

# IMAGES in PAEDIATRIC CARDIOLOGY

Micallef SP, Grech V, Sammut P, DeGiovanni JV. 3 year old with chronic wet cough: Intralobar Bronchopulmonary sequestration. *Images Paediatr Cardiol* 2016;18(2):5-10.  
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## Introduction

Bronchopulmonary sequestration (BPS) is a rare congenital foregut anomaly of the lung, consisting of nonfunctioning primitive tissue that does not communicate with the healthy tracheobronchial tree. The abnormal segment has its own anomalous systemic blood supply, and typically drains into the pulmonary venous system.

It is frequently asymptomatic and may be discovered incidentally. Symptoms may vary, with patients presenting with vague symptoms related to chronic respiratory infection. Early diagnosis and intervention decreases morbidity and mortality.<sup>3,4</sup>

We present a case of intralobar pulmonary sequestration involving the right lower lobe in a male child.

## Case presentation

A three year old male, known case of mild aortic stenosis, was referred to the paediatric respiratory physician with a seven month history of chronic productive cough and recurrent wheeze. The cough was resistant to regular inhaled corticosteroids and salbutamol. Physical examination was unremarkable (longstanding positive finding of a systolic murmur), with no respiratory compromise.

Of note, in the past history, the child had been admitted to the Neonatal Intensive Care Unit at 16 hours of age with mild respiratory distress, which settled after minimal support and intravenous antibiotics.

At three years of age, there was one presentation to hospital with wheezing, resolving on nebulised salbutamol and oral antibiotics. A plain chest radiograph at the time reported “signs of right sided basal pneumonia associated with a small right sided pleural effusion” (Figure 1).

**Figure 1: Plain chest radiograph showing blunting of cardiophrenic and costophrenic angles of the right lung.**



A repeat chest radiograph requested by the respiratory physician showed persistence of the above-mentioned abnormalities. A high resolution contrast enhanced computed tomography (HRCT) scan of the chest was performed, showing an elevated right hemi-diaphragm and a smaller right lung. The right basal lung parenchyma received its blood supply from an anomalous artery arising from the abdominal aorta at the level of the coeliac axis. The veins of the affected lobe drained directly into the pulmonary veins, implying a right-to-left shunt.

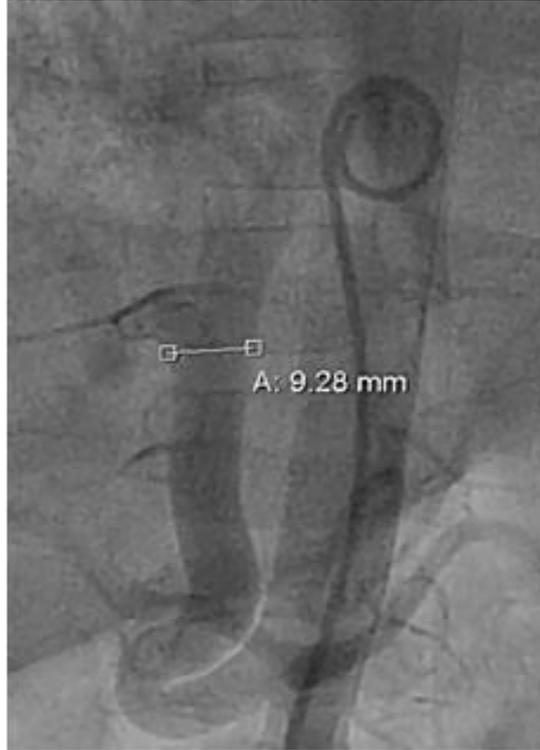
The lobe involved appeared inseparable from surrounding normal pulmonary parenchyma with a discernible reduction in the number of bronchi within the involved lung. The findings were suggestive of a diagnosis of intralobar pulmonary sequestration (Figure 2).

**Figure 2: CT showing anomalous artery**

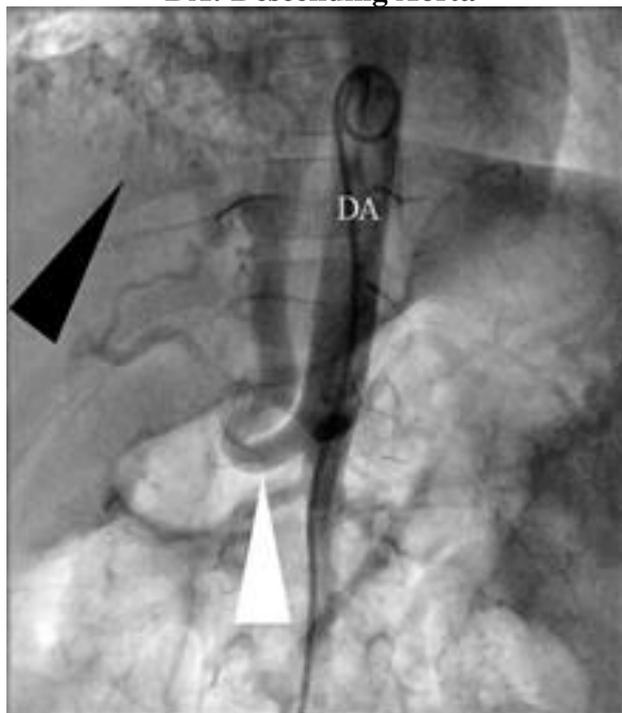


The patient was referred to the paediatric cardiology team for further management. A diagnostic and therapeutic cardiac catheterisation was performed which confirmed the presence of the anomalous vessel, measuring 9mm in diameter, arising close to the coeliac axis (Figures 3,4). This vessel was embolised successfully using a 10 by 8 Amplatzer duct occluder device (Figure 5).

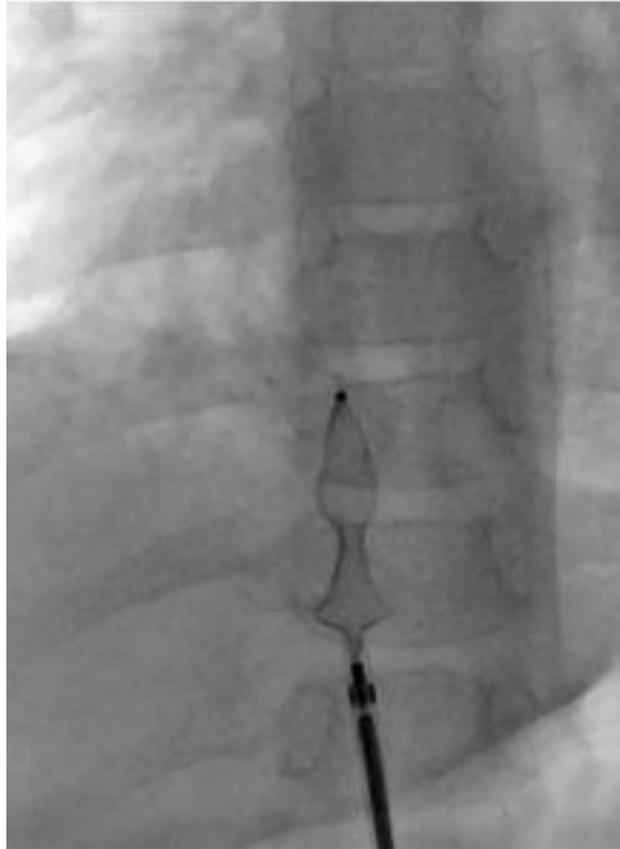
**Figure 3: Angiogram showing aberrant artery measuring approx 9mm**



**Figure 4: Angiogram showing aberrant artery (white) supplying sequestered lobe (black).  
DA: Descending Aorta**



**Figure 5: Device deployed in aberrant vessel supplying sequestered lobe**



The patient had an uncomplicated clinical course, with his symptoms having improved significantly. He continues to receive regular outpatient follow-up, and remains well to date.

### **Discussion**

Intralobar pulmonary sequestration is a rare congenital anomaly, being responsible for 0.15-6.4% of all congenital pulmonary malformations.<sup>1,2</sup> Asymptomatic patients can present with incidental pulmonary lesions on plain chest radiography. Ultrasound may show a hyperechoic chest mass with Doppler ultrasound indicating flow through arteries and veins.<sup>3</sup> Recurrent pneumonia or pleural effusions may render patients symptomatic.

There are two predominant types of BPS: extralobar (ELS), where sequestered lung lies outside the visceral pleura and can be invested in its own external layer, and intralobar (ILS), where abnormal bronchopulmonary tissue shares common visceral pleura with neighbouring lung tissue (Table 1). The final diagnosis is reached via histological examination of resected tissue.<sup>4</sup> Both intrapulmonary and extrapulmonary BPS may be associated with other congenital anomalies.<sup>3</sup>

BPS usually presents at a young age, in particular ELS which can even present as antenatal fetal hydrops, although 10% of ELS and 15% of ILS are diagnosed in adulthood in view of paucity of symptoms.<sup>2,3</sup>

**Table 1. Comparison of intralobar and extralobar pulmonary sequestration**

<b>Pulmonary Sequestration</b>	<b>Extralobar</b>	<b>Intralobar</b>
<b>Age of onset</b>	61% in <6 months; 10% asymptomatic	Rare before 2 years of age unless diagnosed antenatally, 20% by 20 years, 15% asymptomatic
<b>Gender (male:female)</b>	3-4:1	1:1
<b>Location</b>	Lower lobe: 63% Other: intra-abdominal, Mediastinum	Lower lobe: 98%
<b>Affected lung</b>	65% Left	55% Left
<b>Associated anomalies</b>	65%	11%
<b>Symptoms</b>	Respiratory distress Feeding difficulties Fetal hydrops Congestive cardiac failure	Recurrent infections (frequent) Pneumothorax, Haemothorax Haemoptysis Congestive cardiac failure

Diagnosing pulmonary sequestration is fraught with difficulty, depending on the location and type of pathology and the presence or absence of symptoms. Contrast enhanced HRCT is usually sufficient to provide a diagnosis, as exemplified in this case report, giving detailed anatomical information, clearly demonstrating pertinent anatomy. The gold standard for diagnosing pulmonary sequestration is CT angiography, delineating arterial supply and venous drainage, with the benefit of being less invasive than conventional angiography. This modality allows for safer surgical planning with more detailed visualisation of vascularisation of sequestered tissue.<sup>4,5,6,7</sup>

BPS can be diagnosed on prenatal ultrasound, although it may be difficult to differentiate from other lung abnormalities such as congenital cystic adenomatoid malformation, bronchogenic cysts, enteric duplication cysts and neuroblastoma.<sup>8</sup> A precise antenatal diagnosis can only be achieved if a systemic feeding artery is visualised.<sup>8</sup>

There have been varying clinical techniques used to manage this pathology. BPS can be managed conservatively or operatively, depending on the clinical setting. Many authors advocate no intervention in the case of asymptomatic patients as such lesions rarely cause clinical problems.<sup>3,8</sup>

In symptomatic patients, anomalous lung may be removed surgically or embolised with endovascular intervention. With regards to surgery, sequestrectomy or pulmonary lobectomy can be performed.<sup>2,5,7,9</sup> Lobectomy is preferred when sequestered tissue cannot be distinguished from functioning parenchyma. Sequestrectomy allows the preservation of as much lung tissue as possible.<sup>2,10</sup> In children, surgery may be associated with a high risk of skeletal and muscular deformities such as asymmetry of the chest wall, scoliosis caused by rib fusion or scapular winging.<sup>3,4</sup> Endovascular intervention may therefore be preferred to avoid the morbidity associated with surgery.<sup>4,11,12</sup> Embolisation, however, carries a significant risk of subsequent pulmonary infections.<sup>4</sup>

In conclusion, there is a wide range of manifestations associated with pulmonary sequestration, ranging from complete asymptotology to those reporting various cardiac or pulmonary complaints. Diagnostic imaging must be dependent on the clinical scenario but must delineate

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vascularity, which will guide subsequent treatment. In this case report, a child with chronic cough had pulmonary sequestration involving the right lower lobe, with early diagnosis and prompt treatment leading to resolution of symptoms, which should be the standard of care in all patients with this pathology.

## References

1. Savic B, Bertel FJ, Tholen W, et al. Lung sequestration: report of seven cases and review of 540 published cases. *Thorax* 1979;34:96-101.
2. Andrade CF, Paiva da Costa Ferreira H, Bueno Fischer G. Congenital lung malformations. *J Bras Pneumol.* 2011;37(2):259-271
3. Corbett HJ & Humphrey GM. Pulmonary sequestration. *Paediatr Respir Rev.* 2014; 5(1):59-68
4. Marine LM, Valdes FE, Mertens RM, Bergoeing MR, Kramer A. Endovascular treatment of symptomatic pulmonary sequestration. *Annals of Vascular Surgery* 2011; 25(5):11-15
5. Gezer S, Tastepe I, Sirmali M, Findik G, Turut H, Kaya S, et al. Pulmonary sequestration: a single-institutional series composed of 27 cases. *J Thorac Cardiovasc Surg.* 2007;133(4):955-9.
6. Jesch NK, Leonhardt J, Sumpelmann R, Gluer S, Nustede R, Ure BM. Thoracoscopic resection of intra- and extralobar pulmonary sequestration in the first 3 months of life. *J Pediatr Surg.* 2005;40(9):1404-6.
7. Azizkhan RG, Crombleholme TM. Congenital cystic lung disease: contemporary antenatal and postnatal management. *Pediatr Surg Int.* 2008;24(6):643-57.
8. Adzick NS, Harrison MR, Crombleholme TM et al. Fetal lung lesions: management and outcome. *Am J Obstet Gynecol* 1998; 179:884–889.
9. Shanmugam G, MacArthur K, Pollock JC. Congenital lung malformations, antenatal and postnatal evaluation and management. *Eur J Cardiothorac Surg.* 2005;27(1):45-52.
10. Hertenberg C, Daon E, Kramer J. Intralobar pulmonary sequestration in adults: three case reports. *J Thorac Dis* 2012;4(5):516-519
11. Lee KH, Sung KB, Yoon HK, et al. Transcatheter arterial embolization of pulmonary sequestration in neonates: long-term follow-up results. *J Vasc Interv Radiol* 2003;14: 363-367.
12. Nayar PM, Thakral CL, Sajwani MJ. Congenital lobar emphysema and sequestration treatment by embolization. *Pediatr Surg Int* 2005;21:727-729

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