

Congenital diaphragmatic hernia: prevalence and management

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Summary:

Background: The morbidity and mortality of congenital diaphragmatic hernia is related to the severity of lung hypoplasia ,pulmonary hypertension and associated anomalies.

Patients and methods: fifty nine consecutive patients with surgically proved diaphragmatic hernia (excluding hiatal hernia) were included in this prospective study. All patients were seen at Al-Kadhymia hospital for children and Al-Mustansyria hospital, in the period from January 2003 to December 2009.Plain chest x-ray was done for all the patients understudy, but barium study was arranged for only nine patients.

Results: There were fifty nine child with congenital diaphragmatic hernia , Bochdalek hernia forms 81.4% of all congenital diaphragmatic hernia, the peak incidence at presentation was in the neonatal period (80,2%) of patients. The chest roentgenographic appearance was diagnostic in 87.5% of the patients with Bochdalek while Morgagni hernia in 62.5% of patients was diagnostic. Morgagni hernia constitutes 13.6% of the congenital variety, commonly to the right side (87.5% of patients).The incidence of presentation beyond the neonatal period was 50% of cases . Absence of hemidiaphragm was found in 5% of the congenital variety , the chest x-ray was diagnostic in all patients.

Conclusion: The chest roentgenograph (frontal view) is a very helpful diagnostic tool for the diaphragmatic hernia with a high diagnostic accuracy (73.7%).

Keywords: Congenital diaphragmatic hernia(CDH), Bochdalek hernia (BH) , Morgagni hernia (M H.).

Fac Med Baghdad
2010; Vol. 52, No. 4
Received Oct., 2010
Accepted Nov., 2010

Introduction:

Congenital diaphragmatic hernia(CDH) refer to the developmental defect in the diaphragm that allows abdominal viscera to herniated into the chest during the critical period of lung development when the bronchi and pulmonary arteries are undergoing branching[1]. 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 [2].Although, the diagnosis can be made antenatal [3,4,5] the presentation may be delayed [6].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[7]. 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 [8,9].

Patients and methods:

Fifty nine consecutive patients with surgically proved diaphragmatic hernia (excluding hiatal hernia) were included in this prospective study.

These patients were seen and operated upon at Al-Kadhymia Hospital for children and AL-Mustansyria Hospital, in the period from January 2003 to December 2009. The clinical details were reviewed, including the presenting symptoms, original diagnosis, associated anomalies and operative findings. The roentgenographic findings were reviewed and evaluated by radiologists

All patients underwent plain chest roentgenogram (frontal view, whether erect or supine). From those fifty nine patients, nine of them underwent barium studies (six barium follow through and three barium enema).

Results:

From the fifty nine patients with diaphragmatic hernia, forty eight patients were proved to have congenital Bochdalek hernia, eight patients were proved to have Morgagni hernia &in three cases absence of diaphragm .

Bochdalek hernia (Boch, H) forms 81.4% of all congenital hernias, more in the left side (87,5%)(Table I), with males preponderance (62.5%) , the ages ranged from birth to fourteen years. The peak incidence of presentation was in the first 24 hours of life (62.5% of cases). The incidence in the neonatal period was 79.2% of patients, while delayed presentation, i.e., after neonatal period was only reported in 20.8% of patients (Table II). The latent asymptomatic period ranged between three months to fourteen years (median 11 months),. Shortness of breath (SOB) was the main presentation of Boch. H (93.8% of patients)other clinical presentation were cyanosis(52%),vomiting(16.7%)

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and cough (16.7%) of the patients. Combination of both, upper and lower parts of gastrointestinal tract (GIT) were the main contents of hernia (87.5% of patients). In one case, the liver was the only herniated organ (Table III). Volvulus of the stomach was encountered in one case of left sided Boch. H. 12.5% of patients were associated with other congenital anomalies, and the commonest was 13 pairs of ribs.

The most common roentgenographic appearance (table IV) was mediastinal shift to the contra lateral side, an ill defined ipsilateral hemidiaphragm with multiple ring shadows or wide tubular shadow. This appearance was found in forty one patients (85%) and it was diagnostic of diaphragmatic hernia.

Three patients (6.3%) were presented with large single ring shadow with or without air-fluid level in the lower 1/3 of the thoracic cavity, an ill defined ipsilateral hemidiaphragm and mediastinal shift to the contralateral side except in one case in which there was a central mediastinum. In this latter case the stomach was the only herniated viscus. These roentgenographic appearances were suggestive of diaphragmatic hernia.

One patient (2.1%) presented with patchy opacities (simulating an inflammatory process), with the mediastinal shift to the contralateral side and an ill defined ipsilateral hemidiaphragm. This roentgenographic appearance was suggestive of diaphragmatic hernia and was proved to be due to herniated bowel loops.

One patient (2.1%) presented with complete opacification of the left hemithorax with the mediastinal shift to the contralateral side and ill defined ipsilateral hemidiaphragm (simulating massive pleural effusion). This appearance was in supine position and it was suggestive of diaphragmatic hernia because of high position of the colonic gas shadow in the left upper abdomen while in the erect position the appearance has changed to multiple rings and tubular shadow which was diagnostic of hernia.

One patient (2.1%) presented with soft tissue density mass occupying the lower 2/3 of the right hemithorax with the mediastinal shift to the contralateral side. This was associated with an ill defined ipsilateral hemidiaphragm and a high position of the colonic gas shadow in the right upper abdomen. In this case the diagnosis was suggestive of diaphragmatic hernia and it was proved to be due to intrathoracic herniation of the liver only.

The chest roentgenographic appearance, in general, was diagnostic in forty two (87.5%) of patients and was suggestive in six (12.5%) patients & the diagnosis was proved by barium study (four patients barium meal & follow through, other two cases barium enema were done)

Morgagni hernia (Morg. H.) constitutes 13.6% of the congenital variety, commonly in the right side (87.5%) of the patients (table I). The incidence was almost equal in both sexes the ages ranged from birth to 13 years. The peak incidence of presentation was found in the first 24 hours of life (25% of

patients). The incidence in the neonatal period was 50% of patients, while the incidence of the delayed presentation (2 month +) was 50% of patients (table II). The latent asymptomatic period ranged between 2 months to 13 years. The most common presenting symptom was SOB (75% of patients) (The combination of both upper and lower parts of GIT were the main herniated viscera (50% of cases)).

In one patient, the liver was the only herniated viscus in right sided hernia, while volvulus of the stomach encountered in two patients of right sided hernia (table III). 25% of patients were associated with other congenital anomalies (Downs syndrome and ventricular septal defect).

The most common roentgenographic appearance (table IV) was, mediastinal shift to the contralateral side, an ill defined ipsilateral hemidiaphragm and multiple ring shadows or wide tubular shadow and it was found in 4 out of 8 patients (50%), this appearance was diagnostic of diaphragmatic hernia.

Two patients (25%) were presented with soft tissue density mass obliterates the right cardiophrenic angle with mediastinal shift to the contralateral side and a normal appearance of the diaphragm, This was suggestive of Morgagni hernia. In these two patients the liver was the only herniated viscus in the first, and the colon in the second.

Two (25%) were presented with large single ring shadow in the lower 1/3 of the thoracic cavity. In the first patient there was an air-fluid level, and it was associated with a central mediastinum, and an ill defined ipsilateral hemidiaphragm. This appearance was proved to be due to herniated stomach with volvulus. The second patient was associated with mediastinal shift to the contralateral side and a normal appearance of the diaphragm with obliteration of the right cardiophrenic angle. This was proved to be due to herniated stomach and bowel. These appearances were suggestive of Morgagni hernia.

The chest roentgenographic appearance was diagnostic of Morgagni hernia in five (62.5%) of patient and was suggestive in three (34.5%) of patients and then the diagnosis was proved by barium meal with follow through and barium enema.

Absence of hemidiaphragm (Abs. diaph.) was found in 5% of the congenital variety, and only in the left side with males preponderance (66%) of patients. In all patients the symptoms started since birth (table II). The main presenting symptom was SOB (100%). In all patients both upper and lower parts of GIT were herniated to the thoracic cavity. No associated anomalies could be identified.

The roentgenographic appearance (table IV) was, mediastinal shift to the contralateral side, an ill defined lateral hemidiaphragm and multiple ring shadows. This appearance was diagnostic of diaphragm hernia. The appearance of intestinal gas shadow crossing to the thoracic cavity was found in 66.7% of patients.

The chest roentgenograph was diagnostic in all patients, No barium study was done.

Table I. Type & Site of diaphragmatic hernia.

Type	Site		Total No. %	
	Right	Left		
Boch. H.	6	42	48	81.4
Morg. H.	7	1	8	13.6
Abs. H.	0	3	3	5
Total	13	46	59	100

Table II. Age distribution of presentation of congenital diaphragmatic hernias.

Age group	Boch. H. No. %	Morg. H. No. %	Abs. Diaph No. %	Total No. %
1 st 24 hr.	30 (62.5)	2 (2.5)	3100	35 (59.32)
1-7 days	3 (6.25)	1 (12.5%)		4 (6.78)
1-8 weeks	5 (10.42)	1 (12.5)		6 (10.17)
2-12 months	5 (10.42)	1 (12.5)		6 (10.17)
1-4 years	3 (6.25)	1 (12.5)		4 (6.8)
5-9 years	--	1 (12.5)		1 (1.7)
10-15 years	3 (4.16)	1 (12.5)		3 (5.08)
Total	48	8	3	59

Table III. hernial contents according to herniated viscus

Content	Boch. H. No. %	Morg. H. No. %	Abs. Diaph No. %
Small bowel	43 (84.6)	5 (62.5)	3 (100)
Large bowel	42 (87.5)	6 (75%)	3 (100)
Spleen	15 (31.3)	--	3 (100)
Stomach	14 (29.2)	2 (25)	3 (100)
Kidney	1 (2.11)	--	--
Liver	2 (4.2)	1 (12.5)	2 (66.7%)
Omentum	2 (4.2)	2 (25%)	--
Appendix	1 (2.1)	--	--
Pancrease	1 (2.1)	--	1 (33.3%)

Table IV. The plain chest roentgenographic findings (frontal view).

Radiological CXR Findings		Type of Diaph. Hernia		
		Boch. H. No. %	Morg. H. No. %	Abs. Diaph No. %
Mediastinum	Central	1 (2.1)	1 (12.5)	--
	Shift	47 (97.9)	7 (87.5)	3 (100)
	Widening			--
Diaphragm	ill defined	48 (100)	6 (75)	3 (100)
	Normal	--	2 (25)	--
Thoracic cavity	a	41 (85.1)	4 (50)	3 (100)
	b	3 (6.3)	2 (25)	
	c	1 (2.1)		
	d	1 (2.1)		
	E	7 (14.6)	1 (12.5)	2 (66.7)
	f	1 (2.1)	2 (25)	
abdominal intestinal shadow crossing to the thoracic cavity in diaphragmatic hernias		30 (62.5)	4 (50)	2 (66.7)

- a. = Multiple rings or wide tubular shadow
b.= large single ring shadow in the lower 1/3 of the thoracic cavity (with or without air-fluid level)
c.= Patchy opacities
d.= Complete opacification of the hemi thorax

e.= Herniation of bowel loops to the contra lateral side of the thoracic cavity
f.= Soft tissue density mass

Discussion:

Congenital diaphragmatic hernia constitutes a major surgical emergency in the new born. The key of survival lies in the prompt early diagnosis and treatment, and with out surgical intervention the mortality rate approaches 100% especially when it presents during the first 24 hours of life due to a lethal combination of pulmonary hypoplasia and pulmonary hypertension [10, 11]

From the total of in fifty nine patients the peak incidence was found in the first 24 hours of life was 59% of all patients, Clacks reported 60% of cases presented within the first 24 hours of life[7] which were in agreement with present study.

bochdalek hernia: constituted 81.4% in our study and it was reported also to be the commonest [7,8], that is in agreement with the present work.

Left sided Boch. H. in our patients were considerably more frequent than right sided, that is compatible with other studies [12, 13], 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 [7,8,14.] The sex preponderance varies in different studies. Male preponderance in our patients 62.5% is in agreement with the most of other studies [7,13].

In this study the peak incidence was found in the first 24 hours of life (62.5% of cases), while only 20% of cases were reported beyond the neonatal period. A previous study conducted by Forshall [15] reported that nearly all cases were presented within the first 48 hours of life, other studies found that 5%-25% of cases were presented beyond the neonatal period [13]. The latter studies, in general, were in agreement with our study.

In this study we found that 12.5% of cases were associated with congenital anomalies. 13 pairs of ribs may be encountered in 11% of patients, Slavotinek encountered this anomaly in 13% of patients [16]. The associated congenital anomalies are well recognized conditions, Its incidence varied from 17% to 50% of cases [13, 14]. The reasons for these differences are not clear but probably due to numerous factors, including the region of the study, techniques used for the diagnosis of the anomalies, and sever defects may be missed if they resulted in still births without autopsy

In this study the most common roentgenographic appearance was, a mediastinal shift to the contralateral side, an ill defined ipsilateral hemidiaphragm with multiple rings or wide tubular shadow represent a herniated gas filled bowel loops. This appearance was found in 85.1% of cases and it was diagnostic. Kerley et al. [17] reported that

herniation of the large or small intestine into the thorax irrespective of the defect is anterior or posterior, congenital or acquired, has a characteristic appearance. The hernial contents were recorded to be mostly small bowel [6,7] which was in agreement with our study.

The herniated liver alone was seen in one case only in the present series which is a recognised condition reported by others [15]. In addition, the appendix was found within the hernial contents in one case of the current series. This condition is of clinical importance because of the risk of the appendicitis with subsequent perforation, and appendicular abscess at the hemithorax [18].

Morgagni hernia is less common than Boch. H. in our patients (13.6%). The incidence is variable with different studies ranging from 2%-23% [1,2]. These findings run in parallel with the current work.

Both sex are equally affected in our patients in contradictory to study by Clark, et al reported male preponderance [7].

This type of hernia usually presented much later than the Boch. H. 50% of our patients were found beyond the neonatal period which were in agreement with this study [4,10].

The clinical presentation of this hernia involve respiratory and gastrointestinal systems of varying quantities and qualities from one series to another [6]. In our work we found that the main presenting symptom was shortness of breath which occurs in 75% of the patients. Chronic gastric volvulus within the foramen of Morgagni is very rare, leading to partial or complete closed loop obstruction [19].

Associated congenital anomalies were more frequent with Morg. H. than Boch. H. and it is extremely variable in different series. The most common one was Down's syndrome [17]. These findings were in full agreement with our data.

Roentgenographic diagnosis is difficult because of the presence of the heart shadow. It may be diagnosed when an air- fluid level was noted in the lower mediastinum [20], or when gas containing bowel is present in the hernial mass. The chest film reveals a round shadow of variable size in the right cardiophrenic angle, The hernia might present with a solid mass in the right cardiophrenic area [20], and can be difficult to differentiate it from an epicardial fat pad, pleuropericardial cyst, right middle lobe pathology and right atrial or pericardial tumour [21]. Our findings were in agreement with other studies. Absence of hemidiaphragm was seen in three patients (5%). and in all patients the left hemidiaphragm was affected, with males preponderance is (66.6%). Jackson found that it constituted 7.1% of all congenital forms, It occurs usually on the left side and is more common in males.

Barium studies were performed for seven patients and they were of value in excluding the presence of the bowel in the thoracic cavity as most of these hernias will contain either small or large bowel or both [22].

The mortality rate was 12% (7 patients), four patients, who died before surgery, had associated major malformations, which affected their outcome, other three patients had a difficult postoperative course and died because of respiratory insufficiency. In other series a mortality rate of 30-60% has been reported [7,23] which is not comparable with this study, which might be due to death of critical patients before the diagnosis was established and the autopsy not done routinely in our hospitals.

Conclusion:

The commonest type of congenital (excluding hiatal) hernia is Bochdalek hernia. It occurs mostly in the left side and presents mainly in the neonatal period, Morgagni hernia is less common, mostly to the right side and presents mainly beyond the neonatal period. The chest roentgenograph (frontal view) is a very helpful diagnostic tool for the diaphragmatic hernia with a high diagnostic accuracy (73.7%).

Barium studies should be restricted to those patients in whom the plain chest film was suggestive of a diaphragmatic hernia.

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