CONGENITAL HERNIAS OF THE DIAPHRAGM IN CHILDREN

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Background: Congenital diaphragmatic hernia (CDH) is a major congenital malformation. Different types have been described. Bochdalek hernia (BH) remains most prevalent with high mortality rates. Other variants are less common and carry good prognosis. Although, the diagnosis can be made antenatally, the presentation may be delayed. There is paucity of national literature on CDHs. We present our experience with these challenging paediatric malformations. **Methods:** Medical records of 18 patients (≤14 years) treated by the group of authors between October 1998 and April 2002 were retrospectively reviewed and demographic data, clinical presentation, morbidity and outcome were studied. **Results:** There were 13 (72%) children with Bochdalek hernia, 2 (11%) with eventration of the diaphragm, 2 (11%) with hiatus hernia and 1(6%) with a Morgagni hernia. The lesions were more common in girls and all the defects were left sided. The average age at the time of presentation of BH was 23 hours (2 to 72 hours) commonly presenting with cyanosis and respiratory distress. Associated anomalies were documented in 10 (77%) cases; six had multiple malformations and four died before surgery. In other types of CDHs, the mean age at presentation was 39 (18-60) months and they mostly presented with recurrent respiratory tract infections and/or mild gastrointestinal symptoms. Diagnosis was made on history, physical examination, plain x-ray chest, and gastrointestinal contrast study, when required. 14 (78%) children were operated and a sac was present in 5. Post-operative complications occurred in 5 (55%) patients with Bochdalek hernia, which were managed conservatively. The overall survival rate was 67% (n=12). **Conclusion:** The different types of CDHs presented from neonatal age to later childhood with distinct symptoms. Surgery was safe and effective. Higher morbidity and mortality was observed in newborns with Bochdalek hernia.

Keywords: Congenital diaphragmatic defects; Bochdalek hernia; Congenital diaphragmatic eventration; Morgagni hernia; Hiatus hernia

INTRODUCTION

Congenital diaphragmatic hernia (CDH) occurs when the abdominal viscera migrate upward through a defect in the diaphragm. The different variants include posterolateral (Bochdalek) defect, congenital eventration of diaphragm, retrosternal (Morgagni) hernia and hiatus hernia; each is defined by its specific location and characterized by unique clinical features. CDH occurs 1 in 2000 to 4000 births and account for 8% major congenital malformations[1]. Although, the diagnosis can be made antenatally[2-4] the presentation may be delayed[5]. In spite of the modern advances in diagnosis and the management, the mortality rate for Bochdalek hernia (BH) remains high; however the neonates that present late have a better survival than those who present early in the postnatal period[6]. Congenital eventration of diaphragm (CED) may occur as a focal lesion or affect the entire diaphragm and its clinical course varies with the extent of involvement[7,8]. Morgagni hernia (MH) and Hiatus hernia (HH) are less common variants and comprise 2% of CDHs with better survival rate; they often present in late childhood with gastrointestinal and/or respiratory symptoms[9].

There is paucity of national literature on CDHs in children and in this report we present our experience with various types of CDHs in children from southern Sindh, Pakistan, focusing on the clinical spectrum, morbidity and outcome.

MATERIAL AND METHODS

This study is based upon a retrospective review of clinical charts of patients treated by the authors with a diagnosis of congenital diaphragmatic hernia during October 1998 to April 2002 inclusive, in Hyderabad, Pakistan. Collected data consisted of demographic information, type of hernia, associated malformations, onset of symptoms, age at presentation, methods of diagnosis, management and outcome. The institutional review boards granted approval of this study.

RESULTS

The study population comprised of 18 patients with various types CDHs including BH, CED, MH and HH. The lesions were more common in girls (M: F ratio 1:3.5) and all the defects were documented on the left side. Their clinical data is summarized in Table 1.

There were 13 newborns with BH; eleven (85%) were born at home and two (15%) in a hospital.

Table 1. Clinical data of patients with the congenital hernias of the diaphragm

No	Age*	Sex	Presenting Symptoms	Side	Diagnostic tools	Surgical treatment	AM	Outcome
Bochdalek hernia (BH)								
1.	1 ½ d	F	Respiratory distress	L	Plain X-ray chest	Laparotomy, direct closure of defect	N	Survived
2.	2 hrs	F	Cyanosis and respiratory distress	L	Plain X-ray chest	No surgical treatment	Y	Expired
3.	18 hrs	F	Respiratory distress and dusky color	L	Plain X-ray chest	Laparotomy, direct closure of defect	Y	Survived
4.	1 day	M	Respiratory distress	L	Plain X-ray chest	Laparotomy, direct closure of defect	Y	Survived
5.	8 hrs	F	Cyanosis and respiratory distress	L	Plain X-ray chest	Laparotomy, closure of defect by inner muscular flaps	Y	Expired
6.	4 hrs	F	Cyanosis and respiratory distress	L	Plain X-ray chest	No surgical treatment	Y	Expired
7.	1 day	M	Respiratory distress	L	Plain X-ray chest	Laparotomy, direct closure of defect	N	Survived
8.	22 hrs	F	Respiratory distress	L	Plain X-ray chest	Laparotomy, direct closure of defect	Y	Survived
9.	10 hrs	F	Cyanosis and respiratory distress	L	Plain X-ray chest	No Surgical Intervention	Y	Expired
10.	12 hrs	F	Respiratory distress	L	Plain X-ray chest	No Surgical Intervention	Y	Expired
11.	16 hrs	F	Cyanosis and respiratory distress	L	Plain X-ray chest	Laparotomy, direct closure of defect	Y	Expired
12.	2 day	F	Respiratory distress	L	Plain X-ray chest	Laparotomy, direct closure of defect	N	Survived
13.	3 day	F	Respiratory distress	L	Plain X-ray chest	Laparotomy, direct closure of defect	Y	Survived
Congenital eventration of the diaphragm (CED)								
1.	2 yr	F	Recurrent chest infections	L	Plain X-ray chest, Fluoroscopy	Laparotomy, plication of diaphragm	Y	Survived
2.	5 yr	M	Recurrent chest infections	L	Plain X-ray chest, Ultrasound, Fluoroscopy	Laparotomy, plication of diaphragm	X	Survived
Hiatus hernia (HH)								
1.	18 mo	F	Vomiting and Recurrent chest infections	X	Upper GI contrast, fluoroscopy	Narrowing of hiatus, Ashcraft procedure	X	Survived
2.	3 yr	F	Recurrent chest infections	X	Upper GI contrast, fluoroscopy	Narrowing of hiatus, Ashcraft procedure	X	Survived
Morgagni hernia (MH)								
1.	5 yr	M	Vague abdominal pain, Respiratory infections	L	Upper GI contrast study	Laparotomy and repair of defect	Y	Survived

AM= Associated Malformations
Age*= Age at presentation

Only one mother had antenatal ultrasound examination, at 32 week gestation, but the defect was not detected. The mean birth weight was 2800 g (1500 – 3300 g). Average age at the time of presentation was 23 hrs (range 2 hrs – 72 hrs). Cyanosis and respiratory distress were the common symptoms at presentation. 10(77%) had associated anomalies (Table 2) and of these, 06 had multiple malformations. The diagnosis was made on the basis of history, scaphoid abdomen, chest auscultation and plain radiograph of chest (Figure 1). Pre-operative stabilization with nasogastric tube, vascular access, oxygenation and supportive medical treatment was extended in all the cases by consultant pediatrician. 04 (31%) patients died before any surgical intervention, due to severe respiratory distress and multiple malformations and 09 (69%) were operated through an abdominal approach. A sac was identified in 04 (45%). In eight (89%) patients, the defects were 3-5 cm in diameter; thus a direct closure with non-absorbable suture was possible. In one patient, the defect required closure with pedicled abdominal wall muscle flaps because of the non-availability of synthetic mesh. Ventral hernia was created in 5 (56%) newborns to accommodate return of the herniated contents into the relatively small abdominal cavity and also to overcome the respiratory insufficiency in absence of mechanical ventilation facilities.

Table 2. Associated anomalies in children with the congenital hernias of the diaphragm

Anomalies	No.
Bochdalek hernia & Congenital eventration of the diaphragm:	
VSD	5
ASD	4
Dextrocardia	3
Meningocele	1
Down Syndrome	2
Hydroureter	1
Hypospadias	2
Cryptorchidism (Bilateral)	1
Amelia (lower extremity)	1
Malrotation of gut	1
Morgagni hernia:	
Undescended testis (L)	1

Five patients with other types of CDHs (CED, MH and HH) presented with recurrent respiratory infections and/or mild gastrointestinal symptoms and the diagnosis was confirmed with plain x-ray chest and gastrointestinal contrast study (Figure 2 & 3). All these patients were operated upon through abdominal approach. In children with CED transabdominal plication of the attenuated diaphragm was done. In HH, the esophageal hiatus was reinforced and a partial anterior wrap fundoplication was performed and in the child with MH, the defect was repaired and diaphragm was sutured to the underside of the posterior rectus sheath at the costal margin after reduction of its contents and excision of the sac.

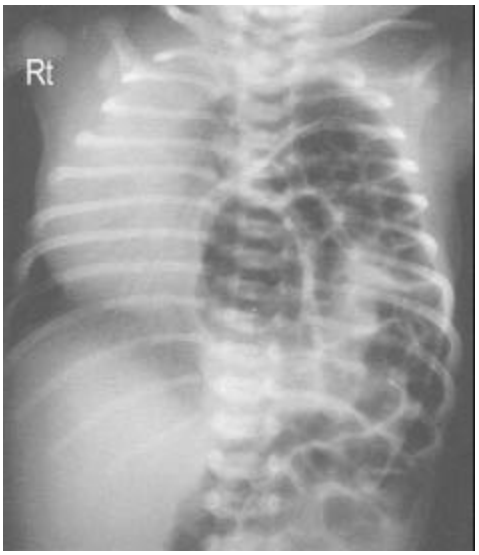

Figure 1. Plain X-ray chest showing bowel loops In the left hemithorax

Post-operative complications occurred in 05 (55%) children with BH; wound sepsis in 02, chest infection in 02 and ipsilateral pneumothorax in 01 patient. All were managed conservatively. 02 children with BH died after surgery from respiratory difficulty. There were no other complications or deaths. The overall morbidity was 36% and survival 67% (Table 1). In follow-up from 6 to 28 months, 4 children (3 with BH and 1 with CED) had ongoing respiratory tract infections and delayed growth and development.

DISCUSSION

The majority of the poster-lateral congenital diaphragmatic hernias occur on the left side, presumably because of late closure of the left pleuroperitoneal canal, and the relative protection offered by the liver on the right. Clark and Vanamo reported left side occurrence from 78-88% in their series [6-10], whereas in this study, all BH defects were documented on the left side. The occurrence was common in girls than boys, which parallel the findings reported by others [11].

The association of congenital anomalies is common and the prevalence varies with each type; 20-39% of BH[12] and 67-82% for MH[13,14]. In this report, the associated anomalies were documented in 10 (77%) children with BH, in 01 with CED and in 01

with MH. The associated malformations played an important role in the final outcome of the children with BH.

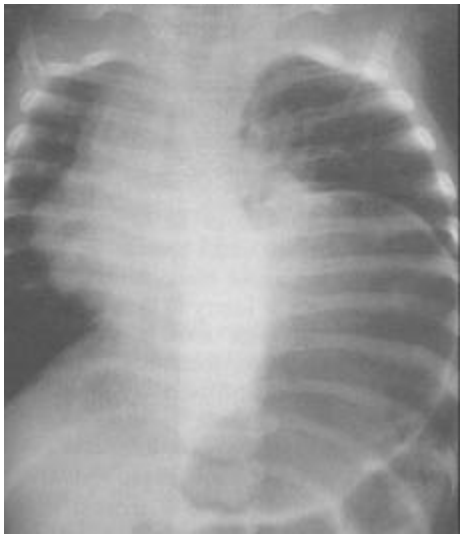

Fig 2. Plain x-ray chest showing left sided congenital eventration of the diaphragm

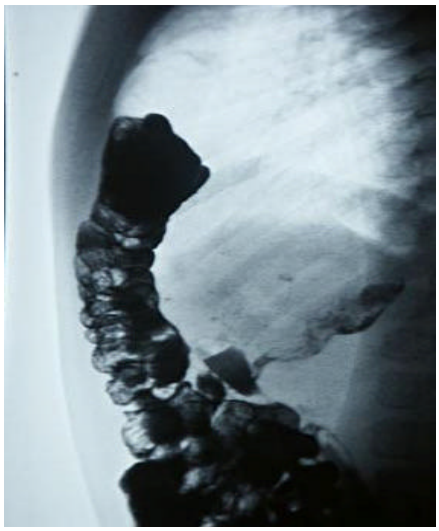

Fig 3- Upper GI contrast study (Lat view) showing bowel in anterior chest through sub costosternal defect

Besides the underlying pathology, associated malformations and early diagnosis plays an important role in the management and outcome of children with CDHs. Prenatal diagnosis occurs in approximately 50% of cases [2], however in our series only 01 mother had prenatal ultrasound assessment but the defect was not noted. Majority of our patients were residents of rural areas, where such facilities are not available.

Depending upon the underlying pathology and compression of lungs, the presentation of CDH varies from neonatal period to later adulthood [15]. In BH the presentation is usually early (within first 24 hours of life), however they may present late. In our series, presentation of patients with BH ranged from 2-72 hrs (average 23 hrs) and 08 (60%) neonates had symptoms in first 24 hours, whereas average presentation of children with other types of CDHs was 3.3 years. In this study, 11 (85%) neonates with BH were born at home and had early onset of symptoms but the referral and definite diagnosis was delayed, which affected their outcome (Table 1). Similarly in 03 (60%) children with other types of CDHs, the respiratory symptoms persisted for long time before they were diagnosed and 01 received anti-tuberculosis drugs for a period of 6-9 months before diagnosis. Thus, unawareness about the condition among family practitioners may have contributed to the delay in diagnosis and referral.

Surgery is recommended treatment for all types of CDHs and the time and approach for surgical intervention vary in each type of CDHs [14,16-19]. In BH, since 1980s, delayed surgery has become accepted method of treatment after Sakai et al [20] reported that early surgical repair worsens the cardiopulmonary functions. In the multi-center CDH study (62 centers with 461 patients), the average age of operated newborns was 73 hrs (1-445 hrs) [6]. In this study, the cases of BH were operated once they were stabilized at an average age of 38 hrs (6-98 hrs). Children with CED, MH and HH were operated after their diagnosis was made. Traditional surgical approach for CDHs includes thoracotomy or laparotomy. Recently, minimally invasive techniques have been described to repair CDHs [21,22]. In our series, transabdominal approach was preferred in all the patients as this facilitated us in different ways. In BH, this facilitated by creating ventral hernia in 05 (56%) children because of their relatively small abdominal cavity to accommodate the herniated viscera from chest in the absence of mechanical ventilation facility, enabling these babies to survive. A ventral hernia is not required when ventilation facilities are available. Also in a child with associated gut malrotation, it was beneficial as the malrotation was corrected at the same time. Same approach was appropriate for MH as well, as the reduction of herniated bowel was easy and so was its inspection later on to look for any injury during the procedure, as observed by Kimmelstiel et al[23]. In children with HH, besides the repair of the hiatus, an anti-reflux procedure can be done through an abdominal approach.

The morbidity and mortality is higher in children with BH than with other types. In this series, post-operative complications occurred in 05 (55%) children with BH, similar to as reported in other studies [10,24]. In spite of recent innovations, mortality

rate of 40-60% has been reported in BH [6,10,24,25]. In this series, a mortality rate of 46% (06 patients) was documented in same group, although the population demographics might not be comparable (Table 1). 04 infants, who died before surgery, had associated major malformations, which affected their outcome. 02 others had a difficult post-operative course and died because of respiratory insufficiency. Survival rate (54%) for children with BH, in this report, parallels other local studies [26]. Long term follow up of children with CDHs has shown that 12-61% of long term survivors had continuing problems with pulmonary function, nutrition and growth, neuro-development and musculoskeletal system [27,28].

In this series, during the follow-up period (6 to 28 months), 04 children, 03 with BH and 01 with CED, had ongoing respiratory tract infections and delayed growth and development.

CONCLUSION

CDHs in children vary in presentation. The overall morbidity (36%) and survival (67%) in the present set of available facilities is encouraging. However, early diagnosis, quick referral and availability of advanced neonatal care will serve a lot towards improvement in the outcome.

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