

Congenital Diaphragmatic Hernia – Our Institutional Experience

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Abstract

Aim: To evaluate the cases of congenital diaphragmatic hernia in neonates.

Objective: To study the age of presentation, gender distribution, relation of birth weight and post-operative outcome, operative findings and their surgical management, pre-operative condition in terms of respiratory distress, cyanosis, chest retraction or ventilator support and post-operative outcome of neonates who were admitted our institute.

Material and methods: This study was conducted in 51 neonates who presented with complaints of respiratory distress, cyanosis, chest retraction etc. between December 2017 and November 2018 in the department of paediatric surgery at our institute.

Results: The male female ratio was 1.55:1 in this study. There was 4 cases of right side congenital hernia and remaining 47 were left sided. There were 29 neonates who discharge with normal respiratory pattern. The

average weight of discharge neonates was 2.65 kg. Total mortality was 43.13% at our busy centre.

Conclusion: Congenital diaphragmatic hernia is a medical emergency rather than surgical emergency. Mortality of neonates will be improved surgically after medical management of portal hypertension. Early onset of respiratory distress leads to worse outcome in neonates of congenital diaphragmatic hernia.

Keywords: Congenital, Diaphragmatic Hernia, Neonate, Pulmonary Hypoplasia, Pulmonary Hypertension.

Introduction

Congenital diaphragmatic hernia is defined as defect in the diaphragm, so abdominal contents herniate in pleural cavity causes pulmonary hypoplasia and pulmonary hypertension. Normal born child have decrease in pulmonary vascular resistance with respiration started. But neonates with congenital diaphragmatic hernia have high pulmonary vascular resistance leads to extra pulmonary right to left shunting [1]. Extra pulmonary

Right to left shunting causes altered pulmonary circulation and pulmonary hypoplasia [2]. Congenital Diaphragmatic hernia is a surgical disease, but it is a medical emergency rather than surgical emergency. Surgical repair of congenital diaphragmatic hernia is not a lifesaving emergency, present accepted concept is to establish the normal physiology of cardio-pulmonary system before surgery [3]. The incidence of CDH is 1: 2000–5000 of live births and associated with high mortality rate [4].

Material and methods

This is a retrospective study conducted in neonates who came with complaints of respiratory distress, cyanosis, chest retraction etc. This study was conducted between December 2017 and November 2018 in the department of paediatric surgery at our institute.

This study was done in 51 neonates (31 male and 20 female) between December 2017 and November 2018 in the department of paediatric surgery at our institute. A detailed history, thorough clinical examination was done in all neonates. All routine and needed investigations were done, like blood investigation, blood cross match, arterial blood gas analysis, x-ray chest and abdomen. Vital signs, hydration status, abdominal contour, history of vomiting, cyanosis, chest retraction, respiratory pattern were noted through clinical examination. Systemic examination of cardiovascular system, respiratory system, central nervous system was done.

Results

Left side is the most common site of congenital diaphragmatic hernia in our study (47 cases of left CDH, 92.15%). 7.84% neonates present with right CDH (4 cases out of 51 cases). Survival rate in our study was 56.86% and mortality rate was 43.14%.

There were 22 cases of CDH contains sac and 29 cases were without sac. Large bowel and small bowel is the

most common content which herniates in thoracic cavity in left CDH, while liver is the common content in right CDH.

In our study 29 neonates of CDH showed diaphragmatic hernia with hernial sac and 22 neonates were without hernial sac. There were 29 neonates of CDH discharge and 22 were expired. Average weight was 2.68 kg. at the time of admission of neonates who discharge and admitted on average 6.38 days of life. Average weight of expired neonates was 2.46 kg. and age 1.59 days at the time of admission.

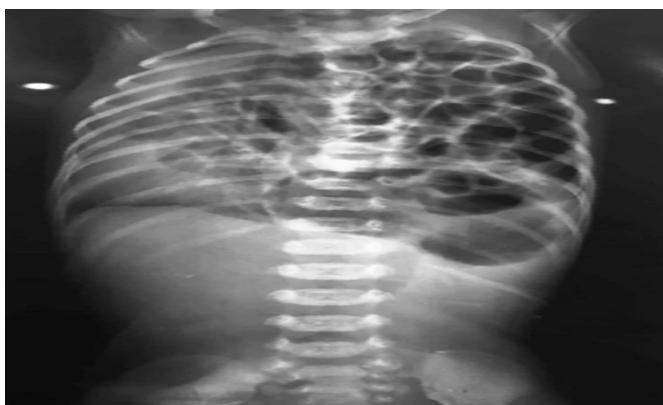


Figure 1: X-ray chest and abdomen showed multiple air-fluid level in left thoracic cavity and shifting the mediastinum towards right side.

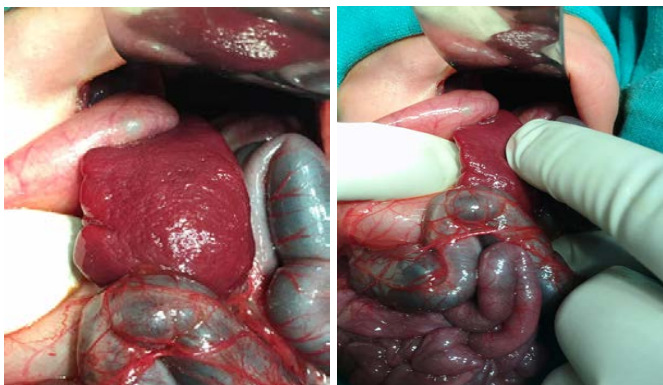


Figure : 2

Figure : 3

Figure 2 & 3: Showed intra-operative photograph of left congenital diaphragmatic hernia.

Discussion

At the end of 1980, it was the thought that CDH was the surgical emergency; surgery will provide space to lung

expansion. There was some temporary improvement and this time period of improvement was known as “honeymoon” period. But patients became deteriorate [5,6].

Congenital diaphragmatic hernia (CDH) is associated with high mortality due to pulmonary hypoplasia, pulmonary hypertension, and hidden congenital heart disease. Outcome of CDH cases varies centre to centre, availability of skilled neonatal care unit, and neonatal associated anomalies. Worldwide reported mortality rate is 20% to 60 % [7]. Mortality rate in our study was 43.13 %. Congenital diaphragmatic hernia was described by Vincent Bochdalek in 1848, and neonatal CDH was first successfully repair by Gross in 1946 [8].

The age on admission and onset of respiratory distress were high-risk factors related to the high risk factor in neonates of congenital diaphragmatic hernia. Neonates of CDH who admitted earlier due to early onset of severe respiratory distress have higher mortality [9]. In our study Mean age of admission of survived neonates was 6.38 days while mean age of expired neonates was 1.59 days. Our study data also support that neonates who presents late due to less severity of illness have low mortality compare to neonates with early presenting symptoms.

Wung et. al. reported the concept of gentle ventilation (permissive hypercapnea) strategies to prevent the iatrogenic barotrauma of lung. This novel concept characterized by securing the spontaneous ventilation of neonate, permissive levels of hypercapnea (paCO₂ 60 to 65mmHg or 9kPa) and avoidance of high inspiratory airway pressures (ideally not exceeding 25 cm H₂O). High frequency oscillatory ventilation (HFOV) has also been utilized in the perinatal management of CDH both as a ‘rescue therapy’ prior to extracorporeal membrane oxygenation (ECMO) and as a primary ventilator

modality in an attempt to reduce pulmonary barotrauma [7,10]. ECMO facility is not available at our centre.

The presence of a hernial sac significantly improves the prognosis in CDH neonates. The reported incidence of presence of sac in CDH is 20 % [11]. In our study 56.86% neonates of CDH had hernial sac. Aihole JS et. al. reported 100% survival in case of CDH with sac [9]. In our study neonates of CDH with sac had 62.07% survival. While, there was 50% survival present in neonates CDH without hernial sac in our study.

Conclusion – Early onset of respiratory distress leads to worse outcome in neonates of congenital diaphragmatic hernia. Congenital diaphragmatic hernia is a medical emergency rather than surgical emergency. Mortality of neonates will be improved surgically after medical management of portal hypertension. Early onset of respiratory distress leads to worse outcome in neonates of congenital diaphragmatic hernia.

Table 1 – Showed incidence of right CDH and left CDH with gender distribution

Diagnosis	Male	Female	Total
Right CDH	03	01	04 (7.84%)
Left CDH	28	19	47 (92.15%)
	31	20	51

Table 2 – Showed final outcome of neonates of CDH with gender distribution

Outcome	Male	Female	Total
Discharge	13	16	29 (56.86%)
Expired	18	04	22 (43.14%)
	31	20	51

Table 3 – Showed relation between final outcome, average age and average weight of neonates at the time of admission

s. no.	Outcome	Average age of neonates at admission	Average weight at admission
1	Discharge	6.38 days	2.68 kg.
2	Expired	1.59 days	2.46 kg.

Table 4 – Showed presence or absence of hernial sac in CDH

Hernial sac	Right CDH	Left CDH	Total	Discharge	Expired
Present	3	26	29 (56.86%)	18	11
Absent	1	21	22 (43.14%)	11	11
	4	47	51	29	22

Table 5 – Showed the distribution of hernial contents in right and left CDH

s.no.	Hernial contents	Neonates of right CDH	Neonates of left CDH
1	Liver only.	1	00
2	Liver and large bowel, small bowel.	3	02
3	Liver, spleen, large bowel, small bowel.	0	01
4	Spleen, large bowel, small bowel, stomach.	0	21
5	Spleen, large bowel, small bowel,	0	18
6	Large bowel, small bowel, stomach.	0	03
7	Large bowel, small bowel.	0	01

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