

Study of the Spectrum of Anomalies of Intestinal Malrotation in Children

SIBGHAT ULLAH¹, MUHAMMAD ABID AZIZ², SAJJAD HUSSAIN³, MANZOOR HUSSAIN⁴, ASHIQ MUHAMMAD⁵

^{1,2,5}Medical Officer, ³District Surgeon, Surgical B Unit District Headquarters Hospital Mardan

⁴Junior Registrar, Medical A Ward Mardan Medical Complex Mardan

Correspondence to: Muhammad Abid Aziz e-mail: surgeonabidaziz@gmail.co Cell +923475785848

ABSTRACT

Objective: To assess the spectrum of malrotation in relation to age groups in terms of symptoms as well as the anatomical types of malrotation.

Study Design: Prospective study

Place and Duration of Study: Children's Hospital, PIMS, Islamabad from 1st June 2019 to 31st January 2020.

Methodology: All patients presenting to the outpatients' department or the emergency room were included in the study. Patients who were unstable or had inconclusive evidence for malrotation were excluded. All participants underwent barium studies and Doppler ultrasonography which confirmed the diagnosis. The data was collected through a proforma incorporating clinical presentation, physical examination, investigations, and operative findings.

Results: The mean age of the patients was 8.6±20.3 months. Majority was male. In the neonatal age group, 10 (31.3%) patients had pain, 32 (100%) had vomiting and 28 (87.5%) had abdominal distension. In the patients under 1 year of age, 7 (70%) had pain, 9 (90%) had vomiting and 7 (70%) had abdominal distention. Of the patients between 12 to 59 months, 6 (100%) had pain 1 (16.7%) had vomiting and 4 (66.7%) had abdominal distention. 31 (62.0%) of patients each had dilated gut loop and air fluid level on x-ray. The Doppler ultrasound found out reverse pattern of superior mesenteric artery and vein in 27 (54.0%) cases.

Conclusion: The intestinal malrotation presents in early life within one month of life in two third of cases, however, a substantial number of cases can present in infancy and few even present later in life.

Keywords: Anomaly, Duodeno-jejunal loop, Intestinal malrotation, Pediatric

INTRODUCTION

Bowel malrotation is a developmental anomaly which affects the peritoneal attachments of small and large bowels in foetal life during organogenesis. The rotation and fixation of the embryo intestine around an axis formed by the superior mesenteric artery is defined as missing or incomplete. In the beginning, the stomach is above the artery and in adulthood it continues. The duodeno-jejunal Loop must be swung from above to the right 90° and below 90° to its final location to the left for a further 90°, a total of 270 degrees. The duodeno-jejunal Loop has to pass around the superior mesenteric artery. The rotation direction is in the opposite direction. From its starting point below the artery, the cecocolic embryo moves counterclockwise, to the left of the 90-degree artery, and a further 90 degree to the right of the artery. Finally, because of its weight, the cecocolic loop descends retroperitoneally to its final adult location of right iliac fossa. Different subtypes classify this spectrum of abnormalities based on the midgut development phase.^{1,2} It is, however, helpful to simplify non-rotation and incomplete rotation categorisation.³ Returning rotation may also take place, but is rare.³

Rotational anomalies that can be symptomatic or not occur between 1/200 and 1/500 live births.^{4,5} It is estimated that 1/6000 live births are symptomatic malfunction.⁶ About two-thirds of children who need malrotation surgery are younger than 1 months of age, 18-25% are 1 month to 1 year of age and 10-18% are older than 1 year.^{4,7,8}

There are different factors and malrotation associated with it. Between 30 and 62% of children who suffer from intestinal abnormalities.^{7,9,10} Diaphragmatic diaphragms, gastroschisis, and Omphalocele are the most common

rotating defects in infants. Approached malrotation can include up to 17% of children with duodenal atresia and 33% of children with jejunoileal atresia.^{11,12}

Some patients with acute gut obstruction due to midgut volvulus in neonatal life, rely on the symptoms and the form of malrotation. They have gallstone vomiting, gastrointestinal disease, utterly stubbornness and dehydration, with or without rectal bleeding. The midgut volvulus can be intermittently twisted and untwisted in other mode of presentation. Recent episodes of vomiting and constipation in these patients have occurring. The signs and symptoms of duodenal blockage caused by the Ladd's bands can be present in patients. Patients can have chronic stomach pain vomiting or inability to prosper in later age groups (5-15 years). Patients with localised or diffuse abdominal distension may experience recurrent episodes of abdominal pain or vomiting in the age group.^{13,14}

Dilated gut loops can be present in plain X-ray abdomen and in lateral positions, with levels of air fluid [15]. However, X-ray abdominal findings are based on patient type and age. We conducted this study in order to study, the various pattern of malrotation in relation to the age of the patient, the different presentations, their diagnosis and treatment options.

MATERIALS AND METHODS

This prospective study was conducted at Department of Paediatric Surgery, The Children's Hospital, PIMS, Islamabad during from the period 1st June 2019 to 31st January 2020. Total 50 suspected malrotation patients were collected from outpatients department, emergency and underwent barium studies and Doppler ultrasonography which confirmed the diagnosis. Patient not

willing to be a part of the study, those already operated for malrotation in other hospitals who come with recurrence or a digestive obstruction were excluded. All patients were treated as inpatients and they were prepared for possible laparotomy after getting informed consent. After taking permission from the hospital ethics committee and informed written consent from parents, the patients underwent laparotomy version 10. Descriptive statistics was used to calculate mean and standard deviation for numerical variables i.e. age. Frequency and percentages were presented for qualitative variables i.e. gender. Graphs and tables were used for data presentation. The nominal variables were reported as frequency and percentages. The numerical data was reported as means and standard deviation. The data was entered and analyzed through SPSS-20.

RESULTS

The mean age of the patients were 8.6±20.3 months ranging from 0.02 to 96 months. There was slight dominance by male gender in our study. The studied patients were male (58.0%) while remaining (42.0%) were females. In our study some of the patients (60.0%) were included in the trial through the emergency department while (40.0%) were selected in the study through the outpatient department. The main presenting features of the patients were analyzed. Out of the total 50 study cases, 25 (50.0%) were having pain at presentation, 44 (88.0%) had vomiting while all the patients (100.0%) had abdominal distension (Table 1)

As per objective we measured the presentation of the patients according to different age categories. It was found out that in neonatal age group, 10 (31.3%) patients had pain, 32 (100%) had vomiting and 28 (87.5%) had abdominal distension. In the patients under 1 year of age, 7 (70%) were having pain, 9 (90%) had vomiting and 7 (70%) had abdominal distension. Of the patients between 12 to 59 months, 6 (100%) had pain 1 (16.7%) had vomiting and 4 (66.7%) had abdominal distension. Similarly, of the patients of 60 months or above age, 2 (100.0%) each had pain and vomiting while abdominal distention was present in none of these cases (Table 2).

Table 1: Baseline details of all the patients

Variable	No.	%
Mean age (years)	8.6±20.3	
Gender		
Male	29	58.0
Female	21	42.0
Clinical presentation		
Pain	25	50.0
Vomiting	44	88.0
Abdominal distension	50	100.0
Mode of admission		
Emergency	30	60.0
Outpatient	20	40.0

In the current study x-ray, ultrasound and barium enema were done find out the probable cause of abdominal distension. We found out that 31 (62.0%) of patients each had dilated gut loop and air fluid level on x-ray. The Doppler ultrasound found out reverse pattern of superior mesenteric artery and vein in 27 (54.0%) cases. In 17

(34.0%) of the cases the barium enema found normal location of duodenojejunal junction and c-shape of duodenum (Table 3). The management of the study patients was surgical Ladd procedure in all 50 (100.0%) cases (Table 4). One (2.0%) patient each in the study had small gut adhesions, numerous worms and Meckel's diverticulum per operatively. Congenital band other findings were showed in Table 5.

Table 2: Presentation of the study patients according to age (months)

Variables	0-1 (n=32)	1-11 (n=10)	12-59 (n=6)	>60 (n=2)
Pain	10 (31.3%)	7 (70%)	6 (100%)	2 (100%)
Vomiting	32 (100.0%)	9 (90.0%)	1 (16.7%)	2 (100.0%)
Abdominal distension	28 (87.5%)	7 (70.0%)	4 (66.7%)	-

Table 3: Radiological and other investigational findings of the study patients (n = 50)

Variable	No.	%
X-ray		
Dilated gut loop	31	62.0
Air fluid level	31	62.0
Doppler U/S		
Reverse pattern of superior mesenteric artery and vein	27	54.0
Normal location of duodenojejunal junction & C-shape of duodenum	17	34

Table 4: Management of the study patients (n = 50)

Variable	No.	%
Surgical Ladd procedure	50	100.0
Laparoscopic method	-	-

Table 5: Congenital bands Other findings (n = 50)

Variable	No.	%
Ladd band	22	44.0
Malrotation of gut	21	42.0
Mid gut volvulus	20	40.0
Congenital bands	11	22.0
Caecum in left iliac fossa	9	18.0
ARM	4	8.0
Pseudo bands	2	4.0
Short mesentery	2	4.0
Dilated deudenum	2	4.0
Complete malrotation	2	4.0
Perforated duodenojejunal junction	2	4.0
Small gut adhesions	1	2.0
Numerous worms obstruction	1	2.0
Meckel's	1	2.0

DISCUSSION

Malrotation the incomplete or abnormal rotation of the GI tract about the superior mesenteric artery is a common condition with diverse outcomes. Bilious emesis is its most common presenting sign.¹⁶ Majority of the intestinal malrotation cases 50% to 70% are diagnosed during the neonatal period. While a significant proportion of cases present beyond the neonatal period, most present in infancy but up to 20% become symptomatic after the first year of life. Keeping in mind the complications and severity

associated with this anomaly we planned to carry out a prospective cross sectional study to find out the spectrum of intestinal malrotation cases presenting at our hospital. A total of 50 cases were enrolled in the current study. In our study the mean age of patients was 8.6 ± 20.3 months ranging from 0.02 to 96 months. Majority of our cases (64.0%) were below 1 month of age while a small proportion 4% were above 5 years. In the studies by Millar AJW and colleagues it was noted that almost 60% patients presented in neonatal age while a substantial proportion (20.0%) presented after 1 year of age.¹⁶

A similar trend was noted by Filston and Kirks.⁹ These findings are similar to our results, however, 2 of our cases presented late after 5 years of age which could be the cause of deviating the average of study patients. In the current study male gender was in dominance with 58% proportion. This is in continuation with other studies on this topic. A study by Moldrem AW and colleagues found out that almost 55% of their cases were male¹⁷. In the studies by Millar and colleagues⁹ and Filston et al¹⁶ as well the male gender was in dominance. Others have also reported that male infants are more frequently affected than the female infants.¹⁸

All the patients (100%) presented with abdominal distension in our study. Moldrem et al¹⁷ reported that 40% of their cases had abdominal distension. In another study by Nahvi and Khorangami¹⁸ it was found out that only 18% of their patients had abdominal distension. Compared to these studies our results show high incidence of abdominal distension. Similarly, in our study (50%) patients had abdominal pain. A comparable finding had been reported by Moldrem and colleagues¹⁷ where they also found out that (50.0%) patients presented with acute abdominal pain. A similar trend was reported by Nahvi and colleagues¹⁸ where they found out that (54%) of their patients had abdominal pain.

Majority of our patients presented with vomiting (88.0%). A similar trend was seen by Nahvi and colleagues¹⁸ where they saw that (100%) of their patients presented with bilious vomiting. In the study by Moldrem and colleagues¹⁷ one of the most frequent presenting complaints was nausea and vomiting.

The presentation of intestinal malrotation patients was measured according to age. It was found out that in neonatal age group very few cases had pain compared older ages. Vomiting was a frequent clinical indicator along with abdominal distension in the neonatal age group. In the patients up to 1 year vomiting was more frequent followed by pain and abdominal distension. In 1 to 5 years age group (100.0%) patients had pain while abdominal distension was quite frequent with vomiting in very few. In the current study patients above 5 years of age had pain and vomiting but no abdominal distension.

We evaluated our study patients with x-ray and ultrasound. On x-ray (62%) of our patients had dilated gut loop and air fluid level while on ultrasound reverse pattern of superior mesenteric artery and vein was found in (54%) patients. Abdominal radiograph may show a dilated duodenum with a fluid level and some gas in the distal bowel; however, it may be interpreted as normal in 20% of cases¹⁹. Similarly, modern ultrasound examination is very helpful in the diagnosis of intestinal malrotation. The

position of the superior mesenteric vein left to artery suggests malrotation in almost all the cases.

Abdominal radiographs may be normal in appearance or may show distention of the stomach and proximal duodenum by air, with little distal bowel gas. The sensitivity of the upper GI series for the diagnosis of malrotation has been reported as 93% and 100%, but a sensitivity of only 54% was reported for the diagnosis of midgut volvulus²⁰.

In the current study on barium enema (34%) cases had normal location of duodenojejunal junction and c-shape of duodenum. The most frequent per operative findings in the current study were Ladd bands in (44%) cases, malrotation of gut (42%), mid gut volvulus (40%), congenital bands (22%) and caecum in iliac fossa in (18%) of the cases. The surgical Ladd procedure was used in all the study cases (100.0%) for the management of intestinal malrotation in the current. Ladd's procedure has been described as choice of treatment for malrotation. There are found components to this operation namely; lysis of Ladd's bands, widening of the root of the mesentery, replacement of bowel into the abdomen and appendectomy.

Von Flue and colleagues²¹ reported about 10 cases with malrotation. Of their 10 cases, 8 were symptomatic and 6 (80%) were managed with Ladd's procedure. In another study by Maxson et al who reported 22 patients of (> 2 year) paediatric population in which Ladd's procedure was successfully performed.²²

The current study is one of the very few trials done in the local as well as national level in recent times. The information regarding the spectrum of intestinal malrotation will help the paediatric surgeons to understand the presenting sign and symptoms and probable investigational specially, radiological and barium enema findings. Timely diagnosis of this anomaly and prompt and appropriate surgical management leads to less morbidity and mortality.

CONCLUSION

On the basis of findings, we conclude that intestinal malrotation presents in early life within one month of life in two third of cases, however, a substantial number of cases can present in infancy and few even present later in life. Male gender was dominant, which validates the previous reports on this topic. Abdominal distension, pain and vomiting were the most frequent presenting complaints in our patients. Keeping in mind the complications and severe morbidity associated with intestinal malrotation it can be suggested that it is imperative to timely diagnose and promptly manage this anomaly so that resultant morbidity and mortality may be prevented.

REFERENCES

1. Torres AM, Ziegler MM. Malrotation of the intestine. *World J Surg* 1993; 17:326-31.
2. Zissin R, Rathaus V, Oscadchy A. Intestinal malrotation as an incident finding on CT in adult. *Abdom imaging* 1999; 24:550-5
3. Marcson RT, Franklin PA, Wagner CW. Malrotation in the older child: Surgical management, treatment and outcome. *Am Surg* 1995;61:135-8
4. Warner B. Malrotation. In: *Surgery of infants and children; Scientific principles and practice*. Oldham KT, Colombani PM, Foglia RP (Eds), Lippincott Williams and Wilkins, Philadelphia. 1997; p1220.

5. Dilley AV, Pereira J, Shi Ec. The radiologist says malrotation: does the surgeon operate? *Pediatr Surg Int* 2000; 16:45.
6. Berseth CL. Disorders of the intestines and pancreas. In: *Avery's Disease of the Newborn*, 7th ed. Taeusch WH, Ballard RA (Eds). WB Sanders, Philadelphia 1998, p568-85.
7. Stewart DR, Colodny AL, Daggett WC. Malrotation of the Bowel in infants and children - A 15 year review. *Surgery* 1976; 79: 716.
8. Bass KD, Rothenberg SS, Chang JH. Laparoscopic Ladd procedure in infants with malrotation. *J Pediatr Surg* 1998; 33:279.
9. Filston HC, Kirts DR. Malrotation the ubiquitous anomaly. *J Pediatr Surg* 1981; 16:164.
10. Ford EG, Senac MO Jr, Srikanth MS, Weitzman JJ. Malrotation of the intestine in children. *Ann Surg* 1992; 215:172.
11. Smith EI. Malrotation of the intestine. In: *Pediatric Surgery*, 4th ed. Welch KJ, Randolph JG, RauichMR(eds), Year Book Medline Publisher, Chicago 1986; p882.
12. Glover DM, Barry FM. Intestinal obstruction in the newborn. *Ann Surg* 1949; 130:148.
13. Uba AF, Chirdan LB, Edino ST. Intestinal malrotation: Presentation in the older child. *Niger J Med* 2005; 14(1): 23-6.
14. Applegate KE, Anderson JM, Klatte EC. Intestinal malrotation in children: a problem-solving approach to the upper gastrointestinal series. *Radiographics* 2006;26(5): 1485-500.
15. Oh SK, Han BK, Leuin TL. Gastric Volvulus in children: the twist and turns of an unusual entity. *PediatrRadiol* 2008; 38: 297-304
16. Moldrem AW, Papaconstantinou H, Broker H, Megison: Malrotation in Pediatrics. 2008;34:1425-32.
17. Millar AJW, Rode H, Brown RA, Cywes S. The deadly vomit: malrotation and midgut volvulus. *Pediatr Surg Int* 1987;2:172-6.
18. Nahvi, H & Khorgami, Zhamak. (2012). Midgut malrotation in older children. *Acta Medica Iranica*. 46. 133-136.
19. Aslanabadi S, Ghalehgolab-Behbahan A, Jamshidi M, et al. Intestinal malrotations: a review and report of thirty cases. *Folia Morphol (Warsz)* 2007; 66:277.
20. Taghavi M, Alamdaran S A, Feizi A. Diagnostic Value of Ultrasound and Gastrointestinal Series Findings in Detection of Pediatric Intestinal Malrotation, *Iran J Radiol*. 2018 ; 15(2):e15089.
21. Von Flue M, Herzog U, Ackermann C, et al: Acute and chronic presentation of intestinal nonrotation in adult. *Dis Colon Rectum*. 1994, 37: 192-198. 10.1007/BF02047549.
22. Maxson RT, Franklin PA, Wagner CW. Malrotation in the older child: surgical management, treatment, and outcome. *The American Surgeon*. 1995 Feb;61(2):135-138.