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Case Report

Malrotation with Midgut Volvulus Beyond Infancy Requiring an Extensive Ileal Resection: A Case Report

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ABSTRACT

Midgut malrotation is a congenital anomaly resulting from abnormal rotation and fixation of the midgut during embryogenesis. It can be complicated by volvulus and intestinal obstruction. It rarely presents after the first year of life, which makes it a diagnostic challenge. A 5-year-old Arab female presented to the emergency department (July 2024) with repetitive frothy vomiting and diffuse abdominal pain. Laboratory findings showed leukocytosis, metabolic acidosis (pH 7.04, 13 HCO3), and deteriorating renal and liver functions. Abdominal x-ray and ultrasound suggested intestinal obstruction (the presence of distended bowel loops) without a definitive source for the obstruction. An upper gastrointestinal study wasn't done due to continuous vomiting and worsening acidosis. Resuscitation and immediate surgical exploration to identify and treat the source of obstruction were done on the same day. It revealed an intestinal malrotation with distal jejunal volvulus and borderline ischemia of the ileum. First operation included detorsion of the volvulus, excision of the gangrenous segment (a 6cm segment located 6cm proximal to the ileocecal valve) with anastomosis of the remaining gut, and appendectomy. Postoperatively, the patient improved progressively for a week, but then developed an acute abdominal distension with tenderness, necessitating a relaparotomy with a near-total resection of the ileum (150 cm) and stoma formation (double-barreled ileostomy and jejunostomy). Finally, the stoma was closed, and the patient had a jejunoileal anastomosis. The patient recovered well and was discharged on a regular, pureed diet (weight 15.6 kg, passing soft stool twice/day) after 83 days of inpatient observation.

1. Introduction

Midgut malrotation (usually referred to as "malrotation") is a congenital rotation and fixation anomaly of the embryonic intestines, leading to a narrow mesenteric root attachment **(Figure 1)**. Malrotation itself is asymptomatic, but it may then be complicated by bowel obstruction from Ladd bands or midgut volvulus at any age [1]. Midgut volvulus on top of malrotation is a pediatric surgical emergency. The use of ultrasonography (US) in the diagnosis of malrotation is increasing. Still, the fluoroscopic upper gastrointestinal series has historically been the preferred imaging modality for diagnosing both volvulus and midgut malrotation [2]. A Ladd procedure is the ideal surgical management for malrotation with volvulus. It includes detorsion of the volvulus in an anticlockwise direction, division of the Ladd bands, and widening of the mesenteric base. The small bowel is placed on the right abdomen, and the large bowel on the left of the abdomen (nonrotation positioning), and an appendectomy is performed [3]. This manuscript was prepared following the CARE guidelines [4].

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2. Case Presentation

A 5-year-old, 17 Kg, Arab female child was escorted to the emergency department, complaining of persistent non-bilious frothy vomiting that started a day ago, and diffuse abdominal pain. A venous blood gas (VBG) showed a rapidly declining pH from 7.2 to 7.04, 13 HCO3, negative acetone, negative ketones, and negative glucose on urine dipsticks, which ruled out diabetic ketoacidosis (DKA). After a few hours, the patient developed coffee ground vomitus, 50CC in total. The patient was lethargic, and general examination showed tachycardia (194 bpm), tachypnea (40 bpm), hypotension (106/56), cold extremities with a thready pulse, and poor perfusion (prolonged CRT). Abdominal examination showed a generalized tender abdomen with diffuse distension and no hepatosplenomegaly. Renal function tests showed high creatinine levels (78.17 µmol/L) rising rapidly to (94.36 µmol/L), and lactate levels (3 mmol/L). The patient was sent for pediatric surgery assessment. A nasogastric tube was placed, with minimal frothy gastric aspiration. Digital rectal examination showed normal-appearing stool with no blood or mucus. Past medical history and family history didn't include any data of medical importance.

2.1. Radiology

Plain abdominal X-ray showed dilated bowel, and no pneumoperitoneum **(Figure 2)**. Ultrasound showed dilated bowel loops all over the abdomen with mild to moderate ascites without a definitive cause for the obstructive symptomatology. Contrast CT abdomen wasn't done due to deteriorating kidney functions and the child's

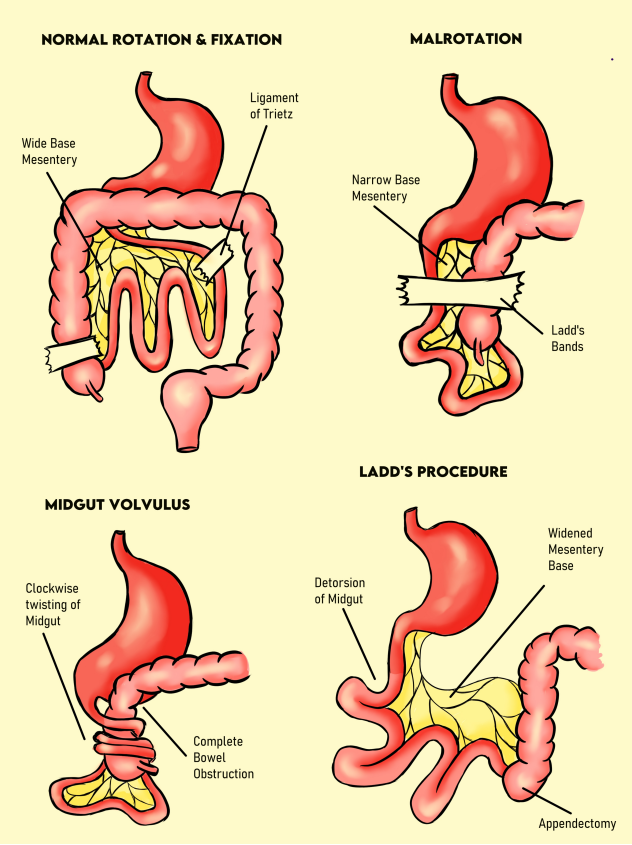


Figure 1: A diagram explaining the process of malrotation, volvulus, and Ladd's operation.

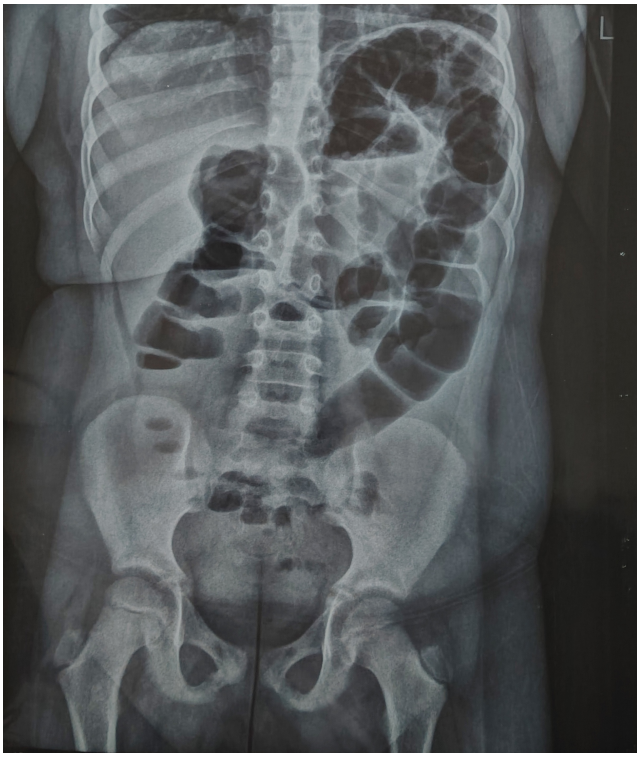


Figure 2: An abdominal plain X-Ray showing dilated bowel loops.

poor general condition. The rising lactate levels, continuous vomiting, and deteriorating kidneys necessitated urgent exploration after resuscitation, and an upper gastrointestinal contrast study (UGI) wasn't done. The patient was taken to the operating room on the same day for urgent surgical exploration due to suspicion of intestinal obstruction of unspecified cause.

2.2. Management

A supra-umbilical transverse incision was done. Peritoneum was filled with bloody serous fluid with no intestinal content; the entire ileum was dusky-to-black, with an intestinal malrotation and distal jejunal volvulus **(Figure 3)**. Reduction of the volvulus was done. 100% oxygen supply was given, and warm saline packs were applied to the dusky ileum for 30 minutes. The color remained dusky, and a 6 cm black gangrenous ileum at a distance of 6cm from the ileocecal valve was excised. An ileoileal anastomosis between the seemingly viable ileum (dusky in color) was performed, appendectomy was done along with lysis of Ladd bands, and a pelvic drain (size 16) was placed. Closure was in layers.

2.3. Post-operative Diagnosis

Malrotation with midgut volvulus

2.4. Post-operatively

Serum creatinine dropped from (52.31 µmol/L) to (37.65 µmol/L) to (30 µmol/L) to (24.45 µmol/L) over the span of 4 days postoperatively and stayed within normal limits till the sixth day. CBC values returned to normal, and vital signs improved. On the 7th day, the patient developed marked diffuse distension accompanied by mild tenderness. The pelvic drain was left in place to monitor leaks from the anastomosis. It was clear, without change in color or amount, and the vital signs showed tachycardia (143/min), tachypnea (52/min), and hypotension (102/51 mmHg). The patient was referred for a relaparotomy due to suspicion of peritonitis secondary to ischemic perforation.

2.5. Re-laparotomy surgery

The previous supra-umbilical transverse incision was used to open. The whole abdomen and pelvis were filled with blood-stained fluid with adhesions all over. The peritoneal adhesions were separated, and all fluid collections were sucked out. Small intestines were mobilized, revealing that most of the ileum had multiple gangrenous patches **(Figure 4)**, with a thinned gut wall. Surprisingly, the previous anastomosis site (at 6cm from the ileocecal valve) from the first look surgery was found to be intact without perforation. Almost the whole of the ileum was removed (150 cm), except for a small portion of the terminal ileum (6cm). This 6cm of terminal ileum was dusky gray during the first surgery, necessitating close follow-up to preserve it. A viable part of the jejunum with the small portion of the terminal ileum was brought out through separate abdominal wall incisions to form a double-barreled ileostomy and jejunostomy. The preservation of the valve was due to its viability and its importance in maintaining stool caliber (avoiding future short-bowel syndrome symptoms).

2.6. Post-operative follow-up

The patient remained on NPO and IV fluids for 2 weeks, during which she weighed 16.5 kg, before starting a pureed diet. After a month postoperatively, a distal loopogram was done to assess the ileostomy for closure. Then, a laparotomy was done to make a jejunoileal stoma resection and a double-layer jejunoileal anastomosis. A week later, a barium meal follow-through was done,

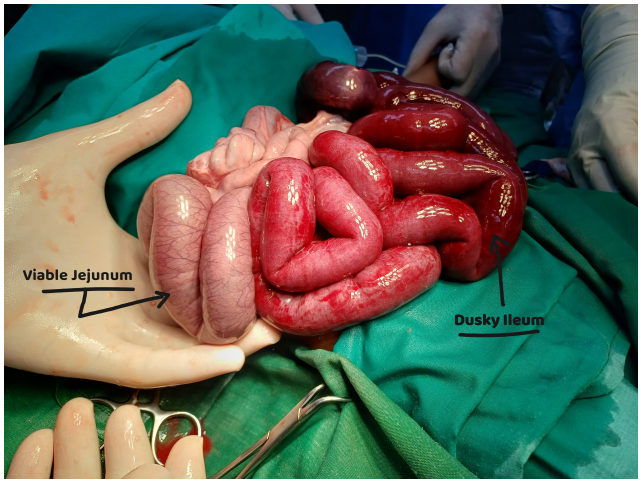


Figure 3: First-look surgery showing dusky ileum.



Figure 4: Second-look surgery showing gangrenous patches all over the ileum.

showing a normal Jejunum and ileal pattern, with adequate transit time. The patient was discharged after 83 days from presentation, on a soft diet, passing semi-loose stools, with no vomiting or dehydration, and weighing 15.6 kg. After 3 months of follow-up, the serum vitamin B12 was 300pg/ml, and stool frequency remained twice to three times per day. A timeline of the patient's presentation and management **(Figure 5)** summarizes the multiple obstacles encountered in treating this patient.

2.7. Pathology report

Small bowel segment measuring 86 cm in length. Externally, the serosa appears green to dusky, congested, and thinned out, with no perforation. The cut section shows gangrenous patches. Five lymph nodes were examined, and no malignant changes were noted.

3. Discussion

In 1897, Franklin Mall was the first to describe the position and development of the human intestine [5]. Between the fourth and eighth weeks of gestation, the small intestines typically revolve counterclockwise around the superior mesenteric artery (SMA) axis. Intestinal malrotation arises when the midgut rotates insufficiently and fixates abnormally during embryogenesis [2, 3].

After the first year of life, malrotation appears to occur less frequently. Thus, the diagnosis of "malrotation beyond infancy", like in our case, is uncommon [6]. Malrotation with volvulus in a neonate presents with upper abdominal distension that goes away with nasogastric tube aspiration and vomiting that is typically green or yellow in color. These are identical to those of duodenal stenosis. Older children present with bilious vomiting, diarrhea, and abdominal pain, which encompasses a much broader differential diagnostic spectrum. This makes the diagnosis of complicated malrotation more challenging [7].

Malrotation appears on ultrasound as an inversion of the SMA/SMV (superior mesenteric artery/ superior mesenteric vein) relationship. Another sign is the presence of a retro-mesenteric D3 segment of the duodenum. On a CT scan, both signs are also detectable [8]. When the diagnosis is difficult, the preferred test is a pediatric upper gastrointestinal contrast study [9]. The choice of diagnostic imaging modality depends on the patient's hemodynamic stability, which wasn't in our favor in this case. Hemodynamic instability may preclude a definitive diagnosis, motivating an early exploration once a bowel obstruction is suspected. The surgery of choice is the Ladd procedure, in which volvulus is reduced by anticlockwise detorsion, Ladd bands are divided, and the mesenteric base is widened. The small bowel is placed on the right abdomen, and the large bowel on the left of the abdomen (nonrotation positioning), and an appendectomy is performed [10, 11]. To accurately determine the degree of necessary resection, bowel viability is evaluated by performing a second-look laparotomy 12 to 24 hours following the initial one. Our case had a relaparotomy after excising the gangrenous segment on day seven post-operatively due to the seemingly reassuring laboratory and clinical state of the child [12]. The second-look surgery wasn't done at 12-24 hrs. because the entire ileum was dusky in the first operation. The medical team decided to give the Ileum more time to re-oxygenate rather than resecting the whole segment immediately. The gradual improvement of the child's state further supported this approach until the acute deterioration on day 7, necessitating a relaparotomy and near-total ileal resection.

The first limitation of this case report is that Cheng et al. [13] presented the initial case demonstrating that the Jejunum can undergo structural and functional adaptive processes in the absence of the ileum in a prepubertal child, following a 4-year follow-up. We can't prove the same finding in our case due to an inadequate follow-up period. However, serum vitamin B12 levels (30 pg/mL) and adequate stool frequency (soft stools twice/thrice a day) at 3-month follow-up were reassuring. It excluded short-bowel syndrome and vitamin B12 deficiency. The patient is now under outpatient clinic follow-up in a different hospital. The second limitation is the underutilization of fluoroscopy in radiology due to the patient's

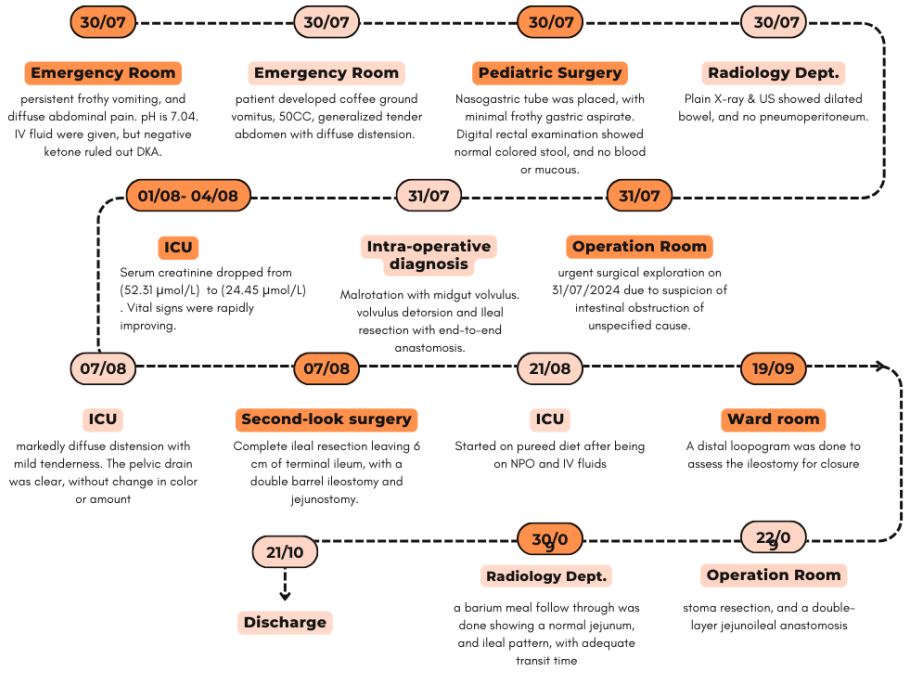


Figure 5: A timeline depicting the management steps of this patient.

deteriorating renal profile and rising lactate levels. However, the ultrasound and x-ray diagnosed an intestinal obstruction, which motivated early surgical exploration.

4. Conclusions

In older children, midgut malrotation complicated with a volvulus is an uncommon but serious pediatric emergency that requires prompt diagnosis and a high index of suspicion. As discussed in this case report, when ultrasound and abdominal x-ray diagnose small bowel obstruction without a clear source, prompt surgical exploration can be lifesaving. The patient's management plan illustrated the diagnostic challenges, addressed significant intestinal ischemia, and outlined post-operative follow-up. When the bowel viability is borderline, it is advisable to resect gangrenous parts and anastomose the rest of the gut, with the intention of having a second-look surgery. This may salvage important parts of the small bowel. Additionally, near-total ileal resection requires close observation and frequent follow-ups to monitor the risk of vitamin B12 deficiency and short-bowel syndrome.

Conflicts of Interest

The authors declare that they have no competing interests

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None

Informed consent

Written informed consent was obtained from the patient's parents/legal guardian for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Large Language Model

None

Authors Contribution

EMA and REA were primarily responsible for the conceptualization, methodology, and execution of the research study. EMA led the data collection and analysis, with critical contributions from REA. EMA also provided significant support during surgical procedures and the interpretation of clinical outcomes, while LEA contributed to the literature review and assisted with data management. REA and LEA participated in drafting and revising the manuscript critically for important intellectual content. EMA supervised all aspects of the work and guided throughout the research process.

Data Availability

No datasets were generated or analyzed for this case report. All relevant clinical information is included in the published article. Additional de-identified data may be made available from the corresponding author on reasonable request, in accordance with institutional policies and patient privacy regulations.

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