

Malrotation of Gut in Children: Initial Experience from a New Pediatric Surgery Facility

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ABSTRACT

Introduction: Malrotation of gut is a defect in the embryonic rotation and fixation of intestine during first trimester of pregnancy. This results in a spectrum of anatomical defects leading to varied clinical presentation ranging from episodes of proximal bowel obstruction to catastrophic midgut volvulus with gangrene. This paper highlights our experience with clinical profile, diagnosis and management in such patients.

Material and methods: The retrospective study was carried out in the Department of Pediatric Surgery, Superspecialty Hospital Government Medical College Srinagar, India. Data of all the patients admitted with the diagnosis of intestinal malrotation over last 5 years was analyzed. The parameters studied included age and sex of patients at the time of presentation, signs and symptoms, investigations, management and final outcome.

Results: A total of 128 patients of intestinal malrotation were admitted during the study period. There were 79 males and 49 females. The age at presentation ranged from newborn to 14 year. Most of the patients of intestinal malrotation presented in the early neonatal period. Classical Ladd's procedure was performed in 96 (21.09%) patients and was curative. The rest of the patients either needed resection and anastomosis or exteriorization of the bowel. The most common post-operative complication was ileus.

Conclusion: It is vital to interpret the symptoms correctly so that early diagnosis is made and the corrective surgery is performed in time and fatal complications like mid gut volvulus are prevented. A high index of suspicion followed by upper GI contrast is essential to diagnose patients of malrotation in older children with atypical presentation. Laparoscopic Ladd's procedure is associated with more complications than open, especially in infants.

Keywords: Malrotation; Ladd's procedure; Midgut volvulus; Bowel obstruction

INTRODUCTION

Rotation and fixation of gut occurs in a typical fashion during fetal development.^{1,2} Any deviation from the normal can result in a spectrum of anatomical abnormalities giving rise to various clinical presentations ranging from chronic subacute high intestinal obstruction to the dreadful midgut volvulus with gangrene. Though the true incidence of malrotation is reported to be between 1 in 200 to 1 in 500 live births, symptomatic malrotation occurs only in 1 in 6000 live births.³⁻⁵ The more common acute and severe presentations predominate in neonates and infants, while chronic and intermittent complaints are frequently encountered in bigger children and adults. The surgical management has not

changed much since it was first described by Ladd in 1932.⁶ We aim to present our experience with the presentation and management of patients with malrotation over last 5 years at a new facility in our institution.

MATERIAL AND METHODS

This is a retrospective study involving the analysis of all the patients operated, with an informed consent from the parents or guardians, for intestinal malrotation over a period of last 5 years from December 2015 through November 2020. The study was conducted in the Department of Pediatric Surgery, Superspecialty Hospital, Government Medical College Srinagar, India with an approval from the institutional Ethics Committee. The cohort included all the cases diagnosed in the department or referred from the department of pediatrics or other hospitals with a prior diagnosis. Exclusion criteria included: a) Re-explorations for the incomplete previous surgery or its complications and b) cases of malrotation associated with other anomalies like diaphragmatic hernia or eventration, situs inversus etc. The data sheets of all the patients were revisited and information was collected regarding the following: a) demographic profile (age, sex, weight, height and similar history in the family, if any); b) clinical presentation (signs and symptoms); c) diagnosis (clinical examination, investigative modalities used); d) details of surgery (approach like open or laparoscopic, operative findings, type and extent of surgical procedure, intra-operative complications); e) hospital stay e) final outcome, including early or late postoperative complications and mortality and f) post-operative follow-up.

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RESULTS

A total of 128 patients were admitted and underwent operation for intestinal rotational anomalies from December 2015 to November 2020 in our department. The results of this study are shown in the tables (Tables 1 to 5).

About two-third of patients presented in the first year of life, while more than half presented in the neonatal period. Males constituted 61.7% (79, n=128) while females constituted 38.3% (49, n=128). The most common presentation was vomiting (bilious or non-bilious), present in about 3/4th of patients (94/128). Bilious vomiting was the predominant symptom in neonates and infants while non-bilious vomiting was more common in older children. Abdominal pain was the second most important complaint. Forty-two patients (32.8%, n=128) had acute abdomen with 27 infants (21.09%) having signs of bowel ischemia like blood in stools, tender abdomen and abdominal wall erythema. Chronic or recurrent abdominal pain was found in 18% of patients.

In 101 (79.0%) of the patients, classical Ladd's procedure was performed. Open approach was opted in most of the cases due to minimal expertise available for laparoscopic Ladd's procedure, although diagnostic laparoscopy was performed as and when needed to confirm the diagnosis. In those situations where irreversible bowel ischemia was present, resection of gangrenous bowel with primary anastomosis or exteriorization of bowel as double barrel stoma (where there was gross contamination of peritoneal cavity) was performed. Postoperative management included

Age group (Months)	No of patients	Percentage (%)
0-1	51	39.8
1-12	35	27.3
12-60	21	16.4
60-120	15	11.8
>120	06	4.7
Total	128	100

Table-1: Age of presentation

Male	79	61.7%
Female	49	38.3%

Table-2: Sex distribution

Clinical feature	N	%
Vomiting	94	73.4
Acute abdominal pain	42	32.8
Chronic abdominal pain	23	18.0
Failure to thrive + Nonspecific complaints	11	8.6
Acute abdomen with signs of ischemia	27	21.1

Table-3: Clinical presentation

Procedure	N	%
Ladd's procedure	101	78.9
Resection with primary anastomosis	12	9.3
Resection with double barrel stoma	15	11.7

Table-4: Operative procedures

Complication	N	%
Mortality	5	3.9
Wound infection	12	9.4
Delayed peristalsis	18	14.0
Adhesion obstruction	08	6.25
Incisional hernia	01	0.8

Table-5: Complications

intravenous fluids, intravenous antibiotics and passive Ryle's tube suction. Injectable antibiotics were given for 3 to 5 days which was followed by oral antibiotics. Total duration of antibiotic therapy in post-operative period was 7-10 days. Ryle's tube was removed once the aspirates became nonbilious and bowel sounds were heard (average 3 days). Oral feeds were initiated after removal of Ryle's tube, starting with liquid diet. Before the time of discharge all patients were on normal diet.

In this series, 5 patients died (5.5%) which included 3 neonates with midgut volvulus with intestinal gangrene with shock; and 2 infants who died one each of apnea and sudden cardiopulmonary arrest in the postoperative period. Other main postoperative complications included prolonged ileus (14%) and wound infection (9.4%). The most common long-term complication was adhesion obstruction (6.25%), which needed exploratory laparotomy and adhesiolysis in all of them.

DISCUSSION

The malrotation of gut is a surgical condition mostly seen in neonates but can also present in older children. It includes a wide range of rotational and fixation anomalies ranging from small bands obstructing the duodenum to complete non-fixation of intestine in the peritoneal cavity. The exact incidence of the disease is difficult to determine as many patients are asymptomatic. Classically the diagnosis of intestinal rotational abnormalities is primarily by ultrasonography and upper GI contrast study. In neonates and infants, given the classical clinical presentation aided by ultrasound and/or UGI contrast x-rays, diagnosis is more or less straightforward. The real challenge is diagnosing older children with atypical features. Furthermore, incidental diagnosis in asymptomatic children is not uncommon. UGI contrast study remains the most important tool to diagnose such cases.⁷ The management of asymptomatic patients with diagnosis of malrotation is not clear.⁸ But, surgical correction seems to be a more valid option for two reasons – (a) risk of volvulus and its sequelae, (b) atypical clinical features as the age advances which in turn may render diagnosis inconspicuous. Midgut volvulus is the most worrisome complication of the disease and can lead to infarction of variable length of small intestine if not treated urgently. Though more common in infants with malrotation, volvulus should be kept in mind even in older children with atypical clinical features like failure to thrive, feed intolerance, recurrent abdominal pain and hospitalizations. A high clinical suspicion followed by diagnostic imaging can result in timely diagnosis and surgical management.⁹ Ultrasonography is the

initial screening test while UGI contrast study is the gold standard in diagnosis of malrotation. The sensitivity of upper GI contrast in diagnosing malrotation is around 85%.¹⁰ The role of Contrast Enhanced Computed Tomography (CECT) has been described in literature especially in older children and adults.¹¹ In our series, 23 patients had CECT done; 15 were referral cases wherein it was done beforehand. In the rest 8 cases, CECT was done for non-specific recurrent abdominal pain and malrotation was an incidental finding. The standard surgical technique for malrotation is Ladd's procedure with appendectomy. Classical approach was an open exploration via right transverse incision on the abdomen. Laparoscopic Ladd's procedure has been in vogue in the recent past, particularly in stable and older children. The disadvantages of laparoscopic surgery are long learning curve, high conversion rate, safety concerns in neonates and low threshold to conversion if malrotation is associated with volvulus.^{12,13} We tried laparoscopic approach in 8 patients; out of which 5 needed conversion and 1 needed redo on seventh postoperative day due to persistence of obstructive symptoms. In cases of malrotation with volvulus where intestinal gangrene is present, resection of gangrenous bowel and primary anastomosis or exteriorization of bowel is done depending on the condition of patient, bowel and contamination of peritoneal cavity.

CONCLUSION

Early surgical correction should be considered in all cases of malrotation irrespective of symptoms and age at presentation. In older children with atypical features, upper GI contrast should be performed to confirm the diagnosis. In doubtful cases, surgical intervention, open or laparoscopic should not be delayed. Laparoscopic Ladd's procedure is associated with more complications and adverse outcome in infants and has a longer learning curve.

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