Urology in the Nursery/NICU

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No disclosures to report
<table>
<thead>
<tr>
<th>Organs Affected</th>
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<tbody>
<tr>
<td>Upper Urinary Tract</td>
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</table>
Problem # 1

“My OB told me that my baby had-

• “swollen kidney(s)”
• “cyst(s) on the Kidney(s)”
• “big bladder”
• “cyst in the pelvis”

• Some will get better on their own
  which babies to worry about??
Clinical Significance of Hydronephrosis

- UTI / Loss of Kidney Function
- Spontaneous Improvement
- Risks / Cost / Discomfort of Testing
Upper Urinary Tract Anomalies

- Hydronephrosis
- Vesicoureteral Reflux
- Obstruction of the ureter at the kidney or bladder
  - Ureteropelvic junction obstruction - UPJO
  - Ureterovesical junction obstruction
    - UVJO (1° Megaureter)
  - Ureterocele with single or double ureter(s)

- Multicystic kidney
Limitations of Prenatal Diagnosis - Genitourinary Disorders

Modalities

- Ultrasound - difficulty distinguishing cysts versus dilatation single vs duplex ureters
- MRI - cost, availability, need for sedation
- Amniocentesis / chorionic villus sampling for genetic testing - risk to mother and fetus
- Vesicocentesis (bladder aspirate) for urine electrolytes - risk to fetus
Extent of Hydronephrosis
Anterior-Posterior Renal Pelvic Diameter (APRPD) Grading System

• During the second trimester, APRPD is defined as mild - 4 to <7 mm, moderate - 7 to ≤10 mm, and severe >10 mm.
• During the third trimester, APRPD is defined as mild 7 to <9 mm, moderate - 9 to ≤15 mm, and severe >15 mm.
• Larger APRPD ➔ More likely caused by obstructive uropathy
  ➔ Greater risk of surgery postnatally
  ➔ Lower spontaneous resolution rate
**Society of Fetal Urology Grading System**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Central Renal Complex</th>
<th>Parenchyma</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>1</td>
<td>Slight splitting</td>
<td>Normal</td>
</tr>
<tr>
<td>2</td>
<td>Splitting within kidney borders</td>
<td>Normal</td>
</tr>
<tr>
<td></td>
<td>Only minor dilatation of calyces</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Splitting outside renal borders</td>
<td>Normal</td>
</tr>
<tr>
<td></td>
<td>&amp; dilated calyces</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Marked splitting &amp; dilated calyces</td>
<td>Thin</td>
</tr>
</tbody>
</table>

![Images of different grades showing the progression from normal to marked splitting and dilated calyces.]
Prenatal Hydronephrosis

Multidisciplinary consensus on the classification of prenatal and postnatal urinary tract dilation NGUYEN HT ET AL J PED UROL 2014, 10: 982
## 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, Duplex ureters, Urethral atresia, Prune Belly syndrome, Polycystic kidney diseases, renal cysts</td>
<td>Uncommon</td>
</tr>
<tr>
<td>Vaginal anomalies – urogenital sinus / cloaca</td>
<td></td>
</tr>
</tbody>
</table>

Urinary Tract Abnormalities in the Newborn

AP Pelvis diameter

Kidney cyst

Bladder
Timing of Postnatal Ultrasound

• Up to 48 hours after birth – Baby is oliguric - tendency to underestimate the severity of hydronephrosis 2°dehydration

  ➔ Delay postnatal ultrasound for 48hrs unless...
  ◆ oligohydramnios
  ◆ UTI
  ◆ urethral obstruction / urinary retention
  ◆ bilateral severe hydronephrosis
  ◆ poor parental compliance with postnatal evaluation
Risk Stratification

>48hr Ultrasound

POSTNATAL PRESENTATION

- > 48 hours
  - APRPD
  - 10 to < 15mm

  Central calyceal dilation
  - Parenchymal thickness normal
  - Parenchymal appearance normal
  - Ureters normal
  - Bladder normal
  - UTD P1: LOW RISK

- > 48 hours
  - APRPD
  - ≥ 15mm

  Peripheral calyceal dilation
  - Parenchymal thickness normal
  - Parenchymal appearance normal
  - Ureters abnormal
  - Bladder normal
  - UTD P2: INTERMEDIATE RISK

- > 48 hours
  - APRPD
  - ≥ 15mm

  Peripheral calyceal dilation
  - Parenchymal thickness normal
  - Parenchymal appearance abnormal
  - Ureters abnormal
  - Bladder abnormal
  - UTD P3: HIGH RISK
RISK-BASED MANAGEMENT, POSTNATAL DIAGNOSIS

UTD P1: LOW RISK
- FOLLOW UP US: 1 to 6 months
- VCUG: Discretion of clinician
- ANTIBIOTICS: Discretion of clinician
- FUNCTIONAL SCAN: Not recommended

UTD P2: INTERMEDIATE RISK
- FOLLOW UP US: 1 to 3 months
- VCUG: Discretion of clinician
- ANTIBIOTICS: Discretion of clinician
- FUNCTIONAL SCAN: Discretion of clinician

UTD P3: HIGH RISK
- FOLLOW UP US: 1 month
- VCUG: Recommended
- ANTIBIOTICS: Recommended
- FUNCTIONAL SCAN: Discretion of clinician

The choice to utilize prophylactic antibiotics or recommend voiding cystourethrogram will depend on the suspected underlying pathology.
Should the Ultrasound Be Repeated??

Reliability of a Single Postnatal Ultrasound

• Initial normal postnatal US may be misleading.
• Aksu et al. observed that 21–28% of children with prenatal urinary tract dilation had a normal initial postnatal US but \(\Rightarrow\) 45% had abnormal repeat US
• Approximately 15% with prenatal UT dilation develop later worsening or recurrent hydronephrosis after an initial normal postnatal US
  \(\Rightarrow\) Consider a repeat US ultrasound even if first postnatal US is normal e.g. at 3-6 months
Summary - Natural History of Hydronephrosis

• Moderate and severe hydronephrosis - earlier and more frequent postnatal US evaluations compared to mild UT dilation.

• In a meta-analysis, SFU Grade 2 resolved in 70% of the cases and SFU Grade 1 and 2 stabilized in 98% of the cases★

• Sencan et al - prenatal UT dilation and mild (SFU Grade 1 & 2) hydronephrosis on the first postnatal US,
  ➔ follow-up US demonstrated resolution of UT dilation - 67%, improvement -13 %, stabilization - 16%, worsening - 3%✪

• ≈20% of ureteral obstruction UPJO / UVJO will require surgery

Prophylaxis of UTI - Antenatal Hydronephrosis

• Incidence of UTI - SFU Grade 1–2 - 5%,
  - SFU Grade 3–4 - 23%★

• Risk of UTI with / without prophylaxis with SFU Grade 1 & 2 or APRPD < 15 mm was similar (2.2% vs. 2.8%) but SFU Grade 3 and 4 or APRPD ≥15 mm - 14.6% (95% CI: 9.3-22) vs. 28.9% (95% CI: 24.6-33.66), p < 0.01)♦

• Estimated number to treat to prevent 1 UTI was 7 (with SFU 3 or 4)♦

• Circumcision reduces risk of UTI

Antibiotic use, cesarean birth and formula feeding all affect infants’ microbiomes, studies indicate.

The AP (6/15, Neergaard) reports, “Two new studies” published in Science Translational Medicine “are offering some of the clearest snapshots yet of how babies build up protective gut bacteria, adding to evidence that antibiotics and birth by C-section may disrupt that development.” After

Low dose, once daily antibiotic therapy seems safe
Vesicoureteral Reflux in Babies

• My perspective - low threshold for ordering VCUG for moderate hydronephrosis - definitely for severe hydronephrosis
  - low risk of causing UTI / minimal discomfort for baby

• AAP Guidelines for screening post UTI at age 2-24 months
  ➔ all neonates with UTI warrant VCUG -30-40% risk of VU Reflux

• Review of images by Pediatric Urologist / Radiologist to R/O other anomalies eg Posterior Urethral Valves / Ureteral Duplication or Obstruction - UPJO / UVJO
Controversy - Prophylaxis with VUR

- Baby’s kidney has higher incidence of Intrarenal Reflux
  - bacteria penetrate kidney more easily
  - highest risk of renal scarring in first 2 years

My opinion - Prophylaxis warranted for Grades 2-5 VUR in neonate
Natural History of VUR in Babies

• 1° or 2° to subtle voiding dysfunction
  - overactive bladder or contraction of sphincter during void

• Highest VUR resolution in first 2 years - related to subtle voiding dysfunction which improves with neurologic maturation

• Even grade 4 or 5 VUR may resolve in infants

• Early surgery not warranted unless chronic pyelonephritis
Management of Ureteral Obstruction
UPJO or UVJO

• Diuretic Renal Scan
  Normal - 50% / 50%
  with early uptake & drainage

• Mild - Continue ultrasound surveillance until resolves
  – less frequent studies if improving

• Moderate - diuretic renal scan is normal ➔ ultrasound Q3-4mo
  - If diuretic renal scan is abnormal, alternate with ultrasound

• Severe – Monitor more closely
Indications for Surgery - UPJO / UVJO

Surgery rarely required in neonate

Indications
• Decreasing function < 40% on 2 renal scans
• Worsening hydronephrosis
• Symptoms – pain / UTI

Surgery - UPJO –open pyeloplasty for babies
versus laparoscopic / robotic -older
    - UVJO – cystoscopic dilatation & stenting -versus open repair
URETERAL DUPLICATION

- More common in girls >> boys
- Unilateral or bilateral
- Asymmetrical hydronephrosis
  - upper vs lower pole
- Upper pole ureter can be ectopic / obstructed
- Lower pole ureter can reflux
Ureterocele

- Cystic lesion in bladder - distal end of ureter hydroureteronephrosis of 1 or 2 ureters dilatation in 1/2/ or all of kidney

- Presentation - prenatal detection, UTI/VU reflux urinary retention 2° bladder outlet obstruction

Treatment

- Antibiotic prophylaxis
- Observation - some with dysplastic kidney may involute or Procedure - puncture to decompress especially if UTI & temporize
- Possible need for reconstructive surgery later - defer in baby due to small size of bladder
Multicystic Dysplastic Kidney

- Total ureteral obstruction → dysplasia
- Prenatal diagnosis - may be confused with UPJO
- Classic ultrasound appearance & non-function on renal scan
- Contralateral kidney is usually normal
  - but also may have UPJO or VU reflux
- Rare hypertension, very rare tumor - less than 1 /≈ 1700
- Monitor BP at well visits
- No treatment / surveillance for classic cases
POSTERIOR URETHRAL VALVES

• Presentation - prenatal hydroureteronephrosis, UTI, urinary retention /renal failure, lower abdominal mass
• Diagnosis - Ultrasound
  - VCUG - dilated posterior urethra
• Management - catheter - feeding tube -not foley
  - antibiotics / rehydration
  ✪ watch for post-obstructive diuresis
  avoid nephrotoxic antibiotics eg gentamicin
• Treatment - usually endoscopic ablation
  vs vesicostomy if small/premature or persistent ↑ creatinine
• 25% deteriorate to ESRD
Prenatal Consultation – Pediatric Urology

• Parents seek information - may not be accurate
• Useful to explain likely diagnosis & allay parental anxiety
• Plan postnatal evaluation with pediatrician & family
Problem # 2

• “I took Progesterone in the first trimester - did that affect by baby? ”
• “Is it a boy or a girl?”
Disorders of the Genitalia - Neonate

- Minor Penile Anomalies eg Hidden / Webbed Penis
- Hypospadias
- Cryptorchidism
- Acute Scrotum
- Epispadias / Exstrophy
- Disorders of Sexual Development
Concealed / Webbed Penis

Concealed / Hidden Penis –
- Penile shaft / corpora are retracted due to fibrous bands
- Foreskin is inverted with little external skin
- Appears like a nozzle/pyramid
- Confused with Micropenis but corpora / glans normal to palpation

Webbed Penis – attached to scrotum

- Defer circumcision
  ➔ reconstructive surgery at 6 months
Hypospadias

Incidence increasing 2° environmental factors
Reconstructive surgery at 6 months
  unless medical or hormonal issues
  • Outcome enhanced by expertise
  • 90% amenable to single stage repair
    except for severe – scrotal / perineal
Micropenis

• Less than 2 cm stretched length in fullterm baby

• Causes - Hypo or Hyper gonadotropin Hypogonadism - 75%
  - Growth Hormone Deficiency - 15%
  - Idiopathic - 10%

• Evaluation - Karyotype, Endocrine testing - LH, FSH, Pituitary function testing, Testosterone, HCG stimulation MRI Brain

• Treatment - Maintain male gender, Testosterone / Androgen stimulation, Growth Hormone replacement if deficient
Cryptorchidism

- Physical exam is most accurate
  beware of looping vas with higher testis
  ➔ Misdiagnosis as descended testis
- Ultrasound not cost-effective /accurate for impalpable testis★ - exam by pediatric urologist or laparoscopy at 6 months
- Testosterone surge from 1-3 months
  Spontaneous descent stops at 6 months
  ➔ Surgery at 6-9 months
- Bilateral impalpable testes
  ➔ concern for anorchia or DSD
  obtain testosterone & MIS levels, karyotype

Normal Sexual Development

Chromosomal Sex

Gonadal Sex

Phenotypic Sex
Newborn – Concerns for DSD

• Clinical Findings That Raise the Possibility of DSD

Apparent male

• Bilateral nonpalpable testes (full-term infant)
• Severe hypospadias with bifid scrotum
• Unilateral or bilateral undescended testis with hypospadias
Newborn – Concern for DSD

• Clinical Findings That Raise the Possibility of DSD

Apparent female

• Clitoral hypertrophy of any degree
• Abnormal vulva with a single opening
• Inguinal hernia containing a gonad
Disorders of Sexual Development - Formerly Intersex / Ambiguous Genitalia

<table>
<thead>
<tr>
<th>Formerly</th>
<th>Current</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female Pseudohermaphrodite</td>
<td>46, XX DSD</td>
</tr>
<tr>
<td>Male Pseudohermaphrodite</td>
<td>46, XY DSD</td>
</tr>
<tr>
<td>True Hermaphrodite</td>
<td>Ovotesticular DSD</td>
</tr>
<tr>
<td>XX Male</td>
<td>46, XX Testicular DSD</td>
</tr>
<tr>
<td>XY Sex Reversal</td>
<td>46, XY Complete Gonadal Dysgenesis</td>
</tr>
</tbody>
</table>
Evaluation of DSD

- Collaboration with Pediatric Urology, Endocrinology, Genetics
- Physical Exam – Size & Configuration of Phallic Structure / Gonad(s)
- Testing – Hormonal, Radiologic, Genetic/Chromosomal
  ◆ All neonates have 17 hydroxyprogesterone measured through LA state lab to screen for 21-OH CAH
    17OH-P threshold set low, but may miss some non-classical cases
  ◆ Typically also obtain LH, FSH, Testosterone, Dihydrotestosterone, electrolytes
  ◆ Routine karyotype / chromosomal testing
  ◆ Ultrasound of Abdomen/Pelvis, Genitogram assess internal genitalia – gonads & uterus / adrenals
Principal Causes of DSD – According to Gonadal Pattern

- Ovary - CAH, Placental aromatase deficiency, Maternal source of virilization
- Testis - Leydig cell hypoplasia, Testosterone biosynthetic defect, 5-alpha reductase deficiency, Androgen Insensitivity
- Ovary and Testes – Ovotesticular DSD
- Dysgenetic gonads - Gonadal Dysgenesis, Denys-Drash and Frasier syndromes, Smith-Lemi-Opitz syndrome, or Campylomelic dwarfism
Sex of Rearing - DSD

• Involves extensive family counseling and input
• Based on potential sexual and reproductive function but also intrauterine hormonal exposure and brain imprinting.
• Almost all CAH - 21 hydroxylase deficiency babies are raised according to genetic sex
  (except rare cases of severely virilized females 2° androgen imprinting)
• Reconstructive surgery - Girls - Clitoroplasty / Vaginoplasty at 6 months if hormonal status stable
  -Boys - hypospadias repair +/- orchiopexy / gonadectomy
Problem #3

- Nurse pages you -
- “Baby Boy Fontenot has a blue / swollen scrotum”
Differential Diagnosis of Acute Scrotum in Neonate

- Testicular torsion
- Congenital hydrocele
- Meconium periorchitis
- Birth trauma of the scrotum
- Adrenal hemorrhage with patent processus
- Testicular tumor +/- torsion (teratoma*)
- Paratesticular neuroblastoma

Issues Related to Perinatal Torsion

• Prenatal (already infarcted) vs Postnatal torsion
• Possible delay in diagnosis of torsion
  2° intensity / variability of symptoms
• Accuracy of diagnostic testing
  -15% false negative ultrasound
• Potential for bilateral torsion ➔ anorchia
  simultaneous or asynchronous
• Yerkes* -18 pts with unilateral torsion,
  but 22 % had asymptomatic,
  unsuspected contralateral torsion
• Risks of neonatal anesthesia

Perinatal Torsion

• unsuspected contralateral torsion - hydrocele or normal exam or “normal” flow on doppler ultrasound

• **Contralateral torsion can not be excluded accurately**
  ➔ urgent exploration is warranted, unless there is increased anesthetic risk.
Problem # 4

• Nurse pages you “Baby Boy Thibodeaux hasn’t voided”
Urinary Retention in the Newborn - Tips

• VS oliguria - physiologic or due to hypovolemia / sepsis – check ultrasound

• Urinary retention due to
  occult neurogenic bladder – spinal anomaly eg sacral agenesis -IIDDM
  Boys -urethral obstruction – posterior urethral valves / atresia
  Girls- hydrometrocolpos - 2°urogenital sinus / cloaca

Cath insertion - some will pick smallest catheter - causes urethral trauma
  ➔ pick the largest appropriate size & don’t inflate balloon until all the way in

• Trial of voiding and then check residuals post void < 20ml (depending on GA)
REFERENCES

- Roth CC, Mingin GC, Ortenberg J. Salvage of Bilateral Perinatal Testicular Torsion. J Urol 185:6 2464-2468