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Original Article

ANATOMICAL VARIATIONS OF CONGENITAL DIAPHRAGMATIC HERNIA DURING THORACOSCOPIC REPAIR :A TWO EGYPTIAN CENTERS EXPERIENCE.

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Submit Date	2019-02-20
Revise Date	2019-03-19
Accept Date	2019-03-20

ABSTRACT

Background: Congenital diaphragmatic hernia is a major problem in pediatric patients specially neonates with subsequent high morbidity and mortality. Thoracoscopic repair is now being considered the primary choice in management of such patients. The present work aims at evaluating the different anatomical characteristics and variations of congenital diaphragmatic defects during thoracoscopic repair.

Methods: Thirty patients with congenital diaphragmatic hernia were included in the study. Thoracoscopic inspection of the defect and its anatomical characters was followed by repair. Data were recorded regarding symptoms, radiological findings, anatomic landmarks and variations during the procedure itself.

Results: A higher incidence of congenital diaphragmatic hernia in infantile male population was documented with a higher incidence of left sided defect (96.7%). Postero-lateraldefects were common (86.7%) compared to lateral defect. Hernial sac was absent in 90% of cases. Most of cases had anterolateral muscular rim (86.7%). Liver herniation occurred in only two cases (6.7%). The majority of patients had moderate organ displacement (70%). Thoracoscopic repair was successful in 90% of cases. Primary repair was done in all cases with no need of synthetic patches.

Conclusion: The use of thoracoscope helps in creation of a strict phenotyping system that will enhance the ability to predict the clinical course of diaphragmatic defects and will aid in identifying developmental pathways responsible for the disease.

Keywords: Congenital diaphragmatic hernia, Thoracoscope, Diaphragmatic defect, Hernial sac, Liver herniation.

INTRODUCTION

The diaphragm is the muscle separating the thoran cic and abdominal cavities and is essential for the respiratory process. Birth defects of the diaphragm structure and function reveal its critical function.[1]Congenital Diaphragmatic Hernia (CDH) represents less than 5 per 10000 births.[2] The anatomical variation in the diaphragmatic defects are mainly in the region in which they form and in size.[1] No previous study including the detailed anatomy of Congenital Diaphragmatic Defect (CDD) has been performed, and current classification of diaphragmatic defects incompletely describe the diaphragmatic defect regarding its location and extent. Almost all congenital diaphragmatic defects (CDDs) are described to be posterolateral (Bochdalek), anterior (Morgagni), and central and anterior.[3] A recent study from the CDH Study Group

showed that the size of the diaphragmatic defect was the main factor influencing the outcome of infants with CDH and that it is the major factor associated with higher mortality rate.[4] The main cause of high morbidity and mortality associated with CDH is the lung hypoplasia. Respiratory failure is the leading cause of death in the majority of these cases despite intervention.[1] Now, the minimally invasive techniques are introduced in many complex pediatric surgery including successful repair of CDH whether with the use of laparoscope or thoracoscope.[5]

PATIENTS AND METHODS

Written informed consent was obtained from all participants and the study was approved by the research ethical committee of Faculty of Medicine, Zagazig University. The work has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for studies involving humans. This cross sectional study was conducted between July, 2015 till July, 2018 in pediatric surgery departments in Zagazig and Cairo universities. All pediatric patients diagnosed with congenital diaphragmatic hernia and cardio-respiratory stable were included in the study. Patients with acquired and recurrent diaphragmatic Hernia were excluded from the study. Thoracoscopic repair of CDH in neonates was carried out under general anesthesia and in lateral decubitus position. We introduced a 5-mm trocar below the tip of the scapula or in the midaxillary line at the level of the nipple and insufflate the thorax with 4–7mmHg of pressure. After introduction of the scope, we placed two additional trocars, between 3 and 5 mm, under vision.[5] The contents were reduced and the edge of the diaphragmatic defect was identified followed by primary repair with interrupted 2-0 Ethibond or silk sutures tied intracorporeally. A chest tube was inserted when required. When open surgery was required, a subcostal incision was done followed by reduction of the contents and primary repair of the defect.[5] Demographic data of the patients were recorded including age, sex. Careful history was obtained from parents regarding symptoms and duration. Clinical data were collected including the presence of vomiting, dyspnea, recurrent chest infection and radiological finding by X-ray or CT chest. Regarding the procedure itself, details about the anatomy of the defect and its location **(Fig. 1a, 1b**), the residual muscle rims, contents of the hernia and the presence of sac or not as well as the type of repair were recorded.

RESULTS

Twenty-three (76.6%) patients were males and seven patients (23.3%) were female (Male to Female ratio 3.3:1). Regarding the age of patients enrolled in the study, the mean age was 313.8 days (ranging from 3 days to 5 years). More than half of the patients were in the infantile age group (Table 1). In assessing the disease characteristics, it was important to document the location of the diaphragmatic defect and if it was a Rt. or Lt. sided defect. Notably, the postero-lateral defect was the commonest (86.7%) type of hernia in the studied patients and the defect was left sided in the majority of patients (93.3%) **(Fig 2)**. Regarding the pathological type of the hernia, all the patients had a true communication between the chest and abdominal cavities with no eventeration detected in any of the studied cases. However, only three patients (10%) had a hernial sac **(fig.3a)** and the sac was absent in the majority of patients (90%). To evaluate the diaphragmatic muscle available for repair, the muscular rim around the edges of the defect was evaluated in each patient (Table 2). Most of the cases (86.7%) had anterolateral muscular rim, all the patients had anteromedial muscular rim,

posterolateral muscular rim was only found in four cases each (13.3%). Fortunately, all of the patients had non-aplasia type of diaphragmatic hernia and primary repair was achieved. Similarly, the majority of patients (86.7%) did have posterior rim unfoldable from retroperitoneum (Table 3). The prognosis of congenital diaphragmatic hernia cases depends on many factors among which is the presence of associated anomalies and the herniation of the liver in the chest. In our study the liver was herniated into the chest in two cases (6.7%) **(fig.3b**). The majority of patients (70.0%) had moderate organ displacement, while mild organ displacement was found in 23.3% **(fig.4a)** and only a small group of patients (6.7%) had severe organ displacement**(Fig. 4b**). Patients diagnosed with congenital diaphragmatic hernia were admitted and managed in two different centers including; Cairo University Hospitals (Abu El-Reesh) and Zagazig University Hospitals. Regarding the operative technique for repair, in most patients (90.0%), the hernia was successfully repaired thoracoscopically and only 10% of cases needed conversion to open repair. Primary repair was successfully performed in all patients and synthetic patches were not required in any patient

Table 1 Patient demographic characteristics (age and sex).

Variable	**Patients*(n= 30)*** **mean ± SD**	
Age (days)	313.8±80	
– **Neonates**	11	36.7%
– **Infants**	16	53.3%
– **Children**	3	10%
Sex	**Number *(n=30)***	**%**
– **Male**	23	76.7%
– **Female**	7	23.3%

Table 2: Presence of muscular rim in the studied patients.

Site of Rim	**Present**		**Absent**	
	No.*(n= 30)*	**%**	**No.*(n= 30)***	**%**
Anterolateral	26	86.7%	4	13.3%
Anteromedial	30	100%	0	0
Posterolateral	4	13.3%	26	86.7%
Posteromedial	30	100%	0	0

Table 3 Severity of defect and posterior rim unfoldable from retroperitoneum in the studied group.

Variable	Number *(n= 30)*	Percent (%)
Severity of defect according to size	30	100%
– Non-aplasia with primary repair (A&B)	0	
– Non- aplasia with patch repair (C)	0	
– Aplasia with patch repair (D)		
Posterior rim unfoldable from retropertonium		
– Non	6	20%
– Lateral	24	80%

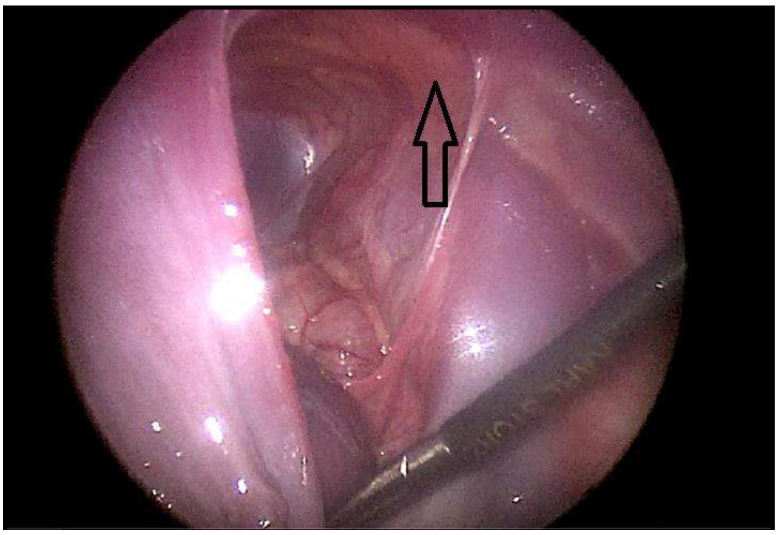

Fig. 1a: Location of defect: antrolateral with defect in lateral muscular rim

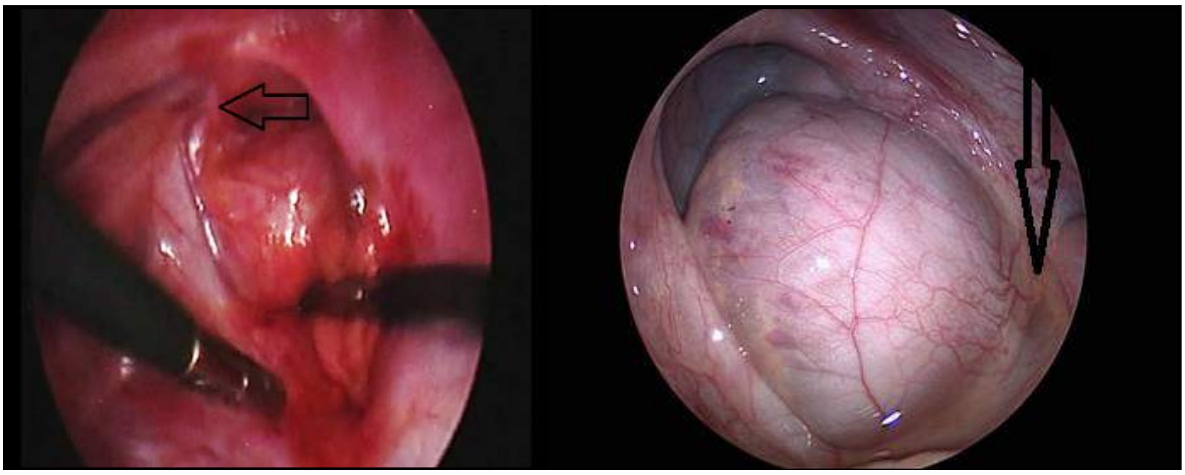

Fig.1b Location of defect: posterolateral defect (A) lt. sided CDH & (B) Rt. sided CDH.

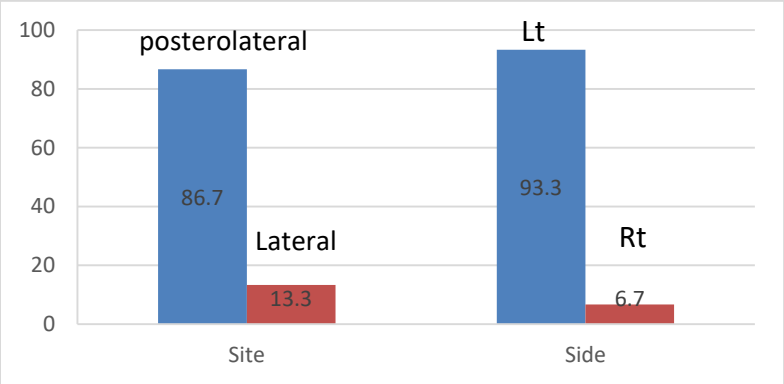

Fig. 2 Bar chart showing site and side of defect in the study group.

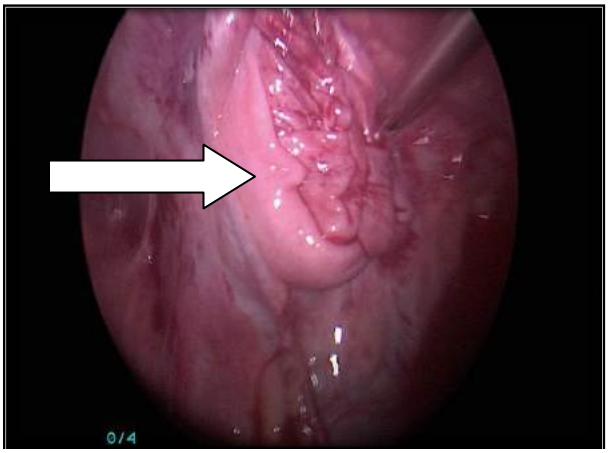

Fig. 3a The arrow pointing at the hernia sac

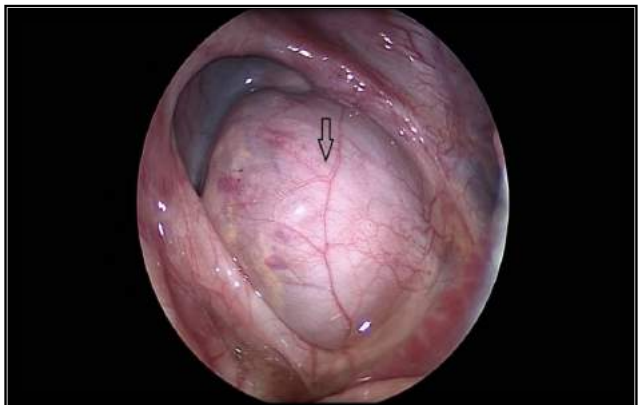

Fig. 3b Liver herniation into chest.

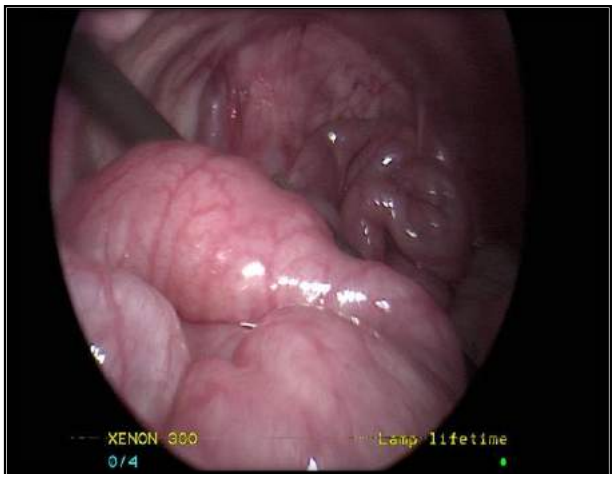

Fig. 4a Detection of organ displacement: Small bowel herniated into the chest (> 1/3 of thoracic cavity).

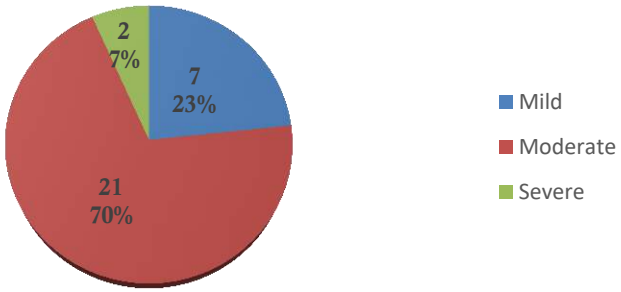

Fig. 4b Pie chart for severity of organ displacement in the patients.

DISCUSSION

Congenital diaphragmatic defects remain a significant cause of morbidity and mortality in neonates, and prognosis continues to be difficult to be interpreted in many cases. Although comorbid congenital malformations or genetic syndromes significantly affect survival, it is also reported that larger-sized defects of the diaphragm are associated with worse outcome.[3]The present study showed a male predominance with male to female ratio of 3.1:1, which is close to the results obtained by Chandrasekharan et al.[6]who reported a male to female ratio 2:1.Moreover, in a study done by Georgescu et al.[7], they mentioned a ratio of 6:1 and Tovar[2]also reported a male predominance in his results. Late-presenting congenital diaphragmatic hernia (CDH) has been defined a CDH diagnosed after the neonatal period due to initial symptoms after the neonatal period or asymptomatic CDH found in the course of routine X-ray examination of the chest beyond the neonatal period.[8] The incidence of late-presenting CDH has been quoted to be as high as 5-45.5% of all cases of CDH. Despite the highly quoted overall mortality for CDH, the prognosis for late-presenting CDH is usually favorable due to milder or absent associated complications such as pulmonary hypoplasia and hypertension.[8]

Most of patients included in this study presented in the infancy period (53.3%) while only (36.7%) presented during neonatal period. This was in contrast to the result obtained by Kim and Chung[8]which showed that the incidence of late-presenting congenital diaphragmatic hernia was (18.9%) and Tsao and Lally[9]who also reported that most patients with CDH will be diagnosed within the first 24 hours of life, and that as little as (20%) may present outside the neonatal period. The higher incidence of CDH in infants reported in the present study may be due to the fact that the inclusion criteria required cardiorespiratory stability, which is needed for successful thoracoscopic repair as Bliss et al.[10]reported that the sensitivity of neonates to acidosis and hypercapnia due to impaired elimination of Co_2 is more than in infants resulting in more respiratory distress symptoms in neonates than infants which may contraindicate a trial of thoracoscopic repair. Another explanation for the older age at presentation may be attributed to delayed diagnosis of CDH cases who were born asymptomatic and were diagnosed later incidentally by pediatricians during routine investigations of infants with repeated chest infection. Regarding the Disease Characteristics, this study showed predominant Lt. sided defect (96.7%) with only (3.3%) of defect on the Rt. side which is near to the results reported by Kosiński and Wielgoś[11]; Malekzadegan and Sargazi[12]reporting that the hernia occurs on the right side in15–20% of cases and in 80–85% of subjects on the left side. Several studies have shown that infants with right-sided CDH have a lower survival rate than those with left-sided lesions (50 vs. 75%) which may be attributed to that infants born with Rt. CDH may demonstrate increased ECMO requirements when compared with neonates with Lt. CDH. Moreover, surgical repair may be more challenging in Rt. CDH because of near-universal liver herniation. These operative challenges are implied by the increased requirement for prosthetic material in right sided CDH vs left sided CDH repair (76% vs 41% respectively).[13]Our study showed variations in the location of the defect with predominant posterolateral defect in (86.7 %) and lateral defect in (13.3%) which are similar results to those published by Ackerman et al.[3]including 41 patients with34cases (90%) with posterolateral defect and 2 cases (4%) with lateral defect and 3 cases with central defect (12%). The present study showed absence of a posterolateral muscular rim in 26 cases (86.7%)and anterolateral in 4 cases(13.3%)with intact posteromedial and anteromedial muscular rim in all cases (100%).The posterior rim of diaphragm was unfoldable from retro-peritoneum in (80%) of cases. Furthermore, the hernial sac was present only in 3 cases (10 %) that is in agreement with the results of Panda et al.[14];Spaggiari et al.[15] and Zamora et al.[16]with incidence of 14.3%,5.6%20% respectively. Presence of hernial sac is associated with good postoperative outcome as it prevents the upward movement of the abdominal organs and decreases the risk of compression of the lungs and thereby there is less risk and less degree of pulmonary hypoplasia.[14] Regarding the size of defect, according to congenital diaphragmatic hernia study group (CDHSG) classification, all cases (100%)were in class B which was classified in the past, as non-aplasia without patch repair. According to Morinietal.[4], the incidence was (15%) in class A,(40%) in class B, (32%) in class C and (13%) in class D. The discrepancy in results may be due to the limited number of cases in our study and patient selection in inclusion criteria which require patient with cardio-respiratory stability that leads to absence of cases in class C and D. Great relation between size

of defect and GIT, pulmonary and neurological morbidities were reported[17]with increase morbidity with increasing size of defect. Liver herniation was found in 2 cases (6.7%), both on the right side of the diaphragm. The presence of liver herniation into the chest (liver up) is considered to be a poor prognostic indicator. In a recent systematic review which included 407fetuses with liver up infants and 303 with the liver down infants, statistically significant worse survival was found in liver up infants (45.4%) compared with liver down infants (73.9%). Also, fetuses with the liver up required extracorporeal membrane oxygenation (ECMO) in 80 % of cases, compared with 25% for those with liver down, and overall survival rate was 45%, compared with 93% for those with liver down.[18] In the present study, the majority of patients (70.0%) had moderate organ displacement, while mild organ displacement was found in 23.3% and only a small group of patients (6.7%) had severe organ displacement . Bedoyanet al.[19]reported that the worst indicators of severity were pulmonary hypoplasia and the presence of liver, stomach, or spleen in the chest; the survival of these cases ranged from 41% to 48%.The average survival of patients with herniation of liver, stomach, or spleen was 46%, which is significantly less than the 61% survival of CDH cases with bowel herniation. Regarding the repair Characteristics 50% of patients were admitted and operated at Abu El-Reesh Cairo University Hospital and 50% in Zagazig University Hospital. Regarding the type of repair done, thoracoscopic repair was successfully done in 90% of cases while conversion to laparotomy was required in 10% of cases due to both technical and ventilatory problems. Costerus et al.[20]reported conversion rate of thoracoscopic surgery to open surgery (3.4–75.0%) and Kim[8]reported conversion to open repair in 20% of patients because of the need for patch closure or intraoperative instability. All defects in our study were repaired primary without need for synthetic patch. On the other hand, Tsai et al.[22]reported that 53% of cases required patch repair and Jawaid et al.[23]mentioned that 31% of their patients required a patch repair of their defect. This discrepancy of results may be attributed to smaller sample size in our study in comparison to other studies and patient selection criteria

CONCLUSION

Congenital diaphragmatic defects show significant anatomical variations.Left sided defects are predominant in 93.3% of patients, mainly posterolateral (86.7%). Hernial sac is rarely present in CDH. Moreover, Liver herniation is a rare event in CDH overall (6.7%), but it is common with right side defects.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

Funding information

None declared

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To cite this article: Ahmed Osama Hassan[1], Ahmed Ezzat Mohamed Rozik[1], Ismail Mohamed Ali Tantawy[1], Mohamed Magdy El Barbary[2]. Anatomical Variations Of Congenital Diaphragmatic Hernia During Thoracoscopic Repair :A Two Egyptian Centers Experience..,Egypt.ZUMJ 2019;25(3);430-438,DOI: 10.21608/zumj.2019.9781.1062