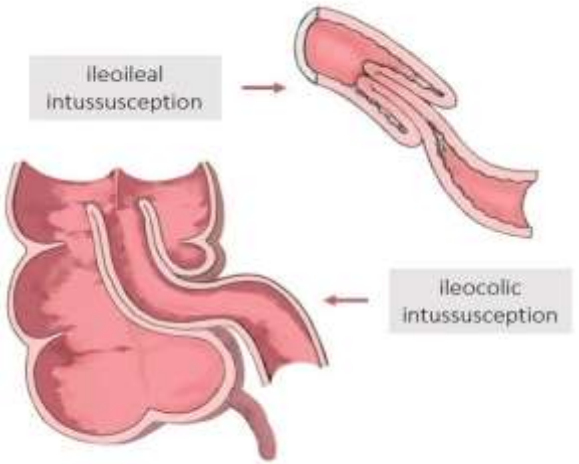
PEDIATRIC SURGERY

INTUSSUSCEPTION

RULE OF 6

1) Peak incidence

6 months of age



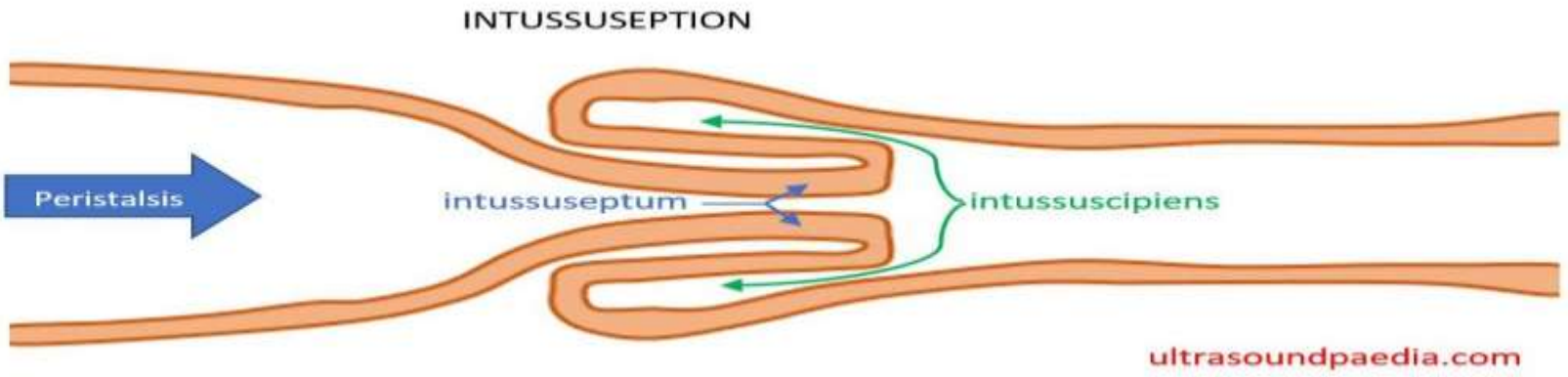
2) Aetiology

6% secondary & 94% idiopathic

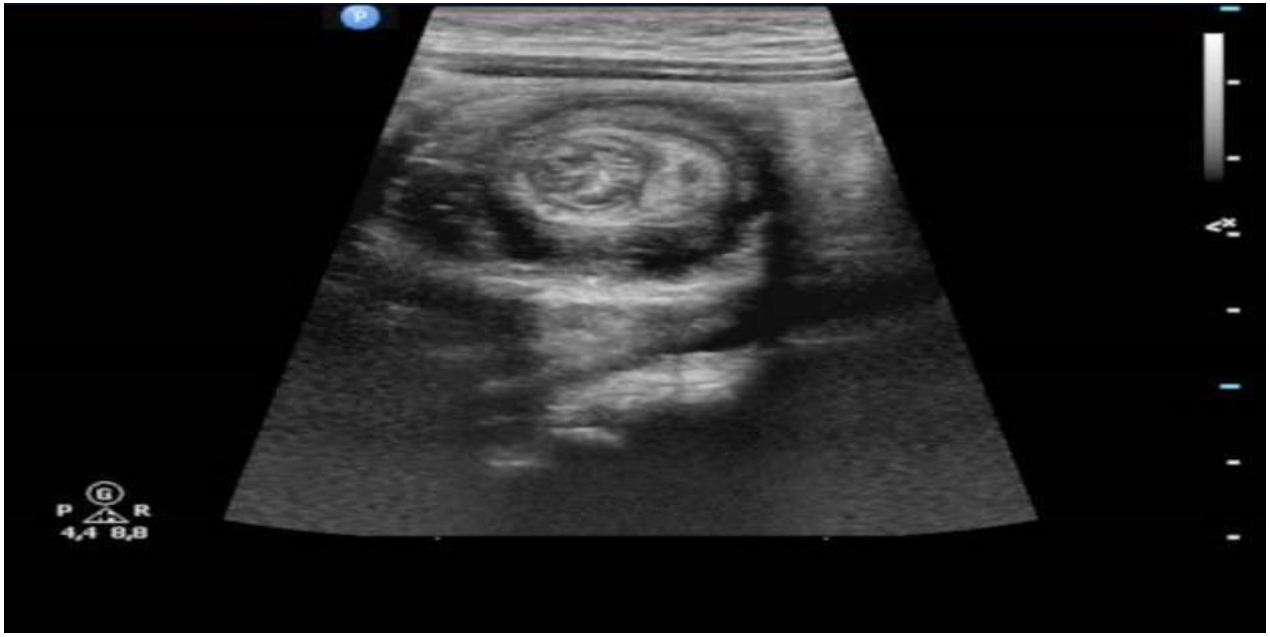
- **3**) **6** symptoms

1. Abdominal pain (incessant cry)
2. Abdominal lump (sausage shaped mass)
3. Abdominal distension
4. Vomiting
5. Bleeding per rectum (red currant jelly stools)
6. Prolapse per rectum (rare)

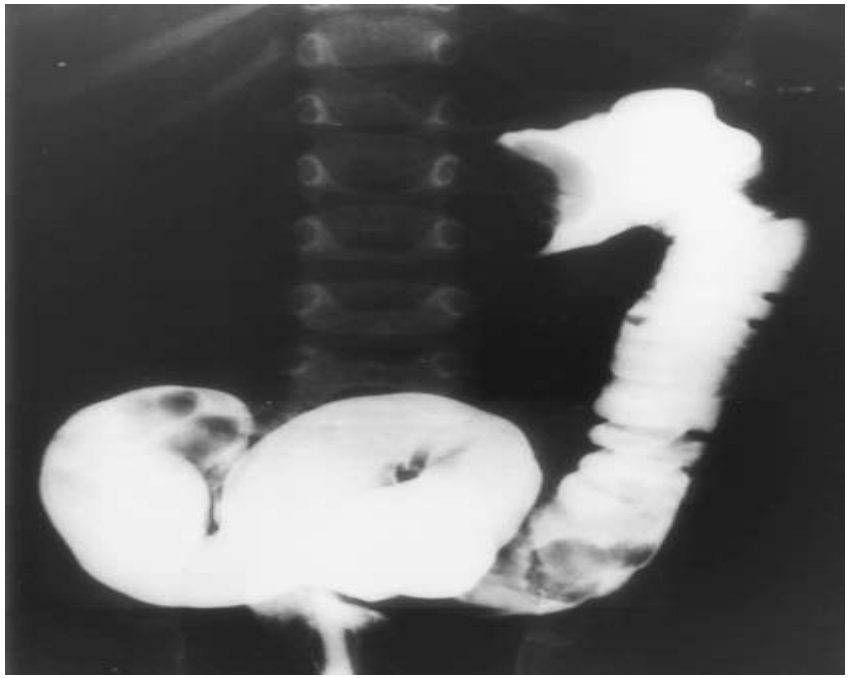
- **4**) **6** signs
- **2** Clinical
  1. Signe de Dance (emptiness of RIF)
  2. Sausage shaped mass (palpable, because **6** layers of intestine are involved circumferentially)



- **2 sonological**
  1. Target sign
  2. Pseudo kidney sign



- **2** radiological
  1. Claw / Pincer sign
  2. Coiled spring sign in Barium enema study.



5 Management

- **6** methods

1. Conservative – parenteral fluids for hydration & withhold oral feedsbecause **6%**of intussusceptions will resolve spontaneously.
2. Hydrostatic reduction with saline enema under Ultrasound guidance
3. Barium enema reduction under fluoroscopic guidance
4. Pneumatic reduction under fluoroscopic guidance
5. Operative manual reduction by squeezing / milking
6. Resection& end to end anastomosis

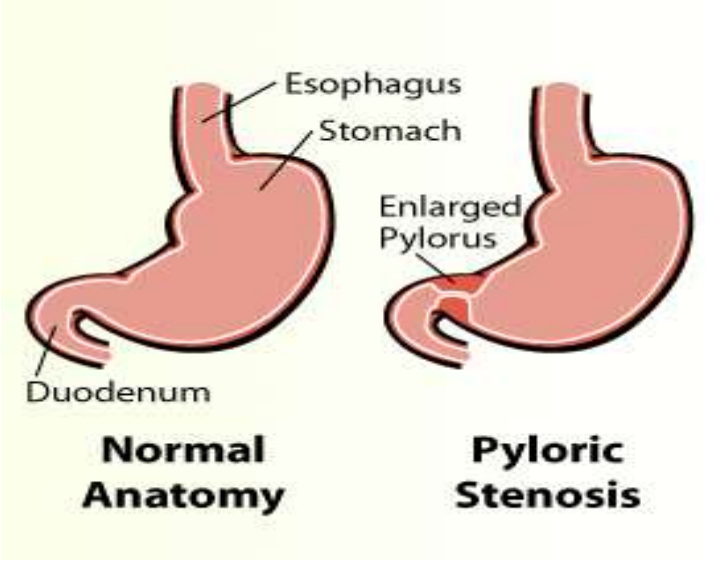
- **6**) Recurrence is **6%**

IHPS

RULE OF 4

Incidence

- Upto 4 per 1000 live births



- Peak at 4 weeks of age (can appear between 4 days to 4 months)

- Male: female is 4:1

- Etiology – 4 in no. -Hereditary& environmental

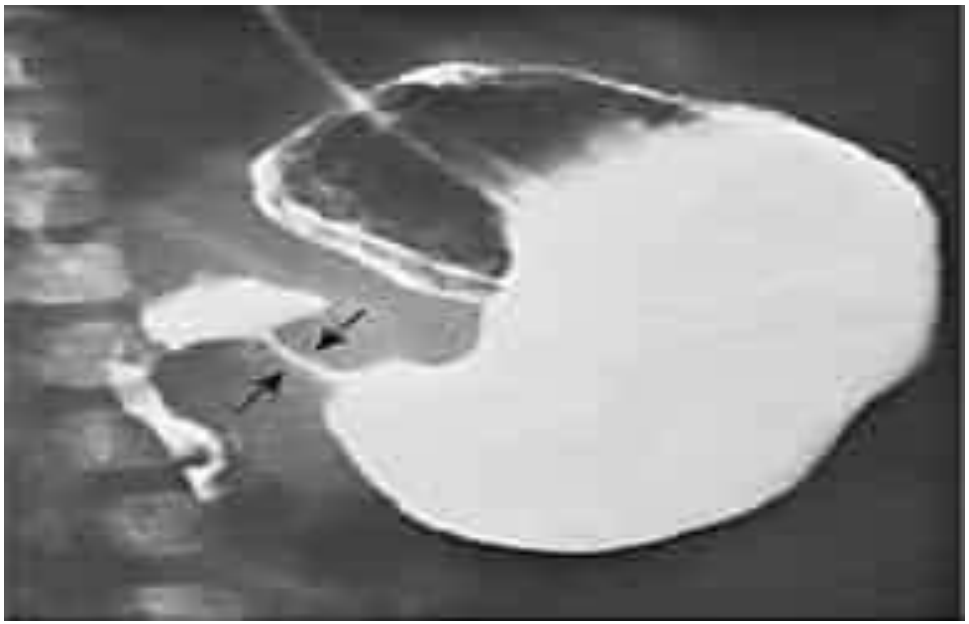
1. Infantile hypergastrinemia& milk curd theory
2. Reduced nitric oxide synthase & thereby reduced nitric oxide locally
3. Immaturity & hypodensity of ganglion cells
4. Miscellaneous – VIP & other neuropeptides

- Clinical Features – 4 in no.
  1. Non bilious, progressive & projectile vomiting
  2. Visible gastric peristalsis from left to right
  3. Palpable pyloric mass(olive shaped) – pathognomonic in **Rt** hypochondrium
  4. Voracious appetite

- Loss due to vomiting – 4 in no.
  1. Na^+
  2. Cl^-
  3. K^+
  4. H^+

- Investigations – 4 in no.

1. Biochemical tests to prove the metabolic variations
2. Plain X-ray Abdomen – grossly distended stomach
3. Ultrasound abdomen –
    1. pyloric mass >**4**mm thickness
    2. Pyloric canal length >**14**mm
4. Barium meal X-ray – 4 findings
    1. Shoulder sign
    2. Beaking
    3. String/double track sign
    4. Mushroom/umbrella sign



- Treatment - 4 in no.
  1. Nasogastric decompression & correction of dehydration
  2. Correction of metabolic abnormalities
  3. Antispasmodics/ parasympatholytics
  4. Definitive treatment – **Ramstedt's Pyloromyotomy**

- 4 Abdominal approaches
  - Midline through the linea alba
  - Paramedian rectus splitting
  - Lateral to right rectusabdominis
  - Circumumbilical/cosmetic/laparoscopic
-

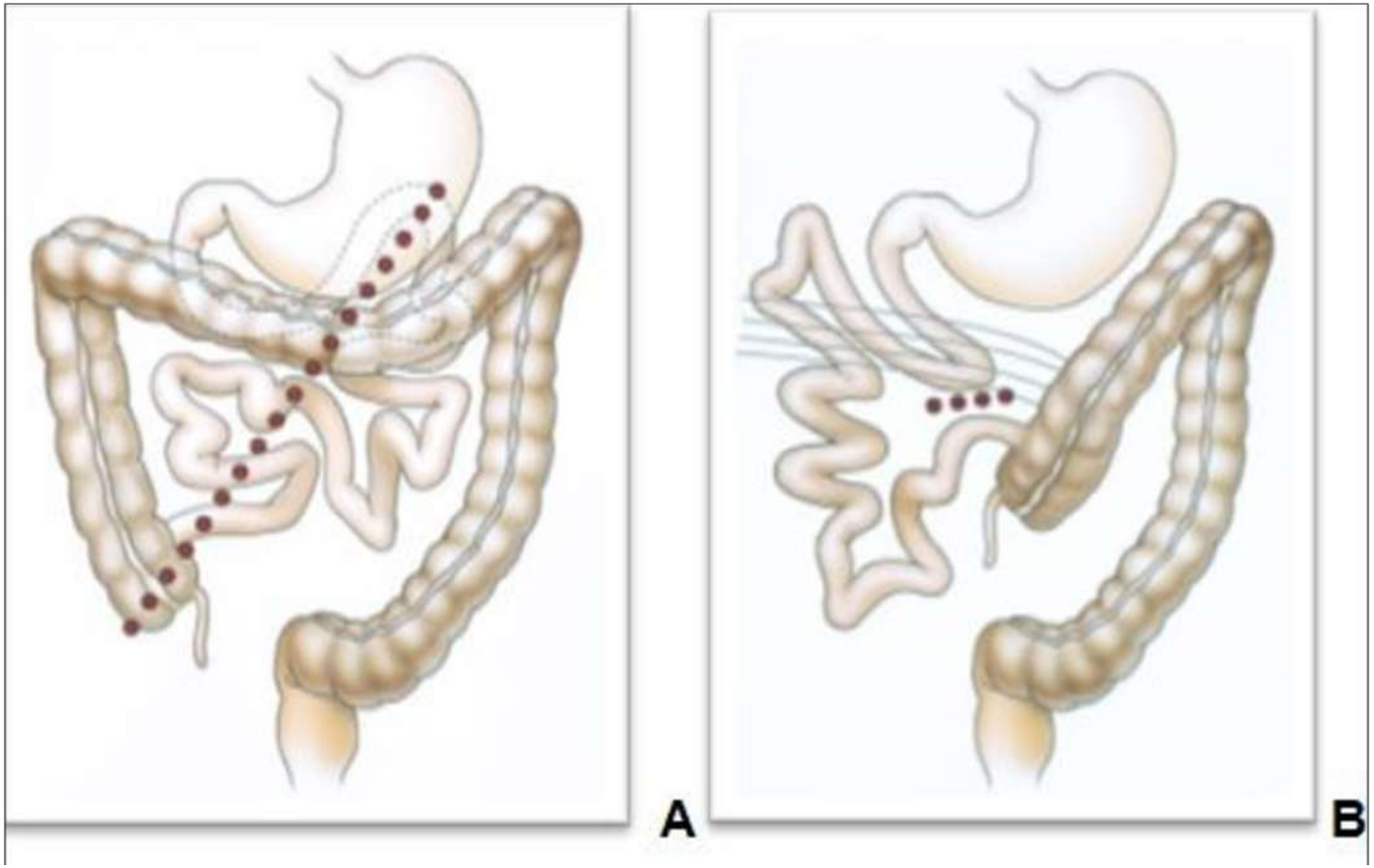
- Recurrence of symptoms in 4% mainly due to incomplete myotomy
- Complications < 4%
- Mortality after treatment is 0.4%

MALROTATION

- Malrotation involves a spectrum of anatomic abnormalities of incomplete rotation and fixation of intestine during fetal development

- Normal rotation of the midgut occurs in fetal life in such a way that duodenum turns anticlockwise through 270^0 across the axis formed by the superior mesenteric artery (SMA) so that DJ flexure goes posterior and reaches the (L) hypochondrium, while the whole midgut comes anterior especially the transverse colon. Thus the caecum comes to the (R) iliac fossa,

- thereby getting the widest root of mesentery stretching from the DJ flexure in (L) hypochondrium to ileocecal junction in (R) iliac fossa.

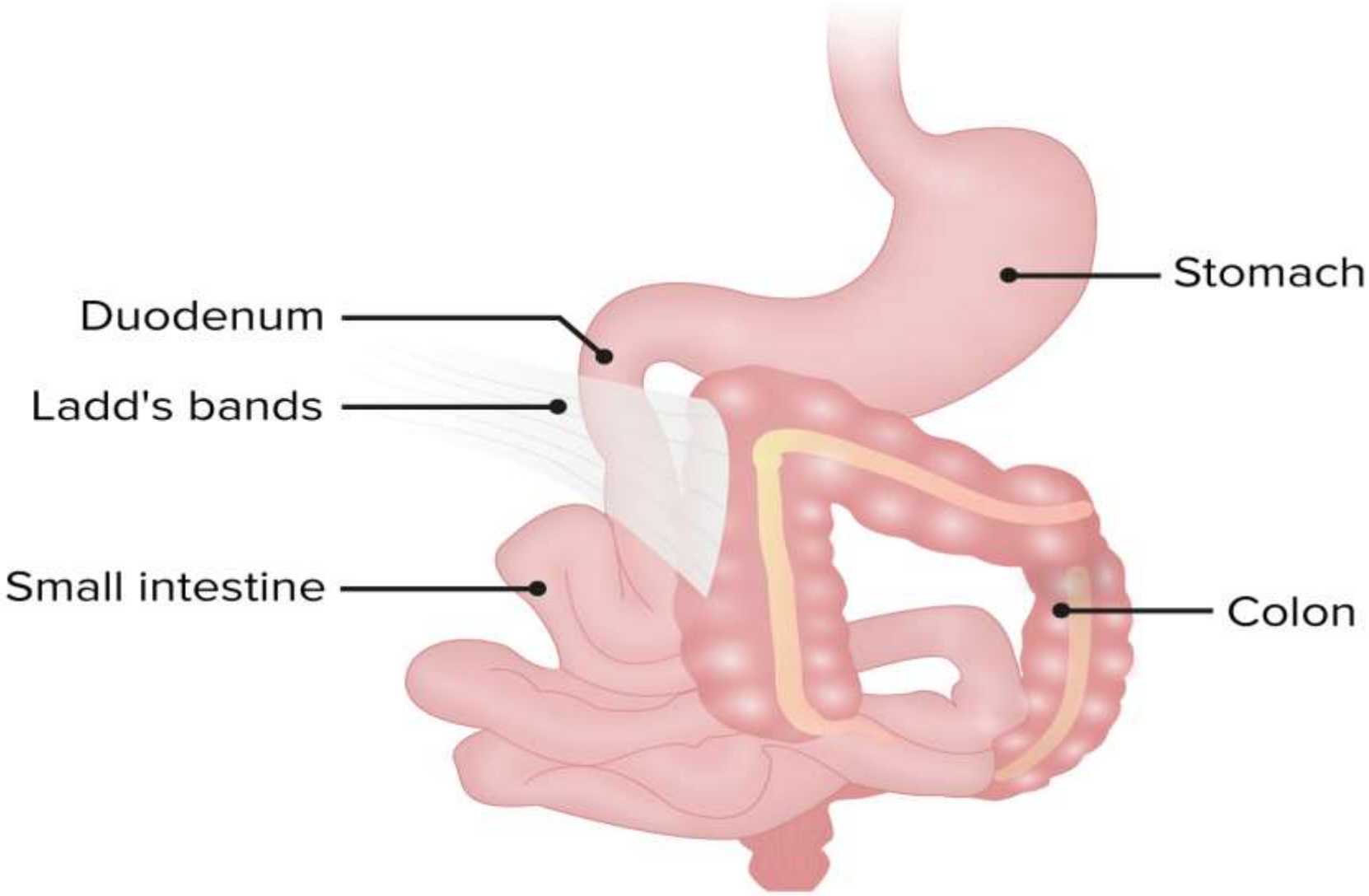

A
B

- **Classified as**
  1. Non rotation
  2. Incomplete rotation
  3. Mixed variant
  4. Reverse rotation
  5. Atypical rotation

In malrotation

- Cecum is to the left of midline in upper abdomen, DJ limb is to the right of midline and the superior mesenteric vascular pedicle will be the narrowest.

- The fixation of mesentery is absent and there is a very high risk of midgut volvulus
- The peritoneal bands (Ladd's bands) extending from the posterior abdominal wall to the cecum cross the duodenum in their course to the abnormally located cecum causing extrinsic duodenal obstruction.


Stomach
Duodenum
Ladd's bands
Small intestine
Colon

- Partial intestinal obstruction will present as intermittent bilious vomiting whereas complete intestinal obstruction will present as frank bilious vomiting.

ASSOCIATED ANOMALIES

- Upto 50% patients with duodenal atresia have malrotation
- Upto 33% patients with jejunoileal atresia have malrotation

Modes of presentation

- Acute midgut volvulus – is a surgical emergency because any delay in attempting surgical detorsion can lead to gangrene of the entire midgut.
- Chronic midgut volvulus – off and on

- Typical malrotation in asymptomatic or minimally symptomatic patients
- Acute duodenal obstruction secondary to Ladds bands
- Chronic duodenal obstruction

- Reverse rotation with colonic obstruction
- Internal hernias (Right and Left mesocolic hernias)
- Volvulus of the caecum

INVESTIGATIONS

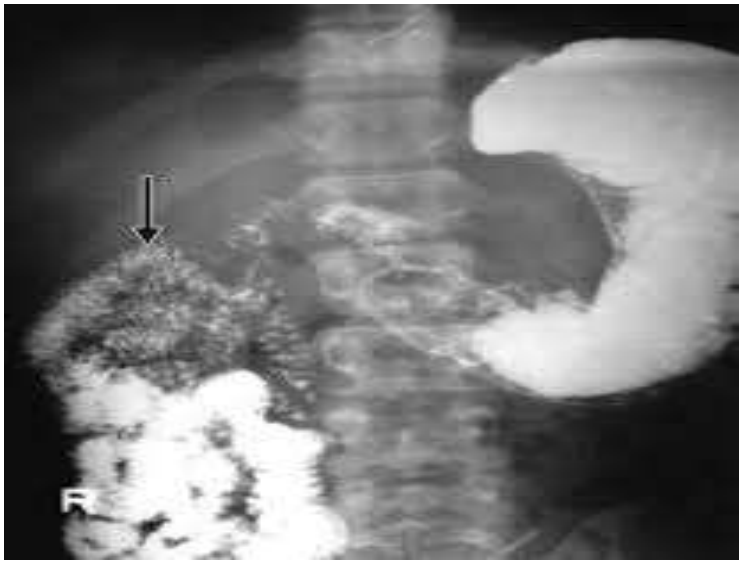
PLAIN XRAY ABDOMEN

1. “double bubble sign” of acute duodenal obstruction
2. “gasless” abdomen in volvulus of midgut

•

UPPER GI CONTRAST STUDIES

- Barium / water soluble contrast instilled via NGT



DOPPLER Ultrasound

- Reversal in relation of SMA to SMV
- Normally SMV is to the right of SMA
- In malrotation, SMV is aberrantly located – anterior or to the left of SMA

Volvulus is highly probable if

- the SMV is shown to be coiling around the SMA
- presence of fixed midline bowel loops and
- duodenal dilation " Whirlpool sign "

OPERATIVE TECHNIQUE

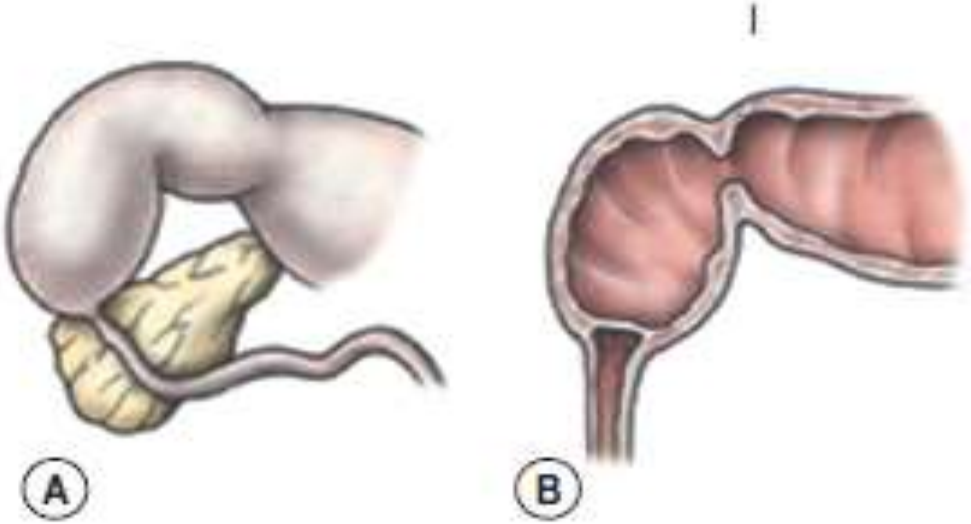
- Original description by **William Ladd – 6 steps**

LADDS PROCEDURE

DUODENAL ATRESIA

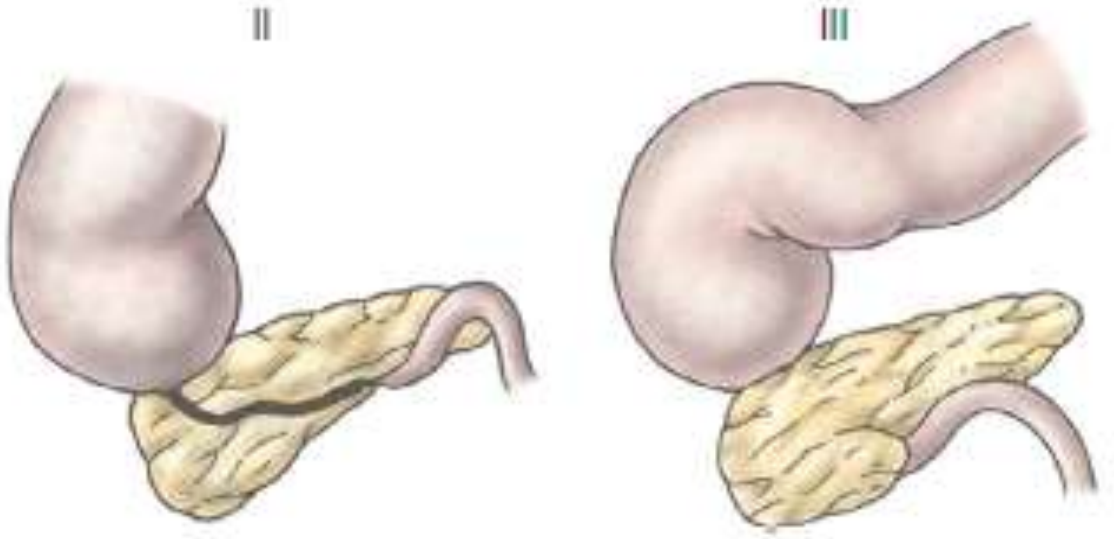
TYPES

- Type I – Continuity maintained, may be normal on the exterior, but obstructed by a intact membrane or diaphragm



- Type II – Continuity not maintained, but connected by a fibrous cord

- Type III – Continuity not maintained, with V shaped defect and absent mesentery



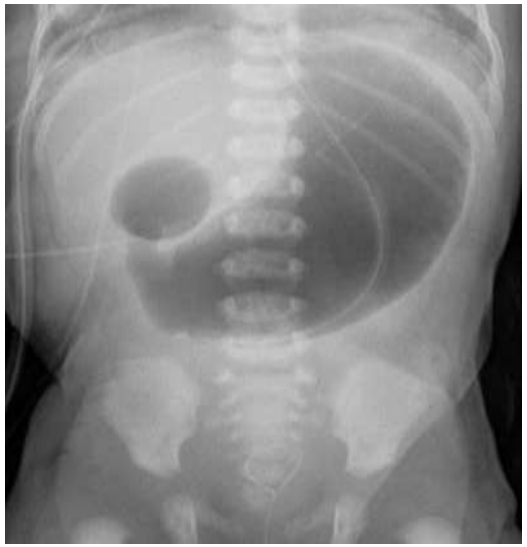
CLINICAL FEATURES

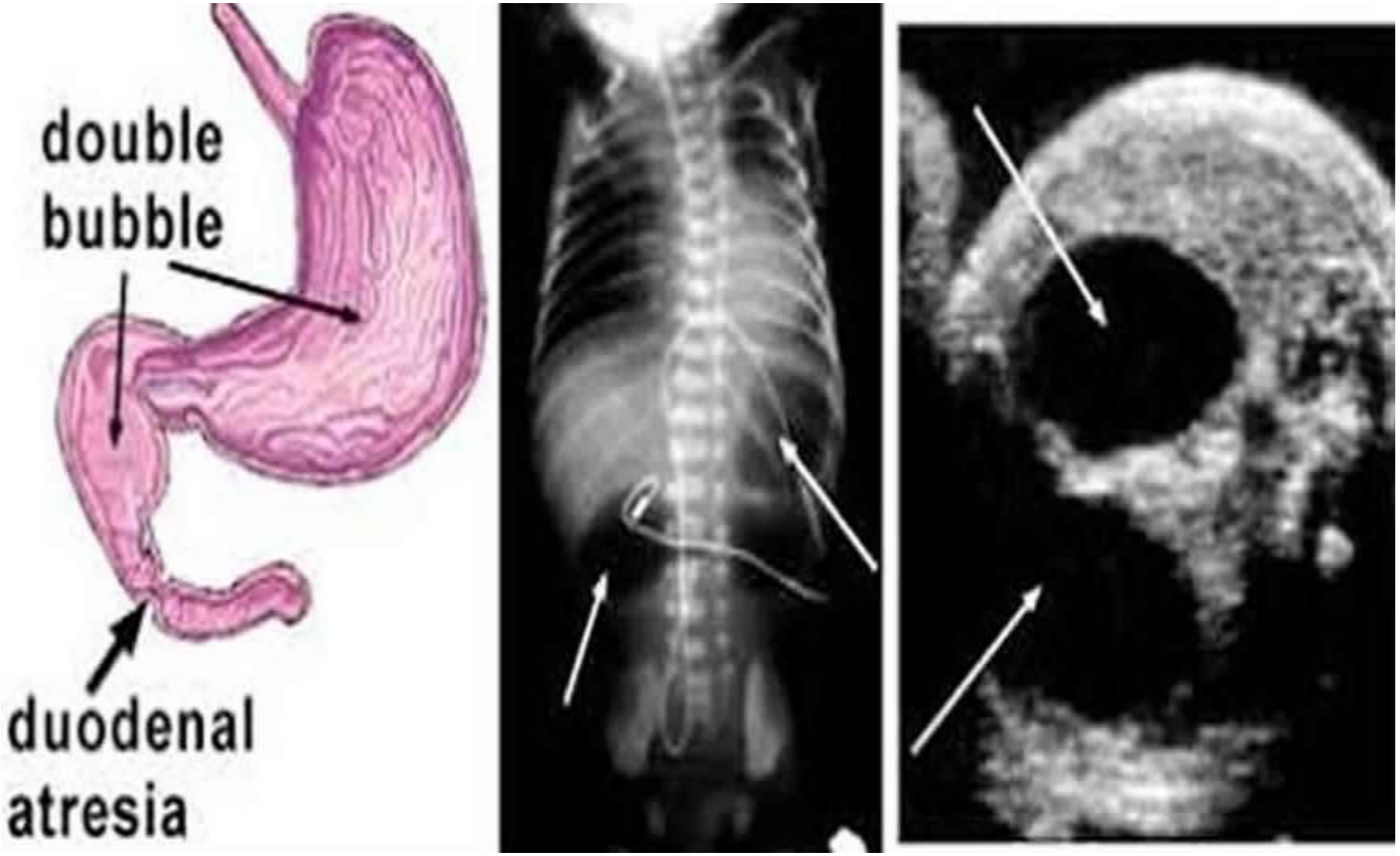
- Ante Natal
  - Polyhydraminos – 50 %
  - Ante Natal detection by ultra sound
- Post Natal
  - Bilious Vomiting from birth
  - Vomiting may be blood stained
  - Emaciation and dehydration
  - High Intestinal Obstruction
  - Associations: Down’s syndrome, Cardiac anomalies etc.

INVESTIGATIONS

- A/N USG - double bubble sign- due to fluid

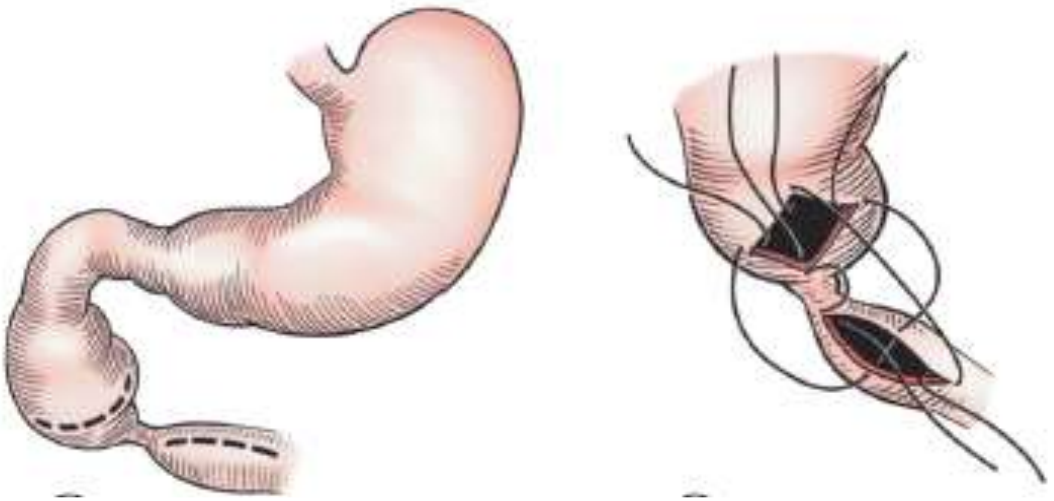
- P/N X RAY- double bubble sign due to air




double
bubble
duodenal
atresia

TREATMENT

- Type 1 Duodenoplasty (longitudinal-transverse)
- KIMURA'S DUODENODUODENOSTOMY



- Type 2 - Duodenoduodenostomy

JEJUNO ILEAL ATRESIA

- intrauterine mesenteric vascular accidents like bands, intussusception or internal hernias

CLINICAL PRESENTATION

- Maternal polyhydramnios due to upper GI obstruction in the foetus.
- Bilious vomiting
- Abdominal distension
- Jaundice
- Failure to pass meconium on the first day of life

INVESTIGATIONS

Prenatal imaging

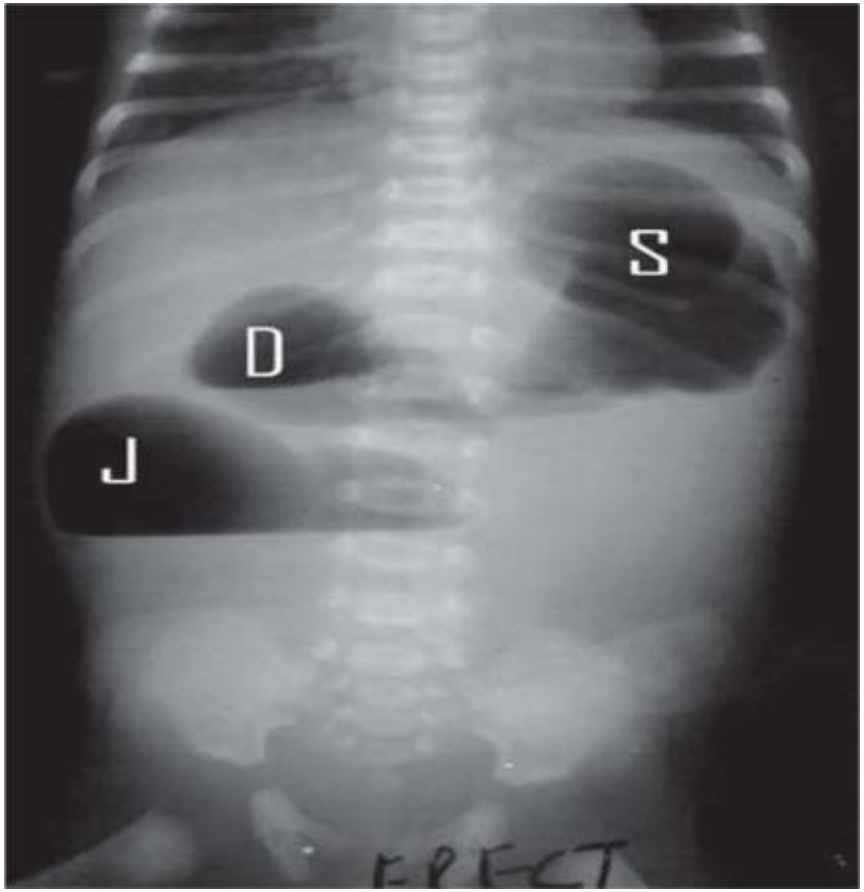
Antenatal USG

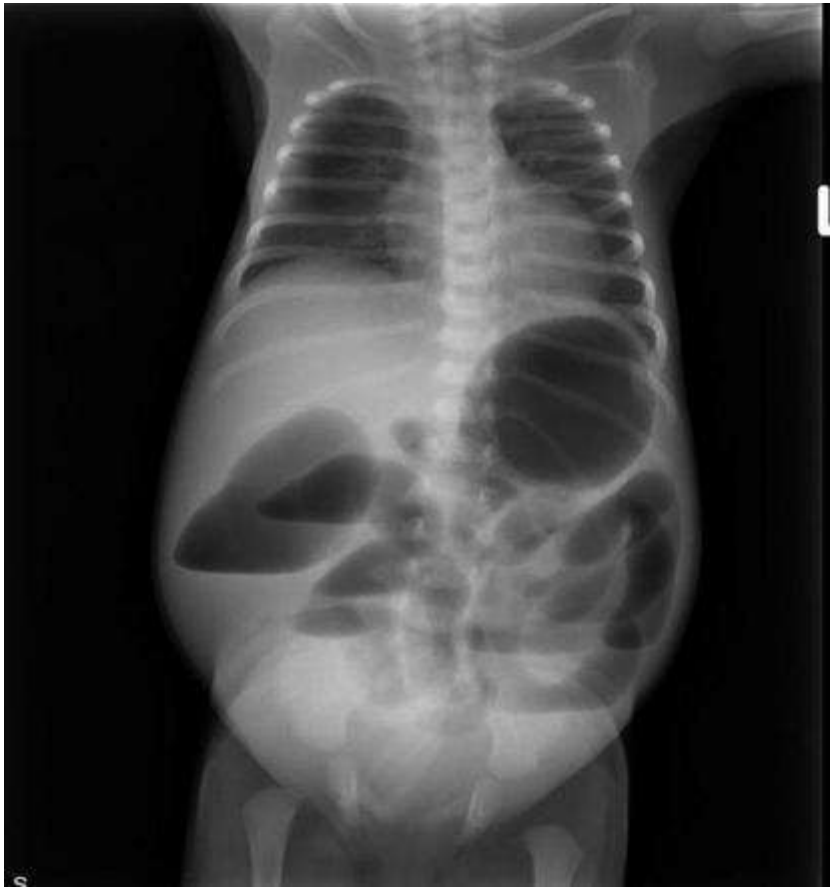
- Polyhydramnios due to upper GI obstruction in the foetus.
- Multiple distended loops of proximal bowel with vigorous peristalsis

Fetal MRI

Postnatal radiographic findings

- X ray babygram in erect posture - thumb sized intestinal loops and air fluid levels are highly suggestive of intestinal obstruction


S
D
J

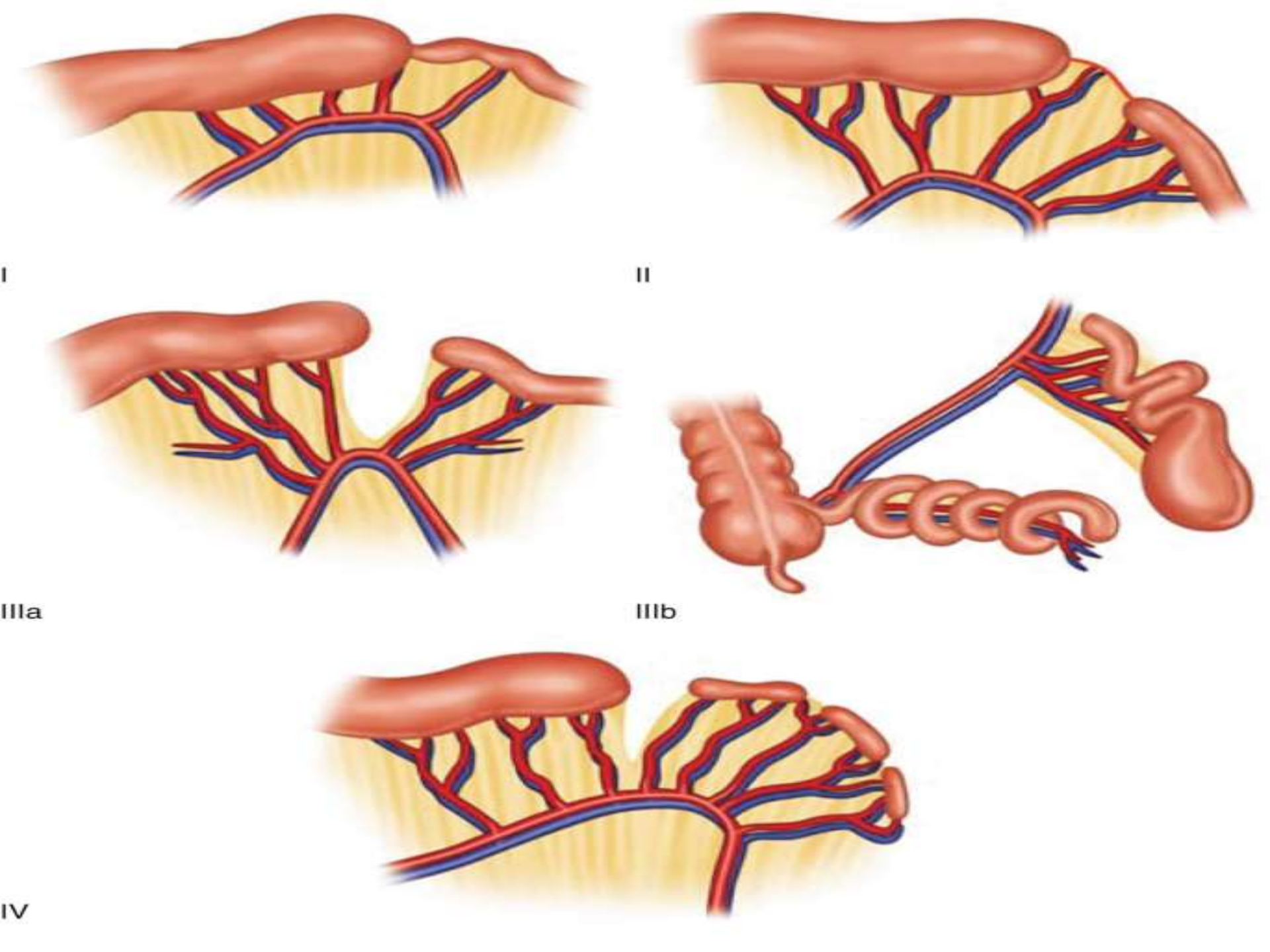


DIFFERENTIAL DIAGNOSIS

- Malrotation with or without volvulus
- Meconium ileus
- Intestinal duplication
- Internal hernia
- Colonic atresia
- Adynamic ileus secondary to sepsis
- Total colonic aganglionosis

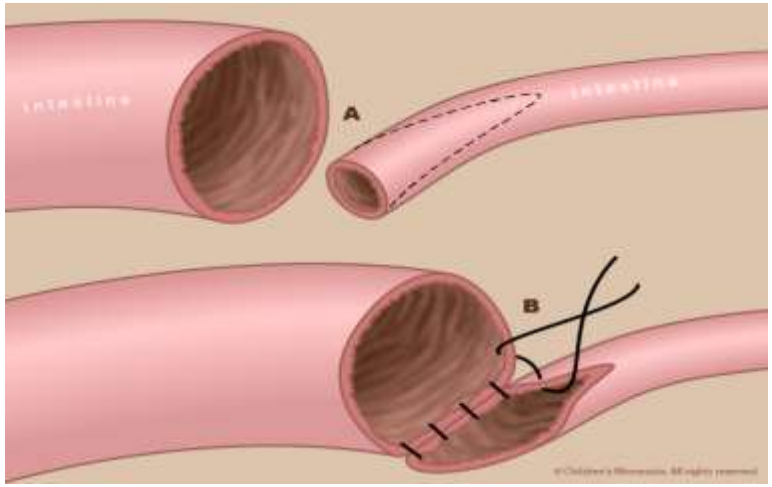
TYPES

- **Type1-**mucosal /septal atresia with an intact bowel wall (19%)
- **Type2-**two atretic ends connected by a fibrous cord with an intact mesentery (31%)
- **Type 3a-**two separated segments of bowel with a V shaped gap with in the mesentery
- **Type3b-**apple peel or Christmas tree appearance (46%)
- **Type 4-**multiple atresias “string of sausages” appearance (6-20%)


I
II
IIIa
IIIb
IV

- # TREATMENT

- Laparotomy, excision of web or resection and primary anastomosis as the case may be.



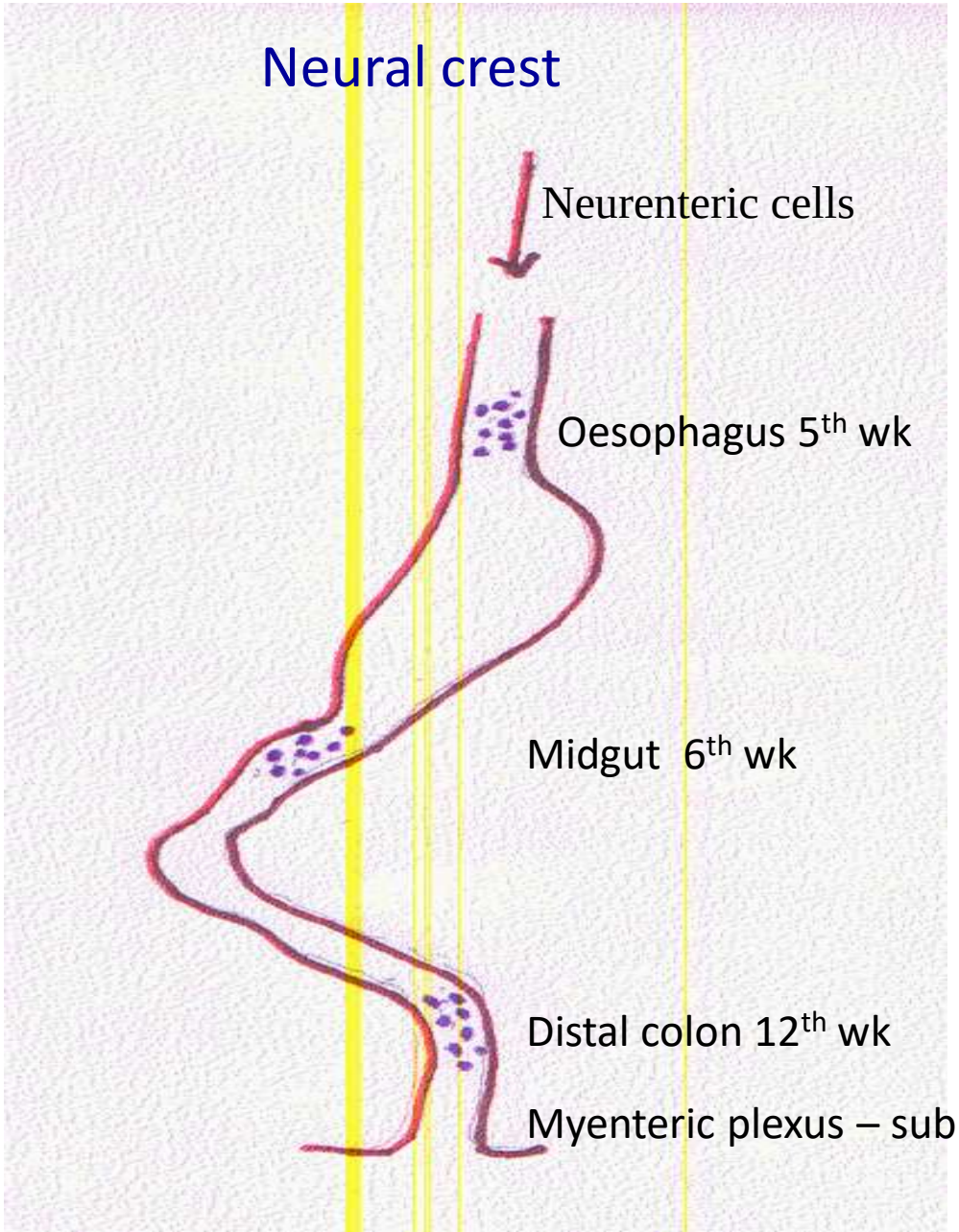
HIRSCHSPRUNG'S DISEASE (HD)

Other name: Congenital intestinal aganglionosis

ETIOLOGY

Failure of migration of neural crest cells (precursor of ganglion cells) during foetal life.

It can be syndromic (Down's syndrome, Waardenburg syndrome) or non- syndromic (90%).


Neural crest
Neurenteric cells
Oesophagus 5th wk
Midgut 6th wk
Distal colon 12th wk
Myenteric plexus – submucosal plexus

TYPES

- Ultra short segment HD – aganglinosis only at the internal sphincter of rectum
- Classic HD (most common) - the transition zone (TZ) is in the recto-sigmoid and aganglinosis extends from the internal sphincter of rectum up to the rectosigmoid junction
- Long segment HD - TZ is proximal to the sigmoid colon
- Total colonic HD - Aganglionosis affects the entire colon

PRESENTATION

1) Failure/delay in passage of meconium within the first 48h of life
2) Vomiting and abdominal distension
3) Chronic constipation and failure to thrive in older infants and children
4) Hirschsprung associated enterocolitis (HAEC) is a severe form - fever, foul smelling diarrhoea, lethargy and shock

CLINICAL FEATURES

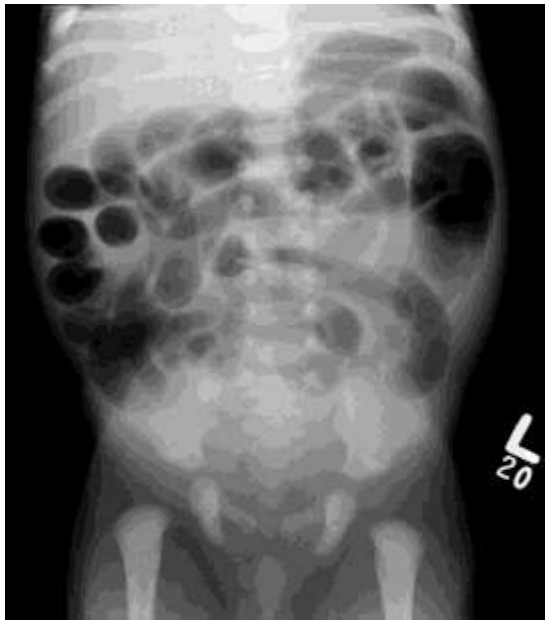
- Abdominal distension, bowel loops may be palpable
- Empty rectal vault or forceful expulsion of faeces during rectal examination
- Can be very sick if HD eneterocolitis ensues
- Malnutrition on late presentation (failure to thrive)

DIFFERENTIAL DIAGNOSIS

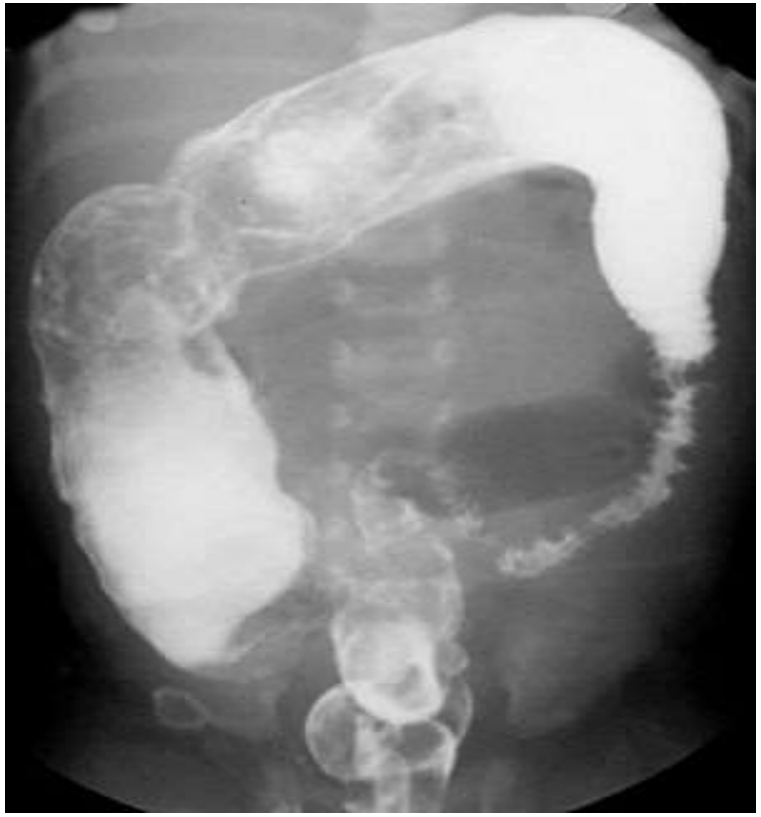
- hypothyroidism,
- intestinal atresia,
- motility disorders,
- inspissated meconium,
- habitual constipation,
- prematurity,
- anorectal malformation.

DIAGNOSIS

- Total white count may be elevated in enterocolitis. TSH should be normal
- Plain abdominal X-ray may show dilated loops with absent rectal gas shadow



- Barium enema can show funnel shaped TZ in most cases. The distal bowel is narrow and proximal distended



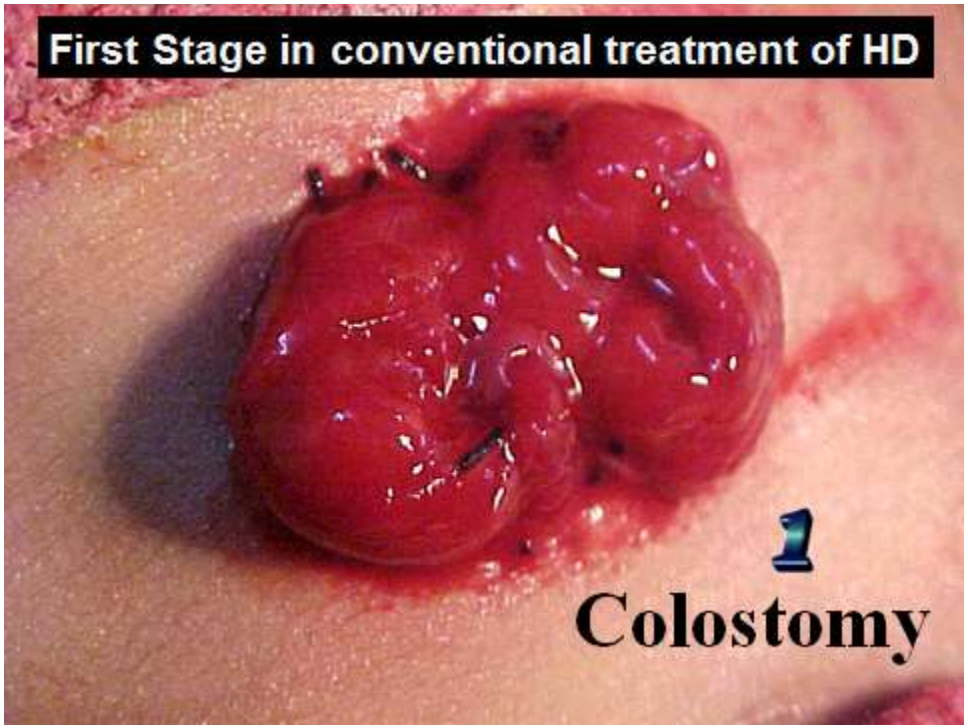
- Rectal biopsy (open or suction) is the gold standard - absence of ganglion cells and presence of hypertrophied nerve fibres are typical.
- Special stains like AChE (acetyl choline esterase) and Calretinin may be used

- Ano-rectal manometry is often performed for older children in some centres

TREATMENT

Aims at removal of diseased segment partially or in Toto in three staged procedures

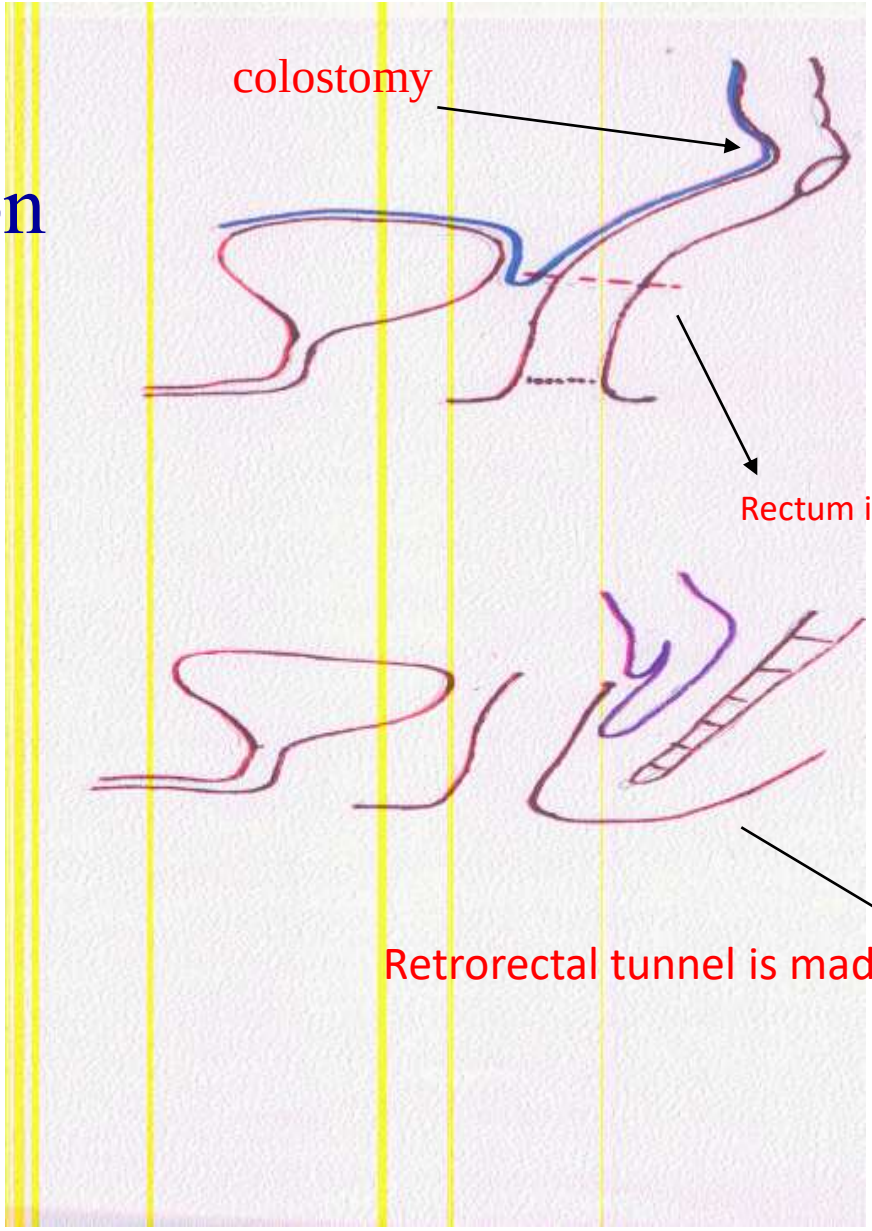
- Colostomy
- Definitive procedure,
- Colostomy closure

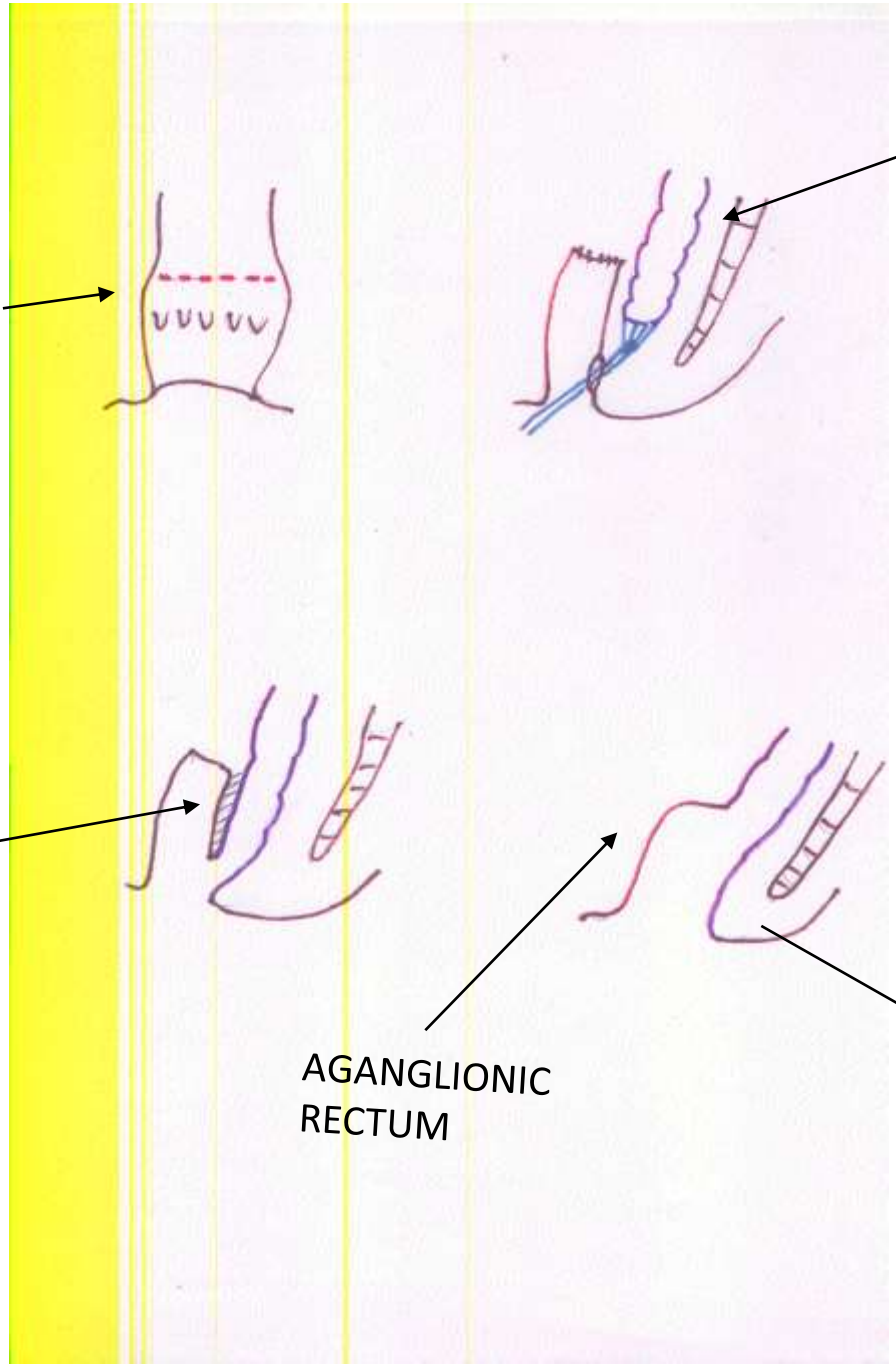

First Stage in conventional treatment of HD
1
Colostomy

Modified Duhamel's is the most popular one.

- The proximal ganglionic colon is brought behind the aganglionic rectumand the intervening wall is either crushed with a clamp or cut and anastomosed with intestinal staplers to create a common chamber formed by the rectum anteriorly and colon posteriorly

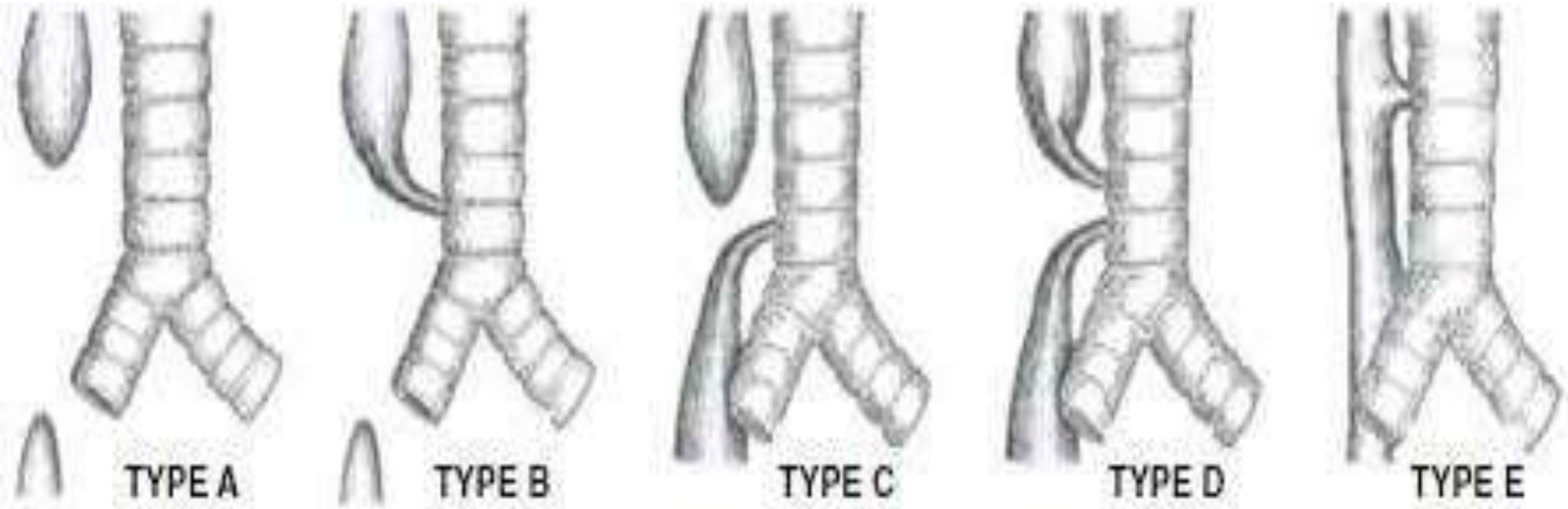
Duhamel's operation




GANGLIONATED
BOWEL
PULLED
THROUGH
LEVEL OF INCISION
SEPTUM
AGANGLIONIC
RECTUM
GANGLIONATED
BOWEL

TEF

- **CLASSIFCATION**



ASSOCIATED ANOMALIES

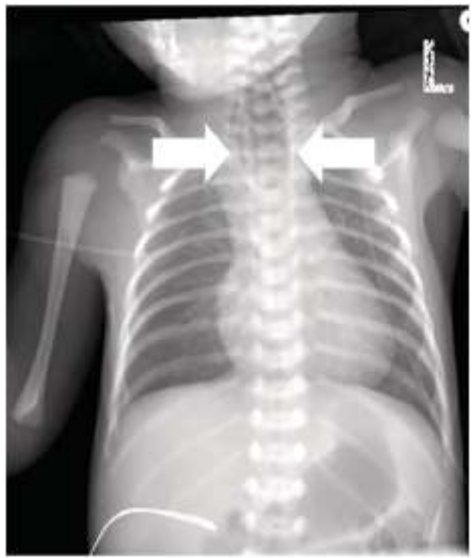
- **V** **V**ertebral
- **A** **A**norectal
- **C** **C**ardiac
- **T** **T**racheal
- **E** **E**sophageal
- **R** **R**enal
- **L** **L**imb

CLINICAL FEATURES

- Antenatal-Polyhydramnios
- Postnatal presentation is characterized by drooling of saliva, choking, coughing, and cyanotic attacks – withhold feeds
- Inability to pass 10F Feeding Tube (orogastric)
- Associated malformations must be looked for

INVESTIGATIONS

- Plain X-ray including the neck, the thorax, and the abdomen
  - ✓FT coiled in upper pouch
  - ✓Gasless abdomen- oesophageal atresia without fistula
  - ✓Lung for signs of aspiration



- Cardiologic assessment
- Abdominal and urogenital ultrasound
- Examination of vertebra
- Chromosomal analysis

- **PREOPERATIVE MANAGEMENT**

- Nursed in prone propped up position with head down(avoids aspiration and GER)
- Low pressure frequent suction/ Replogle tube
- Intubation and ventilation only if necessary
- Parenteral fluids, Inj. Vit K

OPERATIVE MANAGEMET

Esophageal Atresia with Distal Tracheoesophageal Fistula (85%)

- Right Thoracotomy - divide the fistula, to close it on the tracheal side, and to perform an end-to-end anastomosis

Esophageal Atresia with Proximal and Distal Tracheoesophageal Fistula (1.5%)

-fistula is closed flush to the trachea and esophagus.

Esophageal Atresia with a Proximal Tracheoesophageal Fistula Only: A Long Gap Problem (1%)

-gastrostomy and closure of fistula, Replogle tube allows continuous suction

H-Type Fistula (4%)

-ligation of fistula, mostly cervical in position. Recurrent laryngeal nerve injury or neuropraxia should be considered.

Isolated Esophageal Atresia: The Long Gap Problem (8.5%)

-placement of a feeding gastrostomy or feeding jejunostomy, with or without cervical oesophagostomy

-either the preservation of the patient's own esophagus with delayed repair or esophageal replacement with small bowel, large bowel or stomach

OMPHALOCELE

- Omphalocele is a large defect (>4cm) at the base of the umblical cord covered by amniotic membrane that contains midgut and other abdominal organs including liver and often the spleen and gonads.

- Umblical cord is usually attached eccentrically to the herniation.
- Usually associated with malrotation
- Small abdominal cavity and pulmonary hypoplasia

- 1 to 2.5 in 5000 live births
- Male preponderance
- Increased risk with advanced and very young maternal age.
- Maternal obesity
- Nutritional factors are failure to use multivitamins during pregnancy, lack of folic acid fortification and alteration in glycemic control.

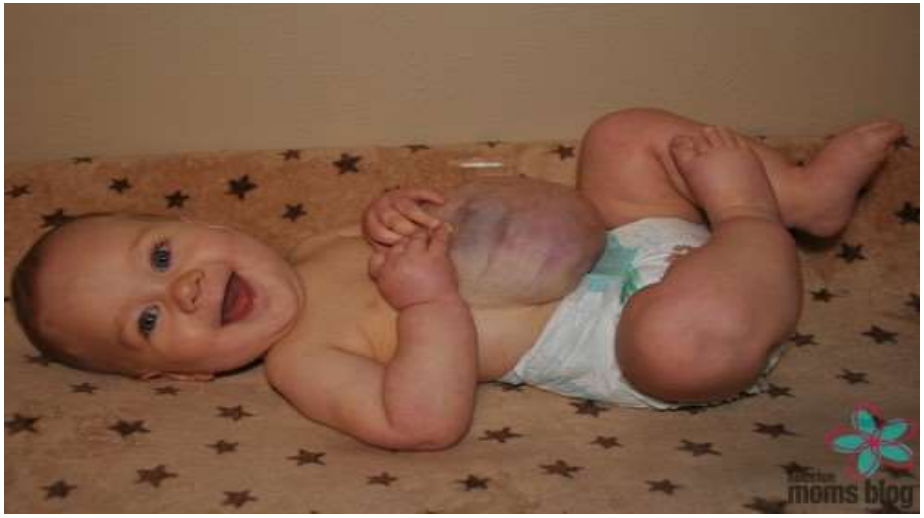
- **Associated syndromes and anomalies are very common (45-55%) unlike as in gastroschisis**

INVESTIGATIONS

- Antenatal USG
- Elevated AFP in both maternal serum and amniotic fluid
- Prenatal MRI can be used to screen for anomalies such as complex cardiac defects and nervous system anomalies

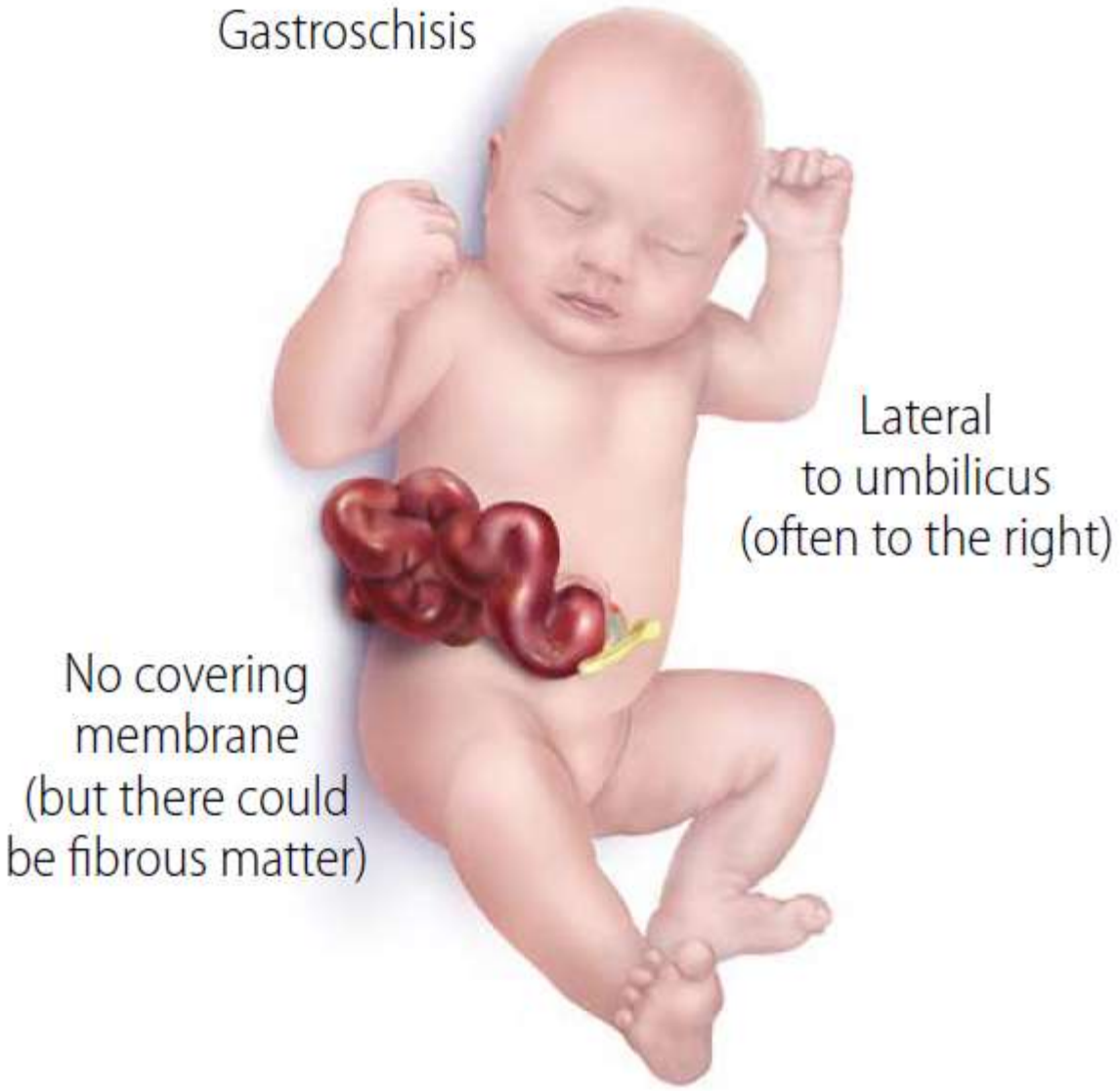
MANAGEMENT

- Conservative management- painting the surface of the intact omphalocele with spirit, povidine iodine, mercurochrome etc for several weeks to stimulate epithelialization followed by delayed ventral hernia repair, gives better prognosis.



GASTROSCHISIS

- Located lateral to attachment of umbilical cord
- Defect less than 4cm
- Sac is absent
- Cord attachment is normal
- Occasionally, a skin bridge may be present between the cord and the defect.


Gastroschisis
Lateral
to umbilicus
(often to the right)
No covering
membrane
(but there could
be fibrous matter)

- Usually contains midgut and stomach
- Thickened,foreshortened,atretic, and possibly ischemic bowel
- Bowel oedematous and leathery as the herniation occurs in fetal life into the amniotic fluid.

- Liver not herniated
- Abdominal cavity space is normal unlike as in omphalocele
- Associated anomalies are rare
- Bowel atresia is common

TREATMENT

- Silo