Prenatal and Postnatal Assessment of Posterior Urethral Valve

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Outline

• Normal kidney and urinary tract development
  • Normal fetal kidney and urinary tract on ultrasound
  • Types of kidney and urinary tract malformations
  • Case of posterior urethral valve
    - prenatal evaluation
    - postnatal evaluation
• Interventions and long-term outcome
## Development of the Kidney

<table>
<thead>
<tr>
<th>Part of kidney</th>
<th>Appears</th>
<th>Functional</th>
<th>Gives rise to</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pronephros</td>
<td>4&lt;sup&gt;th&lt;/sup&gt; week</td>
<td>Never functional</td>
<td>Pronephric ducts used by mesonephros</td>
</tr>
<tr>
<td>Mesonephros</td>
<td>4&lt;sup&gt;th&lt;/sup&gt; week</td>
<td>Functions for 4 weeks</td>
<td>Contributes to male genital structures and bladder</td>
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<tr>
<td>Metanephros</td>
<td>5&lt;sup&gt;th&lt;/sup&gt; week</td>
<td>Starts functioning at 9&lt;sup&gt;th&lt;/sup&gt; week</td>
<td>UG bud gives rise to ureters and collecting ducts</td>
</tr>
<tr>
<td>Urogenital (UG) Sinus</td>
<td></td>
<td>Bladder and urethra</td>
<td></td>
</tr>
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</table>

Image from UpToDate. Overview of CAKUT. Waters, A. and Rosenblum, N. http://www.uptodate.com/online/content/topic.do?topicKey=pedineph/18797&selectedTitle=1%7E150&source=search_result. 7/15/10.
Development of Urethra

- Mesonephric (Wolffian) duct fuses with UG sinus and contributes to urethra
- Caudal UG sinus (genital tubercle) forms phallic urethra
- Urethra formation is complete by 14 weeks

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Normal Fetal Kidney on Ultrasound

• Cannot reliably see fetal kidney on ultrasound during 1st trimester.
• Visualized by 16-22 weeks.
• Can visualize cortex and pyramids by 23-26 weeks, and fine anatomy by 30 weeks.
• Fetal kidney length in mm is roughly equal to the fetal menstrual age in weeks.
Normal Fetal Bladder on Ultrasound (18 wks)
Normal Fetal Kidney on Ultrasound (18 wks)

Transverse View

Spine
Kidneys
Ultrasound Evaluation of Kidney Function

- Fetal kidney function is assessed by
  - visualization of urine in the bladder by 13-16 weeks (the ureters are not normally seen)
  - measurement of amniotic fluid (subjective measures appear to be as good as summing the depth of amniotic fluid pockets in four quadrants)
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Types of Kidney Malformations:

Parenchymal

• Hypoplasia
• Dysplasia
• Renal agenesis

Gross Hypoplastic Kidney


Multicystic Kidney

• Multicystic kidney
• Genetic cystic disease (ARPKD, ADPKD, NPH)

Multicystic Kidney on Ultrasound

PACS, BIDMC
Types of Kidney Malformations: Migration

- Ectopia
- Fusion

Horseshoe Kidney on IVU

Crossed Fused Ectopic Kidney on CT with contrast
PACS, BIDMC
Types of Kidney Malformations: Collecting System

- Pelvis: UPJ obstruction
- Ureter: megaureter, ectopic ureter, VUR
- Bladder: exstrophy
- **Urethra: posterior urethral valve (PUV)**

Ectopic ureter on CT

PACS, BIDMC
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Patient 1: Posterior Urethral Valve

- Congenital defect
- Affects males
- The ‘valve’ is a membranous fold between urethral wall and verumontanum (crest where seminal vesicles enter urethra) that obstructs the urethra
Patient 1: Posterior Urethral Valve

The valve may originate from incomplete canalization or an aberrant insertion of the Wolffian duct into the cloaca.
Posterior Urethral Valve

Key Findings on Ultrasound

- Dilated posterior urethra
- Dilated bladder with possible wall thickening
- Hydronephrosis
Patient 2: Male Fetus on Ultrasound (18 wks)

Genitalia
Legs
Patient 2: “Keyhole Sign”: Dilated Bladder and Prostatic Urethra on Ultrasound

Dilated bladder
Dilated prostatic urethra
Patient 2: Bladder Dilatation on Ultrasound (20 wks)

Bladder (20 wks) measures 4.14 by 2.04 cm on sagittal view.

Normal bladder (18 wks) measures 0.44 cm on transverse view.

Anechoic space demonstrates no flow on Doppler imaging, suggesting a dilated bladder rather than a vascular structure.
Patient 2: Enlarged Hydronephrotic Kidneys on Ultrasound (20 wks)

Right kidney measures 2.80 cm

Left kidney measures 3.42 cm

Normal kidney size: 1 mm/week x 20 weeks = 20 mm or 2 cm
Patient 2: Dilated Renal Pelvis on Ultrasound (20 wks)

Dilated Pelvis of Left Kidney (0.53 cm)

Dilated Pelvis of Right Kidney (0.66 cm)

At 20 wks: Pelvis <5 mm is normal but can be 6-9 mm and normal. Here, pelvis>5 mm plus other findings suggest obstruction.
Patient 2: Amniotic Fluid Measurement on Ultrasound

Amniotic Fluid estimate: 14.23 mL at 20 weeks

5-25 mL is the normal range for most of pregnancy
Patient 3: Visible Ureters and Dilated Renal Pelvices on Ultrasound

Image courtesy of Dr. Tejas Mehta, BIDMC
Differential Diagnosis for Ultrasound Findings

- Mechanical obstruction
  - PUV
  - Urethral atresia
  - Caudal regression syndrome
  - Megacystis-microcolon-intestinal hypoperistalsis syndrome
  - Prune Belly Syndrome (hypotonic abdominal wall, megacystis, ureterectasis, cryptorchidism).

- Functional obstruction
  - Abnormality in the sphincter, innervation, or musculature of the bladder

PUV is suggested by the keyhole sign, the absence of other associated findings, and the severity of obstruction.

Diagnosis is confirmed postnatally.
Accuracy of Ultrasound Evaluation

- In one study:
  - 6 fetuses had catheters placed for dilated urinary tracts thought to be caused by PUV (5) or obstructed megaureter (1)
  - 2 were found to have PUV postnatally
Prognostic Indicators

Patients with PUV often have chronic kidney disease, VUR, and bladder dysfunction.

Risk factors for poor renal function:

• Diagnosis of PUV before 24 weeks
• Oligohydramnios – when present in 2\textsuperscript{nd} trimester, mortality is 90-95%  
• Serum creatinine >1.0 mg/dL
• Bladder dysfunction
Renal Outcome

It is unclear whether PUV and renal dysplasia are associated because there is:

- A common developmental injury
- Damage caused by high back pressure
- Recurrent infection causing scarring
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Clinical Presentation

**Neonates**: respiratory distress, dilatated bladder, urinary ascites, difficulty urinating

**Infants**: Failure to thrive, urosepsis, difficulty urinating

**Boys**: UTIs, incontinence, difficulty urinating
Voiding Cystourethrogram (VCUG) is the study of choice
- Contrast is injected via a catheter into the bladder
- During micturition, the flow of urine is observed on fluoroscopy

Ultrasound can be used to assess hydrouteronephrosis and renal parenchyma

If VCUG is not diagnostic, cystourethroscopy can be used

MRI is a developing technology
  - fast sequences or patient sedation required in pediatric population
Patient 4: VCUG Demonstrates Obstruction and Unilateral Reflux Postnatally

1/3-1/2 of patients with PUV have reflux, which may be unilateral or bilateral.

Dilated renal pelvis
Dilated and tortuous ureter
Dilated bladder
Dilated prostatic urethra
Trabeculations
Diverticulum

Children’s Hospital Boston
Unilateral or Bilateral Reflux

• In one retrospective study of 200 patients with PUV, 27% had unilateral vesicoureteral reflux on VCUG and 37% had bilateral reflux

• Unilateral reflux may be due to protective mechanisms increasing reflux in one kidney in order to preserve contralateral kidney function (vesicoureteral reflux and dysplasia syndrome)
Patient 4: Lateral VCUG shows Dilated Bladder and Prostatic Urethra

Dilated bladder with trabeculations and diverticula

Dilated prostatic urethra
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## Interventions

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<tr>
<th>Prenatal</th>
<th>Vesicoamniotic shunt</th>
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<tbody>
<tr>
<td>In general, prenatal intervention is avoided because the benefits have not been shown to outweigh prenatal and maternal morbidity</td>
<td>Open fetal surgery</td>
</tr>
<tr>
<td></td>
<td>Cystoscopic ablation</td>
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<thead>
<tr>
<th>Newborn</th>
<th>Bladder drainage via feeding tube or catheter</th>
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<tbody>
<tr>
<td></td>
<td>Cystoscopic ablation</td>
</tr>
<tr>
<td></td>
<td>Vesicotomy if needed</td>
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</tbody>
</table>

| Older boys                | Cystoscopic ablation                       |
Cystoscopy

The cystoscope allows visualization of the urinary tract.

Surgical instruments may be passed through the cystoscope.
Long-Term Outcome

• Perinatal mortality, caused by pulmonary hypoplasia and sepsis, has decreased to <10% due to better management
• Long term outcome depends on renal parenchymal function
• Renal transplant has been shown to be effective in a setting where the bladder permits transplant survival
References

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