Differential Diagnosis of Cystic Abdominal Masses

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Disclosure

- Medical Editor Amirsys Content, Elsevier

Learning Objectives

- Form a more specific differential diagnosis using
  - Location
  - Imaging appearance
  - Fetal gender
  - Gestational age at presentation

Axial
Coronal

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Embryology

- Stages of development similar to those found in more primitive animals
- 3 successive nephric structures of increasing advanced design
  - Pronephros, mesonephros, metanephros
- The 3rd, and definitive, kidney forms when ureteric bud induces metanephrinic blastema to form metanephros

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Embryology

- Ureteric bud and metanephric blastema exert reciprocal inductive effects on each other
- Ureteric bud induces metanephric blastema to form nephrons
- In turn, metanephric blastema induces ureteric bud to bifurcate into developing calyces

Multicystic dysplastic kidney

- Multiple non-communicating cysts
- Reniform shape often lost
- Bilateral in 20%
- Contralateral renal anomaly (non-MCDK) in 40%
- Can enlarge dramatically
  - Follow size and amniotic fluid

Cystic Abdominal Mass DDx

- GU
  - UPJ Obstruction
Obstructive renal dysplasia
- Tubular destruction and cyst formation due to increased pressure in collecting system
- Most commonly posterior urethral valves
  - UPJ, UVJ obstruction less common

Obstructive renal dysplasia
- Imaging appearance variable
  - Hyperechoic kidney
  - Loss of corticomedullary distinction
  - Cortical cysts
  - Hydronephrosis
- Renal size may be ↑, ↓, or normal
  - ↓ size suggests late finding (poor prognostic sign)

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Cystic Abdominal Mass DDx
- Duplicated collecting system ureterocele

Cystic Abdominal Mass DDx
- Enlarged Bladder
  - Posterior urethral valves
  - Look for “keyhole”
  - Prune belly syndrome
  - Urethral atresia
Cystic Abdominal Mass DDx

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Prune Belly Syndrome

- 3 components
  - Dramatic collecting system dilatation, including urethra
    - No keyhole sign
  - Deficiency of abdominal musculature
  - Cryptorchidism

Cystic Abdominal Mass DDx

GU
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  - Intraabdominal portion of allantois
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Cystic Abdominal Mass DDx

Ovarian Cyst

- Most common abdominal cyst in female fetus
- Most occur in 3rd trimester
- Off midline
- Unilateral or bilateral
- “Daughter cyst” diagnostic
- Normal GI and urinary tracts

Complex
- Concern for hemorrhage or torsion
- Septated
- Fluid-fluid level
- Solid-appearing
Cystic Abdominal Mass DDx

**Ovarian Cyst**
- Most show substantial regression by 6 months of age
  - 64% complete resolution if simple
  - 40% if complex
- Complex cysts more likely to require excision

**GI**

**Bowel Atresia**
- Can occur anywhere along gastrointestinal tract
- Distal atresias have a tubular, "sausage-shaped" appearance
- Peristalsis within a cystic mass is pathognomonic

**Meconium Pseudocyst**
- Wall-off bowel perforation
- Irregular, thick walls
- Look for other signs of meconium peritonitis
  - Intraperitoneal calcifications
  - Dilated bowel
  - Ascites

**Enteric Duplication Cyst**
- Solitary, thick-walled cyst
  - Round or elongated
- Look for "gut signature"
  - Layered appearance with echogenic mucosa, hypoechoic muscular layer, echogenic serosa
  - Rarely bowel dilatation from obstruction
Cystic Abdominal Mass DDx

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- Choledochal Cyst
  - Cystic dilatation of extrahepatic bile duct
  - Unilocular, simple, right upper quadrant cyst
  - Round in axial plane and fusiform in longitudinal plane
  - Following bile ducts into cyst confirms diagnosis

- Lymphangioma (mesenteric cyst)
  - Lymphovascular malformation
  - Thin-walled cyst
  - May be unilocular or multilocular, with one or multiple septations
  - Can be very complex, insinuating around organs and extending out of abdomen
Cystic Abdominal Mass DDx

- Lymphangioma (mesenteric cyst)

- Fetus-in-Fetu
  - Complex, with a large solid component encapsulated within a cyst
  - Calcifications, including well-formed bones, most specific finding

- Neuroblastoma
  - Most common congenital malignancy
  - 90% occur in fetal adrenal
  - Varied appearance
    - Solid
    - Partially cystic
    - Completely cystic
      - Probably involuting
      - Excellent prognosis

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  - Rare benign cysts which generally involute postnatally

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Cloacal Fun Facts

- Cloaca is Latin for sewer
  - Cloaca maxima is sewer system of ancient Rome

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- Birds are most common animal to have a cloaca

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Sharks and rays have a cloaca

Cloacal Fun Facts

Montremes (Greek for single hole) have a cloaca
- platypus and spiny anteater

Cloacal Fun Facts

Cloacal Kiss

Mel: Are those birds rubbing their butts together?

Nico: No they’re just giving each other a cloacal kiss. The male and female press their cloacals, a posterior opening, to mate.

Mel: I’d rather French kiss
Cloacal Fun Facts

Do birds pee?? I have never seen one do so, just wondering?

Bird urine and poop come out of the same area - it's mixed together, which explains the liquidy, gooey mess. Even more annoying, the eggs that are laid come out of the same hole...

Luckily, eggs are washed off before they get to the store.

Cloacal Fun Facts

Not so fun fact

- In only 6% of prenatal cases with a cloaca was the diagnosis suspected
- Retrospective review showed suspicious findings in 62%


Anorectal Malformations

- Complex group of anomalies involving the distal anus and rectum and frequently urinary and genital tracts
- Classified by position of rectal pouch relative to puborectal sling and presence/absence of fistulas
- More common in males
- Cloacal malformation special class of ARM
- Communication between bladder, vagina and rectum with single external orifice
- Exclusively in females
- 1 per 20,000 live births


Embryology
By 5 weeks the cloaca is a common chamber receiving the intestinal, genital and urinary tracts.

Cloaca is then divided by the urorectal septum (URS) into:
- Urogenital sinus (U) anteriorly
- Anorectum (A) posteriorly

Cloacal membrane dissolves once it comes in contact with the urorectal septum.

Urogenital sinus divides to form:
- Urinary bladder and urethra anteriorly
- Portion incorporates into lower portion of vagina and hymen posteriorly

Separation complete by about 6 weeks gestational age.
**Cloacal malformation**

- Failure of cloacal division
- Classic cloaca
  - 1 perineal orifice: common channel connecting the urethra, vagina and rectum
  - May have variants
- Seldom isolated finding
- Other GU/GI tract and lumbosacral anomalies common

**Cloacal malformation**

- Cystic pelvic mass with fluid-debris level
- Result of mixing urine with vaginal secretions or meconium
- Vagina often most distended structure
- Uterine and vaginal duplication in 60%
- Creates longitudinal septated cystic mass

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No rectum, smooth perineum

Vagina

Bladder

Ch saca
**Key Findings**

- Cystic pelvic mass *especially if vertical septum and/or fluid-debris level*
  - Midline location
  - Funnels to perineum
  - Urinary tract abnormalities
  - Oligohydramnios
  - No anal dimple
  - Lumbosacral anomalies
  - Bowel dilatation
Cystic Abdominal Mass DDx

- Bladder outlet obstruction
- MCDK
- UPJ obstruction
- Urinoma
- Renal duplications
- Patent urachus

- Ovarian Cyst

- Bowel atresia
- Meconium pseudocyst
- Enteric duplication
- Choledochal cyst
- Splenic cyst
- Lymphangiomma
- Fetus-in-fetu
- Neuroblastoma

- GI Tract
- Genital Tract
- Misc.