

# Late Presentation of Congenital Diaphragmatic Hernia- Anaesthetic considerations

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## Abstract

Delayed presentation of congenital diaphragmatic hernia (CDH) is not uncommon and can represent 5-30% of total CDHs.<sup>1</sup> Time before the diagnosis may be prolonged, sometimes to the adult period. Respiratory and gastrointestinal symptoms are frequent but not specific. Children presenting with gastrointestinal symptoms have been shown to be significantly older than those presenting with respiratory symptoms.<sup>2</sup> Late presentations can be misleading to a clinician. However, when a diagnosis of CDH is established, it must be promptly treated surgically in order to avoid complications such as strangulation or bowel perforation. Outcome is usually favourable after surgery. We hereby report a case of congenital diaphragmatic hernia presenting as acute chest pain due to midgut volvulus in left thoracic cavity.

## CASE REPORT

A previously healthy 14 year old male child was admitted to the hospital with a history of acute pain in the left side of chest and grade 1 dyspnoea for 4 days. Pain was constant, dull aching, more upon deep inspiration. Pain was much severe and was associated with vomiting on the day of admission. There was decreased air entry at the left posterior side of the chest, but no other abnormality on auscultation. The abdomen was scaphoid but non tender.

A plain erect chest radiograph (Fig. 1) showed that left dome of diaphragm and left costophrenic angle were obscured by moderate pleural effusion along with loss of left cardiac border silhouette. Mediastinum was displaced to the right. Ultrasonography (USG) of the chest revealed left sided consolidation with mild pleural effusion.

**Figure 1**

Figure 1

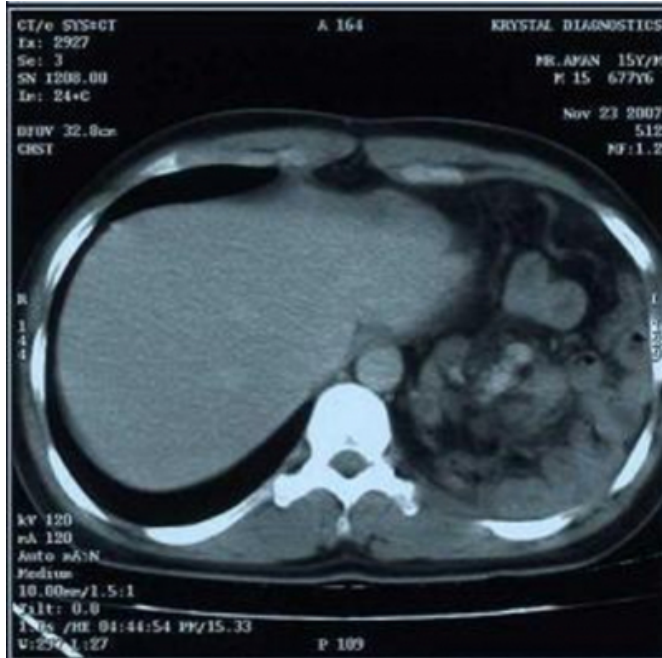


For further evaluation, Computed Tomography (CT) was done and it was extremely helpful for evaluating this patient.

CT findings (Fig. 2) were suggestive of possible congenital left bochdalek hernia with herniation of small gut loops and passive collapse of left lower lobe.

**Figure 2**

Figure 2



A diagnosis of late onset left congenital diaphragmatic hernia (LCDH) was made and he was posted for operation.

Following premedication with glycopyrrolate ( $0.01\text{mg kg}^{-1}$ ) and midazolam ( $0.05\text{mgkg}^{-1}$ ) intravenously, the child was preoxygenated with 100% oxygen. Pulseoximeter and ECG monitors were attached and intravenous fluids were started. Cricoid pressure was applied and rapid sequence induction was performed using thiopentone 250mg and succinylcholine 75 mg. As this patient presented to emergency operation theatre and because of no availability of double lumen tube, he was intubated with a standard 8.0 mm cuffed orotracheal tube and the right mainstem bronchus was intentionally intubated by inserting the tube to a distance of 24.5 cm at the upper incisors. The cuff was inflated, one lung ventilation was confirmed by auscultation and cricoid pressure was removed. Anaesthesia was maintained using halothane 0.5% in 100% oxygen, supplemented with i.v tramadol.

A left subcostal laparotomy showed a large congenital defect of left hemidiaphragm (10x5cm). The left hemithorax contained the whole of the small and part of large gut. There was malrotation of large gut and spleen was absent. The left hemithorax was decompressed and malrotation was

corrected by returning the viscera to the abdominal cavity. The tracheal tube was withdrawn to 21 cm at the upper incisors and two lungs ventilation was commenced. Intercostal drain was inserted and the peritoneal cavity was closed. A postoperative roentgenogram showed (Fig. 3) showed ipsilateral pneumothorax and another x-ray after 7 days was normal and confirmed uneventful recovery.

**Figure 3**

Figure 3



## DISCUSSION

The incidence of CDH is 1:2000 to 1:5000 with equal gender preference.<sup>3,4,5,6</sup> Up to 20% of cases are associated with other congenital anomalies (e.g. mental retardation, Down's syndrome, cleft lip, heart defects).<sup>4,5,6</sup>

The defect arises during embryological development because of incomplete closure of the diaphragm or early migration of the midgut from the umbilical coelom into the abdominal cavity before the diaphragm is fully formed (around 10 weeks); thus allowing the midgut to herniate into the chest cavity. Related abnormalities include complete agenesis of one or both hemidiaphragms and diaphragmatic eventration, where the diaphragm mainly consists of pleuroperitoneal membrane with little or no muscular component, allowing it to be pushed upwards by the abdominal contents.<sup>3,4,5,6</sup>

Herniation may occur at three sites: 1) posterolaterally through the foramen of Bochdalek (78-90% of cases); 2) retrosternally via the foramen of Morgagni (1.5-6% of cases) ; 3) via the oesophageal hiatus (14-24% of cases). Left sided

herniation is more common, as the right hemidiaphragm develops slightly earlier than the left, and is somewhat protected by the liver.<sup>3,5,6</sup> The lung on the affected side is hypoplastic, resulting in right to left shunting of blood.

If CDH does not present acutely in the neonatal period, diagnosis of the defect is more difficult as the signs and symptoms become chronic, vague and inconsistent. Symptoms can be either cardiorespiratory or gastrointestinal, with the latter becoming more prominent as the age of onset of symptoms increases. Chronic cardiorespiratory symptoms include coughing, dyspnoea and recurrent chest infections. Chronic gastrointestinal symptoms include epigastric or subcostal discomfort, nausea and vomiting, fullness and bloating, cramping or diarrhoea.<sup>4,5,6</sup>

A total of 70-95% of cases are diagnosed in the neonatal period based on a triad of: (i) respiratory distress, often severe with cyanosis and decreased breath sounds on the affected side; (ii) scaphoid abdomen (iii) contralateral mediastinal shift. Auscultation of bowel sounds over the involved hemithorax is an unreliable sign. Emergency correction is required in these cases.<sup>3,7</sup>

A plain chest radiograph following passage of a nasogastric tube is very useful in arriving at a correct diagnosis since there is misinterpretation of diaphragmatic hernia with pleural effusion or pneumothorax (as it was seen in our case) with the potential hazard of inserting a chest drain tube into herniated viscus.<sup>8</sup>

Acute herniation of viscera through a congenital diaphragmatic defect in the adult is usually precipitated by increased intra-abdominal pressure because of pregnancy, postural changes or trauma. Once in the thoracic cavity, the viscera usually remain there because of abdomino-thoracic pressure gradient. Small defects tend to present with strangulation of the viscera leading to necrosis, perforation and peritonitis; whereas larger defects present with respiratory distress and compromise of the circulation. Surprisingly, this patient had mild symptoms but defect was large.

The mass effect of the intra-thoracic viscera causes cardiovascular impairment by direct compression of the heart and mediastinal shift, which can kink the vena cava and pulmonary veins, impair venous return to the heart, and cause cardiac output to decrease. A plain chest radiograph will confirm the presence of bowel loops in the thoracic cavity. Once the diagnosis has been established, urgent

surgical correction is required. Anaesthetic management of CDH presenting acutely in the adult is poorly described, but the principles resemble management of late diagnosed traumatic diaphragmatic hernia.

Large bore i.v. access should be gained and fluid resuscitation should be commenced. Invasive arterial and central pressure monitoring should be considered. The patient is at high risk of aspiration because of gastrointestinal obstruction, and therefore antacid premedication should be given, and a nasogastric tube should be inserted and aspirated if possible before rapid sequence with cricoid pressure.<sup>9</sup> If difficult intubation is anticipated, awake fibreoptic intubation or tracheostomy under local anaesthesia should be considered. Expansion of the viscera is likely to worsen the mass effect and impair circulation and respiration. Face-mask ventilation, with potential gastric insufflation; and nitrous oxide anaesthesia should therefore be avoided.

In theory, positive pressure ventilation might preferentially ventilate the normal lung rather than the collapsed lung. However, any re-expansion of the collapsed lung may exacerbate the mass effect, with rapid and disastrous worsening of the circulation. The collapsed lung should therefore be isolated and ventilation of the normal lung started with small tidal volumes and pressures using a double lumen tube, until the affected hemithorax has been decompressed. If this is not possible, a single lumen tube with the bronchial blocker should be considered. As this child presented to emergency operation theatre with acute chest pain and considering the risk of perforation single lumen tube was used but, ideally is not recommended.

In conclusion, CDH in an adult is a diagnostic and anaesthetic challenge. The diagnosis should be considered in any patient presenting with acute chest, abdomen and chronic, vague and inconsistent cardiorespiratory symptoms.

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