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Outcomes of surgical treatment of malrotation in children

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ABSTRACT

Background: Abnormalities of rotation and fixation of the intestines are of intense interest to the pediatric surgeon, as they are frequently associated with volvulus which has catastrophic consequences when diagnosis is delayed or not even considered. This study evaluates the outcomes of surgical management of intestinal malrotation (IM) in children. **Materials and Methods:** The medical records of all patients with symptomatic malrotation, who underwent surgery between January 2000 and September 2009, were reviewed. Patients' characteristics, management, complications, and survival were evaluated. **Results:** Nine patients (eight boys and a girl) underwent surgery for malrotation at a median age of 15 days. Eight presented with acute symptoms and one with chronic symptoms. All the patients had symptoms of intermittent or complete upper intestinal obstruction, and malrotation was documented by an upper gastrointestinal contrast study in two of them. Volvulus was found at the time of surgery in seven patients, five of whom were neonates. One patient also had associated mesentery cyst. Seven patients were treated by Ladd's operation. One patient with massive bowel gangrene due to volvulus had right hemicolectomy. There was one perioperative death from anastomostic leak. Median length of stay was 9 days. Postoperative bowel obstruction was seen in two patients (one died), resulting in an overall mortality of 22.2%. **Conclusions:** Bowel gangrene from volvulus contributes to mortality, and small bowel adhesive intestinal obstruction is a cause of morbidity and mortality following surgery for IM. Neonates with bilious vomiting should raise the suspicion of malrotation until proven otherwise and given prompt intervention. There is a need for high index of suspicion in babies with bilious vomiting especially when recurrent to prevent devastating complications when present.

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INTRODUCTION

Intestinal malrotation (IM) refers to all the abnormalities of intestinal position and attachment.[1] Normal embryologic development of the midgut results in rapid growth and umbilical herniation during the sixth week. During week 10 through 11, the gut begins its 270° counterclockwise rotation about the superior mesenteric artery axis and reenters the abdomen. By the 12th week, fixation occurs. The incidence of symptomatic malrotation is reported to be 1 in 6000 live births.[2] The incidence of this condition in African population is not known. More than half of the patients present with the symptoms during the first month of life, and virtually all have bile-stained vomiting.[3] Because the consequences of malrotation associated with midgut volvulus may be so catastrophic, an understanding of the anatomy, diagnostic criteria, and appropriate therapy for this putative emergency illness is imperative. This report reviews the outcomes of surgical management of intestinal malrotation in a Nigerian Teaching Hospital.

MATERIALS AND METHODS

The records of nine pediatric patients with symptomatic malrotation of the intestine, seen from January 2000 to September 2009, were reviewed. Patients' characteristics, presentation, imaging investigations, operations performed, complications, and outcomes were evaluated.

RESULTS

There were nine children (eight boys and one girl) of age in the range 5 days–13 years (median: 15 days). Three

patients (33.3%) presented within the first 7 days of life, 5 (55.6%) presented at less than 1 month of age, and 75% presented under 1 year of age [Table 1]. There were five termed neonates with weights ranging from 2.9 to 3.5 kg (mean: 4.2 kg). Preoperative symptoms included bilious emesis (87.5%), fever (50%), and abdominal pain (37.5%). Bilious vomiting was the predominant sign in neonates. Abdominal pain followed by bilious emesis was evident in all the children older than 1 year of age.

Plain abdominal radiograph showed double bubble sign in one patient and dilated stomach, duodenum, and paucity of gas in the distal bowel in three patients [Figure 1], suggesting duodenal obstruction. Four patients had upper gastrointestinal series (UGI) which suggested malrotation in two patients and jejunal atresia and stenosis in one case each. However, one patient had associated mesenteric cyst detected with ultrasound scan. Eight patients had emergency exploratory laparotomy and one was done electively. Intraoperative findings included Ladd's band and midgut volvulus [Figure 2] in eight patients, five of whom were neonates. Eight patients were treated by Ladd's operation. One patient with massive gangrene of the small bowel due to volvulus had right hemicolectomy. A 21-day-old neonate with associated mesentery cyst stretching part of the jejunum had segmental resection and end to end anastomosis in addition to the Ladd's procedure. A 3-year-old boy with duodenal obstruction from Ladd's band without volvulus had adhesiolysis. One patient developed anastomotic leak and died of septicemia resulting in the only perioperative death. Two patients (22.2%) were readmitted with small bowel intestinal obstruction within 1 month of initial discharge. One did well with nonoperative management and the other one died of sepsis few hours after readmission, resulting in an overall mortality of 22.2%. Median hospital stay was 9 days (7–17 days).

DISCUSSION

Most of the patients (55.6%) in this study presented during the neonatal period. This is in keeping with the literature reports of 50–80%,[2,4-6] but presentation can occur at any age. Males are said to be slightly more affected than females.[2,6] Our finding of eight boys

Table 1: Patients' characteristics, procedure, complications, and outcomes of intestinal malrotation

Patients	Age at surgery	Sex	Procedure	Complication(s)	Initial outcome	Readmission	Final outcome
1	6 days	F	Ladd's procedure	Nil	Survived	No	Survived
2	21 days	M	Ladd's procedure*	Nil	Survived	No	Survived
3	5 days	M	Ladd's procedure	Nil	Survived	No	Survived
4	3 years	M	Adhesiolysis	Nil	Survived	No	Survived
5	10 days	M	Ladd's procedure	SBO	Survived	Yes	Survived
6	5 days	M	Ladd's procedure	SBO	Survived	Yes	Died
7	2 months	M	Right hemicolectomy	Anastomotic leak	Died	No	Died
8	13 years	M	Ladd's procedure	Nil	Survived	No	Survived
9	4 months	M	Ladd's procedure	Nil	Survived	No	Survived

SBO, small bowel adhesive intestinal obstruction; *Patient also had segmental resection of jejunum with excision of mesentery cyst



Figure 1: Plain abdominal radiograph of a 5-day-old neonate, showing dilated stomach and proximal duodenum with paucity of gas in the distal bowel

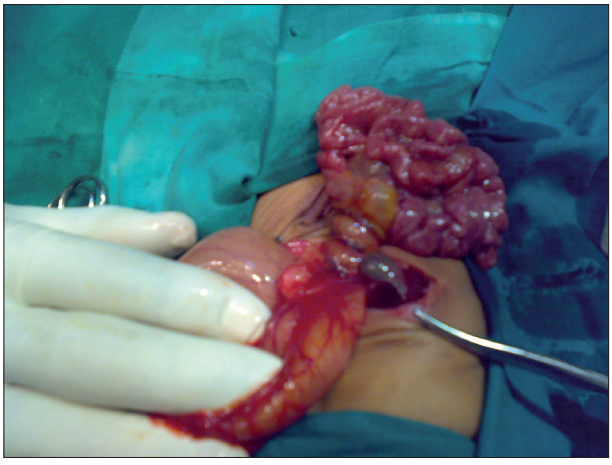


Figure 2: Laparotomy findings of intestinal malrotation and midgut volvulus in a 5-day-old neonate

against one girl is in keeping with this fact but is in contrast to the finding of equal sex distribution reported by Ameh *et al*, in Zaria.[5]

The most dreaded complication of malrotation is volvulus. Volvulus, a rotation of the gut along its mesenteric stalk occurs in 60–70% of neonates diagnosed with IM, with ultimate strangulation in about 15%.[6,7] Delay in diagnosis may lead to intestinal ischemia, intestinal necrosis, septicemia, and short bowel syndrome.[8] All the five neonates in this report had volvulus, giving a vovulus rate of 100% in the neonates. This compares to 77% in a report,[9] but it is higher than 14-42% in other reports.[5,10] This may be due to improved awareness of pediatric surgical services in our environment or difference in the number of patients reported in each series.

The clinical features in the neonates are indistinguishable from those of duodenal stenosis with bilious vomiting and epigastric distension which resolves after vomiting or aspiration by nasogastric tube. Pain or irritability is not a prominent clinical feature in the neonate, but is a common feature in the toddler and older child. This finding has been replicated in our study with all the neonates presenting with bilious vomiting, and abdominal pain in two children of 3 and 13 years of age. Bilious vomiting in a neonate should raise the suspicion of IM with midgut volvulus until proven otherwise. Plain abdominal radiographs are often normal but may show a dilated duodenum with a fluid level and paucity of gas in the distal bowel,[2,6] as it was found in three of our patients.

An upper gastrointestinal contrast study is the investigation of choice for any child presenting with bilious vomiting and should be done immediately. The contrast examination with a volvulus will show a dilated duodenum with a typical corkscrew appearance projecting forward away from the posterior abdominal wall on an oblique view.[6] It was done in four of our patients and it was suggestive of malrotation in two and jejunal atresia and stenosis in one case each. Assessment of the position and relationship between the superior mesenteric vessels by Doppler ultrasound is characteristic, and may be a useful adjunct in the diagnosis of IM, but this is not a readily available tool in developing countries.

The pathologic effects of anomalies of rotation arise from excessive mobility, compression, or kinking of bowel and predisposition to torsion, volvulus, and intussusceptions from the narrow stalk.[6] Ladd's procedure is the surgical treatment of choice for all malrotations. This involves derotation of the bowel if torsion is present, division of the peritoneal attachments lying across the duodenum from cecum to right upper quadrant, widening of the base of the mesentery, appendectomy, and taking down the ligament of Treitz and moving the duodenum to the right and, finally, returning the bowel to a position of nonrotation with the cecum placed into the left upper quadrant. The morbidity and mortality of malrotation are mostly due to midgut vovulus and bowel adhesion. Delayed presentation may lead to bowel gangrene necessitating resection and anastomosis with a resultant increase in morbidity and mortality. The only perioperative mortality in this study was a result of anastomotic leak following right hemicolectomy for bowel gangrene in a 2-month-old child.

In developing countries where total parenteral nutrition, let alone bowel transplantation, is not feasible, bowel necrosis poses a great challenge. If ischemic bowel is found at laparotomy for volvulus, every attempt should be made to preserve the bowel length and if any doubt exists about viability, a second-look laparotomy should be done 12–24 hours later without initial resection or with a very conservative resection. This has been shown to improve the outcomes.[6] In our environment, the morbidity and mortality is compounded by delayed presentation and referral. These problems can be addressed by educating the parents, traditional birth attendants, midwives, general practitioners, and pediatricians on the need for suspicion of malrotation in any child with bilious vomiting and early referral.

Two (22.2%) patients were readmitted for adhesive intestinal obstruction in this report. This compares to 7–24% incidence of postoperative adhesions in previous reports.[5,8,10,11] The steps of the Ladd's procedure required much handling and manipulation of the bowel increasing the risk of adhesion. The adhesions resulting from the operation are thought to be advantageous because they are felt to stabilize the bowel in its new position. However, this can be a major cause of morbidity and mortality. The newly evolving laparoscopic approach for Ladd's procedure may reduce the incidence of postoperative adhesion.[12-14] However, laparoscope is not readily available in a poor resource setting like ours. The mortality from malrotation has been reported to be 6.9–16.7%[4,5,15] compared to perioperative mortality of 11% in this report. The 22.2% overall mortality in this study is lower than 50% reported in Benin, Nigeria.[16]

CONCLUSION

We report a 22.2% risk of postoperative bowel obstruction after a Ladd's procedure, 11% perioperative mortality, and 22.2% overall mortality. A high index of suspicion in the neonates with bilious vomiting, rapid diagnosis, and appropriate operative therapy results in a favorable outcome for children with malrotation. Malrotation with its propensity for volvulus is no doubt a time bomb lying within the abdomen. Detailed education about the potential for postoperative small bowel obstruction and the need for early presentation must be given to the parents of these children on discharge.

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