Congenital Diaphragmatic Hernia: A Single Center Experience Over Seven Years

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Abstract

The congenital diaphragmatic hernia (CDH) is a birth defect in the fetal diaphragm allowing abdominal contents to protrude to the thoracic cavity. Presence of abdominal contents in the thoracic cavity will affect the respiratory reserve leading to respiratory symptoms mainly tachypnea. Congenital diaphragmatic hernia can be presented alone or with other congenital anomalies including cardiac, respiratory, or other anomalies. CDH can be classified as Bochdaleck, or Morgagni hernia. In 75% of cases CDH is on the left side. The aim of this study is to view the management of patients with CDH in terms of diagnosis, treatment, and survival rate. Forty- six cases were studied over seven year period from January 2012 till January 2018. They were diagnosed based on clinical examination, radiological investigations. 18 (39%) cases were 2-7 days of age. 29 (63%) of cases were presented with tachypnea. Bochdaleck's CDH is found in 44 patients (96%). 18 patients (39%) were having pulmonary hypoplasia as an associated anomalies with CDH, such anomalies affect the pulmonary reserve by putting the child in a danger of pulmonary hypertension. Treatment was via surgical repair done by team of thoracic and pediatric surgeons. In this study, we found delaying surgical repair after stabilizing the respiratory reserve leads to good postoperative survival, this was proved by high mortality rate (8 patients 67%) in patients with less than 24 hours age. The aim of this study is to show the way of management of congenital diaphragmatic hernia in terms of diagnosis, treatment, and the survival rate.

Keywords: *Congenital diaphragmatic hernia (CDH), Congenital anomalies, Bochdaleck's CDH, Morgagni's CDH.*

Introduction

The diaphragm is the muscular structure that separate the thoracic cavity from the abdominal cavity and it is essential for the respiratory process, any birth defects in this muscular structure will lead to respiratory insufficiency due to herniation of the abdominal contents up into the thoracic cavity in a spectrum of congenital anomaly known as congenital diaphragmatic hernia. It has been found that genetic and environmental factors play a role in the pathogenesis of this anomaly; nevertheless, still pathogenesis is not well understood.(1)

Congenital diaphragmatic hernia (CDH) is a relatively common malformation in which there is failure of complete fusion of the diaphragm during the prenatal period. This leads to protrusion of abdominal organs into the thoracic cavity which can result in lung hypoplasia and respiratory failure at birth. Anatomically, CDH can be classified as postero-lateral (Bockdalek,70–75%), anterior (Morgagni, 23-28%), or central (2-7%) defects.(2)

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In about 75% of cases CDH can be presented in the left side leading to the possibility of herniation of the small, or large bowel, the stomach, the spleen, the left lobe of liver, or rarely the kidney. CDH on the right side can include the right lobe of the liver, the possibility of the bowel and/or the kidney.[3] CDH occurring bilaterally is a rare presentation.[4]

The classical presenting symptoms are respiratory distress in terms of tachypnea, dyspnea; reluctant for feeding; and failure to thrive. The diagnosis is clinical and radiological. Surgical correction still the treatment of choice for CDH. The survival depends primarily on the severity of pulmonary hypoplasia, and the amount of fixed pulmonary hypertension, and secondly on the severity of associated anomalies.[5]

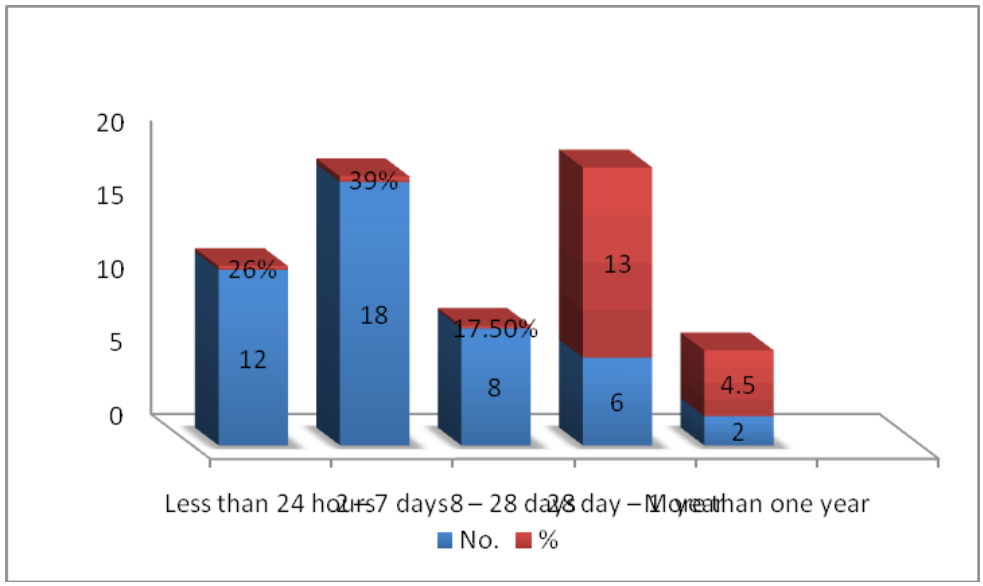
The aim of this study is to show the way of management of congenital diaphragmatic hernia in terms of diagnosis, treatment, and the survival rate.

Patients and Method

This study was carried out prospectively including forty- six children who admitted to Al-Kadhymiya Teaching Hospital with congenital diaphragmatic hernia which were proved by imaging studies (chest x-ray, Barium meal, and chest CT-scan) over a period of seven year from January 2012 to January 2018. Nine children died preoperatively despite optimal respiratory ventilator support, they were excluded from our study. Forty – six children underwent surgical correction of congenital diaphragmatic hernia. Once patients with CDH received in our outpatient unit, full investigations underwent in terms of blood investigations, radiological studies confirming the CDH and any associated anomalies; then the planning for surgical repair is ensured. The surgery is performed by an abdominal subcostal approach, the herniated abdominal viscera are reduced, and the diaphragmatic defect is inspected for the presence of any sac that should be excised. The defect then closed by interrupted sutures in case of small ones, and synthetic patches are used to close the defects in large ones. Post operatively, child is followed up in terms of respiratory failure, and mortality.

Results

Age Distribution: The majority of cases (18 cases) 39% were in their first week of age, while only (2 cases) 4.5% were more than first year of age. As shown in the graph below.



Graph No.(1) : Age distribution among patients with congenital diaphragmatic hernia

Clinical Presentation: Clinically, (29 cases) 63% with CDH were presented having tachypnea, only (12 patients) 26% presented with cyanosis. The gastrointestinal symptoms as vomiting, constipation, and abdominal distension were in (2 patients) 4.5% only. As shown in the table below.

Table No. (1) : Clinical presentation of patients with CDH

Presentation	No. of patients	Percentage
Tachypnea	29	63%
Cough(respiratory infection)	27	58%
Reluctant to feeding	23	50%
Cyanosis	12	26%
Gastrointestinal symptoms (vomiting, constipation, abdominal distension)	2	4.5%

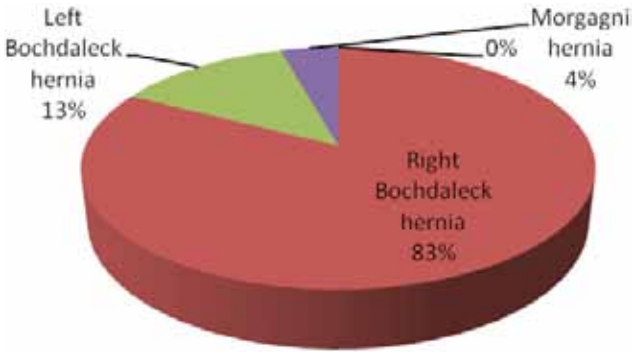


Chart No. (1) : Types of congenital diaphragmatic hernia.

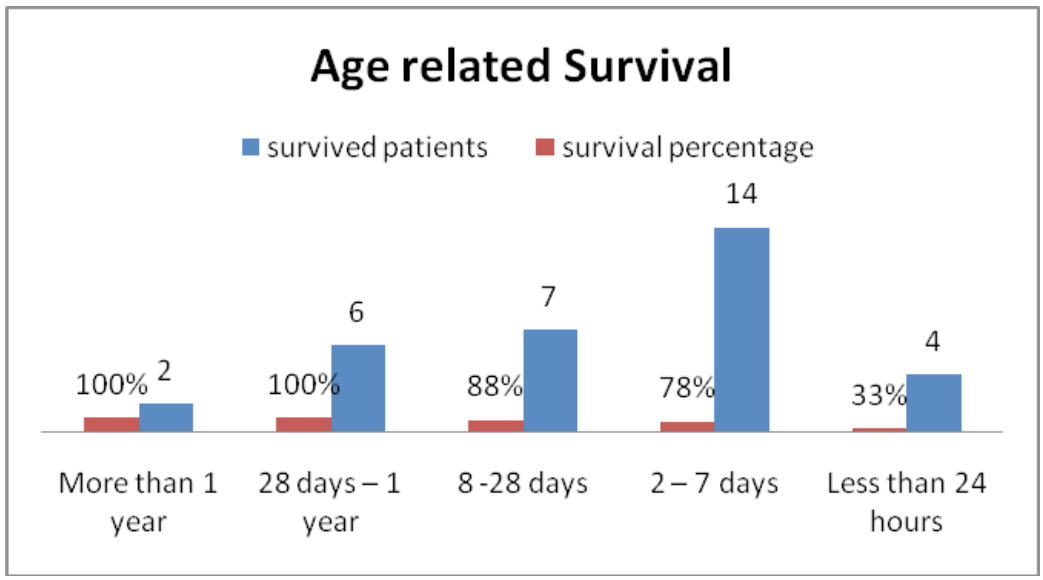
Type of congenital diaphragmatic hernia: Bochdaleck's hernia is found in 44 patients (96%), the majority (38 patients) 83% is on the right, and (6 patients) 13% are on the left. While Morgagnitype of CDH is in 2 patients only (4%). As we can see below in the Pie chart.

Associated anomalies: There are many congenital anomalies associated to congenital diaphragmatic hernia. The most common associated anomalies are pulmonary hypoplasia (18 cases) 39%, congenital heart disease is in (11 patients) 24%. See table no. 2

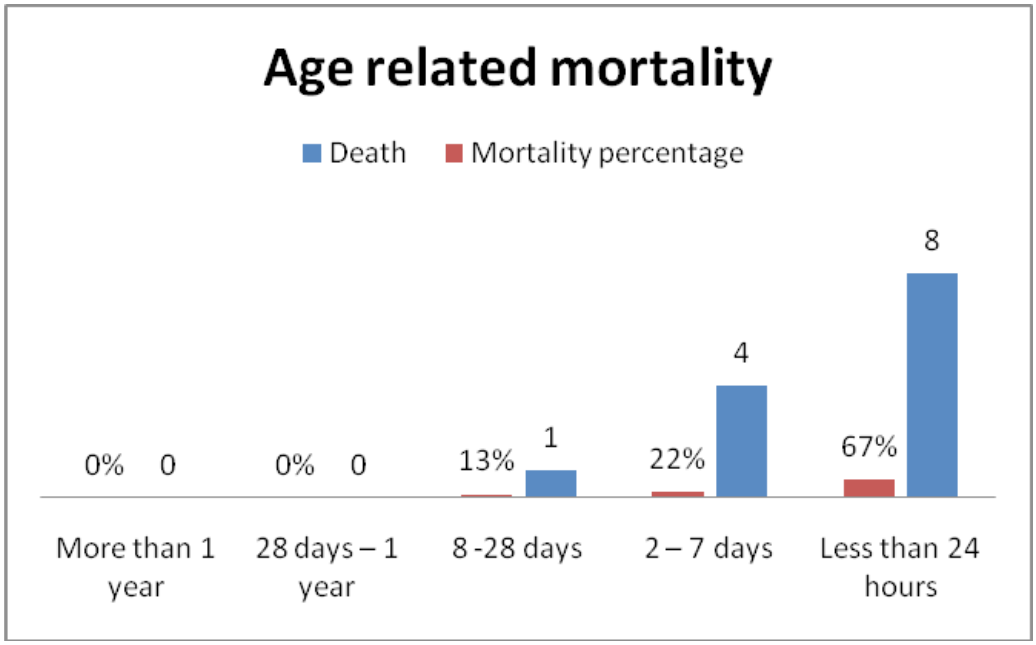
Table No. (2) Associated congenital anomalies in patients with congenital diaphragmatic hernia.

Type of congenital anomaly	No.	Percentage
Pulmonary hypoplasia	18	39%
Congenital heart disease	11	24%
Malrotation of gut	9	19.5%
Renal anomalies	3	6.5%
Meckel's diverticulum	1	2%

Age related survival and mortality: The survival rate is found to be high in patients aged 2-7 days, while a high mortality rate is found in patients below 24 hours age. As seen below.



Graph No.(2) : Age related survival in patients with congenital diaphragmatic hernia



Graph No.(3) : Age related mortality in patients with congenital diaphragmatic hernia.

Discussion

Congenital diaphragmatic hernia is a birth defect, can be presented as a sole birth defect or associated with other congenital anomalies. In our study, the frequent age of presentation was in the first week of life followed by neonates of less than 24 hours age, while an age of more than one year of age was the least frequent age of presentation. Such figures in our study may be due to lack of proper referral protocol to our pediatric surgical unit. Previously, there was a belief of early surgical repair for CDH as early as possible; now with the presence of advanced respiratory and ventilatory resuscitation delaying the surgical repair increase survival rate allowing time for relaxation of pulmonary vasculature;[6] this is what found to be in our study surgical repair in older age group reflected in lower mortality rate.

In the recent study, the survival rate is found to be higher with higher age group, these results goes with Tracy E. T. et al study.[7]

The predominant clinical presentations in our study is respiratory symptoms and signs, like Beck C, et.al study [8]

In regards to the anatomical typing of CDH, the Bochdaleck's type was the predominant one in 44 patients (96%); out of these only 6 patients (13%) were on the left while the rest 38 patients (83%) were on the right side, these results are not the same as Le, E. D. et.al study [9] the reason for this is the limited number of our patients in a relatively short period of time.

We found that pulmonary hypoplasia is a key factor influencing the survival rate for patients by causing pulmonary hypertension, it present in 18 patients (39%), the same as Pandya KA, et.al study.[10]

There was a belief that early surgical correction is the key in managing patients with CDH, but with time it was found that respiratory stabilization is the key is ensuring high survival rate among CDH patients'. In our study the mortality rate was high among patients aged less than 24 hours (8 patients 67%), so delaying the surgery leads to high survival rate after respiratory stabilization. Its goes with other studies Fallon SC, et.al. [11] and Kumar VSH [12] .

Conclusion

Congenital Diaphragmatic Hernia is a congenital pathology with major health problem, posing a high mortality rate if not diagnosed and managed properly. In our center, despite the limited facilities and capabilities but still have a fair experience in managing such pathologies. We recommend early prenatal diagnosis, genetic diagnosis, and proper postnatal care via the

establishment of well-established neonatal intensive care units (NICU), the availability of extracorporeal membrane oxygenation (ECMO) to provide a proper respiratory support, moreover; we need good training in the field of thoracocscopic surgery as a surgical tool in managing such pathologies.

Ethical Clearance: The Research Ethical Committee at scientific research by ethical approval of both MOH and MOHSER in Iraq.

Conflict of Interest: Non

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