

# Late presentation of Bochdalek hernia in children – experience at a single centre

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**Background:** The aim of this study was to present our experience with late presentation Bochdalek hernia (BH), focussing on clinical presentation diagnostic and therapeutic approaches, and their outcomes.

**Methods:** Patients with late presenting BH 1 month of age between 1983 and 2022 were studied. We retrospectively collected and analysed the following data: age, sex, side of herniation, presenting symptoms and signs, associated anomalies, radiographic findings, intraoperative findings, postoperative course, complications, and mortality.

**Results:** Of 175 diaphragmatic hernias 46 (26%) were late presenting BH. Fifty-seven per cent (26/46) were males. Laparotomy was performed in 85% (39/46) of the patients, and a laparoscopy or thoracoscopy was performed in 15% (7/46). The mean hospital stay was 7.6 days, and the mean surgery time was 131 minutes. When the two groups were compared, the duration of surgery and hospitalisation was significantly shorter in the minimally invasive surgery (MIS) group ( $p < 0.05$ ).

**Conclusions:** The clinical characteristics of late presenting BH are variable, either acute or chronic. A laparoscopic or thoracoscopic approach in selected patients is feasible.

**Keywords:** Bochdalek hernia, diaphragmatic hernia, children

## Introduction

Bochdalek hernias (BH) are recognised as a congenital defect present at birth, and this classification holds regardless of when symptoms appear.<sup>1</sup> While most cases are identified within the first 24 hours of life – during which there's a high mortality risk – about 10% may be detected after the newborn period.<sup>2</sup> Cases identified after one month are considered "late presenting" BH. The concept of late presenting BH has been acknowledged since a pivotal study in 1959, with subsequent research exploring various clinical and diagnostic methods. Patients with late presenting BH tend to have a more favourable outlook, often because there are no significant associated abnormalities, or the hernia has a less severe effect on lung development (such as pulmonary hypoplasia) and doesn't lead to pulmonary hypertension.<sup>1-3</sup> Symptoms in these patients can be mild, including respiratory issues, chronic lung infections, pleural effusions, pneumonia, feeding problems, or gastrointestinal disturbances, which vary based on the extent of organ displacement and other complications.<sup>1-3</sup> X-rays, often performed due to symptoms or discovered by chance, are instrumental in diagnosing some of these cases. However, misdiagnosis can lead to significant morbidity.<sup>3-5</sup>

The aim of this study was to present our experience by examining the clinical characteristics of BH with late presentation, as well as diagnostic and therapeutic approaches, and the outcomes of associated anomalies.

## Materials and methods

One hundred and seventy-five patients with congenital diaphragmatic hernia were diagnosed and treated over the past four decades (1983-2022) at our institution. Forty-six of these patients with late presenting BH (over 1 month of age) are the subject of the study. Diaphragmatic eventration, and hiatal, traumatic, and Morgagni hernias were excluded.

Patients with suspected late presenting BH underwent chest radiographs. Computed tomography (CT) scans or upper gastrointestinal (GI) contrast studies were performed if there was diagnostic doubt. Figure 1 describes the diagnostic and therapeutic pathway.

We recorded the following data – maternal demographics, infants age, sex, side of herniation, and presenting symptoms and signs, which were defined as acute or chronic. (Acute – sudden onset of short-term symptoms. Chronic – slow-onset and long-term symptoms). Associated anomalies, radiographic findings, intraoperative details, postoperative course, complications, and mortality were also recorded. Right and left late presenting BH were compared according to various parameters. Age, gender, diagnosis, sac, additional anomaly, malrotation, surgical method, surgery time (min), length of hospital stay (LOS, days), recurrence, mediastinal shift, complications, and height ratio. The height ratio was calculated as the length of the red line, the upper end of the hernia sac to the apex of the lung, divided by the length of the yellow line, from the lower end of the hernia sac to the apex of the lung. As the height ratio increases, the amount of lung

tissue under pressure also increases, and it is hypothesised that this ratio may correlate with the prognosis, which is why this ratio was calculated.

Outcomes were compared between patients who underwent open surgery and minimally invasive surgery (MIS).

### Statistical analysis

Statistical analysis of patients' data, involving descriptive statistics, frequency, and other characteristics was performed for all items. Continuous data are written as mean ± standard deviation. To determine whether the data had a normal distribution, the continuous variables were analysed with Shapiro-Wilk and Kolmogorov-Smirnov tests. Continuous, and normally distributed variables were compared using t-test. Non-parametric tests were chosen when the data did not fit the normal distribution. The categorical variables were evaluated by the chi-square test and some data by Fisher's exact test, as needed. Analyses were performed using SPSS Statistics for Windows, Version 21.0 (IBM Corp., Armonk, NY, USA). All *p*-values were two-sided and *p* ≤ 0.05 was considered statistically significant.

Ethics committee approval of the study was approved by Dicle University Ethical Committee. Approval was provided for the study (Approval No: 07.11.2022-223). This

retrospective study has been conducted according to the principles of the Helsinki Declaration.

### Results

Data on the 46 late presenting BH were analysed. There were 57% (26/46) males and 43% (20/46) females among these patients. While the patients' mean age was 14.7 months (range 1 month–14 years), 85% (39/46) were left-sided and 15% (7/46) were right-sided (Table I and Figure 1).

The most common symptoms were respiratory. Respiratory symptoms were detected more frequently in patients with right as opposed to left sided BH. In all right-sided BH patients, there was liver herniation. GI symptoms were more common on the left. Right late presenting BH was not statistically significant due to the small number of patients (*p* > 0.05). When various parameters of patients with right and left late presenting BH were compared, no significant difference was found between the sides (*p* > 0.05). No significant difference was found when age at presentation was compared to symptoms (*p* > 0.05) (Tables II, III, IV).

Chest radiographs were performed on 46 patients. In instances where chest radiographs yielded ambiguous results and to facilitate differential diagnosis of other pathologies, CT scans and GI contrast studies were performed on 33% and 20% of patients respectively.

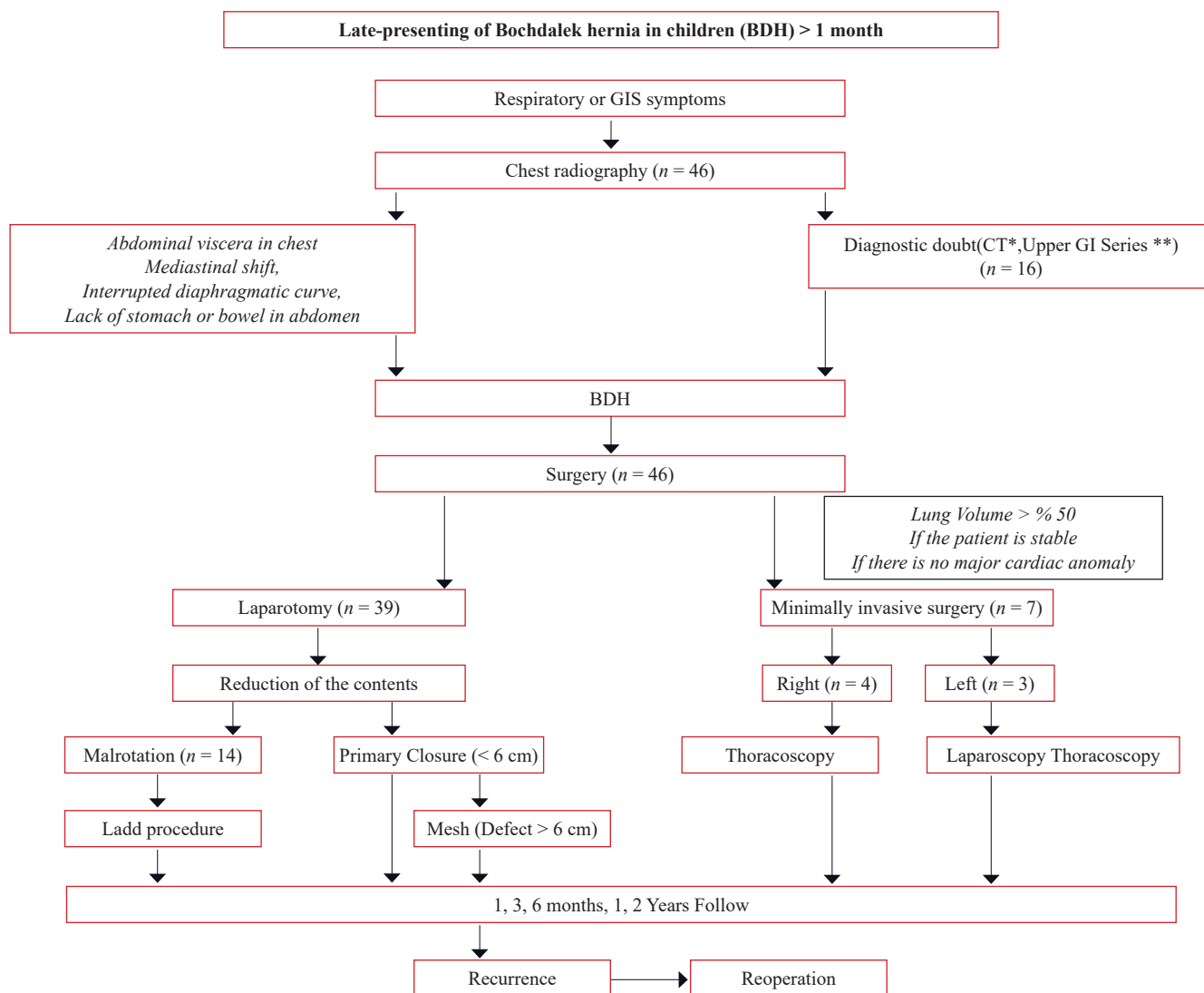


Figure 1

**Table I: Demographic data**

Variable	n (46)	%
<b>Sex</b>		
Male	26	57
Female	20	43
<b>Symptoms duration</b>		
Acute	16	35
Chronic	30	65
<b>Symptoms</b>		
Respiratory	31	67
GIS	9	20
Asymptomatic	4	7
GIS + Respiratory	2	4
<b>Diagnostic method</b>		
Chest radiograph (CR)	46	100
CR + CT scan	15	33
CR + Upper GI series	9	20
<b>Side</b>		
Left	39	85
Right	7	15
Sac (yes)	15	33
Malrotation (yes)	14	30
Additional anomaly (yes)	4	9
<b>Surgical method</b>		
Laparotomy	39	85
MIS	7	15
Mediastinal shift (yes)	14	30
<b>Morbidity</b>		
Recurrence	2	4
Adhesion	1	2
Mortality (yes)	1	2
	<b>Mean</b>	<b>Range</b>
Hospital stay (days)	7.6	3–20
Surgical time (minutes)	131	75–240

LOS – Length of hospital stay

Laparotomy was performed in 39 (85%) patients, and MIS in 7 (15%). A thoroscopic repair was performed in 4 patients with 2 right and 2 left BHs. A laparoscopic repair was performed in 3 patients with a left BH. The mean hospital stay was 7.6 (range 3–20) days, and the mean surgery time was 131 min. When the two groups were compared, the duration of surgery and hospitalisation was significantly shorter in the MIS group ( $p < 0.05$ ). There was no difference in both groups in terms of recurrence or complications ( $p > 0.05$ ) (Table IV).

Malrotation was the most common type of accompanying anomaly, occurring in 30% of patients. The remaining four anomalies were cardiac (2), gastric outlet obstruction (1) and neurological (1).

The small intestine was the most commonly herniated organ, occurring in 30 patients (63%). The other herniated organs in descending frequency were the colon in 22 patients (48%), the stomach in 19 (40%), the spleen in 15 (33%), the liver in 7 (15%), and the kidney in 3 (7%).

Re-operation was required in two patients – one required and biological prosthetic patch repair and the other required

adhesiolysis. One patient with a recurrence was not reoperated and died from multiple anomalies in the sixth postoperative month.

There was no difference in complications and recurrence between patients who underwent open and MIS. Due to multiple anomalies, one patient died in the third postoperative month.

## Discussion

The phenomenon of late presenting BH, initially documented by Kirkland in 1959, represents a relatively uncommon condition, with its frequency among all diaphragmatic hernias estimated to be between 2.6 and 45%.<sup>1-3</sup> In our research, we observed that 26% of cases fell into the category of late presenting Bochdalek diaphragmatic hernia (BDH). The body of literature on this topic is limited, with one notable study in 2005 compiling data on 70 BH patients from 30 different centres, yet not offering a comparative analysis. In contrast to this, our study provides a comparative analysis encompassing a group of 46 patients, examining variables such as the side of the hernia, the age at diagnosis, and the surgical techniques employed.

In a multicentre study by Kitano et al. involving the gender distribution, 50 males (65%) and 27 females (35%) was similar to that in our cohort where 26 (57%) were male, and 20 (43%) were female. The mean age at diagnosis in Kitano's study of 14 months (range: 32 days to 15 years)<sup>5</sup> is also similar to the mean age at diagnosis in our cohort of 14 months (30 days–14 years). In our study, 85% were on the left, which was a higher percentage than the 69% reported by Kitano.<sup>5</sup>

Patients with late presenting BH can exhibit a range of symptoms from acute to chronic, while others may show no symptoms at all.<sup>6</sup> The underlying causes of these symptoms are thought to be the pressure of intra-abdominal organs against the lung and the dislocation of GI organs due to the hernia. In the study conducted by Kitano et al., they found that respiratory complaints were present in 43% of patients, GI issues in 33%, both types of symptoms in 13%, and no symptoms in 11%. The rarity of GI symptoms in patients with a right-side late presenting BH is thought to be due to the liver impeding the movement of the hollow organs into the right thoracic cavity.<sup>5</sup> Our research revealed a higher prevalence of respiratory symptoms, with 67% of patients affected, suggesting that lung compression might be the primary symptomatic driver, more so than reported in Kitano's study. Nonetheless, it's noteworthy that over 10% of patients in both studies did not exhibit symptoms.

Although clinical findings are variable, chest radiographs (CRs) in children with late presenting CDH are the initial imaging modality choice. Often done in combination with an abdominal radiograph, the combination will establish the diagnosis in half the cases. Baglaj et al. studied 107 (49%) of 218 children who were diagnosed with the first chest radiograph.<sup>2</sup> However, in late-present BH, misdiagnosis occurs in more than 25% of these patients with pneumonia and pneumothorax being the most frequent misdiagnoses.<sup>2,7-11</sup> Hence 50% of the patients will require additional radiological evaluation to make the final diagnosis.<sup>2,5</sup> In our study, 47% of our patients were diagnosed with chest radiographs alone, while 53% of them had an additional CT scan and/or upper GI contrast studies examinations performed to establish the diagnosis.<sup>12</sup>

**Table II: Right and left Bochdalek hernia comparison**

	Left (n = 39)		Right (n = 7)		p-value
	Mean	SD	Mean	SD	
Age (month)*	17.6	6.9	17.7	6.3	> 0.05
Maternal Age*	29.6	5.2	26	4.9	> 0.05
Surgery Time* (min)	115	12	110	10	> 0.05
LOS (day)*	7.23	2	4	3	< 0.05
Height ratio (%)*	51.8	6.9	43.2	8.3	> 0.05
	<i>n</i>	%	<i>n</i>	%	
<b>Gender</b>					
Male	23	58.9	3	42.9	> 0.05
Female	16	41.0	4	57.	
<b>Symptom</b>					
Acute	14	35.9	2	28.6	> 0.05
Chronic	25	64.1	5	71.4	
<b>Diagnosis</b>					
Chest radiograph	39	100	7	100	> 0.05
CT scan	16	41.0	4	57.1	
<b>Sac</b>					
Yes	12	30.8	3	42.9	> 0.05
No	27	69.2	4	57.1	
<b>Additional anomaly</b>					
Yes	17	43.59%	1	14.3	> 0.05
No	22	56.41%	6	85.7	
<b>Malrotation</b>					
Yes	13	33.3	1	14.3	> 0.05
No	26	66.7	6	85.7	
<b>Surgical method</b>					
Laparotomy	35	89.7	4	57.1	< 0.05
MIS	4	10.3	3	42.9	
<b>Suture</b>					
Prolene	15	38.5	1	14.3	
Ethibond	19	48.7	3	42.9	> 0.05
<b>Recurrence</b>					
Yes	2	5.1	0	0%	> 0.05
No	37	94.9	7	100%	
<b>Complication</b>					
Yes	3	7.7	0	0%	> 0.05
No	36	92.3	7	100%	
<b>Mediastinal shift</b>					
Yes	10	25.6	4	57.1	> 0.05
No	29	74.4	3	42.9	
Gastric gas status	6	15.4	4	57.1	> 0.05

LOS – Length of hospital stay

**Table III: Comparison of age groups**

	0–1 month (n = 34)	1–3 month (n = 6)	> 3 month (n = 6)	p-value
<b>Side</b>				
Right	5	1	1	> 0.05
Left	29	5	5	
<b>Gender</b>				
Male	18	3	5	> 0.05
Female	16	3	1	
<b>Symptom</b>				
Acute	11	3	2	> 0.05
Chronic	23	3	4	
<b>Diagnosis</b>				
Chest radiograph	20	2	4	> 0.05
CT scan	14	4	2	
<b>Sac present</b>				
Yes	10	2	3	> 0.05
No	24	4	3	
<b>Additional anomaly</b>				
Yes	15	2	1	> 0.05
No	19	4	5	
<b>Malrotation</b>				
Yes	12	2	0	> 0.05
No	22	4	6	
<b>Surgical method</b>				
Laparotomy	28	5	6	> 0.05
MIS	6	1	0	
<b>Suture</b>				> 0.05
Prolene	9	5	2	
Ethibond	19	1	2	
<b>Recurrence</b>				> 0.05
Yes	1	0	1	
No	33	6	5	
<b>Complication</b>				< 0.05
Yes	1	0	2	
No	33	6	4	
<b>Mediastinal shift</b>				> 0.05
Yes	9	2	3	
No	25	4	3	

LOS – Length of hospital stay, MIS – Minimally invasive surgery

Today, a CT scan is the best imaging method in patients with the right BH. It is more useful in the differential diagnosis of intrathoracic masses or suspicious findings than chest radiograph.<sup>13</sup> In our study, CT examination was performed in 33% of patients. GI contrast studies are performed in one-third of patients with diaphragmatic anomalies before surgery,<sup>2,14</sup> to detect additional pathologies such as malrotation, which is less than the 20% of our study and reflects modern trends to use CT scans as the next investigation when there is diagnostic doubt.

Patients with right and left late presenting BH have been compared in a small number of studies in the literature. In all of our right-sided hernia, the liver was herniated. Among the left sided BH, the small intestine was the most commonly

**Table IV: Comparison of surgical techniques**

Variable	Laparotomy (n = 39)		MIS (n = 7)		p-value
	Mean	SD	Mean	SD	
<b>Age (months)</b>	18.8	± 4.3	9.2	± 6.9	< 0.05*
<b>Surgery Time (min)</b>	121	± 12	95	± 17	< 0.05*
<b>LOS (day)</b>	7.5	± 2	4	± 3	< 0.05*
	<b>n</b>		<b>n</b>		
<b>Gender</b>					
Male	24		2		> 0.05
Female	15		5		
<b>Symptom</b>					
Acute	13		3		> 0.05
Chronic	26		4		
<b>Hernia sac</b>					
Yes	12		3		> 0.05
No	27		4		
<b>Malrotation</b>					
Yes	14		0		< 0.05*
No	25		7		
<b>Suture</b>					
Prolene	14		2		> 0.05
Ethibond	19		3		
<b>Recurrence</b>					
Yes	2		0		> 0.05
No	37		7		
<b>Complication</b>					
Yes	3		0		> 0.05
No	36		7		

LOS – Length of hospital stay, MIS – Minimally invasive surgery

herniated organ, accounting for 30 cases (63%). The colon was found to be herniated in 22 patients (48%), the stomach in 19 patients (40%), the spleen in 15 patients (33%), the liver in 7 patients (15%), and the kidney in 3 patients (7%).

In our study, the incidence of hernia sac was 33%, which is at the upper end of the 7–32% range reported in the literature.<sup>15</sup>

In the study by Kim et al.,<sup>11</sup> additional anomalies were reported in 23%. In our study, while malrotation was seen in 30% of the patients, additional anomalies were observed in only 9% of the patients.

Early surgical intervention is crucial to prevent complications as soon as a diagnosis is confirmed. Patients with late presenting BH often have a better prognosis due to less pulmonary hypoplasia.<sup>16,17</sup> The preferred approaches for this are either laparotomy or MIS. In our study, we primarily conducted interventions via laparotomy. However, in recent years, we have chosen MIS approach for selected cases. In patients with right-sided late-presenting BH, we opted for thoracoscopy as the MIS. For patients with left-sided late presenting BH, we utilised either thoracoscopy or laparoscopy. The limitation of our study is the low number of patients who underwent MIS surgery so there is a need for more extensive studies involving a larger patient population to better define the role of these MIS surgical options.

In this study recurrence was observed in 2 patients one of whom died from associated multiple anomalies. This single



death mortality rate is in keeping with the lowest mortality rates reported for delayed presentation of BH which ranges from 0–18%.<sup>4,5,18,19</sup> We believe the low mortality rate in our series is due to a lower rate of misdiagnosis, a low rate of pulmonary hypoplasia and appropriate multidisciplinary paediatric surgical management.

## Conclusions

The clinical characteristics, either acute or chronic, of late presenting BH are variable. The herniated side and the affected organs have different symptoms. There is no correlation between symptoms and age of presentation. Generally, the condition can be diagnosed with a CR, but it should be evaluated by CT scan or GI contrast studies when diagnostic doubt persists in suspected cases. When the right and left late presenting BH hernias were compared, no significant difference was found between them. Surgery can be performed with a low mortality rate by laparotomy, and by a laparoscopy or thoracoscopy approach in selected patients.

## Conflict of interest

The authors declare no conflict of interest.









## Funding source

None.

## Ethical approval

Ethical approval was obtained from the Dicle University Medical Faculty Ethics Committee for Noninterventional Studies.

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