# 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)

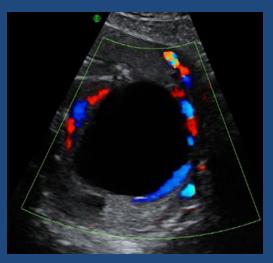
 Even in the presence of severe GU anomalies, usually amniotic fluid volume is normal in the 1<sup>st</sup> Trimester

# Background

- 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)

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- Evaluation of the umbilical arteries to define megacystis as opposed to other abdominal cyst/ hydrocolpos

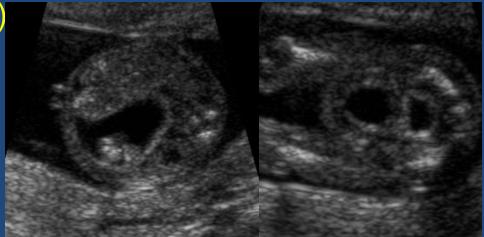




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- External genitalia: Gender/ ambiguous/ incompletely formed

### **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 (3<sup>rd</sup> trimester)
- Bowel: Anorectal region/colon

 After 20w we expect to see meconium filled rectum.



28w

 Around the 27w the whole colon is filled with meconium

 Assessment for microcolon (Megacystis microcolon intestinal hypoperistalsis syndrome)





 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 (Calvo-Garcia M.A.

- cloacas have:
  - Dilated rectum
  - High cul-desac

(Calvo-Garcia M.A. et al Ped Radiol 2011)



**NORMAL RECTUM** 

et al Ped Radiol



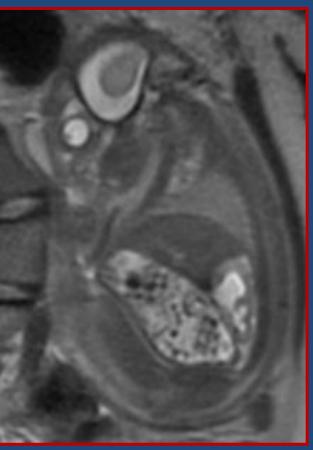
Cloaca

 and imperforate anus with RU fistula:

Can present rectal dilatation with fluid content and enteroliths



**NORMAL RECTUM** 



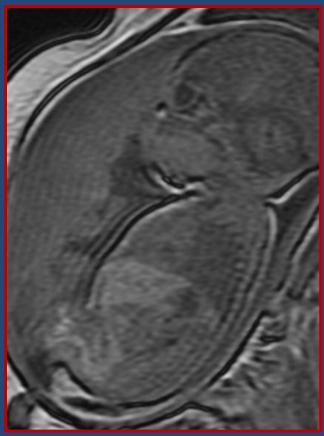
CLOACA

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

(Calvo-Garcia M.A. et al, Ped Radiol e-pub 2012, DOI 10.1007/s00247-012-2571-3)



**NORMAL RECTUM** 

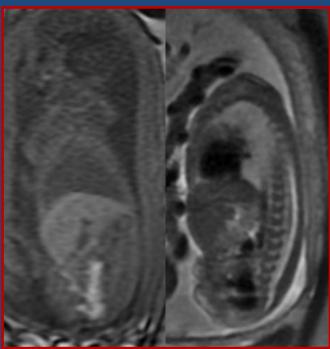


CLOACAL EXSTROPHY

- Bladder exstrophy:
  - Normalmeconium intherectum/colon



**NORMAL RECTUM** 



BLADDER EXSTROPHY

#### **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)

- 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

 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!!!!

- Inability of the bladder to store urine (no visible bladder): normal amniotic fluid
  - Infraumbilical wall defect
    - Bladder exstrophy: usually normal rectum and spine
    - Cloacal exstrophy (OEIS): "elephant trunk-like" image sometimes (but not always!)

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Fetal Anomalies page 536"

Pediatr Radiol DOI 10.1007/s00247-012-2571-3.

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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

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## Thank you!