PEDIATRIC SEMINAR

Congenital Cystic Abdominal Lesions

Thank you to Angela Levy, MD and David Biko, MD for their contributions to this presentation
Illustrations by Aletta A. Frazier, MD
GOALS

- Discuss Location, Origin, and Differential
- Build knowledge on Common Entities
- Expose some Rare Syndromes
- Learn Treatment Options and future Implications when possible
Congenital Cystic Abdominal Lesions

• Arise from any organ system
  – Gastrointestinal
    • Bowel obstruction, duplication cyst, Meckel’s diverticulum, meconium pseudocyst
  – Hepatobiliary
    • Choledochal cyst, liver cyst
  – Genitourinary
    • Pelviectasis (obstructive or nonobstructive), renal dysplasia, urachal cyst, ovarian cyst, hydrometrocolpos, cloacal malformations
  – Other
    • Lymphangioma
Regional Diagnostic Considerations

RUQ:
- Hepatic cysts
- Choledochal cysts
- Adrenal lesions
- Renal lesions

RLQ:
- Meckel’s
- Bowel/Urinary Tract
- Ovarian Cyst

LUQ:
- Atresia
- Renal lesions
- Adrenal lesions
- Extrapulmonary Sequestration

LLQ:
- Bowel/Urinary tract
- Ovarian cyst

Anywhere:
- Duplication cysts, Urinary Tract
- Lymphatic Malformation, Pseudocyst
- Congenital Tumor
Left Ovarian Cyst on Screening Exam
CASE 1
3 year old with vomiting and pain
Diagnosis?

1. Lymphangioma
2. Enteric Duplication Cyst
3. Abscess
4. CSFoma
5. Meconium Pseudocyst
Duplication Cyst - Location

- Ileum - 40%
- Thorax – 20%
- Jejunum – 10%
- Stomach - 10%
- Colon – 10%
- Multiple – 5-10%
Duplication – Clinical Features

- Thoracic – respiratory symptoms, incidental finding
- Abdominal
  - Incidental on prenatal US
  - Obstruction
  - Mass
  - Pain
  - GI bleeding
Enteric Duplication Cyst

- Developmental tubular or cystic structures adjacent to the GI tract
- Usually round and do not communicate with bowel lumen
- Occasionally tubular, communicating with GI tract at one end and blind-ending at the other
Duplication Cyst - Pathology

- Often share muscularis layer with adjacent bowel (intramural)
- Also share blood and nerve supply
- Mesenteric border
- Filled with mucoid material
- Histologically, recapitulate normal GI tract
- May contain ectopic gastric mucosa (20%) or pancreatic tissue
Communicating tubular
Duplication Cyst - Imaging

- Plain film – as soft tissue mass if large
- Ultrasound
  - Preferred study
  - “Rim” sign – gut signature in the wall
  - May contain debris
Duplication Cyst

prenatal

postnatal
Treatment

• Surgery
CASE 2:
Abdominal pain
<table>
<thead>
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<th>Diagnosis</th>
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<td>1</td>
<td>Ovarian Cyst</td>
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<td>Giant Meckel</td>
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<td>5</td>
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Which of the following may have increased activity on a 99mTc-pertechnate scan?

1. Ovarian Cyst
2. Meckel Diverticulum
3. Enteric Duplication Cyst
4. Meconium Pseudocyst
5. Both 2 and 3
Giant Meckel Diverticulum

- Most common anomaly of the GI tract
  - Short tubular structure but may be giant resembling a cyst
- Omphalomesenteric duct anomaly
- 2% - 3% of the population
- 45% present under age 2
- Within 2 feet of ileocecal valve
- About 2 inches long
- 2% present with complications
- Giant if greater than 5-6 cm
Omphalomesenteric (Vitelline) Duct

- Embryonic connection between yolk sac and midgut
- 10th week of embryogenesis
  - Midgut returns to abdomen
  - Duct is a thin fibrous band connecting midgut to umbilicus
  - Disintegration
  - Absorption-failure-may continue to grow
Omphalomesenteric (Vitelline) Duct Anomalies

- Umbilico-ileal fistula
Omphalomesenteric (Vitelline) Duct Anomalies

- Umbilico-ileal fistula
- Umbilical sinus
- Umbilical cyst
Omphalomesenteric (Vitelline) Duct Anomalies

- Umbilico-ileal fistula
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- Persistent fibrous cord
Omphalomesenteric (Vitelline) Duct Anomalies

- Umbilico-ileal fistula
- Umbilical sinus
- Umbilical cyst
- Persistent fibrous cord
- Meckel diverticulum
  - With a fibrous cord
  - With a portion of mesentery
Meckel Diverticulum - Presentation

• Most commonly become symptomatic in first years of life; 60% before reaching 10 years of age.

• Present due to complications
  – **Bleeding** – most common-”brick red”, “currant jelly”
  – **Obstruction** – usually due to inverted Meckel serving as lead point for intussusception
  – **Diverticulitis**- F/nausea/vomiting
  – **Vitelline band** can serve as a fulcrum for volvulus
  – **Littre hernia**- Meckel’s entrapped in a hernia-50% inguinal region
Meckel Diverticulum Pathology

- Antimesenteric side of distal ileum
- True diverticulum - all layers of the small bowel wall
- Most discovered incidentally are lined by small bowel epithelium
- Heterotopic tissue
  - 50% of resected diverticula
  - Gastric most common (23% - 50%)
  - Pancreas (5% to 16%)
  - Rare - Brunner glands, colonic, biliary
Meckel Diverticulum
Pathology
Meckel Diverticulum Pathology
Meckel Diverticulum
Heterotopic Pancreatic Mucosa
Meckel Diverticulum - Imaging

- Plain film –
  - May be visible if giant – mottled air collection
  - May be filled with air or enterolith
Meckel Diverticulum - Imaging

• Ultrasound
  – Gut signature – resembles normal bowel
  – When inflamed, mimics appendicitis
Hemorrhage in Meckel Diverticulum

- Most frequent complication
- Tc$^{99m}$-pertechnetate
  - Localizes in ectopic gastric mucosa
  - Modality of choice in pediatric population
  - Sensitivity 85%, specificity 95% in kids
  - Sensitivity 63%, specificity 2% in adults
Small Bowel Obstruction due to Meckel Diverticula

• Second most common complication of Meckel

• Etiology
  – Inversion with intussusception
  – Diverticulitis
  – Volvulus from attachment to umbilicus
  – Congenital mesodiverticular bands
  – Foreign body impaction
  – Inclusion of Meckel in a hernia (hernia of Littre)
  – Neoplasm
Inflamed Meckel with Small Bowel Obstruction
Meckel Diverticulum - Imaging

- CT
  - Blind-ending pouch
  - Connection to ileum
Meckel Diverticulitis
Etiology

- Luminal obstruction
  - Enterolith
  - Foreign body
  - Edema of orifice
- Peptic ulceration
- Torsion
Meckel Diverticulitis - CT

- Blind-ending pouch
- Mural contrast enhancement
- Connection to ileum
- Midline location
- Associated SBO
Treatment

• Surgery
CASE 3: 2 year old with protruberant abdomen
CASE 3: 2 year old with protruberant abdomen
Diagnosis

1. Ovarian Cyst
2. Lymphangioma
3. Enteric Duplication Cyst
4. Giant Meckel Diverticulum
5. Meconium Pseudocyst
Lymphangioma-Clinical Presentation

- May be discovered incidentally if small
- May be pain, palpable mass, anorexia, vomiting or fever
- More commonly occurs in the head and neck-95%
- Mesentery, retroperitoneum, lung, and mediastinum-5%
- Lymphangiomatosis-rare, multifocal parenchymal organ involvement
Lymphangioma - Pathology

- Benign lesions of vascular origin
- Thin walled cystic masses
- Macroscopic versus microscopic
- Contains chylous, serous, and/or hemorrhagic fluid
- Histopathologically dilated lymphatic spaces
Lymphangioma - Imaging

- US
  - Multilocular cystic masses
  - +/- debris

No gut signature on ultrasound
Lymphangioma - Imaging

- **CT/MRI**
  - Homogenous fluid
- **MRI:** Low T1, high T2
- Changes if hemorrhage or infection
  - Cyst wall or septa may enhance.
Treatment

- Surgery
CASE 4: Newborn who presents for evaluation of line placement
Diagnosis

1. Ovarian Cyst
2. Lymphangioma
3. Enteric Duplication Cyst
4. Giant Meckel Diverticulum
5. Meconium Pseudocyst
Meconium Pseudocyst—Clinical presentation

• Results from in utero bowel perforation
  – May have underlying disorder such as bowel atresia or meconium ileus
  – Extruded bowel contents provoke peritoneal inflammation and rapidly calcify
  – May form cyst
Meconium Pseudocyst-Clinical Presentation

• If perforation never heals-pseudocyst forms.
• Born with abdominal distension with or without obstruction
Pathology
Meconium Pseudocyst Imaging

• Radiograph
  – Well calcified intra-abdominal mass
  – Scattered calcifications due to meconium peritonitis
Meconium Pseudocyst Imaging

- **Ultrasound**
  - Primary imaging modality pre- and post-natal
  - Thick well-circumscribed echogenic cyst walls
  - May have echogenic viscous contents
  - May see dilated bowel due to obstruction
Meconium Pseudocyst Imaging

- CT:
  - Large fluid filled cyst-usually no septations
  - Calcified, thick rim
  - Nonenhancing
Meconium Pseudocyst – Imaging

- MRI
  - High T1 and T2 signal
  - Rim of low signal
Treatment

- Surgery
CASE 5: Female fetus, prenatal evaluation
Case 5:
Postnatal water soluble enema
What is this syndrome associated with?

1. Wilm’s Tumor
2. Malrotation
3. Lymphangioma
4. Posterior Urethral Valves
5. Choledochal Cyst
Megacystis-Microcolon-Intestinal Hypoperistalsis Syndrome (MMIHS)  
Berdon Syndrome

- Rare congenital anomaly (182 cases described)
  - Abdominal distention due to distended non-obstructed bladder
  - Microcolon
  - Decreased or absent intestinal peristalsis
  - May look prune-belly like

Females 3:1 Males, but males more severe
Pathology

- Etiology still unknown
- GU system enlarged, obstructive picture
- Hard to understand uniting cause of autonomic nerve dysfunction to both GI and GU
- Colon looks Hirschsprung-like, but + ganglion cells—actually, hypertrophic
- Vacuolar degeneration bowel and bladder smooth muscle
• Fetal MRI
  – Enlarged bladder
  – Hydronephrosis
  – No meconium visualized within the bowel
    • Rectum should have meconium (high T1) @ 20-24 weeks
  – polyhydramnios
- BE shows microcolon
- Eventually chronically dilated
MMIHS - Associations

- Short bowel syndrome (20%)
- Malrotation (45%)
- Undescended testes
- Omphalocele
- Cardiac anomalies
Prognosis/Treatment

- Usually lethal in the first year despite enteric tubes and multivisceral transplantation
- Parenteral nutrition
CASE 6:
1 year old female with urinary tract infection
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CASE 6:
1 year old female with urinary tract infection
CASE 6:
1 year old female with urinary tract infection
What species may normally possess a spectrum of this abnormality?

1. Rats
2. Dogs
3. Cats
4. Chickens
5. Horses
Persistent Urogenital Sinus

- **Spectrum of cloacal malformation**
  - Cloaca – Latin for “sewer”
    - Common channel for urinary, genital, and distal GI tract
- **Only in phenotypic girls**

**Cloaca** – urinary, genital, and GI tracts not separate

**Urogenital Sinus** – Urinary and genital tracts not separate

**Normal Division** – Urinary, genital, and GI tracts all separate
Embryology - Cloaca

- Common channel of the hindgut and allantois
- 6 weeks
  - Divides into urogenital sinus anteriorly and hindgut posteriorly
- 8 weeks
  - Mesonephric duct is absorbed
  - Paramesonephric ducts form upper 1/3 vagina

Illustrations by David Humphrey, MD, MBA
Embryology

- Differentiate from Exstrophy of the cloaca
  - failure of the lower abdominal wall closure
    - males and females
Pathology

- Sigmoid colon
- Hemiuterus/FT
- Bladder
Cloaca - Pathology

- Resection with reproductive, urinary and GI tract with common opening
- Associated with spinal cord and bony pelvis anomalies
Treatment

• Early intervention
  – Divert GI tract
  – Urinary tract drainage/check renal function
  – Internal vaginal and urethral anatomy-scope
    To measure length of common channel

**SHORT CHANNEL** (<3cm)-posterior sagittal anorectovaginourethroplasty (PSARVUP)

**LONG CHANNEL** - more complicated
Conclusion

- Presentation of a congenital cystic lesion is often nonspecific or lesion may be incidental

- Differential diagnosis for congenital cystic lesion can be variable
  - Categorize cystic lesion into location and organ system to limit differential
  - Ultrasound is most important imaging tool to help differentiate these lesions