

ADULT MIDGUT MALROTATION WITH LADD'S BAND: A RARE CASE REPORT WITH REVIEW OF LITERATURES

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Background: Midgut malrotation is an anomaly of intestinal rotation that occurs during fetal development and usually presents in the neonatal period. It is rare for midgut malrotation to present in adulthood. **Method:** We present a case of 15 year old girl presenting with acute abdomen of 15 days duration. Clinical evaluation and investigations revealed features of acute small gut obstruction with peritonitis. Patient underwent exploratory laparotomy which revealed Ladd's band with midgut malrotation with gangrenous and perforated duodeno-jejunal segment. Derotation with release of band with appendectomy and resection of gangrenous segment with end to end anastomosis and anatomical repositioning of the bowel was performed. **Results:** Complete resolution of symptoms seen in 4 months follow-up. We report this case for the rarity of the case with an unusual presentation of adult midgut malrotation due to Ladd's band presenting with gangrene of the affected gut with perforation, which was managed successfully.

Keywords: Mid gut malrotation; Ladd's band; Ladd's procedure.

INTRODUCTION

Almost 100 Years ago Mr. Zachary Cope delivered an Arris and Gale lecture entitled "The nerve supply of the parietal peritoneum and sub peritoneal tissues with remarks on its clinical application". This lecture and the subsequent writings of Sir Zachary Cope have greatly influenced our understanding of acute abdominal disease; the concepts of clinical observation enunciated in the lecture of 1922 remain valid today, they are part of the heritage of British surgery and form the basis for today's lecture on midgut malrotation.¹

Malrotation is typically diagnosed in the first few months of life, and 90% of cases are diagnosed during the first year. However, adult midgut malrotation is very rare and its incidence has been reported to be between 0.0001% and 0.19%.^{2,3} Most adult diagnoses of midgut malrotation are made in asymptomatic patients; either on imaging investigations for unrelated conditions or at operations for other pathology.

This scenario of incidental diagnosis is becoming increasingly common, particularly with improvements, and increased use, of diagnostic imaging techniques in modern practice. However, there are a small proportion of affected adults who may present with acute or chronic symptoms of intestinal obstruction or intermittent and

recurrent abdominal pain. The true diagnosis in this age group is fraught with immense difficulty, especially because the typical presentation is with non-specific symptoms and the fact that adult Surgeons usually have low index of suspicion and may not consider the diagnosis a possibility in the initial evaluation of adult patients with abdominal pain.

We present the case of a symptomatic 15-year-old female patient complaining of abdominal pain found to have an incidental diagnosis of midgut malrotation with small gut gangrene that was successfully treated with exploratory laparotomy, resection and anastomosis of the gangrenous bowel and Ladd's procedure with anatomical repositioning of the bowel. Also, the present case reminds us that midgut malrotation should be considered in the differential diagnosis of a wide variety of symptoms and should be treated promptly once the diagnosis has been confirmed. A detailed review of literatures has been done in this study.

Case report

A 15 year old girl presented to the emergency department with the chief complaints of pain abdomen for 15 days and vomiting for 7 days. The pain abdomen was diffuse and colicky in nature. The patient had also 2-3 episodes of bilious vomiting per day. There was no history of haematemesis and projectile vomiting. Pain abdomen was associated with low grade and continuous type of fever for 4 days. The patient was not passing stools for 4 days and there was no

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history of trauma and no history of loss of weight and appetite. There was no history of similar complaints in the past and no history of any past surgeries. She was born through normal full term vaginal delivery with no complications and the developmental milestones were also normal. She attained menarche at the age of 13 years. On examination the patient was conscious and coherent with low volume radial pulse of 120/minute and blood pressure of 100/60 mmHg. Her temperature was 101⁰ F. She had sunken eyes and dry tongue. There was no pallor, icterus, pedal edema and lymphadenopathy. On Per abdomen examination there was diffuse tenderness with guarding, rigidity and rebound tenderness. There was evidence of free fluid in the peritoneal cavity and of bowel sounds were absent. On investigation, hemogram showed neutrophilic leucocytosis with WBC count 16000/cmm. Renal and liver function tests were normal. Widal test for Salmonellosis was negative. Plain X-ray abdomen in erect posture was showing a dilated small bowel segment with no free gas shadow below right dome of diaphragm with some air fluid levels. Ultra sonogram revealed dilated small bowel loops with free fluid in the peritoneal cavity. The patient was initially resuscitated with 2L of crystalloids and was taken up for exploratory laparotomy with the preoperative diagnosis of intestinal obstruction with peritonitis. The intra operative findings included the presence of dilated, thickened, gangrenous segment of fourth part of duodenum and adjoining jejunum with a perforation over the gangrenous jejunum (Figure 1).The appendix and caecum were found in left iliac fossa. A Ladd's band was found which was responsible for obstruction of duodeno-jejunal segment (Figure 2).The band was released and the ligament of Treitz was divided and the duodenum was mobilized. The gangrenous part was resected and end to end anastomosis of third part of duodenum and jejunum was done. An appendectomy was done as a part of Ladd's procedure. Anatomical repositioning of the gut was done with a cecopexy on right side. She had an uneventful post operative recovery.



Figure 1

Dilated, thickened and gangrenous 4th part of duodenum and jejunum with perforation.

Patient was discharged on 10th post-op day. She was followed up for a period of 4 months and was found to be asymptomatic.



Figure 2
Ladd's band division

DISCUSSION

Initial presentation of symptomatic midgut malrotation is rare in adults. However, a significant number of cases remain quiescent during childhood. Incidental diagnosis may then occur in adulthood; when imaging investigations are carried out for other symptoms or, during surgery for unrelated pathology. It has been reported that the incidence of malrotation in adults is approximately 0.2%. However, it is probable that this rate will rise with future developments in diagnostic imaging. It is difficult to ascertain the true incidence, but evidence from post mortem studies suggest that gut malrotation may affect up to 1 in 6000.^{3,4} Midgut malrotation is broadly considered a deviation from the normal 270 degree counterclockwise rotation of the gut during embryonic development. During 4th week of fetal development, the embryonic gut, consisting of a straight endodermal tube, develops vascular pedicles to be divided into the foregut, midgut and hindgut based on the anatomical blood supply. The midgut is supplied by the superior mesenteric artery (SMA) and by the fifth week of embryonic life, it begins a process of rapid elongation and outgrows the capacity of the abdominal cavity. This leads to a temporary physiological herniation into the umbilical cord at about the sixth week of life with return to the abdominal cavity about 4 to 6 weeks later. During this period, the midgut undergoes a 270 degree counterclockwise rotation around the SMA axis. This process leads to the formation of the duodenal C-loop, placing it behind the SMA in a retroperitoneal position and emerging at the ligament of Treitz. The progressive reduction of the physiological midgut herniation commences at about week 10 of embryonic development. The duodeno-jejunal flexure (DJF) and jejunum are to reduce first and lie to the left. The distal small bowel then follows and lies progressively to the right of the abdominal cavity. The descent of the caecum from its higher position in the right upper quadrant forms the latter part of this complex

rotational development; it becomes positioned in the right lower abdomen. The ascending colon then assumes a retroperitoneal position, also on the right side. The base of the small bowel mesentery subsequently fuses with the posterior peritoneum in a diagonal fashion, from the ligament of Treitz at the DJF to the caecum, completing the whole process at about the eleventh week of fetal development.³⁻⁶

The failure of the normal physiological rotation of the midgut leads to various degrees of anomaly including the entire small bowel remaining on the right side of the abdomen, the caecum, appendix and colon on the left and an absent ligament of Treitz. In addition, the small bowel mesentery may develop a narrow vertical attachment and the peritoneal fibrous bands fixing the duodenum and caecum to the abdominal wall may persist. These congenital bands extend from the right lateral abdominal wall, across the duodenum and attach to the undescended caecum and are known as Ladd's bands.^{3,5,7,8} Ladd's bands compress the duodenum and can potentially cause duodenal obstruction. The malrotation of the gut and abnormal location of the caecum produces a narrow superior mesenteric vascular pedicle, as opposed to the normally broad based small bowel mesentery. This narrow SMA takeoff and lack of posterior peritoneal fusion predispose to subsequent midgut volvulus and obstruction with potential vascular catastrophe.^{7,9}

Midgut malrotation in adults presents in numerous ways and the symptoms are non-specific. The clinical diagnosis in adolescents and adults is difficult because it is rarely considered on clinical grounds. Many patients remain asymptomatic and the diagnosis is discovered incidentally during investigations or laparotomy for other unrelated problems in adult life. Wang and Welch showed that 24 of 50 patients were clinically asymptomatic in their case series of adolescents and adults with malrotation.³ Adults with a rotational abnormality of the gut usually present differently to pediatric patients. Two distinct patterns of adult presentations have been reported in the literature: acute and chronic.^{4,7,10} Chronic presentation is more common in adults. This is characterized by intermittent crampy abdominal pain, bloating, nausea and vomiting over several months or years. The symptoms may be highly nonspecific. However, the range of clinical presentations underlines the need for a high index of suspicion of midgut malrotation, when investigating the cause of intermittent and varying abdominal symptomatology in a healthy young adult.^{4,7}

Dietz et al studied a series of 10 adults with bowel obstruction caused by intestinal malrotation.⁴ They reported that 5 adults presented with chronic features and that the duration of symptoms extended to 30 years. Fu et al reported that 6 of 12

patients in their series presented with chronic intermittent abdominal symptoms.⁷ Diagnostic delays are common in this group of patients because of the nonspecific nature of the presentations. The pathophysiology of these chronic symptoms may relate to the compression effect of Ladd's bands running from the caecum and ascending colon to the right abdominal wall.^{4,11}

The other group of symptomatic adults typically presents with symptoms of acute bowel obstruction and these patients may or may not report a previous history of abdominal symptoms, as with our patient. These patients may on occasion, have symptoms and signs of an impending abdominal catastrophe. Moldrem et al reported that 48.5% of their thirty-three patients presented with an acute abdomen.¹⁰ Acute presentation may be due to volvulus of the midgut or ileo-caecum, reported as the most common cause of bowel obstruction in adults with gut malrotation. Other causes of acute presentation may be related to internal herniation caused by Ladd's bands. There is also a subgroup of acutely presenting adult patients with malrotation. They are identified when affected by other common abdominal diseases. Their unusual intestinal anatomy results in atypical signs and symptoms. These patients may present with localized peritonitis in the right upper quadrant or on the left side of the abdomen if their appendix becomes inflamed. The atypical presentations may lead to confusion, as one of the common abdominal pathology may mimic another, leading to incorrect diagnosis of conditions such as acute appendicitis, cholecystitis, pancreatitis, perforated peptic ulcer disease and left colonic diverticulitis. Several authors have reported observing atypical presentations of this nature before discovering gut malrotation with abnormal location of the caecum and appendix at surgery.^{4,7}

We can expect an increase in the incidental diagnosis of gut malrotation with increasing and widespread use of radiological investigations. Diagnostic features of midgut malrotation can be identified using plain abdominal radiograph, ultrasound scan (USS), computed tomography (CT) scan, magnetic resonance imaging (MRI) scan and mesenteric arteriography.^{10,12} Conventional plain radiography is neither sensitive nor specific in the diagnosis of gut malrotation although right-sided jejunal markings and the absence of a stool-filled colon in the right lower quadrant may be suggestive, leading to further investigation. Abdominal color Doppler USS may reveal malposition of the SMA, raising the suspicion of gut malrotation with or without the abnormal location of the hollow viscous.^{10,12,13} Characteristic USS findings of midgut volvulus were first described by Pacros et al and these findings include duodenal dilatation with distal tapering and fixed midline bowel and mesentery twisted around the

SMA axis. These features classically present as the 'whirlpool' sign.¹⁴ The reported gold standard for diagnosis of gut malrotation is an upper gastrointestinal (UGI) contrast study, particularly in the pediatric age group.^{4,11,12} This will generally show the duodenum and duodeno-jejunal flexure located to the right of the spine. The use of a contrast enema in conjunction with the UGI study has also been advocated as it can be used to demonstrate an abnormally located ileocaecum and right colon. However, contrast study findings may be nonspecific and a normal study does not exclude the possibility of gut malrotation.^{4,1,11,12}

CT scan with or without UGI contrast study is increasingly used preferentially as it is now considered as the investigation of choice; providing diagnostic accuracy of 80%.^{4,10,12} CT and MRI scans may show the SMV to be in an anomalous position; posterior and to the left of the SMA. In addition, they may show the abnormal anatomical arrangements of the midgut with the duodenum not crossing the spine. Deviation from the normal positional relationship of SMV and SMA was originally described by Nichols and Li as a useful indicator of the diagnosis of midgut malrotation.¹⁵ However, abnormal orientation of the SMA-SMV relationship is not entirely diagnostic of malrotation; it can also be seen in some patients without the pathology and a proportion of patients with malrotation may have a normal SMA-SMV relationship.¹²

Patients with gut malrotation will often have an underdeveloped or absent uncinat process of the pancreas. This is possibly due to the failure of the SMA to migrate to the left of the SMV.^{10,12} The CT appearance of midgut volvulus is diagnostic of malrotation. The shortened mesentery allows the small bowel and mesentery to twist and wrap around the narrowed SMA pedicle to create a distinctive 'whirlpool' appearance on CT scan. This pattern was first described by Fisher in a patient with midgut volvulus.¹⁶ It can be detected with both abdominal USS and CT scan. CT scan findings of gut malrotation and small bowel obstruction without volvulus may show internal herniation secondary to Ladd's bands.

Mesenteric angiography was previously used but is now rarely indicated in the evaluation of malrotation. It has the capacity to demonstrate the abnormal relationship between, and detect the patency of, the mesenteric vasculature. Angiography was used to demonstrate the characteristic corkscrew appearance of a whirling SMA and its branches; the 'barber pole sign' as well as extensive collaterals caused by proximal SMA occlusion.¹⁷ However, its role has been superseded by the CT scan which has the overall advantage of not only detecting the abnormal location of the midgut but also the reversed mesenteric anatomical relationship as well as any

other intra-abdominal anomalies associated with malrotation.

Symptomatic midgut malrotation undoubtedly requires surgical intervention although the management of asymptomatic patients is more controversial. Choi et al reviewed 177 patients over a 35-year period.¹⁸ They found that asymptomatic patients had a low risk of intestinal volvulus and therefore advised that routine investigations, screening and elective surgery were not necessary with close follow-up. However, it is increasingly argued that all suitable patients with intestinal malrotation should undergo surgical correction regardless of age as it is impossible to predict which patients will develop catastrophic complications.⁹ Several small case series have recommended that elective Ladd's procedure should be performed in all patients with intestinal malrotation. The authors of the studies that include cases of life threatening small bowel ischemia argue this point particularly strongly.^{2,4,7,10} Of course, the operative policy should be based on the presentation and suspected diagnosis; the potential risks of the procedure need to be weighed against the benefits.

The surgical management of intestinal malrotation was first described by William Ladd in 1936 and this remains the mainstay of treatment.⁵ The classical Ladd's Procedure consists of 4 parts: division of Ladd's bands overlying the duodenum; widening of the narrowed root of the small bowel mesentery by mobilizing the duodenum and division of the adhesions around the SMA to prevent further volvulus; counterclockwise detorsioning of the midgut volvulus if present and appendectomy to prevent future diagnostic dilemma of an abnormally located appendix.^{5,22} The original Ladd's procedure was described for the pediatric population group and the full components of this procedure may not be offered in the adult group.^{3,5,10} Most authors are of the opinion that Ladd's procedure is an adequate treatment for intestinal malrotation. Fu et al reported a complete resolution of symptoms in 9 and near complete resolution in 2 of 11 patients.⁷ Dietz et al also reported a complete symptomatic resolution in 8 of their 10 patients treated surgically.⁴ Variations of the techniques used to manage intestinal malrotation have been introduced to prevent recurrent volvulus. These include re-establishment of the normal gut anatomy by duodenopexy, cecopexy and suture fixation of the ascending colon to the right abdominal wall, in the retroperitoneal position.^{3,4,19} We offered this modified procedure to our patient by division of Ladd's band anatomical repositioning of the bowel, appendectomy and cecopexy. Of course our patient had to undergo resection and anastomosis of gangrenous perforated portion of the 4th part of duodenum and proximal

jejunum. She was completely symptom free at 4 months follow up.

There are recent reports of the use of the laparoscopic approach in the surgical treatment of intestinal malrotation. The technique appears to be safe and effective when performed by experienced laparoscopic surgeons, especially in the absence of volvulus.^{7-9,19,20} Laparoscopic Ladd's procedure in pediatric groups is increasingly reported in the literature. It is becoming more accepted as an initial approach to surgical correction of intestinal malrotation, resulting in shorter hospital stays. There are few reports of this approach in adults. The laparoscopic approach can be technically challenging and conversion to open procedure is common.^{7-9,21} A few published works have indicated that the laparoscopic approach can be successful in patients with malrotation and midgut volvulus.^{9,20} A retrospective analysis of both open and laparoscopic Ladd's procedures by Stanfill et al performed at the Children's Hospital of Illinois, USA noted that short-term results were superior with the laparoscopic approach and can be achieved without any increase in the duration of the operation.²¹

Although most of the literature consists of occasional case reports or small case series, we searched for literature published between 1983 and 2012 using PubMed and Web Japan Medical Abstracts Society and found 37 reported cases of teenage patients (ages 13 through 19) with intestinal malrotation.²³ Twenty patients were male and seventeen were female. The diagnosis could be made by radiographic studies in all these patients. Patients presented with a variety of gastrointestinal disorders. Abdominal pain was the most frequent symptom (30/37). Other symptoms were nausea, feeding intolerance, reflux, and respiratory problems. The Ladd procedure was performed on 27 patients; on 12 patients the procedure was conducted laparoscopically.

CONCLUSION

Intestinal malrotation is a rare condition but is considered an important cause of bowel obstruction in adults. The diagnosis of malrotation after childhood is difficult and usually not readily considered as the cause of intra abdominal symptoms. The presentation is usually nonspecific and this often leads to diagnostic and treatment delay with possible bowel ischemia and necrosis, with high degree of morbidity and mortality. Therefore, a high index of suspicion needs to be maintained and prompt surgical intervention must be considered in order to prevent an abdominal catastrophe and fatality. There are no reliable means of identifying which group of patients with intestinal malrotation will develop subsequent complications. In the light of this, many authors including us are now advocating early surgical

intervention in the form of a standard and modified Ladd's procedure. There is evidence in the literature that the use of Ladd's procedure with anatomical repositioning of the bowel relieves symptoms and in fact, prevents recurrence in the majority of patients.

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