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Right Lung Agenesis with Dextrocardia in a 4-Month-Old Infant: A Case Report and Literature Review

Yenny Framela, Didik Hariyanto

Department of Child Health, Faculty of Medicine, Universitas Andalas / Padang/ West Sumatra, Indonesia

Corresponding Author: Yenny Framela

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ABSTRACT

Background: Pulmonary agenesis is a rare congenital malformation characterised by complete absence of the lung parenchyma, bronchus, and pulmonary vasculature on the affected side. It arises from a disruption of the lung bud during the fourth to fifth week of gestation. Right-sided agenesis is less common than left-sided agenesis but carries a worse prognosis, owing to a greater degree of mediastinal and cardiac shift, distortion of the airway and remaining lung, and a higher risk of recurrent respiratory infection and tracheal compression.

Case presentation: We report a 4-month-old female infant who was referred with fever, cough, and progressive shortness of breath requiring continuous positive airway pressure (CPAP) support. Physical examination revealed asymmetrical chest movement with diminished breath sounds, scanty coarse crackles, and hyperresonance over the right hemithorax, while the cardiac impulse and heart sounds were displaced to the right hemithorax. Laboratory investigations were within normal limits. Chest radiography showed complete opacification of the right hemithorax with mediastinal shift. Computed tomography (CT) confirmed the absence of the right lung parenchyma, the right main bronchus, and the right pulmonary artery, together with compensatory hyperinflation of the left lung. Echocardiography showed dextrocardia with preserved biventricular size and function and no associated structural cardiac defect.

Conclusion: Right lung agenesis with dextrocardia should be considered in any infant presenting with persistent unilateral chest opacification, especially when accompanied by a contralateral mediastinal shift and a displaced cardiac impulse. Cross-sectional imaging is essential for accurate diagnosis and for excluding associated cardiovascular, gastrointestinal, and skeletal anomalies. In the absence of life-threatening associated malformations, management is largely conservative, supported by chest physiotherapy and aggressive treatment of intercurrent respiratory infections, with long-term outcome depending mainly on the functional capacity of the contralateral lung.

Keywords: Dextrocardia; Pulmonary agenesis; Congenital lung anomaly; Infant; Case report

INTRODUCTION

Pulmonary agenesis is defined as the complete congenital absence of one or both lungs, including the bronchus, pulmonary parenchyma, and pulmonary vasculature on the affected side. Population-based estimates suggest an incidence of approximately one in

10,000 to one in 100,000 live births, although the true prevalence is likely underestimated because many affected infants do not survive the neonatal period or remain undiagnosed if asymptomatic into adulthood [1,2].

Embryologically, the lung bud arises as a ventral diverticulum of the foregut at approximately the fourth week of gestation, subsequently dividing into the two primary bronchial buds that give rise to the right and left lungs. A disturbance of this process during the fourth to fifth gestational week, before the pseudoglandular stage, can result in failure of one bronchial bud to develop, producing unilateral agenesis. Because the heart also undergoes its leftward looping during this same critical window, an early developmental insult can simultaneously disrupt cardiac positioning, explaining the frequent association between unilateral lung agenesis and dextrocardia or dextroposition of the heart [3].

Right-sided agenesis is reported less frequently than left-sided agenesis, yet it is associated with a worse prognosis. This has been attributed to a more pronounced mediastinal shift, herniation and rotation of the hyperinflated contralateral lung across the midline, distortion of the trachea and remaining bronchus, and a higher rate of associated tracheal stenosis and gastrointestinal anomalies, all of which are independent predictors of mortality [1,4,5].

Because the clinical presentation of unilateral lung agenesis-recurrent respiratory distress, unilateral chest opacification on radiography, and a shifted mediastinum-closely mimics more common conditions such as pneumonia, pleural effusion, massive atelectasis, or a foreign-body aspiration, the diagnosis is frequently delayed or missed on initial presentation [4,6,7]. We report a case of right lung agenesis with dextrocardia in a young infant, with the aim of increasing clinical awareness of this rare anomaly and outlining a structured diagnostic and management approach.

CASE PRESENTATION

A 4-month-old female infant was referred to our tertiary hospital with a five-day history of fever, cough, and progressively worsening shortness of breath. On admission, the patient was tachypnoeic and required continuous positive airway pressure (CPAP) to maintain adequate oxygenation. There was no history of cyanotic spells, feeding difficulty prior to the current illness, or recurrent hospitalisation. The pregnancy and delivery history were unremarkable, and there was no family history of congenital heart disease or other major birth defects.

On physical examination, the patient was in moderate respiratory distress with subcostal and intercostal retractions. Inspection and palpation of the chest revealed markedly reduced movement of the right hemithorax compared with the left. Auscultation revealed diminished breath sounds with scanty coarse inspiratory crackles over the right hemithorax, while percussion over the left hemithorax was hyperresonant, consistent with compensatory hyperinflation. Notably, the apex beat (ictus cordis) and heart sounds were best heard over the right hemithorax, raising suspicion of dextrocardia or dextroposition of the heart. The abdomen was soft and non-distended, and no other dysmorphic features or limb, vertebral, or anorectal anomalies were noted on initial examination.

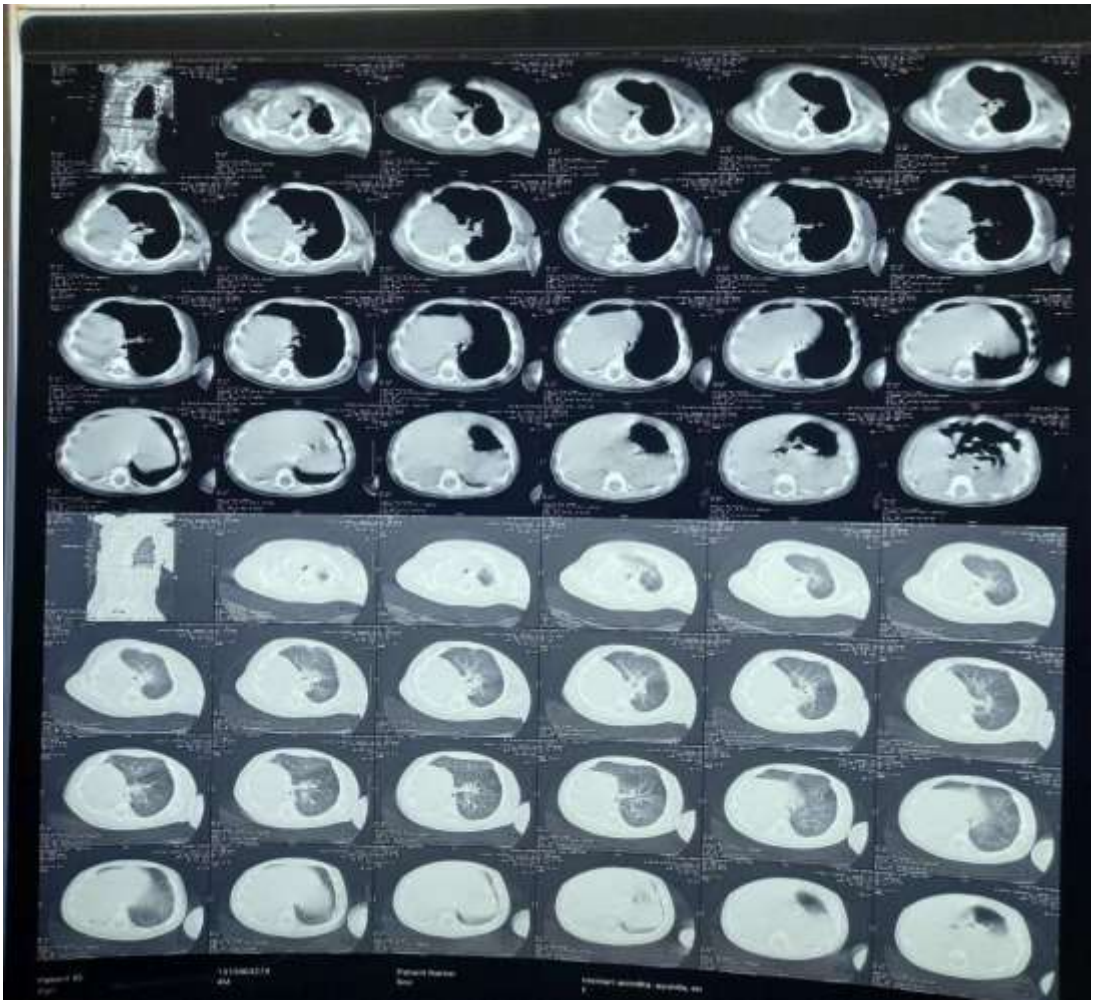
Routine laboratory investigations, including a complete blood count, C-reactive protein, electrolytes, and renal function, were all within normal limits for age, making an active bacterial pneumonia with parapneumonic effusion less likely as the sole explanation for the radiographic findings.

Chest radiography demonstrated a homogeneous opacification of the right hemithorax with a shift of the mediastinum and cardiac silhouette towards the right side, and compensatory hyperlucency of the left lung field. Given the discrepancy between the clinical findings (right-sided dullness/crackles with hyperresonant left

lung, and a right-sided cardiac impulse) and the plain radiographic appearance, a contrast-enhanced computed tomography (CT) of the thorax was performed.

CT of the thorax confirmed complete absence of the right lung parenchyma and the right main bronchus, with marked hyperinflation of the left lung parenchyma extending across the midline into the right hemithorax, and absence of the right pulmonary artery. There was no evidence of tracheal stenosis, oesophageal compression, or an associated diaphragmatic hernia. Transthoracic echocardiography confirmed dextrocardia with the cardiac apex pointing to the right, while biventricular chamber size and systolic function were within normal limits, and no significant intracardiac structural defect, such as an atrial or ventricular septal defect, was identified.

Based on the clinical, radiological, and echocardiographic findings, a diagnosis of right lung agenesis with dextrocardia, complicated by an acute lower respiratory tract infection, was established. The patient was managed with supplemental oxygen via CPAP, empirical broad-spectrum intravenous antibiotics, adequate hydration, and chest physiotherapy directed at the functioning left lung. Surgical intervention was not indicated, as there was no associated tracheal stenosis, vascular compression, or life-threatening structural cardiac anomaly. The patient showed gradual clinical improvement, with resolution of respiratory distress and successful weaning from CPAP, and was discharged in good clinical condition on a structured chest physiotherapy programme, with arrangements for outpatient follow-up in the paediatric pulmonology and cardiology clinics to monitor growth, respiratory function, and pulmonary arterial pressure.



DISCUSSION

Embryology and Pathogenesis

The respiratory system originates from the laryngotracheal groove of the foregut endoderm at approximately day 26 of gestation. This diverticulum elongates and bifurcates into the right and left lung buds, which subsequently undergo branching morphogenesis to form the bronchial tree. Concurrently, the primitive heart tube undergoes rightward looping (D-looping) and the developing lungs influence the final position of the cardiac apex within the thorax. An insult occurring during this narrow developmental window—whether vascular, genetic, or related to a teratogenic exposure—can arrest the growth of one lung bud while sparing the contralateral side, and may simultaneously alter the normal leftward orientation of the heart, producing the combination of unilateral pulmonary agenesis and dextrocardia or dextroposition observed in our patient [3].

Clinical Presentation and Differential Diagnosis

Infants with unilateral lung agenesis may remain asymptomatic for a variable period, particularly when the contralateral lung provides adequate compensatory function, and many cases are only unmasked during an intercurrent respiratory infection, as occurred in our patient [6,7]. The classic clinical triad—asymmetrical chest expansion, a shifted cardiac impulse, and a unilaterally opaque hemithorax with hyperinflation of the opposite lung on chest radiography—should raise suspicion for this diagnosis. However, because these radiographic features overlap considerably with massive pleural effusion, complete lung collapse from a foreign body or mucus plug, and severe pneumonia with consolidation, lung agenesis is frequently misdiagnosed initially as pneumonia or, in older children, as a retained foreign body [4,5,6]. The discordance in our patient between a radiographically opaque right hemithorax and the clinical findings of a right-sided cardiac impulse together with a hyperresonant left hemithorax was the key clue prompting further cross-sectional imaging.

Diagnostic Approach

Plain chest radiography is usually the first imaging modality obtained, but it is rarely sufficient to confirm the diagnosis and may be misleading, as in the present case. Contrast-enhanced CT of the thorax is considered the investigation of choice, as it can directly demonstrate the absence of the bronchus, lung parenchyma, and pulmonary artery on the affected side, while also delineating the position and degree of herniation of the contralateral lung and excluding associated tracheal or oesophageal abnormalities [1,4,5]. Echocardiography remains essential in every case, both to confirm the cardiac position (dextrocardia versus dextroposition secondary to mediastinal shift) and to identify any associated structural cardiac defects, which are reported in up to 40% of patients with unilateral pulmonary agenesis [1]. In our patient, echocardiography demonstrated true dextrocardia without an associated structural cardiac lesion, which is a relatively favourable finding given the higher prevalence of cardiac anomalies reported in right-sided agenesis.

Associated Anomalies and Prognostic Factors

Unilateral pulmonary agenesis is an isolated finding in only around one-quarter of reported cases, with the remainder showing associated cardiovascular, skeletal, gastrointestinal, genitourinary, or tracheal anomalies [1]. A large systematic review found that right-sided agenesis was significantly more often associated with tracheal stenosis than left-sided disease, and that both tracheal stenosis and gastrointestinal anomalies were independent predictors of mortality, whereas the overall two-year survival of unilateral agenesis approached 62%, with no deaths reported beyond 13 years of age once a child has survived early infancy [1]. A more recent

systematic review similarly emphasised the wide anatomical heterogeneity of unilateral lung agenesis and the importance of a thorough search for associated malformations once the diagnosis is suspected [2]. In line with these findings, our patient underwent a comprehensive evaluation that did not identify tracheal stenosis, oesophageal compression, diaphragmatic hernia, or major structural cardiac disease, all of which are favourable prognostic indicators.

Management and Follow-up

Surgical intervention is rarely required for isolated unilateral lung agenesis and is generally reserved for cases complicated by significant tracheobronchial compression, severe gastro-oesophageal reflux with recurrent aspiration, or progressive pulmonary hypertension [8,9]. In the absence of these complications, management remains predominantly supportive and conservative, focusing on prompt treatment of intercurrent respiratory infections, optimisation of nutrition and growth, and a structured chest physiotherapy programme aimed at maximising the function of the remaining lung [6,7]. Several recently published case reports of right lung agenesis with dextrocardia, including cases diagnosed in adulthood, have similarly described favourable outcomes with conservative management when no major associated anomalies were present [4,5]. Long-term follow-up should include serial assessment of growth and respiratory symptoms, periodic echocardiography to monitor for the development of pulmonary hypertension secondary to the redistribution of the entire pulmonary blood flow through the single remaining lung, and pulmonary function testing once the child is old enough to cooperate, as obstructive ventilatory patterns have been described even in adults who survive into later life with this condition [4].

CONCLUSION

Right lung agenesis with dextrocardia is a rare congenital anomaly that should be considered in any infant presenting with an opaque hemithorax accompanied by a contralateral hyperresonant lung and a displaced cardiac impulse, particularly when the clinical findings appear discordant with the plain chest radiograph. Contrast-enhanced CT and echocardiography are essential for confirming the diagnosis and for systematically excluding associated cardiovascular, tracheal, and gastrointestinal anomalies, which are the principal determinants of long-term prognosis. In the absence of such complications, as in the patient described here, conservative management with supportive respiratory care and chest physiotherapy can achieve a favourable short-term outcome, although lifelong follow-up is warranted to monitor for late complications such as pulmonary hypertension and obstructive lung disease.

Declarations

Ethics approval and consent to participate: Not applicable (single case report).

Consent for publication: Written informed consent was obtained from the patient's parent/legal guardian for publication of this case report and any accompanying images.

Availability of data and materials: The datasets used and/or analysed during the current case are available from the corresponding author on reasonable request.

Competing interests: The authors declare that they have no competing interests.

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Authors' contributions: YF was responsible for clinical management of the patient, conception of the manuscript, literature review, and drafting of the manuscript. DH supervised the clinical management and critically revised the manuscript for important intellectual content. Both authors read and approved the final manuscript.

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