FETAL KIDNEY ASSESSMENT

22ND ANNUAL OBSTETRICAL ULTRASOUND

SETTING THE STANDARD FOR 2019

Sunnybrook
HEALTH SCIENCES CENTRE

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OBJECTIVES

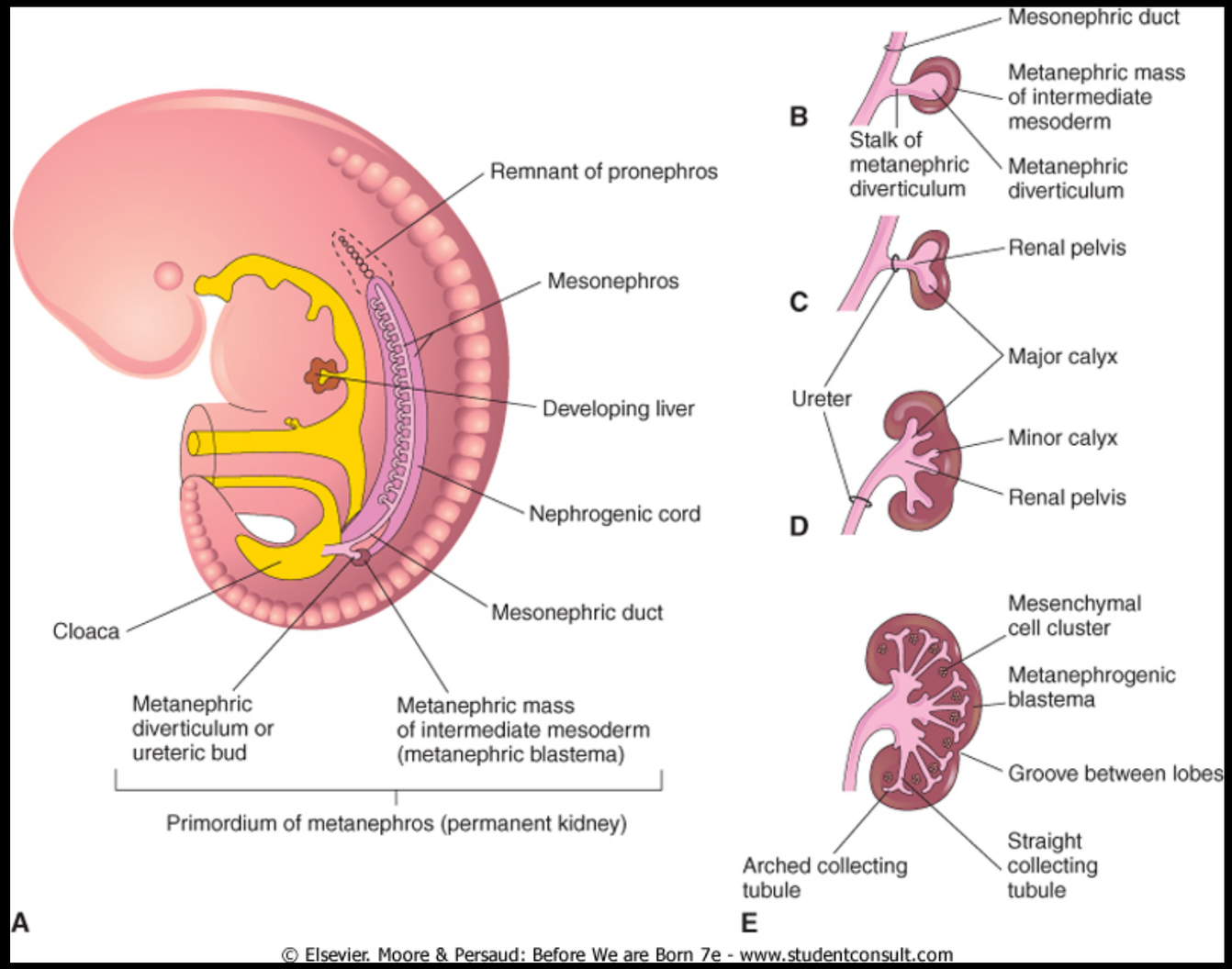
- Normal development
- Approach to Fetal GU system
- Renal Agenesis and Ectopia
- Cystic Renal Diseases
- Echogenic Kidneys
- Urinary Tract Dilatation
- Renal Tumors

NORMAL DEVELOPMENT

FETAL KIDNEYS

DEVELOPMENT OF FETAL KIDNEY

- 3 sets of fetal kidneys:
  - Pronephros
  - Mesonphros
  - Metanephros (permanent)
- 2 key structures are ureteric bud and metanephric mass
- Develops in pelvis and ascends to normal position by 11th week

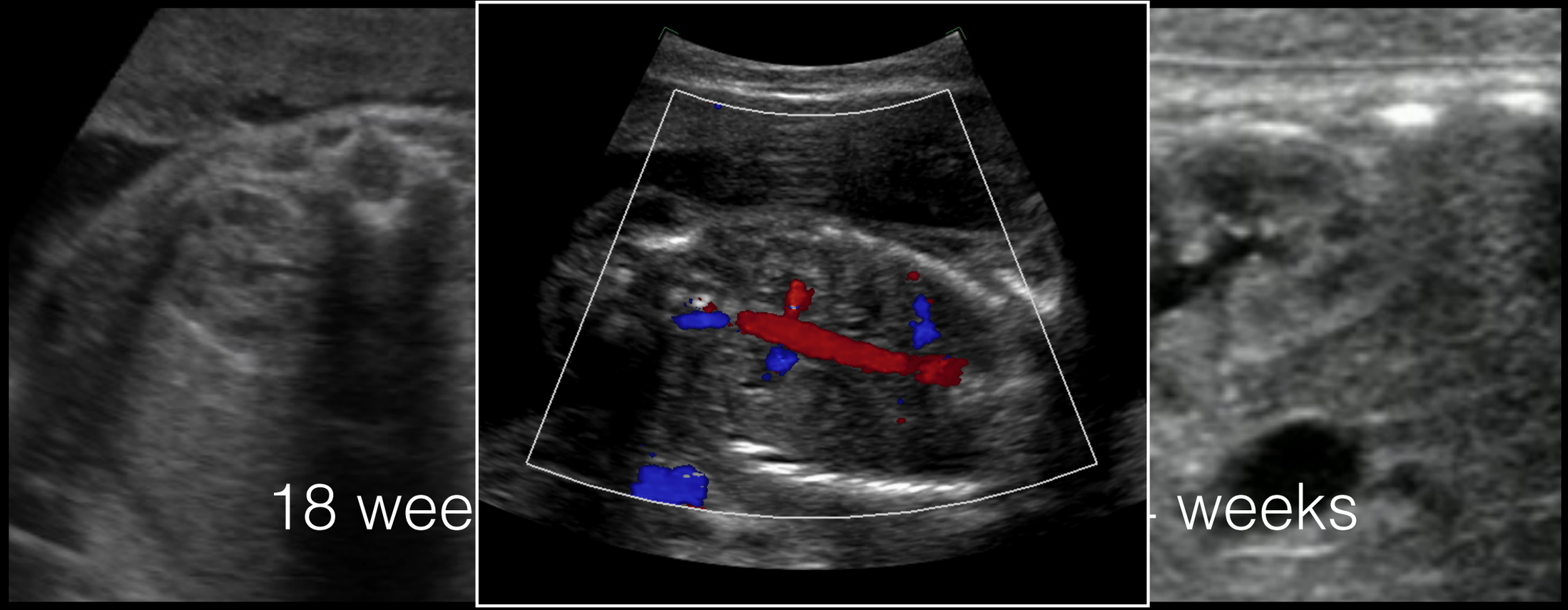


EARLY APPEARANCE FETAL KIDNEYS

- Kidneys visible 99% fetuses by 12-13 weeks
- Early sonographic appearance:
  - Oval hyperechoic paravertebral masses
  - Echogenicity similar to liver or spine
  - Corticomedullary differentiation (CMD) by 14-15 weeks
  - Reference charts for size
  - Consistent renal/abdominal circumference ratio 0.27-0.3

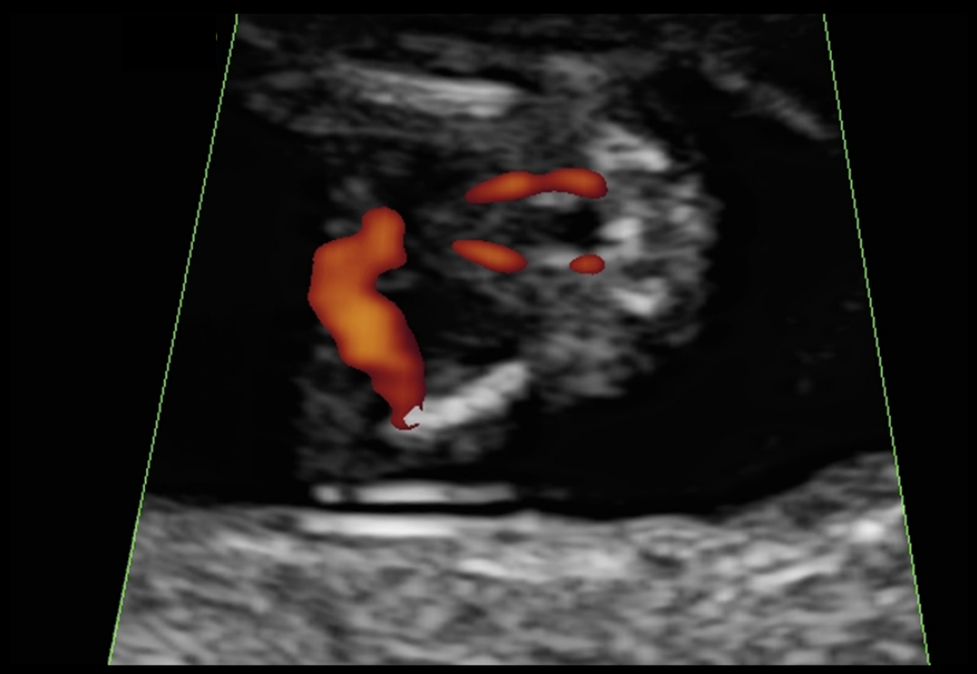
SECOND TRIMESTER APPEARANCE

- More isoechoic (less echogenic than liver/spleen)
- CMD should always be seen > 18 weeks
- CDS can confirm renal arteries



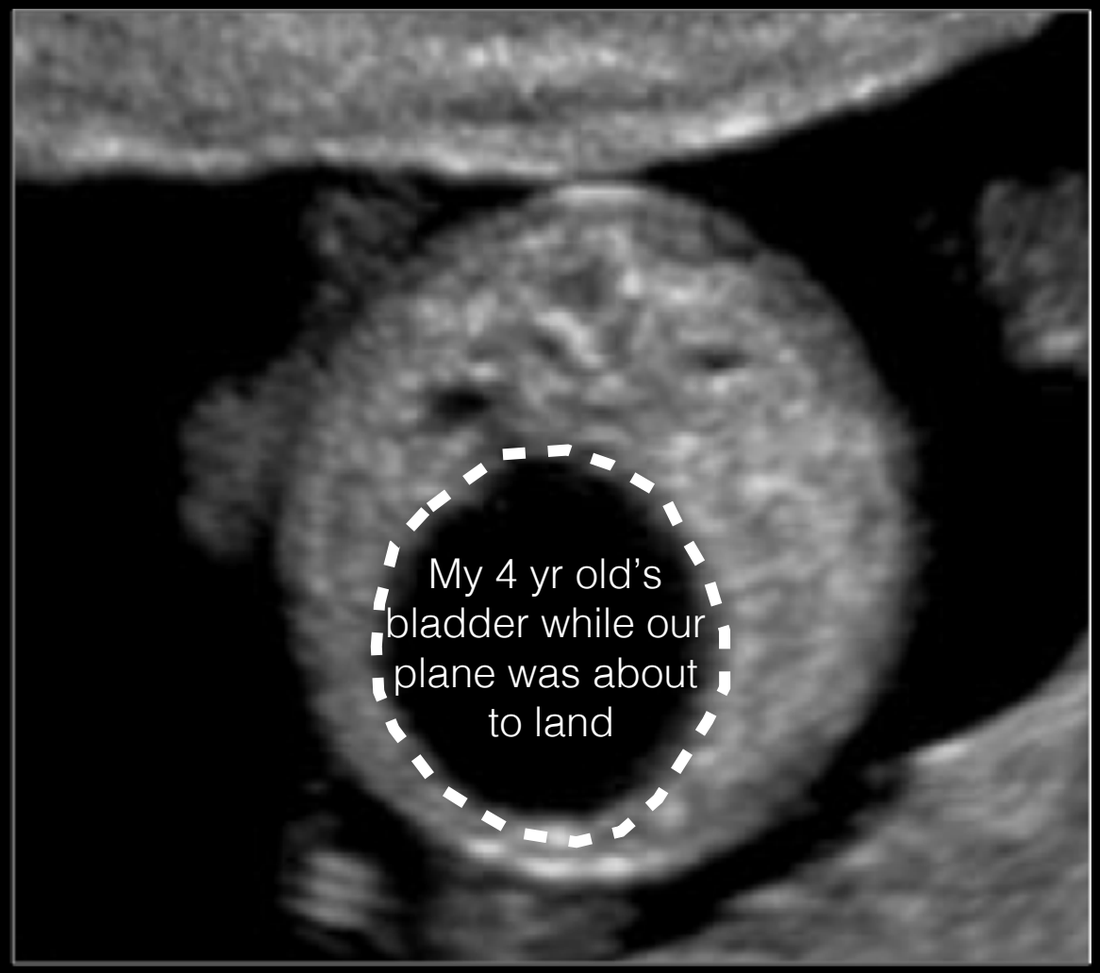
BLADDER AND FLUID

- Kidneys start producing urine around 9 weeks
- By 14-16 weeks, 2/3 of AFV from kidneys
- Bladder visible 98% by 12-13 weeks
- Bladder fills/empties around every 25 min



MEGACYSTIS FIRST TRIMESTER

- Sagittal bladder diameter >7 mm
- 7-15 mm
  - 90% resolve if no chromosomal abnormality
- >15 mm almost all have obstructive uropathy



APPROACH TO FETAL GU ASSESSMENT

THINGS TO CONSIDER

- Is the bladder present?
- Are kidneys present?
  - number, position, size, echogenicity, cysts
- Is urinary tract dilated?
  - level of obstruction
- Unilateral or bilateral?
- Oligohydramnios?
- Fetal gender?

GENITOURINARY ABNORMALITIES

Nephropathy

MCKD
Agenesis
PCKD

Obstructive Uropathy

UPJ
UVJ
PUV
Urethral atresia

Syndromes

Tri 13
Meckel Gruber
Beckwith Wiedemann
VHL

APPROACH

- Are kidneys present and do they look normal?
- Is bladder seen? Normal size?
- Is fluid (ie. lung maturation) compromised?
- Is problem nephropathy or uropathy? or both?
- Hereditary or non-hereditary?
- Antenatal follow up?
- Postnatal follow up?

KEY POINTS TO REMEMBER

- "B-F-F-F"
- Bladder?
- Fluid?
- Familial?
- Followup?

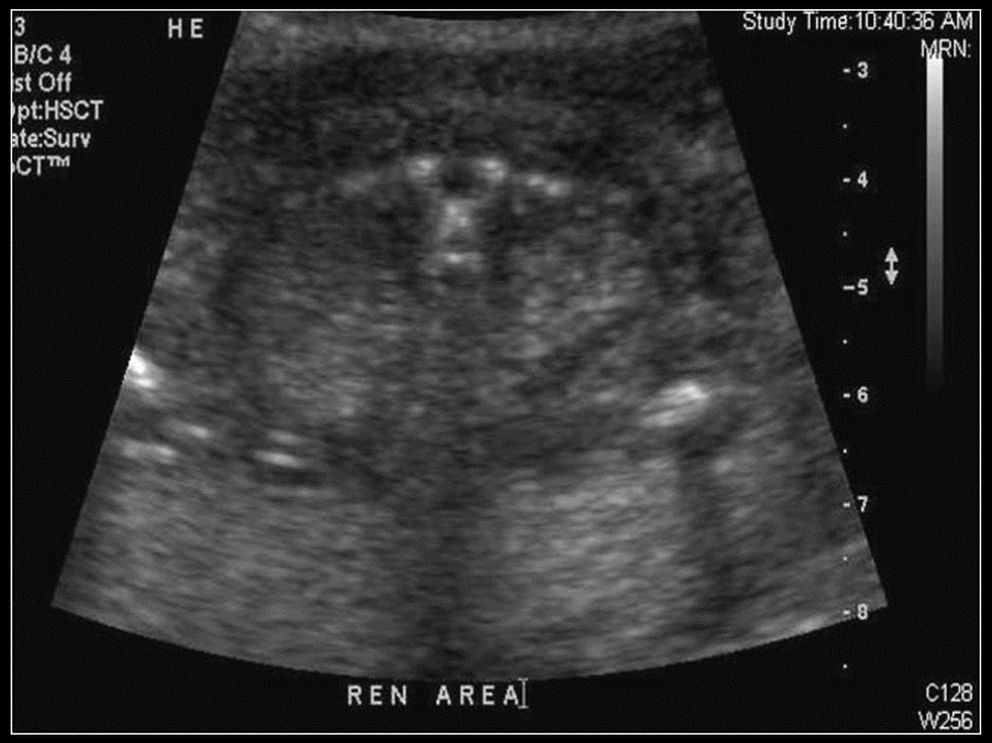
RENAL AGENESIS AND ECTOPIA

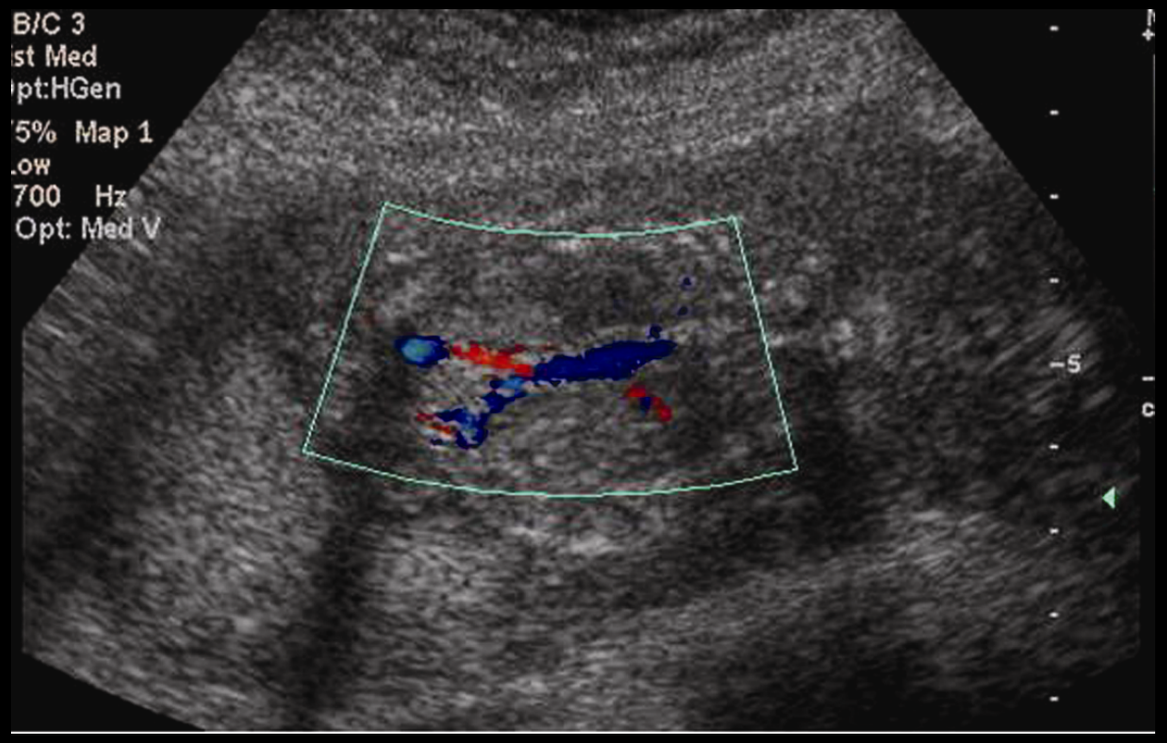
BILATERAL RENAL AGENESIS

- Lethal
- Ureteric bud fails to develop
- 1 in 4000 births
- Recurrent risk 4% for subsequent pregnancies
- Risk of silent renal GU anomalies in family members

BILATERAL RENAL AGENESIS

- Severe oligohydramnios/anhydramnios
- Non visualization of bladder for > 1 hour
  - Warning: Urachal diverticulum or retrograde filling
- Absent renal arteries

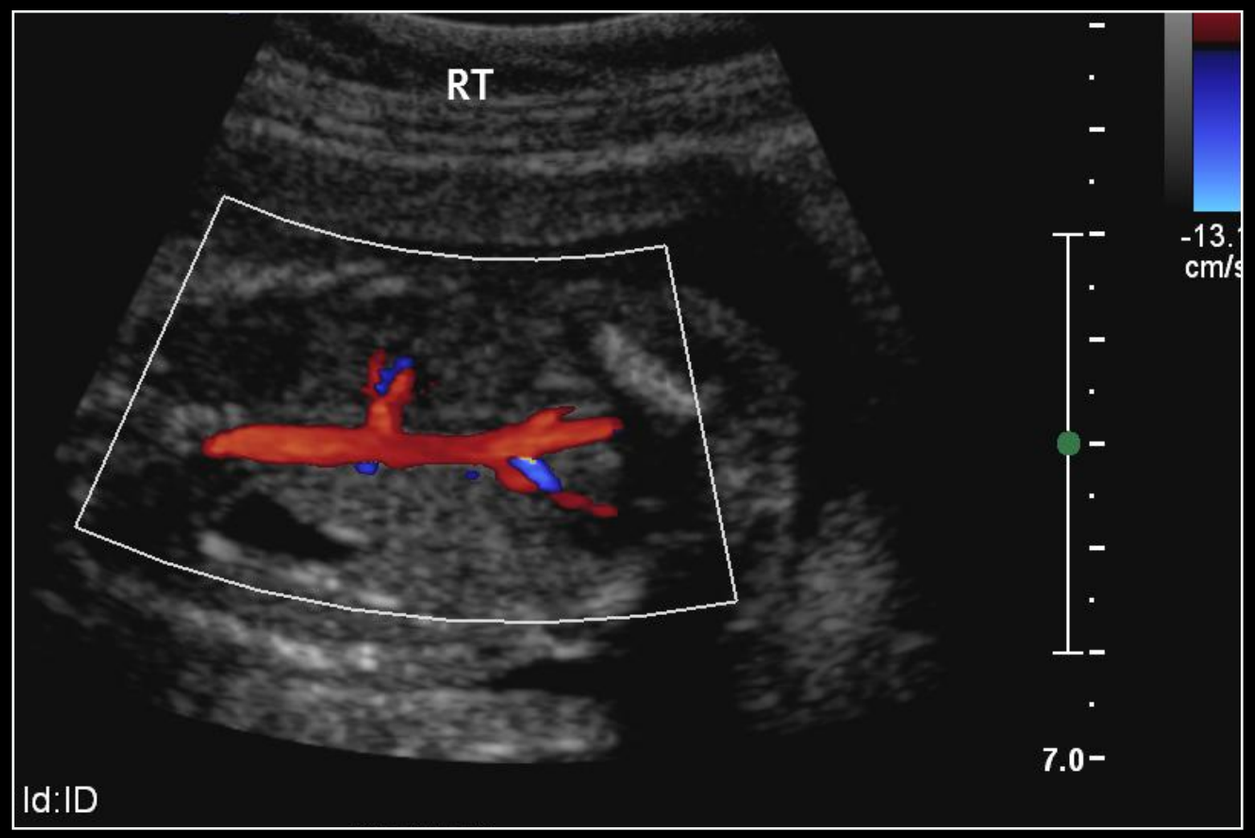


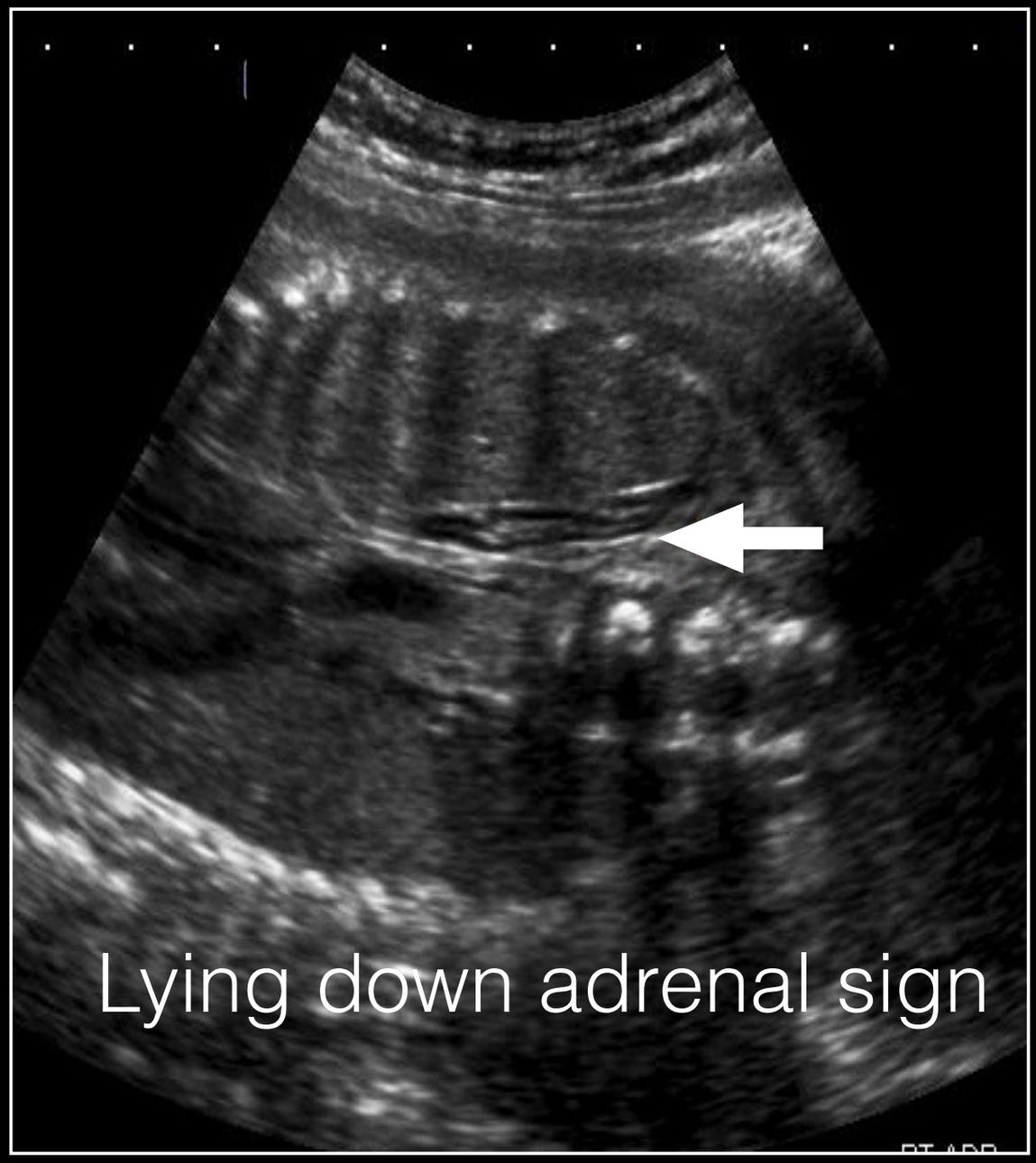


UNILATERAL RENAL AGENESIS

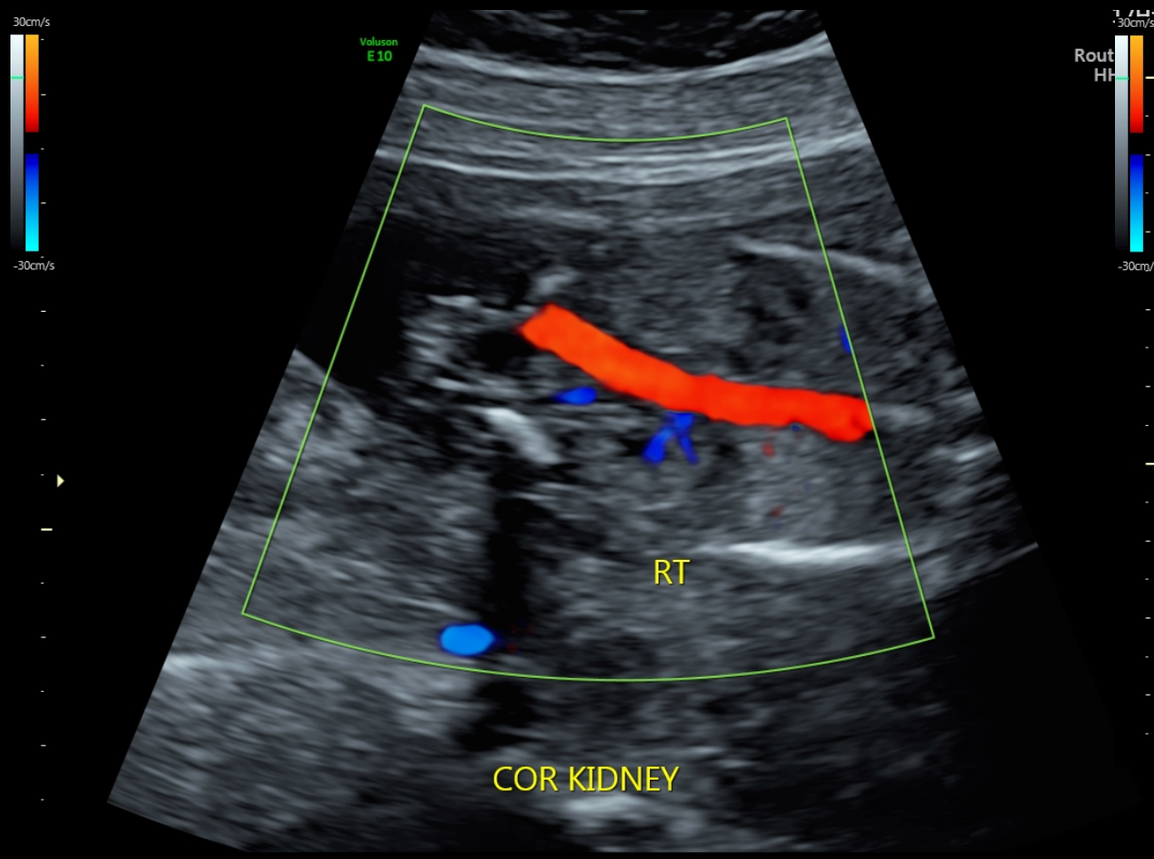
- 1 in 1000 births
- Contralateral kidney may be enlarged
- Contralateral renal anomalies:
  - VUR
  - Associations: Cardiac, skeletal, GI
- If isolated, good prognosis
- Recurrence 1% if parents normal, 7% if one parent has solitary kidney

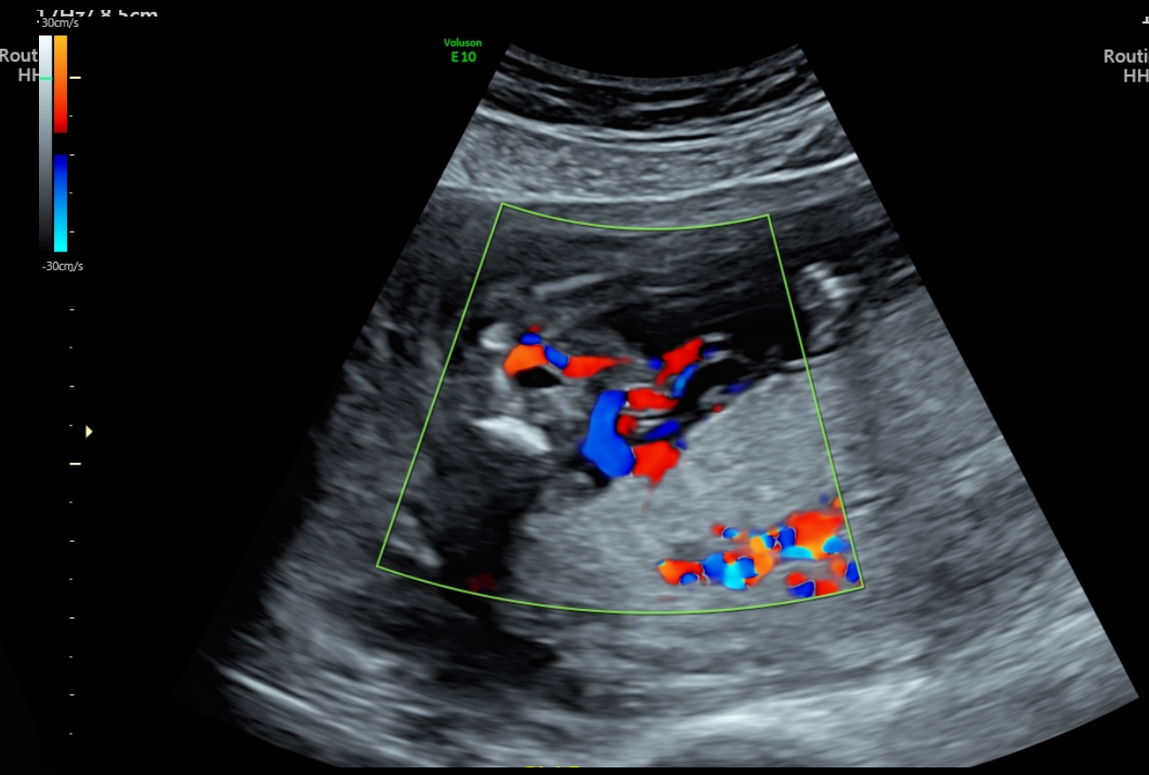
UNILATERAL RENAL AGENESIS





UNILATERAL RENAL AGENESIS

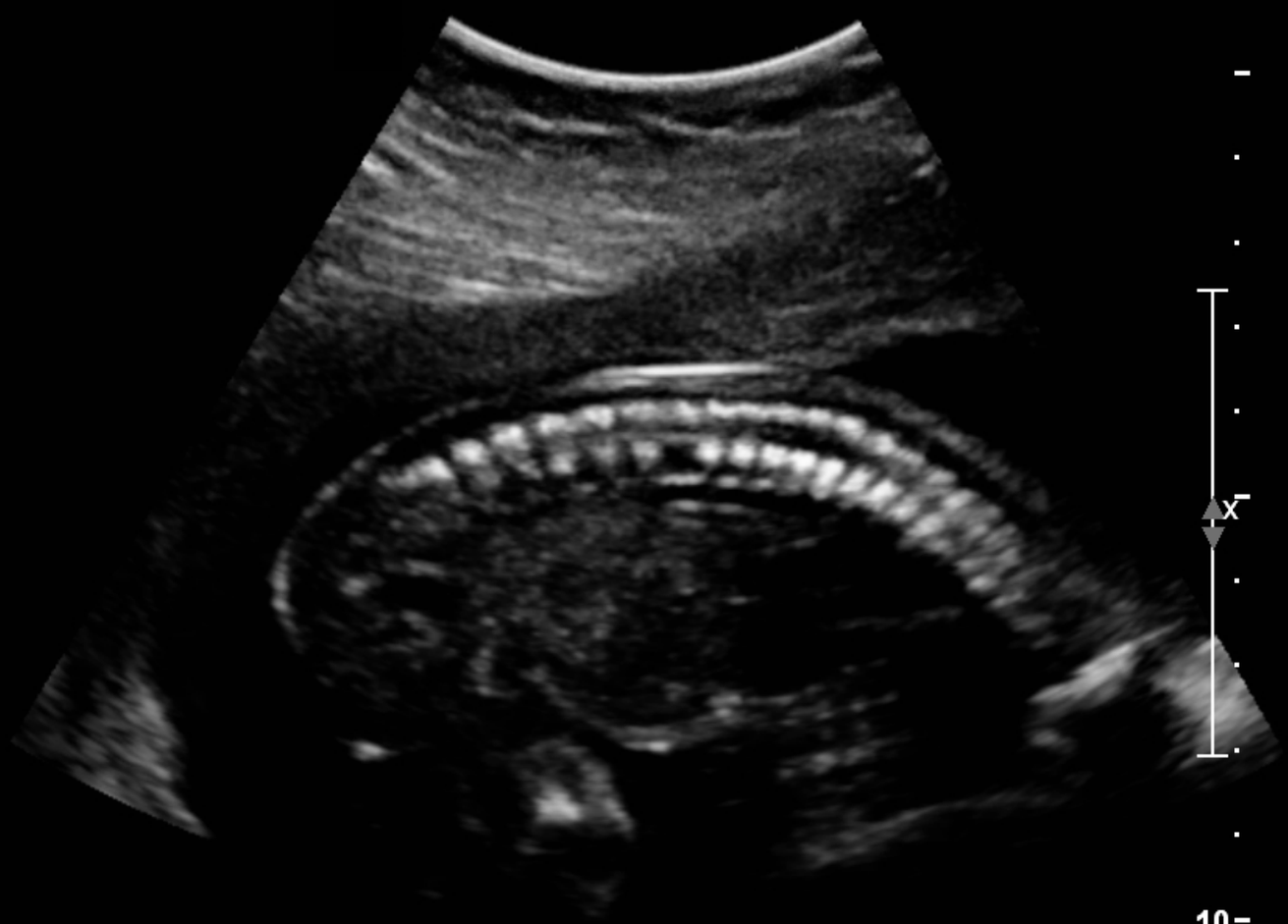


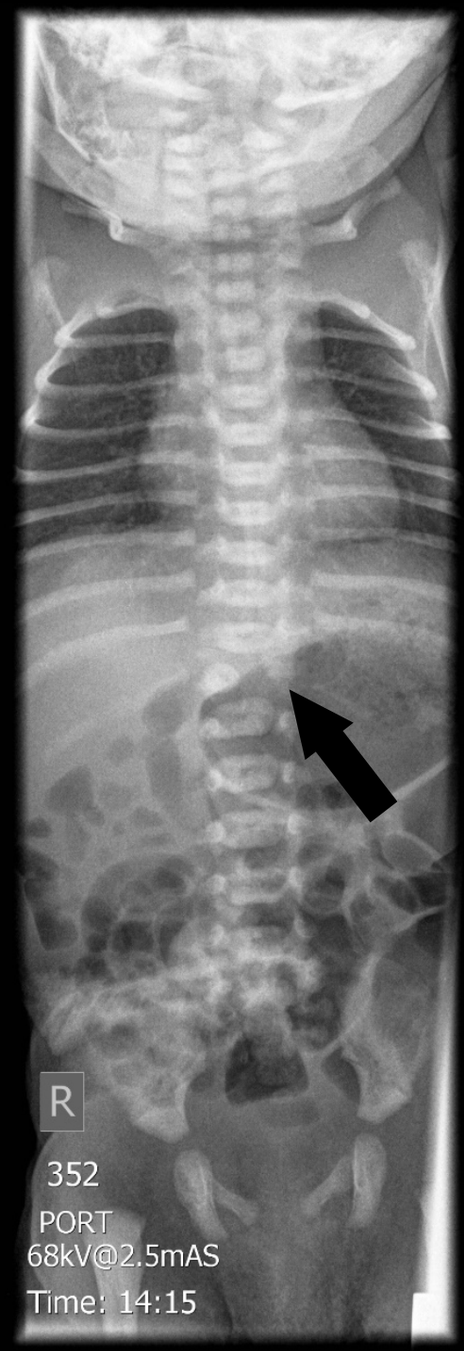


2 vessel cord

UNILATERAL RENAL AGENESIS

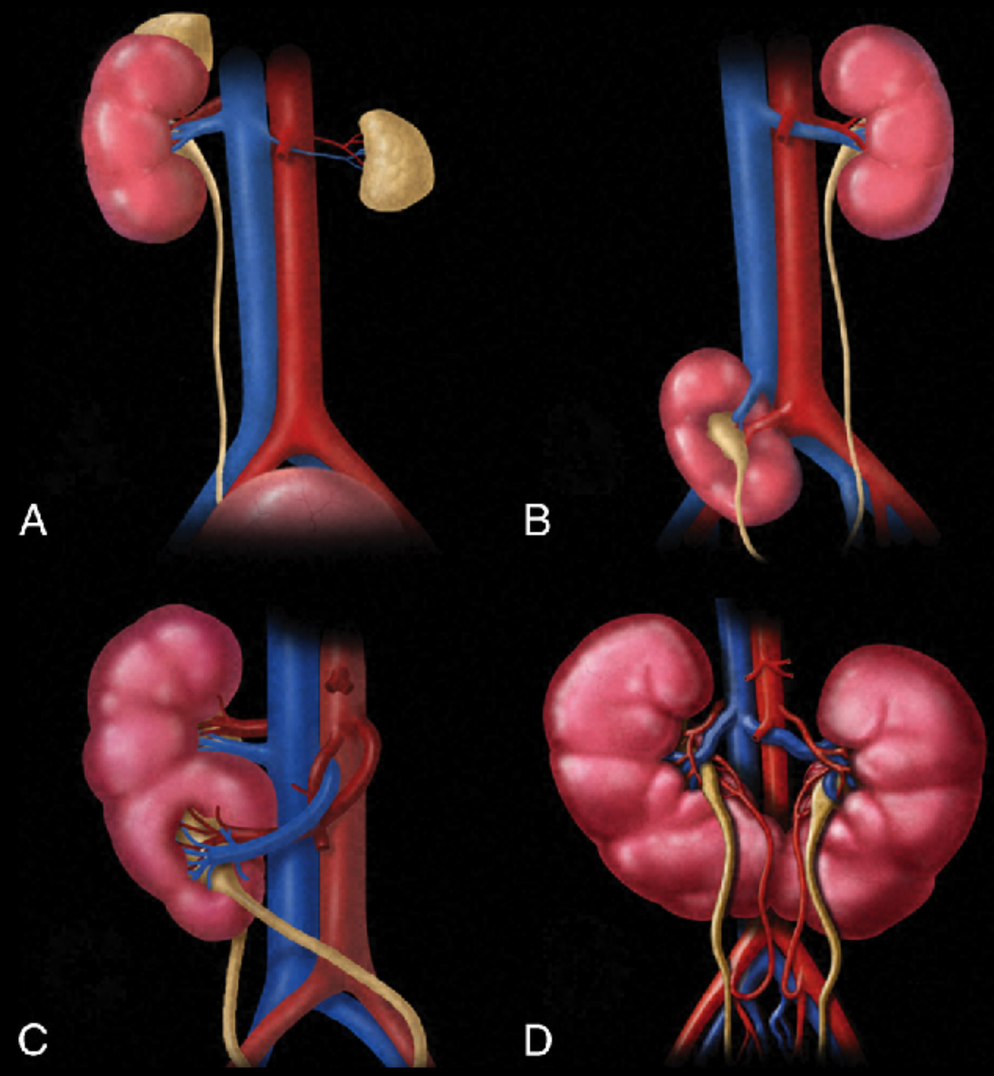
Hemivertebra



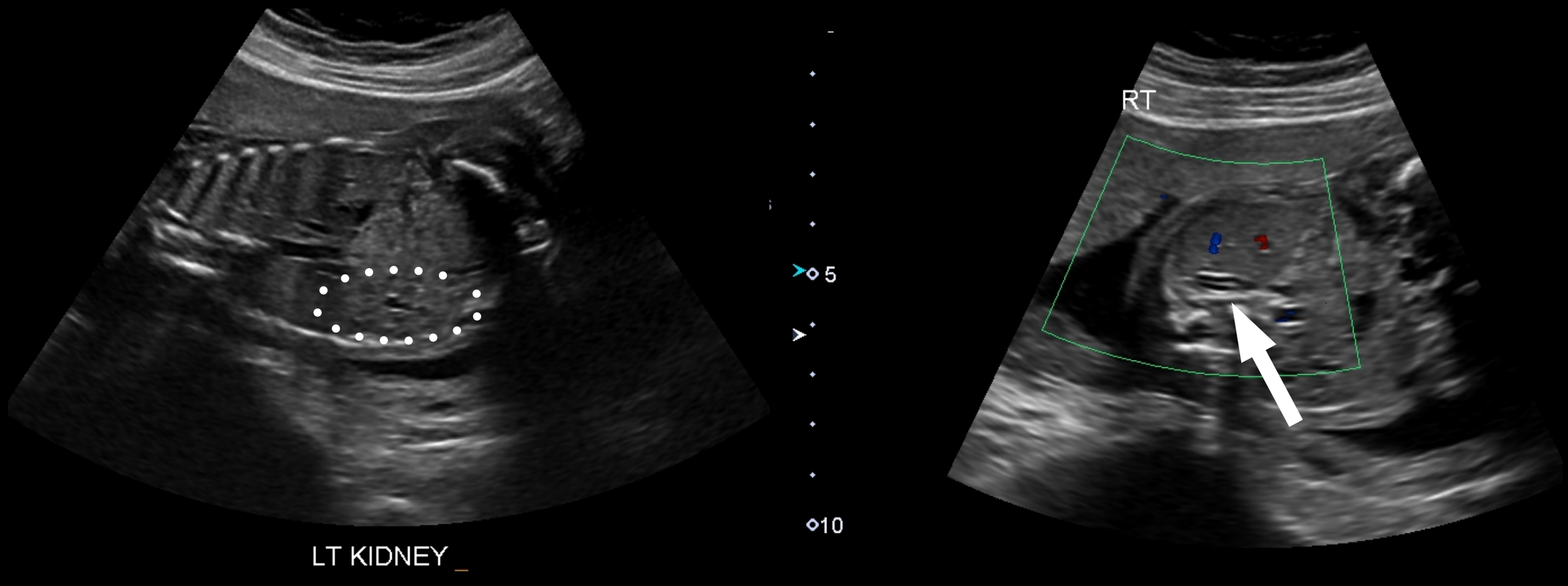


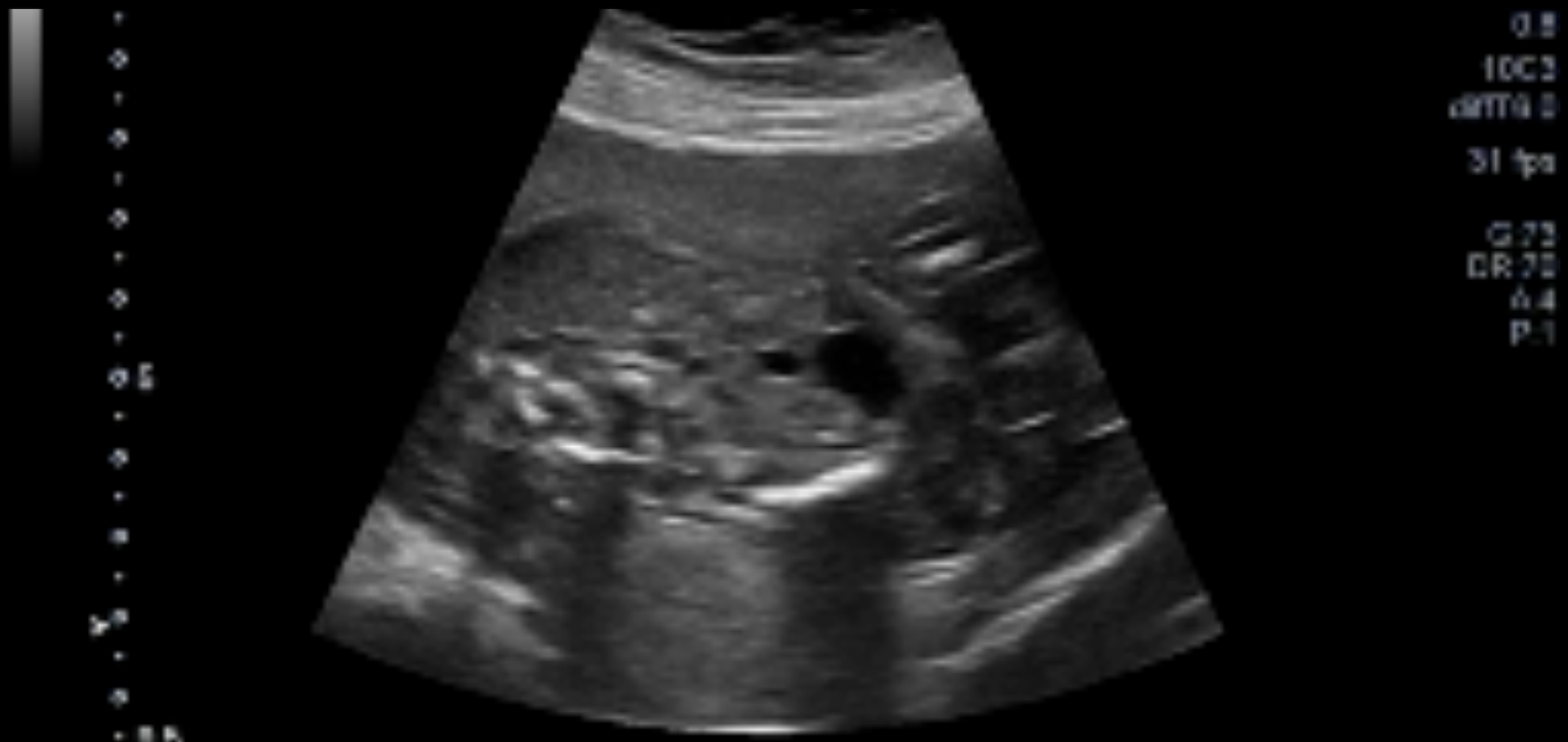
ECTOPIC KIDNEY

- Incidence of ectopic kidney 1 in 1200
- Pelvic most common
  - scan adjacent to bladder and iliacs
- Crossed renal ectopic:
  - with fusion
  - without fusion
  - *distinguished from duplex by angulation between two kidneys
- Associations: Genital, skeletal, GI, VUR

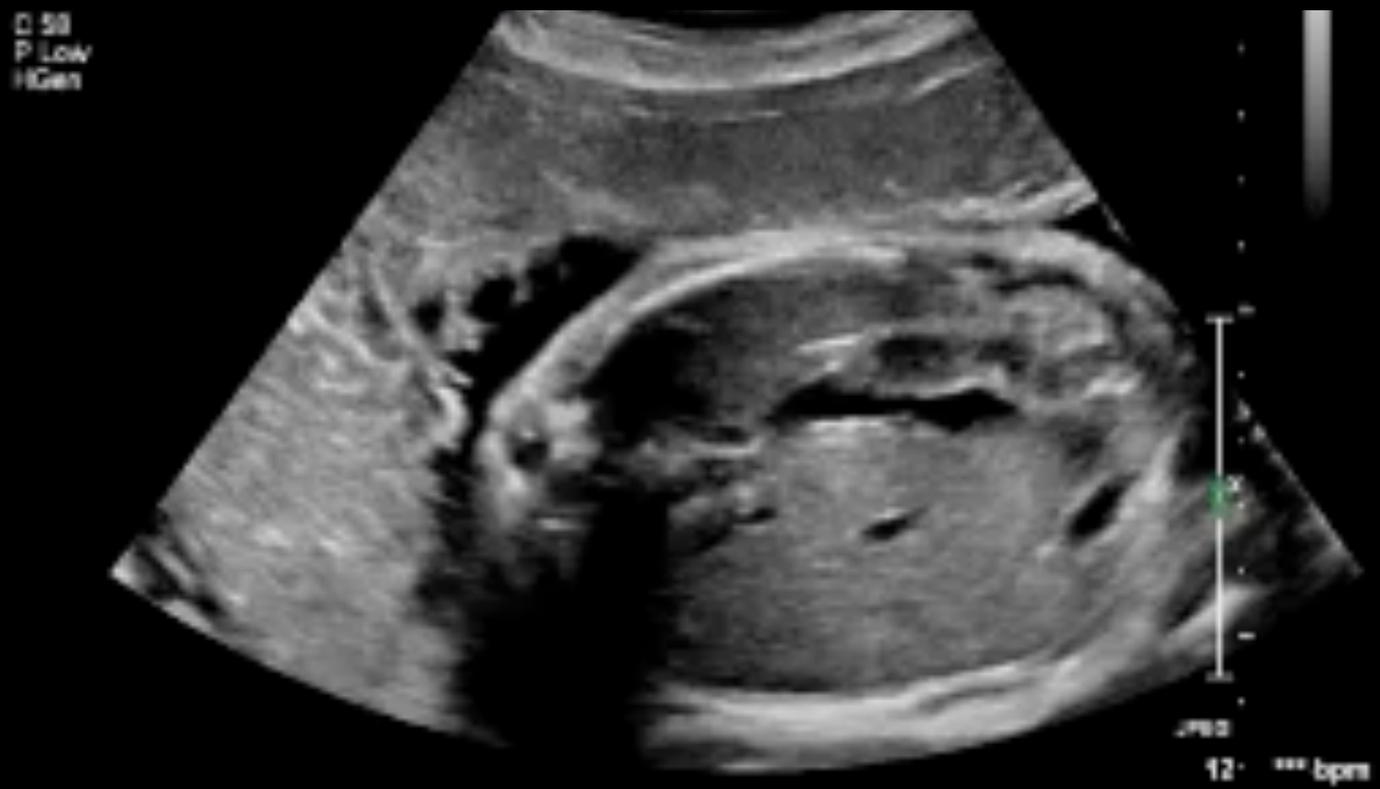


ECTOPIC KIDNEY





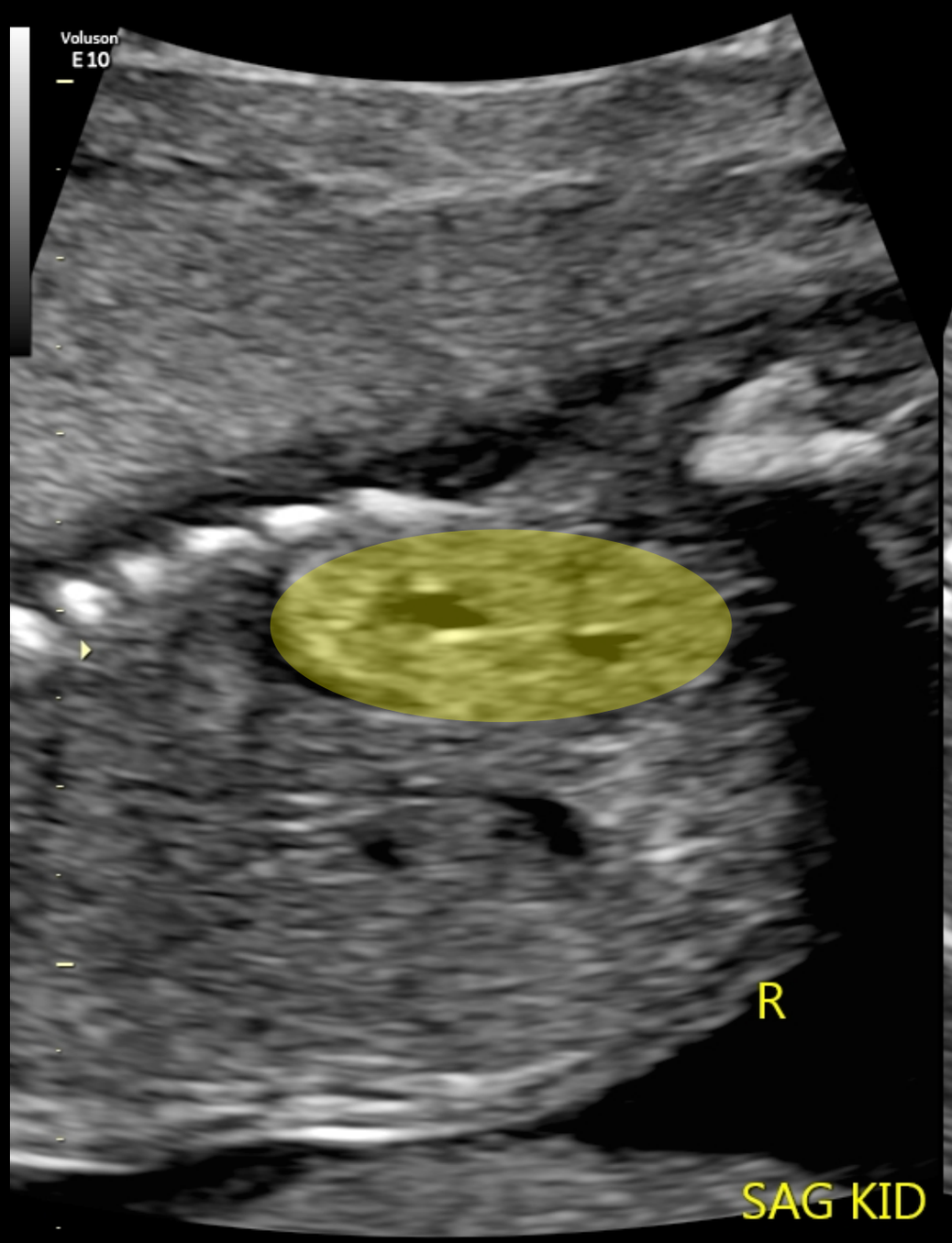
ECTOPIC KIDNEY FOLLOW UP

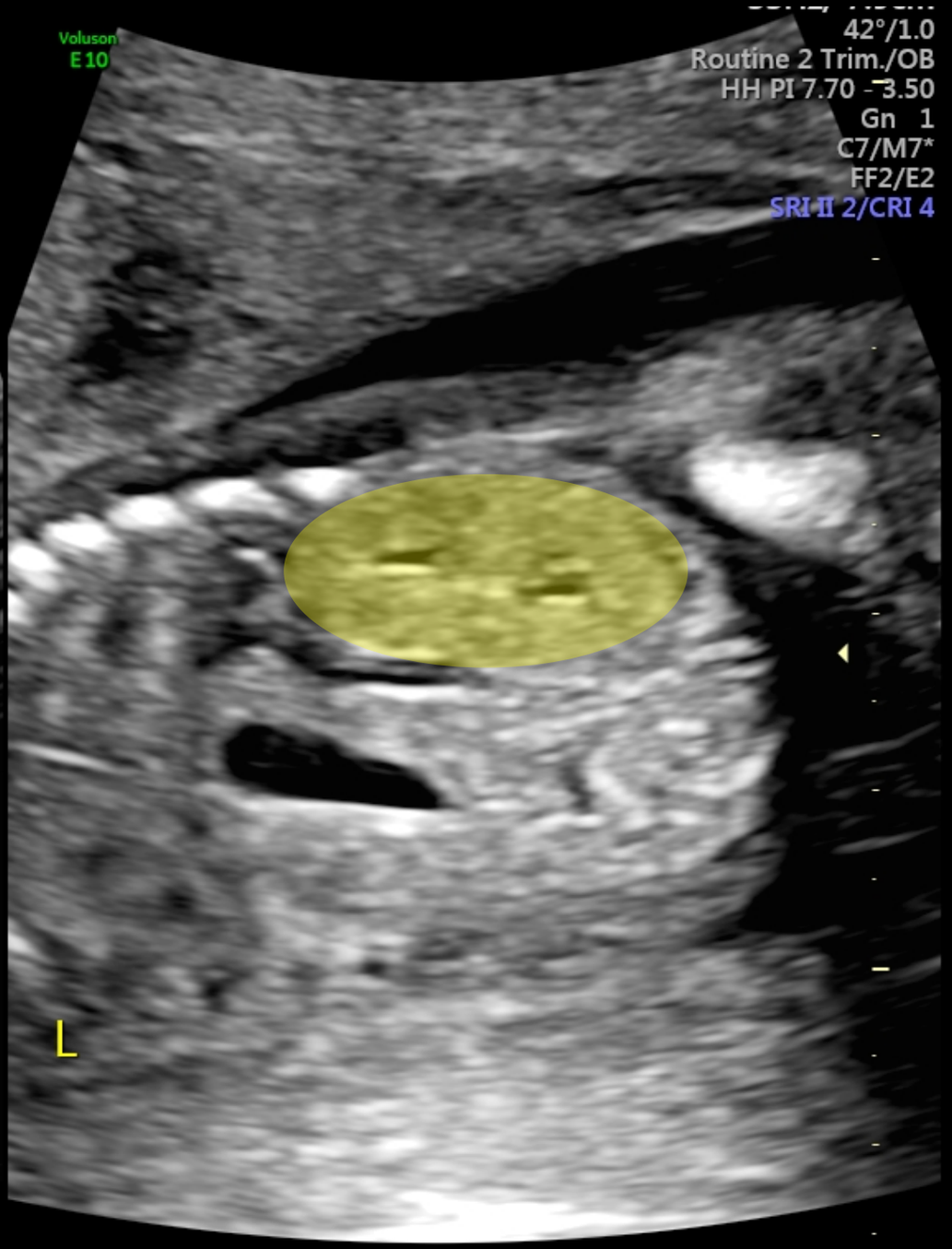


DUPLEX KIDNEY

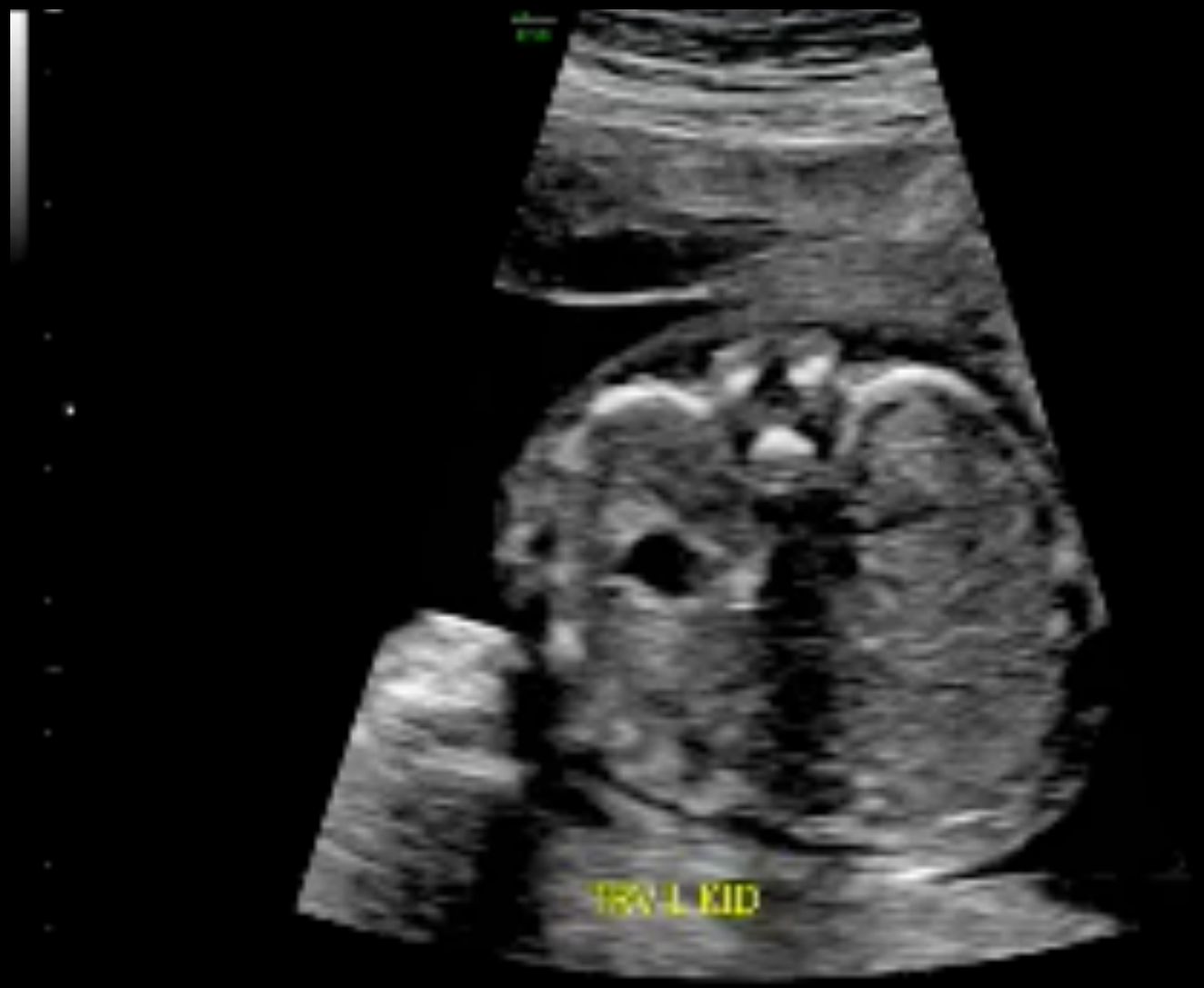
- Two ureteral buds give rise to duplex kidney
- Two pelvicaliceal systems
- Partial or complete ureteral duplication
- Upper and lower pole moieties in same plane
- Obstruction upper pole moiety + ureterocele common
- Reflux common lower pole moiety

DUPLEX COLLECTING SYSTEM





DUPLEX COLLECTING SYSTEM



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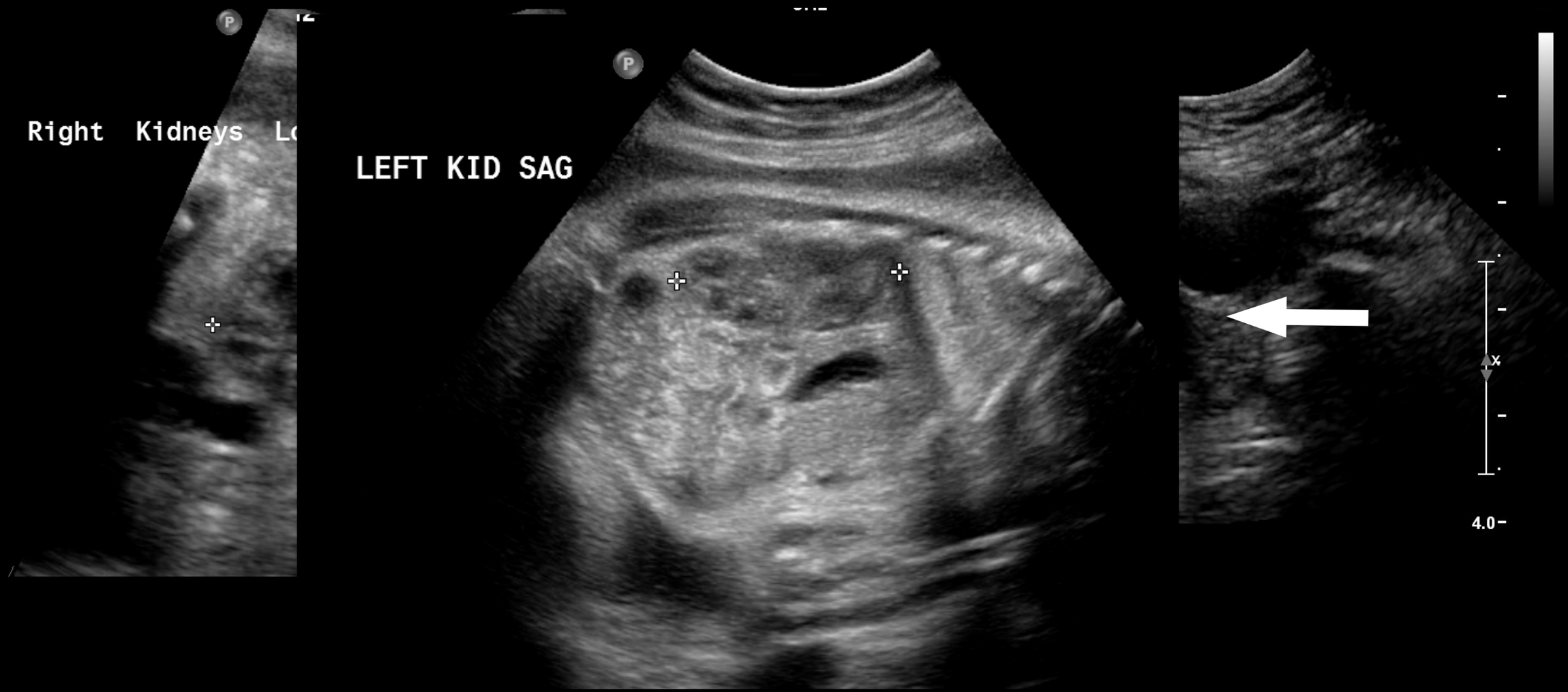


CYSTIC RENAL DISEASES

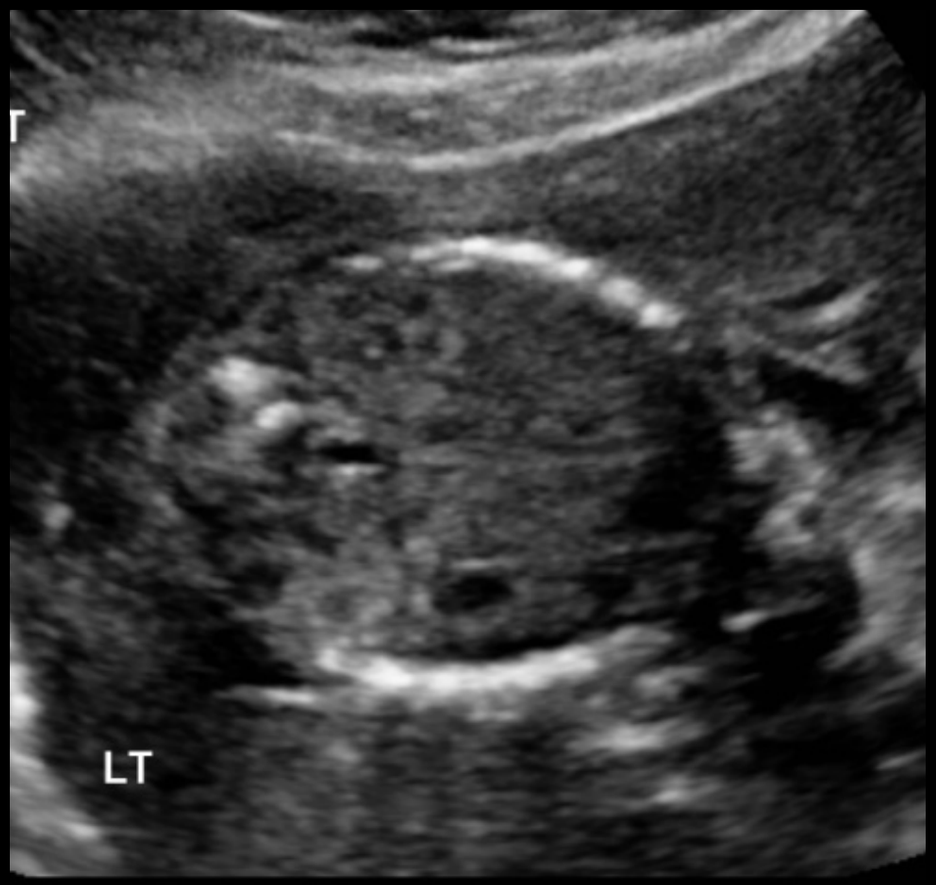
CLASSIFICATION OF CYSTIC RENAL DISEASES

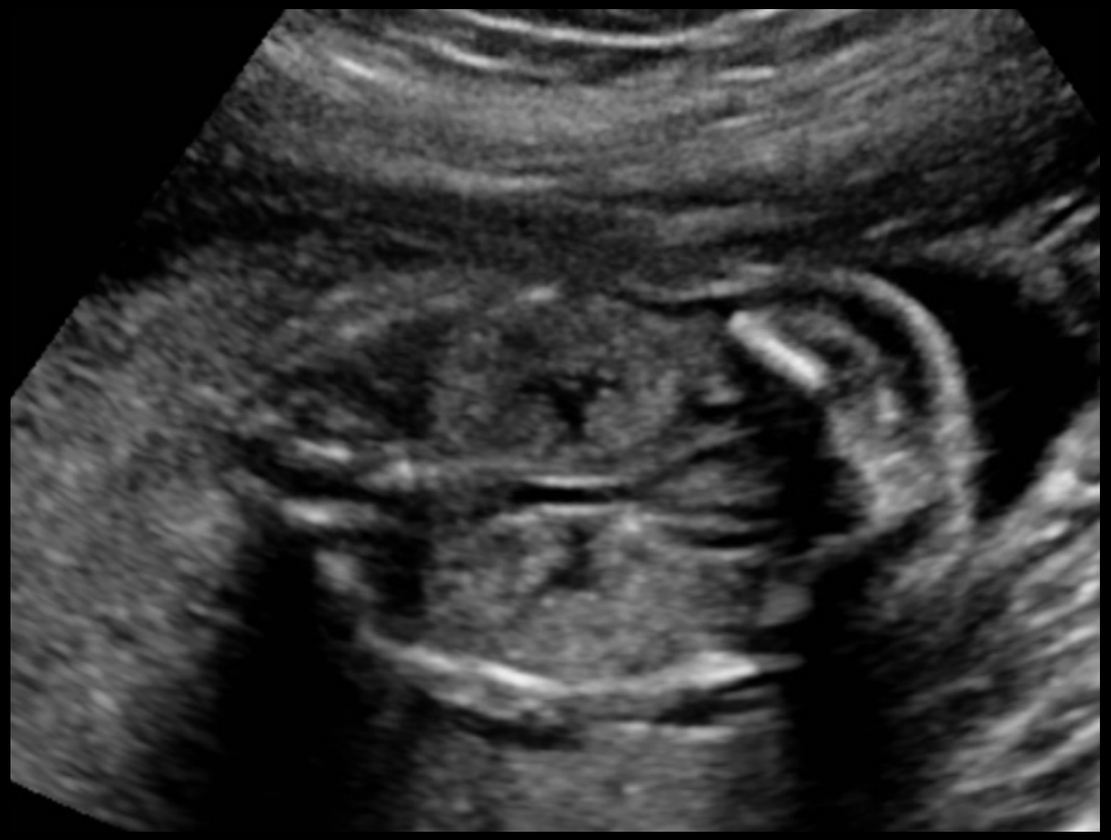
- Hereditary
  - ARPCKD
  - ADPCKD
    - Glomerulocystic disease
    - Medullary cystic dysplasia
- Non-Hereditary
  - MCDK
  - Obstructive dysplasia

ARPCKD

- 1 in 20,000 live births
- Enlarged hyperechoic smooth kidneys
- Evolution of cystic changes through third trimester
- Cortex is spared - hypoechoic rim sign
- Reversal of CMD can be seen
- Low fluid
- Recurrence risk: 25%

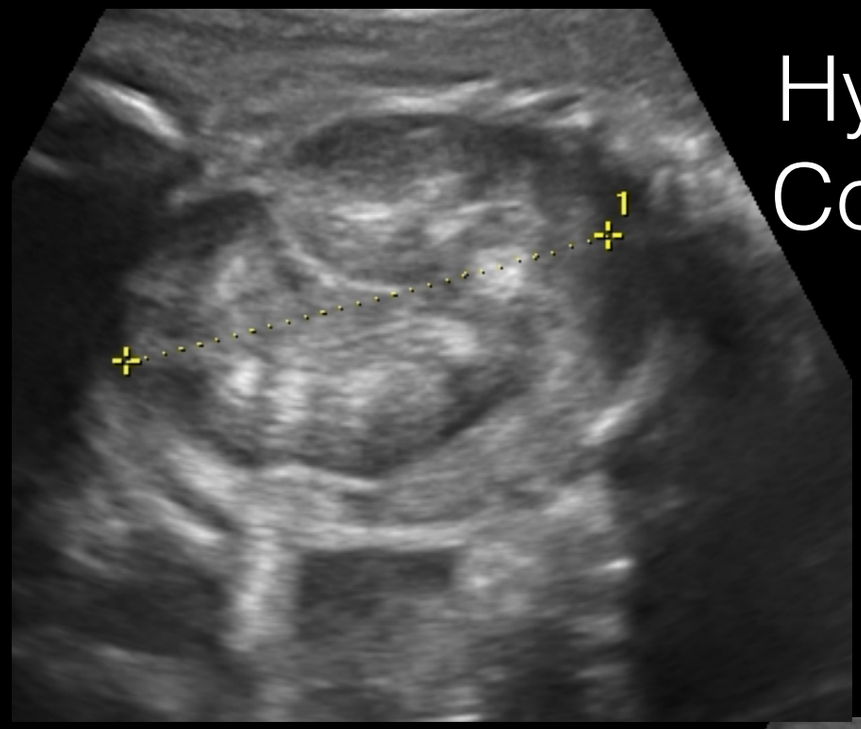
ARPCKD



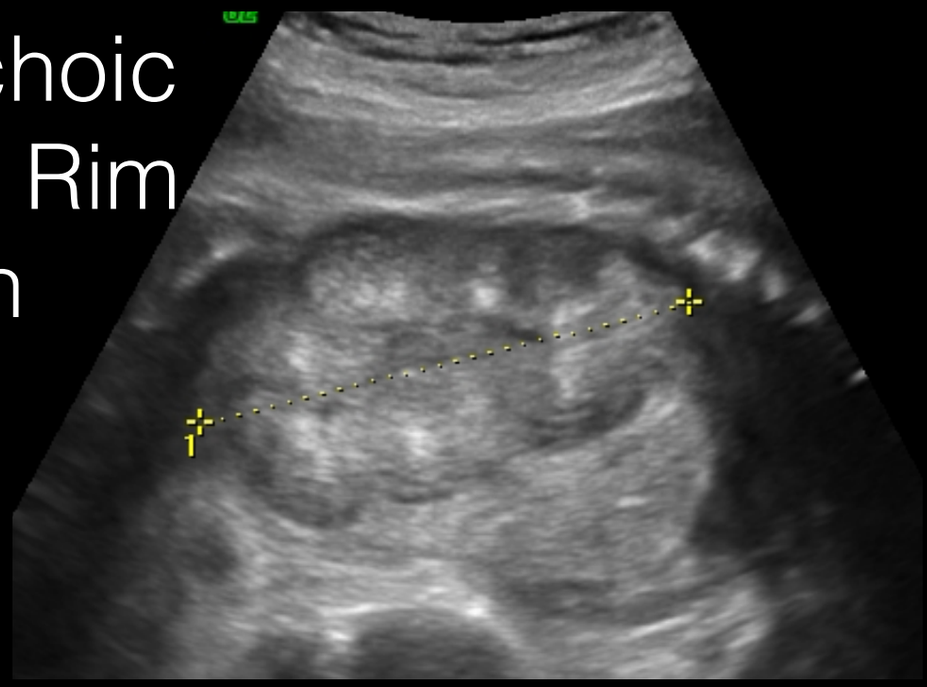


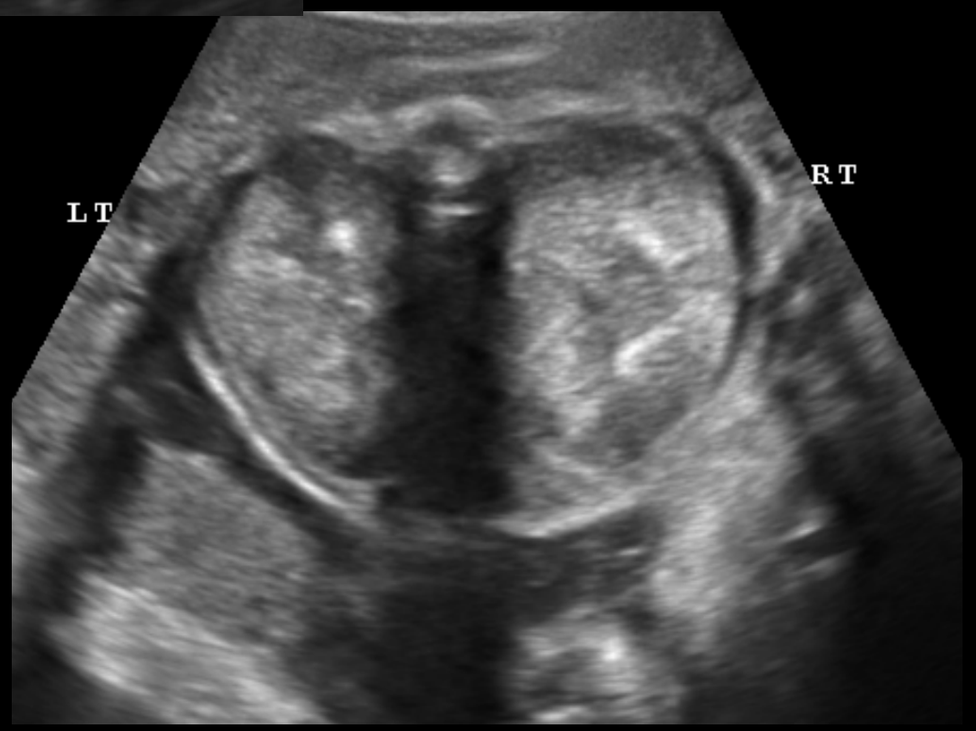
21 week anatomy scan
Enlarged kidneys: 26mm and 28 mm

ARPCKD



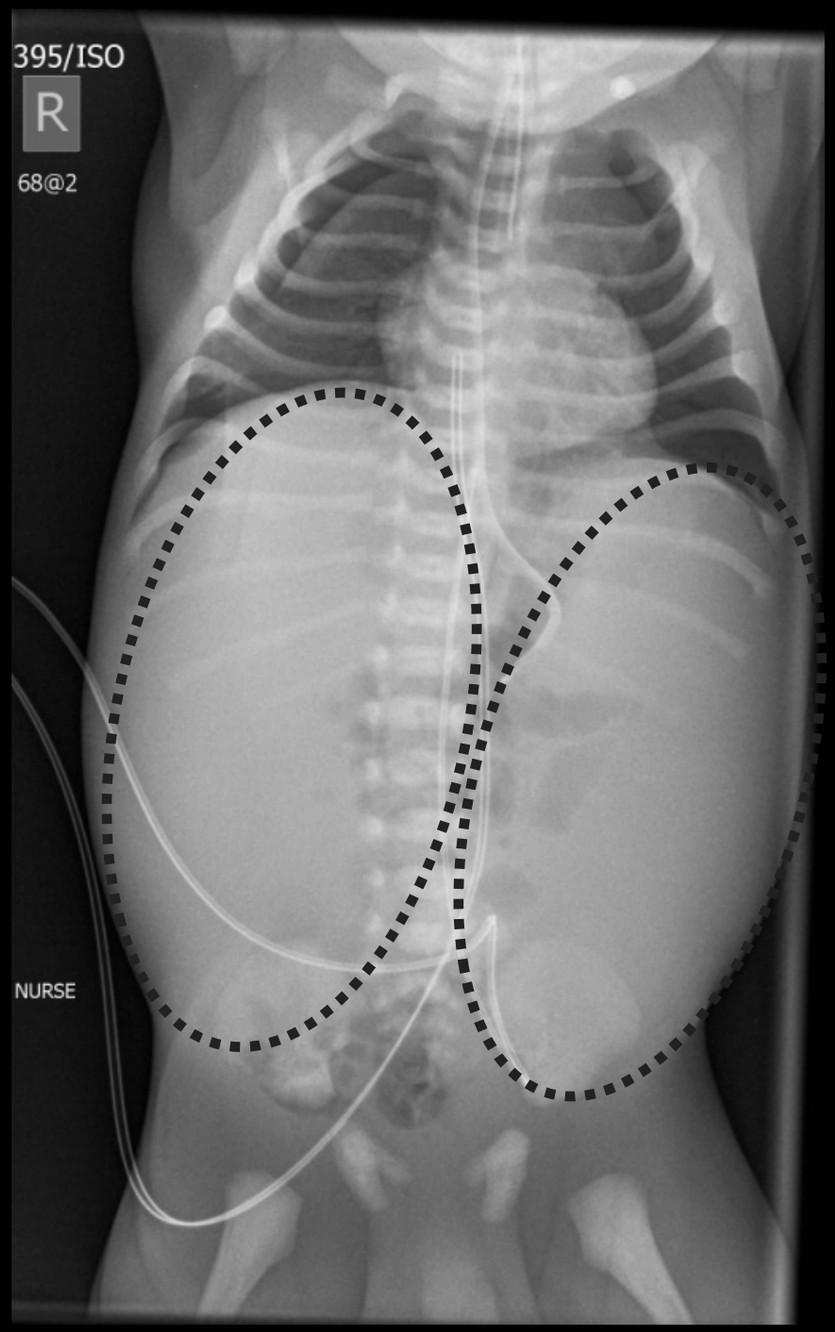
Hypoechoic Cortical Rim Sign

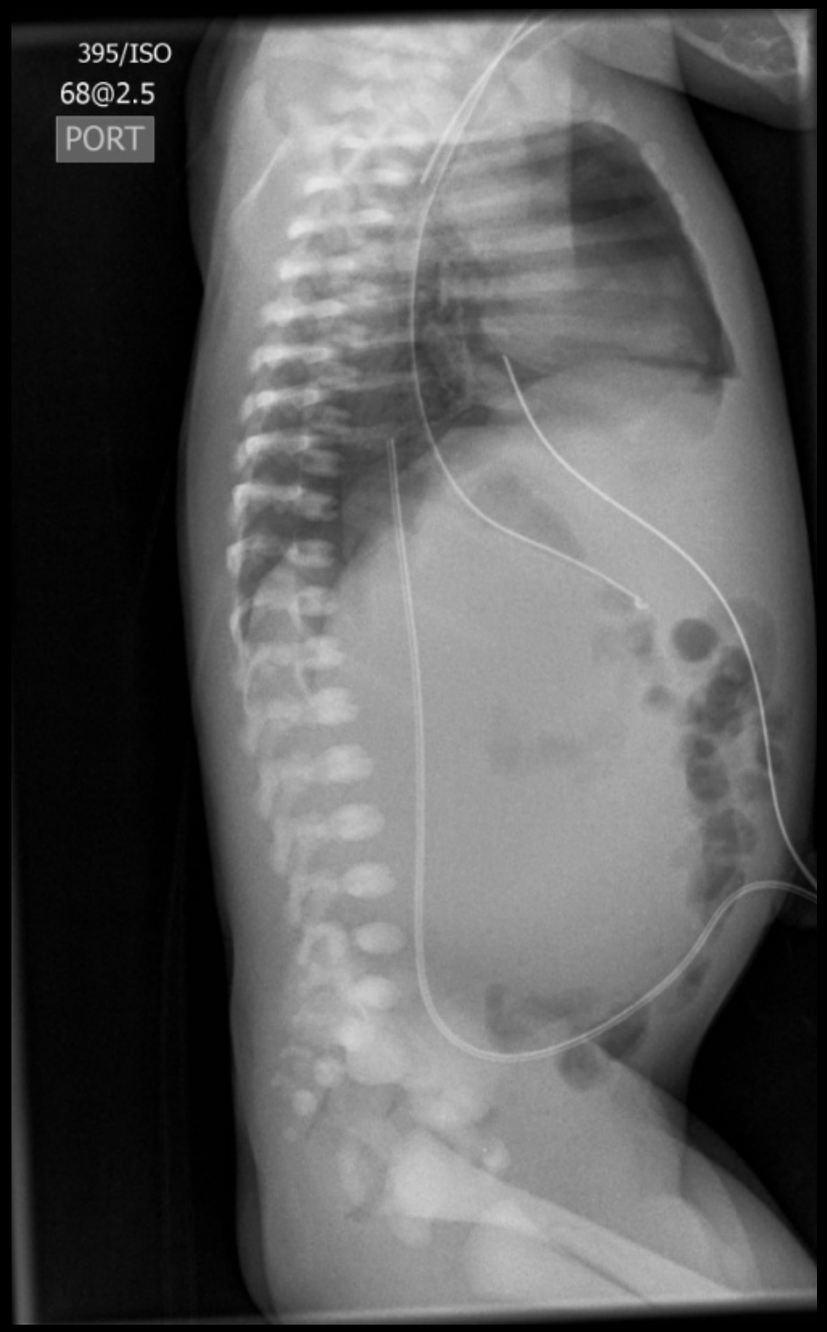




31 weeks: Bilateral enlarged kidneys

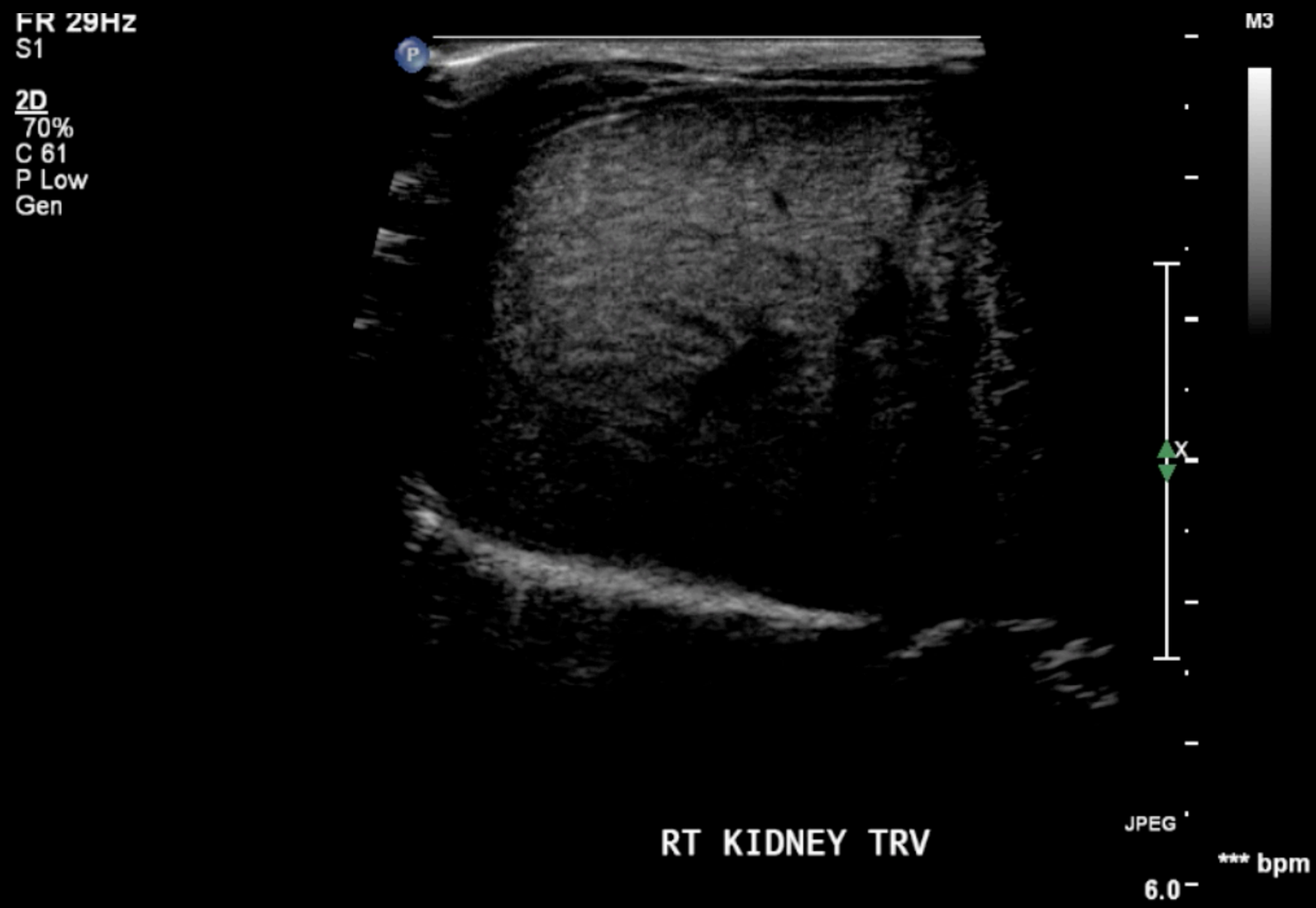
ARPCKD





ARPCKD

Postnatal Images

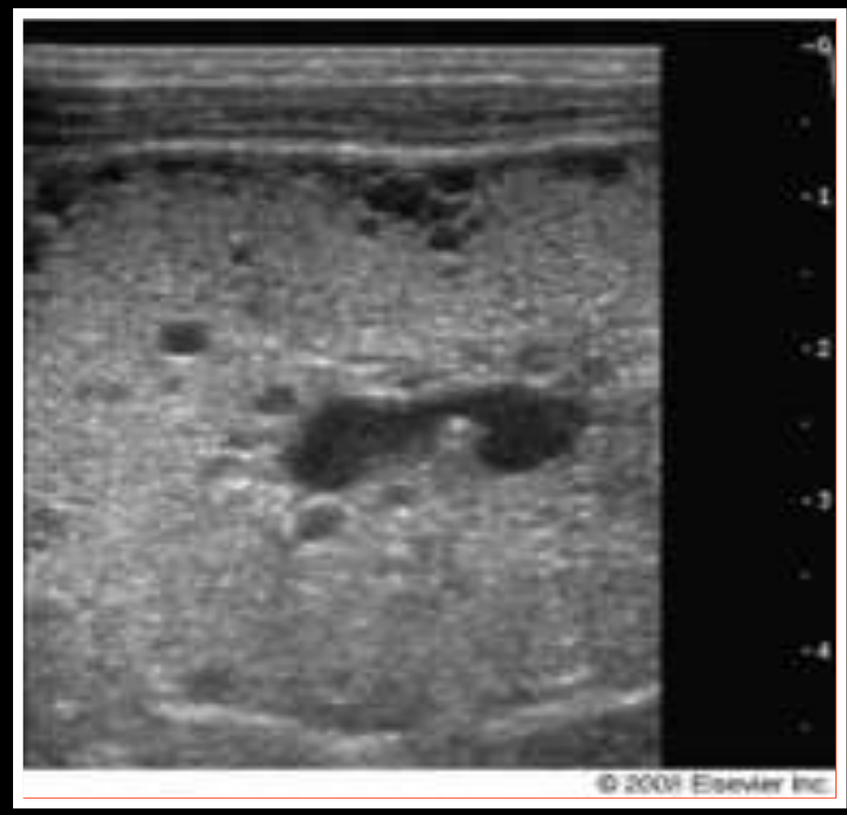


ADPCKD

- 1 in 800 live births
- Moderately enlarged kidneys with hyperechoic cortices
- Usually normal AFV
- Cysts usually don’t develop until adulthood
- Association: duplex, MCDK, UPJ obstruction
- Ask about family history, scan parents

GLOMERULOCYSTIC KIDNEYS

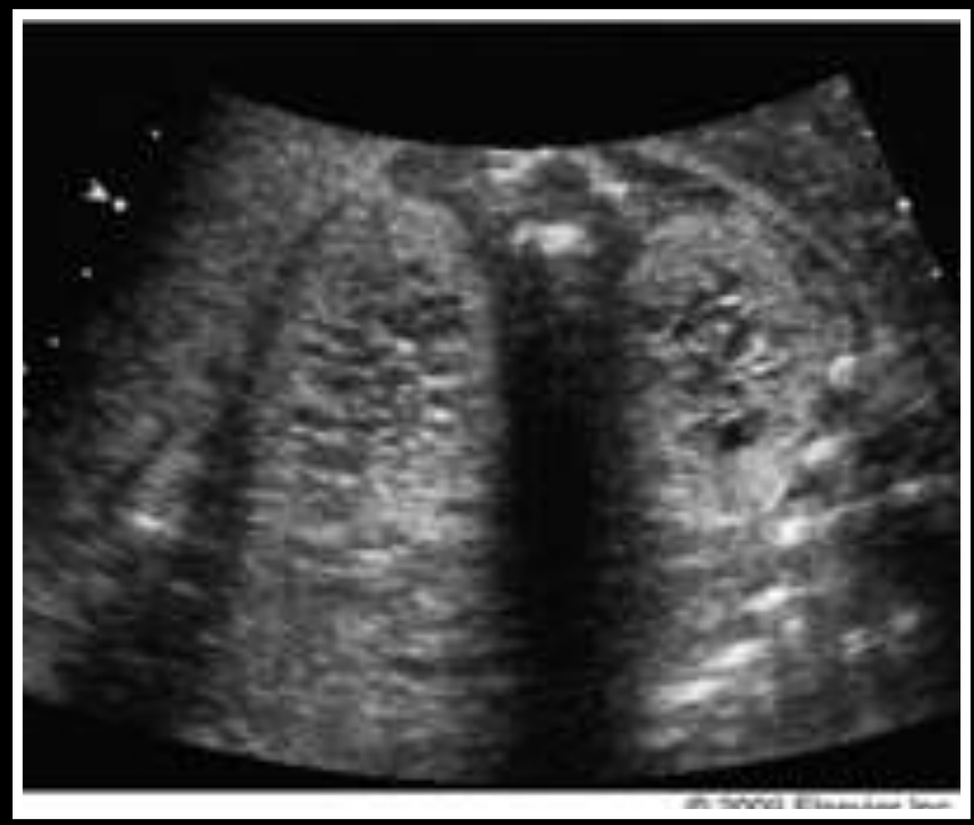
- Cysts corresponding to dilated Bowman spaces
- Nonspecific origin
  - Sporadic
  - Familial
  - Syndromes (TS, Tri 13, Zellwegger, Short rib polydactyl, Orofaciodigital syndrome type 1)
- Classic appearance is enlarged kidney with subcapsular cortical cysts



Callen 2013

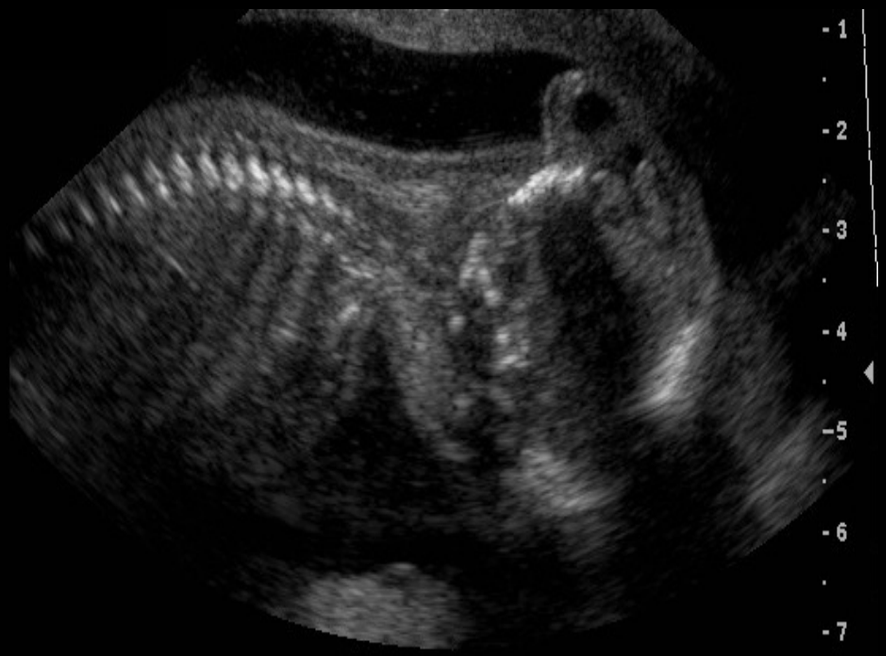
MEDULLARY CYSTIC DYSPLASIA

- Cysts involve medullary tubules
- Associated with many syndromes including Meckel-Gruber
- Present early with enlarged kidney, pseudo CMD, prominent medulla

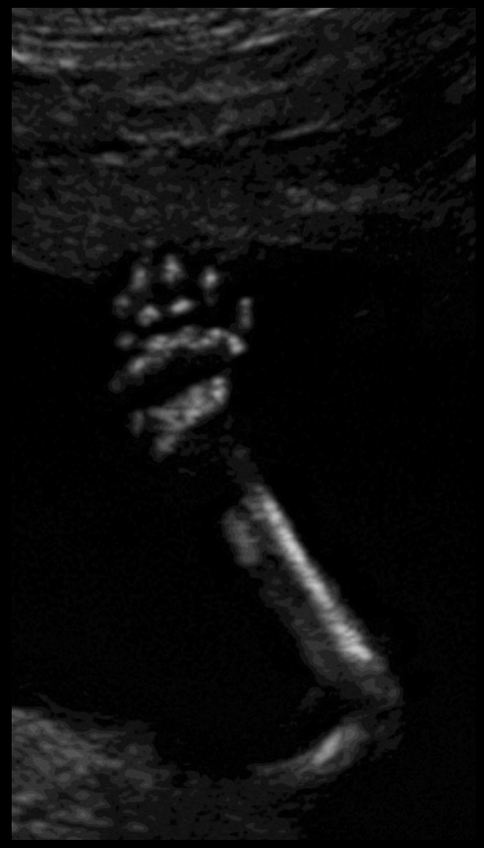


Callen 2013

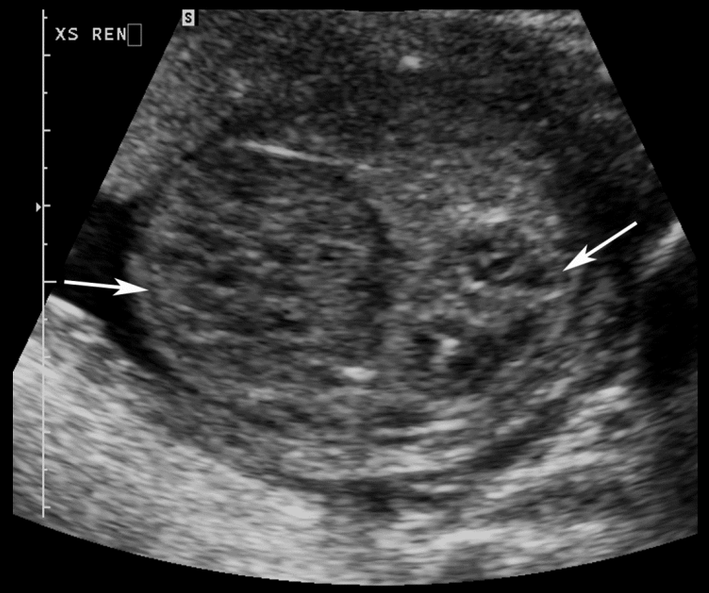
MECKEL GRUBER SYNDROME



Encephalocele



Polydactyly

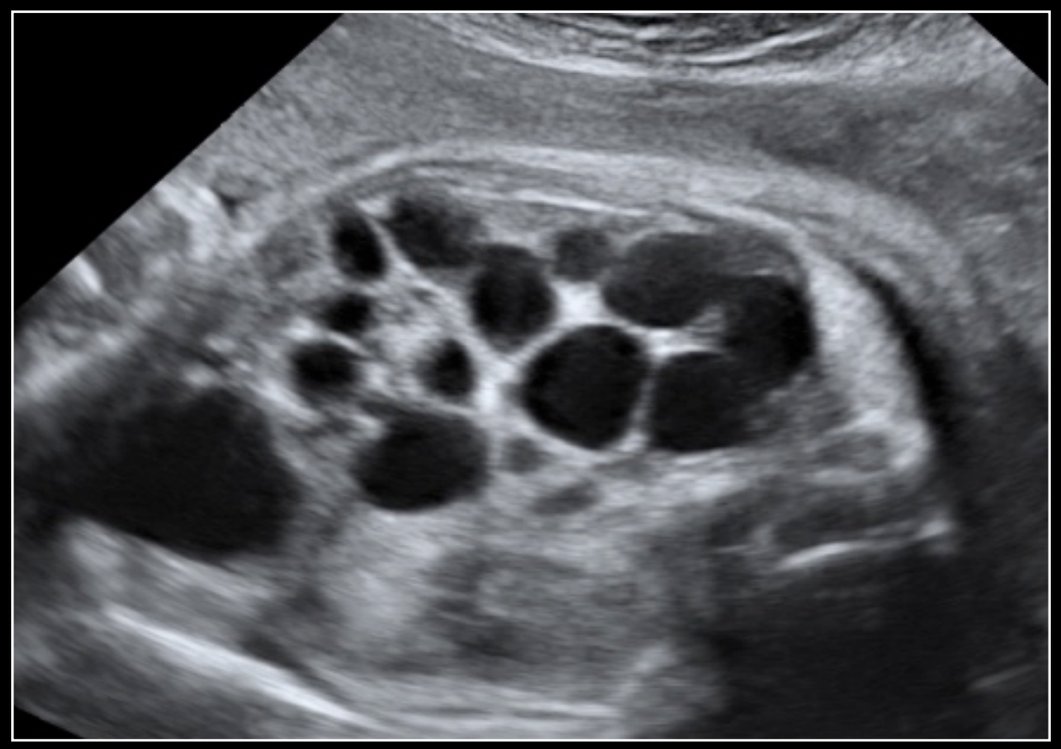


Kidney Cysts

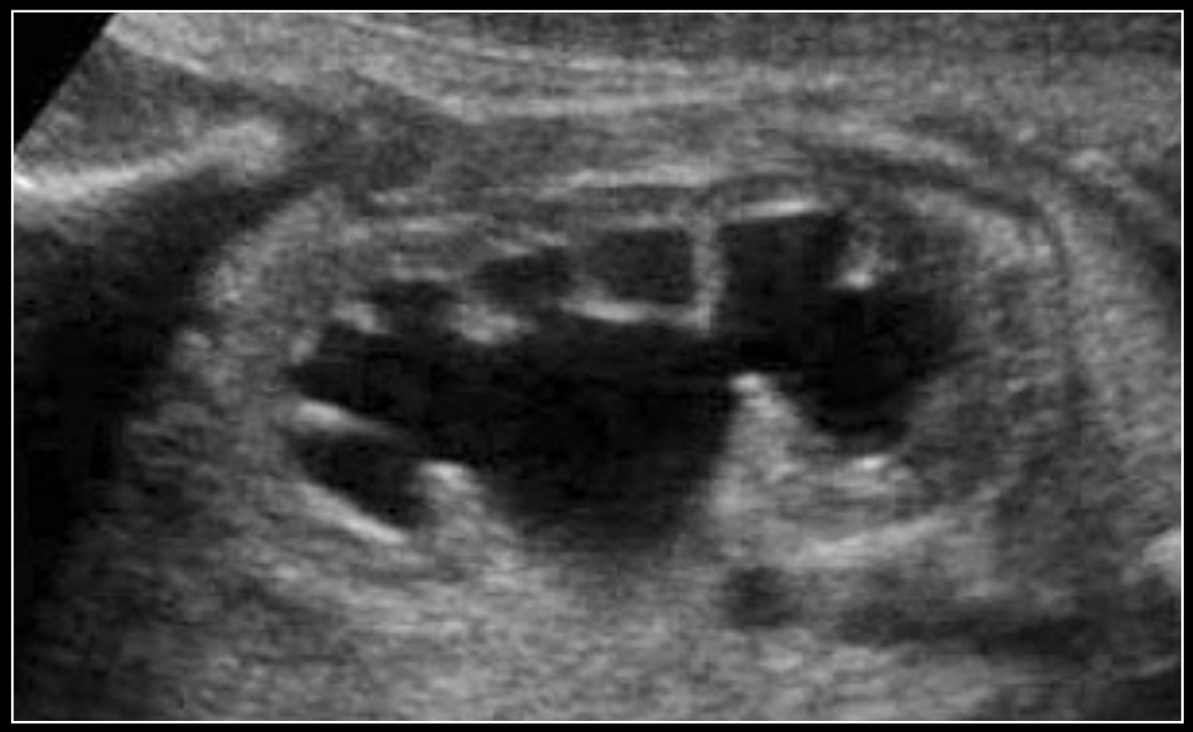
MULTICYSTIC DYSPLASTIC KIDNEY

- Unilateral noncommunicating cyst of varying size/shape
- Irregular contours
- No normal tissue between cysts, no collecting system
- Can occur in upper moiety of duplex
- May involute during pregnancy or after birth
- Association: UPJ, VUR, agenesis
- Unilateral or bilateral (up to 25%)

MCDK VS HYDRONEPHROSIS

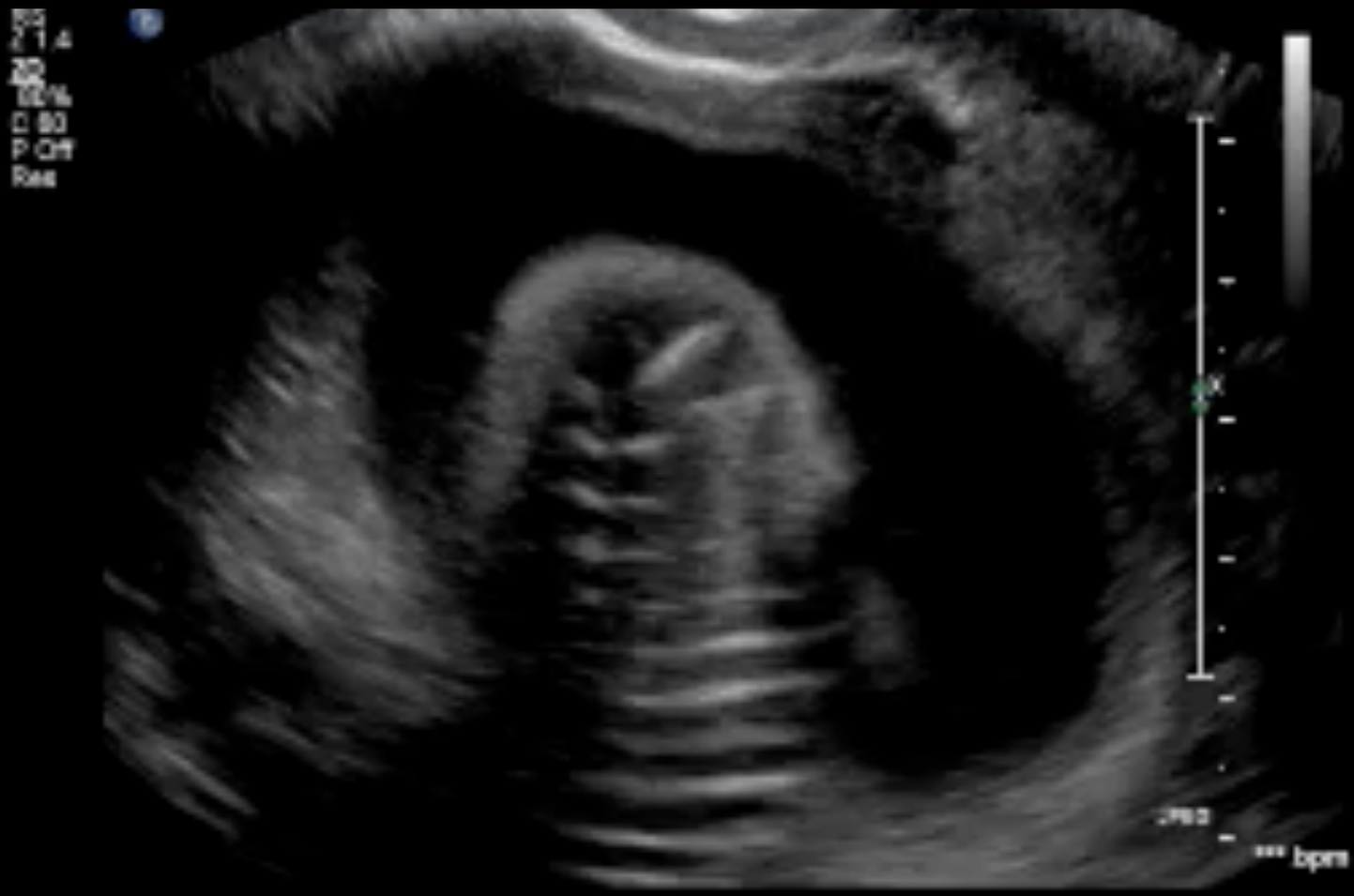


Irregular contour
Cysts vary in size/shape
Noncommunicating
Dysplastic echogenic tissue

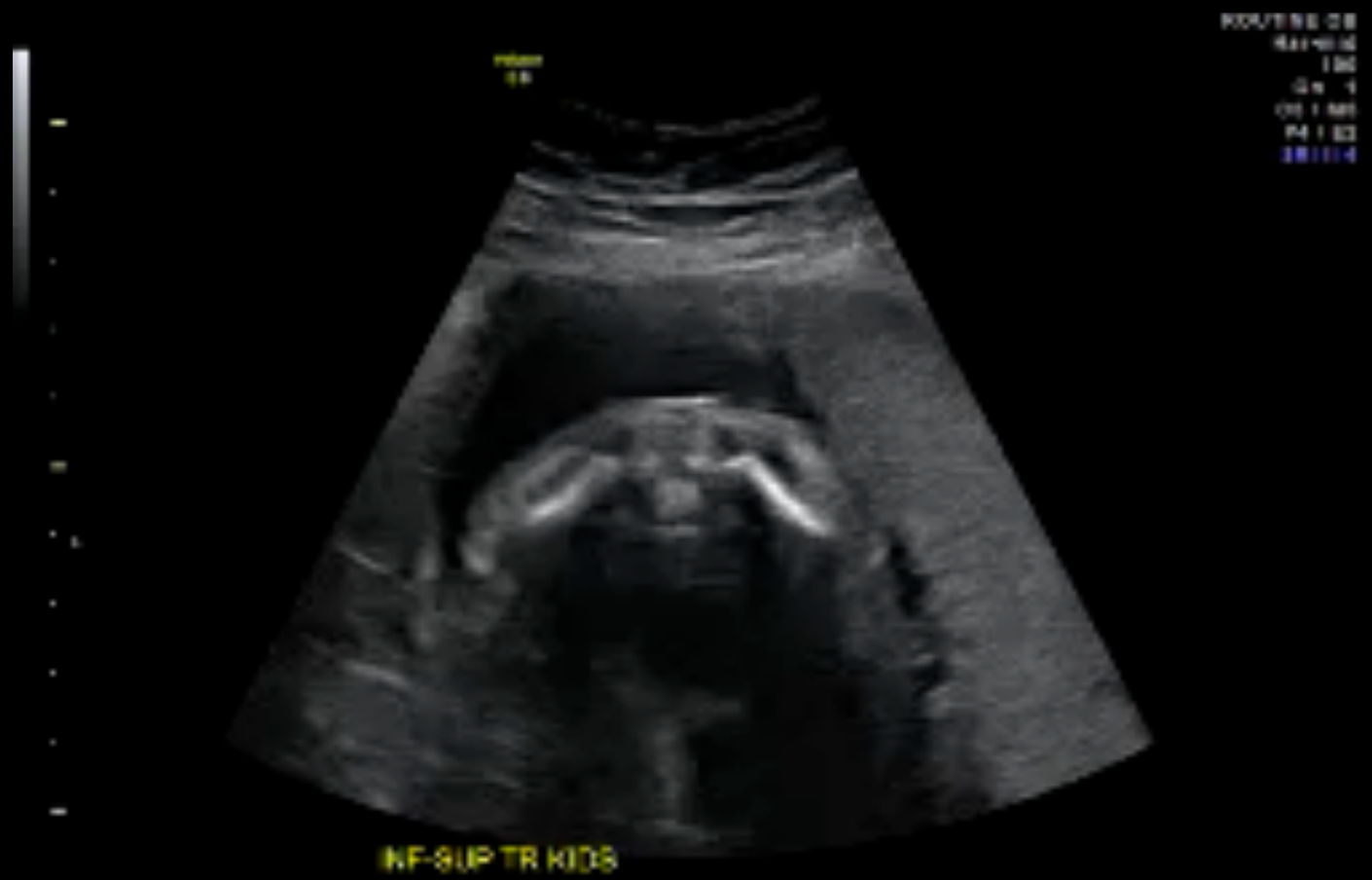


Smooth contour
Communicating ‘cysts’
Rim of renal tissue

QUERY EARLY MCDK



FOLLOW UP: UNILATERAL MCDK



OBSTRUCTIVE CYSTIC RENAL DYSPLASIA

- Unilateral (UPJ or UVJ)
- Bilateral (bladder outlet obstruction)
- Variable appearance
- Usually cysts are subcapsular

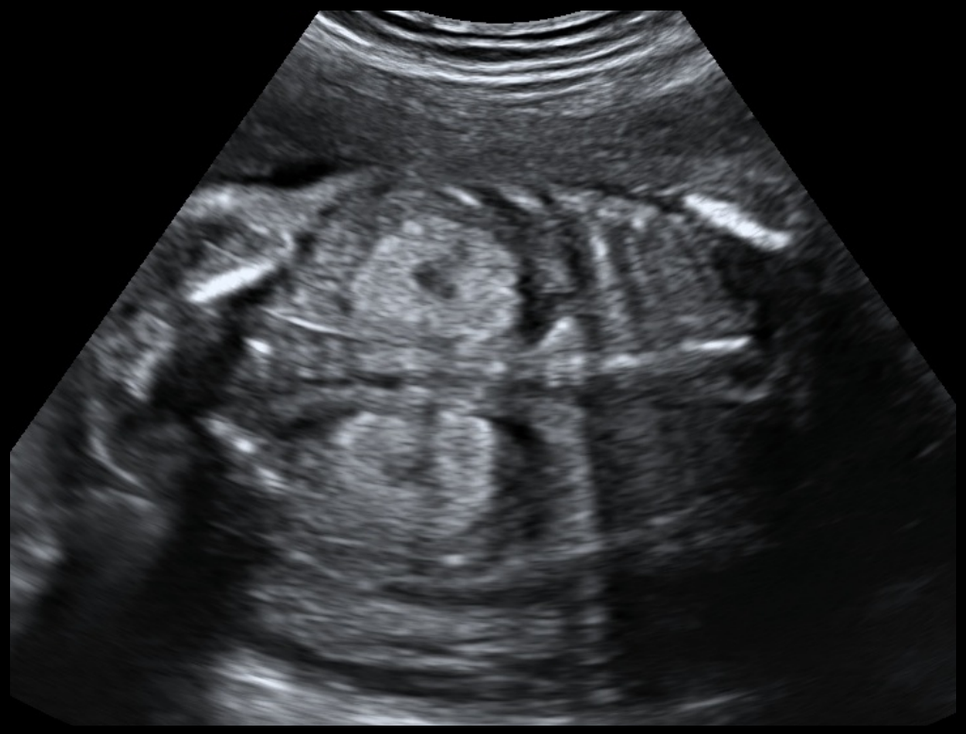
ECHOGENIC KIDNEYS

ECHOGENIC KIDNEYS

- Recall: Early in pregnancy normal kidney is echogenic
- Finding not a diagnosis
- Differential considerations:
  - PCKD (AR and AD)
  - Syndomes: Tri 13, Meckel Gruber, Beckwith Wiedemann, Perlman, Zellwegger, Bardet-Biedl
  - Renal vein thrombosis (risks: maternal diabetes,TTTS)
  - Infection: CMV
  - Congenital nephrotic syndrome (rare**) - assoc. thick placenta
  - May be normal

ECHOGENIC KIDNEYS

- COMMON:
  - Physiologic variation
  - Obstructive
  - ARPCKD or ADPCKD



- UNCOMMON:
  - Meckel-Gruber
  - Bardet-Biedl
  - Aneuploidy
  - Infection
  - RVT
  - Congenital Nephrotic Syndrome
  - Overgrowth Syndromes
    - Beckwith-Wiedeman
  - Vater syndrome

ECHOGENIC KIDNEYS

- Prognosis defined by etiology
- Assess for:
  - Unilateral vs bilateral
  - Oligohydramnios
  - Obstruction
  - Parenchymal changes
    - CMD?
    - Cysts?

ECHOGENIC KIDNEYS

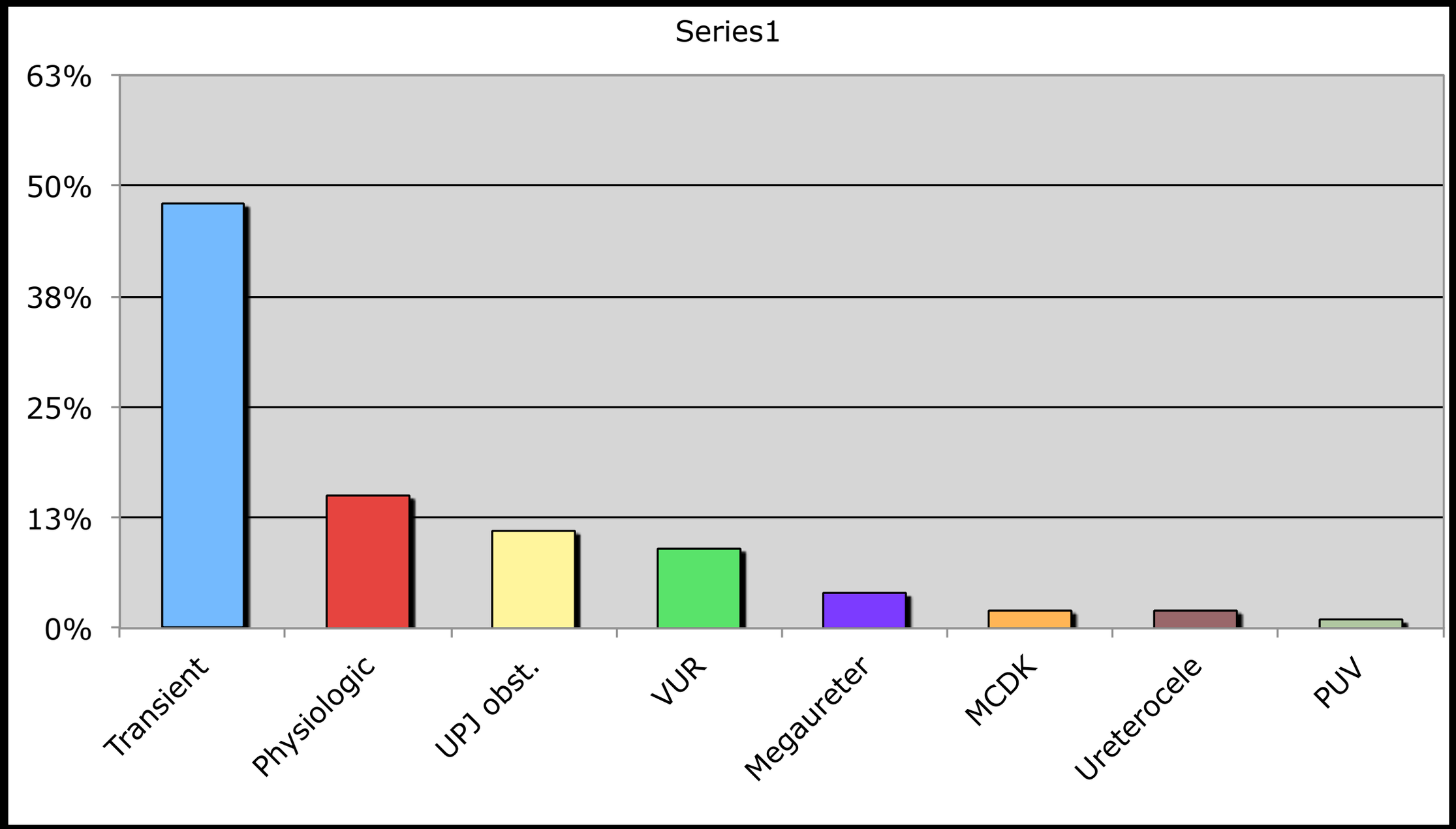
Hyperechoic Kidneys	Corticomedullary Differentiation
Beckwith Weideman, Perlman, Zellwegger, Infection, Intoxication, MCD, maternal, normal variant	Present
Obstructive dysplasia, ARPCKD	Absent
ADPCKD, Diabetes, Bardet Biedl, RVT, Meckel Gruber, Chromosomal, Nephrotic	Variable
Some ARPCKD, metabolic	Reversed

URINARY TRACT DILATATION

UPPER URINARY TRACT DILATATION

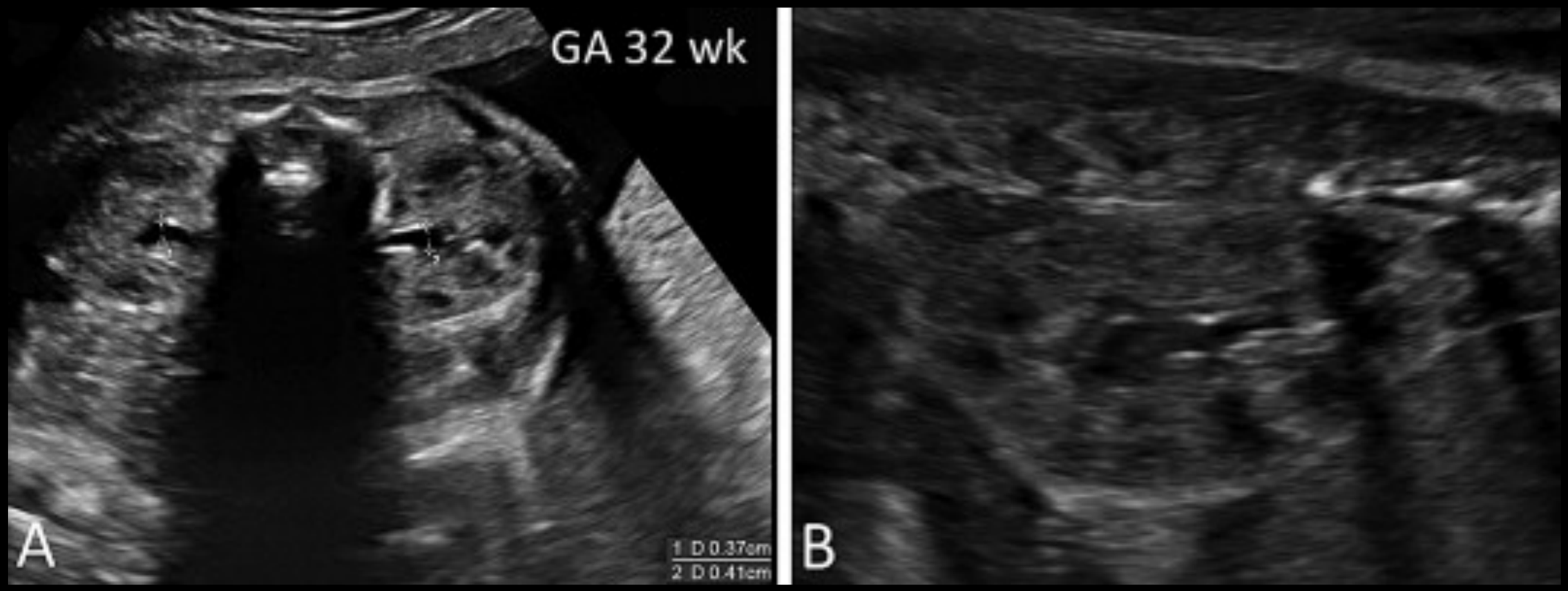
- Upper tract dilatation most common abnormality 1-5%
- Measure AP diameter renal pelvis (APD) in transverse plane
- Hydronephrosis: dilatation of pelvis and calyces
- Pyelectasis: dilatation of pelvis only
- Various etiologies:
  - Transient/physiologic
  - VUR
  - Obstructive

UPPER URINARY TRACT ETIOLOGY



HOW TO MEASURE

- Transverse plane measure AP diameter of renal pelvis:

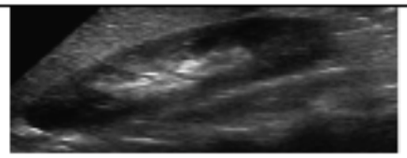
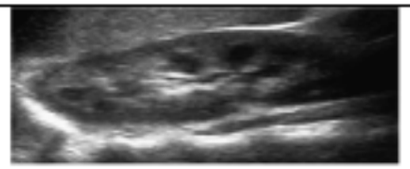
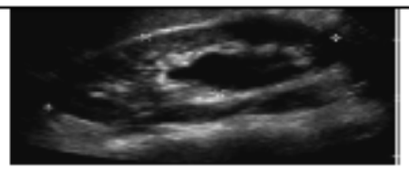
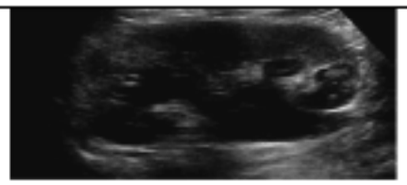
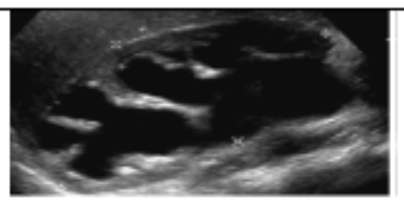


URINARY TRACT DILATATION THRESHOLD

- Langer et al 1996
  - 5 mm < 28 wks
  - 10 mm > 28 weeks
- Gringon et al 1986
  - Grade 1: APD 1 cm no caliectasis
  - Grade 2: APD 1-1.5 cm and no caliectasis
  - Grade 3: APD > 1.5 cm + slight caliectasis
  - Grade 4: APD > 1.5 cm + moderate caliectasis
  - Grade 5: APD > 1.5 cm + severe caliectasis + cortical atrophy

SFU GRADING

Table 1 Society of Fetal Urology grading system of congenital hydronephrosis

Grade	Central renal complex	Renal parenchymal thickness	Ultra sound scan
0	intact	Normal	
I	Slight splitting of pelvis	Normal	
II	Evident splitting of pelvis and calices	Normal	
III	Wide splitting of pelvis and calices	Normal	
IV	Further splitting of pelvis and calices	Reduced	

PYELECTASIS CUT-OFF VALUES

- SFU GRADING SECOND TRIMESTER:
  - Mild 4-7 mm
  - Moderate 7-10 mm
  - Severe > 10 mm
- SFU GRADING THIRD TRIMESTER:
  - Mild 7-9 mm
  - Moderate 9-15 mm
  - Severe > 15 mm
- Lee et al study
  - >15 mm 88% chance post natal pathology

SFU CONSENSUS STATEMENT

- AP diameter of the renal pelvis of > 4 mm in the 2nd trimester and > 7 mm in 3rd trimester warrants further follow up in prenatal period

- Follow up:
  - Repeat US in 3rd trimester
  - Ultrasound at birth and 1 month

PROGNOSTIC FACTORS

- 80% of mild (4-7 mm) spontaneously resolve
- Severity of dilatation correlated to higher risk of pathology EXCEPT for VUR
- Progression of dilatation through to third trimester worse prognosis
  - 12% with isolated 2nd trimester UT dilatation had postnatal pathology
  - 40% if observed in both 2nd and 3rd trimester
- Worrisome findings: oligohydramnios, abnormal cortex, early GA at diagnosis

PROGNOSIS AHN

- Lee et al, 2006
  - Meta-analysis
  - 36% have underlying uropathy
    - 12% mild < 9 mm T3
    - 45% moderate 9-15 mm T3
    - 88% > 15 mm T3

UTD CONSENSUS 2014

- Consensus Statement 2014
- ACR, AIUM, ASPN, SFU, SMFM, SPR, SRU
- Recommendations:
    - Use of term UT dilatation
    - Classify based on:
        - APD, Caliceal dilatation (major/minor), parenchymal thickness, parenchymal appearance, ureter, bladder
    - Cut offs:
        - 16-27 weeks < 4 mm
        - ≥ 28 weeks < 7 mm
        - Postnatal (> 48h) < 10 mm

POSTNATAL WORK UP

- Low Risk:
  - One additional ultrasound > 32 weeks
  - Post natal ultrasound at birth (>48 hr) and 1 month
  - Modify aneuploidy risk if needed
- High Risk:
  - Follow up ultrasound 4-6 weeks after presentation
  - Postnatal ultrasound at birth and 1 month
  - Specialist consultation

OUTCOME ISOLATED ANTENATAL HYDRO

- Sighu Pedatr Nephrol (2006)
  - HSC
  - 7 articles selected that used SFU grading system
    - SFU grade 1-2 (APD < 12 mm)
      - 98% resolve, stabilize or improve
      - Conclusion: minimal investigation require
    - SFU grade 3-4 (APD > 12 mm)
      - more variable outcome

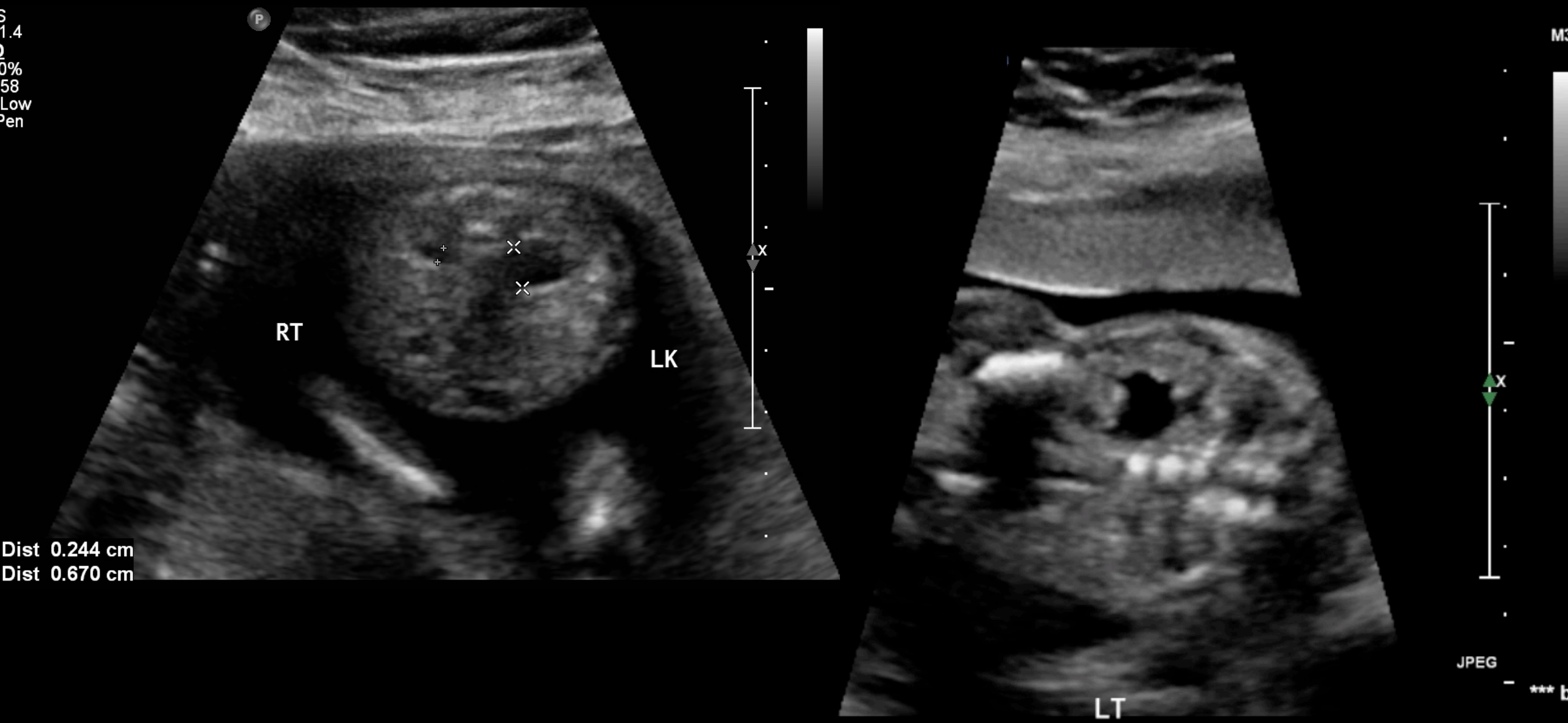
APD AS PREDICTOR FOR POSTNATAL SX

- Zhang et al, Pediatric Urology 2018
  - Systematic review and meta-analysis
  - 5 studies with total 1159 patients
  - Using 15 mm as cut off for APD:
    - Sensitivity 0.81
    - Specificity 0.78
  - Conclusion: APD >15 mm moderate diagnostic value predicting postnatal surgery

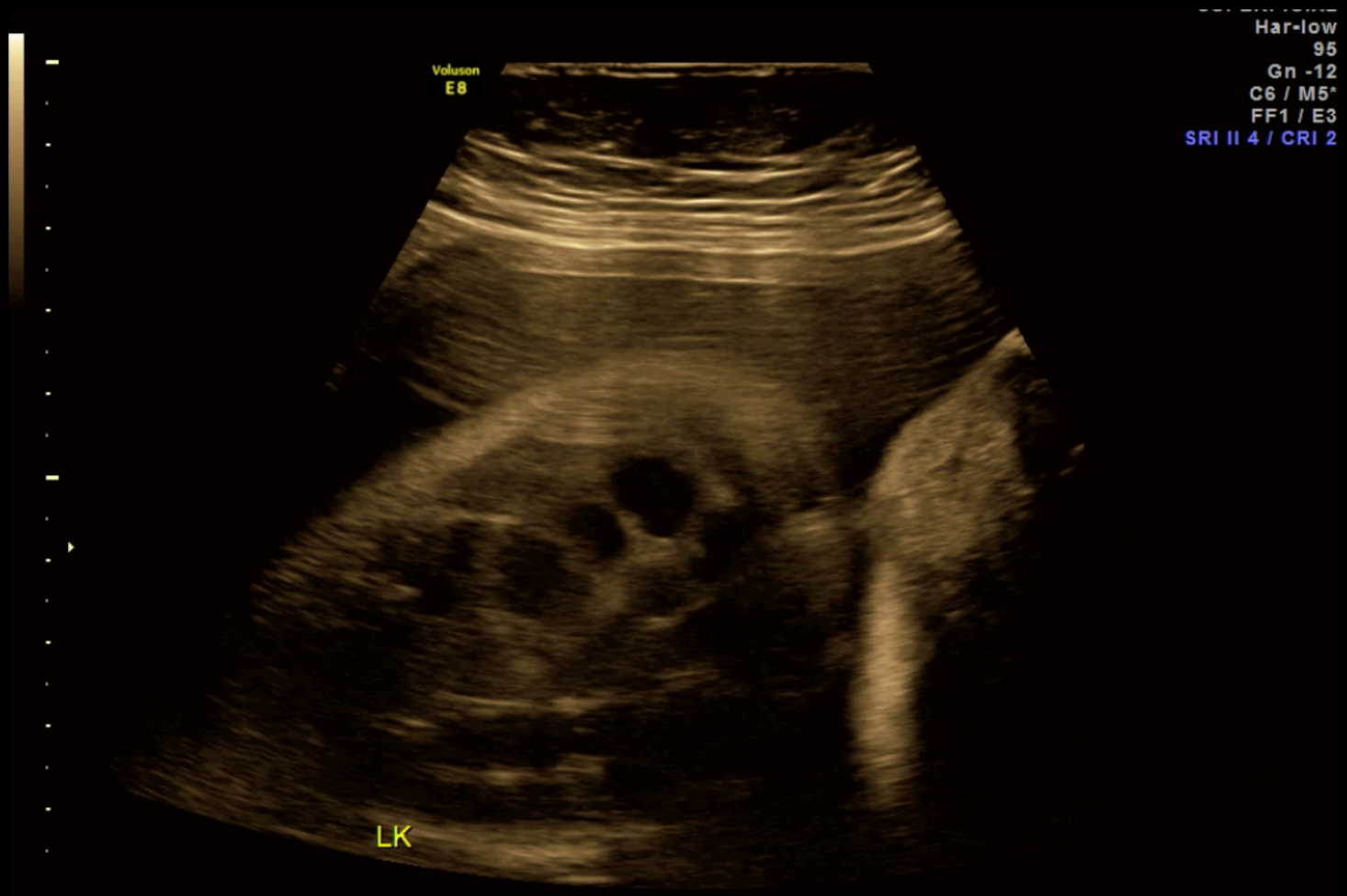
OUR CURRENT PRACTICE

- Isolated Antenatal Hydronephrosis:
  - 18-22 week anatomy APD > 4 mm, repeat at 32 weeks
  - If APD < 10 mm, no postnatal investigation
- For APD ≥ 10 mm:
  - refer to nephrology clinic for postnatal management
  - trimethoprim 2 mg/kg/day started at birth
  - postnatal ultrasound after 48h

ANTENATAL HYDRONEPHROSIS



ANTENATAL HYDRONEPHROSIS

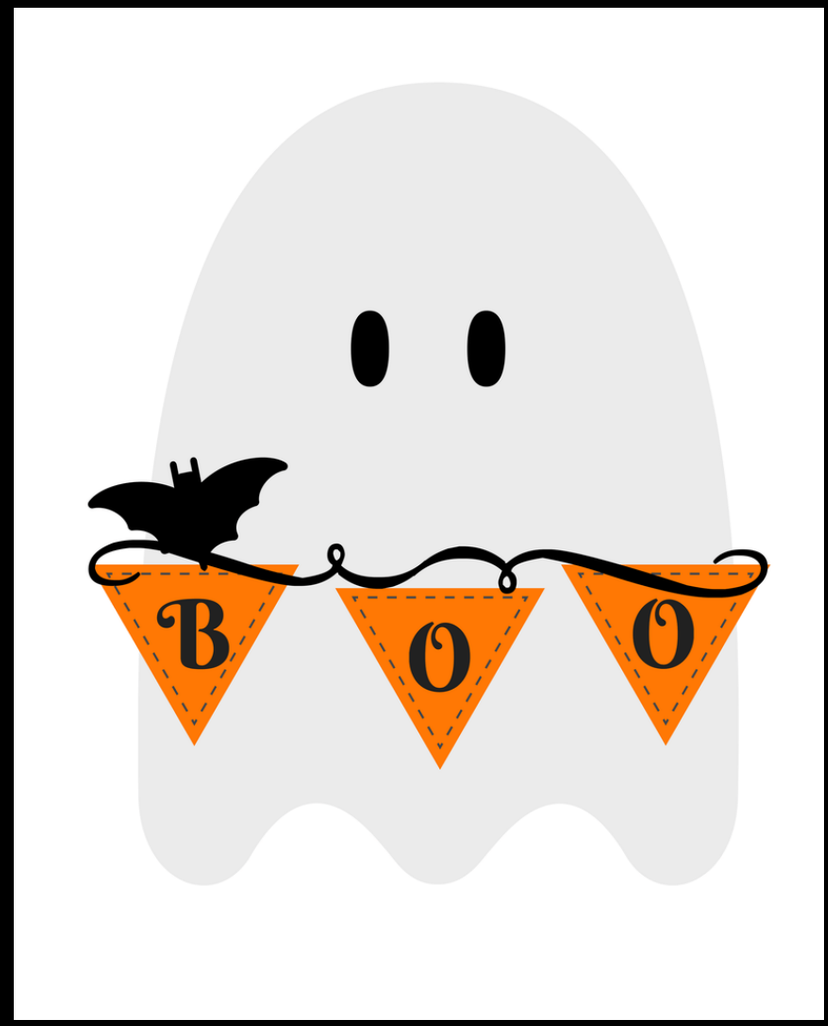


MEGAURETER

- Dilated ureter without bladder dilatation
- Usually result of UVJ obstruction
- May be associated with duplication of collecting system with ectopic ureterocele
- Increased risk of:
  - Unilateral agenesis
  - MCDK
  - Hirschsprung disease

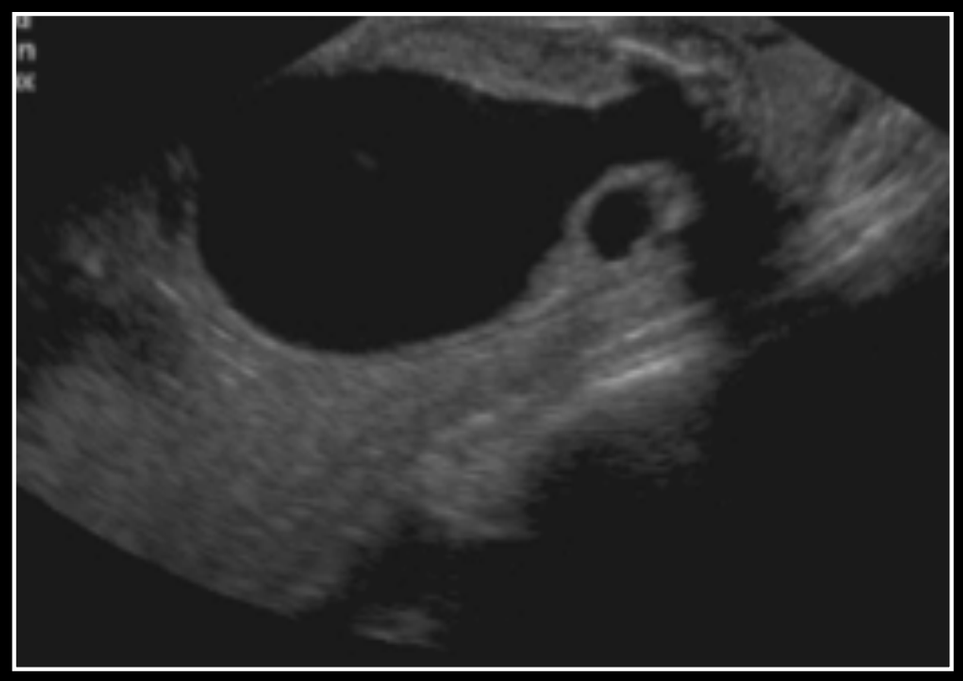
BLADDER OUTLET OBSTRUCTION (BOO)

- 1 in 3000 at 11-15 weeks
- Causes:
  - PUV
  - Urethral atresia/stricture
  - Prolapsed ureterocele
  - Prune Belly Sundrome
  - Megalourethra
  - Megacystic-Microcolon-intesntinal hpoperistalsis syndrome (MMIHS)

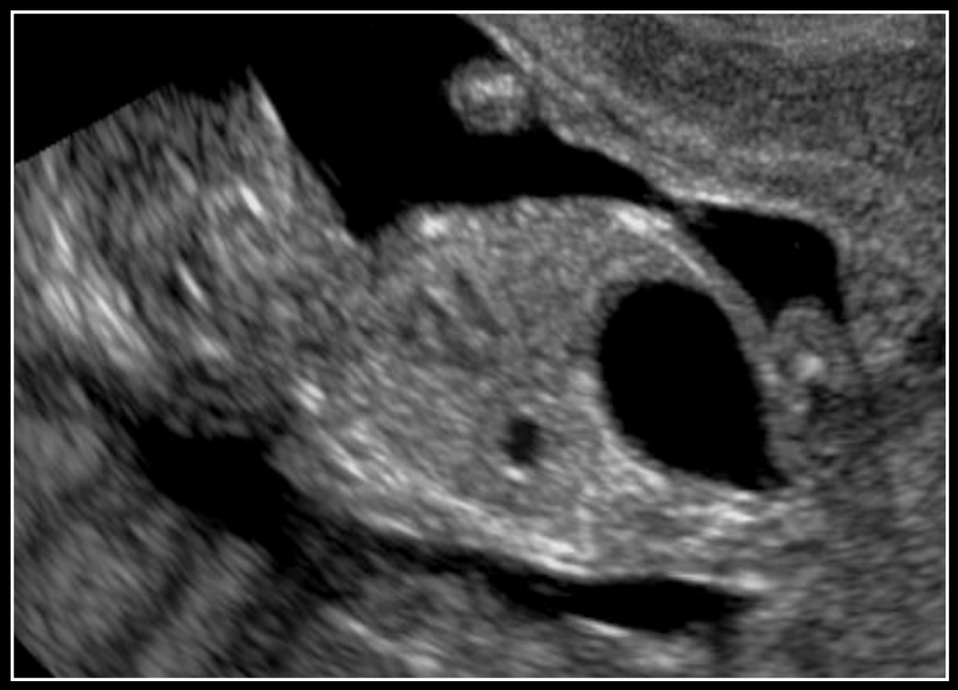


POSTERIOR URETHRAL VALVES

- Membranous folds in posterior urethra
- Most common of bladder outlet obstruction
- Predominantly male

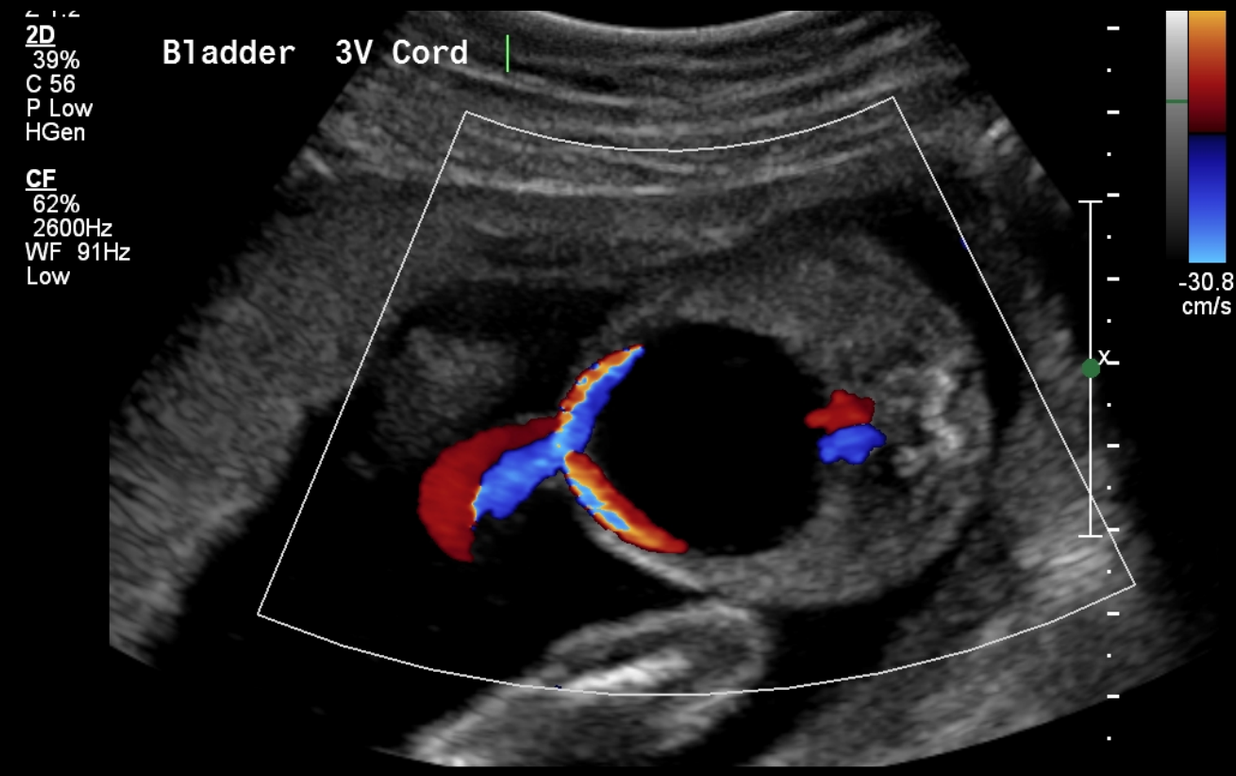


Keyhole Sign



MMIHS

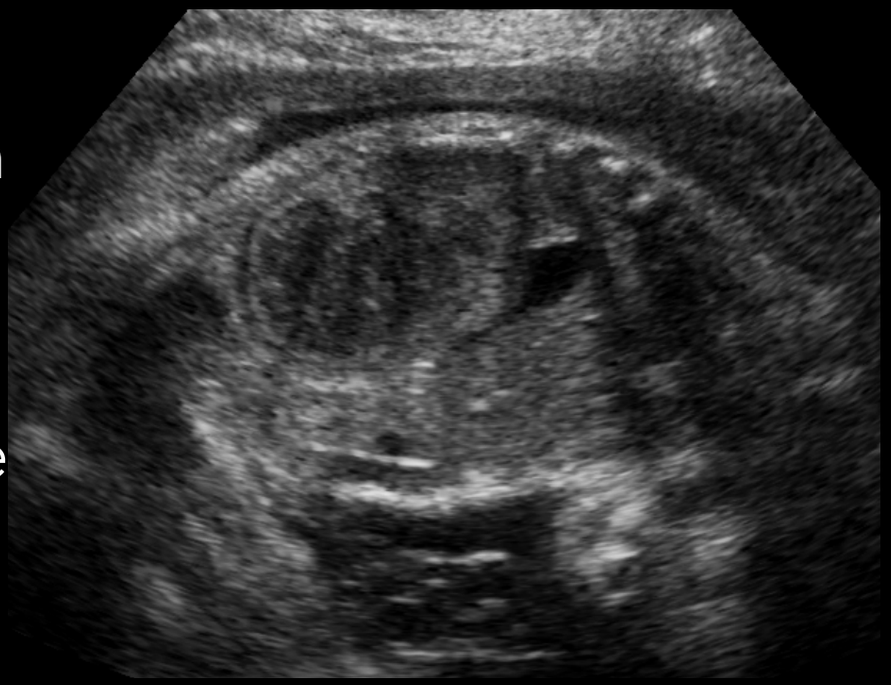
- All female
- Thin walled bladder
- Dilated ureters
- Hydronephrosis
- Normal parenchyma
- Normal AFV clue to diagnosis

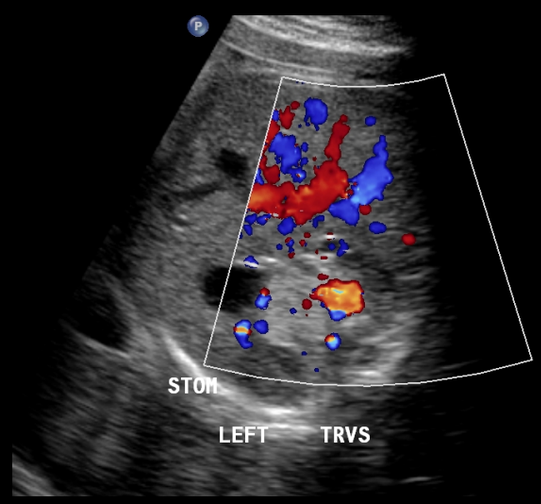


RENAL TUMORS

RENAL TUMORS

- Congenital mesoblastic nephroma most common real neoplasm in fetus and newborn
- Benign hamartoma made up of spindle cells
- Wilms tumor (malignant epithelial lesion) rare
- Features:
  - Echogenic solid mass replacing kidney
  - Assoc. with polydramnios
- Adrenal mass?
  - Neuroblastoma most common





Thank you

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