

## Intestinal Malrotation: Presentation in the Older Child

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### ABSTRACT

**Background:** The clinical diagnosis of intestinal malrotation in the older child is not always easy because of its non-specific presentations. The aim of this study was to determine the pattern of presentation of malrotation in older Nigerian children.

**Methods:** The clinical, radiological and operative records of all the children aged 2 years or above, managed for malrotation at the Jos University Teaching Hospital between March 1992 and December 2002 were retrospectively reviewed.

**Results:** There were 9 patients, with a median age of 5 years (range: 3-14 years). The commonest complaint was intermittent colicky abdominal pain in 9 (100%), followed by recurrent vomiting in 8 (88.9%), haematemesis and constipation each in 5 (55.6%) and repeated episodes of bloody stools and diarrhoea. Other features included abdominal distension in 5 (55.6%) and failure to thrive in 4 (44.4%). Preoperative diagnosis was possible only in 3 patients, through the use of barium meal. Operative findings included obstructing bands of Ladd, partial volvulus and mesocolic hernias. Surgery promptly and satisfactorily relieved the symptoms.

**Conclusion:** The diagnosis of intestinal malrotation should be considered in any child with prolonged history of recurrent colicky abdominal pain, vomiting or diarrhoea, especially if there is associated history of failure to thrive. Surgical intervention provides satisfactory relief of symptoms and should be implemented as soon as the diagnosis is made.

**KEY WORDS:** Intestinal malrotation; older child; diagnosis.

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### INTRODUCTION

Abnormalities of intestinal rotation and fixation are understood in relation to the normal foetal intestinal development. Malrotation of the

intestine occurs when the normal rotational process and fixation of the intestine fail to take place. Intestinal obstruction from midgut volvulus, congenital peritoneal bands, or mesocolic herniation may be a sequel. A wide spectrum of clinical features has been ascribed to malrotation<sup>1-3</sup>. Majority of cases of malrotation presents in the neonatal period, but some may escape diagnosis until later in life<sup>4,5</sup>. In older child, the presentation may be atypical, causing difficult diagnosis and continued morbidity. We wanted to know the pattern of presentation of intestinal malrotation in the older child ( $\geq 2$  years old) over a 10-year period at the Jos University Teaching Hospital, in north central Nigeria.

### PATIENTS AND METHODS

This was a retrospective review of the clinical records of all children aged 2 years and above, diagnosed to have intestinal malrotation at the Jos University Teaching Hospital (JUTH) between March 1992 and November 2002. In the context of the present study, 'older children' refers to those aged 2 years and above and this age group formed the focus of the study. The data were analysed for patient's age, sex, clinical features, investigations, and outcome of definitive management.

### RESULTS

A total of 22 children with intestinal malrotation were managed within the period of the study, out of whom 9 (all males) fulfilled the inclusion criteria for the study. Their median age was 5 years (range: 3-14 years). Their age distribution is shown in Table I.

The clinical features in the 9 children are shown in Table II. The commonest presentation was recurrent colicky abdominal pain or discomfort in 9 (100%). In 7 of these patients, the pain usually started a few hours after meal, with no known relieving factor.

Recurrent vomiting was the complaint in 8(88.9%) patients. The vomiting was bilious in 5 and non-bilious in 3. Occasional haematemesis was present in 4(44.4%)

patients. Vomiting was described as projectile in 5(55.6%) patients and non-projectile in 3(33.3%). Recurrent constipation and episodes of bloody diarrhoeas was present in 5 and 2 patients, respectively. Seven patients were small for their ages.

Plain abdominal x-rays were not diagnostic in any of the patients. The oesophagogastrosopic findings in the 5 patients with haematemesis were normal; barium meal and follow through in these 5 patients confirmed malrotation (duodenojejunal junction on the right side of the vertebral column) only in 3 patients.

All the patients had exploratory laparotomy. Pre-operative diagnosis was certain only in 3 patients. Three patients had incomplete rotation of the duodenojejunal loop with complete rotation of the caecocolic loop. Both the duodenum and the proximal jejunum were entrapped behind the ascending colon and caecum by numerous fibrous bands passing from the caecum and proximal colon across the duodenum to the posterior right upper abdomen. Four patients had partial midgut volvulus with multiple mesenteric lymph adenopathy, peritoneal bands, and narrow mesenteric base. The adhesions were divided and derotation of the intestinal loops was affected in the counter-clockwise direction, followed by an appendectomy to avoid diagnostic difficulty in case there is appendicitis later in life. The caecum and proximal colon were left in the left side of the abdomen without caecopexy. The small bowel loops were left in the right side of the abdomen.

Two patients had internal (mesocolic) hernia. In these patients, the unsupported area of the descending mesocolon between the inferior mesenteric vein and the posterior parietal colonic attachment formed a cul-de-sac that contained small bowel (left mesocolic hernia). The caecum was completely rotated in these 2 patients. About 8cm and 12cm respectively, of the terminal ileum were free between the sac and the ileocaecal valve. In the case of the mesocolic hernia, the bowel was reduced through the neck of the sac. All adhesions were divided. The peritoneum adjacent to the inferior mesenteric vein was stitched to the posterior peritoneum and the neck of the sac was closed.

All the patients showed marked improvement after the procedures and are presently well. There was no postoperative morbidity or mortality.

**Table I. Age distribution of 6 children with malrotation seen at JUTH in the year 1992-2002.**

Age (years)	No. of patients	%
3	2	27.2
4	1	11.1
5	3	33.4
7	1	11.1
11	1	11.1
14	1	11.1
Total	9	100%

**Table II. Clinical features of 9 children with intestinal malrotation at JUTH in the year 1992-2002.**

Clinical features	No. of patients	%
Recurrent colicky abdominal pain	9	100
Intermittent vomiting	8	88.9
Repeated haematemesis	5	55.6
Constipation	5	55.6
Diarrhoea	2	22.2
Failure to thrive	4	44.4
Abdominal distension	3	33.3
Occasional visible peristaltic waves	5	55.6

## DISCUSSION

The accurate incidence of intestinal malrotation is difficult to estimate because of varied presentations. However, an incidence of 1 in 500 live births is commonly reported<sup>6</sup>. All our patients were males. Male dominance in malrotation presenting in the older age group has been reported by other authors<sup>3, 7</sup>.

The clinical features of malrotation in this study were similar to those in earlier reports<sup>3, 8-10</sup>. Majority presented with features of partial obstruction, namely recurrent colicky abdominal pain, intermittent vomiting and constipation. The diagnosis of partial obstruction requires a high index of suspicion, because the features are non-specific. This is particularly important in the tropics where such presentation is likely to be blamed on worm infestations or other bacterial infections.

Vomiting was bilious in 5 (55.6%) patients, indicating upper gastro-intestinal obstruction. The non-bilious vomiting in the remaining 3(33.3%) could have been erroneously attributed to other causes like worm infestation or extra gastro-intestinal causes. This study and others<sup>3,8</sup>, however, showed that non-bilious vomiting does not exclude malrotation. Non-bilious vomiting due to malrotation may be of gastro-intestinal origin when there is a pre-ampular duodenal obstruction or of extra gastro-intestinal origin when there is a systemic irritation or upset from the dilated bowel above the obstruction. Thus relying on the presence of bilious vomiting may lead to delayed diagnosis and catastrophic consequences. Passage of bloody stool or haematemesis in a patient with malrotation has been regarded as an ominous sign because it is an indication of compromise of the intestinal circulation, usually due to a volvulus<sup>3</sup>. This was our experience in four (44.4%) and 2 (22.2%) of our patients who had episodes of haematemesis and bloody diarrhoea respectively. The bowels in our series were, however, viable probably because the patients had partial volvulus. It is possible that the volvulus in these patients were complete initially, but partially untwisted before or at the time of operation. Whatever the case, such patients require prompt and appropriate treatment because delay in intervention may result in bowel infarction, gangrene and loss.

Failure to thrive is a known feature of chronic malrotation and was present in 4 of our patients. Malabsorption has been implicated in failure to thrive patients with malrotation<sup>3,4</sup>. It is thought that malabsorption occurs in patients with intestinal malrotation when chronic volvulus or intrinsic narrowing of the root of the small bowel mesentery results in chronic venous and lymphatic congestion of the bowel wall<sup>4,9,11</sup>; a feature seen in 4 of our patients.

Expedient work up, especially radiological imaging and adequate resuscitation is imperative in management of a patient with disorders of intestinal rotation<sup>1</sup>. Plain radiographs of the abdomen may reveal a proximal gastric or duodenal distension, with or without intra-luminal air or multiple air fluid levels in the central upper abdomen if there is obstruction. A duodenal triangle has been

reported as a plain film sign of midgut malrotation and volvulus. These triangular gas shadows located in the right upper quadrant are produced by the liver edge overlying the air-filled duodenum, and may be best demonstrated by an upright film<sup>1</sup>. We did not see any of these radiographic features in this study.

It is advised that one should proceed with contrast studies in patients where malrotation and volvulus are being suspected, provided there is no evidence of peritonitis<sup>1,3</sup>. Though the use of barium enema has been advocated<sup>6,13</sup>, a barium meal and follow through is considered the diagnostic study of choice in these patients presenting non-acutely<sup>7,14</sup>. This test may show abnormal position of the duodenojejunal junction usually to the right of the midline, but occasionally to the left; obstruction of the second or third part of the duodenum with a "Corkscrew" or "beak" appearance; and a proximal jejunum located on the right side of the abdomen<sup>1,4</sup>. This upper gastro-intestinal series revealed the duodenojejunal junction to the right side of the midline in 5 of our patients. Although barium aspiration is a potential risk with this procedure<sup>3</sup>, none of our children who had this study aspirated.

The use of the barium enema is based on the understanding that it may outline the caecum and the proximal colon in the left flank. The pitfall is that malrotation is associated with a normally positioned caecum in 5-20% of cases<sup>15,16</sup>. This was our experience in this study; 22.2% of our patients with malrotation and mesocolic hernia had their caecum in the right iliac fossa.

Sonography, computed tomography and magnetic resonance imaging have been added to the diagnostic armamentarium of malrotation by some centres. The presence of sonographic "whirlpool or barber pole sign" is diagnostic of malrotation with volvulus<sup>1,17</sup>. However, we have no experience with the use of these imaging modalities in the diagnosis of malrotation.

In 3 of our patients, the diagnosis was certain pre-operatively, but in the remaining 6, it was not possible to diagnose the malrotation before surgery. This is in consonance with other reports where majority of non-

complicated malrotations in the older children are confirmed only at surgery<sup>10, 18</sup>. Division of adhesions and/or release of hernias were sufficient to relieve the chronic symptoms in 5 of our patients without volvulus. However, Ladd's procedure was necessary to achieve relief of symptoms in those with partial volvulus. We routinely did not suture-plex the caecum in its new position in the left lower quadrant, although prophylactic fixation of the intestine in the (malrotated) position has been advocated<sup>19</sup>. It has been shown, however, that such fixation is unnecessary and absolutely offers no benefit<sup>20</sup>.

The pre-operative diagnosis of intestinal malrotation in the older child requires a high index of suspicion based on the history of recurrent abdominal pain, vomiting, constipation or diarrhoea, especially when there is associated failure to thrive.

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