Multicystic Dysplastic Kidney
and Other Congenital GU Anomalies

Graham Lieberman, HMS IV
Maryellen Sun, MD
Agenda

1. Patient Presentation
2. Pregnancy History
3. Fetal Evaluation
4. 30-week Survey and Imaging Findings
5. Patient Differential
6. Features of MCDK
7. Companion Patient Specimens
8. Continued Antenatal Care
9. Birth and Pediatric Follow-up
10. Patient and Child Outcome and Summary
11. Genitourinary Embryology
12. Companion Patients
Our Patient: Presentation

Demographics:

Name: Patient AB
28 year old woman
G1P0, 30 weeks pregnant
New to this city, establishing care

PM/SH: None
PObH: None
Our Patient: Current Pregnancy

“I’ve had good prenatal care”
Took prenatal vitamins
Normal 1st trimester US
Normal genetic testing

But…
Our Patient: Current Pregnancy

“I’ve had good prenatal care”
Took prenatal vitamins
Normal 1st trimester US
Normal genetic testing

But…

“I never had my 18-week ultrasound…”
Prenatal Care: Fetal Evaluation

*Per ACOG, ACR, and AIUM:*

Fetal Surveys (US):

1\textsuperscript{st} trimester survey is standard
2\textsuperscript{nd} OR 3\textsuperscript{rd} trimester survey is standard

“Limited exam” (US) to follow-up

MRI if needed
Our Patient: 30-week Survey

Our patient underwent a 30-week fetal survey.
Fetus: Enlarged Left Kidney on US

Left Kidney

Courtesy: Dr. Mei-Mei Chow
Fetus: Enlarged Left Kidney on US

Enlarged Kidney

Normal Calyces

Left Kidney

Courtesy: Dr. Mei-Mei Chow
Fetus: Abnormal Right Kidney on US

Right Kidney

Courtesy: Dr. Mei-Mei Chow
Fetus: Abnormal Right Kidney on US

Multiple cysts
Absent calyces

Abnormal Kidney

Right Kidney

Courtesy: Dr. Mei-Mei Chow
Fetus: Bilateral Kidneys on US

Courtesy: Dr. Mei-Mei Chow
Our Patient: Fetal Imaging Findings

Slightly enlarged left kidney
Abnormal right kidney
Enlarged
Multiple cysts

Differential...?
**Differential**

**Hereditary**

- ARPKD (1:20,000 live births)
- ADPKD (1:1000)
- Syndromic (very rare!)
  - Glomerulocystic
  - Medullocystic

**Sporadic**

- Renal dysplasia (1:500)
- MCDK (1:2000)
- Obstructive cysts (varies)
- Cystic tumors (varies)
- Simple cysts (common)
Multicystic Dysplastic Kidney (MCDK)

The fetus has imaging findings diagnostic of multicystic dysplastic kidney.
Agenda

1. Patient Presentation
2. Pregnancy History
3. Fetal Evaluation
4. 30 week Survey and Imaging Findings
5. Patient Differential

6. Features of MCDK
7. Companion Patient Specimens
8. Continued Antenatal Care
9. Birth and Pediatric Follow-up
10. Patient and Child Outcome and Summary
11. Genitourinary Embryology
12. Companion Patients
Features of MCDK

1:1000-3000 live births
M>F
Sporadic (aka non-hereditary)
Associated with CONTRALATERAL renal anomalies, heart defects, GI atresia, spinal column defects

MCKD is the most common cause of neonatal abdominal mass
Features of MCDK II

Usually unilateral, L>R
   Bilateral is usually syndromic
Contralateral kidney hypertrophies
   May have anomalies, most commonly VUR
   Note: in utero, affected kidney is usually larger
Typically asymptomatic
   Surgical removal if symptomatic
Natural history: affected kidney involutes by age 3
   Equivalent in adulthood to renal agenesis
Features of MCDK III

Exact cause is unknown
Renal parenchyma fails to develop
Ureter is usually atretic
Renal mass is urine-filled non-communicating cysts
Originally primitive nephrons
Companion Images: Surgical Specimens

Niaudet, P. Renal cystic diseases in children. UpToDate, Jan 2015.
Agenda

1. Patient Presentation
2. Pregnancy History
3. Fetal Evaluation
4. 30-week Survey and Imaging Findings
5. Patient Differential
6. Features of MCDK
7. Companion Patient Specimens
8. Continued Antenatal Care
9. Birth and Pediatric Follow-up
10. Patient and Child Outcome and Summary
11. Genitourinary Embryology
12. Companion Patients
Our Patient: Follow-Up

Informed Patient AB of anomaly
Baby AB is otherwise normal and healthy
Termination usually not suggested
   Also, too late to do so…
Return in 6 weeks for additional US
Our Patient: 36-wk Limited Exam

Our patient underwent a 36-week limited US exam.
Fetus: Enlarged Left Kidney on US

Left Kidney

5.85 cm LT

Courtesy: Dr. Mei-Mei Chow
Fetus: Enlarged Left Kidney on US

Left Kidney

Normal Kidney

Normal Calyces

5.85 cm  LT

Courtesy: Dr. Mei-Mei Chow
Fetus: Abnormal Right Kidney on US
Fetus: Abnormal Right Kidney on US

Multiple cysts

Absent calyces

Abnormal Kidney

Right Kidney

3 7.33 cm

Courtesy: Dr. Mei-Mei Chow
Our Patient: 36-wk US Findings

Both fetal kidneys are enlarged (should be 3-4cm)
  R 25% > L

Otherwise, Baby AB looks normal

Plan for normal birth, pediatric follow-up
Our Patient: Birth

Baby AB born

Healthy, vaginal delivery, no complications

Serial kidney ultrasounds with pediatric visits
Child: Pediatric Follow-Up

Baby AB underwent serial renal ultrasounds during his routine pediatric visits.
Child: 1 Year Old Left Kidney on US

Left Kidney

Courtesy: Dr. Mei-Mei Chow
Child: 1 Year Old Left Kidney on US

- Left Kidney
- Dilated pelvicalyceal system
- Simple cyst

Courtesy: Dr. Mei-Mei Chow
Child: 1 Year Old Renal Scan

Renal Scan, Furosemide+

Courtesy: Dr. Mei-Mei Chow
Child: Pediatric Management

Right kidney is non-functional
Left kidney has mild hydronephrosis (Grade III)
Likely due to mild VUR
Treat with prophylactic antibiotics until potty-trained
Teach proper bowel/bladder regimen after that
Continue to evaluate over time
Child: Pediatric Follow-Up II

Child AB underwent a 2-year-old renal US
Child: 2-Year-Old Left Kidney on US

Left Kidney

Courtesy: Dr. Mei-Mei Chow
Child: 2-Year-Old Left Kidney on US

Normal Kidney

Left Kidney

Courtesy: Dr. Mei-Mei Chow
Child: 2-Year-Old Right Kidney on US

Right Kidney

Courtesy: Dr. Mei-Mei Chow
Child: 2-Year-Old Right Kidney on US

Involuting renal tissue

Right Kidney

Courtesy: Dr. Mei-Mei Chow
Child AB: Imaging Findings

Right kidney has involuted
Left kidney VUR has resolved
Agenda

1. Patient Presentation
2. Pregnancy History
3. Fetal Evaluation
4. 30-week Survey and Imaging Findings
5. Patient Differential
6. Features of MCDK
7. Companion Patient Specimens
8. Continued Antenatal Care
9. Birth and Pediatric Follow-up
10. Child Outcome and Summary
11. Genitourinary Embryology
12. Companion Patients
Child AB: Long-Term Outcome

Child AB at 2 years of age has a healthy, fully functional left kidney. The VUR has fully resolved. The right kidney has involuted and has been asymptomatic throughout childhood. Baby AB is otherwise completely healthy and has the same long-term prognosis as in renal agenesis.
Our Patient: Summary

Our patient, Patient AB, is a 28yo G1P0 30-weeks-pregnant woman establishing new prenatal care.

1\textsuperscript{st} trimester screening US was normal. 3\textsuperscript{rd} trimester screening US at this encounter revealed an abnormal, enlarged, multicystic right kidney, diagnostic of multicystic dysplastic kidney (MCDK).
Our Patient: Summary

MCDK is a sporadic, congenital, dysplastic kidney disease of unknown origin. It results in a non-functional cystic renal mass that fails to develop normal renal parenchyma. The affected kidney usually involutes in early childhood. The unaffected kidney is usually normal, but there is an increased risk of contralateral renal anomalies as well as cardiac, GI, and spinal defects.

MCDK is the most common cause of neonatal abdominal mass.
Our Patient: Summary

Patient AB’s baby was born healthy. He was found to have a nonfunctional kidney and contralateral Grade III VUR. The VUR self-resolved over the subsequent year.

Baby AB continued to grow and develop normally. The prognosis is the same as in renal agenesis.
Agenda

1. Patient Presentation
2. Pregnancy History
3. Fetal Evaluation
4. 30-week Survey and Imaging Findings
5. Patient Differential
6. Features of MCDK
7. Companion Patient Specimens
8. Continued Antenatal Care
9. Birth and Pediatric Follow-up
10. Patient and Child Outcome and Summary

11. Genitourinary Embryology
12. Companion Patients
Urogenital Embryology

5 weeks
- superior mesenteric artery
- adrenal gland & artery
- mesonephric duct
- mesonephros
- inferior mesenteric artery
- common iliac artery
- renal artery
- metanephric blastema
- ureteric bud (from mesonephric duct)
- allantois
- cloaca

6 weeks
- adrenal gland
- mesonephros
- genital ridge
- renal artery
- kidney
- mesonephric duct
- ureter
- bladder
- urogenital sinus

Case courtesy of Dr. Matt Skalski, Radiopaedia.org
Urogenital Embryology

7 weeks

- adrenal gland
- kidney
- site of previous renal artery
- renal artery
- gonad
- mesonephric duct
- ureter
- bladder
- urogenital sinus

8 weeks

- adrenal gland
- renal artery
- kidney
- sites of previous renal arteries
- gonad
- mesonephric duct
- ureter
- bladder
- urogenital sinus

Case courtesy of Dr. Matt Skalski, Radiopaedia.org
Companion Patient (CP): Congenital Anomalies

Horseshoe Kidney
Crossed-Fused Renal Ectopia
Faceless Kidney
Renal Hypoplasia
Renal Malrotation
Thoracic Kidney
Supernumerary Kidney
Urachal Diverticulum
Urachal Abscess
Undescended Testicles
CP1: Horseshoe Kidney on CT and US

Axial C+ CT

Sag IVC Mid

Doppler Sag IVC Mid

Isthmus

Courtesy: Dr. Maryellen Sun
CP2: Horseshoe Kidney with RCC on MRI

RCC R. Kidney
CP3: Horseshoe Kidney with Carcinoid on CT

Liver lesions

Carcinoid R. Kidney

Isthmus

Coronal C+ CT

LK
CP4: Cross-Fused Renal Ectopia on CT

Axial C- CT

Bowel in left renal fossa

Coronal C- CT

 RK

 LK
CP5: Faceless Kidney on CT

Faceless kidney = absence of renal sinus on imaging. It is frequently associated with duplication.
CP6: L Renal Hypoplasia on IVU and CT

Plain Film Pyelo

Axial C- CT

Functioning hypoplastic left kidney
CP7: L Renal Malrotation on CT

Malrotated left kidney

Axial C+ CT

From: Singer A et al. 2008
CP8: R Thoracic Kidney on CT

Axial C- CT, Soft-Tissue Window

Thoracic right kidney

Axial C- CT, Lung Window
CP9: R Supernumerary Kidney on CT

Coronal C+ CT

Two right-sided kidneys
CP10: Urachal Diverticulum on Cystogram

**Frontal Cystogram**

**Lateral Cystogram**

**Urachal diverticulum due to patent urachus**

The urachus runs from the bladder apex to the umbilicus in the fetus and should obliterate after birth.

Courtesy: Dr. Maryellen Sun
CP11: Urachal Abscess on CT

Axial C- CT

Uracchal abscess

Courtesy: Dr. Maryellen Sun
CP12: Undescended Testicles on US

_empty_ Empty

L Scrotal US R Scrotal US

_left_ right undescended testes

L Pelvic US 30 year old male R Pelvic US

Left, right undescended testes
We discussed:

☑ 1. Patient Presentation
☑ 2. Pregnancy History
☑ 3. Fetal Evaluation
☑ 4. 30-week Survey and Imaging Findings
☑ 5. Patient Differential
☑ 6. Features of MCDK
☑ 7. Companion Patient Specimens
☑ 8. Continued Antenatal Care
☑ 9. Birth and Pediatric Follow-up
☑ 10. Patient and Child Outcome and Summary
☑ 11. Genitourinary Embryology
☑ 12. Companion Patients
Acknowledgments

Dr. Maryellen Sun
Dr. Mei-Mei Chow
Dr. Thomas Anderson
Dr. Marty Smith
Dr. Gillian Lieberman
Joe Singer
References


