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Congenital Diaphragmatic Hernia: A Review of Its Management

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Review Article

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ABSTRACT

Congenital Diaphragmatic Hernia (CDH) is considered a defect in diaphragm development which occurs congenitally, its incidence is about 1 in 2500 newborns. Major components of CDH are increased pulmonary pressure with fetal circulation persistence, with significant reduction in lung compliance and its tidal volume. Preoperative stabilization and optimization should preceed the surgical repair which is done usually via laparotomy which is considered to be the standard approach. Minimally invasive procedures for CDH repair provides the patient with the benefit of less pain and avoidance of the consequences of thoracotomy or laparotomy compared with the open repair strategy, inspite of prescence of disadvantages mainly from insufflation and CO_2 absorption, but this can be controlled with lower CO_2 pressure, better insufflation instruments and continuous monitoring of blood gases.

Keywords: Congenital diaphragmatic hernia; abnormal pathophysiology; laparotomy; thoracotomy; surgical repair.

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1. INTRODUCTION

1.1 Epidemiology

CDH prevalence ranges from 1: 1,200 to 1: 12,000 births [1-10]. This difference may be due to deaths before referral which are not included. Studies show that CDH incidene is 1: 2,107 to 1: 3,163 births [1,3-6,8].

In a study done by Torfs a more prevalence of CDH had been detected in rural areas.

1.2 Research Directions

To enhance our knowledge of the abnormal pathophysiology and challenges we face in CDH cases.

To reach international consensus regarding the time and type of surgical intervention for treatment of congenital diaphragmatic hernia.

1.3 Embryological Development of the Diaphragm

The diaphragm is mesodermal in origin which results from five embryonic structures fusion [11]:

1. A central part from Septum transversum.
2. Two membranous structures called pleuro-peritoneal membranes.
3. Chest wall mesoderm.
4. Oesophageal mesentry.
5. Aortic surrounding mesoderm [12].

1.4 Anatomy of the Diaphragm

The diaphragm separates the abdomen from thorax. It is composed of central aponeurotic part and peripheral muscular part [13].

The muscular part is further divided to three parts:

1. A vertebral portion which arises from the crura and the arcuate ligaments. Vertebrae.
2. A costal portion which arises from the interior of the lower six ribs and costal cartilages.
3. A sternal part which arises from the deep surface of the xiphoid process of sternum.

The muscular fibres converges and being inserted centrally into central tendon which is fused partially with the pericardium [13].

1.5 Diaphragmatic Arterial Supply

Inferior phrenic arteries which are aortic or celiac axis branches gives the majority of diaphragmatic arterial supply, pericardiophrenic, internal thoracic , musculophrenic, superior phrenic arteries and two branches from the thoracic aorta also share in diaphragmatic arterial supply [14].

1.6 Diaphragmatic Venous Drainage

Usually the venous drainage of the diaphragm follows the arterial supply [15].

1.7 Diaphragmatic Lymphatic Drainage

Diaphragmatic lymph nodes which lies on its superior surface can be divided into anterior, middle, and posterior groups. They drain superior hepatic surface, junction between oesophagus and stomach, inferior diaphragmatic surface, then these nodes drain superiorly to mediastinal, parasternal nodes, posterior mediastinal and brachiocephalic nodes [15].

1.8 Diphragmatic Nerve Supply

The peripheral portions of the diaphragm takes sensory innervation from the 7th to the 12th intercostal nerves [15].

Central part of diaphragm sensory nerve fibres run in the phrenic nerve [13].

Motor supply of diaphragm arises from the phrenic nerve (C3, C4, C5), [15].

1.9 Pathophysiology of Congenital Diaphragmatic Hernia

CDH pathophysiology consists mainly from pulmonary hypertension together with persistent fetal circulation with decreased lung compliance and tidal volume. Surfactant deficiency is another aspect in CDH pathophysiology [16-19].

Reduction in total alveolar number and lung volume results in lower tidal volume [20-23].

Reduction in the diameter and number of arterial branches, hypermuscularization of the arteries, amplified arterial reactivity and reduced nitric oxide synthesis results in significant increase in pulmonary vascular resistance [24,25].

Increased pulmonary vascular resistance results in pulmonary hypertension [25-27] which results in decreased lungs total blood flow with increased right ventricular end-diastolic pressure, right-to-left and persistent fetal circulation [26, 28]. These events results in hypercapnia, hypoxemia and acidosis, which induces further pulmonary vasoconstriction with pulmonary hypertension worsening with subsequent increase in persistence of the fetal circulation and so on, which results in a vicious circle [25,29].

After birth, deterioration of the baby hemodynamic status with right sided heart overload and potential failure [30-32].

Mediastinal deviation by the herniated content may reduce the cardiac venous return with more worsening of hemodynamic status [33].

1.10 CDH Clinical Manifestations:

Respiratory distress in the first 24 hours afterbirth is considered the main clinical manifestation. Tachypnea, subcostal, sternal, and supraclavicular retraction may be present. Pallor and Cyanosis may also be present. Scaphoid abdomen is usually present because of abdominal viscera herniation into the chest [34].

Asymmetrical and larger hemithorax, diminished or absent respiratory sounds with prescence of bowel sounds may be present on the ipsilateral hernial side may be present. Shifted trachea and heart sounds to the contralateral side or in the middle of chest also may be present. Hemodynamic instability with arterial hypotension due to prescence of pulmonary hypertension [35].

Growth retardation, Sudden death, GIT perforation or strangulation, airway infections or recurrent pneumonias, rupture of a herniated spleen, chest pain, urinary tract obstruction due to ureteric herniation and obstruction, acute abdomen, vomiting, intrathoracic appendicitis, and other rare presentations [34,36,37].

1.11 CDH Diagnosis

Most of CDH cases are being diagnosed antenatally, during routine ultrasound [38]. Polyhydramnios due to reduction in amniotic fluid volume swallowed by the fetus [18,19]. The hernial content detected by prenatal ultrasound can move in and out of the chest [39].

A plain chest radiograph postnatally is usually enough to confirm the CDH diagnosis. Bowel loops within the hemithorax, mediastinal deviation to the contralateral side, and absent or decreased abdominal gas may be present [40].

2. Treatment

2.1 Surgery

2.1.1 Timing of Operation

CDH repair is no longer considered an emergency procedure [41]. A delay of the operative repair until haemodynamic stability of the infant is achieved started since the early 1990s [42,43]. No mortality rate significant difference had been found in studies compared early vs late repairs, including two randomized trials of early (<12 hours) versus delayed repair after 24 hours [44] and after 96 hours [45].

So, CDH repair is usually delayed until achieving cardiopulmonary stability, although the accurate definition of haemodynamic stability remains variable and inconsistent from centre to another [46].

In 1995, Wung et al. revealed advantages with a delayed repair for CDH [42]. Repair had not been performed until the pre- and postductal *Sp*O2 equalized and resolved right-to-left shunting on echocardiogram. survival was 94% with an average of 4.2 days after birth before operation. Overall survival of 84% had been achieved with spontaneous respiration, permissive hypercapnia, and elective repair after 36 hours of life [47].

2.1.2 Operative Approach

Open repair of CDH can be done by thoracic or abdominal approach. Laparotomy advantages include intrathoracic viscera easier reduction, diaphragmatic posterior rim mobilization, easier management of associated intestinal rotational anomalies, and avoidance of thoracotomy related musculoskeletal deformities. Most (91%) of neonatal repairs for CDH are being performed through a subcostal laparotomy incision [48,49]. Less than 10% are being done through a thoracotomy. The herniated abdominal viscera should be reduced out of the thorax giving special attention to the spleen that can be injured [50].

The surgeon should avoid any respiratory compromise and if occured abdomen should be

left open. [51]. Temporary closure can be done using a prosthetic silo or the skin. A trial of delayed closure, should be attempted after resolving of the edema or increased intra-abdominal domain [52].

Chest tubes routine usage after CDH repair to drain pleural fluid has been abandoned [53,54]. Chest tubes can cause injury to the lung especially if it had been connected to suction. [55].

If needed chest tubes should be placed to water seal. Repeated thoracocentesis can been done to treat symptomatic pleural fluid and should be removed early to avoid infectious complications.

2.2 Minimally Invasive Repair

Several morbidities seen after open repair of congenital diaphragmatic hernia pushed the surgeons to search for minimal invasive surgical (MIS) techniques. Both laparoscopic and thoracoscopic techniques had been performed and utilized worldwide since 1995 in many centers [56]. Minimally invasive surgical techniques had been used for both primary prosthetic patch repairs with supposed benefits of avoidance of thoracotomy-associated complications, less postoperative pain, and reduction of surgical stress [57,58].

The suggested benefits of Minimally invasive repairs had been questioned due to: (1) Absorption the CO2 insufflation gas in CDH neonates [59,60]; and (2) Increased intrathoracic pressure which may decrease venous return, organs perfusion, and pulmonary volumes. The added difficulty of lung hypoplasia, Pulmonary hypertension increase the challenges faced by MIS.

Acidosis and hypercapnia may wosen the right to left pulmonary shunting. Careful selection of patient is a must for a safe completion of minimally invasive repair [61].

Congenital diaphragmatic hernia repair using Robot has been shown to be safe due to benefits of articulating instruments free movements for suturing [62].

2.3 Diaphragmatic Alternatives

2.3.1 Nonabsorbable patches

Synthetic patches: Gore-Tex or polytetrafluoroethylene or Marlex are commonly being used as diaphragmatic alternatives for neonates with large defects [63]. Shorter time for preparation, Immediate availability; ease of reshaping for adequate fitting into the diaphragmatic defect; low risk of hemorrhage and lesser extent of dissection of tissues are considered important advantages for these types of diaphragmatic replacements [64].

Chest wall tethering, increased intestinal obstruction incidence, deformities of abdominal wall, splenectomy, patch infections can be considered disadvantages for these types of diaphragmatic replacements [63,65].

2.3.2 Postoperative Care

Mild support with flow synchronization by ventilator should be used [66-68].

Intravenous fluids given to the baby should be controlled with monitoring of the degree of hydration of the baby [69].

If a chest tube had been placed, it should be connected to under water seal [42].

Gastrointestinal obstruction resulting from gastric volvulus, adhesions or volvulus of the midgut can occur, so close follow up of the baby postoperatively should be done. Other complications include chylothorax and chylous ascites, for them a TPN based treatment should be used and medium chained triglycerides feedings [70].

3. CONCLUSION

Congenital diaphragmatic hernia is a congenital diaphragmatic defect which occurs during its development with an incidence of 1: 2500 newborns. Majority of Congenital diaphragmatic hernia babies are symptomatic within the first 24 hours of life. On the other hand Congenital diaphragmatic hernia can manifest at any age or even not detected until later life, or never been diagnosed. Surgical repair is the main stem of Congenital diaphragmatic hernia repair. Surgery should be delayed after stabilization of haemodynamic status. So, Congenital diaphragmatic hernia is considered a neonatal rather than surgical emergency. Congenital diaphragmatic hernia can be repaired thoracoscopically with excellent results in selected and stabilized cases. CDH patients treated by MIS should benefit from less pain and incisional complications, avoidance of the disadvantages of laparotomy thoracotomy with

reduced surgical stress compared with traditional open repair, although there are specific disadvantages mainly from CO_2 absorption during insufflation, yet improved recently with lower CO_2 pressure and better quality of insufflation.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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