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Research Article

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Outcome of Surgical Treatment of Malrotation in Children: A 20 Year Experience in A Tertiary Centre

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Abstract

Aim: *This study evaluated the clinical presentation, diagnostic evaluation, and outcomes of surgical management of intestinal malrotation in children.*

Materials and Methods: *The medical records of all neonates and children with a diagnosis of malrotation, who underwent surgery between January 2000 and December 2019, were retrospectively reviewed.*

Results: *A total of 96 patients, 59 males, underwent surgical correction for intestinal malrotation. The median age at surgery was 11.4 days (3 days –12 years). The most frequently observed symptom of malrotation was bilious vomiting. Diagnostic evaluations included upper gastrointestinal contrast study, Doppler ultrasound scanning of the mesenteric vessels, and oral contrast CT scan of the abdomen. All patients underwent surgical correction. Sepsis was the most common postoperative complication. The short gut syndrome was observed in 7. Mortality was observed in six (6.2%) patients.*

Conclusion: *Neonates uniformly presented with bilious emesis or other clinical and radiographic evidence of high small-bowel obstruction. The diagnosis of malrotation after childhood is difficult. Ultrasound with colour doppler can be used as a routine screening method. Malrotation with its predisposition for volvulus and vascular compromise demands prompt diagnosis and management.*

Keywords: Midgut Volvulus, Intestinal Malrotation, Ladd's Procedure, Short Gut Syndrome

Introduction

The growth of the intestinal tract during the first 2 months of fetal development exceeds the capacity of the abdominal cavity, and the bowel develops outside the abdomen and physiological herniation occurs between about 6-12 weeks of intrauterine life. The intestinal tract returns to its usual intra-abdominal location within 10-12 weeks of gestation. The midgut normally undergoes 270 ° counter-clockwise rotation around the superior mesenteric artery, causing the duodenum to be fixed retroperitoneally and the cecum to the lateral abdominal wall [1].

Upon completion of the rotation, the mesentery is fixed in a broad base to the retroperitoneum from the ligament of Treitz to the ileocecal junction [2]. Midgut malrotation is broadly considered a deviation from the normal 270 degree counter clockwise rotation of the gut during embryonic development. The failure of the natural physiological midgut rotation leads to varying degrees of anomalies [3]. In complete non-rotation of the midgut the entire small bowel remains on the right side of the abdomen, the caecum, appendix and colon on the left and an absent ligament of Treitz [4].

Interruption or incomplete rotation of the intestine and abnormal position of the caecum result in a short superior mesenteric vascular pedicle and the tissue that fix the cecum to the abdominal wall now extend from the cecum and cross over the duodenum to the lateral abdominal wall forming the Ladd's bands [5]. Incomplete fixation of the right and left colon and the duodenum leads to potential space where the small intestine may push out through the unsupported area. This creates an internal hernia with possible entrapment and strangulation of the bowel [3].

Malrotation with abnormal bowel fixation presents an increased risk of intestinal obstruction and volvulus [6]. Any delay in diagnosis can cause bowel necrosis and short bowel syndrome [6,7]. Seventy-five percent of cases present during the neonatal period and up to 90% become symptomatic within the first year of life [6,8]. Malrotation can present acutely in newborns as bowel obstruction and intestinal ischemia associated with midgut volvulus, or in children, adolescents and adults with malnutrition and vague abdominal discomfort like intermittent crampy abdominal pain, bloating, nausea and vomiting chronic diarrhoea, constipa-

tion, and gastrointestinal bleedings over several months or years [9]. The Ladd procedure aims to minimize the risk of volvulus and vascular compromise.

We analyzed our 20-year experience with children who underwent operation for intestinal rotational defects, describing their clinical presentation, surgical procedures, complications and follow-up.

Materials and Methods

We retrospectively studied a cohort of 96 patients who were found to have intestinal malrotation at a tertiary level referral hospital from 1999 to 2019. After obtaining institutional review board approval, patient charts were reviewed for age at diagnosis, gestational age at birth, sex, weight, comorbidities, duration and nature of presenting signs and symptoms, results of preoperative imaging studies, intraoperative findings, postoperative complications, follow-up and resolution or persistence of preoperative symptoms.

Upper GI, lower GI studies, USG with colour doppler and CT scan abdomen were used for confirmation of diagnosis. Upper gastrointestinal contrast studies were reviewed by both paediatric radiologists and authors. Particular attention was paid to location of the ligament of Treitz (LOT) and cecum. The position of the LOT was described based on its relation to the midline and to the gastric outlet. Position of the cecum was described as either normal (right lower quadrant) or abnormal (position elsewhere in the abdomen). Atypical malrotation was defined as LOT location at or to the left of midline and below the level of the pylorus, with or without a highly mobile or high-riding cecum noted on small bowel follow-through or contrast enema.

All ultrasound examinations were performed by an expert paediatric radiologist. The structures between the SMA and the aorta were studied in the axial and longitudinal planes. The position of the third part of duodenum between the SMA, the SMV and the abdominal aorta was evaluated in the longitudinal plane. Once the decision for primary operative treatment was made, the diagnosis of malrotation was confirmed at surgical exploration. Intraoperative diagnosis was confirmed if Ladd's bands were noted to extend across the duodenum and/or the intestinal mesentery was found to be narrowed. All patients found to have malrotation on exploration were treated with Ladd's procedure.

Operative

The steps of operative procedure consist of

- Counter clockwise derotation of the midgut volvulus if present.
- Division of obstructing Ladd's bands overlying the second part of duodenum.
- Widening of the narrowed small bowel mesentery.
- Appendectomy.

A Fogarty embolectomy (Edward Lifesciences) 100 cm length catheter, 4F size with inflated balloon diameter of 9 mm was passed through the orogastric route into the proximal jejunum to enable detection of intraluminal duodenal and jejunal obstruction. In an event of volvulus with gangrene, the necrotic segment was resected with endeavors to preserve the maximum intestinal length and ostomy was created. Continuity was subsequently restored.

Results

A total of 96 patients, 59 were male underwent surgical correction of intestinal malrotation as an isolated procedure or in combination with the repair of other congenital anomalies. The most common age range at presentation was less than 7 days (Table 1). The median age at surgery was 11.4 days (3 days-12 years). The most frequently observed symptoms of malrotation were bilious vomiting (n = 91, 94.7%) (Table 2). The diagnostic evaluation included 82 (85.4%) upper gastrointestinal series (UGI) (Figure 1), ultrasound with doppler in 56 (58.3%) (Figure 2), contrast-enhanced CT scan abdomen in 38 (39.6%) (Figure 3) and lower GI enemas in 27 (28.1%). The intraoperative presentations are shown in (Table 3). Operative procedures included correction of malrotation, appendectomy in 83 patients (86.4%) and correction of congenital anomalies as indicated. None of the patients had recurrent volvulus. The most common postoperative complications are sepsis; other surgical complications are shown in (Table 4). The cause of death was mainly attributed to intestinal gangrene leading to sepsis, severe short bowel effects, respiratory distress syndrome, and multiple organ failure before the enterostomy was reversed. The length of hospital stay was 11–27 days. The average time of enterostomy reversal in this study was 83 days (range, 65-97 days) after the primary laparotomy. When following up patients, we were able to contact 79 patients, which represent an 82.2%, with a mean follow-up of 8 years ranging from 14 months to 11.5 years. The vast majority of patients were incident free.

Table 1: Age of presentation

%	Age	n
50	< 7days	48
27	8-30 days	26
17.7	1 -12 months	17
5.2	> 1 year	5

Table 2: Clinical presentation

%	Clinical Features	n
94.7	Bilious vomiting	91
79.1	Intestinal obstruction with absent passage of stools	76
16.6	Bloody stool	16
11.5	Abdominal distention	11
3.1	Recurrent abdominal pain	3
2.1	Failure to thrive	2

Table 3: Operative findings

%		n
58.3	Malrotation with Ladds band	56
28.1	Malrotation with Ladds band with volvulus	27
13.5	Malrotation with Ladds band with volvulus with gangrene	13

Table 4: Incidence of postoperative complications

%	Clinical Features	n
11.4	Sepsis	91
9.4	Wound infection	76
7.3	Feeding difficulty	16
7.3	Short bowel syndrome	11
5.2	Post op adhesive obstruction	3
3.1	Pneumonia	2

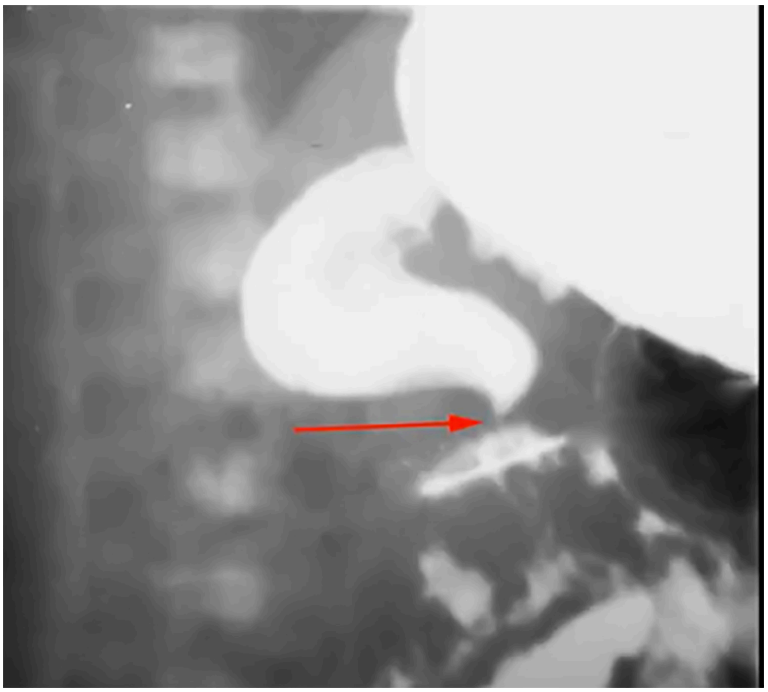

Figure 1: Upper GI contrast study showing complete obstruction with the distinctive conical or beak shape

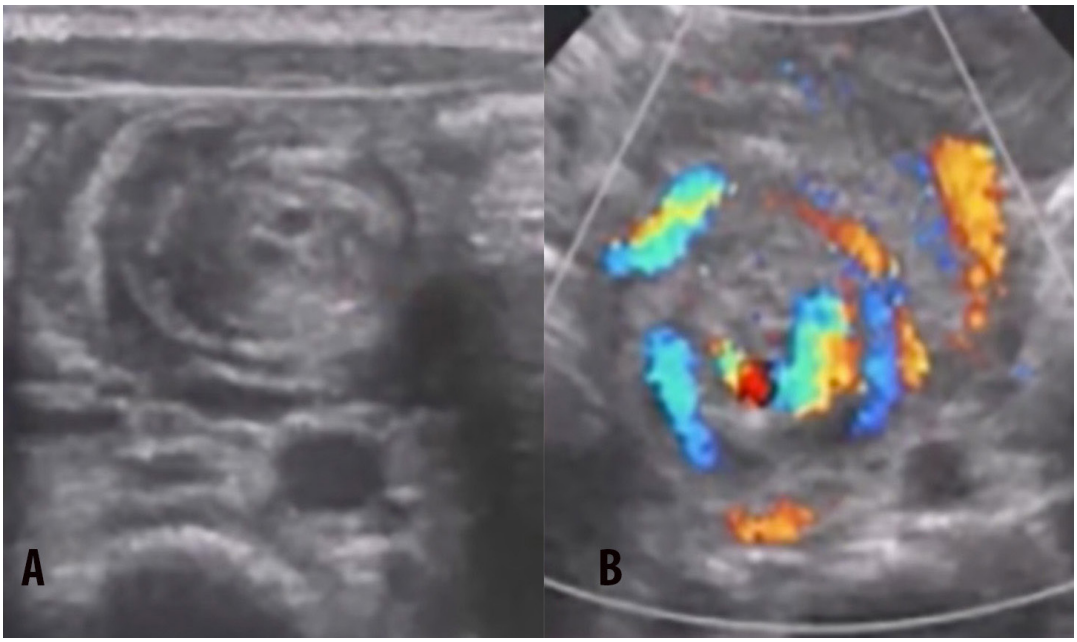

Figure 2: (A) Ultrasound in grey scale showing of 'whirlpool' sign and (B) in colour doppler

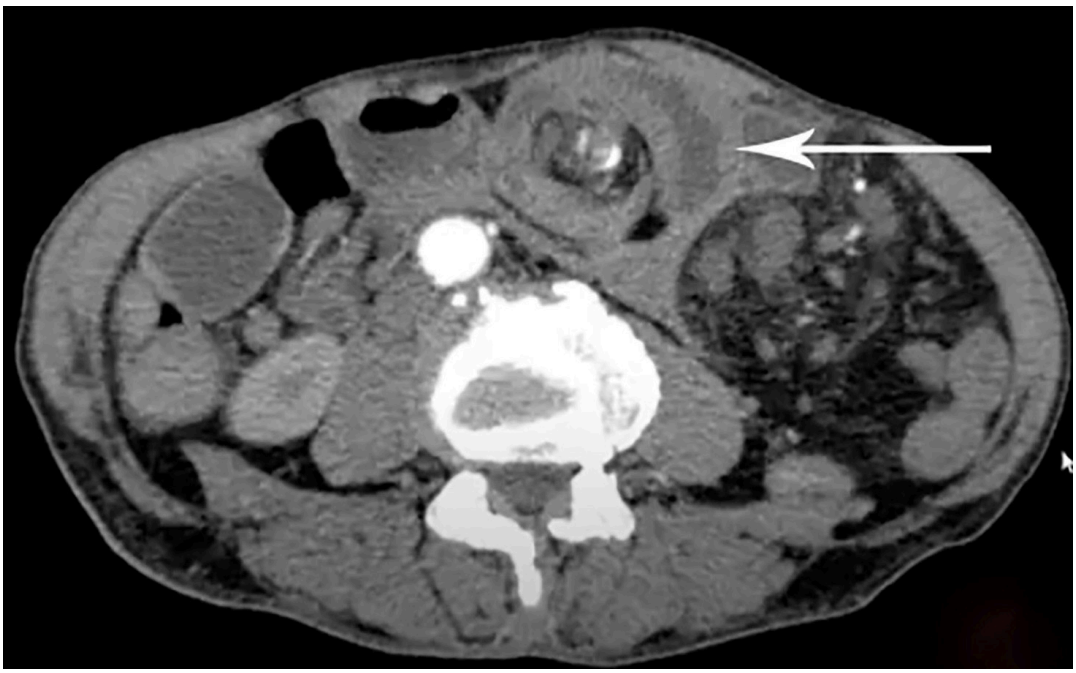

Figure 3: CT scan abdomen showing twisted bowel around central superior mesenteric artery

Discussion

Intestinal malrotation occurs at a rate of about 1 in 500 live births [10]. Up to 40% of patients with malrotation present within the first week of life and up to 75% to 85% have been diagnosed by age 1 year [11]. In our series (n=48, 50%) presented within 7 days and (n=91,94%) by 1 year of age. Conventional plain X-ray for the diagnosis of malrotation is neither sensitive nor specific. Abdominal color doppler may divulge malposition of the SMA, raising the suspicion of gut malrotation [12]. Pacros et al first described the 'whirlpool' sign [13]. It includes duodenal dilatation with distal tapering and a fixed midline bowel and mesentery twisted around the SMA axis. The gold standard for the diagnosis of gut malrotation in the pediatric age group is an upper gastrointestinal contrast study [14]. It demonstrates the duodenum and duodenojejunal flexure located to the right of the spine. A contrast enema has also been recommended for use in combination with UGI studies because it can demonstrate an abnormal location of the ileo-cecum and right colon. However, the possibility of gut malrotation cannot be excluded in a normal contrast enema study [15]. CT scans with oral contrast are increasingly being used as they are now considered the investigation of choice; offering 80 % diagnostic accuracy [16]. CT and MRI scans detect the anomalous position of the SMV, posterior and to the left of the SMA. It also shows the abnormal anatomical arrangements of the midgut with the duodenum not crossing the spine [15]. In this series USG colour Doppler picked (n=62, 64%), UGI studies picked (n=76, 79%) and oral contrast CT scan was required in (n=42, 43.7%).

Neonates uniformly presented with bilious emesis or other clinical and radiographic evidence of high small-bowel obstruction, which was evident in 91 patients (94.1%) in our series. Beyond the neonatal period, malrotation may present with bilious vomiting and bowel obstruction more commonly presents with abdominal pain, diarrhea, vomiting, and failure to thrive [16, 17]. In patients with chronic midgut volvulus, long-standing malabsorption syndrome may occur. The vague nature of these symptoms calls for a high level of clinical suspicion of intestinal malrotation [14, 18].

Some authors recommend routine appendicectomy to prevent confusion with the abnormally located vermiform appendix in the left upper quadrant if the patient presents with abdominal pain [19]. Patients may have an atypical presentation of appendicitis which may increase morbidity and mortality [20]. In our series, we preferred appendectomy and 83 patients (86.4 %) underwent appendectomies. The Ladd's procedure for malrotation is a known risk factor for postoperative adhesion-related obstruction in neonates undergoing laparotomy [21]. The adhesion formation is partly due to the nature of the procedure in which the small bowel mesentery is incised to permit widening. The raw area after widening predisposes for adhesion. Stauffer et al. reported that 5 (12%) of 41patients required surgical intervention for bowel obstruction during follow-up also reported small bowel obstruction in 11% of patients. In this study, 5 (5.2%) children developed bowel obstruction. Two patients (2.1%) required multiple admissions due to small bowel obstruction. Conservative management was attempted in all 5 patients but 1 child required surgery [22, 23].

Recurrent midgut volvulus is uncommon and has been reported in up to 2-7% of children [24]. A history of Ladd's procedure does not exclude recurrence [25]. In this series, we did not encounter recurrent volvulus. Volvulus is the most feared complication, seen in 60-70 % of neonates with malrotation. Delay in diagnosis may cause strangulation in 15% of cases which leads to intestinal ischemia, intestinal necrosis, septicemia, and short bowel syndrome [26, 27]. We encountered 40 patients (41.6%) with volvulus of which 13 (13.5%) were strangulated and gangrenous.

The most serious consequence of midgut volvulus is death or loss of significant length of the intestinal tract resulting to a TPN-dependence for life [28]. Bowel necrosis is a major challenge in developing countries where total parenteral nutrition is out of reach and bowel transplantation is not an option [23]. If the ischemic bowel is discovered during a laparotomy for volvulus, every effort should be made to preserve bowel length, and if viability is in doubt, a second-look laparotomy should be performed 12-24 hours later without initial resection or with a very conservative resection. We have accrued 7 patients with short-bowel syndrome over 20-year experience. Six patients in our series died from intestinal necrosis caused by midgut volvulus with overwhelming septic episodes. The notable improvement in the perioperative mortality rate of children with midgut volvulus is a result of developments in diagnostic techniques, early intervention and aggressive nutritional support.

Conclusion

Our findings demonstrate that neonates uniformly presented with bilious emesis or other clinical and radiographic evidence of high small-bowel obstruction malrotation. The diagnosis of malrotation after childhood is difficult. An ultrasound with color doppler used as a routine screening method can easily show the retroperitoneal position of the third part of duodenum. The diagnosis of malrotation on the basis of an upper GI series may be challenging even for experienced radiologists. Malrotation with midgut volvulus and gangrene is a serious complication. To have a normal functioning gastrointestinal tract, the condition must be corrected completely.

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