

## Case Report

# Midgut malrotation presenting as duodenal obstruction in an adult: a rare entity

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### ABSTRACT

Intestinal malrotation is a congenital anomaly of the small bowel with an incidence of 1 in 500 live births, mostly presenting in infancy. Presentation in adults is rare. A 46-year-old male presented with acute pain in abdomen, obstipation and bilious vomiting. The patient was vitally stable with insignificant clinical examination and normal hematological investigations. On abdominal CT, abnormal right sided small bowel loops with pulled up caecum in the midline, fibrotic bands extending from the DJ flexure towards the caecum, and Whirlpool sign relating to the superior mesenteric vein were seen. The patient was managed conservatively initially, and subsequently a Ladd's procedure was performed. Post operative course was uneventful. Though rare after childhood, symptomatic midgut malrotation may present in adults. Thus, high index of suspicion in patients presenting with non-specific abdominal pain and recurrent bilious vomiting, allows early recognition and management of this rare cause of small bowel obstruction.

**Keywords:** Intestinal obstruction, Midgut malrotation, Ladd's bands, Ladd's procedure

### INTRODUCTION

Midgut malrotation is a rare congenital anomaly, presenting in 0.2-1% of the population, and a mere 0.2-0.5% in adults.<sup>1,3</sup> It occurs in fetal life, around 10<sup>th</sup> week, due to failure of the clockwise rotation of the intestine around the superior mesenteric artery, leading to formation of a narrow-based mesentery and Ladd's bands, predisposing to intestinal obstruction. It has an incidence of around 1: 500 live births, mainly presenting in infancy.<sup>2</sup> It is rare in adulthood, often presents with frequent chronic, non-specific symptoms or as acute cases of upper gastrointestinal obstruction, and therefore may have a delayed diagnosis and management.

### CASE REPORT

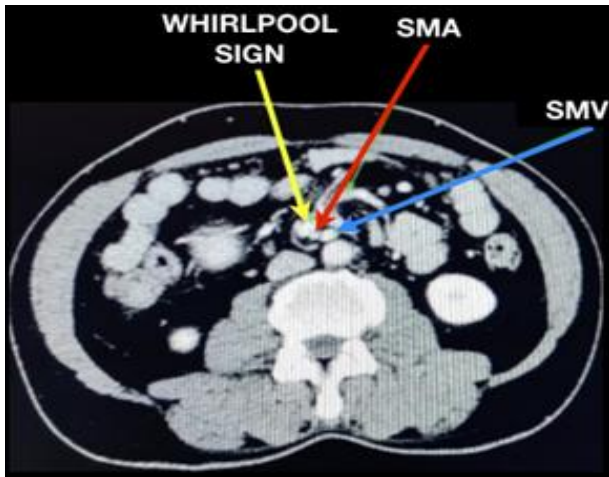
A 46 year old male presented with acute onset generalised colicky abdominal pain, bilious vomiting and obstipation since 2 days. Patient had no significant past history. The

patient was vitally stable with soft and non-tender abdomen, and normal rectal examination. The haematological and biochemical tests were within limits. An erect abdominal X-ray was found to be inconclusive, and an abdominal ultrasound revealed dilated stomach and duodenum.

A contrast enhanced CT scan of the abdomen was done, which showed, dilatation of the 2<sup>nd</sup> part of the duodenum, tapering of the retro mesenteric part of duodenum and superior mesenteric artery to the right of the superior mesenteric vein with swirling of the mesenteric vessels around the SMA, known as the whirlpool sign. The ileocaecal junction and caecum was pulled up and placed in the midline, just lateral to the mesenteric root, with fibrotic bands running across from the caecum to the right lateral abdominal wall, suggestive of Ladd's bands.

Patient was initially managed conservatively and posted for diagnostic laparoscopy in view of CT findings. A fibrous band spanning across the caecum to the right lateral

abdominal wall was seen with narrow, thickened root of mesentery. Adhesiolysis of the bands was done, but IC junction and DJ flexure were plastered to each other with some bowel adhesions which precluded further tracing of partly distended edematous bowel and hence converted to midline laparotomy. Remaining bands were excised and the anterior leaflet of the narrow root of mesentery was opened, DJ flexure and the IC junction freed, thus untwisting and restoring the bowel length. Appendectomy was performed and caecum fixed in the right iliac fossa (Ladd's procedure).



**Figure 1: Transverse section on CT scan showing whirlpool sign.**



**Figure 2: CT scan showing the dilated second part of duodenum.**

Patient tolerated the procedure well, was started on oral diet on the 3<sup>rd</sup> day and discharged 12 days post surgery. He was well on regular follow up over a year and half with no delayed symptoms or complications.

## DISCUSSION

Intestinal malrotation occurs as a result of failure of the 270 degree counterclockwise rotation of the midgut

around the superior mesenteric artery (SMA) axis during the first month of embryonic development. Normally, around the 4<sup>th</sup> to 10<sup>th</sup> week of development, the midgut elongates and herniates out of the abdominal cavity, into the umbilical cord, followed by the anticlockwise rotation of the gut around the SMA and return back into the cavity.<sup>1</sup> This results in a broad based mesentery, extending from the DJ flexure in the left upper quadrant and the IC junction in the right iliac fossa, with the ascending and descending colon to the right and left respectively.

Deviation from this normal process results in malpositioning of the midgut and formation of adhesions between the bowel loops and abdominal wall, known as Ladd's bands. The gut malrotation and shortened mesentery, may allow the formation of midgut volvulus due to twisting of the bowel around the SMA axis, which may lead to ischaemia and gangrene of that segment of bowel, and Ladd's bands if present, may lead to duodenal obstruction.<sup>3</sup>

A number of intestinal malrotation variations may occur, ranging from non rotation, reverse fixation and malrotation. Stringer classified it based on embryologic development as shown in the table.<sup>4</sup> The most common variation seen is the complete non rotation.<sup>2</sup>

**Table 1. Stringer classification for bowel malrotation.<sup>4</sup>**

Type	Variation of malrotation
I a	Non rotation of the colon and duodenum
II a	Non rotation of duodenum only
II b	Reversed rotation of colon and duodenum
II c	Reversed rotation of duodenum only
III a	Duodenum and colon not rotated
III b	Incomplete attachment of hepatic flexure
III c	Incomplete attachment of caecum
III d	Internal hernia near ligament of Trietz

Presentation of intestinal malrotation most commonly occurs in the first month of life, and very few cases are seen in adulthood, or may remain asymptomatic for life.<sup>5</sup> In adults it may present with non specific, chronic symptoms like recurrent abdominal pain, vomiting or intolerance to food with weight loss. Acute or acute on chronic presentation may also be seen, with severe abdominal pain, bilious vomiting, haematemesis or haematochezia, with or without haemodynamic instability, but occurs in only about 20% of the adult cases, and has increased incidence midgut volvulus leading to bowel ischaemia and gangrene, thus more likely fatal.<sup>1,7</sup> Due to its vague presentation in adults, it may lead to a missed or delayed diagnosis.

Diagnosis of intestinal malrotation may be made in symptomatic cases, or as an incidental finding while investigations are carried out for other symptoms or an unrelated pathology.<sup>6</sup> Abdominal X-rays are found to be neither sensitive, nor specific. On ultrasonography, diagnosis of intestinal malrotation is often missed, unless

specific findings are looked for. These may be an inverted SMA-SMV relationship, where the SMA lies anterior or to the right of SMV. A more useful sign is to check for the retro mesenteric part of the duodenum, which lies in a transverse plain between the aorta and SMA normally, and thus rules out intestinal malrotation.<sup>2</sup> The gold standard for diagnosis in adults, remains contrast enhanced computed tomography (CECT) of the abdomen and pelvis, which may show right sided small bowel and the large bowel to the left, displaced caecum, inversion of the SMA-SMV relationship with swirling of the mesenteric vessels around the SMA, Whirlpool sign, seen in case of midgut volvulus.<sup>2,3,5</sup>

Treatment is based on severity of symptoms. Chronic cases may be managed conservatively, but should eventually be taken for elective laparoscopic or open Ladd's procedure. Acute cases may need emergency laparotomy with untwisting of the bowel in case of midgut volvulus, or resection of bowel segment in case of gangrene, along with the Ladd's procedure, after appropriate haemodynamic resuscitation. Ladd's procedure consists of the following steps- division of the fibrotic Ladd's bands to release the compression in the midgut; widening the root the mesentery, mobilising the DJ flexure and caecum with lengthening of the bowel; fixing the caecum in the right iliac fossa, with or without fixation of the DJ flexure, and appendectomy, in case of appendicitis in the future, which may be difficult to diagnose. Ideally, all cases of intestinal malrotation should undergo Ladd's procedure. Hence, all conservatively managed cases and also cases incidentally diagnosed, should undergo elective Ladd's procedure, to prevent the future recurrence of midgut volvulus or intestinal obstruction.<sup>3,4,7</sup>

## CONCLUSION

Through this study, we aim to highlight the importance of keeping the diagnosis of intestinal malrotation in the differential in adults presenting with acute or chronic abdominal symptoms. High index of suspicion, accurate

radiological diagnosis and timely and adequate surgical management is important to prevent complications and future recurrences in cases of intestinal malrotation.

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