THE FETAL GENITOURINARY TRACT

Beverly G. Coleman, MD
Emeritus Professor of Radiology
Perelman School of Medicine
University of Pennsylvania
Director of Fetal Imaging
The Children’s Hospital of Philadelphia
Philadelphia, Pennsylvania

LEARNING OBJECTIVES
After completing this presentation, the learner will be able to discuss:
1. The normal sonographic appearance of all of the organs and structures that constitute the fetal GU tract
2. A systematic approach to analyzing fetal scans referred for suspected GU tract anomalies
3. Tips for recognizing renal developmental variants
4. Distinguishing features of urinary tract obstruction compared to cystic renal disease

OUTLINE
- The Normal Urinary Tract
- Renal Developmental Variants
- Urinary Tract Obstruction
- Cystic Renal Disease

THE NORMAL URINARY TRACT
- Viz at 11-13 wks (TV) and 14-16 wks (TA)
- Corticomedullary differentiation at 16-18 wks
- Exponential in size with GA; RC/AC ratio = 0.27 - 0.30; Nomograms for renal length 14-42 wks & renal volume 15-42 wks
- Increased conspicuity by 3rd trimester with thicker cortex, renal sinus/perinephric fat, best corticomedullary differentiation
- Color Doppler renal arteries & veins

THE NORMAL URINARY TRACT
Normal Fetal Kidneys
- 14 weeks
- 20 weeks
- 30 weeks
- 8 MHz TV
- 9 Mhz
- 12 Mhz
**THE NORMAL URINARY TRACT**

Normal CM Differentiation at 27 weeks

![Image of normal CM differentiation at 27 weeks]

**The Normal Fetal Bladder**
- Anechoic, midline, anterior, pear shaped with a thin wall
- First structure to be visualized at 9-10 wks on both TA & TV scans
- Fills and empties every 25-30 minutes
- Max volume from mean of 1 mL at 20 wks to 36 mL at 41 wks
- Color Doppler —— umbilical arteries

**THE NORMAL URINARY TRACT**

The Normal Fetal Bladder
- **Collapsed**
- **Distended**

**THE NORMAL URINARY TRACT**

Normal Fetal Adrenals
- Disc shaped, “ice cream sandwich”
- Viz at 20-30 wks, length with GA
- Nomograms – length, volume, weight, etc.

**Normal Fetal Gender**
- Male, female identical to 11 wks; phallus direction “angle of the dangle”
- Testicles descend after 25 wks; both >97% after 32 wks; Small hydroceles common in 15% of normal males
- Uterus can be viz late; ovaries rarely ever viz

**THE NORMAL URINARY TRACT**

The Normal Genitalia
- **XY 23 wks**
- **XX 28 wks**

![Images of normal fetal adrenals and genitalia]
**THE NORMAL URINARY TRACT**

**Importance of Fetal Gender Assessment**
- Confirmation for specific structural anomalies
- Family history of X-linked disorders
- Familial syndromes with genital anomalies
- Gender assignment in multiple gestations
- Exclude maternal cell contamination on amnio

**Fetal Urine Production**
- Begins at the 9th week of embryonic life
- Varies with gestational age: 2-5 ml/hr at 20 wks — 10 ml/hr at 30 wks — 28 ml/hr at 40 wks
- After 14 wks, 2/3's of the amniotic fluid is derived from fetal urination and 1/3 from pulmonary fluid

**Amniotic Fluid**
- Before 10 wks, AF = ultrafiltrate maternal plasma
- 10-20 wks resembles fetal plasma; vol c/w weight
- AFV ave 500ml at 20 wks — 1200ml at 32 wks — 800ml at term
- Amniotic Fluid Evaluation
  - Subjective assessment studies indicate this is very reliable by experienced observers
  - Semi-quantitative Criteria
    - AF Nomograms for 4 Quadrants at 16-42 wks (1990)
    - DVP Method: Vertical Depth of largest single pocket (exclude the umbilical cord & fetal parts)

**Fluid Production**
- Kidneys
- Lung
- Membranes & Cord
- Skin

**Fluid Removal**
- GI Tract
- Lung
- Membranes & Cord

**Approach to GU Tract Analysis**
- Kidneys normal – number, size, location, echogenicity?
- If not, is there unilateral versus bilateral disease?
- If obstruction is present, how severe & at what point?
- Are there renal cysts, where and how significant?
- Are the ureters and bladder visible?
- Are the adrenals and genitalia normal?
- Is amniotic fluid volume normal, increased or decreased?

**Renal Developmental Variants**
- Absent/ectopic tissue
- Abnormal fusion/ascent
  - Renal Agenesis
  - Pelvic Kidney
  - Horseshoe Kidney
  - Crossed Fused Renal Ectopia
Renal Agenesis

**Unilateral**
- Excellent Prognosis!
- AFI in normal range
- One “lying down” adrenal
- +/- Compensatory hypertrophy
- Assoc with VACTERL
  - cardiac, genitai, MSK, GI
  - Recurrence risk 1% (in parents)

**Bilateral**
- LETHAL
- Severe oligo/anhydramnios
- Two “lying down” adrenals
- May be isolated; Numerous Assoc anomalies = 15% cardiac
  - 40% non-cardiac
  - Recurrence risk 4%

Renal Developmental Variants

**Unilateral Renal Agenesis**
- Solitary RK normal in size & echotexture

**Bilateral Renal Agenesis**
- Incidence 1:4000 with 2.5:1 male/female ratio
- Imaging Features = anhydramnios, “absent bladder”
- Adrenals may = size of early kidneys; diff shape
- Pulmonary hypoplasia: MUST measure C/T ratio
  and compare TC to gestational age nomograms
- Fetal movement — talipes, other joint contractures
- Potter’s facies = flat nose, low set ears, micrognathia

“Lying Down” Adrenal Glands

Color Doppler for Absent RA’s & Collapsed Bladder
Bilateral Renal Agenesis

- **VACTERL in Di-Di Twins**

RENAL ECTOPIA

- No kidney observed in the flank, usually unilateral RA (54%)
- Most common forms of renal ectopia:
  1. Pelvic kidney (37%)
  2. Horseshoe kidneys (5%)
  3. Crossed fused (4%)
- +/-Mild to moderate dilatation 2° obst/reflux
- Abnormal morphology 2° malrotation, dysplasia

PELVIC KIDNEY

- Empty renal fossa with flattened “lying down” adrenal gland
- Kidney can be missed as often similar to bowel in texture, superior to bladder, ± malrotation
- Normal size or ↑ contralateral kidney
- Color Doppler → follow RA to pelvis

PELVIC KIDNEY

- **Is One Kidney Truly Missing?**

PELVIC KIDNEY

- **Left Kidney with Vescoureteral Reflux**

PELVIC KIDNEY

- **Dysplastic LK with Ureterocele**
**PELVIC KIDNEY**

- **Dysplastic Right Pelvic Kidney & LRA**

**HORSESHOE KIDNEY**

- Lower poles connected by isthmus, parenchymal or fibrous; rare in upper pole
- Kidneys more inferior in location 2˚ ascent
- Malrotation is common, clue=anterior pelves
- Incidence=1:400 in the general population
- Associations – T18, XO, VACTERL, various syndromes, etc

**PELVIC KIDNEY**

- **Association with Multiple Anomalies**

**HS KIDNEY in DiGeorge Syndrome**

- **Isthmus & Anterior Renal Pelves**
**HS KIDNEY** in Cloacal Malformation

- *UTD A2-3 in mono-di Twin A*

**Twins with HS Kidney**

- *MCDK involving Right Moiety*

**Dysplastic HS KIDNEY**

**CROSSED RENAL ECTOPIA**

- Ectopic kidney with both on the same side; unilateral empty renal fossa
- Most often L→R, 95% fused; freq malrotated
- Kidney appears large and bilobed; ureter crosses the midline to insert at bladder
- Renal arteries, one usu more inferior
- Associations – vertebral segmentation anomalies, spina bifida, sacral agenesis, etc

**CROSSED RENAL ECTOPIA**

- *Common Vascular Trunk*
CROSSED RENAL ECTOPIA

LK crossed to RK

CROSSED RENAL ECTOPIA

CFRE asso with MCA

“PANCAKE” KIDNEY

DYSPLASTIC “PANCAKE” KIDNEY

TERMINOLOGY

AVOID nonspecific terms like:

- Hydronephrosis
- Pyelectasis
- Pelviectasis
- Uroplasia
- Urinary tract or pelvic fullness/prominence

Multidisciplinary consensus on the classification of prenatal and postnatal urinary tract dilution (UTD classification system)

American College of Radiology (ACR), Reston, VA, USA
American Institute of Ultrasound in Medicine (AIUM), Laurel, MD, USA
American Society of Pediatric Radiology (ASPR), The Woodlands, TX, USA
American Society for Pediatric Endoscopy (ASPE), Washington, DC, USA
American Society for Pediatric Neurology (ASPNL), Silver Spring, MD, USA
American Society for Reproductive Medicine (ASRM), Fairfax, VA, USA
American Urological Association (AUA), Columbia, MD, USA
American Society for Reproductive Radiology (ASRR), Reston, VA, USA
American Society of Reproductive Medicine (ASRM), Silver Spring, MD, USA
American Urological Association (AUA), Columbia, MD, USA

URINARY TRACT OBSTRUCTION

- Ureteropelvic Junction Obstruction
- Ureterovesical Pathology
- Lower Urinary Tract Obstruction
UPJ OBSTRUCTION

- Most common site for prenatal obstruction, 1:2,000 live births
- Tip: RP + calyces with no dilated ureter or bladder
- Several causes — ischemia, stricture, valves, anomalous vessels or crossing bands, high ureteral insertion, muscle abnormality
- Contralateral renal anomaly, 25%; Extra-renal anomalies occur in 10%
- AFV usually normal; “paradoxical” polyhydramnios occurs in 33%
- Bilateral UPJ, incidence ~10%
BILATERAL UPJ

**A2-3 High Risk, Renal Pelves 9 & 10 mm**

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URETEROVESICLE PATHOLOGY

- Complete ureteral duplication with ectopic ureterocele
- Ectopic ureter with single collecting system
- Ureterovesicle junction stenosis
- Congenital megaureter

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Duplicated Collecting System

- Imaging features: intervening band of renal tissue separates upper and lower pole pelves; bilateral in 10-20% of cases
- Asym renal size in unilateral cases; length >95% on the affected side
- Upper pole → ectopic ureter obstructs
- Lower pole → normotopic ureter refluxes
- Scan bladder several times; check external genitalia

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Duplicated Collecting System???

- Duplicated LRA

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Duplicated Collecting System???

- Bilateral Duplicated Main Renal Vessels

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Duplicated Collecting System

- A2-3 SEVERE UP Obstruction & LP Reflux
UNILATERAL DUPLICATION

• A2-3 High Risk LK Obstruction >> Reflux

• A2-3 High Risk with UP Renal Dysplasia

• Dilated Lower Poles and LUP Dysplasia

UNILATERAL DUPLICATION

• A2-3 Severe RK Reflux >>> Obstruction

• A2-3 with Large Obstructing Ureterocele

BILATERAL DUPLICATIONS

• A2-3 with Focal OCD in Polar Regions

• Dilated Lower Poles and LUP Dysplasia

• A2-3 with Large Obstructing Ureterocele

• A2-3 with Focal OCD in Polar Regions
Bilateral Ectopic Ureters

- Ectopic Ureters without Duplicated Systems

CONGENITAL MEGAURETER

- A2-3 UTD with URETER >> BLADDER

In Utero Voiding

- Imaging features = dilated bladder, renal pelvis, calyces and ureters with oligohydramnios; ± dysplasia with cysts; ± urinary ascites
- Degree of collecting system dilatation does not = degree of renal dysplasia
- Causes = posterior/anterior valves, urethral atresia, meatal stenosis, cloacal malformation, etc
- 43% cases have associated anomalies; aneuploidy

Lower Urinary Tract Obstruction

- “Keyhole” Posterior Urethra
- Bladder Volume of 80 ml
Lower Urinary Tract Obstruction

- Patent Urachus with Urinary Ascites

CAUSES of MEGACYSTIS

- Posterior urethral valves
- Urethral atresia/stricture
- Prune belly syndrome
- Megalourethra
- Cloacal malformation
- Megacystis-microcolon-intestinal hypoperistalsis syndrome

PRUNE BELLY SYNDROME
Megacystis Microcolon Intestinal Hypoperistalsis Syndrome

• GU tract obstruction → renal macrocysts and/or microcysts; most common cause is urethral obstruction; ureteropelvic or ureterovesical junction obstruction less common

• Unilateral, bilateral, rarely segmental

• Renal size may be ↑, ↓ or normal

• Worse prognosis is obstruction → OCD before 20 wks

CYSTIC RENAL DISORDERS

• Obstructive Cystic Dysplasia

• Multicystic Dysplastic Kidney

• Autosomal Dominant Polycystic Renal Disease

• Autosomal Recessive Polycystic Renal Disease

Obstructive Cystic Dysplasia

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Obstructive Cystic Dysplasia

Antenatal Predictors of Poor Postnatal Renal Function

**Ultrasound**
- Severe Oligohydramnios esp if early onset
- Increased renal echogenicity
- Renal Cortical Cysts
- Slow Bladder Filling s/p vesicocentesis

**Fetal Urine**
- Sodium Level (Na⁺)
- Calcium Level (Ca²⁺)
- B₂-Microglobulin
- Osmolality

**Fetal Blood**
- B₂-Microglobulin

ECHOGENIC KIDNEYS

- Obstructive Cystic Dysplasia
- Hereditary Cystic Diseases
- Nonhereditary Fetal Diseases
  - Infection, Renal Vein Thrombosis, Toxic Agents
- Syndromes
  - T13, Meckel Gruber, Beckwith Wiedemann
- Chromosomal Anomalies
- Congenital Nephrotic Syndromes
- Maternal & Metabolic Diseases
- Nephroblastomatosis
- Normal Variant

TRISOMY 13
- Echogenic, Enlarged Kidneys with MCA

Beckwith Wiedeman Syndrome
- Nephromegaly + Echogenic with Macroglossia
NORMAL VARIANT

- Echogenic Kidneys with Normal AFI & Renal Size

12 Mhz Linear

9 Mhz Sector

MULTICYSTIC DYSPLASTIC KIDNEY

- Non functioning kidney in 90%; may be late sequelae of renal obstruction
- Variants=pelvic, horseshoe or duplicated kidney
- MCDK usu involutes; rarely complicated by infection, hypertension, Wilm’s tumor,
- F/U scans to check fluid, contralateral kidney
- Neonatal work up required in EVERY case!

Unilateral MCDK

- 80% of cases, L>R renal involvement

Cysts vary in size & may enlarge dramatically

MCDK

FEATURE | MCDK | HYDRONEPHROSIS
--- | --- | ---
Cysts | Random distribution | Calyces in a row
Cyst Size | Quite variable | Uniform unless compound
Communication | None | Calyces → infundibulum
Parenchyma | Islands between cysts | Intact, peripheral to calyces
Renal Shape | Absent reniform contour | Contour preserved

Unilateral MCDK

- Random Cyst Distribution & Patent MRA
**Bilateral MCDK**
- *Di-di twins with replaced renal parenchyma*

**Bilateral MCDK**
- Completely replaced renal parenchyma = LETHAL

**Segmental MCDK**
- MCDK UP Moiety

**Segmental MCDK**
- UP Cystic Dysplasia without duplication

**Polycystic Kidney Disease**

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<thead>
<tr>
<th></th>
<th>Autosomal Dominant</th>
<th>Autosomal Recessive</th>
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<tbody>
<tr>
<td><strong>Size</strong></td>
<td>Often Enlarged</td>
<td>Markedly Enlarged</td>
</tr>
<tr>
<td><strong>Cysts</strong></td>
<td>Cysts +/- viz, usu late</td>
<td>Most below limits of resolution for US</td>
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<tr>
<td><strong>Parenchyma</strong></td>
<td>Preserved medullary region</td>
<td>Echogenic medullary region</td>
</tr>
<tr>
<td><strong>AFI</strong></td>
<td>Normal</td>
<td>Profound oligo, absent fluid</td>
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<tr>
<td><strong>Prognosis</strong></td>
<td>Normal renal fn at birth</td>
<td>Dimal</td>
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**Autosomal Dominant Polycystic Kidney Disease**
- CMD No cysts

- Maternal Kidney

- Kidney
**Autosomal Dominant Polycystic Kidney Disease**

- Scattered Parenchymal Cysts

**Autosomal Recessive Polycystic Kidney Disease**

- Imaging Features = ↑ hyperechoic kidneys, ± 1-2 mm cysts and echogenic parenchyma; kidneys may look normal up to 20 wks
- Kidney Size > 2 SD above the mean for the expected gestational age
- Oligohydramnios/anhydramnios with non-visualized bladder
- Check for signs of pulmonary hypoplasia such as ↓ TC and ↓ C/T ratio without cardiomegaly

**Autosomal Recessive Polycystic Kidney Disease**

- Parents are Known Carriers

**Autosomal Recessive Polycystic Kidney Disease**

- Severe Oligohydramnios

- Severe Biliary Ectasia
Autosomal Recessive Polycystic Kidney Disease

- “MIRACLE CHILD” with normal AFI, micromelia & polydactyly

CONCLUSIONS

- It is important to examine the entire GU tract using a systematic approach with the aim to make the most specific diagnosis possible and/or to narrow the differential as much as possible.
- A thorough knowledge of what is normal or is a normal GU variant is critical in order to minimize patient anxiety.
- Any associated anomalies involving other organs and structures should be carefully imaged and analyzed as GU tract anomalies are common in cases of aneuploidy and various syndromes.

KEY REFERENCES


Metabolic Polycystic Kidney Disease

CONCLUSIONS

- When GU tract obstruction is expected, the most important aspect of the fetal evaluation is to determine the site of the obstruction in addition to assessing the severity of the degree of obstruction.
- A clear understanding of the numerous sonographic features that help distinguish renal cystic disease from obstruction is crucial in making the correct diagnosis in many cases.
- As much information as possible is critical for parents to make an informed decision regarding pregnancy management with all options fully discussed.

KEY REFERENCES
