ABSTRACT

Background: Intestinal malrotation refers to the partial or complete failure of rotation of midgut around the superior mesenteric vessels in embryonic life. Arrested midgut rotation results due to narrow-based mesentery and increases the risk of twisting midgut and subsequent obstruction and necrosis. Midgut volvulus secondary to intestinal malrotation is a rare presentation in adults more so in the elderly. Its rarity portends a diagnostic dilemma for both the surgeon and radiologist, and CT scan showed dilated large and small intestine segments with air-fluid levels and twisted mesentry around superior mesenteric artery and vein indicating “whirlpool sign”. Case presentation: In our patient 42 years old male who presented to the emergency department with acute onset of pain in the RUQ, vomiting, and absolute obstipation for the last 2 days. This patient had a history of similar two episodes of sub-acute intestinal obstruction in the last 6 months which has been managed conservatively. The patient underwent an elective exploratory laparotomy after 2 days of failed conservative management. Operative findings included the presence of adhesive bands in the RUQ (Ladd’s Band), volvulus of the mid-portion of the small bowel in a clockwise fashion, and multiple intra-abdominal adhesions and empty RIF and caecum pulled up in RUQ. We subsequently performed a Ladd’s procedure, multiple adhesiolysis, and caecopexy and the patient had an uneventful recovery with eventual discharge on postoperative day 12. Conclusion: This case report reviews the incidence of malrotation in adults. It also highlights the difficulty in diagnosing midgut volvulus in the adult population given the nonspecific, insidious symptoms, therefore, prompting awareness of its existence and a high degree of clinical suspicion. Malrotation should be considered in the differential diagnosis in patients presented with acute abdomen and intestinal ischemia.

1. INTRODUCTION:

Intestinal mal-rotation is a rare congenital anomaly that originates from the failure of the normal rotation and fixation of the midgut during embryologic development<sup>[1]</sup>. It has been estimated that it affects approximately 1 in 500 live births<sup>[2]</sup>. However, the true incidence is difficult to determine as a substantial number of cases will go undetected throughout life.

The vast majority of the complications associated with midgut malrotation present in the first month of life and 60-85% of cases are diagnosed in this age group<sup>[3]</sup>. In adults it is a very rare condition and it is more difficult to recognize due to the lack of symptoms<sup>[4]</sup>. This scenario of incidental diagnosis is becoming increasingly common, particularly with improvements, and increased use, of diagnostic imaging techniques in modern practice.

However, there are a small proportion of affected adults who may present with acute or chronic symptoms of intestinal obstruction or intermittent and recurrent abdominal pain, vomiting, constipation, diarrhoea.

We report this case of an adult patient with an acute presentation of midgut malrotation which highlights the dilemmas of preoperative diagnosis, as this patient had a previous history of recurrent sub-acute intestinal obstruction which has been managed conservatively, and this case supported by a review of the literature.

2. CASE PRESENTATION: The patient is a 42 years old male with low social background with a history of sudden onset, dull aching pain in the right upper quadrant (RUQ) for the last 2 days duration. His pain was associated with nausea, vomiting, and obstipation. He also reported significant abdominal distension. At the time of presentation, he had similar two episodes in the past for which he needs sick medical attention and successfully managed conservatively but his symptoms does not improve 48 hours of treatment.

He had similar two episodes in the past for which he needs sick medical attention and successfully managed conservatively but his symptoms does not improve 48 hours of treatment.

On physical exam. He was afibrile and hemodynamically stable. Per abdominal examination revealed a distended abdomen with diffusely tender with the point of maximal tenderness in the upper abdomen.

There was no palpable organomegaly or intraabdominal masses. Ascites was not clinically demonstrable. Bowel sounds were hypoactive with normal digital rectal examination. Further systemic examination was unremarkable.

His hematological investigations revealed an elevated white blood cell count of 12.5 × 10<sup>9</sup>/L with Hb 10.2 gm/dl. His liver enzymes were unremarkable.

His initial management consisted of intravenous (IV) fluids, IV antibiotics, analgesic, catheterization, and a nasogastric tube. Ultrasonography (USG whole abdomen) showed malrotation with midgut volvulus.
We perform an elective laparotomy under GA after 48 hours of conservative management. We put a mid-line vertical incision and the abdomen is opened. The caecum, appendix, and distal part of the small gut are found in the left hypochondrium with empty RIF. Extensive adhesion was found with ascending colon and small gut. Adhesiolysis has done and distortion of the maltreated loops of the small intestine was performed in the anticlockwise direction. Intra-abdominal adhesions and Ladd’s bands as fibrous stalks of peritoneal tissues attaching caecum to the abdominal wall were dissected. Appendectomy and caecopexy were done. On the postoperative 12th day, he was discharged without any complaints. In the 1st month of the follow-up, he had no complaints, abdominal wound healthy and he was taking a regular diet.

4. HISTOPATHOLOGY: The specimen of an appendix on HPE showed chronic appendicitis.

5. COMPLICATION: Though our patient does not have any complications to date. The complications related to localized midgut volvulus are Ischemia of the gut, short bowel syndrome, perforation, bleeding, wound infection, sepsis, and recurrence.

6. DISCUSSION:
The terminology volvulus evolved from the Latin word “volvere” which means “to roll or twist” [5]. The first description of volvulus was mentioned in papyrus Bears, dated 1550 BC as a “natural course of twisting of a segment of the alimentary tract led to either spontaneous resolution or rotting of the intestines”. Hippocrates was the first to treat volvulus by inserting a large quantity of air through the anus and by inserting a 10 digit (22 cm) long suppository [6]. Intestinal malrotation refers to the partial or complete failure of 360°-counterclockwise rotation of midgut around the superior mesenteric vessels in embryonic life [7]. In our patient 42 years old male admitted with a past history of recurrent episodes of subacute intestinal obstruction. Now he has similar symptoms with failure of conservative treatment and diagnosed as midgut malrotation.

Arrested midgut rotation results due to narrow-based mesentery and increases the risk of twisting midgut and subsequent obstruction and necrosis. Most of the patients with malrotation present within the first month of life and intestinal malrotation in adults is in the ratio of 0.2–0.5% [8]. Adult midgut malrotation is very rare and its incidence has been reported to be between 0.0001% and 0.19% [9].

According to stringer classification which is based on the embryological stage of development, there are three different types of Midgut malrotation (MM) as type I (non-rotation), type II (duodenal mal-rotation), and type III (combined duodenal and caecal malrotation) [10].

This patient developed type III mal-rotation. Intestinal rotation and fixation begin at the 8th gestational weeks with a total of 270 degrees anticlockwise rotation. At the 12th weeks, the intestinal segments were fixed as the ascending and descending colon on the right and left quadrants, respectively [11]. Even though clinical symptoms are obscure, adult patients visit the hospital mostly with complaints such as vomiting and recurrent abdominal pain, obstipation, probably due to chronic partial obstruction [12]. In ultrasonography, the superior mesenteric vein (SMV) lies to the left or anterior to the superior mesenteric artery (SMA). Doppler USG may show the whirlpool sign with the rotation of SMV around SMA which is typical for malrotation [13]. Since mal-rotation commonly causes intestinal obstruction all patients deserve elective laparotomy.

This patient has undergone an elective laparotomy followed by Ladd’s procedure, de-rotation of MM, and a prophylactic appendicectomy is added to prevent the future difficulty of diagnosis of appendicitis [14]. Ladd’s procedure aims to dissect

---

**Figure 1.** USG of whole abdomen.
Figure 1. USG whole abdomen a “whirlpool” appearance (blue arrow) of mesenteric root with mesenteric vessels twisted around it. Also “beak” like termination of the duodenum as it crosses the midline. These findings suggest a midgut volvulus.

X-ray abdomen (Plain) showed the empty right iliac fossa with multiple air-fluid levels and scanty air in the rectum as shown in (Figure-2).

**Figure 2.** On abdominal plain X-ray (erect) - shows empty RIF with multiple air fluid levels (yellow arrow) and large intestinal gut loop in the left hypocondrium (red arrow).

We did a CT scan of the abdomen and pelvis which showed a whirls pool appearance of bowel at mid-abdomen (red arrow) & sagital view (blue arrow, B) demonstrated evidence of small bowel malrotation with volvulus causing a "whirlpool"appearance”.

**Figure 3.** Axial contrast-enhanced CT scan of the abdomen (red arrow, A) & sagital view (blue arrow, B) demonstrated evidence of small bowel malrotation with volvulus. (Figure-3)

**Figure 4.** Intra-operative A) On the right De –torsion of MM (blue arrow) B) On the left side, Division of Ladd’s bands (fibrous bands) red arrow.

---

**2. TREATMENT:**
We perform an elective laparotomy under GA after 48 hours of conservative management. We put a mid-line vertical incision and the abdomen is opened. The caecum, appendix, and distal part of the small gut are found in the left hypochondrium with empty RIF. Extensive adhesion was found with ascending colon and small gut. Adhesiolysis has done and distortion of the maltreated loops of the small intestine was performed in the anticlockwise direction. Intra-abdominal adhesions and Ladd’s bands as fibrous stalks of peritoneal tissues attaching caecum to the abdominal wall were dissected. Appendectomy and caecopexy were done. On the postoperative 12th day, he was discharged without any complaints. In the 1st month of the follow-up, he had no complaints, abdominal wound healthy and he was taking a regular diet.

4. HISTOPATHOLOGY: The specimen of an appendix on HPE showed chronic appendicitis.

5. COMPLICATION: Though our patient does not have any complications to date. The complications related to localized midgut volvulus are Ischemia of the gut, short bowel syndrome, perforation, bleeding, wound infection, sepsis, and recurrence.

6. DISCUSSION:
The terminology volvulus evolved from the Latin word “volvere” which means “to roll or twist” [5]. The first description of volvulus was mentioned in papyrus Bears, dated 1550 BC as a “natural course of twisting of a segment of the alimentary tract led to either spontaneous resolution or rotting of the intestines”. Hippocrates was the first to treat volvulus by inserting a large quantity of air through the anus and by inserting a 10 digit (22 cm) long suppository [6]. Intestinal malrotation refers to the partial or complete failure of 360°-counterclockwise rotation of midgut around the superior mesenteric vessels in embryonic life [7]. In our patient 42 years old male admitted with a past history of recurrent episodes of subacute intestinal obstruction. Now he has similar symptoms with failure of conservative treatment and diagnosed as midgut malrotation.

Arrested midgut rotation results due to narrow-based mesentery and increases the risk of twisting midgut and subsequent obstruction and necrosis. Most of the patients with malrotation present within the first month of life and intestinal malrotation in adults is in the ratio of 0.2–0.5% [8]. Adult midgut malrotation is very rare and its incidence has been reported to be between 0.0001% and 0.19% [9].

According to stringer classification which is based on the embryological stage of development, there are three different types of Midgut malrotation (MM) as type I (non-rotation), type II (duodenal mal-rotation), and type III (combined duodenal and caecal malrotation) [10].

This patient developed type III mal-rotation. Intestinal rotation and fixation begin at the 8th gestational weeks with a total of 270 degrees anticlockwise rotation. At the 12th weeks, the intestinal segments were fixed as the ascending and descending colon on the right and left quadrants, respectively [11]. Even though clinical symptoms are obscure, adult patients visit the hospital mostly with complaints such as vomiting and recurrent abdominal pain, obstipation, probably due to chronic partial obstruction [12]. In ultrasonography, the superior mesenteric vein (SMV) lies to the left or anterior to the superior mesenteric artery (SMA). Doppler USG may show the whirlpool sign with the rotation of SMV around SMA which is typical for malrotation [13]. Since mal-rotation commonly causes intestinal obstruction all patients deserve elective laparotomy.

This patient has undergone an elective laparotomy followed by Ladd’s procedure, de-rotation of MM, and a prophylactic appendicectomy is added to prevent the future difficulty of diagnosis of appendicitis [14]. Ladd’s procedure aims to dissect
the bands that apply pressure to the duodenum and jejunum and relieve the small intestine as in the present case

Surgical management of intestinal malrotation at any age is by the Ladd’s procedure (16). This procedure was first described by William Ladd in 1936 and consists of the following steps (14).

i. Division of Ladd’s bands (fibrous bands) lying over the duodenum to the caecum.

ii. Widening of the narrowed root of the mesentery.

iii. Counterclockwise de-torsioning of the midgut volvulus if present and inspecting the bowel to observe if bowel resection is required.

iv. Appendicectomy if required.

v. Placing the small bowel to the right and fixing the colon to the left.

Most adult patients have a resolution of their symptoms of intestinal malrotation after the Ladd’s procedure (17). Other alternatives procedure such as endoscopic untwisting and laparoscopic management has been used in previous cases in the literature. The laparoscopic approach to Ladd’s procedure is safe and effective and promotes the reduction of pain, ileus, length of hospital stay, and better cosmetic aspect (18). Though our patient does not have any gut ischemia even after 48 hours of conservative management so far. Yet, an acute presentation is commonly associates with extensive bowel necrosis and may lead to massive resection.

7. CONCLUSION: In case of intermittent intestinal obstruction in young adults, the physicians should keep in mind that early and accurate diagnosis of MM with appropriate surgical treatment may save patients from unexpected complications. In acute presentations, CT should be chosen as the primary imaging modality and also high suspicion of midgut malrotation after failed conservative management.

FOOTNOTES: Twitter - SUKANTA SIKDAR (@SUKANTASIKDAR3) Twitter.

8. DECLARATION OF COMPETING INTEREST: The authors declare no conflicts of interest pertinent to this case report.

9. FUNDING: No source of funding to be declared.

10. ETHICAL APPROVAL: No institutional review board is required for the publication of a case report at our institution.

11. PATIENT CONSENT: Written consent was obtained from the patient directly.

REFERENCES:


