CRACKCast Episode 172: Pediatric Gastrointestinal Disorders

Episode Overview

Key concepts:

- Physiologic jaundice of the newborn and breast milk jaundice are the most common causes of jaundice in the neonatal period.
  - Direct hyperbilirubinemia in infants is always pathologic and requires a detailed evaluation
  - Hypertrophic pyloric stenosis is associated with gradually progressive nonbilious emesis that becomes projectile.

- Hypochloremic-hypokalemic metabolic alkalosis is the classic electrolyte derangement associated with hypertrophic pyloric stenosis.

- Bilious vomiting in the neonate should initiate a diagnostic evaluation for possible malrotation with volvulus or other intestinal obstructive pathology.
  - If they are toxic call surgery STAT
  - Toxic-appearing infants with bilious emesis should receive an emergent surgical consultation.

- Necrotizing enterocolitis occurs more commonly in premature infants, but 10% of affected infants are full term. Pneumatosis intestinalis—intramural air seen on x-ray—is pathognomonic for NEC.

- Gastroesophageal reflux (GERD) is very common in infants and is usually benign and self-limited. Occasionally, GERD may cause more severe symptoms, including irritability, respiratory distress, and failure to thrive. GERD usually responds to conservative measures (eg, positioning, thickening of formula, smaller and more frequent feedings); pharmacologic interventions are seldom needed.

- The classic clinical triad of intussusception includes colicky, intermittent abdominal pain, a palpable sausage-shaped abdominal mass, and bloody stools; however, the triad occurs in less than one-third of patients.

- Children with intussusception may present atypically, with an altered level of consciousness (eg, lethargy) rather than abdominal pain.

- Hirschsprung’s disease is a common cause of constipation in the neonate and usually manifests as delayed passage of meconium. Occasionally, children may present later in life with symptoms of chronic constipation. Toxic megacolon is the most serious complication.

- Meckel’s diverticulum classically manifests in children younger than 5 years, with massive, painless, brick red rectal bleeding.

- More than 90% of GI foreign bodies pass without complications

- Button batteries lodged in the esophagus may cause serious burns, erosions, and perforations within as little as 2 hours. Of all foreign bodies, button batteries require the most expeditious removal, usually by endoscopy.

- Appendicitis is the most common surgical disease in children. Diagnosis depends on a combination of clinical factors, including history, physical examination, laboratory values, and imaging studies. The ideal diagnostic algorithm remains elusive.
Effective imaging strategies for children with suspected appendicitis include initial ultrasound examination followed by CT scanning of the abdomen for those with equivocal findings.

- Causes of pancreatitis in children include viruses, trauma, drugs, and toxins. Biliary disease in children is more commonly caused by cholestasis rather than biliary obstruction. Pigment gallstones are more common than cholesterol stones in children. Biliary tract disease is usually diagnosed with right upper quadrant ultrasound imaging; management strategies are similar to those for adults.

Core questions:

1) List 8 causes of neonatal jaundice and indicate whether they are conjugated or unconjugated
2) List indications for work-up of a jaundiced infant
3) What are RFs for hyperbilirubinemia? (8)
4) What is the differential diagnosis for vomiting in a child?
5) Describe the typical presentation of each of the following:
   a. Hypertrophic pyloric stenosis
   b. Malrotation with midgut volvulus
   c. NEC
   d. GERD
   e. Intussusception
   f. Hirschsprung’s Disease
   g. Meckel’s Diverticulum
   h. HSP
6) List Xray findings for each of the following:
   a. Malrotation with midgut volvulus (2)
   b. NEC (4)
   c. Intussusception (5)
   d. Hirschsprung’s Disease (2)
7) Describe the conservative management of a patient with GERD.
8) What is the preferred diagnostic test for diagnosis for intussusception?
9) List causes of lead points in pts with intussusception.
10) Describe each of the following signs on physical exam:
    a. Sandifer’s syndrome
    b. Red-currant Jelly Stools
    c. Dance’s Sign
    d. Rovsing’s sign
    e. Psoas Sign
    f. Obturator Sign
11) Describe the “Rule of 2” for Meckel’s Diverticulum
12) What are 3 common locations of lodging in the esophagus
13) List 3 indications for FB removal from stomach. Describe the management of button battery FBs
14) What is HSP? How does it typically present?
15) List three complications of HSP.
16) Why is appendicitis different in very young children?
17) List 10 causes of pancreatitis in children
18) List 10 causes of biliary tract disease in children
   a. List conditions associated with the development of gallstones in children.
Wisecracks
1. What are the risk factors for necrotizing enterocolitis?
2. Describe the proposed pathophysiology of necrotizing enterocolitis?
3. List five pathologic causes of constipation in a child.
4. What is the most concerning complication of hirschsprung’s disease? How does it occur?
5. What is gallbladder hydrops? What conditions is it associated with?

Rosens in Perspective

● GI disorders are tough! Non-verbal children can present with virtually any set of symptomatology!
● It may be helpful to organize these varied GI diseases by patient age or mechanical/inflammatory-infectious/genitourinary/atypical:
  ○ Several disorders occur as a normal variant of early neonatal and infant development = (eg, neonatal jaundice, gastroesophageal reflux, hypertrophic pyloric stenosis). Others result from congenital malformations (eg, malrotation, Meckel’s diverticulum) or genetic abnormalities (eg, Hirschsprung’s disease). Idiopathic or poorly explained disorders include necrotizing enterocolitis (NEC), intussusception, Henoch-Schönlein purpura (HSP), and inflammatory bowel disease (IBD).
  ○ The child’s age can also help identify common causes of abdominal pain. Infants, for example, may have disorders such as NEC, hypertrophic pyloric stenosis, or intussusception, whereas older children are more likely to present with appendicitis, pancreatitis, or biliary tract disease. It can be helpful to consider the top most likely and can’t miss diagnoses in a 3mo old, 13mo, 3yo and 13 yo to round out your age appropriate differential.

Rosens Chapter 171 breaks it down by age. Review table 171.1 for age specific pathology.

Core questions:

[1] List 8 causes of neonatal jaundice and indicate whether they are conjugated or unconjugated

● Unconjugated bilirubin binds to albumin and is carried to the liver, where it is conjugated by glucuronyl transferase and excreted into bile.
● Hyperbilirubinemia and jaundice in the neonate usually result from a combination of three factors—increased production, decreased clearance and excretion, and increased enterohepatic resorption.

Hyperbilirubinemia in the neonatal period is usually unconjugated. Conjugated hyperbilirubinemia, which is always pathologic, is less common.

1. Conjugated causes:
   a. TORCH infections (Congenitally acquired infections including Toxoplasmosis, Other (which isn’t very helpful but can include syphilis, varicella-zoster, parvovirus B19), Rubella, CMV, and Herpes infections)
   b. Sepsis / bacterial infection of any site
   c. Viral infections
d. Drugs/toxins 

e. Bile duct atresia/stenosis 

f. Cystic fibrosis

2. Unconjugated causes:
   a. Physiologic jaundice (>50% of cases of jaundice!) Onset at 2-7 days of life. 
   b. Breast milk jaundice (2nd most common cause) 
      i. hormonally mediated or related to increased enterohepatic reabsorption of 
         bilirubin. Typically presents after the first three to five days of life, peaking 
         within two weeks after birth 
   c. Hemolysis:
      i. ABO incompat. 
      ii. Cephalohematoma 
      iii. Breakdown diseases: spherocytosis, thalassemia, G6PD 
   d. Obstructive: 
      i. Meconium ileus 
      ii. Pyloric stenosis 
   e. Infectious 
   f. Metabolic 
      i. Congenital hypothyroidism

[2] List indications for work-up of a jaundiced infant

So, as a recap, why do we actually care about jaundice?

- Increased risk of bilirubin-induced neurologic dysfunction (BIND).
  - Acute bilirubin encephalopathy (ABE) refers to the early, potentially reversible, signs and 
    symptoms, including somnolence, poor feeding, hypertonia or hypotonia, and a high-pitched 
    cry associated with severe hyperbilirubinemia (>20 mg/dL or 342 mcmol/L - total serum 
    bilirubin). This only occurs from unconjugated bilirubin because it is lipid soluble and can 
    cross the blood brain barrier.
  - If untreated can progress to: neck (retrocollis) and trunk (opisthotonos “opis-thot-onos”), 
    fever, irritability and, ultimately, apnea, seizures, and death. If treated, some or all these 
    symptoms may be reversible. Kernicterus refers to the chronic, long-term neurologic 
    sequelae of BIND.

Box 171.1 Indications for Evaluation of Jaundiced Infants

- Jaundice in first 24 hours of life 
- Any elevated directed serum bilirubin level 
- Rapidly rising total bilirubin level unexplained by history and physical 
- Total serum bilirubin approaching exchange level or not responding to phototherapy 
- Jaundice persisting beyond 3 weeks of age 
- Sick appearing infant 

Jaundice within the first 24 hours of life should be evaluated emergently as it is often from a pathologic 
hemolytic process.
Conjugated (direct) hyperbilirubinemia is always pathologic, resulting from biliary atresia, other biliary obstructive pathology, severe infections, toxins, and inborn errors of metabolism.

**Evaluation should include:**
- CBC
- Peripheral smear
- Reticulocytes
- Coombs test (determine immune-mediated major blood group incompatibility)
- Diagnostic testing in ill-appearing infants includes finger stick blood glucose level measurement, electrolyte panel, urine assay for reducing substances, serum ammonia levels, and evaluation for infection.

The treatment of high unconjugated bilirubin is based on the guidelines for phototherapy graph, which considers the total serum bilirubin level and risk factors for hyperbilirubinemia (which brings us to our next question).

For guidelines on how and when to treat elevated bilirubin:


**[3] What are RFs for hyperbilirubinemia? (8)**

- Prematurity (<37 wks GA)
- Isoimmune-mediated hemolysis
- ABO incompatibility)
- Sepsis
- Cephalohematomas,
- Dehydration,
- Genetically inherited abnormalities
  - hereditary spherocytosis
  - glucose-6-phosphate dehydrogenase (G6PD) deficiency.

**[4] What is the differential diagnosis for vomiting in a child?**

- See Rosens table 171.4 for differential considerations of vomiting by age

HUUUGE! There are a number of ways to remember this so do what works for you but I like to break it down by category:
- Mechanical, inflammatory/infectious, genitourinary, CNS, metabolic, or other.
- Mechanical in infancy includes the variety of pathologies in 171.4
  - As they get older think about constipation as well.
- Inflammatory or infectious think about the variety of pathologies in 171.4
  - Into childhood or adolescence also consider appendicitis, pancreatitis, biliary tract disease.
- Genitourinary is UTI's and as they get older testicular/ovarian torsion and don't forget pregnancy in adolescence.
- CNS can include causes of increased ICP like hydrocephalus, intracranial hemorrhage, tumor.
- Metabolic think about DKA and inborn errors of metabolism
- Other remember to consider non-accidental trauma and toxic ingestions.
[5] Describe the typical presentation of each of the following:

- **Hypertrophic pyloric stenosis**
  - Male (up to 85% male), infant, first-born, 2-6 weeks (can present up to 20wks old), with progressive vomiting
  - Classic findings seen late in the disease:
    - Weight loss, “olive” on exam, hypochloremic-hypokalemic metabolic alkalosis.
    - Projectile vomiting may initially be absent
  - On ultrasound, the pylorus appears thickened (pyloric muscle thickness > 4 mm; pyloric diameter > 14 mm) and elongated (>19 mm), which is diagnostic. A characteristic string sign, reflecting passage of contrast material through the narrowed pyloric sphincter, may also be evident. In advanced stages with complete obstruction at the pylorus; plain films may reveal a distended, air-filled stomach
  - **Differential Diagnosis**
    - Reflux classically begins shortly after birth and remains relatively constant. Infant with pyloric stenosis typically have progressively worsening emesis beginning around 2 or 3 weeks of life. In advanced stages, it occurs with every feed and is often described as projectile.
    - Malrotation with midgut volvulus, duodenal atresia, and necrotizing enterocolitis, reflux and pyloric stenosis, emesis is rarely bilious.
  - **Management:**
    - Hypertrophic pyloric stenosis is not a true surgical emergency but may be a fluid and electrolyte emergency. As long as the blood gas is okay and there’s no signs of FTT or dehydration gen surg often will handle these semi-urgently

- **Malrotation with midgut volvulus**
  - M > F, 50% occur by the age of 1 (30% in the first month), 75% by age 5.
  - **sudden-onset bilious emesis and abdominal distention.** Any pigmented staining of the vomitus suggests the presence of bile. When bile is initially produced, it is bright yellow and turns green only with time and oxidative exposure. **Differential coloring of bile-stained emesis, yellow versus green, is not predictive of a surgical condition.** Infants usually appear quite ill and may present in shock.
  - **DX:** limited upper GI contrast series:
    - Symptoms, including bilious emesis and abdominal distention but, unlike malrotation with volvulus, NEC is characterized radiographically by diffusely dilated loops of small bowel and the presence of air within the bowel walls, termed pneumatosis intestinalis.
  - **Management:**
    - **Emergent Peds GenSx consultation with any infant/neonate with bilious vomiting**
    - In acute midgut volvulus, operative intervention must be rapid to save the bowel from necrosis. Intravenous (IV) access should be obtained, and laboratory studies should include blood glucose level, a CBC with differential, electrolyte values, and renal and liver function tests. Repeated fluid boluses of 20 mL/kg of normal saline should be given until adequate circulation has been reestablished.
    - Broad spectrum Abx.
    - NG/OG insertion with drainage.
Pathophysiology
  - During embryological development, the GI tract rotates around the superior mesenteric artery. As it completes the rotation, the duodenum forms a C-loop and is fixed to the retroperitoneum in the left upper quadrant at the ligament of Treitz. The cecum becomes similarly fixed in the right lower quadrant. Thus, the duodenum and cecum normally come to lie widely separated and are firmly fixed in position by peritoneal attachments called Ladd’s bands. They are only loosely connected by a broad-based mesentery. In cases of malrotation, the duodenum and cecum do not rotate completely, remain closely positioned, and are suspended in the midgut region by the mesenteric vascular stalk. This unusually close proximity results in a short stalk of mesentery that easily twists on itself, resulting in obstruction of the distal duodenum and bowel ischemia and necrosis secondary to compression of the superior mesenteric artery.

NEC
  - Most common GI emergency in the newborn period
  - Most affected infants are premature and acquire the condition in the NICU, NEC usually is not considered a disease of the ED.
    - Multifactorial, but 90% are premature with some injury to the intestinal wall.
    - Infants with NEC first usually develop feeding intolerance and bilious or nonbilious emesis. In the more advanced stages of the disease, infants may appear extremely ill, with hematemesis, hematochezia, fever, and shock.

Management:
  - Airway management
  - IV/IO access
  - fluid/electrolyte replacement
  - NG/OG to suction
  - Labs, cultures,
  - Glucose replacement
  - Vasoactive agents as needed
  - Broad spectrum ABX (See box 171.2)
  - Consult surgery - in consideration of possible

GERD
  - GERD occurs as a result of an incompetent lower esophageal sphincter. GER is classified as a disease aka GERD if complications occur. For example, chronic reflux of gastric contents into the esophagus may result in esophagitis, aspiration, and failure to thrive if it is severe.
  - Begins shortly after birth and resolves with time, usually by the age of 1 year. Improves when upright (sitting at 6mo and often really improves when standing at 12mo).
  - Exists on a spectrum - mild spitting up to FTT. rarely is a progressive disease like pyloric stenosis.
Intussusception

- Intussusception refers to the invagination of part of the intestine into itself. It is the most common cause of intestinal obstruction in children younger than 2 years, occurring most frequently in infants 5 to 12 months of age.
- Etiology still unclear - ?lead point causing telescoping of the bowel?
  1. Note that once the bowel telescopes it remains “stuck”. The episodes of pain are due to peristalsis.
- Ileocolic intussusceptions are most common.
  1. Ileocecal intuss. Is usually due to a preceding viral illness or mesenteric adenitis (e.g. post Kawasaki’s disease, HSP).
- Clinical: abdominal pain, +/- vomiting, currant jelly stools, “sausage-shaped abdominal mass”.
  1. **cyclic episodes of severe abdominal pain as waves of peristalsis cause bowel dilation adjacent to and proximal to the involved bowel. These episodes typically last 10 to 15 minutes and occur in intervals of 15 to 30 minutes. During the painful episode, the child may be irritable and inconsolable, often described as drawing the legs up to the abdomen and screaming in pain.**
  2. Clinically in a pain free period the patient should have a palpable mass or focal findings on exam in the context of a dramatic history of painful episodes.
  3. May present with altered LOC, lethargy, pain out of proportion = ischemic symptoms
    - a. Great ddx on the altered pediatric patient

- Imaging:
  1. **it is not necessary that the kid be in pain for the ultrasound to be positive.

- Mgmt:
  1. NPO, IV fluids for resuscitation, pain control.
  2. Air or contrast enema (pneumostatic or hydrostatic enema)
  3. **Either type of enema requires readily available backup by a pediatric surgeon in case the bowel DOES NOT reduce or iatrogenic perforation occurs.**
  4. The overall success rate for an air-contrast enema or barium enema approaches 90%. Surgical intervention is indicated in cases of prolonged intussusception with signs of perforation or shock or if enema reduction is unsuccessful.
  5. A severe, acutely life threatening complication of air enema reduction is tension pneumoperitoneum. Intussusception recurs in approximately 12% of radiologic reductions. Less than 50% of these recurrences occur within the first 48 hours postreduction. Although admission for observation after reduction has traditionally been recommended, a shorter observation period (6 hours) is safe, and children who are able to tolerate oral fluids may be discharged home.

Hirschsprung's Disease

- Boys > girls. Functional bowel obstruction = stool accumulation in the colon, leading to megacolon and chronic constipation. May present in infancy but approximately 10% can occur over 3 years old. The severity and age of presentation typically depends on the amount of bowel affected so short segment hirschsprung’s may be diagnosed later in life
- Hirschsprung’s disease accounts for approximately 20% of cases of partial intestinal obstruction in early infancy.
• Vomiting, irritability, poor weight gain, failure to thrive, and abdominal distention may be present.

• **congenital aganglionosis of the colon**—that is, an absence of ganglion cells (or types of neurons) in the myenteric plexus of the distal colon.

**Meckel’s Diverticulum**

- **Congenital malformation in the intestines:**
  - Meckel’s diverticulum are remnants of the omphalomesenteric duct and contain bowel wall, with 60% containing heterotopic tissue, usually gastric mucosa. Bleeding occurs when acid secretion from the ectopic gastric mucosa causes ulceration and erosion. Meckel’s diverticulum is the most common congenital malformation of the small intestine.
    - Colour - brick red to melena.

Complications may include severe anemia, intussusception, obstruction, perforation, and peritonitis.

**HSP**

- **Henoch schonlein purpura (”Hen-ock Shawn-line”), aka: anaphylactoid purpura, is a hypersensitivity vasculitis with immune complex deposition with immunoglobulin A; it mainly affects the small vessels (arterioles and capillaries). Although it is most well-known for its characteristic petechial to purpuric rash, HSP is a systemic vasculitis affecting any vessels.**
  - HS = hypersensitivity, PP = pain, purpuric rash
  - H = hematuria, S = sore abdomen, P = PP
  - abdominal pain, palpable purpuric rash, arthralgias, and renal disease. It is most common in children 4 to 11 years of age and occurs during the spring season following viral upper respiratory infections.
    1. +/- N/V/D
    2. Palpable purpura on the buttocks and lower extremities (dependent areas)
    3. 70% have GI bleeding
    4. Some may have ileoileal intussusception
  - Microscopic hematuria occurs in 50%. The syndrome is often relapsing and remitting for several weeks and may be associated with arthralgias. Neurologic involvement is less common in children.
  - Clinical dx. Get a urine, CBC, differ, BC, ESR.
    1. THEY SHOULD NOT HAVE THROMBOCYTOPENIA
  - Mgmt:
    1. Nsaisds, tylenol, prednisone for severe symptoms only. If these kids are well (ie no clinical or laboratory signs of complications and the pain is tolerable) I typically set them up with outpatient peds to follow their symptoms, urine and blood pressures. I admit to hospital if they have ++abdo pain, ++joint involvement limiting ambulation/self care, mental status changes, ++ GI bleed, compromised renal function, HTN or other signs of nephrotic syndrome.
Additionally, a set of six criteria, with the presence of at least one of these criteria, predicts the need for hospitalization—orchitis, moderate or severe abdominal pain, polyarthritis, GI bleeding, and inability to ambulate. Those with compromised renal function should have a nephrology consultation and also should be considered for admission, particularly if presenting with hypertension.

[6] List Xray findings for each of the following:

- **Malrotation with midgut volvulus (2)**
  1. Remember that you can have malrotation without midgut volvulus!
  2. **X-ray findings:**
     1. air-fluid levels suggesting obstruction,
     2. abnormally dilated bowel loops overlying the liver,
     3. a paucity of small bowel gas distally
  3. limited upper GI contrast series showing:
     1. an abnormal position of the duodenal C-loop
     2. small bowel, with a characteristic corkscrew appearance
  4. **Ultrasonography,** usually performed to evaluate for hypertrophic pyloric stenosis, may reveal an abnormal orientation of the superior mesenteric artery and vein (the vein is abnormally positioned anteriorly or to the left of the artery;) or a whirlpool sign caused by the vessels twisting around the mesenteric stalk, causing an echogenic twisting pattern.

- **NEC (4)**
  1. Plain abdominal radiographs are the imaging study of choice in NEC. Radiographs may show:
     1. nonspecific dilated loops of bowel,
     2. intramural air (pneumatosis intestinalis), which is pathognomonic for NEC
     3. air within the portal system and biliary tract,
     4. pneumoperitoneum.
  2. Pneumatosis is present in 75% of patients with NEC. No individual laboratory test is diagnostic or specific for NEC, but may reflect dehydration, electrolyte derangements, and sepsis.
  3. Volvulus is associated with a paucity of small bowel gas, whereas the hallmark of NEC is diffusely dilated loops of small bowel and pneumatosis intestinalis.

- **Intussusception (5)**
  1. X-ray not the best test
  2. If air is visualized throughout the entire colon and cecum, with normal gas patterns Intuss. Is less likely.
  3. soft tissue mass or mass effect,
  4. dilated loops of small bowel and a paucity of gas in a decompressed colon suggesting obstruction,
  5. a target sign (representing air in the intussusceptum as it telescopes into adjacent bowel),
  6. meniscus sign (representing air compressed like a meniscus from invaginating bowel),
  7. free air
  8. Ultrasound is the best test.
     1. Target sign, pseudokidney sign,
- **Hirschsprung's Disease (2)**
  - nonspecific and may reveal evidence of
    1. fecal impaction with proximal obstruction,
    2. air-fluid levels,
    3. dilated colon.

[7] **Describe the conservative management of a patient with GERD.**

  - Mgmt: smaller feedings, frequent burpings, formula thickened with cereal, and a semi-upright position after feeding. Pharmacologic regimens are not recommended for infants with uncomplicated reflux (so-called happy spitters). Although lacking supportive evidence, acid suppression can be used, but should be reserved for those with more severe symptoms, such as esophagitis, weight loss, or significant irritability, in whom more conservative lifestyle modifications have failed.

[8] **What is the preferred diagnostic test for diagnosis for intussusception?**

  - Ultrasound!

- Ultrasonography is the diagnostic imaging modality of choice. In skilled hands, it is highly sensitive and specific. The classic finding is a target sign (also referred to as a bull’s eye or doughnut sign), visualization of the telescoping intestinal wall in the transverse or cross-sectional view. When visualized in the longitudinal plane, it is referred to as the pseudo–kidney sign. See Figure 171.9.

[9] **List causes of lead points in patients with intussusception.**

  - In younger children, lead points are usually the result of enlarged Peyer’s patches secondary to a recent viral infection.
  - Approximately 75% of cases, especially in children older than 5 years, have a pathologic lead point; lesions include HSP vasculitis, Meckel’s diverticulum, lymphoma, polyps, post surgical scars, celiac disease, and cystic fibrosis.
  - Ileoileal intussusception occurs more frequently in children with HSP.

[10] **Describe each of the following signs on physical exam:**

  - **Sandifer’s syndrome**
    - Sandifer’s syndrome, although rare, refers to the stereotypical opisthotonic movements (back arching) highly suggestive of severe GERD.
    - An unusual posturing consisting of arching of the back, torsion of the neck, and lifting up of the chin. Sandifer syndrome is found in typically-developing preschool-aged children
    - GERD may occasionally be associated with brief periods of apnea and pallor, apparent life-threatening events, or respiratory symptoms, including chronic coughing, recurrent stridor, and persistent wheezing.
• **Red-currant Jelly Stools**
  - Rare finding in intussusception
  - **Blood, gross or occult, may be present in the stool.** Diarrhea containing mucus and blood constitutes the classic currant jelly stool, a relatively infrequent and late finding.

• **Dance’s Sign**
  - A rare finding in intussusception:

**FROM UPTODATE:**
Most exam features suspicious for of intussusception include:
- No abdominal tenderness, or only focal tenderness (especially in the right mid or upper abdomen)
- Abdomen not distended
- Lethargy or altered consciousness (often episodic)

Features that are more specific for intussusception, but are present in a minority of patients, include:
- Right lower quadrant that is scaphoid (empty) (Dance’s sign)
- Palpable “sausage-shaped” mass in the right mid or upper abdomen”

• **Rovsing’s sign, Psoas Sign, and Obturator Sign**

The rovsing, psoas, and obturator signs are difficult to assess in young children and should not be relied on due to their poor sensitivities and specificities.

From: [https://www.slideshare.net/kulwant1955/a-p-p-e-n-d-i-c-i-t-i-s](https://www.slideshare.net/kulwant1955/a-p-p-e-n-d-i-c-i-t-i-s)


Meckel’s diverticulum traditionally follow so-called the rule of 2’s.
- The diverticulum is 2 cm wide, 2 cm long, and located within 2 feet of the ileocecal valve.
- Moreover, the condition occurs in 2% of the population, and only 2% of affected patients ever become symptomatic.
• Of symptomatic patients, 50% manifest symptoms by the age of 2 years, and most present by the age of 20 years.

[12] What are 3 common locations of lodging in the esophagus

Foreign bodies commonly become lodged in one of three areas of normal physiologic narrowing—
• Upper esophageal sphincter (cricopharyngeus muscle), thoracic inlet (C6-T1);
• Aortic arch, tracheal bifurcation (T4-6);
• Lower esophageal sphincter, diaphragmatic hiatus (T10-11).

Of objects that have made it into the stomach, 80% to 90% are passed without complications.

[13] List 3 indications for FB removal from stomach. Describe the management of button battery foreign bodies

• See box 171.3, Indications for Emergency Removal of GI FB
  o Signs of respiratory distress
  o Evidence of esophageal obstruction
  o Sharp or long (>5cm) objects in esophagus or stomach
  o High powered magnets
  o Signs or symptoms of intestinal inflammation, obstruction, or perforation
  o Esophageal foreign bodies impacted for >24H or for an unknown amount of time

Specific stomach indications
• Button battery that has not left the stomach after 48 hrs from time of ingestion
• Objects longer than 5 cm
• Objects wider than 2 cm
• Obstructive symptoms
• Febrile, peritoneal findings suggestive of perforation

Button batteries lodged in the esophagus cause severe mucosal erosions, severe burns, and mediastinitis in as little as 2 hours, most likely as a result of the electrical current discharged from lithium batteries.

GI consultation for emergent foreign body removal is necessary. Button batteries in the stomach usually pass without complications and do not require removal unless they fail to pass the pylorus in 48 hrs of ingestion.

[14] What is HSP? How does it typically present?

Henoch schonlein purpura, aka: anaphylactoid purpura, is a hypersensitivity vasculitis with immune complex deposition with immunoglobulin A; it mainly affects the arterioles and capillaries. Although it is most well-known for its characteristic petechial to purpuric rash, HSP is a systemic vasculitis affecting any vessels.
• HS = hypersensitivity, PP = pain, purpuric rash
• H = hematuria, S = sore abdomen, P = PP
Abdominal pain, palpable purpuric rash, arthralgias, and renal disease. It is most common in children 4 to 11 years of age and occurs during the spring season following viral upper respiratory infections.

- +/- N/V/D
- Palpable purpura on the buttocks and lower extremities
- 70% have GI bleeding
- Some may have ileoileal intussusception

Microscopic hematuria occurs in 50%. The syndrome is often relapsing and remitting for several weeks and may be associated with arthralgias. Neurologic involvement is less common in children.

- Clinical dx. Get a urine, CBC, differ, BC, ESR.
  - THEY SHOULD NOT HAVE THROMBOCYTOPENIA

Management:
- Nsaids, tylenol, prednisone for severe symptoms only.

Additionally, a set of six criteria, with the presence of at least one of these criteria, predicts the need for hospitalization—orchitis, moderate or severe abdominal pain, polyarthritis, GI bleeding, and inability to ambulate. Those with compromised renal function should have a nephrology consultation and also should be considered for admission, particularly if presenting with hypertension.

[15] List three complications of HSP.

- Intussusception***
- GI bleeding
- Renal failure
- Hypertension
- Secondary cellulitis of leg/body purpura
- Testicular torsion
- Severe rare ones are CNS bleed, mesenteric ischemia or MI

[16] Why is appendicitis different in very young children?

Uncommon in children < 5. Plus they can’t communicate very well!

Children have an appendix that is more prone to perforation due to weak appendiceal wall, and no omentum to prevent infectious spread. In children, the rate of appendiceal perforation varies inversely with age. Perforation is highest among children younger than 5 years, among whom more than 50% are ruptured at the time of surgery.

Ultrasonographic findings consistent with appendicitis include an enlarged, noncompressible appendix (wall thickness > 2 mm; total appendix diameter > 6 mm) that is painful during scanning

[17] List 10 causes of pancreatitis in children
Uncommon in age < 10 yrs. associated with trauma, infection, structural anomalies, systemic disease, and drugs or toxins. Idiopathic causes account for 30% of cases. Biliary obstruction should be considered in the adolescent.

- Trauma
  - Bicycle crash / handlebar injury
  - Sports tackles
- Infection
  - EBV, CMV
- Structural:
  - Congenital cyst or duct malunion or otherwise
- Systemic disease
  - Autoimmune:
    - Sjogren syndrome, inflammatory bowel disease, or primary biliary cholangitis
    - Cystic fibrosis, Sickle cell disease
- Drugs
  - eg, metronidazole, mercaptopurine, valproate, and isoniazid)
- Toxins
  - Hyperlipidemia
  - Alcohol
- Idiopathic
- Biliary obstruction (more common in adolescents)

[18] List 10 causes of biliary tract disease in children

Usually related to systemic disease
- Hemolytic disease:
  - Sickle cell disease
  - Spherocytosis
- Cystic fibrosis
- TPN
- Infectious causes
  - Sepsis
  - RMSF
  - Salmonella / Shigella infections
  - Recent URTI/GI infections
  - Kawasaki’s
  - Strep. pharyngitis
- Dehydration

Other miscellaneous causes:
- Ceftriaxone use in neonates
- Nephrotic syndrome
- Hydrops of the gallbladder

See UPTODATE for a table of differential diagnosis considerations for etiology of acute pancreatitis

List conditions associated with the development of gallstones in children.
Pigment gallstones stones are more common in childhood; cholesterol stones are uncommon prior to adolescence. They result from the excess breakdown of red blood cells and are usually seen in those with hemolytic anemia, such as sickle cell disease and spherocytosis. Adolescents may form cholesterol gallstones in association with oral contraceptives, pregnancy, or obesity.

Wisecracks.
[1] What are the risk factors for necrotizing enterocolitis?

- Diagnosis in the newborn period (usually kids in the NICU)

Uptodate:
- Prematurity
- Microbial bowel overgrowth
- Milk feeding
- Impaired mucosal defense
- Circulatory instability of the intestinal tract
- Medications that cause intestinal mucosal injury or enhance microbial overgrowth

[2] Describe the proposed pathophysiology of necrotizing enterocolitis?

The exact pathophysiologic mechanism of NEC is unclear but is likely multifactorial. Prematurity is the most common and universally accepted risk factor; 90% of all affected infants are born prematurely. The primary pathologic event may be inflammation or injury to the intestinal wall, beginning in the mucosa and extending transmurally.

[3] List five pathologic causes of constipation in a child.

- Neurogenic
  - Hirschsprung’s disease
  - CP
  - Spinal cord injury
  - Neurofibromatosis
  - Infantile botulism
  - Dysautonomia
  - Duchenne muscular dystrophy
- Endocrine / metabolic
  - Cystic fibrosis
  - Hypokalemia (adynamic ileus)
  - Lead poisoning
  - Hypo / Hypercalcemia
  - Hypothyroidism
  - DM
  - Scleroderma
  - Porphyria
● **Anatomic**
  ○ Atresia, webs, volvulus
  ○ Imperforate anus
● **Other**
  ○ Opiates, anticholinergics, antidepressants, chemotherapy, aluminum containing antacids

[21] **What is the most concerning complication of Hirschsprung's disease? How does it occur?**

Hirschsprung-associated enterocolitis (HAEC)
- Can surgical intervention (preoperative HAEC), in the immediate postoperative period, or long after definitive repair (postoperative HAEC).
- Presentation usually explosive foul-smelling diarrhea, fever, vomiting, and abdominal pain and distension, and sometimes rectal bleeding and shock. Mild cases may be misdiagnosed as gastroenteritis.
- Therefore, HAEC should be considered in any child known to have HD presenting with symptoms suggestive of gastroenteritis. Radiographs show an obstructive picture, with air-fluid levels and dilated and edematous bowel loops.

The mechanisms underlying HAEC are not fully understood. Contributing factors include stasis and reduced production of protective mucin, which lead to bacterial overgrowth in the lumen of the bowel proximal to the involved segment, and possibly translocation of bacteria through the mucosa

[22] **What is gallbladder hydrops? What conditions is it associated with?**

Hydrops of the gallbladder (ie, fluid distention of the gallbladder from chronic cystic duct inflammation or obstruction without inflammation or infection) is associated with viral upper respiratory or GI infections, Kawasaki disease, streptococcal pharyngitis, mesenteric adenitis, nephrotic syndrome, and leptospirosis:

- “This entity occurs specifically in children and is characterized by an acute illness with massive distention of the gallbladder in the absence of stones, bacteria, or congenital malformations.” From: PMID: 6854491