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Comparison between late-presenting and isolated neonatal congenital diaphragmatic hernias

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ABSTRACT

Purpose: Late-presenting posterolateral congenital diaphragmatic hernias (CDH) are anatomically similar to isolated neonatal CDH but are diagnosed and treated after the first month of life. We aim to characterise the clinical manifestations and short-term postoperative course of this entity and compare it with isolated CDH of the neonatal period. **Materials and Methods:** In the 30-year period from 1980 to 2010, 116 children with CDH were treated at the Aghia Sophia Children's Hospital, Athens, Greece. Twenty-three (19%) of these children were late-presenting cases, being diagnosed between the ages of 1 month and 4 years. Ninety-three were neonatal cases, of whom 22 (24%) were excluded due to severe associated anomalies, leaving 71 cases of isolated neonatal CDH. We compared these two groups of patients with regard to preoperative symptoms, postoperative hospital stay, time to complete feeding, overall complication rate, and reoperation rate. **Results:** Isolated neonatal cases presented more often with acute respiratory symptoms (n=25; P= 0.016) and failure to thrive (n= 38; P= 0.03). Late-presenting cases presented more often with chronic respiratory symptoms (n=14; P= 0.0044) or gastrointestinal symptoms (n=12; P= 0.006). Thirty-five cases with minor or serious complications were reported in the neonatal group, whereas only five complications were observed in the late-presenting group (P= 0.028). We did not record any recurrences or reoperations in the late-presenting group, but we had two recurrences and three reoperations in the neonatal group. Time to full feeds and postoperative hospital stay was shorter in the late-presenting group. **Conclusions:** Our data demonstrate differences between the two groups in preoperative symptoms and short-term postoperative complications and short-term outcome. Late-presenting cases of CDH had a greater number of chronic symptoms preoperatively, more favorable postoperative outcomes, and less recurrences and reoperations.

Key words: Complications, congenital diaphragmatic hernia (CDH), isolated CDH, late presen-tation, postoperative outcome

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INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a congenital anomaly with an estimated incidence of 1:2000 to 1:4000 births.[1,2] About 5%–25% of cases are diagnosed after the first month of life and are classified as late-presenting cases.[3-5] This group of patients is thought to have a better postoperative course and prognosis than the early-presenting CDH group.[5-7]

There are few reports in the literature comparing late-presenting cases with isolated neonatal CDH cases,[6,8] and it is not definitely known whether late-presenting cases of CDH have a different set of symptoms, postoperative course, and outcome. We conducted this retrospective cohort study to find out if there were any differences between late-presenting and neonatal CDH cases in our center over the last 30 years. We also try to provide some clues to a better understanding of cases of late-presenting CDH.

MATERIALS AND METHODS

One-hundred and sixteen children with Bochdalek CDH were treated in the Department of Paediatric Surgery, Aghia Sophia Children's Hospital in Athens, Greece, between 1980 and 2010. The hospital is a tertiary referral center for the capital of Greece and covers a population of about five million individuals.

Hospital records for all children treated for CDH were retrospectively reviewed for postoperative complications, reoperations, postoperative time to full feeding, whether there was a normal x-ray at discharge,

and length of hospital stay. All patients were operated on by seven surgeons using a transabdominal approach. Patch repair was not performed in any patient included in the study. All data were collected using Microsoft® Excel™. SPSS® for Windows® was used for statistical analysis. The Student's *t*-test was used to compare continuous variables and Fisher's exact test to analyze categorical variables. All tests for significance were set at the 5% level and all *P*-values were two-tailed.

RESULTS

Twenty-two (24%) cases of neonatal CDH associated with unfavourable prognostic factors were excluded [Table 1]. Eleven other patients were excluded because they had undergone a patch repair.[9] Twenty-three patients (20%) were older than 1 month at the time of diagnosis and were designated as latepresenting CDH. All of them were included in this study. None of these patients had any associated anomalies that could affect postoperative outcomes.

There were two groups, one group with 71 cases of isolated neonatal CDH and a second group with 23 cases of late-presenting CDH. The characteristics of the studied patients are presented in Tables 2 and 3, which also show the symptoms at diagnosis for both groups.

Among the late-presenting cases there appeared to be a fewer left-sided CDH than right-sided CDH, but the difference was not statistically significant (P= 0.09).

Symptoms at presentation [Table 3]

Children with late-presenting CDH showed a wide spectrum of clinical symptoms. There appeared to be two main clinical variants: one with predominantly gastrointestinal symptoms (regurgitations, vomiting, lack of appetite, constipation, and abdominal pain) and another with predominantly respiratory problems (pulmonary infections, cough, dyspnoea, cyanosis, and tachypnoea). According to the clinical course, an acute or chronic type of manifestation was also discerned.

The neonatal group most commonly presented with acute onset of symptoms and most often with acute respiratory distress (35%). The patients in the late-presenting group generally presented with chronic respiratory symptoms (persistent cough, reduced exercise tolerance, chest pain) (66%); acute symptoms were seen in only two of these cases (8%). Acute respiratory symptoms were significantly more common in the neonatal group (P= 0.016), and chronic respiratory symptoms were more common in the late-presenting cases (P= 0.004). There was no difference between the two groups in acute gastrointestinal symptoms (bilious vomiting, bloody stools, acute abdominal pain) (P= 0.225), but chronic symptoms (retching, bloating, occasional vomiting, chronic abdominal pain) were more common in the late-presenting cases than in the neonatal ones (P= 0.006). Also, neonates with isolated CDH presented more often with failure to thrive as compared to their older counterparts (P= 0.03).

Table 1: Causes for patient exclusion

Extreme prematurity (<25 weeks)	10
Patch repair	11
Severe associated anomalies	
Cardiac	9
CNS disorder	5
Overall*	22

*Patients may belong to more than one category

Table 2: Patient characteristics

Patient characteristic	Neonatal	Late-presenting	*P*-value
Mean age at diagnosis	1 day	2 years	-
Left: Right (*n*)	56: 15	14: 9	*P*= 0.01
Male: Female (*n*)	44: 27	13: 10	*P*= 0.80
Liver herniated (*n*)	23	2	*P*= 0.03*

Table 3: Principal symptoms at diagnosis

Symptoms	Neonatal isolated CDH	Late-presenting CDH	*P*-value
Prenatally diagnosed	31 (44)	-	-
Incidental finding (no symptoms)	-	4 (17)	-
Respiratory*			
Acute	25 (35)	2 (9)	*P*= 0.016*
Chronic	18 (25)	14 (66)	*P*= 0.0044**
Gastrointestinal**			
Acute	15 (21)	2 (8)	*P*= 0.225
Chronic	14 (20)	12 (52)	*P*= 0.006**
Failure to thrive***	38 (54)	6 (8.7)	*P*= 0.03*

*Acute respiratory symptoms: chest pain dyspnoea, apnoeic episodes, episodes of desaturation, and other symptoms of <24 hours' duration and necessitating immediate treatment. Chronic respiratory symptoms: recurrent episodes of cough, pain, etc., that lasted for >24 hours and with mild clinical course; **Acute gastrointestinal symptoms: abdominal pain, vomiting, and melena of <24 hours' duration and necessitating immediate treatment. Chronic gastrointestinal symptoms: recurrent episodes of abdominal pain, episodes of vomiting, bloating, and loss of appetite that lasted >24 hours and had mild clinical course; ***Failure to thrive: inability to gain weight or loss of weight.

Complications and outcomes [Tables 4 and 5]

The most common postoperative complication in both groups was postoperative ileus, with no significant difference between the two groups. As per our definition, two or more of the following three findings were necessary in order to establish the diagnosis of postoperative ileus/obstruction in our patients: abdominal pain or distension, inability to pass gas and stools, and air–fluid levels on x-ray. Overall, there

Table 4: Short-term postoperative complications and their rates in the two groups

	Neonatal	Late-presenting	*P*-value
Postoperative ileus/obstruction*	9	2	*P*=1
Postoperative episode of fever/sepsis	5	1	*P*=1
Wound problems	1	1	*P*=.444
Pneumothorax	2		
Lung effusion	7	1	*P*=.67
Recurrence	2	-	
Reoperation**	3	-	
Death	8	-	-
Overall	35	5	*P*=.028*
Reoperation rate	4,2%	-	
Overall complication rate	49%	22%	*P*=.028*

*Two of the following findings were necessary in order to establish the diagnosis of postoperative ileus/obstruction in our patients: abdominal pain or distension, inability to pass gas and stools, and air–fluid levels on x-ray; **One reoperation because of intestinal obstruction and another two because of recurrence.

were 35 reported minor or major complications among the neonatal cases and only five complications among the late-presenting cases. The overall complication rate was 49% for the neonatal group and 22% for the older group. Postoperative episodes of fever/sepsis were seen exclusively in the neonatal group. Seven patients (10%) died postoperatively because of pulmonary hypoplasia and pulmonary hypertension. Three cases in the neonatal group required reoperation. One was reoperated because of intestinal obstruction and the other two because of recurrence. The reoperation rate was 4.2% for the neonatal group, while there was no recurrence in the late-presenting group. Six patients in the neonatal group and one in the late-presenting group had postoperative lung effusion.

The mean postoperative hospital stay and time to full feeds was 22 and 6 days, respectively, for the neonatal group, and 7 days and 3 days, respectively, for the late-presenting group. Thus, late-presenting cases had a significantly shorter postoperative hospital stay ($P< 0.05$) and time to achieve full feeds ($P< 0.05$). Thirty patients (42%) from the neonatal group and seventeen (74%) from the late-presenting group had a normal x-ray at discharge, a statistically significant difference ($P= 0.016$).

DISCUSSION

Late-presenting CDH was first reviewed by Kirkland in 1959.[10] Although his attention was mainly directed toward this congenital anomaly in adults, the review also included children. Since then, late-presenting CDH has been recognized by paediatric surgeons as a separate variant.[11,12] Late-presenting CDH constitutes 5%–45.5% of all CDH cases.[3-5] Although the diaphragmatic defects in delayed CDH are presumably congenital and anatomically similar to the defects of neonatal diaphragmatic hernia, the clinical presentation differs considerably from the more common newborn entity. Late-presenting CDH more commonly presents with chronic respiratory symptoms. Gastrointestinal symptoms are also more common than in neonatal cases.[4,6,7] In fact, the diagnosis of late-presenting CDH may be challenging due to its varying presentations.[13-15]

Table 5: Outcome measures in the two groups

	Neonatal	Late-presenting	*P*-value
Mean time to full feeding (days)	6	3	*P*= 0.048*
Normal chest x-ray at discharge*	30 (42%)	17 (74%)	*P*= 0.015*
Mean postoperative hospital stay (days)	22	7	*P*= 0.036*

*Normal-appearing lung size (in contrast to some neonatal cases that had small lung), no fluid collection, normal lung expansion, no lobe consolidation, and both diaphragmatic crura easily discerned and normal appearing. A consultant radiologist verified all of the above

No previous study has tried to statistically quantify the difference in symptoms between the two groups, despite indications from several reports that such differences do exist. In our study we tried to find out if there was any statistically significant tendency for different presentation and postoperative course between those two clinical entities. We are fully aware that the number of the patients included in the study is small, but we are dealing with a very rare clinical condition and it is difficult to gather enough patients to have the ideal study. In fact, ours is one of the largest ever reported series of late-presenting cases of CDH.

Our findings support the reports that there are differences between neonatal CDH and late-presenting CDH in preoperative symptomatology and clinical presentation. This difference is likely due to differences in the pathogenic course. Neonatal CDH results in immediate symptoms due to severe pulmonary hypoplasia and increased pulmonary resistance. Moreover, the small thoracic space in neonates cannot accommodate the abdominal viscera. On the other hand, late-presenting CDH cases present later because herniation may not occur for many years and when it does the lung is not hypoplastic, has normal reserves, and is more capable of adaptation; thus these patients have chronic symptoms rather than acute ones.[16,17]

In contrast to the high mortality and morbidity rates for newborn CDH, the prognosis for late-presenting CDH is usually more favourable.[5-7,16,17] Partly, this difference may be due to the presence of associated malformations in

neonatal cases: 27%–47% of cases of neonatal CDH have been associated with extrapulmonary malformations.[18-20]

Our data supports the general view that late-presenting CDH represents a distinct clinical entity, not only in terms of clinical presentation but also in terms of postoperative course and short-term outcomes, even when compared to neonatal isolated CDH cases. There seemed to be a tendency towards fewer complications for the late-presenting group and in our study there were no major complications such as death, recurrence, or reoperation. Thus, even after exclusion of cases of neonatal CDH with severe associated anomalies or extreme prematurity, there still was a more favorable postoperative course for late-presenting cases.

The reduced postoperative time to full feeds and shorter postoperative hospital stay also supports previous reports. We also used the last x-ray prior to discharge as a measure of postoperative lung expansion and normalisation. However, we know that this is not always dependent upon an operation, as many patients have lung hypoplasia before operative correction.[14]

This difference in complication rates and postoperative course in favour of late-presenting CDH might be attributable to certain features of these patients. First, in some if not in all late-presenting cases there is no herniation at birth and diagnosis is made only later in life after the abdominal organs have entered the thorax through a persistent congenital diaphragmatic defect. In such cases, as one might expect, there is better, if not entirely normal, lung function and therefore better postoperative outcome. Better lung function might also occur irrespective of the duration of a bowel herniation. Other possible explanations include the fact that a case presenting later in life might be easier to operate on (smaller gap, easier to approximate) and the fact that the child is better developed, better tolerates the stress of an operation, and has better healing.

On the other hand, we know that neonatal patients are transferred to a neonatal unit after a major operation, which might influence the postoperative reporting of complications. Personnel in neonatal units generally hold detailed daily reviews for each patient, making it more likely that minor complications are noticed. Still, a major complication such as a reoperation or recurrence would have been impossible not to record, no matter where the patient received postoperative care.

In conclusion, our data show a tendency towards fewer and less severe postoperative complications and better short-term outcomes for late-presenting cases of CDH. Larger, multicenter studies might help to definitely prove these differences, and we hope that our study provides the initiative for other researchers.

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