



## Case report

## A distinctive case of congenital midgut malrotation with volvulus in an adolescent female managed by Ladd's procedure - A case report

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## ABSTRACT

**Introduction and importance:** Midgut Malrotation (MM) is a potentially fatal and rare congenital anomaly that results from an incomplete rotation of the bowel 270 degrees around the Superior Mesenteric Axis. Newborns are the most affected age group, nevertheless, adult malrotation can manifest, but in a much rarer incidence rate. Clinical awareness must be present when faced with a surgical abdomen in an adolescent patient because this pathology can have several misleading signs and symptoms which may eclipse the true preoperative diagnosis and masquerade as another, and this could result in implementing a different therapeutic approach. Swift clinical informed judgments must be made and acted upon to limit the morbidity and mortality resultant from this pathology.

**Case presentation:** We present the case of a 15-year-old female, who was brought to our Emergency Department (ED) with a 5-day-history of biliary emesis which evolved into obstipation with continuous and excruciating abdominal pain. Computed Tomography (CT) scan indicated gastric and duodenal dilation in addition to a "Whirlpool sign". Exploratory laparotomy was done, and a Ladd's procedure was performed.

**Clinical discussion:** We treated our patient by performing Ladd's procedure and adhesiolysis. Diagnosis is conventionally established pre-/intraoperatively based on radiological imaging and clinical suspicion. Therapeutic methods for this pathology are primarily surgical in nature.

**Conclusion:** Midgut Malrotation is a rare entity, thus, it is crucial to further study this type of clinical presentation and keep it in mind to be able to make an accurate diagnosis to reach the optimal outcome for patients who present with acute surgical abdomen.

## 1. Introduction

Intestinal malrotation is defined as the failure of the fetal intestines to accomplish a full 270 degrees counterclockwise rotation cycle around the superior mesenteric axis by around 10 weeks of intrauterine life [1]. As a result, anatomical and functional aberrations of the small and large bowel will take place. It is remarkably rare to be witnessed, with approximately a 0.2% incidence rate of live deliveries [2]. The mainstream presentation of intestinal malrotation is during the first week of

neonatal life. Nonetheless, the incidence of intestinal malrotation is even rarer in adults, with an approximate incidence rate of 0.2–0.5% [3]. Timely diagnosis and clinical suspicion are paramount in order to diminish the morbidity and mortality of this pathology which may arise from complications such as volvulus, intussusception, and internal herniation [4].

The work has been reported in line with the SCARE criteria and the revised 2020 SCARE guidelines [5].

**Abbreviations:** CT, Computed Tomography; ED, Emergency Department; MM, Midgut Malrotation; IV, Intravenous; IBS, Irritable Bowel Syndrome; PUD, Peptic Ulcer Disease.

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## 2. Presentation of case

### 2.1. Patient information

We demonstrate the case of a 15-year-old Middle Eastern female patient who is a known case of congenital hypothyroidism, who presented to the Emergency Department (ED) with symptoms of acute surgical abdomen. The clinical complaints began 15 days prior to hospital admission when the patient reported a gradual, colicky, intermittent, and periumbilical abdominal pain. She also suffered from nausea, multiple episodes of non-biliary emesis, loss of appetite, and bowel habits alternating between constipation and diarrhea. The pain became more intense 5 days prior to admission and as a result, she was accompanied by her family to the Emergency Department of a rural hospital where she was treated as a case of an Acute Appendicitis by an open Appendectomy. She was discharged home the following day. Symptoms were relieved temporarily postoperatively. However, numerous episodes of biliary emesis began to occur 5 days prior to admission. Additionally, she developed obstipation which was intermittently relieved by the usage of soft enema. 36 h prior to admission, she redeveloped obstipation but this time, it was unresponsive to conservative therapy. On the day of admission, she developed a sudden, continuous, and severe generalized abdominal pain. The pain was not relieved by over-the-counter analgesics. As a result, she was referred to our University Hospital via ambulance with this clinical picture. No fever nor genitourinary symptoms were reported. Her family history involved a sibling who was deceased postpartum due to a congenital heart defect. Her drug history comprised the administration of Thyroxine replacement for her thyroid condition. Her allergic history is insignificant. She is a non-smoker nor a consumer of alcohol. Her BMI is 21 Kg/m<sup>2</sup>.

### 2.2. Clinical findings

Physical examination revealed mild tachycardia and tachypnea. Otherwise, her vital signs were within normal. On inspection, her abdomen was asymmetrically distended, especially above the umbilicus. It moved painfully with respiration. We noted a McBurney's scar incision. Upon palpation, there was generalized guarding and tenderness.

On auscultation, bowel sounds were absent. No masses were palpated. Laboratory tests demonstrated leukocytosis (WBC: 12000/ $\mu$ L) and hypokalemia (Potassium was 2.9 mmol/L), but otherwise, within normal ranges.

### 2.3. Diagnostic assessment

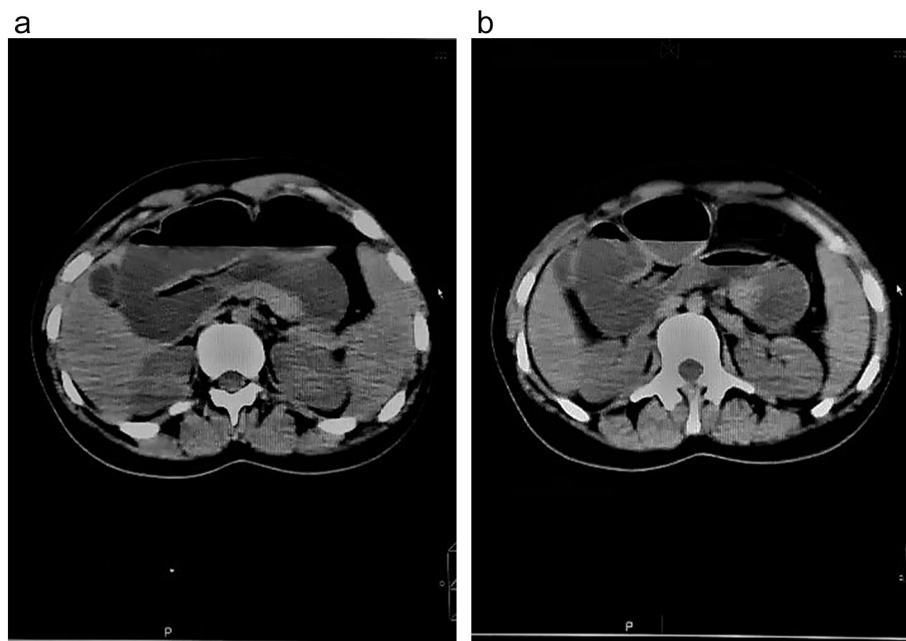
Abdominal Ultrasound demonstrated normal liver, spleen, kidneys, gallbladder, and biliary ducts. No free fluid was present in the abdomen, pelvis, or along the Appendectomy scar. However, moderate dilations of intestinal loops, which were edematous in their walls, was noted. CT imaging of the abdomen and pelvis exposed thickening of gastric and small bowel loops. Furthermore, stenosing at the end of the duodenum was noted (Fig. 1A–B) Additionally, a classical “Whirlpool sign” was shown, which suggested intestinal malrotation (Fig. 2).

Initial management approach involved establishing two large-bore IV cannulas, IV resuscitation, potassium levels adjustment by administering IV potassium chloride, IV analgesics, prophylactic IV antibiotics, and complete laboratory investigations panel including blood sampling and crossmatch.

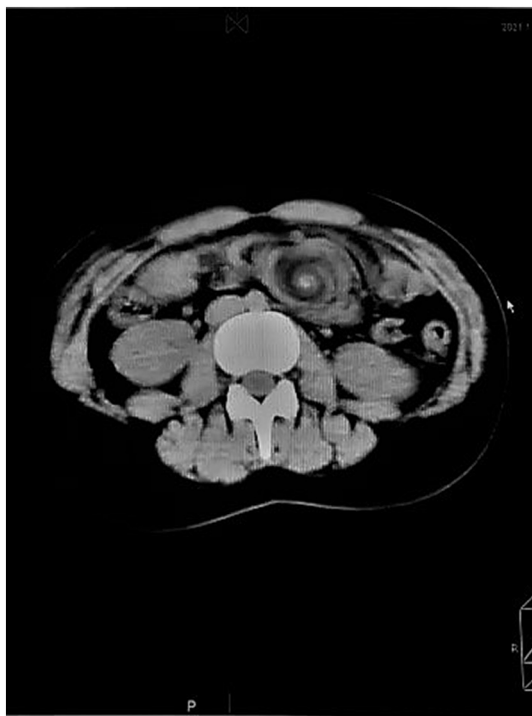
We faced the challenge of the unavailability of a laparoscopic device in the ED at that time.

### 2.4. Therapeutic intervention

An exploratory laparotomy was mandated based upon the given clinical picture. The procedure took place at our tertiary university teaching hospital. It was accomplished by two fifth year senior General Surgery residents with five years of surgical experience and by a General Surgery specialist with 9 years of General Surgery experience. The procedure was carried-out under general anesthesia with no anesthetic complications. Laparotomy had confirmed the radiological findings of the CT imaging. By exploration, dilation of the stomach, first, second, and third parts of the duodenum was noted. Marked stenosing was located at the terminal segment of the duodenum. Furthermore, intestinal malrotation was revealed and Ladd's bands were marked. In addition, vivid adhesions were demonstrated. The procedure involved detorsion of the mal-rotated intestinal loops in an anticlockwise fashion. Additionally, Ladd's bands were dissected, the small intestine was



**Fig. 1.** A-B: Axial view from a CT scan of the abdomen and pelvis revealing dilation of the stomach, 1st, and 2nd parts of the duodenum with air-fluid levels in addition to stenosing at the end of the duodenum, suggesting intestinal obstruction.



**Fig. 2.** Axial view from a CT scan of the abdomen and pelvis revealing the classical “Whirlpool sign” which demarcates the collapsing small bowel loops surrounding the Superior Mesenteric Artery due to malrotation.

folded into the right side of the abdomen, and the colon was placed on the left side (Ladd’s Procedure). Moreover, adhesiolysis was done (Fig. 3A-B-C-D-E).

The patient had successful postoperative recovery and was discharged within a week of the operation. She was informed of multiple lifestyle modifications that will enhance the chances of a full recovery, such as the avoidance of heavy objects lifting, regular sterile wound dressings, and a prescription of vital postoperative prophylactic antibiotics. She was followed-up in the outpatient settings for one month. She has visited the General Surgery clinic regularly to undergo clinical examination and to ensure that a successful uneventful recovery process is taking place. Clinical evaluation involved laboratory investigations, thorough physical examination, and abdominal imaging via ultrasound.

### 3. Discussion

The conventional timeline of the intestinal rotation cycle and its fixation in its normal physiological place commences during the 6th intrauterine week spanning a complete 270 degrees in a counterclockwise manner. When gestational life reaches its 12th week, the ascending colon gets situated in the right quadrant and the descending colon gets situated in the left quadrant whilst the intestines become fixed in their respective location [6]. Based on the gestational developmental status of fetus, the Stringer Classification was set forth in the literature. It states that there are three distinctive types of Midgut Malrotation and they are: Type I (non-rotation), Type II (duodenal malrotation) with subtypes IIa, IIb, and IIc, and finally, Type III (joint cecal and duodenal malrotation) with subtypes IIIa, IIIb, IIIc, IIId [7,8]. Our case was a Subtype IIIa according to the Stringer classification. This type denotes; duodenum is situated right to the midline and the caecum is positioned high, predisposing to volvulus.

Establishing a diagnosis of midgut malrotation in the adult population is genuinely rare. Approximately 90% of such cases get diagnosed during the first year of live births [9] Struggling to diagnose midgut malrotation can be owed to two reasons; the first is the lack of

characteristic clinical findings, whilst the second reason is the diminished adult incidence rate. Adult patient population classically present with a spectrum of non-specific symptoms which correspond to symptoms of other widespread pathologies such as Irritable Bowel Syndrome (IBS), Peptic Ulcer Disease (PUD), pancreatic and biliary diseases, and psychiatric conditions [10].

The chief presenting symptoms of adults agonized by acute malrotation were parallel to those of midgut volvulus and small intestinal obstruction. As the midgut mesentery is truncated, this prompts patients who present with malrotation to suffer from midgut volvulus, mesenteric torsion, and intestinal ischemia. Small intestinal obstruction can be an end-result of an occluded loop of bowel, a strangulation from Ladd’s bands, or intussusception [11]. Diagnosing Midgut Malrotation is chiefly established accidentally intraoperatively, or in postmortem situations -after an autopsy is made- This is due to the non-characteristic symptoms that are reported by patients [11]. There is a diverse scale of non-pathognomonic symptoms inquired from patients who suffer from MM, such as episodes of intermittent ill-defined abdominal pain and episodes of emesis. On the other hand, approximately 10–15% the adult population with MM present with dire episodes of abdominal pain, nausea, bouts of emesis, bleeding per rectum, or hematemesis. This may or may not be accompanied by hemodynamic instability [12,13] MM can manifest chronically with no demarcated symptoms. Furthermore, it is extremely rare. As a result, this can lead to either a misdiagnosis, or even a delay in establishing one. Ultimately, this culminates into a surge in morbidity and mortality [12] Using an imaging modality, specifically CT scanning, in addition to the surgeon’s examination and clinical suspicion of MM, can be considered as the most valuable methods of reaching and establishing a diagnosis in the preoperative state [14].

The pathognomonic radiological sign of intestinal malrotation on imaging modalities such as CT scanning is the classic “Whirlpool sign” which is manifested by the collapsing intestinal loops which mantle the midgut mesenteric vascular structures [12–15,16].

The mainstay treatment modality is via surgery, namely Ladd’s procedure. It is the most prevalently used technique. Next to it, is small intestinal resection. This operation is the gold standard of MM treatment and it is also done on infants with MM [15].

The bull’s eye of Ladd’s operation is to dissect the bands which apply tensile forces to the duodenum and the jejunum and this is to release the small bowel loops [17] -as was done in our case-.

### 4. Conclusion

The clinical manifestations of midgut malrotation varies critically among patients. Symptoms are usually vague and can mimic other pathologies. These obscure and non-pathognomonic findings may result in misdiagnosing the case for another pathology. Moreover, we ought to carefully consider this type of pathology when we are presented with an acute surgical abdomen in an adolescent patient. Although rare, it can occur even in that age group. Keeping an open mind towards the different differential diagnoses will aid us in taking time-efficient clinical judgments and will allow us to perform optimal therapeutic surgical interventions rapidly, so that we can limit the resulting morbidity and mortality.

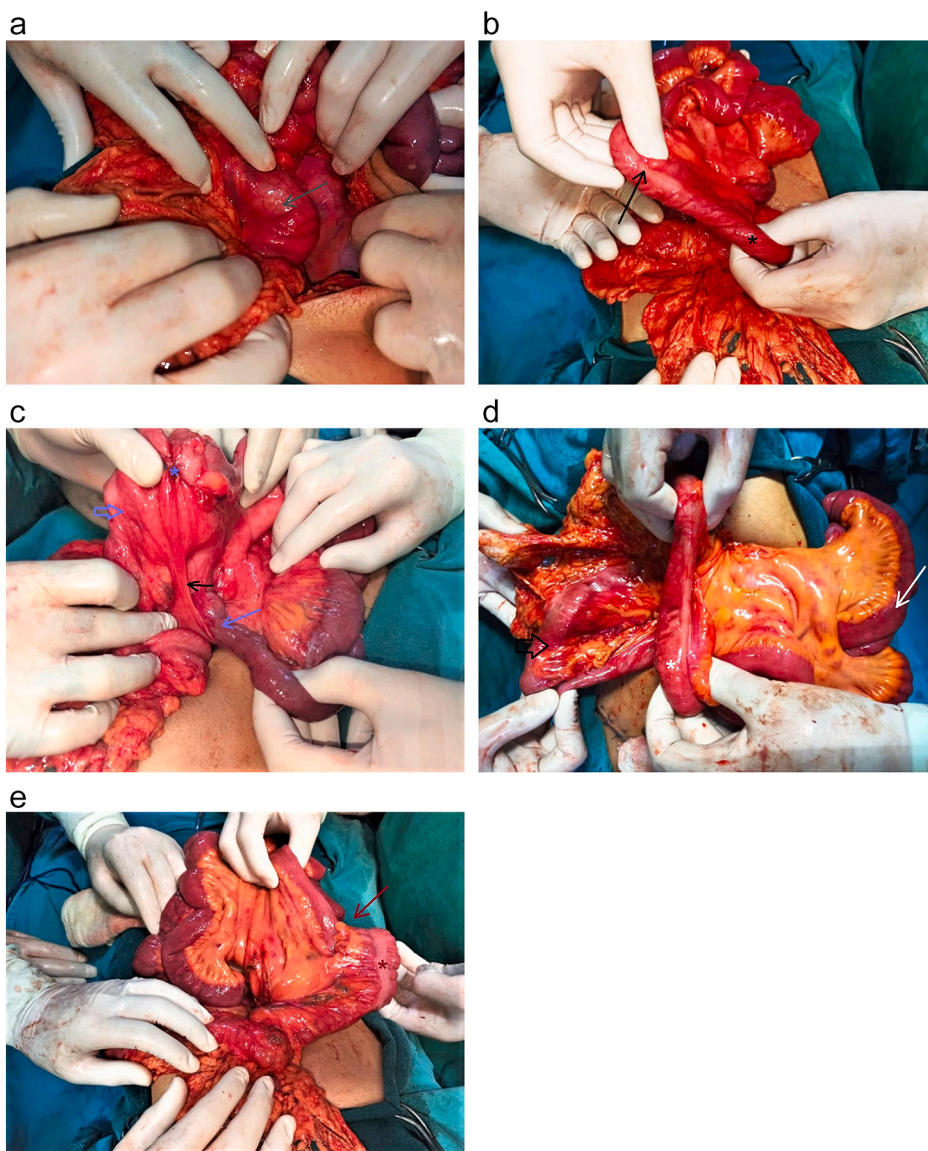
### Ethics approval and consent to participate

Institutional review board approval is not required for deidentified single case reports or histories based on institutional policies.

### Consent of patient

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.





**Fig. 3.** A - Intraoperative Image: Arrow identifies the Duodenum before freeing it from Ladd's bands. We can also note its obvious dilation.

B - Intraoperative Image: Black Arrow identifies the Cecum, and the Star identifies the Ascending Colon.

C - Intraoperative Image: Blue Star identifies the Cecum. Hollow arrow points to the ascending colon. Filled arrow points towards the duodenum. Black Arrow identifies Ladd's Bands; demonstrating how Ladd's Bands span from the Cecum across the Duodenum -causing obstruction and color change indicating ischemia- towards the retroperitoneum.

D - Intraoperative Image: White Star identifies the Cecum and the ileocecal valve. White Arrow identifies the Duodenum. Hollow Black Arrow identifies the Transverse Colon accompanying the Omentum.

E - Intraoperative Image: Red Arrow identifies the Cecum and the position of the ileocecal valve. Red star identifies the Ascending Colon. This photo demonstrates that the Cecum and the Ascending Colon are both free of any attachments. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

#### Availability of data and materials

The datasets generated during and/or analyzed during the current study are not publicly available because the Data were obtained from the hospital computer-based in-house system. Data are available from the corresponding author upon reasonable request.

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#### CRediT authorship contribution statement

OA, RA, IA: who wrote, original drafted, edited, visualized, validated, literature reviewed the manuscript.

FA, JS: supervision, project administration and review of the manuscript.

BA: General Surgery Specialist, who performed and supervised the operation.

OA: conceptualization, resources, and the corresponding author who submitted the paper for publication.

All authors read and approved the final manuscript.

#### Declaration of competing interest

The authors declare that they have no competing interests.

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