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OHSU

Fetal GU Anomalies

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Disclosure

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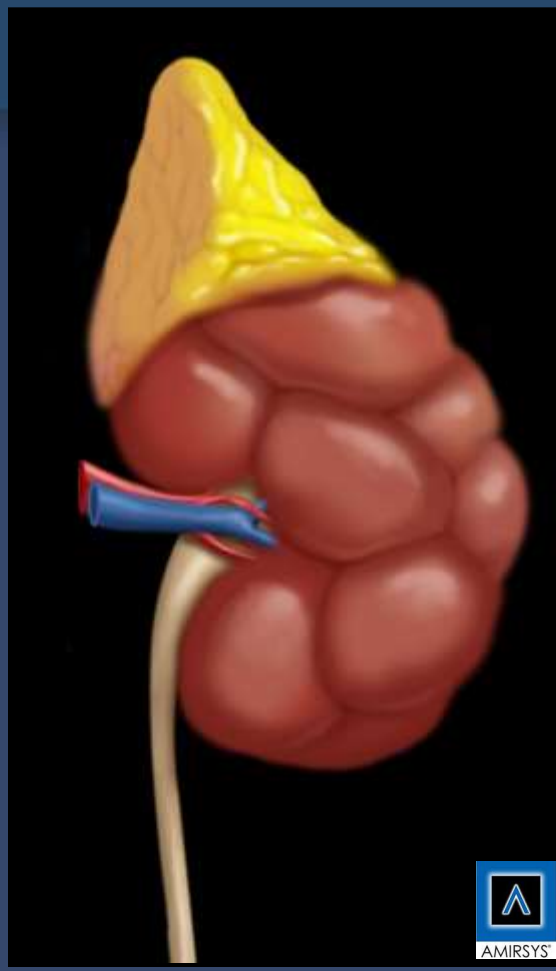
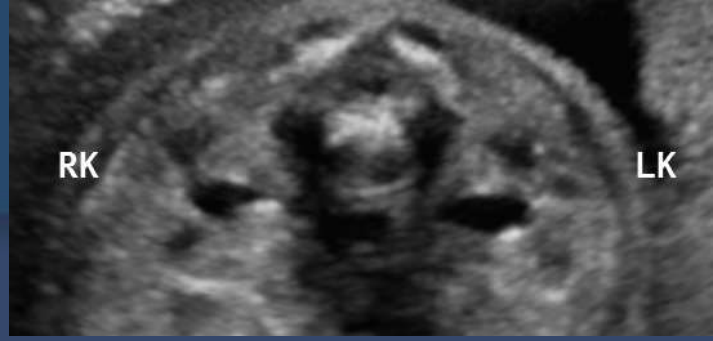
Outline

- Mild hydronephrosis
- Significant hydronephrosis
  - Case-based
- Renal cystic dysplasia
  - Differential diagnosis



Mild Hydronephrosis: causes



- Incidence: 1-5%
- Common benign causes:
  - Maternal progesterone influence on renal pelvis
  - Natural narrowing of UPJ (kinks and folds)
    - Occur early in development & resolve with growth
- Less common significant causes
  - Obstruction and reflux
  - Aneuploidy in 0.3 to 0.9% (most common T21)
    - Likelihood ratio of 1.5-1.9
      - (Society of Genetic Counselors, 2010 guidelines)
    - Amniocentesis?
      - Only in the setting of additional high risk factors/findings

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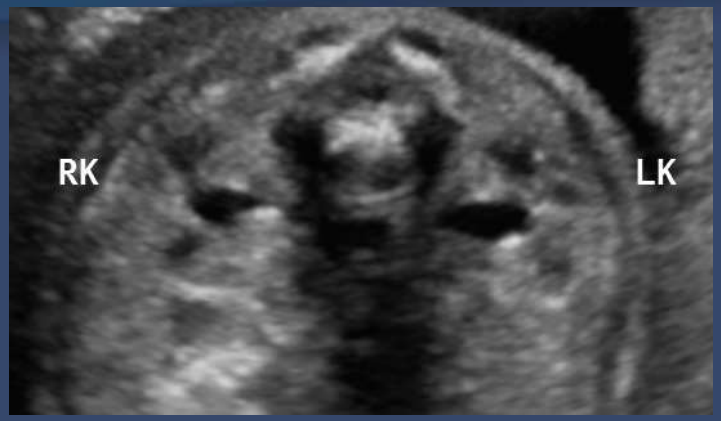
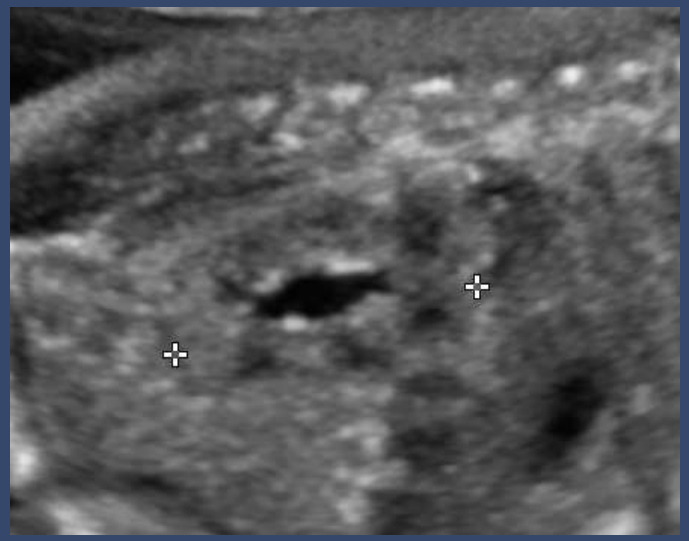
Pelviectasis: Diagnostic criteria

- Increased renal pelvis anterior-posterior diameter
  - Midrenal axial view
- By the numbers:
  - ≥ 3 mm in first trimester
  - ≥ 4 mm at 14-22 weeks
  - ≥ 5 mm at 22-32 weeks
  - ≥ 7 mm after 32 week
- Limitations of using AP diameter only
  - Only evaluating renal pelvis





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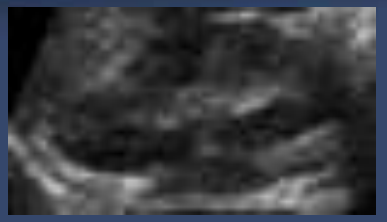
Society For Fetal Urology (SFU) grading system use longitudinal views to asses calyces

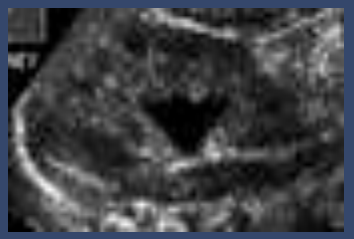
	Descriptors
Grade 0	No fluid in renal sinus fat
Grade 1	Fluid barely splits sinus fat
Grade 2	Urine fills intrarenal pelvis
	Urine fills extra renal pelvis + major calyces
Grade 3	SFU 2 + minor calyces uniformly dilated and parenchyma preserved
Grade 4	SFU 3 + thin or cystic parenchyma

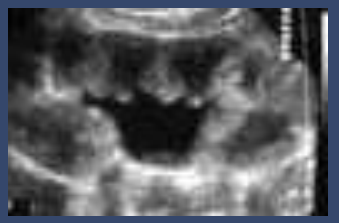
Grade 0



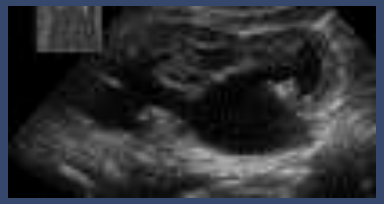
Grade 1

Grade 2

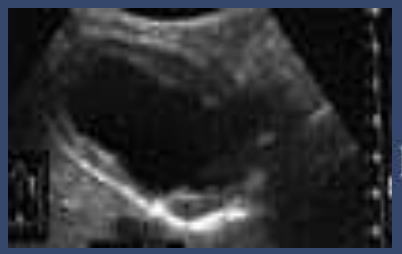




Grade 3



Grade 4



www.uab.edu/images/peduro/SFU/sfu_grading_on_web/sfu_grading_on_web.htm

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Society for Fetal Urology (SFU) grading system

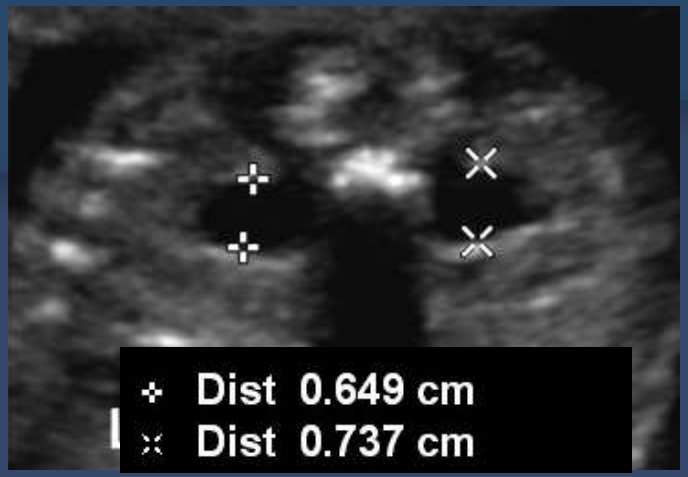
- In pediatric population:
  - Good inter-rater agreement
  - Predictive of outcome

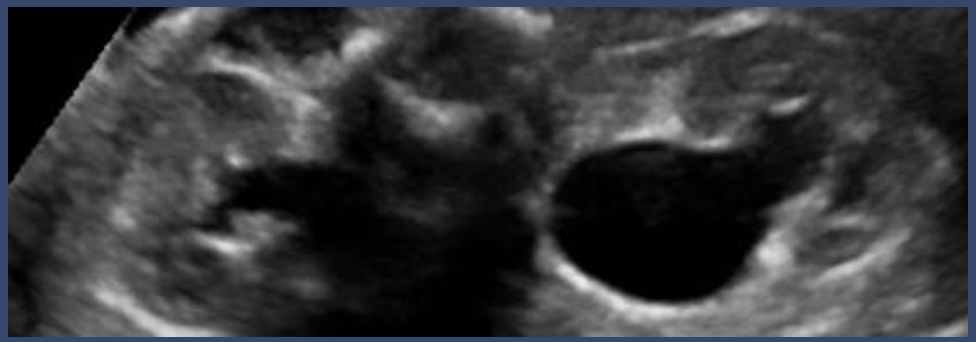
	SFU 1	SFU 2	SFU 3	SFU 4
Stable	100%	87% (144)	30% (37)	0%
Surgery		13%(21)	70% (85)	100%
Outcome	Benign	Benign	Surveillance +/- surgery	

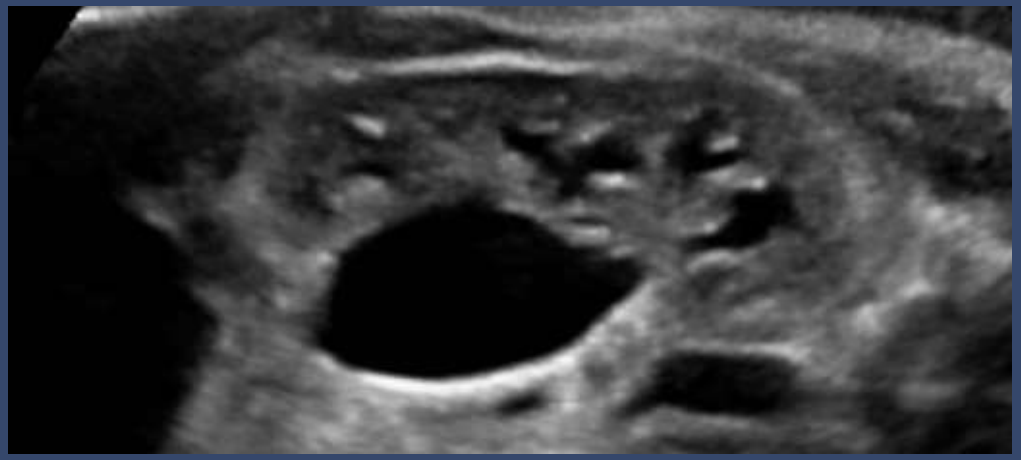
Yang Y, etal, Journal of Ped Surg (2010) 45; 1701-6

- Can we use this grading system for prenatal diagnosis?
  - We don't know yet: good inter-rater agreement
- We should at least use better descriptors

Case example: mild to asymmetric severe







Diagnosis:
UPJ Obstruction

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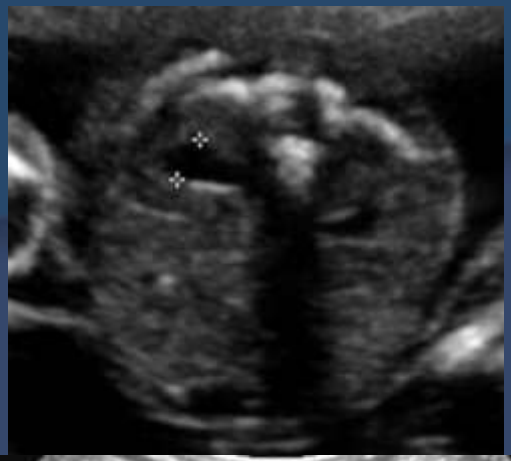
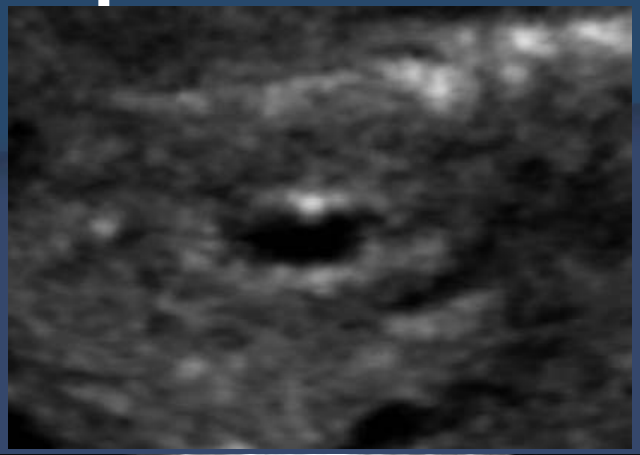
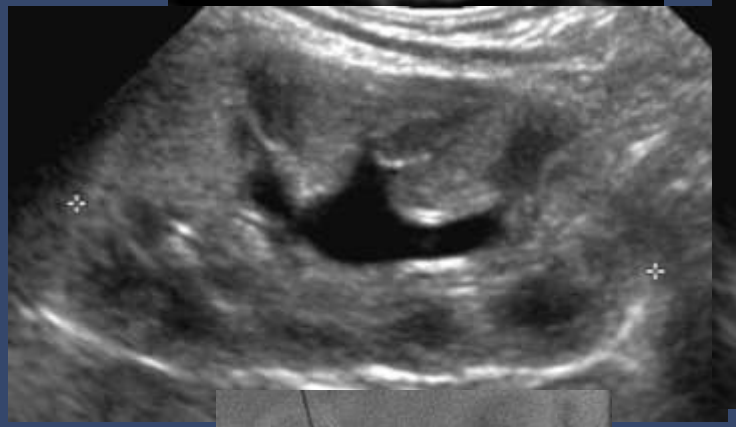
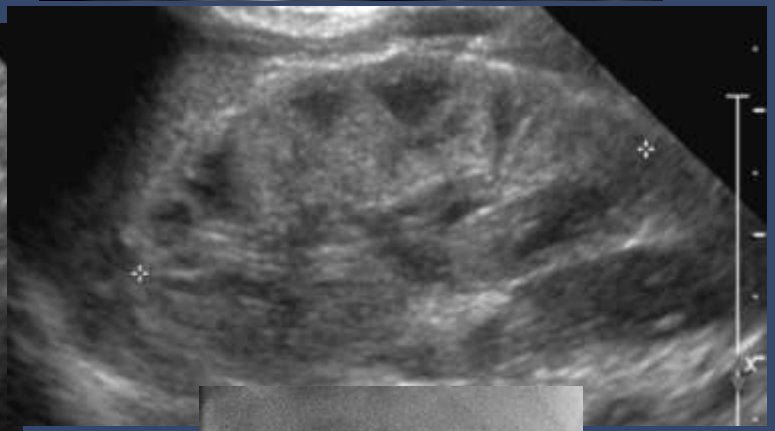
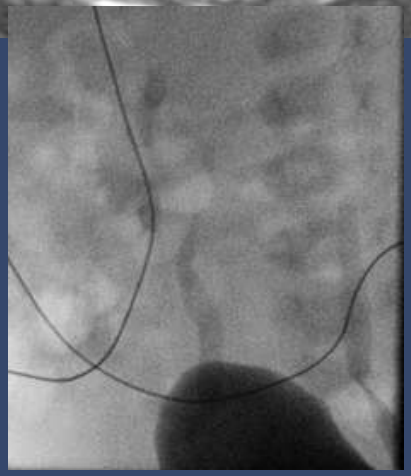
Case Example: Stable persistent mild

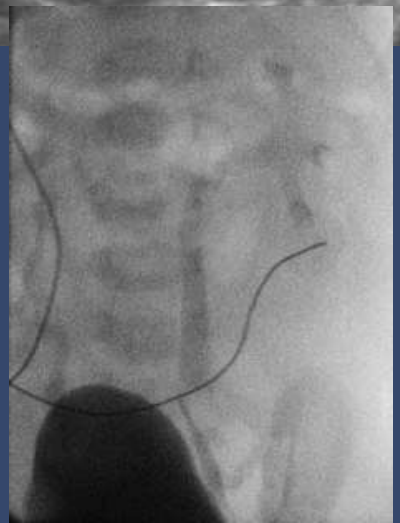












Final Diagnosis: Bilateral reflux

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Pelviectasis Summary

- AP diameter
  - ≥ 4 mm before 22 wks
  - ≥ 7 mm after 32 wks
- Note degree of calyceal dilatation and appearance of parenchyma
  - consider using SFU grading system
- Know your patient
  - Low risk or high risk for aneuploidy and/or obstructive uropathy
- Suggest post natal imaging
  - Ultrasound (preferably after 72 hours)
  - VCUG

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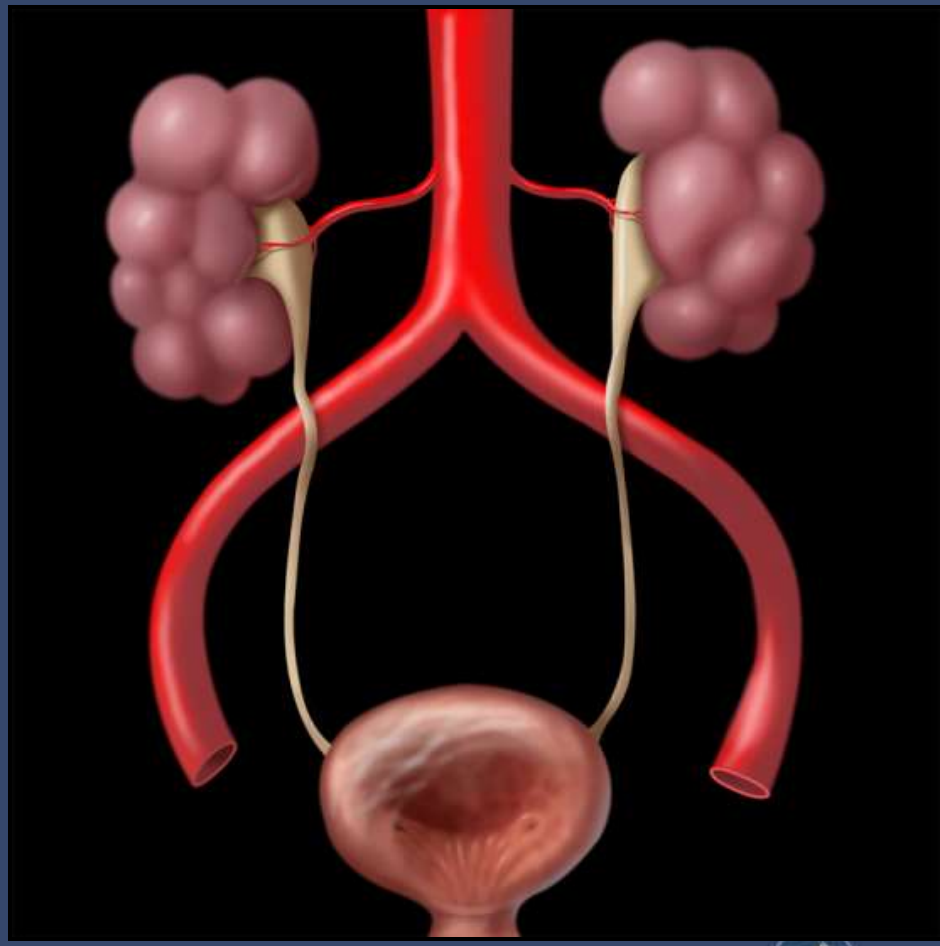
Significant Hydronephrosis: A Case based approach

- Morphology of distention
  - Upper tract
  - Mid tract
  - Lower tract
- Affect on renal parenchyma
- Affect on amniotic fluid



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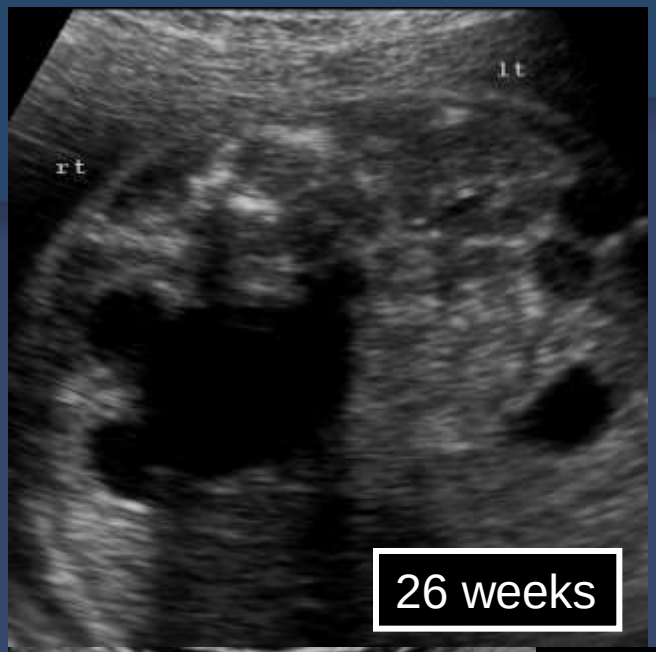
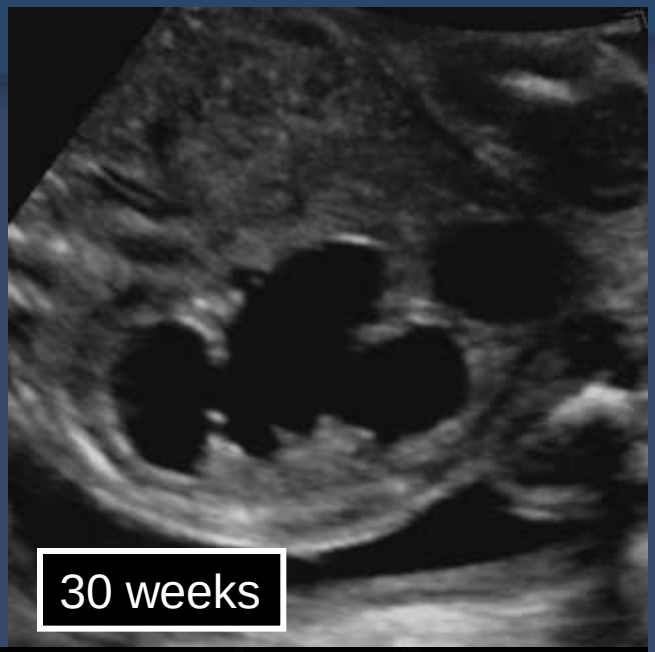
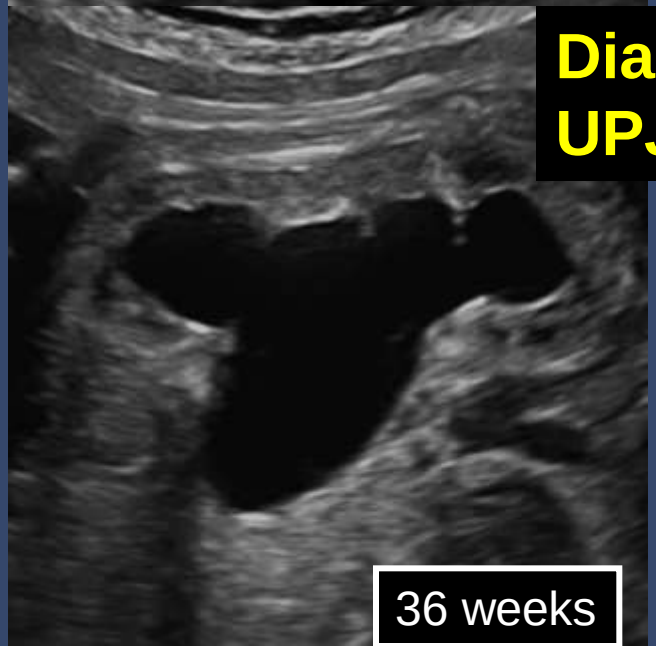
Case Example: severe unilateral hydronephrosis



26 weeks

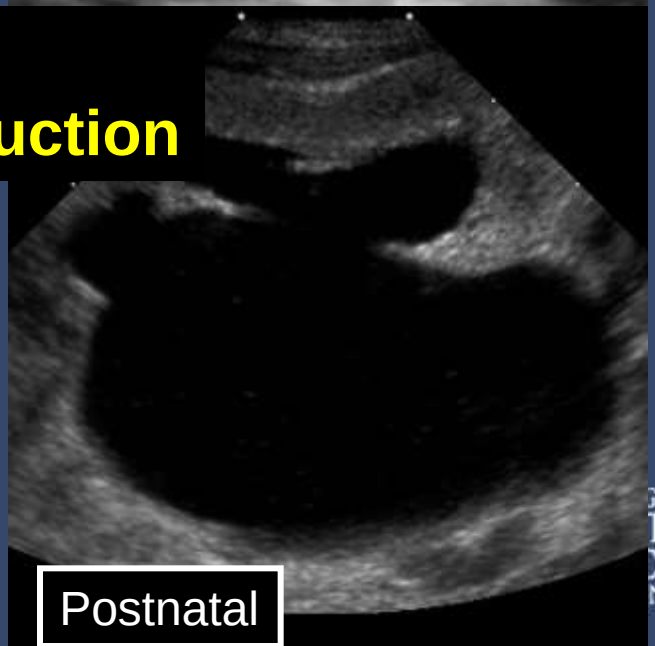


30 weeks



36 weeks

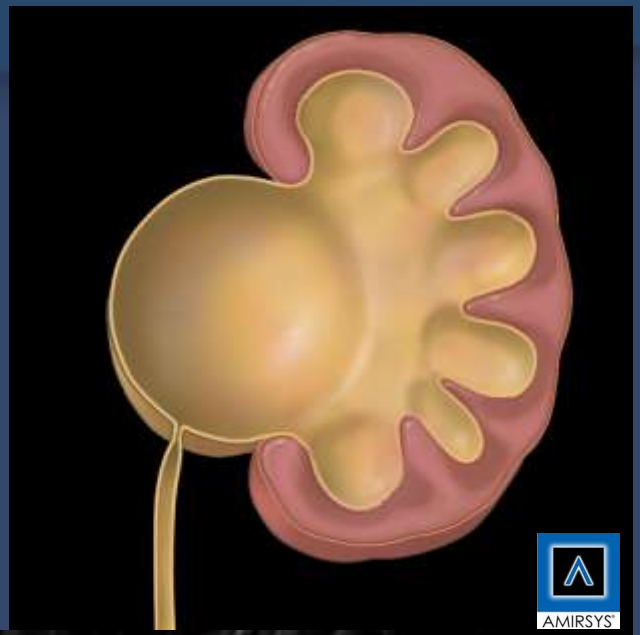
Diagnosis:
UPJ Obstruction

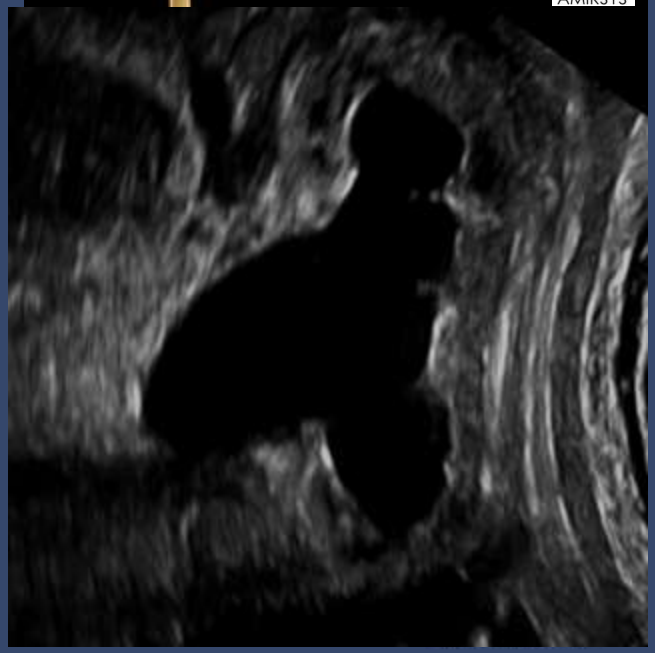


Postnatal

Ureteropelvic junction (UPJ) obstruction

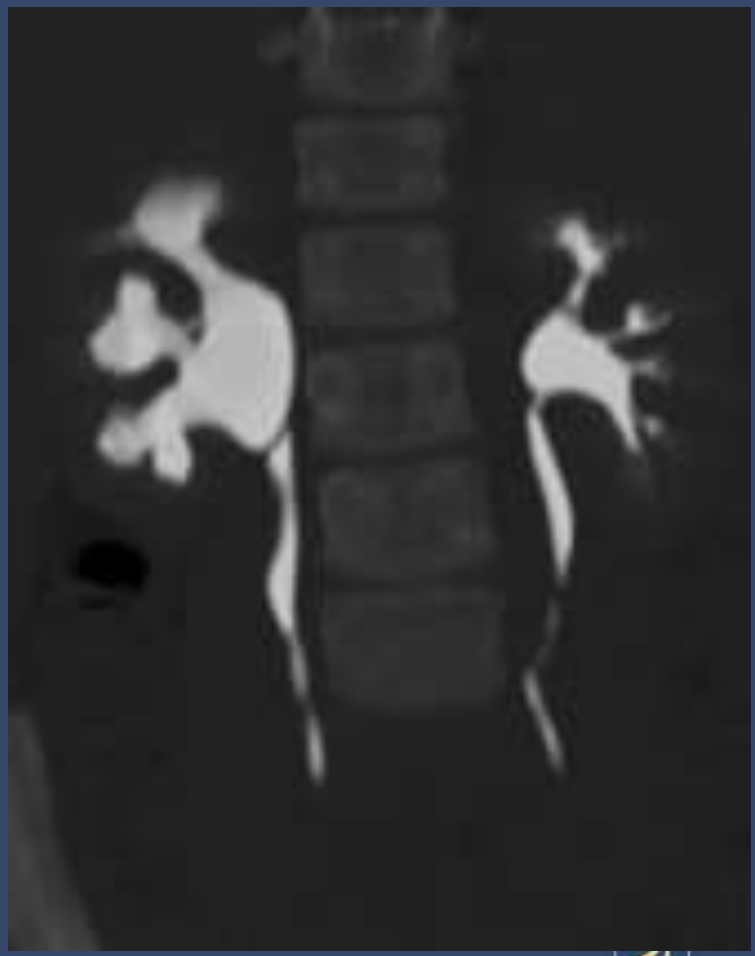
- Imaging Pearls
  - Pelvis + calyceal dilatation only
  - Pelvis ends abruptly “bullet shaped”
- Partial > complete obstruction
- 10% bilateral
- 25% with contra lateral anomaly
- Incidence
  - 1:2000
  - 20% all cases of hydronephrosis
  - Males > females
  - Not associated with aneuploidy or genetic syndromes



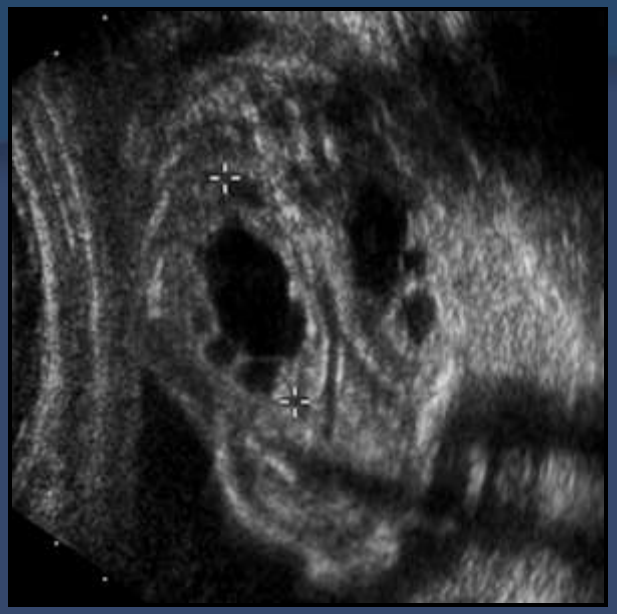


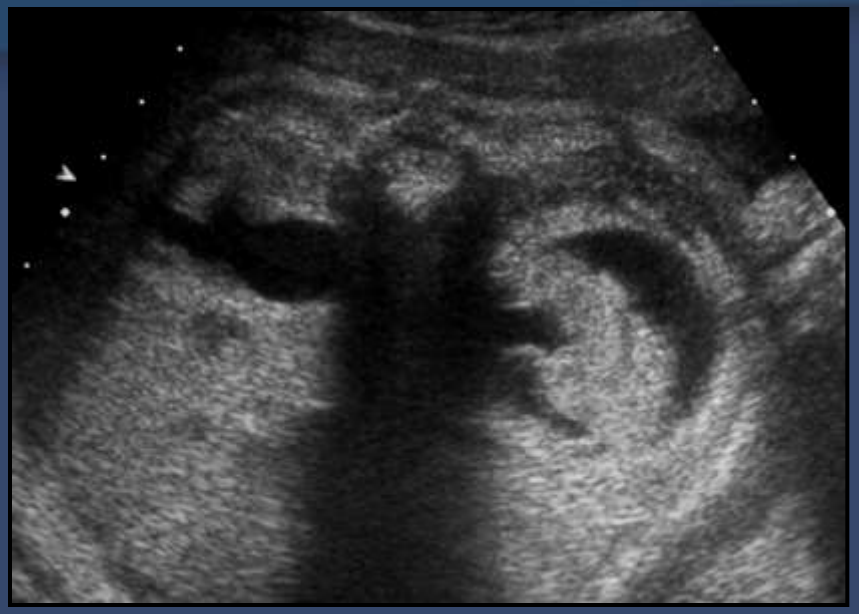
Ureteropelvic junction (UPJ) obstruction

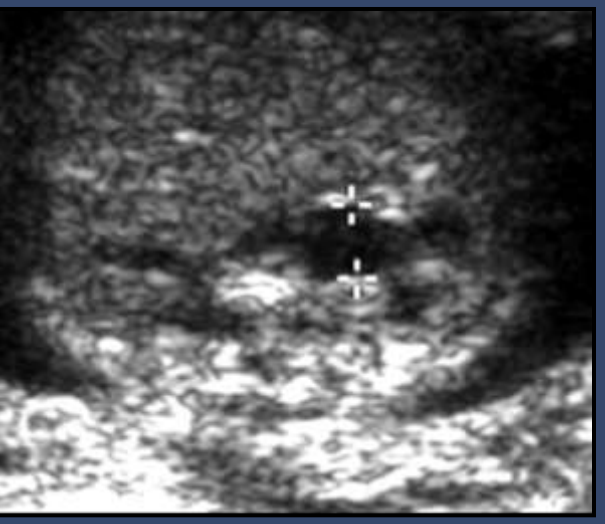
- Possible etiologies
  - Abnormal muscularis layer or neural innervation at UPJ
  - 1/3 may have accessory crossing vessel
- Complications:
  - Obstructive renal dysplasia
  - Urinoma

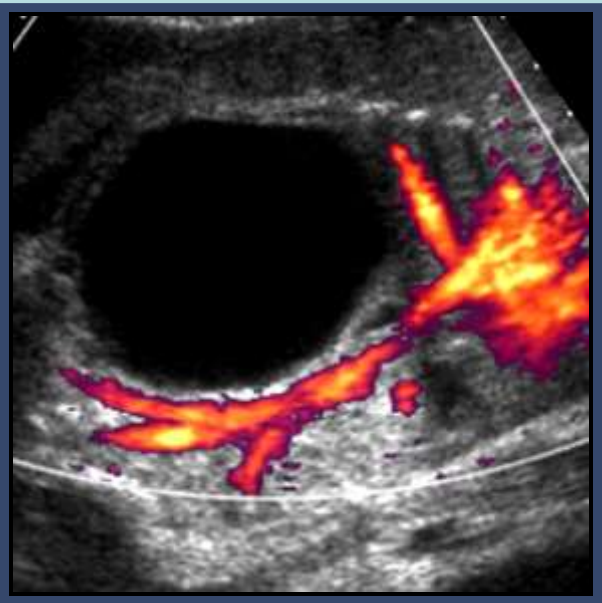


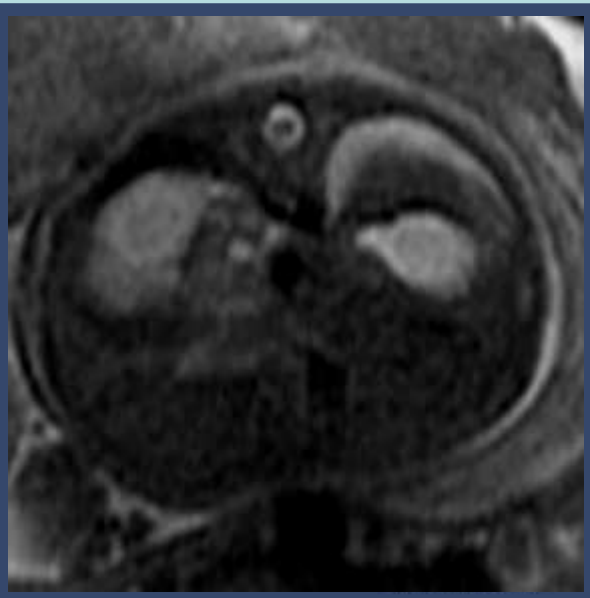
UPJ: 2 cases with urinoma



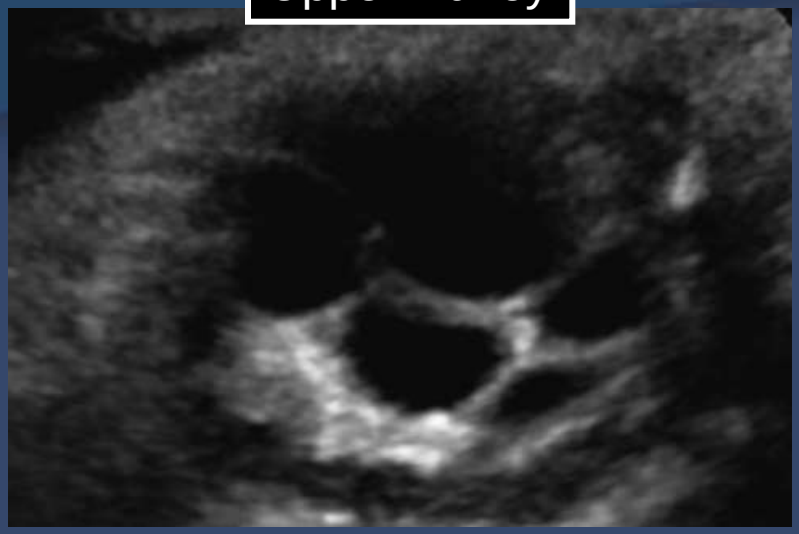


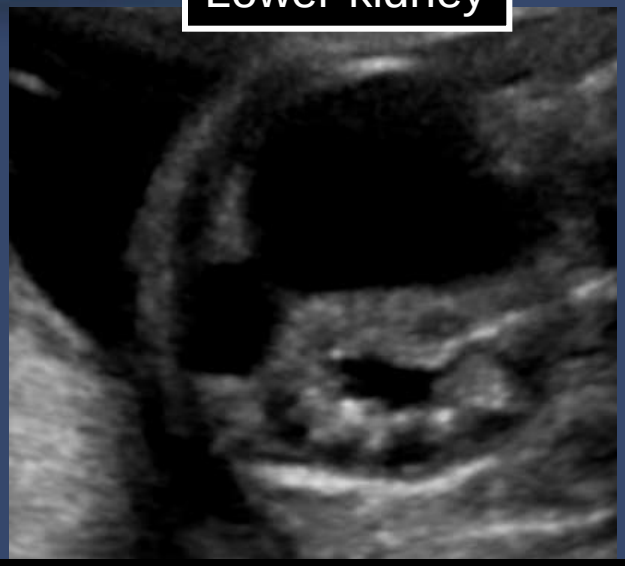




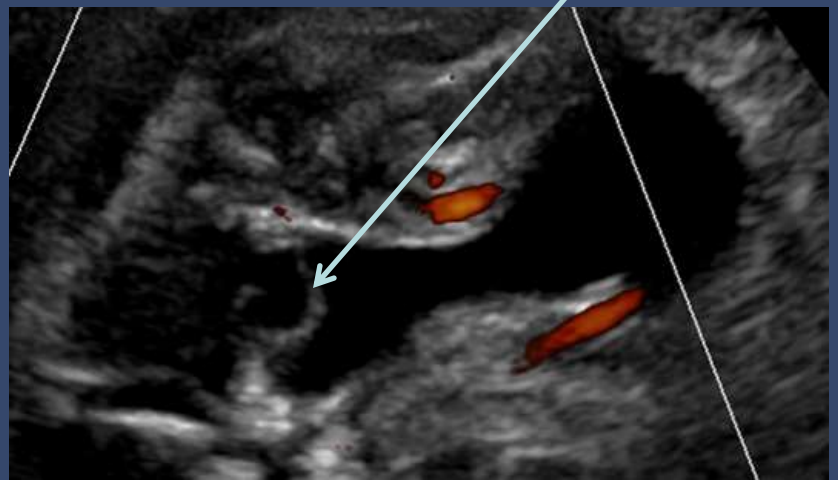


New diagnosis: Case at 21 weeks





Diagnosis: Renal duplication with ureterocele



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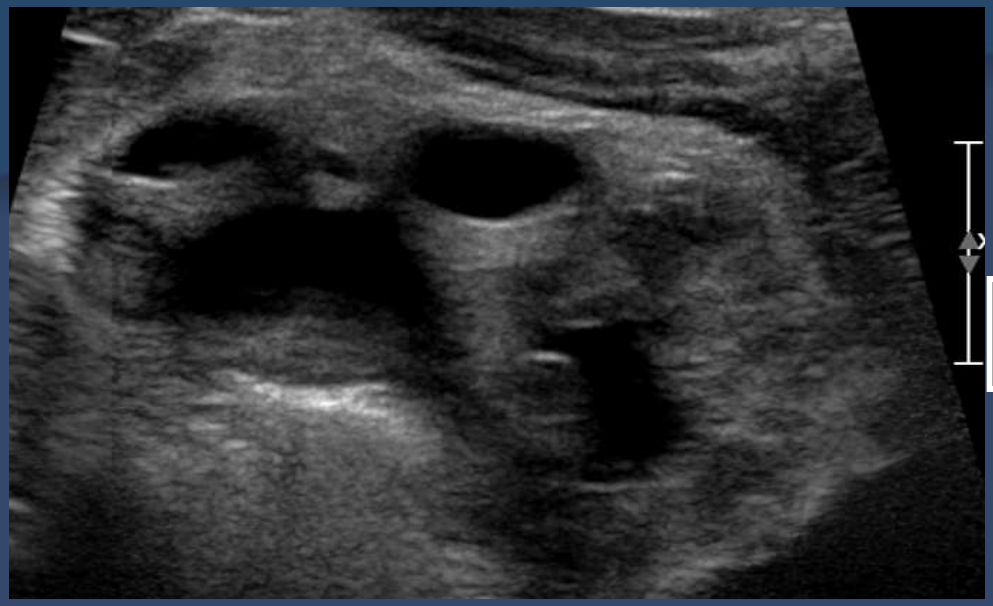
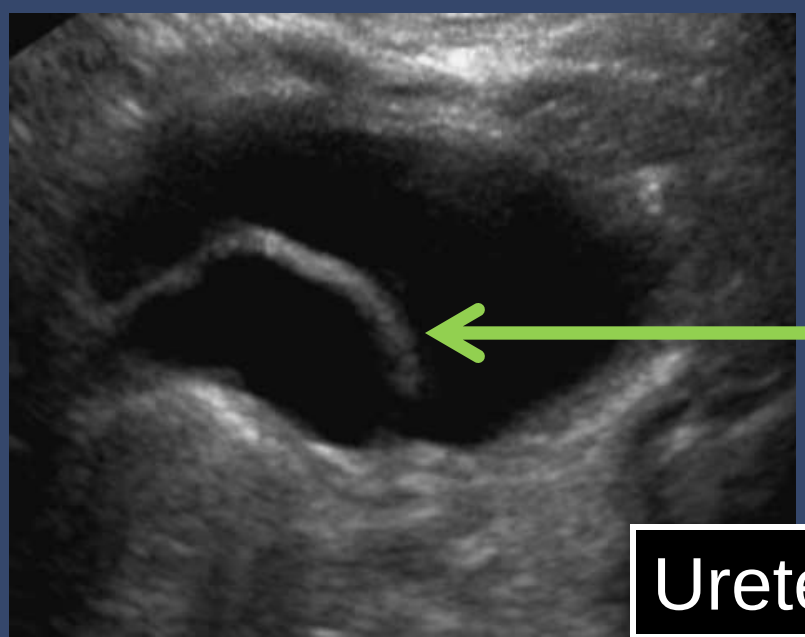
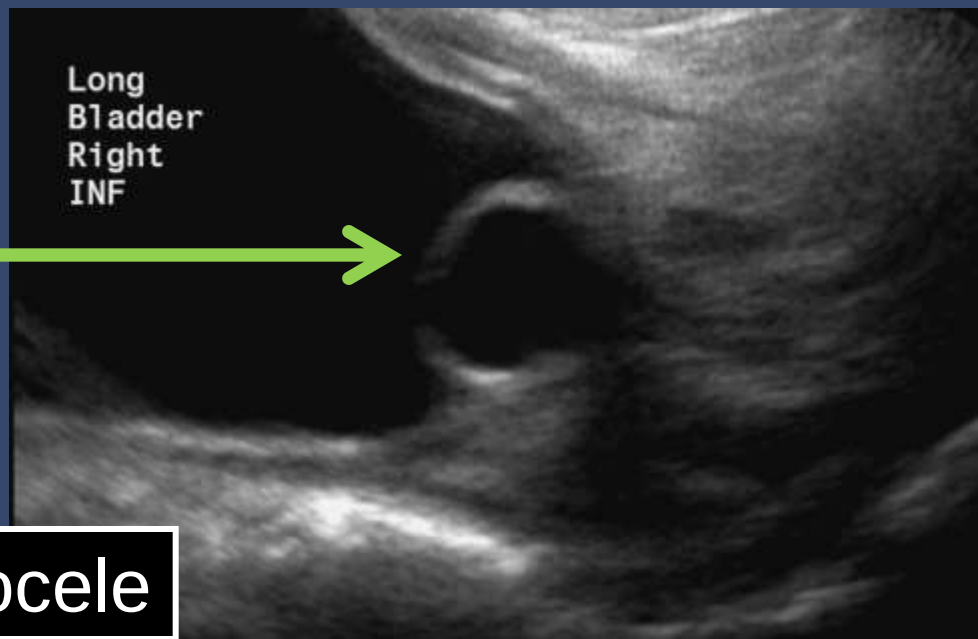
Postnatal Ultrasound



Duplicated kidney

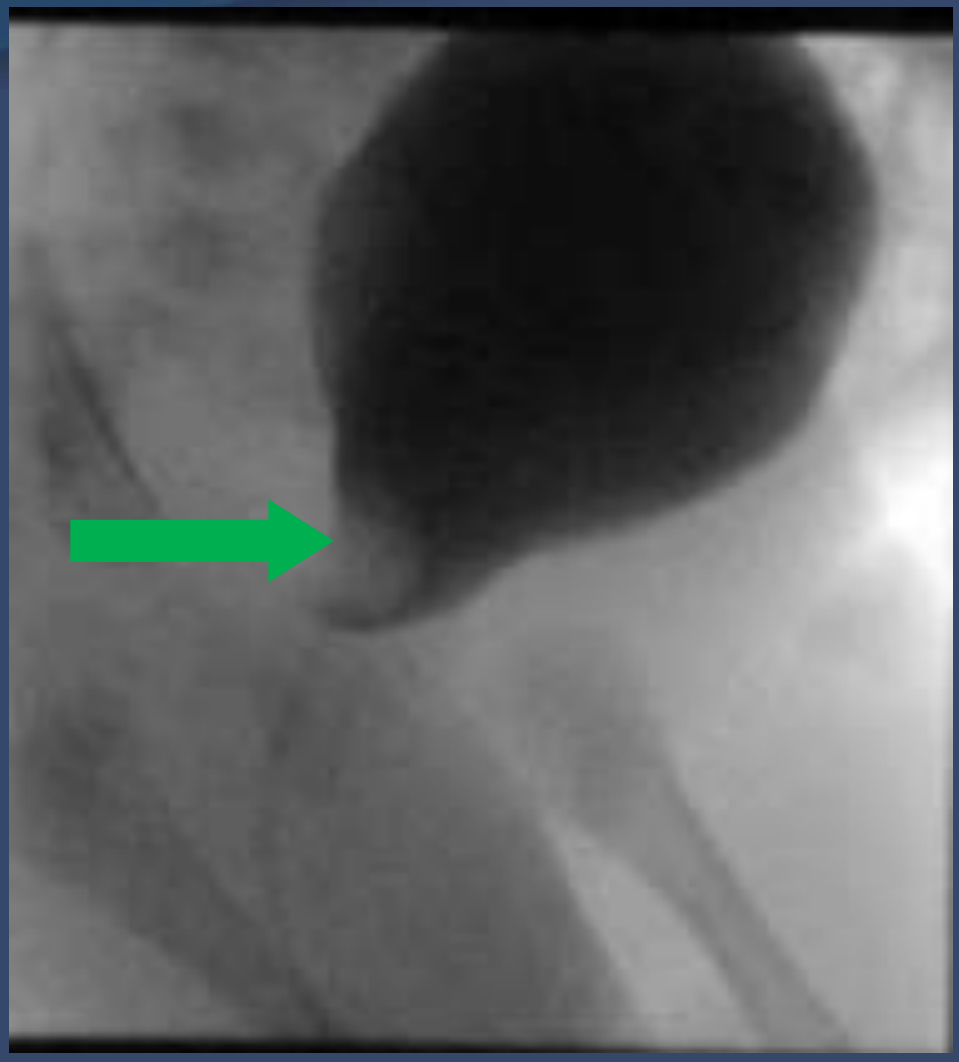


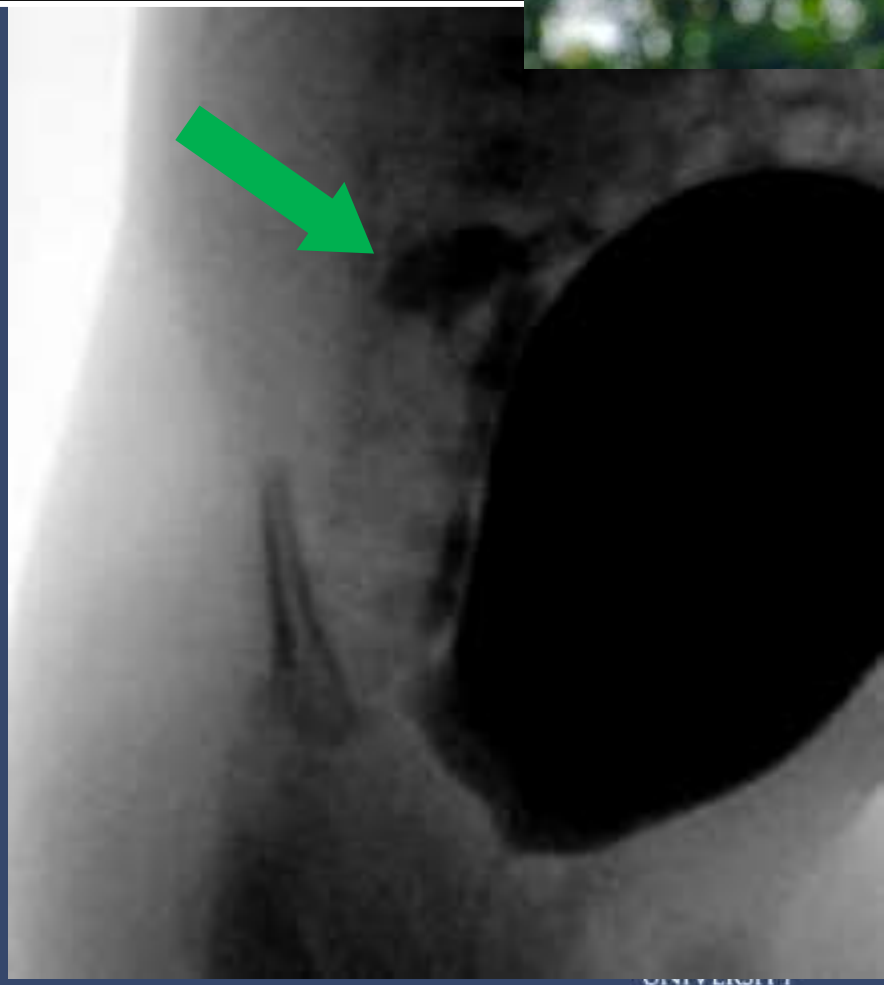


Ureterocele

VCUG– Ureterocele and Reflux

Drooping lily sign

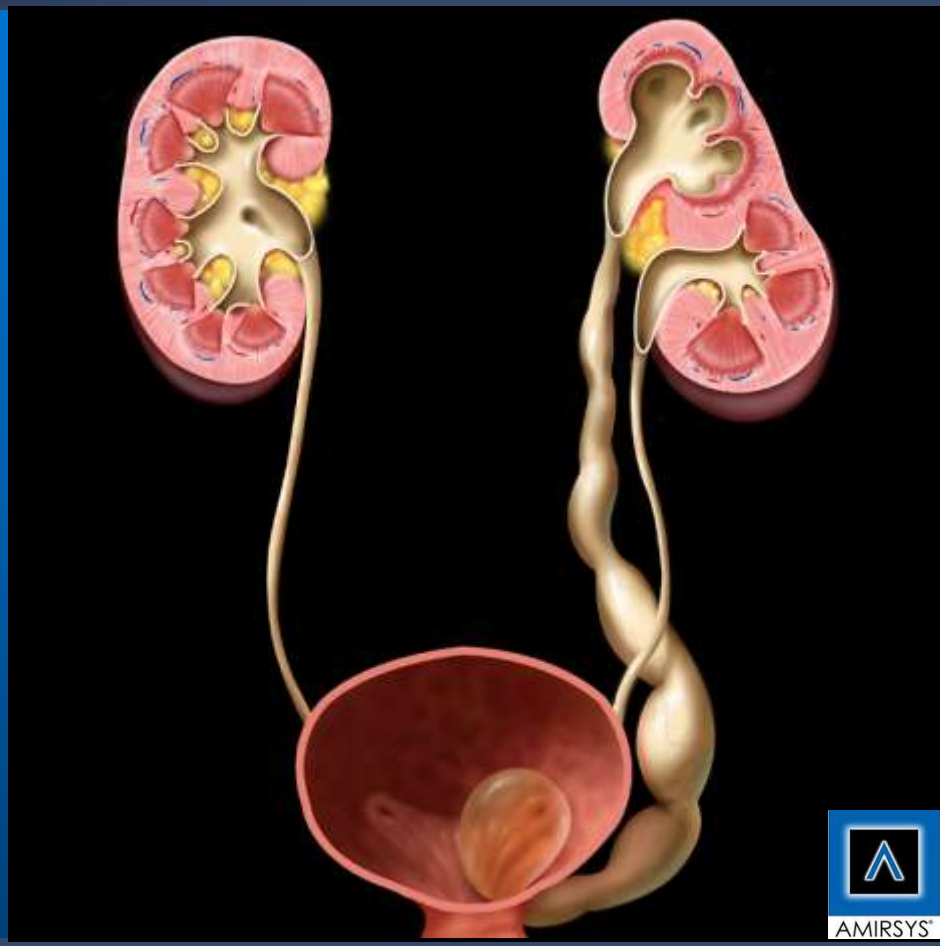




Renal Duplication

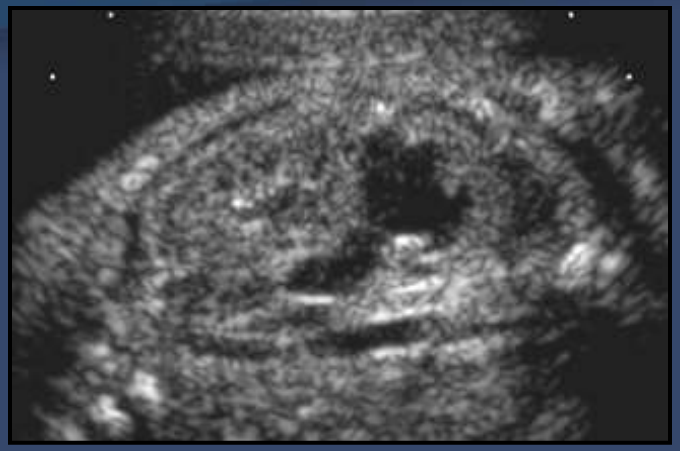
Weigert Meyer Rule

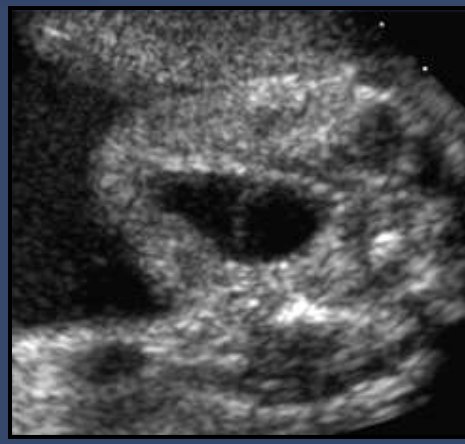
- Upper pole with ectopic ureter
  - Inserts medial and inferior to trigone
- Lower pole with reflux

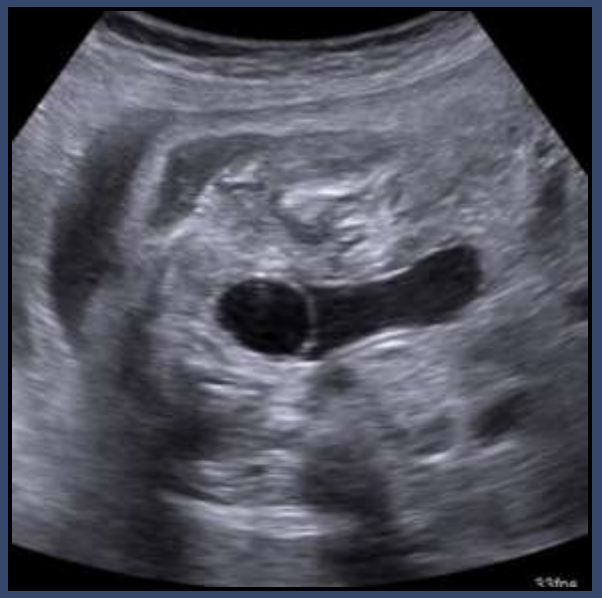


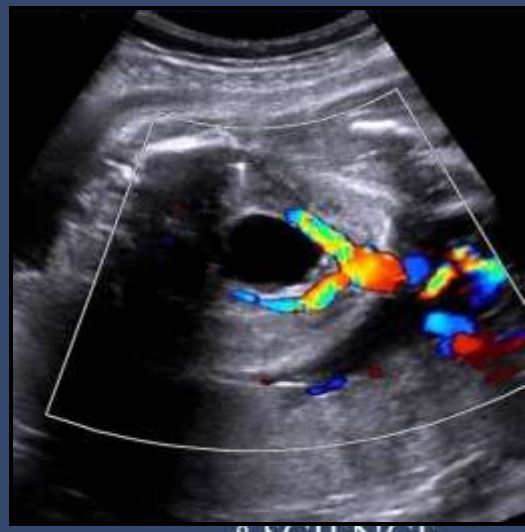
Duplicated collecting system: imaging pearls

- Be suspicious when hydronephrosis is limited to upper pole
  - good longitudinal views
- Ureterocele in bladder is the key finding
  - May be large and can mimic bladder
  - May obstruct urethra or other ureter
- Bilateral hydronephrosis in 10-20%
- Twice as common in female fetuses

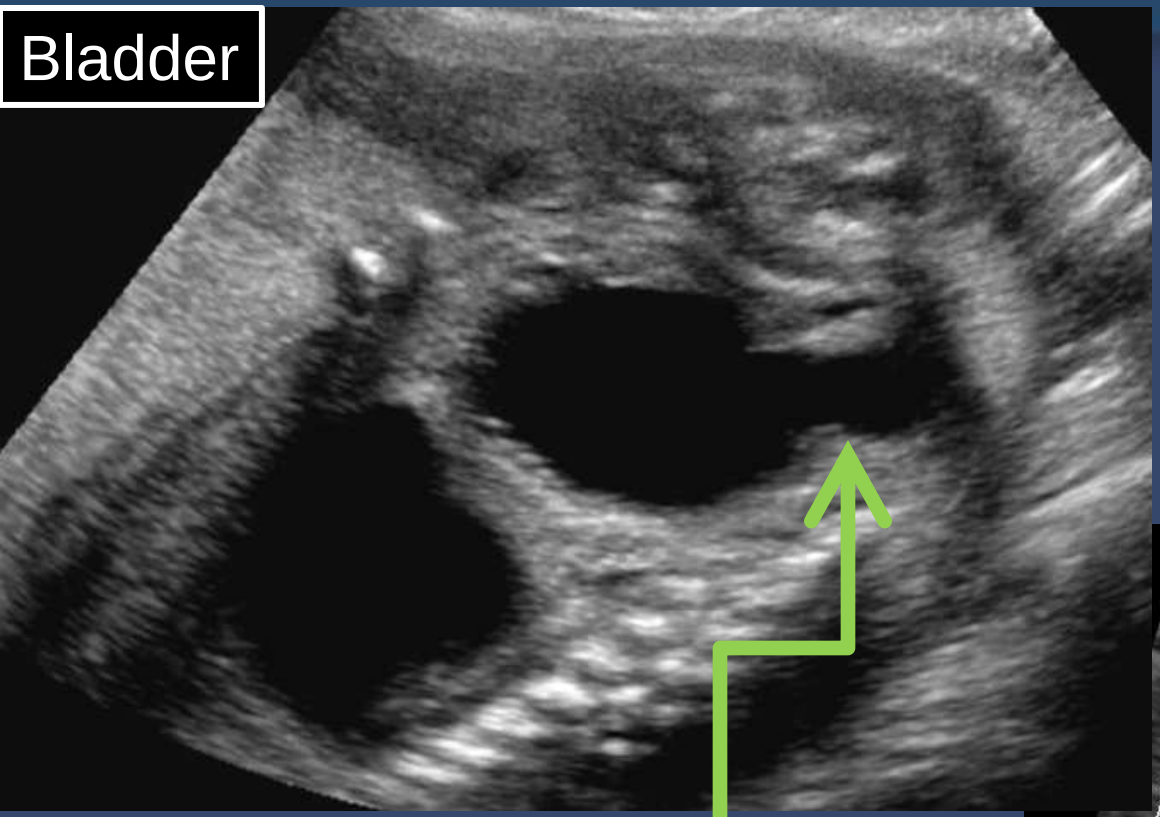






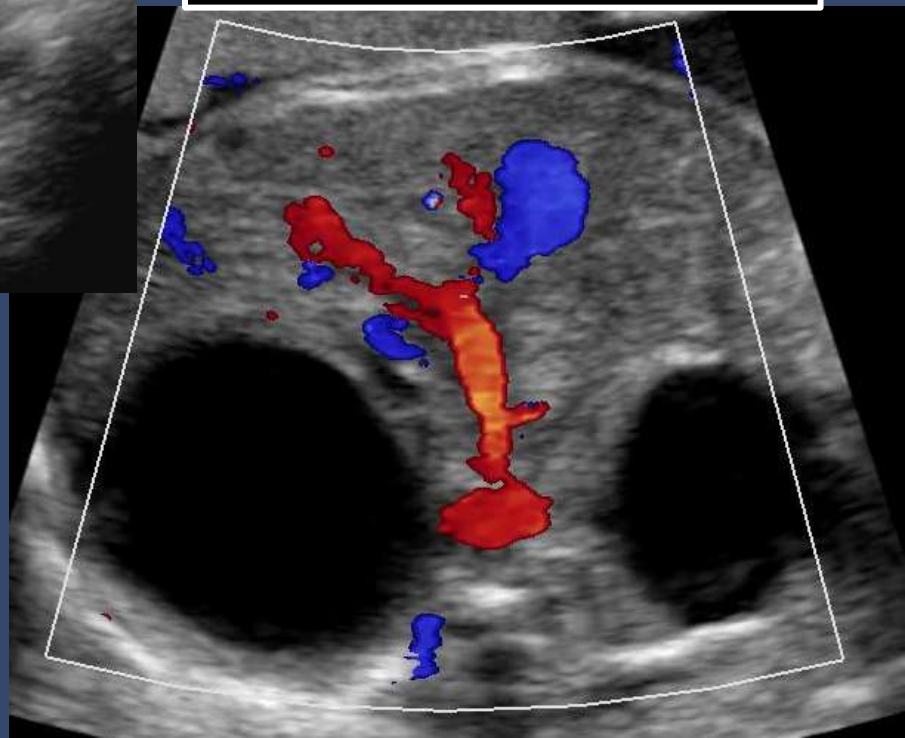


New Diagnosis: Case 20 weeks fetus

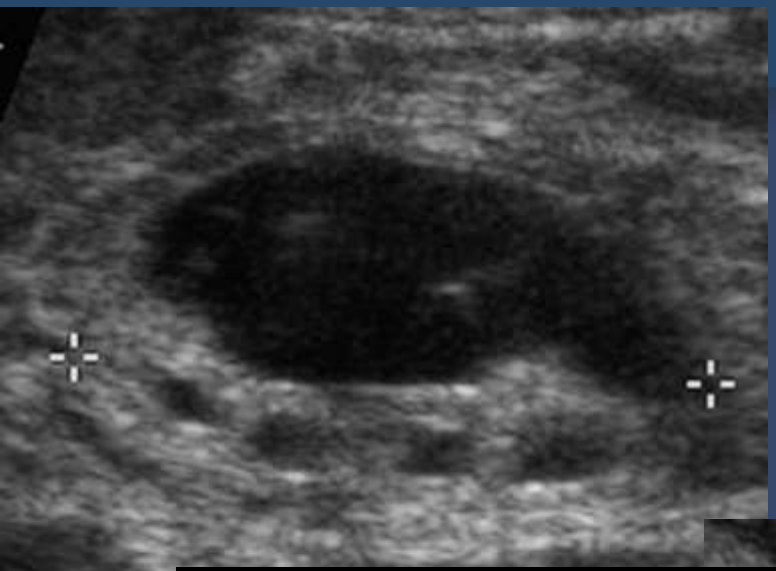


Keyhole appearance

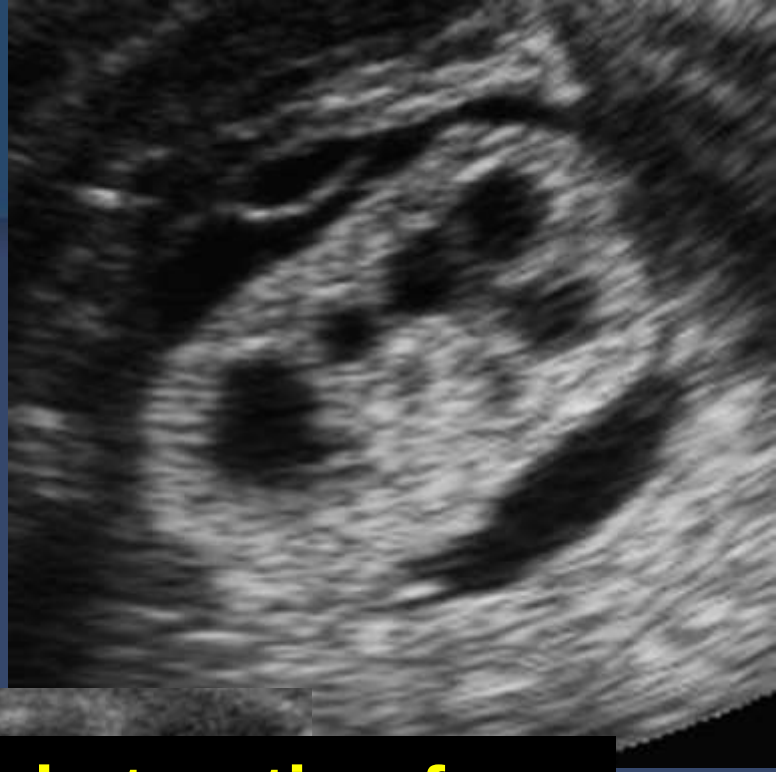
Axial view of kidneys



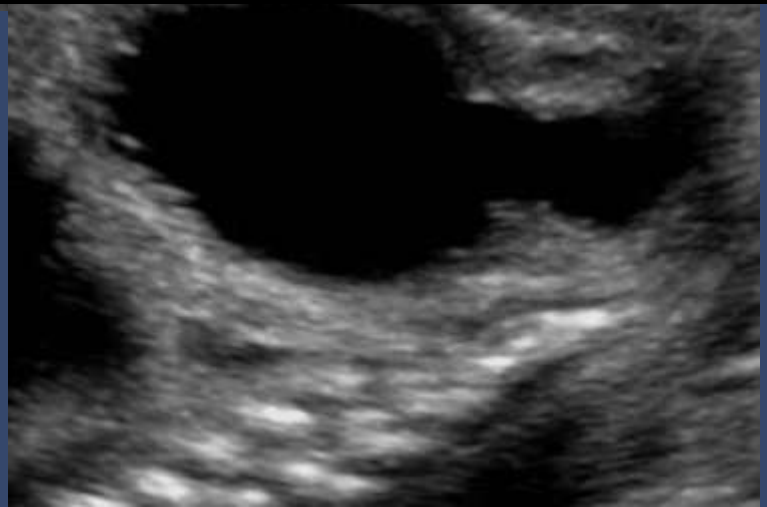
Case f/u 3 wks



Kidneys



Diagnosis: Bladder outlet obstruction from Posterior Urethral valves



Bladder

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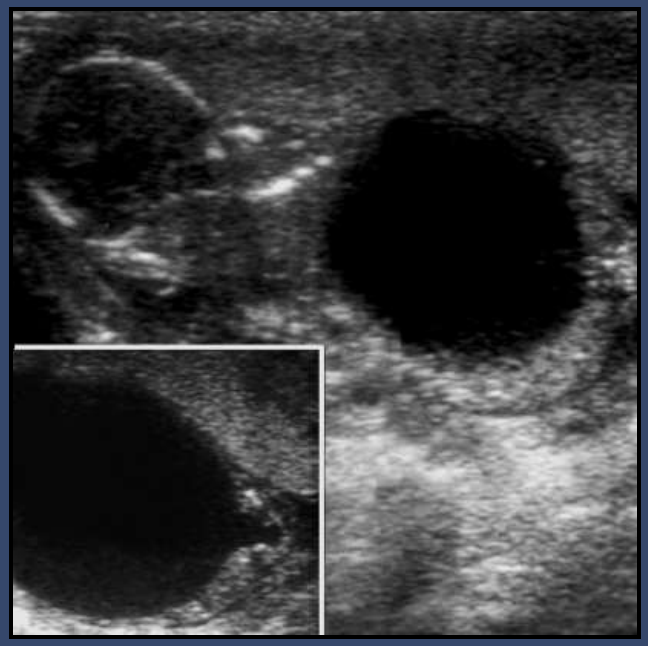
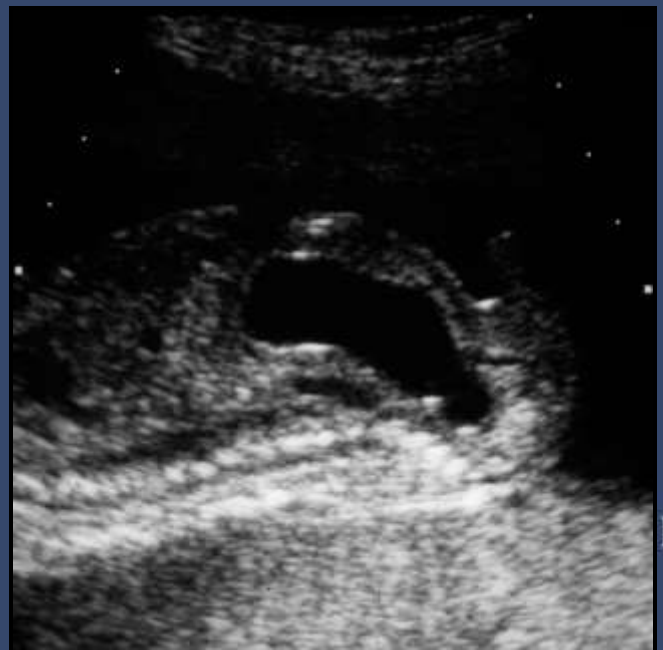
Lower urinary tract obstruction (LUTO)

Posterior urethral valves – Imaging pearls

- Variable hydronephrosis (may see echogenic kidneys) +/- ureter dilatation
- Dilated thick walled bladder
- Dilated posterior urethra: funnel, keyhole
- Decreased or absent Amniotic fluid





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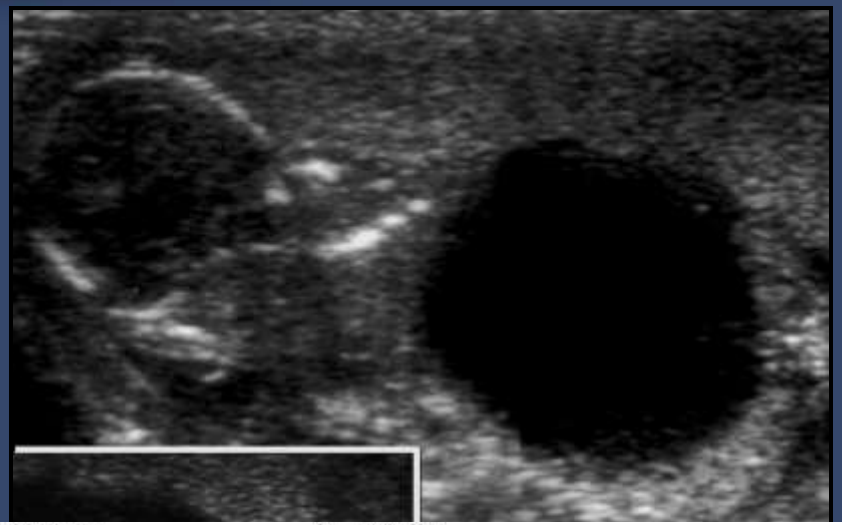
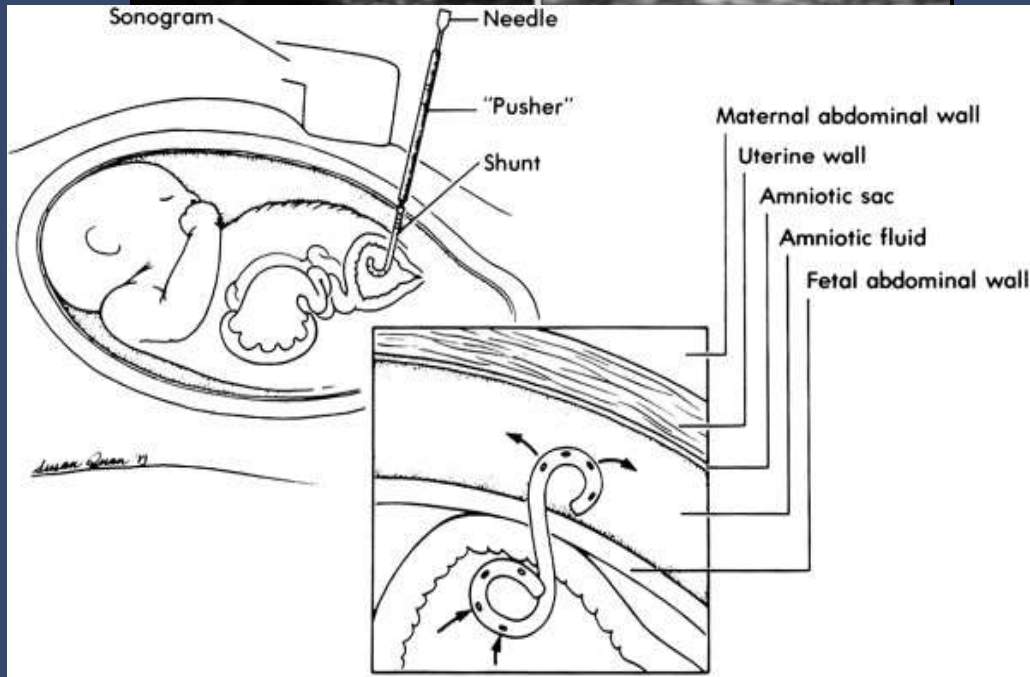
LUTO – urethral obstruction: Antenatal counseling and management

Two main concerns:

- Renal injury
  - 25-30% survivors with ESRD
  - Early dialysis then transplant
- An/Oligohydramnios
  - Single most important prognostic feature
  - Pulmonary hypoplasia
  - 45-55% mortality
- Bypass the urethra?





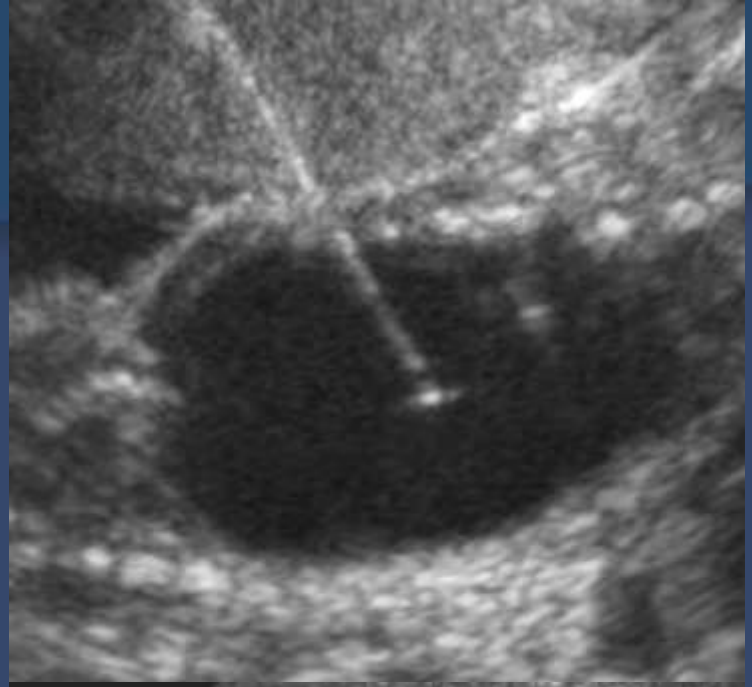
Therapeutic option

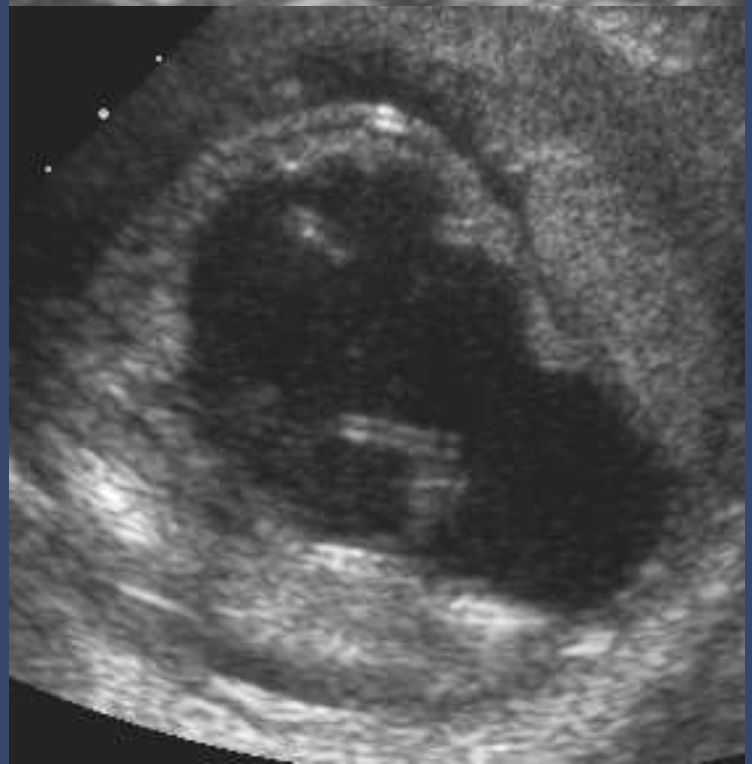
- Vesicocentesis

  Assess electrolytes looking for a favorable profile

  – Na <100, Cl <90, Osm <210 mEq/L, Ca, PO4, & β2 microglobulin all <2mmol/L.

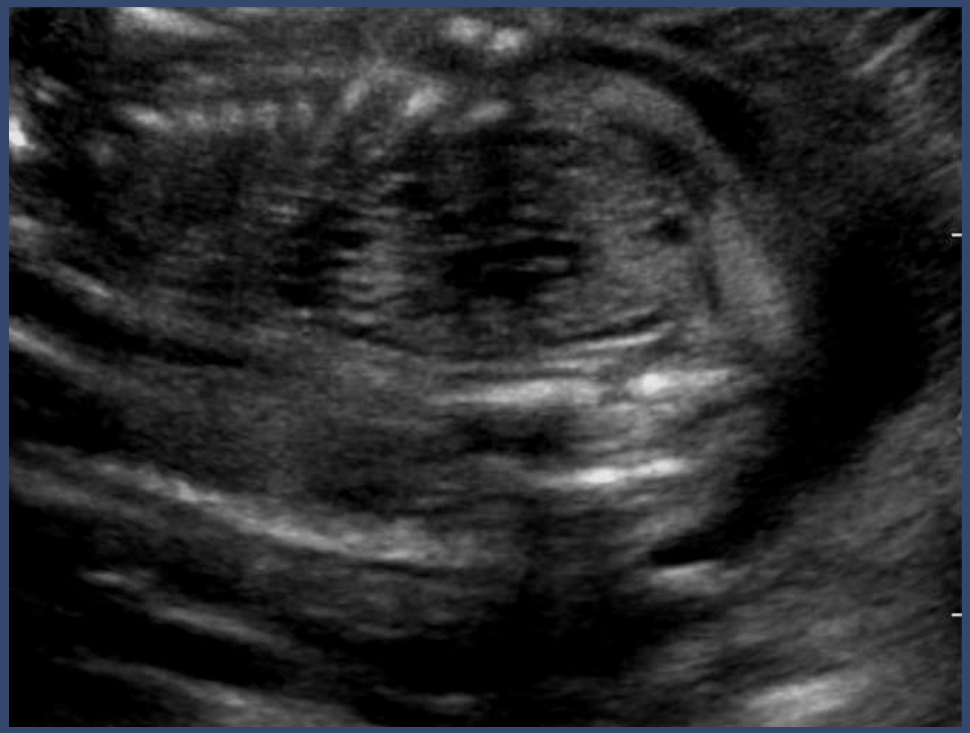
- Vesicoamniotic shunt

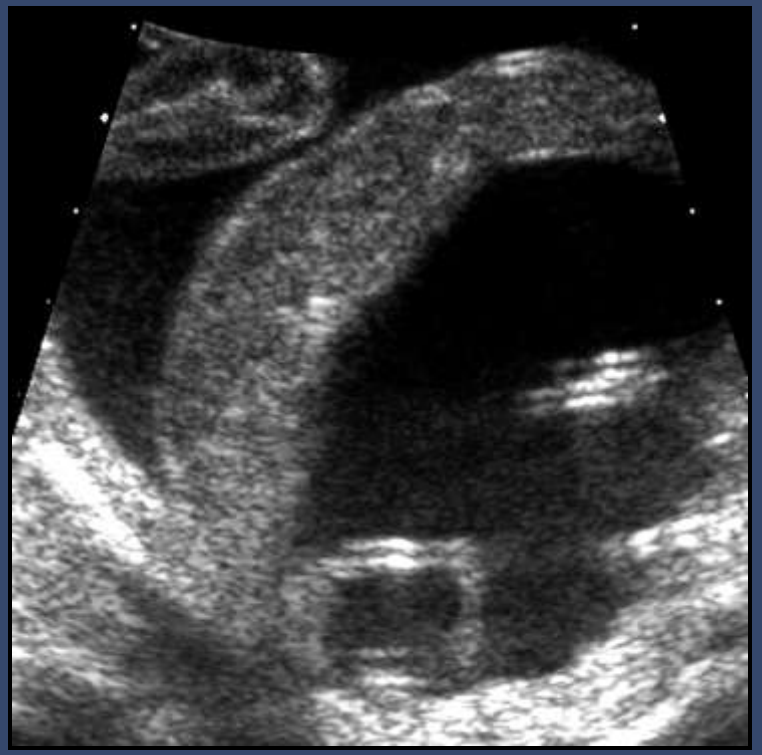




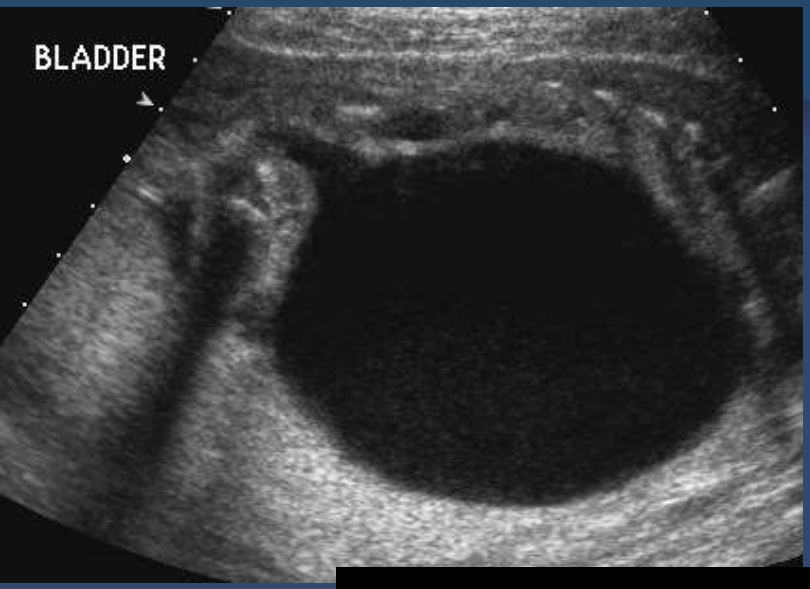
Bladder Shunting

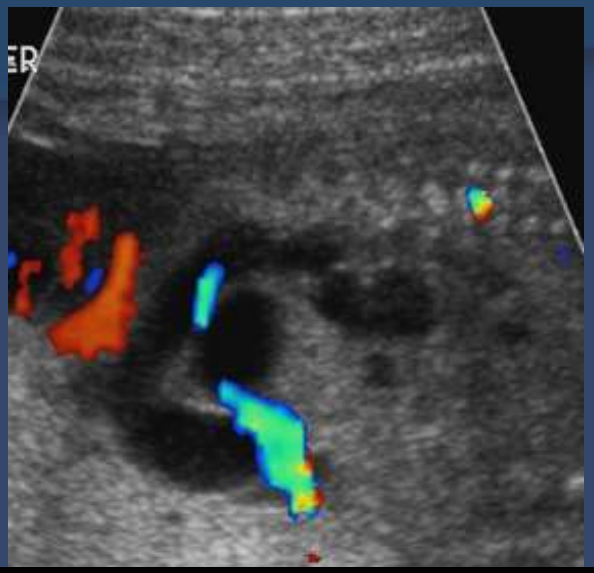
- Complications in 1/3
  shunt dysfunction, PPROM, infection/chorioamnionitis,
- Shunting improves survival – improves pulmonary function
- Residual renal dysfunction – still poor in most cases

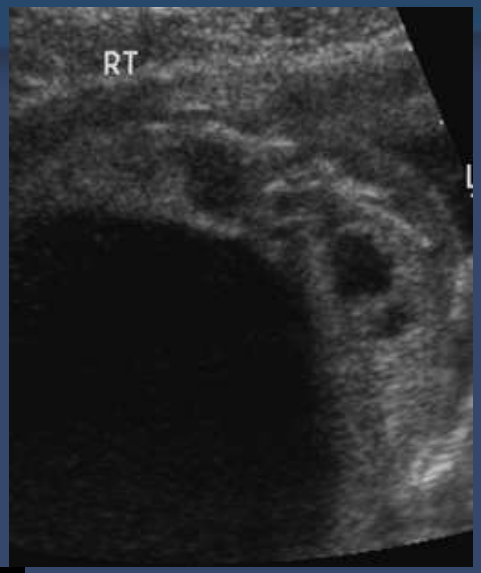




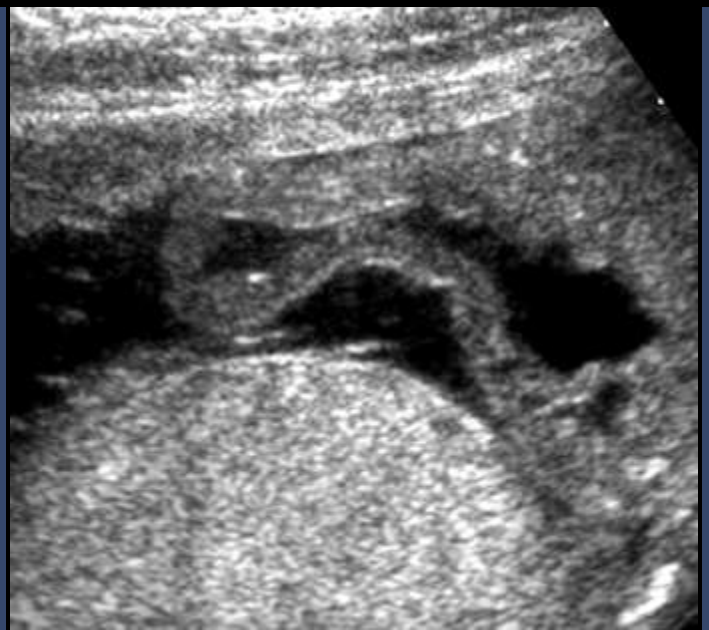
Case: Not all dilated bladders are PUV:







Diagnosis: Prune Belly Syndrome



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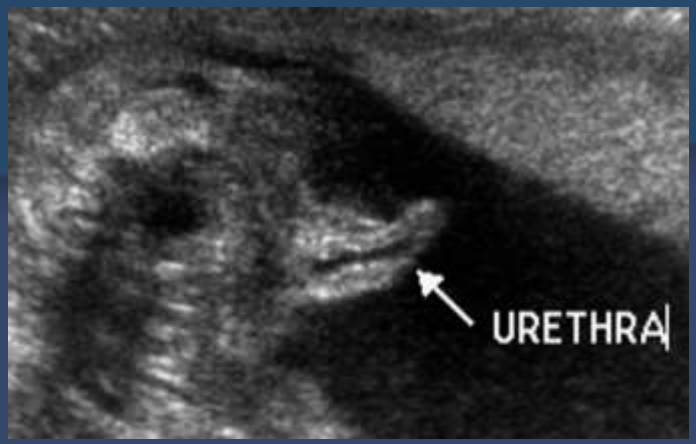
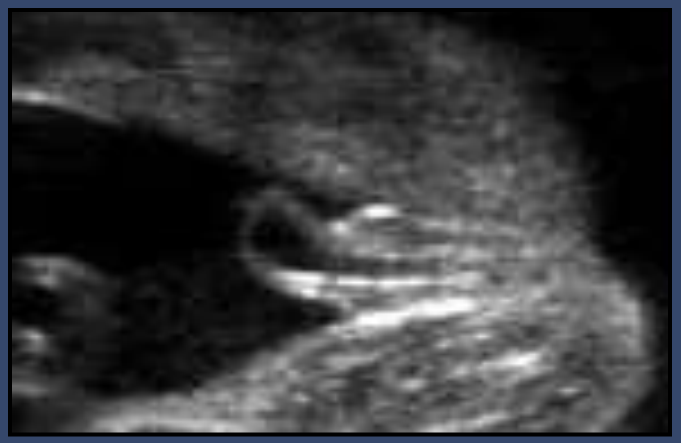
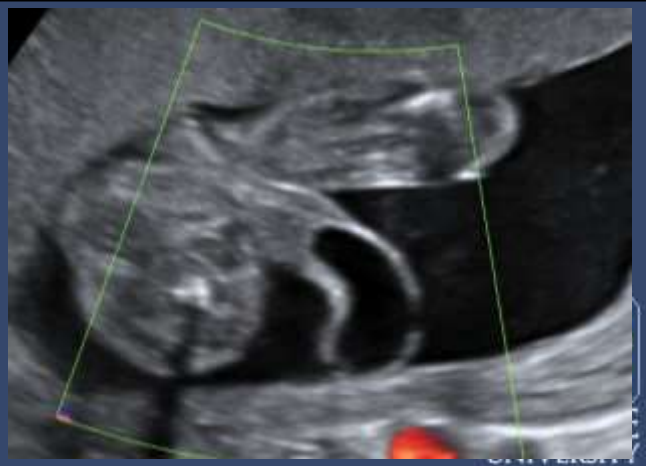
Prune Belly Syndrome

- 3 components
  - Dramatic collecting system dilatation
  - Deficiency of abdominal musculature
  - Cryptorchidism
- Findings overlap w/ PUV
  - Less likely keyhole sign
  - Ureter more likely dilated
  - Entire urethra may be dilated
  - Oligohydramnios often not as severe
  - Small chest but often not as severe as with LUTO

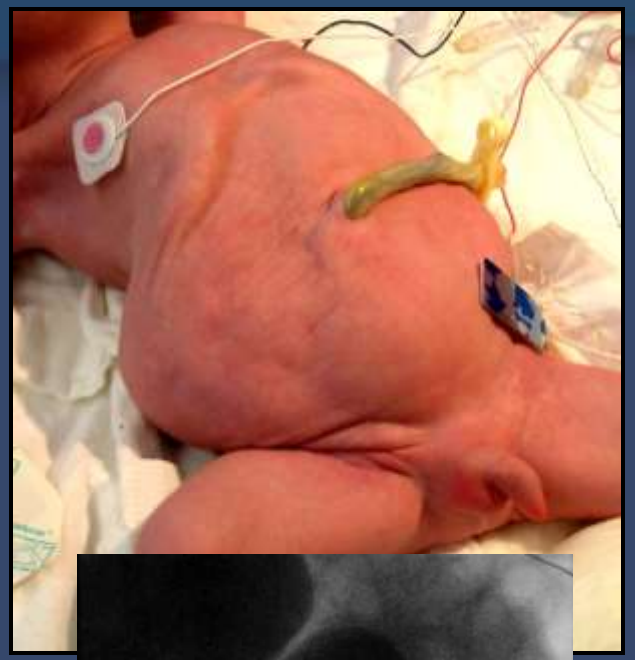


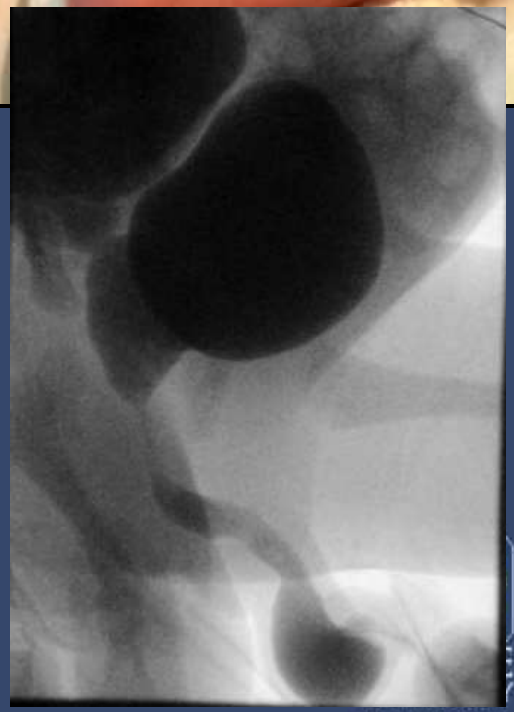




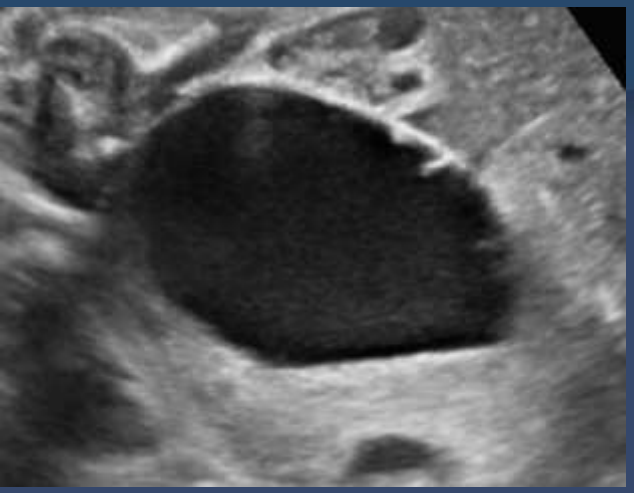
Prune Belly: Counseling and management

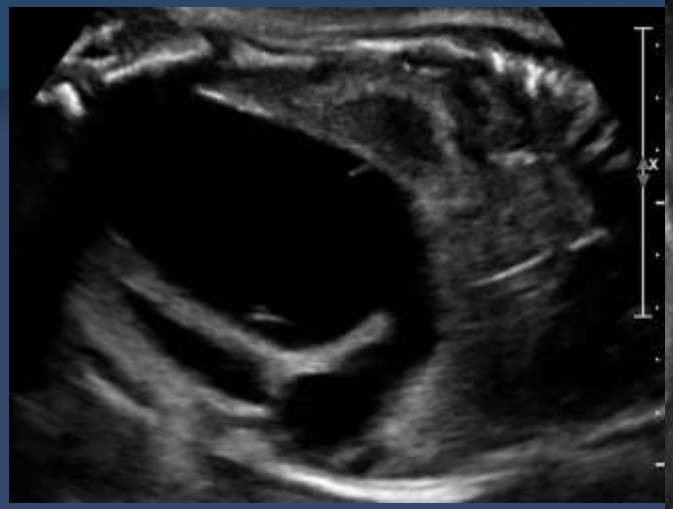
- More rare than PUV
  - ~4/100,000 live births
- Males >> Females
- Inheritance/Etiology?
  - Associated with aneuploidy and other genetic defects
- Neonate issues/anomalies
  - Pulmonary hypoplasia
  - Renal dysfunction (50% ESRD)
  - GI malrotation and anorectal anomalies

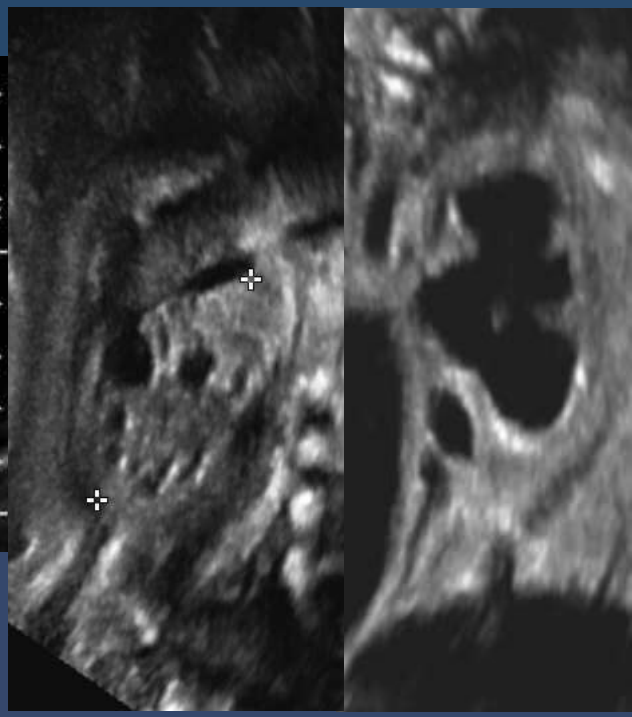


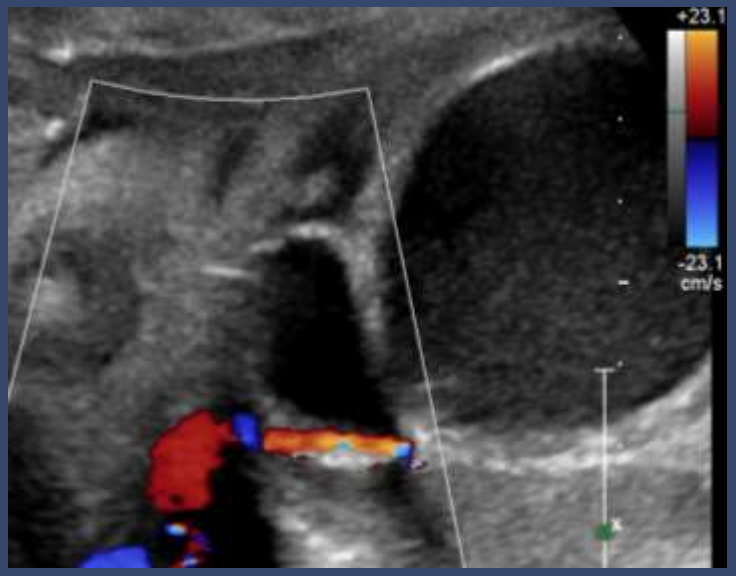


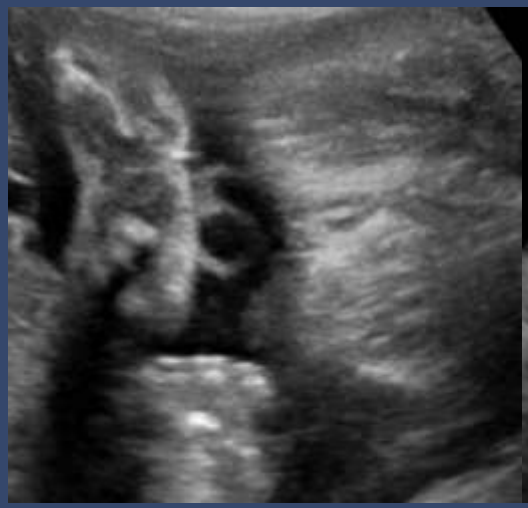
Case: Not all distended pelvic structures are bladder

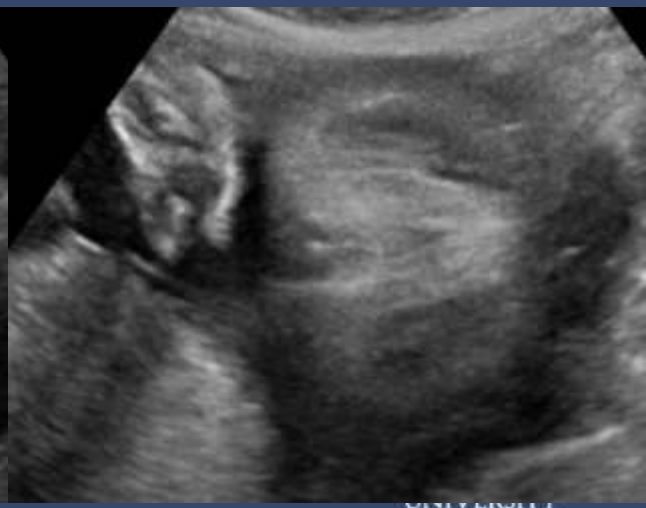




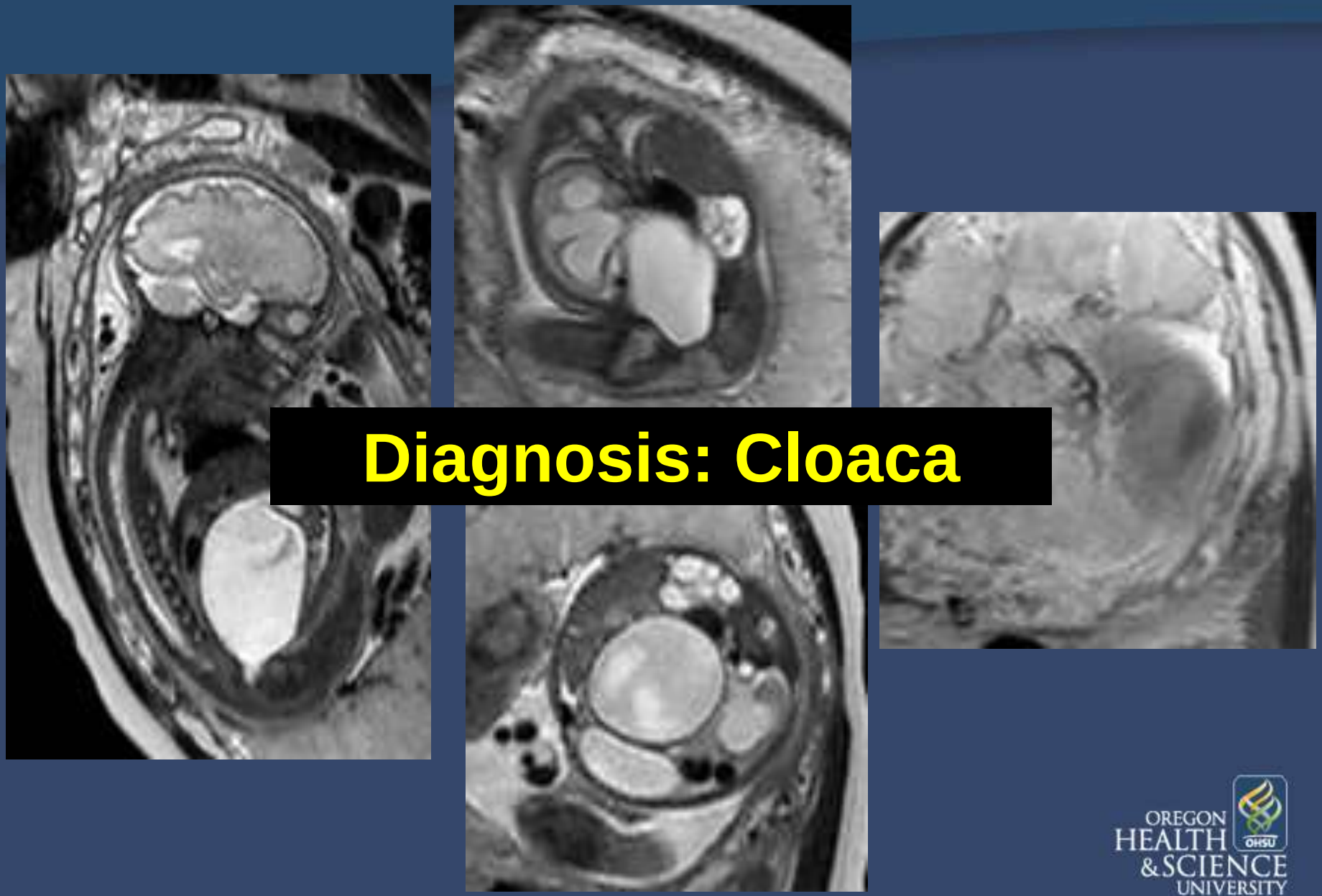






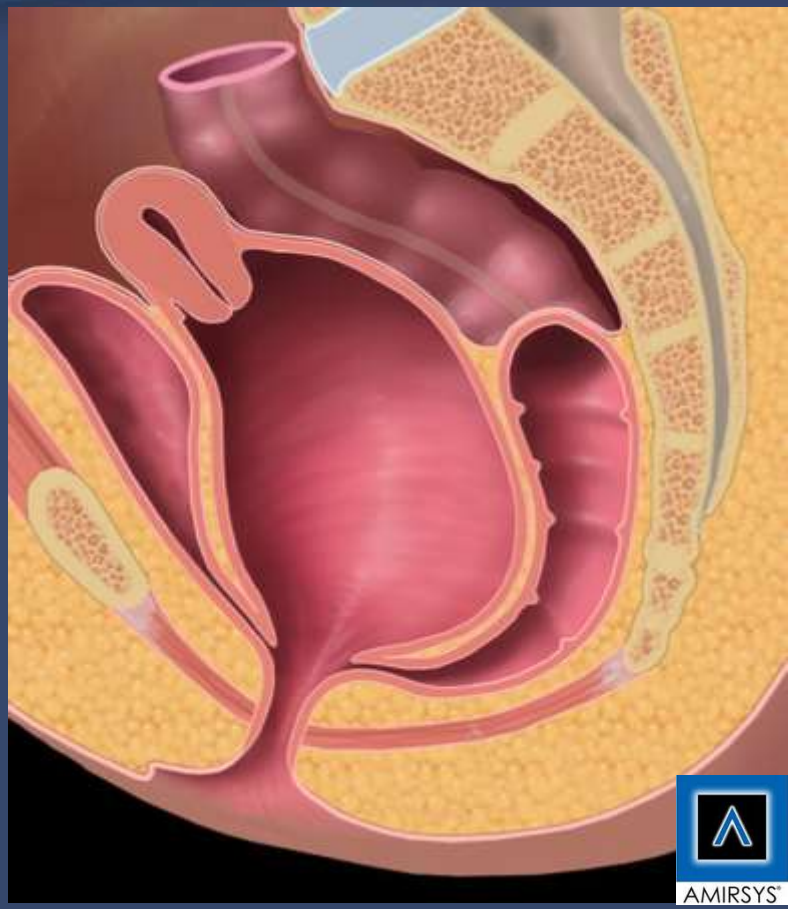


Case: Fetal MR

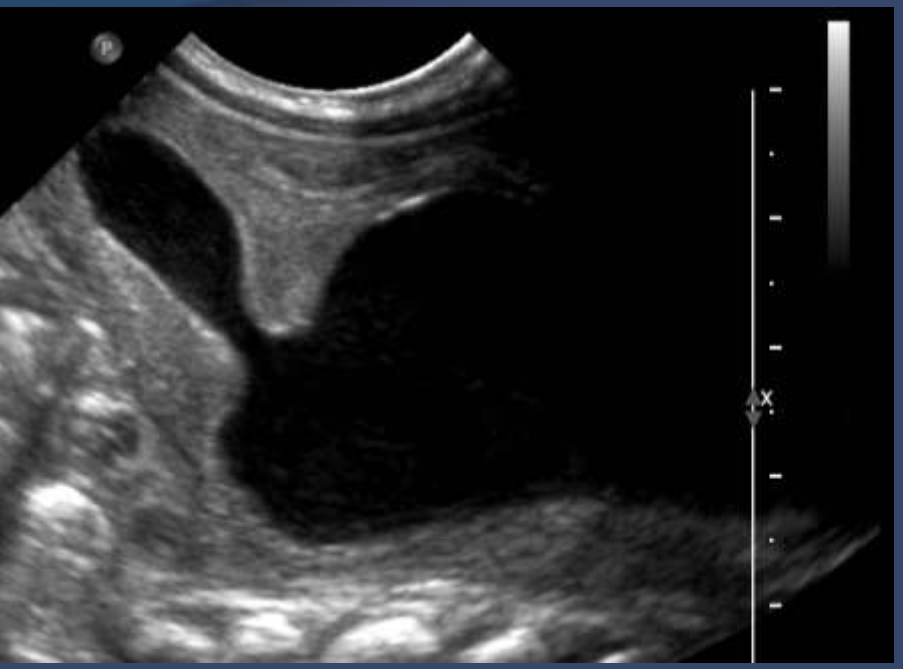


Cloacal Malformation

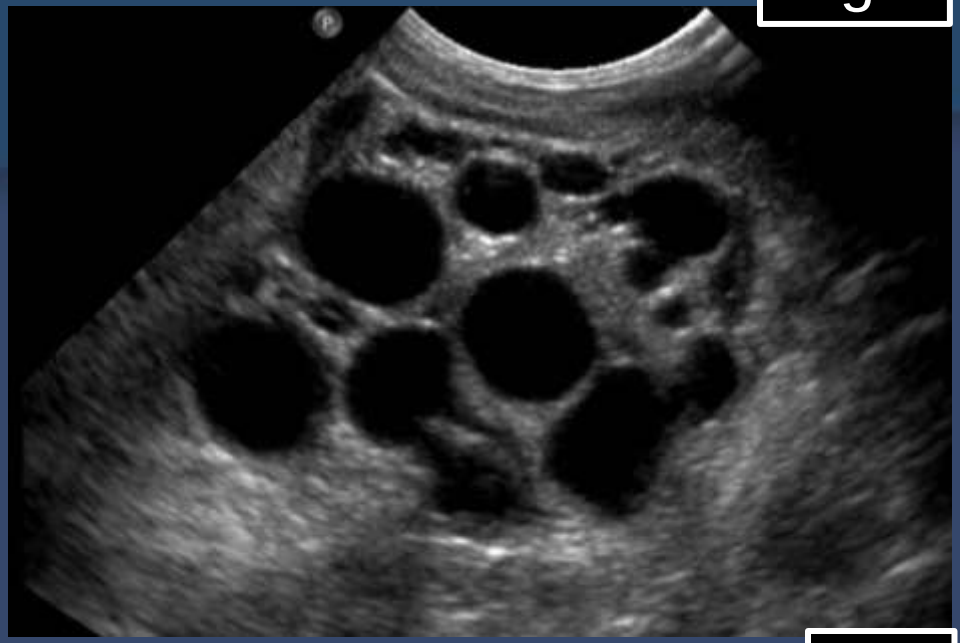
- Failure of early cloaca division
- Spectrum of abnormal anatomy related to time of arrest
- Classic: coalescence of urethra, vagina, and hindgut with single draining perineal orifice
- DDX: hydrocolpus, Urogenital sinus: bladder + vaginal connections only



Postnatal



Right



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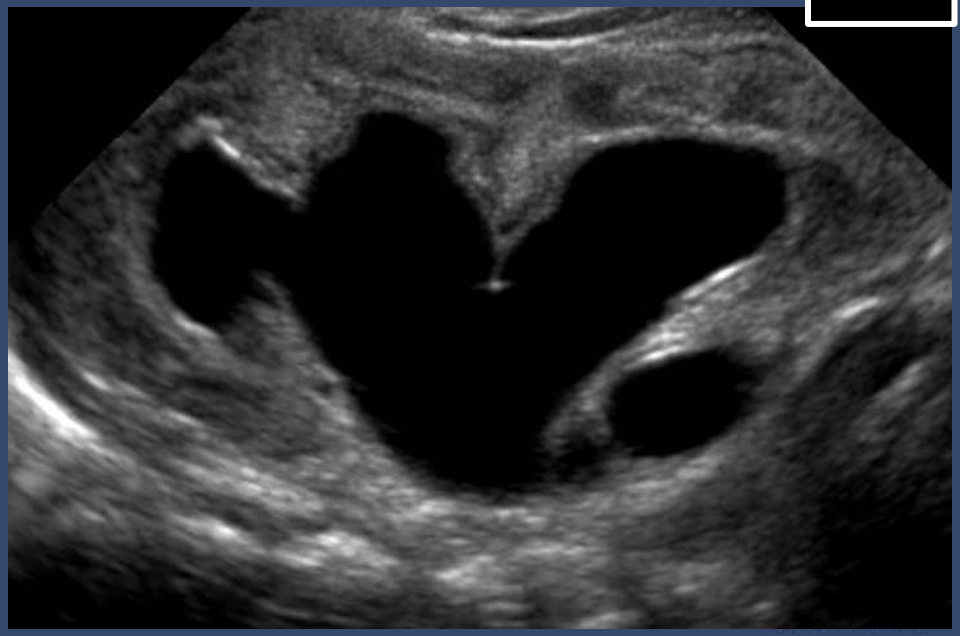
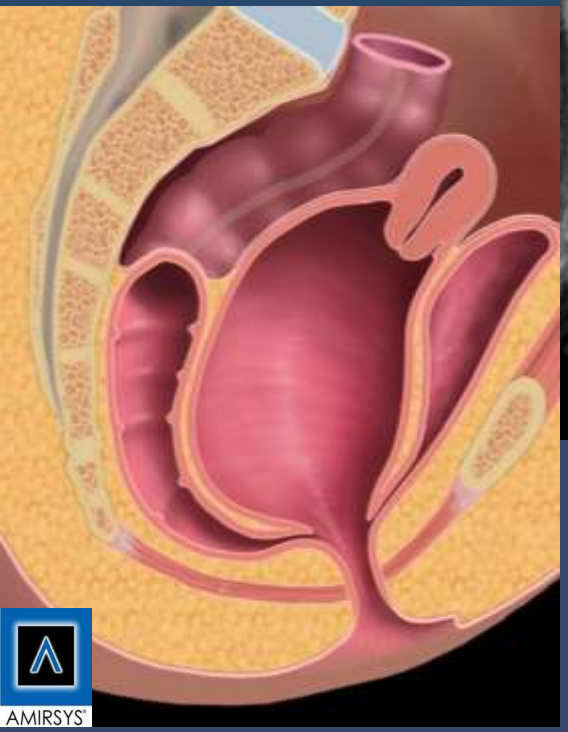
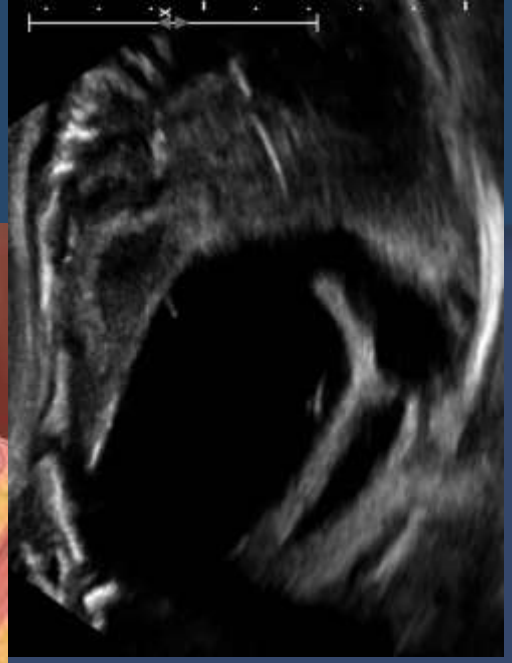
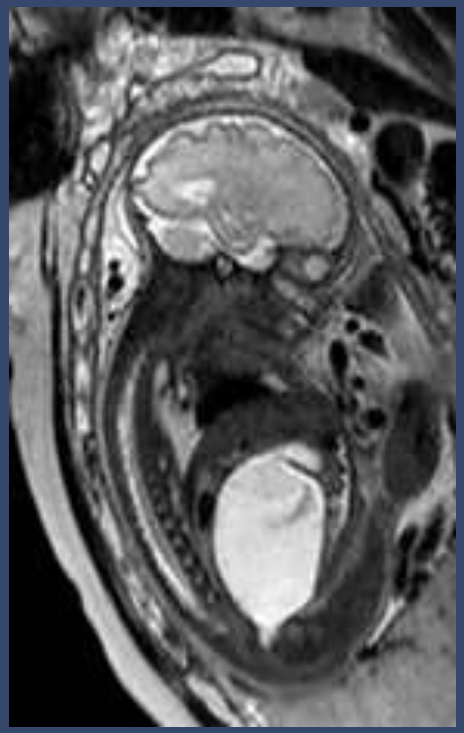
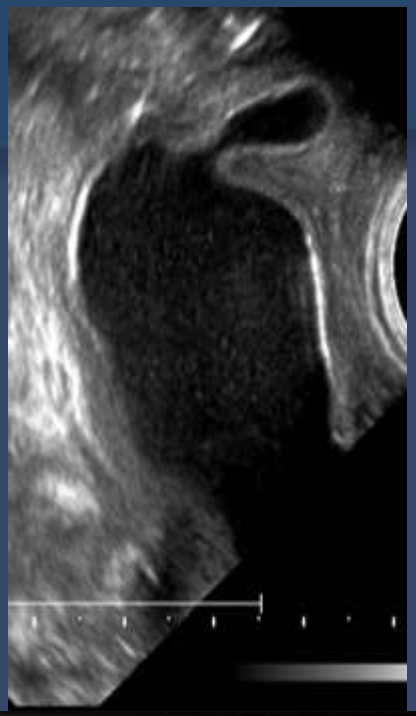
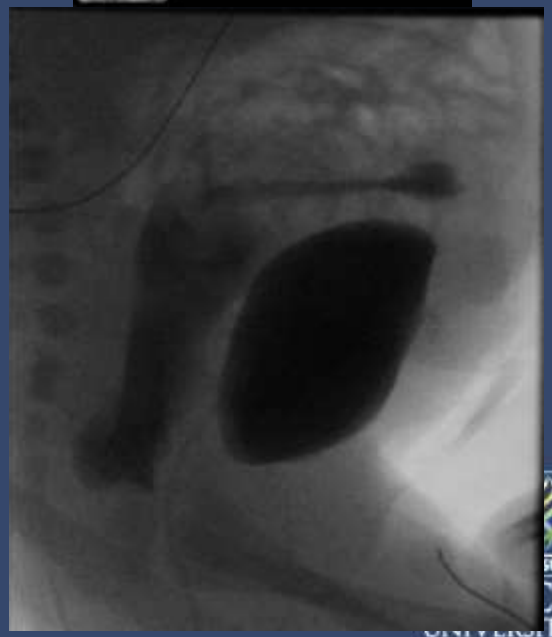


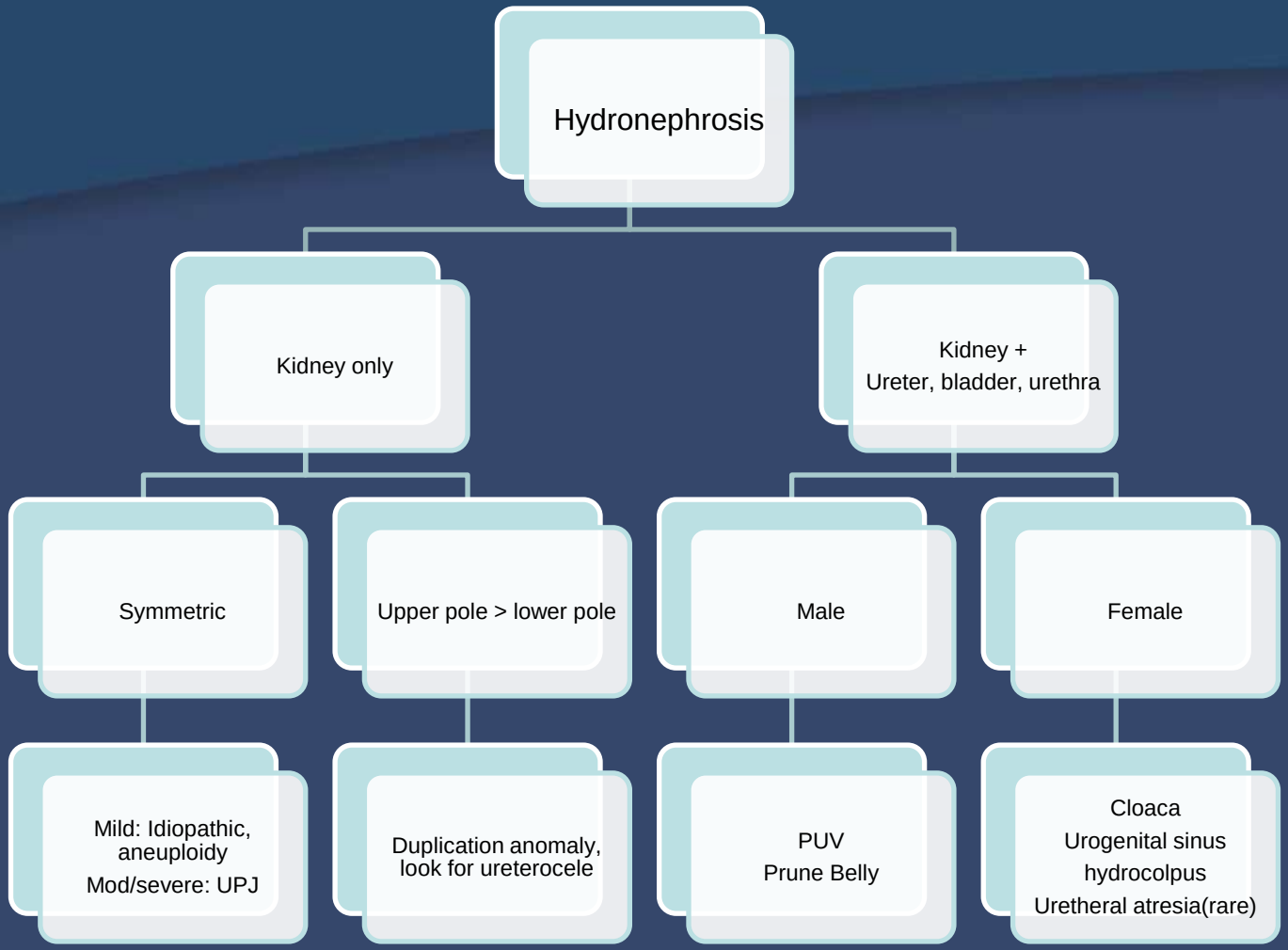

AMIRSYS











Renal Cystic Dysplasia

OBSTRUCTIVE RENAL DYSPLASIA

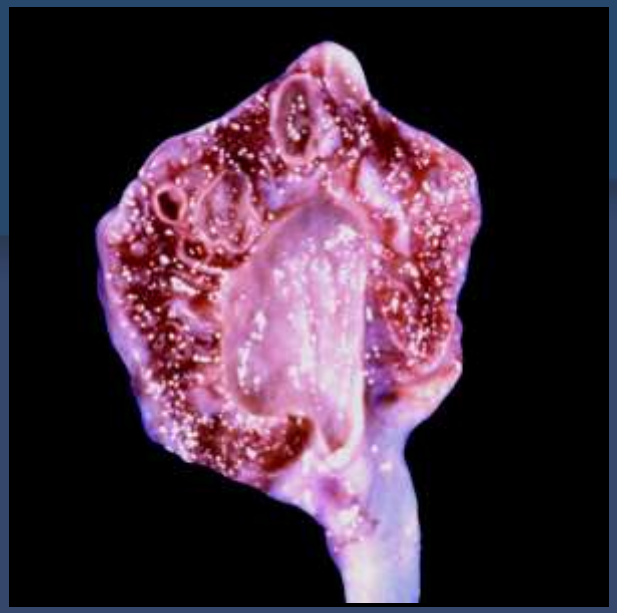
MULTICYSTIC DYSPLASTIC KIDNEY (MCDK)

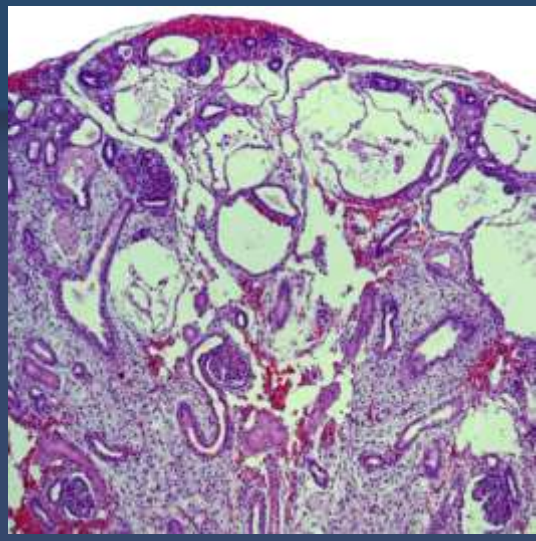
AUTOSOMAL RECESSIVE POLYCYSTIC KIDNEY DISEASE (ARPCKD)

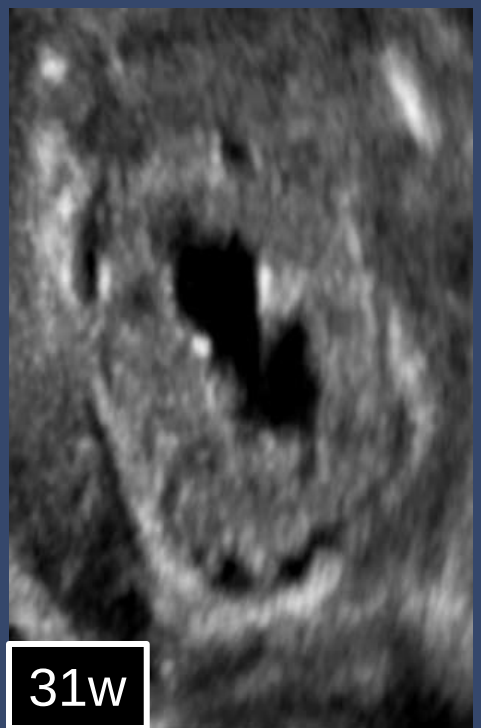
ANEUPLOIDY AND SYNDROMES

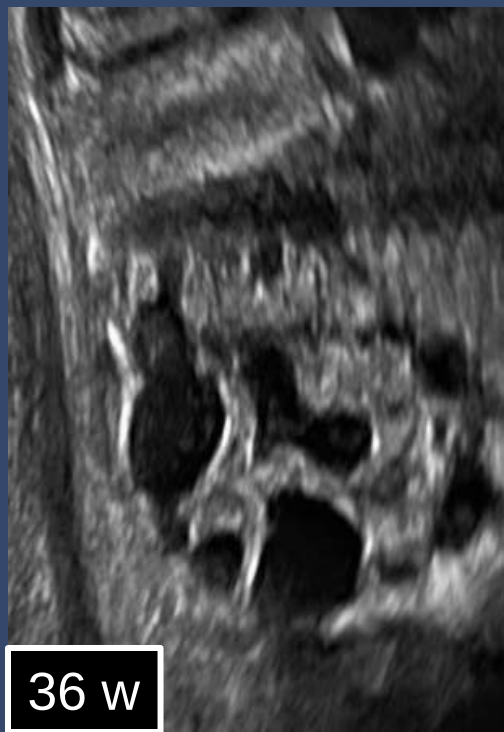
Obstructive Renal Dysplasia

- Tubular destruction and cyst formation due to increased pressure in collecting system
- Most common cause is posterior urethral valves
  - UPJ, UVJ less common





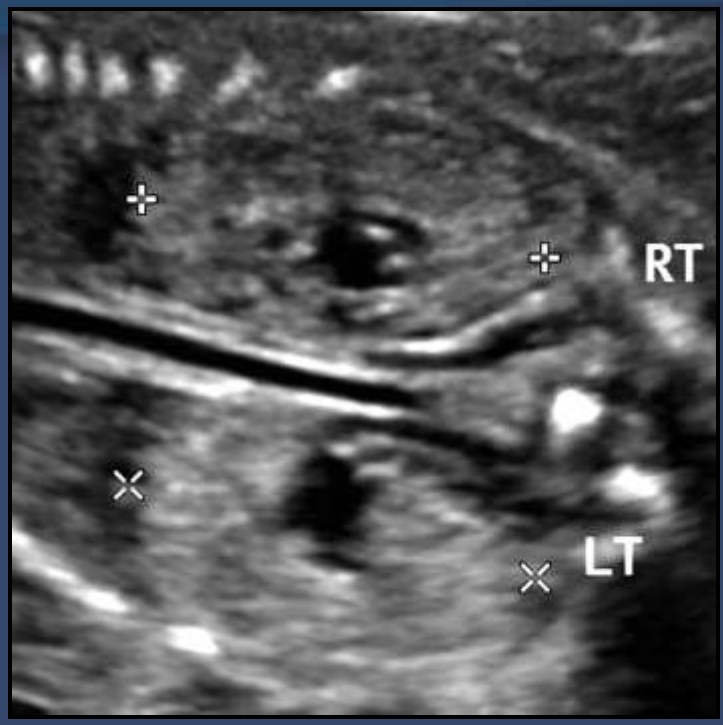


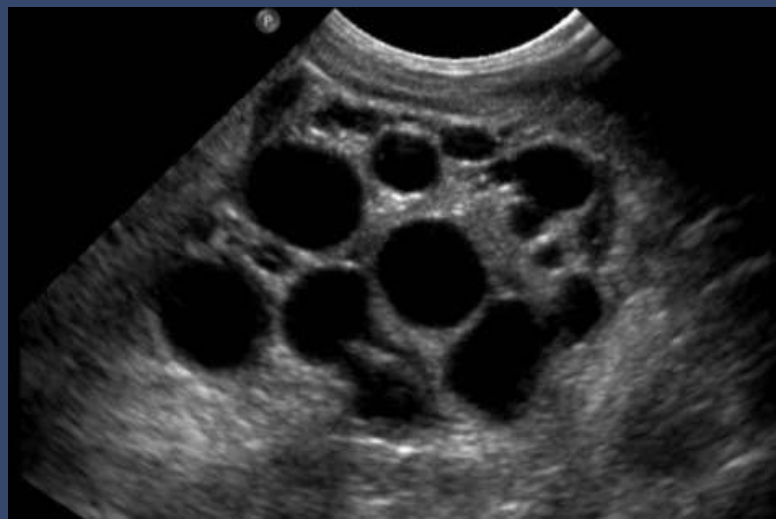


Obstructive renal dysplasia

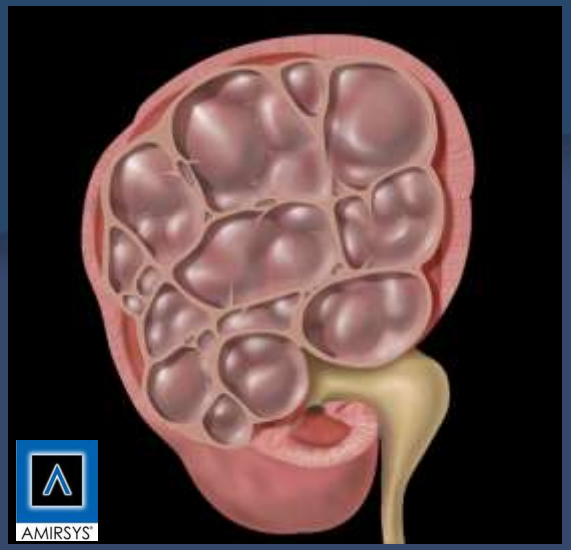
- Imaging Pearls
  - Variable hydronephrosis
  - Hyperechoic kidney
    - Early: Loss of corticomedullary distinction (lose hypoechoic pyramids)
  - Cysts (late)
- Renal size may be ↑, ↓, NI
  - ↓ size suggests late finding (poor prognostic sign)

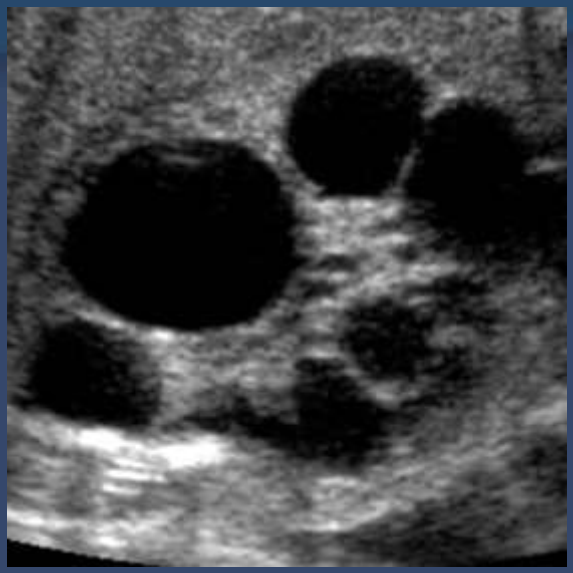
Two fetuses with PUV





Multicystic dysplastic kidney





- Multiple noncommunicating cysts of variable size
- Reniform shape often lost (not always)
- Bilateral MCDK in 20%
- Contralateral other renal anomaly in 40% (UPJ, agenesis)
- Can enlarge dramatically in fetal life
- Tend to decrease in size in neonatal life
- Most of the time, the kidney affected does not work

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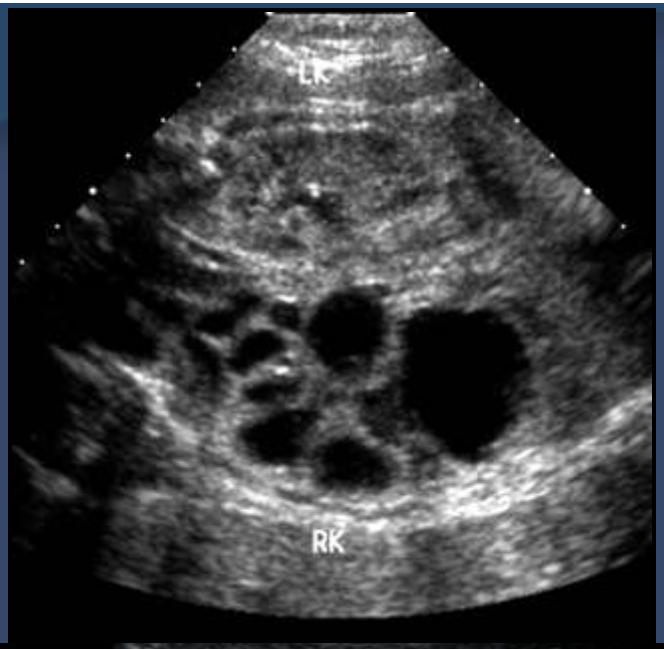
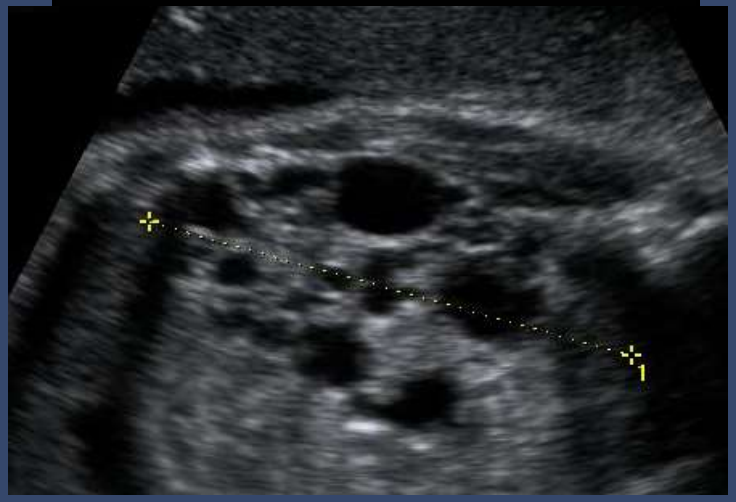
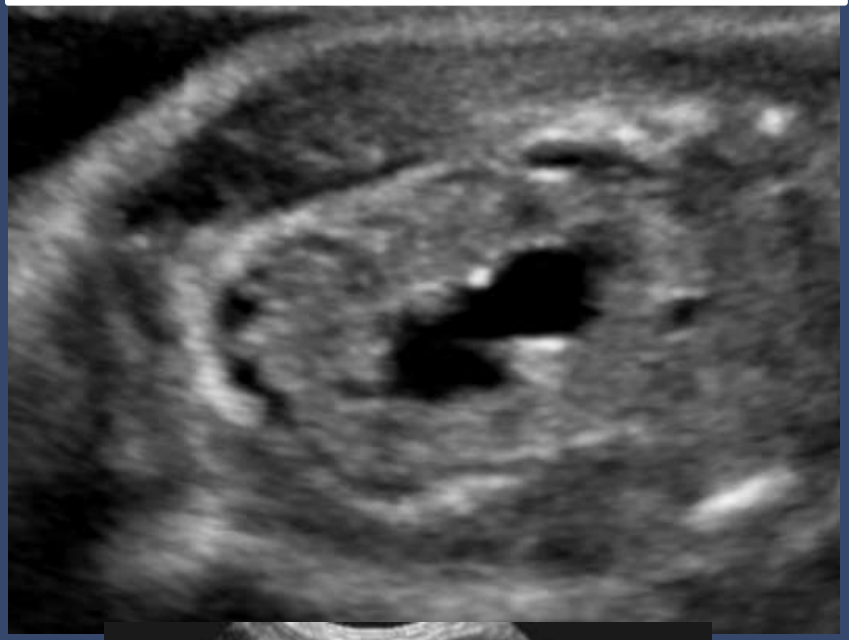
MCDK vs Post obstructive dysplasia ?

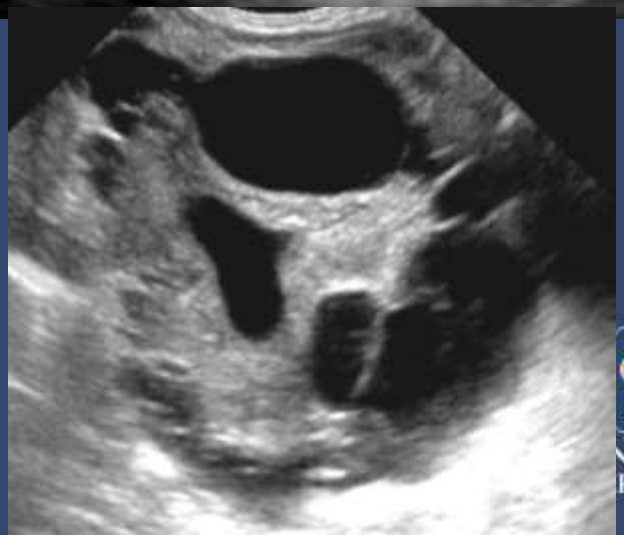
2 cases antenatal dx: MCDK





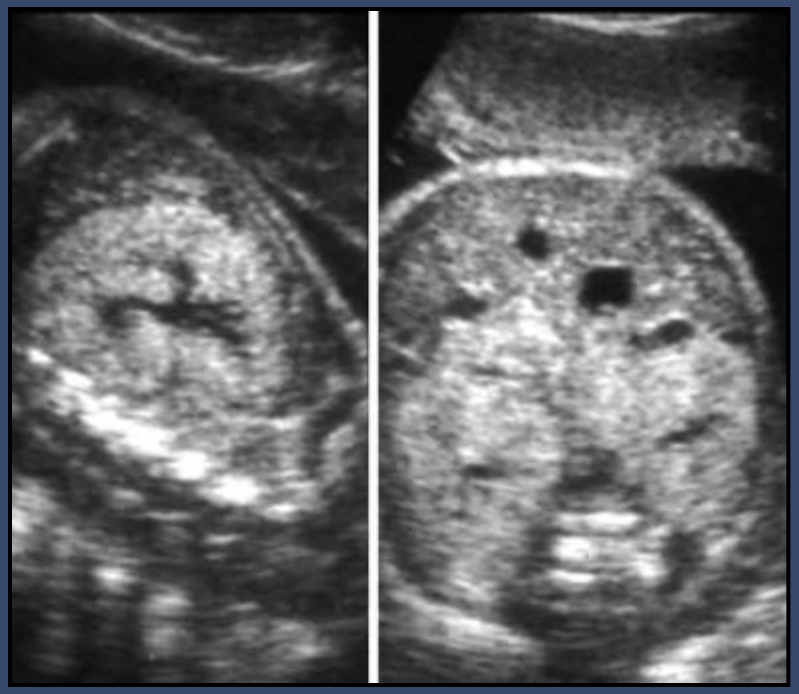
1 case prenatal dx: Post Obst CD
Post natal: MCDK

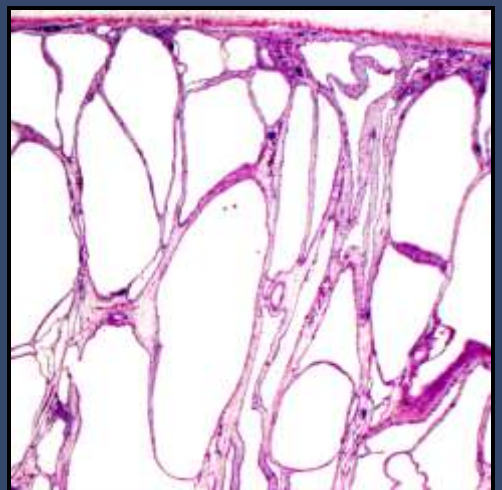




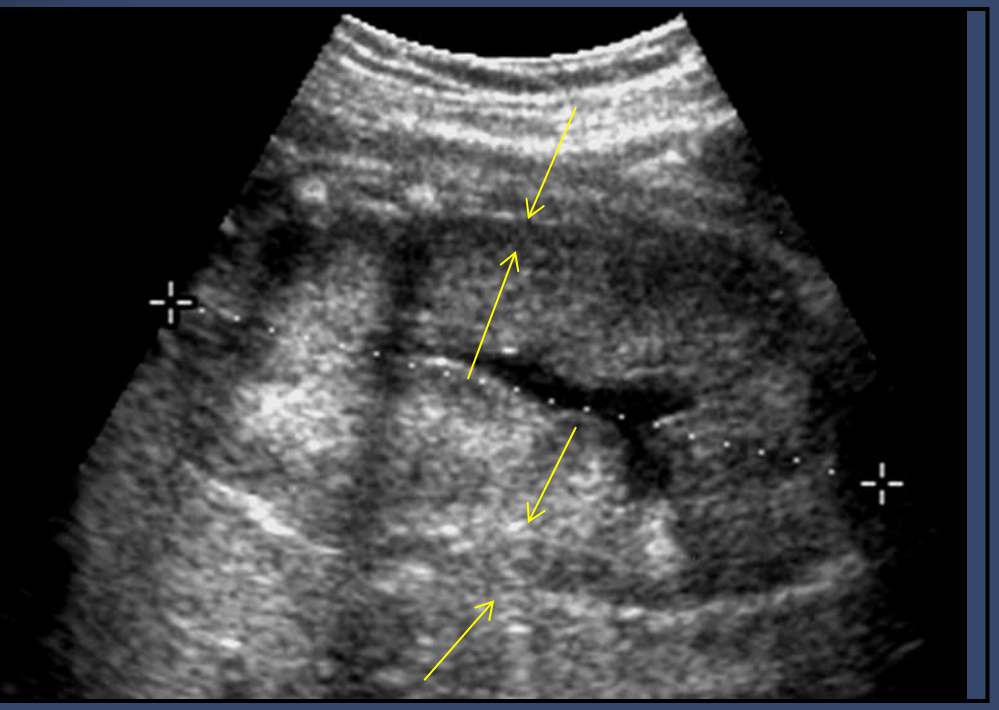
Autosomal recessive polycystic kidney disease (ARPKD)

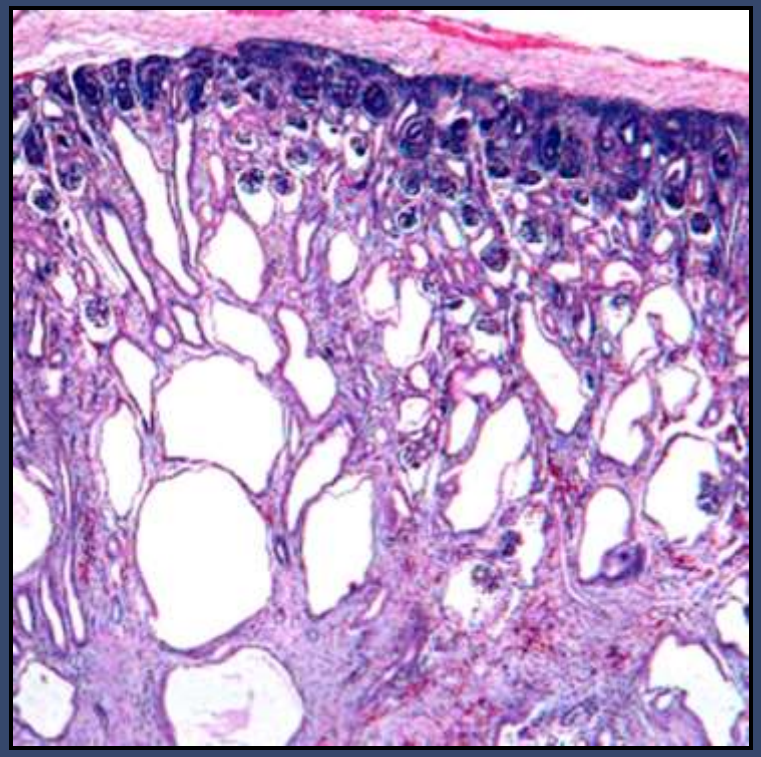
- Ectasia of collecting ducts and distal tubules
- Imaging pearls:
  - Enlarged, hyperechoic kidneys +/- visible cysts
  - Hypoechoic cortex may be seen (affects tubules> cortex)
- Variable oligohydramnios





ARPCKD Imaging pearl: relatively “spared” hypoechoic cortex





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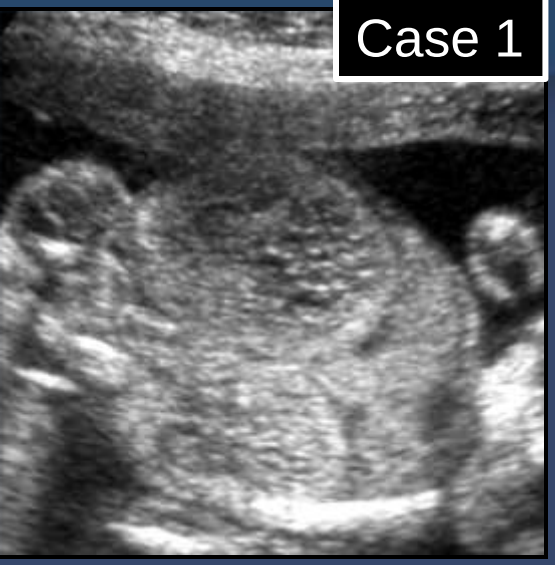
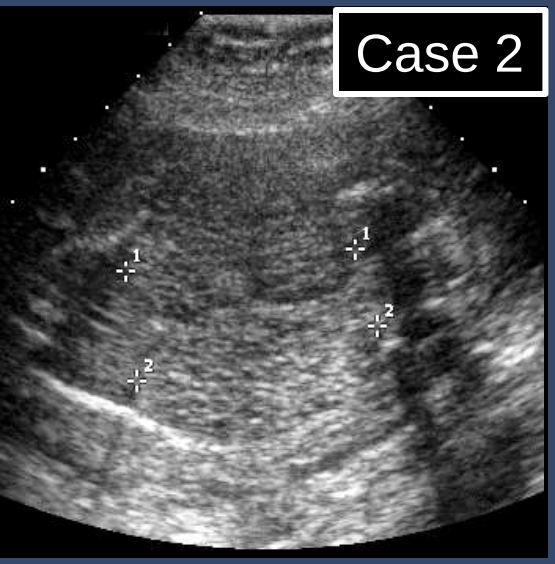
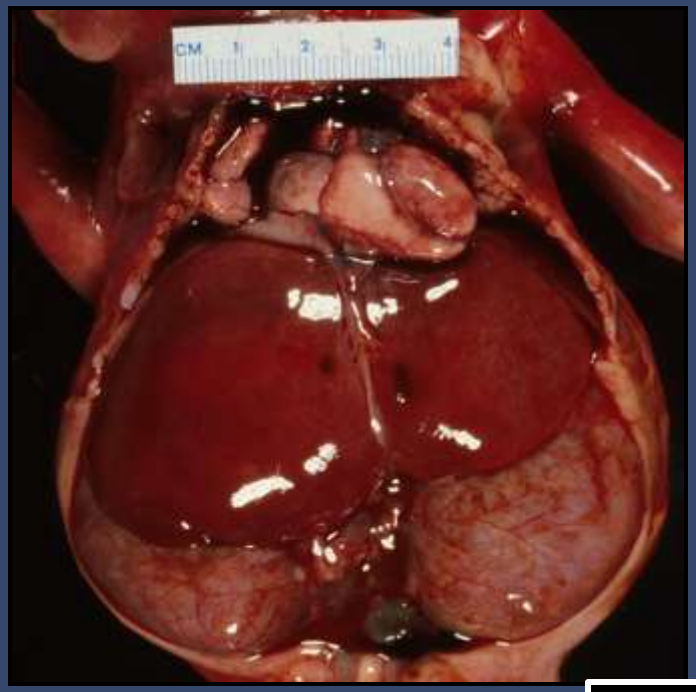
ARPCKD Imaging pearl: Variable fluid: prognosis depends on amount of pulmonary hypoplasia

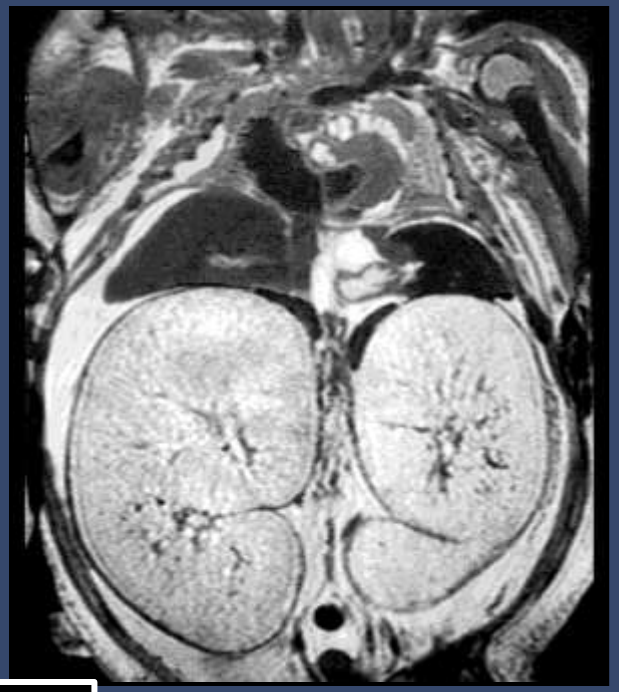


Case 1



Case 2





Case 3

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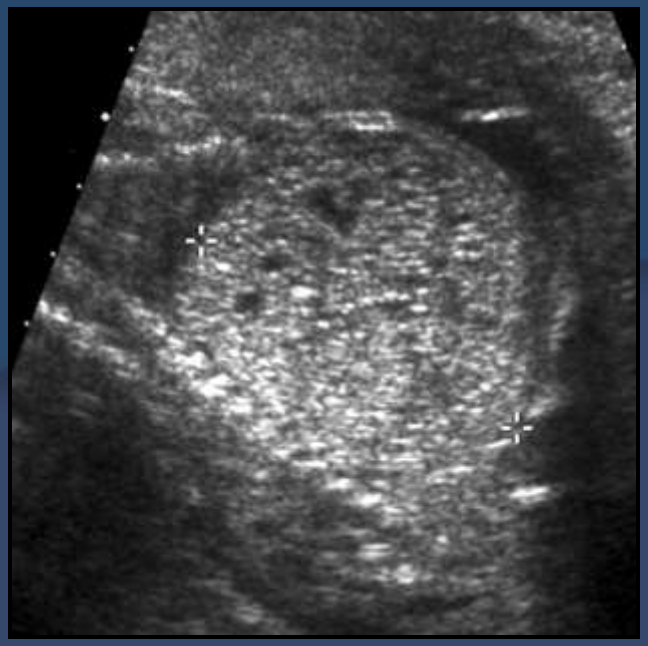
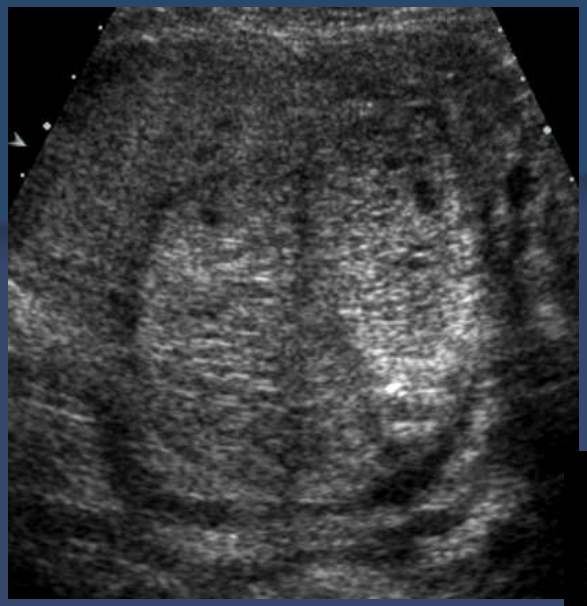
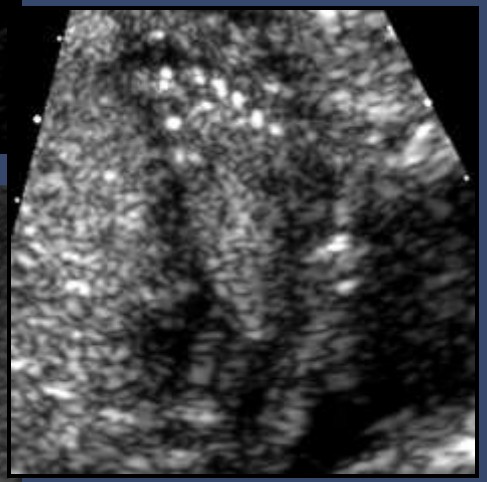
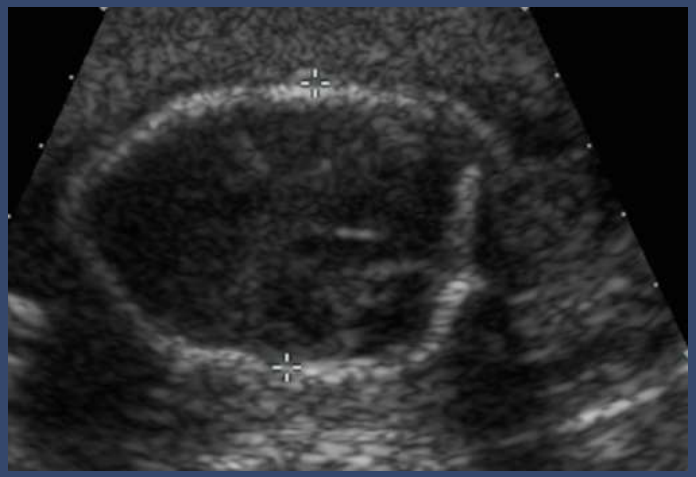
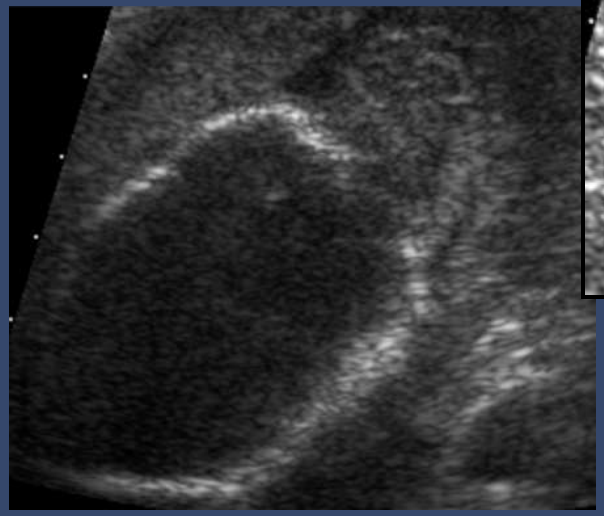
OTHER CAUSES OF BILATERAL CYSTIC KIDNEYS











Diagnosis: Meckel Gruber Syndrome

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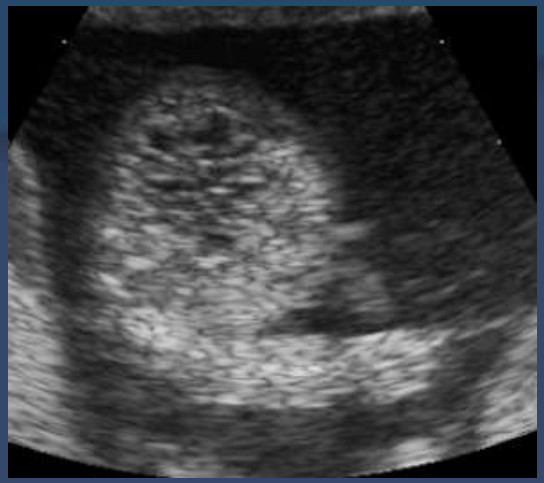
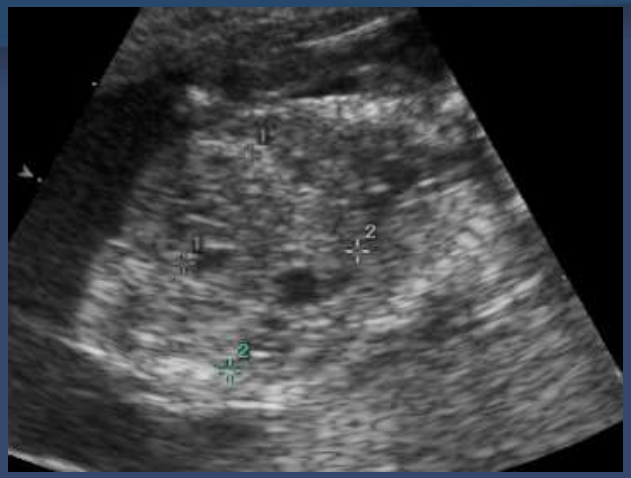
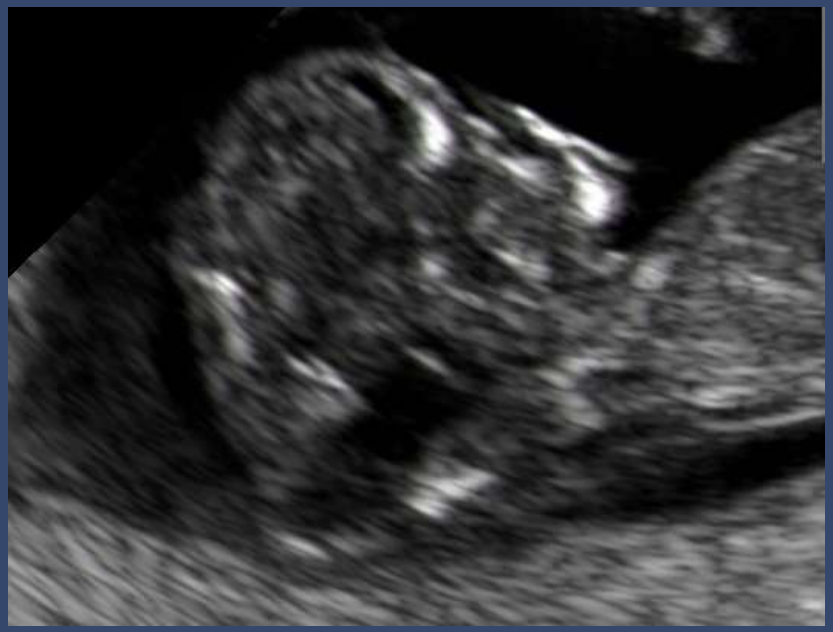
Second pregnancy 2 years later







Meckel Gruber: Autosomal Recessive

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Meckel-Gruber Features

Renal cystic dysplasia in 95-100%

Cephalocele other CNS anomaly in 90%

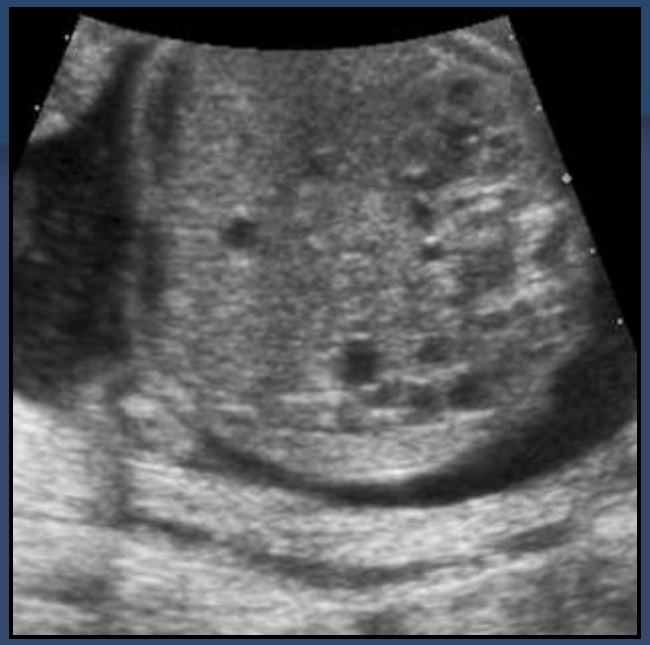
Postaxial polydactyly: 55-75%

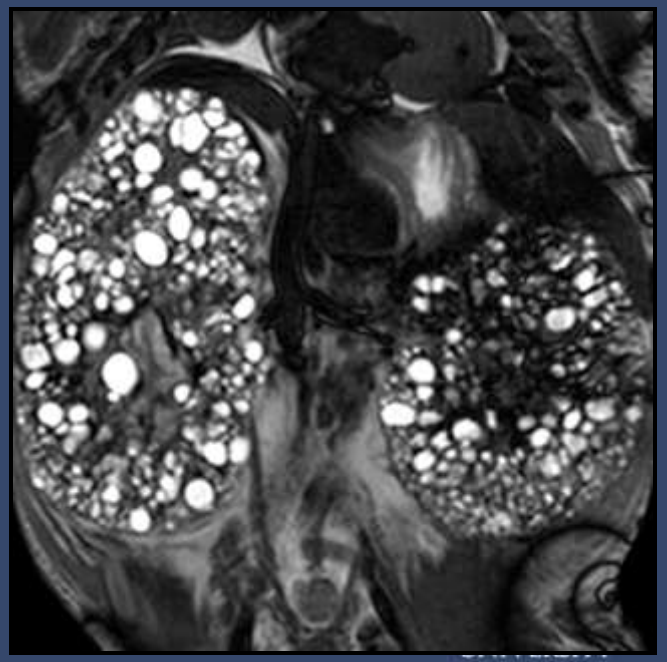
At least 2 of 3 classic features required for the clinical diagnosis

Autosomal recessive pattern

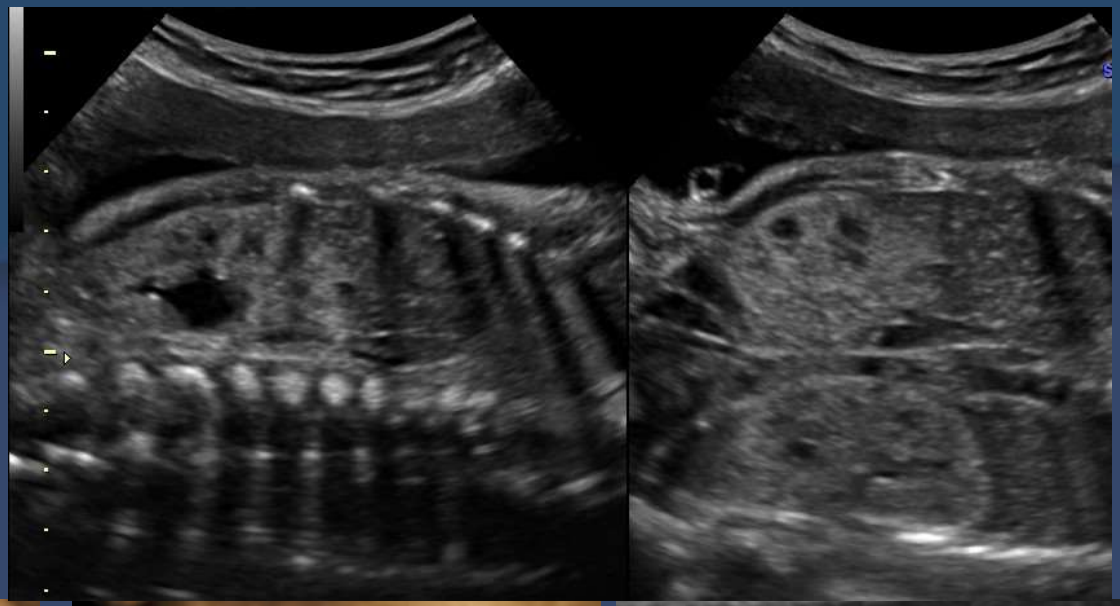
Genetic testing available

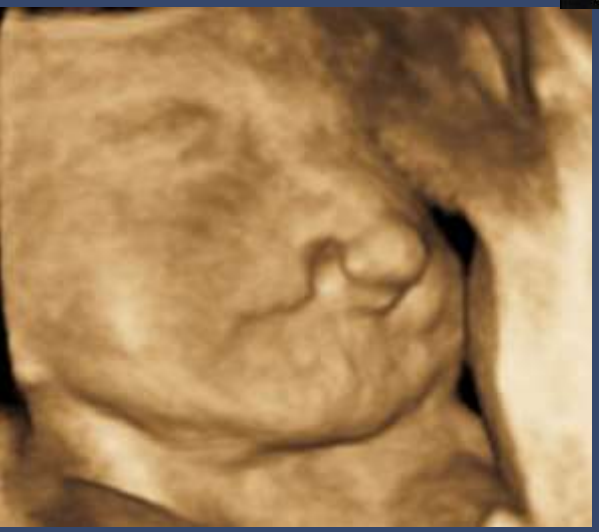
(8 genes identified, 6 gene panel)

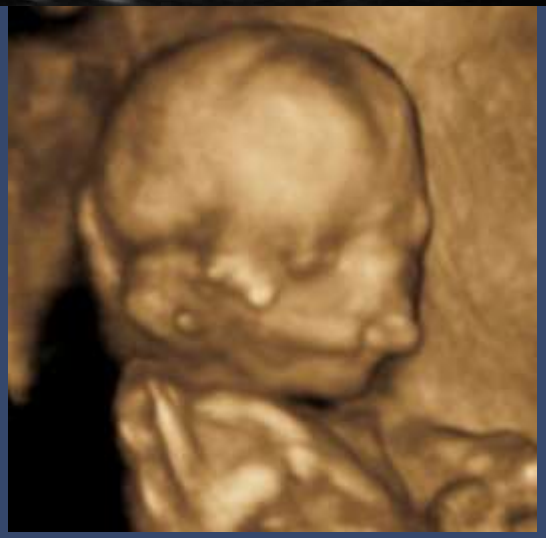


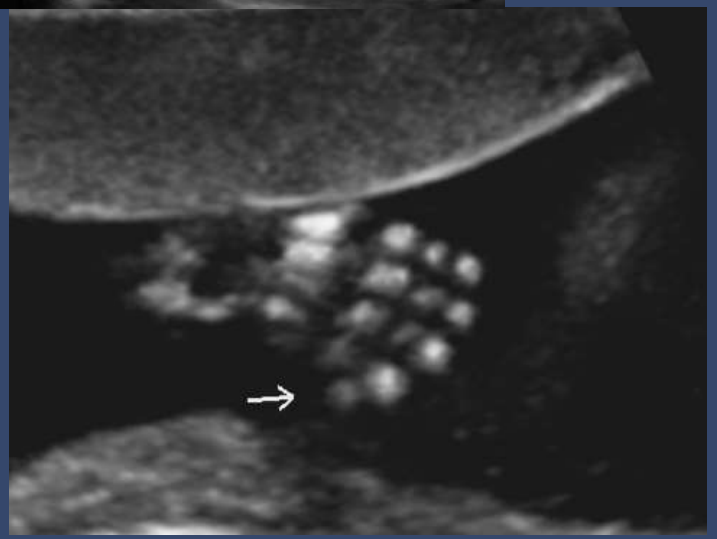


New Case









Trisomy 13

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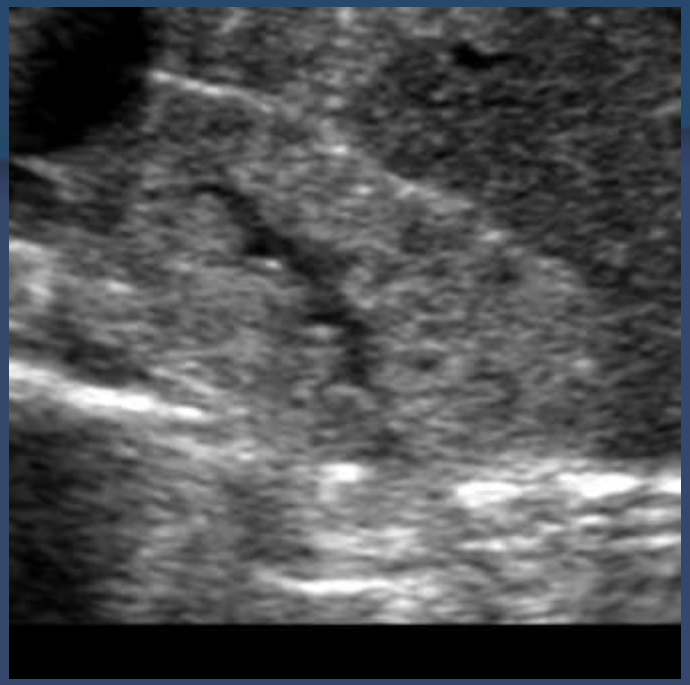
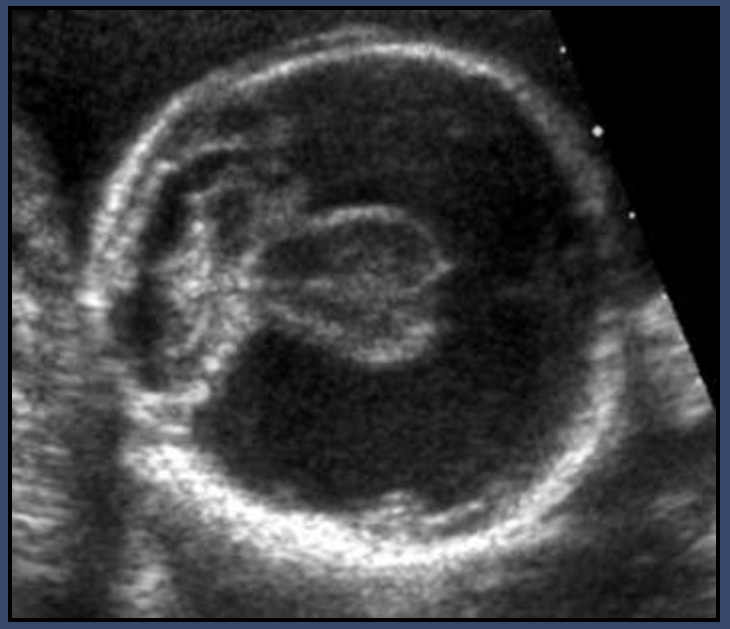
Trisomy 13

- Renal anomalies in 50%
  - Cystic dysplasia
  - Hydronephrosis, duplications
- CNS anomalies in 70%
  - Holoprosencephaly
- Cardiac anomalies 80%
- Facial anomalies 50%
- Skeletal anomalies 50%

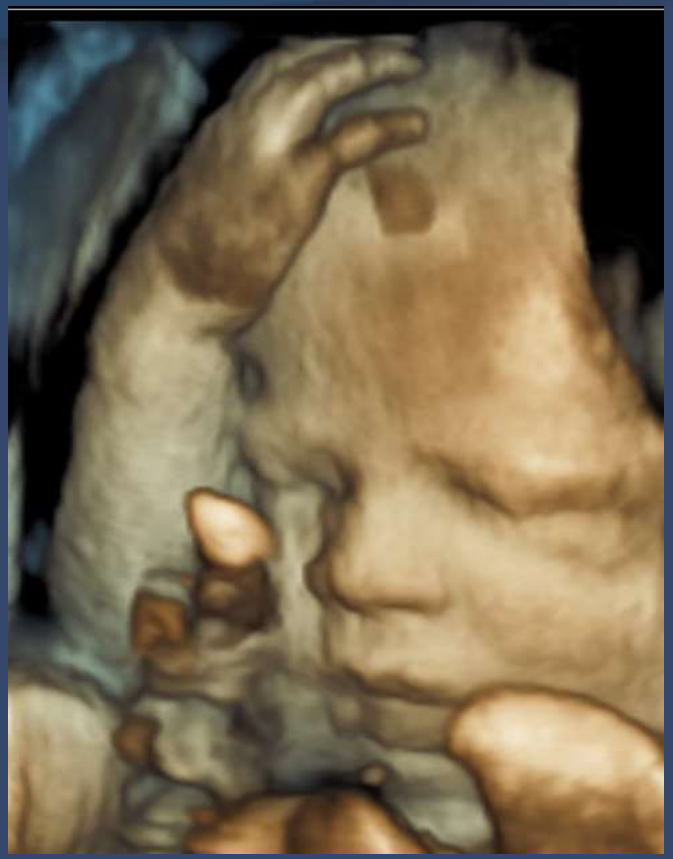




Renal Cystic Dysplasia

Diagnosis	Unilateral or Bilateral	Imaging Pearls	Kidney size	Associations	Prognosis
Post obstructive cystic dysplasia	Bilateral or unilateral	Cortical cysts Partial if duplicated	Normal or small	PUV > UPJ, ureterocele	Excellent if unilateral
MCDK	L>R, 20% Bi	Scattered cysts of variable size	Increased	40% contralateral renal anomaly	Excellent if unilateral
ARPCKD	Bilateral	Echogenic kidney with cortical sparing	Increased	Hepatic disease in pediatric pop	Poor if pulmonary hypoplasia
Meckel Gruber	Bilateral	Echogenic kidneys +/- cysts	Increased	Encephalocele polydactyly	Poor/fatal
Trisomy 13	Bilateral	Echogenic kidneys +/- cysts	Increased	Holoprosencephaly Polydactyly Heart defect	Poor/Fatal

THE END…..MY BRAIN IS FULL!!



THANK YOU FOR YOUR ATTENTION

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