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## Treatment of a congenital diaphragmatic hernia with associated wandering spleen: Case report of a 17-year-old girl

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## ABSTRACT

**INTRODUCTION:** A congenital diaphragmatic hernia (CDH) is rarely diagnosed in adults and can allow passage of abdominal viscera into the chest cavity. A particularly rare association is a wandering spleen due to absence of its diaphragmatic and retroperitoneal attachment which predisposes to elongation of the vascular pedicle with risk of torsion, infarction and rupture.

**PRESENTATION OF CASE:** A 17-year-old girl presented with a two-day history of increasing abdominal pain. Examination identified an abdominal mass. Computer tomography (CT) chest, abdomen and pelvis revealed a significantly enlarged wandering spleen with signs of torsion and an associated large left CDH with viscera in the chest cavity. The patient proceeded to an open splenectomy and repair of CDH. Post-operatively the patient developed ileus and required a temporary chest tube for pneumothorax, but otherwise progressed well.

**DISCUSSION:** Untreated CDH with a symptomatic wandering spleen is an extremely rare diagnosis with only one similar previous case report. Clinical detection is unlikely, making CT scanning the diagnostic test of choice. Surgery is recommended given the high morbidity and mortality of associated complications of both conditions. Splenic preserving options are favoured, however the majority of identified cases require splenectomy because of associated torsion or splenomegaly. Reduction of the CDH should be performed with primary closure of the defect and mesh reinforcement where possible.

**CONCLUSION:** CDH with associated wandering spleen in adults presents an extremely rare but clinically important diagnosis. Prompt surgical management as reported in this case should be performed to minimise immediate and future complications.

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## 1. Introduction

Congenital diaphragmatic hernia (CDH) is a developmental defect of the diaphragm which can allow passage of abdominal viscera into the chest cavity [1]. There are multiple types of CDH with the most common being a Bochdalek hernia, comprising 95 per cent [1]. A Bochdalek hernia involves a posterolateral defect of the diaphragm, usually left sided, compared to the less common Morgagni hernia with an anterior diaphragmatic defect [1]. CDH occur in around 1 in 2000 to 1 in 5000 births, where only 5 per cent go unrecognized into adulthood, usually with few respiratory symptoms [2,3]. The diaphragmatic and retroperitoneal attachments of the spleen may be absent in a CDH, resulting in a wandering spleen [4].

An ectopic or wandering spleen occurs due to increased laxity or absence of the ligaments that fix the spleen in the left upper quadrant of the abdomen [5]. These ligaments include the gastrosplenic

ligament which allows attachment to the posterior aspect of the stomach, the splenorenal ligament attaching to the pancreatic tail and left kidney as well as retroperitoneal and diaphragmatic attachments [5]. Splenomegaly or hormonal changes associated with pregnancy can predispose to ligamentous laxity, whilst absence of splenic attachments is associated with congenital causes [6]. This laxity leads to elongation of the splenic vascular pedicle, with potential complications of torsion leading to infarction and splenic rupture, pancreatitis, bowel obstruction, gastric volvulus or gastric and duodenal compression [6]. The incidence of wandering spleen is less than 0.2%, with around 500 cases reported worldwide [6].

We present an extremely rare case of symptomatic wandering spleen with associated CDH [7]. Our experience can contribute to a better understanding of the rare link between these entities and safe surgical management. This article has been reported in line with the SCARE criteria [8].

## 2. Presentation of case

A 17-year-old girl self-presented to the emergency department with a two-day history of abdominal discomfort, distension and

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A: Enlarged wandering spleen.



B: Viscera in left chest from CDH

Fig. 1. A: Enlarged wandering spleen. B: Viscera in left chest from CDH.

vomiting. She reported acute abdominal pain that was constant and made worse with mobilising or lying on the right side. She also reported left sided abdominal distension. The patient's mother reported no issues during pregnancy of the patient and only a brief period of respiratory concerns after delivery, with no delay in discharge home. The patient had no significant family history; no previous medical or surgical history; no regular medications and no allergies.

Upon initial examination, a firm abdominal mass was detected. Blood tests were unremarkable except for thrombocytopenia with a platelet count of  $67 \times 10^9$  /L. Ultrasound demonstrated a large undifferentiated homogenous mass of the mid abdomen. A CT chest, abdomen and pelvis was performed which demonstrated a 19 cm abnormally positioned spleen in the mid-abdomen (Fig. 1A). There were associated dilated vessels at the splenic hilum consistent with post-systemic collaterals or varices and a swirled appearance of the splenic artery, highly suggestive of splenic torsion that may be longstanding. The spleen enhanced heterogeneously, with no discrete areas of infarction. The CT chest demonstrated a very large congenital diaphragmatic hernia containing small bowel, right colon, transverse colon and most of the pancreas. The left lung and left pulmonary artery appeared hypoplastic (Fig. 1B).

Following the CT findings, further questions to the patient revealed that she had never noticed an abdominal mass until two days prior to presentation. There was no prior history of abdominal pain, chest pain or respiratory issues. The patient reported that during schooling she struggled with cardiovascular endurance exercises.

The following day, the patient underwent an open splenectomy and repair of CDH by a hepatobiliary surgeon, with previous experience in CDH repair in young adults. Following left subcostal incision, massive splenomegaly occupying most of the abdominal cavity was encountered. A splenectomy was performed (Fig. 2) by dividing the ectatic splenic pedicle with EndoGIA Tristapler™ (Minneapolis, Minnesota, USA). Next, the abdominal viscera were reduced from the chest revealing the left posterolateral Bochdalek congenital diaphragmatic hernia (Fig. 3). The diaphragmatic hernia was closed using 0 Ethibond and reinforced with Phasix™ mesh



Fig. 2. Enlarged spleen removed.

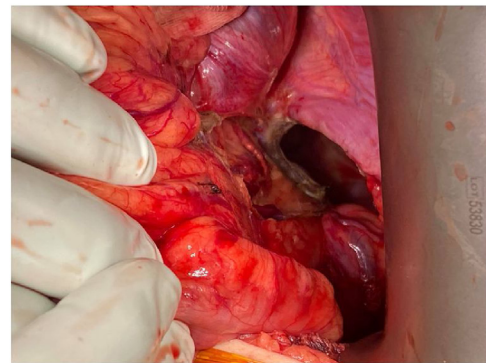


Fig. 3. Intraoperative image of left posterolateral Bochdalek hernia defect.

(Franklin Lakes, NJ, USA) with the use of a foley catheter and Val-salva to remove air from the cavity prior to complete closure. The caecum, base of small bowel mesentery and splenic flexure were fixed to the abdominal cavity with 3-0 PDS to prevent migration. The abdomen was then closed without tension with 0 loop PDS.

Post-operatively the patient was monitored in intensive care. As expected, the patient demonstrated a left pneumothorax on



**Fig. 4.** Chest X-ray prior to discharge showing re-expansion of left lung.

chest x-ray. On day 2 post operatively, the patient developed mild hypoxia and a chest tube was therefore inserted. Bowel movements were slow to progress, but the patient made an otherwise unremarkable recovery with chest tube removed on day 7 and evidence of significant left lung re-expansion on chest x-ray (Fig. 4). Prior to discharge, the patient was registered with Spleen Australia, received the recommended immunisations and was commenced on lifelong amoxicillin.

Histopathology on the spleen reported a weight of 809 g. Microscopic examination showed expanded and congested red pulp but no evidence of lesions or malignancy. The patient was followed up two weeks post-operatively and was recovering well.

### 3. Discussion

Reported cases of wandering spleen with an associated untreated Bochdalek CDH are extremely rare with only one identified published case report [7]. In this case, a 26-year old gentleman presented unwell and was found to have a diaphragmatic hernia with partial bowel obstruction on imaging as well as a palpable suprapubic mass [7]. The patient proceeded to theatre where the mass was identified as a 950-gram torted wandering spleen that required splenectomy. The diaphragmatic hernia, as with our case, was identified as a Bochdalek with a large amount of abdominal viscera herniated and part of the small bowel ischaemic, but viable once reduced. The diaphragmatic defect was closed primarily and reinforced with perinephric fascia.

The majority of CDH are diagnosed in the paediatric population, often with associated pulmonary hypoplasia or other congenital defects leading to severe respiratory distress and a high mortality rate [2,3]. Diaphragmatic development is usually complete by week nine of foetal development. The sealing of the left side of the diaphragm often occurs one week after the right, making left sided CDH more common [1]. Part of the diaphragm forms by fusion of the pleuroperitoneal membrane with the septum transversum, leading to separation of the coelomic cavity into abdominal and thoracic cavities [9]. A persistent pleuroperitoneal defect at 10 weeks of development can lead to portions of the intestines entering the thoracic cavity [1]. Most adults are asymptomatic when diagnosed with CDH, however, symptoms can include abdominal or chest pain, shortness of breath, and vomiting [9]. CT scanning is the gold standard for diagnosing CDH. [10]. Surgical repair is recommended regardless of symptoms; to prevent future complications of strangulation leading to gangrenous viscera in the hernia [1,3].

The approach to CDH repair can be by laparotomy or laparoscopy, with a thoracic approach being less common [3,10]. Laparotomy by a midline or subcostal incision allows better visualisation of the diaphragm and offers the advantage of identifying the position of the viscera once reduced and repairing any malrotation [10]. Thoracotomy allows safer separation of adhesions formed between abdominal viscera and thoracomedial contents, particularly in recurrent hernias [10]. Combined thoracoabdominal approach may be required in particularly challenging cases [10]. Primary closure of the defect with a non-absorbable suture, size permitting, is generally recommended along with mesh reinforcement [3,10].

Pneumothorax is a recognised short-term surgical complication however, there is no consensus within the literature on the recommended preventative strategy. Some surgeons choose to place a prophylactic intercostal chest drain at the time of operation and accept any risks of this procedure. Others implement the approach utilised in our case of evacuating the air from the thorax prior to closure, and reserve the insertion of the drain to patients that develop a significant post-operative pneumothorax [10]. Another potential complication is hernia recurrence which usually occurs in the first two years. A review of adult Bochdalek hernias reported rates of 1.6%, where all initial operations were by open thoracic or thoracoabdominal approach [10,11]. It is recommended that during CDH repairs the spleen should be inspected and a splenopexy performed if retroperitoneal attachment is absent [2].

The diagnosis of wandering spleen is challenging clinically, but with increasing use and availability of CT, is more detectable [6]. When managed conservatively, an asymptomatic wandering spleen is associated with a complication rate of around 65%. This makes operative management by splenopexy or splenectomy the treatment of choice [6]. In our case, a massively enlarged spleen and evidence of torsion necessitated splenectomy. Other factors precluding splenopexy include splenic vessel thrombosis, functional asplenia and suspicion of malignancy [6]. Due to these factors, a splenectomy is required in the majority of cases [6].

### 4. Conclusion

This case report describes an extremely rare clinical presentation of a symptomatic wandering spleen and an untreated Bochdalek CDH in a young adult. The diaphragmatic defect permitted a significant amount of viscera to herniate into the chest and was associated with congenital absence of the retroperitoneal and diaphragmatic splenic attachments of the spleen. The spleen had therefore migrated, torted and enlarged. Both of these conditions are potentially fatal if left untreated and therefore understanding safe surgical approaches is imperative.

### Declaration of Competing Interest

The authors report no declarations of interest.

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### Ethical approval

This article was approved for publication by Barwon Health Research, Ethics, Governance and Integrity Unit.

**Consent**

Written informed consent was obtained from the patient and family for publication of this case report and images.

**Author's contribution**

Jordyn Dangen: conceptualisation, investigation, writing – original draft, writing – review and editing. Steve Lau: conceptualisation, investigation, writing – review and editing, supervision, project administration. Saleh Abbas: conceptualisation, methodology, investigation, writing – review and editing, supervision.

**Registration of research studies**

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