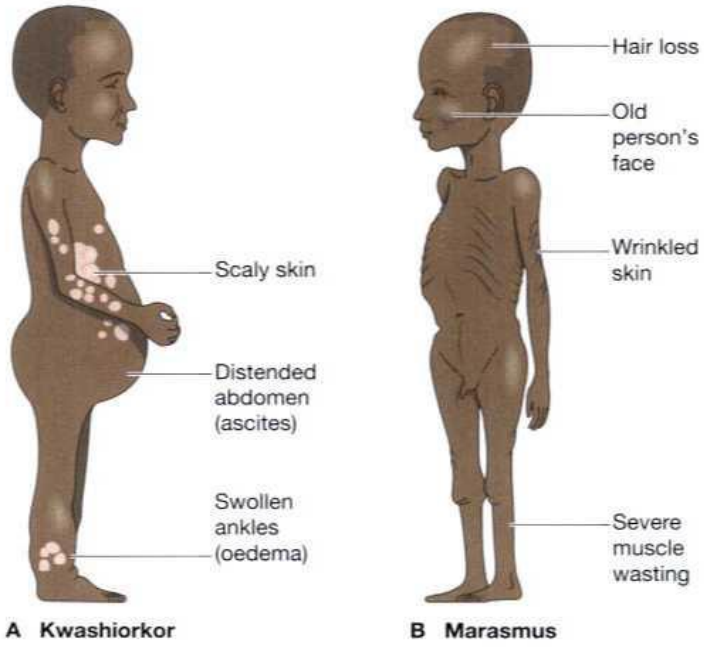
- **Nutrition:**

Essential nutrients	Non-essential nutrients
Are those which can only be obtained from the diet	Can be synthesized by the body (from other compounds) or obtained from the diet

Macronutrients	Micronutrients
Carbohydrates: which are converted in the body to glucose and other monosaccarides that can be digested and absorbed	**Water-soluble vitamins:** • Vitamin C • B-complex: thiamine, riboflavin, niacin, pantothenic acid, pyridoxine, biotin, folic acid, cobalamin
Proteins: which are converted in the body to oligopeptides (2-20 amino acids) or amino acids by pepsinogen and pancreatic proteases. Infants need more protein in their diet (due to faster growth rate)	**Fat-soluble vitamins**: A, D, K and E
Fats: which are converted by the body to fatty acids and glycerol	**Essential trace minerals**: iron, zinc, copper and chromium
Minerals: sodium, potassium, magnesium, chloride, calcium, phosphorus	

Selected vitamin and mineral deficiencies	
Nutrient	**Signs and symptoms**
Vitamin A	Night blindness; xerophthalmia
Vitamin D	Rickets (children); osteomalacia (adults)
Vitamin E	Anemia/ hemolysis
Vitamin K	Coagulopathy/ prolonged PT
Vitamin B1 (thiamine)	Beriberi
Vitamin B3 (niacin)	Pellagra (diarrhea, dementia and dermatitis)
Vitamin B9 (folic acid)	Megaloblastic anemia
Vitamin B12 (cobalamin)	Megaloblastic anemia; sub-acute combined degeneration of the spinal cord
Vitamin C	Scurvy

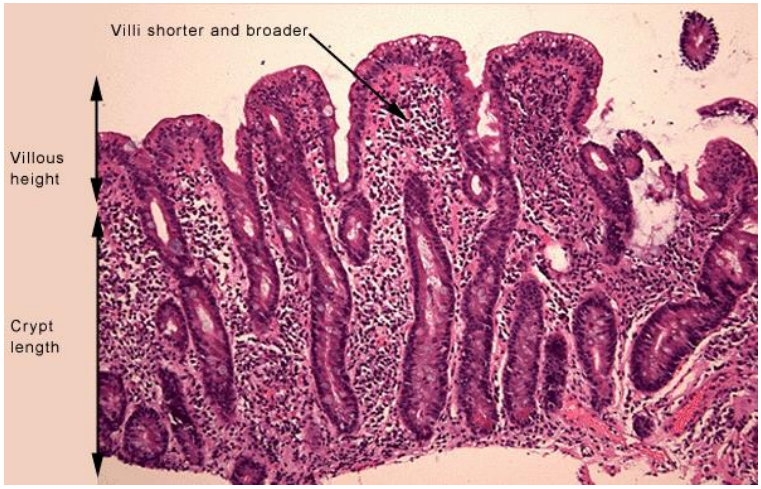
Marasmus	Kwashiorkor
• More common • Resulting from protein and non-protein deficiencies • Features: muscle wasting, loss of body fat, wrinkled skin, prominent ribs	• Less common • In regions where starch is the major element in diet • Resulting from protein deficiency • Features: generalized edema, abdominal distention, thin sparse hair and change in skin pigmentation



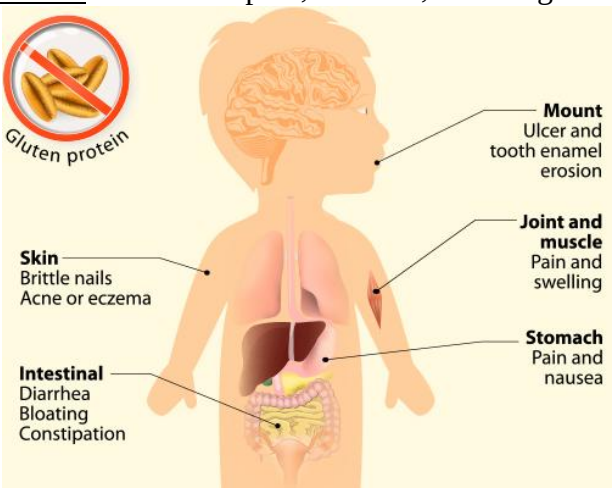
- **Malabsorption**:
  - **Characterized by the triad of**: diarrhea, abdominal distention and impaired growth.
  - **Etiologies:**

Carbohydrates	• **Undigested sugars are**: osmotically active (dragging water into intestinal lumen thus causing diarrhea); fermented by colonic bacteria (thus producing hydrogen, acids and carbon dioxide). An example is lactose intolerance which can be diagnosed by hydrogen breath test or acidic stool pH in the presence of reducing substances. • **Malabsorption results from**: enzyme deficiency or mucosal atrophy
Proteins	• **Malabsorption results from**: congenital enterokinase deficiency; protein-losing enteropathies or inflammatory disorders (e.g. Crohn's disease). • **Fecal α_1-antitrypsin level**: measuring enteric protein losses.
Lipids	• Absorption of fats requires pancreatic lipase and bile salts which form micelles. • **Decreased lipase activity results in**: steatorrhea (foul-smelling greasy stool) and fat-soluble vitamins deficiency. • **Malabsorption results from**: exocrine pancreatic insufficiency (such as in cystic fibrosis and chronic pancreatitis) or bile acid deficiency.

  - **Protein intolerance:**
    - ✓ This occurs in 8% of children due to cow's milk.
    - ✓ Clinical features: abdominal pain, diarrhea, vomiting. In addition, chronic blood loss in the stool might result in anemia.
    - ✓ Investigation: CBC (to rule-out anemia or infections), Electrolytes (to check for disturbances due to dehydration that is resulting from vomiting and diarrhea) and stool (for occult blood and culture). Notice that diagnosis can be made when symptoms resolve as you withdraw the suspected antigen.
    - ✓ Management: hydration, withdrawal of cow's milk.
  - **Celiac disease:**
    - ✓ It is an autoimmune disease which is characterized by gluten intolerance and mucosal damage (mucosal atrophy, crypts elongation and infiltration by lymphocytes).

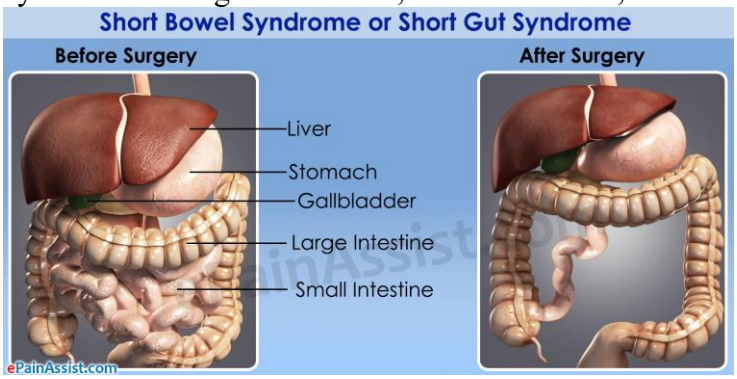


- ✓ It appears between 6 months-2 years when wheat or oats are introduced into the diet of the infant.
- ✓ Clinical features: abdominal pain, diarrhea, vomiting and bloating.



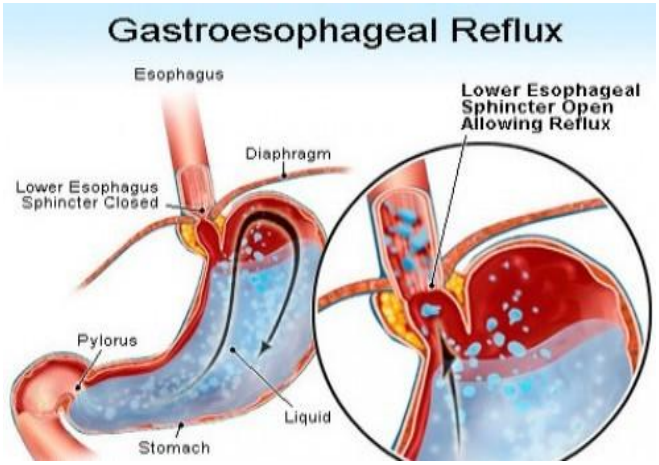
- ✓ Investigations (start with less invasive procedures):
  - ❖ Clinical response when gluten is removed from diet.
  - ❖ Serum IgA-endomysial or serum tissue transglutaminase antibody testing.
  - ❖ But small bowel biopsy remains the gold standard.
- ✓ Management: gluten-free diet for life!

- **Short bowel syndrome (short small intestine):**
  - ✓ Causes: congenital gut lesions (gastroschisis, volvulus or intestinal atresia) all requiring surgical resection which results in smaller surface area for absorption; surgery for necrotizing enterocolitis; Crohn's disease; tumors or radiation.



  - ✓ Clinical features: malabsorption of carbohydrates and fats is common (malabsorption of vitamin B12 and bile acids occurs when ileum is removed); diarrhea and failure to thrive.
  - ✓ Management: Total Parenteral Nutrition (TPN). Small bowel transplantation is reserved for those who have complication of TPN especially liver disease (Notice that liver transplantation might also be required in this condition!).

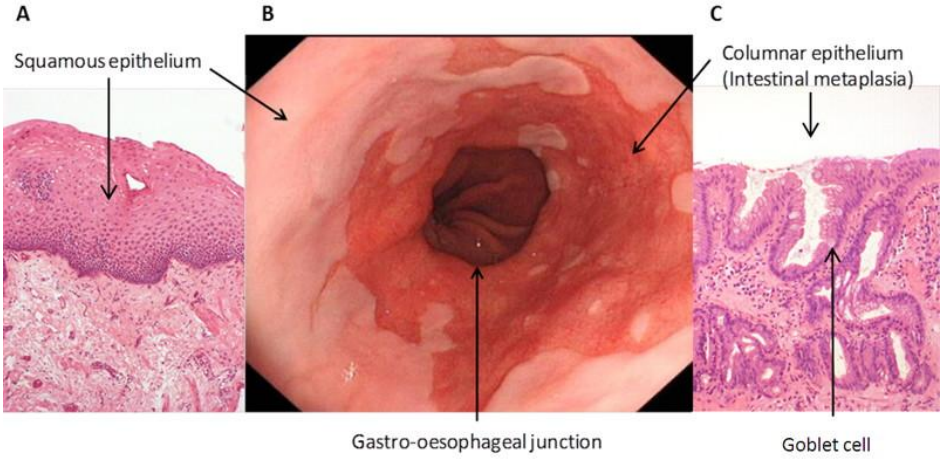
- **Gastroesophageal Reflux Disease (GERD):**
  - It is a pathologic condition in which there is transient relaxation of Lower Esophageal Sphincter (LES) resulting in retrograde of gastric contents into the esophagus and subsequent inflammation of the mucosa.



  - **Clinical features:**

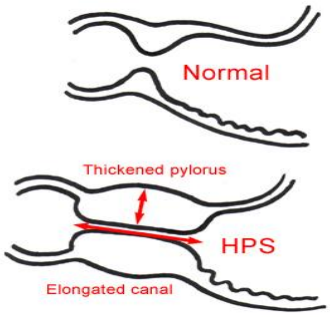
Infants	Older children
• Emesis (most common presentation) with Sandifer syndrome (turning head to one side with arching of the back due to painful esophagitis). • Infant might refuse to feed (due to pain) or have constant hunger (desiring the buffering action of the milk)	• Mid-epigastric pain (heart burn) which is relieved by food or antacids and aggravated by fatty foods, caffeine and supine position. • They might also have nausea on awakening and halitosis.

  - **Complications**: barrett's esophagus (conversion of the normal stratified squamous non-keratinized epithelium of the esophagus to columnar epithelium which predispose to esophageal adenocarcinoma).

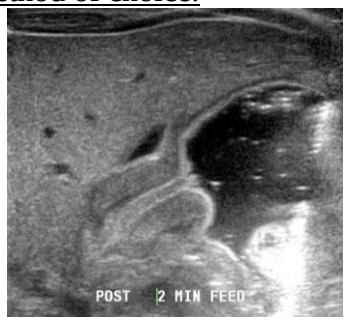


  - **Investigations**: the gold standard is pH probe measurement. Endoscopy only done when diagnosis is uncertain. bronchoscopy is done when aspiration is suspected.
  - **Management**: sitting position; small frequent meals (avoid fatty and spicy food); use of antacids, H_2-blockers (ranitidine) or Proton Pump Inhibitors PPIs (omeprazole). If all of the previous fail, Nissen fundoplication can be done (wrapping the fundus of stomach around the distal 3.5 cm of esophagus).

- **Hypertrophic pyloric stenosis:**

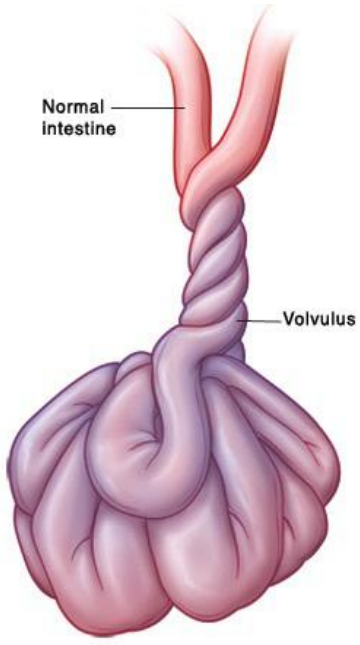


- It is the thickening of pylorus circular smooth muscle which results in obstruction and susbsequent projectile vomiting. It presents in the 2nd week of life and more commonly occurring in first-born males.
- **Clinical features**: projectile nonbilious vomiting immediately after feeding (this might cause dehydration).
- **Investigations**:
  - ✓ Physical examination: an olive-like mass can be felt.
  - ✓ Ultrasound is the method of choice.

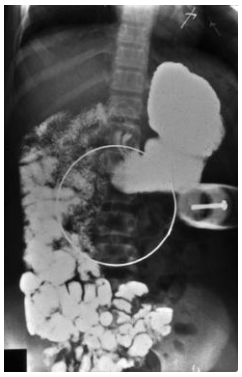


  - ✓ Electrolytes: hypochloremic, hypokalemic metabolic alklaosis.
- **Management**: first correct dehydration and electrolyte disturbances and then do partial pyloromyotomy.

- **Malrotation and midgut volvulus:**

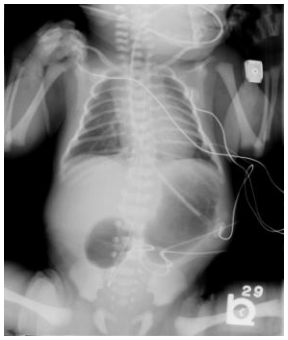


- Midgut will twist around superior mesenteric vessels resulting in ischemia and infarction of bowel. This commonly occurs in males and patients with heterotaxy (liver is on the left side; stomach is on the right side).
- **Clinical features**: bilious vomiting, sudden abdominal pain, abdominal distention and blood in stool.
- **Investigations**: Upper intestinal contrast imaging is the diagnostic tool of choice.



- **Management**: emergency surgery (untwisting of the gut with fixation to prevent recurrence + resection of infracted segments). This will result in short bowel syndrome and TPN may be required.

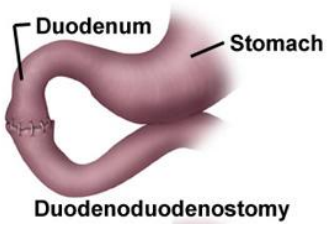
- **Duodenal atresia:**



- Duodenal atresia is caused by failure of the lumen to recanalize at 10 weeks f gestation. It occurs more in males and it is associated with Down syndrome (in 25% of cases).
- **Clinical features:** prenatal ultrasound shows polyhydramnios (fetus is unable to swallow amniotic fluid); physical examination shows scaphoid abdomen with epigastric distention; vomiting, weight loss and failure to thrive are common.
- **Investigations**:
  - ✓ Abdominal X-ray: double-bubble sign.

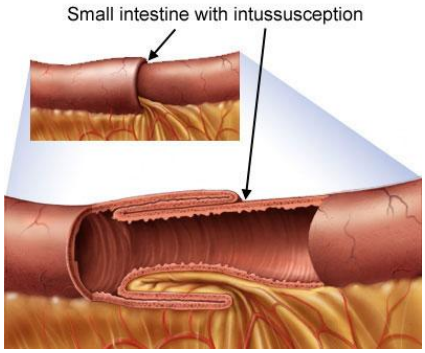
- ✓ Upper intestinal contrast imaging can be used.
- ✓ Electrolytes: hyochloremic metabolic alkalosis.

- **Management**: hydration and correction of electrolyte disturbances followed by duodenoduodenostomy.



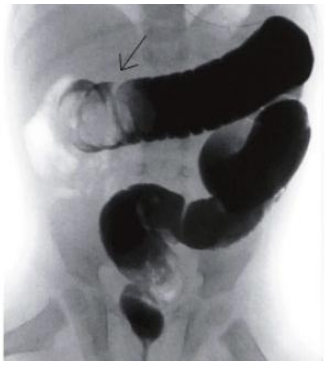
- **Intussusception:**

- It is the telescoping of a proximal part of intestine into a distal part which occurs more in males between the age of 5-9 months. Ileocolic intussusceptions is the most common location. The cause is unknown but lymphoma might drag proximal intestine inward. This is going to result in bowel wall edema, hemorrhage and ischemia.



- **Clinical features**: sudden colicky abdominal pain, vomiting and currant-jelly stool. A sausage-like mass can be felt in the right upper quadrant with physical examination.

- **Investigations:**
  - ✓ Gold standard: contrast enema (showing coil spring sign).
  - ✓ Abdominal ultrasound can also be used.



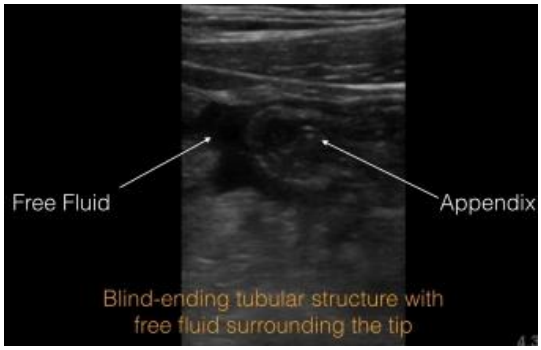
- **Management:**
  - ✓ Contrast enema with air (successful in 80-90% of cases). Recurrence is 5%
  - ✓ If it fails → surgery. Recurrence is 1%

- **Differential diagnosis of acute abdominal pain:**

- **Appendicitis, cholecystitis, pancreatitis, intussusception, volvulus, acute gastroenteritis, hepatitis and testicular torsion.**

- **Appendicitis:**

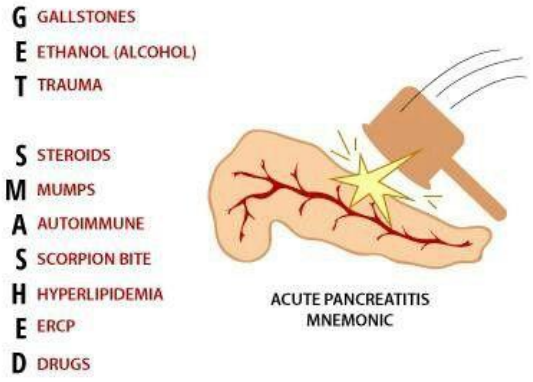
- It is the most common pediatrics emergency operation occurring between the age of 10-12 years and represented by obstruction and inflammation of the appendix. This obstruction results from fecalith or lymphoid tissue with a subsequent ischemia. The pain is referred to T10 dermatome (periumbilical region). If appendix is not surgically removed within 48 hours, this will result in perforation.



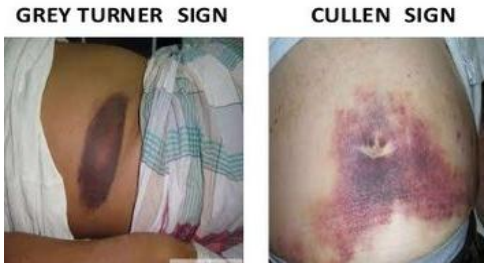
- **Clinical features**: fever, vomiting, abdominal pain which starts periumbilical (as mentioned above) then localizes to McBurney's point (1/3 the distance between anterior superior iliac spine and the umbilicus).
- **Investigations**: CBC (increased neutrophils), Abdominal ultrasound or CT-scan aid in the diagnosis.
- **Management**: hydration, perioperative antibiotics and appendectomy (through laparoscopy).

- **Acute pancreatitis:**

- It is an acute inflammation of the pancreas which is uncommon in children. It results from obstruction of pancreatic duct which leads to premature activation of pancreatic proenzymes that will autodigest pancreatic cells → interstitial edema, necrosis and hemorrhage.
- **Etiology**: Blunt trauma (most common cause), idiopathic (2nd most common cause in 25% of children), infections, obstruction or systemic diseases (such as cystic fibrosis).



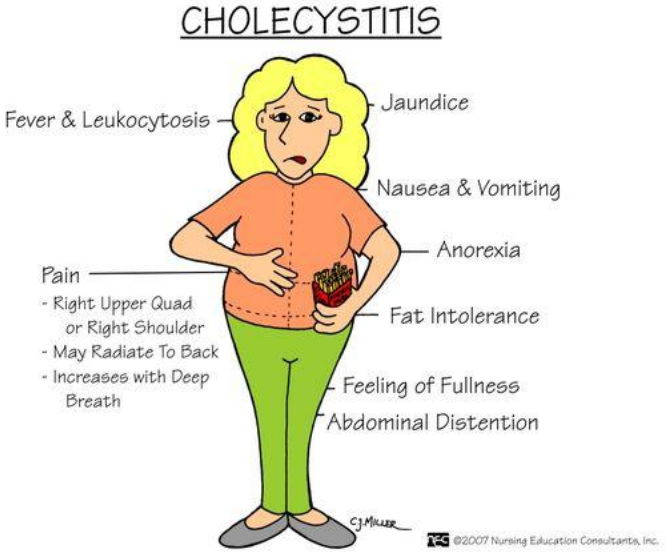
- **Clinical features**: acute epigastric pain that is referred to the back, fever and vomiting. Physical examination might show Gray-Turner sign (bluish discoloration of the left flank) or Cullen sign (bluish discoloration of periumbilical region). If there is severe hemorrhage, hypovolemic shock might occur (indicated by hypotension and tachycardia).
- **Investigations**: serum amylase (paying attention that serum lipase is more specific) and abdominal ultrasound.
- **Management**: hydration, analgesia (IV paracetamol = perfalgan) and TPN. Surgery in early pancreatitis is controversial and it is done to remove the necrotic tissue.

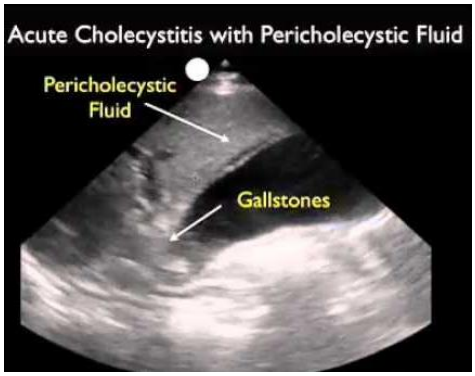


- **Cholecystitis:**

- It is inflammation and transmural edema of the gallbladder that is associated with gallstones. It is uncommon in healthy children. Obstruction of the cystic duct by a stone result in bile stasis with increased pressure. This will result in inflammation with increased risk of infection, necrosis and perforation.

- **Clinical features**: right upper quadrant pain, fever and vomiting. Physical examination reveals Murphy's sign (palpation of right upper quadrant during inspiration causes severe pain).
- **Investigations**: abdominal ultrasound.
- **Management**: hydration, analgesia, antibiotics and then removal of gallbladder (cholecystectomy) via laparoscopy.





- **Constipation and ecopresis:**

- **Constipation**: it is reduction in defecation that is associated with abdominal discomfort difficult/ painful defecation and stool retention. Stools are always dry and hard. This condition is common during childhood.
- **Encopresis**: involuntary defecation which is associated with emotional disturbance or psychiatric illness. There will be passing of liquid stool around a hard retained stool mass. This condition is common in males.
- **Normal stool patterns:**

1st week of life	4/day
By 1 year of age	2/day
By 4 years of age	1/day
Adults	Ranging from 3/day – 3/week

- **Etiology of constipation:**
  - ✓ Functional Fecal Retention (FFR: most common): behavioral pattern of stool with-holding resulting in large amount of stool which is retained and accompanied with: encopresis, abdominal pain, abdominal distention and fecal halitosis.
  - ✓ Organic cause (5% of children): Hirschsprung's disease is the most common cause of organic constipation.
- **Management**: adequate soluble fibers and increased water intake, stool evacuation by using mineral oil to soften and lubricate stool.

- **Inflammatory Bowel Disease (IBD):**

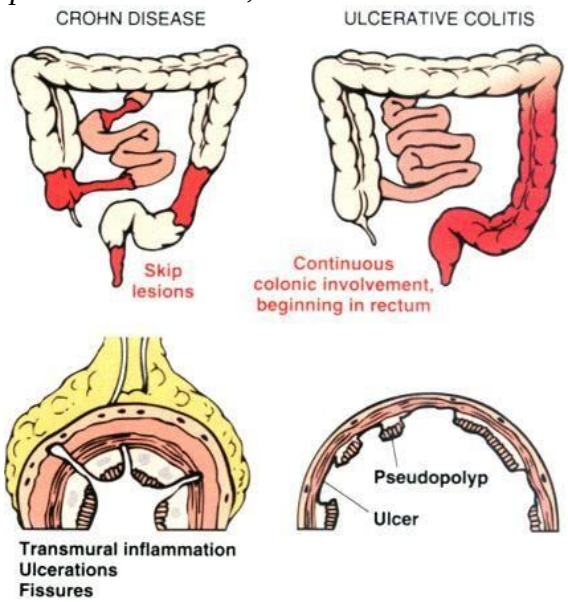
- **IBD involves both**: Crohn's disease (which is more common in males) and ulcerative colitis (which affects males and females equally).
- **Clinical features:**
  - ✓ Ulcerative colitis (UC):
    - ❖ There is diffuse inflammation (with no skip lesions) but only the mucosa is involved. It is limited to the colon and begins in the rectum then extends in a contagious fashion.
    - ❖ If it only affect rectum (ulcerative proctitis); if it affects the whole colon (pancolitis).
    - ❖ Clinical features depend on the severity of the disease:

Mild UC (60%)	Bloody diarrhea and abdominal pain
Moderate UC (30%)	Nocturnal stooling and cramping
Severe UC (10%)	> 6 stools/day, fever and anemia

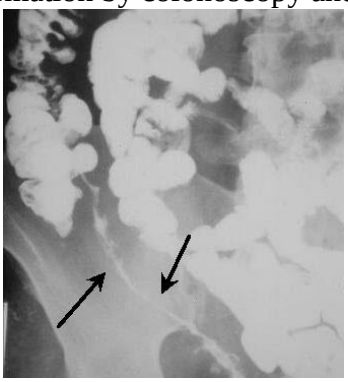
- ❖ *Complications*: toxic megacolon (patient presenting in septic shock) and increased risk of colon cancer.

✓ <u>Crohn's disease (CD):</u>

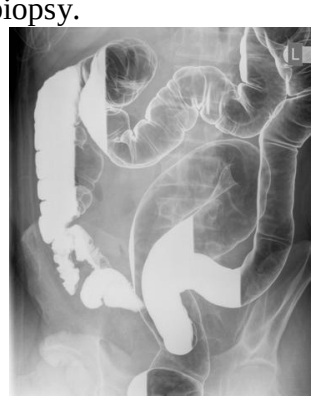
- ❖ There are skip lesions and inflammation is transmural (involving the whole wall of the GI tube). It can occur anywhere in GI tract from mouth to anus but commonly occurring in terminal ileum.
- ❖ *Clinical features*: abdominal pain, diarrhea (±blood) and malabsorption of iron, zinc and vitamin B12.
- ❖ *Complications*: fistulas, adhesions and strictures.



- **Investigations:**
  - ✓ CBC (which usually shows anemia or leukocytosis).
  - ✓ ↑ESR (because this is and inflammatory disease).
  - ✓ Stool for occult blood and culture (to rule-out an infection).
  - ✓ Barium: string-sign (CD); lead-pipe (UC).
  - ✓ Conformation by colonoscopy and biopsy.



String-sign



lead-pipe

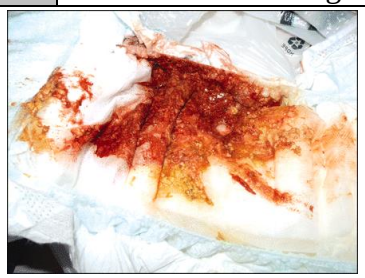
- **Management:**

Pharmacological	Surgery
• Sulfasalazine for UC • Corticosteroids and immunosuppressive agents (azathiprine) for CD	• UC can be treated by total proctocolectomy (removal of the rectum and entire colon)

- **GI bleeding:**

- **Terminologies:**

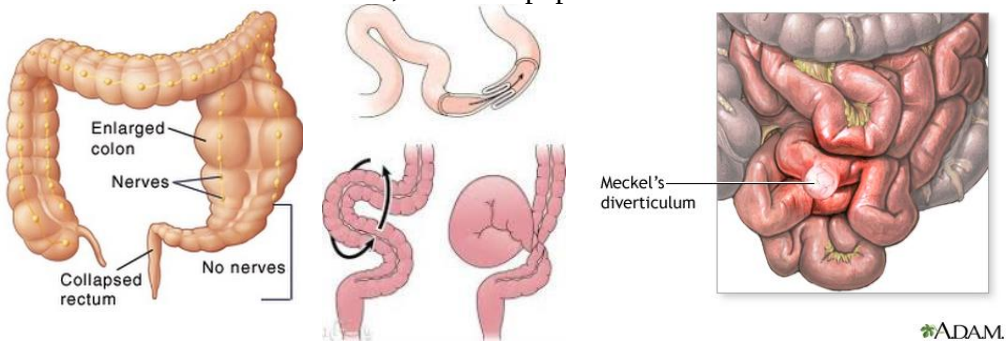
Hematemesis	Vomiting fresh or old blood (coffee ground appearance)
Hematochezia	Fresh blood in stool indicating bleeding from lower GI tract
Melena	Dark stool indicating bleeding from upper GI tract



Hematochezia

Melena

- **Conformation of GI bleeding**: stool for occult blood (= positive guaiac test)
  - ✓ False-positive: ingested iron, beets or rare red meats.
  - ✓ False-negative: large ingested doses of vitamin C.
- **Upper GI bleeding:**
  - ✓ Causes:
    - ❖ *Neonate*: swallowing maternal blood during delivery (especially CS) or feeding from a bleeding nipple.
    - ❖ *Children*: swallowing blood during an episode of epistaxis.
    - ❖ *Esophagus*: esophageal varices (due to portal hypertension) or Mallory-Weiss tear (laceration in esophagus due to forceful vomiting).
    - ❖ *Gastritis or ulcer*: due to burn, medications, severe stress or H.pylori infection.
  - ✓ Investigations: CBC (for Hb and platelet count), coagulation studies and endoscopy.
  - ✓ Management: stabilize hypovolemia and anemia in patient, octreotide (for varices), antibiotics (for H.pylori), H_2-blockers or PPIs for gastritis and ulcers.
- **Lower GI bleeding:**
  - ✓ Causes:
    - ❖ *Neonates*: necrotizing enterocolitis, Hirschsprung's disease or volvulus.
    - ❖ *Infants and children*: infectious colitis (e.g. Salmonella, Shigella, E.coli and Campylobacter), Mickel's diverticulum (outpouching of small intestine in terminal ileum containing ectopic gastric mucosa that produces acid and causes painless rectal bleeding. It is identified by nuclear medicine scan and has to be removed surgically), anal fissure and anal polyp.
    - ❖ *Adolescents*: IBD, Gastritis/peptic ulcer and infectious colitis.

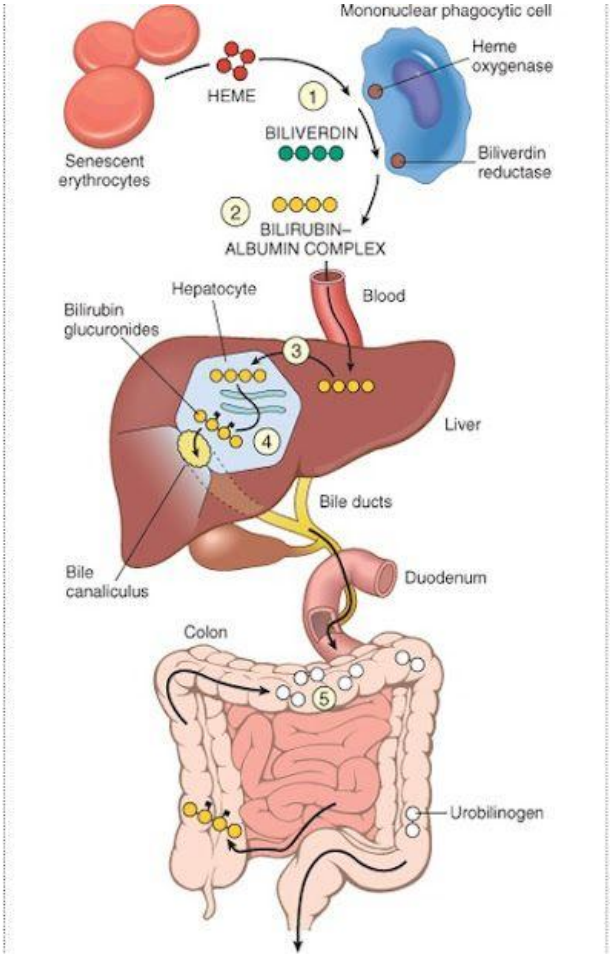


- **General concepts regarding the liver:**

- **Hepatocellular enzymes:**

AST	Sensitive but non-specific marker of liver disease
LDH	Non-specific marker of liver disease
ALT	Very specific marker for liver disease

- **Biliary enzymes**: GGTP and ALP are both elevated with biliary disease.
- **Bilirubin**: it is derived from breakdown of heme. Indirect bilirubin will be conjugated with glucouronide in the liver via the enzyme UDP-glucouronyl transferase to form direct bilirubin.



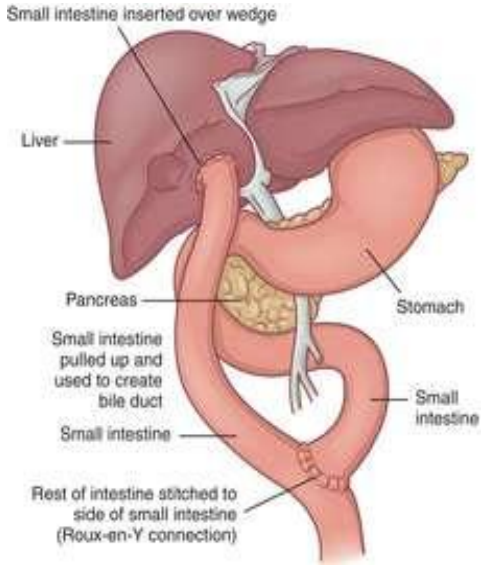
- **Genetic disease causing indirect hyperbilirubinemia:**

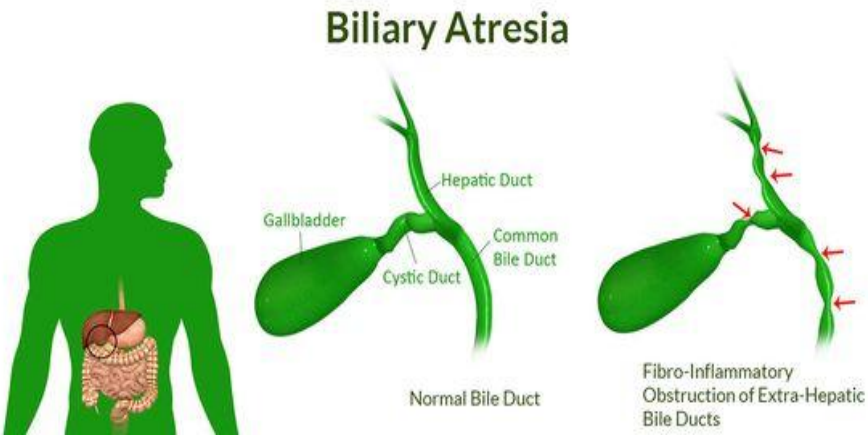
Gilbert's syndrome	• Autosomal Dominant (AD) • Reduction in activity of the enzyme UDP-glucouronyl transferase by 50% resulting in mild jaundice
Crigler-Najjar type I	• Autosomal Recessive (AR) • UDP-glucouronyl transferase is totally absent • Kernicterus will occur due to severe indirect hyperbilirubinemia (deposition of bilirubin in basal ganglia) • Management: repeated exchange transfusion and phototherapy to keep bilirubin < 20 mg/dL in first 2-4 weeks of life. Liver transplantation is the definitive treatment.
Crigler-Najjar type II	• Autosomal Dominant (AD) • 90% of UDP glucouronyl transferase is absent with somehow lower likelihood of kernicterus • Management: phenobarbitone which stimulates glucouronyl transferase enzyme.
Dubin-Johnson syndrome	• Autosomal Recessive (AR) • Defective excretion of conjugated bilirubin into bile resulting in black-liver jaundice.

- **Cholestatic disease of infancy:**

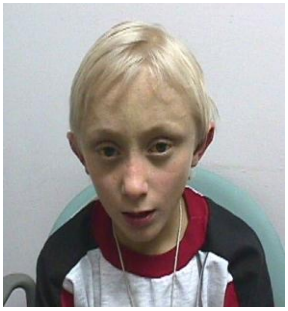
- It is represented by retention of bile within the liver which results in elevation of direct bilirubin.
- **Causes**: infections (hepatitis), idiopathic (neonatal hepatitis) or biliary atresia.
- **Clinical features**: jaundice, pale stool, dark urine and hepatomegaly.

- **Neonatal hepatitis:**
  - ✓ It is an idiopathic inflammation of the liver which commonly occurs in males and is self-limited in 70% of neonates.
  - ✓ Clinical features: ranging from (jaundice, pale stool, dark urine and hepatomegaly) to (liver failure, cirrhosis and portal hypertension).
  - ✓ Diagnosis: clinical presentation and exclusion of other causes of cholestasis. Liver biopsy can be done for confirmation.
  - ✓ Management: nutritional support (medium-chain triglycerides), supplements with fat-soluble vitamins, ursodeoxycholic acid (to enhance bile flow but it is not used until biliary obstruction is excluded).
- **Biliary atresia:**
  - ✓ It is a progressive fibrosclerotic disease affecting extrahepatic biliary tree.
  - ✓ Presentation at 4 weeks of age with jaundice (direct hyperbilirubinemia), dark urine and pale stool. By 4 months of age, bile duct obliteration and cirrhosis will occur.
  - ✓ Diagnosis: intraoperative cholangiogram. Abdominal ultrasound, radionucleotide scan and liver biopsy will be done to exclude other causes of cholestasis.
  - ✓ Management: treatment of choice is Kasai portoenterostomy which must be performed by 50-70 days of age (cholangitis is a complication). After the procedure, there must be nutritional support, supplementation with fat-soluble vitamins and ursodeoxycholic acid (once bile flow is re-established).





- **Allagile syndrome:**
  - ✓ It is an AD disease in which there is lack of intrahepatic bile ducts and multi-system involvement due to abnormality is chromosome 20.
  - ✓ Clinical features:
    - ❖ Cholestatic liver disease: jaundice, dark urine, pale stool and pruritis which can be debilitating in these patients.
    - ❖ *Facial characteristics*: broad forehead, pointed chin, widely-spaced eyes, saddle nose and large ears.
  - ✓ Diagnosis is made by clinical features and management is supportive.



- **Viral hepatitis:**
  - ✓ Clinical features of hepatitis: fever, right upper quadrant pain, fatigue, malaise, jaundice and hepatomegaly.

Hepatitis A (HAV)	• Picornavirus • Transmission: fecal-oral • Most common hepatitis virus causing infection. • Virus shed in stool (at 2 weeks); jaundice (at 4 weeks). Notice that > 70% of children are asymptomatic. • Diagnosis: HAV-IgM (acute disease); HAV-IgG (recovery and immunity) • Management: supportive; prevention: vaccination
Hepatitis B (HBV)	• DNA virus • Transmission: vertical transmission from mother to her fetus or parenteral transmission through: infected blood products, needles or body secretions. • Clinical features will appear after 90 days. Chronic hepatitis is more common in infants acquiring the infection from their mother. This will predispose them to cirrhosis and subsequent hepatocellular carcinoma. • Diagnosis: active disease (HBsAg); protection from vaccination or natural infection (HBsAb); active infection with increased infectivity (HBeAg) • Management: acute infection (supportive), chronic infection (interferon-α and antivirals); prevention: vaccination.
Hepatitis C (HCV)	• RNA virus (flavivirus) • Transmission: vertical transmission or parenteral exposure. • Clinical features: rarely symptomatic in children but it will result in chronic infection in 80% of patients with increased risk of cirrhosis and hepatocellular carcinoma. • Diagnosis: HCV-antibody in blood or HCV-PCR
Hepatitis D (HDV)	• RNA virus. • It need HBsAg to replicate. • Diagnosis: HDV-antibody.
Hepatitis E (HEV)	• RNA virus • Transmission: fecal-oral • 20% mortality when infecting pregnant women. There is no chronic state. • Diagnosis: HEV-antibody

- **Autoimmune hepatitis:**
  - ✓ It is a progressive destructive liver disease which is characterized by: elevated serum transaminases, hypergammaglobulinemia and circulating autoantibodies. It occurs more in females before age of puberty and other non-hepatic autoimmune disease is present in 20%-40% of cases.
  - ✓ Type-I is more common than type-I and is characterized by the presence of ANA or anti-smooth muscle antibody.
  - ✓ Clinical features: 50% present with acute hepatitis; 50% present with chronic liver disease.
  - ✓ Management: corticosteroids and immunosuppressive agents.