

Intestinal Malrotation in Older Children – A Case Series of Delayed Diagnosis

Shrikesh Singh¹, Saurabh Srivastav², Gaurav Raj Singh³, * Tanvir Roshan Khan⁴

¹ Associate Professor, Department of Paediatric Surgery, Dr RML Institute of Medical Sciences, Lucknow, India.

² Assistant Professor, Department of Paediatric Surgery, Dr RML Institute of Medical Sciences, Lucknow, India.

³ Junior Resident, Department of Surgery, NIMS College, Jaipur, India.

⁴ Additional Professor, Department of Paediatric Surgery, Dr RML Institute of Medical Sciences, Lucknow, India.

Abstract

Background: Intestinal malrotation presents with symptoms that can be easily confused with other non-surgical conditions in children. In this case series, we aimed to increase awareness about this condition for its early recognition and management; and contribute to the current knowledge on this condition.

Case report: In this case series, we present five cases of Intestinal malrotation where the diagnosis was not suspected by the primary physicians. They subsequently had delayed surgical referral leading to morbidity and mortality.

Conclusion: Intestinal Malrotation in older children is usually not suspected due to atypical symptoms. Its diagnosis should be considered in older children with recurrent abdomen pain, especially if there is an associated history of malnutrition or poor weight gain. Early identification of malrotation by the attending physicians is of utmost importance. The patient should be referred for early surgical intervention to prevent morbidity and mortality. Education related to the timely diagnosis of malrotation in older children is crucial.

Key Words: Abdomen Pain, Case Series, Delayed Diagnosis, Intestinal Malrotation, Malnutrition, Poor Weight Gain.

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*Corresponding Author:

Tanvir Roshan Khan, Additional Professor, Department of Paediatric Surgery, Dr RML Institute of Medical Sciences, Lucknow, India. Email: doctrk@gmail.com

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1- INTRODUCTION

Intestinal malrotation refers to the abnormal position of the intestine in the peritoneal cavity and usually involves the midgut. It results from abnormal or incomplete rotation of the midgut during embryonic development (1). The incidence is about 1 in 500 live births (2). The majority of the patients with Intestinal malrotation (>85%) are diagnosed in the neonatal or early infancy; however, it is not uncommon to see these patients presenting in childhood or adulthood (3). The real incidence of malrotation of the bowel is difficult to estimate as most children do not have typical presentation or may be asymptomatic (4). The presentation of bilious vomiting in a previously healthy infant should raise the suspicion of malrotation; however, the presentation beyond infancy varies, and may present as life-threatening midgut volvulus (5). In this case series, we present five cases of Intestinal malrotation where the diagnosis was not suspected by the primary physicians. They subsequently had delayed surgical referral leading to morbidity and mortality. We aimed to increase awareness about this condition for its early recognition and management; and contribute to the current knowledge on this condition.

2- CASE SERIES

2-1. Case 1

A 5-year-old Female came to our department with a complaint of recurrent episodes of pain in the abdomen, and vomiting since the age of 3 months which was being managed conservatively. The patient was well till the age of 3 months when she developed the first episode of pain abdomen with vomiting when she was managed on Intravenous (IV) medications and sent back home without any investigation. As per her mother's report, she continued to have similar episodes and she was managed with medications for

each episode. Later at the age of 4 years, when she had less weight for her age and decreased hemoglobin, a blood transfusion was done and was diagnosed with malnutrition associated with the tubercular abdomen. Treatment for tuberculosis was started and continued for 3 months. She was asymptomatic for this duration and again she had similar complaints. Whole abdomen Ultrasonography (USG) was also performed but failed to detect malrotation. She was later referred to our department for further management. Contrast Enhanced Computed Tomography (CECT)abdomen was then done which revealed the following findings: Small bowel is located on the right side and large bowel located on the left side of the peritoneal cavity and reversed relation of Superior Mesenteric vein and artery, suggesting Intestinal Malrotation. Laparotomy was done which revealed the following findings: 1. Appendix placed on the left side; 2. Malrotation of the gut with narrow mesentery (**Fig. 1**). Surgical correction in the form of Ladd's procedure was done, and the child recovered well.

2-2. Case 2

A 4-year-old female presented to the emergency with complaints of fever, nonspecific pain in the abdomen, and bilious vomiting for one month. A detailed history reported by her mother revealed that the patient was asymptomatic till the age of 2 years when she first complained of pain in the abdomen with occasional vomiting. She had nonspecific recurrent episodes of colicky pain in the abdomen relieved by medications. At the age of 3 years, the patient complained of intermittent fever and recurrent abdominal pain associated with vomiting and no passage of stools for around two days which was managed conservatively. Although USG and CT abdomen were done elsewhere and reported multiple mesenteric lymph nodes conglomerated together predominantly in the

periumbilical region with mild ascites, malrotation was not reported. Her distension continued to increase with severe abdominal pain and vomiting; then she had subsequently referred to us with a distended abdomen, constipation, multiple bilious vomiting, and dehydration. We planned an emergency laparotomy which revealed the following findings. 1. Gross

distension of bowel loops; 2. Clockwise rotated bowel along its axis with an appendix on the left side; 3. Narrow mesentery; 4. Polysplenia; 5. Suggestive of Intestinal malrotation with mid-gut volvulus, Ladd's procedure was done without appendectomy (**Fig. 2**). The child recovered well.



Fig. 1: Exploratory laparotomy in the 5-year-old female showing appendix on the left with narrow DJ mesentery with malrotation of gut



Fig. 2: Ladd's procedure is being done in the 4-year-old male child showing a distended bowel with rotated bowel with left side appendix with Polysplenia.

2-3. Case 3

A 13-year-old male was admitted with a history of abdomen pain since the age of 5 years which was insidious in onset, diffuse, and colicky. There was an associated history of multiple episodes of bilious vomiting. On examination, the patient was malnourished. He was afebrile and hemodynamically stable. His pulse and blood pressure were measured as 84 bpm and 110/70 mmHg, respectively. The abdomen was mildly distended with slight tenderness in the epigastrium, umbilicus, and right iliac fossa. A digital rectal examination revealed no significant abnormality. Routine blood investigations were normal. Chest radiograph did not show air under the diaphragm. Abdominal radiograph revealed non-dilated gas-filled loops of bowel in the central and right abdominal regions. The diagnosis was

confirmed when a contrast-enhanced CT was obtained which demonstrated features of malrotation, i.e., SMV was seen encircling and rotated around SMA associated with rotation and whirlpooling of small bowel loops and twisting of mesentery with prominent proximal bowel loops. The patient was taken up for exploratory laparotomy. Intraoperative findings showed the small bowel on the right side of the abdomen. The caecum and ascending colon were present in the midline. The duodenal loop was found making a double counterclockwise rotation around the SMA pedicle at the base of the transverse mesocolon. Also, a jejunal loop was seen making another double counterclockwise rotation around a band at the ileocolic junction. Ladd's procedure was performed with appendicectomy and widening of the mesentery (**Fig. 3**).

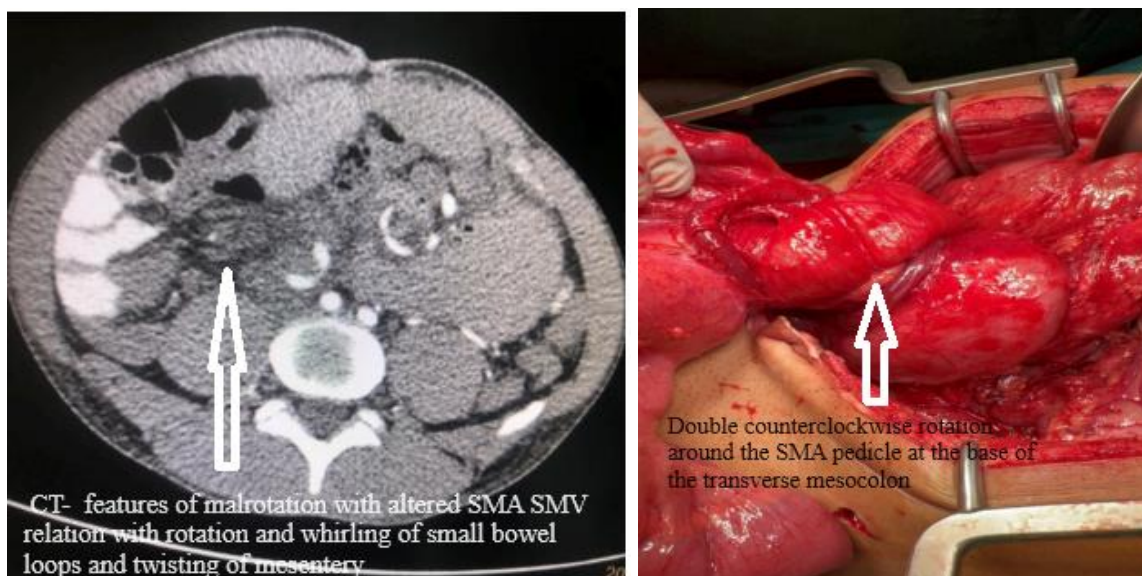


Fig. 3: 13-year-old male with CT suggestive of rotation of the Superior mesenteric Vein along with whirling of small bowel loop and operative finding of double counterclockwise rotation around the SMA pedicle at the base of the transverse mesocolon.

2-4. Case 4

A 7-year-old boy was admitted to our care after experiencing five days of constipation and inability to pass gas. He had previously received treatment

elsewhere but had received no relief. He was also experiencing bilious vomiting and was dehydrated and lethargic at the time of admission. Upon examination, his abdomen was tense and tender with discoloration of the abdominal wall. Both

hernial orifices were intact and no other surgical scars or lesions were found. Per rectal examination revealed mixed blood staining. Routine blood tests were normal, except for slightly elevated prothrombin time. USG abdomen was done and showed twisting of proximal jejunal bowel loop around its mesentery with pulling up SMA within the twisted mass of bowel loop

suggestive of volvulus with associated bowel obstruction. Considering the patient's condition, he was resuscitated and prepared for surgical exploration. On exploratory laparotomy, the peritoneal cavity had blood-filled fluid with multiple dense adhesions with malrotated midgut volvulus with a nonviable bowel (**Fig. 4**).

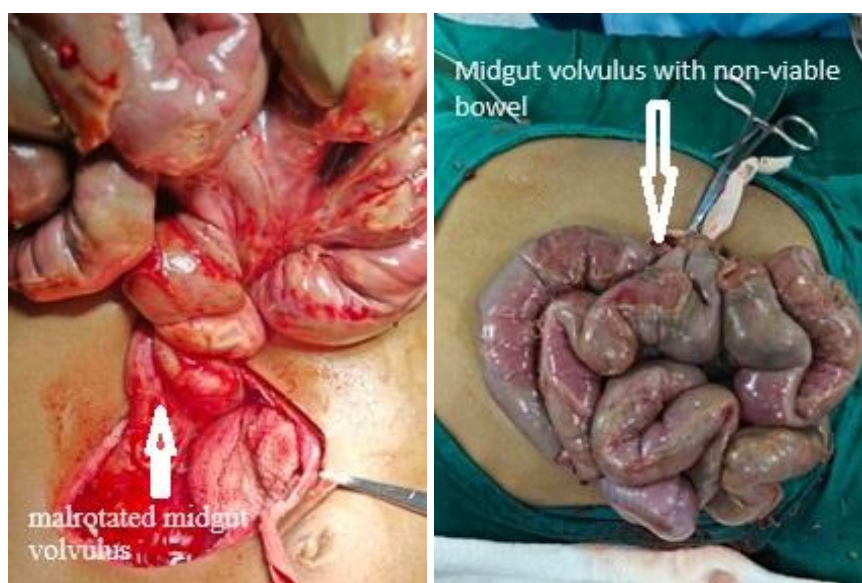


Fig. 4: Clinical picture of the 7-year-old male child with malrotation with volvulus with exploratory laparotomy finding of non-viable bowel.

Derotation of the midgut with peritoneal lavage was done and a peritoneal drain was placed. He was scheduled for a re-look laparotomy after 24 hours. The patient, however, later developed hematemesis disorientation and shock for which he was managed. The patient's parents took the patient against the medical advice from the hospital.

2-5. Case 5

An 11-year-old female was referred to our center in a state of shock and poor general condition. On examination, she had abdominal distension which was tense and tender, and bilious aspirate in the Nasogastric tube. She was resuscitated and shifted to the operation theater for urgent laparotomy which showed midgut volvulus

with gangrene of the bowel. Derotation of the gangrenous bowel with peritoneal lavage was done and a drain was put in for a re-look laparotomy. She, however, died before re-look laparotomy.

3- DISCUSSION

Intestinal malrotation, unfortunately, presents with symptoms that can be easily confused with other non-surgical conditions in children. Most of the patients with intestinal Malrotation present in the neonatal period contrary to those in our series and some of those published by other authors. The first contact physicians are usually unaware of this condition and the child is being treated symptomatically in most of the cases.

The case series presented here highlights the importance of early identification of this problem and appropriate referral by the first contact physicians. The early recognition not only leads to better outcomes but also prevents the morbidity and mortality associated with Intestinal Malrotation and in our series all of the cases had delayed referral for Paediatric Surgical consultation subsequently increasing morbidity and mortality. These patients mostly had nonspecific symptoms like pain in the abdomen, bloating, and vomiting; however, one important thing

that needs to be mentioned is that these patients had a history of bilious vomiting at some point of time in their presentation and had poor nutrition. So, it is recommended to carefully record the history in these cases. Like in our first case, the child was given treatment for tuberculosis for 3 months before an opinion from the Paediatric Surgeon was sought. Similarly, all of these children had delays in the identification of Intestinal Malrotation as a cause of abdominal pain (**Table 1**).

Table-1: Details of the children with delayed diagnosis of Intestinal malrotation

S No.	Age (Years)/Sex	Presentation/duration of symptoms	Management	Outcome
1	5 Year/Female	recurrent episodes of pain in the abdomen, and vomiting since the age of 3 months of age	Surgical correction in the form of Ladd's procedure	Recovered well
2	4 Year/Female	fever, nonspecific pain in the abdomen, and bilious vomiting since 2 years of age	Ladd's procedure was done without appendectomy	Recovered well
3	13 Year/Male	Pain in the abdomen with recurrent episodes of bilious vomiting, since 5 years of age	Ladd's procedure was performed with appendicectomy and widening of the mesentery	Recovered well
4	7 Year/Male	Sudden onset of severe pain in abdomen with bilious vomiting and constipation for 5 days	Exploratory laparotomy with gut derotation and peritoneal drain	Left against medical advice before relook laparotomy.
5	11 Year/Female	Abdominal pain and distension with bilious vomiting for 3 days followed by dehydration and shock at the time of presentation	Urgent exploratory laparotomy with gut derotation and peritoneal drain	Expired, before relook exploratory laparotomy.

Few previous studies suggest that chronic conditions are more common in children above the age of 2 years (6-7). In their case series and literature review of eleven patients with a delayed presentation over 5

years, the authors concluded that the presentation of intestinal malrotation beyond infancy is a rare condition and early intervention and treatment can prevent catastrophic conditions such as

intestinal volvulus and intestinal Ischemia (8). One case report described the delayed presentation in a 6-year-old female child with intestinal obstruction, being treated for recurrent abdominal pain for the last 4 years with surgical opinion sought only when the condition deteriorated (9).

It is a known fact that symptomatic patients with documented malrotation require surgical intervention. However, the management of patients with asymptomatic malrotation is debated. In the study of Stewart et al., 15% of all patients diagnosed with malrotation were asymptomatic (10). The finding in the present series was midgut volvulus with ischemia and bowel loss in two cases and the importance of the surgical opinion and correction is largely emphasized. Most of the current literature supports the surgical correction of malrotation in patients with mildly symptomatic or incidentally discovered cases (11).

Surgical Correction in the form of Ladd's Procedure involves evisceration, derotation, cutting of Ladd's bands, widening of Dudeno-jejunal mesentery, and appendectomy. It has become the standard of care. The use of laparoscopy for the diagnosis or correction of malrotation has been reported in the past 20 years. Laparoscopy helps to determine the location of the ligament of Treitz and fixation of the cecum with the appendix. Relevant studies have emphasized the value of laparoscopy in both the diagnosis and treatment of malrotation, especially in patients with acute abdominal symptoms with questionable diagnoses requiring emergency surgery (12).

Few previous studies on the presentation of Intestinal Malrotation have also reported delayed presentation (13). The reports have also suggested that delayed presentation of these patients results in more incidence of volvulus and intestinal gangrene as the diagnosis is often missed at admission (14). In neonates, intestinal

malrotation presents acutely with bilious vomiting (93%), failure to thrive, and abdominal pain. Around 37% of infants and 12% of older patients have an acute presentation in the form of intestinal volvulus and obstruction leading to bowel gangrene (15). The present series also highlights the importance of early diagnosis and surgical treatment in patients of Intestinal malrotation, so the incidences of volvulus with bowel ischemia and gangrene can be minimized.

4- CONCLUSION

Intestinal Malrotation in older children is usually not suspected due to atypical symptoms. Its diagnosis should be considered in older children with recurrent pain abdomen, especially if there is an associated history of malnutrition or poor weight gain. Early identification of malrotation by the attending physicians is of utmost importance. The patient should be referred for early surgical intervention to prevent morbidity and mortality. Education related to the timely diagnosis of malrotation in older children is crucial.

5- AUTHORSHIP CONTRIBUTIONS

Dr. Shrikesh Singh: Investigations, Data curation, clinical images, Dr Saurabh Srivastav: Visualization, Methodology, Conceptualization, data collection. Dr. Gaurav Raj Singh- conceptualization, original draft, clinical images. Dr. Tanvir Roshan Khan: Writing – review & editing, Supervision, Methodology, Investigation, Data curation.

6- ETHICAL CONSIDERATIONS

Informed consent was obtained from the patient's parents/guardians.

7- COMPETING INTEREST

None.

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