

# CONGENITAL DIAPHRAGMATIC HERNIA WITH LATE PRESENTATION: A CASE REPORT

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## ABSTRACT

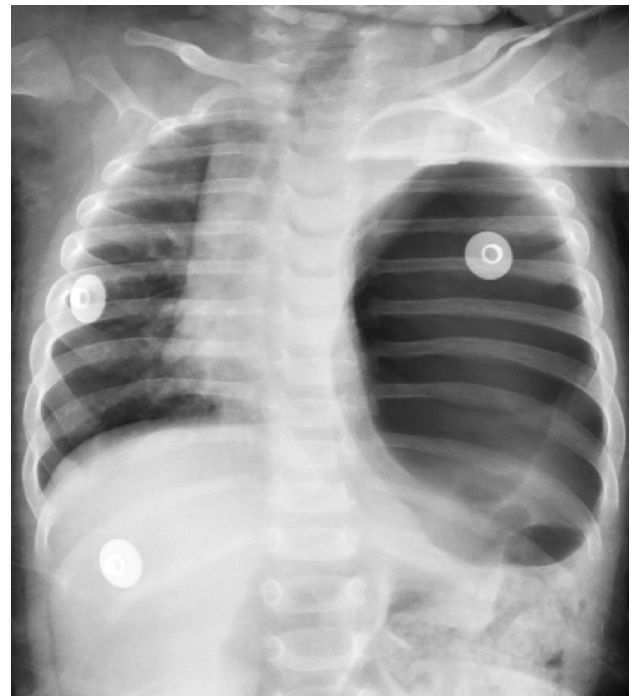
Congenital diaphragmatic hernia (CDH) presenting after neonatal period is extremely varied,<sup>1</sup> also CDH is a neonatal period disease, late presenting CDH may occur after neonatal period which first reported by Kirkland in 1959<sup>2</sup> so more attention to diagnosis of this kind of CDH is done in the past decade.<sup>3</sup> CDH mainly present with acute respiratory distress in neonatal period however late presenting CDH has milder symptoms with favorable outcome.<sup>4</sup> Due to milder and confusing clinical presentation of this kind of CDH, it has considerable diagnostic challenge.<sup>5</sup> Confusing symptoms lead to delay diagnosis of this type of hernia so diagnostic imaging play an important role in diagnosis.<sup>5,6</sup> Chest-x-ray (CXR), upper GI series (UGI) and CT are the most common modalities used for<sup>6</sup> diagnosis.

**Keywords:** Congenital Diaphragmatic Hernia; Bochdalek Hernia; Radiography; CT scan

## Case Report

An 8-month-old male infant admitted to our hospital 6 hours after minor head trauma due to falling down on the back head from 50 centimeter height. Infant was restless and agitated. No loss of consciousness, nausea or vomiting was reported. vital sign in emergency room were as follow; pulse rate (PR) was 150/min, respiratory rate (RR) was 62/min blood pressure was normal. Breath sounds was diminished. Complete blood count (CBC) revealed white blood cell count (WBC) of 21000 in cubic mm with 84% polymorphonuclear (PMN), hemoglobin(Hb) was 10.6 gm/dl with mean corpuscular volume(MCV) of 75 femtoliters and mean corpuscular hemoglobin(MCH) of 23 picograms and platelet count of 428000 in cubic mm. Venous blood gas (VBG) at emergency room revealed  $PCO_2=44$  mm Hg,  $PH=7.25$ ,  $PO_2=42$  mm Hg,  $HCO_3=18$  mm/L and base excess (BE)=-7.8. Patient had no fever. Nasal  $O_2$  is administered however  $O_2$  saturation of patient didn't reach over than 90% by pulse oxymetry. Due to reduce breath sounds and

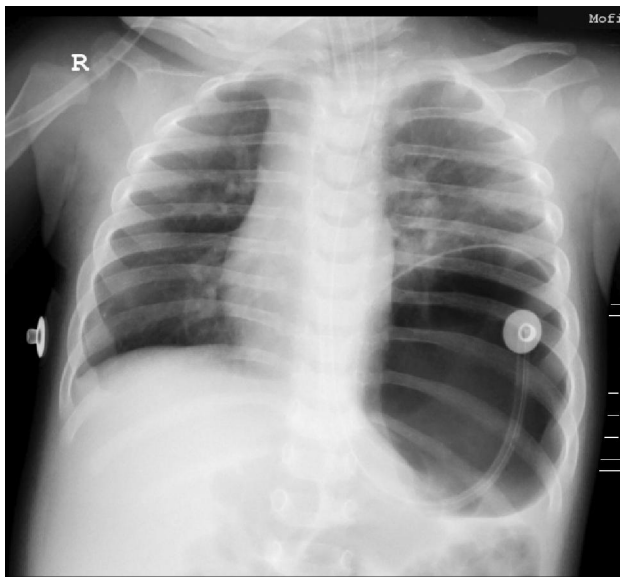
$O_2$  saturation, chest X-Ray (CXR) was taken which revealed lucent left hemithorax with mediastinal shift to right side (Fig-1A).



**Figure 1A:** An 8-month-old infant with respiratory distress; CXR reveals cystic lesion in left hemithorax with mediastinal shift and pushes left hemidiaphragm downward. Gastric gas shadow isn't seen in normal position.

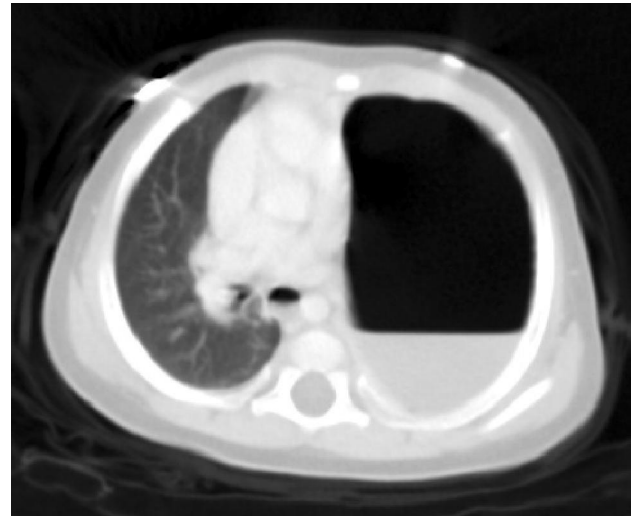
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Surgical consult was done for placement of chest tube with impression of patient being have tension pneumothorax, However pediatric surgery fellow refused that diagnosis and scheduled patient for thoracotomy with diagnosis of congenital lobar emphysema (CLE) or large cystic lung disease. For this reason radiologic consult was requested for preoperative radiographic diagnosis. Radiologist confirmed that the CXR finding is not compatible with pneumothorax; however, this large cystic lesion may be due to a gas filled stomach herniated to left hemithorax due to absence of gastric fundus gas under diaphragm. Nasogastric tube (NGT ) was placed and follow up CXR revealed reduce in size of the lucent lesion in left hemithorax and position of NGT in left hemithorax.(Fig-1B).



**Figure 1B:** An 8-month-old infant with respiratory distress; CXR after placement of NGT reveals position of NGT in left hemithorax and reduce size of mentioned cystic lesion in Fig-1A. these findings are suggestive of herniated stomach to left hemithorax.

Finally patient underwent computerized tomography scan (CT Scan) for confirmation of diagnosis. CT Scan with coronal and sagittal reformatted views revealed distended stomach in left hemithorax (Fig-1C, D) which pass through a defect in posterior part of diaphragm in left side suggestive of Bochdaleck hernia. Patient underwent surgical thoracotomy with surgical impression of cystic lung disease or congenital lobar emphysema, stomach was seen in left hemithorax and pushed to abdomen and diaphragmatic defect was repaired. Pulmonary hypoplasia due to diaphragmatic



**Figure 1C:** An 8-month-old infant with respiratory distress; Thorax CT scan with lung window reveals a cystic lesion in left hemithorax with air-fluid level.



**Figure 1D:** An 8-month-old infant with respiratory distress; Thorax CT scan with mediastinal window reveals a cystic lesion in left hemithorax with air-fluid level.

ragmatic hernia cause some difficulties in patient recovery however patient discharged 4-week after surgery without surgical or respiratory complication.

## Discussion

Congenital diaphragmatic hernia CDH presenting with various clinical presentation.<sup>1</sup> This congenital anomaly still has high mortality.<sup>7,8</sup> Patient representing this anomaly mainly diagnosed within some hours or days after birth in neonatal period However late presenting CDH have incidence of 5-27% of all cases.<sup>1,4</sup> Study by Elhalaby<sup>1</sup> in 33 cases revealed 45.5% diagnosis

between 2 months to 14-year-old more common of Bochdalek and left posterior type. delayed form of presenting diaphragmatic hernia mainly present with chronic respiratory complication and less likely with respiratory distress or gastrointestinal complication.<sup>3,4,5</sup> Study by Chang<sup>9</sup> showed that late presenting CDH had more subtle symptoms and more diagnostic difficulties. CDH is more common in male than female nearly 2:1.<sup>1,3,4,6</sup> Age of patient with in late presenting Bochdalek-type CDH is vary and study by Cigdem<sup>4</sup> in 19 patient revealed 36.8% cases over than 1-year-old. The largest study by Baglaj<sup>6</sup> which review of 122 articles of late presenting CDH up to 2003 he found that 41.8% cases younger than 1-year and 34.8% between 1-5 years. 93.8% of cases in this review article had correct preoperative diagnosis.<sup>6</sup> Our case revealed criteria of other studies, however he represented acute respiratory distress which was less likely seen in other reports. Misdiagnosis is the most important things in management of late presenting CDH which led to delayed diagnosis of patient and appropriate treatment. In study by Baglaj<sup>6</sup> more common wrong diagnosis according to CXR were, pneumothorax, pleural effusion with or without pneumonia, lung cyst, hydro pneumothorax or pyopneumothorax, pneumatocele and even mediastinal or paravertebral mass when the lesion was opaque. Emergency room physician suspected to tension pneumothorax however, surgeon was suspected to cystic lung disease or congenital lobar emphysema. Most frequently diagnostic tool in studies were CXR without or with NGT and upper GI series(UGI).<sup>6</sup> Baglaj<sup>6</sup> in his review article found that CT Scan was done in 7.8% of cases with or without CXR and UGI series. We didn't do UGI series due to scheduling surgery of distress of infant however CXRs and CT scan with reformatted views were sufficient for diagnosis in this case.

## Conclusion

Late presenting congenital diaphragmatic hernia may have varying presentation and may be misleading. Appropriate clinical examination and ordering CXR especially with NGT placement are main stay in diagnosis. If patient condition is permit UGI series is

the second imaging step, however spiral CT scan with reformatted views has valuable information for diagnosis especially when patient is not suitable for UGI series.

## References

1. Elhalaby EA, Abo Sikeena MH. Delayed presentation of congenital diaphragmatic hernia. *Pediatr Surg Int* 2002; **18**: 480–5.
2. Krikland J. congenital posterolateral diaphragmatic hernia in the adult. *Br J Surg* 1957; **47**: 16–22
3. Baglaj M. Presenting congenital diaphragmatic hernia in children: a clinical spectrum. *Pediatr Surg Int* 2004; **20**: 658–69.
4. Cigdem MK, Onen A, Otcu S, Okur H. Late Presentation of Bochdalek-Type Congenital Diaphragmatic Hernia in Children: A 23-Year Experience at a Single Center. *Surg Today* 2007; **37**: 642–5.
5. Zaleska-Dorobisz U, Bagaj M, Sokoowska B, adogórska J, Moroñ K. Late presenting diaphragmatic hernia: clinical and diagnostic aspects. *Med Sci Monit.* 2007 May; **13** Suppl **1**: 137-46.
6. Baglaj M, Dorobisz U. Late-presenting congenital diaphragmatic hernia in children: a literature review. *Pediatr Radiol* 2005; **35**: 478–88.
7. Butler M, Stolar C, Altman P. Contemporary management of congenital diaphragmatic hernia. *World J Surg* 1993; **17**: 350-5.
8. Weinstein S, Stolar C. Congenital diaphragmatic hernia and extracorporeal membrane oxygenation. *Pediatr Clin NAm* 1993; **40**: 1315–33.
9. Chang SW, Lee HC, Yeung CY, Chan WT, Hsu CH, Kao HA, Hung HY, Chang JH, Sheu JC, Wang NL. A twenty-year review of early and late-presenting congenital Bochdalek diaphragmatic hernia: are they different clinical spectra? *Pediatr Neonatol.* 2010 Feb; **51**(1): 26-30.