Journal of Population Therapeutics & Clinical Pharmacology

RESEARCH ARTICLE
DOI: 10.53555/jptcp.v31i7.7227

TWIST IN TALE – DELAYED INTESTINAL MALROTATION – A CASE SERIES

Dr. A. E. Poornima[1], Dr. D.Tamilselvan[2]*, Dr. C. Sabarigirinathan[3], Dr S. Balasiddharth[4], Dr. Dinesh Venkatesan[5]

[1]Assistant Professor, Department of General Surgery, ESIC Medical College, K.K.Nagar, Chennai 600078, Tamilnadu, India
[2]*Assistant Professor, Department of Urology, Kilpauk Medical College, Chennai 600010, Tamilnadu, India
[3]Professor and Vice Principal, Govt. Dental College, Chennai, Tamilnadu, India
[4]Department of Pharmacology, M.K.C.G Medical College, Berhampur, Odisha - 760 004, India
[5]Private Practitioner, Chengalpattu, Tamilnadu, India

***Corresponding Author:** Dr. D. Tamilselvan
*Assistant Professor, Department of Urology, Kilpauk Medical College, Chennai 600010, Tamilnadu, India
*Email: gyam351@gmail.com

ABSTRACT
Irregular rotation of the gut disrupts the establishment of the superior mesenteric artery inside the peritoneum during fetal rotation. A rotational abnormality of the midgut is rare in adults. In most cases, surgery is necessary for the treatment of symptoms. In spite of the challenges of diagnosis and surgery, treatment is usually successful when it is done early. The condition of misrotation of the intestine is usually discovered by accident in an adult without any symptoms. A literature review has been conducted on intestinal malrotations and small bowel obstructions.

KEYWORDS: Intestinal Malrotation, Delayed Presentation, Intestinal Obstruction, Intestinal Volvus, Ischaemia, Ladd's Procedure.

INTRODUCTION
Appendicitis is one of the most common abdominal conditions, often presenting with a varied range of clinical manifestations. However similar range of manifestations arises in an anatomical anomaly like Intestinal Malrotation.

Intestinal Malrotation is a congenital pathology with acute and catastrophic complications, such as Intestinal Volvus, with massive bowel loss[1]. During embryogenesis, intestinal malrotation occurs because the midgut rotates, fixes, and develops abnormally, resulting in a congenital anomoly[2]. This condition is primarily caused by genetic factors. Neonatal illnesses are most common, while adults with sudden onset of vomiting and intestinal obstruction are more likely to suffer from it.

Intestinal Malrotation was first described by Dr. LADD in 1930s, and treatment protocol is well covered in the surgical literature. A new technique was developed by Dr. Kareem called as Kareem's Procedure in 2021. In this article, we deal with our experiences in treating 3 cases of Intestinal Malformation.

An intestinal obstruction in neonates is reliably identified by malrotation with midgut volvulus. Symptoms of the condition can also include gastroesophageal reflux, early satiety, and mild abdominal discomfort[3].

EMBRYOLOGY AND GENETICS

A complex developmental process is taking place with irrigation organs. Gastronomic tracts are formed as straight tubes during embryonic development. Midgut blood is supplied by the superior mesenteric artery (SMA) between the proximal bile duct entry and the mid-distal transverse colon[4]. Across the vitelline duct and SMA, the midgut is divided into two sections: the caudad and cephalad. Distal duodenum, jejunum, and proximal ileum belong to the cephalad midgut while distal ileum, cecum, and appendix belong to the caudad midgut. When embryos are developing, their midguts grow disproportionately to their sizes.

An embryonic coelom emerges from the midgut during pregnancy. Right SMA shows duodenojejunal loop movement, while the left SMA shows cecocolic loop movement [5]. With the progression of pregnancy, the midgut grows primarily in the cephalad region. Cecocolic loops are not affected by small bowel rotation counter clockwise.

In the 10th week of pregnancy, a pregnant woman's bowels reenter the abdominal cavity. Before entering the proximal small bowel, the distal small bowel is rotated 90 degrees counter clockwise, resulting in the C-loop configuration of the duodenum[6]. There is 2 C-loops surrounding the SMA: one on the inferior side and one on the posterior side. After this, the distal ileum, cecum, and proximal colon are rotated 180 degrees counterclockwise. Consequently, the colon follows its normal picture frame path, crossing anteriorly over the SMA and positioning itself to the right. Cecums occupy a different location in the right abdomen during this time.

It extends and descends in the right lower abdomen quadrant throughout pregnancy and early infancy. This stage involves the retroperitoneal fixation of the second, third, and fourth parts of the duodenum, as well as the Treitz ligament in order to anchor the junction between the duodenum and the jejunum. Mesenteric bands connect the mesenteries and retroperitoneum to anchor the bottom and top of colons[7]. There is a transverse attachment of the mesocolin to both the greater omentum and retroperitoneum. The transverse colon is attached to different mesenteric structures than the sigmoid colon. There is a junction between the duodenum and the ileocecal valve in the lower right abdomen[8]. While they are loosely attached to the posterior abdominal walls, their broad mesentery bases prevent them from volvulsing.

An inactivated FOXF1 transcription factor has been linked to intestinal malrotation. The respiratory system of patients with ACD/MPV has severe abnormalities during development. Describes a lung condition in which the intrinsic pulmonary vascular system is not developed adequately.

ABNORMAL EMBRYOLOGY

A variety of outcomes can result from arresting the embryonic midgut at any time during either or both loops. Rather than a single abnormality, malrotation refers to a spectrum of abnormalities in midgut development. A common mistake is to call non-rotation the position of the duodenum and distal colon when rotation is stopped early. Incomplete rotation refers to the small bowel and/or colon not rotating 180 degrees counter clockwise, which can be mistaken for malrotation. From skeletons that are completely non-rotative to nearly normal anatomy, this spectrum is broad[9].

There is often a correlation between volvulus and abnormally rotated bowel because of abnormal mesenteric attachment. In the duodenum, the caudal midgut returns to the abdominal cavity first as it rotates clockwise rather than counter clockwise. Unlike the duodenum, the colon snakes posteriorly rather than anteriorly and posteriorly[9]. Duodenal hernias can cause colonic rotation to reverse during herniation.

An obstruction in the intestine occurs when the midgut twists around the superior mesenteric artery at an angle greater than 720 degrees, thereby creating a volvulus. Symptoms of volvulus include obstructing lymphatic drainage, venous drainage, arterial and venous drainage. However, malrotation serves as a predisposing factor for volvulus, so semi-elective surgery will be necessary

if there is malrotation. It is also possible to cause a fatal outcome with volvulus, so it is considered an acute surgical emergency.

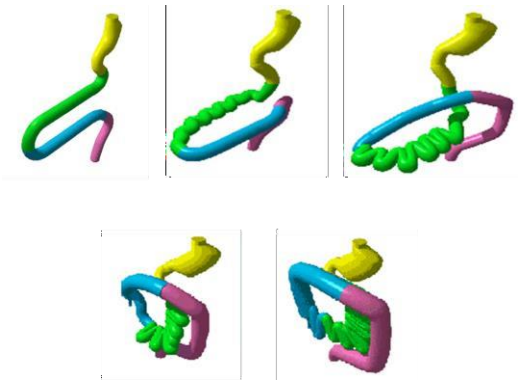


Figure: 1 Normal Intestine Development

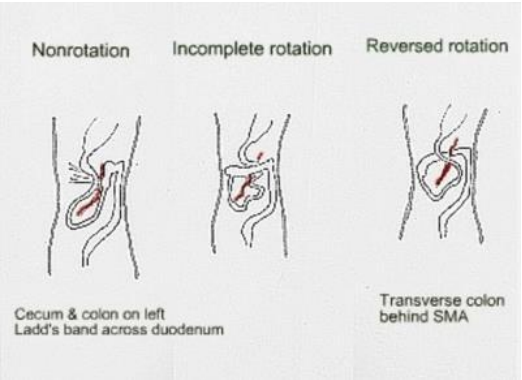


Figure: 2 Abnormal Intestine Development

PRESENTATION

A case of this disease occurs every 500 births, according to estimates. In the first month after birth, 75% of babies will develop a disease, while 15% by the second year. A newborn with sudden onset bilious vomiting should be considered to have a mal-rotation with vertical midgut volvulus until otherwise proven. Aside from mild abdominal pain and early satiety, these conditions also result in delayed development.

Most common Presentation are

- Acute midgut volvulus
- Chronic midgut volvulus
- Acute intestinal obstruction secondary to congenital bands
- Chronic intestinal obstruction secondary to congenital bands
- Reverse rotation with colonic obstruction
- Internal hernia
- Volvulus of caecum
- Asymptomatic/Minimally symptomatic

There is widespread recognition that symptomatic malrotation requires surgical intervention. Ladd procedures have become more popular in recent years. As far as the Ladd procedure is concerned, nothing has changed. Malrotation has been diagnosed or corrected with laparoscopy since the turn of the century[10]. Using laparoscopy, the Treitz ligament can be identified and clamped to the cecum. Delayed presentation of these patients results in more incidence of volvulus and intestinal gangrene as the diagnosis is often missed at admission. Few patients have persistent symptoms even after surgery in the postoperative period[11]. The reason for their persistent complaints is still not known.

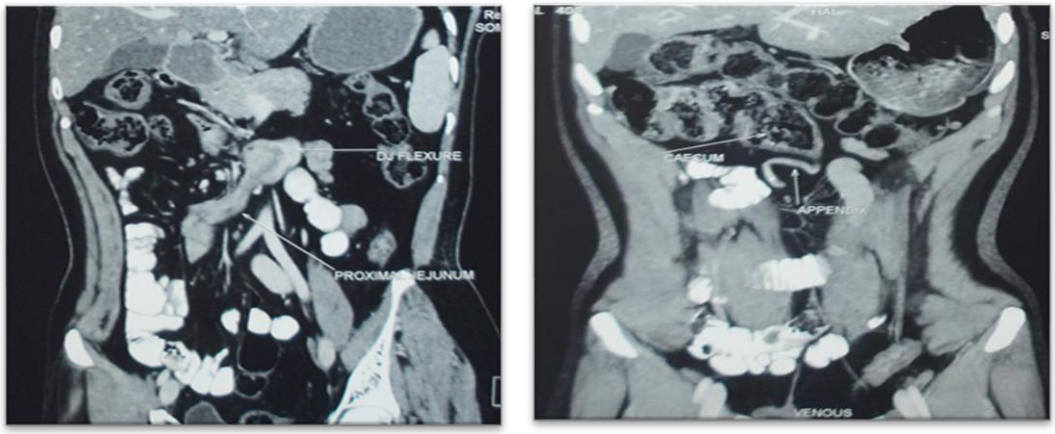
However, postoperative bowel adhesions and long-standing duodenal dilation with stenosis may explain their symptoms.

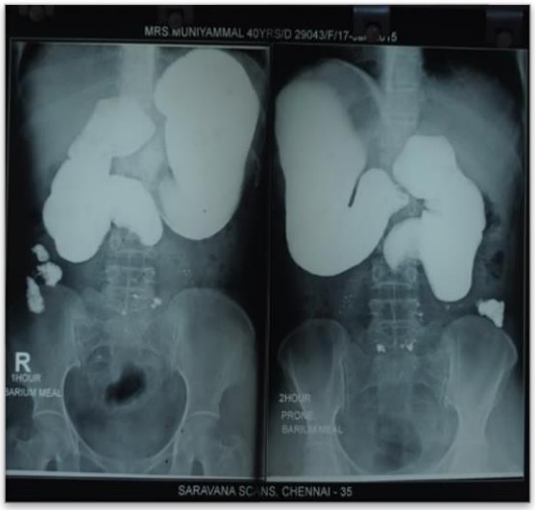
CASE SCENARIOS

CASE 1:

A 40-year-old female, reported Persistent vomiting for 15 days duration of about 4 to 5 post-prandial episodes per day, which is bilious in nature. The patient presents poorly localized cramp-like abdominal pain which gets relieved after vomiting. A previous History of similar illness since childhood was present which was relieved by the Over-the-counter (OTC) anti-emetics. And no previous history of surgery. On Examination Patient was well built and well-nourished who showed stable vital signs. Palpation and auscultation revealed no palpable masses and normal bowel sounds. No VGP / VIP. CECT reveals Intestinal Malrotation leading to secondary volvus. Other Biochemical and OGDScopy are normal. Ladd's Procedure was performed and Post Operatively symptoms were relieved.

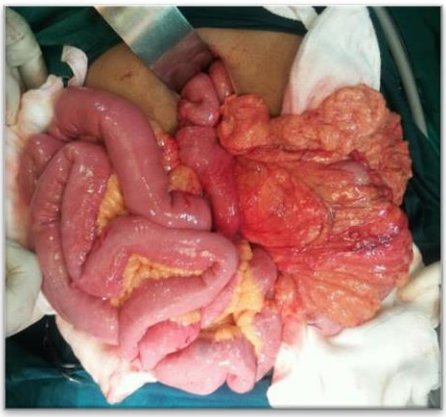
CECT ABDOMEN





BARIUM UGI STUDY

CLINICAL IMAGE

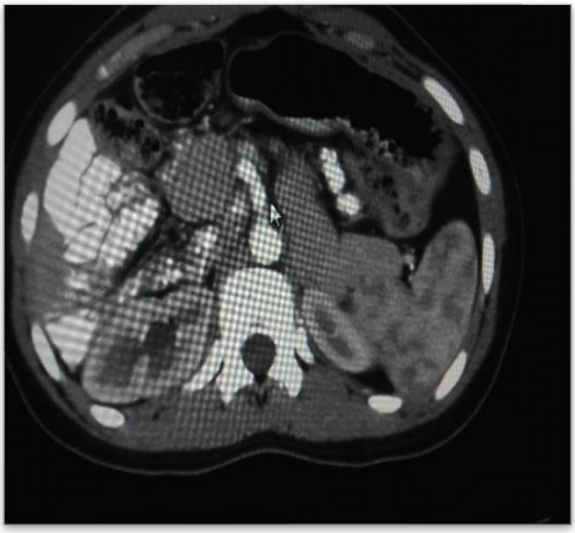


CASE 2

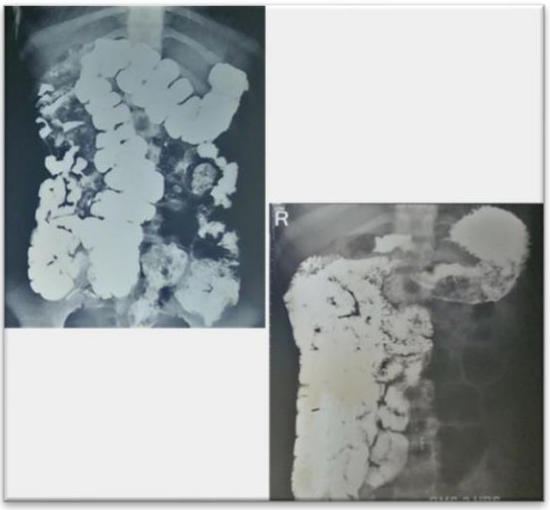
A 15-year-old female reported 2 episodes of haematemesis in a short duration in the past 2 months. History of nausea and dyspepsia are present. There is also a history of early satiety favouring Intestinal Obstruction. On Examination Patient was thin built and moderately nourished who showed stable vital signs. Palpation and auscultation revealed no palpable masses and normal bowel sounds. P/R – NAD. CECT reveals Intestinal Malrotation leading to secondary volvus. Other Biochemical and OGDScopy are normal. Ladd's Procedure was performed and Post Operatively

Appetite got improved, increased oral intake was noticed and Weight gain was achieved. Symptoms were relieved.

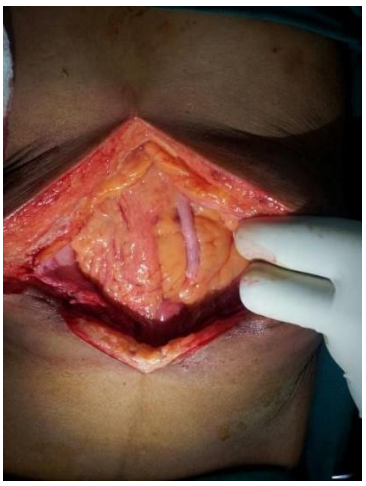
CECT ABDOMEN

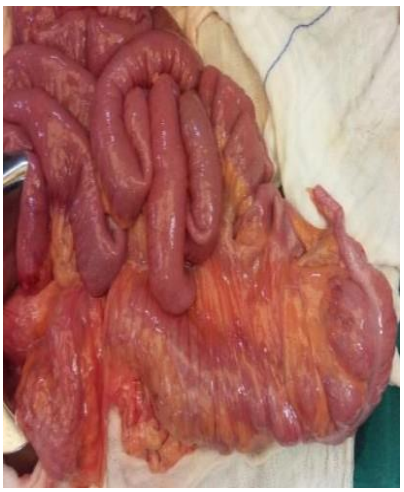


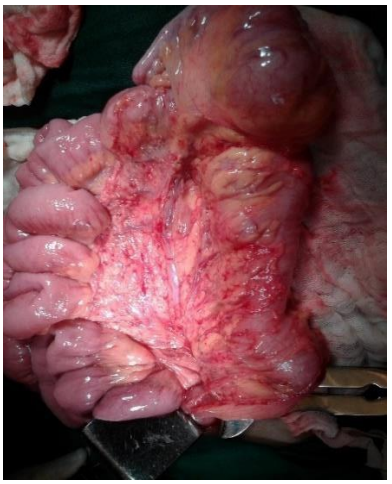
BARIUM UGI STUDY



CLINICAL IMAGE



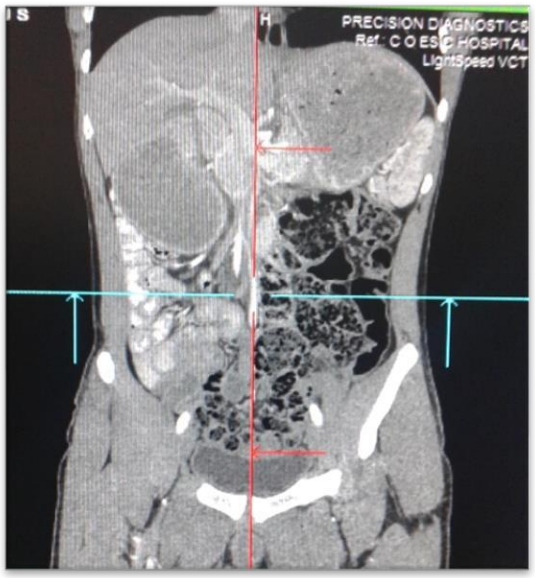


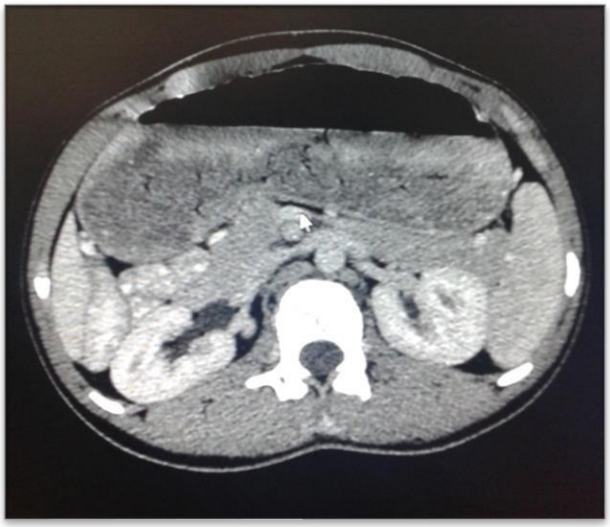


CASE 3:

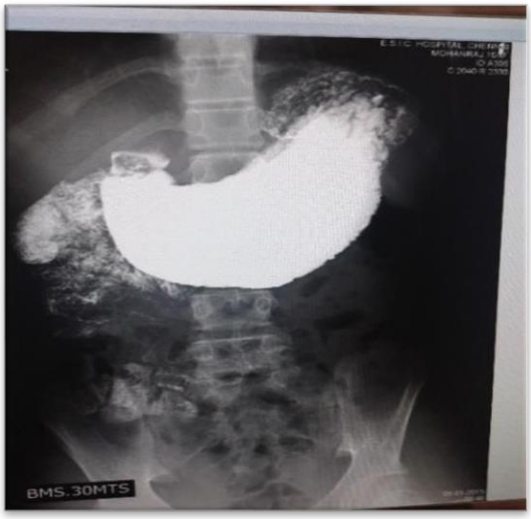
A 16-year-old male reported Persistent vomiting for 2 months duration of about 1 to 2 post-prandial episodes per day, which is bilious in nature. The patient presents with history of long standing dyspepsia. No previous history of surgery. On Examination Patient was well built and well-nourished who showed stable vital signs. Palpation and auscultation revealed no palpable masses and normal bowel sounds. CECT reveals Intestinal Malrotation leading to secondary volvus. Other Biochemical and OGDScopy are normal. Ladd's Procedure was performed and Post Operatively symptoms were relieved.

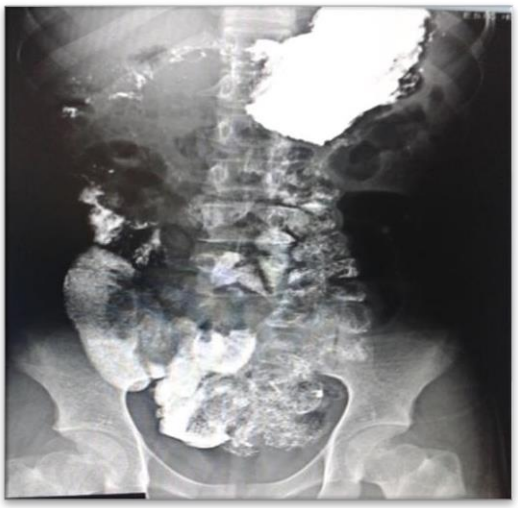
CECT ABDOMEN





BARIUM UGI STUDY





DISCUSSION

There are several reasons why an embryo's midgut can be malrotated. Endodermis develops as early as the fourth week of pregnancy. Three vascular pedicles separate the gut approximately five weeks into pregnancy. An intestinal system consists of three stages that are developed by rotation of the midgut.

Stage 1, The midgut emerges from the fetal abdomen during weeks 5 to 10 and rotates 90 degrees counterclockwise.

Stage 2, It is also common for the abdomen to rotate counterclockwise during week 11 as well. Transverse colons are found above ascending colons, while descending colons are found below. By rotating the colon, the position of the colon will be shifted from right to left to right.

Stage 3 A pregnancy results in fusing of the mesentery with the posterior abdominal wall and descent of the cecum.

Failures in these rotational processes lead to malformations encompassing the entire midgut, resulting in severe mispositioning where the small bowel is located on the right side and the colon on the left side of the peritoneal cavity. Anomalies associated with each stage include

1. omphaloceles in Stage 1 due to gut failure to return to the abdomen,

2. nonrotation, malrotation, and reversed rotation in Stage 2,
3. unattached duodenum, mobile cecum and unattached small bowel mesentery - Stage 3

An abnormal rotation of the midgut causes the duodenojejunal junction to be displaced right of the midline[12], a condition called midgut malrotation1. Adults are rarely affected by midgut volvulus, but it usually occurs acutely within the first month of birth[13]. An adult with malrotation is prone to bowel obstruction from midgut volvulus. Neonatal (usually) and aging patients are more likely to present with acute volvulus of the midgut or ileocecum. There are usually chronic symptoms such as abdominal pain, bloating, vomiting, constipation, and diarrhea associated with chronic symptoms.

Only 0.0001% to 0.19% of adults have midgut malrotation. There is a high rate of asymptomatic diagnosis. Incidental diagnoses can occur after imaging, investigations, or operations for unrelated conditions. Adults who suffer from the disease rarely exhibit symptoms. When abdominal pain recurs, indigestion is inflamed, and intermittent abdominal pain does not disappear, it is hard to diagnose.

CONCLUSION

Delay intestinal malrotation in adults presents significant diagnostic and therapeutic challenges, although it is a rare condition. This congenital anomaly presents in a variety of ways and has many complications, such as volvulus and obstruction of the digestive tract. For positive outcomes, it is imperative to recognize the condition early and to seek surgical intervention, primarily through the Ladd procedure. According to the literature review, adults with unexplained gastrointestinal symptoms should consider intestinal malrotation as a differential diagnosis. In order to improve the management of this condition and the prognosis, continuous advances in diagnostic imaging and surgical techniques are essential.

REFERENCES

1. Martin V, Shaw-Smith C. Review of genetic factors in intestinal malrotation. Pediatr Surg Int 2010;26:769-81.
2. Kapfer SA, Rappold JF. Intestinal malrotation-not just the pediatric surgeon problem. J Am Coll Surg 2004;199:628- 35.
3. Berrocal T, Lamas M, Gutiérrez J, et al. Congenital anomalies of the small intestine, colon, and rectum. Radiographics 1999;19:1219-36
4. Strouse PJ. Disorders of intestinal rotation and fixation, malrotation. Pediatr Radiol 2004;34:837-51
5. Malek MM, Burd RS. The optimal management of malrotation diagnosed after infancy: a decision analysis. Am J Surg 2006;191:45-51.
6. Husberg B, Salehi K, Peters T, et al. Congenital intestinal malrotation in adolescent and adult patients: a 12-year clinical and radiological survey, Springerplus 2016;5:245.
7. Ray D, Morimoto M. Malrotation of the intestine in adult and colorectal cancer. Indian J Surg 2015;77:525-31.
8. Himanshu Menghwani et al Delayed Presentation of Malrotation: Case Series and Literature Review : J Indian Assoc Pediatr Surg. 2023 Jul-Aug; 28(4): 271–277. doi: 10.4103/jiaps.jiaps_2_23: 10.4103/jiaps.jiaps_2_23
9. Peter J. Strouse Disorders of intestinal rotation and fixation (‘‘malrotation’’) Pediatr Radiol (2004) 34: 837–851 DOI 10.1007/s00247-004-1279-4
10. Vicki Martin • Charles Shaw-Smith Review of genetic factors in intestinal malrotation Pediatr Surg Int (2010) 26:769–781 DOI 10.1007/s00383-010-2622-5
11. Susan Nham et al Paediatric intestinal pseudo-obstruction: a scoping review -European Journal of Pediatrics (2022) 181:2619–2632 https://doi.org/10.1007/s00431-021-04365-9
12. Rodríguez et al A new technique in the treatment of intestinal malrotation Cir Pediatr. 2023; 36: 191-194

13. Intestinal malrotation in an adult: case report Turkish Journal of Trauma & Emergency Surgery Ulus Travma Acil Cerrahi Derg 2012;18 (3):280-282 doi: 10.5505/tjtes.2012.60973