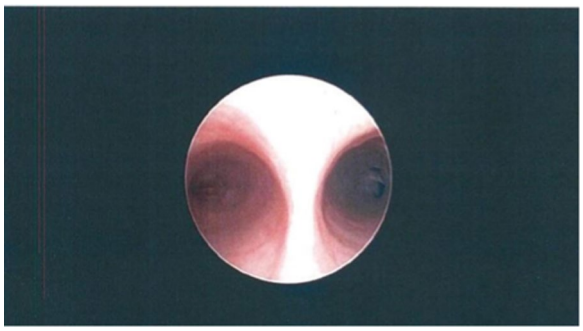

Fig. 3 Intraoperative bronchoscopy image from the first patient demonstrated no signs of airway obstruction

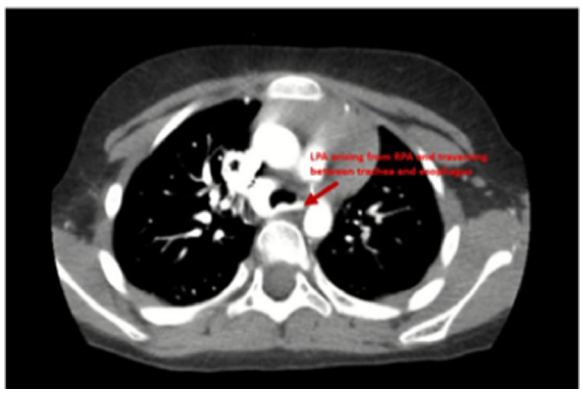

Fig. 4 Preoperative CT scan from the second patient demonstrated an anomalous

circulations. No evidence of pulmonary hypertension was identified on preoperative echocardiogram.

LPA arising from the RPA and traversing between the trachea and the esophagus.

An elective repair was performed via a median sternotomy. The PDA was dissected and ligated using 5–0 prolene sutures. The MPA, RPA, and the entirety of the LPA were then dissected, allowing visualization of the LPA within the hilum of the left lung. The RPA and LPA were clamped. The LPA was transected from the RPA. The remaining short cuff was oversewn with 6–0 Prolene sutures. The LPA was then mobilized from the posterior tracheal wall. The MPA was clamped and incised, and the LPA was spatulated and anastomosed to the MPA with 6–0 prolene sutures. The patient was extubated at the conclusion of the case and discharged home on postoperative day one.

At 1-month follow-up, an echocardiogram showed good biventricular systolic function. At 1-year follow-up, the patient had a widely patent anastomosis with some hypoplasia on the LPA (LPA: 12 mm, RPA: 13 mm). Of note, the patient's RPA was 11 mm and her LPA was 7.8 mm × 7.3 mm beyond the hilum at baseline. At 3-year follow-up, minor LPA stenosis was noted on rigid bronchoscopy (LPA: 11.6 mm, RPA: 13 mm) with mild compression of the left mainstem bronchus 3 mm, which was unchanged from the preoperative scan. A normal posterior trachea was also noted. A Doppler ultrasound performed showed a maximum LPA velocity of 1.8 m/sec. The patient was in excellent health without residual respiratory symptoms.

mainstem bronchus, posterior to the trachea, and anterior to the esophagus, resulting in dynamic compression of the distal trachea to 3 mm in diameter (Fig. 4). By comparison, the proximal trachea measured 8 mm in diameter. The LPA also received blood supply from a collateral vessel off the descending aorta, establishing a left-to-right shunt between the systemic and pulmonary

Case 3

A 4-month-old male weighing 7.3 kg was diagnosed with a type IA PAS with a small mid-muscular ventricular septal defect and a small patent foramen ovale after presenting with a murmur [2, 9]. A CT scan and a rigid bronchoscopy found 2–3 non-stenotic complete tracheal rings in the proximal to mid trachea. The anomalous LPA

branched off the distal RPA just before the bifurcation, then coursed posterior the trachea and anterior to the esophagus. An echocardiogram showed a LPA diameter of 4 mm, which was was smaller than the RPA diameter (5.5 mm) and was mildly hypoplastic.

Surgical repair was performed via median sternotomy. The PDA was dissected and divided. The MPA, RPA, and LPA were extensively dissected deep into the mediastinum posterior to the trachea all the way to the left hilum. After systemic heparinization, the aorta and right atrial appendage were cannulated. CPB was commenced without cooling. The RPA and LPA were clamped. The LPA was transected close to the RPA, leaving a sufficient cuff that was oversewn with 6–0 Prolene sutures. The LPA was then pulled through the sling course posterior to the trachea while maintaining its orientation. The MPA was clamped. An MPA incision was made. The LPA was spatulated to allow a generous anastomosis using 7–0 Prolene sutures. The patient was extubated at the end of the case.

At 2-week follow-up, an echocardiogram showed mild stenosis of the LPA. At 4-month follow-up, the LPA was found to be slightly hypoplastic (RPA: 5.5 mm, LPA = 4 mm) with no change in LPA or RPA size from baseline. At 18-month follow-up, the patient was in excellent health with no respiratory symptoms. A rigid bronchoscopy showed 2–3 tracheal rings without stenosis. A continuous Doppler ultrasound performed showed an LPA velocity of 2 m/sec.

Discussion

Our first case presents a unique course of an anomalous LPA that traveled anterior to the trachea without sling formation. This anatomic configuration caused no compression of the trachea, esophagus, or right mainstem bronchus. However, the distal LPA caused considerable compression of the left mainstem bronchus, necessitating correction of the anomalous LPA. In contrast, our second and third cases highlight more traditional PAS anatomy, where the LPA originated from the RPA immediately proximal to the bifurcation and traveled between the trachea and esophagus. Together, our case reports highlight the importance of thorough preoperative evaluation of anomalous PA courses to differentiate unique morphology from more traditional PAS anatomies.

Furthermore, each case highlights a different tracheal morphology, yet none of these patients underwent bronchoplasty or tracheoplasty at the time of repair. These patients had excellent postoperative outcomes with minimal residual respiratory symptoms, indicating the absence of important tracheal stenosis after PAS repair despite the absence of tracheal reconstruction. The ideal intervention for PAS has remained controversial, with a main debate being the necessity of tracheoplasty or bronchoplasty concurrent with PAS repair. Prior studies have found that PAS repair alone without tracheal or bronchial interventions achieved good long-term outcomes when there was minimal tracheal stenosis [5, 10, 11]. Our cases highlight the importance of thorough preoperative assessment for tracheal stenosis, particularly in the absence of complete tracheal rings, in guiding surgical planning. Noninvasive imaging techniques like optical coherence tomography may be applied to detect the presence of dysplastic or fragmented cartilages within the tracheal wall [12]. Additionally, computational fluid dynamics may assess for the effects of possible structural wall pathologies on flow velocity and wall stress, which may suggest symptomatic tracheal stenosis [13].

Although the small sample size of this report does not allow for definitive conclusions about surgical approach, the outcomes of our cases agree with literature findings that tracheal reconstruction may not be essential in anomalous LPA patients without complete tracheal rings. With careful patient evaluation and selection, anomalous LPA repair may be performed independently of tracheal reconstruction with low to minimal morbidity and mortality. However, despite the excellent early outcomes, patients remain at risk for later complications. For example, Choi et. al followed 22 PAS patients for an average of 6.9 years, during which 5 of patients needed PA reintervention and 7 patients developed respiratory symptoms that required hospital readmission [10]. Long-term follow-up of our patients could provide valuable insights into the quality of life, need for tracheal or PA reinterventions, and survival of young PAS patients as they enter teenagerhood.

Conclusions

Our case reports illustrate the diverse anatomical presentations of anomalous LPA, including a unique case of LPA coursing anterior to the trachea without true sling formation. Cross-sectional CT imaging and bronchoscopy are critical in defining the presence of rings and airway anatomy. Despite findings of airway narrowing or compression, none of our patients underwent bronchoplasty or tracheoplasty at the time of LPA repair to achieve excellent outcomes and relief of respiratory symptoms. Further studies with larger cohorts and long-term evaluations are needed to inform surgical decision-making and optimize care for patients with anomalous LPA.

Abbreviations

CPB	Cardiopulmonary bypass
CT	Computed tomography
LPA	Left pulmonary artery
MPA	Main pulmonary artery
PAS	Pulmonary artery sling
PDA	Patent ductus arteriosus
RPA	Right pulmonary artery

Author contributions
All authors' read and approved the final manuscript. YZ participated in the design and drafting of the manuscript. OM participated in the design and drafting of the manuscript. JAR participated in the design, drafting, and editing of the manuscript. RDS was responsible for patient care and participated in editing of the manuscript. CRP was responsible for patient care and participated in editing of the manuscript. KR was responsible for patient care, obtaining relevant images, and editing of the manuscript. TK was the senior author and was responsible for patient care, design, editing, and oversight of the manuscript.

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Availability of data and materials
No datasets were generated or analysed during the current study.

Declarations

Consent for publication
Verbal and written consent for participation and publication of the cases and any accompanying images was granted by each patient's respective parent/guardian.

Competing interests
The authors declare no competing interests.

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