

**PUBLISHED ABSTRACT**

# Pulmonary Hernia in an Infant with Hepatic Osteodystrophy: A Novel Cause of Dyspnea

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## Background

Pulmonary herniation is a rare condition where the lung parenchyma protrudes beyond the confines of the thoracic cavity. It is categorized as either congenital or acquired, with acquired subdivided into spontaneous, traumatic, and pathologic. In pediatric patients, pulmonary hernias are typically congenital, related to agenesis of the ribs or sternum, or traumatic. While pulmonary hernia has been described in other chest wall diseases, it has not been described in the setting of hepatic osteodystrophy. In this condition, osteopenia results when chronic cholestasis causes vitamin D and calcium malabsorption, resulting in secondary hyperparathyroidism and rickets. We present a case of an infant female with hepatic osteodystrophy undergoing work-up for respiratory distress, incidentally, found to have an anterior thoracic pulmonary hernia contributing to symptoms of dyspnea.

## Case Details

Our patient is a 10-month-old female with a history of extrahepatic biliary atresia who underwent a failed Kasai portoenterostomy at 47 days-of-life, currently listed for liver transplant. She presented with increased work of breathing, and a chest x-ray showed a lucency projecting over the right middle lung as well as rachitic rosaries (**Figure 1**). She was hypophosphatemic (1.6 mg/dL) with normal serum calcium. Transaminases and total and direct bilirubin were elevated but at her baseline (ALT 161 U/L, AST 308 U/L, total bilirubin 23.3 mg/dL, direct 16.9 mg/dL). A respiratory viral panel and COVID-19 testing were negative. She was admitted, and subsequent work-up revealed an undetectable hydroxycholecalciferol level, persistently low phosphorus, and an elevated parathyroid hormone (434 PG/ML), consistent with hepatic osteodystrophy with secondary hyperparathyroidism and rickets. Her hospital course was marked by intermittent periods of increased work of breathing without hypoxia, during which she was treated with high flow nasal cannula. Serial chest x-rays remained unchanged, and, due to an elevated d-dimer, a CTA chest was obtained (**Figure 2**).



**Figure 1:** Chest x-ray with relative lucency projecting over the medial right lung due to anterior thoracic pulmonary herniation.



**Figure 2:** CTA chest demonstrating herniation of portion of right lung anteriorly beyond the anterior margin of the right ribs.

The study was negative for thrombus but demonstrated herniation of a portion of the right lung anteriorly beyond the anterior margin of the right rib as well as multiple healing right rib fractures. While the etiology of her increased work of breathing is believed to be multifactorial, pulmonary herniation causing dyspnea and pain was identified as a contributing factor.

### Discussion

Here we not only present an unusual cause of increased work of breathing in an infant, but a never reported constellation of findings in thoracic pulmonary hernia. Due to higher clinical concern for infection or pneumothorax and the rare nature of this diagnosis, the identification of pulmonary hernia was delayed and incidental. There were no findings of chest wall structure agenesis and the patient had no prior history of respiratory concerns in early infancy, which speaks against the presence of a congenital hernia. Instead, the patient demonstrated osteopenia with resolved rib fractures suggesting either a pathologic herniation due to chest wall weakness or an unidentified chest wall trauma. While more common causes of increased work of breathing must first be considered, pulmonary herniation is a potential contributing factor in infant patients with increased work of breathing, especially if they have risk factors for chest wall weakness.

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