Diagnostic Imaging of Pediatric Gastrointestinal Abnormalities

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Learning Objectives

• By the end of this lecture, you will be able to...
  – Determine when abdominal ultrasound versus fluoroscopy is appropriate for the infant with vomiting.
  – More confidently recognize pneumatosis and indicate two common disease processes that cause it.
  – Differentiate pediatric liver tumors in the infant based on imaging.
Warm Up Case

Audience Response Question:
Is this bowel gas pattern normal?

A. Yes
B. No
Case 1: A neonate presents with vomiting and this abdominal radiograph.
Question 1: What is the appropriate next step?

A. Upper GI Series  
B. Limited abdominal ultrasound  
C. No imaging needed  
D. Fluoroscopic enema  
E. Repeat radiographic series later

GI Tract Obstruction: Upper or Lower?

- Upper tract obstruction DDx:
  - Malrotation with midgut volvulus  
  - Duodenal atresia or stenosis  
  - Intraluminal web  
  - Annular pancreas  
  - Hypertrophic pyloric stenosis

- Distal bowel obstruction DDx:
  - Neonate
    - Meconium ileus  
    - Jejunal/ileal atresia  
    - Colonic atresia  
    - Hirschsprung  
    - Left colon syndrome  
  - Infant to Preschool
    - Intussusception  
    - Hernia  
  - Child and older
    - Appendicitis
What Can We See Radiographically?

Let’s Dig Into This Further...
Question 2: What clinical history should prompt an emergent upper GI series?

A. Failure to pass meconium
B. Abdominal distention
C. Blood in stool
D. Bilious emesis
Malrotation with Midgut Volvulus

- Intestinal malrotation = Failure of rotation and fixation of bowel early in the embryo
- Short fixation of mesentery $\rightarrow$ susceptible to volvulus around a vascular stalk
- Congenital adhesions (Ladd’s Bands) are associated
  - Extend from cecum to liver
  - Externally compress the duodenum

Radiographs: Malrotation with Midgut Volvulus

- Normal
- Double-Bubble Sign
- Paucity of Bowel Gas
- Air-Fluid Levels
• NORMAL Upper GI Series:
Ligament of Treitz (DJ junction) should be:
1. Left of Spine
2. Level of pylorus

Upper GI Series: Malrotation
Upper GI Series: Malrotation with Midgut Volvulus

Malrotation with Midgut Volvulus: Treatment with Ladd Procedures

1. The bowel is untwisted.
2. Ladd’s bands are cut.
3. Mesentery is spread out.
4. Small bowel is placed in right abdomen.
5. Large bowel is placed in left abdomen.
6. Appendix is removed.

Distal Bowel Obstruction: Clinical and Radiographic Presentation

- Abdominal distention
- Vomiting – may be bilious
- Absence of bowel gas to rectum within 24 hours
- Failure to pass meconium within 48 hours

If Distal Bowel Obstruction Is Suspected: Water-soluble Contrast Enema

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Question 3: Which of the following enema findings suggests Hirschsprung Disease?

A. Microcolon
   - Implies lack of colonic distention during fetal development
   - Differential Considerations:
     - Jejunal or ileal atresia
       - Congenital absence of a segment of jejunum or ileum
       - Prognosis depends on amount of residual functional bowel
     - Meconium ileus
       - Obstruction of the neonatal small bowel by abnormally thick meconium.
       - Patients virtually always have cystic fibrosis.
       - Complication: Perforation and Peritonitis

B. 

C. 

D. 

Normal Contrast Enema

- Normal study
  - Rectum morphology → shouldered margin
  - Rectal diameter → larger than sigmoid colonic diameter
  - No caliber narrowing
  - Cecum identified in RLQ

Hirschsprung Disease

- Synonymous with colonic aganglionosis → Lack of intrinsic enteric ganglion cells causes a functional obstruction of bowel
- The denervated colon is small and spasmodic
- Variable length of affected colon always includes the anus
- 90% of cases are diagnosed in the newborn
- 10-15% of patients with HD have trisomy 21
**Small Left Colon Syndrome / Meconium Plug Syndrome**

- A functional immaturity of the colon
- “Meconium plug” syndrome is a misnomer, or at least confusing – different from meconium ileus!
- Etiology is unknown, but seen in hypotonic states:
  - Infants of diabetic mothers
  - Infants of mothers treated with magnesium sulfate for eclampsia
- May relate to immaturity of receptors in bowel
- Transient -- prognosis is excellent

**Do Upper Tract and Distal Bowel Obstruction Coexist?**

Yes!

What approach do we take?

Upper GI series first, then enema.
Question 4: A 4-week-old infant presents with non-bilious emesis and weight loss. What imaging study is appropriate?

A. Abdominal radiograph
B. Upper GI series
C. Limited abdominal ultrasound
D. None needed, surgical consult

Question 4: Non-bilious emesis + weight loss in first 5 weeks of life

Suspect this diagnosis:
Hypertrophic Pyloric Stenosis

• Narrowing of pyloric channel by hypertrophied muscle → hypothesized that Helicobacter pylori is responsible
• Male to female ratio is 4:1
• Presents in weeks 3-5 of life with non-bilious emesis, poor weight gain, dehydration
• Extreme cases: palpable “olive” or small mass at lateral margin of right rectus muscle below liver edge

• Clinical differential diagnosis
  – Malrotation with volvulus: bilious emesis
  – Gastroesophageal reflux: weight gain

Hypertrophic Pyloric Stenosis
Sonographic Diagnosis

• Scanning technique
  – Transverse-oblique positioning of transducer, parallel to right costal margin
  – Use liver as the acoustic window
  – Slight downward angling of transducer
• Measurements:
  – thickness of pyloric wall
  – length of pyloric channel
• \[ \pi = 3.14 \]
  – Thickness > 3 mm
  – Channel > 14 mm
Normal Gastric Pylorus

Normal vs Hypertrophied Gastric Pylorus
Question 5: An infant with abdominal distention and this radiograph most likely has what sort of history?

A. Trisomy 21  
B. Prematurity  
C. Bilious emesis  
D. Anal atresia

Pneumatosis in an infant: Necrotizing Enterocolitis

- Typically presents in premature baby <1000 grams
- 1-3 weeks of age
- Enterally fed
- Pearl: If NEC occurs in premature infant within 24 hours, consider hypoplastic left heart syndrome and recommend emergent pediatric cardiology consultation.
Pneumatosis in an infant: Necrotizing Enterocolitis

• Clinical Presentations:
  – Abdominal Distention
  – Feeding Intolerance
  – Bloody Stools
• Free Air means surgery
• Abdominal radiographic surveys acquired every 6-8 hours

Pneumatosis:
Cystic and Linear Lucencies
Along Curvilinear Path
Pneumatosis: Cystic and Linear Lucencies Along Curvilinear Path
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Question 6: A two-year-old girl with abdominal pain undergoes sonographic evaluation. What is the diagnosis?
Question 6: A two-year-old girl with abdominal pain undergoes sonographic evaluation. What is the diagnosis?

A. Hypertrophic pyloric stenosis
B. Hepatoblastoma
C. Perforated appendicitis
D. Intussusception
E. Rhabdomyoscaroma

Intussusception

• Acquired telescoping of bowel into itself
  – Idiopathic -- hyperplasia of Peyer’s patches thought to be the culprit; ileocolic or ileoileocolic
  – Lead point: Congenital or acquired mass
• Ileocolic intussusception is the most common cause of SBO
• Delayed diagnosis leads to...
  – Bowel obstruction
  – Venous congestion
  – Bowel necrosis and perforation
Intussusception

- Clinical Presentation
  - Peak age of 6 months - 3 years
  - Fussy, intermittent crying, pulling legs up, vomiting
  - Classic triad of colicky abdominal pain, palpable abdominal mass, and current jelly stools is seen less than 50% of the time

- Diagnosis
  - X-ray
  - Ultrasound
  - Enema
  - Incidentally found on CT and MR imaging

Intussusception and Radiography

- Poor sensitivity (45%) for detection of intussusception
- Useful to exclude other entities such as constipation
- Useful to exclude pneumoperitoneum
- Classic x-ray findings:
  - Crescent sign
  - Absence of bowel gas in the ascending colon
  - PITFALL: Gas in small bowel or in sigmoid colon may be present in right lower quadrant
Intussusception and Ultrasound

• Identification of intussusception
  – High accuracy, nearly 100% in experienced hands
  – Sensitivity of 98% and Specificity of 88-100%

• Accurate Diagnosis:
  – Mass is large (usually 3-5 cm)
  – Usually found in right abdomen, but may be anywhere
  – Transverse scan: eccentric semilunar hyperechoic fat and sometimes lymph nodes

• Pitfalls:
  – Any cause of bowel thickening may fool some eyes
  – Stool and psoas muscle might trick the inexperienced technologist or radiologist

Ileocolic Intussusception
Ileocolic Intussusception

Intussusception and Ultrasound: Assessment of Reducibility

Sonographic findings of bowel necrosis and other potential limitations in reduction:

- Little blood flow within bowel walls
- Thick peripheral hypoechoic rim
- Free intraperitoneal fluid
- Trapped lymph nodes (2 or more, > 11 mm)
- Lead point
- Trapped fluid within the lumen between bowel loops
Ileocolic Intussusception: Oligemia

Intussusception with Lead Mass
Question 7: What tissue type is visualized during Meckel diverticulum scintigraphy?

A. Ectopic gastric mucosa
B. Ectopic pancreas
C. Tagged red blood cells
D. Tagged white blood cells
Meckel Diverticulum: Cause of lower gastrointestinal bleeding

• Meckel diverticulum -- most common cause of lower GI hemorrhage in previously healthy infants.
• More than 50% of these patients present with bleeding by the age of 2 years.
• Meckel diverticulum = vestigial remnant of the omphalomesenteric duct → the most common congenital anomaly of the gastrointestinal tract, with an incidence of 1%–3% in the general population.
• It is normally located on the antimesenteric border of the terminal ileum within 80–100 cm of the ileocecal valve and is on average 2 cm in length.

Meckel Diverticulum

• 50-60% of Meckel diverticula contain ectopic gastric mucosa (even higher percentage in children with bleeding) → actively secretes the hydrochloric acid responsible for mucosal ulcerations
• The most common sign of Meckel diverticulum is gross rectal bleeding.
Meckel Diverticulum: 99mTc-pertechnetate Scan

- 99mTc-pertechnetate is taken up by the mucin-producing cells of gastric mucosa and is then secreted into the gut lumen.
- Avid accumulation of 99mTc-pertechnetate in gastric mucosa → scintigraphy with 99mTc-pertechnetate the study of choice for identifying ectopic gastric mucosa in a Meckel diverticulum.

Meckel Diverticulum: 99mTc-pertechnetate Scan

- Pre-examination fasting of 3–4 hours may reduce the size of the stomach and improve sensitivity for the detection of ectopic gastric mucosa.
- Pretreatment options include the following:
  - Histamine H2 blockers (cimetidine, ranitidine, famotidine)
  - Proton pump control acid secretion
  - Glucagon relaxes smooth muscles and slightly suppresses peristalsis and transit of any secreted 99mTc-pertechnetate through the small bowel.
Meckel Diverticulum: $^{99m}$Tc-pertechnetate Scan

**Question 8:** What non-heritable association/syndrome should be considered with this radiograph?

A. Trisomy 13
B. Turner syndrome
C. VACTERL
D. CHAOS
Tracheoesophageal Fistula & Esophageal Atresia

- Esophageal atresia with or without tracheoesophageal fistula incidence is 1 in 3,000-4,000 live births
- Most commonly, proximal esophageal atresia with a distal tracheoesophageal fistula

http://www1.umn.edu/eatef/whatis.html

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<th>Diagnosis</th>
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<td>Multiple malformation</td>
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<td>Total</td>
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Radiographic Assessment

- Bowel gas?
- Vertebral anomaly?
- Cardiac silhouette?

What is VACTERL/VATER?

A non-random association --
At least THREE of the core anomalies required for diagnosis.

V = vertebral anomalies
A = anorectal malformation
C = cardiac anomalies
T = tracheoesophageal fistula
E = esophageal atresia
R = renal anomalies
L = limb anomalies
Question 9: Images from a third-trimester fetal ultrasound and fetal MR imaging study are shown.

Question 9: Which of the following is the most important differential consideration?

A. Mesenchymal hamartoma
B. Congenital hemangioma
C. Hepatocellular carcinoma
D. Hepatoblastoma
Hepatoblastoma

- A malignant tumor, usually presents by age 5...
  - No definite association with chronic liver disease
  - Genetic conditions associated with this tumor:
    - Beckwith-Weidemann (hemihypertrophy syndrome)
    - Familial adenomatous polyposis
  - Tend to be single masses, right lobe predominance
  - Coarse calcifications are classic
- Laboratory evaluation: elevated AFP
- Lung is the most common site of metastases, followed by portal / periaortic lymph nodes
- Resectability is the most important prognostic factor

Mesenchymal hamartoma

- A rare benign developmental anomaly
  - Multicystic mass
  - Disorganized collection of mesenchyma, bile ducts, hepatocytes, and hematopoietic cells
- Median age of presentation is 1 year
Congenital hemangioma

• Infantile Hemangioma
  – Small (and may be undetectable) at birth, grow rapidly in infancy, then involute gradually over childhood
  – Glucose Transporter 1 Positive (GLUT1-positive)
  – Usually multifocal or diffuse

• Congenital Hemangioma
  – Fully grown at Birth
  – GLUT1-negative
  – Rapidly Involuting (RICH), Non-Involuting (NICH), or a mixed pattern
  – Solitary masses

The term “Infantile Hemangioendothelioma” is NO LONGER IN FAVOR

  – Kaposiform hemangioendothelioma and epithelioid hemangioendothelioma are distinct pathologic entities from hepatic hemangioma
  – Lesions previously called “hemangioendothelioma” may, in fact, be a form of angiosarcoma.
**Congenital hemangioma**

- A large benign hemangioma ...  
  - May cause high-output cardiac failure due to arteriovenous shunting  
  - May cause respiratory distress due to large size  
- Suspect this etiology for a solitary liver mass in an infant with negative alpha fetoprotein (AFP)  
- On contrast-enhanced imaging, mass shows interrupted contrast pooling peripherally with gradual enhancement centrally  
- $^{99m}$Tc-labeled RBC scan is highly specific

**Hepatocellular carcinoma**

- Not the right choice because carcinomas do not occur in the fetus or infant  
  - Fetal tumors (in general):  
    - Teratomas  
    - Sarcomas  
    - -blastomas  
- Hepatocellular CA  
  - Most common type of liver cancer in adults  
  - Secondary to viral hepatitis or cirrhosis
Summary

• We have looked at examples of upper and lower intestinal tract obstruction and significance of bilious emesis.
• Pneumatosis is difficult and important to recognize.
• Intussusception is uncommonly diagnosed by radiograph and ultrasound is useful to identify a 3 – 5 cm mass with alternating layers.
• Evaluate bloody stool in an otherwise healthy, asymptomatic infant with $^{99m}$Tc scintigraphy.