SURGICAL MANAGMENT OF

CONGENITAL DIAPHRAGMATIC HERNIA

An Essay
Submitted in Partial Fulfillment of
M.S Degree in general surgery
BY

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2011

العلاج الجراحى للفتق الحجابي الخلقي

خطة بحثية للحصول على درجة الماجستير في الجراحة

بواسطة

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بكالوريوس الطب والجراحة (٢٠٠٤)

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SUMMARY

Congenital diaphragmatic (CDH) a malformation that hernia is results from failure of fusion of the different elements that contribute to the formation of the diaphragm. The hernia may occur alone or may occur as one of multiple malformation syndromes. The hernia may occur through foramen of Bochadalek in the posterior portion of diaphragm or through foramen of Morgagni immediately behind the xiphoid process.

Respiratory dysfunction develops as a result of associated anomalies and lung hypoplasia which range from minimal hypoplasia of one side to severe bilateral hypoplasia as a result of compression and inhibition of lung growth. The congenital hernia usually occurs on left side less common on the right and rarely bilaterally.

Prenatal ultrasound is done routinely during prenatal care allowing for assessment of fetal size, anatomy and the prenatal diagnosis of many anaomalies. After the prenatal diagnosis of CDH is made, predicting outcome for these cases remains challenging, cases associated with multiple congenital anomalies are more risky.

Ninety percent of CDH cases are symptomatic within the 1st 24 hours of life and are considered as early presenting CDH however this disease may manifest later on in life causing what's known as late presenting CDH or go unnoticed until very late in life or even never be diagnosed.

Clinically the infant well be tachypneic with grunting may be cyanotic presenting in the first few hours or days of life but late presentation may be also encountered. Physical signs as mediastinal shift to the opposite side, diminished air entery and scaphoid abdomen also are helpful in diagnosis.

Chest x ray performed may show air filled loops of bowels inchest cavity. Prenatal diagnosis by ultrasonography is another way of early diagnosis.

Numerous interventions have been explored in hopes of improving the outcome in newborns with CDH. Extracorporeal membrane oxygnator and high frequency ventilation are now used routinely in the management of severe respiratory failure.

The congenital diaphragmatic hernia may be considered as a surgical emergency. After proper and adequate resuscitation by nasogastric decompression and ventilatory support which may include extracorporeal membrane oxygenation (ECMO). The patient transferred to operating room. After reduction of the hernia, the defect is closed by continuous prolene suture. Agensis of the diaphragm or large defect may require insertion of prothesis as Gortex membrance or other material. Postoperatively ventilatory support is maintained until the infant has satisfactory blood gases, ECMO may also be required.

It is concluded that endoscopic role in congenital diaphragmatic hernia including thoracoscopic and laparoscopic repair may also be beneficial in diagnosis and management of CDH.

Mortality rate remained high especially in severely affected infant despite improved surgical techniques, ventilatory support and medical care.

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INTRODUCTION

Congenital diaphragmatic hernia refers to a congenital defect in the posterolateral diaphragm at the "foramen of Bochdalek". It is a relatively common cause of neonatal respiratory distress with overall incidence between 1:2000 and 1:5000 live births. Congenital diaphragmatic hernia accounts for about 90% of congenital diaphragmatic defects. Left sided congenital diaphragmatic hernia represent 80-90 % of congenital diaphragmatic hernias. The specific etiology of congenital diaphragmatic hernia is unknown but it is believed to result from a defective formation of the pleuroperitoneal membrane. ***(Bambini , 2000).***

During the last 25 years greater emphasis has been placed on the physiological behaviors of the lungs in CDH. The degree of pulmonary hypoplasia and hypertension is the most important determinant of survival in congenital diaphragmatic hernia. ***(Cilly et al., 2006).***

The mortality rate of infants born with congenital diaphragmatic hernia remain high despite optimal perinatal care. The high mortality rate in congenital diaphragmatic hernia has been attributed to pulmonary hypoplasia an associated persistent pulmonary hypertension. In recent years, newer management strategies such as permissive hypercapnia, high frequency ventilation, extracorporeal membrane oxygenation and delayed surgical repair have emerged in the care of high-risk congenital diaphragmatic hernia patients which offer some hope of improving overall survival. ***(Puri , 2006)***

AIM OF THE WORK

To highlight recent trends in the methods of early diagnosis and management of congenital diaphragmatic hernia including indications and role of surgery. In addition, we will focus on more recent developments in the surgical management of congenital diaphragmatic hernia including the application of laparoscopic techniques, fetal interventions and lung transplantation.

Development of the Diaphragm

The diaphragm is a composite structure that develops from four embryonic components (Fig. 1).

- Septum transversum.
- Pleuroperitoneal membranes.
- Dorsal mesentry of esophagus.
- Lateral body walls.

The diaphragm is a dome-shaped, musclotendinous partition that separates the thoracic and abdominal cavities ***(Keith and Persaud, 2007).***

The Septum transversum :

The transverse septum composed of mesodermal tissue, is the primordium of the central tendon of the diaphragm (Fig. 1 D and E).

The septum transversum grows dorsally from the ventro lateral body wall and forms a semicircular shelf, which separates the heart from the liver (Fig. 2). During its early development, a large part of the liver is embedded in the septum transversum. The septum transversum is located caudal to the pericardial cavity and partially separates it from the developing peritoneal cavity. The septum transversum is first identifiable at the end of the third week as a mass of mesodermal tissue cranial to the pericardial cavity. After the head folds ventrally during the fourth week, the septum transversum forms a thick incomplete partition between the

pericardial and abdominal cavities (Fig. 3).The septum transversum does not separate the thoracic and abdominal cavities completely. A large opening, the pericardio peritoneal canal is found on each side of the esophagus (Fig. IB). The septum transversum expands and fuses with the mesenchyme ventral to the esophagus (primitive mediastinum) and the pleuroperitoneal membranes (Fig. 1C)***(Shields, 2005).***

The pleuroperitoneal Membranes:

These membranes fuse with the dorsal mesentery of the esophagus and septum transversum (Fig. 1C). This completes the partition between the thoracic and abdominal cavities and forms the primordial diaphragm. Although the pleuroperitoneal membranes form large portions of the fetal diaphragm, they represent relatively small portions of the newborn infant's diaphragm (Fig. IE) ***(Keith and Persaud, 2007).***

The dorsal Mesentery of the esophagus :

As previously described, the septum transversum and pleuroperitoneal membranes fuse with the dorsal mesentery of the esophagus. This mesentery constitutes the median portion of the diaphragm. The crura of the diaphragm - a leg like pair of diverging muscle bundles that cross in the median plane anterior to the aorta (Fig IE) develop from myoblasts that grow into the dorsal mesentry of the esophagus ***(Christine and Bryan, 2007).***

The muscular in growth from the lateral body wall:

During the 9th to 12th weeks, the lungs and pleural cavities enlarge "burrowig" into the lateral body walls (Fig. 4). During this excavation process, the body-wall tissue is split into two layers:

- An external layer that becomes part of the definitive abdominal wall.
- An internal layer that contributes muscle to peripheral portions of the diaphragm, external to the parts derived from the pleuroperitoneal membranes (Fig. ID and E).

Further extension of the developing pleural cavities into the lateral body walls form the right and left costodiaphragmatic recesses (Fig. 5),establishing the characteristic dome-shaped configuration of the diaphragm. After birth the costodiaphragmatic recesses become alternately smaller and larger as the lung move in and out of them during inspiration and expiration ***(Keith and Persaud, 2007).***

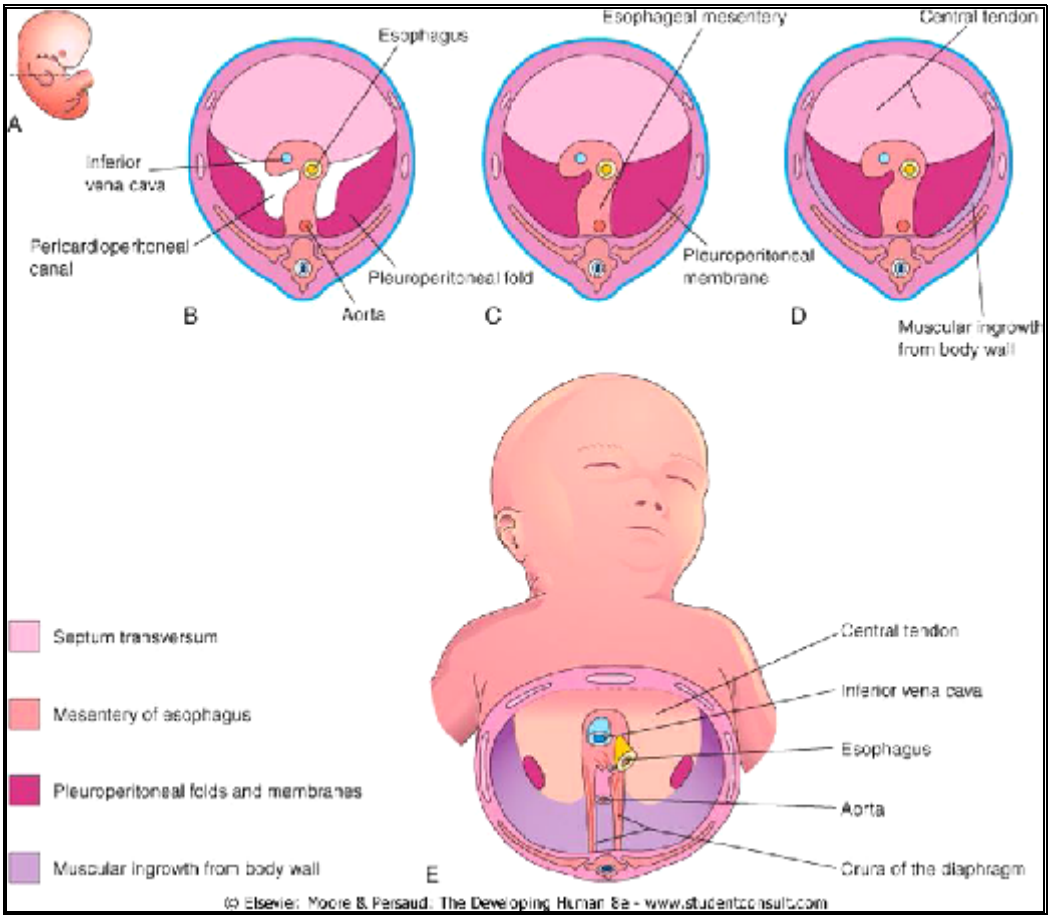

Figure (1) : Illustrations of development of the diaphragm. A, Sketch of a lateral view of an embryo at the end of the fifth week (actual size) indicating the level of sections in B to D. B to E, The developing diaphragm as viewed inferiorly. B, Transverse section showing the infused pleuroperitoneal membranes. C, Similar section at the end of the sixth week after fusion of the pleuroperitoneal membranes with the other two diaphragmatic components. D, Transverse section of a 12-week fetus after ingrowth of the fourth diaphragmatic component from the body wall. E, Inferior view of the diaphragm of a newborn indicating the embryologic origin of its components ***(Keith and Persaud, 2007).***

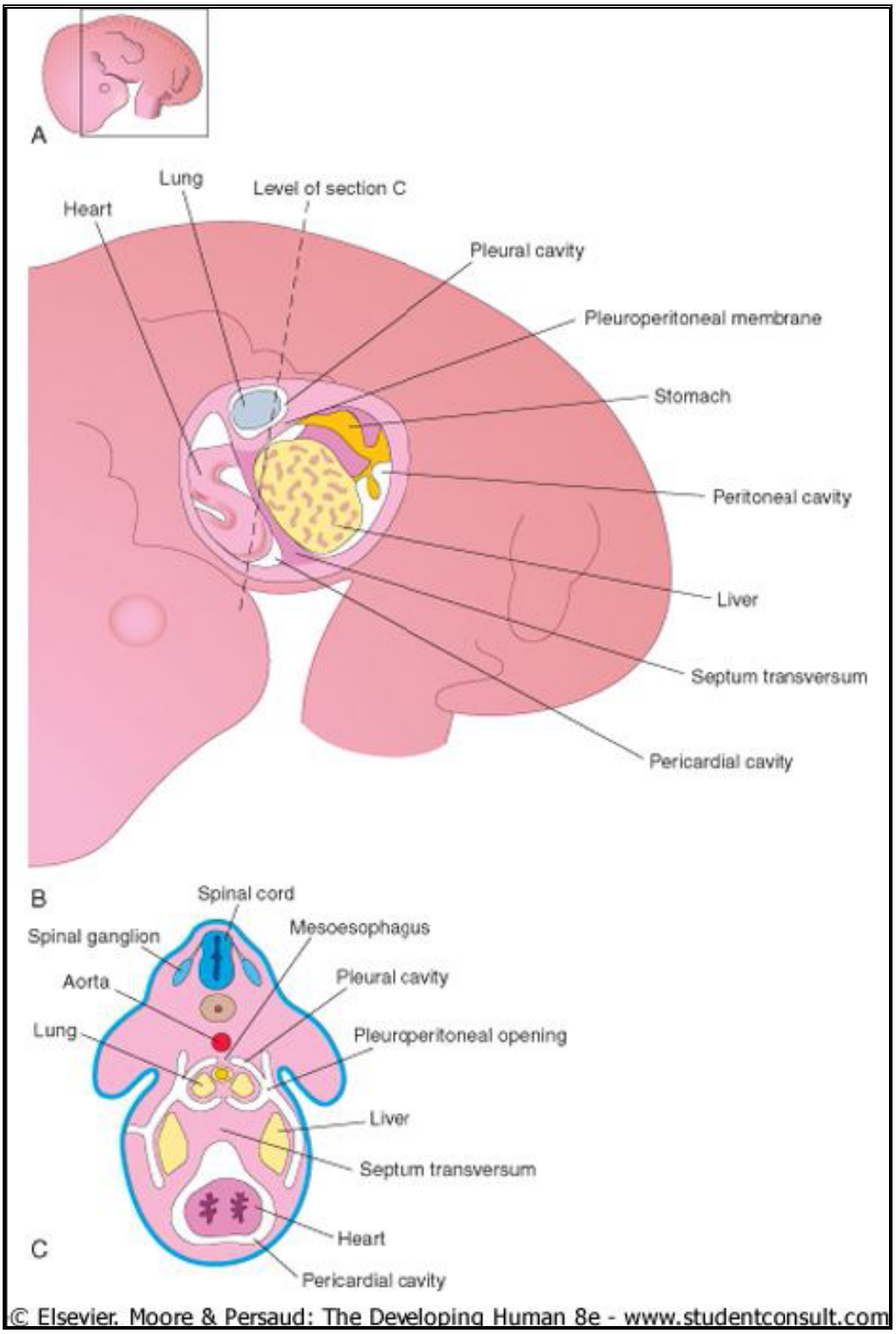

Figure (2): **(A)** Sketch of a lateral view of an embryo (approximately 33 days). The rectangle indicates the area enlarged in **(B)** The primordial body cavities are viewed from the left side after removal of the lateral body wall. **(C)** Transverse section through the embryo at the level shown in **B** ***(Keith and Persaud, 2007).***

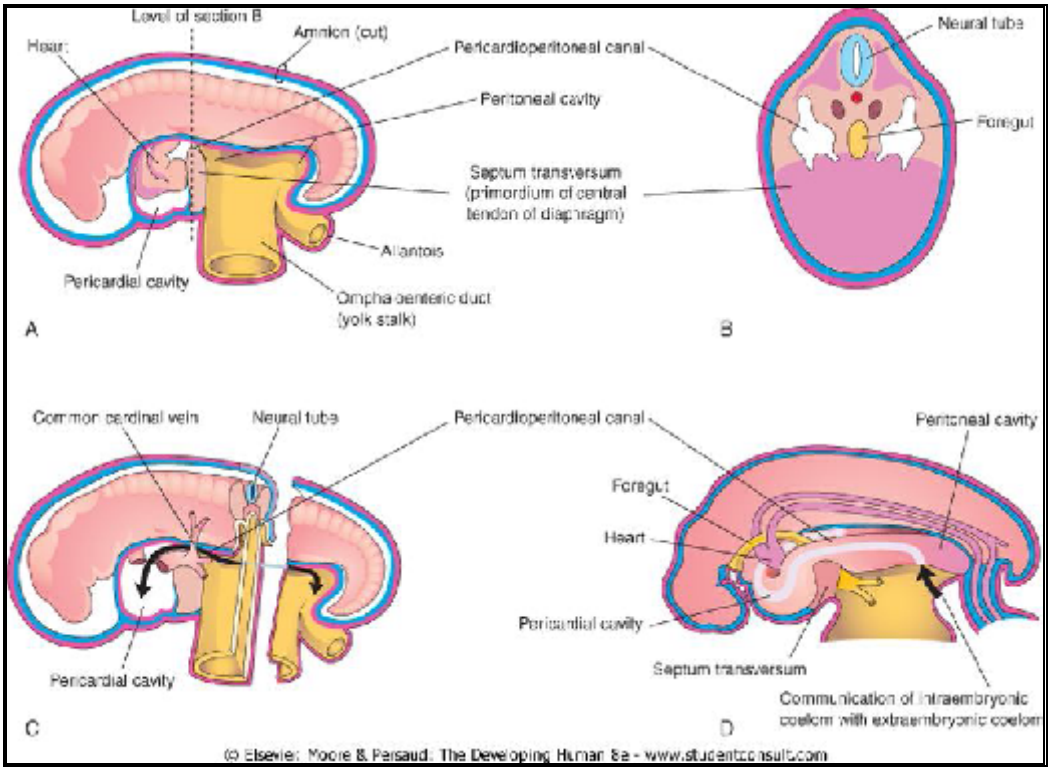

Figure (3): Schematic drawings of an embryo (approximately 24 days). **(A)** The lateral wall of the pericardial cavity has been removed to show the primordial heart. **(B)** Transverse section of the embryo illustrates the relationship of the pericardioperitoneal canals to the septum transversum (primordium of central tendon of diaphragm) and the foregut. **(C)** Lateral view of the embryo with the heart removed. The embryo has also been sectioned transversely to show the continuity of the intraembryonic and extraembryonic coeloms (arrow). **(D)** Sketch showing the pericardioperitoneal canals arising from the dorsal wall of the pericardial cavity and passing on each side of the foregut to join the peritoneal cavity. The arrow shows the communication of the extraembryonic coelom with the intraembryonic coelom and the continuity of the intraembryonic coelom at this stage ***(Keith and Persaud, 2007).***

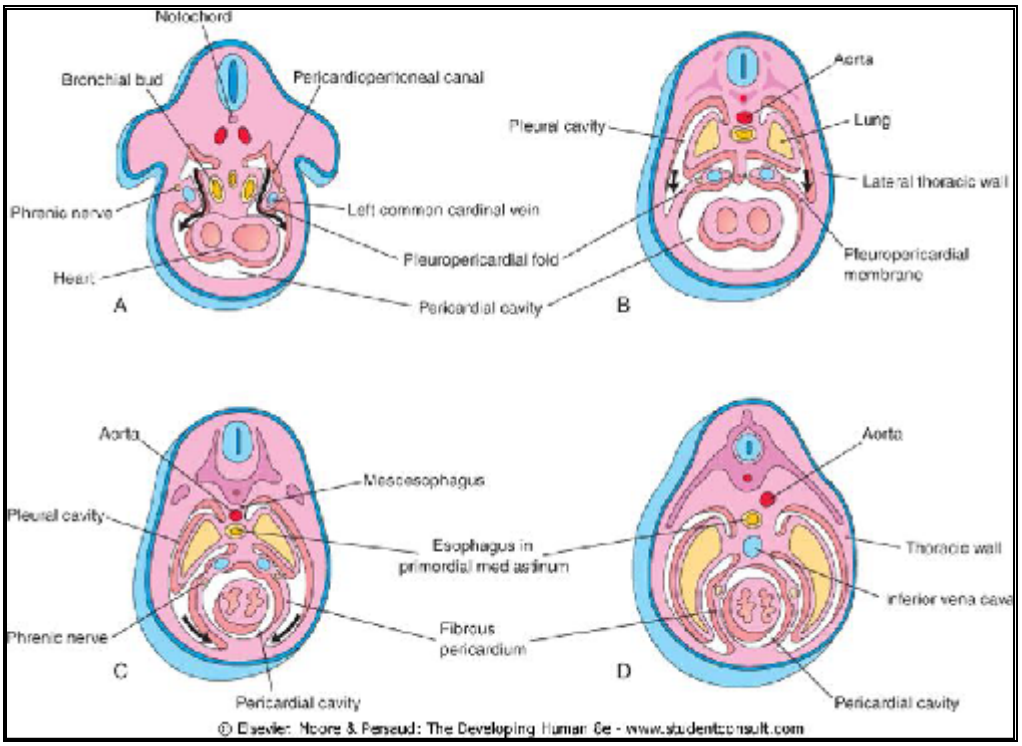

Figure (4) : Schematic drawings of transverse sections through embryos cranial to the septum transversum, illustrating successive stages in the separation of the pleural cavities from the pericardial cavity. Growth and development of the lungs, expansion of the pleural cavities, and formation of the fibrous pericardium are also shown. **(A)** At 5 weeks. The *arrows* indicate the communications between the pericardioperitoneal canals and the pericardial cavity. **(B)** At 6 weeks. The *arrows* indicate development of the pleural cavities as they expand into the body wall.**(C)** At 7 weeks. Expansion of the pleural cavities ventrally around the heart is shown. The pleuropericardial membranes are now fused in the median plane with each other and with the mesoderm ventral to the esophagus. **(D)** At 8 weeks. Continued expansion of the lungs and pleural cavities and formation of the fibrous pericardium and thoracic wall are illustrated ***(Keith and Persaud, 2007).***

Positional changes and innervation of the diaphragm :

During the fourth week of development, the septum transversum, prior to its descent with the heart, lies opposite the third to fifth cervical somites (Fig. 6A). During the fifth week, myoblasts (primitive muscle cells) from these somites migrate into the developing diaphragm, bringing their nerve fibres with them. Consequently, the phrenic nerves that supply motor innervation to the diaphragm arise from the ventral rami of the third, fourth, and fifth cervical spinal nerves. The three twinges on each side join