Abstract:
Reversed rotation of mid gut the most unusual presentation of intestinal malrotation is responsible for only 2 to 4 of all rotational abnormalities of the midgut. It was first described in late 1800 and occurs when the midgut loop rotates wrongly in clockwise 90 rather than normal anti-clockwise 270. As a result, the duodenum and transverse colon are in an inverted position and transverse colon lies behind the superior mesenteric artery and anteriorly placed duodenum. Here we report a rare case of a pregnant lady with reverse rotation of gut presenting as midgut volvulus in second trimester for which surgery was done and the patient recovered uneventfully. Foetal outcome was unaffected by the surgery. We also present a review of the available literature.

Keyword: Reverse rotation, Volvulus, Pregnancy

INTRODUCTION
Congenital anomalies of intestinal rotation may produce curious and perplexing problems in the surgery of intestinal obstruction. Rotational gut abnormalities are generally considered diseases of the neonatal and pediatric populations. However, they may be symptomatic in teenagers and adults, often with disastrous outcomes. Symptomatic bowel malrotation first presenting in the adult is rare. Reversed rotation of mid gut, the most unusual presentation of intestinal malrotation is responsible for only 2% to 4% of all rotational abnormalities of the midgut. Reversed intestinal rotation is probably the rarest and most interesting, for in most of the recorded cases symptoms of the disorder were delayed well into adult life. Midgut volvulus is the most common complication of malrotation in the adult.

CASE REPORT
Fig-1, X-ray Abdomen erect Showing dilated bowel loop
A 25 yr old pregnant lady (G2 P1 L1) with 6 months amenorrhoea presented with Abdominal pain for 3 days which was insidious, progressive, intermittent, colicky type in the epigastric region, non–radiating. She had Vomiting for 3 days, 6-8 episodes / day biliary, non projectile. There was no history of malena, hematemesis, constipation or diarrhoea. There was no history of fever or similar illness in the past. She is not a known Diabetic or Hypertensive. She had not undergone any surgery in the past. Last child birth was 10 months back and it was uneventful. Physical Examination revealed Dehydration and Tachycardia 110/ min. Other vital parameters were normal. On Local Examination abdomen was distended, there was no guarding , Severe Epigastric tenderness was present. Fundus of uterus was palpable at the level of umbilicus. Foetal heart sound heard normally, No other mass palpable, No organomegaly , No free fluid , Bowel sound was Sluggish , Per Rectal Examination was not contributory . Other systems examination was normal. Basic investigations were within normal limits. X- ray abdomen showed markedly Dilated bowel loop in the epigastric region[Fig-1]. The diagnosis remained elusive until an USG Abdomen showed **Whirlpool sign in epigastric region**[Fig-2], SMA, SMV reversed & evidence of bowel mass suggestive of Malrotation of gut with midgut volvulus , with intra uterine foetus corresponding to 20 weeks of gestation.

Pre – op Diagnosis was Acute intestinal obstruction with Malrotation of gut with midgut volvulus. The patient was resuscitated with intravenous fluids and prepared for an emergency exploratory laparotomy. Intraop findings were Midgut volvulus[Fig-3] twisted clockwise thrice around its axis. Straight duodenum, **Transverse colon lying behind the duodenum & superior mesentric pedicle with absent mesocolon** (Retroduodenal/ Retroarterial)[Fig-4,5] Unduly mobile caecum & ascending colon with long mesocolon lying in right hypochondrium & epigastrium. Multiple bands between colon & small bowel[Fig-6]

**Fig-2,USG abdomen Showing Whirlpool Sign**
Detorsion of midgut volvulus , Adhesiolysis , Caecopexy[Fig-7] and Appendectomy were done.Retroduodenal transverse colon was left as such since transverse Colon was not constricted moved freely in the retroduodenal tunnel and symptoms were only due to volvulus.Patient had a good post op recovery started on liquids on POD 3, Semisolids POD 5, Passed stoolson POD 7, Sutures removed and discharged on POD 10. Post op foetal scan was normal. Foetal outcome was unaffected during the entire course. Patient is on regular follow up.
DISCUSSION
Midgut malrotation is a congenital anomaly in the embryological development of the foetal intestinal rotation. It has been estimated that it affects approximately 1 in 500 live births [1]. However, the true incidence is difficult to determine as a substantial number of cases will go undetected throughout life. The vast majority of the complications associated with midgut malrotation present in the first month of life and 60-85% of cases are diagnosed in this age group [1,2]. It is reported that more than 90% of patients will present by the time of their first birthday [3]. Adult midgut malrotation is very rare and its incidence has been reported to be between 0.0001% and 0.19% [3,4].

Normal embryonic development
Normal embryology of human intestine is based on studies of MALL, FRAZER & ROBBINS[26].

Fig-3, Intraop picture showing midgut volvulus
Normal midgut development consists of 3 recognized stages:

Physiological umbilical herniation, which begins in the fifth week of gestation. The midgut, supplied by the superior mesenteric artery, elongates and protrudes into the vitelline sac. While in the umbilical cord, the midgut loop normally rotates 90° counterclockwise around the axis of the superior mesenteric artery, placing the duodenum posterior to the transverse colon.

The physiological midgut hernia returns to the abdominal cavity in the tenth week. There the intestine revolves another 180° counterclockwise, completing a net 270° counterclockwise rotation to place the duodenum behind the superior mesenteric artery.

The cecum-ascending colon migrates to the right flank and the descending colon fixes to the left paracolic gutter.

The key features of normal intestinal rotation include (1) the duodenum describes a C-loop with concavity to the patient’s left and the third portion of the duodenum (at the ligament of Treitz) to the left of the midline; (2) the superior mesenteric artery runs in front of the third portion of the duodenum, which is in a retroperitoneal position; (3) the mesentery is attached posteriorly along a broad line that runs from the ligament of Treitz in the left upper quadrant to the cecum in the right lower quadrant, thereby preventing torsion of the mesentery on its axis, and (4) the colon describes a frame with the cecum and ascending colon fixed along the right side of the abdomen and the descending colon fixed along the left side.

As rotation of gut occurs in three stages, Derangement can occur in each stage leading to abnormalities. In first stage Exomphalos major, in Second stage Non rotation , Incomplete rotation & Reverse rotation and In Third stage Subphrenic caecum , Right lumbar caecum , Pelvic caecum ,Mobile proximal colon can occur.
REVERSE ROTATION
First reported by Tscerning in 1883[25]. Reversed rotation of the midgut, the rarest presentation of intestinal malrotation, accounts for only 2%–4% of all embryonic midgut rotational anomalies[5]. It is twice as common in women as men[6]. Reversed rotation occurs when the midgut loop erroneously rotates in a clockwise direction[7]. As a result, the duodenum and the transverse colon are reversed in position; the cecum and transverse colon lie posterior to the superior mesenteric artery and the duodenum lies anterior.

Fig-4, Showing retroduodenal transverse colon
Etiology of reversed rotation
In reversed rotation, a 90° clockwise rotation replaces the normal 270° counterclockwise rotation in the second stage. During fixation, the small bowel mesentery forms a narrow attachment to the posterior parietal peritoneum, and the transverse colon passes through the retroarterial tunnel, leading to a high risk of obstruction. The cecum and right colon incompletely attach to the peritoneum. Two variations of reversed rotation have been described in the literature[29]. In the more common Retroarterial type, the postarterial (caudal) segment returns to the abdomen first, positioning the transverse colon in a retroarterial tunnel behind the superior mesenteric vessels and duodenum with the duodenum crossing right to left anterior to the artery and the cecum placed on the right. The cause for this may be Any factor which allows caecum to slip back first into the peritoneal cavity, before small bowel returns – Dott [20]. Large umbilical opening will cause caecum to slip in first – Hunter [21]. Failure of DJ and hindgut loops to rotate normally and the extra – abdominal gut follows passively – Snyder and choffin[22] However, if the prearterial (cranial) segment is reduced first, it enters the abdominal cavity on the left, pushing the hindgut to the right, causing the entire colon to lie on the right side of the abdomen, and the cecum in the midline. In either case, the duodenum usually remains anterior to the superior mesenteric artery[8]. The second type has only been described in 6 patients[9], and in 2 cases was associated with situs inversus of other abdominal viscera. Reversed rotation has been found to coexist with other anomalies. One report was of a congenital heart condition in association with reversed rotation[9]. Another author encountered reverse rotation in association with anomalies of the biliary duct and gallbladder[10].

Fig-5, Showing Retroduodenal/Retroarterial Transverse colon Rare in adults
Abnormalities of midgut rotation, including nonrotation, malrotation, and reversed rotation, are most frequently found in neonates and children, accounting for 1% of all intestinal obstruction
They are rarely the cause of acute abdominal emergencies in adults. The infant with reversed rotation presents with acute intestinal obstruction requiring immediate surgical repair. Incomplete fixation of the cecum and the ascending colon predisposes to volvulus of the ileocecal segment on its narrow mesenteric attachment. Pressure of volvulus may cause obstruction of the duodenojejunal junction.

**Adult presentation** Unlike the other types of malrotation, most cases of reversed rotation present in adults[11]. Most adults with reversed rotation have a history of chronic intermittent abdominal pain, often with vomiting. They may also present with an acute episode of bowel obstruction in the transverse colon as it courses through its retroarterial tunnel or Acute presentation may be due to volvulus of the midgut or ileocaecum, reported as the most common cause of bowel obstruction in adults with gut malrotation as in our case. Vegetarian diet and pregnancy may be factors which lead to volvulus; a theory based on a number of cases found in vegetarian Indian villages and in women during pregnancy and puerperium [30]. Duodeno-jejunal obstruction has been rarely described. It is most commonly due to Ladd’s bands.[11]

**Diagnostic Tools**

Preoperative diagnosis of reversed rotation is rarely possible, although most cases are diagnosed surgically. Oblique and lateral films may show the duodenum anterior to the transverse colon. A mobile cecum and ascending colon should be visualized, due to incomplete mesenteric attachments. Upper GI series will show the duodenum located anterior to the transverse colon and right-sided course of the duodenum. Obstruction may be observed at the ileocecal segment from volvulus. Rarely, the duodenojejunal junction will demonstrate obstruction by periduodenal bands or herniation into the retroarterial tunnel.

**Fig-6, showing bands**

Characteristic USG findings of midgut volvulus was first described by Pacros et al. When reverse rotation is associated with midgut volvulus, duodenal dilatation with distal tapering and fixed midline bowel and mesentery twisted around the SMA axis is found on an USG. These features classically present as the ‘whirlpool’ sign [15]. Use of water-soluble contrast enema is appropriate to diagnose retroarterial reversed rotation[12]. The mid-transverse colon often has an area of narrowing where it passes behind the superior mesenteric artery. The transverse colon might display redundancy or kinking. The CT appearance of midgut volvulus is also diagnostic. The small bowel wrapped around the SMA creates a distinctive whirl pattern [15,16] as the volvulus, which causes the mesenteric veins and lymphatics to become congested. Mesenteric edema can also develop. The relationship of the superior mesenteric vein (SMV) to the SMA as shown by CT can suggest the diagnosis of bowel malrotation, although it is not pathognomonic [18].

**OPERATIVE MANAGEMENT**
If obstruction is at DJ level due to bands - Ladd’s procedure is done. Instead, if it was due to due to volvulus - De torsion of the volvulus and ladd’s procedure is done.

Fig-7, Caecopexy completed
The surgical management of intestinal malrotation was first described by William Ladd in 1936 [28] and this remains the mainstay of treatment. The classical Ladd’s Procedure consists of 4 parts: division of Ladd’s bands overlying the duodenum; widening of the narrowed root of the small bowel mesentery by mobilising the duodenum and division of the adhesions around the SMA to prevent further volvulus; counterclockwise detorsioning of the midgut volvulus if present and appendicectomy to prevent future diagnostic dilemma of an abnormally located appendix[28]. If obstruction is at transverse colon, Many operations have been devised to release the constriction at the retroarterial tunnel; these include colonic resection, displacement of the transverse colon anterior to the duodenum after colonic transsection and side-to-side anastomosis of the colon. Butler has suggested that the whole midgut be rotated through 180 degrees in an anticlockwise direction; this would produce the condition of intestinal nonrotation and would be an improvement[19]. Estrada and Gurd point out, patients with intestinal nonrotation are particularly liable to symptoms of intestinal obstruction and to volvulus[13]. Assuming that the retroarterial tunnel is formed post-partum and therefore has a bloodless plane of cleavage, they dissected this area and were able to turn the whole of the midgut through 360° in an anticlockwise direction, thus restoring the normal anatomy. The gut was then fixed to the posterior abdominal wall in its usual position.

This procedure would seem to be indicated in all cases where there is obstruction behind the superior mesenteric vessels. Manual dilatation of retroarterial tunnel and caecopexy was suggested by Truesdale[23].

CONCLUSION
Intestinal malrotation though being a rare condition should be considered in differential diagnosis of bowel obstruction in adults. The diagnosis of malrotation after childhood is difficult and usually not readily considered as the cause of intraabdominal symptoms. The presentation is usually nonspecific and this often leads to diagnostic and treatment delay with possible bowel ischaemia and necrosis. Evidence of which portends a poor prognosis and death. In our scenario where it was associated with pregnancy diagnosis becomes even more difficult. Therefore, a high index of suspicion needs to be maintained and prompt surgical intervention must be considered in order to prevent an abdominal catastrophe and fatality.

REFERENCES


