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Case Report

A RARE CASE OF INCOMPLETE ROTATION OF MIDGUT (STAGE II ARREST)

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ABSTRACT
Abnormalities in midgut rotation occur during the physiological herniation of the midgut between the 5th and 10th week of gestation (Warner, 1997). The most significant abnormality is a narrow small bowel mesentery which is prone to volvulus. This occurs most frequently in the neonatal period. Less commonly (Nehra and Goldstein, 2011), midgut malrotation presents in adulthood with either acute volvulus or chronic abdominal symptoms. It is the latter group that represents a diagnostic challenge. We report a case of a 19-year-old female patient who presented with a 6-months history of non-specific gastro-intestinal symptoms. After extensive investigation the patient was diagnosed with midgut malrotation following an upper gastro-intestinal series. The patient was treated with a open Ladd's procedure and at 3 months was gaining weight and had stopped vomiting. This case presented for its rarity.

Keywords: *Malrotation, Adult, Ladd's, Midgut*

INTRODUCTION
Malrotation is considered any deviation of the normal rotation of the midgut in embryological development (Warner, 1997), causing intermittent episodes of gastrointestinal obstruction or acute events of midgut volvulus. Although mainly a paediatric diagnosis, some cases do present in adult life. Radiologic investigations include: upper gastrointestinal contrast studies, Doppler sonography and a contrast enhanced CT of the abdomen. If a true malrotation is diagnosed or found by coincidence, a Ladd's procedure is advised (Bachur, 2006), even if the patient is asymptomatic. Here we report the stage II arrest of rotation of midgut and its management.

CASES
This 19 years old student was admitted to female medical ward for vomiting and pain abdomen. She was treated for acid peptic disease. She was referred to department of surgery for UGI scopy, which revealed severe distal esophagitis stomach filled with bilious fluid, no further examination was possible. She was advised conservative management and review. However she failed to improve and had persistent abdominal pain and vomiting with deterioration in general condition. Intestinal obstruction was thought of and she was sent for evaluation and repeat endoscopy. UGI scopy was repeated and findings were same and a differential of PARADUODENAL ILEUS? WILKE'S SYNDROME OR CONGENITAL MAL ROTATIONOF GUT was suggested to be ruled out. Patient was taken over to surgical side. She was ill built and nourished, with features of hypovitaminosis. Vital signs were within normal limits. Nutritional supplements were started.
Her abdomen was distended in the upper part. There was obvious prominent VGP. There was tenderness in upper abdomen, but no guarding or rigidity. There was no mass, or organomegaly. Hernia orifices were free. Bowel sounds were normal. Rectal examination was normal. Her laboratory investigation revealed hypokalemia which needed correction with KCl infusions. Other biochemical and haematological parameters were essentially normal. CXR was normal and revealed no dextrocardia. AXR was essentially normal except for large gastric air bubble. Thin barium meal series was done which revealed dilated stomach and barium filling 1st and 2nd part of duodenum which was distended (Figure 1.1). No barium found descending beyond 3rd part of duodenum. Barium was aspirated after the study and stomach wash given. Ultrasonogram was inconclusive due to air shadows (sonologist advised CT evaluation) CT scan abdomen revealed findings of midgut malrotation with duodenum lying anterior to superior mesenteric

artery (Figure 1.2). There was superimposition of superior mesenteric vein over superior mesenteric artery. No evidence of volvulus. She was taken up for surgery after informed consent.

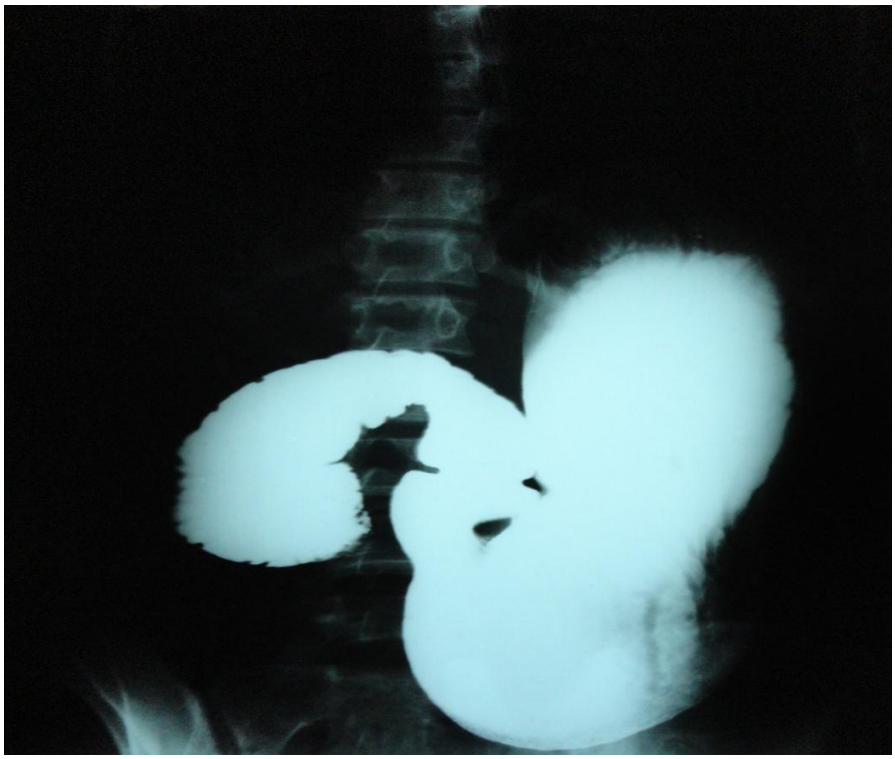

Figure 1.1: Shows the image of upper GI series with dilatation stomach and first part of duodenum

Figure 1.2: CECT image of midgut malrotation

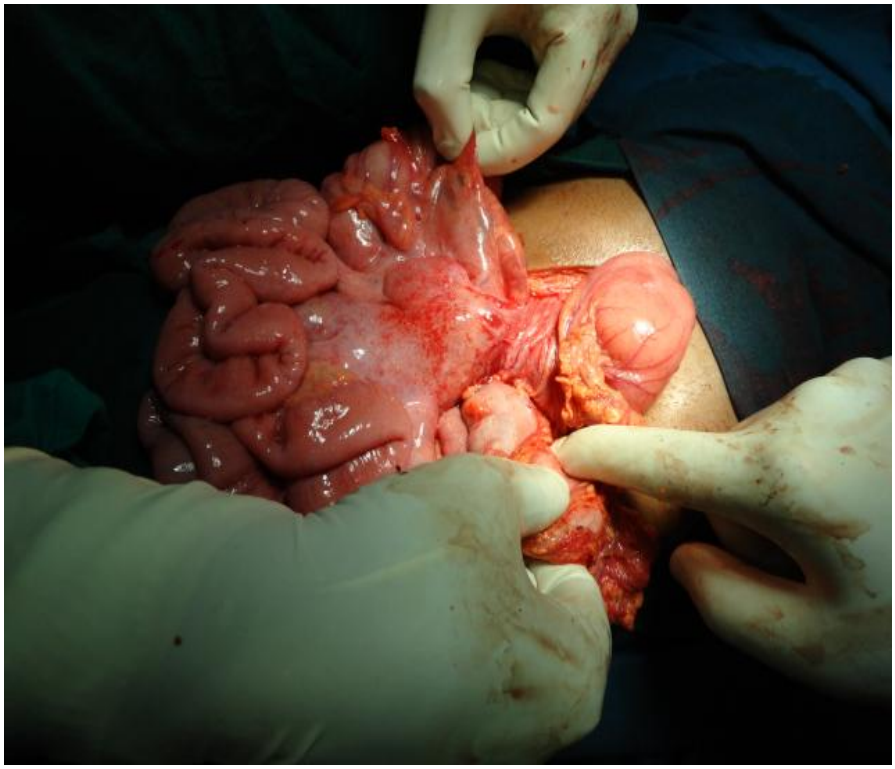

Figure 1.3: Shows intraoperative image of Ladd's band

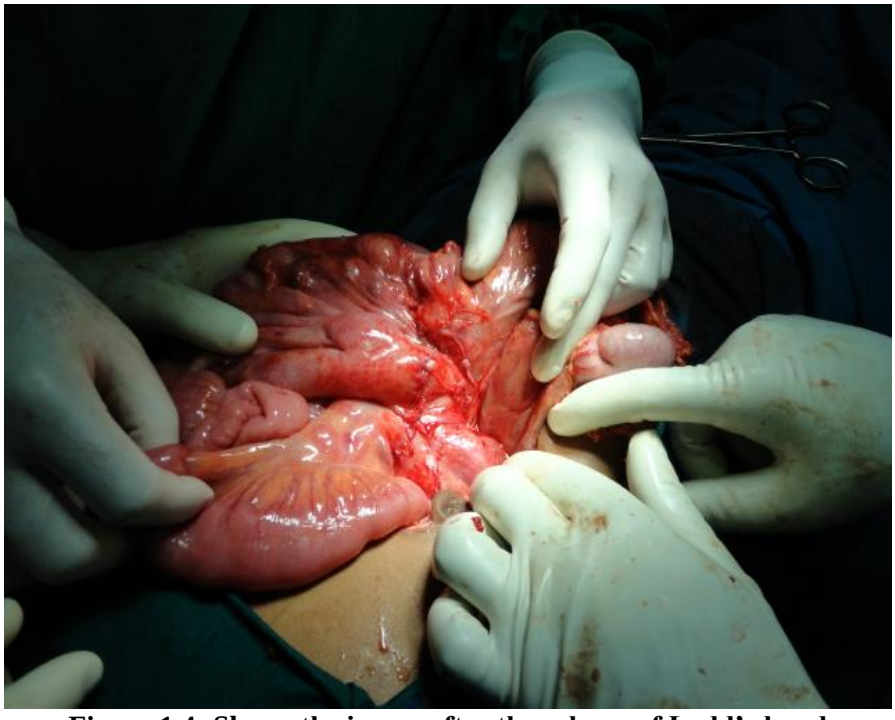

Figure 1.4: Shows the image after the release of Ladd's band

Exploratory laprotomy through midline incision was done after stabilising the patient under GA. Stomach and 1st and 2nd part of duodenum was found dilated, Ladd band was found running across 3rd part of duodenum compressing it. Distal jejunum and ileum were found anterior to superior mesenteric artery,

collapsed with no color changes suggestive of vascular compromise. Further distally ileocaecal junction was found lying in midline ending in mobile caecum and ascending colon. Transverse colon was found entering a narrow tunnel below superior mesenteric vessels. Splenic flexure and descending colon was found on left of abdomen with loose mesenteric attachment fixing it to postero lateral wall (Figure 1.4 & 1.5). Ladd band was released and superior mesenteric vein was found lying to the left of superior mesenteric artery. 3rd part of duodenum was completely released and mobilised to lie on right side of superior mesenteric artery. The transverse colon was carefully released from underneath the superior mesenteric pedicle to which it was adherent and was fixed to posterior peritoneum. Released colon was left lying on left of abdomen while small intestine was allowed to lie on right side. Appendicectomy was done. Abdominal wound was closed in layers. Post operative recovery was uneventful. NGT was removed on 2nd POD following establishment of bowel sounds. She was started on liquids and weaned to semisolids and solids. Patient recovered well. Sutures were removed on 10th POD.

DISCUSSION

Intestinal malrotation is defined as intestinal nonrotation or incomplete rotation around the superior mesenteric artery (SMA). It involves anomalies of intestinal fixation as well. Interruption of typical intestinal rotation and fixation during fetal development can occur at a wide range of locations; this leads to various acute and chronic presentations of disease. The most common type found in pediatric patients is incomplete rotation predisposing to midgut volvulus, which can result in short-bowel syndrome or even death. In 1936, William E. Ladd wrote the classic article on treatment of malrotation, and his surgical approach (ie, Ladd procedure) remains the cornerstone of practice today. Intestinal malrotation occurs at a rate of 1 in 500 live births. Most infants with gastroschisis, omphalocele, or congenital diaphragmatic hernia present with intestinal malrotation. Approximately 50% of patients with duodenal atresia and 33% of patients with jejunoileal atresia have a malrotation as well.

Also, intestinal malrotation occurs in association with Hirschsprung disease, gastroesophageal reflux, intussusception, persistent cloaca, anorectal malformations (imperforate anus), and extrahepatic anomalies. Male predominance is observed in neonatal presentations at a male-to-female ratio of 2:1. No sexual predilection is observed in patients older than 1 year. As many as 40% of patients, with malrotation, present within the first week of life. 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; many are recognized intraoperatively during other procedures or at autopsy. The history of present illness and clinical features varies in patients with intestinal malrotation according to acute or chronic presentation, as well as according to type of rotational defect. A full understanding of normal development aids in understanding the etiology of malrotation. Normal embryology: Normal rotation takes place around the superior mesenteric artery (SMA) as the axis.

It is described by referring to 2 ends of the alimentary canal, the proximal duodenojejunal loop and the distal cecocolic loop, and is usually divided into 3 stages. Both loops make a total of 270° in rotation during normal development. Both loops start in a vertical plane parallel to the SMA and end in a horizontal plane. ***Nonrotation***: Arrest in development at stage I results in nonrotation. Subsequently, the duodenojejunal junction does not lie inferior and to the left of the SMA, and the cecum does not lie in the right lower quadrant. The mesentery in turn forms a narrow base as the gut lengthens on the SMA without rotation, and this narrow base is prone to clockwise twisting leading to midgut volvulus.

The width of the base of the mesentery is different in each patient, and not every patient develops midgut volvulus. ***Incomplete Rotation***: Stage II arrest results in incomplete rotation and is most likely to result in duodenal obstruction. Typically, peritoneal bands running from the misplaced cecum to the mesentery compress the third portion of the duodenum. Depending on how much rotation was completed prior to arrest, the mesenteric base may be narrow and, again, midgut volvulus can occur. Internal herniations may also occur with incomplete rotation if the duodenojejunal loop does not rotate but the cecocolic loop does rotate. This may trap most of the small bowel in the mesentery of the large bowel, creating a right mesocolic (paraduodenal) hernia.

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***Incomplete Fixation*:** Potential hernial pouches form when the mesentery of the right and left colon and the duodenum do not become fixed retroperitoneally. If the descending mesocolon between the inferior mesenteric vein and the posterior parietal attachment remains unfixed, the small intestine may push out through the unsupported area as it migrates to the left upper quadrant. This creates a left mesocolic hernia with possible entrapment and strangulation of the bowel. If the cecum remains unfixed, volvulus of the terminal ileum, cecum, and proximal ascending colon may occur.Other rare anomalies of rotation can occur (Warner, 1997). These include. **Reverse rotation of the duodenojejunal limb resulting in a duodenum that rests anterior to the superior mesenteric artery. Reverse rotation of the cecocolic limb resulting in a transverse colon that is posterior to the superior mesenteric artery** Reverse rotation of the duodenojejunal limb with normal rotation of the cecocolic limb resulting in a paraduodenal hernia. In this anomaly, the duodenum is located anterior to the superior mesenteric artery. Anterior to the duodenum, the cecocolic limb rotates normally, and the mesentery of the right colon creates a pouch into which the small bowel can herniate. **Imaging Studies**-The following imaging studies may be helpful: 1.Plain abdominal radiography Plain radiography has limited use for defining obstruction because infants may have a gasless abdomen or one that is almost normal. The classic pattern for duodenal obstruction, if present, is the double-bubble sign produced by an enlarged stomach and proximal duodenum with little gas in the remainder of the bowel. Distended bowel loops and possibly pneumatosis intestinalis may be observed if necrotizing enterocolitis is present. If free air in the abdomen is a concern, obtain a left lateral decubitus radiograph as well. b) Upper GI series Upper GI series is the study of choice in patients who are stable. Normal rotation is present if the duodenal C-loop crosses the midline and places the duodenojejunal junction to the left of the spine at a level greater than or equal to the pylorus. If contrast ends abruptly or tapers in a corkscrew pattern, midgut volvulus or some other form of proximal obstruction may be present. Barium is the contrast of choice in patients who are stable or have chronic symptoms. Contrast studies may not be possible in patients who are actively vomiting or are otherwise unstable and need immediate surgical exploration. Water-soluble agents should be used if the study must be performed prior to imminent surgery. In this upper GI series with abnormal results, the duodenum does not cross the midline, and the small bowel is present only in the right side of the abdomen. C) Lower GI series (contrast enema) Occasionally, upper GI series findings may be indeterminate for the location of the duodenojejunal junction. In these cases, lower GI series may be used to identify location of the cecum. Lower GI series can also rule out colonic obstruction and ileal atresia. However, a normally placed cecum does not unequivocally rule out a malrotation, and clinical judgment must be exercised. D) Ultrasonography -In the hands of experienced ultrasonographers, ultrasonography has been shown to be very sensitive (approximately 100%) in detecting neonatal malrotation. Highest sensitivity is achieved when inversion of the superior mesenteric artery (SMA) and the superior mesenteric vein (SMV) is shown. Other diagnostic findings are fixed midline bowel loops and duodenal dilation with distal tapering. Also, volvulus is highly probable if the SMV is shown to be coiling around the SMA. All features are enhanced if water is instilled first by nasogastric (NG) tube. The presence of ascites and thickened bowel wall were not found to be statistically significant predictors of malrotation with midgut volvulus.

E) CT scanning -CT scanning is not well developed for diagnosing malrotation and midgut volvulus in children. In adults, abdominal CT is favored by experts because in addition to findings that indicate malrotation (eg, the third part of the duodenum does not pass between the superior mesenteric artery and the aorta), it also provides information concerning bowel location and perfusion, and the presence of volvulus. Furthermore, it is the preferred test for a variety of other causes of surgical abdominal pain in adults. In our case the diagnosis was clinched by CT. All the above findings could be detected and image recorded. **Medical Care**: Medical care of intestinal malrotation is directed toward stabilizing the patient.Maintain patients on nothing by mouth (NPO) and adjust nasogastric (NG) or orogastric tube to low intermittent suction. Correct fluid and electrolyte deficits. Administer broad-spectrum antibiotics prior to surgery, if possible. If a patient has signs of shock, administer appropriate fluids, blood products, and vasopressor medications to improve hypotension. In some centers dopamine is used as first-line

therapy because of its possible effects to increase splanchnic blood flow. It is advisable to routinely start dopamine at an infusion rate of 3 mcg/kg/min intravenously (IV) and continue it postoperatively even if the patient is not hypotensive. If the patient is unstable, do not delay surgical intervention for upper GI and laboratory studies. Quick surgical intervention, not prolonged medical management, produces the best results if midgut volvulus is suspected. **Surgical Care** -The Ladd procedure remains the cornerstone of surgical treatment for malrotation today. Prior to William Ladd's publication in 1936, surgical treatment for malrotation with or without volvulus had a mortality rate higher than 90%. In fact, at Ladd's own institution, the mortality rate was 100% before the development of his new technique. A classic Ladd procedure is described as reduction of volvulus (if present), division of mesenteric bands, placement of small bowel on the right and large bowel on the left of the abdomen, and appendectomy. Published reports for laparoscopic Ladd procedure are now appearing in the literature as well. Midgut volvulus- If midgut volvulus is present, the entire small intestine along with the transverse colon is delivered out of the abdominal incision, where the volvulus can be reduced. Because the volvulus usually twists in a clockwise direction, reduction is accomplished by twisting in a counterclockwise direction. After the blood supply has been restored by detorsion, the surgeon must make a decision about viability of the involved bowel. The outcome is better when no gangrenous bowel is present or when a small localized gangrenous segment is present, which can be resected and a primary anastomosis performed. Enterostomy is performed when questionable viability is observed at the ends of a gangrenous area that is resected. If multiple areas of questionable viability are present, many surgeons choose to leave the areas and perform a second-look operation in 12-24 hours if the patient is not showing clinical recovery. Duodenal obstruction After the volvulus is reduced or if no volvulus was present, identify any extrinsic obstruction to the duodenum. If peritoneal bands crossing the duodenum are found, ligate them with careful attention to protecting the superior mesenteric vessels. The bands may also obstruct the ileum or the jejunum and sometimes run to the gallbladder and liver. Extrinsic obstruction may also be due to the cecum, colon, or superior mesenteric artery (SMA) impinging on the duodenum; relief is obtained by placing the cecum with its mesentery in the left upper quadrant and exposing the anterior duodenum through its entire length. After extrinsic obstruction has been relieved, determine that no intrinsic obstruction exists by passing an NG tube through the duodenum. Appendectomy -Frequently, dissection of the peritoneal bands causes damage to the appendiceal vessels. Therefore, all pediatric surgeonsperform an appendectomy prior to closure. Appendectomy is also advisable because the normal anatomical placement of the appendix is disrupted when the cecum is placed on the left side of the abdomen. Laparoscopy: Laparoscopy has been used to repair malrotation with signs of duodenal obstruction but no midgut volvulus. The Ladd procedure, including widening of the mesenteric base and dissection of peritoneal bands, has been performed successfully and has resulted in shorter hospital stays. Laparoscopic Ladd procedure has been reported more frequently in the literature and is becoming more accepted as an initial approach to surgical correction. It has been noted that short-term results were superior with the laparoscopic approach and can be achieved without any increase in operative duration. Complications include the following: 1.Short-bowel syndrome: Short-bowel syndrome is the most common complication of midgut volvulus. These patients have longer delays to recovery of bowel motility and function. They are at high risk for malabsorption and can require very long-term parenteral nutrition. Furthermore, they have more complications from treatment and much longer hospital stays than patients with malrotation and no volvulus. Infection: Wound infections and sepsis can occur in the immediate postoperative period, requiring extended treatment with intravenous antibiotics. Patients may also have infection because of long-term placement of central venous catheters. Translocation of enteric bacteria and superimposed candidal infection further complicate the hospital course. Postsurgical: long-term complications after Ladd procedure to correct intestinal malrotation. Adhesive small bowel obstruction, require operative adhesiolysis, recurrent volvulus, and 3 patients died. The Ladd procedure has a low postoperative morbidity rate. A separate review of long-term sequelae after Ladd procedure found that only 14 of 161 (8.7%) patients developed complications following surgery; 9 patients developed adhesive small bowel obstruction, 5 patients required operative adhesiolysis, 1 patient developed recurrent volvulus, and 3

patients died. Reoperation: including volvulus of the cecum, recurrence of midgut volvulus, bowel obstruction due to adhesions, insertion of central venous catheter, abdominal wall cyst, and wound dehiscence. Persistent GI symptoms: including constipation, intractable diarrhea, abdominal pain, vomiting, and feeding difficulties.

Conclusion

Our case is a rare presentation of incomplete rotation of midgut (**Reverse rotation of the duodenojejunal limb resulting in a duodenum that rests anterior to the superior mesenteric artery. Reverse rotation of the cecocolic limb resulting in a transverse colon that is posterior to the superior mesenteric artery)** in a teenager who presented with abdominal pain and vomiting. Initially due to rarity of this condition malrotation of gut was never entertained as differential diagnosis. Barium and CT scan was very useful in confirming the diagnosis. This patient underwent Ladd procedure and presently is doing well. She need frequent monitoring for possible later complications including volvulus, adhesive obstruction.

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