

# Malrotation of the Midgut Associated with Horseshoe Kidney Presenting as Gastric Outlet Obstruction in a 15-year-old boy

Chigbundu Collins Nwokoro, E. A. Emmanuel<sup>1</sup>, A. A. Olatunji<sup>2</sup>, B. A. Salami, L. O. Amosu, I. O. Ogundele

Department of Surgery, Paediatric Surgery Unit, Olabisi Onabanjo University Teaching Hospital, Departments of <sup>1</sup>Anaesthesia and Intensive Care and <sup>2</sup>Radiology, Olabisi Onabanjo University Teaching Hospital, Sagamu, Ogun State, Nigeria

## Abstract

Malrotation occurs in approximately 1 in 500 live births. However, the true incidence of malrotation is unknown since many asymptomatic patients fail to present. Approximately 90% of patients with malrotation are diagnosed within the 1<sup>st</sup> year of life. Eighty per cent of them are diagnosed within the 1<sup>st</sup> month of life. Nevertheless, there are recent reports of manifestations later in life both as emergency conditions and more chronic gastrointestinal symptoms. The relationship between malrotation and horseshoe kidney has not been fully understood, but few case reports have highlighted their occurrence in the same patient. The mode of presentation of this case and its association with a horseshoe kidney is the reason for this report. This was a case of malrotation associated with horseshoe kidney. He had exploratory laparotomy and Ladd's procedure. Malrotation is associated with horseshoe kidney which presented as gastric outlet obstruction. He responded well to treatment after Ladd's procedure.

**Keywords:** Gastric outlet obstruction, horseshoe kidney, malrotation

## INTRODUCTION

Malrotation of the intestinal tract is a congenital anomaly referring to either lack of or incomplete rotation of the foetal intestines around the axis of the superior or mesentery artery during foetal development. The malrotation of the gut and abnormal location of the caecum produces a narrow superior mesenteric vascular pedicle, as opposed to the normally broad-based small bowel mesentery. Approximately 85% of malrotation cases present in the first 2 weeks of life.

It is estimated to occur in one out of every 500 live birth. However, presentation of intestinal malrotation is rare and its incidence has been reported to be between 0.2% and 0.5% in adulthood.<sup>[1]</sup>

The majority of children with malrotation develop symptoms within the 1<sup>st</sup> year of life. Intestinal malrotation is most often recognised in infancy, as most infants develop symptoms of acute bowel obstruction within the 1<sup>st</sup> week of life.<sup>[2]</sup>

The true incidence of malrotation in older children or adults is unclear, because a number of patients may be asymptomatic,

and in adults or older children, the difficulty of diagnosis results from both the absence of specific physical findings and the low frequency in these groups of patients.<sup>[3]</sup>

Overall, about 50% present in the 1<sup>st</sup> week of life and more than 60% before the end of the 1<sup>st</sup> month; the most frequent symptom is bile-stained emesis. Malrotation occurs equally in boys and girls. However, more boys become symptomatic by the 1<sup>st</sup> month of life than girls. This condition is diagnosed in 50% of patients by age 1 month and is diagnosed in 75% by age 1 year. The remaining 25% of patients present after age 1 year and into late adulthood. Some may be recognised intraoperatively during other procedures or at autopsy.<sup>[4,5]</sup>

In about 70% of children with intestinal malrotation, there are other associated congenital malformations, including the following: gastroschisis, omphalocele and congenital diaphragmatic hernia, Hirschsprung's disease,

**Address for correspondence:** Dr. Chigbundu Collins Nwokoro,  
P. O. Box 445, NIPOST, Sagamu, Ogun State, Nigeria.  
E-mail: collinsnwokoro@gmail.com

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gastro-oesophageal reflux, intussusception and anorectal malformation. Approximately 50% of patients with duodenal atresia and 33% of patients with jejunoileal atresia have malrotation as an association.<sup>[4,6]</sup>

Malrotation may present with acute midgut volvulus, and most of the patients with acute midgut volvulus present in the 1<sup>st</sup> year of life. Although more than 60% have an acute onset, chronicity of symptoms for more than a year is a frequent finding in about 12% of patients.<sup>[7]</sup>

The primary presenting sign of acute midgut volvulus is a sudden onset of bilious emesis and is due to intermittent or partial twisting that results in lymphatic and venous obstruction. This may be associated with failure to thrive, especially when chronic torsion of the root of the mesentery leads to lymphatic obstruction and malabsorption. Other clinical features that the patients may present with include recurrent bouts of diarrhoea alternating with constipation, intolerance of solid food, obstructive jaundice and gastro-oesophageal reflux.<sup>[7,8]</sup>

Acute duodenal obstruction may be present, and this is usually seen in infants and is due to compression or kinking of the duodenum by peritoneal bands while the older child may present with chronic duodenal obstruction. The most common symptom in chronic duodenal obstruction is vomiting which is usually bilious. Internal herniation may also be a feature, and patients with this condition usually have recurrent abdominal pain, which may progress from intermittent to constant, vomiting and constipation. Findings on physical examination will depend on the type of malrotational defect, the presence of complications and other congenital malformations.<sup>[9,10]</sup>

The age at diagnosis ranges from infancy to preschool age, however, some people who have malrotation go through their entire life without having any symptoms and are never diagnosed, while others may not have symptoms until adolescence or adulthood.<sup>[11]</sup>

The diagnosis of malrotation and volvulus should always be kept in mind when assessing any infant or child with symptoms of vomiting and pain.<sup>[11]</sup> Investigations that may contribute to early diagnosis of this condition include plain abdominal radiograph, barium meal, abdominal ultrasound and abdominal computerised tomography scan. Occasionally, barium enema could be used as an investigative tool because it could help in confirming the location of the caecum. However, barium meal is more accurate in defining the size, shape, rotation and presence of obstruction in the duodenum or upper jejunum.<sup>[12,13]</sup>

Abdominal computerised tomography scan, ultrasound and Doppler flow help to characterise the positional relationship of the mesenteric artery and vein. Laparoscopy may also be used to confirm or exclude the diagnosis in selected cases and may also be used for treatment.<sup>[14]</sup>

Diagnosis in the older child becomes more difficult, as the differential diagnostic spectrum of symptoms of abdominal pain, vomiting and causes of acute abdomen becomes wider.

However, there are a small proportion of affected adults who may present with acute or chronic symptoms of intestinal obstruction or intermitted and recurrent abdominal pain. The true diagnosis in adolescents and adult age group is more difficult, especially because the typical presentation is with non-specific symptoms and the fact that adult surgeons usually have low index of suspicion and may not consider the diagnosis in the initial evaluation of adult patients with abdominal pain.<sup>[15,16]</sup>

The treatment of choice is Ladd's procedure of derotation of the bowel if twisted, division of the peritoneal attachments lying across the duodenum from caecum to right upper quadrant, taking down of the ligament of Treitz and moving the duodenum to the right, separation of any adhesions between bowel loops. Since the appendix is located in an unusual position, it would be difficult to diagnose acute appendicitis in the future; therefore, an appendectomy is also usually performed.<sup>[17]</sup> We are presenting this case because of the unusual mode of presentation and its association with horseshoe kidney.

## CASE REPORT

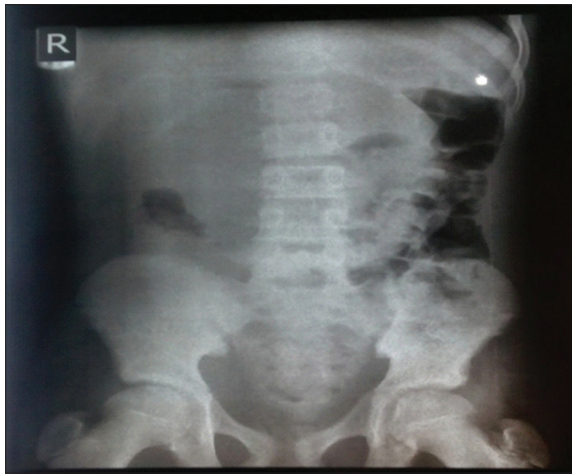
The index patient was a 15-year-old male who presented at the children emergency room with a 3-week history of recurrent colicky epigastric abdominal pain. The pain progressively increased in intensity and frequency. The pain was usually associated with epigastric abdominal swelling which disappeared once the pain subsided. There was associated projectile vomiting which was most times bilious. There were weight loss and anorexia, no abdominal distension, constipation, nor fever. He was not a known peptic ulcer disease patient.

On examination, he was acutely ill-looking with intermittent painful distress, not pale, but mildly dehydrated. His pulse rate was 80 beats per minute and respiratory rate was 22 cycles per minute. Chest and cardiovascular examinations were not remarkable.

The examination of the abdomen revealed fullness in the epigastrium with visible peristaltic waves in the epigastrium during episodes of colics. There was mild tenderness in the epigastric region with hyperactive bowel sounds. Rectal examination was not remarkable. A provisional diagnosis of gastric outlet obstruction was made.

The patient was resuscitated with intravenous crystalloid, parenteral analgesics and nasogastric decompression. Serum electrolytes were within normal limits. Plain abdominal radiograph showed a paucity of bowel gases [Figure 1]. Abdominal ultrasound scan suggested jejunojejunal intussusception. Barium meal and follow through showed grossly dilated stomach and duodenal C-loop with duodenojejunal loop on the right side of the midline [Figure 2].

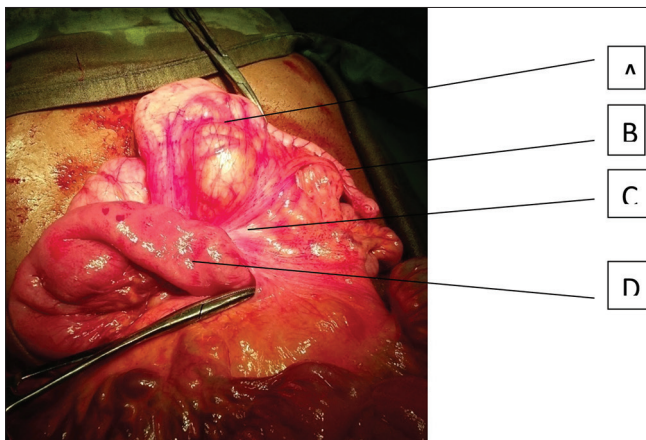
The patient had exploratory laparotomy done via upper midline incision. Intraoperative findings were dilated stomach and malrotated midgut with caecum in the epigastrium.



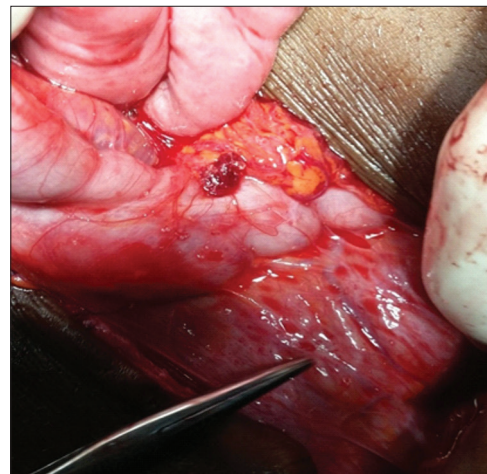
**Figure 1:** Plain abdominal radiograph showing paucity of intra-abdominal gases



**Figure 2:** Barium meal and follow through showing grossly dilated stomach and duodenal C-loop with duodenojejunal loop on the right side of the midline



**Figure 3:** Caecum (A), appendix (B) in the epigastrium and congenital band (C) obstructing the proximal jejunum (D)



**Figure 4:** The horseshoe kidney

Duodenojejunal junction was found on the midline in the epigastrium, congenital bands between the caecum and proximal jejunum 6 cm from duodenojejunal junction [Figure 3] and a horseshoe kidney [Figure 4]. The congenital bands were divided, twisted bowel was derotated and appendicectomy was done.

He had an uneventful post-operative care and was discharged home to the paediatric surgical outpatient clinic for follow-up. His parents were also counselled about the horseshoe kidney which would require close monitoring and prolonged follow-up.

## DISCUSSION

The index case was a case of malrotation associated with horseshoe kidney who presented with features of gastric outlet obstruction. Physical examination and results of plain abdominal X-ray, abdominal ultrasound and barium meal were suggestive of gastric outlet obstruction. However, emergency exploratory laparotomy revealed malrotation and horseshoe kidney. He had a Ladd's procedure and removal of

the appendix. His post-operative outcome was uneventful, and he had been followed up at the paediatric surgical outpatient clinic for about 10 months after discharge from the hospital and has remained symptom free.

Intestinal malrotation can be defined as any deviation from the normal 270° counterclockwise rotation of the midgut. During foetal development, the midgut supplied by the superior mesenteric artery grows too rapidly to be accommodated in the peritoneal cavity. Prolapse into the umbilical cord occurs around the 6<sup>th</sup> week. Between the 10<sup>th</sup> and 12<sup>th</sup> weeks, the midgut returns to the abdominal cavity having undergone a 270° counterclockwise rotation around the superior mesenteric artery.<sup>[18]</sup>

In malrotation, the midgut does not complete its normal lengthening and rotation and thus is incorrectly positioned within the peritoneal cavity. Normally, the process of lengthening and rotation begins between the 4<sup>th</sup> and 5<sup>th</sup> weeks of gestation. From this time until about week 10, the midgut is outgrowing the abdominal cavity and is forced to herniate

through the umbilicus to continue unhindered growth. During weeks 10 and 11, the intestine returns to the peritoneal cavity. From the 11<sup>th</sup> week forwards, the small bowel undergoes fixation.<sup>[19]</sup>

The small intestine is a straight tube early in development that derives its blood primarily from the Superior Mesenteric Artery. This vessel divides the midgut into two parts: the cephalad or pre-arterial portion and the caudal or post-arterial portion. The pre-arterial portion is made up of duodenojejunal loops, while the post-arterial portion is cecocolic loops. The SMA is important not only because it supplies the majority of blood flow to the small intestine, but also because it serves as the axis for the normal embryologic rotation of the bowel during development.

When the bowel herniates through the umbilicus, the pre-arterial portion rotates 180° counterclockwise around the axis of the SMA, while the post-arterial portion rotates 90° counterclockwise.

During the 10<sup>th</sup> and 11<sup>th</sup> weeks, the pre-arterial portion of the gut re-enters first followed by the post-arterial portion. While the bowel returns into the abdominal cavity, both segments complete a total turn of 270°. This configuration places the normal anatomy of the C-loop of the duodenum posterior to the superior mesenteric artery and the transverse colon anterior to the superior mesenteric artery.<sup>[20]</sup>

There is no typical set of symptoms that are ascribed to this clinical syndrome. The diagnosis of intestinal malrotation can be made by radiographic studies. Conventional radiography is neither sensitive nor specific for malrotation. Other different imaging modalities have become available to help guide the diagnosis and treatment of this surgical emergency. Plain abdominal radiographs are often the initial step in the imaging evaluation of patients with suspected malrotation. This relatively inexpensive and widely available test allows the radiologist and surgeon to exclude other potential diagnoses. Ultrasonography can also be used as an adjunct to plain film radiography by determining the position of the superior mesenteric vessels and the relationship to the third portion of the duodenum. In normal anatomy, the superior mesenteric artery lies left of the superior mesenteric vein; reversal of this relationship may suggest malrotation.<sup>[21,22]</sup>

The standard upper gastrointestinal barium series is considered the gold standard test to detect malrotation by most paediatric surgeons and remains accurate for detection with accuracy over 80%.

Important features in this text which are suggestive of malrotation or volvulus include low duodenojejunal junction position, absence of the duodenojejunal junction from its typical anatomical position which is to the left of the vertebral body or midline, jejunum located on the right side of the midline. Barium enema has also been employed to visualise the position of the caecum. The normally rotated caecum is found in the right lower quadrant of the abdomen, and up to 20%

of patients with malrotation will have a normally positioned caecum. This limitation makes barium enema an unpopular investigative modality.<sup>[17,21,23]</sup>

Computed tomography imaging can also be used to evaluate the position of the third part of the duodenum, the duodenojejunal junction and the anatomical relationship between the superior mesenteric artery and superior mesenteric vein.<sup>[24]</sup>

The standard treatment has remained Ladd's procedure of derotation of the bowel if torsion is present, division of the peritoneal attachments lying across the duodenum from caecum to right upper quadrant, separation of any addition between bowel loops, widening of the mesenteric base and finally returning the bowel to a position of non-rotation with the caecum placed into the left upper quadrant and removal of the appendix.<sup>[25]</sup>

These principles have remained the same since it was described by Ladd. In general, symptomatic patients with malrotation should be treated surgically. However, Spigland *et al.* recommended that all patients with malrotation are candidates for laparotomy, even if they are asymptomatic, because the complications associated with intestinal malrotation are based on anatomical reasons that do not alter with age, thus the potential to develop sudden onset of acute midgut volvulus in an asymptomatic patient at any age exist.<sup>[25,26]</sup>

Recently, laparoscopic techniques for treating malrotation in both infants and adults have been described. Laparoscopic management has been successfully performed recently in selected cases. Laparoscopic Ladd's procedure is feasible, safe and as effective as the standard open Ladd's procedure.

The need to apply any fixation sutures has been debated over the years. Some authors practice cecopexy, while others believe that adhesions and the broad-based mesentery would stabilise the bowel.<sup>[27]</sup>

Post-operative complications include recurrence and adhesive obstruction. However, these are treated surgically whenever they presented.<sup>[28]</sup>

The horseshoe kidney which was discovered incidentally at laparotomy was left untouched. However, its presence was discussed with the patient's parents in view of the higher incidence of associated pathologies such as vesicoureteric reflux disease, pelviureteric junction obstruction, urinary system calculi and urinary tract infection.<sup>[29]</sup>

## CONCLUSION

Malrotation is a congenital abdominal condition which usually manifests in infants but may also present during adolescence and adulthood as shown in the index case. Some patients with malrotation may live their entire lives without manifesting. The association between malrotation and horseshoe kidney has not been established, hence the need to report the index case.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

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