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PRESENTATION AND OUTCOME OF SURGICAL TREATMENT OF MALROTATION IN CHILDREN

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Summary

Introduction : Malrotation is a common anomaly in the pediatric age group which includes a wide spectrum of anomalies of Rotation. Both acute and chronic presentations are common. Atypical malrotation not having all the features of classic malrotation is frequently found which a diagnostic dilemma and the management varies from centre to centre. Materials and Methods : The medical records of all patients with symptomatic malrotation, who underwent surgery between July 2001 to June 2011, were retrospectively reviewed. Patients' presentation, management, operative findings and complications were evaluated. Results : 68 patients underwent surgery for malrotation at a median age of 2 years. Male to female ratio was 2:1. 28(41%) presented with acute symptoms and 40(59%) with chronic symptoms. 54(79%) patients had vomiting, 36 (53%) presented with abdominal distension, 19(28%) had recurrent abdominal pain. Diagnostic laparoscopy was done in 7(10%) patients. Ladd's band was found in 16(24%) patients and Volvulus was found at the time of surgery in 5(7%) patients. 5(7%) patients also had associated anomalies. Ladd's procedure was done in 15(22%) patients and 23(34%) patients needed resection and anastomosis. Median length of hospital stay was 10 days. Postoperative bowel obstruction was seen in 4(6%) patients and 2(3%) patients had post operative intussusception. There was 2(3%) death due to septicaemia with volvulus and gangrenous gut. Conclusion : The clinical presentation and anatomy of malrotation occurs along a wide clinical and anatomic variations and a high index of suspicion is required to prevent a delay in diagnosis.

Key words

Malrotation; intestinal obstruction; vomiting; ladd's band

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Introduction

Malrotation is a common anomaly in the pediatric age group that results from abnormal or incomplete rotation of the mid gut during embryonic development[1,2]. The incidence has been estimated at 1 in 6000 live births but an increased incidence of 0.2 % has been found in barium swallow studies and autopsy studies estimate that the true incidence may be as high as 1% of total population[3]. Malrotation includes a wide spectrum of anomalies of Rotation. The classic presentation of malrotation occurs in the newborn period with bilious vomiting and abdominal distension[1]. Up to 75% of patients present during the 1st month of life, another 15% within the 1st year, but presentation in children or adults is not uncommon[3]. Typically, the mid gut develops acute vascular congestion and arterial insufficiency secondary to torsion of the superior mesenteric vessels but chronic volvulus with malrotation may present with more intermittent symptoms and vague clinical findings, making diagnosis more difficult and the treatment of malrotation found incidentally in this group is more controversial. Patients with chronic malrotation and intermittent volvulus usually have significant collateral mesenteric circulation and avoid the bowel infarction and occlusion of venous outflow occurs before arterial occlusion in these patients[2,4,5]. Some advocates conservative treatment based on the patient's age or selective surgical treatment according to the type of malrotation diagnosed by radiology, others recommend aggressive treatment regardless of age, the type of malrotation, or the presence of symptoms[6-10]. Currently, children whose anatomy is somewhere between completely normal and classically malrotated are called "atypical malrotation" or "malrotation variant" and are being increasingly diagnosed by radiologists and referred for surgery[11,12] . These patients are a clinical and medicolegal challenge to the surgeon who faces the dilemma of operating on patients who do not have typical malrotation, and who may or may not be symptomatic, but in whom the risks of not operating are unknown[1].

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Materials and methods

Records of all patients with symptomatic malrotation who underwent surgery or in whom malrotation was diagnosed at the time of operation during the period of July 2001 to June 2011 in Department of Pediatric Surgery, Chittagong Medical College Hospital were retrospectively reviewed. The operations were done by different surgeons. Patients in whom no surgery was done and patients who had abdominal wall defects (omphalocoele, gastroschisis) and diaphragmatic hernias were excluded from this study because these patients represent unique subsets of intestinal rotational disorders. Clinical information including age at operation, sex, presenting symptoms, radiologic studies (plane and contrast abdominal X-ray, abdominal ultrasound and computed tomography), operative findings, and outcome were documented. The position of the ligament of Treitz was described relative to the midline (at or to the left of midline versus to the right of midline). Malrotation was diagnosed based on the position of the ligament of Treitz, bands extending across the duodenum and the bowel rested on a narrow mesentery without retroperitoneal fixation and position of the caecum. No attempt was made to classify retroperitoneal attachment of the duodenum because lateral views were not available in all cases. Caecal position was classified as either right lower quadrant (normal) or somewhere other than the right lower quadrant (abnormal). Follow up was from 15 days to 1 year.

Results

During the 10-year study period, 68 paediatric patients (Neonate 33, Infant 16, and Children 19) underwent operation for malrotation, none of whom had abdominal wall defects or diaphragmatic hernias. The mean age at operation was 2 yrs (range 3 days to 12 years), with a higher portion of boys undergoing operation (45 boys, 23 girls; ratio 2:1). Symptoms were acute (less than 48 hours duration) in 28 (41%) patients and chronic in 40 (59%). The majority of children presented with vomiting (54 patients, 79%); of those with emesis, it was bilious in 69% (37 of 54) and nonbilious in 31% (17of 54). Other common symptoms on presentation included abdominal distension (36 patients, 53%), constipation (31 patients, 46%) and recurrent abdominal pain (19 patients, 28%). Bilious vomiting and abdominal distension were more frequent in patients who presented with acute symptoms and more in neonate and infants than with chronic symptoms whereas recurrent abdominal pain predominated in chronically symptomatic patients and in children.

Malrotation was found incidentally in 12 (18%) patients, 8 were operated with the clinical diagnosis of acute surgical abdomen and 4 were operated for Intussusception.

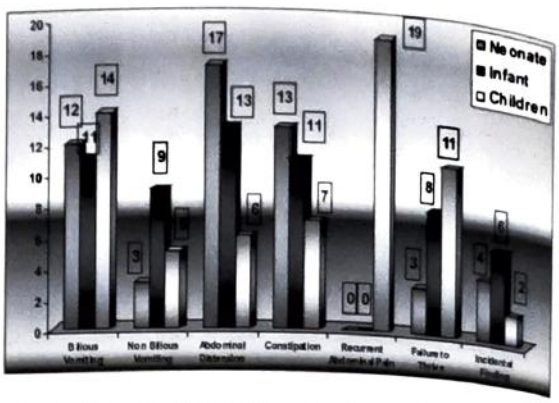

Fig 1 : Presentation of malrotation at different ages

The diagnosis of patients presenting with acute complaints was mostly straight forward and Plane X-ray abdomen (35 patients) and abdominal USG (51 patients) were sufficient in most cases. 21 patients underwent Ba study and malrotation was seen in 13 patients. Abdominal CT was done in 9 patients and Diagnostic Laparoscopy needed in 7 patients. Most children had chronic symptoms and needed more investigations.

Table I : Investigations done to diagnose malrotation

Investigations	Neonate	Infant	Children
Plane X-ray abdomen	15	12	8
Ba meal follow through	0	3	18
USG of whole abdomen	20	12	19
CT scan of abdomen	0	0	9
Diagnostic laparoscopy	0	0	7

Peroperatively typical malrotation was found in only 16 (24%) patients who had Ladd's band, Superior Mesenteric artery on the left of Duodenojejunal flexure, abnormal position of Caecum and short mesentery. 44 had atypical malrotation. Caecal position was abnormal in 60 patients, Bands from lateral wall upto Caecum, duodenum or small gut was found in 55 patients, short mesentery in 40 patients and Superior Mesenteric artery on the left of Duodenojejunal flexure was in 48 patients. 45 patients had features of intestinal obstruction and necrosed gut was found in 19 patients with 5 having volvulus. Associated anomaly was fond in 5 patients (Ileal Atresia 2, Cardiac anomaly 1, Meckel's diverticulum 1, Down's syndrome 1). Typical Ladd's procedure was done in 15 (22%) patients and 23 (34%) patients needed resection and anastomosis of gut. Release of bands was done in all patients having bands (55) and Appendectomy was done in 20 patients.

Table II : Operative findings in patients of malrotation

Operative Findings	Neonate	Infant	Children
Ladd's band	4	6	6
Atypical malrotation	13	14	17
Associated anomaly	2	3	0
Gut necrosis	7	11	1
Intestinal obstruction	17	13	15
Volvulus	2	3	0
Internal hernia	0	1	3

Post operative prolonged ileus (16) was the most common complication and all improved without any consequences. 4 patients developed adhesive bowel obstruction and all were conservatively treated and improved but 1 needed 3 admissions. 5 patients (7%) had wound dehiscence with 2 needed secondary repairs. 2 patients developed post operative Jejuno-ileal intussusception and needed re-exploration and manual reduction 1 at 3rd POD and the other at 5th POD. 2 patients died which developed sepsis following resection and anastomosis of gut following volvulus and gangrenous gut. Follow up period was 15 days to 1 year with mean follow up period of 2 months. 5 patients were lost to follow up.

Table III : Complications in patients underwent surgery for malrotation

Complications	Neonate	Infant	Children
Sepsis	2	0	0
Prolonged ileus	4	5	7
post operative adhesive bowel obstruction	1	2	1
Post operative intussusception	0	2	0
Respiratory failure	1	1	0
UTI	0	0	1
Wound dehiscence	2	2	1

Discussion

Malrotation is the term used to denote an interference with the normal process of orderly return of the fetal intestine from the physiologic hernia to the abdominal cavity during which it undergoes systematic rotation and fixation[13]. During the second month of embryonic period the gut undergoes rapid elongation, exceeds the capacity of the abdominal cavity and herniates out into the extracoelomic umbilical cord. The intestines then return to the abdomen during the third month and undergo a counter-clockwise rotation about the axis of the superior mesenteric artery. This rotation brings the duodeno-jejunal loop under the superior mesenteric artery and affixes it to the retroperitoneum in the left upper quadrant; the ceco-colic portion rotates and affixes to the right lower quadrant. The effect of this rotation is to secure the pedicle of the superior mesenteric artery across a broad mesenteric base, thereby preventing postnatal rotation and possible vascular compromise of the bowel. Abnormalities of this process occur along a spectrum and were classified by Rescorla and Grosfeld as "nonrotation," which is an early arrest of rotation of the duodenojejunal loop when the gut rotates only 180^0 instead of the normal 270^o leaving the small gut in the right side of the abdomen and large gut on the left; and "incomplete rotation," which is an arrest of the duodenojejunal loop after it has partially rotated around the superior mesenteric artery but has not ascended to a normal position, the duodenal loop lacks 90 ° and the cecocolic loop lacks 180 ° of their normal 270 ° counter-clockwise rotation [1,13-17]. Associated anomalies are common which includes intestinal atresia, imperforate anus, cardiac anomalies, duodenal web, Meckel's diverticulum, Diaphragmatic and other Hernia, exomphalos and Trisomy 21[3,13]. The clinical presentation and anatomy of malrotation occurs along a wide clinical and anatomic spectrum and there is little data about the medicolegal dilemma of whether to operate on children who present with atypical symptoms or atypical anatomy. Malrotation may be difficult to diagnose after the neonatal period as these patients usually have intermittent symptoms and vague clinical findings, but malrotation with midgut volvulus is usually diagnosed straightforward. Failure to recognize this condition promptly can result in not only acute ischemia but also chronic mesenteric vascular compromise[4]. Patients with chronic malrotation typically present with obstruction, chronic abdominal pain, or malabsorption with diarrhea[7]. Failure to thrive, Mild abdominal discomfort, constipation, abdominal distention caused by ascites, obstructive jaundice, peritonitis with septic shock, solid food intolerance, dyspnea, gastrointestinal bleeding, early satiety and gastroesophageal reflux are among the other presentations described in the literature[3,4,7,18-22]. After a lengthy period of medical evaluation, some of these patients are labeled with functional or psychiatric disorders[3].

Other patients with malrotation in the absence of volvulus may remain asymptomatic for life [4,22]. Although malrotation may be asymptomatic in many older children and adults, it has been shown that older children presenting with malrotation frequently have potentially life-threatening complications of this anomaly[23] .The discordant incidence of malrotation between cases and autopsy studies suggest that many patients are asymptomatic but anatomically at risk for midgut volvulus[3].

Diagnosis is usually made by barium meal and follow through, abdominal ultrasound scan with special attention the relative positioning of the superior mesenteric artery (SMA) and vein and rarely angiography[24]. Abnormal findings in barium meal include positioning of duodenojejunal flexure to the right of the spine, duodenal obstruction, and the "coil spring", "corkscrew", or "beak" appearance of the obstructed proximal jejunum[3]. Malrotation may cause the SMV to lie directly anterior or to the left of the SMA, instead of in the normal right-sided position and produce a "whirlpool" pattern in color flow Doppler imaging[25]. Angiography may show the same pattern[26]. Although both the abnormal course and position of the intestines and the abnormal orientation of the mesenteric vessels may be identified on CT scans, it is not as reliable as the above[27]. Laparoscopic approach is an alternate technique if the other investigations are not conclusive and we are using it increasingly to diagnose, confirm, or treat the malrotation. Laparoscopy can confirm whether the cecum is fixed, the root of the mesentery is wide, and the presence of Ladd's bands obstructing the duodenum[28-31].

Most centres do the Ladd procedure for malrotation which consists of (1) evisceration of the bowel and inspection of the mesenteric root; (2) counterclockwise derotation of the midgut volvulus; (3) lysis of Ladd's peritoneal bands, with straightening of the duodenum along the right abdominal gutter; (4) appendectomy; and (5) placement of caecum in the left lower quadrant[12]. We usually do not do appendectomy routinely with the argument that it may be used for reconstructive purpose later on if needed.

Conclusion

As noted by Ladd in 1932, malrotaion is a condition rare enough that is likely to escape the mind, and it is common enough to be important[2,3]. A high index of suspicion is required to prevent a delay in diagnosis.

Based on our review, we support the policy that regardless of age and even in asymptomatic patients, any malrotation diagnosed should have particular surgical attention. When an anomaly of fixation is suspected in an asymptomatic patient, a more thorough investigation is indicated. We suggest ultrasonography of abdomen, barium meal follow through and abdominal CT if necessary. If doubts still remain, laparoscopy can be both diagnostic and therapeutic. How to handle the different forms of intestinal malrotation will continue to be puzzling. The idea that one operation can resolve all types of symptoms and alleviate the risk of volvulus in this widely varied population oversimplifies the problem.

Disclosure

All the authors declared no competing interestes

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