

Antenatal Diagnosis of Extralobar Pulmonary Sequestration And its Early Perinatal Surgical Management

DAMINI NAGPAL¹, BHAGYAJYOTHI B KHANAGAVI², SANTOSH BABURAO KURBET³, MANOJ TOGALE⁴

¹Dept of General Surgery, J. N. Medical College, KLE Academy of Higher Education and Research, Deemed to be University, Belagavi, Karnataka, India-590010

²M.S Associate Professor, Dept of Ophthalmology, KAHER's J N Medical College, Belagavi

³Dept of Paediatric Surgery, J. N. Medical College, Belagavi KLE Academy of Higher Education and Research, Deemed to be University, Belagavi, Karnataka, India-590010

⁴Dept of General Surgery, J. N. Medical College, KLE Academy of Higher Education and Research, Deemed to be University, Belagavi, Karnataka, India-590010

*Corresponding Author

Article History

Received: 01.10.2025

Revised: 14.10.2025

Accepted: 10.11.2025

Published: 11.11.2025

Abstract:

Pulmonary sequestration is an uncommon congenital lung malformation characterized by non-functioning pulmonary tissue with systemic arterial supply and absent tracheobronchial communication. It may present as intralobar or extralobar sequestration, with intralobar type being more frequent. We report the case of a female neonate diagnosed antenatally at 21 weeks of gestation with a wedge-shaped echogenic intra-thoracic lesion consistent with pulmonary sequestration. The pregnancy was otherwise uneventful, and the infant was delivered at term via Caesarean section due to meconium-stained liquor. Postnatal CT thorax confirmed extralobar pulmonary sequestration in the left lower lobe with arterial supply from the thoracic aorta and mild bilateral pneumothorax. The neonate underwent left thoracotomy and excision of the sequestered segment on day four of life. Histopathology confirmed the diagnosis. The postoperative course was uneventful, and the baby was discharged on exclusive breastfeeding with regular follow-up. This case highlights the importance of antenatal detection, multidisciplinary planning, and early surgical management to optimize neonatal outcomes in pulmonary sequestration.

Keywords: Pulmonary sequestration, tracheobronchial communication, thoracic aorta.

INTRODUCTION

Congenital pulmonary malformations are rare developmental anomalies of the lower respiratory tract that may present variably, ranging from incidental radiological findings to life-threatening neonatal respiratory compromise (1). Among these anomalies, pulmonary sequestration represents a distinct entity characterized by non-functioning lung tissue that lacks communication with the native tracheobronchial tree and derives its blood supply from aberrant systemic arteries, most frequently from the thoracic or abdominal aorta. The venous drainage may be into the pulmonary veins, azygos system, or directly into the inferior vena cava, depending on the type of sequestration (2).

Pulmonary sequestration is traditionally classified into intralobar sequestration (ILS) and extralobar sequestration (ELS). ILS, which accounts for approximately 75–85% of cases, is enclosed within the visceral pleura of the normal lung and usually drains into the pulmonary venous system. In contrast, ELS has its own pleural covering, often drains into systemic veins, and may be associated with other congenital anomalies such as diaphragmatic hernia or congenital heart disease, and a distinct aetiology (3). Clinically, ILS is more often diagnosed later in childhood or adolescence, typically after recurrent pulmonary infections, whereas ELS tends to present earlier due to mass effect or associated anomalies (4).

The reported incidence of pulmonary sequestration is between 0.15% and 6.4% of all congenital pulmonary malformations, with a slight male predominance in extralobar types (5). Although uncommon, the true incidence is likely underestimated because some cases remain asymptomatic and undiagnosed until adulthood. With the widespread use of antenatal ultrasonography and foetal anomaly scans, prenatal diagnosis of pulmonary sequestration has become increasingly feasible (6). High-resolution ultrasonography often demonstrates an echogenic, homogeneous intrathoracic mass, while colour doppler aids in identifying the characteristic systemic feeding vessel, which is the hallmark feature distinguishing sequestration from other congenital lung lesions such as congenital pulmonary airway malformation (CPAM), congenital lobar emphysema, and bronchogenic cyst (7).

The clinical presentation of pulmonary sequestration in the neonatal period is unusual. When symptomatic, neonates may present with respiratory distress, recurrent pneumonia, hemoptysis, hemothorax, or features of high-output cardiac failure secondary to large systemic shunts (8). However, many cases remain stable initially and are identified only through prenatal or incidental postnatal imaging. Management typically involves surgical resection, which is considered curative and prevents long-term complications such as recurrent infections, abscess formation, or rarely malignant transformation (5).

In recent years, thoracoscopic approaches have gained popularity in older children and adults; however, in neonates and young infants, thoracotomy remains the traditional approach because of limited operative space and the need for meticulous dissection of aberrant systemic vessels. The timing of surgery is individualized based on clinical presentation, lesion size, and presence of complications (9).

Here, we report a rare case of antenatally diagnosed extralobar pulmonary sequestration that was confirmed postnatally and managed with thoracotomy and resection in the early neonatal period. This case underscores the importance of antenatal diagnosis, multidisciplinary coordination, and timely surgical intervention in optimizing neonatal outcomes.

RESULTS AND OBSERVATIONS:

Antenatal Findings

A 25-year-old primigravida underwent an anomaly scan at 21 weeks of gestation, which revealed a wedge-shaped, pleural-based echogenic lesion in the left hemithorax, measuring 20×16 mm. Colour Doppler demonstrated a systemic feeding vessel arising from the thoracic aorta, consistent with pulmonary sequestration (Figure 1). A follow-up ultrasonography at 29 weeks confirmed persistence of the lesion, now measuring $2.7 \times 2.9 \times 2.6$ cm with internal vascularity. There were no signs of hydrops foetalis, and foetal growth was appropriate for gestational age.



FIG 1: Antenatal ultrasound (21 weeks) showing echogenic intra-thoracic lesion in the left lung with systemic vessel from thoracic aorta.

Perinatal and Neonatal Course

The baby was delivered at 40 weeks and 1 day by lower-segment Cesarean section due to meconium-stained liquor. Birth weight was 2800 g, and Apgar scores were 7, 8, and 10 at one, five, and ten minutes, respectively. The neonate cried immediately and was admitted to the NICU for observation. On admission, she was hemodynamically stable with normal cry, activity, and no signs of respiratory distress.

Investigations

- **Baseline laboratory tests:** Hemoglobin 14.6 g/dL, WBC $14,400/\text{mm}^3$, platelets $2.62 \times 10^5/\text{mm}^3$, electrolytes within normal limits, and total bilirubin 5.18 mg/dL.
- **Imaging:**

CECT thorax (Day 1): Revealed a well defined hyperdense lesion in the posterobasal segment of the left lower lobe, measuring $2.8 \times 2.8 \times 1.9$ cm, with arterial supply directly from of the thoracic aorta at T9–T10 level (Figure 2). Venous drainage was into the inferior vena cava. Associated findings included mild bilateral pneumothorax and reduced left lung volume.

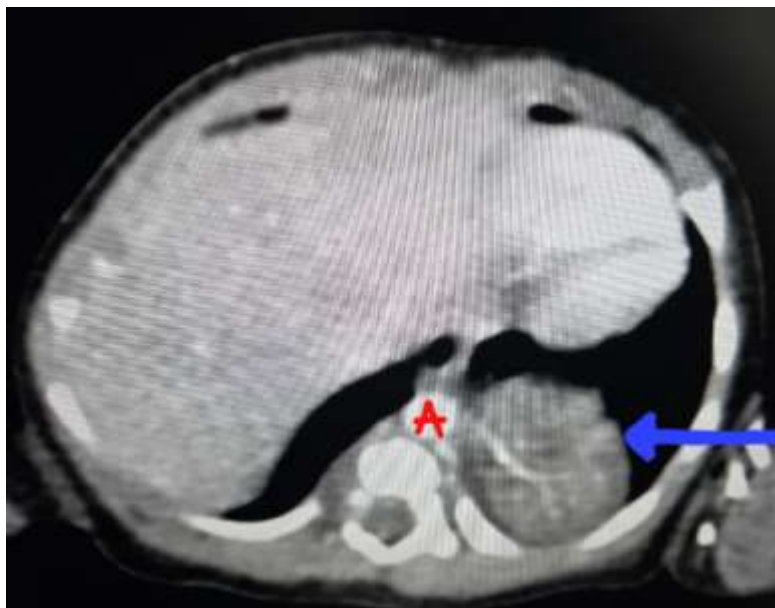


FIG 2: CT thorax (axial section) showing arterial supply to sequestered lung tissue directly from thoracic aorta (A = aorta, arrow = feeding artery).

Treatment and NICU Management

The neonate was stabilized in the NICU and managed conservatively with intravenous fluids and broad-spectrum antibiotics (amikacin and cefotaxime) for four days. Supportive care included close monitoring, oxygenation, and gradual feeding initiation.

Surgical Intervention

On day 4 of life, the infant underwent a **left posterolateral thoracotomy**. Intraoperatively, an abnormal segment of the left lower lobe supplied by a large aberrant systemic artery was identified, ligated and cut. The sequestered segment was carefully dissected and excised (FIG 3).



FIG 3 : Excised sequestered lobe of lung in the left lower chest.

Histopathology

The excised specimen measured $4 \times 2 \times 1.5$ cm. Microscopy showed pleura-covered lung tissue containing cystic spaces lined by ciliated columnar epithelium and dilated alveoli, consistent with **extralobar pulmonary sequestration**.

Postoperative Outcome

The neonate tolerated the procedure well. Oral feeds were initiated and accepted without difficulty. She gained weight (discharge weight: 2820 g) and remained clinically stable. The baby was discharged on exclusive breastfeeding with advice for regular pediatric surgery follow-up. The infant is doing well with good weight gain and no respiratory issues at 2 mths follow up period (FIG 4)



FIG 4 : Follow up photograph at 2 months post surgery

DISCUSSION

Pulmonary sequestration is a rare congenital malformation in which non-functioning lung tissue receives direct systemic arterial supply without communication to the native tracheobronchial tree(1,2). It accounts for less than 6% of congenital lung anomalies, but its clinical importance lies in its association with recurrent infections, respiratory compromise, and, rarely, malignant transformation(1,2). The aetiology of these lesions is unclear, with theories explained by Rokitansky's fraction theory as early as 1861 explains a normal lung tissue development which later becomes non-functional and recent theory suggesting the chronology of development at a pseudoglandular phase of pulmonary growth between 5-17 weeks of gestation also explaining its foregut communication in some cases (3). Two forms exist: intralobar sequestration, which shares pleura with normal lung and usually presents later in childhood, and extralobar sequestration, which has a separate pleural covering and often presents earlier. Neonatal presentation of extralobar sequestration, as in this case, is uncommon(11).

M:F ratio is 1.6:1, cough, fever, hemoptysis are the common modes of presentation. Radiological imaging, starting with use of doppler studies and MRI in antenatal period and post-natal use of CT usually clinch the diagnosis. Rt side sequestrations have foregut communication as high as in 15-20%, and may warrant use of contrast studies to demonstrate GI communication. Associated anomalies were seen in 50% of cases, which included CDH, CHD, ectopia

cordis, pericardial defects, vertebral defects in ELPS cases (12). A study by Wei et.al concluded that early age of onset of symptoms, recurrent respiratory infections, mass or cystic lesion located in lower chest, and aberrant systemic blood supply are indicators of pulmonary sequestration.(12) The increasing use of antenatal ultrasonography has improved the recognition of pulmonary sequestration. A homogenous echogenic mass with systemic arterial supply on Doppler is highly suggestive, as seen in our patient at 21 weeks of gestation. Postnatal CT imaging is essential for confirming the diagnosis and mapping vascular anatomy, which is critical for safe surgery. In our case, CT confirmed the left lower lobe lesion with systemic arterial supply from the thoracic aorta, allowing precise operative planning.

Infants who were less than 6 months had poor intra-operative tolerance of single lung ventilation during resection (13). In addition, the probability of infection increases over one year of age, leading to more perioperative complications after surgery beyond this age group. Surgical resection remains the standard treatment to prevent infection, hemorrhage, or progressive lung disease. However intra-arterial catheterisation and embolization is being done in older children(14). While some advocate delayed surgery in asymptomatic infants, early intervention is considered safe and effective when the lesion is large or vascular needing early surgery. Our patient underwent thoracotomy on day four of life with an uneventful recovery. Thoracoscopy is now increasingly used in older children, but thoracotomy has been traditionally

used so far in neonates due to limited space and the need for meticulous vessel control (15).

Histopathology typically demonstrates lung tissue with cystic changes lined by respiratory epithelium, confirming sequestration, as seen in this case. Compared with published literature, where most intralobar sequestrations are diagnosed later in childhood, this report adds value by documenting antenatal detection and successful neonatal surgical management. The outcome underscores the importance of early diagnosis, detailed imaging, and multidisciplinary care in ensuring excellent prognosis.

CONCLUSION

This case highlights the value of antenatal ultrasonography in identifying pulmonary sequestration and guiding perinatal management. Early neonatal surgical intervention can be performed safely, ensuring excellent outcomes. A multidisciplinary approach involving obstetrics, neonatology, radiology and pediatric surgery is crucial in the management of such rare congenital lung anomalies.

REFERENCES

1. Gen O, Gurkok S, Dakak M, Gozubuyuk A, Ozkan M, Caylak H. Pulmonary sequestration and surgical treatment. *Asian Cardiovasc Thorac Ann*, 2006, vol14,3-6.
2. Gabelloni M, Faggioni L, Accogli S, Aringhieri G, Neri E. Pulmonary sequestration: What the radiologist should know. *Clin Imaging*. 2021;73:61–72. doi:10.1016/j.clinimag.2020.12.011
3. Bratu I, Flageole H, Chen MF, Di Lorenzo M, Yazbeck S, Laberge JM. The multiple facets of pulmonary sequestration. *J Pediatr surg*. 2001, 36:784-90.
4. Dhanju G, Goubran A, Kirkpatrick I, Wiebe S, Fogel J. Antenatal diagnosis of bronchopulmonary sequestration: A case report and review of the literature. *Radiol Case Rep*. 2023;19(2):604–9. doi:10.1016/j.radcr. 2023.01.010
5. Torres LFB, Jacob GVV, Noronha L de, Seade M. Congenital pulmonary extralobar intra-abdominal sequestration. *J Pediatr (Rio J)*. 1997;73(1):45–8. doi:10.2223/JPED.559
6. Wong KKY, Flake AW, Tibboel D, Rottier RJ, Tam PKH. Congenital pulmonary airway malformation: advances and controversies. *Lancet Child Adolesc Health*. 2018;2(4):290–7. doi:10.1016/S2352-4642(18)30035-X
7. Gautam MS, Naren Satya Srinivas M, Prathyusha IS, Hema Chandra Reddy K, Mayilvaganan KR, Raidu D. An Atypical Presentation of Congenital Pulmonary Airway Malformation (CPAM): A Rare Case with Antenatal Ultrasound Findings and Review of Literature. *Pol J Radiol*. 2017;82:299–303. doi:10.12659/PJR.901217
8. Nagar R, Bütter A, Brahm G, Price A. Pulmonary sequestration causing severe cardiac failure requiring lobectomy in an extreme preterm infant. *J Pediatr Surg Case Rep*. 2015;3(10):415–8. doi:10.1016/j.epsc.2015.08.001
9. Bawazir O. Thoracoscopy in pediatrics: Surgical perspectives. *Ann Thorac Med*. 2019;14(4):239–45. doi:10.4103/atm.ATM_133_19
10. De Vreede I, Bilardo CM, Van Rijn RR, Clur SAB, Heij HA. Intrapericardial Extralobar Pulmonary Sequestration Presenting as a Prenatal Intrathoracic Mass. *Pediatr Cardiol*. 2008;29(5):980–2. doi:10.1007/s00246-007-9112-6
11. Kumbasar U, Uysal S, Doğan R. Congenital pulmonary malformations. *Türk Gogus Kalp Damar Cerrahisi Derg*. 2024;32(Suppl1):S60–7. doi:10.5606/tgkdc.dergisi.2024.S1.09
12. Wei Y, Li F. Pulmonary sequestration: a retrospective analysis of 2625 cases in China. *Eur J Cardiothorac Surg*. 2011 Jul;40(1):e39-42.
13. Wani SA, Mufti GN, Bhat NA, Baba AA: Pulmonary sequestration: early diagnosis and management. *Case Rep Pediatr*. 2015, 2015:454860. 10.1155/2015/454860
14. Lee K.H, Sung, K.B, Yoon H.K, Ko G.Y, Yoon C.H, Goo H.W, Kim E.A, Kim K.S, Pi S.Y. Transcatheter arterial embolization of pulmonary sequestration in neonates: long-term follow-up results. *J Vasc Interv Radiol* 2003, vol. 14. 363-367.
15. Wan I.Y, Lee T.W, Sihoe A.D., Ng C.S, Yim A.P. Video-assisted thoracic surgery lobectomy for pulmonary sequestration. *Ann Thorac Surg*, 2002 , vol.73. 639-640.