Right-sided congenital diaphragmatic hernia masqueraded as right lobar pneumonia in a term newborn infant

Menghao Duan ¹, Teo Eu-Leong Harvey James,^{2,3} Suresh Chandran ^{3,4,5,6}

¹Department of Pediatrics, KK Women's and Children's Hospital, Singapore

²Department of Diagnostic and Interventional Imaging, KK Women's and Children's Hospital, Singapore

³Paediatric Academic Clinical Programme, Duke NUS Medical School, Singapore

⁴Department of Neonatology, KK Women's and Children's Hospital, Singapore

⁵Paediatric Academic Clinical Programme, Lee Kong Chian School of Medicine, Singapore

⁶Paediatric Academic Clinical Programme, Yong Loo Lin School of Medicine, Singapore

Correspondence to

Professor Suresh Chandran; profschandran2019@gmail.com

Accepted 2 October 2021

DESCRIPTION

A full-term male infant born by normal vaginal delivery developed respiratory distress at 15 min of life. Antenatal history was unremarkable. The last fetal scan at 32 weeks gestation did not reveal any congenital anomaly. There was no maternal history of leaking liquor, fever, or group B streptococcus colonisation.

Physical examination showed tachypnoea, chest retractions and decreased air entry in the right lung. Oxygen saturation was 80%–82%, and blood gas showed respiratory acidosis. He was placed on continuous positive airway pressure of 7 cm water and FiO₂ 0.3. Vital signs were within normal limits. Blood count and C reactive protein levels were unremarkable. Chest X-ray (CXR) showed opacity in the lower zone of the right chest, suggesting lobar consolidation ([figure 1](#)). Antibiotics were initiated promptly. At 6 hours of life, with worsening respiratory distress and echocardiography showing evidence of pulmonary hypertension, he was intubated. Repeat CXR revealed multiple radiolucent bowel loops in the right hemithorax ([figure 2](#)), consistent with a diagnosis of right-sided congenital diaphragmatic hernia (R-CDH) as confirmed by ultrasound. Karyotyping was normal.

The infant remained haemodynamically stable while intubated with FiO₂ of 0.3. Surgical repair was performed on day 2 via the transabdominal approach. Intraoperatively, viable loops of intestines were found in the chest and mobilised back into the abdomen. There was no significant liver herniation. The diaphragmatic defect was repaired

Figure 2 A follow-up chest radiograph at 6 hours of life shows the opacification in the right lung to have increased to involve almost the entire right hemithorax. There are now bowel loops seen in the right lower chest consistent with the diagnosis of a congenital diaphragmatic hernia.

by primary closure. Postoperatively, he was extubated on day 7, initiated on feeds on day 9, and discharged home on day 17 of life.

Congenital diaphragmatic hernia (CDH) occurs in 0.08–0.45 per 1000 births, attributed to the failure of fusion of pleuroperitoneal membranes during fetal development.¹ Antenatal ultrasound screening identifies more than 60% of CDH by second trimester and prognosticates using serial lung-to-head ratio assessment and degree of liver herniation. Fetal MRI estimates fetal lung volumes and liver position.²

CDH occurs on the right in only 13% of cases.³ Infants with R-CDH often present with respiratory distress at birth due to lung hypoplasia and persistent pulmonary hypertension of newborn.⁴ Diagnosis of R-CDH can be challenging as clinical presentation and radiological findings may masquerade as lung pathology.^{5,6} In R-CDH, the liver may obstruct the diaphragmatic defect and prevent extrusion of abdominal contents into the thorax, hence not visualised on plain radiographs and delaying diagnosis.

Initial CXRs of R-CDH may be misinterpreted as pneumonia, as demonstrated in our case. Pleural effusion may occur due to hepatic venous outflow obstruction causing congestion. Differentials include pneumatoceles, cystic adenomatoid malformation, congenital lobar emphysema and diaphragmatic eventration.⁷ When diagnosis is equivocal, alternative-imaging modalities such as

Figure 1 The chest radiograph shows the presence of an opacity in the lower zone of the right chest. Some non-specific air lucencies are noted in the periphery of the right lower chest.

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To cite: Duan M, James TE-LH, Chandran S. *BMJ Case Rep* 2021;**14**:e244975. doi:10.1136/bcr-2021-244975

Images in...

ultrasonography can be complementary. Ultrasound identifies parenchymatous organs and peristaltic movement of bowels in the hemithorax, and the absence of hyperechoic diaphragm and pulmonary A-lines in CDH.⁸ Doppler vascular flow aids in diagnosing thoracic liver herniation.

Intestinal malrotation is reported in 39% of infants during initial CDH repair and those with a missed association have higher risk of future abdominal complications.⁹

Overall survival in R-CDH is approximately 63%.¹⁰ The severity of pulmonary hypoplasia and pulmonary hypertension

is a significant factor determining R-CDH outcome. The timing of surgical repair also contributes to subsequent morbidities.⁴ Fortunately, the initial stabilisation at birth, early diagnosis and surgical intervention gave our patient a favourable outcome.

Contributors MD and SC were involved in the conception and drafting of the manuscript. HT provided interpretation of radiograph findings and input for figure captions. All authors contributed substantially to the article revision.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Consent obtained from parent(s)/guardian(s)

Provenance and peer review Not commissioned; externally peer reviewed.

ORCID iDs

Menghao Duan <http://orcid.org/0000-0002-6596-7920>

Suresh Chandran <http://orcid.org/0000-0001-5961-7441>

Patient's perspective

I work as a paramedic and dealing with life-and-death situations is part of my daily job. However, when my newborn baby became unwell, I felt so helpless. I am very grateful to the doctors for their prompt diagnosis and surgery to ensure my baby had the best outcome.

Learning points

- Clinical and radiological features of congenital diaphragmatic hernia (CDH) may be misleading at birth and more often suggest other common diagnoses. Delayed diagnosis could lead to increased morbidity and mortality.
- The presence of gas-filled bowels high in the right upper abdominal quadrant migrating through the right subdiaphragmatic plane in the radiogram suggests a diagnosis of R-CDH.
- Use of continuous positive airway pressure respiratory support at birth in undiagnosed CDH could worsen the intrathoracic migration of gut loops. Hence, fetal diagnosis, elective intubation at birth and stabilisation followed by surgical repair are paramount to achieve an optimal outcome.

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