## Case Report

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# Reverse rotation of gut with small bowel volvulus

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#### **ABSTRACT**

Malrotation of the midgut is generally regarded as paediatric pathology and is rare in adults leading to delay in the diagnosis and management. High suspicion is therefore required in patients with abdominal symptoms. We present a case of a 25-year old female who presented with an acute abdomen with preoperative computed tomography scan and operative findings confirming midgut malrotation. The duodenum, small bowel, caecum and appendix were not located in their anatomical site, with the presence of Ladd's bands and intestinal volvulus. Emergency laparotomy was done with patient having an uneventful postoperative recovery. Review on malrotation and volvulus is being presented to highlight its rarity and the controversies in its management.

**Keywords:** Midgut malrotation, Acute abdomen, Ladd's bands, Laparotomy

## INTRODUCTION

Intestinal malrotation is a congenital anomaly occurring in the development of the foetal gut formation. It has been estimated that it affects approximately 1 in 500 live births. The vast majority of the complications associated with midgut malrotation present in the first month of life and 60-85% of cases are diagnosed in this age group. It is reported that more than 90% of patients will present by the time of their first birthday. Adult midgut malrotation is very rare and its incidence has been reported to be between 0.0001% and 0.19%. 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.

We report a case of an adult patient with an acute presentation of midgut malrotation, highlightening the dilemmas in the preoperative diagnosis and its management.

## **CASE REPORT**

A 25 years female presented to the emergency department with upper abdomen colicky pain of three days duration, associated with episodes of bilious vomiting. She gave no history of hematemesis, jaundice, fever, trauma, malena or hemetochezia. General examination showed dehydration with tachycardia. Abdomen examination revealed epigastric tenderness without guarding or rigidity, without organomegaly or any other mass palpable. Rectal examination showed normal fecal staining without malena. Plain radiography showed dilated bowel loops (Figure 1). Ultrasound abdomen showed "Whirlpool sign" in the epigastric region with Superior Mesenteric Artery (SMA) and Superior Mesenteric Vein (SMV) reversed with a bowel mass suggestive of malrotation of gut with midgut volvulus (Figure 2). With the preoperative diagnosis of acute intestinal obstruction with malrotation and midgut volvulus, patient was resuscitated and taken for emergency laparotomy. Intraoperative findings were

midgut volvulus twisted clockwise thrice around its axis, Straight duodenum, transverse colon lying behind the duodenum & superior mesentric pedicle with absent mesocolon, unduly mobile caecum & ascending colon with long mesocolon lying in right hypochondrium & epigastrium and multiple bands between colon & small bowel (Figure 4-6). Detorsion, adhesiolysis, caecopexy and appendicectomy were done (Figure 7). Retroduodenal transverse colon was left as such. Her postoperative period was uneventful.



Figure 1: X-ray abdomen showing dilated bowel loops.

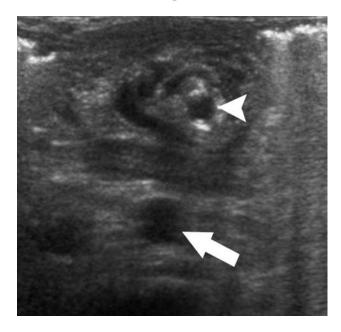


Figure 2: Ultrasound abdomen showing Whirlpool sign.

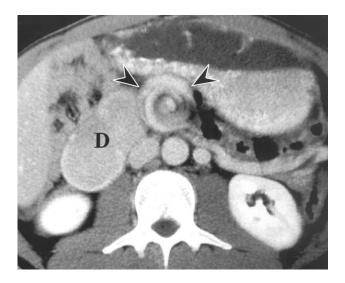


Figure 3: CECT abdomen showing Whirlpool sign.



Figure 4: Intraoperative picture showing abnormally mobile caecum and appendix.

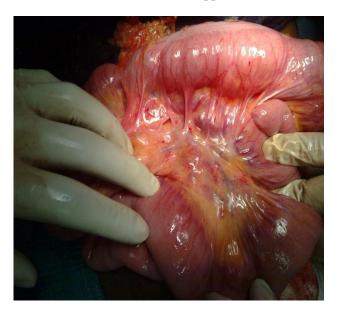


Figure 5: Intraoperative picture showing multiple bands.



Figure 6: Intraoperative picture showing straight duodenum.



Figure 7: Intraoperative picture showing caecopexy.

#### **DISCUSSION**

Normal embryology of human intestine is based on studies of Frazer & Robbins. In 1923 Dott published the first article on abnormalities of rotation of gut. In 1932 William Ladd described the surgical management of malrotation of gut. Incidence of malrotation leading to clinical disease has been reported to occur 1 in 6000 live births. Reverse rotation of gut was first reported by Tscerning in 1883. Midgut malrotation presenting with symptoms is rare in adulthood. Incidental diagnosis may be done, when imaging for other gastrointestinal symptoms or, during surgery for other pathology.

Midgut malrotation is broadly considered a deviation from the normal 270 degree counterclockwise rotation of the gut during embryonic development. Physiological herniation of bowel into the umbilical cord occurs at about the sixth week of life with return to the abdominal cavity occurring about 4 to 6 weeks later. Dott (1923) hypothesized that factors which allows caecum to slip back first into the peritoneal cavity, before small bowel returns leads to reverse rotation. Hunter (1922) explained that large umbilical opening will cause caecum to slip in first causing reverse rotation. Failure of duodeno-jejunum and hindgut loops to rotate normally with the extra-abdominal gut following passively was explained by Snyder and Choffin (1954).

The progressive reduction of the physiological midgut herniation commences at about 10 weeks of embryonic development. The Duodeno-Jejunal Flexure (DJF) and jejunum reduces first and lie to the left of the abdomen followed by the distal small bowel which lies progressively to the right and then the ascending colon which assumes a retroperitoneal position. The base of the mesentery subsequently fuses with the posterior peritoneum in a diagonal fashion, completing the whole process at about the eleventh week of fetal development. 1,4,7,11

The failure of the normal physiological rotation of the midgut leads to various degrees of anomaly. 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. The malrotation of the gut produces a narrow superior mesenteric vascular pedicle associated with a normal broad based small bowel mesentery. This narrow SMA take off predispose to subsequent midgut volvulus and obstruction with potential vascular catastrophe. <sup>12,13</sup>

Many patients remain asymptomatic and the diagnosis is discovered incidentally during investigations or laparotomy for other unrelated problems in adult life. Wang and Welch4 showed that 24 of 50 patients were clinically asymptomatic in their case series of adolescents and adults with malrotation. Two distinct patterns of adult presentations have been reported in the literature: acute and chronic. 11,12,14 Chronic presentation is characterized by intermittent crampy abdominal pain, nausea and vomiting, which may be highly nonspecific. Dietz et al.11 studied a series of 10 adults with bowel obstruction due to intestinal malrotation and reported that 5 adults presented with chronic features with the duration of symptoms extending to 30 years. Fu et al. 12 reported that 6 of 12 patients in their series presented with chronic intermittent abdominal symptoms. 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. 11,15 Acute presentation may be due to volvulus of the midgut or ileocaecum or may be related to Ladd's bands.

The atypical presentations in gut malrotation may mimics many intraabdominal pathology. Several authors have reported discovering gut malrotation with abnormal

location of the caecum and appendix at surgery. 11,12 Doppler ultrasound may reveal malposition of the superior mesenteric artery, raising the suspicion of gut malrotation. 14,16,17 Characteristic ultrasound findings of midgut volvulus were first described by Pacros et al., which includes duodenal dilatation with distal tapering and mesentery twisted around the SMA axis. These features classically present as the 'whirlpool' sign. 18 The gold standard investigation for the diagnosis of gut malrotation is upper gastrointestinal (UGI) contrast study. 11,16,17 However, contrast study findings may be nonspecific and a normal study does not exclude the possibility of gut malrotation. 11,12,15,16 Computed tomography scan with or without UGI contrast study is increasingly used preferentially as it is now considered the investigation of choice; providing diagnostic accuracy of 80%. 11,14,16 Deviation from the normal positional relationship of SMV and SMA was originally described by Nichols and Li<sup>19</sup> as a useful indicator of the diagnosis of midgut malrotation. 'Whirlpool' appearance on CT scan was first described by Fisher in a patient with midgut volvulus.<sup>20</sup> 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.<sup>21</sup>

Symptomatic midgut malrotation undoubtedly requires surgical intervention although the management of asymptomatic patients is more controversial. Choi et al.<sup>22</sup> reviewed 177 patients and found that asymptomatic patients had a low risk of intestinal volvulus and close follow-up alone is needed. The surgical management of intestinal malrotation was first described by William Ladd in 1936.6 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.6 There are recent reports of the use of the laparoscopic approach in the surgical treatment of intestinal malrotation, which appears to be safe and effective when performed by experienced laparoscopic surgeons, especially in the absence of volvulus.<sup>2,12,13,23,24</sup> Surgical intervention depends on site of obstruction. If obstruction is at DJ level and due to bands, Ladd's procedure is the surgery of choice. If it is due to volvulus, Detorsion with Ladd's procedure is done. If obstruction is at the level of transverse colon, colonic resection with displacement of the transverse colon anterior to duodenum and colocolic anastomosis is done.

### **CONCLUSION**

Intestinal malrotation is a rare condition in adulthood but is considered an important cause of bowel obstruction. The presentation is usually nonspecific and this often leads to diagnostic and treatment delay with possible bowel ischaemia and necrosis, with a poor prognosis and death. 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.

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