

Bochdalek Hernia: A Comprehensive Review of Pathophysiology, Clinical Presentation, Diagnostic Modalities, and Contemporary Management Strategies

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ABSTRACT

Bochdalek hernia, a rare congenital diaphragmatic anomaly, has garnered substantial attention due to its intricate pathophysiological underpinnings and multifaceted clinical implications. This article endeavors to provide an exhaustive analysis of Bochdalek hernia, encompassing its embryological origins, intricate pathophysiology involving the posterolateral diaphragmatic defect, and the subsequent herniation of abdominal contents into the thoracic cavity. A meticulous exploration of the clinical presentation underscores the considerable variation in symptomatology, ranging from asymptomatic cases incidentally detected on imaging, to severe respiratory distress and life-threatening consequences in neonates.

The diagnostic armamentarium, including radiographic, ultrasonographic, and tomographic modalities, is scrutinized with a focus on their respective sensitivities and specificities in discerning this anatomical aberration. In light of contemporary advancements, the article elucidates the expanding role of three-dimensional reconstructions and minimally invasive techniques in achieving accurate diagnosis and surgical planning.

Furthermore, the management landscape is delineated, encompassing conservative approaches for asymptomatic cases, urgent surgical interventions for acute presentations, and the nuanced perioperative considerations for neonates and pediatric patients. A comprehensive overview of surgical techniques, ranging from traditional open procedures to the evolving realm of robotic-assisted and laparoscopic repairs, is provided, with emphasis on individualized strategies tailored to patient-specific factors.

In summation, this article offers a comprehensive and sophisticated exploration of Bochdalek hernia, amalgamating embryology, pathophysiology, clinical presentation, diagnostic modalities, and contemporary management paradigms. It is envisaged that this review will serve as a valuable resource for medical practitioners, surgeons, and researchers, fostering an enriched understanding of this intricate condition and facilitating informed decision-making in its management.

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INTRODUCTION

Bochdalek's hernia, also known as congenital posterolateral diaphragmatic hernia or pleuroperitoneal hernia, is a developmental defect in the posterolateral diaphragm caused by a lack of closure of the pleuroperitoneal cavity due to incomplete development of the diaphragm during the 8th to 10th week of pregnancy, before the intestine returns from the yolk sac to the abdomen. This defect was first

described in 1848 by the Czech anatomist and pathologist, Vincent Alexander Bochdalek (1801–1883). 1,2,3

Bochdalek hernia (BH) usually presents on the left side in 85% of cases. Right-sided hernia are rare (13% of cases), since the right pleuroperitoneal canal closes earlier and the liver supports the right diaphragm, minimizing the opportunity for herniation into the right thoracic cavity There are also descriptions in the literature of bilateral located hernias (2%). 4,5

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Bochdalek hernia is a type of diaphragmatic hernia commonly diagnosed in neonatal and postnatal patients with acute pulmonary symptoms. When the BH goes undiagnosed during the antenatal period, it is usually detected within 1 month after birth. The clinical manifestation of symptoms and diagnosis in adults is rare, with the majority of cases found incidentally.^{6,7}

Symptoms manifest differently in children and adults. BH in children is characterized by cyanosis, tachycardia, respiratory failure, asymmetric growth in the chest cavity, pulmonary hypertension with a risk of congenital developmental anomalies in 10–15%. Adults tend to present with vague or nonspecific complaints such as pleural effusion, chest pain, dyspnea, abdominal pain, dysphagia, and postprandial fullness. Compared with the acute manifestations of neonatal BH, most patients with adult BH had more chronic symptoms, 14 % of adult BHs are asymptomatic.^{8,9,10}

Diagnosis can be made with a simple chest X-ray, CT scan, MRI, or upper GI series. On a chest radiograph, BH may appear as gas and fluid-filled viscera or as a pleural effusion. The gold-standard for diagnosis of BH remains a double-contrast axial CT. The presence of a soft tissue contour in the chest CT, in addition to opaque, filled, dilated bowel segments above the diaphragm, establishes definitive diagnosis. CT was reported to have sensitivities of 78% for left-sided hernias and 50% for right-sided hernias. ^{11,12}

The surgical repair is recommended in both symptomatic and asymptomatic adult patients with BH to prevent life-threatening complications due to the incarceration of hernia, obstruction, strangulation, and perforation. For surgical treatment of BH, the transthoracic, transabdominal and combined thoracoabdominal approaches have been described. ¹³

The laparotomy facilitates the intra-abdominal manipulation of reduced abdominal organs, observation of perfusion defects in prolapsed organs, and management of injury to the reduced organs. Minimal invasive approaches by were also reported and provide better visualization and working space than an open approach especially when the right hepatic lobe obstructs view in right-sided BH. There is no established consensus for choosing an approach, therefore surgical treatment needs to be individualized based on findings on CT imaging. ^{14,15}

During the procedure, contents are reduced to the peritoneal cavity, and the diaphragmatic defect is closed using non-absorbable sutures with implantation of a prosthetic mesh, or use of a muscle flap to construct a repair that is reinforced because of the continuing stress on the diaphragm that results from respiratory movements. However, if the diaphragm defect is not too large, it may be better to repair the diaphragm by direct suturing to avoid infection and postoperative adhesions. ¹⁶

EPIDEMIOLOGY

Congenital diaphragmatic hernia (CDH) is a congenital abnormality of the diaphragm that affects approximately 1 in 2000 to 1 in 3000 births. BH is the most frequent CDH, affecting 1 in 2,200 to 2,500 live births with a male to female ratio of 3:1. There are currently fewer than 100 cases of Bochdalek hernias reported in adults in the literature, and only about 20 cases involving right-sided hernias.^{14,15}

The true prevalence of BH in adults remains unknown, with estimates ranging from 0.17% to 12.7% based on reviews of imaging studies. Mullins et al. reported a review of 13138 abdominal CT reports that the incidence of adult BH was 0.17 %, with 68 % being right-sided, 18% were on the left, and 14% were bilateral and 77 % of patients being female with a ratio of 17:5, mean age was 66.6. ¹⁶

Ramspott et al., in their systematic review showed that Peak ages for right-sided Bochdalek hernias were 40 to 50 and 70 to 80 years, 61% of the patients were women, 39% were men. This suggests right sided BH are also under reported due to lack of clinical symptoms. ¹⁷

On the other hand Misaka et al., in their literature review from 1999 to 2019 in Japan, showed that adult BH has a female predominance, and 60% of patients were female, with a mean age of 58 years; 56% of patients were above 60 years and 29% of patients were aged 70–80 years. These findings may suggest that adult BH is more common in elderly persons. ¹⁸

PATHOPHYSIOLOGY

In general, the pathophysiology of Bochdalek's hernia is poorly understood. The human diaphragm begins to form in multiple folds at about 4 weeks of gestation. The central tendon region of the diaphragm develops from the septum transversum, which separates the thoracic and abdominal cavities. Posterolateral infoldings form the pleuroperitoneal membranes. The pleuroperitoneal canal, in turn, communicates between the pleura and the peritoneal cavity through the foramen of Bochdalek in the posterior part of the developing diaphragm. This communication closes around week 8 of gestation. Failure to close this foramen allows a BH to develop. ^{17,18}

Given the possibility of an acquired diaphragmatic hernia, it is difficult to determine the exact cause of Bochdalek hernia in adults. It can be associated with several genetic syndromes with chromosomal abnormalities, such as Cornelia de Lange syndrome and Pallister-Killian syndrome. ^{17,18}

An adult Bochdalek hernia is often precipitated by conditions of increased intra-abdominal pressure, such as physical exertion (including sexual intercourse), pregnancy, childbirth, sneezing or coughing, surgery under the pneumoperitoneum, or even heavy meals. In addition, adult BH is believed to be closely related to body mass index. ^{18,19}

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Adults typically develop it due to late rupture of the peritoneal sac containing the viscera or plugging of the diaphragmatic defect by a solid organ and subsequent herniation after increased intra-abdominal pressure, brought on by conditions like obesity and pregnancy. 19

CASE PRESENTATION

An 80-year-old female presented to the emergency room with complaints of a history of abdominal pain and chronic constipation. She had a previous medical history of systemic arterial hypertension and chronic kidney disease, stage 4. She was medicated with olanzapine and fluoxetine due to anxiety and depression diagnosed a few years earlier. She had a history of cataract surgery and no history of upper abdominal surgery. She had fallen several times from a standing height, but never suffered a serious chest or abdominal trauma.

She had 5 months of history of chronic constipation, worsening over the last month and relieved with home remedies. She also complained of mild left-sided chest pain, 1 month prior to the admission, being medicated with NSAID with a complete relief. Four days before the hospitalization she presents to emergency room with generalized abdominal pain, nausea and vomiting of gastro-alimentary content. She received medical treatment with omeprazole and metimazole with improvement of the symptoms and discharge home with painkillers therapy.

Four days later, she returned to the emergency room with severe abdominal pain. The pain was generalized, accompanied by abdominal distension, and inability to defecate and unable to lie flat. No vomiting and nausea were associated. On arrival vital signs were stable. Abdominal examination revealed a slightly distended abdomen with decreased bowel sounds. Mild right upper quadrant pain was present with aggravation on deep palpation. A nasogastric tube was placed draining gastrobiliary content. First lab investigation showed leukocyte count 7,900 cells/mm³ (neutrophils: 82.2%, lymphocytes:10.6%), serum creatinine 1.08 mg/dL, serum sodium 137 mEq/L, serum potassium 4.03 mEq/L and serum chloride 98 mEq/L.

Chest X-ray was performed and revealed intestinal gas over the liver and an elevated right hemi-diaphragm (Fig. 1) meanwhile an abdominal x-ray showed gas-fluid levels (Fig. 2).



Fig. 1. Chest X-ray shows intestinal gas over the liver and an elevated right hemidiaphragm (white arrow).



Fig. 2. Abdominal X-ray showing gas-fluid levels.

CT scan detected a continuity defect in the posterolateral right hemidiaphragm showing slippage of the ascending colon loop with air-fluid level and secondary dilatation of all the loops (Fig. 3, 4 and 5). These findings were suggestive of the Bochdalek hernia.

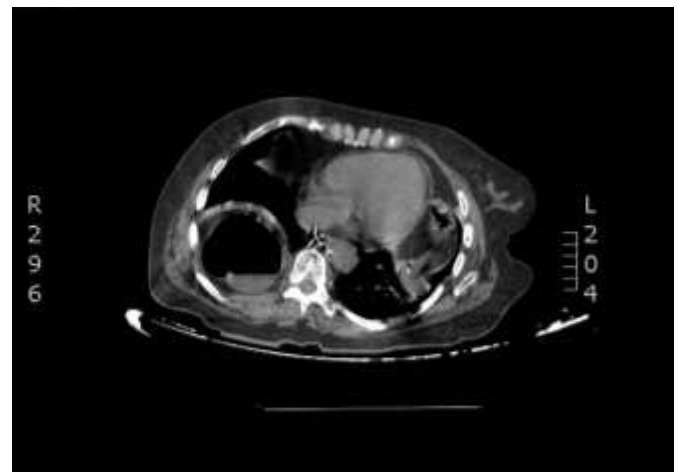


Fig. 3 Axial CT-scan showing a posterolateral defect in the right hemidiaphragm and the presence of herniated colon in the right thoracic cavity.



Fig. 4 Coronal CT-scan revealed herniation of the ascending colon.

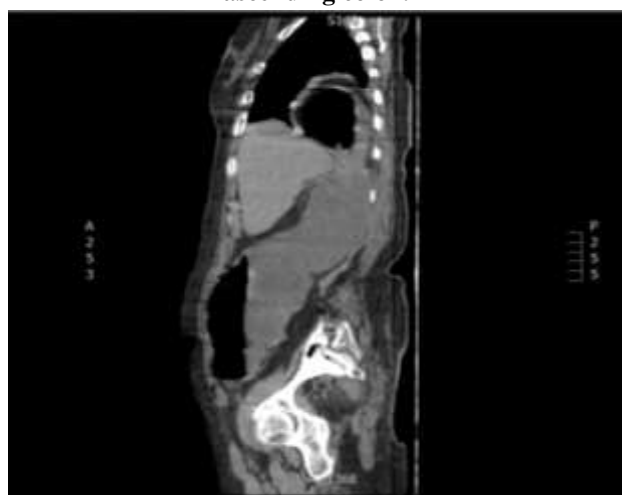


Fig. 5 Sagittal CT-scan showing ascending colon loop passing through a defect in the posterolateral right hemidiaphragm.

The patient underwent to transabdominal open surgical repair using an enlarged Kocher-type incision. A 5 cm hernia orifice was found in the right posterolateral diaphragm with a 10x5x5 cm hernia sac containing ascending and transverse colon loop with multiple adhesions to the sac (Fig. 6,7). There were no ischemic changes. Adhesions were released and the herniated contents were reduced to the peritoneal cavity. The hernia defect was closed primarily with 3 interrupted prolene stitches (Fig. 8). A chest tube was placed due to the pneumothorax caused by the surgical dissection. The chest tube is removed 2 days later. The patient was sent home on the fourth post-op day following an uneventful recovery.

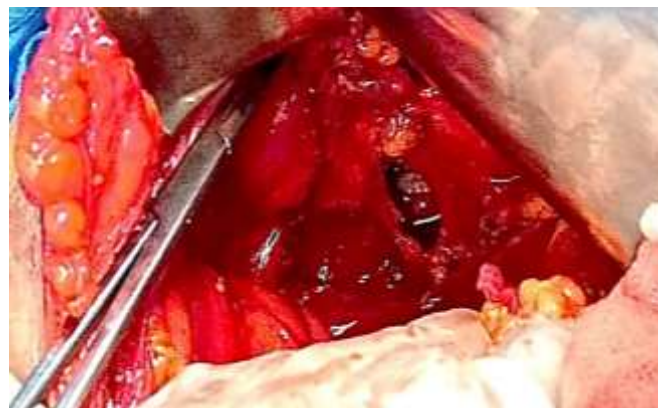


Fig. 6. Intraoperative vision of the wall defect.



Fig. 7. Hernia sac containing ascending and transverse colon loop with multiple adhesions to the sac.



Fig. 8. Diaphragm defect repair with interrupted prolene stitches (white arrow).

CONCLUSIONS

In conclusion, the intricate tapestry of Bochdalek hernia, with its intricate embryological origins, multifaceted pathophysiological mechanisms involving posterolateral diaphragmatic defects, and the subsequent herniation of abdominal viscera into the thoracic cavity, underscores the need for heightened clinical vigilance and diagnostic acumen. The considerable heterogeneity in clinical presentation, ranging from latent asymptomatic cases to emergent scenarios marked by severe respiratory compromise, mandates a nuanced approach to diagnosis and management.

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The diagnostic landscape, replete with radiographic, ultrasonographic, and tomographic modalities, has witnessed evolving sophistication, wherein the amalgamation of traditional imaging paradigms with cutting-edge three-dimensional reconstructions has expanded diagnostic precision. This contemporary diagnostic refinement synergizes with an increasingly diverse armamentarium of surgical interventions, from classical open repairs to minimally invasive techniques, necessitating tailored approaches aligned with patient-specific parameters.

The synthesis of embryological insights, mechanistic understanding, and clinical acumen has engendered a paradigm shift in the management ethos, affirming the indispensability of individualized strategies in optimizing patient outcomes. As technological innovation continues to advance, it is anticipated that the diagnostic and therapeutic landscape of Bochdalek hernia will further evolve, presenting both challenges and opportunities to the medical community.

Ultimately, the comprehensive investigation presented in this article serves not only to deepen our comprehension of Bochdalek hernia, but also to underscore the importance of interdisciplinary collaboration among clinicians, radiologists, and surgeons in addressing this intricate congenital anomaly. By leveraging a sophisticated understanding of its complexities, clinicians are better poised to navigate the diagnostic intricacies and curative nuances of Bochdalek hernia, thus elevating the standard of care for affected individuals across the lifespan.

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