

Surgical Management of Intestinal Malrotation in Adults

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Abstract

Objective The aim of this study was to review our experience with diagnosis and surgical management of intestinal malrotation in adult patients.

Patients and Methods A retrospective review of the surgical outcome of adults with intestinal malrotation was performed. Twelve patients were observed and treated between July 1996 and July 2006 (4 women and 8 men; the mean age of the patients was 28.5 years). Surgical outcomes, including postoperative complications, deaths, and resolution of preoperative symptoms, were measured.

Results A diagnosis of malrotation was made preoperatively in five patients by upper gastrointestinal contrast study, barium enema, or computed tomography scan. The anomaly was discovered incidentally at laparotomy in seven patients. All cases were proved to be malrotation intraoperatively. Nine patients underwent laparotomy and three underwent laparoscopic surgery (one converted to an open procedure). Follow-up ranged from 2 months to 118 months. Three patients had complications: one had wound infection, one had delayed gastric emptying, and one developed adhesive ileus. There were only two recurrences detected and one patient with recurrence required reoperation. No one died.

Conclusions Intestinal malrotation is a rare but important cause of abdominal pain in adults. It may present with chronic or acute symptoms. Laparotomy and laparoscopy are alternative and feasible techniques with low rates of

complications for the treatment of intestinal malrotation in adults.

Intestinal malrotation is an anomaly of fetal intestinal rotation that places the patient at risk of chronic or acute complications [1]. The disease is usually diagnosed in the neonatal period, but in rare cases the diagnosis is not made until adulthood [2–6]. Indeed, most patients are asymptomatic and are discovered incidentally at surgery for other conditions. However, some adults may present chronically with symptoms of intermittent intestinal obstruction or vague abdominal complaints, and others present acutely with small bowel obstruction. The true percentage of patients who will ultimately present with complications attributable to malrotation remains unknown. An upper gastrointestinal (GI) contrast study (UGI) may demonstrate an abnormality of intestinal rotation. Surgical treatment with the Ladd procedure will relieve symptoms and prevent recurrence in most cases. Surgeons may always not consider this possible diagnosis when evaluating adult patients with abdominal pain because of the rare incidence of malrotation in adults [4]. We assessed the evaluation and treatment of a group of adults with intestinal malrotation with the aim to emphasize the significance of this diagnosis.

Patients and Methods

Twelve patients with complications of intestinal malrotation underwent surgical operations between July 1996 and July 2006. Data were collected retrospectively via personal records of the staff surgeons. The mean age of the 12 patients was 28.5 years (range: 22–58 years). Eight of them were men.

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Eligibility was determined by the following criteria: (1) proven intestinal malrotation: location of the duodenojejunal flexure to the right of the midline spinal column with or without location of the cecum in the left abdomen; (2) presence of complications: acutely with bowel obstruction and intestinal ischemia, or chronically with vague abdominal pain, or with atypical symptoms.

Technique of laparotomy

Based on the diagnosis, the patient was asked for informed consent for exploratory laparotomy and the Ladd procedure. The right colon was mobilized and then reflected to the left (Fig. 1). Ladd's bands (Fig. 2) were lysed and the C-loop was straightened. The duodenum was completely mobilized. Adhesions surrounding the superior mesenteric artery were also lysed, and a prophylactic appendectomy was performed.

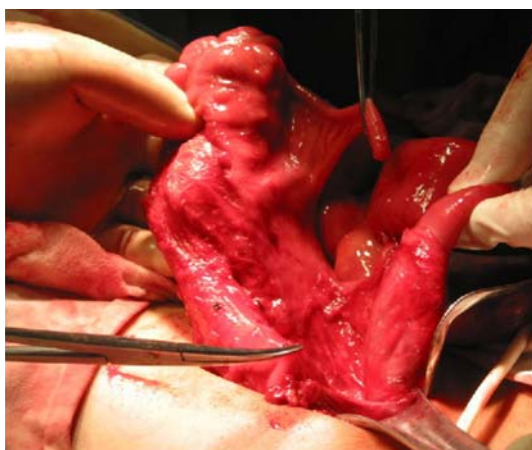


Fig. 1 The right colon was mobilized

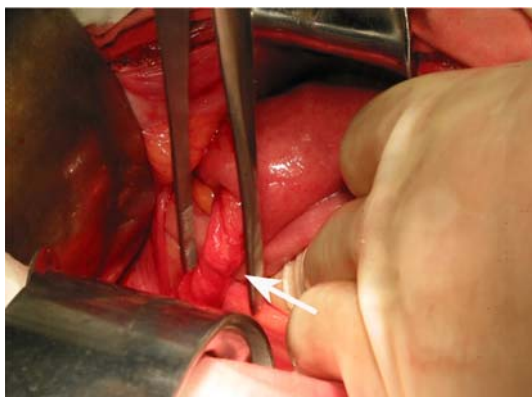


Fig. 2 Ladd's bands (arrow) were identified which extended from the colon to the duodenum

Technique of laparoscopy

Following the induction of general endotracheal anaesthesia, an open insertion technique was performed using a Hasson trocar through a periumbilical incision. The abdomen was insufflated to a pressure of 12–15 mmHg. One 10-mm trocar was placed below the umbilicus, and two 5-mm trocars were placed below the ribs on each side in mid-clavicular line. Exploratory laparoscopy was performed, and the intestinal rotation abnormality was evaluated. Under direct vision, the colon was untwisted completely and positioned in the left upper abdominal quadrant; Ladd's band was divided and broadened. The small intestine was also untwisted and pulled to the anal side. The base of the mesentery was spread out. Finally, appendectomy was performed. The trocars were removed and the fascial defects were closed with absorbable sutures.

Results

Four of the 12 patients presented with acute obstructive symptoms and signs of impending abdominal catastrophe. Six patients had a comparatively long history (from 2 months to 16 years) of intermittent, vague abdominal pain. These patients had been extensively evaluated and treated with several medical regimens without relief. One patient subsequently presented with an acute exacerbation of abdominal pain with the development of peritoneal signs requiring urgent medical attention. At explorative laparotomy malrotation with midgut volvulus was confirmed. The other two patients presented with atypical symptoms. One had right upper quadrant peritonitis, and the other had left lower quadrant peritonitis.

A diagnosis of malrotation was made preoperatively in five patients by upper UGI contrast study, barium enema (BE), or computed tomography (CT) scan. Nine patients underwent UGI (Figs. 3–6). Several of these series had been previously read as normal before referral. Four of these patients were believed to have superior mesenteric artery compression syndrome and their malrotation had not been recognized. Only three of these studies were read as diagnostic of malrotation. Three patients underwent BE, which revealed that the intestines of one patient were in a state of volvulus (Fig. 7), those of another patient were in a state of malrotation, and the other's intestines were normal. In 5 of the nine patients, CT scan was also done, which supported the diagnosis of malrotation in three patients (Fig. 8).

Nine patients underwent laparotomy and 3 underwent laparoscopic surgery (one converted to an open procedure). All cases were proved to be malrotation intraoperatively. Laparotomy or laparoscopic surgery revealed the "classical" form of malrotation, which consists of complete



Fig. 3 Upper gastrointestinal contrast study showing obstruction by Ladd's bands



Fig. 4 Upper gastrointestinal contrast study showing obstruction by Ladd's bands and a right-sided duodenojejunal junction

failure of rotation of both the proximal and distal loops resulting in a duodenojejunal flexure located to the right of the spine, the entire small bowel residing in the right abdomen, and the cecum and colon in the left abdomen, in 8 patients and the “unclassical” form of malrotation, which has less common partial forms of malrotation, in the remaining 4 patients. The anomaly was discovered incidentally at laparotomy in 4 patients. One of them, who presented with right upper quadrant peritonitis, was found to have a subhepatic cecum, and one with peritonitis was found to have perforated appendicitis localized to the left lower quadrant. One of the two patients underwent an appendectomy and a prophylactic Ladd procedure, and the other just underwent an appendectomy because of poor physiological status.



Fig. 5 Upper gastrointestinal contrast study revealing a duodenal-jejunal junction that is located below the level of the duodenal bulb (arrow demonstrating “whorl sign”)



Fig. 6 Upper gastrointestinal contrast study demonstrating nonrotation. Small bowel (SB) is located entirely in the right abdomen, and large bowel is located in the left abdomen, with the cecum (C) in the left lower quadrant

There were no major intraoperative difficulties or cardiopulmonary, cerebrovascular, or thrombotic complications. There was wound infection in a patient. One patient had symptomatic delayed gastric emptying but has since recovered. One patient had an ileus and a single episode of pancreatitis. The mean postoperative hospital stay was 6 (range: 3–13) days. One patient had several recurrent episodes of partial intestinal obstruction, which were believed to be adhesions, and required modification to diet and no reoperation. Another patient who underwent laparoscopic surgery developed a single episode of small bowel obstruction that required laparotomy and lysis of adhesions. Follow-up ranged from 2 months to 118 months.



Fig. 7 Barium enema showing the typical “beak” sign of volvulus

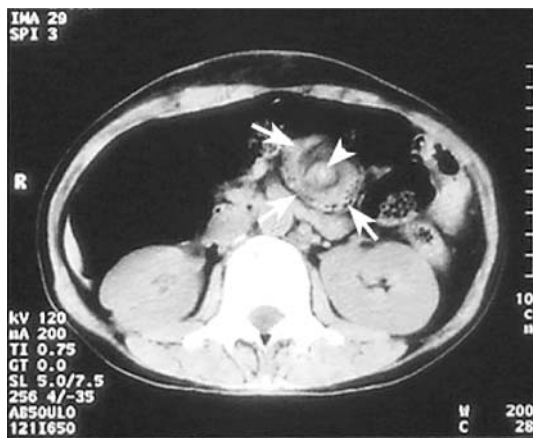


Fig. 8 Computed tomography scan of the upper abdomen demonstrating the “whirling” pattern of jejunal loops (arrow) encircling the mesenteric vessels (arrowhead)

There were no deaths in this series. Patient data are summarized in Table 1.

Discussion

Intestinal malrotation with complications is often seen in infants and children, but few cases have been reported in adults [6–8]. Most often (60%–85%) the condition is diagnosed in the first month of life [6–8]. Forrester et al. examined the epidemiology of malrotation in Hawaii during 1986–1999, finding a rate of 2.86 per 10 000 live births and fetal deaths [9]. Compared with whites, the malrotation rate was significantly higher among Far East Asians, Pacific Islanders, and Filipinos. The reported incidence of malrotation in adulthood is 0.2% [1]. Most adult cases are silent throughout life and are discovered incidentally unless

they cause acute or chronic abdominal pain. Many patients have ill-defined abdominal complaints from childhood [10].

During embryonic development, the midgut herniates into the umbilical sac. At 4 week’s gestation, the embryo’s gut is in the form of a straight tube. During the fifth week, a loop develops with the superior mesenteric artery (SMA) at its axis. The SMA supplies blood to the midgut. Intestinal rotation primarily involves the midgut. The rotation involves three stages. The first stage occurs in weeks 5–10. It is a 90-degree counterclockwise rotation of the midgut, and the bowel returns to the abdominal cavity. The second stage occurs at week 11 and involves a further 270-degree counterclockwise rotation within the abdominal cavity so that the duodenal “C” loop and small bowel become attached to the posterior abdominal wall with the ascending colon to the right, the transverse colon above, and descending colon to the left. In the third stage, the cecum descends, and the ascending and descending colon attach to the posterior abdomen, with fusion and anchoring of the mesentery [2, 4]. Disturbances during any of the individual stages result in anomalies of rotation and abnormalities of mesenteric fixation. Intestinal malrotation can cause shortness of the mesenteric root and narrowness of the resulting suspensory pedicle of the gut. The shortened mesentery allows the small bowel to twist around the narrowed pedicle of the superior mesenteric artery, which places patient at high risk of volvulus of the midgut and intestinal ischemia. Even though there is no volvulus, congenital adhesive bands (Ladd’s bands) may compress the duodenum and cause chronic obstruction [11]. The spectrum of abnormalities can be classified into various subtypes based on the stage of midgut. But for practical purposes, it is useful to simplify categorization into non-rotation and incomplete rotation. “Nonrotation” is the “classic” form of malrotation, it represents most cases identified in adults [12].

Unlike the pediatric population, most adult patients with malrotation lack other congenital anomalies and are free of symptoms related to their malrotated bowel [11]. The few adults who do have symptoms typically present with a prolonged history of abdominal complaints suggestive of obstruction. Symptoms in the adult patient are often mistaken for irritable bowel syndrome, peptic ulcer disease, biliary and pancreatic disease, and psychiatric disorders. In general, there are three distinct patterns of presentation of malrotation in adults [13].

The first group of patients will present with acute obstructive symptoms and signs of impending abdominal catastrophe. On careful questioning some of these patients may also describe a prior history of intermittent, but milder symptomatology. In our series, four patients presented in this manner. The second group will present with chronic

Table 1 Summary of clinical history, diagnostic procedures, operative details, and outcomes in the twelve patients with intestinal malrotation

Patient no.	Gender	Age	Symptoms	Length of history	Diagnostic procedures	Surgical findings	Treatment procedures	Postoperative length of stay	Complications	Recurrence
1	F	35	Abdominal pain	2	months	UGI, CT, BE	Nonrotation	Ladd procedure	(Laparoscopic surgery)	7
2	M	29	Epigastric pain, nausea, vomiting, upper gastrointestinal bleeding	10 days	UGI	Incomplete rotation	Ladd procedure (laparoscopic surgery, converted to open surgery)	5		No
3	M	33	Epigastric pain, tender, weight loss, etc.	16 years	UGI, CT	Nonrotation	Ladd procedure	9	Ileus and pancreatitis	No
4	M	24	Hypochondriac and lower abdominal pain, weight loss	3 days	UGI	Nonrotation	Ladd procedure (laparoscopic surgery)	5		Yes
5	F	26	Upper abdominal pain, weight loss	1 year	UGI, CT	Nonrotation	Ladd procedure	11	Symptomatic delayed gastric emptying	No
6	F	22	Abdominal pain, vomiting	1 week	UGI, CT	Nonrotation	Ladd procedure	3		No
7	M	58	Abdominal pain	3 days		A perforated appendix at the left lower quadrant, and nonrotation	Appendectomy	13	Wound infection	No
8	M	23	Abdominal pain, distention	3 years	UGI, CT, BE	Nonrotation	Ladd procedure	3		No
9	M	23	Abdominal pain, nausea and vomiting	6	months	UGI, BE	Incomplete rotation	Ladd procedure	4	
10	M	22	Crampy abdominal pain, vomiting, and diarrhea	5 days	UGI	Nonrotation	Ladd procedure	3		Yes
11	M	22	Chronic abdominal pain	12 years	UGI	Incomplete rotation	Ladd procedure	5		No
12	F	25	Right upper abdominal pain, nausea, vomiting	10 h		A subhepatic inflamed appendix, and incomplete rotation	Ladd procedure	4		No

UGI upper gastrointestinal contrast study with small-bowel follow-through; BE barium enema

symptoms of bowel obstruction characterized by a history of recurring but remitting episodes of nausea, bilious vomiting, early satiety, and crampy abdominal pain. Associated symptoms may include intermittent diarrhea, hematochezia, constipation, malabsorption, weight loss, and headache. In our series, six patients presented in this manner. The third group will present with atypical symptoms from a common abdominal disease. These patients with common problems, who repeatedly have a diagnosis that is completely unrelated to an accordant malrotation, present with uncommon symptoms because of their unusual intestinal anatomy. In our series, two patients presented in this manner. One with right upper quadrant peritonitis was found to have appendicitis, and the appendix was located under the liver, and the other with perforated appendicitis was found to have a cecum localized to the left lower quadrant.

Plain abdominal radiographs are neither sensitive nor specific. The gold standard for diagnosis of malrotation is UGI combined with BE [13]. Upper gastrointestinal contrast study often reveals a duodenal-jejunal junction located to the right of or overlying the spine and below the level of the duodenal bulb (Figs. 3–5). In the case of midgut volvulus, a corkscrew taping of the duodenum or jejunum will be found (Fig. 5). Barium enema has been used to identify the position of the cecum, but the results have been reported as normal in 40% of patients with confirmed malrotation [14]. The BE may show malposition of the right colon and cecum and may reveal a beaklike stenosis of the intestine in patients with midgut volvulus (Fig. 7) [15]. A normal UGI does not exclude the possibility of malrotation; abnormalities of rotation of the cecocolic loop may occur in the face of normal rotation of the duodenojejunal loop. Similarly, a normal BE does not always reveal the converse condition. Therefore UGI and BE should be performed sympathetically.

Computed tomography (CT) is used increasingly today. Malrotation can be diagnosed on CT by a right-sided small bowel, a left-sided cecum, an inverse relationship between SMA and the superior mesenteric vein (SMV), and aplasia of the uncinate process [12]. The inverse positions of SMV in the setting of malrotation was first described by Nichols and Li. It refers to the superior mesenteric vein lying to the left of the SMA instead of to the right [16]. A distinctive whirlpool-like pattern on CT due to encircling of the loops of the bowel around the SMA was first described by Fisher in a patient with midgut volvulus [17]. Intestinal malrotation can cause shortness of the mesenteric root and narrowness of the resulting suspensory pedicle of the gut. The shortened mesentery allows the small bowel and mesentery to twist around the narrowed pedicle of SMA with midgut volvulus, which may be seen on CT as the so-called whirlpool sign (Fig. 8). Although most patients with malrotation will not have all of the character-

istics described here, identification of any one abnormality warrants closer scrutiny and consideration of other diagnostic modalities [13]. In our series, five patients underwent CT scans. Two of them were false-negatives.

Ultrasonography has also been reported as a diagnostic tool in intestinal malrotation. Characteristic signs of midgut volvulus include duodenal dilation with distal tapering, fixed midline bowel, the whirlpool sign [18], and dilation of the SMV. It also allows an accurate estimate of the number of twists the mesentery has undergone, which may be helpful when the surgeon attempts to reduce them at operation [19]. In addition, the relative positions of the SMA and SMV can indicate malrotation even in the absence of volvulus. Because abdominal ultrasonography and CT can readily demonstrate the relative positions of the SMA and SMV, they are valuable tools for diagnosing midgut malrotation and volvulus.

The classical treatment for intestinal malrotation is the Ladd procedure, which entails counterclockwise detorsion of the midgut volvulus (if present), division of the abnormal coloduodenal Ladd's bands tethering the midgut and causing extrinsic compression, mobilization of the duodenum, division of adhesions around the SMA to broaden the mesenteric base to prevent further volvulus, and an appendectomy [20]. For our patients, the Ladd procedure was highly effective in that it led to complete resolution of symptoms for 9 of the 11 patients available for follow-up and near-complete resolution for the remaining 2 patients.

The question of whether asymptomatic patients with a documented malrotation require surgery is still controversial. Some authors have advocated operative intervention only in patients who are symptomatic. Choi et al. [21] reviewed 177 patients over a 35-year period and found that asymptomatic patients had a low risk of intestinal volvulus and advised that routine investigation, screening, and elective surgery were not necessary with close follow-up.

In contrast, other authors have argued that any risk of midgut volvulus, no matter how small, warrants operative intervention. Many authors recommend an elective Ladd procedure in all patients with intestinal malrotation [22, 23]. Clearly, the potential for volvulus is lifelong and its sequelae do not alter with age. Additionally, not all asymptomatic patients are truly without symptoms.

Not all patients are correctly diagnosed before operation. Occasionally, malrotation is found coincidentally at laparotomy. In our series, the anomaly was discovered incidentally at laparotomy in 4 patients. For instance, 2 patients with allotropic appendicitis were found to have intestinal malrotations. Appendectomy and prophylactic Ladd procedure were applied in one patient and an appendectomy was applied in the other. The patients were discharged, respectively, at day 10 and day 6 postoperatively. Both of them were asymptomatic during follow-up. We believe intestinal

malrotation should be reduced as far as possible, because no reliable means exists to predict which patients will later experience complications [11]. Of course, policy should be based on the primary diagnosis. Operative intervention for underlying malrotation should be undertaken only if it does not add undue risk to the procedure.

Recently, laparoscopic diagnosis and treatment for intestinal malrotation in both infants and adults have been described [11, 24]. It has been confirmed that the laparoscopic Ladd procedure was safe, feasible, and effective. Conversion to an open procedure is common because of the difficulties encountered [24]. In our series, three patients underwent laparoscopic surgery, one of them was converted to an open procedure because of the difficulty assessing the anatomy. This patient was the first one chosen to undergo a laparoscopy for this indication, and the surgeon's learning curve at that time may have reduced the chance of success. In laparoscopy, any incomplete procedure, such as residual mesenteric torsion or remaining Ladd's bands, should be avoided. Otherwise, a delayed laparotomy may be required.

Several literature reports have concluded that midgut volvulus with malrotation can be managed laparoscopically [11, 24, 25]. Nevertheless, we do not think laparoscopy is suitable for patients with midgut volvulus. The procedure may be difficult because of intestinal distension. Moreover, grasping and pulling the edematous or necrotic segments with forceps may cause damage to the intestine.

Conclusion

Intestinal malrotation is an uncommon yet important cause of acute or chronic abdominal pain in adults. Its etiology is relative to disturbances of any individual stages during embryonic development. Patients with malrotations will present with acute obstructive symptoms, with chronic abdominal complaints, or with atypical symptoms. The gold standard for diagnostic study is UGI combined with BE. Malrotation may also be diagnosed by CT scan or ultrasonography. The classic treatment for intestinal malrotation is the Ladd procedure, whether via an open procedure or laparoscopically. Our satisfying results stress the importance of a high index of suspicion for malrotation, appropriate diagnostic studies, quick recognition of the disease process, and timely and aggressive definitive treatment.

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