Original Article

Laparoscopic Diagnostic Finding in Atypical Intestinal Malrotation in Children with Equivocal Imaging Studies

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Abstract

Background: Atypical presentation of intestinal malrotation provide a diagnostic and therapeutic dilemma for the surgeon to answer the question is it wisdom to operate or not? Upper gastrointestinal (UGI) contrast study is essential for diagnosis; however, 'soft' radiologic findings place the responsibility of excluding malrotation directly on the surgeon. Recently, minimally invasive surgical techniques would be able to accomplish the identical evaluation and treatment of this group of patients. **Patients and Methods:** A total of 40 patients (25 male, 15 female), age of 2-14 years, presented with symptoms of chronic abdominal pain, intermittent upper intestinal obstruction, recurrent bilious vomiting and failure to thrive. On clinical examination, none of the patients had signs of acute abdominal emergency. UGI contrast study was done and it was equivocal. All patients underwent laparoscopic evaluation. **Results:** A total of 36 patients (90%) were found on laparoscopy to have a discrepant finding of chronic intestinal malrotation. With narrow mesenteric base which put them at significant risk of midgut volvulus. Two patients (5%) were found to have chronic appendicitis with extensive adhesion at the right iliac fossa, one patient (2.5%) has annular pancreas and one patient has negative laparoscopic exploration. Laparoscopic findings of chronic intestinal malrotation includes, huge dilated stomach and the first part of duodenum, ectopic site of caecum central in the abdomen or under the liver, medial and low position of duodenojejunal junction, congested mesenteric veins with lymphatic oedema, generalised mesenteric lymphadenopathy, reversed relation of superior mesenteric artery and vein, right-sided small bowel. **Conclusion:** Laparoscopic diagnostic finding permits direct evaluation and treatment of undocumented malrotation in children, with equivocal UGI contrast study. Furthermore, laparoscopy provides an excellent opportunity to assess the base of the mesentery. Those children with a narrow base should undergo laparoscopic Ladd procedure to avoid catastrophic midgut volvulus.

Keywords: Ladd procedure, laparoscopy, undocumented malrotation

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 patients present during the first month of life. Another 15% will present within the 1st year. Many other cases will present less dramatically. Failure to thrive, gastroesophageal reflux, early satiety and mild abdominal discomfort are routinely reported. Partial volvulus leads to mesenteric venous and lymphatic obstruction and subsequently impairs nutrient absorption. 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 discordant incidence of malrotation between clinical cases and autopsy studies suggests that many patients are asymptomatic but 'anatomically at risk' for midgut volvulus.[2] The differential diagnosis of duodenal obstruction on contrast study includes malrotation with or without midgut volvulus, duodenal stenosis, web or atresia. The diagnosis can be more difficult to establish in a child who has vague, intermittent or chronic symptoms. The

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upper gastrointestinal (UGI) contrast study may demonstrate equivocal findings, with an abnormal duodenal sweep, which may fail to cross the midline or follow a wandering path. In such cases, laparoscopy provides a minimally invasive alternative method of diagnosis and treatment.[3] The role of laparoscopy in the paediatric patient with suspected volvulus or an acute abdomen remains a controversy. However, diagnostic laparoscopy with Ladd's procedure has been shown to be effective, even in the context of volvulus.[4] Our goal was to describe our experience with laparoscopy in the diagnosis of undocumented malrotation, in an attempt to more clearly define its role in this group of patients.

Patients and Methods

From May 2007 to October of 2012, 40 patients (25 male, 15 female), aged from 2 to 14 years, presented with symptoms of chronic abdominal pain, intermittent upper intestinal obstruction, recurrent bilious vomiting and failure to thrive. On clinical examination, none of the patients had signs of acute abdominal emergency. Pre-operative UGI contrast study was equivocal or inconclusive (We consider it equivocal if the final report explicitly stated uncertainty or if differential diagnosis was considered). No pre-operative definitive diagnosis. All patients underwent full clinical examination, admitted, hydrated with intravenous fluids and scheduled for laparoscopic exploration. Single intravenous dose of third generation cephalosporin was given before surgery. The study was approved by the research ethics committee at our hospital.

Results

Clinical presentation

There were 40 patients (25 male, 15 female) aged 2-14 years. Presentation was to the clinic (85%), emergency department (15%), the presenting symptoms as shown in Figure 1, with the most common being chronic abdominal pain, failure to thrive, intermittent upper intestinal obstruction and episodic bilious vomiting. All patients' symptoms were dating since birth. No pre-operative definitive diagnosis, with equivocal or inconclusive UGI contrast study. All patients were haemodynamically stable and had no findings to suggest an acute abdomen at the time of surgery. All patients underwent laparoscopic evaluation. Oral intake was started on the second post-operative day. Hospital stays ranged from 1 to 3 days (average, 2 days). There were no operative or post-operative complications. All patients had resolution of their symptoms, post-operative follow-up ranged from 10 to 12 months.

Laparoscopic ›nding

A total of 36 patients (90%) were found on laparoscopy to have a discrepant finding of chronic intestinal malrotation as shown in Figures 2 and 3, with narrow mesenteric base which put them at significant risk of midgut volvulus. Two patients (5%) were found to have chronic appendicitis with

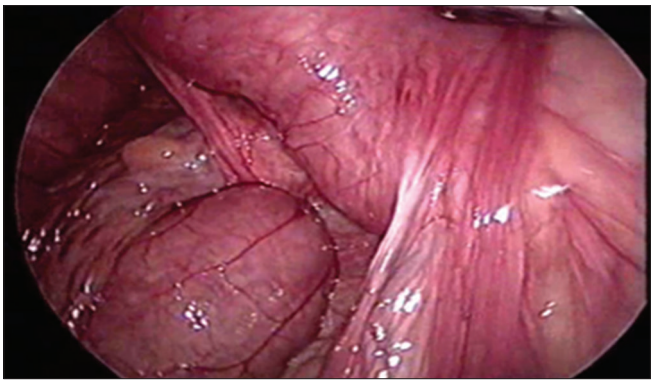

Figure 1: Presenting symptoms

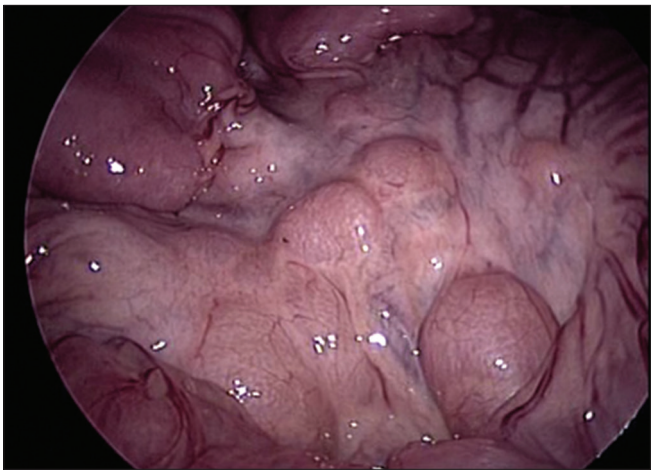

Figure 2: Laparoscopic ›nding

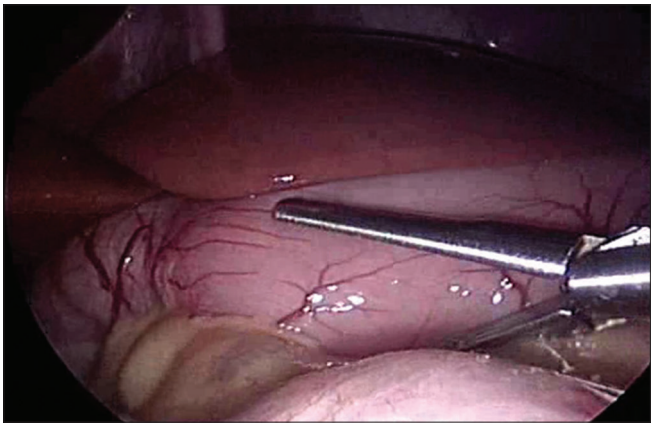

Figure 3: Ladd's bands

extensive adhesion at the right iliac fossa, one patient (2.5%) has annular pancreas and one patient has negative laparoscopic exploration, laparoscopic findings includes huge dilated stomach and the first part of duodenum [Figure 4], ectopic site of caecum central in the abdomen or under the liver [Figure 5], medial and low position of duodenojejunal junction (DJJ), congested mesenteric veins with lymphatic oedema [Figure 6], generalised mesenteric lymphadenopathy [Figure 7], reversed relation of superior mesenteric artery and vein, right-sided

small bowel. No patients had intestinal ischemia requiring resection. Laparoscopic Ladd procedure with appendectomy was done for patients with malrotation. All procedures completed laparoscopically.

Discussion

A typical presentation of intestinal malrotation is 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 non-specific gastrointestinal complaints. These patients with vague abdominal complaints provide a diagnostic and therapeutic dilemma for the surgeon. Because of the risk of midgut volvulus, these patients must be expeditiously evaluated and treated appropriately.[5] Patients with even a question of this diagnosis has historically undergone laparotomy with Ladd procedure if the mesenteric base is narrow and appendectomy if the caecum is in an abnormal position. Although, there have been a number of small series and case reports describing the use of laparoscopy to diagnose and correct malrotation.[6] Minimally invasive surgical techniques would be able to accomplish the identical evaluation and treatment of this group of patients, but without the associated morbidity of a laparotomy.[7] In our patients, the presentation was a typical in all patients, the most common symptom was chronic intermittent abdominal pain, followed by failure to thrive, intermittent upper intestinal obstruction and episodic vomiting. Nearly, 85% of patients presented to the clinic and all patients had their symptoms dating since birth. Patients presenting this way are more likely to represent diagnostic and management challenges. Despite that UGI studies being the standard radiographic modality in diagnosing rotation abnormalities, UGI studies may have significant false results. Prasil *et al.*[8] reported a false-negative rate of 6% in their series of 90 patients. Long *et al.*[9] reported that causes of these false results are multiple and attributed to redundant duodenum, bowel distension and not recognising normal duodenal variants. Dilley *et al.*[10] 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.[11] 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.[1,12] In our series, pre-operative UGI studies for all patients were equivocal or inconclusive and differential diagnosis was considered. 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.

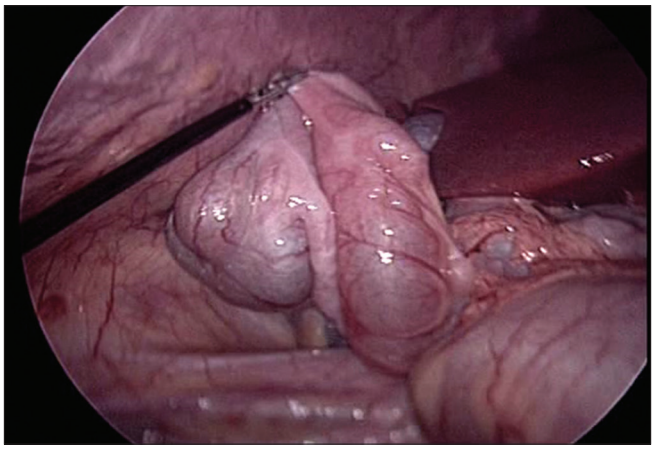

Figure 4: Dilated stomach

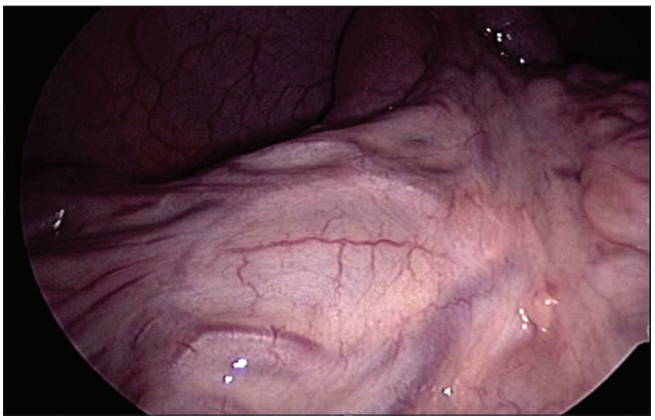

Figure 5: Subhepatic caecum

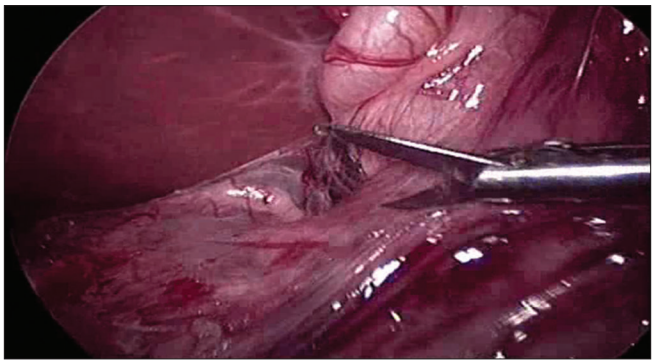

Figure 6: Congested mesenteric vessels with dilated lacteals

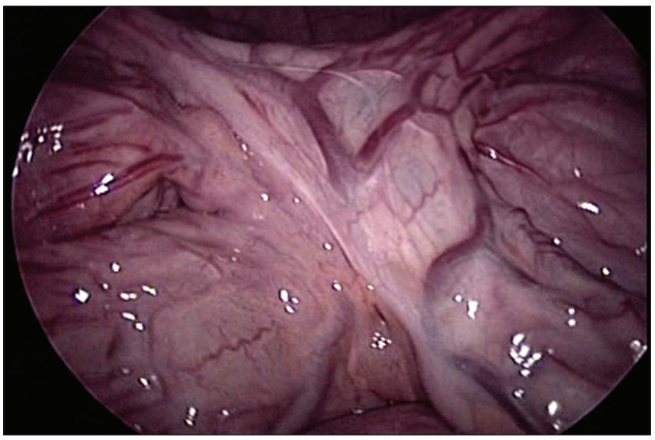

Figure 7: Mesentric lymphadenopathy

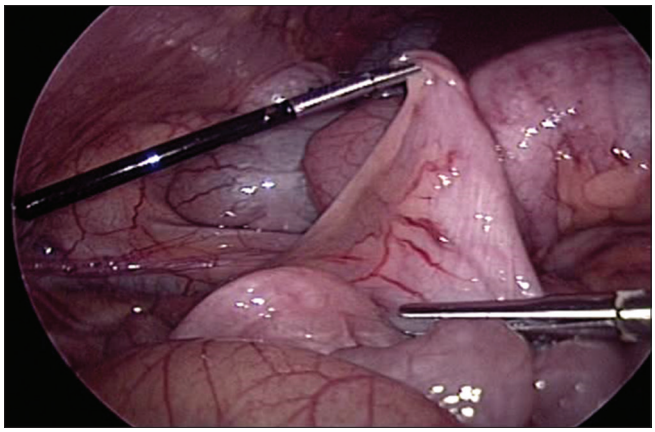

Figure 8: Laparoscopic adhesiolysis

Other important factors that can confuse the issue are the relative increased mobility of various structures in the infant and child. For instance, the caecum generally becomes fixed in the right lower quadrant, but in up to 36% of patients it can be on a mobile mesentery. Furthermore, 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 non-fixed intestines.[13] Thus, because of the laxity of the various retroperitoneal attachments, it can be difficult to distinguish an abnormality at risk for midgut volvulus from an abnormality not at risk using radiologic studies alone. Patients with a mobile caecum and caecal volvulus can present similarly. Patients presenting with chronic abdominal complaints typically describe recurrent, but remitting episodes of nausea, bilious vomiting and crampy abdominal pain. Associated symptoms may include intermittent diarrhoea, haematochezia, constipation, malabsorption, weight loss and headache. Symptoms may have been present since childhood or they may be of more recent onset. This collection of symptoms should be considered a surgical problem rather than a functional or a psychological problem until proved otherwise. Unfortunately, delays in diagnosis are common for most patients.[14] In our series, during the laparoscopic exploration, two patients show a picture of chronic appendicitis with extensive adhesion in the right iliac fossa and managed with laparoscopic adhesiolysis [Figure 8] with appendectomy. One patient with annular pancreases managed with laparoscopic duodenoduodenostomy. A total of 36 patients show the seven laparoscopic diagnostic finding of chronic intestinal malrotation and managed with laparoscopic Ladd procedure without appendectomy. The author notices that during the laparoscopic exploration, the DJJ and caecum has lax peritoneal attachments, with relative 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 pre-operative imaging. Radiographic terms such as 'wandering duodenum', 'low ligament of treitz' and 'abnormal rotation' are frequently encountered in the radiologist's vernacular. Such "soft" radiologic findings place the responsibility of excluding malrotation directly on the surgeon. In equivocal cases, laparoscopy may be used to determine the position and fixation of the caecum and the overall breadth of the mesenteric pedicle. In addition, when performing open exploration based on this pre-operative descriptive, a surgeon may find relatively normal rotation that is not suggestive of malrotation or prone to volvulus. The laparoscopic approach allows excellent visualisation 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 to an open procedure at this point.[3,15] The author notices that 36 patients out of 40 patients, found on laparoscopy to have seven discrepant finding of intestinal malrotation. Our laparoscopic findings includes huge dilated stomach and the first part of the duodenum as results of intermittent obstruction, ectopic site of caecum central in the abdomen or under the liver, medial and low position of DJJ indicating malrotation, congested mesenteric veins with lymphatic oedema, generalised mesenteric lymphadenopathy, reversed relation of superior mesenteric artery and vein and right-sided small bowel. Acute midgut volvulus is a true surgical emergency. Torsion of the narrow mesenteric pedicle produces an acute closed-loop intestinal obstruction and vascular insufficiency. Intestinal ischemia and necrosis may proceed rapidly unless treated promptly. Midgut volvulus is present in up to 50% of patients operated on for malrotation.[1] However, midgut volvulus may be intermittent or incomplete and chronic.[6,15,16] Chronic (intermittent or partial) midgut volvulus results in lymphatic and venous obstruction, with enlargement of the mesenteric lymph nodes. This situation is more commonly encountered in children older than 2 years. The author notices that congested mesenteric veins with lymphatic oedema generalised mesenteric lymphadenopathy was one of key of laparoscopic diagnostic finding and it more prominent in older children. Maxson *et al.* report that the most frequent complaints are chronic vomiting (68%), intermittent colicky abdominal pain (55%), diarrhoea (9%), haematemesis (5%) and constipation (5%). 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.[17] We noticed that, all patients in our series, are complaining of weight loss, failure to thrive and on examination they are underweight with anaemia and hypoproteinema. Bilious vomiting in conjunction with abdominal pain should be considered a surgical problem until proved otherwise. Non-bilious vomiting may also occur as a neurogenic response

to gastroduodenal distension. In our series, all patients presented with recurrent bilious vomiting.

Conclusion

Patients with undocumented malrotation, may present with acute or chronic abdominal complaints and have a diagnosis that is completely unrelated to a coincident malrotation. This group includes patients with common problems who, because of their unusual intestinal anatomy, present with uncommon symptoms. For example, appendicitis that occurs in a patient with a subhepatic caecum will localise to the right upper quadrant and a caecal perforation in a patient with non-rotation will localise to the left lower quadrant. Pre-operative evaluation may need to be modified in these patients, both to reach a diagnosis and to adequately define the malrotation.[14] 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 favour of surgery and offers an alternative approach to routine open Ladd procedure for these children. We believed that minimally invasive surgical techniques with these key diagnostic finding would be able to accomplish the identical evaluation and treatment of this group of patients.

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Confiicts of interest

There are no conflicts of interest.

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