Adult Intestinal Malrotation: A Case Report
B Papaziogas, A Patsas, G Paraskevas, P Tsiaousis, I Koutelidakis, C Christoforakis, K Atmatzidis

Citation

Abstract
Midgut malrotation is an anomaly of intestinal rotation which occurs during fetal development and usually presents in the neonatal period. It is rare for malrotation to present in adulthood. We present a case of malrotation in an adult female patient who presented with cramping generalized right abdominal pain and vomiting of one day duration. A computed tomography abdominal scan and upper gastrointestinal contrast studies showed malrotation of the small bowel. The patient was consented for exploratory laparotomy during which typical Ladd's bands and a distended flabby third and fourth duodenal portion extrinsically obstructing the misplaced duodeno-jejunal junction were recognized. Detorsion of the twisted mesentery, lysis of the bands, appendectomy and a side-to-side duodeno-jejunal anastomosis were performed. Complete resolution of symptoms is noted in a two-year follow-up period.

INTRODUCTION
Midgut malrotation is an anomaly of fetal intestinal rotation and fixation that usually presents within the first month of life [1]. It is rare for malrotation to present in adulthood [2]. Indeed, most adult patients are asymptomatic and incidentally are discovered later in life at surgery for other conditions [3,4]. However, some adults may present acutely with bowel obstruction and intestinal ischemia due to midgut or cecal volvulus, or chronically with symptoms of intermittent bowel obstruction or vague abdominal complaints. Because presentation is nonspecific and the index of suspicion for malrotation progressively decreases in the older population, the clinical diagnosis is usually not considered in the initial evaluation. Findings diagnostic of malrotation are described using several modalities such as barium studies, computed tomography (CT) scans and angiography [1]. Treatment remains the Ladd procedure originally described by Dr. Ladd in 1936 [6]. Complete resolution of acute obstruction or chronic abdominal pain is the result of a high index of suspicion for malrotation, appropriate diagnostic studies and aggressive definitive surgical treatment.

We present a case of malrotation in an adult who presented with a cramping right abdominal pain, as an escalation of a chronic vague intermittent abdominal one and underwent emergency laparotomy.

CASE REPORT
A 20-year-old young woman presented to our emergency department with cramping generalized right abdominal pain. Her symptoms began 24 hours prior shortly after eating. This cramping pain recurred intermittently at the beginning (every 1 to 2 hours), but, as time went by, the recurrences were more scarce. The duration of these pain episodes was 30 to 40 minutes. She vomited once prior to arrival without relief of nausea. Her last bowel movement was 2 days prior and it was described as normal. Her history consisted of multiple similar episodes of postprandial abdominal pain and vomiting since her teenage years which remained unidentifiable as regards to the cause. She had no other history of abdominal surgery. She was on no current medications and admitted cannabis abuse.

On physical examination, the patient’s vital signs were: pulse 88/min, blood pressure 120/70mmHg, temperature 36.8°C and respiratory rate 16/min. She was a well-developed, well-nourished woman in an acute distress. Her abdomen was not distended, but intestinal peristalsis was obscure. She exhibited peritoneal signs on the right side of her abdomen, with intense guarding even on superficial palpation. Her rectal examination was normal. Hemoglobin, white blood cell count and basic chemistry panel were all within normal values. Abdominal x-rays revealed two air-fluid levels in the right abdomen and air within the colon. A sonography of the upper right quadrant revealed nothing
irregular. She was admitted and managed with nasogastric tube decompression and intravenous fluid hydration under continuous re-evaluation.

On hospital day 1, the admittance cramping pain continued recurring intermittently. Axial contrast-enhanced computed tomography scan obtained through mid-abdomen showed inverted relationship between superior mesenteric vein (SMV) and artery (SMA) – the vein was lying to the left of the artery. Opacified small bowel was present almost entirely on the right side, the duodenjejunal junction was right-sided and the third and fourth duodenal portion did not cross the midline (Figure 1).

**Figure 1**
Figure 1: Axial contrast-enhanced computed tomography scan obtained through mid-abdomen shows inverted relationship between superior mesenteric vein (B) and artery (A) – the SMA is lying to the left of the SMV. Opacified small bowel present almost entirely on the right side

Upper gastrointestinal contrast studies showed malrotation of the small bowel without evidence of the duodenum crossing the lumbar spine. Almost the entire small bowel was noted to be sequestered on the right side of the abdomen (Figure 2).

**Figure 2**
Figure 2: Spot radiograph from barium upper gastrointestinal series shows contrast agent-filled duodenum and jejunal loops that remain right-sided without crossing the spine to the left

Based on the diagnosis of malrotation, the patient was consented for exploratory laparotomy and Ladd’s procedure. Upon entering the abdomen, the colon was noted to be completely mobilized (Figure 3) and the duodenum intraperitoneal in all of its portions. The ligament of Treitz was absent and the duodeno-jejunal junction was right-sided. A distended flabby third and fourth duodenal portion formed a steep angle, extrinsically obstructing the misplaced duodeno-jejunal junction (Figure 4). The jejunal loops were misplaced, slightly distended and oedematous, while the rest of the small bowel was normal. Cecal bands attaching to the duodenum but not to the right lateral abdominal wall were also noted (Figure 5). Bands were sharply lysed, a move that brought the cecum into the left lower quadrant. A laborious effort was exerted in order to mobilize the duodenum, eliminate the steep angle between the third and the fourth portion of it and thus treat the obstruction of the duodeno-jejunal junction. The effort proved to be unsuccessful, forcing us to perform a side-to-side duodeno-jejunal anastomosis, between the flabby duodenal portion and the first inches of the jejunum (Figure 6). Finally, an appendectomy was performed, to avoid diagnostic errors in later life. No cecopexy was performed.

Postoperatively, the patient did well, tolerated a regular diet
on postoperative day 7, and was discharged home on postoperative day 8. She has been in follow-up, which now extends to twenty months, without any signs of recurrence.

**Figure 3**
Figure 3: Fully mobilized right colon

**Figure 4**
Figure 4: The mobilized fourth portion of the duodenum and its steep angle

**Figure 5**
Figure 5: Fibrous adhesions between the jejunal loops and the large bowel

**Figure 6**
Figure 6: The side-to-side duodeno-jejunal anastomosis, between the flabby fourth duodenal portion and the first inches of the jejunum.

**DISCUSSION**

Midgut malrotation has been estimated to occur in approximately one in 500 live births [7]. Approximately 85% of malrotation cases present in the first two weeks of life [8]. However, it is difficult to accurately ascertain the true incidence because this condition will go undetected during childhood in a substantial subset of patients. In adulthood, intestinal malrotation is estimated to occur in 0.2% to 0.5% [9,10]. A literature review by von Flue et al. cites 40 cases from 1923 to 1992 [5].

Intestinal malrotation can be broadly defined as any deviation from the normal 270 degree counterclockwise
rotation of the midgut. During fetal development, the midgut supplied by the superior mesenteric artery grows too rapidly to be accommodated in the peritoneal cavity. Prolapse into the umbilical cord occurs around the sixth week. Between the tenth and twelfth week, the midgut returns to the abdominal cavity, having undergone a 270 degree counterclockwise rotation around the superior mesenteric artery. This rotation of intestinal development has been divided into 3 stages. Stage I occurs in weeks 5 to 10. It includes extrusion of the midgut into the extraembryonic cavity, a 90° counterclockwise rotation and return of the midgut into the fetal abdomen. Stage II occurs in week 11 and involves further counterclockwise rotation within the abdominal cavity completing a 270 degree rotation. The duodenum rotates caudal to the artery, and its C-loop traces this path. The transverse and ascending colon demonstrate the path of rotation of the cecum cephalad to the artery. Stage III involves fusion and anchoring of the mesentery. The duodenum becomes fixed retroperitoneal in its third portion, emerging at the ligament of Treitz, and the cecum becomes fixed to the lateral abdominal wall by peritoneal bands. The takeoff of the branches of the superior mesenteric artery elongates and becomes fixed along a line extending from its emergence from the aorta to the cecum in the right lower quadrant [11].

Intestinal anomalies can be categorized by the stage of their occurrence. Stage I anomalies include omphaloceles caused by failure of the gut to return to the abdomen. Stage II anomalies include nonrotation, malrotation, reversed rotation and paraduodenal hernias. Stage III anomalies include an unattached duodenum, mobile cecum, and an unattached small bowel mesentery [11]. If rotation is incomplete, the cecum remains in the epigastrium but the bands fixing the duodenum to the retroperitoneum and cecum continue to form. These bands (Ladd’s bands) [12], extending from the cecum to the lateral abdominal wall and crossing the duodenum, have the potential to cause obstruction. The mesenteric takeoff remains confined to the epigastrium, resulting in a narrow pedicle suspending all the branches of the superior mesenteric artery and the entire midgut. A volvulus may occur around the mesentery, obstructing the jejunum and also cutting off the blood supply to the midgut. Intestinal obstruction and the potential for total vascular compromise of the midgut supervene unless the condition is corrected.

Not all patients with malrotation present with symptoms. Indeed, most adult patients are asymptomatic and incidentally discovered later either at surgery for other conditions or at autopsy [4]. However, some may present with chronic symptoms of intermittent bowel obstruction or vague abdominal complaints. Even fewer may report acute episodes of agonizing abdominal pain [13]. Symptoms can arise from acute or chronic intestinal obstruction that may be caused by the presence of the Ladd bands and/or a volvulus.

There is no typical set of symptoms that are ascribed to this clinical syndrome. The location of the pain may vary from epigastric to left upper abdominal quadrant and it may be described as either intermittent cramping or persistent aching pain. It most often occurs postprandially and may last several minutes to an hour. Others have described severe abdominal cramping followed by diarrhea suggestive of chronic volvulus [14]. Vomiting may or may not be bilious and it is variable in duration and frequency. Another well-described presentation is a malabsorption pattern associated with diarrhea, nutritional deficiencies and failure to thrive [14]. Some authors postulate that diarrhea and malabsorption may be caused by bowel lymphedema resulting from lymphatic obstruction by chronic volvulus and resulting in loss of proteins into the bowel lumen [13]. Rare presentations of chronic volvulus include cases of obstructive jaundice by mechanical compression of the biliary tract [15], chylous ascites and superior mesenteric vein thrombosis, secondary to long-standing lymphatic and venous obstruction [16]. Lymphatic hypertension and disruption have been postulated to occur secondary to torsion of the small bowel mesentery. Other reported symptoms include constipation, solid food intolerance and gastroesophageal reflux [14].

The diagnosis of malrotation in adulthood is often delayed, because of the wider and more obscure constellation of clinical symptoms observed in adult patients, which leads clinicians and patients to attribute symptoms to the wrong diagnosis. All too often, such patients undergo numerous investigational tests and carry diagnostic labels such as irritable bowel syndrome, peptic ulcer disease or psychogenic disorder [15].

The diagnosis of intestinal malrotation can be made by radiographic studies [1]. Conventional radiography is neither sensitive nor specific for malrotation, although right-sided jejunal markings and the absence of a stool-filled colon in the right lower quadrant may be suggestive of this finding [12].

The standard upper gastrointestinal barium series remains
accuracy for detection (accuracy over 80%) and the rules familiar to pediatric radiology also apply for adults – that is, the duodeno-jejunal junction fails to cross the midline and lies below the level of the duodenal bulb \([24]\). An abnormal junction in an adult should not be dismissed as a normal variant.

Contrast enema examination usually shows malposition of the right colon with the ileum entering the cecum from the right, but the cecum may assume a normal location in up to 20% of patients. This normal location may cause malrotation to be missed on this type of study \([1122]\). The contrast enema findings are also non-specific because cecal location can be variable without malrotation.

Many cases of quiescent malrotation in adults are currently being detected on cross-sectional imaging (particularly CT) performed for various unrelated reasons \([24]\). CT not only shows the intestinal malpositioning seen on barium studies but also depicts associated extraintestinal findings not evident on conventional examinations. For example, deviation from the normal relationship between the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) is a useful indicator of malrotation \([33]\). In most patients with quiescent malrotation, the SMA and SMV will assume a vertical relationship or show left-right inversion \([33]\). Abnormalities of SMA-SMV orientation are not entirely diagnostic, however, because some patients with malrotation will have a normal relationship, and a vertical or inverted relationship can also be seen in patients without malrotation \([33]\). Therefore, isolated detection of such an abnormality is not sufficient for diagnosis but should warrant closer examination of the bowel. Finally, inspection of the pancreas in malrotation will reveal underdevelopment or absence of the uncinate process.

Midgut volvulus, a clockwise twisting of the bowel around the SMA axis because of the narrowed mesenteric attachment, is the most feared complication of intestinal malrotation and a clear indication for emergent surgery. The clinical diagnosis of midgut volvulus in adolescents and adults is difficult because the presentation is usually non-specific and malrotation is rarely considered. Recurrent episodes of colicky abdominal pain with vomiting over a period of months or years are typical and may eventually lead to imaging \([33]\). Diarrhea and malabsorption from chronic venous and lymphatic obstruction may also occur. Findings on abdominal radiographs in midgut volvulus are usually abnormal but non-specific. Upper gastrointestinal examination shows the typical corkscrew appearance of the proximal small bowel. However, in older patients with acute symptoms, CT is generally performed instead of a barium examination. Fortunately, the CT findings of malrotation with midgut volvulus are characteristic. The CT whirl or whirlpool sign describes the swirling appearance of bowel and mesentery twisted around the SMA axis \([1223]\). A similar appearance can be seen on sonography. Additional CT findings include duodenal obstruction, congestion of the mesenteric vasculature, and evidence of underlying malrotation. The presence of intestinal ischemia or necrosis is an ominous sign.

The classic treatment for incomplete intestinal rotation is the Ladd procedure, which entails counterclockwise detorsion of the midgut volvulus (if present), division of the abnormal coloduodenal Ladd bands tethering the midgut and causing extrinsic compression, widening of the mesenteric base to prevent further volvulus and removal of the malpositioned appendix \([124]\). These principles have remained the same since Ladd’s address to the New Hampshire Medical Society in 1936 \([40]\). Generally, symptomatic patients with malrotation should be treated surgically. Moreover, Spigland et al. recommended that all patients with malrotation are candidates for laparotomy, even if they are asymptomatic \([4]\), because the complications associated with intestinal malrotation are based on anatomical reasons that do not alter with age, thus the potential to develop sudden onset of acute midgut volvulus in an asymptomatic patient, at any age, exists.

Recently, laparoscopic techniques for treating malrotation in both infants and adults have been described. The laparoscopic experience with adults has consisted primarily of isolated case reports \([1072929]\), till Mazziotti et al. recently reported a series of malrotation patients managed successfully with laparoscopic intervention \([10]\). Moreover, Matzke et al. published their comparative results for open and laparoscopic Ladd procedures, concluding that the laparoscopic Ladd procedure is feasible, safe and as effective as the standard open Ladd procedure for the treatment of adults who have intestinal malrotation without midgut volvulus \([10]\). Similar conclusions are presented by Seymour et al. \([10]\), who also noted that long-term follow-up will be required to determine whether patients are at risk for either new symptoms or recurrent malrotation-related problems such as midgut volvulus.

Conclusively, the clinical diagnosis of malrotation after
Adult Intestinal Malrotation: A Case Report

childhood is usually not considered, because of the rare incidence of the disorder. These patients often present with obstruction and ischemia associated with a volvulus, or with chronic abdominal pain with elations, as in our case. Diagnosis requires a high index of suspicion. Specific findings that are diagnostic of malrotation can be detected through the use of both upper and lower gastrointestinal tract barium studies, angiography of the superior mesenteric artery, CT scan and often emergency laparotomy. Treatment remains as it was originally described by Ladd in 1936: mobilization of the right colon and duodenum, division of Ladd’s bands, division of adhesions around the superior mesenteric artery and appendectomy. Complete resolution of acute obstruction or chronic abdominal pain is the result of a high index of suspicion for malrotation, appropriate diagnostic studies and aggressive definitive treatment.

CORRESPONDENCE TO
Dr. B. Papaziogas Fanariou Str. 16, 551 33 Thessaloniki, Greece Tel: 0030-2310-992562, Fax: 0030-2310-992563 Email: papaziog@med.auth.gr

References
Author Information

B. Papaziogas
2nd Department of Surgery, “G. Gennimatas” General Hospital, Aristotle’s University of Thessaloniki Medical School

A. Patsas
2nd Department of Surgery, “G. Gennimatas” General Hospital, Aristotle’s University of Thessaloniki Medical School

G. Paraskevas

P. Tsiaousis
2nd Department of Surgery, “G. Gennimatas” General Hospital, Aristotle’s University of Thessaloniki Medical School

I. Koutelidakis
2nd Department of Surgery, “G. Gennimatas” General Hospital, Aristotle’s University of Thessaloniki Medical School

C. Christoforakis
2nd Department of Surgery, “G. Gennimatas” General Hospital, Aristotle’s University of Thessaloniki Medical School

K. Atmatzidis
2nd Department of Surgery, “G. Gennimatas” General Hospital, Aristotle’s University of Thessaloniki Medical School