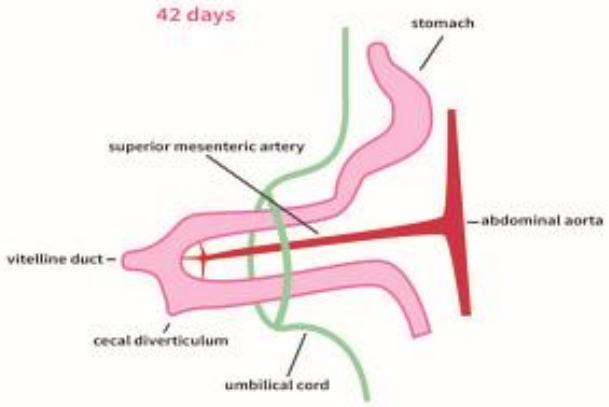
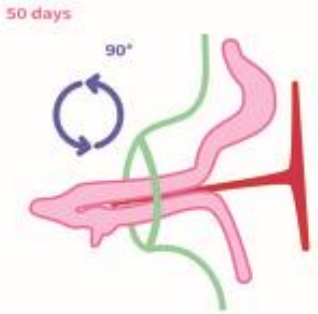
MALROTATION OF GUT EMBRYOLOGY AND ETIOPATHOGENESIS

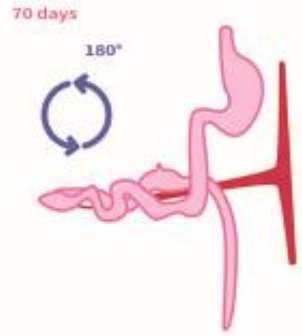
CONTENTS

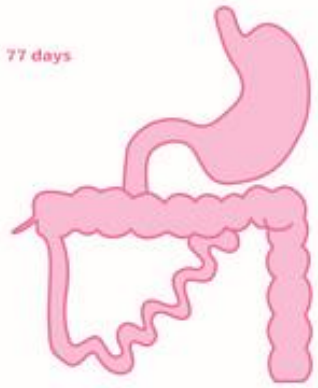
- Embryology
- Rotational anomalies
- Epidemiology
- Signs and symptoms
- Diagnosis
- Management
- Post operative care
- Outcome

Embryology


42 days
stomach
superior mesenteric artery
abdominal aorta
vitelline duct
cecal diverticulum
umbilical cord


50 days
90°


70 days
180°


77 days


later fetal period

Key Points in embryology

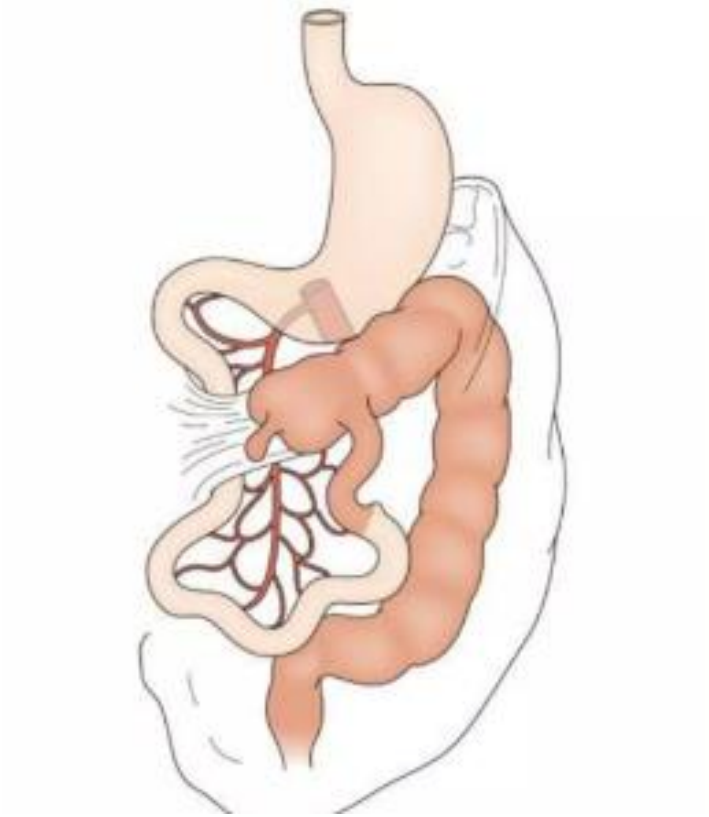
- Intestinal rotation starts at 5 weeks (herniation)
- Rotation takes place around SMA as axis
- 180 degree counter-clockwise rotation of pre arterial and post arterial limb take place intra abdomen at around 10 weeks of gestation.
- Duodeno-jejunal junction is anchored in left upper quadrant, cecum in right lower quadrant.
- Ladds bands attach to cecum irrespective of its position at the end of rotation to the retroperitoneum.
- Ligament of treitz connects and supports duodeno-jejunal junction.

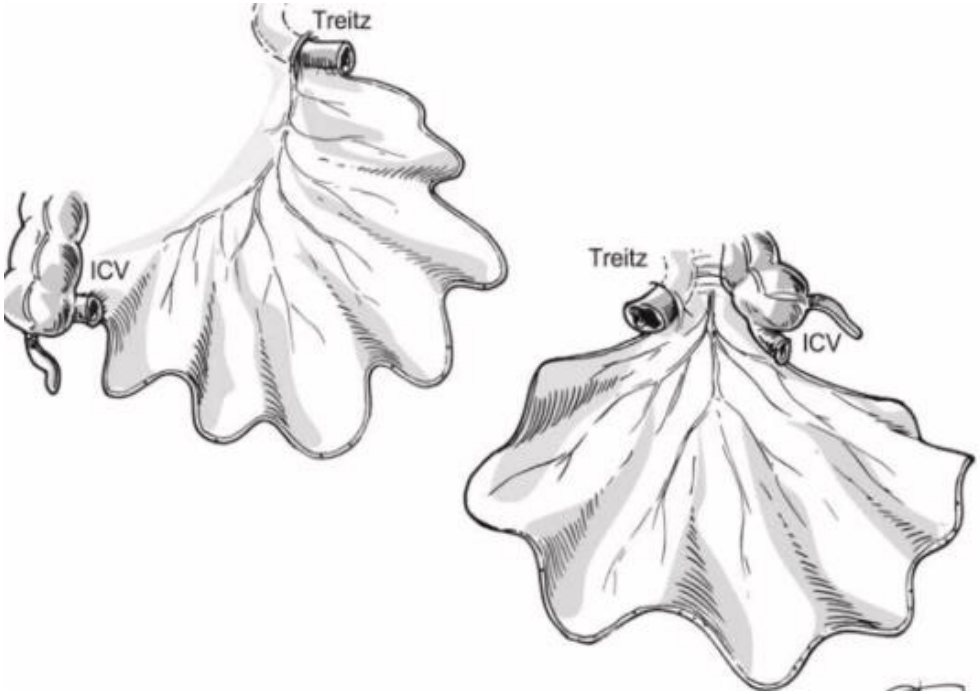
ROTATIONAL ANOMALIES

Rotational anomalies occur when gut fails to rotate or rotation is incomplete.

1. **Incomplete rotation (classical malrotation)**

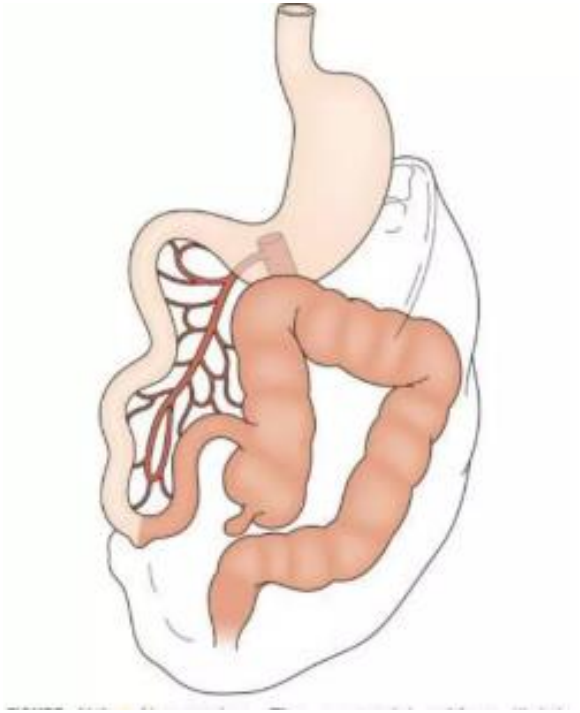
- Normal rotation is arrested at 180 degrees
- DJ junction mid abdomen
- Ileo-cecal valve in right upper quadrant/ mid abdomen
- Narrow pedicle at the base of mesenrety, more likely to rotate.
- Ladd's bands cross the DJ loop and obstructs the duodenum

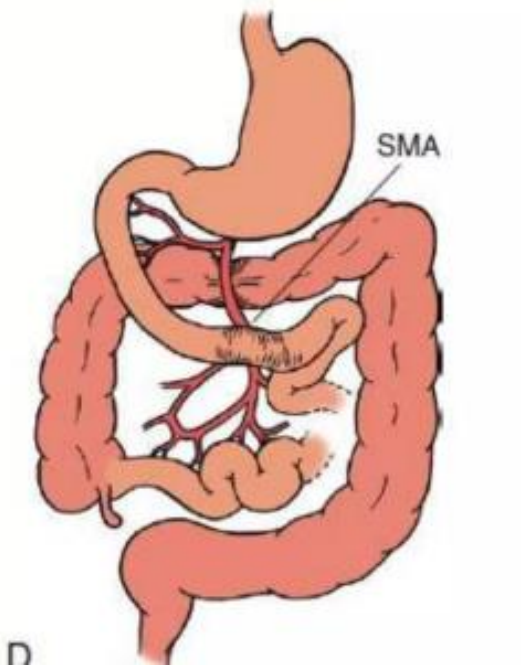



Treitz
ICV
Treitz
ICV

Non rotation and reverse rotation

- DJ junction in right upper quadrant
- Ileo-cecal region is free of attachments and far from duodenum , mesentery is not having a narrow pedicle





EPIDEMIOLOGY

- The incidence of mid-gut malrotation is estimated to be around 1 in 6000 live births.
- Most common type of rotational anomalies is nonrotation, with a rate of 2 per 1000 contrast studies of the upper gastrointestinal tract.

Presentation

1. Bilious / non bilious emesis
2. Midgut volvulus

- Abdominal pain
- abdominal tenderness
- distension of abdomen
- rapid deterioration in general condition
- Mesenteric ischemia (shock/ sepsis)

3. Chronic duodenal obstruction in later life.

- Failure to thrive.
- Recurrent mild abdominal discomfort
- Gastroesophageal reflux (GERD)
- Recurrent partial volvulous with lymphatics and venous obstruction leading to malabsorption and nutritional deficiency.
- Incidental detection in children and adults during routine radiographic evaluation of the gut.

- Presentation by one month of age: 30 percent
- Presentation before one year of age: 58 percent
- Presentation before five years of age: 75 percent

Analysis of over 2700 cases of intestinal rotation in children up to 17 years of age obtained from a national hospital discharge database : Aboagye J, Goldstein SD, Salazar JH, Papandria D, Okoye MT, Al-Omar K, Stewart D, Lukish J, Abdullah F. Age at presentation of common pediatric surgical conditions: Reexamining dogma. J Pediatr Surg. 2014 Jun;49(6):995-9. doi: 10.1016/j.jpedsurg.2014.01.039. Epub 2014 Feb 11. PMID: 24888850.

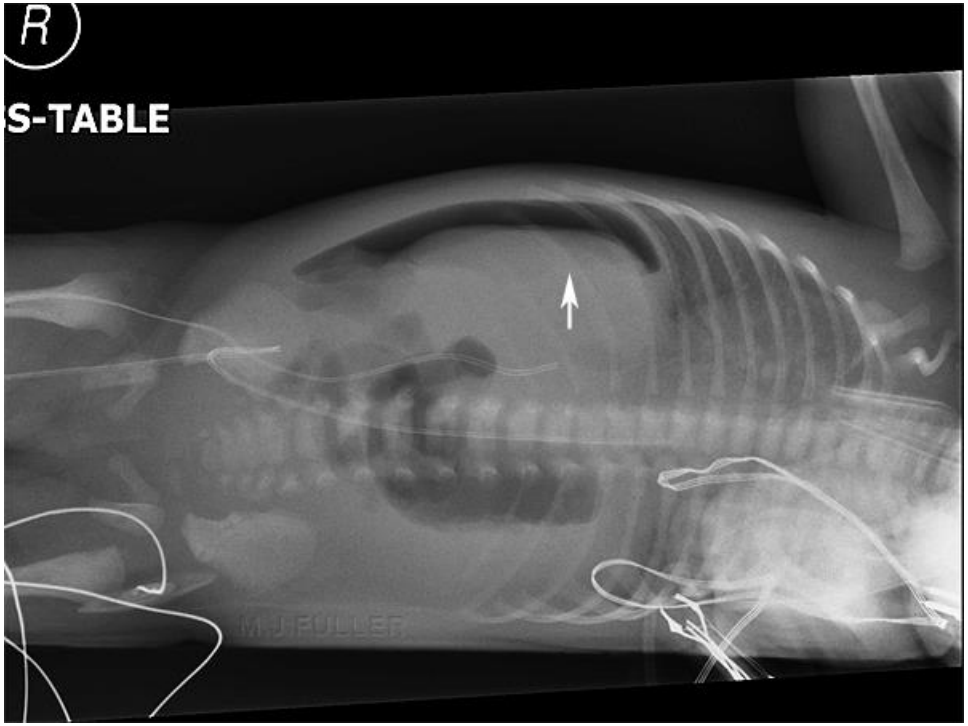
DIAGNOSIS

- **IMAGING STUDIES**

1. **Abdominal Radiography**

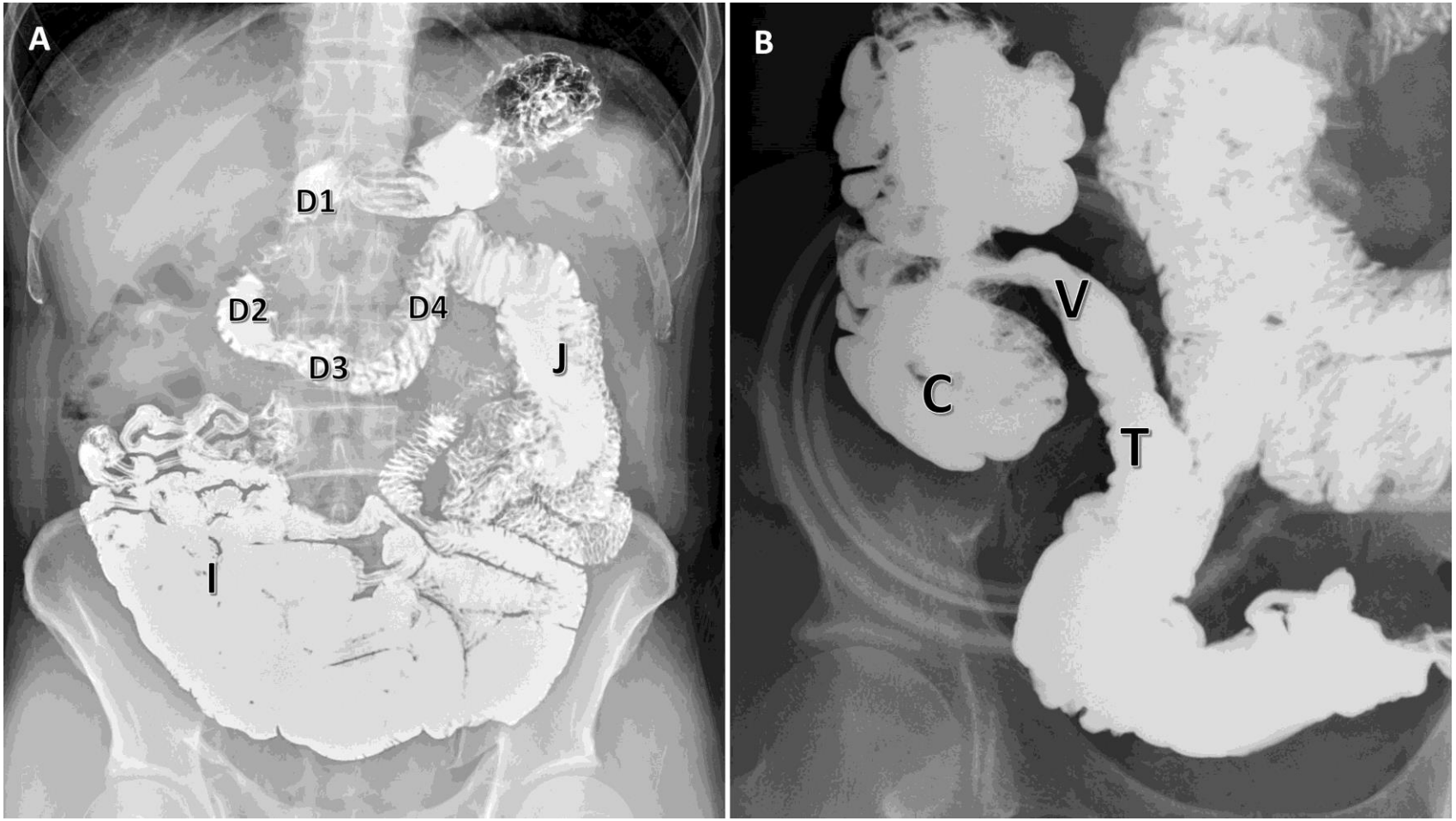
Double-bubble sign with no or little gas in the rest of the GI tract / multiple air fluid levels .

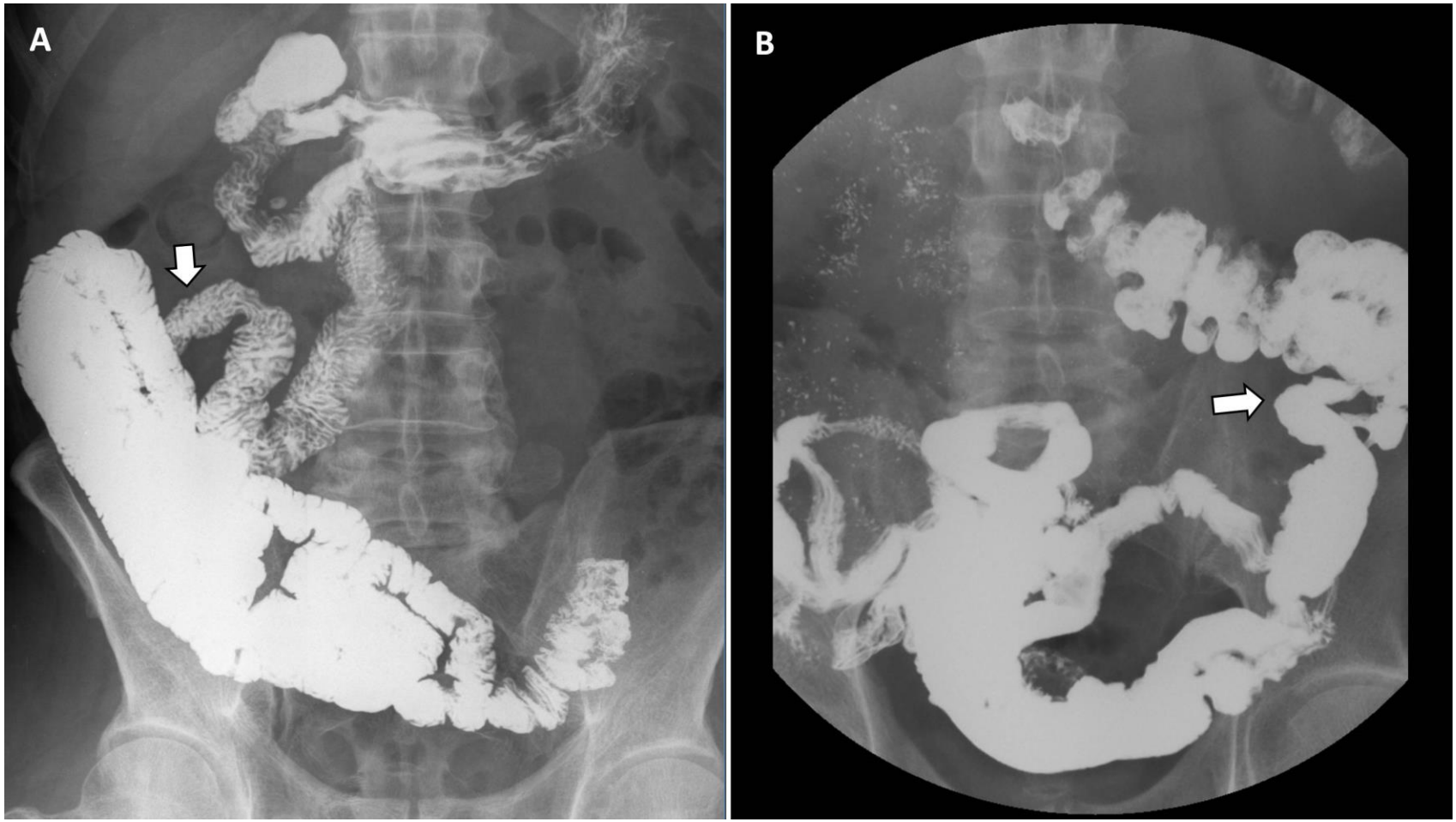
Left lateral decubitus x-ray can be obtained to visualize better if pneumoperitoneum is suspected.


R
S-TABLE

2. **Upper GI Contrast study**

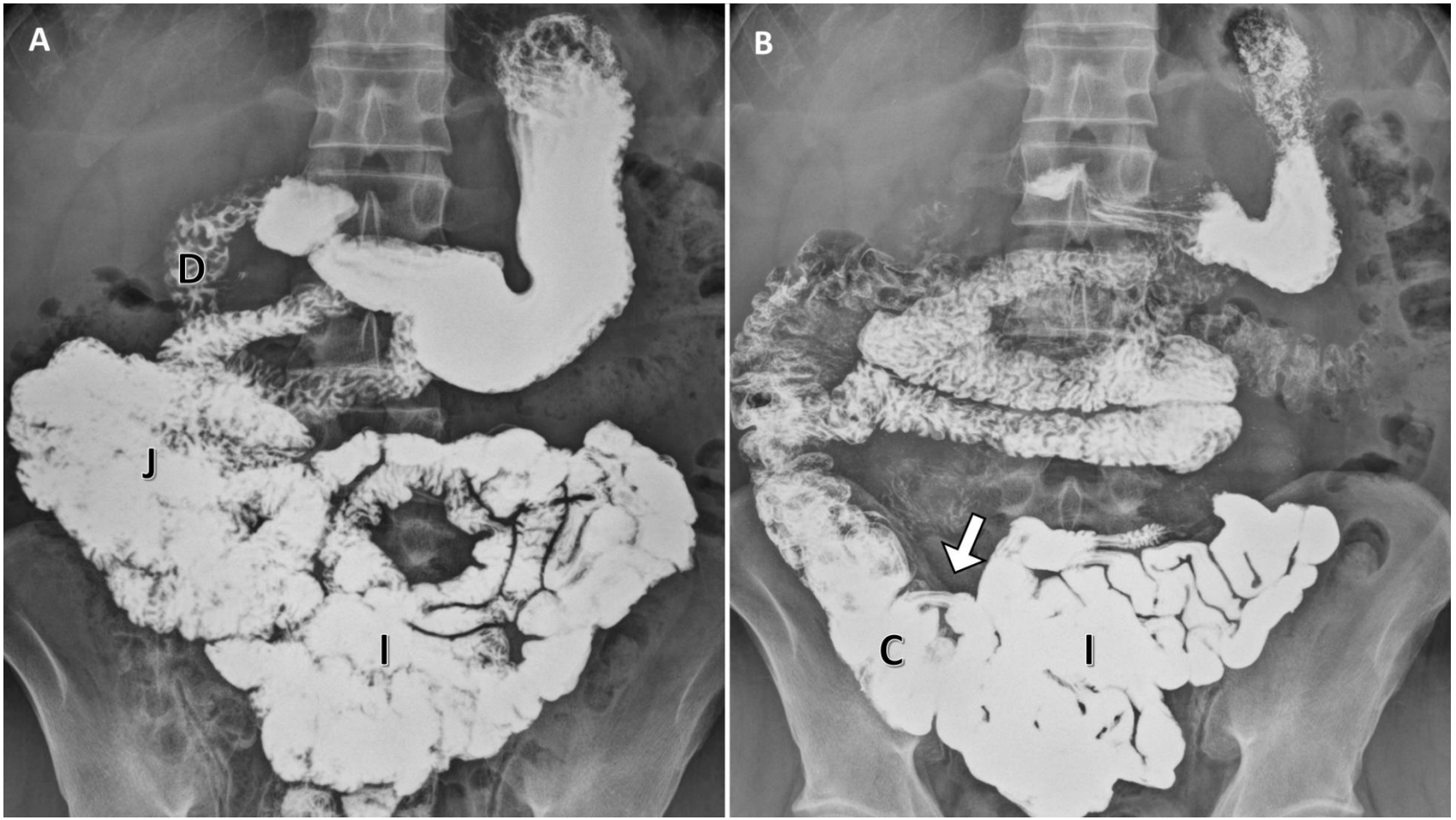
- Duodeno-jejunal junction at the left side of the spine shadow at the level of the pylorus.
- Findings of malrotation include ligament of Trietz on the right side of the abdomen instead of the left


A
D1
D2
D3
D4
J
I
B
V
C
T



Non rotation of the bowel

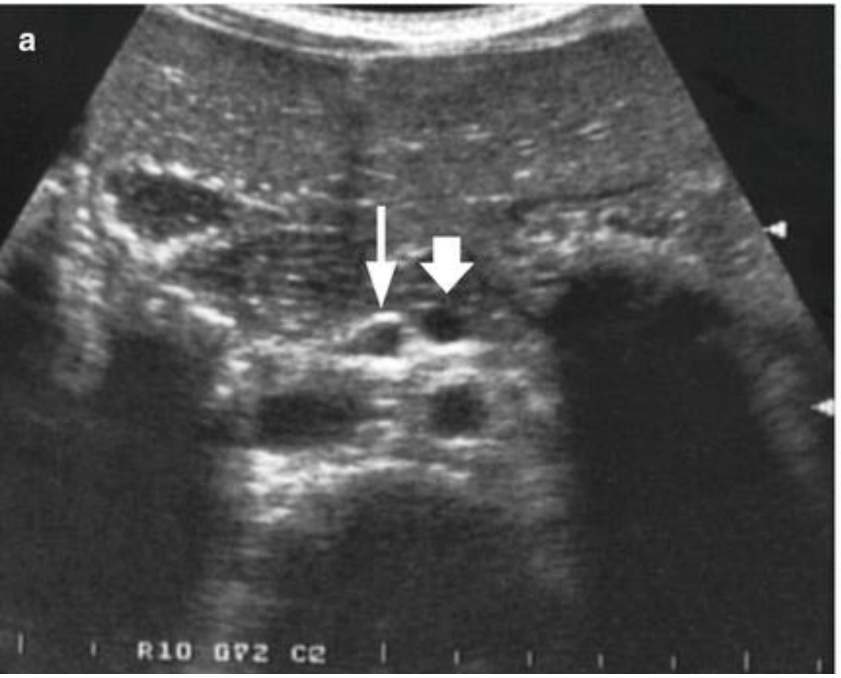
Incomplete rotation

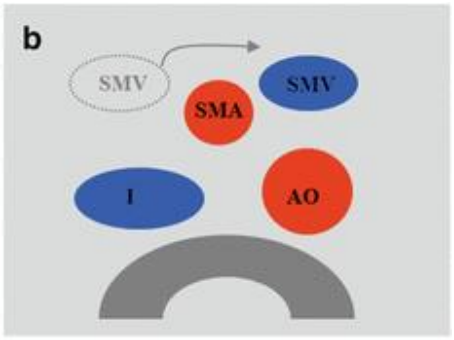


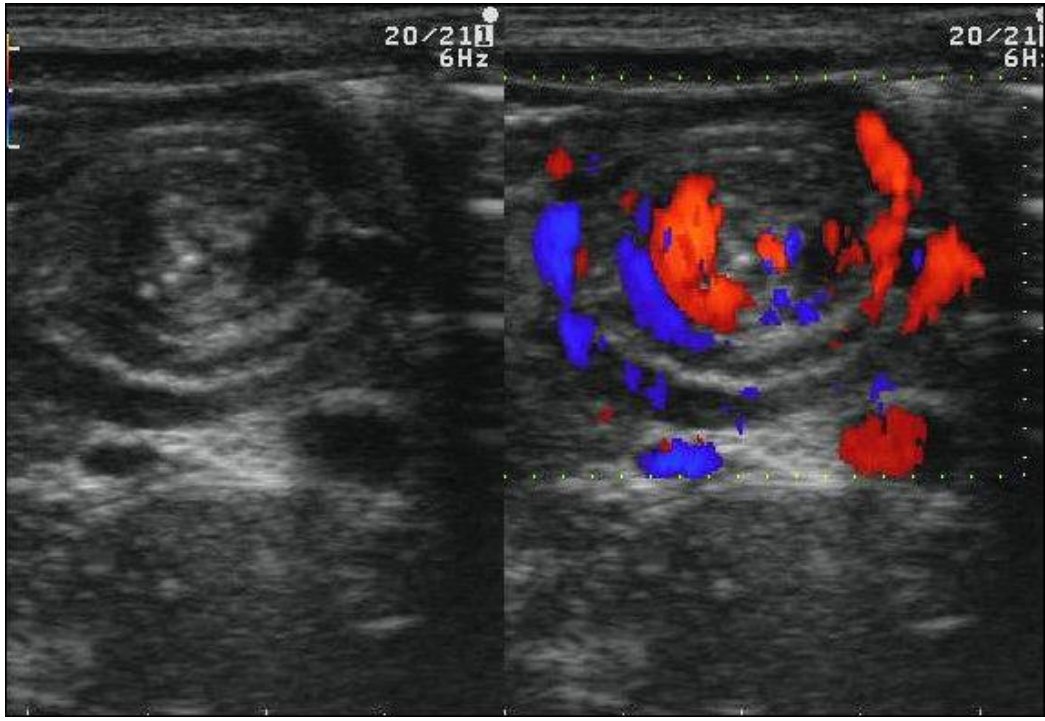
3. **Ultrasonography**

- Reversal of normal anatomical relation between SMA and SMV.
- Whirlpool sign – midgut volvulus

Sensitivity of 94% and a specificity of 100% for the use of ultrasonography to diagnose intestinal malrotation either with or without midgut volvulus : Nguyen HN, Kulkarni M, Jose J, Sisson A, Brandt ML, Sammer MBK, Pammi M. Ultrasound for the diagnosis of malrotation and volvulus in children and adolescents: a systematic review and meta-analysis. Arch Dis Child. 2021 Dec;106(12):1171-1178.


a
R10 G72 C2


b
SMV
SMA
SMV
I
AO


20/21
6Hz
20/21
6H

4. Computed Tomography

- Absence of a retro-mesenteric/retroperitoneal segment of the duodenum (the third part of the duodenum).
- abnormal SMA /SMV relationship (SMA is smaller and more circular).
- Large intestine predominantly on the left side and small bowel on the right side of the spine shadow
- 'whirlpool' sign present indicating twisting of blood vessels around the mesenteric pedicle

5. **Barium enema**

- Misleading and should not be used
- Final fixation of colon upto term gestation
- Cecum is poorly fixed, can mimic malrotation

MANAGEMENT

- **Volvulus not resulted in significant ischemia** -> Ladd procedure

- **Midgut has significant ischemia but not frankly necrotic.**

1. Abdomen closed loosely
2. 24hours apart, re explored

- **Frankly necrotic midgut and non recoverable** -> Affected gut is removed

Volvulus not resulted in significant ischemia

Ladd procedure:

- Devolvulizing the midgut
- Widening the mysentery, remove all adhesions to separate duodenojejunal junction as far as possible from ileoceacal region
- Small bowel on the right abdomen, large bowel on the left of the abdomen
- Appendectomy.

Post operative management

- **Parenteral nutrition** (PN) is the cornerstone of management until **intestinal** adaptation returns and Enteral Nutrition can be started.
- Parenteral nutrition may be necessary for an extended period of time if there is malabsorption or if a long segment of ischaemic intestine has been removed.
- Protein 3.5-4mg/kg, Lipid 3.5-4g/kg, GIR 10-14mg/kg/min. Electrolytes as per lab reports.
- Target calories: 100-120kcal/kg/day

COMPLICATIONS

- Early postoperative small bowel obstruction from adhesive bands.
- Wound infections
- Short bowel syndrome and vitamin deficiencies due to bowel loss.

OUTCOMES

- **Resolution of symptoms:** 89 % of patients who undergo operative intervention.
- **Mortality :** overall mortality rate after surgery for malrotation ranges from 3 to 10%
- **Recurrent volvulus :** Risk of recurrent volvulus after laparoscopic versus open Ladd procedure was 3.5 and 1.4 percent, respectively

Nehra D, Goldstein AM. Intestinal malrotation: varied clinical presentation from infancy through adulthood. Surgery. 2011 Mar;149(3):386-93. doi: 10.1016/j.surg.2010.07.004. Epub 2010 Aug 17. PMID: 20719352.

THANK YOU