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# Primary adult midgut volvulus mimicking acute appendicitis: A case report and review of the literature



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## ABSTRACT

**INTRODUCTION:** Adult midgut volvulus is a rare surgical disease but remains an important cause of small bowel obstruction. It is most commonly secondary to postoperative adhesions. Primary cases may be due to congenital malformations.

**PRESENTATION OF CASE:** We report the case of an adult primary midgut volvulus in a 35-year-old Chinese male. Based on the clinical presentation and investigations, the patient was presumed to have acute appendicitis. An open appendectomy was performed. During the operation, the appendix appeared normal. Instead a jejunal volvulus was found secondary to extensive adhesions.

**DISCUSSION:** The epidemiology, embryology and etiology of jejunal volvulus are highlighted. Clinical presentation includes acute colicky abdominal pain usually in the peri-umbilical or epigastric regions, with possible signs of small bowel obstruction. Radiologic diagnosis is confirmed with the ultrasonic 'whirlpool' sign or via computed tomography (CT) scan. Early surgical intervention is necessary to avert bowel ischemia in the presence of a closed loop obstruction.

**CONCLUSION:** While rare, adult midgut volvulus has the potential for morbidity and mortality if not diagnosed early with intestinal ischaemia and related sequelae. Clinicians should consider this and if in doubt, perform an early contrast CT scan.

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## 1. Introduction

Midgut volvulus occurs when there is a rotation of small bowel around its mesenteric vascular pedicle [1]. It is more commonly seen in the pediatric population, with approximately 85% of malrotation cases presenting in the first 2 weeks of life, and more than 90% of patients will present by their 1st birthday [2,3]. In adults, midgut volvulus is a rare surgical disease, with reported incidences between 0.00001% to 0.19% [1]. Despite its rarity, midgut volvulus remains an important cause of small bowel obstruction and more importantly it can compromise blood supply to the small intestine resulting in mesenteric ischemia and infarction [4]. As such, it requires a high index of clinical suspicion and a low threshold for operative intervention to prevent or treat the development of intestinal ischemia and/or gangrene, as this scenario carries a high morbidity and mortality rate. Most cases of adult midgut volvulus are secondary to postoperative adhesions. Those without any underlying or identifiable cause can be classified as primary cases, and are even less commonly seen. In this case report, we present a rare case of adult primary midgut volvulus mimicking acute appendicitis.

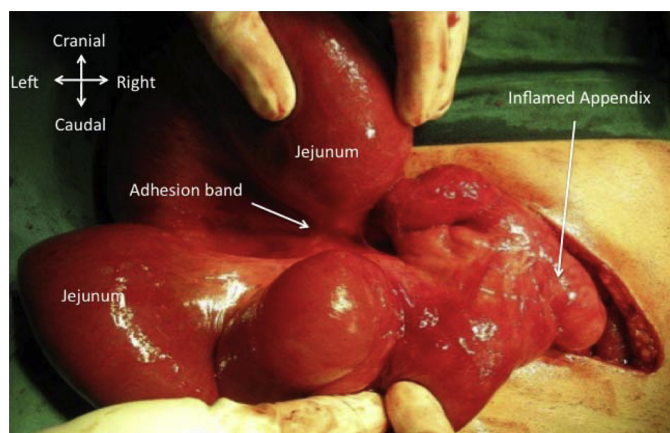
## 2. Presentation of case

A 35-year-old Chinese male presents with 1-day history of intermittent and worsening right iliac fossa pain. He denies any nausea, vomiting, fevers, or diarrhea. Past medical history includes renal stones previously treated in Japan with shock wave lithotripsy a few years ago. He denies any previous surgeries. On examination, he was afebrile, and haemodynamically normal (blood pressure 123/79 mm Hg, heart rate 68/min, oxygen saturation 98% on room air). He was tender maximally in the right iliac fossa with involuntary guarding. There were no palpable masses. Per rectal exam revealed soft brown stools. Initial investigations revealed slightly raised white cell count of 9700/mL<sup>3</sup>. Chest radiograph did not reveal any free air under diaphragm, excluding a possibility of a perforated viscus.

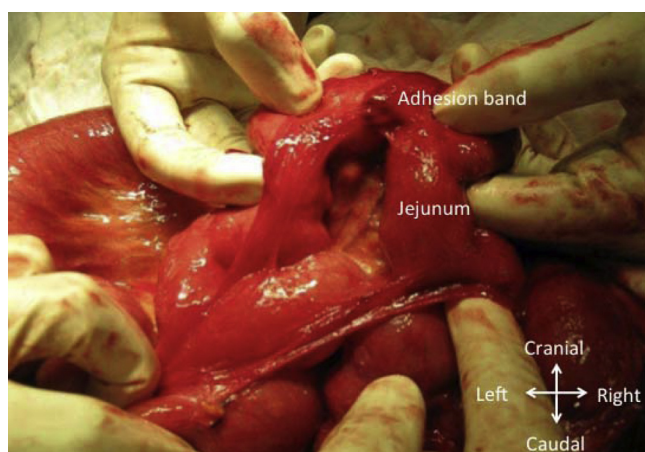
The patient was clinically diagnosed to have acute appendicitis, and appendectomy was promptly arranged. After an incision was made in the McBurney's area, a moderate amount of serous fluid and dilated small bowel loops were seen on entry into the peritoneal cavity. The appendix only appeared mildly inflamed, and upon running the bowel a jejunal volvulus was noted (Figs. 1 and 2) after extension of the Gridiron incision. This appeared secondary to extensive adhesions between the small bowel loops, and extensive adhesiolysis was performed. The jejunal volvulus was reduced

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**Fig. 1.** Intraoperative finding showing jejunal volvulus secondary to adhesion band. Inflamed appendix on the right.



**Fig. 2.** Intraoperative finding showing jejunal volvulus and location of adhesion band.

with no evidence of intestinal ischemia. Routine appendectomy was performed.

Postoperatively, the patient developed ileus but this resolved spontaneously. He made an uneventful recovery and was discharged on 10th postoperative day.

### 3. Discussion

#### 3.1. Epidemiology

The frequency and mode of presentation for midgut volvulus in adults differ by geography. In Western countries, the annual incidence of midgut volvulus has been estimated to be about 1.7–5.7 per 100,000 population [5], and 3–6% of patients present with intestinal obstruction (IO). In Africa and Asia, about 24–60 per 100,000 population suffer from midgut volvulus annually [6], with 20–50% of these presenting with IO [7].

#### 3.2. Embryology

To understand intestinal rotation and its anomalies, we should consider intestinal embryology [8]. The embryo's gut is formed as a straight tube in the fourth week of fetal life. During the fifth week, a vascular pedicle develops. The superior mesenteric vessel supplies the midgut, and is defined by a rapidly enlarging loop with the superior mesenteric artery running out to its apex. The cephalad portion of the loop gives rise to the first 20 feet of small bowel, and

the remainder of the loop forms the distal small bowel and colon up to the splenic flexure. Intestinal rotation primarily involves the midgut. During the development of the intestine, rotation of the intestine occurs in 3 stages. Stage I occurs in weeks 5–10, involving midgut extrusion into the extra-embryonic cavity, a 90° counterclockwise rotation, and return of the midgut into the fetal abdomen. Stage II occurs in week 11 and involves further counterclockwise rotation within the abdominal cavity completing a 270° rotation, bringing the duodenal “c” loop behind the superior mesenteric artery with the ascending colon to the right, the transverse colon above, and descending colon to the left. Stage III involves fusion and anchoring of the mesentery, where the cecum descends, and the ascending and descending colon attach to the retroperitoneum [8,9].

Intestinal anomalies can be categorized by the stage of their occurrence. Stage I anomalies include omphaloceles caused by failure of the gut to return to the abdomen. Stage II anomalies include nonrotation, malrotation, and reversed rotation. Stage III anomalies include an unattached duodenum, mobile cecum, and an unattached small bowel mesentery [9].

#### 3.3. Etiology

Primary midgut volvulus is more common in children and young adults, and cannot be attributed to any predisposing anomaly found intra-operatively. In a previous study of midgut volvulus, the small bowel in high-risk populations were shown to have a corresponding longer mesentery, with deficient mesenteric fat, and has a narrower insertion [10]. A fiber-rich diet has also been implicated, particularly a large amount, with resultant forceful small bowel peristalsis and eventual volvulus [6,10]. In this case report, our patient did not have any of these known predisposing factors, with the midgut volvulus likely secondary to congenital adhesion bands.

The leading secondary cause of midgut volvulus is postoperative adhesions. Other predisposing conditions that could increase the likelihood of small bowel volvulus include a narrow mesenteric root, malrotation, internal hernias, diverticulosis, congenital bands, jejunal lipomatosis, colostomy, fistula, Meckel's diverticulum, calcified mesenteric lymph nodes [11], mesenteric lipomas [12], pregnancy, endometriosis, abscess, mycobacterial disease, aneurysms, and hematomas, and tumors such as mesenteric lymphangiomas or jejunal adenocarcinomas [1,7,9].

#### 3.4. Clinical presentation

Midgut volvulus in adults can present with acute colicky abdominal pain usually in the peri-umbilical or epigastric regions. They may then develop small bowel obstruction and present with nausea, vomiting and/or abdominal distension. In patients without prior history of abdominal surgery or other obvious causes like hernias, there should be a high index of suspicion of midgut volvulus as a cause of the intestinal obstruction [13].

Patients at risk of midgut volvulus such as those with congenital midgut malrotation are usually asymptomatic [14]. Many remain so, with the anomaly discovered only at autopsy or incidentally during CT scans performed for another indication [15]. However, some may have chronic and unexplained abdominal discomfort, and even fewer may report acute episodes of severe abdominal pain [9,13].

#### 3.5. Radiological diagnosis

Diagnosis with plain abdominal radiographs is difficult. Localization of small intestinal loops predominantly in the right side or absence of cecal gas shadow should arouse suspicion of malrotation; this was not present in our patient. Ultrasonography has been

reported to be helpful in diagnosis of intestinal malrotation in the literature, with the classically described ‘whirlpool sign’ [16]. Standard upper gastrointestinal series may show a vertical duodenum not crossing the midline, with the entire small bowel found in the right half of the abdomen. Accuracy of the upper gastrointestinal series is reported to be >80% [17].

The gold standard of diagnosis is a computed topography (CT) scan of the abdomen. Malrotation can be diagnosed with CT with the anatomic location of a right-sided small bowel, a left-sided colon, an abnormal relationship of the superior mesenteric vessels, and aplasia of the uncinate process [15]. The classical ‘whirlpool sign’ is also visible on CT imaging in patients with or without malrotation signifying volvulus [18]. CT imaging has a diagnostic accuracy of >83% [19].

Diagnostic imaging should not be considered in a distressed patient with a rigid abdomen, as further delay will increase the likelihood or worsen any developing bowel ischemia, causing unnecessary morbidity. In our case, we elected to operate on the patient immediately with a clinical diagnosis of appendicitis. To our surprise, a jejunal volvulus was found at operation. A delay in operative intervention by performing a CT imaging might have compromised blood flow to bowel and could have resulted in bowel ischemia necessitating bowel resection.

### 3.6. Management

The classic treatment for incomplete intestinal malrotation is the Ladd’s procedure, which entails division of Ladd’s bands and mobilization of the duodenum, division of adhesions around the SMA to broaden the mesenteric base, placing the small bowel primarily to the right and the large bowel to the left of the midline, and an appendectomy. These principles have remained the same since Ladd’s address to the New Hampshire Medical Society in 1936 [3]. Generally, symptomatic patients with malrotation should have surgical intervention. Spigland et al. recommended that all patients with malrotation are candidates for laparotomy, even if asymptomatic [2]. Mazziotti et al. recently reported a series of malrotation patients managed laparoscopically [20].

In patients with volvulus secondary to other causes, the principles of management include early surgical intervention to decompress bowel, resection of non-viable bowel and addressing the underlying cause, e.g. adhesiolysis for adhesions.

## 4. Conclusion

Adult midgut volvulus is a rare cause of small bowel obstruction but has the potential for morbidity and mortality if not diagnosed early with intestinal ischaemia and related sequelae. As such, clinicians should maintain a reasonable index of suspicion and in diagnostic dilemmas, we recommend performing a contrast CT scan early. Management is primarily surgical. In presence of gangrenous bowel, resection is almost unanimously agreed upon, with or without anastomosis. The underlying cause should be addressed.

### Conflicts of interest

None.

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### Ethical approval

None.

### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

### Author contribution

Study concept/design: all.

Data collection: all.

Writing the paper: all.

Critical revision: all.

### Guarantor

Jeffrey J. Leow.

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