

## DELAYED PRESENTATION OF CONGENITAL DIAPHRAGMATIC EVENTRATION IN A 12-YEAR-OLD GIRL: A RARE CASE REPORT.

**Dr. Harsh Rajeev Mehta\*** - junior resident, general surgery, GMC Aurangabad, Maharashtra, India.

**Dr. Sriranjani Iyer** - junior resident, general surgery, GMC Aurangabad, Maharashtra, India.

**Dr. Sarojini P. Jadhav** - Professor and head, general surgery, GMC Aurangabad, Maharashtra, India.

**Dr. Junaid Shaikh** - Associate Professor, general surgery, GMC Aurangabad, Maharashtra, India.

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### Article Info

#### \*Corresponding Author:

Dr. Harsh Rajeev Mehta

Email:

harshees81@gmail.com

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### Abstract

**Introduction-** A diaphragmatic hernia is a protrusion of the abdominal viscera through the diaphragm with or without a hernial sac.

Its estimated incidence has been reported to be 1 in 2000 to 5000 live births. Late presentation of congenital diaphragmatic hernia is uncommon, accounting for 5-30% of all cases.

**Case presentation-** A twelve-year-old female child presented with chief complaints of non-specific, intermittent, vague abdominal pain with no other complaints. CECT of abdomen was suggestive of left sided congenital diaphragmatic hernia.

Intraoperatively, there was evidence of: -

1-Eventration of left hemidiaphragm

2-Evidence of stomach, spleen, splenic flexure and transverse colon in left hemithorax.

3-Hypoplasia of left lung.

Posterior half of diaphragm was approximated with posterior peritoneum with polydioxanone sutures and strengthening done.

**Discussion-** This type of hernia usually presents later in life with respiratory or gastrointestinal problems that may be mistaken for acquired disease. Beyond infancy, diaphragmatic hernia is an unusual finding. There are different causes other than congenital diaphragmatic hernia like trauma, phrenic nerve palsy and longstanding acquired hiatus hernia.

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Conclusion- Clinical suspicion and chest radiograph remain key in initial diagnosis. Late-presenting congenital diaphragmatic hernias should be included in the differential diagnosis of any child with persistent gastrointestinal or respiratory problems associated with abnormal chest X-ray film findings.

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### Introduction:

A diaphragmatic hernia is a protrusion of the abdominal viscera through the diaphragm with or without a hernial sac. Congenital diaphragmatic hernia (CDH) is the result of incomplete closure of the normal pleuroperitoneal canal during fetal development [1]. Its estimated incidence has been reported to be 1 in 2000 to 5000 live births [2]. Congenital diaphragmatic hernia usually presents with respiratory distress during the neonatal period and is associated with pulmonary hypoplasia.

Late presentation of congenital diaphragmatic hernia is uncommon, accounting for 5-30% of all cases in several studies [3]. The types of congenital diaphragmatic hernias with late presentation include foramen of Morgagni (retrosternal defect), foramen of Bochdalek (posterolateral defect), paraesophageal hernia and eventration [3]. Although diaphragmatic defects in delayed congenital diaphragmatic hernia are presumably congenital and anatomically similar to the defects of neonatal diaphragmatic hernias, clinical presentation, operative management and complications differ considerably from the more common newborn entity [4]. Clinical presentation of the late presenting diaphragmatic hernia is different from the newborn form and that a diaphragmatic hernia in a previously healthy child can be the cause of various abdominal and thoracic symptoms [5,6,7]. The clinical presentation can vary from a longstanding and intermittent nonspecific course to a life-threatening acute event. In contrast to the high mortality and morbidity rates for newborn congenital diaphragmatic hernias, prognosis for late-presenting congenital diaphragmatic hernias is usually favourable [8-10]. About 80% of cases occur on the left side, 19% on the right, and less than 1% are bilateral [11].

### Case Presentation:

A twelve-year-old female child presented with chief complaints of non-specific, intermittent, vague abdominal pain for one month.

There were no associated complaints of pain, fever, vomiting, constipation, diarrhoea, passage of blood in urine or stools, loss of weight or appetite, breathlessness or chest pain.

There was no history of trauma to abdomen or chest.

Patient was not a known case of any major surgical or medical illnesses with no history of previous hospital admissions or undergone any operative procedures.

On physical examination: -

Per abdomen - was soft to touch, with no tenderness, guarding, rigidity or distention felt. Bowel sounds were heard normally.

Respiratory system examination - air entry was reduced in lower lobe of left lung with no adventitious sounds.

Rest of systemic examination was within normal limits.

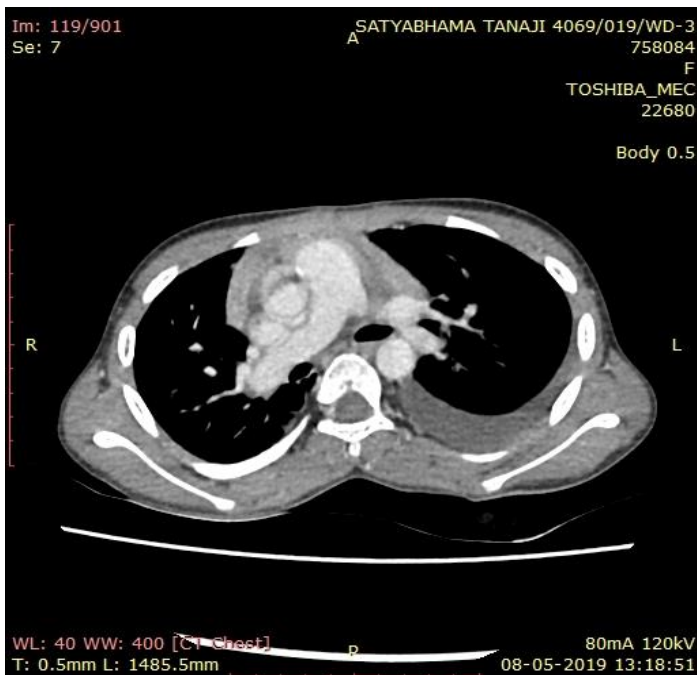
Routine laboratory investigations were within normal limits.

On 2-D Echo - normal findings were noted.

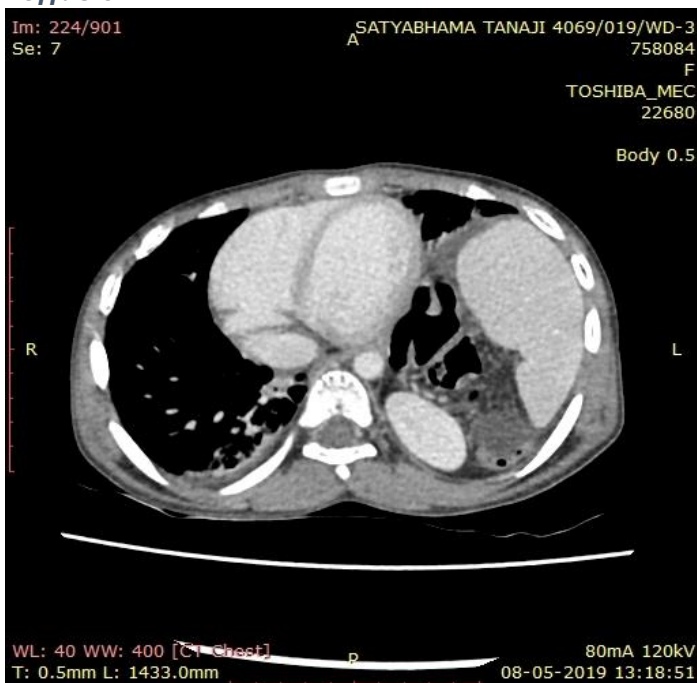
Ultrasonography of abdomen and pelvis showed evidence of upward migration of spleen, splenic flexure of colon and stomach upto 5<sup>th</sup> and 6<sup>th</sup> intercostal space with thickening of left hemidiaphragm suggestive of diaphragmatic eventration.

On contrast enhanced computed tomography scan of abdomen and pelvis, a defect of size 8.2cm was noted in posterior 2/3<sup>rd</sup> of left hemidiaphragm, with thickening of anterior aspect of diaphragm while posterior half could not be visualised. There was herniation of distal one-third of transverse

colon, splenic flexure, spleen and left kidney noted in left hemithorax with mild left sided pleural effusion and collapse of lower lobe of right lung. CECT findings were suggestive of left sided congenital diaphragmatic hernia.



**Figure 1-**shows left lung collapse with pleural effusion



**Figure 2-** shows left kidney, adrenal gland, spleen herniation into left hemithorax



**Figure 3-** shows bowel loops in left hemithorax

Patient was planned to undergo an elective surgery by abdominal route. Following complete workup and anesthesia fitness, an exploratory laparotomy was performed.

Intraoperatively, there was evidence of: -

- 1- Eventration of left hemidiaphragm
- 2- Evidence of stomach, spleen, splenic flexure of colon, left kidney, left adrenal gland and transverse colon in left hemithorax.
- 3- Hypoplasia of left lung.

Abdominal organs were healthy with no evidence of gangrenous changes. All the contents were reduced back into abdomen.

Posterior half of diaphragm was approximated with posterior peritoneum with polydioxanone sutures and strengthening done. Patient tolerated procedure uneventfully. She was extubated within the first two hours and maintained oxygen saturation levels on low oxygen supply.

Postoperative ultrasound of abdomen and chest xray were done on the third day having evidence of collapse consolidation of left lung parenchyma with mild to moderate left sided pleural effusion for which pleural tapping was done serially over the next four days.

Lung expansion gradually improved on the left side following pleural tapping, nebulisation and chest spirometry exercises.

Patient was vitally stable and resumed routine activities approximately six days postoperatively

with no complaints of breathlessness or chest pain.

On the tenth day postoperatively, patient was discharged.

At follow up two and four weeks later, patient was comfortable with no complaints.

### **Discussion:**

Congenital diaphragmatic hernias usually present in the newborn period with little difficulty in diagnosis, especially when gas-filled bowel loops are present in the hemithorax. Occasionally this type of hernia will present later in life with respiratory or gastrointestinal problems that may be mistaken for acquired disease. Beyond infancy, diaphragmatic hernia is an unusual finding. There are different causes other than congenital diaphragmatic hernia, for example, trauma, phrenic nerve palsy and longstanding acquired hiatus hernia.

Gastrointestinal symptoms predominate in left sided hernias whereas respiratory symptoms are more common in right-sided lesions [12].

Left-sided congenital diaphragmatic hernias are more frequent than right-sided hernias and commonly produce severe respiratory distress and cyanosis in the newborn infant. In older children they are usually not discovered until they become large enough to cause symptoms referable to the respiratory or gastrointestinal tract.

Clinical suspicion and chest radiograph remain key in initial diagnosis.

The accuracy of the plain radiograph in evaluating a suspected case of late-presenting CDH has been debated in the literature [13]. Ultrasound should be the complementary imaging modality of choice in all suspected diseases of the diaphragm in children, including congenital diaphragmatic hernias with delayed presentation.

The hernia may be intermittent and the presence of a normal chest radiograph does not exclude the diagnosis [14]. Further imaging evaluation is warranted if clinical suspicion is high like contrast series radiographs or CT scan. The diagnosis of a delayed diaphragmatic hernia is of importance because of the high non-operative mortality of

this condition [13].

Following diagnosis, the management of acute presentation of congenital diaphragmatic hernia should be aimed to achieve urgent bowel decompression. This is critical to prevent complications such as gastric volvulus, bowel strangulation or respiratory complications.

There has been some debate concerning management of congenital

eventration of the diaphragm (16). Symptomatic cases require imbrication through a thoracotomy or laparotomy approach. The presence of associated ipsilateral defects should be considered. Acute symptoms necessitate urgent management including contrast studies such as barium meal or barium enema, followed by prompt surgical intervention.

Some researchers have given little consideration to operative timing in the patients who present beyond the neonatal period: Whittaker suggested that correction is not urgent in older children. (17) Kirkland and MacDougall et al advised immediate surgery even if the presentation is a chance finding. (18)

Though in some cases it may be difficult to differentiate between

congenital diaphragmatic hernia and eventration, embryologically,

the latter results from failure of muscular buttressing without early intestinal herniation. Histologically, it usually consists of peritoneum, fibrous connective tissue which may contain some muscle fibers, and pleura (19, 20). Differentiation between an eventration and diaphragmatic hernia may be impossible without surgery.

Some researchers suggested that correction of diaphragmatic defects is not urgent in older children [17]. However, the vast majority of pediatric surgeons advise surgical correction without delay once the diagnosis has been established [21-27]. Emergency surgery is considered if symptoms are acute, otherwise the patient is scheduled for the next routine operative list [27]

The contents of late-presenting CDH vary considerably, ranging from only colon to nearly the entire bowel, spleen, liver, and left kidney. The presence of the stomach in the chest during the

prenatal diagnosis of CDH was reported to be a poor prognostic sign [28].

Prognosis of late-onset congenital diaphragmatic hernia is excellent once the correct diagnosis is

made. However, making the correct diagnosis is challenging because of its diverse clinical presentations.

**Table 1 below shows various reported cases with diaphragmatic pathologies.**

**Table 1** Reported cases with simultaneous multiple ipsilateral congenital diaphragmatic pathologies (excluding hiatus hernias)

Author	Age	Sex	Congenital pathologies	Defect contents	Surgery	Outcome
Epstein et al (7)	56 years (adult)	F	multiple defects in right central tendon (no sacs)	part of the liver	right thoracotomy, reduction and repair	good
Appelquist et al (1)	65 years (adult)	F	multiple defects in right central tendon (no sacs)	part of the liver	right thoracotomy, reduction and repair	good
Kubota et al (9)	10 months	M	two defects in left diaphragm: Anterior and paravertebral (no sacs)	parts of the liver and spleen	laparotomy, reduction and repair	good
Kataria et al (8)	8 years	M	eventration of left diaphragm and anteromedial defect (no sac)	omentum	left thoracotomy, reduction and repair	good
Kataria et al (8)	1 year	M	eventration of left diaphragm and defect in apex of dome (no sac)	small bowel	left thoracotomy, reduction and repair	good
Urushihara (12)	newborn	F	right posterolateral D.H. (no sac). Right anterolateral D.H. (with sac containing pulmonary sequestration)	small bowel and right liver lobe	laparotomy, reduction and repair	good

**Conclusion:**

This case depicts the diagnostic difficulties encountered with late-onset congenital diaphragmatic hernia due to its rarity and vague presentation. It emphasises that congenital diaphragmatic hernia should be considered as one of the differential diagnosis in all age groups when clinical suspicion is raised with such clinical features.

Congenital diaphragmatic hernias that present shortly after birth usually pose few diagnostic problems, although they are associated with several management difficulties. In contrast, the diagnosis of delayed presenting congenital diaphragmatic hernia may be challenging due to its vague presentation. The symptoms and signs are non-specific and may overlap with other pathologies. The risk of misdiagnosing delayed presenting congenital diaphragmatic hernia as pneumothorax or pleural effusion with subsequent insertion of a chest drain has been reported and reflects the significant diagnostic difficulties and emphasizes the fact that late-presenting congenital diaphragmatic hernia

should be suspected in every patient when diaphragm is not clearly visible on radiography.

Late-presenting congenital diaphragmatic hernias should be included in the differential diagnosis of any child with persistent gastrointestinal or respiratory problems associated with abnormal chest X-ray film findings. Although the mortality in these patients is small but the morbidity may be significant. A high index of suspicion is required to avoid delay in appropriate management.

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