CASE REPORT

Pulmonary Artery Sling with Bridge Bronchus in a Premature Infant: A Diagnostic and Management Challenge in Malaysia

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ABSTRACT

Pulmonary artery sling with bridge bronchus is a rare congenital vascular anomaly presenting significant diagnostic and management challenges, especially in resource-limited settings. This report details the case of a premature infant born at 34 weeks' gestation, diagnosed with PA sling and bridge bronchus, compounded by severe tracheal stenosis and bronchopulmonary dysplasia. Despite intensive medical care, including ventilation and permissive hypercarbia, the infant remained critically ill, with recurrent desaturation episodes and hospital-acquired infections. Definitive surgical intervention was deferred due to low birth weight and resource limitations. The infant succumbed to death at day 50 of life. This case emphasizes the complex interplay of prematurity, congenital anomalies, and healthcare limitations in developing countries. Multidisciplinary approaches and targeted research are essential for improving outcomes in such challenging cases.
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INTRODUCTION

Pulmonary artery (PA) sling is a rare congenital vascular anomaly classified as a type of vascular ring (VR), frequently associated with tracheobronchial stenosis (1). The association with bridge bronchus is a more rare and complex condition. This anomaly involves the middle and lower lobes of the right bronchus originating from the left main bronchus(2). A PA sling is characterized by an aberrant left PA arising from the posterior segment of the right PA, passing between the trachea and oesophagus to supply the left lung(1). Approximately 50%- 65% of cases present with complete tracheal rings, forming the "ring-sling complex". Clinically, PA sling with "ring-sling complex" manifests with significant respiratory distress, often severe enough to cause sudden death(2). This case report highlights the diagnostic and management challenges encountered ina developing country with centralised congenital cardiac surgery at the National Heart Institute (IJN), and constrained by limited surgical expertise, logistical hurdles, and prioritisation of critical surgical cases.

Case Presentation

A female premature infant was born to a 42-year-old mother with a prolonged history of subfertility. The pregnancy was complicated by gestational diabetes mellitus and preterm premature rupture of membranes with inadequate antenatal corticosteroid coverage. The infant was delivered at 34 weeks of gestation via emergency lower-segment caesarean section due to breech presentation and anhydramnios. A gestational age assessment score corresponds to 34 weeks of gestation. She required immediate positive pressure ventilation for poor respiratory effort.

Postnatally, she was stable under low-setting ventilation with no hemodynamic compromise. Examination showed a temperature of 36.8°C, pulse rate of 150 beats per minute, respiratory rate of 45 breaths per minute, and blood pressure of 55/40 mmHg. Initial chest radiography revealed a ground-glass appearance, and she was successfully extubated to non-invasive ventilation (NIV) on day two of life. Upon cardiac auscultation on day seven of life, a grade 3/6 pansystolic murmur was detected, and echocardiography confirmed peri membranous ventricular septal defect (VSD) with preserved ventricular function (Figure 1a and 1b).

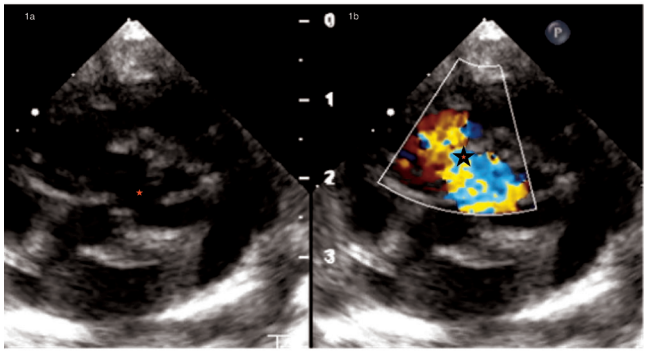


Figure 1a and 1b: Parasternal short axis view of echocardiography revealed the presence of peri membranous ventricular septal defect (red asterisk) measuring 3mm with left to right shunt

She remained stable under NIV and developed prematurity-related complications, including NIV dependency which progressed to bronchopulmonary dysplasia, rickets of prematurity and apnoea of prematurity. Nevertheless, she required reintubation on day 40 of life due to severe respiratory distress and recurrent desaturation episodes. During this time, she was tachypnoeic with respiratory rate of 70 breaths per minute, there was presence of moderate subcostal and intercostal chest recession with desaturation down to 70% under NIV. Lung finding revealed inspiratory stridor and expiratory wheezing. Following reintubation, the infant required high-frequency oscillatory ventilation and elevated positive end-expiratory pressure (PEEP) settings to address persistent hypercarbia, with arterial blood gas analysis revealing partial pressure of carbon dioxide (PaCO2) levels between 90 and 120 mmHg. Repeated echocardiographic evaluation of the previously identified small subaortic VSD confirmed its presence as a minor defect without other structural anomalies, alongside good biventricular function. A follow-up chest radiograph revealed an atypical low T-shaped carina (Figure 2), raising concerns about potential airway obstruction.

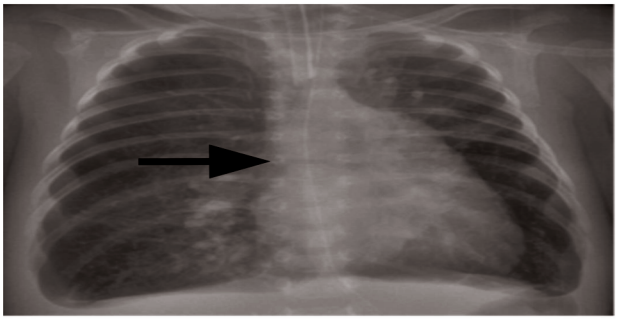


Figure 2: An anteroposterior chest radiograph revealed a low-shaped T carina (black arrow) , suggestive of a pseudo carina at the T7-T8 vertebra level, accompanied by horizontally branching bronchi

Contrast enhanced computed tomography (CT) of the thorax (Figure 3a) and volume-rendered multiplanar reconstruction images (Figure 3b and 3c) were performed on day 45 of life which was confirmed a type IIA left PA sling, tracheal stenosis, and bridge bronchus. A retrospective review of previous echocardiographic studies revealed that the PA branches were non-confluent, with the left PA originating from the right PA which had been missed during the initial assessment (Figure 4a and 4b). Although the infant was referred for definitive surgical interventions, including left pulmonary artery reimplantation and tracheobronchoplasty, the procedure was postponed due to her low weight of 2100 g, which was below the required threshold of 3000 g. Despite the implementation of an aggressive ventilation strategy, the infant remained in critical condition, with persistent hypercarbia and ongoing challenges in reducing ventilator support. Additionally, she experienced recurrent hospital-acquired infections, further exacerbating her already fragile state. Ultimately, she succumbed to death at day 50 of life, due to a combination of complications arising from her congenital anomalies, prematurity, and infections.

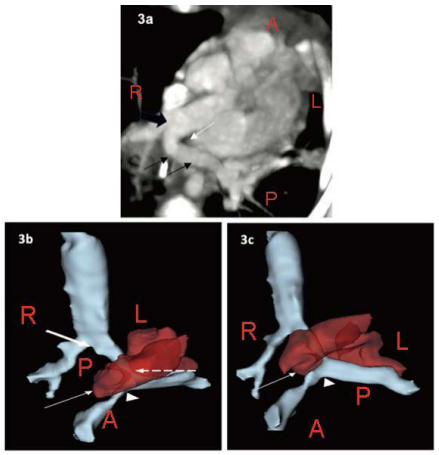
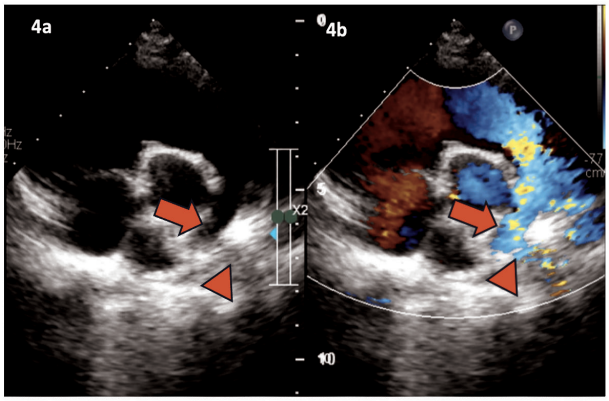


Figure 3a: An axial reformatted contrast-enhanced CT scan at the level of left pulmonary artery showing left pulmonary arterial sling (thin black arrows) arising from right pulmonary artery (thick black arrow). It compresses the left main bronchus (white arrow). Figure 3b and 3c is a volume-rendered multiplanar reconstruction images showing left pulmonary artery sling (thin arrow) partially encircle the markedly narrowed left main bronchus (dashed arrow). The true carina (thick arrow) is at its normal location, however, the pseudo carina (arrowhead) which gives rise into right lower lobe bronchus (bridging bronchus) is located at a lower position. A (anterior);P (posterior);R(right);L(left)



Figures 4a and 4b: Show a parasternal short axis view of echocardiography demonstrating that the pulmonary artery branches are non-confluent. The left pulmonary artery (indicated by a red arrowhead) arises from the right pulmonary artery (marked by a red arrow).

Discussion

PA sling is a variant of VR in paediatrics, leading to partial compression of the trachea and/or oesophagus. First identified in 1989, PA sling is characterized by the abnormal course of the left PA, which partially encircles the trachea, forming a "sling" shape(3). This condition accounts for one to three percent of congenital heart defects, with a true prevalence of approximately 1 in 17,000 school-aged children(4). PA sling is frequently associated with other congenital cardiac abnormalities and tracheobronchial stenosis but rarely associated with bridged bronchus.

The bridge bronchus anomaly, first described by Gonzales-Crussi in 1976, occurs when the right bronchus arises abnormally from the left main bronchus(5). This bridging bronchus typically traverses the mediastinum to connect to the right lung, forming a pseudo carina. The condition originates during the fifth week of gestation and is frequently associated with high mortality, particularly when there is significant tracheobronchial stenosis. Tracheobronchial stenosis associated with PA sling is a rare cause of airway obstruction in neonates and infants, resulting from complete cartilage rings encircled by the PA sling(1). Tracheomalacia, due to compression from the vascular ring, further increases the risk of mortality. The prognosis in this patient is poor, primarily due to low body weight, severe clinical manifestations, and the concurrent “ring-sling” complex.

In this case, the clinical manifestations and chest radiograph findings indicative of airway stenosis became apparent only on day 40 of life. This suggests that the PA sling causes a gradual compression of the airway, a phenomenon supported by the literature, which notes that most cases of PA sling present during infancy due to the chronic progression of the disease. The patient exhibited classic signs of tracheal stenosis, including inspiratory stridor, expiratory wheezing, severe cyanosis, and recurrent chest infections(2).

Diagnostic evaluation typically begins with echocardiography, which may reveal the absence of the usual pulmonary artery bifurcation. Chest radiograph showing a low T-shaped carina often prompt further imaging, such as CT or magnetic resonance imaging (MRI) (1).

CT imaging was pivotal for diagnosis. Although CT involves radiation exposure, it remains the best modality for anatomical imaging in this setting due to its rapid acquisition time and non-invasive nature. MRI was not feasible due to prolonged scan time and sedation needs. Bronchoscopy provides dynamic airway information but was not performed due to clinical instability. Bronchography is theoretically useful but its safety in premature infants is questionable (2).

In 1988, Wells classified PA sling into two types, further subdivided into subtypes A and B depending on whether a bronchus supplies the right upper lobe. Type II, as observed in this case, is marked by an infracranial sling at the T7 to T8 vertebral level and is often accompanied by airway obstruction that necessitates intervention(1). The management of PA sling and associated airway anomalies requires personalized surgical planning, typically involving both tracheal and vascular reconstruction. However, for asymptomatic patients, a conservative approach is preferred. There is no definitive consensus on managing the "ring-sling" complex anomaly. A diameter/length ratio (DLR) of less than 5.9 is a reliable indicator for tracheal surgery. DLR is defined as the ratio of the narrowest diameter of the stenotic segment to the total tracheal length, measured via CT (4). In this case, the DLR was not measured because the CT scan covered only up to the C7 vertebral level. Measuring the DLR requires imaging of the entire trachea, which extends up to the C4 vertebral level.

Although airway surgery was considered, there was uncertainty about its necessity. Literature suggests that in some cases, bridging bronchus anomalies may not require immediate surgical correction. Off-pump left pulmonary artery reimplantation could have been considered to alleviate airway compression, potentially deferring tracheoplasty until the infant grew larger (5). However, such procedures demand high surgical expertise, which is limited in our setting. Moreover, the CT scan did not include the entire trachea, preventing the calculation of the diameter/length ratio (DLR), a key metric to assess the need for tracheal surgery. The scan field was limited due to the infant's critical state and sedation risk, and only reached the C7 level.

Surgical management typically involves left PA reimplantation and tracheoplasty under cardiopulmonary bypass (CPB). Off-pump surgery is technically demanding but could reduce CPB-related risks in premature infants (5). In Malaysia, CPB is generally required due to limited expertise in off-pump neonatal procedures. Early surgery

was considered, but due to the patient's low weight (2100 g), the decision was made to defer intervention until reaching the institutional threshold of 3000 g. A palliative approach was adopted in the interim, though the prognosis remained poor.

To date, this is the first reported case of a PA sling with a bridge bronchus in a premature infant, highlighting the rarity of such a complex condition in the neonatal population, particularly in preterm infants.

Conclusion

This case report highlights the diagnostic and management challenges of PA sling and associated airway anomalies in premature infants. Early intervention remains constrained by clinical fragility, prematurity, and resource limitations. Multidisciplinary collaboration is crucial for optimal outcomes, and further research is needed to refine management protocols for this rare and severe condition.

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