



Evaluation of Cases of Intestinal Malrotation with Midgut Volvulus

¹Dr.Santosh V.Patil, ²Dr. Priya S. Patil, ³Dr.Vibhu Sharma, ⁴Dr.Sudhakar S. Jadhav

¹Dr. Santosh V.Patil, ³Dr.Vibhu Sharma, ⁴Dr.Sudhakar S. Jadhav

Department of Paediatric Surgery, Paediatric Surgery Centre and PG Institute, Sangli, Maharashtra, India.

²Dr. Priya Santosh Patil, Associate Professor, Department of Anatomy, Bharati Vidyapeeth (Deemed to be University)

Medical College & Hospital, Sangli, Maharashtra, India.

Corresponding Author: Dr. Priya Santosh Patil, Associate Professor, Department of Anatomy, Bharati Vidyapeeth (Deemed to be University) Medical College & Hospital, Sangli, Maharashtra, India.

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Introduction

Congenital intestinal malrotation mostly presents as a paediatric surgical emergency. Patients most commonly present during childhood with acute abdominal symptoms. Intestinal malrotation can remain undetected and present at a variable age even during adulthood. ^[1] The patients may also present as intestinal obstruction or volvulus which is a surgical emergency. The accurate diagnosis and prompt surgical intervention can be lifesaving in such a scenario. The diagnosis of malrotation by imaging modalities is often challenging. ^[2] The knowledge of intestinal malrotation along with associated complications, embryological basis can help the surgeon in better understanding and prompt management of such cases. Keeping this in mind the current study focusses on identifying the cases of intestinal malrotation retrospectively in a period of three years. The aim was to analyze the data in order to highlight the age of presentation, symptoms and signs, diagnostic tests used to diagnose cases of intestinal malrotation. We have discussed the surgical intervention and complications in such cases with their clinical implications.

Material and Methods

The study was conducted at Paediatric Surgery Centre and P.G.Institute, in a period of three years from January 2015 to March 2018. It was a retrospective, observational, institute based study. All the patients who were diagnosed of intestinal malrotation and confirmed on X-ray and USG with Doppler study were included in the study. The patients who had come for other acute abdominal emergencies apart from malrotation and associated anomalies like congenital diaphragmatic hernia or internal hernia were excluded from the study.

The patients' medical records were evaluated. The symptoms and details of physical examination were noted. For diagnosis of intestinal malrotation plain radiograph and USG with Doppler study of abdomen were used. The patients' operative notes were studied and recorded. The follow up of these patients was obtained through the case papers. The patients or the parents were asked standard questions regarding the growth of the child, general development, abdominal complaints and any other episode of bilious vomiting in the post-operative period.

Observations

In a period of three years a total of 15 cases of intestinal malrotation were identified at our institute. Out of these 8 were male patients and 7 were females. The age wise distribution of cases is shown in table 1.

Table 1: Age wise distribution of 15 cases

	Neonate(0 to 1 month)	Infant (1 month to 1 year)	More than 1 year
Male	3	3	3
Female	2	1	3
Total	5	4	6

Symptoms: The symptoms at presentation according to their severity are mentioned in table 2. The most common symptom was bilious vomiting seen in 93.3% cases which was followed by distension of abdomen and abdominal pain. Other symptoms like constipation and weight loss were less common. One case presented with per-rectal bleeding.

Table 2: Symptoms of patients

Symptoms	Number of patients out of 15	% of patients
Vomitting	14	93.3 %
Abdominal pain	4	26.6 %
Distension of abdomen	6	40 %
Constipation	2	13.3 %
Weight loss	5	33.3 %

Investigations: The diagnostic modality used for confirmation of intestinal malrotation was Plain X-Ray , Contrast- Barium meal and USG abdomen with doppler. [Fig:1,2,3] Investigations like Barium meal was done in 4 cases and MRI of abdomen done in 1 patient. The choice of investigations depended on the clinical presentation and severity of the patient condition. 13 out of 15 patients had midgut volvulus.

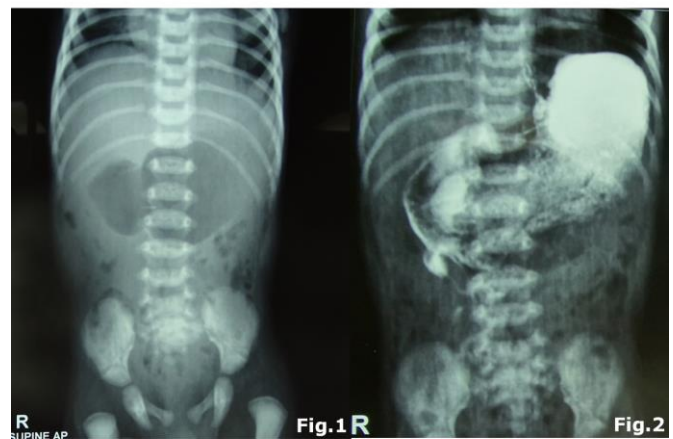


Fig:1 – Showing Plain X-ray and Fig 2 showing Barium meal. Dilated stomach and duodenal loop with reduced gas shadows in bowel are seen



Fig. 3 USG showing dilated stomach and duodenum with midgut volvulus

Surgical Management: The choice of surgical management was exploratory laparotomy with Ladd’s procedure.[Fig:4,5] Most of the patients (13 out of 15 cases) 86.6% had associated volvulus which was corrected. Some of the patients (2 cases) 13.3 % had additional intestinal bands which were corrected. In cases where the mesentery was narrow, widening of the mesentery was done. Appendicectomy was performed in all 15 cases. The vascularity of the intestine was checked and the abdomen was closed.

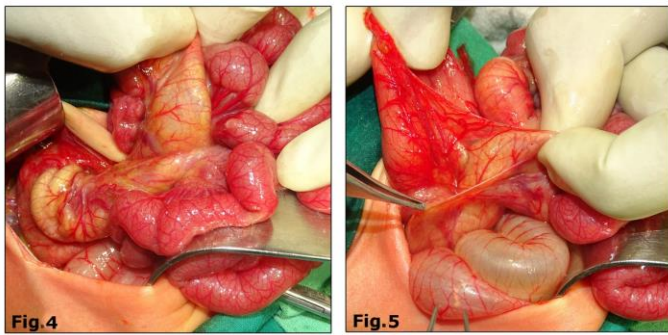


Fig. 4,5 : Intraoperative photograph showing midgut volvulus and intestinal bands respectively

Complications: Only one patient had very short gut and total gangrene of the gut which had presented with per rectal bleeding, succumbed and expired due to septicemia, 8 days after surgery.

Follow-up: Post-operative follow up showed good recovery and weight gain within three months. One patient operated in 2015 had intestinal obstruction due to adhesions in 2018 and underwent laparotomy again for adhesionolysis.

Discussion

The development of small intestine mainly occurs outside the abdominal cavity during 5th to 10th weeks of intrauterine life. The growth and development of intestine can be divided into three stages: [3]

First stage: Stage of physiological herniation wherein the rapidly elongating gut pushes out of the abdominal cavity through the umbilicus. (6th to 10th week)

Second stage: Reduction of hernia occurs in the tenth week when the gut reenters the abdominal cavity and assumes definitive positions.

Third stage: This is the stage of fixation which takes place from eleven weeks onwards and comprises of the fixation of parietal peritoneum and descent of caecum to its adult position.

The anomalies of rotation can be described as maldevelopment occurring during the above stages. The primitive midgut is divided by the centrally located

superior mesenteric artery into a proximal pre-arterial segment and a distal post-arterial segment. [4] The normal rotation of the gut occurs for about 270 degrees counterclockwise around the axis passing through superior mesenteric artery. First 90 degree rotation brings the midgut loop back to the abdominal cavity. In the next 90 degree counterclockwise rotation the coils of jejunum and ileum lie behind the superior mesenteric artery. The third rotation brings the postarterial segment with the caecum and appendix to the right side and the transverse colon crosses the superior mesenteric artery. The elongation of postarterial segment makes the caecum descend and lie in the right iliac fossa.

An error or disruption of the rotation of gut can lead to complete nonrotation or incomplete rotation of the gut leading to malrotation anomalies. Depending upon the errors in degree of rotation the intestinal anomalies are divided into types such as nonrotation, mixed rotation, reversed rotation, hyperrotation and encapsulated small intestine or internal omphalocele. [3] The anomalies due to maldevelopment during third stage result in abnormalities of position of caecum alongwith its complications. [3,4] Malrotation are are divided into two main types : Nonrotation and mixed rotation. Nonrotation is the most common type when the gut rotates only through 90 degrees and the postarterial segment is first to enter the abdominal cavity. In this condition the colon lies on the left side, caecum in the midline and small intestine on the right. This condition is common in males than females in a ratio of 2:1. It is frequently associated with volvulus.

The incidence of rotational anomalies is difficult to estimate due to the fact that most of them remain asymptomatic and may be diagnosed as an incidental finding. The relative incidence of nonrotation is about 0.5 percent in autopsy specimens. Some have reported an incidence of 1:200 in anatomical specimen and clinical

incidence of 1:25,000 admissions. In some researches there were more cases of malrotation on radiography while very less number of them were symptomatic.^[3]

While in a review article by S.Aslanabadi the incidence reported was 1 in 500. There was a male predominance with a male-to-female ratio of 2:1. They also mention that 40% of patients with malrotation present within one week, 50% within a month and 75% by the end of one year of their life. The patients who remain asymptomatic can even present in adult life in about 25% cases.^[4]

It is clear from above that intestinal malrotation is mostly asymptomatic. It presents usually with an associated complication such as volvulus or other abdominal diseases like appendicitis or diverticulitis.^[5] In the present study out of total cases of malrotation all presented with intestinal obstruction and 93.3 % had bilious vomiting as the presenting symptom. The review article by S. Aslanabadi in 2007, shows the presenting complaints and clinical picture of thirty cases of malrotation. They too had bilious vomiting as the main complaint in majority of patients followed by abdominal distension and failure to thrive.^[4,6] The mainstay investigations carried out in suspected cases of malrotation were Plain or contrast radiograph and USG. In rare cases of complications a CT scan was also done. Use of CT is debatable in cases of malrotation.^[5] In our institute we prefer Radiograph and USG as choice of investigation. Surgical treatment in our cases of intestinal malrotation was a standard Ladd's procedure. In Ladd's procedure the intestinal bands are cut and mesentery is widened.^[7] The duodenum is straightened and small intestine placed on right side while large bowel on left side. Hence caecum and appendix come to lie in left hypochondrium as a part of this procedure. In future if such patients suffer from appendicitis, they may be misdiagnosed. Hence to avoid this, appendectomy is done routinely at our institute during Ladd's procedure for

all cases of malrotation. However not all surgeons do appendectomy in every patient of malrotation as they prefer conservation of appendix for future use as conduit.

^[5] Intestinal malrotation is commonly associated with volvulus.^[8] This further increases the risk of torsion, ischaemia, necrosis and gangrene of the bowel which makes it a life threatening emergency. In this study 86.6% (13 out of 15) cases presented with midgut volvulus which was corrected during the surgery. There were bands seen intraoperatively which were taken care of. In (7 out of 15 cases) 46.6 % cases the mesentery had a narrow root which was widened during surgical procedure. All 13 out of 15 operated patients were followed up for 6 months and showed good recovery, weight gain and no recurrence. One patient presented after three years with intestinal obstruction and underwent adhesiolysis. One patient died due to volvulus leading to gangrene of intestine and septicaemia.

Intestinal malrotation may very rarely present as mesocolic hernia – internal hernias.^[9] This is because of the mesentery which fails to fix to the abdominal wall thus creating potential space for internal herniation. The herniated loop is prone for ischemia and necrosis. In our study we did not come across cases with internal hernias.

Intestinal malrotation can present in any age group from neonate to adult. The presentation, diagnosis and surgical treatment is much easier with less complications and recurrence in paediatric age group.^[10] In adults it is often misdiagnosed or present as complex syndromes with surgical emergency or chronic abdominal symptoms.^[6,11] The incidence of this condition is also impossible to estimate exactly due to the variable age of presentation. Nowadays there are more cases diagnosed due to advanced modalities of investigations, thus depicting a higher incidence. The imaging modalities for diagnosis also differ and are debated. In this scenario prior

knowledge of embryological basis of malrotation, high degree of clinical suspicion of presence of malrotation and proper choice of investigations can definitely be valuable for prompt diagnosis and during surgical procedure. The presence of malrotation, bands, narrow vascular pedicle, narrow root of mesentery and or volvulus can be taken care of during surgery to avoid life threatening complications.

Conclusion

Intestinal malrotation may be undetected and most commonly presents with complications like midgut volvulus. Timely investigations as indicated with proper interpretation of findings will be helpful in correct diagnosis and prompt surgical management with reduction in morbidity and mortality. Treatment of paediatric cases of malrotation show better results than adults with less complications and recurrence.

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