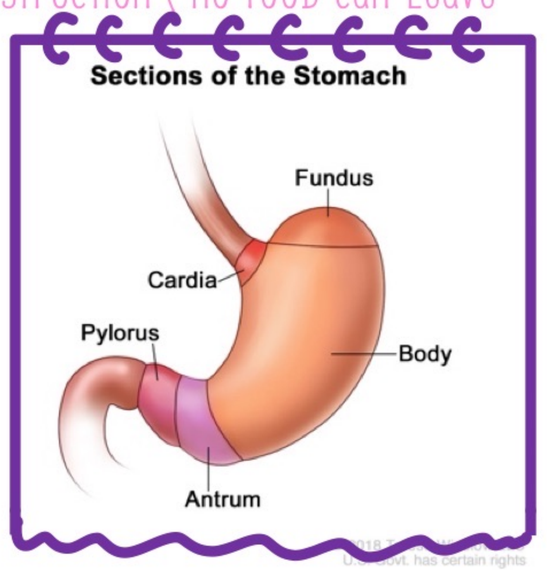
Sereen Draghmeh

Hypertrophic Pyloric Stenosis

Brief Explanation:

From its name we can conclude that there is hypertrophy (increase in size) of the pylorus (the opening between the stomach and the duodenum) , so this causes gastric outlet obstruction (no food can leave the stomach)



Risk Factors:

1. Male > Female : 4:1
2. Family History
3. Younger Maternal Age
4. Being a first born infant
5. Maternal feeding patterns

Etiology:

1. Genetic Factors:
    a. Race discrepancies
    b. Increased frequency in males
    c. First born infants with a positive family Hx
2. Environmental Factors:
    a. Method of feeding (breast vs formula)
        i. Breast feeding decreases the likelihood of developing as it contains protective factors that reduce the risk of inflammation or muscle hypertrophy
    b. Seasonal variability
    c. Exposure to erythromycin
    d. Transpyloric feeding in premature infants
        i. Due to developing irritation or hypertrophy from the transpyloric tube
3. Other Factors: (PONNG)
    a. Excessive Substance P
        i. Substance P is a neuropeptide involved in smooth muscle contraction so increased levels would lead to increased contraction of the pyloric muscle
    b. Decreased neurotrophins
        i. Neurotrophins regulate nerve and muscle development. Deficiencies may lead to abnormal pyloric muscle innervation and hypertrophy.
    c. Deficient nitric oxide synthase
        i. Nitric oxide is involved in the smooth muscle relaxation which includes the relaxation of the pyloric muscle so a deficiency in it can cause impaired relaxation of the pyloric muscle which can promote hypertrophy too
    d. Gastrin hypersecretions
        i. Elevated levels of gastrin will cause increased gastric acid secretion which promotes hypertrophy of the pyloric muscle

Hx and PEx: (Think of it like a story)

Hello Doctor, my baby is **2 weeks old** and has been vomiting whenever I feed him **formula milk**. The **vomit is not green in colour and sometimes is brown in colour** more like **coffee ground in appearance**. His vomiting occurs **within 30-60 mins after I feed him** and the vomiting is **forceful and reaches further away from me.** And even though I feed him, he is **still hungry**.

From this History we can highlight a few key points that help us think of Hypertrophic Pyloric Stenosis as a DDx:

1. 2 weeks
    a. Usually it presents between 2-8 weeks
2. Formula fed:
    a. As we said above being formulae fed is a risk factor
3. **Vomit is not green in colour + coffee ground:**
    a. That means its not bilious, so you have to think of everything that can be affected before the ampulla of vater (which is where the bile is secreted)
4. **Vomiting occurs within 30-60 mins after eating:**
    a. keep in mind that if it was immediate then you should think of esophageal atresia
5. **Vomiting is forceful and reaches far away:**
    a. indicates that it's projectile vomiting
6. Still hungry

After the patient comes and give you their Hx you start PEx: here are the things that you would see:

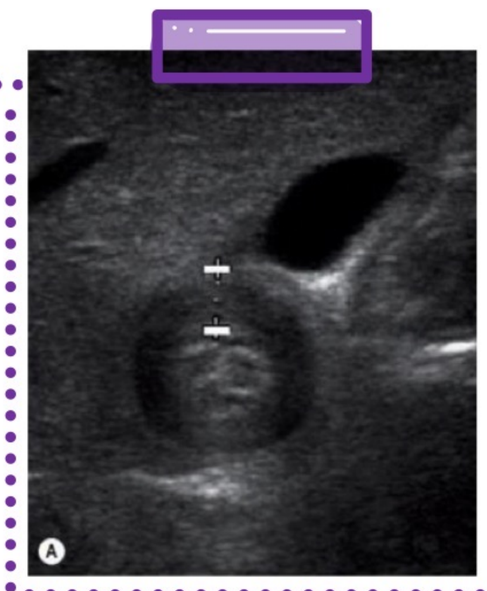
1. First of all you will notice that the patient is **very thin (weight loss)** especially if this has been going on for a long period of time
2. You can also notice that the patient is **dehydrated**:
    a. you can check that by checking urine output or assessing the skin turgor (they would have poor skin turgor)
3. You may also see **visible peristaltic waves**
4. You can also **palpate a mass, this is called the olive sign**
    a. you are palpating the pylorus- remember there is hypertrophy here so logically speaking you may be able to feel it
    b. present in 70-90% of pts

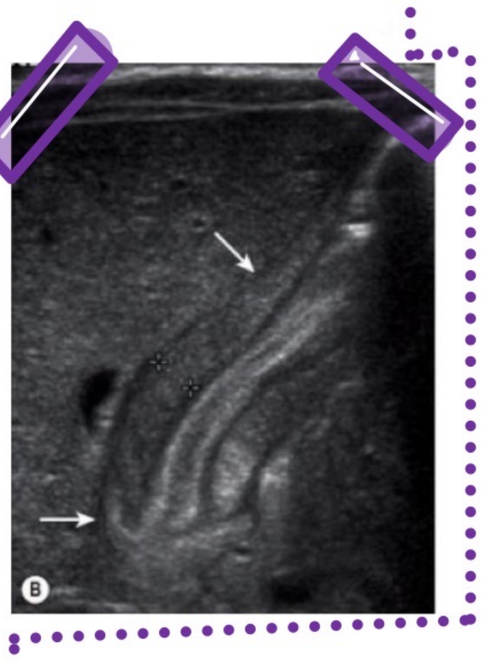
Investigations:

1. Labs: **Hypochloremic, Hypokalemic, Metabolic Alkalosis**
   a. Chloride: Hypochloremic
   b. Potassium: Hypokalemic
   c. ABGs : Metabolic Alkalosis
      i. pH> 7.45
      ii. HCO3- > 28 mEq/L – hallmark of metabolic Alkalosis

Explanation: Persistent vomiting leads to the loss of gastric contents, which are rich in hydrochloric acid (HCl). When the H+ is lost it will cause the blood to become more alkaline, increasing the bicarbonate levels causing metabolic alkalosis , and when the Cl- is lost it will lead to hypochloremic.To compensate for low hydrogen ion levels in the blood (from vomiting), hydrogen ions move out of cells into the bloodstream. To maintain electrical neutrality, potassium (K^+) shifts into cells, further lowering serum potassium levels.

2. Imaging:
   a. **Ultrasound**
      i. **Muscle thickness of >= 4 mm**
         - Picture A shows that there is an increase in the thickness of the muscle
      ii. **Pyloric length >= 16 mm**
         - Picture B shows that there is increase in pyloric length
      iii. **Donut Sign**
         - Picture A shows a donut shape





Management

1. Preoperative:
   a. NPO
   b. NG tube
      i. For gastric decompression
   c. IV fluid Resuscitation
   d. Correction of Electrolytes
2. Surgery:
   a. Correction by **Laparoscopic/Open Pyloromyotomy**
      i. Operation involves cutting through the hypertrophied muscle of the pylorus while leaving the inner mucosal lining intact.

Complications:

1. Mucosal perforation
2. Postoperative emesis – occurs in most infants
3. Prolonged postoperative emesis – due to GERD or incomplete myotomy

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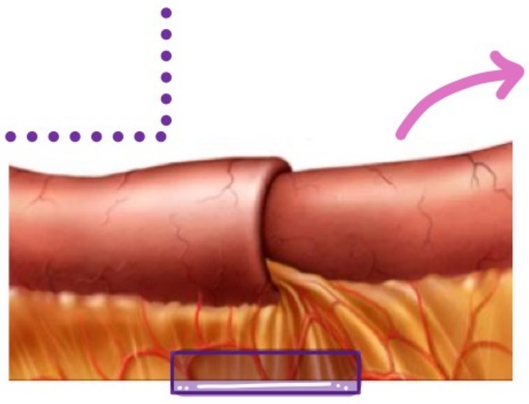
Intussusception

Brief Explanation

Intussuspection is when a part of the intestine slides into another part of the intestine; think of it like a telescope how the parts enter each other

Definition:

Its 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 which leads to arteriual insufficiency, ischemia and bowel wall necrosis



So basically what happens is that when the proximal intestine enters the distal intestine it will pull along with it its mesentery, this mesentery contains the blood vessels that supplies that part of the intestine so pulling it inside the distal intestine will compress it leading to venous obstruction first (venous obstruction is first cause as we know the veins have a thinner wall compared to arteries so they are easier to compress) . This venous compression will lead to edema in the bowel wall. This compression and edema will lead to arterial insufficiency which will cut the blood supply to the intestine leading to ischemia then bowel wall necrosis

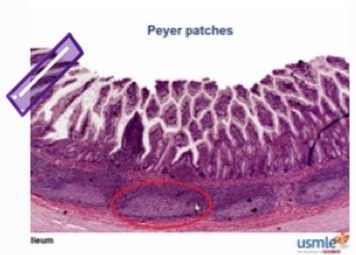
So summary:

[Intussusception → Mesentery compressed → venous congestion → edema → arterial insufficiency → ischemia → necrosis] important !!

Types:

MOST COMMON LOCATION IS ILEOCOLIC

1. **Primary – Idiopathic**
   a. There is no leading point
   b. Likely due to hypertrophied Peyer patches within the bowel wall



i. Peyer's patches are small masses of lymphoid tissue located in the lining of the small intestine. In some infants, these patches can become hyperplastic (enlarged) in response to minor infections or inflammation, which increases their bulk. This hypertrophy is thought to be a primary factor leading to the intussusception because it facilitates the telescoping of one segment of the bowel into another.

   c. Age: 4-9 months
   d. 2/3 are boys
2. **Secondary**
   a. Reasons:
      i. Meckel's diverticulum
      ii. Polyps and Duplications
      iii. Appendix
      iv. Hemangiomas
      v. Carcinoid tumors
      vi. Foreign bodies
      vii. Ectopic Pancreas/ Gastric mucosa
      viii. Hamartomas from Peutz Jeghers syndromes
      ix. Lipomas and Lymphomas
      x. Small bowel tumors
      xi. HenochSchonelein purpura
      xii. Cystic Fibrosis
      xiii. Celiac Disease
      xiv. Clostridium Difficile colitis

Hx and PEx: (Think of it like a story)

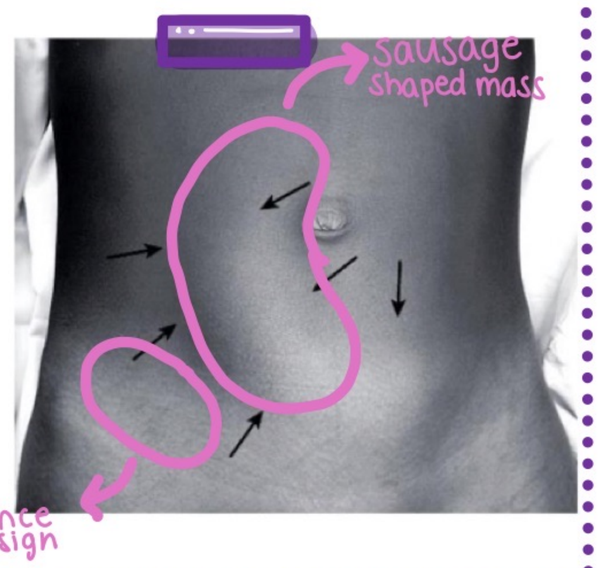
History:

Hello doctor my son is **5 months** and suddenly has been having **cramping like abdominal pain** every **15-30 mins** which is causing him to cry and **pull his legs to his abdomen** whenever the attack occurs. His pain has also been associated with **vomiting** that was not green in colour in the beginning but then became green in colour. He also passes stool that like **jelly in consistency and red in colour**

Physical Examination:

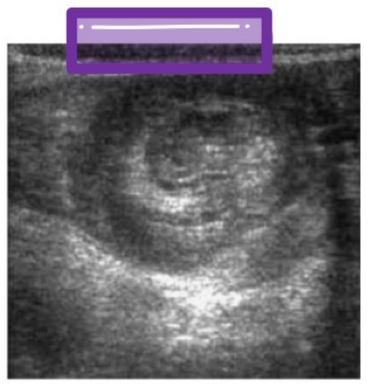
1. Signs of **dehydration** (you can see that by seeing his urine output or having poor skin turgor)
2. A late sign would be **hypotension, fever and tachycardia**
3. RUQ mass can be palpated – **Sausage shaped or curved mass**
4. RLQ would be empty – **Dance sign**
    a. empty since if we are talking about the most common type which is ileocolic, the terminal ileum would be inside the colon so that part would be empty
5. **Audible Peristaltic rushes** (early sign)
6. On PR examination there would be **blood stained mucus or blood** (late sign)
7. You may also see **prolapsing of the intussusceptum through the anus** (late sign)

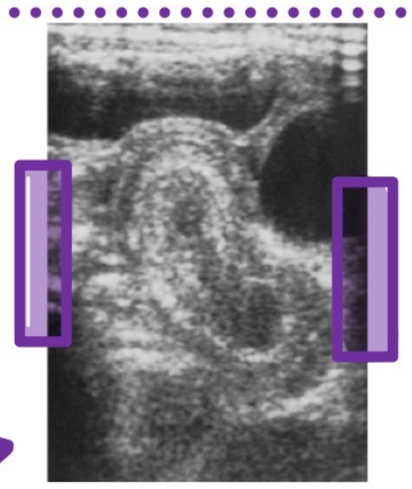
- Do a testicular examination to rule out testicular torsion



Investigations:

1. Imaging:
    a. Ultrasound:
        i. **Target/ donut lesions** – in transverse plane
        ii. **Pseudo-kidney sign** – in longitudinal plane
    b. X ray:
        i. often normal early in the disease, but later they may show small bowel obstruction, perforation, or a soft tissue mass.





Management:

1. **Initial**:
    a. NG tube
    b. NPO
    c. IV fluid Resuscitation and maintenance of IVF
    d. Correct electrolyte disturbances
2. **Non-Operative Management**
    a. **Hydrostatic/ pneumatic reaction:**
        i. Its basically using a fluid or air to try and separate the bowels
        ii. Usually under fluoroscopy or ultrasound guidance
        iii. Contraindications:
            1. Perforation
            2. Peritonitis
            3. Persistent hypotension/ tachycardia
            4. Sepsis
3. **Operative management (laparoscopic or open)**
    a. Indications:
        i. Nonoperative reduction is unsuccessful or incomplete
        ii. Signs of peritonitis/pneumoperitoneum
        iii. Presence of a lead point (secondary intussusception)

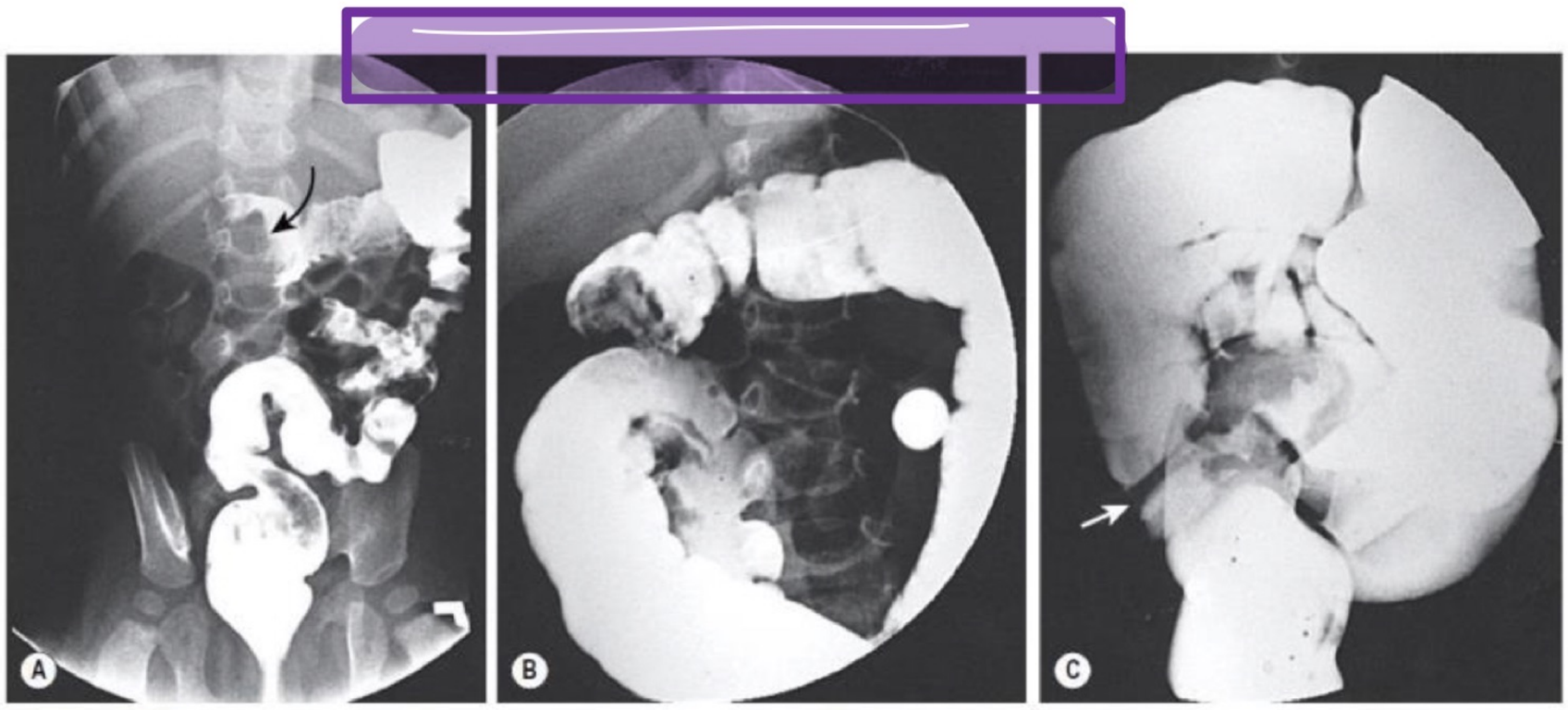


FIGURE 38-7 ■ Fluoroscopic examination using isotonic contrast for hydrostatic reduction of intussusception. **(A)** Intussusception (arrow) seen in midtransverse colon. **(B)** Reduction has occurred to the hepatic flexure. **(C)** Complete reduction with reflux of contrast medium into the terminal ileum. Note the edematous ileocecal valve (arrow).

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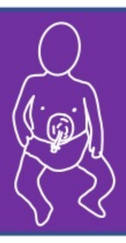


Omphalocele and Gastroschisis

GASTROSCHISIS →

Brief Explanation

ITS CONGENITAL DEFECT OF THE ABDOMINAL WALL WHERE THE INTESTINES AND SOMETIMES OTHER ABDOMINAL ORGANS HERNIATE THROUGH AN OPENING

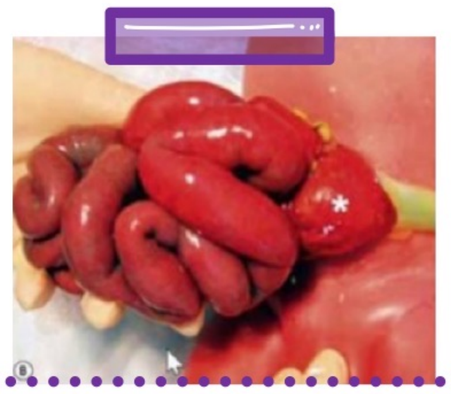
Epidemiology:

1. 1 in 4000 liver births
2. Higher incidence in mothers younger than 21 years of age
3. Delivery should be in a tertiary center

Pathology and associated features

a. Gastroschises is associated with a vibrable degree of inflammatory thickening of the visceral bowel walls, which result in the **characteristic appearance of matted intestines**

b. Associated with
   a. intestinal motility disorders
   b. rotational diseases
   c. UDT 15
   d. bowel atresia



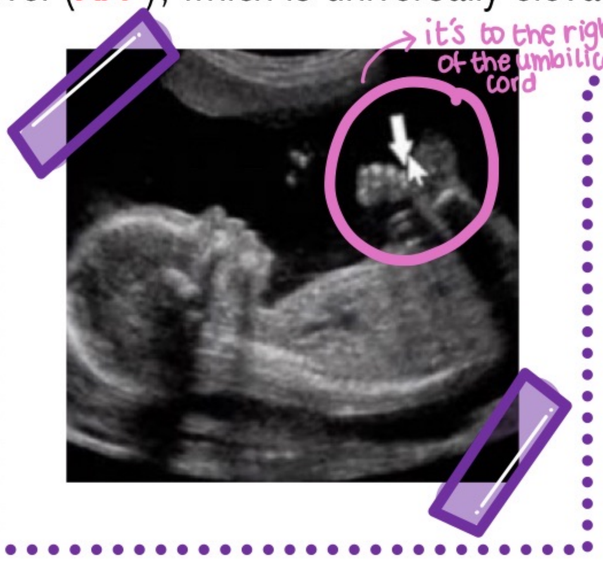
Classification:

a. **Simple:**
   a. Intact bowel without other complications.

b. **Complicated:**
   a. Includes atresia, short bowel syndrome, or ischemia.

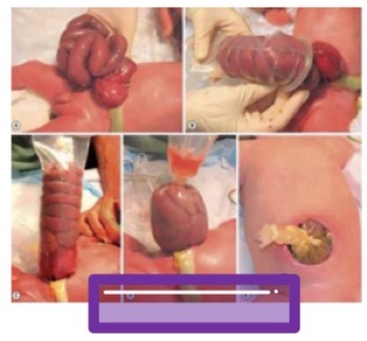
Diagnosis:

1. **AN US by 20 weeks gestation**
   a. **Bowel loops freely floating** in the amniotic fluid
   b. defect in the **abdominal wall to the right of a normal umbilical cord**
   c. abnormal maternal serum alpha fetoprotein level (**AFP**), which is universally elevated +/- intrauterine growth restriction (**IUGR**)



Management:

1. Resuscitation:
    a. NPO
    b. NG
    c. IVF
    d. Rectal tube to decompress
2. Bowel should also be wrapped in warm **saline soaked gauze** and placed in a **central position** on the abdominal wall
3. Surgery:
    a. **primary closure**
        I. Normally used when only a small part of the bowel is obstructed
        II. Here I just try and put the bowel back into the abdomen and close it
    b. **staged closure (with silo)**
        I. Here I put the bowel in a silo bag and then I place it perpendicularly and let gravity run its course, in which the bowel will slowly enter back into the abdomen (this normally take 7 days) ; then I remove the silo bag and close the abdomen wall



Long term outcome:

a. Generally they are excellent
b. Morbidities are related to prematurity, bowel motility and length

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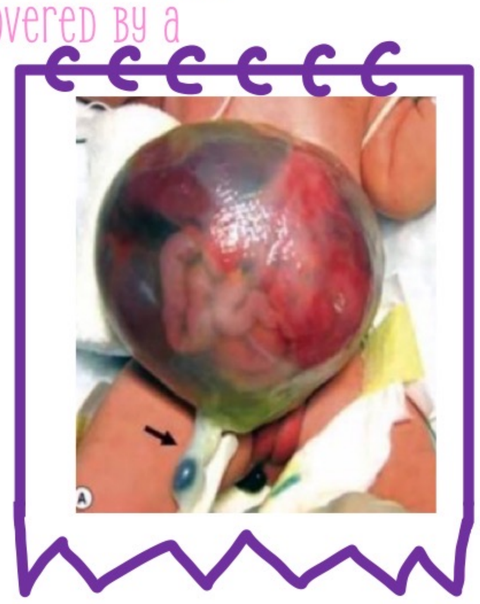
Omphalocele →

Brief Explanation

Omphalocele is a congenital defect in the abdominal wall where abdominal organs (intestines, liver, or other viscera) herniate through the base of the umbilical cord. Unlike gastroschisis, the organs are covered by a protective membrane consisting of peritoneum and amnion.



Epidemiology and Associations:

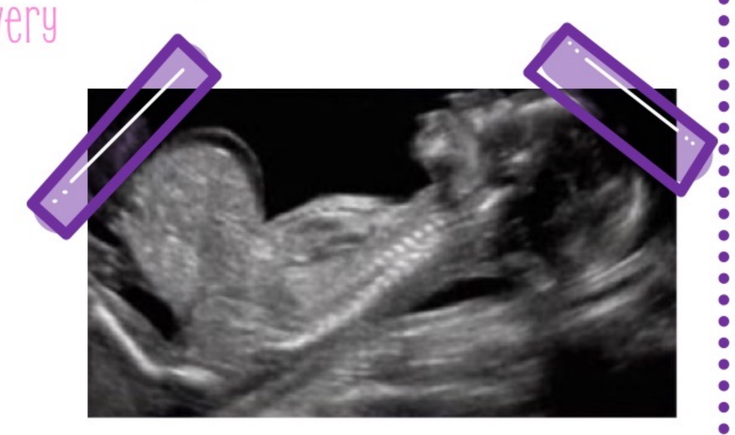
1. 1 in 4000-6000
2. Associated with genetic defect and other anomalies
    1. Trisomy's 13.18,21 and 45X
    2. Beckwith-Weideman
    3. Pentalogy of Cantrell
    4. Cardiac (14-47% incidence of anomalies)
    5. Central Nervous (3-33% anomalies)
3. Outcomes depends on associated anomalies
4. Delivery in tertiary center at term, normal vaginal delivery (except if it is a giant omphalocele and containing the liver – to avoid shoulder dystocia, sac rupture and bleeding)

Long term Morbidities

1. GERD
2. Pulmonary insufficiency
3. Recurrent lung infections or asthma
4. Feeding difficulty with failure to thrive

Diagnosis:

1. **Antenatally US – 18 week evaluation**
    a. Prognostic factors:
        i. Omphalocele diameter compared with abdominal circumference (**O/AC**)
            1. To know if the sac will return back to the abdomen after birth
        ii. Omphalocele diameter compared with the femur length (**O/FL**)
        iii. Omphalocele diameter compared with the head circumference (**O/HC**)
            1. To see if you are able to do a vaginal delivery
        iv. Organ contained inside the sac



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Management:

1. Resuscitation:
    a. NPO
    b. NG
    c. IVF
    d. Rectal tube to decompress
2. Sac should also be wrapped in warm **saline soaked gauze** and placed in a **central position** on the abdominal wall
3. Surgery:
    a. **primary closure**
        I. Normally used when only a small part of the bowel is obstructed
        II. Here I just try and put the bowel back into the abdomen and close it
    b. **staged closure (with silo)**
        I. Here I put the bowel in a silo bag and then I place it perpendicularly and let gravity run its course, in which the bowel will slowly enter back into the abdomen (this normally take 7 days) ; then I remove the silo bag and close the abdomen wall

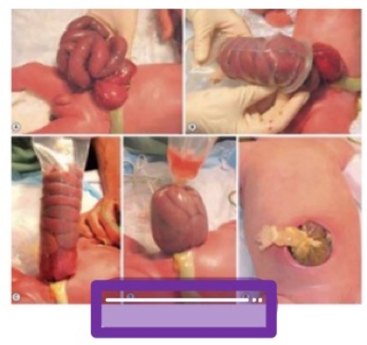

Table 48.1 Differentiating Characteristics Between Gastroschisis and Omphalocele

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

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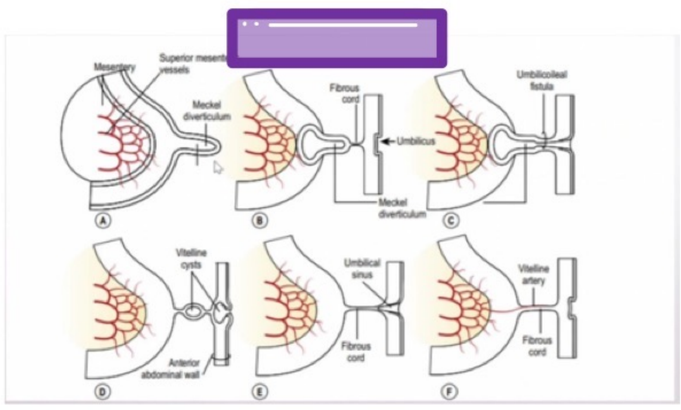
Meckel's Diverticulum

Brief Explanation

Meckel's diverticulum is a congenital outpouching of the small intestine caused by incomplete closure of the vitelline duct during fetal development

Incidence and Epidemiology:

1. True incidence of Meckel's diverticulum is unknown because most patients are asymptomatic
2. Estimated to affect the population at approx. 2 % of the population
3. 4 % of cases will become symptomatic
4. M:F of 2:1



Rule of 2s:

1. Occurs in **2% of the population**
2. **2:1 male to female ratio**
3. Discovered by **2 years of age**
4. Located **2 feet** (60 cm) **from the ileocecal valve** , commonly 2 cm in diameter and 2 inches (5 cm) long
5. Contains **2 types of heterotopic mucosa** (gastric is the most common followed by pancreatic)

Clinical Presentation:

1. The three most common presentation in children are
   a. **Intestinal bleeding (30-56%)**
      i. Since the most common type of heterotopic mucosa is gastric , it will produce acid there that will cause ulceration causing bleeding
   b. **Intestinal obstruction (14-42%)**
      i. Either by:
         1. Acting as a leading point causing intussusception
         2. Causing volvulus if there is a cord which will cause the bowel to twist around bowel
         3. Cause internal hernia
   c. **Diverticular inflammation (6-14%)**
      i. Mimicking appendicitis
2. Less common signs include
   a. cystic abdominal mass
   b. newborn with an umbilical fistula resulting from a patent vitelline duct
   c. in elderly, neoplasia can develop within the Meckel diverticulum (carcinoid is the most common tumor)

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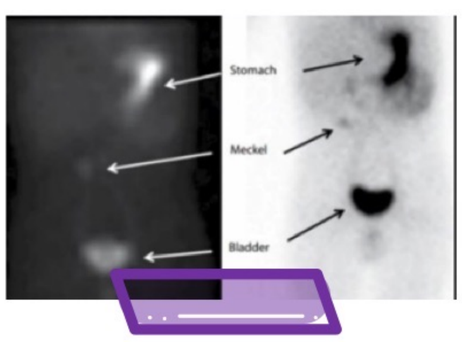
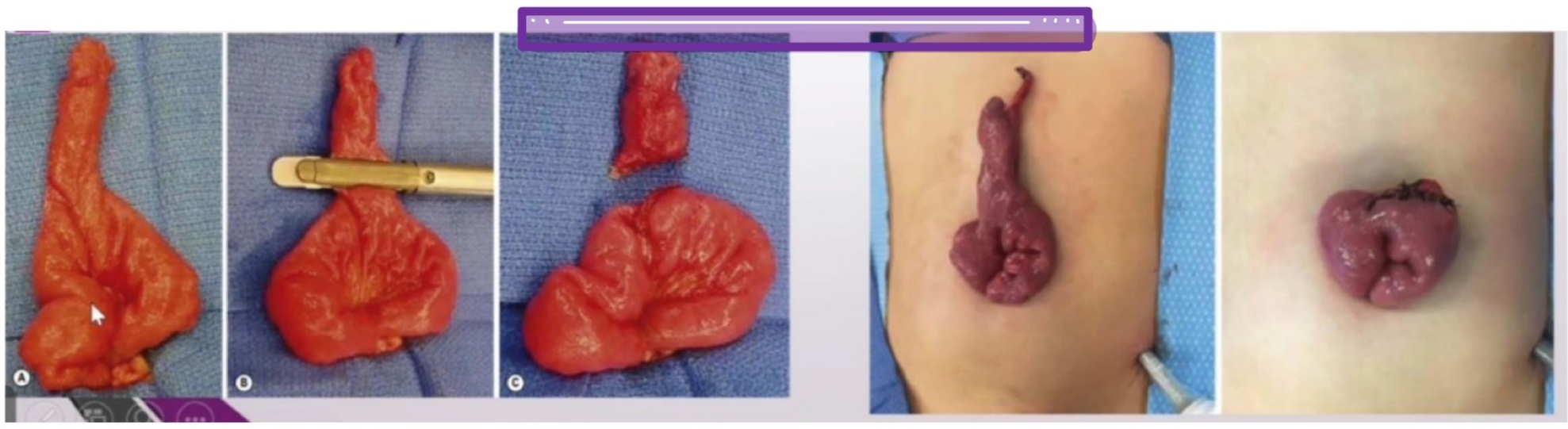
Diagnosis:

1. In patients presenting with obstruction or inflammation, the diagnosis of a Meckel diverticulum is not usually definitively determined preoperatively
2. Imaging
    a. **US and CT might be helpful**
    b. In case of bleeding diverticulum, **technetium – 99m pertechnetate radionuclide study** (Meckel scan)- false negative occur in up to 25%
        i. Technetium - 99m has a high affinity to gastric mucosa so anywhere that has gastric mucosa will absorb the technetium - 99m
        ii. The reason why 25% are false negatives is because not all Meckel's diverticulum contain gastric heterotopic mucosa, some are pancreatic



Management

1. Stabilize the patient in case of bleeding
2. Surgery:
    a. open or laparoscopic diverticulum resection or segmental bowel resection + anastomosis



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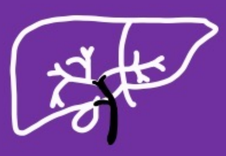
Biliary Atresia



Brief Explanation

Biliary atresia is a rare condition in newborns where the bile ducts, which carry bile from the liver to the intestine, are blocked. This leads to bile build-up in the liver, causing damage, jaundice, and eventually liver failure if untreated.

Definition:

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**

Epidemiology:

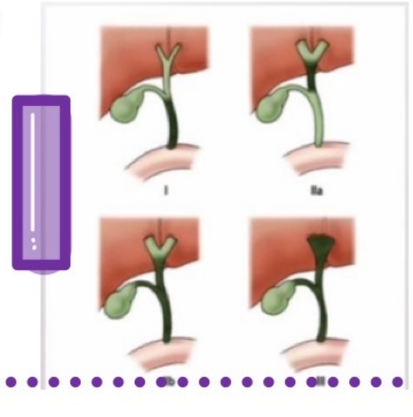
1. The incidence of BA varies around the world (Europe : 1 in 18000 live births; France: 1 in 19500 live births; UK and Ireland: 1 in 16700 live births; Japan: 1 in 9640 live births)
2. The highest recorded incidence is in French Polynesia (1 in 3000 live births)
3. There is a slight female predominance
4. Isolated vs Syndromic:
    a. Its an isolated diseases of term infants in 85% of cases.
        i. In 85% of cases, biliary atresia occurs on its own, without being part of a larger set of abnormalities.
    b. In the remainders 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)

Etiology:

1. The etiology is multifactorial:
    a. intrauterine or perinatal viral infection
    b. immunologically mediated inflammation
    c. autoimmune/genetic factors
    d. exposure to toxins
    e. abnormal ductal plate remodeling
    f. a vascular or metabolic insult

Classification:

1. BA is classified according to anatomic and cholangiography findings
    a. Type I is atresia of the common bile duct
    b. Type IIa is atresia of the common hepatic duct
    c. Type IIb is atresia of the common bile duct and the common hepatic duct and cystic duct
    d. Type III is atresia of all extrahepatic bile ducts up to the porta hepatis



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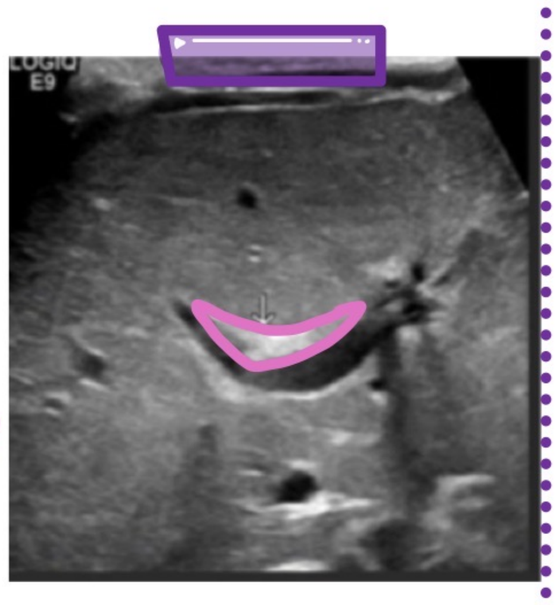
Clinical Presentation:

1. Signs suggestive of BA
    a. Jaundice
    b. Pale stools
    c. Hepatomegaly
2. Consequences:
    a. Anemia
    b. Malnutrition
    c. growth retardation ensue because of malabsorption of nutrients and fat- soluble vitamins

Diagnosis:

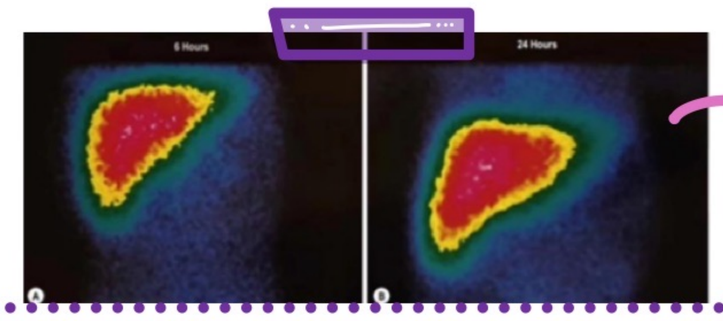
1. Routine Assessments
    a. Stool colour
        i. Pale stools suggest impaired flow
    b. Consistency of the liver on palpation
        i. A firm, enlarged liver can indicate liver disease.
    c. Conventional liver function tests plus gamma-glutamyl transpeptidase
        i. Elevated in bile duct obstruction.
    d. Coagulation (Prothrombin time, activated partial thromboplastin time)
    e. Ultrasonography
        i. Pathognomonic sign is the triangular sign
    f. Hepatobiliary scintigraphy
        i. A nuclear scan to assess bile flow. Failure of bile to reach the intestine suggests obstruction.



2. Specific investigations:
    a. Histobiochemical
        i. To evaluate liver cell damage and function.
    b. Hepatitis A,B,C serology
    c. TORCH titers
    d. Alpha 1 antitrypsin
        i. To rule out deficiencies causing liver disease
    e. Serum lipoprotein X
        i. Indicator of cholestasis
    f. Serum bile acids
        i. Indicator of cholestasis
    g. Confirmation of extrahepatic bile duct patency
    h. Duodenal fluid aspiration
        i. Detects bile in the duodenum
    i. ERCP
        i. Visualizes bile ducts
    j. Near infrared reflectance spectroscopy
        i. Advanced imaging for bile structures
    k. Needle biopsy
        i. Examines liver tissue for fibrosis or bile duct abnormalities.
    l. Direct observation (open or laparoscopically)
    m. Surgical cholangiography

Sereen Draghmeh

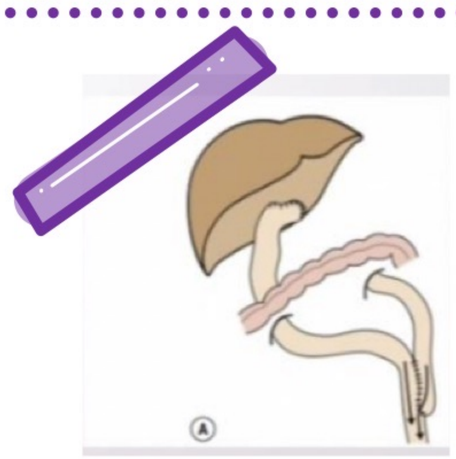
n. HIDA scan
   I. The test involves injecting a radioactive tracer into the bloodstream, which is taken up by the liver and excreted into the bile ducts. Imaging tracks the flow of bile from the liver into the intestines.
   II. In biliary atresia the tracer accumulates in the liver but does not reach the intestine, suggesting bile duct obstruction.



Shows bile obstruction since the liver still contains the tracer

Surgery:

1. Roux-En-Y and enterotomy for portoenterostomy (kasai procedure)
2. **Liver transplantation**
   a. Indications for it following portoenterostomy:
      i. Lack of bile drainage
      ii. Signs of developmental retardation or its sequelae
      iii. Presence of socially unacceptable complications/side effect



Outcome:

1. Classically the major determinants of satisfactory outcome after portoenterostomy are:
   a. **Age at initial operation**
      i. **MOST IMPORTANT FACTOR**
   b. Successful achievement of postoperative bile flow
   c. Presence of microscopic ductal structures at the porta hepatis
   d. The extent of liver parenchymal disease at the time of diagnosis
   e. Technical factors involving the portoenterostomy anastomosis
   f. CMV status, syndromic or isolated
2. Postoperative milestones:
   a. Following a successful kasai operation, pigmented stool is usually seen within 2-3 weeks
   b. Such success is typically seen in 2/3 of patients, but is maintained into adulthood in only ½ of the patients with initial jaundice clearance
3. Liver transplantation will be required in 2/3 of patients at some point in life

Post-Op Complication:

1. Cholangitis
2. Fat, protein, and mineral absorption
3. Failure to thrive
4. Portal hypertension
5. Hepatopulmonary syndrome and Porto-pulmonary hypertension
6. Intrahepatic bile lake cysts
7. Hepatic malignancy