

## Case Reports

# Acute Complications of Late-presenting Congenital Diaphragmatic Hernia in Children: Report of 2 Cases and Review of Literature

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### Abstract

Congenital diaphragmatic hernia can present beyond the neonatal period. This article reports 2 cases of late-presenting congenital diaphragmatic hernia (CDH) in children presenting with symptoms of acute abdominal pain and respiratory distress. The diagnosis was confirmed on chest X-ray which showed herniation of abdominal organs into the left hemithorax and mediastinal shift to the right. In one child, the diagnosis was further confirmed with limited contrast follow through. The hernia defects were repaired through laparotomy and laparoscopy respectively. Late-presenting CDH poses diagnostic difficulties and children presenting with non specific respiratory or abdominal symptoms may actually suffer from acute complications of CDH.

### Key words

Bochdalek; Complications; Congenital Diaphragmatic Hernia; Late-presenting; Morgagni

### Introduction

Congenital diaphragmatic hernia (CDH) occurs in about 1 in 3000 births. Majority of patients are diagnosed antenatally or present with respiratory distress within the first few hours of life. Late presenting CDH is a milder form that presents beyond the neonatal period and accounts for 5-25% of cases.<sup>1,2</sup> It can manifest through a wide range of acute or chronic, gastrointestinal or respiratory symptoms and poses diagnostic difficulties. As a result of diagnostic delay, there are risks of inappropriate treatment, potential morbidity and mortality.<sup>1-3</sup> We herein report 2 children with acute complications resulting from late-presenting diaphragmatic hernia.

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### Case 1

A previously healthy 5-year-old girl presented with acute onset of dyspnoea and upper abdominal pain. It was associated with vomiting of undigested food. There was no chest pain or previous respiratory illness.

On examination, she was dyspneic with respiratory rate up to 36/min. The oxygen saturation (SaO<sub>2</sub>) was 96-98% on room air. Trachea was deviated to the right and left chest was dull to percussion and air entry was reduced. Abdomen was soft and non tender.

Baseline investigations including complete blood counts, liver and renal function and blood gases were normal. Haemoglobin was 12.3 g/dl, white blood cell 15.9 x 10<sup>9</sup>/l, platelet 283 x 10<sup>9</sup>/l and pH was 7.37. Chest X-ray (Figure 1a) showed a large gas filled shadow in the left hemithorax. The tip of the nasogastric tube was also in the left hemithorax. In view of stable vitals and to further confirm the diagnosis, limited contrast follow through (Figure 1b) was performed and confirmed the stomach in the left hemithorax with persistent hold up of contrast in the stomach despite different positioning.

In view of the possible strangulation of stomach, emergency laparotomy was performed. A left subcostal incision was made. It revealed a left Bochdalek defect with

the body of stomach, small and large bowel and spleen herniated to the left hemithorax. Abdominal contents were all reduced into the abdomen and were all healthy. The diaphragmatic defect was primarily closed without the need of synthetic mesh. She was discharged on post-operative day 11. She remains well 6 months post-operatively.

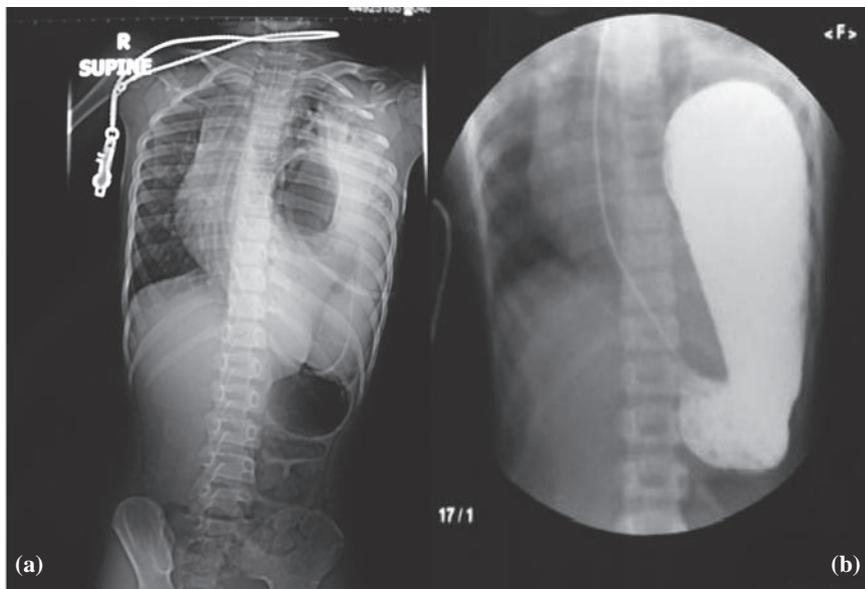
## Case 2

A 3-year-old boy complained of sudden onset of colicky periumbilical pain. There was no vomiting or gastrointestinal bleeding. On examination, the SaO<sub>2</sub> was 97% on room air. The abdomen was soft with central

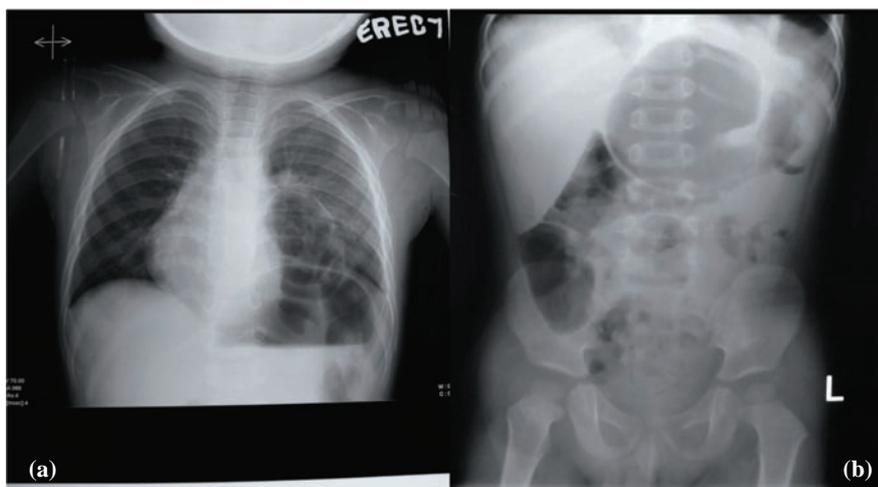
tenderness. Air entry was equal on both sides of the chest.

Baseline investigations including complete blood count, renal and liver function, and clotting profile were obtained. Haemoglobin was 12.5 g/dl, white cell count  $19.1 \times 10^9/l$ , platelet  $481 \times 10^9/l$ , renal function and clotting profile were normal. Chest X-ray (Figure 2a) showed a bowel loop in the left hemithorax with mediastinal shift to the right. Abdominal X-ray (Figure 2b) showed dilated bowel loops. The provisional diagnosis was diaphragmatic hernia or hiatus hernia with strangulation of intestine.

Emergency laparoscopy was performed. Laparoscopy revealed a Morgagni diaphragmatic hernia with the omentum and transverse colon in the left hemithorax. Hernia contents were reduced and omentum was divided



**Figure 1** (a) A gas filled structure in the left hemithorax with mediastinal shift to the right. (b) Limited contrast follow through showing stomach in left hemithorax with persistent contrast hold up in stomach despite different positioning.



**Figure 2** (a) Loop of bowel in the left hemithorax with mediastinal shift. (b) Abdominal X-ray showing dilated bowel loops.

from the rim of hernia defect. The reduced intestine was healthy. The defect was about 2 cm in size and was successfully repaired laparoscopically.

He was discharged on post-operative day 5. He remains well on follow up 9 months after the operation.

## Discussion

95% of late-presenting congenital diaphragmatic hernias have a posterolateral defect, with 80% being left sided and the rest being right sided, central or bilateral. The condition is male predominant with male to female ratio being 2:1.<sup>1</sup> Patients can present acutely or chronically through a wide range of non-specific gastrointestinal or respiratory symptoms. Baglaj, in his review of 125 articles<sup>1</sup> including 362 children, reported that 43% present with respiratory symptoms, 33% with gastrointestinal symptoms, 13% with a mixture of the 2 groups of symptoms and 11% being asymptomatic. Common acute symptoms include pneumonia, upper respiratory infection, respiratory distress, vomiting and abdominal pain. Chronic symptoms include chronic respiratory difficulties, silent aspiration, cough and failure to thrive. Most of the left sided CDH present acutely (60.5%) while most right sided delayed CDH present with chronic symptoms (57.4%) Right sided CDH present younger than left sided defects and 2/3 present within the first year.<sup>1</sup> As compared to neonatal hernias, late-presenting CDH have several features, namely a high incidence of right sided defects, presentation with respiratory symptoms in younger patients and gastrointestinal symptoms in older ones and younger age of patients with right CDH as compared with left sided defects.<sup>2</sup> Contrary to neonatal CDH manifesting uniformly with respiratory distress, no single symptom is pathognomonic for the disease and this presents challenging diagnostic difficulties, especially in right sided hernias.<sup>1</sup> Misdiagnosis of right sided CDH as pneumonia or emphysema had been reported.<sup>1-3</sup> However, the overall prognosis of late presenting CDH is better than neonatal ones.<sup>2</sup>

There is a lower incidence of associated anomalies as compared with neonatal CDH, including cardiac and great vessel anomalies, pulmonary anomalies (lung sequestration, accessory lobe, lung cyst), funnel chest, chromosomal anomalies, urogenital anomalies (hypospadias, horseshoe kidney) and meningomyelocele.<sup>1,2</sup>

Commonly employed investigations include chest X-ray (with or without nasogastric tube insertion), contrast studies, computer tomography (CT) scan and laparoscopy.<sup>3</sup> The

diagnostic adequacy of plain radiographs is estimated at 27-98%<sup>3</sup> CT scan can show diaphragmatic discontinuity, intrathoracic herniation of abdominal contents, and the "collar sign", which refers to the waist like constriction of mesenteric folds.<sup>3</sup>

Contents of hernia differ between left and right sided defects. The stomach, small and large bowel and the spleen are frequently encountered in left sided hernias. The liver is most often found in right sided defects.<sup>1</sup>

Complications of CDH can result from bowel perforation or strangulation, gastric volvulus and perforation, omental infarction, splenic torsion and incarceration. It has been reported in a number of cases that late-presenting CDH has been misdiagnosed as pneumothorax or pleural effusion and a chest drain was inserted, leading to gastric perforation, liver trauma and colonic perforation.<sup>1,2</sup> Causal factors for postoperative mortality include sepsis, respiratory insufficiency and bowel strangulation. The mortality risk has been estimated at 2.2%.<sup>1</sup>

Both laparotomy and laparoscopic repair were respectively employed in our 2 patients. The choice of surgical approach depends on the patient's condition as well as the availability of surgical expertise and facilities. Traditionally, repair of congenital diaphragmatic hernia is by laparotomy. This is technically less demanding and enables easy reduction of viscera, especially for large defects. Laparoscopic repair confers the benefit of minimally invasive surgery of less wound pain and faster recovery.<sup>4</sup> Laparoscopic repair was reported to have a reduced post operative stay and is increasingly employed in practice.<sup>5,6</sup> There were few reports on laparoscopic repair of Bochdalek type CDH in the literature,<sup>4,7</sup> Laparoscopic primary and mesh repair of Morgagni and traumatic CDH have also been described.<sup>5,8</sup>

But this is technically more demanding with a longer learning curve. Thoracoscopic approach is preferred for Bochdalek hernias because of potential technical difficulties in the laparoscopic approach, namely the risk of injuring the spleen and splenic vessels while trying to pull it into the abdomen and poor view of diaphragmatic defect when the abdominal viscera have been successfully relocated in the peritoneal cavity. Thoracotomy is also used, especially in chronic hernias in which adhesions between lung and abdominal organs are frequently encountered.<sup>4</sup>

## Conclusion

Correct diagnosis is crucial in the management of

children with acute complications of late-presenting congenital diaphragmatic hernia. Diagnosis can be challenging given the wide spectrum of clinical manifestations. Basic investigations such as chest X-ray can provide adequate information in the diagnosis as illustrated in our cases. Early surgical repair should be performed in view of the possibility of strangulation.

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