Hydronephrosis in the Neonate

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Objectives

• Review kidney development and embryology
• Be familiar with etiologies of hydronephrosis
• Understand the risk factors, evaluation and management of hydronephrosis in the neonate
• Know when referral is appropriate

Case #1

38 week infant admitted to the NICU for known severe prenatal hydronephrosis
  • Routine prenatal history, normal amniotic fluid levels
  • Possible etiologies?
  • Management?

Case #2

Former 28 week premature infant now 4 weeks old, feeding and growing in the NICU, diagnosed with a febrile UTI
  • Possible etiologies?
  • Management?

Case #3

10 do female in your office for routine newborn check
  • Prenatal finding of ‘cystic kidney’
  • Possible etiologies?
  • Management?

Case #4

Parents in your office for a prenatal visit
  • 30 weeks gestation, severe oligohydramnios diagnosed at 18 weeks
  • Fetus with severe bladder distention
  • Bladder stent placed at 22 weeks
  • Possible etiologies?
  • Management?
Embryology

Gross Anatomy

• Hilum
  o Pelvis
  o Arteries
  o Veins
• Parenchyma
  o Cortex
    • Majority of glomeruli
  o Medulla
    • Collecting tubules

*Embryology illustrations courtesy of *The Developing Human*, Moore & Persaud

Parenchyma

1. Nephron
   - Glomerulus and Bowman’s capsule
   - Tubules
     o PCT, loop of Henle, DCT
2. Collecting Tubule
   - Cortical
   - Medullary

Development of the Kidneys and Ureters

• Embryonic development includes 3 sets of excretory organs
  o 2 sets of primitive kidneys
    • The pronephros
    • The mesonephros
  o 1 set of permanent kidneys
    • The metanephros

Primitive Kidneys

• Pronephros
  o Quickly degenerate
  o Majority of the ducts are preserved and utilized by the next set of kidneys, the mesonephros
• Mesonephros
  o Appear late in the 4th week of gestation
  o Function as interim kidneys until the permanent kidneys develop

Permanent Kidneys

• Metanephros
  o Begin to develop early in the 5th week
  o Start to function in the 9th week
  o 2 sources
    • Metanephric diverticulum (ureteric bud)
      o Origin of ureter, pelvis, calices and collecting tubules
    • Metanephric mass of intermediate mesoderm
      o Origin of nephrons
Nephrogenesis

Glomerular development
- Week 8
  - Initiation
- Week 10 - 18
  - Gradual increase in glomeruli
- Week 18 - 32
  - Rapid increase in glomeruli
- Week 32
  - Upper limit is reached
  - No further increase in number, only size and function

Nephrogenesis Pearls
- Number of nephrons nearly doubles between 20 and 38 weeks
  - Premature infants may not have a full set of nephrons at birth
- Impact of premature delivery on nephron development is under investigation

Newborn Kidneys
- Term kidneys each contain 800,000 – 1,000,000 nephrons
- Nephron formation is completed by birth (term)
- Function does not reach adult levels until 2 years

Hydronephrosis

Epidemiology
- Prenatal hydronephrosis identified in 1-5% of all pregnancies
  - 35% go on to have post-natally detected pathology
- Majority detected at 20 weeks of later
  - Can be seen as early as 12-14 weeks
- Of those detected in the 2nd trimester, 33% resolve by the 3rd trimester

When to worry
- No consensus on correlation with post-natal anomalies
- Most likely to see post-natal anomalies when
  - Anterior-Posterior Renal Pelvis Diameter (AP diameter)
    - >6 mm at <20 weeks
    - >8 mm from 20-30 weeks
    - >10 mm at >30 weeks
Terms

- Hydronephrosis
  - Renal collecting system dilatation of any degree
- Pyelectasis or Pelviectasis
  - Dilatation of the renal pelvis
- Caliectasis / Pelvicaliectasis
  - Dilatation of the renal calyces / pelvis
- Mild, moderate, severe

Radiologic Grading

- Hydronephrosis / VUR grading
  - Grade 1: Renal pelvis diameter ≤ 6 mm
  - Grade 2: Renal pelvis diameter > 6 mm and ≤ 9 mm
  - Grade 3: Renal pelvis diameter > 9 mm

New Nomenclature

- New proposed nomenclature to describe dilatation of the urinary tract
- Uses descriptive terms to allow for standardization
- Applies to both pre and post-natal imaging


PRE-NATAL RISK STRATIFICATION

- UTD P1: LOW RISK
- UTD P2: INTERMEDIATE RISK
- UTD P3: HIGH RISK

POST-NATAL RISK STRATIFICATION

- UTD P1: LOW RISK
- UTD P2: INTERMEDIATE RISK
- UTD P3: HIGH RISK

POST-NATAL MANAGEMENT

- UTD P1: LOW RISK
  - VUR: Not recommended
  - Renogram: Not recommended
  - Abx: Not recommended

- UTD P2: INTERMEDIATE RISK
  - VUCG: Recommended
  - Antibiotic prophylaxis: 1. > grade 3 VUR 2. Pending VCUG
  - If VUCG negative and persist >3 months of age, proceed with Renogram

- UTD P3: HIGH RISK
  - VUCG: ASAP
  - Antibiotic prophylaxis: 1. > grade 3 VUR 2. Pending VCUG
  - IF VUCG negative, then proceed with Renogram at 2-3 months of age
Parenchymal thinning
Bladder wall thickening
Etiologies
1. Vesicoureteral Reflux
2. Obstruction
3. Duplications
4. Multicystic Dysplasia – frequently mistaken for hydronephrosis

Vesicoureteral Reflux (VUR)
• Retrograde flow from the bladder into the ureter
  • Primary:
    o VUR with otherwise normal lower tract
  • Secondary:
    o VUR due to an obstructed or poorly functioning lower tract
Epidemiology of Reflux

- 1-2% of population
  - Likely an underestimate
- 17-37% prenatally diagnosed hydronephrosis
- 25-33% of siblings with VUR
  - Unclear inheritance pattern

Physiology of Reflux

- The ureterovesicular junction fails to act as a one-way valve
  - VUR risk is related to the length of the ureter within the bladder wall:
    1. Reflux likely
    2. Reflux possible
    3. Reflux unlikely

Reflux Grading

Reflux graded 1-5
1. Ureter only
2. Ureter and pelvis, no dilatation
3. Mild dilatation of pelvis
4. Moderate dilatation pelvis and calyces, mild blunting; ureter may be dilated
5. Tortuous dilated ureter with severe dilatation of the pelvis and significant blunting of calyces

Imaging

- Reflex can be present in the absence of hydronephrosis
  - Renal ultrasound is insufficient
- Voiding Cystourethrogram
  - Scout KUB
  - Bladder catheterization, fill with contrast
  - Fluoroscopy before and after catheter removal, ideally including voiding phases

VCUG

- Right grade 5
- Left grade 2

Natural History of VUR

- VUR often resolves with time
- As the bladder and ureter grow the intravesicular (bladder) portion of the ureter grows and reflux may resolve
**VUR and UTI's**

- Places child at risk of recurrent UTI, pyelonephritis, and kidney damage
  - 25-40% of children with a UTI who underwent VCUG had reflux

**Morbidity of Reflux**

- Repeated UTI’s can lead to renal scarring
- Significant renal scarring can lead to decrease in renal function
- NAPRTCS 2008 – 5.2% transplanted patients had reflux nephropathy

**Obstruction**

Blockage may occur at:
- Ureteropelvic Junction (UPJ)
- Ureterovesicular junction (UVJ)
- Urethra (Posterior Urethral Valves)

**Ureteropelvic Junction Obstruction**

- Most common cause of antenatal obstruction
- 1:500 (with routine prenatal ultrasounds)
- Male:Female ratio 3:1
- Left > Right
- 10% bilateral

**UPJ**

- **Etiology**
  - Intrinsic narrowing
    - Ureteral stenosis or hypoplasia (upper segment)
    - Kinked ureter
  - Extrinsic compression (10%)
    - Crossing vessel entrapment
  - Renal rotation or ectopy
- **Diagnosis**
  - Suspected on ultrasound
  - Confirmed on Iodixanol renogram
UPJ on lasix renogram

Ureterovesical Junction Obstruction

- Second most common cause of prenatally detected urine obstruction
- 1:10,000 births
- Male:Female ratio
  - 1.2-4.8:1.0
- L > R Ureter
- 10-20% Bilateral

UVJ

- Etiology
  - Narrowed distal ureter with normal UV junction
  - Abnormal ureteral peristalsis
  - Primary megaureter
  - Blockage with stone or mass
- Diagnosis
  - Suspected on ultrasound
  - Confirmed on lasix renogram

UVJ on lasix renogram

Posterior Urethral Valves

- Reports of incidence vary
  - 1: 5,000-8,000 live births
  - Up to 1:25,000
- Retention of embryonal mesonephric duct tissue in the prostatic urethra
- Most common cause of male lower urinary tract obstruction

PUV

- Most often diagnosed prenatally
  - Oligohydramnios
  - Distended bladder
- May present with UTI after birth
- May have associated reflux
- May have significant renal impairment
Multicystic Dysplasia

- Most common cause of an abdominal mass in newborns
- 1:4000 live births, sporadic
- Variant of renal dysplasia
  - Multiple non-communicating cysts of varying size
  - Absence of normal parenchyma
  - Results in non-functional kidney

MCDK

- Results from abnormal interface between ureteral bud and Metanephric mass of intermediate mesoderm
- Usually involutes (54% within 5 yrs)
- Associated anomalies in 50%
  - Reflux, obstruction
  - Rare associations
    - Hypertension
    - Malignancy

Management

Repeat ultrasound after birth (within 48 hours), if hydronephrosis persists:
1. Consider need for VCUG urgently
2. Check BUN / Cr, electrolytes if appropriate
   - VUR:
     - Prophylactic abx (if ≥ grade 3 or UTI): amoxicillin 10-20 mg/kg PO QD
     - Urology / Nephrology consult
   - Multicystic Dysplastic kidney:
     - Nephrology consult for VCUG or renogram
   - Obstruction (UPJ, UVJ, or PUV):
     - Urology consult, consider prophylactic abx, possible Foley placement

Delivery considerations

- Where does the baby need to deliver?
  - Will there be a need for urgent urological intervention?
- Local delivery
  - Renal ultrasound within 48 hours after birth
  - VCUG based on ultrasound findings

Case #1

38 week infant admitted to the NICU for known severe prenatal hydronephrosis
- Routine prenatal history, normal amniotic fluid levels
- Possible etiologies?
  - Reflux, UPJ/UVJ
- Management?
  - Renal ultrasound and VCUG
  - Check serum creatinine
Case #2

- Former 28 week premature infant now 4 weeks old, feeding and growing in the NICU, diagnosed with a febrile UTI
- Possible etiologies?
  - reflux
- Management?
  - Renal Ultrasound and VCUG (after treatment)

Case #3

- 10 day female in your office for routine newborn check.
  - Prenatal finding of “cystic kidney”
- Possible etiologies?
  - MCDK, hydronephrosis, other cystic disease
- Management?
  - Renal Ultrasound
  - Consideration of VCUG or I.V. contrast renogram based on US findings

Case #4

- Parents in your office for a prenatal visit
  - 30 weeks gestation, severe oligohydramnios diagnosed at 18 weeks
  - fetus with severe bladder distention
  - Bladder stent placed at 22 weeks
- Possible etiologies?
  - Posterior Urethral Valves, neurogenic bladder
- Management?
  - Delivery at center with urology and nephrology available

Questions?