Diagnosis and Management of Common Perinatal Urologic Problems

Dr. Jonathan VB Riddell MD FRCP ABU (Dipl.)
Assistant Professor – Pediatric Urology

CME Disclosure

• I have no conflicts of interest or relevant financial relationships with any commercial entities.

Brief Overview

• Fetal urologic abnormalities
• Evolution into perinatal period
• Implications for provider
• Unrecognized hypospadias at time of circumcision

CHU Ste. Justine – Montreal Quebec
Hopital Mere-Enfant
Impact of Technology (US)
-Kidneys, bladder and amniotic fluid can be visualized as early as 12 weeks
-Renal dysplasia, duplication, hydronephrosis and hydroureter can be detected
-Bladder cycling, megacystis and trabeculation
-Urologic anomalies can be isolated or exist with syndromes
-Definitive diagnosis usually still made neonatally

Prenatal Urinary Tract Dilation
- Occurs in 1-2% of pregnancies
- 80,000 children per year in USA
- Reflects spectrum of etiologies from physiologic (majority) to others with significant morbidity even mortality

Table 1: Etiology of urinary tract dilation detected on antenatal ultrasound.

<table>
<thead>
<tr>
<th>Etiology</th>
<th>Incidence (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Transient/physiologic</td>
<td>50–70</td>
</tr>
<tr>
<td>Ureteropelvic junction obstruction</td>
<td>10–30</td>
</tr>
<tr>
<td>Vesicoureteral reflux</td>
<td>10–40</td>
</tr>
<tr>
<td>Ureterovesical junction obstruction/megaureter</td>
<td>5–15</td>
</tr>
<tr>
<td>Multicystic dysplastic kidney disease</td>
<td>2–5</td>
</tr>
<tr>
<td>Posterior urethral valves</td>
<td>1–5</td>
</tr>
<tr>
<td>Ureterocele, ectopic ureter,</td>
<td>Uncommon</td>
</tr>
<tr>
<td>duplex system, urethral atresia,</td>
<td></td>
</tr>
<tr>
<td>Prune belly syndrome, polycystic</td>
<td></td>
</tr>
<tr>
<td>kidney diseases, l cysts</td>
<td></td>
</tr>
</tbody>
</table>

Adapted from Nguyen et al. 2010 [16].

Controversies in Prenatal Urology
- Evidence co-relating severity of prenatal UT dilation with postnatal pathology lacking
- No uniformity on how to define, classify and grade UT dilation both pre- and postnatal
- UT dilation is dynamic; hydration, bladder volume and pt position all play a role
- Uropathies represent a spectrum
Multidisciplinary consensus on the classification of prenatal and postnatal urinary tract dilation (UTD classification system)

Hlep T. Nguyen, Carol B. Benson, Bryann Bromley, Jeffrey B. Campbell, Jeanne Chow, Beverly Coleman, Christopher Cooper, Jude Crino, Kassa Darge, C.D. Anthony Herndon, Anthony O. Odibo, Michael J.G. Somers, Deborah R. Stein

Table 2 US parameters included in the Urinary Tract Dilation Classification System.

<table>
<thead>
<tr>
<th>US parameters</th>
<th>Measurement/findings</th>
<th>Note</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anterior-Posterior Renal Pelvic Diameter (APRPD)</td>
<td>(mm)</td>
<td>Measured on transverse image at the maximal diameter of intrarenal pelvis</td>
</tr>
<tr>
<td>Calyceal dilation</td>
<td>Central (major calyces)</td>
<td>Yes/No</td>
</tr>
<tr>
<td></td>
<td>Peripheral (minor calyces)</td>
<td>Yes/No</td>
</tr>
<tr>
<td>Parenchymal thickness</td>
<td>Normal/Abnormal</td>
<td>Subjective assessment</td>
</tr>
<tr>
<td>Parenchymal appearance</td>
<td>Normal/Abnormal</td>
<td>Evaluate echogenicity, corticomedullary differentiation, and for cortical cysts</td>
</tr>
<tr>
<td>Ureter</td>
<td>Normal/Abnormal</td>
<td>Dilatation of ureter is considered abnormal; however, transient visualization of the ureter is considered normal postnatally</td>
</tr>
<tr>
<td>Bladder</td>
<td>Normal/Abnormal</td>
<td>Evaluate wall thickness, for the presence of ureteroceles, and for a dilated posterior urethra</td>
</tr>
</tbody>
</table>

Table 3 Normal values for Urinary Tract Dilation Classification System.

<table>
<thead>
<tr>
<th>Ultrasound findings</th>
<th>Time at presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>16–27 weeks</td>
</tr>
<tr>
<td>Anterior-Posterior Renal Pelvis Diameter (APRPD)</td>
<td>&lt;4 mm</td>
</tr>
</tbody>
</table>
PRENATAL PRESENTATION

<table>
<thead>
<tr>
<th>Term</th>
<th>10-17 wks AP/RPO</th>
<th>18-20 wks AP/RPO</th>
<th>20-26 wks AP/RPO</th>
</tr>
</thead>
<tbody>
<tr>
<td>Central or subcalyceal dilation*</td>
<td>4 to &lt;7mm</td>
<td>7 to &lt;10mm</td>
<td>≥ 10mm</td>
</tr>
<tr>
<td>Parenchymal thickness normal</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Parenchymal appearance normal</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ureters normal</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bladder normal</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No unexplained oligohydramnios</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>UTD A-1</td>
<td>LOW RISK</td>
<td></td>
<td></td>
</tr>
<tr>
<td>UTD A-2-3</td>
<td>INCREASED RISK</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Central and peripheral calyceal dilation may be difficult to evaluate early in gestation.
**Oligohydramnios is suspected to result from a UG cause.
The good
Isolated UT dilation – Implications for Provider

- Arrange ultrasound after 48h
- If unilateral pathology or bilateral low risk one offers antibiotic prophylaxis and VCUG
- Arrange non-urgent referral to pediatric urologist/nephrologist (if not involved) within 1 month

Top 3 for unilateral pathology

- Uretero-pelvic junction obstruction
- Vesico-ureteral reflux
- Megaureter

Do low risk patients need VCUG?

- Guidelines: discretion of clinician
- Circumcision is more effective than CAP
- CAP most effective in dilating reflux
- Is family willing to give CAP?
- Is family reliable for surveillance?
- Education for urine cultures

Correlation of Prenatal and Postnatal Ultrasound Findings With the Incidence of Vesicoureteral Reflux in Children With Fetal Renal Pelvic Dilatation

Douglas E. Coplen,* Paul F. Austin,+ Yan Yan and Jeffrey M. Dicken
From the Division of Urology, Department of Pediatrics and Surgery, Washington University School of Medicine, St. Louis, Missouri.

- Children with prenatal UT dilation have rate 12-38% VUR
- If dilation persists postnatally, rate >40%
- Poor correlation between severity of dilation and grade of reflux
Unilateral High Risk

- Often UPJ syndrome or primary nonrefluxing megaureter
- VCUG helpful to R/O reflux – sets FU to monitor function as opposed to UTI

- Over 2/3 of former SFU 3/4 managed conservatively improve or remain stable over months to years

Operative indications in <6 months of age

- Renal insufficiency
- Recurrent febrile UTIs or sepsis
- Bilateral severe disease with thinning and renal scan evidence of obstruction or progression
### The bad...

### Exceptions to guidelines

- The grading system is designed to be used in cases of isolated UT dilation and not to be applied to unique situations or anomalous kidneys such as solitary, ectopic, multicystic dysplastic kidneys (MCDK) or other cystic diseases of the kidney.
- * Certain situations (e.g., Posterior urethral valves, bilateral severe hydronephrosis) may require more expedient follow up.
- In these cases, preservation of renal function becomes paramount.

### Implications to provider

- Earlier urologic involvement (preferably prenatally)
- Ultrasound can be done <48h
- Consider serial serum Cr
- Consider antimicrobial prophylaxis

### The ugly...
Failure to Visualize Bladder

Lack of urine

- Multicystic dysplastic kidney (MCDK)
- Bilateral renal agenesis (Potter’s syndrome)

Exstrophy-epispadias complex

- Non-visualization bladder
- Lower abdominal mass splaying umbilical arteries
- Small penis/anterior scrotum
- Low set umbilicus with widened symphisis

Megacystis differential

- Severe bilateral reflux
- Posterior urethral valves
- Prune Belly Syndrome
- MMIH (megacystis-microcolon-intestinal hypoperistalsis)
- Urethral atresia
Approach to Megacystis

- Sex of infant
- Renal parenchyma – degree of dysplasia
- Degree of hydronephrosis/obstruction
- Serial evaluation of amniotic fluid
- Presence of other fetal anomalies

Fetal Ultrasound Criteria for Megacystis Differential

<table>
<thead>
<tr>
<th>Keyhole sign</th>
<th>Severe VUR</th>
<th>Prune Belly</th>
<th>Posterior Urethral Valves</th>
<th>MMIH</th>
<th>Urethral Atresia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bladder wall</td>
<td>Thin</td>
<td>Thin</td>
<td>Trabeculated with dilated prox. urethra</td>
<td>Thin</td>
<td>Symmetrical and cystic</td>
</tr>
<tr>
<td>AF</td>
<td>Normal</td>
<td>Oligo</td>
<td>Normal -&gt; oligo</td>
<td>Normal -&gt; polyhydranmos</td>
<td>Severe oligo -&gt; anhydramnios</td>
</tr>
<tr>
<td>Other US findings</td>
<td>Ureteral tortuosity</td>
<td>Pulmonary hypoplasia</td>
<td>Renal dysplasia/VURD</td>
<td>Colon not visible; proximal dilation</td>
<td>Small chest/huge abdomen</td>
</tr>
<tr>
<td>Pathophysiology</td>
<td>Yo-yo voiding</td>
<td>Abnormal smooth muscle</td>
<td>Sail-like valves at UG membrane</td>
<td>Decrease in ganglion cells</td>
<td>Survive if fistula develops</td>
</tr>
</tbody>
</table>

In Utero Intervention for Posterior Urethral Valves

- Holmes et. al (UCSF 2001) reviewed 36 cases of fetal intervention (open surgery) for suspected valves
- 43% mortality rate
- Long-term outcomes did not improve for postnatally confirmed PUV pts
- And only 14/36 actually had PUV!

Vesico-Aminotic Shunts

- PLUTO trial (2013) randomized 31 singleton pregnancies with LUTO to shunt vs. conservative management
- Only 20% enrolment reached; 2/3 pts had PUV and 1/3 urethral atresia
- Eight from VAS group survived to 28 days vs. four in conservative group (p=0.27 ITT, p=0.03 based on treatment received)
Shunt complications

- SROM after shunt insertion (3/15)
- Dislodged (3/15)
- Blocked (1/15)
- Other series observe bowel injury, errant placement, entero-colitis, chorio-amnionitis, urinary ascites
- Many infants delivered prematurely with respiratory complications

Take home message

- Multidisciplinary consensus going forward relies heavily on AP diameter of renal pelvis to stratify risk
- Low risk patients can be managed without VCUG, antibiotics or invasive studies
- Intermediate and high risk cases benefit from a multidisciplinary approach
- A huge academic opportunity awaits...

• 111 fetuses with severe LUTO; fetal cystoscopy (34) vesicoamniotic shunting (16) and no fetal intervention in 61.

**Conclusion** Fetal cystoscopy and vesicoamniotic shunting improve the 6-month survival rate in cases of severe LUTO. However, only fetal cystoscopy may prevent impairment of renal function in fetuses with posterior urethral valves.
Discovery of MMIP

- Mega-meatus intact prepuce
- Pass 5Fr catheter to protect urethra
- If too close/thin apply vaseline gauze
- Stitch only to control bleeding
- Stitching prepuce can lead to pathologic phimosis