Edited by: Tasnim Ahmed

40 pages

Miscellaneous topics

Dr. Abeer Diab

Topics

. Hypertrophic Pyloric Stenosis (HPS)

. Intussusception

. Congenital Abdominal Defects (2 types)

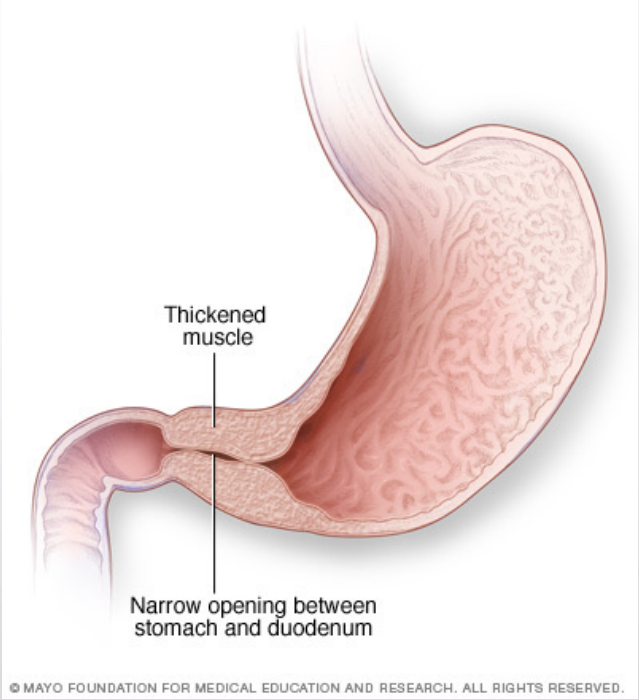
. Meckel diverticulum

. Biliary Atresia

1- Hypertrophic Pyloric Stenosis (HPS)

- M:F = 4 : 1

- Risk factors:
  - Family history
  - Male gender M > F
  - Younger maternal age
  - Being a first-born infant
  - Maternal feeding patterns



Aetiology

Unknown (multifactorial with environmental influences)

- Genetic factors
  - race discrepancies
  - increased frequency in **males**
  - **first-born infants** with a positive family history)
- Environmental factors
  - method of feeding (breast vs **formula**)
  - seasonal variability
  - exposure to erythromycin
  - **transpyloric feeding** in premature infants
- Other factors
  - excessive substance P
  - decreased neurotrophins
  - deficient nitric oxide synthase
  - gastrin hypersecretion

Presentation :

Nonbilious, progressive projectile vomiting (of recent feedings)
full-term neonate
2-8 weeks old

On exam : Usually appears well (early) but if late presentation , they will show signs of dehydration
Visible gastric peristaltic waves
Palpable pylorus "olive sign" (70–90% of patients)

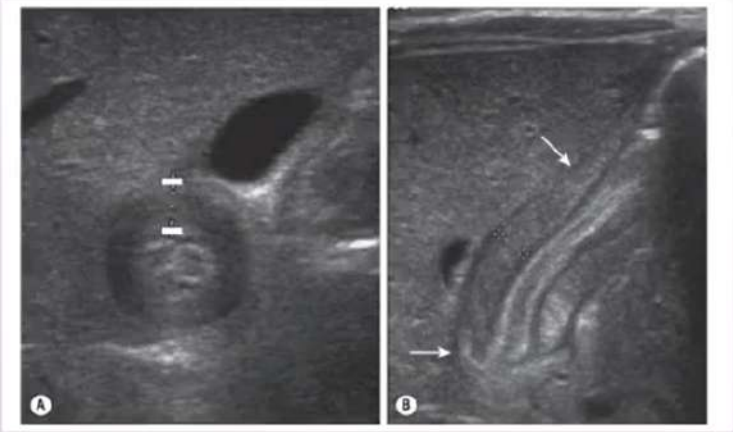
Investigations : Hypochloremic hypokalemic metabolic alkalosis

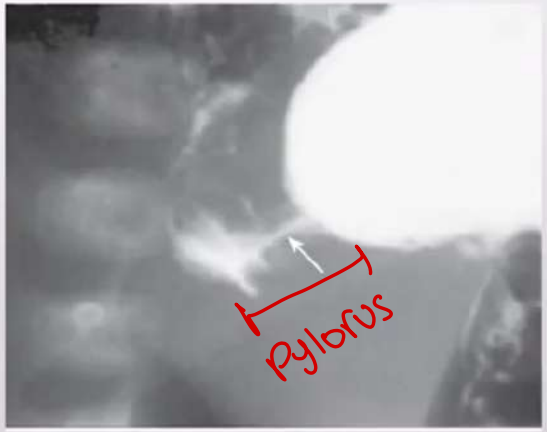
Diagnosis

- US: muscle thickness of ≥4 mm and a pyloric length of ≥16 mm

When US findings are equivocal, then do Upper gastrointestinal series

narrowed pyloric lumen

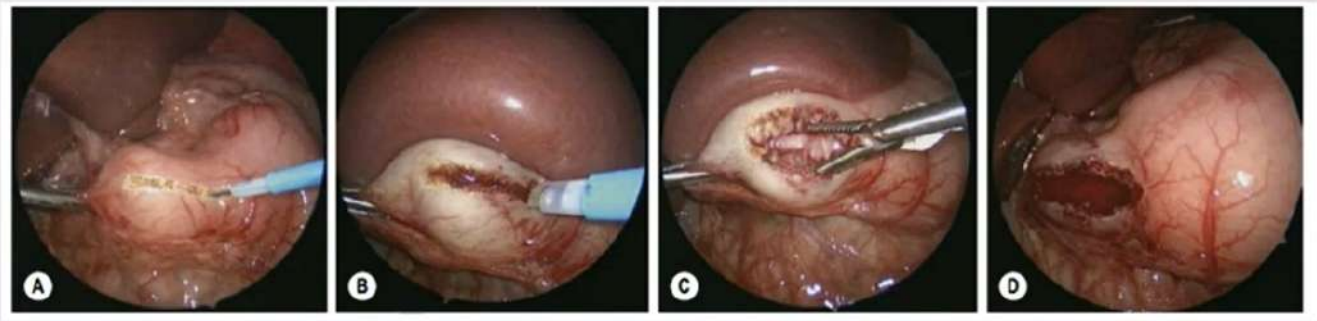




Management

- Preop. supportive measures:
  - NPO+/- gastric decompression
  - IV fluid resuscitation
  - Correction of electrolytes

- Surgery:
  - Non-emergent
  - Laparotomy or laparoscopic Pyloromyotomy don't reach mucosa, just cut part of the muscle



Complication

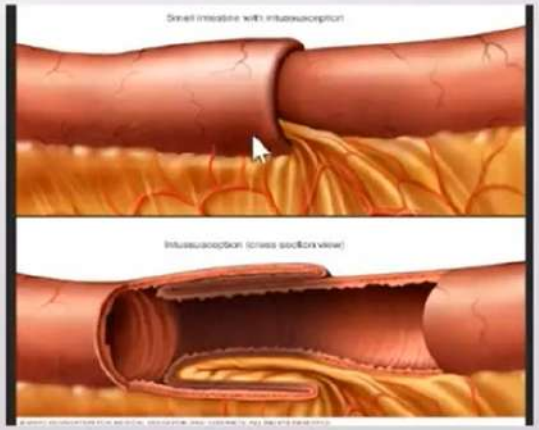
- Mucosal perforation (1-2%)
- Postoperative emesis (occur in most infants)
- Prolonged postoperative emesis

(less common | due to GER or incomplete myotomy)

2- Intussusception

An acquired invagination of the proximal bowel (intussusceptum) into the distal bowel (intussuscipiens) that will compress the mesentery resulting in venous obstruction and bowel edema → into arterial insufficiency, ischemia and bowel wall necrosis

It is the most common cause of small bowel obstruction in this age group



- **Primary** : no leading point , likely due to hypertrophied Peyer patches within the bowel wall.

  mostly: distal & proximal ileum

  between ages 4 and 9 months

  ¬ 2/3 are boys

- **Secondary** :Meckel diverticulum , polyps and duplications, appendix, hemangiomas, carcinoid tumors, foreign bodies, ectopic pancreas or gastric mucosa, hamartomas from Peutz– Jeghers syndrome and lipomas, lymphomas and small bowel tumors.Henoch–Schönlein purpura and cystic fibrosis, celiac disease and Clostridium difficile colitis

colicky

- The classic presentation is an infant or a young child with intermittent, cramping abdominal pain every 15-30 min associated with "currant jelly" stools and a palpable mass on physical examination (seen in <25%)

↳ late sign due to sloughing of mucosa

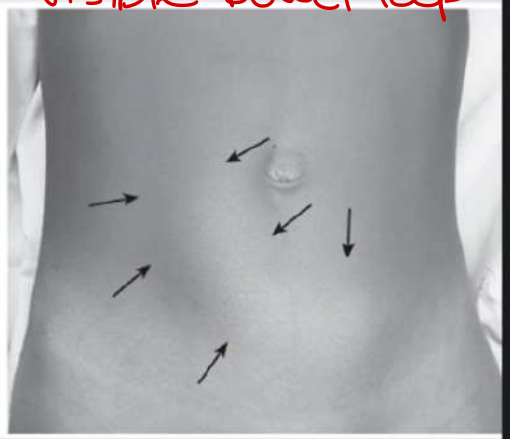
- Pain is associated with: Vomiting (gastric early | bile later) , Abdominal distension , Lethargy (later) , Red currant jelly stools (later) , hyperextension and flexion of the knees up

- On exam :

↳ Rt iliac fossa

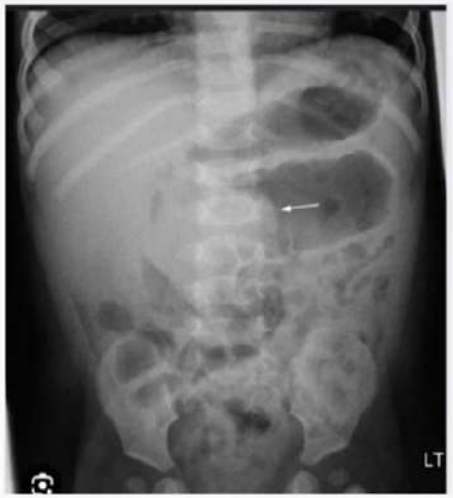
Signs of dehydrations , Abdominal distension , RUQ mass . Empty RIF (Dance sign) in quick succession as a result of bacteremia and bowel necrosis.

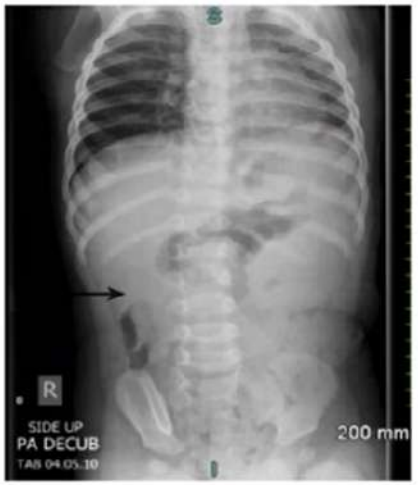
visible bowel loop



Diagnosis

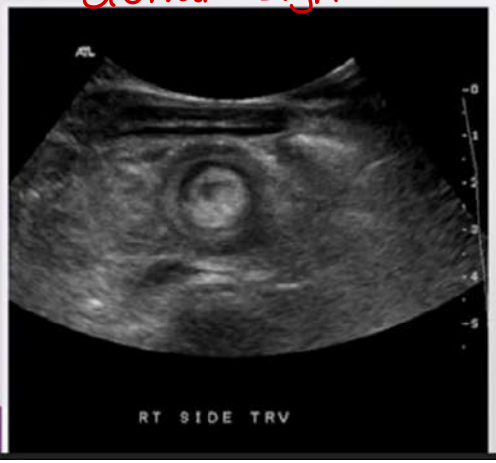
- Xray :



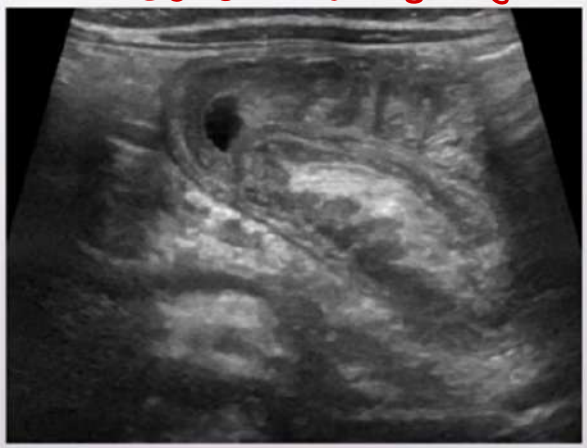


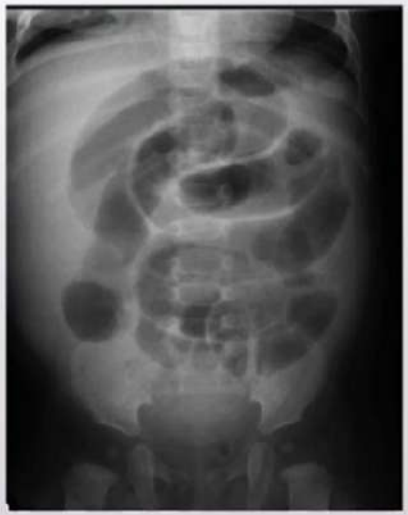
- US : Target' or 'donut' lesion (in transverse plane) , Pseudokidney' sign (on longitudinal plane)

donut sign



Pseudokidney sign





Management

Initial management :

- NGT (to decompress the stomach)
- NPO
- IV fluid resuscitation and maintenance IVF
- correct electrolytes disturbances

Non operative management (if patient is stable & no signs of peritonitis)

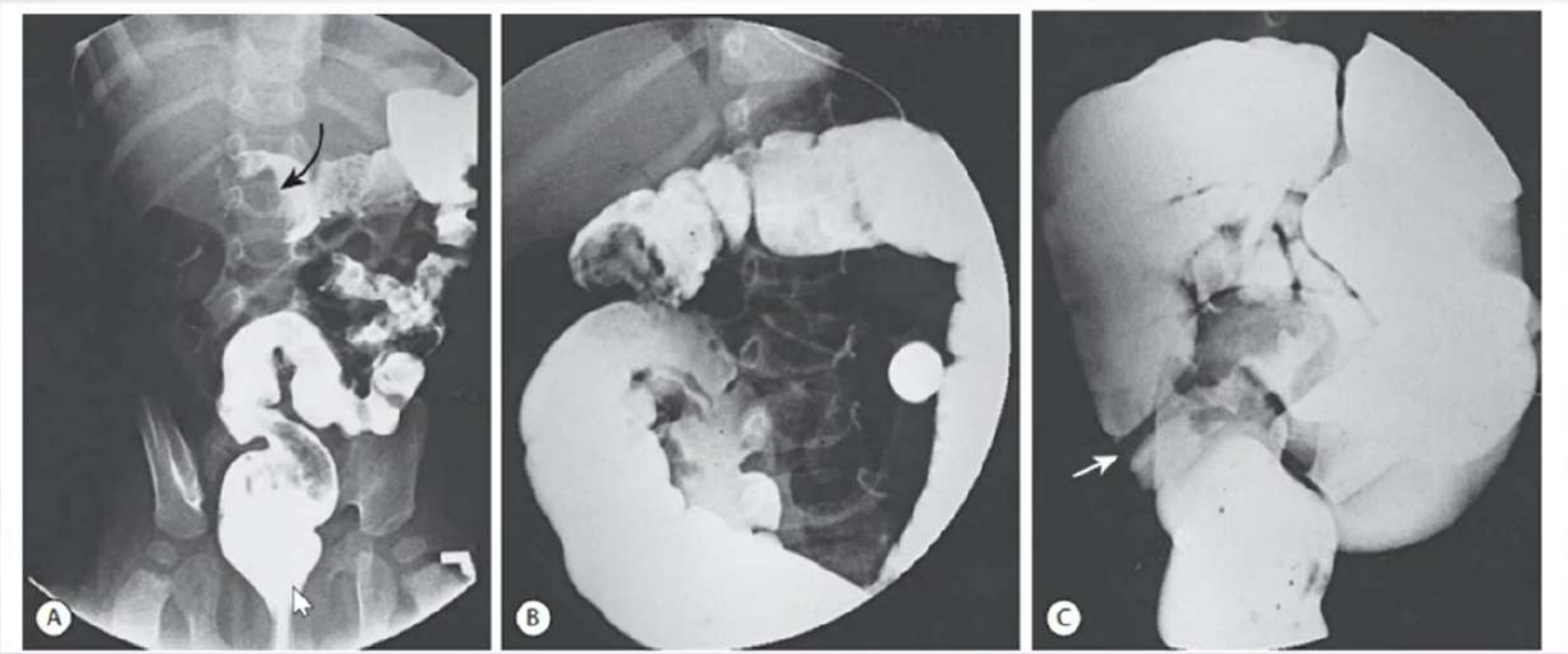
Hydrostatic/ Pneumatic Reduction (Under fluoroscopy or ultrasound guidance)

fluid ← | → air

C/I : perforation /peritonitis , persistent hypotension/tachycardia-sepsis

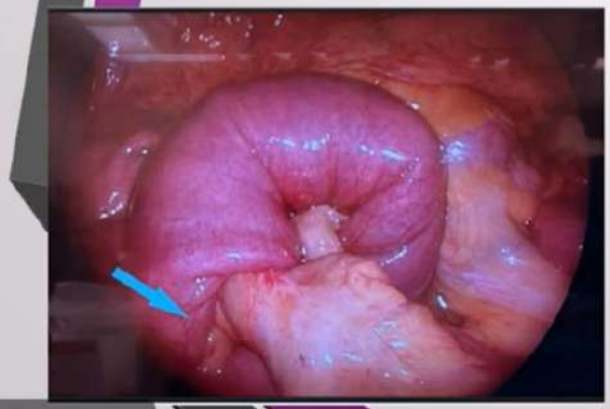
Success rate ¬85%

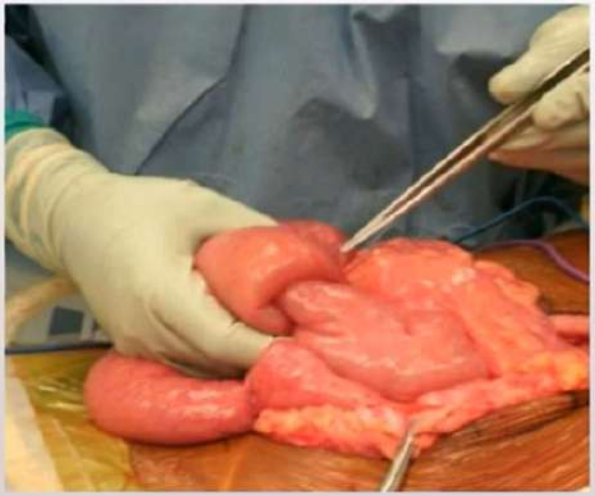
→ contraindications



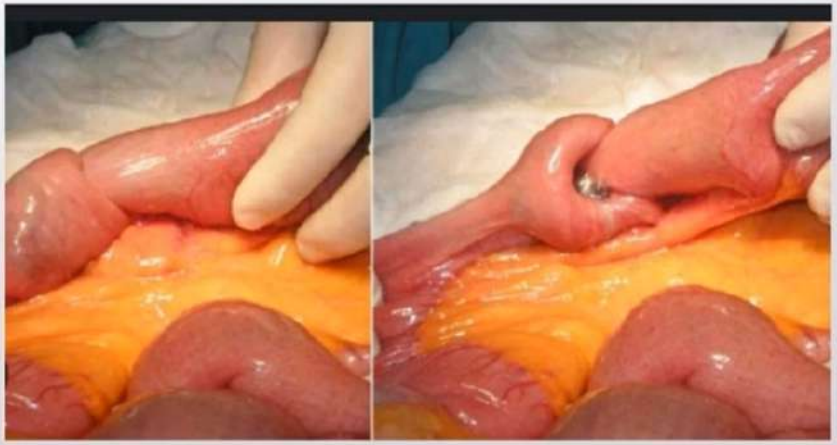
bowel inside bowel

- **Operative management (laparoscopic or open)**
- Indications:
  - Nonoperative reduction is unsuccessful or incomplete
  - Signs of peritonitis/ pneumoperitoneum
  - Presence of a lead point (secondary intussusception)

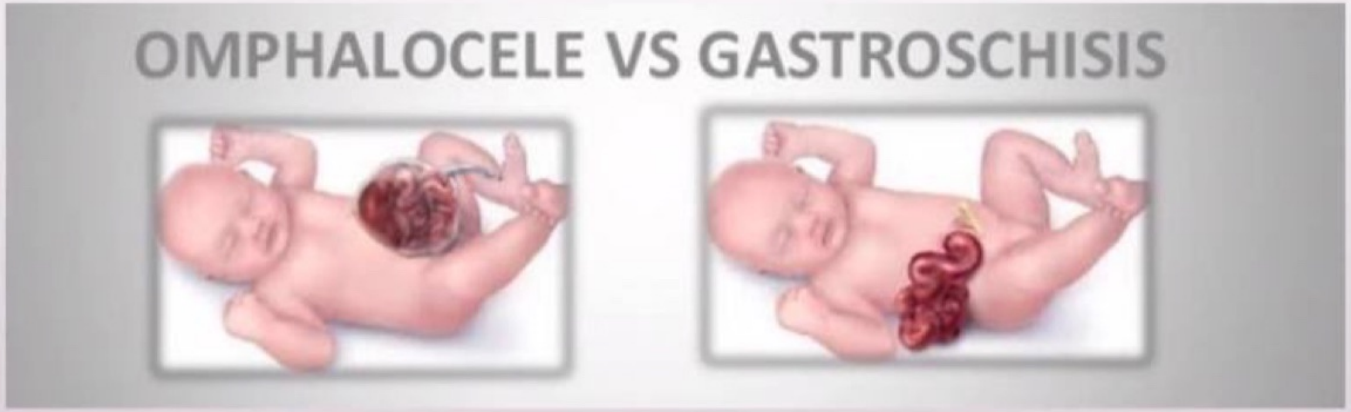




lead point



3- Congenital Abdominal Wall Defects (Omphalocele and Gastroschisis)



happens due to failure of retraction embryonically or failure of closure during rotation of the bowel

1. herniation 2. rotation 3. retraction 4. fixation

A-Gastroschisis

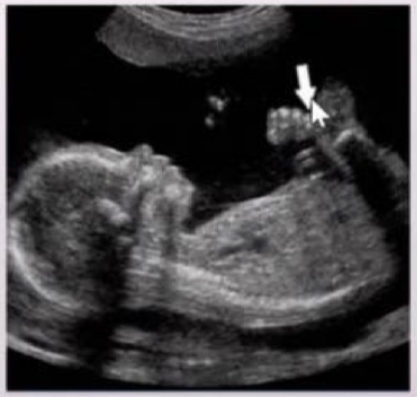
congenital abdominal wall defect characterized by the extrusion of abdominal contents (typically the intestines) through a small opening, usually to the right of the umbilicus. Unlike omphalocele, there is no protective membrane covering the exposed organs, leaving them directly exposed to amniotic fluid, which can cause inflammation and damage.

- 1 in 4000 live births
- Higher incidence in mothers younger than 21 years of age → associated with substance abuse
- Diagnosis : AN US by 20 weeks' gestation

Bowel loops **freely floating in the amniotic fluid** and a defect in the abdominal wall to **the right of a normal umbilical cord**) +**abnormal maternal serum α-fetoprotein (AFP) level,** which is universally elevated +/-Intrauterine growth restriction (IUGR)

Rt to umbilical cord

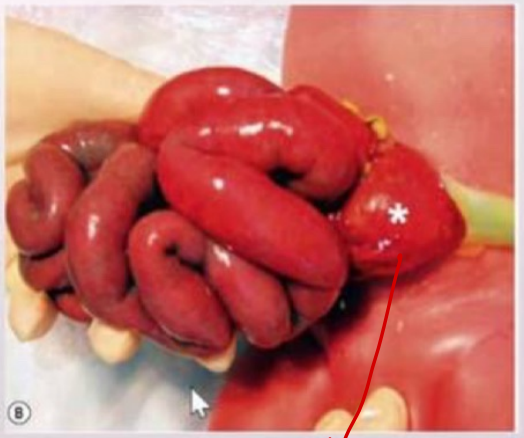
- Delivery should be in a tertiary centre



- Gastroschisis is associated with a variable degree of inflammatory thickening of the visceral bowel walls, which results in the characteristic appearance of "matted" intestines
- Associated with intestinal motility disorder , rotational disease , UDT 15-25% , bowel atresia

un-descendant testis

- Simple VS complicated (atresia ,Short bowel)



Stomach

Management

- Resuscitation

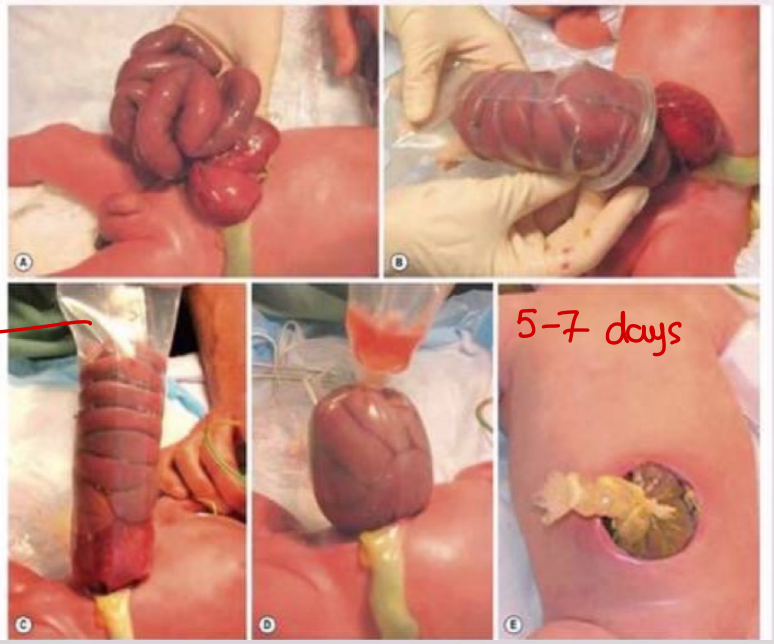
(NPO, NG, IVF, rectal tube to decompress)

- bowel should be wrapped in warm saline-soaked gauze and placed in a central position on the abdominal wall
- Surgery :

Either Primary closure or Staged closure

↳gradual reduction

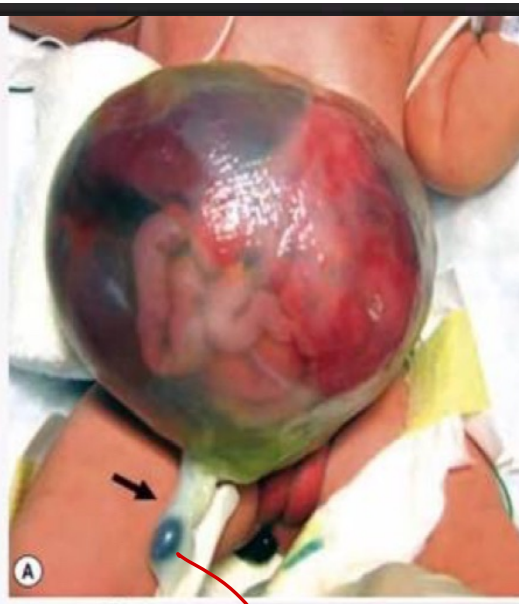
(with silo)



Long term outcome

- Long-term outcomes for infants born with gastroschisis are generally excellent
- Morbidities related to prematurity , bowel motility and length

B- Omphalocele



- 1 in 4000–6000.

  umbilical cord centered
- Associated with genetic defect and other anomalies (Trisomies 13, 18, 21, and 45 X ,Beckwith–Weideman, pentalogy of Cantrell , cardiac (14–47% incidence of anomalies) and central nervous (3–33% anomalies)
- Outcomes depend on associated anomalies
- Long term morbidities : gastroesophageal reflux disease (GERD), pulmonary insufficiency, recurrent lung infections or asthma, and feeding difficulty with failure to thrive

Diagnosis

Antenatally :

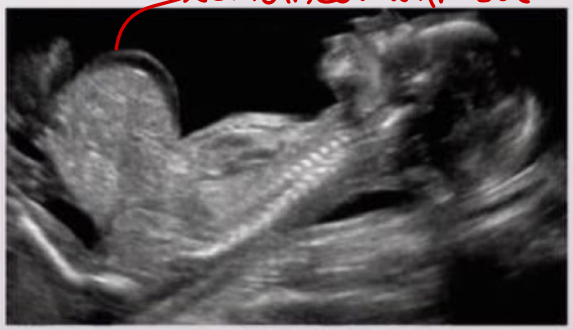
- ¬ 18-week US evaluation , elevated AFP

→most prognostic

(prognostic factor :omphalocele diameter compared with abdominal circumference (O/AC, or omphalocele ratio), the femur length (O/FL), and the head circumference (O/HC), , organ contained inside the sac

do karyotyping, renal US, fetal echo → associated anomalies

- Deliver in a tertiary centre , at term , normal vaginal delivery (except if it is giant omphalocele and containing liver (to avoid shoulder dystocia , sac rupture and bleeding)



Management

- **Resuscitation**

NPO, NG, IVF, rectal tube to decompress)

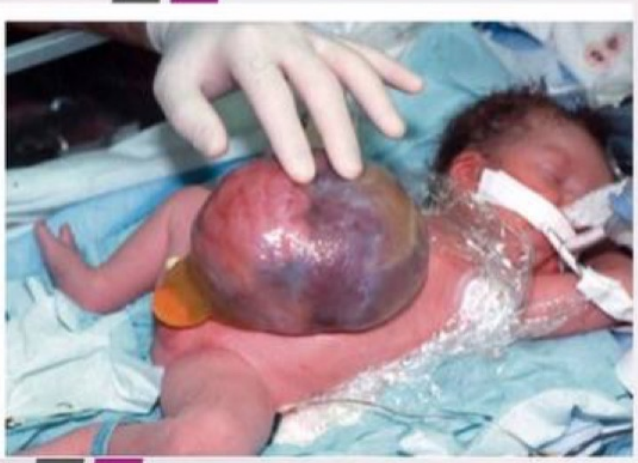
- **sac should be wrapped in warm saline-soaked gauze** and placed in a central position on the abdominal wall

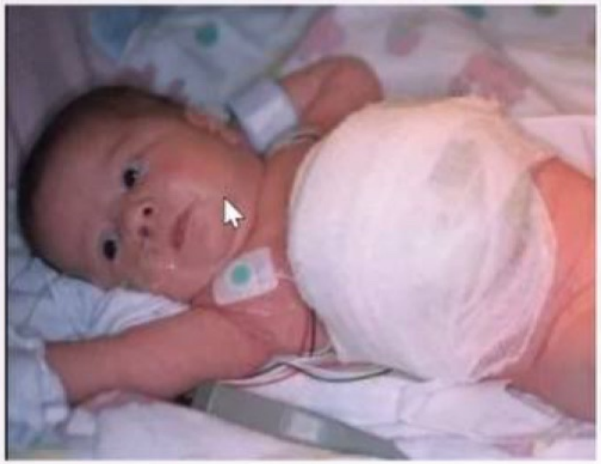
- **Surgery :**

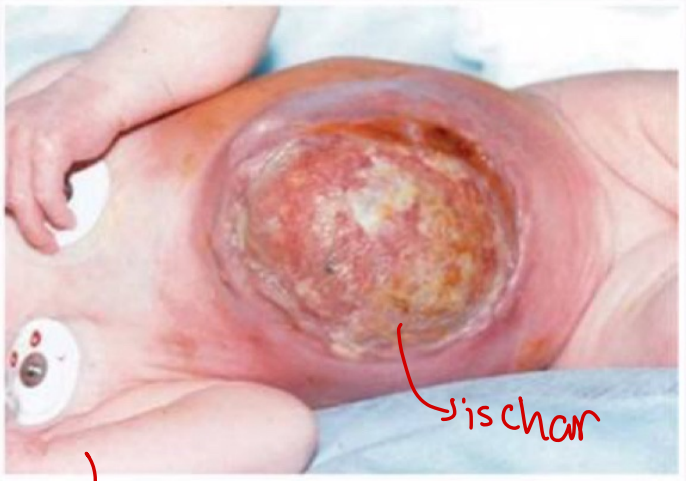
1 Primary closure : in small defect , consists of excision of the sac and closure of the fascia and skin over the abdominal content

2 Staged closure using a mesh or using a silo with serial reduction then closure if large sac

3 paint and wait/ Scarification technique), in case of giant omphalocele , associated cormobidities)







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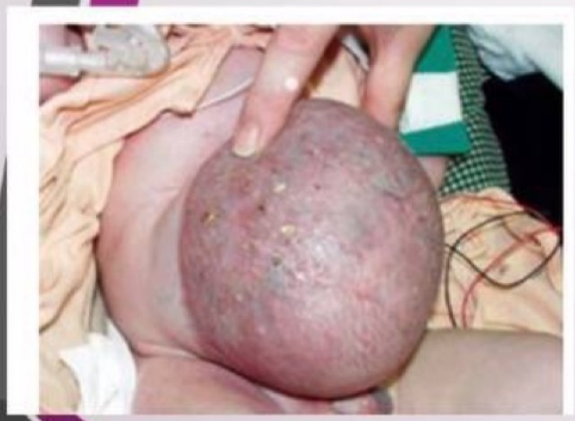
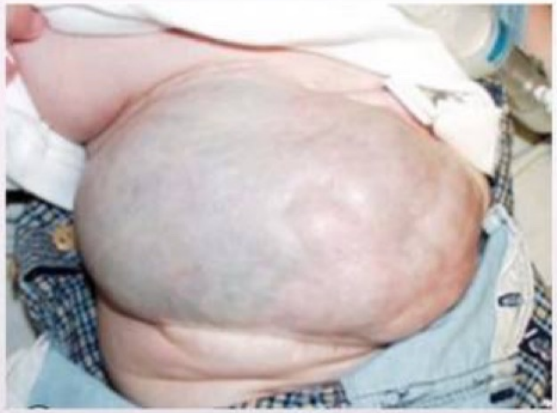




hernia

important :

Table 48.1 Differentiating Characteristics Between Gastroschisis and Omphalocele

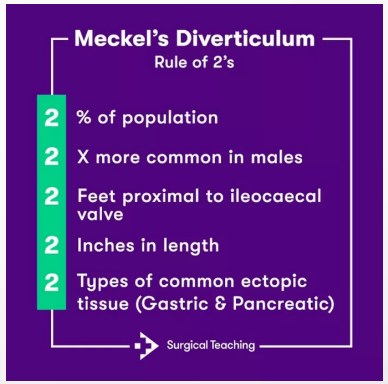
Characteristic	Omphalocele	Gastroschisis
Herniated viscera	Bowel ± liver	Bowel only
Sac	Present	Absent
Associated anomalies	Common (50%)	Uncommon (<10%)
Location of defect	Umbilicus	Right of umbilicus
Mode of delivery	Vaginal/cesarean	Vaginal
Surgical management	Nonurgent	Urgent
Prognostic factors	Associated anomalies	Condition of bowel

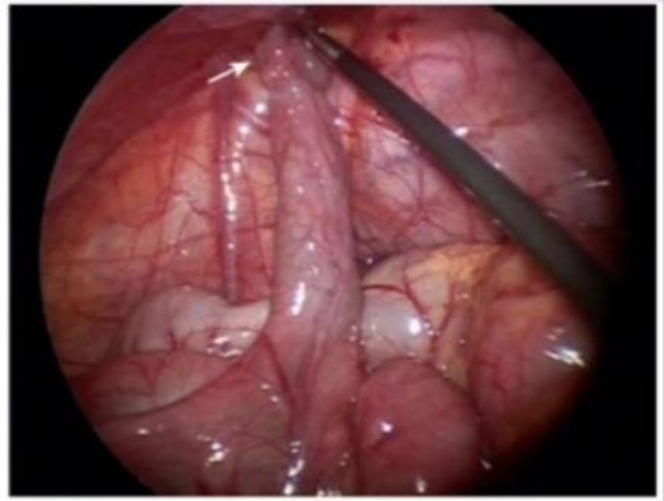
↳length & motility

4- Meckel diverticulum

- True incidence of Meckel diverticulum is unknown because most patients are asymptomatic.
- Estimated at approximately 2%,
- ¬ 4% will become symptomatic
- M:F of 2:1
- Rule of 2s : occurs in 2% of the population
  2:1 male-to-female ratio
  discovered by 2 years of age
  located 2 feet (60 cm) from the ileocecal valve
  commonly 2 cm in diameter and 2 inches (5 cm) long
  contain two types of heterotopic mucosa (Gastric is the most common followed by pancreatic)

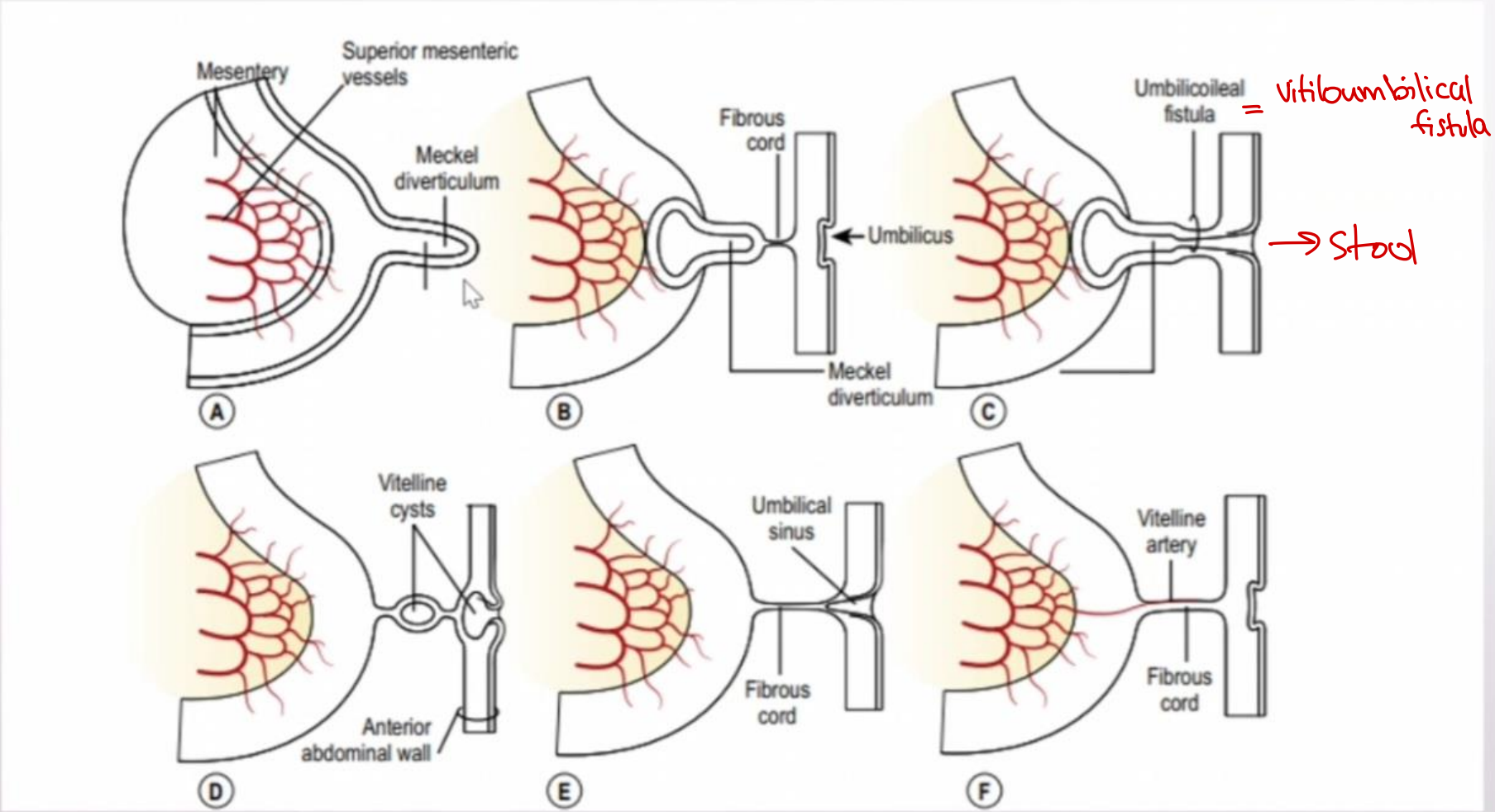
Rule of 2:





• true diverticulum : contains all layers of the wall

Vitilo-intestinal duct



↳ bc gastric acid causes erosion of the wall → bleeding

- The three most common presentations in children are **intestinal bleeding (30–56%), intestinal obstruction (14–42%), and diverticular inflammation (6–14)**
  - ↳ intessusception
  - ↳ volvulus
  - ↳ internal hernia
  - ↳ diverticulitis
- Less common signs include a cystic abdominal mass and a newborn with an umbilical fistula resulting from a patent vitelline duct, In elderly, neoplasia can develop within the Meckel diverticulum. (Carcinoid is the most common tumor)

Diagnosis

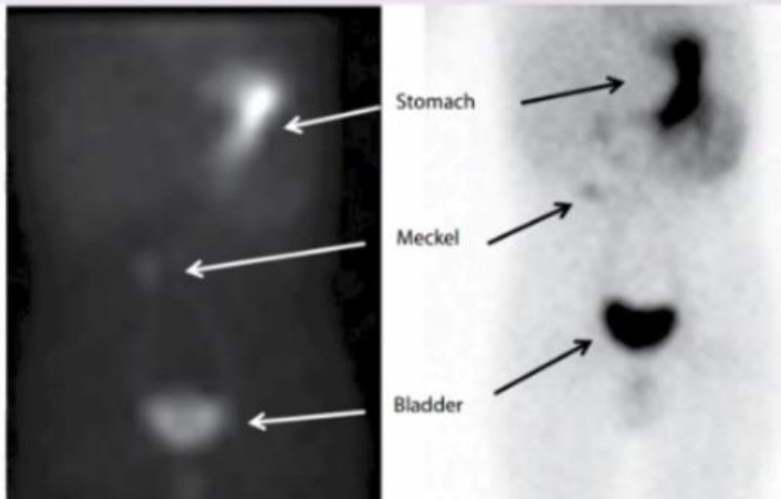
- In patients presenting with obstruction or inflammation, the diagnosis of a Meckel diverticulum is not usually definitively determined preoperatively
- US and CT might be helpful
- In case of bleeding diverticulum , technetium-99m pertechnetate radionuclide study (Meckel scan) , false negative 25%

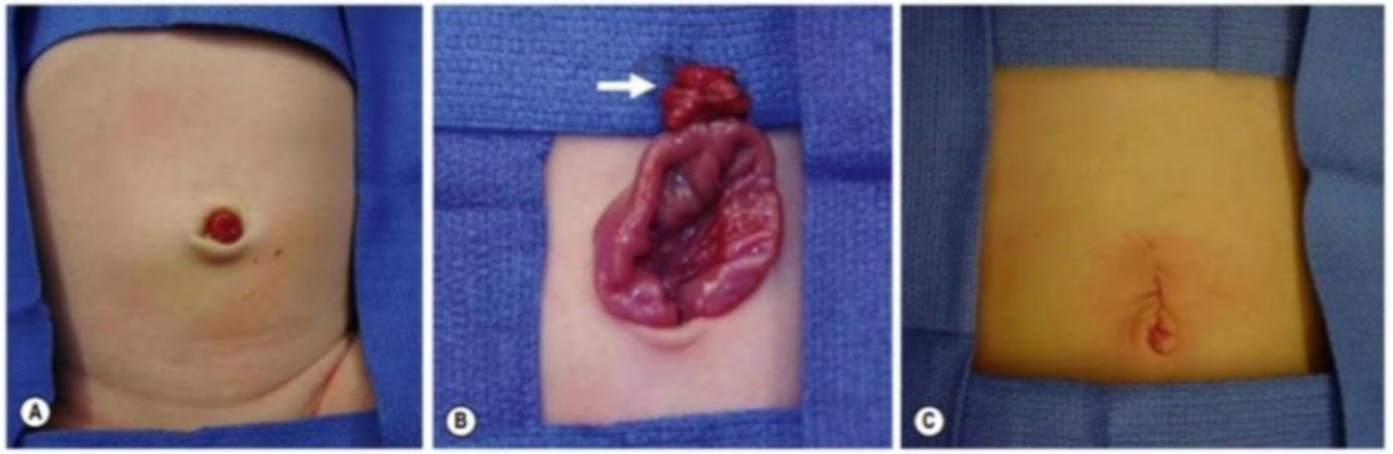
↳ can be pancreatic mucosa

Principle

- The scan uses **technetium-99m pertechnetate**, a radiotracer that is preferentially taken up by gastric mucosa.
- If a Meckel's diverticulum contains ectopic gastric tissue, it will concentrate the tracer, making it visible on the scan.

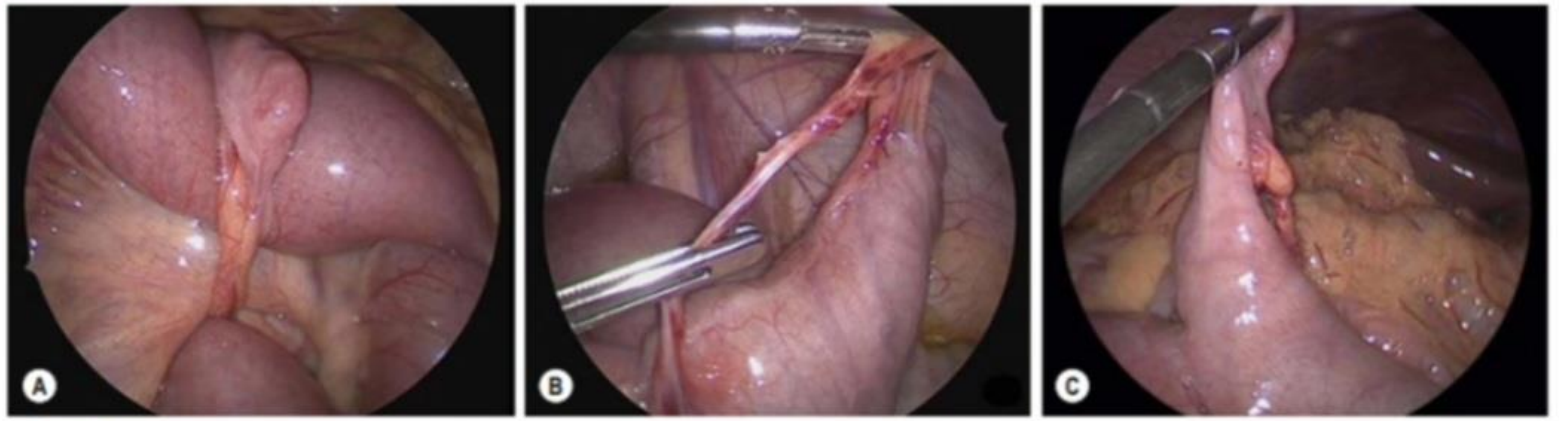
excretion through kidneys
normal people: stomach + ureter + bladder
meckel's: stomach + diverticulum + ureter + bladder





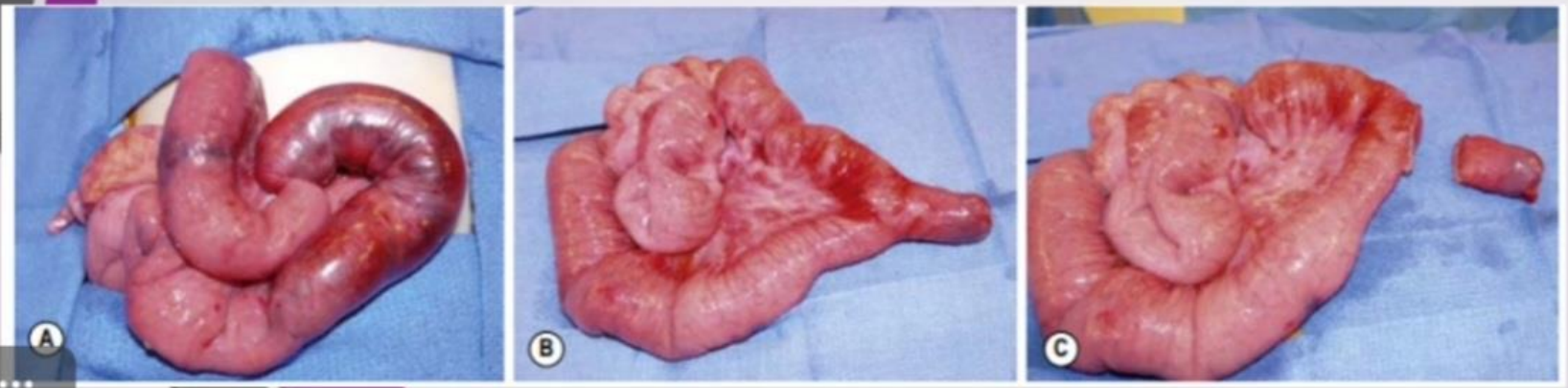
Vitiloumbilical fistula

Open



MIS

intestinal obstruction



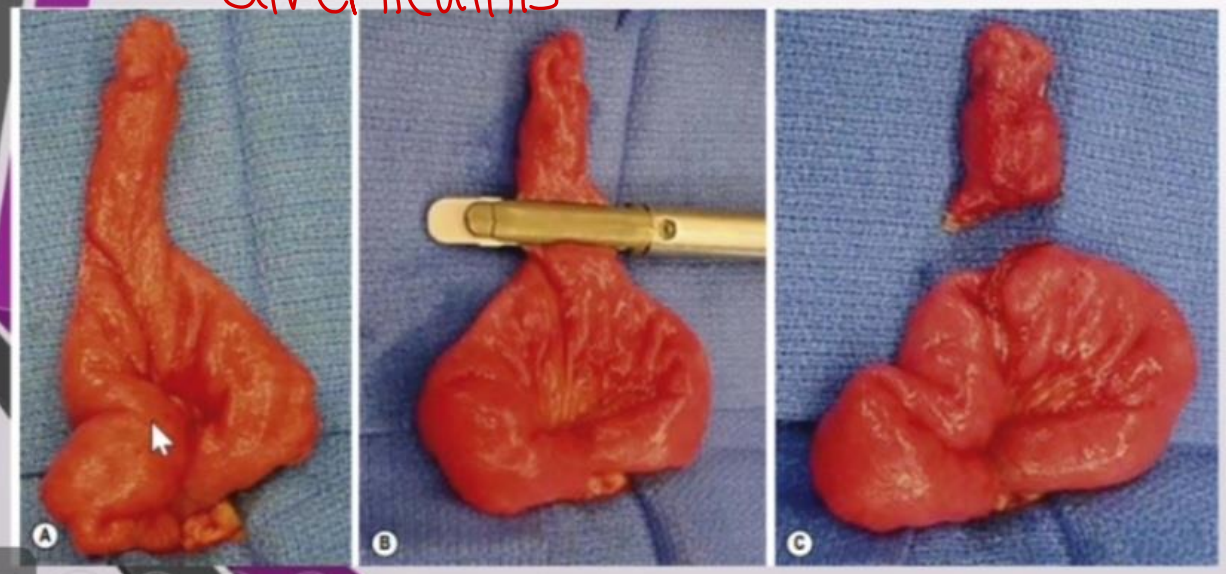
open

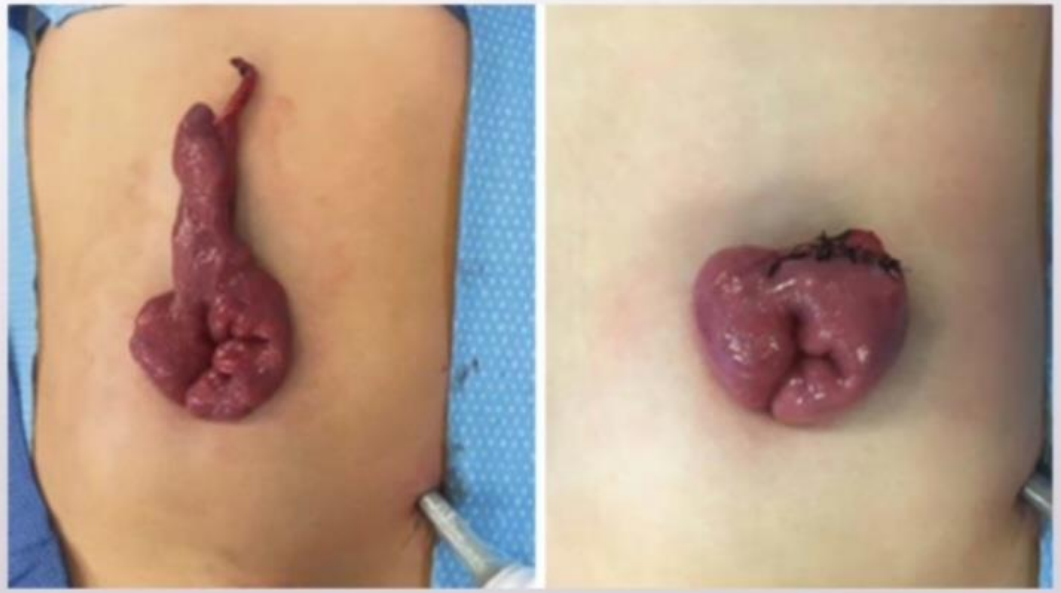
intussusepsion

Management

- Stabilize the patient in case of bleeding
- Surgery : open or laparoscopic diverticulum resection or segmental bowel resection + anastomosis

diverticulitis (open)





5- Biliary atresia

- Biliary atresia (BA) is a relatively rare obstructive condition of the bile ducts causing neonatal jaundice
- It is a sclerosing cholangiopathy that represents the most common cause of end-stage liver disease and the most common indication for liver transplantation in children
- The incidence of BA varies around the world (Europe: 1 in 18,000 live births; France: 1 in 19,500 live births; UK and Ireland: 1 in 16,700 live births; Japan: 1 in 9640 live births
- The highest recorded incidence is in French Polynesia (1 in 3000live births).
- There is a slight female preponderance F > M

- It is an isolated disease of term infants in 85% of cases. In the remainder of affected patients, it occurs as part of a syndrome, the most common of which is BASM (biliary atresia, splenic malformation (asplenia or polysplenia) and malrotation).

unknown etiology these are hypothesis

- The etiology is multifactorial (intrauterine or perinatal viral infection, immunologically mediated inflammation and other autoimmune/ genetic factors, exposure to toxins, abnormal ductal plate remodeling, a vascular or metabolic insult)

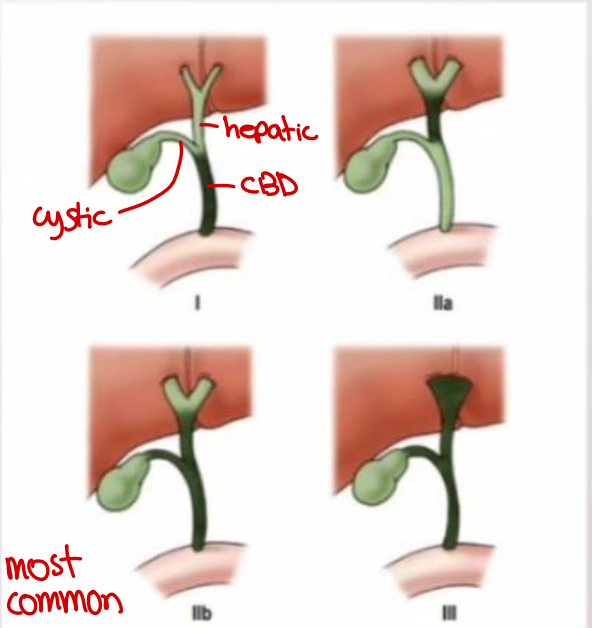
don't memorize
only know it's multifactorial

- BA is classified according to anatomic and cholangiographic findings.

Type I is atresia of the common bile duct

type IIa is atresia of the common hepatic duct, type IIb is atresia of the common bile duct and the common hepatic duct

Type III is atresia of all extrahepatic bile ducts up to the porta hepatis



Presentation

Key is early dx.

- Signs suggestive of BA are jaundice, pale stools, and hepatomegaly.
- Anemia, malnutrition, and growth retardation ensue because of malabsorption of nutrients and fat-soluble vitamins.

any jaundice >2 weeks is not considered physiological especially if direct bilirubin is increased (obstructive jaundice)

Box 43.1 Diagnosing Biliary Atresia

Routine Assessments

Stool color
Consistency of the liver on palpation
Conventional liver function tests plus γ-glutamyl transpeptidase
Coagulation (prothrombin time, activated partial thromboplastin time)
Ultrasonography
Hepatobiliary scintigraphy

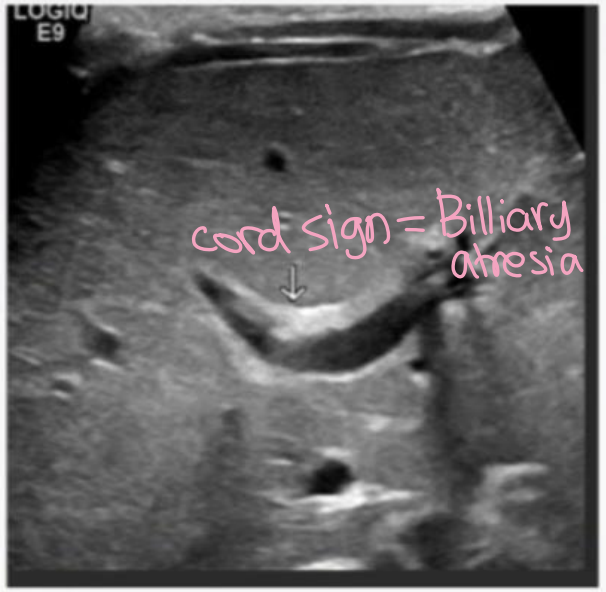
ggt >>>

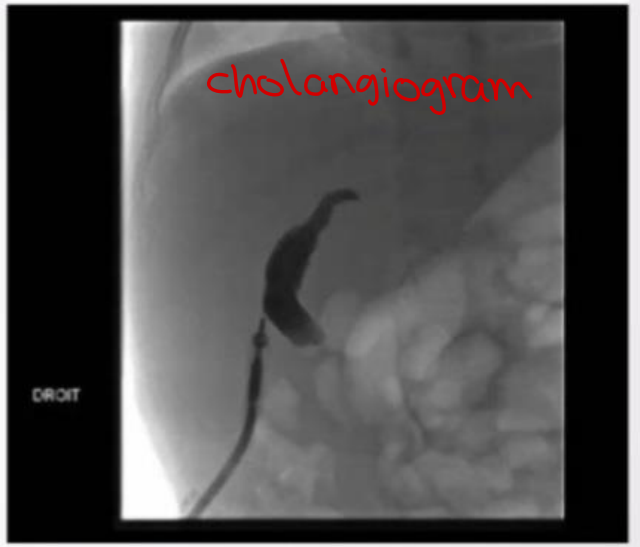
Pt & Ptt >>>

Specific Investigations

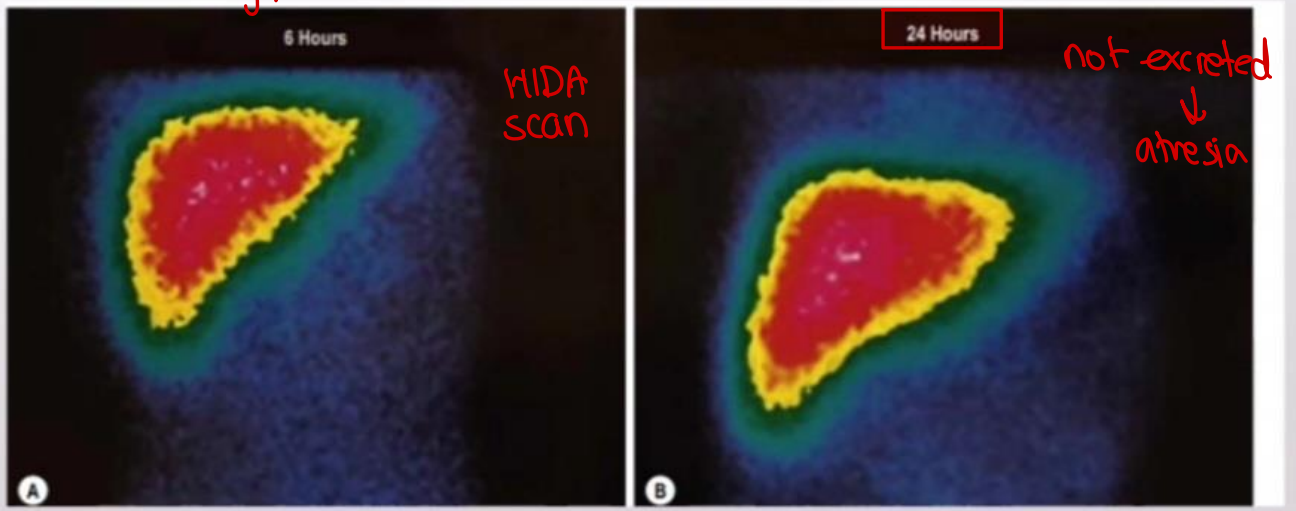
Histobiochemical
Hepatitis A, B, C serology
TORCH titers
α1-Antitrypsin
Serum lipoprotein-X
Serum bile acids
Confirmation of extrahepatic bile duct patency
Duodenal fluid aspiration
Endoscopic retrograde cholangiopancreatography (ERCP) hard for babies
Near-infrared reflectance spectroscopy
Needle biopsy → cirrhosis + proliferation of biliary duct
Direct observation (open or laparoscopic)
Surgical cholangiography

VS





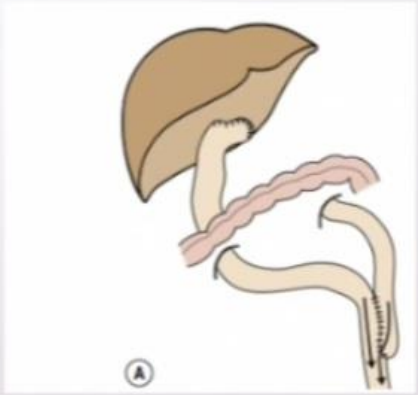
rough liver



cord sign + asplenia + atretic gallbladder → biliary atresia

Surgery

1
- ROUX-EN-Y LIMB AND ENTEROTOMY FOR PORTOENTEROSTOMY (kassi procedure) connect bowel to porta hepatis



2
- Liver tx
- The indications for liver transplantation following portoenterostomy are: (1) lack of bile drainage; (2) signs of developmental retardation or its sequelae; and (3) presence of socially unacceptable complications/side effects.

outcome

- Classically, the major determinants of satisfactory outcome after portoenterostomy are
  (1) **age at** initial operation
  (2) successful achievement of postoperative bile flow
  (3) presence of microscopic ductal structures at the porta hepatis
  (4) the extent of liver parenchymal disease at the time of diagnosis
  (5) technical factors involving the portoenterostomy anastomosis
  (6) CMV status , syndromic or isolated

- Following a successful Kasai operation, pigmented stool is usually seen within 2–3 weeks

- Such success is typically seen in 2/3 of patients, but is maintained into adulthood in only 1/2 of the patients with initial jaundice clearance.
- liver transplantation will be required in 2/3 of patients at some point in their life.

Post op complications

- Cholngitis
- Fat , protein , and mineral malabsrbtion
- Failure to thrive
- Portal hypertension
- HEPATOPULMONARY SYNDROME AND PORTOPULMONARY HYPERTENSION
- INTRAHEPATIC BILE LAKE CYSTS
- HEPATIC MALIGNANCY

NOW THIS IS

Remarkable