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Original Research Article

Types and Outcome of Congenital Diaphragmatic Hernia in Children in Basrah

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Abstract

A prospective study that review the types of congenital diaphragmatic defects in pediatric age groups in Basra and their outcome, to describe the demography of diaphragmatic defects and to assess their mortality and morbidity. This study was conducted at the neonatal intensive care unit and surgical ward of the Basra children specialty hospital. The medical records of 67 diaphragmatic defects patients, admitted to the hospital from July 2013and July 2015. Data for patient demographics, associated congenital anomalies, and mortality were collected in this study in addition to the types of diaphragmatic defects. In this study Bochdalek hernia is the commonest type of diaphragmatic defects in all age groups (64.2%), followed by diaphragmatic event ration (13.4%), hiatus hernia (11.9%), Morgagni hernia (4.5%), congenital central hernia (4.5%), and finally absent hemi diaphragm (1.5%). Male is affected more than female in all age groups. Shortness of breath is the presenting feature in all age groups with predominance in neonates. Vomiting is the second presenting feature, especially in infant and older children. Associated anomalies occur in about 19.4%. Overall complication rate was 24.1 and the survival rate was 80.3%. It may not reflect the real survival because many fetal and post-delivery deaths occurred and not registered in our society.One should have a high index of suspicion regarding diaphragmatic defects especially those neonates presenting with shortness of breath. Most neonates with diaphragmatic defects are diagnosed by plain chest x-rays (88.2%) so that we should avoid injudicious use of contrast study or CT scan for the diagnosis of diaphragmatic defects. High survival rate may not reflect the real event because many patients died before, during, or just after delivery so that prenatal diagnosis and management of diaphragmatic defect must be encouraged.

Key words: Congenital diaphragmatic hernia, hiatus hernia

الخلاصة

يهدف البحث لاستعراض انواع عيوب الحجاب الحاجز الخلقية في الفئات العمرية المختلفة ودراسة أثارها السريرية بالإضافة الى وصف التكوين الديموغرافي لعيوب الحجاب الحاجز و تقييم معدل الوفيات والاعتلال.

أجريتهذه الدراسة في وحدة العناية المركزة وقسم الجراحة في مستشفى البصرة التخصصي للأطفال وتمت دراسة 67 حالة لمرضى العيوب الخلقية للحجاب الحاجز للفترة من حزيران 2013الى حزيران 2015. تم جمع البيانات الديموغرافية, التشوهات الخلقية المرتبطة, والوفيات للدراسة والتحليل بالإضافة الى دراسة الأنواع في الفئات العمرية المختلفة.تبين ان فتق بوكد الك (Bochdalek) هو الاكثر شيوعا في جميع الفئات العمرية (64.2%) يليه رخاوة الحجاب الحاجز (13,4%). كما تبين تاثر الذكور اكثر من الاناث في جيع الفئات العمرية. ضيق التنفس هو العلامة الرئيسية لهذه العيوب في جميع الفئات العمرية وخاصة حديثي الولادة يليها التقيء بالمرتبة الثانية خاصة في الأطفال الرضع والأكبر سنا. تحدث الحالات الخلقية المرتبطة بنسبة 19,4% ومعدل المضاعفات المصاحبة 24,1%ز اما معدل البقاء على قيد الحياة فهو 80,3% الذي قد لا يعكس النسبة الحقيقية للوفاة لان العديد من الوفيات الجنينية وما بعد الولادة مباشرة غير مسجلة في مجتمعناما يتطلب تشجيع التشخيص للعيوب الخلقية للحجاب الحاجز قبل الولادة وأحالتها الى المراكز المتخصصة.

الكلمات المفتاحية:فتق الحجاب الحاجز الولادي, فتق الفرجه الحجابية.

Introduction

Congenital diaphragmatic hernias are complex and life-threatening lesions that are not just anatomicd efects of the diaphragm, but represent a complex set ofphysiologic derangements of the lung, the pulmonary vasculature, and related structures, diaphragmatic hernias remain an important cause of perinatal morbidity and mortality worldwide [1, 2].The management of posterolateral congenital diaphragmatic hernia remains a challenge for pediatric surgeons[1,2].The incidence of congenital diaphragmatic hernia has been reported between 1 in2000 to 5000 births [3].Approximately,80% of congenital diaphragmatic hernia are left sided [1,4].Although previously thought to be low, the incidence of associated malformations in infants with a congenital diaphragmatic hernia ranges from 10-50%. Cardiac anomalies have been found in 24% of infants[5].The diaphragm is derived from the septum transversum, the two pleuroperitoneal membranes, muscular components from somites at cervical segments three to five, and the mesentery of the esophagus [6].Newborns with congenital diaphragmatic hernia typically present with respiratorydistress. The infants will often have a scaphoid abdomen and anincreased chest diameter.[1]Small diaphragmatic hernias may present with respiratory or gastrointestinal symptoms in later childhood [7].The diagnosis of congenital diaphragmatic hernia is typically made by chestradiography. In rare circumstances, a contrast radiographis necessary. Occasionally, congenital diaphragmatic hernia may beasymptomatic and discovered incidentally [8,9].

Overall survival was 64% in which birth weight and 5-minute Apgar scores had the strongest correlation [1].Anterior diaphragmatic hernias of Morgagni account for less than 2% of all congenital diaphragmatic hernias [1].Eventration of the diaphragm is an abnormal elevation ofthe entire hemi diaphragm or, more commonly, the anterior aspect of the hemidiaphragm. This elevation produces a paradoxical motion of the affected hemidiaphragm[10].

The aim of this study was to describe the epidemiological characteristics of diaphragmatic defects in pediatric age groups in Basrah, to study the types of diaphragmatic defects in pediatric age groups and their clinical implications, and to analyze the mortality rate of different types of congenital diaphragmatic defects.

Materials and Methods

This study was conducted at the neonatal intensive care unit and surgical ward of the Basra children speciality hospital. The medical records of 75 diaphragmatic defect patients, admitted to the hospital from July 2013 and July 2015. Seven cases were excluded due to incomplete data and one traumatic patient also excluded, so that a total of 67 cases were studied.Data collecting form for age ,sex, patient demographics, associated congenital anomalies, and mortality were collected in addition to the type of diaphragmatic defects.

Results

Bochdalek hernia is the commonest type of diaphragmatic defects in all age groups (64.2%); furthermore it is the predominant type in neonates (91.2%). The incidence of diaphragmatic eventration and Morgagni hernia increases with advancing age. Hiatus hernia has higher presentation in infancy (30% of all defects in infants, and 75% of hiatus hernia occurred in infants), followed by older children (25%) with no reported occurrence in neonates. Overall, the frequency as follow: Bochdalek hernia (64.2%), diaphragmatic eventration (13.4%), hiatus hernia (11.9%), Morgagni hernia (4.5%), congenital central hernia (4.5%), and finally absent hemidiaphragm (1.5%).

Table 1: Age related types of diaphragmatic defects

AGE	TYPES						
	Bochdalek	Eventration	Morgagni	Central	Absent hemi-diaphragm	Hiatus hernia	TOTAL
Neonates	31 91.2%	2 5.9%	0 0%	1 2.9%	0 0%	0 0%	34 50.7%
Infants	7 35%	4 20%	1 55	1 5%	1 5%	6 30%	20 29.9%
>12 months	5 38.5%	3 23.1%	2 15.4%	1 7.7%	0 0%	2 15.4%	13 19.4%
Total	43 64.2%	9 13.4%	3 4.5%	3 4.5%	1 1.5%	8 11.9%	67 100%

Male was affected more than female in all age groups with an overall ratio of 1.39·The majority of patients were term 97%.

Shortness of breath was the presenting feature in all age groups (76.1%) with predominance in neonates (97%). Vomiting is the second presenting feature, especially in infant and older children (16.4%). Four patients presented with recurrent chest infections (2 infants and 2 older children).One patient only presented with hematemesis (infant).

Table 2: Age related presentation of diaphragmatic defects

AGE	Presenting features							
	Dyspnoea		Vomiting		hematemesis		Recurrent respiratory infections	
	No.	%	No.	%	No.	%	No.	%
Neonates	33	97%	1	3%	0	0%	0	05
Infants	11	55%	6	30%	1	5%	2	10%
>12 months	7	53.8%	4	30.8%	0	0%	2	15.4%
Total	51	76.1%	11	16.4%	1	1.5%	4	6%

Associated anomalies occur in about 19.1% (4 cases had no comments about associated anomalies), especially affecting neonates 22.8%. Congenital heart disease occurred in 8 cases (12.7%), and represent 66.7% of all associated anomalies. Otheranomalies included renal, Meckel`s diverticulum, pectus excavatum and congenital cataract, one for each (1.6%).

Table3:Age related associated anomalies

AGE	Associated anomalies					
	Present		Absent		Total	
	No.	%	No.	%	No.	%
Neonates	7	22.8%	24	77.4%	31	100%
Infants	2	10%	18	90%	20	100%
>12 months	3	15%	9	75%	12	100%
Total	12	19.1%	51	80.9%	63	100%

Overall complication rate was 24.1 (14 cases). No significant differences in complication rate found in different age groups (neonates =24.1%, infants=23.5%, older =25%). Simple wound infection occurred in 2 neonates (3.4%), while burst abdomen in one infant (1.7%), requiring re-exploration and did survive. Early adhesions, lung collapse, uremia in 2 caseeach (3.4%). Severe respiratory infections, gastric volvulus, anemia, and hypoglycemia in one case each (1.7%).

Table 4: age related complications

AGE	Complications				
	Present		Absent		Total
Neonates	7	24.1 %	22	75.9%	29
Infants	4	23.5 %	13	76.5%	17
>12 months	3	25%	9	75%	12
Total	14	24.1 %	44	75.9%	58

Most neonates with diaphragmatic defects are diagnosed by plain chest x-rays (88.2%). Overall 44 patients (65.7%) were diagnosed by plain chest x-rays only. The need for contrast study or CT scan is increased by advancing age. Sixteen patients (23.8%) required contrast study to confirm diagnosis, while 7 cases (11%) were diagnosed by CT scan.

Table 5: Age related diagnostic modalities

AGE	Diagnostic modality					
	Chest x-rays		Contrast study		CT scan	
	No.	%	No.	%	No.	%
Neonates	30	88.2%	2	5.9%	2	5.9%
Infants	7	35%	9	45%	4	20%
>12 months	7	53.9%	5	38.5%	1	8%
Total	44	65.7%	16	23.8%	7	11%

Survival rate was 80.3% (n= 53).High mortality occurred in neonates and older children (23.5%, 25% respectively), while best survival encountered in infancy mortality 10%.

Table 6: Age related survival

AGE	Survival					
	Alive		Died		Total	
	No.	%	No.	%	No.	%
Neonates	26	76.5%	8	23.55	34	100%
Infants	18	90%	2	10%	20	100%
>12 months	9	75%	3	25%	12	100%
Total	53	80.3%	13	19.7%	66	100%

Mortality rate also differ according to type of the defects and was as follows: Bochdalek hernia 21.5% (n=9), Morgagni hernia 33% (n=1), congenital central hernia (0), absent hemidiaphragm(0), diaphragmatic eventration 22.2% (n=2), and hiatus hernia 12.5% (n=1).

Table 7: Type related survival

AGE	Survival				
	Alive		Died		Total
	No.	%	No.	%	No.
Bochdalek	33	78.65	9	21.4%	42
Eventration	7	77.8%	2	22.2%	9
Morgagni	2	66.7%	1	33.3	3
Central	3	100%	0	0%	3
Absent hemidiaphragm	1	100%	0	0%	1
Hiatus hernia	7	87.5%	1	12.5%	8
Total	53	80.3%	13	19.7%	66

One patient discharged on family response

Discussion

Bochdalek hernia is the commonest type of diaphragmatic defects in all age groups (64.2%), followed by diaphragmatic eventration (13.4%), hiatus hernia (11.9%), Morgagni hernia (4.5%), congenital central hernia (4.5%), and finally absent hemidiaphragm (1.5%). Hiatal hernias account for only 9% of diaphragmatic hernias in infants younger than 1 year [11].About half of congenital diaphragmatic defects occur in neonates (50.7%). Infants constitute 29.9% of these defects, while older than 1 year old child are affected by 19.4%.

Bochdalek hernia occur in 31 neonates (72.1%), 7 infants (16.3%), 5 children older than 1 year (11.6%). The younger age is 4 hours and the oldest is 4 years. So that one should keep in mind even old child may have asymptomatic Bochdalek hernia or may present with gastrointestinal symptoms[11].The mean age of presentation is as follows: absent hemidiaphragm (2.9 months), Bochdalek hernia (3.8 months), central diaphragmatic hernia (1.1 year), diaphragmatic eventration (1.2 years), hiatus hernia (1.5 years), and Morgagni hernia (1.6 years).

Regarding congenital diaphragmatic hernia (49 cases), Bochdalek hernia constitutes 87.8%, similar to other study [1].Morgagni hernia and central diaphragmatic hernia occur in 3 cases, each (6.1%), Morgagni hernia occurs in 9% to 12% [11].The majority of Bochdalek hernia are left sided (36 of 43, 83.7%). Right sided Bochdalek hernia occurs in 7 cases (16.3%),which are similar to other studies[1,4], although no bilateral Bochdalek hernia encountered in this study. Male are affected more than female in a ratio of 1.3, similar to [1].All affected patientsare term which is different from other study where preterm predominate [12].About 60% of patients have body weight of more than 2.5 kg., differ from other[12]. Shortness of breath is the presenting feature in all age groups with predominance in neonates.

Vomiting is the second presenting feature, especially in infant and older children which is similar to other study[1].CXR is the preferred imaging study for diagnosing congenital diaphragmatic defects especially in neonates. Contrast study and CT scan may be mandatory for the diagnosis especially in infant and older children.

Associated anomalies occur in about 19.4%, which is relatively lower than found in other studies[1,11], and may be attributed to the fact that many newborns with associated anomalies died before achieving surgical correction. Congenital heart disease occurred in 8 cases (12.7%), lower than other series.[5,11](Arnold, 2012; George A. *et al*, 2008) Congenital heart disease represents 66.7% of all associated anomalies, higher than other study[13].Other anomalies included renal, Meckel`s diverticulum, pectus excavatum and congenital cataract, one for each (1.6%).

Hernia sac present in 7 cases of Bochdalek hernia (16.3%), all on the left side. Sac present in all Morgagni hernia and absent in central diaphragmatic hernia.

Overall complication rate was 24.1%. No significant differences in complication rate found in different age groups (neonates =24.1%, infants =23.5%, older =25%).

Survival rate was 80.3% which is relatively high as compared with other studies[1,14,15,16].It may not reflect the real survival because many fetal and post-delivery deaths occurred and not registered in our society. High mortality occurred in neonate and older children (23.5%, 25% respectively), while best survival in infancy mortality 10%which is similar to other study [17].

The highest mortality was in patient with Morgagni hernia may be due to rarity of the defect and less experience to deal with or due to late presentation with recurrent or severe chest infections. Again high mortality found in patient with diaphragmatic eventration that demand attention.

Conclusions

1. Many patients died during delivery or just after that so we encourage prenatal diagnosis with aggressive safe resuscitation

and early referral to specialized centre to save those neonates with low Apgar score.

2. Shortness of breath is the main presenting feature of diaphragmatic defects so that one should have a high index of suspicion regarding diaphragmatic defects in any neonate presenting with shortness of breath.

3. Unexpectedly high mortality noticed in patient with Morgagni hernia and eventration that demand extra care and prolonged detailed study to find out the exact causes and the best way to manage.

4. Avoid injudicious use of contrast study or CT scan for the diagnosis of diaphragmatic defects in order to avoid respiratory aspiration or unnecessary exposure to radiation.

References

1. KuoJenTsao, Kevin P. Lally; congenital diaphragmatic hernia and eventration; George W. Holcomb III, Patric J. Murphy, Daniel J. Ostlie. Ashcraft's Pediatric Surgery, 6 E (2014); 24: 327.
2. PremPuri; congenital diaphragmatic hernia and eventration; P. Lumley J. R. Siewer. Springer surgery atlas series (2006); Ch. 13, 114.
3. Harrison M.R, deLorimier A.A: Conge nital diaphragmatic hernia. *SurgClin North Am* 1981; 61:1023-1035.
4. Pediatric surgery, F. Charles Brunicardi, MD, FACS, Schwartz's Principles of Surgery, Ninth Edition (2010); 39:2728.
5. Charles J. H. Stolar and Peter W. Dillon; congenital diaphragmatic hernia and eventration; Arnold G. Coran, MD, pediatric surgery, 7E (2012); Ch. 63, 810.
6. The gut tube and the body cavity; T.W. Sadler, Ph. D langman`s, 12 E (2012); Ch. 7, 94.
7. Principle of pediatric surgery, Norman S. Williams MS FRCS FMed Sci. Bailey and Love Short Practice of Surgery 26th E (2010); 8: 119
8. Berman L., Stringer D., Ein S.H., et al: The late-presenting pediatric Bochdalekhernia: A 20-year review. *J PediatrSurg* 1988; 23:735-739.
9. Weber T.R., Tracy Jr. T., Bailey P.V., et al: Congenital diaphragmatic hernia beyond infancy. *Am J Surg* 1991; 162:643-646.
10. Waldemar A. Carlo; respiratory tract disorder; Robert M. Kliegman, MD; Nelson Textbook of pediatrics; 19th edition; 2011; Ch. 95: 1075
11. George A. Taylor &Omolola M. Atalabi& Judy A. Estroff, Imaging of congenital diaphragmatic hernias, 8 July 2008, # Springer-Verlag 2008
12. García AM, Machicado S, Gracia G, Zarante IM, Risk factors for congenital diaphragmatic hernia in the Bogota birth defects surveillance and follow-up program, Colombia, PediatrSurg Int. 2015 Nov 16.
13. Gischler SJ, van der Cammen-van Zijp, M.H., Mazer P, et al. A prospective comparative evaluation of persistent respiratory morbidity in esophageal atresia and congenital diaphragmatic hernia survivors.
14. J Pediatr Surg. 2009; 44:1683-1690. Langham MR Jr[1], Kays DW, Ledbetter DJ, Frentzen B, Sanford LL, Richards DS, congenital diaphragmatic hernia, Epidemiology and outcome, ClinPerinatol. 1996 Dec; 23(4):671-88. 48.
15. Trachsel D, Selvadurai H, Bohn D et al (2005) Long-term pulmonary morbidity in survivors of congenital diaphragmatic hernia. Pediatric Pulmonol 39:433–439
16. Brown RA, Bösenberg AT (2007) Evolving management of congenital diaphragmatic hernia. PaediatrAnaesth 17:713–719
17. Cigdem MK, Onen A, Otcu S et al (2007) Late presentation ofBochdalek-type congenital diaphragmatic hernia in children: a23year experience at a single center. Surg Today 37:642–645.