New Modalities In The Management of Congenital Diaphragmatic Hernia

An essay

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Presented by:

Memy Mohamed Khedre

M.B.B.Ch

Supervised By :

Professor Dr./ Hatem Abdel Kader saafan

Professor of Pediatric Surgery
Faculty of Medicine,
Ain Shams University

Dr./Hesham Mohamed Abdel kader

Assistant Professor of Pediatric Surgery
Faculty of Medicine,
Ain Shams University

DR./ Khaled Mohamed Abdel Salam

Lecturer of Pediatric Surgery
Faculty of Medicine,
Ain Shams University

Ain Shams University
Faculty of Medicine
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List of abbreviations :

- **CDH :** Congenital Diaphragmatic Hernia.
- **LH :** Lung Hypoplasia.
- **HFOV :** High-Frequency Oscillatory Ventilation.
- **ECMO :** Extracorporeal Membrane Oxygenation.
- **PPHN :** Persistent Pulmonary Hypertension of Newborn**.**
- **MIS :** Minimally Invasive Surgery**.**
- **iNO :** Inhaled Nitric Oxide.
- **LHR :** Lung-to-Head Ratio.
- **FETENDO:** Fetal endoscopy.
- **FETO :** Fetoscopic Endoluminal Tracheal Occlusion.
- **GERD :** Gastroesophageal Reflux Disease

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Introduction

Congenital diaphragmatic hernia (**CDH**) is a condition that is classically described as a "failed closure of the pleuroperitoneal canals between the eighth and tenth week of gestation" ***(Montedonico et al., 2008)***. This creates an opening between the supraperitoneal cavity and the inferior thoracic wall early in development. The intestine protrudes into the chest cavity and encroaches upon the lungs and heart; leading to pulmonary hypoplasia and pulmonary hypertension ***(Keckler and Schropp 2010).***

The vast majority of these cases (90% to 95%) occur posterolaterally; Bochdalek-type; ***(van Loenhout et al., 2009).*** In addition to this, CDH occurs once out of every 2,500 births and is responsible for 8% of all major congenital anomalies ***(Clugston et al., 2006).***

The aetiology of congenital diaphragmatic hernia is unknown, however, 2% of cases have been noted to be familial and another 15% of patients have associated chromosomal abnormalities ***(King and Booker 2005).***

Bochdalek popularized the concept of herniation due to failed closure of the primitive communication between the pleura and the peritoneal cavity (pleuroperitoneal duct), while Morgagni described a hernia that occur through a defect resulting from failure of the anterior

pleuroperitoneal membrane to fuse with the sternum and costal cartilages during embryogenesis ***(Akhavan-Heidari et al., 2006; Yavuz et al., 2006*).**

Anatomically, herniation through the right sternocostal triangle is actually the Morgagni hernia while the one through the left sternocostal triangle is Larry Hernia ***(Gilkeson et al., 1997).***

An infant with CDH typically presents at birth with an increased work of breathing, tachypnea, tachycardia, and cyanosis ***(Keckler and Schropp 2010)***.

Inspection of the chest and abdomen may reveal asymmetrical chest rise and an unusually "caved-in" belly (scaphoid) abdomen. Borborygmus may sometimes be appreciated on the affected side of the chest ***(Black 2010)***.

Diagnosis of congenital diaphragmatic hernia can be made prenatally or after birth. In developed countries, a high prenatal detection rate of congenital diaphragmatic hernia (59%) has been reported and the gestational age at diagnosis was greater than 24 weeks in half of the prenatally diagnosed cases ***(Garne et al., 2002)*.**

However, prenatal detection of CDH is rare in developing countries due to inadequate facilities ***(Abubaker et al., 2011).***

Ultrasonographic features in keeping with this condition include maternal polyhydramnios, an absent or intrathoracic stomach bubble, a mediastinal and cardiac shift away from the side of the herniation, a small fetal abdominal circumference, and rarely fetal hydrops ***(Spina et al., 2003).***

Useful prognostic tool that can be measured include the fetal lung area to head circumference ratio, the position of the liver and stomach using 3D-ultrasound scan, and measurement of fetal lung volume using magnetic resonance imaging. Jani et al. revealed that a low lung area to head circumference ratio (less than 1) and herniation of the liver are predictive of poor survival ***(Jani et al., 2006).***

Currently, in-utero endoscopic (fetoscopic) surgery offers a promising route for delivery of therapeutic agents to the fetus ***(Deprest et al., 2010).***

Fetal surgery has been explored in experimental studies and has been applied clinically. In experimental studies, prenatal tracheal occlusion has been shown to induce lung growth with reduction of herniated viscera and a dramatic improvement in lung compliance and gas exchange ***(Adzick et al., 2003).***

Fetal surgery has revolutionized from open surgical repair to tracheal occlusion techniques: open surgical tracheal occlusion, endoscopic external tracheal occlusion, and endoscopic endoluminal tracheal occlusion ***(Abubakar et al., 2011).***

The management of CDH can be summarized in four stages. The first stage is to stabilize the infant immediately following birth. This includes any rescue efforts. If an infant is suspected of having CDH during a resuscitation attempt, endotracheal intubation is done immediately ***(Keckler and Schropp 2010; Black 2010)***.

The concept has changed from performing emergency repair to delaying repair for at least 24-48 hours to allow for clinical stabilization and a fall in pulmonary vascular resistance ***(King and Booker 2005)***. Depending on the clinical condition of the patient, surgery can be delayed for up to 7-10 days to allow maximal relaxation of the pulmonary vasculature ***(Benjamin et al,. 2011).***

The second stage involves critical care management. The infant is admitted to the neonatal intensive care unit (NICU) where stabilization can be maintained and improvement monitored. Central to this strategy is the management of the patient's ventilation. Currently, mechanical ventilation (MV), high-frequency oscillatory ventilation (HFOV), and

extracorporeal membrane oxygenation (ECMO) are all viable options for CDH ***(Keckler and Schropp 2010)***.

The third stage of CDH management can be employed: surgical repair ***(Keckler and Schropp 2010).***

Minimally invasive surgery(MIS) for infants and children continues to grow. MIS was first introduced for the treatment of congenital diaphragmatic hernia(CDH) in **1995;Silen et al**. used thoracoscopy,while **Van der Zee and Bax(1995)** used laparoscopy. However, Most pediatric surgeons have hesitated to apply MIS to CDH because of the associated fragile respiratory status and pulmonary hypertension(PH). Nevertheless, thoracoscopic repair under high-frequency oscillatory ventilation(HFOV) ***(Liem et al., 2010)*** Or after extracorporeal membrane oxygenation(ECMO) therapy have been reported recently ***(Kim et al., 2009)***.

And MIS appears to be gaining added acceptance for CDH repair without any fixed selection criteria being established ***(Kim et al., 2009; Yang et al., 2005; Guner et al., 2008; Cho et al., 2009).***

A fourth stage of management is employed: immediate and long-term postoperative care ***(Keckler and Schropp 2010).***

Despite all these advances in neonatal care and surgical management, congenital diaphragmatic hernia (CDH) remains a condition with a significantly high mortality rate ***(Doherty et al., 2006)***.

Aim of the work

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This work aiming to discuss the new modalities in the management of congenital diaphragmatic hernia With great emphasizing on the fetal management, perioperative management and thoracoscopic repair of the defect.

Chapter 1

Anatomy and embryology of diaphragm and congenital diaphragmatic hernia

I. *Epidemiology:*

The congenital diaphragmatic hernia occurs one in every 2,500 live births ***(van den Hout et al., 2009).*** The true incidence remains unknown because of early deaths among severely affected fetuses and infants, which is commonly described as the "hidden mortality" of CDH. In one study there is no sex association in CDH ***(Stege et al., 2003).***

II. *Diaphragm Anatomy and Function*

The word diaphragm is derived from the Greek dia (in between) and phragma (fence). The diaphragm is a musculo-fibrous dome-shaped membrane between the thoracic and abdominal cavities ***(Maish 2010).***

The diaphragm is a curved musculofibrous sheet that separates the thoracic from the abdominal cavity. Its convex upper surface faces the thorax, and its concave inferior surface is directed towards the abdomen. The positions of the domes or cupolae of the diaphragm are extremely variable because they depend on body build and the phase of ventilation. Thus the diaphragm will be higher in short, fat people than in tall, thin people, and over inflation of the lung, as occurs for example in emphysema, causes marked depression of the diaphragm. Usually, after

forced expiration the right cupola is level anteriorly with the fourth costal cartilage and therefore the right nipple, whereas the left cupola lies approximately one rib lower. With maximal inspiration, the cupola will descend as much as 10 cm, and on a plain chest radiograph the right dome coincides with the tip of the sixth rib. In the supine position, the diaphragm will be higher than in the erect position, and when the body is lying on one side, the dependent half of the diaphragm will be considerably higher than the uppermost one ***(Gray's 2008).***

III. ***Embryology of diaphragm***

The diaphragm is formed by the fusion of the following embryonic structures :

1) Septum transversum

2) Pleuroperitoneal membranes

3) Dorsal mesentery of esophagus

4) Dorsal and dorsolateral body wall

The intraembryonic coelom is initially a single space which is later divided into pericardial, peritoneal and pleural space. The diaphragm separates the thoracic cavity from the peritoneal cavity ***(Saddler 2006).***