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## **Management of congenital diaphragmatic hernia : Review of 50 cases in Thi-qar / Iraq (2013-2022)**

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# Management of congenital diaphragmatic hernia : Review of 50 cases in Thi-qar / Iraq (2013-2022)

## Abstract

**Background:** Congenital diaphragmatic hernia (CDH) occurs when there is a hole in the diaphragm, which is the thin sheet of muscle separating the chest from the abdomen.

The first description of diaphragmatic hernia appeared in 1575. In 1848, Bochdalek described congenital diaphragmatic hernia (CDH) occurring through a posterolateral defect. Successful surgical treatment of CDH in an infant was first performed in 1902, whereas the first neonate operated within 24 hours of life was reported in 1946. Pulmonary hypoplasia and pulmonary hypertension were early recognised as important reasons for the high mortality rate.

**Aim:** Analysis of the clinical profile and outcome of 50 cases of congenital diaphragmatic hernia and determination of risk factors for mortality and difference in age, gender, indications, presentations & complications of surgery.

**Patients and Methods:** A retrospective & prospective observational study was conducted in 50 patients of congenital diaphragmatic hernia for different indications over a period of 9 years from May 2013 till May 2022 in Thi-Qar governorate. On admission, an informative history was taken from the family of every patient including age, gender, sex, onset and nature of clinical features related to disease itself. Other data included possible chronic diseases, associated anomalies & presence of previous surgical operation and history of drug allergy. Neonates with clinical and intraoperative diagnosis of diaphragmatic eventration were not included in this study.

## Results:

Of the 50 cases of congenital diaphragmatic hernia, 27 of them were males and 23 were females aged from less than one month up to more than one year. Different investigations were done preoperatively. In addition, different complications were identified during and after surgery.

## Conclusion & recommendation:

CDHs are common on the left side with fairly good prognosis. Though, the right-sided CDH are rare; they do carry a good prognosis, as it was seen in our experience. On the other hand pulmonary hypertension is a very bad prognostic

sign and low birth weight and the presence of other associated congenital malformations lead to increased perioperative mortality.

**KEYWORDS:** Congenital diaphragmatic hernia, neonatal outcome, pulmonary hypertension, Bochdalek hernia, Morgagni hernia.

## **INTRODUCTION :**

The first description of diaphragmatic hernia appeared in 1575. In 1848, Bochdalek described congenital diaphragmatic hernia (CDH) occurring through a posterolateral defect. Successful surgical treatment of CDH in an infant was first performed in 1902, whereas the first neonate operated within 24 hours of life was reported in 1946. However, early surgery did not improve survival rates and the mortality was in the region of 50%. One reason for this was that more neonates underwent surgery who previously would have died without the repair of the CDH. Pulmonary hypoplasia and pulmonary hypertension were early recognised as important reasons for the high mortality rate.<sup>[1]</sup> In recent years, an enormous effort has been made by research groups all over the world to describe the pathogenesis and pathophysiology of CDH, and apply these findings to clinical practice. Attempts have been made to define prognostic factors. Extracorporeal membrane oxygenation (ECMO) has produced encouraging results. Fetal surgical therapy remains an option in selected cases despite huge technical and ethical problems.<sup>[2]</sup> Recently, several new therapeutic methods have been suggested, such as high frequency oscillatory ventilation, partial liquid ventilation, nitric oxide inhalation, surfactant therapy, and fetal tracheal ligation. However, more experience is required before the value of these approaches is clear. Despite these efforts, the mortality remains unacceptably high. The challenge for the future is to continue development of therapeutic approaches in order to improve survival of neonates with CDH.<sup>[3][4]</sup>



Figure(1): Chest X ray shows of patient left side CDH.

Bochdalek hernia make up the majority of cases of CDH. The major problems in these hernias are posterolateral defects of the diaphragm, which result in either failure in the development of the pleuroperitoneal folds or improper or absent migration of the diaphragmatic musculature. <sup>[5][6]</sup>

Morgagni hernia is a less common entity, accounting for only 5-10% of CDH cases. The foramen of a Morgagni hernia occurs in the anterior midline through the sternocostal hiatus of the diaphragm, with 90% of cases occurring on the right side. <sup>[7][8]</sup>

### **Presentation**

Early diagnosis - Right-side heart; decreased breath sounds on the affected side; scaphoid abdomen; bowel sounds in the thorax, respiratory distress, and/or cyanosis on auscultation;

CDH can often be diagnosed in utero with ultrasonography (US), magnetic resonance imaging (MRI), or both

Late diagnosis - Chest mass on chest radiography, gastric volvulus, splenic volvulus, or large-bowel obstruction. <sup>[9][10]</sup>

Congenital hernias (neonatal onset) - Respiratory distress and/or cyanosis occurs within the first 24 hours of life.

Congenital hernias (childhood or adult onset) - Obstructive symptoms from protrusion of the colon, chest pain, tightness or fullness the in chest, sepsis following strangulation or perforation, and many respiratory symptoms occur. <sup>[11]</sup>

## **Work-up**

### Laboratory Studies

Low levels of maternal serum alpha-fetoprotein (AFP) have been associated with congenital diaphragmatic hernia (CDH). However, decreased AFP also is observed with trisomy 18 and trisomy 21; thus, a low AFP level, by itself, is not diagnostic of CDH.

### Imaging Studies

By chest radiography, CDH may be signaled at an early stage by a finding of bowel and stomach in the chest cavity and shifting of the mediastinum (usually to the right). At a later stage, CDH may be signaled by a suspicious mass incidentally found on a chest radiograph. <sup>[12][13]</sup>

## **Medical Therapy**

Resuscitation with ventilatory support is of prime importance in patients born with a CDH. There has been a trend toward switching from conventional mechanical ventilation to high-frequency oscillatory ventilation (HFOV). HFOV serves to minimize airway pressure and, in conjunction with permissive hypercapnia, helps those with CDH suffer less traumatic lung injury and fewer long-term complications. Mortality has been shown to decrease from 49% to 20% when HFOV is used early in the treatment course. ECMO has been shown to decrease the mortality of CDH significantly but is currently reserved for individuals whose condition fails to improve with both HFOV and conventional mechanical ventilation. The decision to utilize ECMO is made early in the disease process, usually within 24 hours of birth. <sup>[14]</sup> <sup>[15]</sup>

With the addition of HFOV, more reports exist of decreased morbidity and mortality with early surgical intervention. However, there remains some controversy regarding this practice. The classical teaching is that patients need to be stabilized (often with the use of ECMO) and that repair should be delayed until the infant is better prepared to survive the operation. With the addition of HFOV, more reports exist of decreased morbidity and mortality with early surgical intervention. <sup>[16]</sup>

## **Preoperative management**

### Preoperative Medical Optimization and Evaluation

Preoperative evaluation ensures the patient is adequately optimized and stable for the procedure. The key medical goals for the neonate in preparing for surgery include improving gas exchange in the lungs and oxygen delivery to the periphery, lowering pulmonary arterial pressures to acceptable levels, correcting acid-base imbalances, and addressing any significant comorbidities. Additionally, it is important to ascertain at what point the hernia occurred in gestation, as early herniation is associated with worse pulmonary development and requires greater ventilatory assistance when compared to late herniation.<sup>[17]</sup>

Pertinent tests and imaging to be performed prior to surgery include a complete blood count, complete metabolic panel, lactate, coagulation assays, arterial blood gas, chest X-ray, echocardiography, and a head ultrasound. The CDH Euro Consortium suggested the following as indicators that the neonate is stable for surgery:

Normal MAP for gestation

Preductal SpO<sub>2</sub> between 85% to 95% on FiO<sub>2</sub> < 50%

Lactate < 3 mmol/L

UOP > 1 cc/kg/hr.<sup>[18]</sup>

### **Surgical Therapy**

With regard to treatment of CDH in the perinatal period, significant issues remain, and many questions are yet to be answered. However, the possibility of maternal-fetal surgical treatment of CDH is on the horizon. For treatment of CDH in the neonatal period, a subcostal incision is used. An attempt should be made to carry out a primary repair of the hernia. The abdominal contents are reduced, and the edges of the diaphragm are then approximated with nonabsorbable suture. In some cases, such as when the defect is large or the repair is being made while the patient is on ECMO, a prosthetic material such as expanded polytetrafluoroethylene (ePTFE) or polypropylene is used. For CDH related to traumatic rupture, the surgical approach depends on the timing of the diagnosis with the surgical intervention. Minimally invasive approaches (eg, involving video-assisted thoracoscopy or laparoscopy) are being explored. Putnam et al found that a minimally invasive approach was associated with a shorter hospital stay and reduced small-bowel obstruction but also with higher recurrence rates. Criss et al found that in low-risk patients born with small or moderate-sized defects, a thoracoscopic approach was associated with a shorter hospital stay, reduced mechanical ventilation days, and decreased time to feeding but also with a trend toward higher recurrence rates. Robotic approaches are being explored as well.<sup>[19]</sup>

## **Procedure**

### **Congenital diaphragmatic hernia repair**

Surgical repair of CDH after delivery depends on your individual baby's progress in the days following birth.

The multidisciplinary team, led by your baby's pediatric surgeon and neonatologist, will determine the timing of repairing the defect in the diaphragm. In our experience it is best to wait until the lungs show less resistance from the pulmonary hypertension and the baby has transitioned off ECMO. Our experienced team utilizes various medications and inhaled therapy to enhance this process.<sup>[20]</sup>

Babies with CDH are extremely sensitive to noise and movement, so surgical repair of CDH is often performed in the N/IICU. When it is time for the repair, the surgical team transforms your baby's N/IICU bedside into an operating room. This eliminates the additional stress to the baby of having to be transported.

Your baby will receive general anesthesia and will be monitored by a pediatric anesthesiologist. An incision is made just below your baby's rib cage, the organs in the chest are guided back down into the abdomen and the hole in the diaphragm is sewn closed. The space created in the chest allows the lungs to continue to grow.

For babies with large defects or completely lacking a diaphragm, the hole is closed with a GORE-TEX® patch or muscle flap. As your child grows, the condition of the patch will be regularly monitored by doctors to ensure that it remains intact.<sup>[21]</sup>

## **Complications**

It is extremely important to keep in mind that there is an increased risk of concomitant congenital defects in an infant with CDH. If a diaphragmatic hernia is diagnosed in utero, a high index of suspicion for concomitant abnormalities must be maintained. It should always be remembered that infants with CDH are at increased risk for postnatal respiratory failure, incarceration or strangulation of bowel, and pulmonary hypoplasia.

In cases of traumatic or adult repaired CDH, recurrence of the hernia is possible; thus, follow-up with chest radiography is important.<sup>[22]</sup>

### **Aim of study :**

Analysis of 50 cases of congenital diaphragmatic hernia and determination of difference in age, gender, indications, presentations, side effects & complications of surgery .

**Patients and methods :** A A retrospective & prospective observational study was conducted in 50 patients of congenital diaphragmatic hernia for different indications over a period of 9 years from May 2013 till May 2022 in Thi-Qar governorate. On admission,

an informative history was taken from the family of every patient including age, gender, sex, onset and nature of clinical features related to disease itself . Other data included possible chronic diseases, presence of associated anomalies, history of drug allergy, etc.....

Different investigations done including chest X-ray, ultrasonography chest CT scan, routine blood investigations, renal functions & echocardiography.

### Results:

Of the 50 cases of congenital diaphragmatic hernia, 27 of them were males and 23 females aged from less than one month up to more than one year and this sample was obtained from Bent alhuda teaching hospital. Every patient was exposed for clinical examination especially auscultation of chest. Different investigations were done including chest X-ray, chest CT scan and blood investigations, renal function test , electrocardiography ,echocardiography , bronchoscopy .

Table 1 (Sex of patients) : The incidence was slightly higher among males ( 54% ) than femles (46% )

sex	Number of patients	Percentage %
Male	27	54%
female	23	46%

Table 2 ( Age distribution ):

Age	Number of patients	Percentage %
Less than 1 month	20	40%
1 mon to 1 year	20	40%
More than 1 year	10	20%

Table 3 ( Type of hernia ):

Type of hernia	Number	Percentage %
Bochdalek	45	90 %
Morgagni	5	10%



Table 4 ( Different Presentations of patient )

Presentation	Number of patients	Percentage %
Dyspnea	30	60%
Tachypnea	30	60%
Poor oral intake	20	40%
Repeated chest infection	20	40%
Scaphoid abdomen	20	40%
Deleyed bowel motion	20	40%
Failure to thrive	20	40%
Marasmus	20	40%
Poor response to external stemuli	20	40%
Repeated vomiting	10	20%

Table 5 ( Different types of incisions )

Type of incision	Number	Percentage %
Left kocher	40	80%
Right kocher	5	10%
Upper midline	5	10%

Table 6 ( Different Perioperative Complications )

Complications	Number	Percentage %
Bradycardia during & afer surgery	5	10%
Cardiac arrest during & after surgery	5	10%
Sepsis after surgery	10	20%
Hypoxia during & after surgery	20	40%
Upper GIT bleeding	2	4%
Abdominal compartment syndrome	2	4%
Decreased left lung volume	10	20%
Mortality	5	10%

Table 7 ( Different Preoperative investigations )

Investigations done before surgery	Number	Percentage %
Chest x-ray	50	100%
Chest ct-scan	40	80%
Complete blood count	50	100%
Blood urea	50	100%
Serum creatinine	50	100%
Random blood sugar	50	100%
Abdominal U/S	20	40%

## Discussion:

In our analysis twenty seven patients were males and twenty three patients were females and both genders were nearly equal with a slight higher number of males. This differs from the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues <sup>[23]</sup> which was done over a period of 12 years with a male to female ratio of 4:1 whereas 65/83 (78%) of patients were males and 18/83 (22%) were females.

In our study, we did not encounter any neonate with bilateral CDH but Morgagni hernia or Larrey's hernia could not be identified while the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues <sup>[23]</sup> did not encounter Morgagni hernia or Larrey's hernia nor bilateral CDH. This means that our study included variable types of hernias in contrast to the Indian study which included only the posterolateral type of CDH. In our study, none of the neonates had any lethal or cyanotic congenital heart diseases but pulmonary hypertension was observed in a significant number of patients. This is similar to the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues <sup>[23]</sup> regarding congenital heart disease and pulmonary hypertension. Among the nonlethal cardiac malformations, atrial septal defect, ventricular septal defect, and patent foramen ovale predominated. Medical treatment by Sildenafil was not applied in our study and this differs from the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues <sup>[23]</sup> in which Sildenafil was attempted in moderate-to-severe degrees of PPHN with a good outcome.

Mean length of hospital stay was  $22.64 \pm 4.9$  days (median 27.5 days, range 13–32 days) among the survivors in the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues <sup>[23]</sup> but was much shorter in our study.

All of our patients underwent open surgery ( laparotomy ) and this differs from the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues <sup>[23]</sup> in which four of their patients underwent either laparoscopy or thoracoscopy.

The mortality rate in our study was 10% of those who underwent surgery and this was different from the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues <sup>[23]</sup> in which a total of 3/73 (4%) deaths was recognized. This is due to the well equipped tertiary centres in India.

All of our patients were exposed to surgery and all had been given the chance for survival by surgical intervention and those who died before surgery could be applied on them had not been recorded in our study and this differs from the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues <sup>[23]</sup> in which about ( 12% ) of neonates had been recorded in the study but had not been selected for surgery and died preoperatively due to the high mortality rate perioperatively and very low chance for survival after surgery.

ECMO facility was not available in our study and this is similar to the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues<sup>[23]</sup>.

None of the neonates required ventilation postoperatively in the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues<sup>[23]</sup> while there are several patients who required ventilation temporarily in our study.

The operation for CDH is no longer an emergency procedure. It is increasingly being recognized that stabilization of labile physiology is paramount and delayed repair is now frequently employed in most of the surgical centers. This goes with our study and with the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues.<sup>[23]</sup>

An operative correction was generally performed through an upper transverse or subcostal abdominal incision. Minimally invasive approaches such as thoracoscopy and laparoscopy have also been described in the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues<sup>[23]</sup> in only 4 patients. On the other hand, only upper midline incision, right and left subcostal incisions were applied in our study. Thoracoscopy and laparoscopy were not applied in our study.

The size of the diaphragmatic defect has been described by previous studies to be a risk factor for poor outcome of CDH. It has been shown to correlate well with mortality as well as morbidity in live born infants with CDH. Defect size is likely to be a marker for the degree of pulmonary hypoplasia. The larger the defect the more severe the pulmonary hypoplasia and pulmonary hypertension and the worse the prognosis.

No mesh was used in our study. This does not agree with the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues<sup>[23]</sup> in which polypropylene mesh in two left-sided CDH cases were used due to large diaphragmatic defect sizes, but unfortunately, they did not recover most possibly due to severe pulmonary hypoplasia and pulmonary hypertension. None of the right-sided CDH cases required mesh repair in the Indian study.

The presence of a hernial sac significantly improves the prognosis in CDH neonates which is formed of parietal peritoneum and lung pleura and has been reported in approximately 20% of cases in the literature. In our study, we found intraoperatively, the presence of sac in several neonates; all of them survived. This agrees with the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues.<sup>[23]</sup>

With regard to the contents of organs in the chest cavity, the presence of the spleen, stomach and liver in the chest cavity had been associated with mortality rate after surgery.

No recurrence of hernia was encountered in our study. This does not agree with the Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues <sup>[23]</sup> in which single case of recurrent left-sided CDH presenting as para-esophageal hernia at 9 months of age, which was successfully repaired and is doing well on 4 years follow-up. No intestinal obstruction was observed in our patients and this differs from Indian study made by [Jayalaxmi Shripati Aihole](#) and colleagues <sup>[23]</sup> in which 2 cases of adhesive small bowel obstructions were encountered postoperatively at 2 months and 8 months, respectively, which were managed by adhesiolysis

Most cases had a left diaphragmatic hernia and this agrees with the study made by [Jayalaxmi Shripati Aihole](#) and colleagues <sup>[23]</sup> in which the Left-sided CDH predominated with 75/83 (90%) and right-sided CDH were 8/83 (10%).

The prenatal detection rate for CDH varies enormously in published studies, from 10% to 79%. Most are detected after 24 weeks of gestation. However, prenatal detection of CDH is rare in developing countries due to inadequate facilities where is in our study diagnosis after birth and this agrees with the study made by [Jayalaxmi Shripati Aihole](#) and colleagues. <sup>[23]</sup>

## **Conclusions :**

CDHs are common on the left side with fairly acceptable outcome. Although, the right-sided CDH are rare; they do carry a good prognosis, as it was seen in our experience. The overall survival rate among neonates with CDH in our institutional experience was good. Males were affected by congenital diaphragmatic hernia more than females. Dypnea and tachypnea were the most common presentation before surgery. Bochdalek was more common than Morgagni hernia. Recurrence has not been recorded after surgery.

## **Recommendations:**

1-Sophisticated investigations may be mandatory for persistent respiratory symptoms in order to detect CDH which has an indication for surgery.

2-Early diagnosis of congenital diaphragmatic hernia is very important in order to improve prognosis.

3-Good preoperative preparation is crucial to prevent unwanted complications during and after surgery.

4-Careful monitoring during and after surgery may decrease morbidity and mortality.

5- Conservative measures may treat most complications during and after surgery.

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