Malrotation beyond infancy☆,☆☆,★

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Pediatric;
Ladd procedure;
Atypical presentation

Abstract
Objective: The aim of this work was to study the various presentations of malrotation and management in patients older than 1 year.
Materials and Methods: Medical records of patients operated on over the last 6 years who were older than 1 year with a diagnosis of intestinal malrotation were evaluated retrospectively. Data about age at presentation, sex, presenting symptoms, time to diagnosis, radiographic imaging performed, surgical intervention, complications, and postoperative follow-up were collected and evaluated.
Results: The study population included 35 children and 3 adults. About three-fourths of pediatric patients were younger than 5 years, and about half of these presented in the second year of life. All patients who presented in the second year of life had a classical presentation of malrotation. Older patients presented more commonly with atypical symptoms. Of these, 5 older children were previously treated for suspected diagnosis of abdominal tuberculosis. Two patients were referred for acute pancreatitis and 1 for severe gastroesophageal reflux disease. Three adults presented with acute small intestinal obstruction and were diagnosed to have malrotation intraoperatively.

Upper gastrointestinal contrast study was diagnostic of malrotation in all pediatric patients. Ultrasound and color Doppler study of the abdomen revealed abnormal relationship of the superior mesenteric artery and vein in about one-third of the patients. All patients underwent a standard Ladd procedure. Midgut volvulus was present in about one-fourth of patients. Forty percent of patients with atypical presentation had persistence of preoperative symptoms postoperatively. Two adults developed complications postoperatively. There was no mortality in the present study.

Conclusion: Malrotation should be suspected in all patients with varied acute or chronic abdominal symptoms, and the upper gastrointestinal contrast study should be conducted. If the existence of typical malrotation is confirmed, surgical correction is mandatory to avoid volvulus and intestinal obstruction independently of the patient’s age.

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Contributors: N.G.N. has planned the study, managed the patients, and finalized the manuscript and will stand as guarantor. P.D.B., A.M.Q., and S.K.B. helped in the management of patients and in the preparation of the manuscript.

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Intestinal malrotation is a congenital anomaly that results from abnormal or incomplete rotation and fixation of the midgut during embryonic development. It consists of a spectrum of abnormalities ranging from normal rotation to typical malrotation to complete nonrotation. The condition occurs at a rate of about 1 in 500 live births [1,2]. About 75% to 85% of these patients are diagnosed during infancy [3], whereas the diagnosis in the rest can be delayed to childhood or even to adulthood. The true incidence of malrotation is difficult to estimate in the older population because most will remain asymptomatic throughout their lives [4]. Many are recognized only intraoperatively during other procedures or at autopsy [5].

Sudden onset of bilious vomiting in a previously healthy infant is the hallmark of intestinal malrotation. Nevertheless, the presentation may vary from life-threatening acute ischemic midgut volvulus to innocuous abdominal complaints like nonspecific pain and failure to thrive. Often, these symptoms are attributed to psychogenic causes, gastrointestinal allergies, or milk intolerance [6]. Incongruent presentations, especially in older patients, and limited awareness of the condition as a clinical entity after infancy, especially among primary clinicians, may lead to delay in diagnosis and treatment. Recently, Nehra and Goldstein [7] have stressed the need for an increased awareness of this entity and its varied presentations at different ages, in an attempt to reduce time to diagnosis leading to better outcome.

The objective of this retrospective study is to focus on varied modes of presentation of malrotation beyond infancy and its management.

1. Materials and methods

This retrospective data analysis of patients of malrotation was done in the Department of Surgery at Government Medical College and Hospital, Nagpur, India. It included patients older than 1 year with an established diagnosis of intestinal malrotation treated over a 6-year period, from February 2006 to January 2011. Diagnosis was established on clinicoradiologic grounds and confirmed by surgical findings. Radiologic diagnosis of malrotation was inverted when there was a lack of a duodenal C-sweep and the duodenojunal junction had not crossed the midline and/or not ascended to the level of the pylorus (Fig. 1). Malrotation was defined as “typical” if the duodenojunal junction was to the right of the midline and “atypical” when it was on the left side but not ascended to the level of pylorus. Surgical findings of malrotation included presence of midgut volvulus, ligament of Treitz at abnormal position, presence of Ladd bands, and presence of narrow mesenteric root (Fig. 2).

A total of 178 patients were managed for malrotation. Of these, 38 patients (21.3%) who were older than 1 year were studied. Data were obtained from the patients’ medical records and included information about age at presentation, sex, presenting symptoms, time to diagnosis, radiologic investigations performed, intraoperative details, complications, and postoperative follow-up. The age of presentation was then correlated with patients’ presentation and intraoperative findings (whether the malrotation was typical, atypical, or midgut volvulus). The data were compared by \( \chi^2 \) analysis and Mann-Whitney \( U \) test (nonnormalized data).

2. Results

2.1. Demography

The study population included 35 children and 3 adults (Table 1). About three-fourths of pediatric patients were younger than 5 years, and about half of these presented in the second year of life.
2.2. Clinical presentations

Table 2 shows various clinical presentations of the patients in this study. All patients who presented in the second year of life had a classical presentation with a sudden-onset bilious vomiting and upper abdominal distention. However, beyond that period, only about one-fourth of the patients had classical presentation. Statistical analysis of this data showed significant correlation ($P < .01$; Karl-Pearson correlation coefficient = −0.455).

A subgroup analysis of presenting symptoms in patients younger and older than 2 years at the time of diagnosis revealed a significant increase in chronic symptoms in the older subset. Furthermore, the atypical symptoms were significantly more common in patients older than 2 years ($P = .001$, highly significant). Atypical symptoms in our study group included recurrent colicky abdominal pain (61.9%), nonbilious vomiting (38.1%), failure to thrive/weight loss (33.3%), early satiety, and abdominal bloating. Five older children (age group, 6-12 years) had previously received antituberculous treatment (ATT) for suspected diagnosis of abdominal tuberculosis. Chest radiographs were unremarkable in all of these patients. One of these patients had documented positive Mantoux test result, and in the rest, ATT was started empirically. All these patients did not show any response to ATT. Upper gastrointestinal (UGI) contrast study in all these patients was suggestive of malrotation.

Two patients were referred to us with a provisional diagnosis of acute pancreatitis. These patients had severe abdominal pain, bilious vomiting, fever, and tenderness all over the abdomen. One of these patients had a similar episode 3 months back, which had been managed conservatively. In both patients, serum lipase was elevated (>180 U/L). These patients were managed conservatively. However, the bilious vomiting persisted. Because of discordance of clinical presentation, UGI contrast study was performed, which was diagnostic of intestinal malrotation. Magnetic resonance cholangiopancreatography in both these patients revealed normal, biliary, and pancreatic ductal anatomy. Another 2½-year-old patient presented for management of severe gastroesophageal reflux disease. He was found to have malrotation on UGI contrast study.

Three adults in our group presented with acute small intestinal obstruction and were diagnosed to have malrotation intraoperatively. None of these patients had any previous abdominal symptoms. There were no other associated congenital anomalies in any of these patients.

2.3. Radiologic investigations

Abdominal radiographs typically showed features of partial duodenal obstruction in half of the patients in our study. Most of these patients presented with classical features of malrotation. Three adults had multiple air-fluid levels. Plain x-ray abdomen in the remaining patients (42.1%) was inconclusive. The ultrasonography abdomen and color Doppler study was performed in all patients. It revealed abnormal relationship of the superior mesenteric artery and vein in 12 patients (31.5%). The classical whirlpool sign of midgut volvulus was present in 5 of these patients. Ultrasonography abdomen was inconclusive in adults because of bowel gas. Upper gastrointestinal contrast study was diagnostic of malrotation in all pediatric patients. The study diagnosed atypical malrotation in 5 patients, which was confirmed at laparotomy. Contrast study was not performed in adults.

<table>
<thead>
<tr>
<th>Initial presentation</th>
<th>Patients age</th>
<th>Total</th>
<th>$P$</th>
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<tr>
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<td>Pediatric patients &gt;2 y</td>
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<td>0</td>
</tr>
<tr>
<td>GERD</td>
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<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Acute small bowel obstruction</td>
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<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
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<td>18</td>
<td>3</td>
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</table>

HS, highly significant; NS, nonsignificant; GERD, gastroesophageal reflux disease.

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Patient demographics</th>
</tr>
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<tr>
<td>Demographic character</td>
<td>Pediatric patients up to 2 y (n= 17)</td>
</tr>
<tr>
<td>Age, mean ± SD</td>
<td>1.45 ± 0.44</td>
</tr>
<tr>
<td>Male (%)</td>
<td>10</td>
</tr>
<tr>
<td>Female (%)</td>
<td>7</td>
</tr>
</tbody>
</table>
2.4. Surgical management

The diagnosis of malrotation was confirmed at laparotomy. Table 3 illustrates intraoperative findings and its correlation with patients’ age. It can be noted that atypical malrotation was observed only in older children \( (P < .05) \), whereas midgut volvulus was seen uniformly at all ages \( (P = .169, \) not significant). All patients underwent a standard Ladd procedure with de-rotation of bowel, division of the Ladd bands, broadening of the mesentery, appendectomy, and placement of the bowel in a nonrotated orientation. Midgut volvulus was present in about one-fourth (9/38) of patients. However, none of these patients had a vascular compromise. These included 2 of the 3 adults. In addition, the adults had thick vascular Ladd bands and megaduodenum. All patients older than 2 years revealed dilated mesenteric vessels and thickened mesentery. The pancreas was found to be edematous at laparotomy in 2 patients with pancreatitis. However, there was no pseudocyst formation or pancreatic necrosis. In 5 patients who were previously diagnosed as having abdominal tuberculosis, mesenteric lymph node biopsy was done. Nissen fundoplication was added to the Ladd procedure in a patient who presented with gastroesophageal reflux. Correlation between clinical presentation of patients and intraoperative findings is given in Table 4.

2.4.1. Postoperative period

Patients with classical presentation required gastric decompression for a significantly longer period than did those with atypical presentation (mean: 5.59 ± 2.04 days [median: 6] vs mean: 3.94 ± 2.57 days [median: 2]; Mann-Whitney \( U \) test: \( z = 4.011, P = .001 \), highly significant). Duration for resumption of oral intake and time to discharge were also longer in patients with classical presentation (mean: 6.68 ± 1.84 days [median: 9] vs 5.81 ± 3.69 days [median: 5]; \( z = 3.278, P = .0010 \), highly significant). However, postoperative complications were not observed in the typical malrotation group. Persistence of symptoms after surgical correction was common with atypical presentation (seen in 4/10 patients). Of these, 2 patients each were included in the typical (2/10) and atypical malrotation groups (2/5). Follow-up UGI contrast study, however, was unremarkable. Four of the 5 patients who initially were misdiagnosed as having abdominal tuberculosis were relieved of their symptoms after surgery. The result of histopathologic examination of lymph nodes in all of these patients was negative for tuberculosis. One 12-year-old boy, however, had recurrent episodes of severe abdominal pain. Reexploration in this patient revealed small bowel adhesions and thick mesentery. This patient continued to have symptoms even after the second exploration and required long-term antispasmodic medications. Two patients with pancreatitis were asymptomatic for a mean period of 6 months. Two of the 3 adult patients developed complications, including 1 patient with duodenal obstruction, which was diagnosed on postoperative contrast study. On exploration, she was found to have a stricture at the second portion of the duodenum from dense scar tissue and required gastrojejunostomy. The other patient had postoperative adhesive intestinal obstruction and required adhesiolysis. There was no mortality in this study.

3. Discussion

Symptomatic malrotation appears to present with decreasing frequency after the first year of life; hence, “malrotation beyond infancy” is a rare diagnosis. In its

<table>
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<th>Initial presentation</th>
<th>Age of patients</th>
<th>Total</th>
<th>( P ) value</th>
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<tr>
<td></td>
<td>1-2 y</td>
<td>Pediatric patients &gt;2 y</td>
<td>Adults</td>
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<tr>
<td>Typical malrotation</td>
<td>13</td>
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<tr>
<td>Atypical malrotation</td>
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<td>0</td>
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<tr>
<td>Typical malrotation with volvulus</td>
<td>4</td>
<td>3</td>
<td>2</td>
</tr>
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NS, nonsignificant; S, nonsignificant.

<table>
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<tr>
<th>Presentation</th>
<th>Classical malrotation</th>
<th>Atypical malrotation</th>
<th>Classical malrotation with volvulus</th>
</tr>
</thead>
<tbody>
<tr>
<td>Classical presentation</td>
<td>15</td>
<td>0</td>
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</tr>
<tr>
<td>Atypical presentation</td>
<td>5</td>
<td>5</td>
<td>0</td>
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<tr>
<td>Acute pancreatitis</td>
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<tr>
<td>GERD</td>
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<td>0</td>
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<tr>
<td>Acute small bowel obstruction</td>
<td>1</td>
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<td>2</td>
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GERD, gastroesophageal reflux disease.
typical form, it can be diagnosed easily and corrected safely and effectively with surgery. However, the clinical presentation in older children is less dramatic with a broad array of symptoms. Such presentations in children and adults are likely to consist of more chronic symptoms such as postprandial bloating, intermittent cramping, and occasional vomiting or acute presentations such as intestinal obstruction. Intermittent volvulus or partially obstructing bands may cause these vague gastrointestinal symptoms, and a surgical correction is rewarding in these cases. Unfortunately, many patients are never referred to surgeons and are instead labeled as functional disorders. Our study corroborates with the findings of Penco et al [8] and Durkin et al [9], which suggest that chronic symptoms are more common in children older than 2 years.

In this study, all patients of malrotation presenting in the second year of life had a classical presentation with a sudden-onset bilious vomiting and upper abdominal distention. However, about two-thirds of patients beyond this period had atypical presentation, and in most of these patients, the diagnosis of malrotation was missed initially. In an Indian scenario, abdominal tuberculosis is considered one of the common causes for chronic abdominal symptoms, especially in the older pediatric age group. Of 6 referred children in this study, 5 were initially treated with ATT empirically. Nevertheless, they were found to have malrotation, and most of them were relieved of their symptoms after surgery.

In 2 patients, malrotation was diagnosed on UGI contrast study after an episode of pancreatitis. Persistent bilious vomiting in these patients forced us to investigate further. Both these patients had normal hepatic function and radiologically normal biliary tree. An MRCP done in these patients did not reveal any pancreatic abnormalities such as pancreaticobiliary malunion or pancreatic divisum. There are only isolated reports of malrotation causing pancreatitis in adults [10,11]. Anatomical distortion of the duodenal bulb by Ladd bands resulting in rotation of the pancreatic head with subsequent pancreatic duct obstruction has been postulated to cause recurrent pancreatitis. Jaundice caused by obstruction of the distal common bile duct by chronic volvulus is also reported [12,13].

A literature review of symptomatic malrotation in adults revealed less than hundred cases in the last 80 years [14,15]. Adults with malrotation can present in several ways. Presentation is often heralded only by vague, nonspecific abdominal complaints. Patients with these chronic symptoms typically present with recurrent intermittent cramping, nausea, and vomiting. Less common symptoms include intermittent diarrhea, hematochezia, constipation, malabsorption, and weight loss [16]. There is also a subset of patients in whom intestinal malrotation is truly asymptomatic and is diagnosed incidentally during a laparotomy or laparoscopy done for other conditions such as gastric bypass or during imaging (computed tomography and magnetic resonance imaging) done for other medical issues. 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 [17,18]. Such acute presentations caused by midgut volvulus or internal hernias have been reported even in adults.

On UGI contrast study, normal rotation is said to be present if the duodenal C-loop crosses the midline, placing the duodenojejunal junction to the left of the spine at the level of the gastric outlet. Upper gastrointestinal contrast study has a high sensitivity for intestinal rotational disorders and has been considered the criterion standard. Applegate et al [19] published 7 keys for obtaining an accurate UGI series for malrotation. It is important to note that normal cecal position does not exclude malrotation. Ultrasound has also been discussed widely as an adjunctive investigation in the diagnosis of malrotation [20,21]. Findings on ultrasound include inverse orientation of the superior mesenteric artery and vein in patients with malrotation and the whirlpool sign of midgut volvulus. In the present study, the ultrasound was diagnostic of malrotation in nearly one-third of the patients, whereas the diagnosis of midgut volvulus was missed in about half of these patients. Three-dimensional computed tomography reconstructions have also been reported to be used to diagnose malrotation.

It is generally accepted that symptomatic patients with documented malrotation require surgical intervention. However, the management of patients with asymptomatic malrotation is controversial. In the study of Stewart et al [22], 15% of all patients diagnosed as having malrotation were asymptomatic. Supporters of conservative approach argue that several studies have reported a reduced risk of midgut volvulus in cases presenting beyond the neonatal period [23,24]. Furthermore, the traditional Ladd procedure is not an innocuous. Others have agreed that surgery is not justified unless abdominal disorders can be directly linked to the malrotation [25,26]. A recent study that used a statistical model to assess the need for surgery in asymptomatic adults concluded that watchful waiting has resulted in increased life expectancy in adults older than 20 years [27]. Operative findings from the present study depict that 30% patients with typical malrotation older than 2 years had midgut volvulus. Thus, it reaffirms the importance of surgical correction in this group. All the 3 adults in our study were asymptomatic before the present episode, and 2 of them were found to have midgut volvulus at laparotomy. There are reports of midgut volvulus even in the older group of patients [28]. Thus, it supports the notion that the volvulus can occur in an asymptomatic patient with malrotation at any time. At present, it is impossible to predict the risk of midgut volvulus based on age, chronicity of symptoms, or manner of presentation [29]. Most of the current surgical literature favors operative correction of malrotation in patients with even minor symptoms or in those discovered incidentally [15,29,30].

Operative treatment (Ladd procedure) has been considered standard therapy since its first description in the 1930s.
The principles of the Ladd procedure remain unchanged; however, the use of laparoscopy to diagnose or correct malrotation has been reported over the last 2 decades. Laparoscopy is useful in determining the position of the ligament of Treitz and also the fixity of cecum. Palanivelu et al [31] highlighted the value of laparoscopy in both diagnosis and therapy, especially in patients with acute abdomen symptoms with doubtful diagnosis, which warrant urgent surgery. In their series of 73 children with acute abdomen symptoms in whom diagnostic laparoscopy was urgent surgery. In their series of 73 children with acute abdomen symptoms in whom diagnostic laparoscopy was done, malrotation was found in 9.5% of patients. In 1995, van der Zee and Bax [32] reported, for the first time, laparoscopic surgery for malrotation in a neonate. Since then, there have been many reports of successful correction of malrotation, laparoscopically [33,34]. A laparoscopic Ladd procedure has been shown to have low morbidity and can be safely accomplished in individuals with or without volvulus. Recently, Stanfill et al [35] compared the outcome between open and laparoscopic Ladd procedure in 156 children. They concluded that laparoscopic Ladd procedure can be performed safely in selected patients without an increase in complications with better short-term results compared with open procedure without any increase in operative duration. Another important use of laparoscopy, as shown by Mazziotti et al [36], is to evaluate the mesenteric base and to determine the adequacy of its width to prevent midgut volvulus. In their study of 7 patients with rotational abnormalities who underwent laparoscopic assessment, classical Ladd procedure was required in only 2 patients according to this criterion of width of mesenteric base. This criterion might be advantageous in a subset of patients with atypical presentations, in patients with atypical malrotation, or incidentally diagnosed truly asymptomatic patients because the procedure would avoid associated morbidity of a laparotomy. However, more studies are required to substantiate it further.

In the present study, preoperative symptoms persisted even after correction in 40% patients of the atypical malrotation group. On the contrary, about 80% patients of the typical malrotation group were relieved of their symptoms after surgery. Furthermore, 40% of the patients with initial atypical symptoms had persistence of symptoms postoperatively. Thus, the present study corroborates with the observations of Devane et al [37] that persistence of gastrointestinal symptoms is common in patients with atypical presentation. Intestinal dysmotility has been postulated for such symptoms. Several investigators have grouped various clinical presentations based on age and suggested that malrotation in older patients is a distinct clinical entity [12,38,39].

To conclude, malrotation should be suspected in all patients presenting with varied acute or chronic abdominal symptoms, and a UGI contrast study should be conducted. If the existence of typical malrotation is confirmed, surgical correction is mandatory to avoid volvulus and intestinal obstruction, irrespective of the patient’s age. Nevertheless, how to deal with the different atypical presentations of intestinal malrotation will continue to be puzzling both diagnostically and therapeutically. Perhaps, further studies based on the laparoscopy as a diagnostic tool would help in formulating a policy for definitive management in such cases.

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References