Fetal Urinary Tract Anomalies: Common and Uncommon

- GU tract – 1:250 – 1:1000 the most common site (30%) of anomalies in fetus
- Close to 90% of GU anomalies detected in Eurofetus were detected antenatally with US. (Levis, S, USOG, 2003)

Ruth B. Goldstein, MD
Chief, Ultrasound
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Embryology
- 5th week – ureteric bud arises from lower mesonephric ducts
- Metanephric blastema induced by ureteric bud (reciprocal)
  - Ureteric bud → collecting system
  - Metanephric mesenchyme → nephron

New nephrons form till 34 wks

Amniotic Fluid Homeostasis
- 1st trimester – transudative fluid
- 2nd trimester – skin keratinizes fetal functions (GU and GI)

Fetal Urination and Fetal Swallowing control
Reconstitution of amniotic fluid depends on GU tract

Necessary for life and survival
- 1 functioning kidney, and a way to get urine into sac

Standard Obstetrical Sonogram
1. Normal amniotic fluid
2. Fluid filled bladder
3. 2 kidneys (in the flank)

Kidneys (and renal arts) ascend and rotate 6-9th week

Anomalies of Rotation + Position

sprojects.mni.mcgill.ca
**Normal Anatomy and Development**

- Tubular function starts ~ 14th week
- Fetal kidneys identified on sono as early at 12-14 wks
- Bladder usually identified by 12 weeks
- Normal AFV reflects normal urination after 16 wks
- After 16 wks, normal amniotic fluid implies at least 1 kid

**Renal Agenesis**

- Unilateral – 1:500 live births (compatible w/ life)
- Bilateral – 1:4000 live births

Anhydramnios after 17 wks
No kidneys, no bladder
“Lying down adrenals”

**Urine production incr. 11X with GA 20-41 wks**
(5 ml/hr to 56 ml/hr at 41 wks)

Kidney size incr. with GA
(~ 1 mm per week)

Lee RS, et al.

**Fetal HYDRONEPHROSIS**

1:100 pregnancies

Degree Fetal HN | ANY Postnatal Path.
--- | ---
Mild | 11.9%  
Moderate | 45.1%  
Severe | 88.3%  

Only 36% of fetuses with prenatally diagnosed hydroneph. had postnatal pathology confirmed.
Society of Fetal Urology 1993
Grading of Postnatal Hydronephrosis

0 - intact central renal complex plus normal parenchyma
1 - slight central renal complex splitting
2 - central renal complex splitting within renal border, major calyces
3 - wide splitting of central renal complex + pelvis dilated outside renal border + calices uniformly dilated (normal parenchyma)
4 - large dilated calices + renal pelvis + parenchyma thin (ie, less than half that of the contralateral kidney).

If bilateral hydro, <4 mm is the cut off for parenchymal thinning.

Longpre, et al, J Ped Urol, 2011 (Vancouver)
Outcome after Antenatal Hydronephrosis: A Multivariate Analysis.
Fetal hydronephrosis in 100 pts w f/u up 34 mos Outcome retrospectively correlated w antenatal dx APD (mm), Soc. Fetal Urol grade

- 53% resolved, 23% uncomplicated persistent hydro- followed
- 24/100 children had surg. for UPJ
- APD < 1.93 – 83% resolved
- Resolved hydro --- Mean APD = 0.94 cm
- Pyeloplasty --- Mean APD = 2.9 cm

Rule #1 – Mild hydronephrosis creates needless worry

Hydronephrosis – figure out the level

1. What is dilated? (ureters? bladder?)
2. Bilateral vs. Unilateral
3. Is the amniotic fluid adequate?
4. Repeat sono later if possible (many resolve)

UPJ Obstruction
(hydro, no ureter, normal UB + AFV)

1. Males > females
2. 10-30% of prenatal hydro
3. 90% of significant unilateral hydro
4. Cause may be intrinsic or extrinsic
5. Many resolve postnatally (69%)
6. Only 19-28% of prenatally Dx UPJ obst. require surg. interv.**

Rule #2 - Not much danger from a unilat. UPJ obstruction.

Paranephric urinoma

Featureless cystic mass “touching” the spine

- UPJ most common
- Slightly better if PUU/male.

Rule #3 – These kidneys are usually damaged

Ureterectasis

Causes:
- UVJ obstruction, primary megaureter
- Reflux
- Ureterocele
- Bladder outlet obstruction
Duplicated Kidney

Weigert-Meyer Rule

Upper pole obstructs
Lower pole refluxes

Ureterocele

Bilateral Ureterectasis

Bladder Outlet Obstruction (BOO)
(Lower Urinary Tract Obstruction (LUTO))

2.2/10,000 live births

Bladder Outlet Obstruction

Causes –
* Posterior urethral valves, urethral atresia, cloaca, ureterocele
* Females – cloaca, megacystis microcolon (dysfn sm. m. UB, SB)
* In 1st trimester – more likely to be atresia than valves
* Urethral atresia – worse outcomes than PUV

Males – 60-70 % PUV, 40% atresia and others**

Prognosis

Amniotic fluid – Oligo assoc. with worse prognosis
Hydronephrosis/ureters (worse hydro ≠ worse outcomes)
Renal cortex

Hyper-echogenic, loss of CM diff., cysts

Path = Fibrosis + reduced # glomeruli

1:5000-8000 births, the most common cause of chronic lkd ds due to LUTO

Posterior Urethral Valves

“Keyhole” posterior urethra

Posterior urethra

Valve


Kilby et al 2007


13w2d
Prognosis of fetuses with LUTO

Best performers:
- Renal cortex (echogenic or cystic) c/w bx or autopsy
  70-87% sensitive, 78-100% specific
- Oligohydramnios – LR 1.3-9, Cortical appearance LR 1.3-9
- Less predictive - GA at Dx, megacystis, degree of hydro –

Cysts are 100% predictive of dysplasia


Posterior Urethral Valves

- 25-50% of survivors will develop renal failure by 10 yrs
- 60% of children undergoing renal transplant had PUV

Most important prognostic feature = Renal Dysplasia

Fetal Bladder Tap

Normal fetal urine is hypotonic to serum.
(Na< 100 mEq/L, Cl < 90 mEq/L, osm < 210 mOsm/L)
- Na+ > 100 mgl/dL
- Calcium > 8 mgnL
- B2 microglobulin > 6 poor, >13 mg/L = fatal

Serial taps recommended to assure “fresh” urine.
We will not predict dysplasia with 100% accuracy.

Fetal Intervention for BOO

1. Vesico-amniotic shunt
2. Vesicostomy
3. Puncture of valves endoscopically

Experimental -- Mixed results

Congenital Megalourethra

Promosnhti and Visesindh Fetal Diagn Ther 2010
- 28 cases reported
- 12 (43%) had assoc. anomalies (most imperf anus/anal atresia)
- 4 vertebral/ limb anomalies
- 4 neonatal deaths
- 7 TOP

2 (7%) resolved completely = Normal
16(57%) had favorable outcomes at delivery

Prognosis ➔ AFV and other anomalies, esp. kidneys
Normal amniotic fluid but no bladder visualized

Bladder Exstrophy
- 1:10,000 – 50,000 live births
- Males>females 4:1
- External mass in. f to cord insertion
- Low cord insertion
- Kidneys and ureters usually normal
- Epispadius (Epispadius Extrophy complex)

Congenital Cystic Disease of Kidney

Potter’s Classification
I. AR - PCKD (Infantile)
II. MCDK
III. AD – PCDK (adult)
IV. Dysplasia assoc. with LUTO

90% are assoc. with obstruction

Dysplasia
- Abnormal metanephric differentiation
- Disorganized tissue
- Often metaplastic tissues (ie, cartilage)

MCDK
- Non functioning kidney
- Size = proportional to size/# cysts

• Bilateral MCDK – lethal
• Unilateral MCDK – compat w/normal life if other is ok
• Most involute in 1st year – no reason to remove.

MCDK / empty bladder/ oligo
== MCDK + renal agenesis

Sponge Kidney

Disease of collecting tubules
I. AR - PKD (Infantile)
II. MCDK
III. AD – PKD (adult)
IV. Dysplasia assoc. with LUTO
Autosomal Recessive Polycystic Kidney Ds
1:20,000 live births

- Temporal penetrance
- Some not associated with oligo
- Hepatic fibrosis and kidney ds associated

ARPCKD – a Ciliopathy

- Mutation polycystic kidney hepatic ds 1 (PKHD1) on chrom 6p12
- PKHD1 encodes the protein fibrocystin-polyductin
- Affects primary cilia of epithelial cells
  (antennae for orienting dev cells and mitosis)
  -- Lining renal tubules/collection ducts and intrahepatic bile ducts
  -- Dysfn leads to abnl ciliary signaling resulting in abnl architecture

Ciliopathies
ARPKD – renal collecting duct
ADPKD – renal tubule homeostasis
Others – Nephronophthisis, glomerulocystic ds, medullary sponge
Liver – fibrocystic ds caused by ductal plate malformation
  CHF in ARPKD, Caroli syndrome (with CHF), polycystic liver ds

Thank you!