Original Research Article

DOI: http://dx.doi.org/10.18203/2349-2902.isj20184076

Malrotation presenting beyond infancy: a clinical study

Anil Balraj T., S. Srinivas*, K. Ramesh Reddy, Manisha Reddy G., Anusiri Inugala

Department of Paediatric Surgery, Niloufer hospital, Hyderabad, Telangana, India

Received: 09 September 2018 **Accepted:** 14 September 2018

*Correspondence: Dr. S. Srinivas.

E-mail: shantisrinivas@gmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Background: Intestinal malrotation is a congenital anomaly that results from abnormal or incomplete rotation and fixation of midgut during embryonic development. This study was done at Niloufer Hospital and Institute for Women and Child Health, Hyderabad with an aim of evaluating the clinical presentation, diagnosis, management and outcome of malrotation presenting beyond 1 year of life

Methods: Overall 50 cases of malrotation presented to the department of pediatric surgery over a period of 2 year (April 2015 to April 2017). 12 patients of these 50 (12/50) whose age was above 1 year i.e. who were beyond infancy were studied in this study

Results: Out of the 12 patients, 6 were female and 6 were male. Age of patients ranged from 15 months to 13 years. Younger patients below 2 years commonly presented with typical symptoms of bilious vomiting and patients above 2 years of age presented with more varied and vague symptoms. 1/12 of patients presented with a poor general condition and succumbed during the course of treatment

Conclusion: Small intestinal obstruction due to malrotation is relatively common in neonatal period. Malrotation beyond infancy is an uncommon diagnosis. Malrotation in grown up children usually is not suspected due to varied symptoms. High index of suspicion is needed as early intervention and treatment in the form of Ladds procedure can prevent any catastrophic events like volvulus and bowel ischemia.

Keywords: Malrotation, Volvulus

INTRODUCTION

Intestinal malrotation is a congenital anomaly that results from abnormal or incomplete rotation and fixation of midgut during embryonic development. Anomalies of intestinal rotation may present as spectrum of abnormalities ranging from normal rotation to typical malrotation to complete non-rotation. The incidence of intestinal malrotation is about 1 in 500 live births.^{1,2} About 75-85% of these patients are diagnosed in early infancy before one year of age whereas the diagnosis of rest can be delayed to childhood or even adulthood.³ The real incidence of mal-rotation is not possible to estimate in the older population as most children do not present

with any symptoms throughout their lives.⁴ Malrotation presenting beyond infancy may also be diagnosed as an operative surprise at laparotomy.

Sudden onset of bilious vomiting in a previously healthy infant is the most common presentation of intestinal malrotation.⁵ Diagnosis of malrotation is relatively easy in neonatal period because most of the new-borns present with bilious vomiting. The presentation of malrotation beyond infancy may vary from life threatening acute ischemic mid-gut volvulus to nonspecific abdominal complaints like non-specific pain, non-bilious vomiting and failure to thrive. In most of these children abdominal symptoms are labelled as gastrointestinal allergies, milk

intolerance and even as abdominal tuberculosis. Unusual presentations in the older children and limited awareness of the condition as a clinical entity after infancy, may lead to delay in diagnosis and treatment. The objective of this retrospective study is to increase the awareness about malrotation as a cause of intestinal obstruction in children beyond infancy and focus on various modes of presentation of malrotation occurring beyond 1st year of life and its management.

METHODS

This retrospective study of small intestinal obstruction in children above 1 year due to malrotation was conducted in the Department of Pediatric Surgery Niloufer Hospital Institute for Women and Child health, Hyderabad, Telangana, India.

It included patients older than 1 year treated with an established diagnosis of intestinal malrotation. Data included patients treated from April 2015-2017 (2 years). Diagnosis was made by clinical symptoms, radiological investigations and findings of malrotation were confirmed by surgery. Radiological diagnosis was inferred on upper GI contrast when there was gastric dilatation, a lack of duodenal C-loop and the duonenojejunal junction had not crossed the midline and/or not ascended to the level of pylorus. Malrotation was defined as "typical" if the duodeno-jejunal flexure is to the right of mid-line and "atypical" when it was on the left side but not ascended to the level of pylorus. After proper resuscitation, all these patients were subjected to exploratory laparotomy and Ladds procedure. Surgical findings of malrotation included presence of mid-gut volvulus, ligament of treitz at abnormal position, presence of ladds bands and presence of narrow mesentry root. The steps of Ladds procedure included, derotation of volvulus if present, release of ladds bands, straightening of the duodenum, widening of duodenocolic mesentry, ruling out intrinsic duodenal obstruction, appendectomy and placing the bowel in position of nonrotation.

A total of 50 patients were managed for malrotation during the period of study. Of these 12 (12/50) patients who were older than 1year were studied. Data was obtained from patient's medical records and included information about age at presentation, sex, presenting symptoms, time of diagnosis, radiological investigations performed, intra-operative findings, complications and post-operative follow-up. The age of presentation was then correlated with patient's presentation and intra-operative findings.

RESULTS

Clinical presentation

A total of 50 patients were treated in our institute for small intestinal obstruction due to malrotation of whom

12/50 (24%) were more than 1 year of age. 3/12 patients were less than 2 years and rest of 9 were more than 2 years (Figure 1).

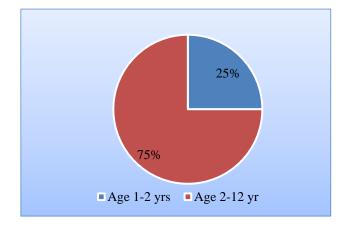


Figure 1: Age distribution.

Compared to neonates older patients whose age was more than 1 year presented with a spectrum of varied symptoms which ranged from typical to atypical. 3/12 patients who presented within the 2 year of life typically presented with bilious vomiting. An analysis of presenting symptoms in patients younger and older than 2 years at time of diagnosis revealed a significant increase in atypical symptoms in the older subset of patients (Figure 1).

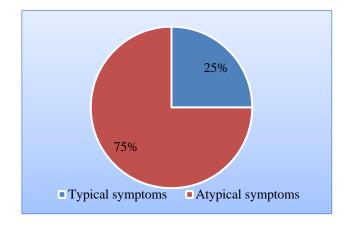


Figure 2: Incidence of typical versus atypical symptoms.

Atypical symptoms in our study included recurrent colicky abdominal pain, non-bilious vomiting, and failure to thrive/weight loss, early satiety and abdominal bloating. One patient was treated with ATT for suspected diagnosis of abdominal tuberculosis. Patient did not show any response to ATT. Upper GI contrast in this patient was suggestive of malrotation.

Radiologic investigations

Plain abdominal radiographs showed the features of partial duodenal obstruction in 7 patients (Figure 3). 1

patient had multiple air fluid levels, suggesting acute intestinal obstruction. Plain X-ray abdomen in rest of the patients (4/12) was inconclusive.



Figure 3: Plain X-ray abdomen suggestive of partial duodenal obstruction.

Upper gastrointestinal contrast series was diagnostic in all cases (Figure 4).

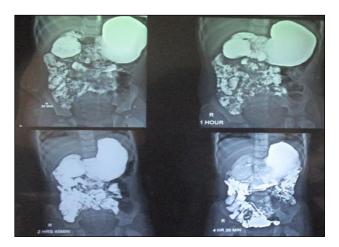


Figure 4: Upper GI series suggestive of malrotation.

Surgical management

The diagnosis of malrotation was confirmed at laparotomy in all 12 cases. All patients underwent a standard Ladds procedure with de-rotation of bowel, division of ladds bands, broadening of mesentry, Appendectomy, placement of bowel in a non-rotated orientation. Midgut volvulus was seen in 4/12 patients (Figure 5). Vascular compromise gangrene of bowel was seen in 1 patient. 8 patients showed presence of lacteals, thickened bowel wall, mesenteric nodes suggesting the chronic nature of obstruction. In one patient in whom

abdominal TB was suspected initially mesenteric lymph node biopsy was done and TB was ruled out on histopathology.

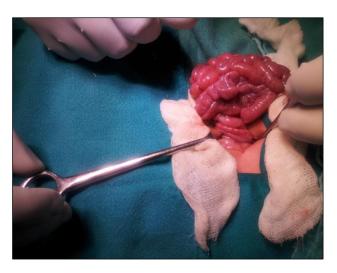


Figure 5: Malrotation with midgut volvulus.

Post-operative period

The post-operative period was un-eventful in 11 patients, 1 patient who had presented with acute intestinal obstruction and underwent extensive resection anastomosis of small intestine died postoperatively owing to the poor general condition, bad nutritional status and delayed presentation. The overall mortality noted in the present study was 1/12 (8.2%).

DISCUSSION

Small intestinal obstruction due to malrotation is a relatively common diagnosis. Malrotation is uncommon after 1 year of age in the present study 12/50 cases of malrotation were diagnosed after 1 year of age. Small bowel obstruction due to malrotation presenting after 1 year of age is associated with varied symptoms, which are often non-specific and are associated with delay in diagnosis. In its typical form which commonly is seen in younger children it can be suspected and diagnosed easily, however the symptomatology in older children is less dramatic and atypical thus delaying the diagnosis. Such presentations in children older than 1 year consists mainly of atypical symptoms such as failure to thrive, weight loss, non-bilious vomiting, chronic colicky abdominal pain, abdominal bloating caused by intermittent volvulus and obstructing bands. Present study corroborates with Penco et al; Durkin et al and N.G Nagdev et al which suggest that chronic symptoms are more common in children older than 2yrs of age. 5,6,4

The diagnosis of malrotation is difficult after neonatal period because of the wider and more obscure constellation of clinical symptoms in older patients. In this study patients had classical presentation of sudden onset of bilious vomiting and upper abdominal distension

within 2nd year of life. Those patients who present with acute duodenal obstruction are diagnosed rapidly, and those who presented with vague abdominal symptoms, diagnosis was delayed upto years.8,9 The older patients presented more commonly with atypical symptoms and diagnosis was delayed for even about 6 years owing to the non-specific nature of the symptoms. 1 patient in the present study was being treated empirically with antituberculous therapy for abdominal kochs however he was diagnosed with malrotation which was confirmed at surgery. There are reports of malrotation in adults even being treated as pancreatitis. 10,11 Another well described presentation is mal-absorption pattern associated with diarrhea, nutritional deficiencies and failure to thrive caused by bowel lymphedema resulting from lymphatic obstruction due to chronic volvulus resulting in loss of proteins from bowel lumen.^{8,12,13,10} It is important to note that although many patients diagnosed as having malrotation beyond the neonatal period may be asymptomatic, they can still present with intestinal obstruction with or without strangulation at any age. 14,15 High morbidity is associated with both acute and chronic volvulus, thus surgical correction of documented malrotation is the treatment of choice.

CONCLUSION

Thus, we conclude that malrotation beyond infancy is an uncommon diagnosis which presents atypically. It should be suspected even with the most vauge abdominal symptoms and upper GI contrast has to be done to exclude it. If proven to be malrotation then surgical management with Ladds procedure is the treatment of choice owing to the risk of volvulus which can occur at any age.

Funding: No funding sources Conflict of interest: None declared

Ethical approval: The study was approved by the

Institutional Ethics Committee

REFERENCES

- 1. Filston HC, Kriks DR. Malrotation: the ubiquitous anamoly. J Ped Surg. 1981;16(4):614-20.
- 2. Torres AM, Ziegler MM. Malrotation of the intestine. World J Surg. 1993;17(3):326-31.

- 3. Andrassy RJ, Mahour GH. Malrotation of the midgut in infants and children. Arch surg. 1891;16:158-60.
- 4. Nagdeva NG, Qureshi AM, Bhingare PD, Shinde SK. Malrotation beyond infancy. J Ped Surg. 2012;47:2026-32.
- 5. Penco JM, Murrillo JC, Hernandez A, et al. Anamolis of intestinal rotation and fixation, consequences of late diagnosis beyond 2yrs of age, Pediatr Surg Int. 2007;23:723-30
- 6. Durkin ET, Lund DP, Shaabam AF. Age related differences in diagnosis and morbidity of intestinal malrotation. J Am Coll Surg. 2008;206:658-63.
- 7. Spigland N, Brandt ML, Yazbeck S. Malrotation presenting beyond the neonatal period. J Pediatr Surg. 1990;25:1139-42.
- 8. Brandt ML, Pokarny WI, Mc Gill CW. Late presentations of midgut malrotation in children. Am J Surg. 1985;150:767-71.
- Kullendorf CM, Mikaelsson C, Ivaneev K. Malrotation in children with symptoms of gastrointestinal allergy and psychosomatic abdominal pain. Acta pediatr Scand. 1982;74:469-72.
- Sasaki T, Soh H, Kimura T. Recurrence acute pancreatitis caused by malrotation of the intestine and effective treatment with laparoscopic ladds procedure. Paed Surg Int. 2005;21:994-6.
- 11. Kirby CP, Freeman JK, Ford WDA. Malrotation with recurrent volvulus presenting with cholestasis. Paed surg Int. 2000;16:130-1.
- 12. Firor HV, Steiger E. Morbidity of rotational abnormalities of gut beyond infancy. Clece Clin Quarterly. 1983;50:303-9.
- 13. Howell CG, Vozza F, Shaw S. Malrotation, malnutrition and ischemic bowel disease. J Pediatr Surg. 1982;17:469-72.
- 14. Fukaya T, Brown BP, Lu CC. Midgut volvulus as acomplication of intestinal malrotation in adults. Dig Dis Sci. 1993;38:438-44.
- 15. Rowson JT, Sullivan SN, Girvan DP. Midgut volvulus in the adult, a complication of intestinal malrotation. J Clin Gastroenterol. 1987;9:212-6.

Cite this article as: Balraj AT, Srinivas S, Reddy KR, Reddy MG, Inugala A. Malrotation presenting beyond infancy: a clinical study. Int Surg J 2018;5:3288-91.