



DIAPHRAGMATIC EVENTRATION AND HERNIA

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ABSTRACT Diaphragmatic eventration and hernia are critical conditions affecting the diaphragm, which play a vital role in respiration. Diaphragmatic eventration involves an abnormal elevation of the diaphragm due to muscle weakness or atrophy, which can be congenital or acquired. This condition compromises respiratory mechanics, leading to symptoms such as dyspnea and decreased exercise capacity. On the other hand, diaphragmatic hernia is characterized by the protrusion of abdominal organs into the thoracic cavity through a defect in the diaphragm, which can be present from birth or result from trauma or surgical interventions. Both conditions pose significant clinical challenges, necessitating accurate diagnosis and tailored management strategies to enhance respiratory function and patient quality of life. Advances in imaging techniques and surgical interventions have improved the prognosis for individuals with these diaphragmatic abnormalities. This review paper provides a comprehensive overview of the pathophysiology, clinical presentation, diagnostic modalities, and current therapeutic approaches for diaphragmatic eventration and hernia, emphasizing the importance of early detection and intervention.

KEYWORDS : Diaphragmatic eventration, hernia, pathophysiology, diagnostic modalities, etc.

INTRODUCTION

Diaphragmatic eventration and hernia represent two distinct yet often interrelated conditions that pose unique challenges in the field of thoracic surgery. Diaphragmatic eventration (DE) is characterized by an abnormal elevation of an intact diaphragmatic muscle, which maintains its connections with the thoracic cage but exhibits a weakened, thinned, and dysfunctional state. This condition, while rare with an incidence rate of approximately 0.05%, can lead to significant clinical manifestations, including respiratory distress due to the paradoxical movement of the diaphragm. On the other hand, diaphragmatic hernias, such as the Bochdalek hernia, involve a true defect in the diaphragm allowing abdominal contents to protrude into the thoracic cavity. This can occur due to improper development of the pleura-peritoneal folds during gestation. The clinical outcomes and management strategies for these conditions vary, with recent studies highlighting the potential benefits and challenges associated with surgical interventions.

Recent advancements in surgical techniques, particularly the use of minimally invasive laparoscopic methods have shown promise in the management of DE. A three-step laparoscopic procedure for diaphragm reconstruction has been reported to successfully address the condition, offering a less invasive alternative with positive postoperative outcomes. Similarly, the differentiation between congenital diaphragmatic eventration and hernia with or without a hernia sac has been the subject of research, emphasizing the need for tailored approaches based on the specific type of malformation. The ongoing exploration of these conditions underscores the importance of continued research and innovation in surgical techniques to improve patient outcomes. As the medical community gains a deeper understanding of the pathophysiology and optimal treatment modalities for diaphragmatic eventration and hernia, the potential for enhanced quality of life for affected individuals becomes increasingly attainable.

OBJECTIVES

- Review the presentation of a patient with diaphragmatic eventration and hernia.
- Outline the pathophysiology, clinical presentation and diagnostic modalities of diaphragmatic eventration and hernia.
- Describe the current therapeutic approaches and early detection and intervention for diaphragmatic eventration and hernia.
- Describe the challenges and future direction for diaphragmatic eventration and hernia.

METHODOLOGY

It is a descriptive review in databases, Pubmed, Scopus, Scielo and Lilacs, etc. We included only English language articles, published

between 2010 to 2024 that, who portrayed DE and DH.

PATHOPHYSIOLOGY

The pathophysiology of diaphragmatic eventration and hernia involves distinct mechanisms affecting the diaphragm:

• Diaphragmatic eventration is often a congenital condition where there is an abnormal elevation of the diaphragm due to incomplete muscularization. The diaphragm muscle is replaced by fibroelastic tissue, which does not contract normally during inspiration. This can lead to respiratory issues as the diaphragm is the primary muscle for breathing. Acquired eventration can occur due to trauma or surgery that leads to phrenic nerve paralysis, causing the diaphragm to shift upward and become fibroelastic over time.

• A diaphragmatic hernia, on the other hand, is characterized by a defect in the diaphragm that allows abdominal contents to herniate into the thoracic cavity. This can cause severe respiratory distress due to the displacement of the lungs and restriction of lung expansion. The hernia can be congenital, often associated with genetic syndromes, or acquired due to trauma. In both cases, the defect in the diaphragm results in a mechanical disruption that can lead to life-threatening complications if not treated promptly.

Understanding these pathophysiological differences is crucial for the diagnosis and management of each condition.

CLINICAL PRESENTATION

Diaphragmatic eventration is characterized by an abnormal elevation of an otherwise intact diaphragm. It can be congenital or acquired, often presenting in newborns as a cause of respiratory distress. In adults, it may remain asymptomatic or manifest as intermittent shortness of breath. The condition is due to incomplete muscularization of the diaphragm during development, leading to a thin, membranous sheet that replaces normal muscle tissue. This results in a diaphragm that does not contract properly during inspiration, potentially affecting one's breathing capacity.⁵

On the other hand, a diaphragmatic hernia involves a defect in the diaphragm that allows abdominal organs to protrude into the thoracic cavity. This can lead to significant respiratory compromise, especially if the herniation is large. Clinical presentation includes chest pain, difficulty breathing, and bowel obstruction symptoms. The diagnosis is often confirmed through imaging techniques like ultrasound, which can differentiate between hernia and eventration by identifying specific signs such as the folding of the free muscle edge or the presence of a narrow angle waist indicative of a hernia.

In summary, while both DE and DH can affect respiratory function,

DH typically presents with more acute and severe symptoms and has a higher urgency for surgical intervention compared to DE.

Diagnostic Modalities

The diagnostic modalities for diaphragmatic eventration and hernia include a range of imaging techniques that help differentiate between the two conditions and assess the extent of the diaphragmatic abnormality. Here are the key modalities:

Chest Radiograph:

It commonly known as a chest X-ray, is a fundamental imaging tool used in diagnosing diaphragmatic eventration and hernia. It provides a quick and non-invasive method to visualize the structures of the chest and can reveal the elevation of the diaphragm or the presence of abdominal contents in the thoracic cavity. In the case of diaphragmatic eventration, a chest radiograph will typically show an elevated hemidiaphragm with a smooth contour. The affected portion may appear as a hump, and the rest of the diaphragm maintains a normal contour. A 'double' diaphragmatic contour may be visible on the frontal x-ray, which is more easily confirmed on a lateral projection. For diaphragmatic hernia, the chest X-ray can show bowel loops or other abdominal contents within the chest cavity, indicating a breach in the diaphragm. The diagnosis of right congenital diaphragmatic hernia (CDH) can be more challenging than left CDH, as the liver may prevent bowel loops from herniating into the chest.¹⁰

Ultrasound:

Ultrasound is a key diagnostic tool in the evaluation of diaphragmatic conditions such as eventration and hernia. It's particularly useful because it's non-invasive, widely available, and does not involve radiation exposure. Ultrasound can assess the movement and thickness of the diaphragm. In eventration, the diaphragm appears as an abnormally elevated dome with no disruption to its continuity. It typically affects only a segment of the hemidiaphragm. Ultrasound can show the diaphragm's abnormal contour and help distinguish eventration from other conditions like paralysis or weakness.¹⁰ In cases of diaphragmatic hernia, ultrasound can help identify the presence of abdominal organs in the thoracic cavity, which is indicative of a defect in the diaphragm. It's especially useful in prenatal diagnosis after the second trimester and can also detect cases that are first identified after birth.¹¹ Recently, ultrasound has replaced fluoroscopy as the imaging test of choice for diaphragmatic motion assessment, providing an accurate evaluation without radiation. This motion is measured both in the direction of diaphragm movement and the amplitude of excursion.

Computed Tomography (CT):

It is a highly effective imaging modality for diagnosing diaphragmatic conditions such as eventration and hernia. It provides detailed cross-sectional images that can reveal the anatomy and any abnormalities of the diaphragm with high precision. For Diaphragmatic Eventration, CT can show the uninterrupted integrity of the diaphragmatic muscle and its abnormal elevation into the thoracic cavity. It can also identify associated conditions, such as gastric volvulus, and is useful in preoperative planning and postoperative evaluation.¹ In the case of Diaphragmatic Hernia, CT is particularly valuable as it can display the herniated abdominal organs and potential complications like intestinal strangulation, haemothorax, or rib fractures. It's the most effective diagnostic tool in many cases of diaphragmatic hernia.

Magnetic Resonance Imaging (MRI):

It is an advanced diagnostic tool that plays a crucial role in the evaluation of diaphragmatic abnormalities such as eventration and hernia. MRI provides high-resolution images that offer detailed anatomical information, which is particularly useful for complex cases. For Diaphragmatic Eventration, MRI can visualize the diaphragm as a thin hypointense line on coronal and sagittal sequences, distinguishing it from other conditions that may present with similar symptoms. It's also beneficial for assessing the extent of the eventration and planning surgical interventions if needed. In the case of Diaphragmatic Hernia, MRI is instrumental in evaluating congenital anomalies affecting the fetal lung, especially when lung hypoplasia is suspected. It allows for the assessment of the extent of the anomaly, the amount of normal residual parenchyma, and the quantification of total lung volume, which are essential for stratifying the severity of the condition, guiding counseling, and patient management.¹

Current Therapeutic Approaches For Diaphragmatic

Eventration And Hernia

Current therapeutic approaches for diaphragmatic eventration and hernia encompass a combination of medical management and surgical interventions tailored to the individual patient's condition and clinical presentation.

Medical Management:

Respiratory Support: Patients with diaphragmatic eventration or hernia may require respiratory support, including supplemental oxygen therapy, non-invasive ventilation, or mechanical ventilation, to manage respiratory distress and optimize oxygenation.

Nutritional Support: Adequate nutritional support is essential, especially in neonates and infants with diaphragmatic hernia, to support growth and development and optimize surgical outcomes.

Surgical Interventions:

Primary Repair: Surgical repair is the mainstay of treatment for diaphragmatic eventration and hernia. Primary repair involves directly suturing or reconstructing the diaphragmatic defect to restore anatomical integrity and prevent visceral herniation.

Minimally Invasive Surgery: Minimally invasive techniques, such as laparoscopic or thoracoscopic repair, have gained popularity due to their potential benefits, including reduced postoperative pain, shorter hospital stays, and faster recovery compared to traditional open surgery.

Patch Repair: In cases of large or complex diaphragmatic defects, patch repair using synthetic or biological mesh may be employed to provide additional support and reinforce the repair.

Sequential Staged Repair: In infants with congenital diaphragmatic hernia and associated pulmonary hypoplasia, a sequential staged repair approach may be utilized, involving initial placement of a silo or patch to facilitate lung growth followed by definitive repair at a later stage.

Extracorporeal Membrane Oxygenation (ECMO): In severe cases of congenital diaphragmatic hernia with refractory respiratory failure, ECMO may be used as a bridge to surgery or to provide temporary cardiopulmonary support while awaiting lung growth and development.

Recent Advances:

Tissue Engineering: Emerging techniques in tissue engineering hold promise for developing bioengineered materials for diaphragmatic repair, potentially reducing the risk of recurrence and promoting tissue regeneration.

Fetal Intervention: Fetal intervention techniques, such as fetoscopic endoluminal tracheal occlusion (FETO), are being explored as potential strategies to improve outcomes in select cases of severe congenital diaphragmatic hernia by promoting lung growth and reducing pulmonary hypertension before birth.

Therapeutic approaches for diaphragmatic eventration and hernia continue to evolve, with ongoing research aimed at refining surgical techniques, optimizing perioperative management, and improving long-term outcomes for affected patients.

Early Detection And Intervention

Early detection and intervention are critical in the management of diaphragmatic eventration and hernia for several reasons:

1. Improved Outcomes: The improved outcomes of diaphragmatic eventration and hernia are largely attributed to advancements in medical and surgical management. Here's a summary of the key points:

Congenital Diaphragmatic Hernia (CDH): With advances in the management of CDH, the overall survival has improved. It has been reported to be 70-90% in non-ECMO infants and up to 50% in infants who undergo ECMO. The main focus of management includes gentle ventilation, hemodynamic monitoring, and treatment of pulmonary hypertension followed by surgery.⁸

Complicated Diaphragmatic Hernia: The laparoscopic approach is the preferred technique in hemodynamically stable patients without significant comorbidities because it facilitates early diagnosis of small

diaphragmatic injuries and reduces postoperative complications. Open repair is considered necessary in the majority of unstable patients, where Damage Control Surgery can be life-saving.

Presence of Hernia Sac: Overall survival was significantly better in patient groups with a hernia sac. The requirement for patch repair of the defect was more common among those without a hernia sac. Pulmonary hypertension was manifested more often among CDH babies without a hernia sac.

2. Reduced Mortality: The reduction in mortality rates for patients with diaphragmatic eventration and hernia can be attributed to several factors, including advancements in surgical techniques, better preoperative care, and improved postoperative management. Here's a summary of the key points:

Surgical Advancements: The development of minimally invasive surgical techniques, such as laparoscopic diaphragm reconstruction, has contributed to reduced mortality rates. These procedures are associated with fewer complications and quicker recovery times.¹

Early Diagnosis: The use of diagnostic imaging, like CT scans, has allowed for earlier detection of these conditions, leading to timely surgical intervention and better outcomes.¹⁷

Specialized Care: The management of congenital diaphragmatic hernia (CDH) often involves specialized neonatal care units that provide comprehensive treatment, including the use of extracorporeal membrane oxygenation (ECMO) and targeted neonatal echocardiography, which have improved survival rates.

Improved Postoperative Care: Enhanced postoperative care, including better pain management, respiratory support, and nutritional support, has also played a role in reducing mortality.

Research And Guidelines: Ongoing research and the development of clinical guidelines have led to standardized care protocols, which ensure that patients receive the most effective treatments based on the latest evidence.¹⁷

CHALLENGES AND FUTURE DIRECTIONS

Diagnosing diaphragmatic eventration and hernia poses significant challenges, particularly due to the condition's varying presentations and the difficulty in distinguishing it from other respiratory or gastrointestinal disorders. In infants, symptoms may be subtle and nonspecific, leading to delays in diagnosis and potential complications. Additionally, the rarity of these conditions contributes to limited awareness among healthcare providers, further complicating early detection and intervention. Furthermore, the complexity of surgical repair, especially in cases of large defects or associated anomalies, presents clinical challenges and risks, necessitating careful preoperative planning and postoperative management.

Future research in diaphragmatic eventration and hernia is poised to address these challenges and advance the field. One promising direction involves leveraging advanced imaging modalities, such as high-resolution ultrasound and MRI, to improve diagnostic accuracy and facilitate early intervention. Moreover, the development of predictive biomarkers, including genetic markers and serum biomarkers, holds potential for identifying at-risk individuals and guiding personalized treatment approaches. Additionally, the refinement of minimally invasive surgical techniques, such as laparoscopy and robotic-assisted surgery, offers the promise of reduced morbidity and faster recovery times for patients. Collaborative efforts among multidisciplinary teams, including surgeons, pulmonologists, and geneticists, will be crucial in developing standardized treatment protocols and optimizing long-term outcomes. Furthermore, patient education and support initiatives can empower individuals and families affected by these conditions, enhancing treatment adherence and overall quality of life. Through these concerted efforts, the future of diaphragmatic eventration and hernia management holds promise for improved patient care and outcomes.

CONCLUSION

In conclusion, the pathophysiology of diaphragmatic eventration and hernia involves the abnormal elevation or protrusion of the diaphragm, which can lead to respiratory compromise. Clinically, these conditions may present with symptoms ranging from asymptomatic to severe

respiratory distress. Diagnostic modalities include imaging techniques such as chest X-rays, ultrasound, and CT scans, which are crucial for accurate diagnosis and surgical planning. Current therapeutic approaches focus on surgical intervention, with techniques varying from open surgery to minimally invasive procedures like laparoscopic and robotic-assisted repairs. The choice of procedure depends on the patient's condition, the severity of the diaphragmatic defect, and the surgeon's expertise. The importance of early detection and intervention cannot be overstated, as timely management can prevent complications such as respiratory failure and improve the overall prognosis. Future directions in this field may involve refining surgical techniques, improving perioperative care, and developing guidelines for the management of these conditions to ensure better patient outcomes. Early and accurate diagnosis, coupled with appropriate therapeutic interventions, remains the cornerstone of managing diaphragmatic eventration and hernia effectively.

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