# Late presentation of a right Bochdalek hernia with a right intrathoracic stomach and organo-axial torsion

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## ABSTRACT

A postero-lateral hernia through the foramen of Bochdalek is a rare type of congenital diaphragmatic hernia (CDH). The incidence of Bochdalek hernia on the right side is 10-20% compared to the left side, and herniation of the stomach into the right pleural cavity is extremely rare. We report a case of right-sided Bochdalek hernia with a right intrathoracic stomach and organo-axial torsion misdiagnosed initially, and treated as a case of hyperactive airway disease. The child had a right thoracotomy, excision of the hernia sac that contained the stomach, greater omentum and part of the liver, reduction of the viscera into the abdominal cavity and simple closure of the diaphragmatic defect. Recovery was uneventful. This case highlights the consequences of late diagnosis and the effectiveness of surgical relief. A new clinico-anatomical classification of Bochdalek hernia is presented.

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Bochdalek congenital diaphragmatic hernia results from failure of proper fusion of the pleuroperitoneal membrane with the central portion of the diaphragm and the body wall between the third and eighth week of embryonic life. The resulting persistent pleuroperitoneal canal in the posterolateral region of the diaphragm permits intrathoracic herniation of abdominal contents.[1–4] This type of congenital diaphragmatic hernia (CDH) occurs in approximately 1 in 4000 live births. It is very rare beyond infancy, with approximately 105 reported cases occurring outside the neonatal period in the English literature to date.[1–3,5-7] It is usually left-sided and unilateral, but rarely may be right-sided or bilateral.[8] It predominates in males 3 times more than females.[1,2,4] The most common presenting symptoms include abdominal or chest pain with associated vomiting, dyspnea or intestinal obstruction.[1-4] Most patients present as an emergency and diagnosis before surgery is not correct in over 30%.[1-3] Delayed surgical intervention results in high morbidity and mortality from respiratory failure, bowel necrosis, infarction of abdominal organs, visceral perforation or shock.[1,2] There is an indication for surgical management in all cases. Patients with CDH may suffer from significant airway obstruction, particularly in childhood, when clinically detectable airway obstruction and airway hyperactivity may be a predominant feature.[9,10] In infants with CDH, both the ipsilateral and contralateral lungs are hypoplastic culminating in reduced lung volume, impaired bronchial development and branching, reduced alveolar number and size. Thickening of the muscular layer of the arteriolar vasculature is also a prominent

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finding as well as a decrease in the number of vessels per unit lung.[11] Our objectives in reporting this case are to highlight the need for awareness and thorough differential diagnosis in children who present with prolonged respiratory symptoms, and to document a rare case of right Bochdalek hernia with a right intrathoracic stomach and organo-axial torsion.

Case Report. The patient, a 2-and-a-half-year old Saudi female infant from Southwestern Saudi Arabia, was the product of an uneventful pregnancy and normal delivery with a birth weight of 3.2 kg. There was no history of neonatal complications. She thrived well until the age of 5 months, when she began to have recurrent chest symptoms characterized by cough and breathing difficulty. There was an associated history of intermittent vomiting. She was initially diagnosed and treated as a case of bronchial asthma until the age of 2-and-a-half-years, frequently receiving bronchodilators, oral and inhaled steroids, and antibiotics at her local hospital. The child was subsequently referred to the Assir Central Hospital (ACH), Abha, Kingdom of Saudi Arabia as case of chronic hyperactive airway disease, which was difficult to control, and a right chest mass. Physical examination on admission showed the following: weight was 11 kg, which was below the 3rd percentile; height was 95 cm, which was in the 10th percentile; the head circumference was 47.5 cm, which was in the 10th percentile. The vital signs were: respiratory rate - 28/minute, heart rate - 105/minute, temperature - 37°C. There was no sign of respiratory distress and there was no digital clubbing, eczema or other signs of allergy. The trachea was central. Chest movements were less on the right side, air entry was decreased and crepitations were present in the right lower zone of the chest posteriorly. Breath sounds were broncho-vesicular in nature. There were no wheezes. Bowel sounds were audible in the right lower chest on auscultation. Examination of the abdomen, cardiovascular, neurological and musculoskeletal systems were unremarkable. Arterial blood gases values were: pH = 7.34 mm Hg, $PaO_2$ = 65 mm Hg, $pCO_2$ = 38 mm Hg, HCO3 23 mmol/l. $SaO_2$= 94% (on room air). Complete blood count, serum urea, creatinine, electrolytes and liver function tests were within normal limits. Plain chest x-ray (postero-anterior view) **(Figure 1)** showed a double cystic mass 6 x 7 cm occupying the right middle and lower lung fields with the absence of the gastric air bubble. A plain computed tomography (CT) scan of the chest **(Figure 2)** showed 2 posterior cavities in the right middle and lower lung zones posteriorly. The CT scan of the chest preceded the barium studies, as the provisional

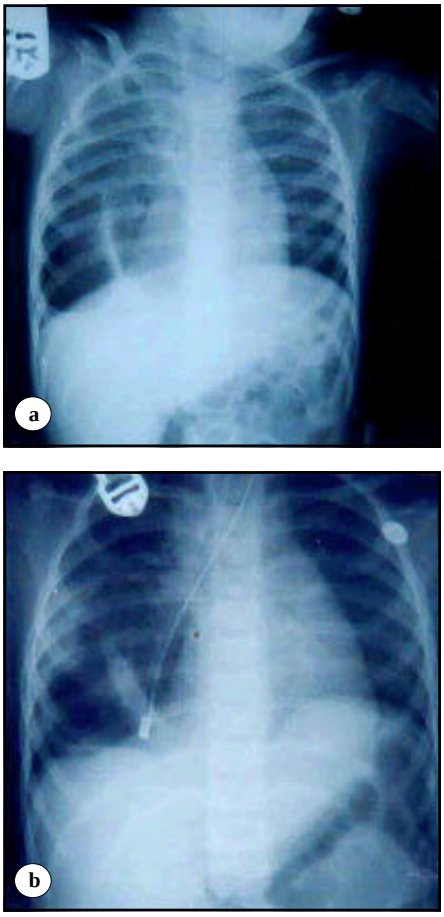


Figure 1 - Plain chest radiograph showing **(a)** (postero-anterior view) a large double cystic mass in the right middle and lower lung zones. The gastric air bubble is absent from under the left hemi diaphragm. **(b)** A nasogastric tube inserted into the esophagus fails to pass into the stomach.

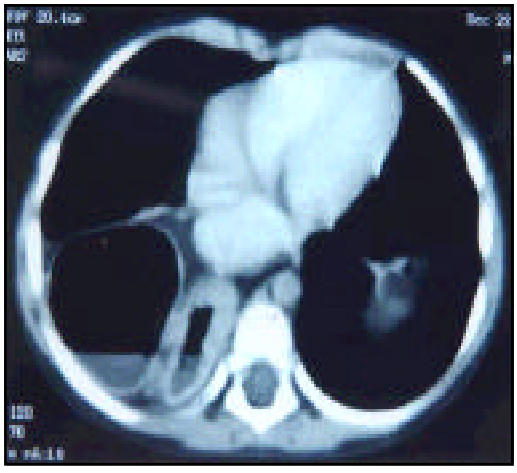

Figure 2 - Computed tomography scan of the chest showing 2 cystic cavities in the right middle and lower zones posteriorly.

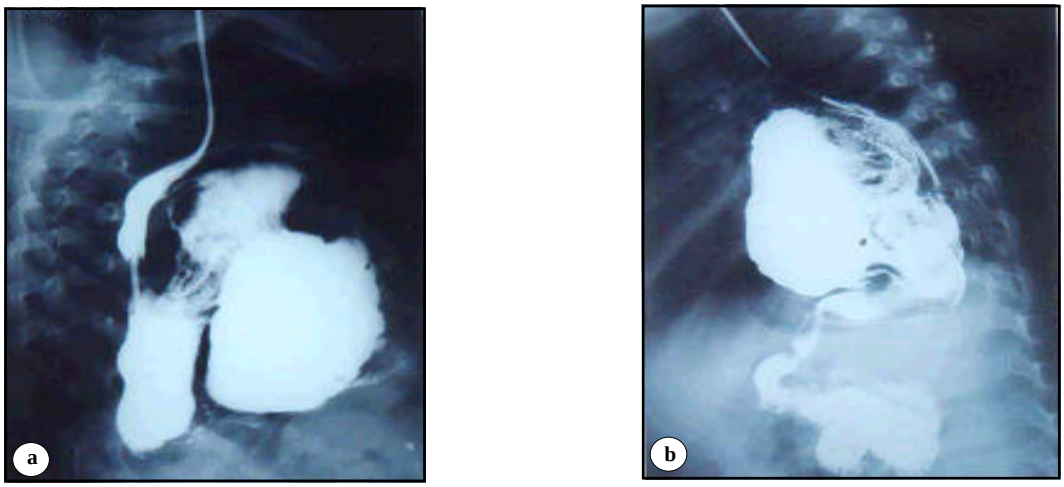


Figure 3 - Barium study: **(a)** (right lateral view) carried out through a nasogastric tube placed in the esophagus. There is a right intrathoracic stomach with organo-axial torsion (upside down stomach). **(b)** Barium flowed freely into the duodenum and jejunum.

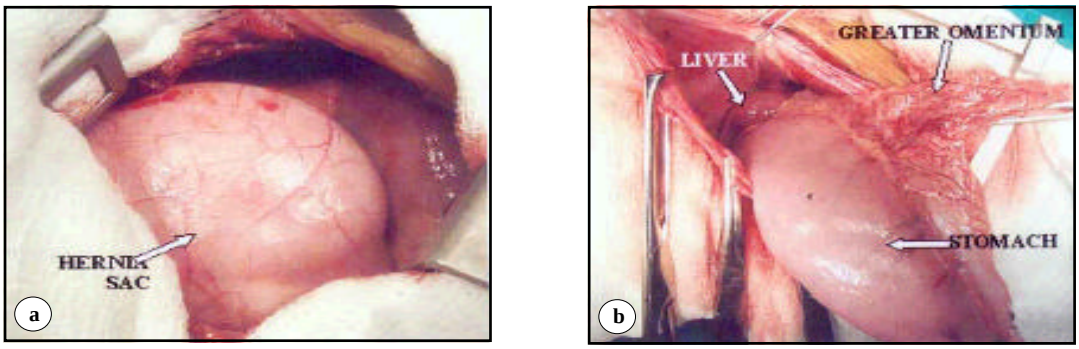


Figure 4 - Intraoperative picture of: **(a)** the right pleural cavity showing a large hernia sac. **(b)** The hernia sac contained the stomach, greater omentum and a part of the liver.

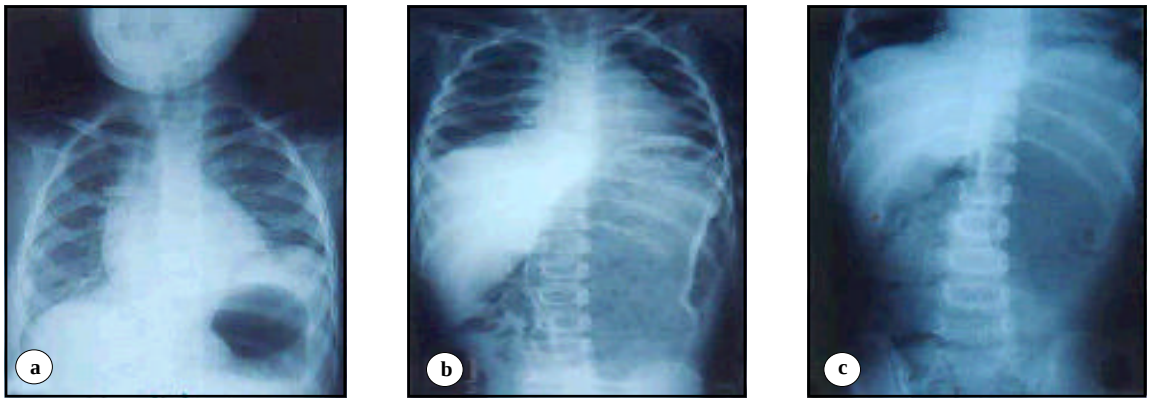


Figure 5 - Postoperative plain radiographs of the chest **(a)** and chest/abdomen **(b)** showing clear lung fields, full expansion of the right lung and gastric dilatation. **(c)** Plain chest and abdominal radiograph (supine) showing marked reduction in the side of the stomach by the 12th postoperative day.

Table 1 - Clinico-anatomical classification of Bochdalek hernias.

| Types | Anatomical Features | Clinical Features |
|---|---|---|
| 1 A | No hernia sac is present.<br>Right or left posterolateral diaphragmatic defect.<br>Pulmonary hypoplasia of the ipsilateral and contralateral lungs.<br>Reduced lung volume.<br>Impaired bronchial development and branching.<br>Reduced alveolar number and size.<br>Thickening of the muscular layer of the arteriolar vasculature<br>Decrease in the number of vessels per unit lung. | Patients present acutely in the neonatal period due to compression of the ipsilateral lung and maldevelopment of the affected lung.<br>Vomiting may be due to organo-axial torsion.<br>Hollow viscera herniation less frequent on the right.<br>Mortality is high. |
| 1 B | Hernia sac is present.<br>Right or left posterolateral diaphragmatic defect.<br>Pulmonary hypoplasia of the ipsilateral and contralateral lungs.<br>Reduced lung volume.<br>Impaired bronchial development and branching.<br>Reduced alveolar number and size.<br>Thickening of the muscular layer of the arteriolar vasculature.<br>Decrease in the number of vessels per unit lung. | Patients present acutely in the neonatal period due to compression of the ipsilateral lung and maldevelopment of the affected lung.<br>Hollow viscera herniation less frequent on the right.<br>Vomiting may be due to organo-axial torsion.<br>Mortality is high. |
| 2 A | No hernia sac is present.<br>Right or left posterolateral diaphragmatic defect.<br>Normally developed lungs. | Delayed herniation or very small hernias may present late.<br>Vomiting may be due to organo-axial torsion.<br>Hollow viscera herniation less frequent on the right.<br>Outcome of surgical repair is excellent. |
| 2 B | Hernia sac is present.<br>Right or left posterolateral diaphragmatic defect.<br>Normally developed lungs. | Delayed herniation or very small hernias may present late.<br>Vomiting may be due to organo-axial torsion.<br>Hollow viscera herniation less frequent on the right.<br>Outcome of surgical repair is excellent. |

diagnosis was an intrathoracic cystic lesion. A barium study **(Figure 3)** revealed a right intrathoracic stomach with organo-axial torsion (upside down stomach). The patient underwent a right lateral thoracotomy through the bed of the fifth rib at which there was a posterolateral hernia sac, which contained a grossly dilated stomach, greater omentum and part of the liver **(Figure 4)**. There were adhesions between the hernia sac and the right lower lobe of the lung. The sac was resected; the sac contents were reduced into the abdominal cavity after a partial Nissen's fundoplication and gastric fundopexy to the underside of the diaphragm lateral to the left rim of the diaphragmatic defect. The edges of the diaphragmatic defect were well developed. The diaphragmatic defect was closed with a double layer of interrupted 0-Prolene sutures (Mayo-type repair). Postoperatively, the patient was admitted into the Pediatric Intensive Care Unit for 24 hours for cardio-respiratory monitoring. She did not require mechanical ventilation, and she was discharged to the pediatric ward with stable vital signs. Oral feeds were gradually introduced from the second postoperative day and were well tolerated. There was no dysphagia or vomiting. A chest radiograph was carried out immediately after surgery. The gastric dilatation noted postoperatively **(Figures 5a & 5b)** started to regress postoperatively **(Figure 5c)**, and the child was discharged from the hospital on the 12th postoperative day without any medications.

Discussion. Bochdalek hernias usually present in the neonatal period with severe respiratory distress due to the herniation of abdominal organs into the pleural cavity, and the mal-development of the lung on the affected side.[1-3] In patients who present late it is thought that the herniation of abdominal viscera occurs later in life[5-7,14] as was the case reported here.

Our patient was misdiagnosed initially as a case of bronchial asthma and given asthmatic treatment with a poor response, though the patient did not have a personal or family history of atopy, there was no wheeze on chest auscultation and there were crepitations and decreased air entry in the right lower zone of the chest. In patients with Bochdalek hernia who present late, misdiagnosis as tension pneumothorax,[1] hemothorax, pneumonia, pulmonary cyst, foregut duplication cyst, or pulmonary sequestration have been reported.[6,7,12,13] The history and physical findings in our patient pointed to the need to consider other differential diagnoses rather than asthma, the child should have had at least a chest x-ray at her initial presentation. This would have pointed to the need for an earlier referral to

ACH, which would have saved the child from having to take long-term asthma medications. Previous chest radiographs from the referring hospital, carried out just prior to referral, showed a cystic mass in the right chest, thus, alerting the physician to an intrathoracic pathology.

There is a consensus that Bochdalek hernias occurring on the left side are best approached through the abdomen,[6,15-17] but right hernias, especially with a herniated stomach, are best approached through a right thoracotomy.[18] The presence of adhesions between the hernia sac and the lung justify this approach in our patient. Laparoscopic[2] and thoracoscopic[18] in older children and adults have been documented. Yamaguchi et al,[18] mentioned the presence of adhesions as a contraindication for thoracoscopic repair. This finding also precludes the abdominal approach.

A hernia sac, such the one present in our patient is not rare in Bochdalek hernia. Thomas and Kapur[1] reported the presence of a hernia sac in 9 out of 34 patients reviewed (26.5%). Organo-axial torsion has also been reported[18] in Bochdalek hernia. In cases when the stomach herniates through the hernia defect, the stomach may be rotated along a horizontal axis giving rise to the so called upside down stomach,[18] as seen in our patient. Organo-axial torsion may lead to hollow viscus ischemia, necrosis, or obstruction which may precipitate vomiting.[7] Intrathoracic gastric dilatation has been reported in Bochdalek hernias.[16,17] This has sometimes been misdiagnosed as tension pneumothorax, and on this basis managed by thoracentesis and tube thoracostomy with resulting perforation of the dilated intrathoracic stomach.[6,16] From a review of cases, Bochdalek hernias reported in the literature, we propose a new classification of the condition considering the variations in presentation and clinico anatomical features **(Table 1).**

In conclusion, we report a rare case of right-sided Bochdalek hernia with a right intrathoracic stomach and trans-axial torsion presenting beyond the neonatal period with recurrent chest symptoms and intermittent vomiting, which were misdiagnosed initially as chronic hyperactive airway disease. We recommend that physicians should have a high index of suspicion and early, appropriate investigations such as barium swallow if child presents with persistent respiratory and gastrointestinal symptoms. This may reveal the appropriate diagnosis, and the subsequent management prompt surgical intervention, thus, the child would be saved from the side effects of unnecessary medications of inappropriate clinical diagnosis.

## References


1. Thomas S, Kapur B. Adult Bochdalek hernia: clinical features, management, and results of treatment. ***Jpn J Surg*** 1991; 21: 114–119.
2. Al-Emadi M, Helmy I, Abu Nada M, Al-Jaber H. Laparoscopic repair of Bochdalek hernia in an adult. ***Surg Laparosc Endosc*** 1999; 9: 423–425.
3. Mullins ME, Stein J, Saini SS, Mueller PR. Prevalence of incidental Bochdalek's hernia in a large adult population. ***AJR Am J Roentgenol*** 2001; 177: 363-366.
4. Moore KL, Persaud TVN. The Developing Human: Clinically Oriented Embryology. 6th ed. Philadelphia (PA): WB Saunders; 1998. p. 206–211.
5. Hosgor M, Karaca I, Karkiner A, Ucan B, Temir G, Erdag G, et al. Associated malformations in delayed presentation of congenital diaphragmatic hernia. ***J Pediatr Surg*** 2004; 39: 1073-1076.
6. Dalton AM, Hodgson RS, Crossley C. Bochdalek hernia masquerading as a tension pneumothorax. ***Emerg Med J*** 2004; 21: 393-394.
7. Niwa T, Nakamura A, Kato T, Kutsuna T, Tonegawa K, Kawai A, et al. An adult case of Bochdalek hernia complicated with hemothorax. ***Respiration*** 2003; 70: 644-646.
8. Sener RN, Tugran C, Yorulmaz I, Dagdeviren A, Orguc S. Bilateral large Bochdalek hernias in an adult. CT demonstration. ***Clin Imaging*** 1995; 19: 40–42.
9. Falconer AR, Brown RA, Helms P, Gordon I, Baron JA. Pulmonary sequelae in survivors of congenital diaphragmatic hernia. ***Thorax*** 1990; 45: 126-129.
10. Nakayama DK, Mutich R, Motoyama EK. Pulmonary dysfunction in surgical conditions of the newborn infant. ***Crit Care Med*** 1991; 19: 926-933.
11. Levin DL. Morphologic analysis of the pulmonary vascular bed in congenital left-sided diaphragmatic hernia. ***J Pediatr*** 1978; 92: 805-809.
12. Ng CP, Lo CB, Chung CH. Congenital diaphragmatic hernia masquerading as pneumonia. ***Emerg Med Australas*** 2004; 16: 167-169.
13. Kaur G, Singh T. Right-sided diaphragmatic hernia masquerading as staphylococcal pneumonia. ***Indian J Pediatr*** 2003; 70: 743-745.
14. Gheibi S, Mahmoodzadeh A. Delayed presentation of right-sided diaphragmatic hernia after mechanical ventilation in a newborn with hyaline membrane disease: A case report. ***Medical Journal of Islamic Academy of Sciences*** 2001; 14: 21-24.
15. Vukic Z. Left Bochdalek hernia with delayed presentation: Report of two cases. ***Croat Med J*** 2001; 42: 569-571.
16. Quah BS, Hashim I, Simpson H. Bochdalek diaphragmatic hernia presenting with acute gastric dilatation. ***J Pediatr Surg*** 1999; 34: 512-514.
17. Goldenberg IS. Acute intrathoracic gastric dilatation associated with Bochdalek hernia in an infant. ***AMA J Dis Child*** 1957; 93: 548-550.
18. Yamaguchi M, Kuwano H, Hashizume M, Sugio K, Sugimachi K, Hyoudou Y. Thoracoscopic treatment of Bochdalek hernia in the adult: Report of a case. ***Ann Thorac Cardiovasc Surg*** 2002; 8: 106-108.