

## Research Article

# Profile of children with intestinal malrotation: A tertiary hospital experience in a developing country

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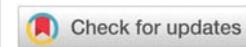
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## Abstract

**Background:** Intestinal malrotation is a congenital disorder resulting from abnormal rotation of the intestine during fetal development. The aim of this study was to evaluate our experience in the management of pediatric patients with intestinal malrotation.

**Materials and methods:** This was a retrospective study of children aged 3 years and below who were treated for intestinal malrotation between January 2014 and December 2018 at the pediatric surgery unit of Enugu State University Teaching Hospital (ESUTH) Enugu, Nigeria.

**Results:** Sixty-one patients had laparotomy for intestinal malrotation during the study period. There was predominance of male patients and the age range was 10 days to 3 years. Abdominal pain and upper gastrointestinal tract contrast study were the most common symptom and investigation respectively. Intestinal obstruction caused by congenital (Ladd's) bands was the most common intra-operative finding and Ladd's procedure was the most performed surgical procedure. Most patients did not develop any post-operative complications. However, surgical site infection was the most common complication following surgery. Mortality occurred in 8 (13.1%) patients.

**Conclusion:** Intestinal malrotation symptoms occur more in males and abdominal pain is a common and consistent symptom. Upper gastrointestinal contrast study is usually diagnostic. There are morbidity and mortality associated with treatment of intestinal malrotation.

## Introduction

Intestinal malrotation is a congenital disorder resulting from abnormal rotation and fixation of the intestine during fetal development [1]. Intestinal malrotation is a spectrum of anomalies that ranges from non-rotation to normal positioning [2]. Depending on the position of ligament of Treitz, malrotation may be classified into typical or atypical: Typical malrotation is when the ligament of Treitz is to the right of the midline while atypical malrotation is when ligament of Treitz is to the left of the midline [2]. Intestinal malrotation occurs in between 1 in 200 and 1 in 500 live births [3]. Symptoms of intestinal malrotation manifest in the early weeks of postnatal life. About 40% of malrotation is diagnosed within 1 week after birth and 75% to 85% within 1 year after

birth [4]. Embryologically, intestinal rotation and fixation of the fetal bowel occur between the 4<sup>th</sup> and 10<sup>th</sup> week of intra-uterine life. The fixation places the duodenojejunal loop to the left of the midline and the caecum in the right lower quadrant [5]. Variations or deviations from this normal process of intestinal development result in malrotation. Symptoms of intestinal malrotation may vary from one patient to another. In newborns, bilious vomiting is a typical presentation [6]. Older children may present with crampy abdominal pain, nausea and/or bilious vomiting [7]. An upper gastrointestinal contrast study is necessary for the diagnosis of intestinal obstruction [8]. Surgery for incidentally detected asymptomatic malrotation is controversial. In the presence of midgut volvulus and intestinal obstruction, surgery is mandatory [7]. The aim of this study was to evaluate our experience in the management of pediatric patients with intestinal malrotation.

## Materials and methods

This was a retrospective study of children aged 3 years and below who were treated for intestinal malrotation between January 2014 and December 2018 at the pediatric surgery unit of Enugu State University Teaching Hospital (ESUTH) Enugu, Nigeria. ESUTH is a tertiary hospital located in Enugu, South East Nigeria. The hospital serves the whole of Enugu State, which according to the 2016 estimates of the National Population Commission and Nigerian National Bureau of Statistics, has a population of about 4 million people and a population density of 616.0/km<sup>2</sup>. The hospital also receives referrals from its neighboring states. Patients with incomplete medical records and those who have had previous intestinal surgery were excluded from the study. Information was extracted from case notes, operation notes, operation register, and admission-discharge records. The information extracted included age, gender, presenting symptoms, interval between onset of symptoms and presentation, investigations performed, intra-operative finding, definitive operative procedure performed, complications of treatment, duration of hospital stay and outcome of treatment. Ethical approval was obtained from the ethics and research committee of ESUTH. Statistical Package for Social Science (SPSS) version 21, manufactured by IBM Cooperation Chicago Illinois, was used for data entry and analysis. Data were expressed as percentage, median, mean and range.

## Results

### Patients' demographics

A total of 65 cases of symptomatic intestinal malrotation in pediatric patients were seen during the study period but only 61 patients had complete medical records and form the basis of this report. Demographic features are shown in Table 1.

### Presenting symptoms

The patients presented with symptoms in various combinations (Table 2).

### Investigations performed

All the patients had plain abdominal radiograph and abdominal ultrasound. Forty-nine (80.3%) patients had upper gastrointestinal contrast study. None of the patients had abdominal CT scan. Plain x ray showed features of duodenal obstruction in 6 (9.8%) patients, abdominal ultrasound was suggestive of malrotation in 11 (18.0%) patients. A radiological diagnosis of malrotation was made through upper gastrointestinal contrast study in 47 (47/49) (95.9%) patients.

### Intra-operative finding and operative procedure performed

The intra-operative findings and operation performed are depicted in Table 3.

### Complications of treatment

Post-operative complications of the patients are shown in Table 4.

## Outcome of treatment

Fifty-three (86.9%) patients recovered and were discharged home. Eight (13.1%) patients expired. The causes of death were overwhelming sepsis and extensive bowel resection resulting in short bowel syndrome. Mortality occurred in 7 neonates and 1 infant. More males died, male: female ratio (5:3). Mortality was more in patients with intra-operative finding of midgut volvulus. Two of the patients that died had intestinal resection while the other 6 patients presented after a mean period of 6 months from the onset of symptoms and they died from fulminant respiratory system infections. The high mortality recorded in the present study could be attributed to late

**Table 1:** Demographic profile of the patients.

Gender	Male	46 (75.4%)
	Female	15 (24.6%)
Age range	10 days to 3 years (median 12 months)	
Interval from symptom onset to presentation	24 hours to 12 months (mean 7 days)	
Age groups		
	Neonates (< 1 month)	36 (59.0%)
	Infants (1-12 months)	18 (29.5%)
	12 months to 3 years	7 (11.5%)
Mean duration of hospitalization	15.3 days (range 11-23)	

**Table 2:** Presenting symptoms of the patients.

Presenting symptoms	Number of patients (%)
Abdominal pain + vomiting	35 (57.4)
Abdominal pain + distension	15 (24.6)
Abdominal pain + lack of weight gain	10 (16.4)
Non-specific symptoms	1 (1.6)

**Table 3:** Findings at surgery and operation performed.

Findings at surgery	Number of patients (%)	Operation performed
Intestinal obstruction by bands	37 (60.7)	Ladd's procedure
Midgut volvulus	Viable	Volvulus reduction
	Non-viable	Bowel resection and anastomosis
Incomplete rotation	7 (11.5)	Ladd's procedure
Non-rotation	1 (1.6)	Ladd's procedure
Incomplete fixation	1 (1.6)	Ladd's procedure

**Table 4:** Post-operative complications.

Complications	Number of patients (%)
None	52 (85.3)
Surgical site infection	4 (6.6)
Adhesive intestinal obstruction	3 (4.9)
Anastomotic leak	1 (1.6)
Recurrent volvulus	1 (1.6)

presentation. In a resource constrained environment like ours, lack of parenteral nutrition and lack of intensive care facilities for close monitoring of patients may also be responsible.

## Discussion

The first reports of intestinal malrotation were surgical and autopsy findings. The first description of the embryologic process of intestinal rotation and fixation was published in 1898 [9]. In 1923, Dott described the relationship between embryologic intestinal rotation and surgical treatment [10]. In 1936, William Edward Ladd wrote the classic article on treatment of malrotation. His surgical approach, Ladd's procedure, remains the cornerstone of the treatment of intestinal malrotation [11]. The clinical significance of intestinal malrotation is duodenal obstruction and midgut volvulus.

In the present study, there is male predominance. This is consistent with the report of other authors [12, 13]. However, Husberg, et al. reported female predominance [14]. Ameh, et al. reported equal number of males and females [15]. The reason for the gender variations is not known. Most cases of intestinal malrotation become symptomatic within 1 year of age. The median age of our patient was 1 year. A series from a single institution involving 170 patients reported that majority of their patients were infants [16]. Howbeit, one study from Lagos, Nigeria reported a median age of 7 years [17]. Seventy-five percent of symptomatic cases of intestinal malrotation occur in newborns and up to 90% of cases occur within 1 year of life [18]. Symptoms of intestinal malrotation can occur at any age: Hence, the need for high index of suspicion. Time of presentation of patients with intestinal malrotation is of essence. For instance, in midgut volvulus, it is a race against time. The late presentation of our patients is evident in the 7 days lag period before presentation. Lack of awareness and poverty on the part of the parents may explain the delayed presentation. More than 50% of our patients were neonates. Spigland et al also reported that about 50% of their patients who underwent surgery for malrotation were in the first month of life [19]. Another study documented that up to 80% of their patients were neonates [20]. The predominance of neonates in intestinal malrotation may be explained by the congenital nature of intestinal malrotation. The duration of hospitalization of our patients is comparable to the report of Ooms, et al. [21]. Post operatively, the length of time patients with malrotation stay in the hospital may depend on the modality of treatment and extent of the operative procedure performed. Laparoscopic treatment is associated with reduced period of hospitalization [21].

Abdominal pain was the most common and consistent symptom in our patients. Other series on malrotation also reported abdominal pain as the most common symptom [14,17]. The abdominal pain may be chronic and crampy due to partial intermittent intestinal obstruction or may be severe and acute in cases of midgut volvulus [22]. Non-specific symptoms such as vomiting, diarrhea, bloating, dyspepsia and early satiety have been reported in patients with intestinal malrotation. Some patients have been labeled as having functional or psychiatric disorders [23].

In cases of duodenal obstruction by Ladd's band, the double bubble sign is seen on plain abdominal radiograph. This double bubble sign was observed in one-tenth of our patients and is produced by an enlarged stomach and proximal duodenum. About one-fifth of our patients had reversal of the relationship between superior mesenteric artery and vein (Whirlpool) sign. This sign is mostly seen in midgut volvulus due to the twist of the superior mesenteric vessels. The upper gastrointestinal series is the criterion standard required for the diagnosis of intestinal malrotation [24]. The high sensitivity of upper gastrointestinal series reported by other studies is in line with the report of the present study [24, 25]. It is important to note that upper gastrointestinal series is to be obtained in patients who are hemodynamically stable. Hemodynamically unstable patients with midgut volvulus should be operated on promptly. Computed Tomography (CT) scan was not obtained in our patients because of its non-availability, cost and high risk of radiation exposure.

Intestinal obstruction caused by congenital bands or volvulus was the most common finding at surgery. Dekonenko et al. also reported intestinal obstruction as the most common sequel of intestinal malrotation [26]. Intestinal obstruction may be acute like in volvulus or chronic like in chronic duodenal obstruction caused by congenital (Ladd's) bands. Incomplete fixation of the bowel (malrotation) may predispose to intussusception. This association between malrotation and intussusception is called Waugh syndrome [1]. Reversed rotation, one of the rarest form of intestinal malrotation, in which the jejunum and transverse colon lie posterior to the superior mesenteric vessels was not seen in any of our patients.

The operative procedure mostly performed for malrotation was the Ladd's procedure. Ladd's procedure summarily entails counterclockwise detorsion of the bowel, surgical division of Ladd's bands, widening of the small bowel mesentery, performing an appendectomy and reorientation of the small bowel on the right and cecum/colon on the left side of the abdomen.

Majority of our patients did not develop any complications. Surgical site infection was the most common complication in the current series. Other studies also recorded surgical site infections following surgery for malrotation [15,27]. The incidence of wound infection is less in laparoscopic surgery than in open surgery [28]. As recorded in one of our patients, recurrent volvulus may occur following treatment for midgut volvulus. Alkan et al. also reported recurrent midgut volvulus [29]. Recurrent midgut volvulus requires an emergency redo-laparotomy for untwisting of the volvulus.

Mortality of 13.1% recorded in the present study is similar to a report from northern Nigeria [15]. Howbeit, Anand et al. reported a mortality of 2.5% in their series [18]. Mortality following treatment for intestinal malrotation may depend on age at presentation, presence of associated anomalies, interval between onset of symptoms and surgery and presence of necrotic bowel [30].



## Limitations of the study

1. Small number of patients. A larger number of patients would have availed better analysis
2. This a single center experience which may not be generalizable.
3. Detailed statistical analysis such as chi square test and t test were not done.

## Conclusion

Intestinal malrotation symptoms occur mainly in males and abdominal pain is a common and consistent symptom. Upper gastrointestinal contrast study is usually diagnostic. There are morbidity and mortality associated with the treatment of intestinal malrotation. Early presentation and referral is recommended for patients with symptoms suggestive of intestinal malrotation to avoid bowel gangrene.

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