

Laparoscopic diagnostic findings in atypical intestinal malrotation in children with equivocal imaging studies

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Background Atypical presentations of intestinal malrotation are more common in older children with a diagnostic and therapeutic challenge. Upper gastrointestinal (UGI) contrast study is essential for the diagnosis of the majority of cases. Recently, laparoscopy has been used in the management of malrotation. We present our experience with laparoscopic management of atypical presentations of intestinal malrotation in children, describing laparoscopic findings in these cases.

Patients and methods A total of 40 patients with atypical presentations of malrotation were included in this study. The main presentations were recurrent abdominal pain, intermittent intestinal obstruction, recurrent bilious vomiting, and failure to thrive. They all were subjected to thorough history taking, clinical examination, routine laboratory investigations, and UGI contrast study. No preoperative definitive diagnosis of malrotation was performed and all patients underwent laparoscopic evaluation.

Results Forty patients (25 males and 15 females) with a mean age of 7 ± 2.8 years were subjected to laparoscopy. Thirty-six patients (90%) were found to have definite laparoscopic findings in the form of markedly dilated stomach and first part of duodenum, ectopic site of cecum, medial and low position of duodenojejunal junction,

congested mesenteric veins with lymphatic ectasia, generalized mesenteric lymphadenopathy, reversed relation of superior mesenteric artery and vein, and right-sided small bowel and narrow mesenteric base. Four patients had differed laparoscopic diagnosis. All the procedures were completed laparoscopically. All the patients achieved full recovery without intraoperative or postoperative complications.

Conclusion Laparoscopy permits direct evaluation and treatment of undocumented malrotation in children, with equivocal UGI contrast study. These newly described laparoscopic findings are the key for the diagnosis of malrotation with atypical presentation. *Ann Pediatr Surg* 11:181–184 © 2015 Annals of Pediatric Surgery.

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Introduction

Intestinal rotation abnormalities occur when normal embryologic intestinal rotation and/or fixation of intestinal mesentery fails to take place. Patients with malrotation have a shortened mesenteric base that makes them susceptible to midgut volvulus, a condition that can result in significant morbidity and mortality. Accurate and prompt diagnosis and timely surgical correction of malrotation are crucial to prevent catastrophic outcomes [1]. Classic malrotation with midgut volvulus is often discovered in a previously healthy term neonate. Up to 75% of the patients present during the first month of life. Another 15% present within the first year. Many other cases will present less dramatically. Failure to thrive, gastroesophageal reflux, early satiety, and mild abdominal discomfort are routinely reported. The diagnosis becomes more challenging with the older child or teenager because the symptoms are often very vague and seemingly unrelated to the abdomen. The role of laparoscopy in the pediatric patient with suspected volvulus or an acute abdomen remains a controversy. However diagnostic laparoscopy with Ladd's procedure has been seen to be effective, even in the context of volvulus [2]. We present our experience with laparoscopic

management of atypical presentations of intestinal malrotation, describing its laparoscopic findings. Some of these laparoscopic findings, to the best of our knowledge, have not been described before.

Patients and methods

This study was conducted at Al-Azhar University Hospitals and affiliated insurance hospitals in the period from May 2007 to October of 2012. It included 40 children presented with symptoms suggestive of intestinal malrotation. They were subjected to clinical examination, laboratory investigation, and upper gastrointestinal (UGI) series. All patients were admitted, hydrated with intravenous fluids, and scheduled for laparoscopic exploration. Single intravenous dose of third generation cephalosporin was given before surgery.

Laparoscopic procedure

The procedure was performed under general endotracheal tube anesthesia with the patient positioned supine. A 5 mm incision was made in the supraumbilical site through which the first 5 mm trocar (for camera) was placed into the abdominal cavity by open Hasson's

technique. The intra-abdominal pressure was maintained from 8 to 12 mmHg according to age. Two additional 3 mm trocars were placed under direct vision in the right and left mid to lower abdominal wall for the working instruments. Exploratory laparoscopy was performed, and the intestinal rotation abnormality was evaluated. Special attention was paid to identify the position of the cecum, duodenojejunal junction (DJJ), and the size of the stomach. The operative table was tilted to different positions during the procedure according to the need. Ladd's bands were identified and divided with laparoscopic adhesiolysis. Care must be taken throughout the procedure, carefully identifying and preserving blood supply without causing torsion. Then the entire bowel is examined, to exclude other obstructive bands or kinks. The base of the mesentery is assessed, and if the length was perceived as less than half of the transverse diameter of the peritoneal cavity, base was widened by incising the peritoneum and gaining length between the vascular arcades of the superior mesenteric artery. The small bowel is then placed on the right of the abdomen and the colon placed on the left.

This study was approved by the research ethical committee at our hospital, with a detailed written informed consent signed by the parents.

Results

Clinical presentation

A total of 40 patients with atypical symptoms of intestinal malrotation were included in this study. They were 25 males and 15 females, with a mean age of 7 ± 2.8 years (range = 2–14 years). The demographic data and laparoscopic diagnostic findings of patients are showed in Table 1. All patients' symptoms were dating since birth. Preoperative UGI contrast study was equivocal (we consider it equivocal if the final report explicitly stated uncertainty or if differential diagnosis was considered). No preoperative definitive diagnosis could be reached.

Laparoscopic findings

Thirty-six patients (90%) were found undergoing laparoscopy having definitive laparoscopic findings in the form of huge dilated stomach and first part of duodenum, ectopic site of cecum central in the abdomen or under the liver (Table 1 and Fig. 1a), generalized mesenteric lymphadenopathy (Fig. 1b), medial and low position of DJJ, Ladd's band (Fig. 2), congested mesenteric veins with lymphatic edema (Fig. 3), reversed relation of superior mesenteric artery and vein, and right-sided small bowel with a narrow mesenteric base. Two patients (5%) were found to have chronic appendicitis with extensive adhesion at the right iliac fossa, one patient (2.5%) was found to have annular pancreas, and one patient (2.5%) had negative laparoscopic exploration. No patients had intestinal ischemia requiring resection. Laparoscopic Ladd's procedure was performed on 36 patients with laparoscopic adhesiolysis (Fig. 2). All procedures completed laparoscopically without conversion. Mean operative time was 50 ± 3.2 min (range = 45–70 min). All patients achieved full recovery without intraoperative or postoperative complications. Oral intake started on the second postoperative day. Hospital-stays ranged from 1 to 3 days (mean = 2 ± 3.2 days).

Table 1 The demographic data and laparoscopic diagnostic findings of patients

Number of patients	40
Sex	
Male	25
Female	15
Age (years)	
Mean	7 ± 2.8
Range	2–14
Presenting symptoms	N (%)
Recurrent abdominal pain	30 (75)
Intermittent intestinal obstruction	36 (90)
Recurrent bilious vomiting	36 (90)
Failure to thrive	40 (100)
Laparoscopic findings	
Ectopic site of cecum	36 (90)
Subhepatic	30 (7)
Central	5 (12.50)
Left sided	1 (2.50)
Hugely dilated stomach	30 (75)
Abnormal position of DJJ	36 (90)
Ladd's bands	30 (75)
Mesenteric lymphadenopathy with congested mesenteric vessels	36 (90)
Chronic appendicitis	2 (5)
Annular pancreas	1 (2.5)
Negative exploration	1 (2.5)

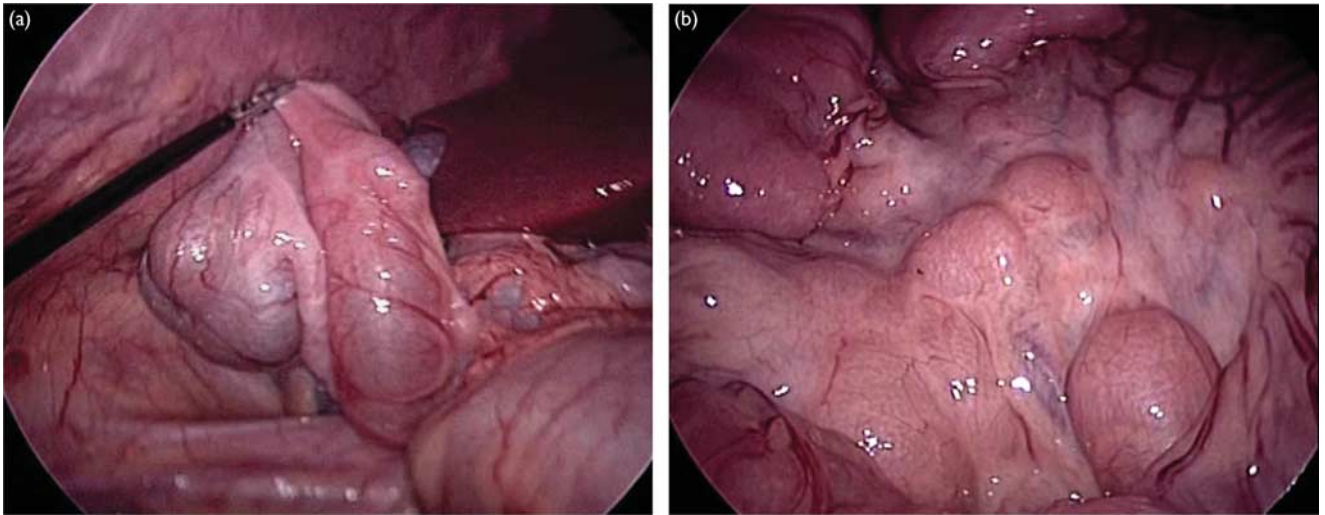
DJJ, duodenojejunal junction.

Discussion

Atypical presentations of intestinal malrotation are more commonly seen in older children. The symptoms are more chronic, intermittent, and result from partial or intermittent duodenal obstruction or a chronic midgut volvulus. These include abdominal pain and vomiting associated with weight loss or failure to thrive, and other nonspecific gastrointestinal complaints. These patients with vague abdominal complaints provide a diagnostic and therapeutic challenge [3]. Maxson and colleagues report that the most frequent complaints of intestinal malrotation are chronic vomiting (68%), intermittent colicky abdominal pain (55%), diarrhea (9%), hematemesis (5%), and constipation (5%) [4]. Partial volvulus leads to mesenteric venous and lymphatic obstruction and subsequently impairs nutrient absorption [2]. Absorption and nutrient transport can be impaired by venous and lymphatic stasis, leading eventually to protein-caloric malnutrition in severe cases of long-standing incomplete rotation with partial obstruction. An increased predisposition to infection has also been observed. Failure to suspect this diagnosis has resulted in dietary manipulation and even psychiatric evaluation in some patients [5]. We noticed that all patients in our series complained of weight loss, failure to thrive with anemia, and hypoproteinemia. Patients with undocumented malrotation may present with acute or chronic abdominal complaints and have a diagnosis that is completely unrelated to a coincident malrotation. Associated symptoms may include intermittent diarrhea, hemochezia, constipation, malabsorption, weight loss, and headache. This group includes patients with common problems who, because of their unusual intestinal anatomy, present with uncommon symptoms. Preoperative evaluation may need to be modified in these patients, both to reach a diagnosis and to adequately define the malrotation [4].

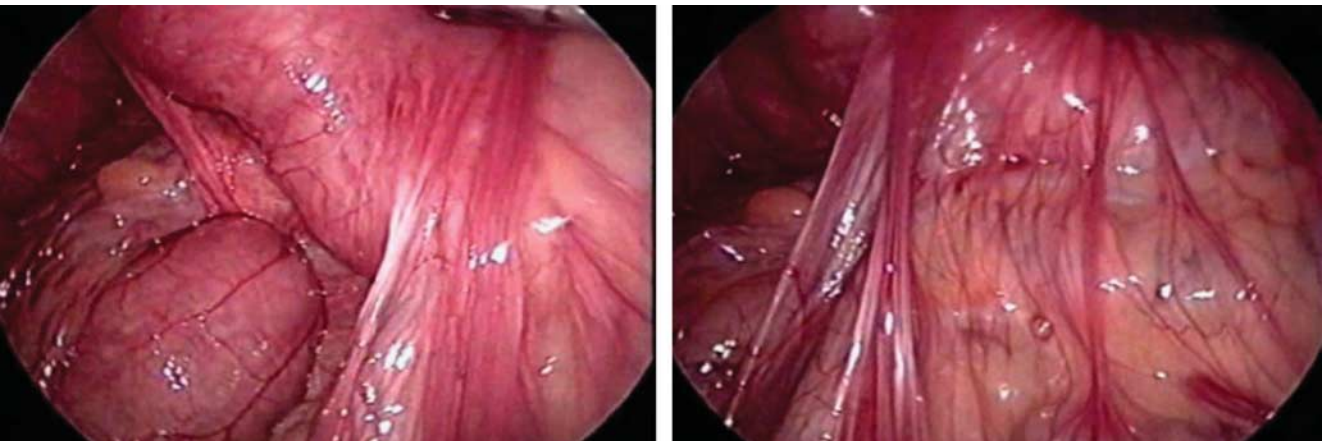
In our patients, the presentation was atypical in all patients; the most common symptom was chronic intermittent

Fig. 1



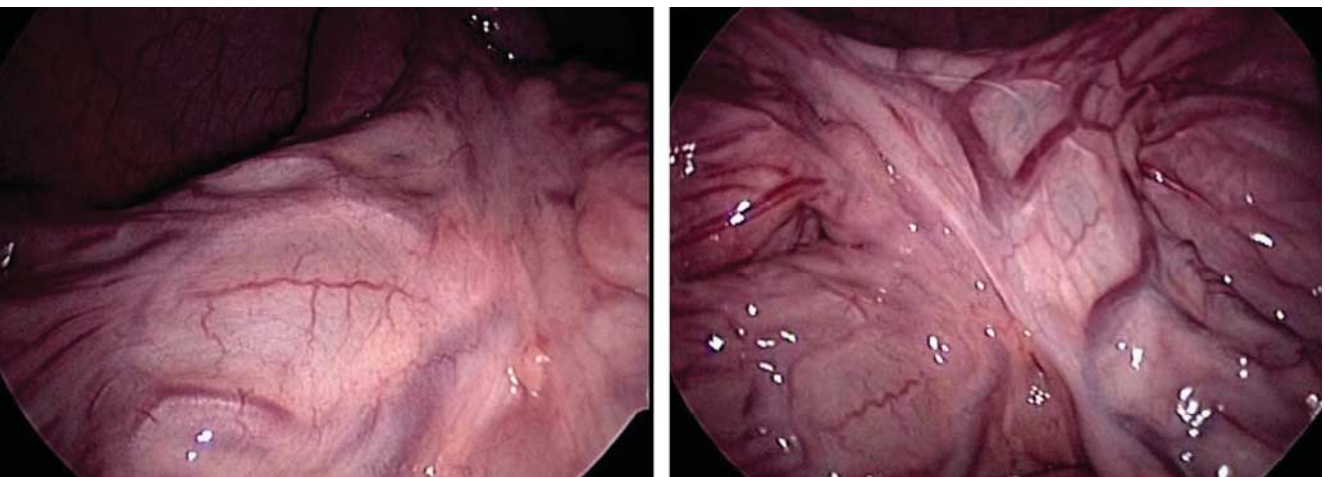
(a) Subhepatic cecum. (b) Mesenteric lymphadenopathy.

Fig. 2



Ladd's bands.

Fig. 3



Congested mesenteric vessels with dilated lymphatics.

abdominal pain, followed by failure to thrive, intermittent upper intestinal obstruction, and episodic vomiting. In all, 85% of patients presented to the outpatient clinic, and all patients had their symptoms dating since birth. Patients presenting this way are more likely to represent diagnostic and management challenges. Despite UGI studies being the standard radiographic modality in diagnosing rotation abnormalities, it may have significant false results. Prasil *et al.* [6] reported a false-negative rate of 6% in their series of 90 patients. Long *et al.* [7] reported that causes of these false results are multiple and attributed to redundant duodenum, bowel distension, and failure to recognize normal duodenal variants. Dilley *et al.* [8] also reported a false-positive rate of 15% in their retrospective series of 72 patients. The DJJ position is an important landmark for UGI studies and is normally located on the left of the vertebral column at the level of the inferior margin of the duodenal bulb [9]. Because the DJJ is mobile in infants and young children secondary to lax peritoneal attachments, many factors can interfere with the DJJ position leading to false-positive and false-negative results [1,10]. In our series, preoperative UGI studies for all patients were equivocal and inconclusive. The only finding was dilated stomach, with equivocal duodenal course, and in some patients the duodenal course is suspected to be a variant of normal. Other important factors that can confuse the issue are the relatively increased mobility of various structures in the infant and the child. For instance, the cecum generally becomes fixed in the right lower quadrant, but in up to 36% of the patients it can be on a mobile mesentery. Also, the laxity in the attachments of the distal duodenum to the retroperitoneum in neonates and in children up to 4 years of age can result in a significant mobility of the DJJ, including mobility to the right of the spine. Others have reported chronic abdominal pain in patients with normally rotated but nonfixed intestines [11]. Recently, there have been a number of small series and case reports describing the use of laparoscopy to diagnose and correct malrotation [12]. Laparoscopy is able to accomplish the identical evaluation and treatment of this group of patients but without the associated morbidity of a laparotomy [13]. In our series, during laparoscopic exploration, 36 patients showed the definite laparoscopic diagnostic findings in the form of huge dilated stomach and first part of duodenum, ectopic site of cecum central in the abdomen or under the liver, medial and low position DJJ, congested mesenteric veins with lymphatic edema, generalized mesenteric lymphadenopathy, reversed relation of superior mesenteric artery and vein, and right-sided small bowel, with a narrow mesenteric base. These laparoscopic findings, to the best of our knowledge, have not been described before during lap management of malrotation. Two patients showed picture of chronic appendicitis with extensive adhesion in the right iliac fossa, and managed with laparoscopic adhesiolysis with appendectomy. One patient with annular pancreas managed with laparoscopic duodenoduodenostomy. The authors notice that during laparoscopic exploration, the DJJ and cecum have lax peritoneal attachments, with relatively increased mobility. The value of laparoscopy is found in these cases of diagnostic confusion when the ligament of Treitz is in an equivocal position on preoperative imaging. In equivocal cases, laparoscopy may be used to determine the position and fixation of the cecum and the overall breadth of the

mesenteric pedicle. The laparoscopic approach allows excellent visualization of the width and fixation of the mesentery and the presence of Ladd's bands. Questionable cases are thus stratified regarding whether operative correction is required. If the mesentery is noted to be narrow, the patient will be prone to volvulus and requires operative correction. The procedure may continue with laparoscopic correction, or the surgeon may wish to convert it to an open procedure at this point [14,15]. Chronic (intermittent or partial) midgut volvulus results in lymphatic and venous obstruction, with mesenteric lymphadenopathy. This situation is more commonly encountered in children older than 2 years. In this study, the authors notice that congested mesenteric veins with lymphatic edema, generalized mesenteric lymphadenopathy was one of the keys of laparoscopic diagnostic finding, and it was more prominent in older children.

Conclusion

The availability of laparoscopy as a less invasive diagnostic and therapeutic modality with these very clear laparoscopic diagnostic finding for malrotation changes the benefit-harm analysis in favor of surgery and offers an alternative approach to routine open Ladd's procedure for these children. We believed that minimally invasive surgical techniques with these described laparoscopic diagnostic findings would be able to accomplish the identical evaluation and treatment of patients with atypical presentation of malrotation.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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