



## Case Report

# Congenital diaphragmatic hernia presenting as bowel obstruction in an infant: A case report

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### ARTICLE INFO

#### Article history:

Received 14-05-2024

Accepted 02-07-2024

Available online 03-07-2024

#### Keywords:

Congenital Diaphragmatic hernia

Bowel obstruction

Infant

Gastrointestinal

### ABSTRACT

**Background:** Congenital Diaphragmatic Hernia (CDH) is commonly diagnosed in antenatal scans or presents in the neonatal age group. Presentation of CDH in infants or older children with respiratory symptoms is less common. Similarly, the presentation of CDH with primarily gastrointestinal symptoms is also quite rare and poses a diagnostic and therapeutic challenge. An atypical case of CDH with later presentation with gastrointestinal symptoms is described. A failure to diagnose and correct it in a timely fashion could have a deleterious impact on the outcome.

**Case Presentation:** A case of a five-month-old child presenting with features of bowel obstruction is described. On evaluation, the child had rapidly progressive respiratory distress and gross abdominal distension, which was found to be due to left-sided CDH. A closed-loop obstruction with impending strangulation was managed with prompt surgery. The patient had a favorable outcome.

**Conclusions:** Late presenting CDH may present with primarily gastrointestinal symptoms with impending respiratory compromise. A high index of suspicion is necessary to ensure early diagnosis and management.

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## 1. Background

Congenital Diaphragmatic hernia (CDH) is being increasingly diagnosed antenatally with the increasing use and sensitivity of antenatal ultrasonography.<sup>1</sup> Most cases not diagnosed antenatally present immediately after birth or in the neonatal period with respiratory distress. Only a few cases escape detection early in life and present late, usually in the form of worsening respiratory symptoms or sometimes as an entirely incidental finding. CDH presenting with primary gastrointestinal complaints due to the diaphragmatic defect is very rare<sup>2–4</sup>. We present a 5-month-old male child, who presented with symptoms and signs suggestive of bowel obstruction and was found to have CDH and had a fulminant preoperative and postoperative

course.

## 2. Case Presentation

### 2.1. History and physical examination

A five-month-old child presented to the Emergency with complaints of progressively increasing abdominal distension for two days along with multiple episodes of non-projectile bilious vomiting and non-passage of stools. There was no history of associated pyrexia. No antenatal ultrasound was done and the child was born via vaginal delivery in a primary care hospital uneventfully. The child was doing fine from birth till the present episode of abdominal distension. On examination, the child had tachycardia, hypotension, tachypnea, subcostal retractions, and low saturation on pulse oximetry (85 %) on room air. There was decreased air entry on the left side with gross

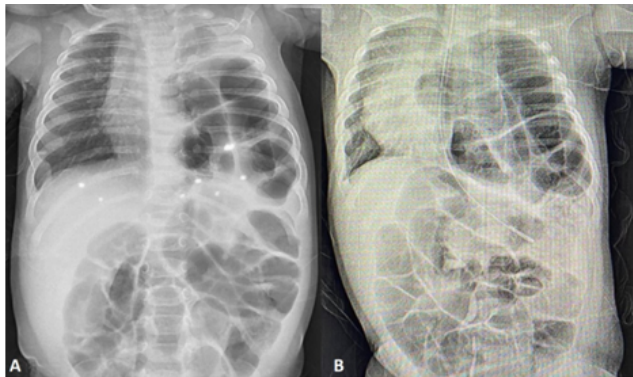
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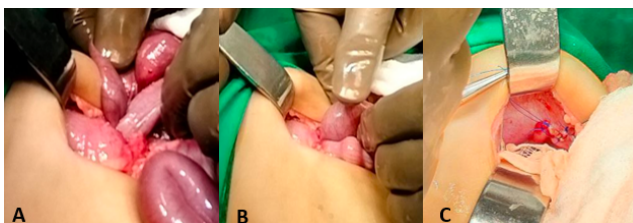
abdominal distension.

## 2.2. Investigations and Immediate management

Immediate resuscitation was started despite which there was persistent deterioration. The child was intubated because of clinical deterioration, following which an urgent bedside radiograph was ordered. The radiograph revealed Left left-sided diaphragmatic hernia with bowel herniation into the left hemithorax. The patient was resuscitated and was admitted to the Paediatric intensive unit for further management. An urgent bedside echocardiogram ruled out pulmonary hypertension and was grossly normal. The child had persistently increasing ventilatory requirements with worsening acidosis on blood gas even after adequate fluid and inotropic support. On auscultation, right-sided breath sounds were also decreased. There was no decrease in abdominal distension and no output in the nasogastric tube. A repeat chest radiograph was ordered which revealed massively dilated bowel loops in the left hemithorax with compression of the opposite side as well.



**Figure 1:** Supine radiograph depicting (A) bowel loops in left hemithorax suggestive of left CDH, with no mediastinal shift and left-expanded right lung fields. (B) Repeat radiograph done after 4 hours depicting progressive dilatation of bowel loops inside left hemithorax with mediastinal shift and compression of right lung fields.



**Figure 2:** Intraoperative image showing (A) small diaphragmatic defect with constriction ring causing closed loop obstruction and impending strangulation. (B) After the extension of the diaphragmatic defect bowel being reduced into the abdomen. (C) Post-complete reduction, completed primary repair of CDH.

## 2.3. Operative treatment

The patient was urgently taken up for emergency surgery and a laparotomy was done. Intra-operatively almost the whole of the distal small bowel was found in the thoracic cavity through a small 2 cm defect in the left hemidiaphragm with a constriction ring effect at the site of the defect (Figure 1). The bowel was stuck at the defect and reduction was not possible. The defect was extended by incision laterally, following which the bowel was reduced. The diaphragmatic defect led to a closed-loop obstruction of the bowel inside the left hemithorax. The proximal stomach and proximal small bowel were also dilated. The stomach, liver, spleen, and other bowel were intraabdominal. The diaphragmatic defect was repaired primarily and the abdomen closed (Figure 2).

## 2.4. Outcome

The patient had post-operative ventilator-associated pneumonia requiring prolonged intubation. Nasogastric feeds were started on postoperative day 3 with a gradual incremental increase. A tracheostomy was done on postoperative day 14 and the patient was subsequently discharged in stable condition on oral feeds. The patient is doing well on follow-up after 6 months.

## 3. Discussion

Congenital Diaphragmatic hernia (CDH) results from a defect in the formation of the diaphragm causing herniation of abdominal organs in the thorax invariably with a component of pulmonary hypoplasia, pulmonary hypertension, and loss of abdominal domain. CDH is a relatively common congenital anomaly seen in neonatal surgical intensive care units with an incidence ranging from 1.5 to 1.7 per 10,000 live births.<sup>1</sup> Bochdalek type (posterior defect) is far more common (more than 20 times) than the Morgagni type (anterior defect). The left-sided defect is seen in almost 85-90 % of cases of Bochdalek CDH.<sup>1</sup>

CDH presenting in the neonatal period or those antenatally diagnosed present a whole battery of management challenges but can be easily diagnosed based on the clinical picture, index of suspicion, and imaging. However, the diagnosis of those presenting late is often delayed due to a low index of suspicion. This is also partially because of the wide variety of clinical presentations of late presenting CDH (LP-CDH). Presenting symptoms in LP-CDH can be acute or chronic in onset.<sup>5</sup> Acute symptoms include acute respiratory distress which may be diagnosed wrongly as either pneumothorax or pleural effusion. There could be acute intestinal obstruction due to bowel constriction at the site of diaphragmatic hernia leading to bowel ischemia and gangrene. Chronic symptoms could range from chronic respiratory symptoms (recurrent cough, dyspnea, shortness

of breath, chest infections), failure to thrive, and chronic gastrointestinal symptoms (recurrent abdominal pain, vomiting, perforation, gastroesophageal reflux, or vague symptoms). The predominant reason for gastrointestinal symptoms presenting as LP-CDH could be attributed to the small defect in the diaphragm leading to the relative paucity of cardiorespiratory compromise.<sup>6</sup> Other reasons for LP-CDH could be a delayed rupture of the peritoneal sac or occlusion of the defect by a solid organ due to raised abdominal pressure.<sup>3</sup>

The index case had an initial presentation with features suggestive of bowel obstruction only. However, the constriction at the level of CDH caused a closed-loop obstruction phenomenon leading to sudden and exponential dilatation of the bowel inside the thoracic cavity. This led to secondary respiratory symptoms with increasing compression of the ipsilateral followed by contralateral lung and mediastinal shift. Timely surgery on an urgent basis meant that the bowel ischemia was halted and no bowel ischemia leading to perforation or necrosis occurred. Koh. Et al.<sup>4</sup> described a similar case in an adult in which bowel obstruction further led to perforation in the transverse colon due to CDH. Bowel obstruction due to LP-CDH has been described in both right and left-sided CDH.<sup>7,8</sup> Bharani et al. described a similar 8-year-old child who presented with respiratory distress and bilious vomiting, who was found to have CDH, and who succumbed even after emergency laparotomy and reduction.<sup>9</sup>

Based on the time of herniation, severity of pulmonary hypoplasia, time of presentation of symptoms, and sequelae, a classification system of CDH has been described by Wiseman et al.<sup>10</sup> Invariably early surgery is required in LP-CDH to prevent more sinister complications as these patients have an otherwise good prognosis.<sup>11</sup> Any delay in surgery for medical stabilization as practiced in neonatal CDH carries the risk of bowel strangulation and worsens prognosis.<sup>12,13</sup>

#### 4. Conclusion

A combination of gastrointestinal symptoms with respiratory distress in any infant or even older child should be suspicious for CDH. Early evaluation and diagnosis followed by expeditious surgery is paramount to prevent complications. Prompt surgical intervention is

necessary for a satisfactory outcome.


#### Acknowledgements


All people that participated in this paper are listed as co-authors. No acknowledgement.

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**Cite this article:** Mahajan D, Midha H. Congenital diaphragmatic hernia presenting as bowel obstruction in an infant: A case report. *IP J Surg Allied Sci* 2024;6(2):69-71.