

## Intestinal Malrotation: An Insight in the Difficulties in the Diagnosis and Management

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### Abstract

*Background:* Various clinical presentations, ranging from chronic abdominal pain to acute midgut volvulus with ischaemic bowel injury, may result from intestinal malrotation. In general it is possible to detect the intestinal malrotation, however there are many variables and exceptions. The purpose of this study was to retrospectively analyse the patients of intestinal malrotation at a single surgical centre in the last 10 years and to identify the spectrum of presentation, factors responsible for the delay affecting the diagnosis and outcome in these patients.

*Methods:* The study was conducted in a tertiary referral centre with separated Department of Paediatric Surgery. Patients admitted and operated from July 2005 to July 2015 with complains of intestinal malrotation were included in this study. Case records were analysed with special emphasis on age, mode of presentation, any delay in treatment if any and the reason for it was noted. The operative records were retrieved to know the exact operative findings, the procedure performed and the outcome of surgery. Follow up was done for at least 1 year, after surgery and postoperative complications if any were analysed.

*Results:* Total 220 patients of intestinal malrotation were included in this study. Male were 143 and 77 were female. Neonates constituted the largest group (n=87) followed by infants (n=56). There 40 patients

who had delay in diagnosis. Biliious vomiting was the commonest presentation (n=62/153) in early age group while chronic abdominal pain was seen in older age group (n=37/77). Ladd's procedure was the commonest surgical procedure (n=167) followed by intestinal resection and stoma formation (n=28) and resection and anastomosis (n=21). There were 14 mortalities and 17 complications needing admission in follow-up period of 1 year.

*Conclusion:* Intestinal malrotation is a surgical problem mostly of neonates but it is more likely to be ignored in older children who are otherwise being treated for some unrelated conditions and continue to develop chronic abdominal pain and intermittent distension, and poor growth. A high index of clinical suspicion is needed especially in these patients. The problem if not diagnosed properly leads to bowel loss, increased morbidity and mortality.

**Keywords:** Intestinal Malrotation; Diagnosis; Management.

### Introduction

Malrotation is a spectrum of anatomic abnormalities due to incomplete rotation and fixation of intestine during fetal development. Since the first description of normal embryology of rotation and fixation of human intestine given by Mall in 1898, it's a diagnostic dilemma for surgeons. There may occur anomalies in rotation and positioning, absence of development or defects of the organ and vessels of digestive tract [1,2].

Rotational anomalies are in form of non-rotation, incomplete rotation to reverse rotation [2,3]. Apart from rotation anomalies, there may be defect of fixation

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and final positioning of the intestine and hernia formation, most commonly paramesocolic [4]. Intestinal malrotation may be associated with various anomalies, including the major defect of the abdominal wall and congenital diaphragmatic hernia, upto 30% to 60% [5]. The presentation of this surgical problem ranges from neonate to adults and varies from bilious or non bilious vomiting, vague abdominal discomfort to life threatening midgut volvulus and ischemic infarction of intestine. The surgical management is not changed much since it was described by Ladd in 1932. Although up to 90% present before 1 year of age but some cases diagnosed very late or frequently in emergency situation and with serious clinical consequences. There are some patients who even getting antipsychotic treatment for recurrent abdominal pain without the diagnosis of intestinal malrotation.

This paper retrospectively reviewed all cases of malrotation highlighting the difficulties in management with delayed and unusual presentation in this part of world.

## Material and Methods

This study was conducted in the Department of Paediatric surgery King George's Medical University Lucknow, India. 10 year data for the patients admitted and operated from July 2005 to July 2015 with complains primarily due to malrotation were

included in this study. The patients in whom malrotation was incidentally found during operation for some other pathology were excluded. The diagnosis was suspected primarily on the basis of clinical presentation and confirmed by upper G I contrast studies. The clinical criteria to suspect a patient having malrotation were vomiting bilious or nonbilious, recurrent abdominal pain and distension or non-specific abdominal symptoms where all other pathology were ruled out. The major criteria for diagnosing malrotation on Upper G I contrast are a dilated stomach, a dilated C loop of duodenum, and right sided duodenojejunal flexure to the midline. No other diagnostic modality was used to diagnose malrotation. Those patients who presented as acute intestinal obstruction with very high index of suspicion for malrotation or with signs of bowel ischemia were subjected to operative procedure directly after resuscitation and plain abdominal radiograph without further diagnostic evaluation and diagnosis was confirmed intraoperatively. The data thus obtained was analysed by stratifying the patients according to age, sex, primary symptoms at the time of presentation, operative procedure and surgical outcome.

## Results

Total 220 patients are included in this study. The results in the form of age of presentation, mode of

**Table 1:** Age and mode of presentation

Age	No. of Patient	Mode of presentation
0-1 month	87	Bilious vomiting- 43 Non-Bilious vomiting- 29 Chronic pain and Distension- 4 Non specific abdominal complaints & failure to thrive-2 Acute intestinal obstruction (Mid gut volvulus)- 5 Signs of bowel ischemia and shock- 4
1-12 month	56	Bilious vomiting- 19 Non-Bilious vomiting- 12 Chronic pain and Distension- 8 Non specific abdominal complaints & failure to thrive-11 Acute intestinal obstruction (Mid gut volvulus)- 3 Signs of bowel ischemia and shock- 3
1-5 year	37	Bilious vomiting- 2 Non-Bilious vomiting- 5 Chronic pain and Distension- 13 Non specific abdominal complaints & failure to thrive-4 Acute intestinal obstruction (Mid gut volvulus)- 6 Signs of bowel ischemia and shock- 7
5-10 year	27	Bilious vomiting- 1 Non-Bilious vomiting- 2 Chronic pain and Distension- 18 Non specific abdominal complaints & failure to thrive-2 Acute intestinal obstruction (Mid gut volvulus)- 9 Signs of bowel ischemia and shock- 8
>10 year	13	Chronic pain and Distension- 18 Non specific abdominal complaints & failure to thrive-2 Acute intestinal obstruction (Mid gut volvulus)- 9 Signs of bowel ischemia and shock- 8

**Table 2:** Sex distribution

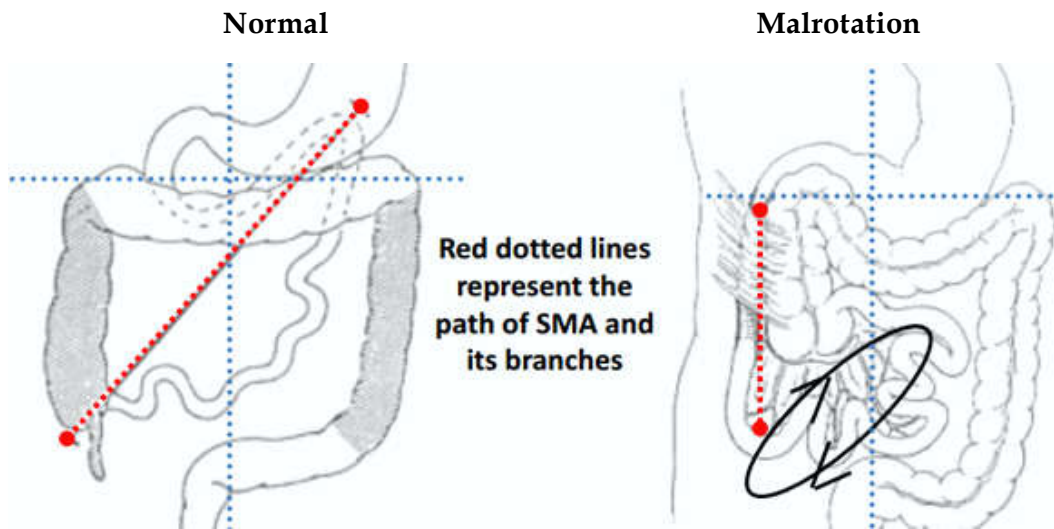
Male	143
Female	77

**Table 3:** Operative procedure

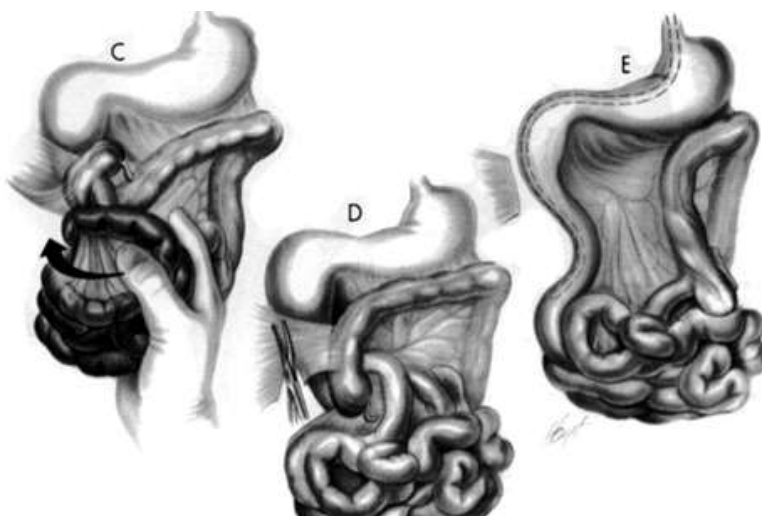
Ladd's procedure(evisceration and derotation of bowel, release of caecalbands,widening of mesentry,)	167
Resection with primary anastomosis	21
Resection with double barrel stoma	28

**Table 4:** Complications

Mortalities	14
Wound infection	12
Delayed peristalsis	27
Incisional hernia	02
Post operative adhesive obstruction	15



**Fig. 1:** Normal intestinal arrangement and intestinal malrotation. SMA- Superior mesenteric artery, courtesy;APSA Committee of Education 2015-2016



Reference:  
Ladd WE, Gross RE:  
Abdominal surgery of  
infancy and  
childhood.  
Philadelphia, WB  
Saunders, 1941.

**Fig. 2:** Steps of Surgery (Ladd's Procedure)

presentation and treatment offered of this study is summarised in the following tables.

### Discussion

Intestinal malrotation is one of the most frequent embryonic malformation related to the digestive tract with presentation either, complete absence of rotation (commonest), incomplete rotation – less than 270° – or inverse rotation (Figure 1). The incidence rate of malrotation is 3-5% of surgical obstruction [2-5]. Males are more commonly affected than females [6]. Some study shows racial variation in incidence of intestinal malrotation, Mongolians are more commonly affected than Caucasians [7].

Most of the patients of malrotation presented in first year of life (n=143)(Table 1) and among those presented in their first year more than half presented in first month(n=87)(Table 1). Even though most of them presenting before the age of 1year, these congenital anomalies are still being diagnosed late in 40 patients (Table 1). According to largest series about 10-40% of cases have a late clinical appearance (6,8-11). In Our study 40(18.8%) patient were having late presentation, > 5 years (Table 1) . About 65% of the patients were male (n=143) (Table 2).

The most common presentation was vomiting either bilious or non-bilious (51.36%) (Table 1), with bilious vomiting mostly seen in neonates. Second most common presentation was recurrent abdominal pain and distension particularly among the child presenting late (n=37/77) (Table 1). The cause is thought to be the recurrent midgut volvulus which derotates spontaneously to relieve the symptoms. Some authors have describe upto 50% of intestinal volvulus in their patients under 8 year of age [12]. Considering high morbidity/mortality rates of patients who present with a volvulus, and reduction of this rate among patients who undergo a preventive Ladd's procedure, the need for early diagnosis and treatment is evident [13,14]. Some older patients presents with non specific abdominal symptoms like anorexia nausea constipation and failure to thrive. Malabsorption and malnutrition in intestinal malrotation could possibly be due to occurrence of lymphoedema or mesenteric venous stasis [15]. This group of patients are most difficult to diagnose. A high index of suspicion, a careful history followed by upper G I contrast is essential to diagnose these patients .Some authors have reviewed several radiological findings and attributed the cause to malrotation depending the position of Treitz ligament at right(typical) or at left, above or below of 12<sup>th</sup> thoracic vertebra(atypical) [16]. Apart from upper GI

contrast study, Doppler ultrasound is also recommended, looking for inversion of the mesenteric vessels - the so called whirlpool sign [17,18]. Positive Doppler ultrasound finding are reliable even in neonates, with an 85% accuracy rate in cases of complete malrotation, and they are useful to indicate other, more reliable, tests(19). About 20%(n=41) (Table. 1) of our patients presented as acute abdomen, and about half of them had signs of bowel ischemia at the time of admission(n=18)(Table 1). These patients after a quick resuscitation and plain abdominal radiograph were subjected to operative intervention and diagnosis is confirmed intraoperatively.

In 167 (75.9%) (Table 3) of our patients classical Ladd's procedure was performed (Figure 2). In those situations where irreversible bowel ischemia was present resection of gangrenous bowel with primary anastomosis or exteriorisation of bowel as double barrel stoma was done. In this series 14 patients died (6.3%) (Table 4). Among those who expired 9 were presented with bowel ischemia and shock with very low general condition, 2 neonates aspirated in postoperative period 1 premature infant developed apnoea and cardiopulmonary arrest, one patients went in bradycardia during surgery leading to cardiopulmonary arrest and one patient expired at home a week after discharge from unknown cause.

The most common post-operative complication was delayed peristalsis 27(12.27%) (Table 4) which was diagnosed by excessive bilious aspirate from ryles tube or vomiting when trial to feed was given. All the patients with this complication were managed conservatively by delayed oral feeds and prokinetic drugs. Persistence of symptom could be due to an intrinsic innervation defect associated with some of these malformations [20] and not to the surgical technique that were used. These symptoms have been describe as affecting 25% of patients who were operated on as neonates affected by volvulus, with vomiting being the most frequent symptoms (45%) and sub-obstructive clinical manifestation with abdominal distention being the least frequent (21%)[21]. It therefore seems necessary to advise family members of the possibility of a partial reappearance of the previous symptoms, that this situation may be long-lasting, and that it could be associated with a defect of innervation of the digestive tract. But above all, they must be advised that surgery is absolutely necessary in order to avoid an episode of intestinal volvulus [22]. 12(5.4%) (Table 4) patients developed wound infections and two developed incisional hernia. 15 patients came

with post-operative adhesion obstruction out of which 10 managed conservatively and 5 required surgical correction (Table 4).

### Conclusion

It can be said that being a surgical problem mostly of neonates Intestinal malrotation can also present in older children. Not uncommonly the patients of malrotation remain undiagnosed for very long time. The diagnosis can be made on clinical suspicion and confirmed by upper GI contrast studies. The common presentation is vomiting but it can also present as a surgical emergency where prompt surgical interventions is required to prevent intestinal infarction. The problem if not diagnosed properly lead to bowel loss and increased mortality in neonates and a high index of suspicion is needed to identify these cases in older children, and it's not rare to see these patients being treated for tuberculosis or having antipsychotic treatment owing to ignorance, which is not uncommon in this part of world.

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### References

- Dott NM: Anomalies of intestinal rotation. Their embryology and surgical aspects, with report of five cases. *Br J Surg* 1923; 11:251.
- Snyder WH, Chaffin L: Embryology and pathology of the intestinal tract: presentation of 48 cases of malrotation. *Ann Surg* 1954; 140:368-377.
- Kantor JL: Anomalies of the colon: their Roentgen diagnosis and clinical significance. Resume of ten years study. *Radiology* 1934; 23:651-662.
- Estrada, R.L: Anomalies of Intestinal Rotation and Fixation. Charles C Thomas, Inc, Springfield, Ill; 1958.
- Warner BW, Malrotation. In: Oldham KT, Colombani PM, (Foglia RP (eds) *Surgery of infants and children: scientific principles and practice*. lippincott-Raven, Philadelphia, 1997. p.1229-1240.
- Ford EG, Senac MO Jr, Srikanth MS, Weitzman JJ: Malrotation of the intestine in children. *Ann Surg* 1992; 215:172-178.
- Forrester MB, Merz RD, Epidemiology of intestinal malrotation, Hawaii, 1986-99. *Paediatr Perinat Epidemiol* 2003; 17:195-200.
- Janik JS, Ein SH: Normal intestinal rotation with non-fixation: a cause of chronic abdominal pain. *J Pediatr Surg* 1979; 14:670-674.
- El-Gohari MA: Cook RCZ, Intestinal malrotation beyond the neonatal period. *Kinderchir* 1984; 39: 237-241.
- Rescorla FJ, Shedd FJ, Grosfeld JL, Vane DW, West KW: Anomalies of intestinal rotation in childhood: analysis of 447 cases. *Surgery* 1990; 108:710-715 (discussion 715).
- Spigland N, Brandt ML, Yazbeck S: Malrotation presenting beyond the neonatal period. *J PediatrSurg* 1990; 25:1139-1142.
- Fernandez Sanchez A, Lopez Pereira P, Diez Pardo JA, Utrilla J: Intestinal malrotation in children. *An EspPediatri* 1987; 27:375- 378.
- Stewart DR, Colodny AL, Daggett WC: Malrotation of the bowel in infants and children: a 15 year review. *Surgery* 1976; 79:716-720.
- Andrassy RJ, Mahour GH: Malrotation of the midgut in infants and children: a 25-year review. *Arch Surg* 1981; 116:158-160.
- Iida F, Wada R, Sato A, Yamada T: Clinicopathologic consideration of protein-losing enteropathy due to lymphangiectasia of the intestine. *Surg Gynecol Obstet* 1980; 151:391-395.
- Mehall JR, Chandler JC, Mehall RL, Jackson RJ, Wagner CW, Smith SD: Management of typical and atypical intestinal malrotation. *J PediatrSurg* 2002; 37:1169-1172.
- Weinberger E, Winters WD, Liddell RM, Rosenbaum DM, Krauter D: Sonographic diagnosis of intestinal malrotation in infants: importance of the relative positions of the superior mesenteric vein and artery. *AJR* 1992; 159:825-828.
- Yeh WC, Wang HP, Chen C, Wang HH, Wu MS, Lin JT: Preoperative sonographic diagnosis of midgut malrotation with volvulus in adults: the "whirlpool" sign. *J Clin Ultrasound* 1999; 27:279-283.
- hao HC, Kong MS, Chen JY, Lin SJ, Lin JN: Sonographic features related to volvulus in neonatal intestinal malrotation. *J Ultrasound Med* 2000; 19:371-376.
- Devane SP, Coombes R, Smith VV, Bisset WM, Booth IW, Lake BD, Milla PJ: Persistent gastrointestinal symptoms after correction of malrotation. *Arch Dis Child* 1992; 67:218-221.
- Coombs RC, Buick RG, Gornall PG, Corkery JJ, Booth IW: Intestinal malrotation: the role of small intestinal dysmotility in the cause of persistent symptoms. *J PediatrSurg* 1991; 26:553-556.
- Maxson RT, Franklin PA, Wagner CW: Malrotation in the older child: surgical management, treatment, and outcome. *Am Surg* 1995; 61:135-138.