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Neonatal left lung agenesis presenting as pneumonia in a resource-limited setting: a case report

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Case Report

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Abstract

Background

Pulmonary agenesis is a rare congenital malformation that occurs due to a defect in lung bud development. It presents with respiratory distress and is often mistaken for common respiratory conditions, including pneumonia, particularly in resource-limited settings. This leads to delayed diagnosis and inappropriate care. Therefore, reporting such cases from resource-limited settings is important to enhance clinical awareness and improve outcomes.

Clinical Case

We present a term female neonate who experienced severe respiratory distress shortly after birth. Initially, the patient was treated for pneumonia at a health center. However, her condition worsened, and she was referred to a tertiary hospital.

At admission, the patient was in severe respiratory distress. Physical examination revealed severe intercostal and subcostal retractions as well as absent air entry on the left hemithorax. Chest radiography showed a complete homogeneous opacification of the left hemithorax with ipsilateral rib crowding; and hyperinflation of the contralateral lung. Contrast-enhanced chest tomography confirmed the absence of the left lung, left main bronchus and left pulmonary artery. Transthoracic echocardiography revealed enlargement of the right-sided heart chambers, a moderate patent ductus arteriosus with bidirectional shunting, and severe pulmonary hypertension. The patient was supported with continuous positive airway pressure (CPAP) and medical therapy for pulmonary hypertension and cardiac support. At 6 months of age, her clinical condition gradually improved but remained oxygen-dependent.

Conclusion

Clinicians should consider this rare congenital anomaly in children presenting with persistent respiratory distress, asymmetric chest findings and order appropriate imaging.

Background

Pulmonary agenesis is a rare congenital anomaly caused by failure of lung bud development(1, 2). Its incidence is about 1–2 per 10,000 live births (2). Bilateral agenesis is incompatible with life, whereas unilateral agenesis can present with variable severity (3). It is frequently associated with other congenital anomalies which significantly influence prognosis (1, 3).

Patients commonly present with respiratory distress, wheezing or asymmetric chest findings (3, 4). Others are diagnosed incidentally during evaluation for common conditions (5, 6).

Pulmonary agenesis has previously been described in some literature, but reports from resource-limited countries, particularly in sub-Saharan Africa, remain rare (1, 2). It is often misdiagnosed as common conditions such as pneumonia or sepsis (6, 7). To date, only one case of unilateral pulmonary agenesis has been reported from Ethiopia, diagnosed incidentally in adolescence (7). To our knowledge, this is the first reported neonatal case, highlighting the diagnostic challenges in resource-limited settings.

Clinical Case

A term female neonate was born to a 30-year-old gravida 3, para 3 mother by elective cesarean section due to two previous cesarean deliveries. Birth weight was 2.5 kg with an Apgar score of 5, 6, and 8 at 1^{st}, 5^{th}, and 10^{th} minutes, respectively. The mother had adequate antenatal follow-up, and serial prenatal ultrasonography did not detect fetal anomalies.

Immediately after delivery, the neonate developed respiratory distress and required bag-mask ventilation for five minutes before spontaneous respiration was established.
At one week of age, the neonate developed tachypnea and was admitted to primary health center. Laboratory evaluation revealed leukocytosis (WBC: 41.47 ×10^3/μL) with neutrophil predominance (Neutrophil: 80%) and elevated C-reactive protein (CRP: 147 mg/L). Chest radiography showed focal right lung opacities and treated as neonatal pneumonia.

However, the respiratory distress progressively worsened with increasing oxygen requirements. The neonate was referred to a tertiary care center for advanced respiratory support.

On admission to a tertiary center, the neonate was alert but in severe respiratory distress. On physical examination there was intercostal and subcostal retractions and absent air entry over the left hemithorax. Chest radiograph revealed complete opacification of the left hemithorax with ipsilateral rib crowding, mediastinal shift to the left and compensatory hyperinflation of the right lung as well as right middle and lower lobe opacity with air bronchogram **(Figure 1)**

Contrast-enhanced chest computed tomography (CT) confirmed absence of the left lung, left main bronchus, and left pulmonary artery (Figure 2)

Transthoracic echocardiography showed displacement of heart to left side, right atrial and ventricular dilation, moderate patent ductus arteriosus (PDA: 2.5 mm) with bidirectional shunting, and severe tricuspid regurgitation.

Based on these findings a diagnosis of left lung agenesis complicated by congenital heart disease and severe pulmonary hypertension was established. The patient was supported with continuous positive airway pressure (CPAP) and was started on sildenafil, furosemide, hydrochlorothiazide, and spironolactone.

Her condition gradually improved and discharged at two months of age on home oxygen (1 L/min via nasal cannula) and continued on medical management.

At six months of follow-up, the infant remained oxygen-dependent. Repeat echocardiography demonstrated dilatation of all cardiac chambers, enlargement of the main pulmonary artery and its branches, and progression to a large patent ductus arteriosus measuring 6–7 mm with bidirectional shunting.

Discussion

Unilateral pulmonary agenesis is a rare congenital anomaly characterized by complete absence of pulmonary parenchyma, bronchial structures, and pulmonary vasculature (1, 2). Left-side pulmonary agenesis, as observed in this patient, generally has a better prognosis than right-side, likely because of less severe mediastinal distortion and better compensatory capacity of the remaining lung (1, 3).

Clinical diagnosis of unilateral lung agenesis is often challenging because its presentation often overlaps with common neonatal respiratory conditions such as pneumonia, transient tachypnea, or respiratory distress syndrome (4, 5). Furthermore, the solitary functioning lung is predisposed to recurrent infections, which may further obscure the underlying congenital abnormality and delay recognition (4).

Our patient was treated as presumed pneumonia at a primary health center which masked the primary structural abnormality. Such diagnostic challenge is observed in many case reports particularly in low-resource settings where access to advanced imaging and specialist interpretation is limited (7).

Chest radiography remains an important initial diagnostic tool and may demonstrate unilateral hemithorax opacification, mediastinal shift, rib crowding, and compensatory hyperinflation of the contralateral lung (1, 8). However, these findings are not specific, and can be misinterpreted as common pulmonary pathology (6, 7). In infection-endemic regions, unilateral lung pathology on chest radiography is commonly interpreted as pneumonia, lung collapse, or post-infectious changes (9). This epidemiological context further reduces clinical suspicion of rare congenital anomalies (2).

In our patient, the initial laboratory findings were suggestive of infection. However, the radiological features indicated a more serious underlying pathology, with pneumonia unlikely to be the primary diagnosis. Pneumonia more commonly presents with patchy or lobar infiltrates without significant volume loss rather than complete opacification of hemithorax (10). Furthermore, the lack of clinical improvement despite antibiotic therapy and the progressive respiratory distress raised suspicion for an underlying structural abnormality.

Contrast-enhanced chest computed tomography is the definitive imaging modality for confirming pulmonary agenesis and delineating associated bronchovascular anatomy (11, 12). It also facilitates identification of coexisting anomalies and is essential for management planning (11, 12).

Unilateral pulmonary agenesis is frequently associated with other congenital anomalies, most commonly cardiovascular defects (50–60%), skeletal (10–15%), gastrointestinal (5–10%), and genitourinary systems (< 5%) (1, 2). These anomalies should be assessed systematically as they significantly impact the clinical presentation, management decisions and prognosis (11, 13, 14). Therefore, early echocardiography assessment and other imaging modalities should be considered in such patients (1, 2).

Management of unilateral lung agenesis is primarily supportive. Non-invasive ventilation, such as CPAP, is preferred to mechanical ventilation to minimize barotrauma to the solitary functioning lung (1, 2, 4).

Furthermore, pulmonary hypertension is the major determinant of the management and prognosis in patients with unilateral pulmonary agenesis with left to right shunts (1, 2). While early closure of the defect may be beneficial in selected cases, evidence suggested that early closure imposes increased risk of right ventricular failure if severe pulmonary hypertension is already established (1, 4, 15). In low-income settings where access to advanced cardiac interventions is limited, conservative medical management with pulmonary vasodilators and diuretics remains the most feasible approach (1, 4). Early recognition of pulmonary agenesis, systematic assessment for associated anomalies, and timely initiation of targeted therapy are essential to improve outcomes (16).

Limitations

As this is a single case report, the findings may not be generalizable to the broader population of patients with pulmonary agenesis. The diagnosis was made after referral to a tertiary center, reflecting the delayed recognition that commonly occurs in primary care settings. Additionally, limited access to advanced cardiac catheterization and surgical intervention, due to resource limitations restricted therapeutic options.

Conclusion

This case highlights that neonatal lung agenesis is frequently missed and concealed by more common respiratory diseases including pneumonia, particularly in resource-limited settings where access to advanced imaging is limited. Therefore, clinicians should maintain a high index of suspicion and consider congenital pulmonary anomalies in children presenting with persistent respiratory distress and asymmetric chest findings.

Abbreviations

CPAP Continuous positive airway pressure

CRP C-reactive protein

CT Computed tomography

PA Posteroanterior

PDA Patent ductus arteriosus

WBC White blood cell count

Declarations

Ethics approval and consent to participate

Ethics approval was not required for case report at our institution. All procedures were performed in accordance with institutional guidelines.

Consent for publication

Written informed consent was obtained from the parents of patient for publication of the case as a case report and any accompanying images. A copy of the written consent is available for review by the editor-in-chief of this journal.

Availability of data and materials

All data relevant to this case report are included in the manuscript. Anonymized patient data are available from the corresponding author upon reasonable request.

Competing interests

The authors declare no competing interests.

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Authors' contributions

Beniam Tadewos Lamboro, MD: Conceptualization, writing original draft (first and second versions), literature review, manuscript revision and editing, corresponding author.

Samuel Mesfin Girma, MD: Critical revision of manuscript, comment on the first and second draft, imaging interpretation, data verification, supervision and final approval.

Nathanael Elias Temesgen, MD: Clinical diagnosis and management, laboratory data acquisition and interpretation, outpatient follow-up management.

Anteneh Girma Mengistu, MD: Literature review, reference management, figure labeling and formatting, submission assistance, review and editing.

All authors have read and approved the final manuscript.

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References

1. Orellana-Donoso M, Barrenechea-Salvador M, Caro-Navarro J, Cervela-Díaz M, Chacón-Ortiz C, Claudet-Córdoba N, et al. Unilateral Lung Agenesis: A Systematic Review of Prevalence, Anatomical Variants, and Clinical Implications. Diagnostics. 2025;15(17):2272. 10.3390/diagnostics15172272.
2. Fukuoka S, Yamamura K, Nagata H, Toyomura D, Nagatomo Y, Eguchi Y, et al. Clinical outcomes of pulmonary agenesis: A systematic review of the literature. Pediatr Pulmonol. 2022;57(12):3060–8. 10.1002/ppul.26135.
3. Schaffer AJ, Rider RV. A note on the prognosis of pulmonary agenesis and hypoplasia according to the side affected. Trans Am Clin Climatol Assoc. 1956;68:25–31.
4. Muensterer O, Abellar R, Otterburn D, Mathew R. Pulmonary Agenesis and Associated Pulmonary Hypertension: A Case Report and Review on Variability, Therapy, and Outcome. Eur J Pediatr Surg Rep. 2015;3(1):33–9. https://doi.org/10.1055/s-0034-1395986.
5. Dinamarco PVV, Ponce CC. Pulmonary agenesis and respiratory failure in childhood. Autopsy Case Rep. 2015;5(1):29–32. 10.4322/acr.2014.046.
6. Kissou SA, Adjaba SBW, Tamini J, Traore H. Unilateral Pulmonary Agenesis about a Case Revealed by an Acute Respiratory Infection in a Young Infant. Case Rep Pediatr. 2023;2023:8110952. 10.1155/2023/8110952.
7. Alwan A, Mohammoud S, Assefa AZ. Incidental pulmonary agenesis with multiple associated anomalies: A case report. Radiol Case Rep. 2023;18(10):3724–8. https://doi.org/10.1016/j.radcr.2023.08.002.
8. Khurram MSA, Rao SP, Vamshipriya A. Pulmonary agenesis: A case report with review of literature. Qatar Med J. 2013;2013(2):38–40. https://doi.org/10.5339/qmj.2013.14.
9. Chawla RK, Madan A, Chawla A, Arora HN, Chawla K. Pulmonary Agenesis. Indian J Chest Dis Allied Sci. 2015;57(4):251–3.
10. Mondal AR, Majumder BK, Chowdhury RB, Musharraf M, Albani SA, Azad AZ. Comparison of WHO Clinical Criteria and Radiological Findings of Pneumonia in Children Aged 02–59 Months. Mymensingh Med J. 2021;30(3):684–9.

11. Sadiqi J, Hamidi H. CT features of lung agenesis - a case series (6 cases). BMC Med Imaging. 2018;18(1):37. https://doi.org/10.1186/s12880-018-0281-5.
12. Ahmed EK, Tajeddin A, Farfour A, Alshammary M, AlKattan K. Right unilateral lung agenesis with dextroposition; the first case in Saudi Arabia. Respir Med Case Rep. 2020;29:100994. https://doi.org/10.1016/j.rmcr.2019.100994.
13. Weber J, et al. Unilateral Lung Agenesis: A Case Series and Review of Literature. AJP Rep. 2023;13(3):e40–3. https://doi.org/10.1055/a-2107-0651.
14. Rahman SMT, Shahriar T, Akhter KM, Tarannum A, Hossain M. A rare variant of right sided pulmonary agenesis presenting in adulthood: 1st reported case from Bangladesh. Respir Med Case Rep. 2022;37:101629. https://doi.org/10.1016/j.rmcr.2022.101629.
15. Wei F, et al. Embolization treatment of right pulmonary artery agenesis with patent ductus arteriosus causing pulmonary hypertension and hemoptysis: a case report and literature review. J Cardiothorac Surg. 2024;19(1):391. https://doi.org/10.1186/s13019-024-02883-9.
16. Nandan D, Bhatt GC, Dewan V, Pongener I. Complete right lung agenesis with dextrocardia: an unusual cause of respiratory distress. J Clin Imaging Sci. 2012;2:81. https://doi.org/10.4103/2156-7514.105140.

Figures

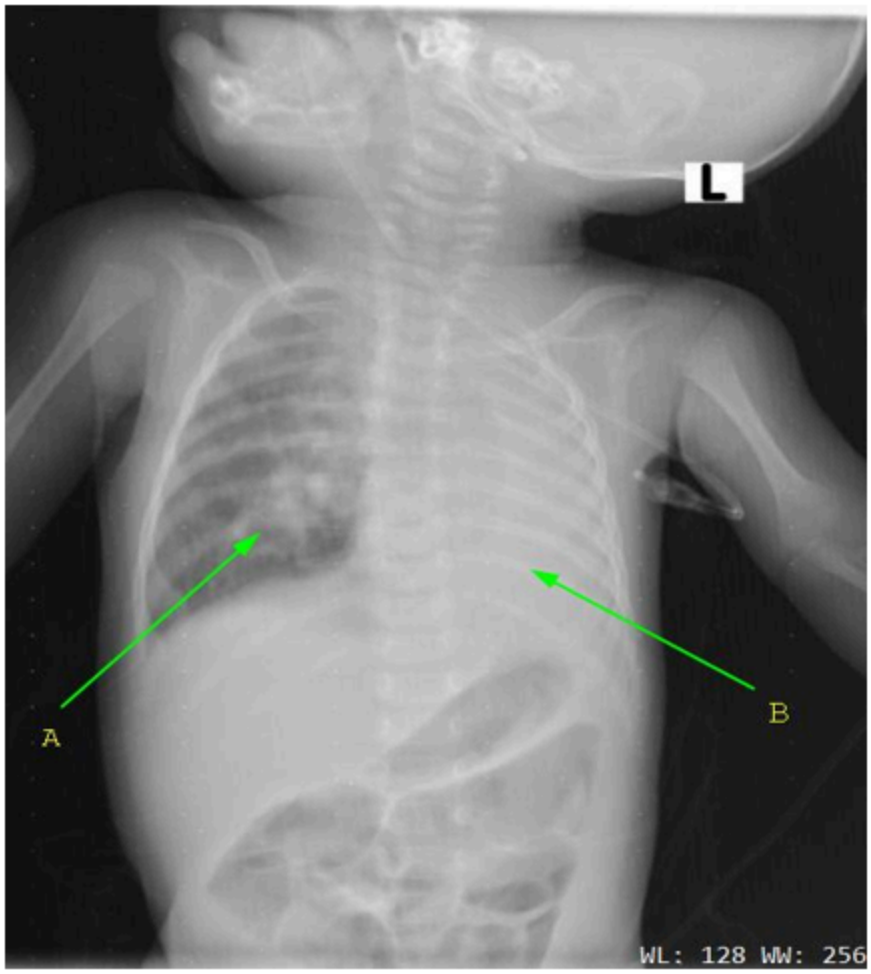


Figure 1

Posteroanterior (PA) chest X ray shows (A) right middle and lower lobe opacity with air bronchogram. (B) whiteout left hemithorax with associated cardiomediastinal shift.

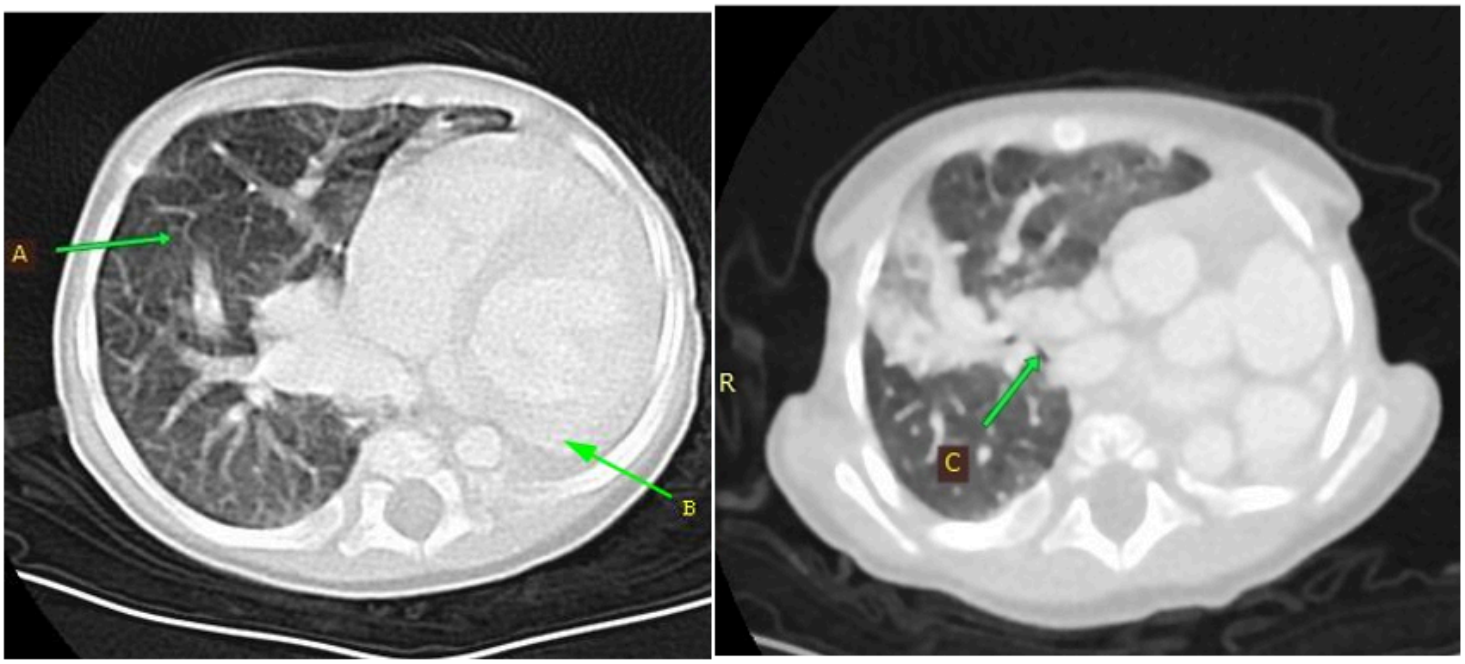


Figure 2

Contrast enhanced chest CT shows (A) right lung parenchyma with compensatory hyperinflation. (B) left hemithorax occupied by mediastinal structures and heart. (C) Right main bronchus with no left counterpart.

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