Complex fetal genitourinary anomalies-how can MRI help?

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Goals & Objectives

• To review prenatal imaging approach to assess GU anomalies

• To discuss the differential diagnosis in the setting of megacystis and absent bladders
  – (most frequent scenarios for potential underlying complex GU malformations)
PRENATAL IMAGING APPROACH
Background

- Fetal urine production starts at 8-10 weeks’ gestation

- The fetal bladder will first be seen at around 10-12 weeks (diameter: no more than 6-8 mm)

- Even in the presence of severe GU anomalies, usually amniotic fluid volume is normal in the 1st Trimester
• Congenital GU abnormalities are common (14-40% of prenatal US abnormalities detected): broad spectrum from mild to severe

• Severe GU abnormalities will most likely present amniotic fluid volume changes, megacystis or other major associated malformations including abdominal wall and spinal defects.
  
  – In other cases, the findings are more subtle (high index of suspicion + improved knowledge of potential associations + Fetal MRI will help)
US Imaging Targets/check list

– Accurate assessment of amniotic fluid volume
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- Evaluation of the umbilical arteries to define megacystis as opposed to other abdominal cyst/hydrocolpos
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- Abnormal content in the bladder /hydrocolpos/bowel? (anorectal malformation)
- Wall defect and/or absence of bladder with normal AFI
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- Wall defect and/or absence of bladder with normal AFI
- External genitalia: Gender/ambiguous/incompletely formed
US Limitations

- In the setting of oligohydramnios, US imaging can be challenging
- Cystic renal dysplasia can be difficult to detect in early stages
- Ectopic vs. absent kidney (Color Doppler can help)
- Anorectal malformations
Fetal MRI Imaging Targets

- Kidneys

- Bladder/posterior urethra (bladder cycles, potential dilatation of Posterior Urethra)/ infraumbilical abdominal wall

- External genitalia

- Spine

- Calculation of lung volumes (3rd trimester)

- Bowel: Anorectal region/colon
Fetal MRI: Assessment of the colon

- After 20w we expect to see meconium filled rectum.
Fetal MRI: Assessment of the colon

- Around the 27w the whole colon is filled with meconium

- Assessment for microcolon (Megacystis microcolon intestinal hypoperistalsis syndrome)
Fetal MRI: Assessment of the colon

- The rectum is close to the bladder and its cul-de-sac at least 10mm below the bladder base

(Saguintaah M. et al. (2002) Ped Radiol)
Fetal MRI: Assessment of the colon

- Long channel cloacas have:
  - Dilated rectum
  - High cul-de-sac

(Calvo-Garcia M.A. et al Ped Radiol 2011)
Fetal MRI: Assessment of the colon

- Cloaca

and imperforate anus with RU fistula:

- Can present rectal dilatation with fluid content and enteroliths. 
Fetal MRI: Assessment of the colon

- **Cloacal exstrophy:**
  - Absent meconium in the rectum/colon

Fetal MRI: Assessment of the colon

- **Bladder exstrophy:**
  - Normal meconium in the rectum/colon
DIFFERENTIAL DIAGNOSIS
Etiology of Megacystis

- **Bladder obstruction**: Overtime oligohydramnios

- **Non-obstructive bladders**: not true or persistent mechanical obstruction. Amniotic fluid usually normal, and in some cases, increased.
Etiology of Megacystis

- **Bladder obstruction:** (overtime oligohydramnios)
  - *Males:*
    - Posterior urethral valves
    - Urethral atresia (early presentation)
    - Complex anorectal malformations
  
  - *Females:*
    - Urethral atresia
    - Cloacal malformations
  
  - *No gender specific:*
    - Extrinsic or intrinsic pathology leading to obstruction:
      - SCT with BOO/ Everted ureterocele
Etiology of Megacystis

- **Non-obstructive bladders** (Amniotic Fluid usually normal, sometimes increased)

  - *Prune Belly Syndrome (PBS)*, more frequent in males

  - *Megacystis Microcolon Intestinal Hypoperistalsis Syndrome (MMIHS)*, more frequent in females. Common development of poly after 30 weeks (presumably owing to GI malformation associated)

  - *Megacystis-megaureter association* (No gender specific-severe vesicoureteral reflux)
Etiology of Non-visualization of the Fetal Bladder

- **Lack of fetal urine production/obstruction**
  - oligo/anhydramnios (maybe a small bladder present)

- **Inability of the bladder to store urine** (no visible bladder)
  - normal amniotic fluid
Etiology of Non-visualization of the Fetal Bladder

- **Lack of fetal urine production/obstruction**
  - oligo/anhydramnios (maybe a small bladder present)

- Pre-renal failure (IUGR): we should see kidneys

- Renal (bilateral renal agenesis, Bilateral MCDK, Bilateral renal dysplasia)
  - In that situation you might encounter anorectal malformations as end-stage bladder outlet obstruction!!!!
Etiology of Non-visualization of the Fetal Bladder

- **Inability of the bladder to store urine** (no visible bladder): normal amniotic fluid
  - **Infraumbilical wall defect**
    - Bladder extrophy: usually normal rectum and spine
    - Cloacal extrophy (OEIS): “elephant trunk-like” image sometimes (but not always!)
Etiology of Non-visualization of the Fetal Bladder

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Nyberg “Diagnostic Imaging of Fetal Anomalies page 536”
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  - **No wall defect is seen but low-set umbilicus** (+ males with short, broad penis):
    - Epispadias (In both males/females the bladder neck is often inadequate: urinary dribbling)

- **If no malformations seen:**
  - Bilateral single ectopic ureters
Key Points

• **Megacystis**
  – Enlarged bladder versus other cystic lesions
    • Relationship with umbilical arteries
  – Assess bladder and adjacent bowel content
  – Always check colon/rectum (fetal MRI)
  – AFV: Oligohydramnios/polyhydramnios

• **Absent bladder**
  – AFV: normal versus decreased
  – Wall defect /low ACI/ prolapsed terminal ileum
  – +/- meconium in colon/rectum
Selected References


• McHugo J, Whittle M. Enlarged fetal bladders: aetiology, management and outcome. Prenatal Diagnosis (2001); 21: 958-963


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• Phillips TM. Spectrum of cloacal extrophy. Seminars in Pediatric Surgery (2011) 20,113-118

Thank you!