Chapter 26

NEONATAL & PEDIATRIC ADRENAL AND URINARY SYSTEM

OBJECTIVES:

Upon completion of this presentation, students will be able to:

- Describe exam preparation and protocols for the neonatal/pediatric urinary system.
- Discuss normal anatomy and sonographic findings of neonatal/pediatric kidneys, adrenals, and urinary bladder.
- Recognize congenital urinary tract anomalies and distinguish from pathology.
- Identify bladder outlet obstructions and their sonographic appearances.
- Explain the different malformations of the genitalia.

INDICATIONS

- Numerous indications for a renal ultrasound in the newborn period:
  - Hydronephrosis
  - Palpable mass
  - Abdominal distention
  - Anuria
  - Oliguria
  - Hematuria
  - Sepsis
  - Myelomenigocele
  - Chromosomal anomalies
  - Abnormal external genitalia
  - Prune-belly syndrome

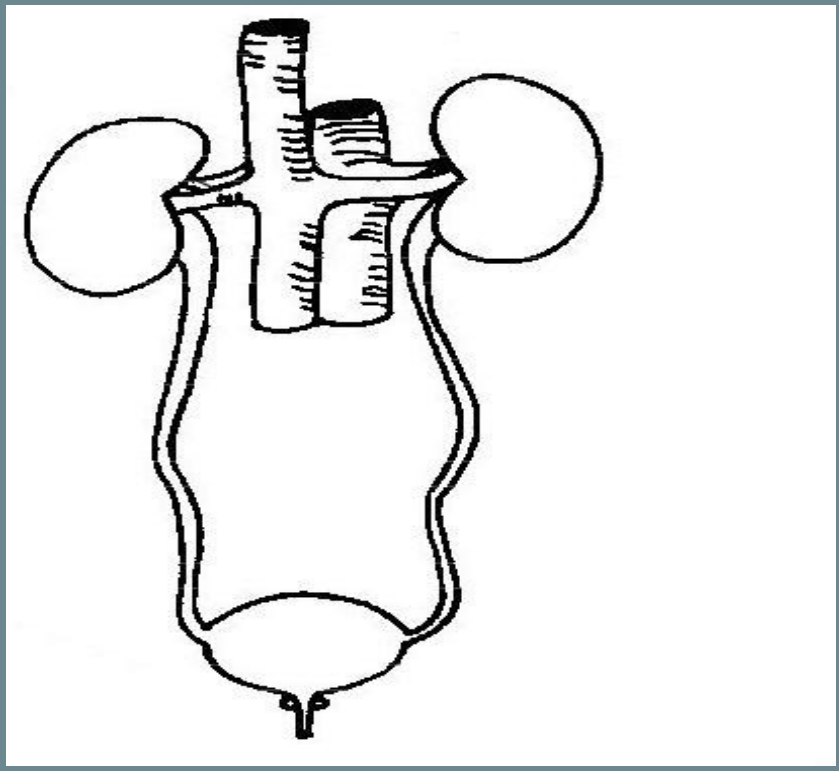
EXAMINATION PREPARATION

- Patient should be evaluated with a full bladder
- Multiple transducers can be used depending on age and size of the patient:
  - 10 MHz tightly curved array for neonates and infants
  - 7.5 MHz tightly curved array for toddler/thin children
  - 9 – 12 MHz linear array for premature infants
  - 3 – 5 MHz curved array for adolescents

EXAMINATION PREPARATION (CONT'D)

- Begin imaging at the level of the bladder first as infant's urinate spontaneously and young children have difficulty holding their bladder
- Obtain images of the pre-void bladder including wall measurement
- With the bladder still full, survey the kidneys and perirenal areas to assess for fluid and to document echogenicity compared to liver/spleen
- Allow the patient to void and re-examine the bladder post-void
- Fully examine the kidneys to include longitudinal and transverse views showing in detail the superior, mid, and inferior poles
  - infants and young children- usually start prone
  - older children and adolescents- usually decubitus

LET'S REVIEW!!



KIDNEYS

- Fetal lobulation may be visualized
  - Remnants of lobes with incomplete fusion
- Glomerular filtration rate is low right after term birth; increases rapidly thereafter
- Renal pyramids remain large in comparison to surrounding cortex
  - Cortex will continue to grow as pyramids regress in size
- Less cortical fat in neonates allows for clear distinction of cortical-medullary differentiation

KIDNEYS (CONT'D)

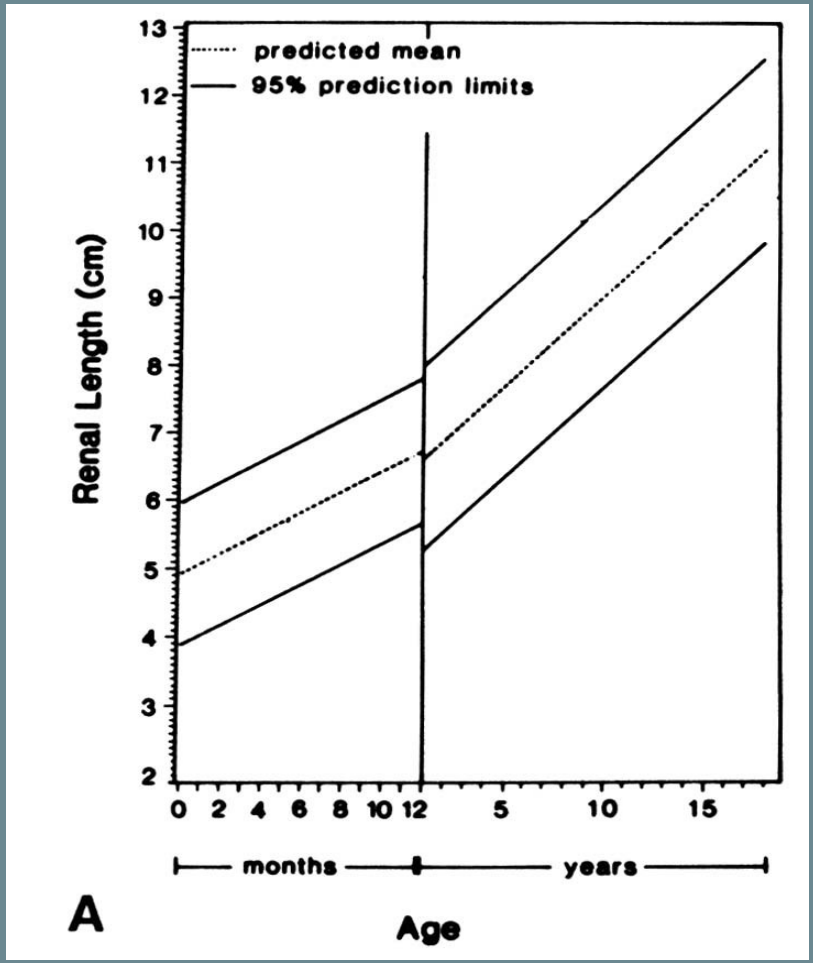
- Medullary pyramids large and hypoechoic
- Surrounding cortex quite thin; echogenicity essentially similar to or slightly greater than normal liver parenchyma
- Renal cortical echogenicity normally decreases to less than liver parenchyma, usually by 4 to 6 months of age
- Renal parenchyma consists of peripheral cortex, glomeruli, and several extensions to edge of renal sinus (column of Bertin)
- Medulla is more central- adjacent to calices
- Normal cortex produces low-level, back-scattered echoes

KIDNEYS (CONT'D)

- Medullary pyramids- hypoechoic and arranged around central, echo-producing renal sinus
- Arcuate vessels- intense specular echoes at corticomedullary junction
- Increased cortical echogenicity results from glomeruli occupying larger proportion of cortical volume and location of 20% of loops of Henle within cortex as opposed to medulla

KIDNEYS (CONT'D)

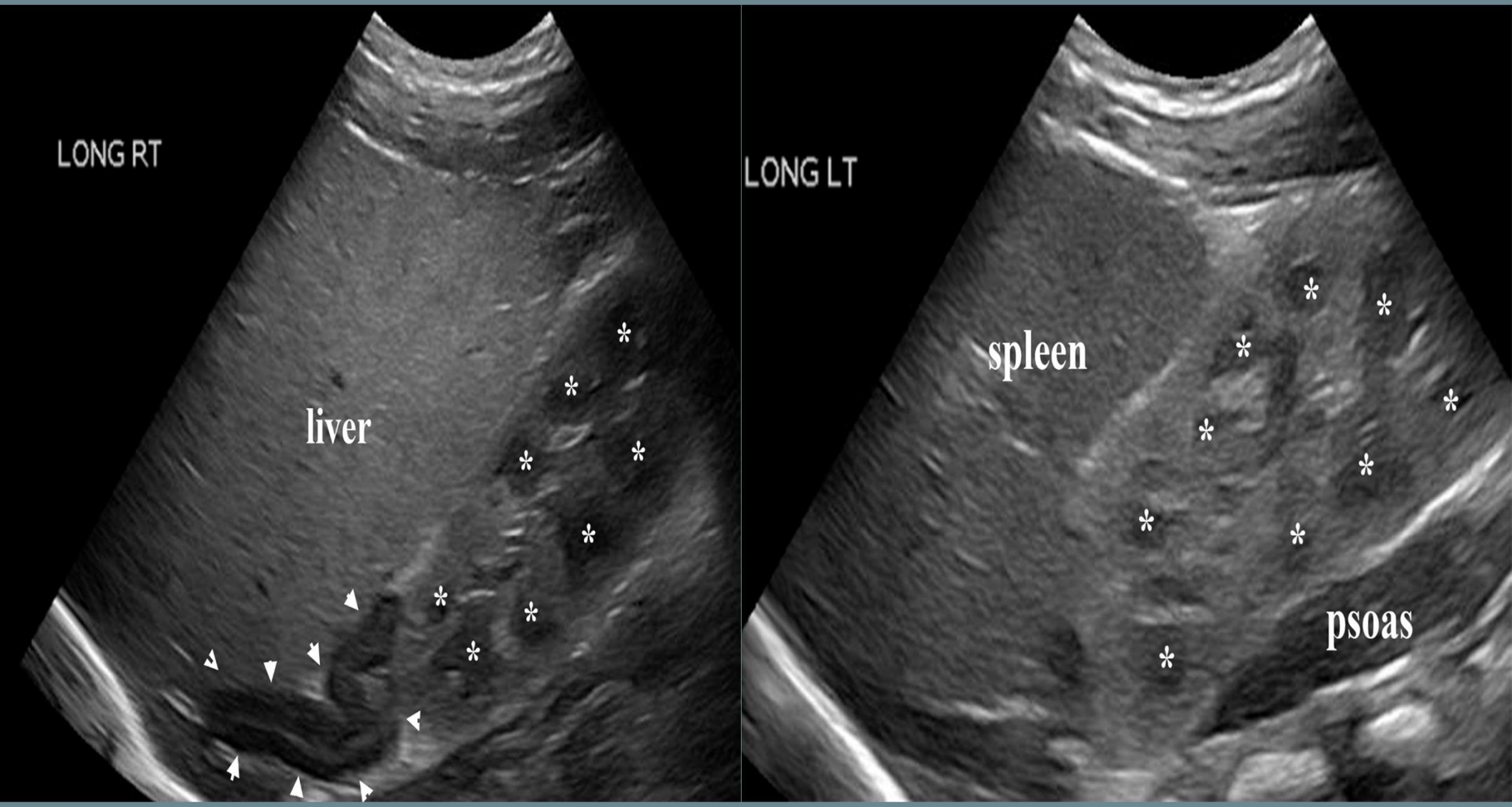
- Normal renal length varies with the age of the neonate or pediatric patient
- Left kidney is somewhat longer
  - Kidney measurement greater than 1 cm side to side should be monitored closely, and may indicate infection, scarring, or congenital abnormalities, such as hypotrophy or duplicated renal system



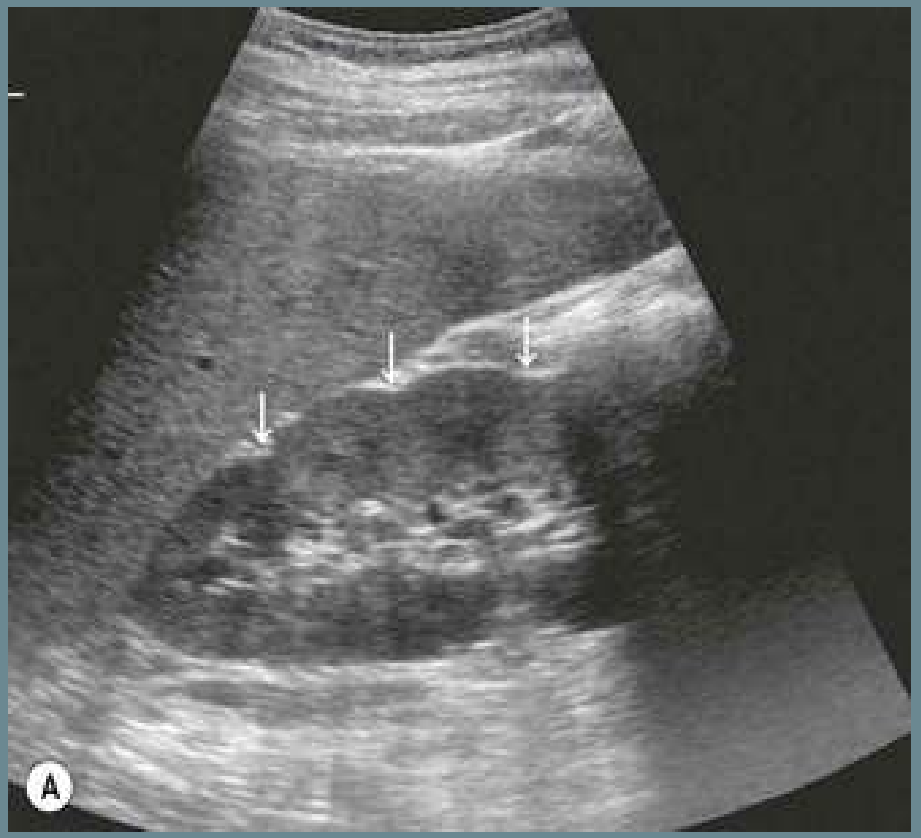
Kidney length in cm

Age	Length (sd)	No	Age	Length (sd)	No
0 – 1 wk	4.5 (0.3)	10	8 – 9 y	8.9 (0.9)	18
1 wk – 4 mo	5.3 (0.7)	54	9 – 10 y	9.2 (0.9)	14
4 – 8 mo	6.2 (0.7)	20	10 – 11 y	9.2 (0.8)	28
8 – 12 mo	6.2 (0.6)	8	11 – 12 y	9.6 (0.6)	22
1 – 2 y	6.6 (0.5)	28	12 – 13 y	10.4 (0.9)	18
2 – 3 y	7.4 (0.5)	12	13 – 14 y	9.8 (0.8)	14
3 – 4 y	7.4 (0.6)	30	14 – 15 y	10.0 (0.6)	14
4 – 5 y	7.9 (0.5)	26	15 – 16 y	11.0 (0.8)	6
5 – 6 y	8.1 (0.5)	30	16 – 17 y	10.0 (0.9)	10
6 – 7 y	7.8 (0.7)	14	17 – 18 y	10.5 (0.3)	4
7 – 8 y	8.3 (0.5)	18	18 – 19 y	10.8 (1.1)	8

https://radiologyassistant.nl/pediatrics/normal-values/normal-values-ultrasound


LONG RT
liver
LONG LT
spleen
psoas

https://quizlet.com/155060220/pediatrics-chap-27-28-flash-cards/



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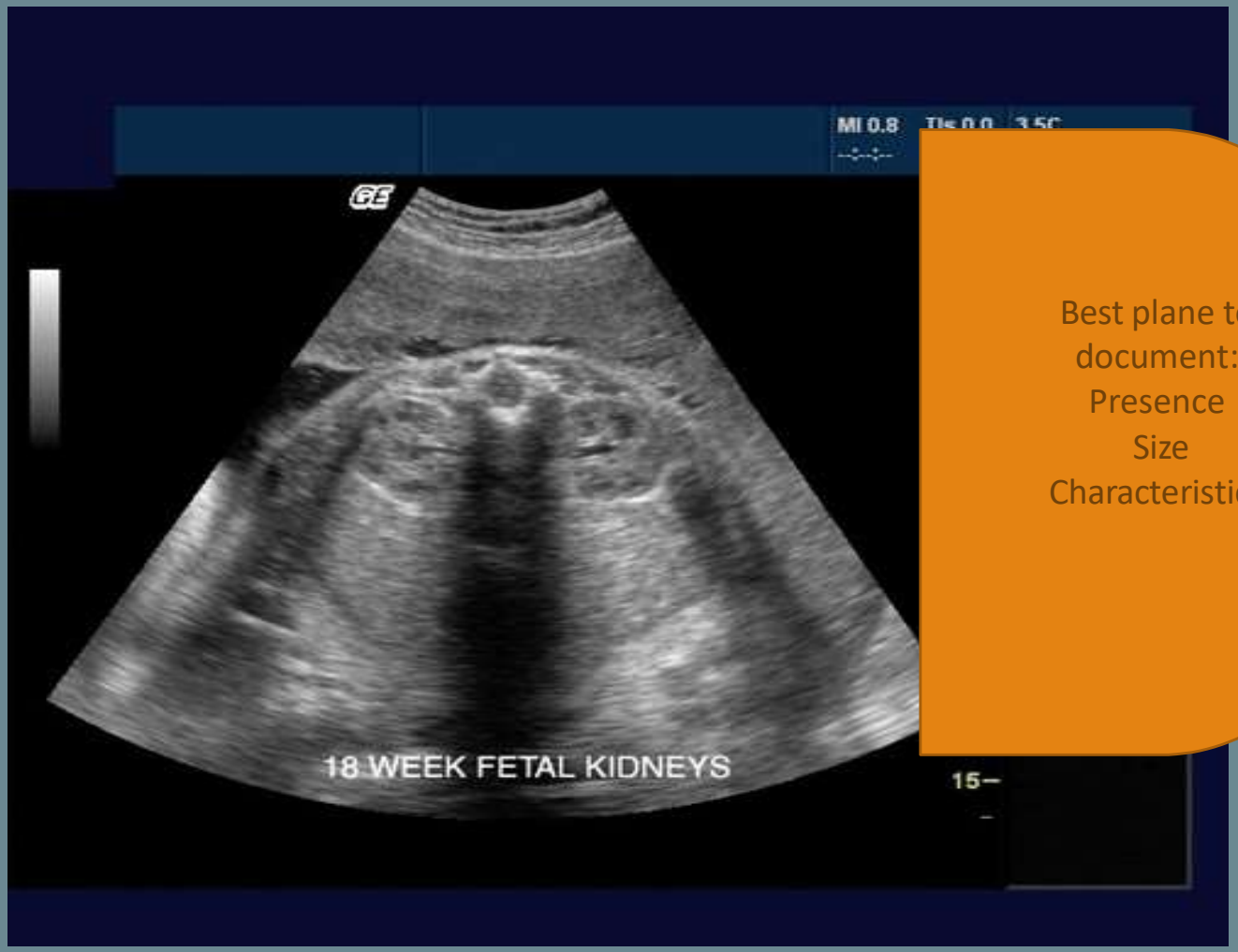


Best plane to document:
Presence
Size
Characteristics

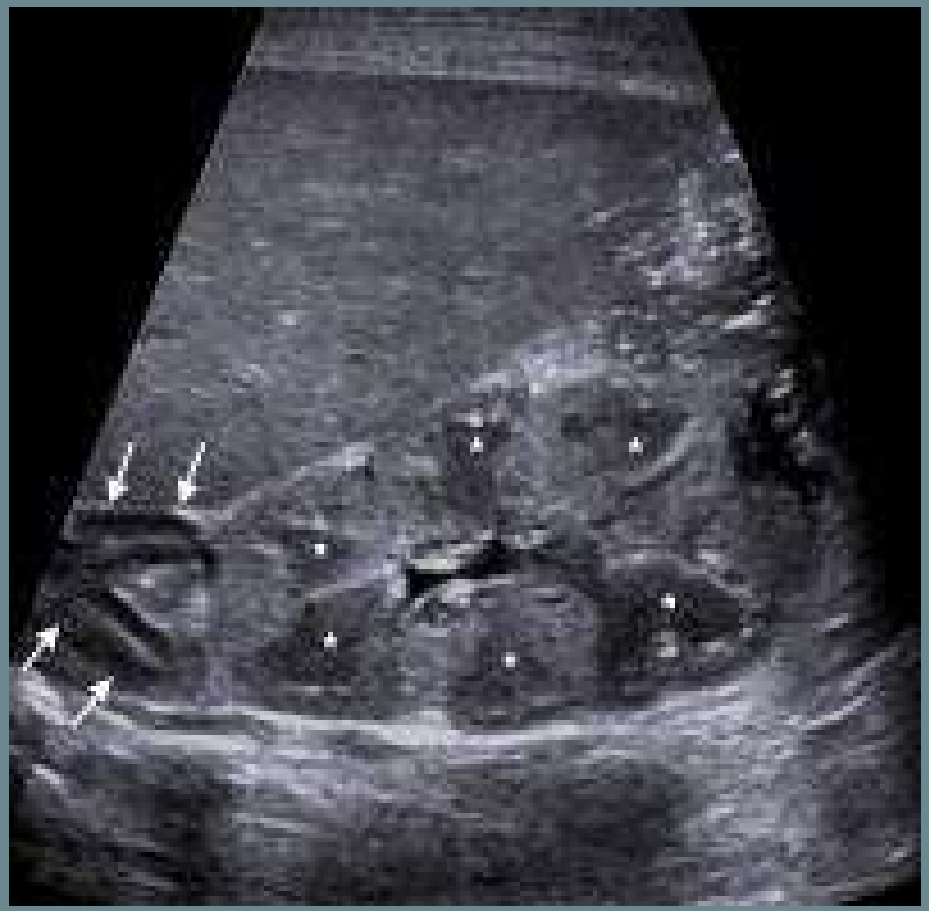
KIDNEYS (CONT'D)

- Anomalies of position, form, orientation:
  - ✓ Pelvic kidney
  - ✓ Horseshoe kidney
  - ✓ Crossed ectopy
  - ✓ Renal duplication

WHAT CAN YOU DO TO IMPROVE VISIBILITY?

ADRENAL GLANDS

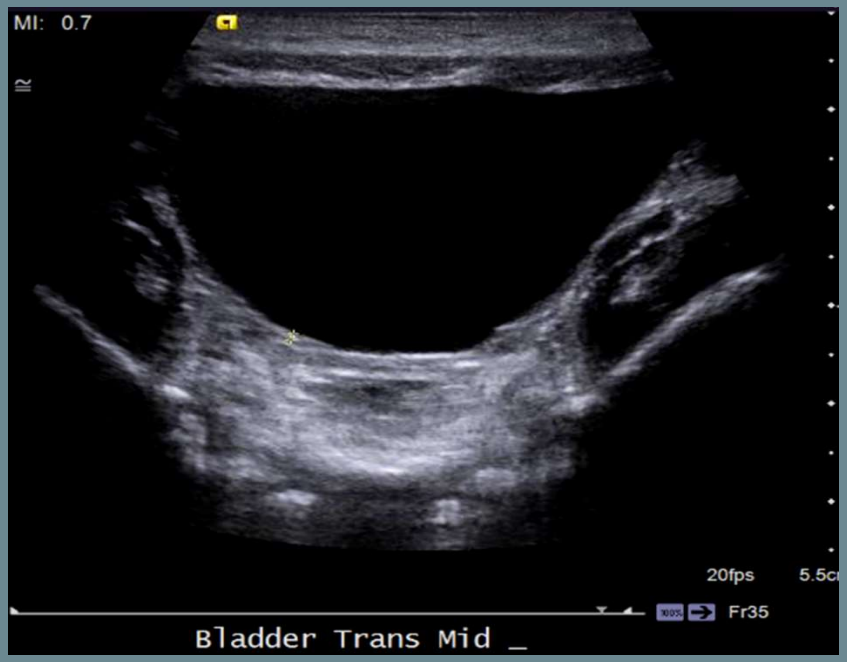
- Larger and more easily identified in the neonate than in older infant or young child
- Left adrenal gland extends slightly more medial than the right
- Sonographically, the gland has an inverted "V" or "Y" shape in the longitudinal plane
  - Central medulla appears as a thin echogenic stripe surrounded by less echogenic cortex
- When the kidney is absent, ipsilateral adrenal gland may have an altered configuration



URINARY BLADDER

- The normal urinary bladder is thin-walled in the distended state and should measure less than 3 mm (with a mean of 1.5 mm) in AP

- When empty, the wall thickness increases but remains less than 5 mm



Normal bladder in 1 day old infant

RENAL, ADRENAL, AND BLADDER PATHOLOGY

CONGENITAL URINARY TRACT ANOMALIES

- Hydronephrosis
- Patent urachus
- Multicystic dysplastic kidney disease (MCDK)

HYDRONEPHROSIS

- Aka. Urinary tract dilatation (UTD)
- Dilation of the urinary collecting system

*Most common urinary tract anomaly in children

* Most common cause of palpable mass in the neonate

- Causes of dilation of the collecting system:
  - Obstruction
  - Reflux
  - Abnormal muscle development

HYDRONEPHROSIS (CONT'D)

- Determine the severity of the hydronephrosis, whether the condition is unilateral or bilateral, if the ureters and bladder dilated, and the status of the renal parenchyma

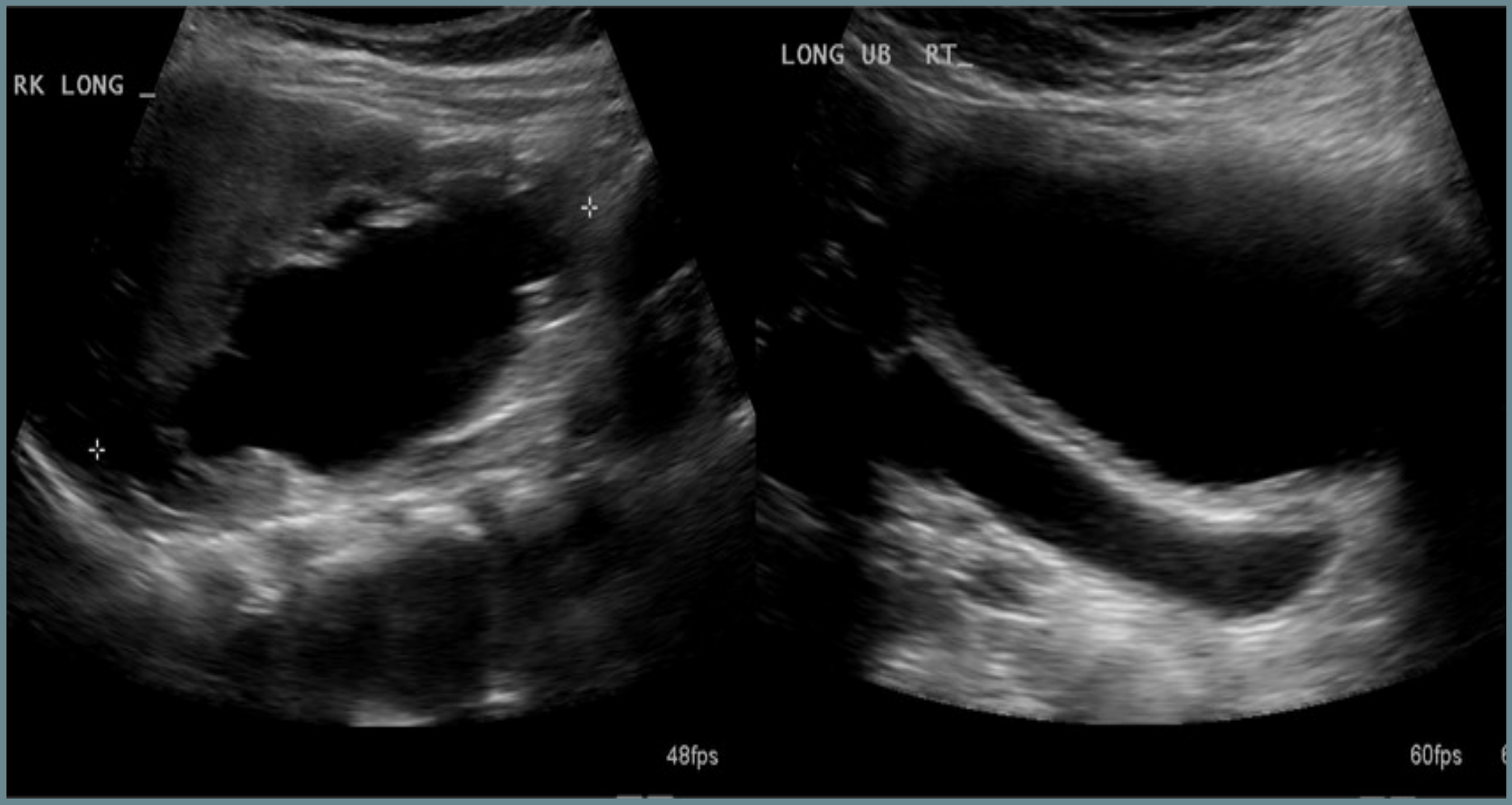
- Sonographic features:
  - Visible renal parenchyma surrounding central cystic component
  - Small peripheral cysts budding off large central cyst
  - Dilated ureter

VESICOURETERAL REFLUX (VUR)

- Common, nonobstructive cause of hydronephrosis
- VUR is abnormal refluxing of urine from the urinary bladder through the ureters and into the kidney
- Five different grades: 1- least severe, 5- most severe
- Treated conservatively because it is nonobstructive and most resolve within 2 years
- Low urine output is normal at birth, typically waiting 7-10 days is preferential

VUR (CONT'D)

- Sonographic findings:
  - Often nonspecific
  - May or may not have hydronephrosis
  - Pelvic or ureteral wall thickening
  - Dilation of collecting system
  - Megaureter
  - Displaced ureteral jet in bladder


RK LONG _
48fps
LONG UB RT_
60fps

URETEROPELVIC JUNCTION(UPJ) OBSTRUCTION

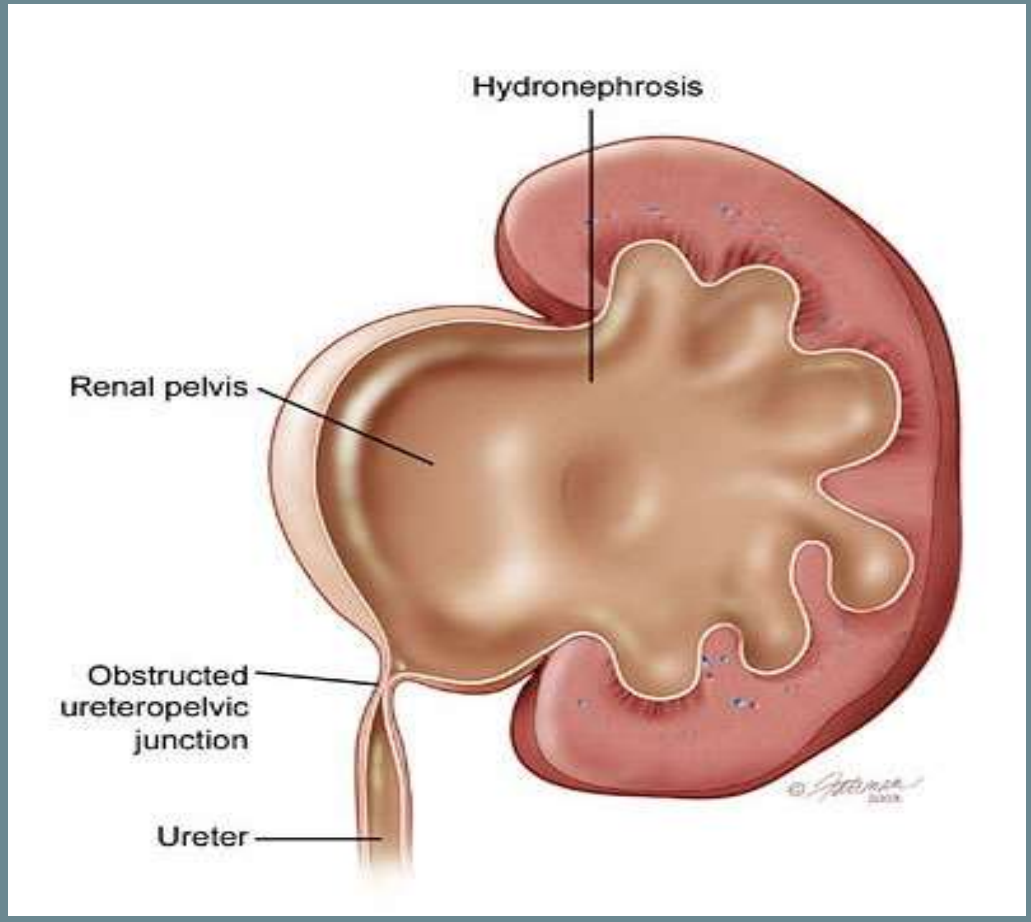
*Most common type of obstruction causing hydronephrosis

- 3:1 male prevalence
- Results from intrinsic narrowing or extrinsic vascular compression at the level of the UPJ
  - Obstruction produces proximal dilation of the collecting system
  - Ureter is often normal in caliber unless vesicoureteral reflux (VUR) or megaureter is present
- Increased incidence of abnormalities in contralateral kidney

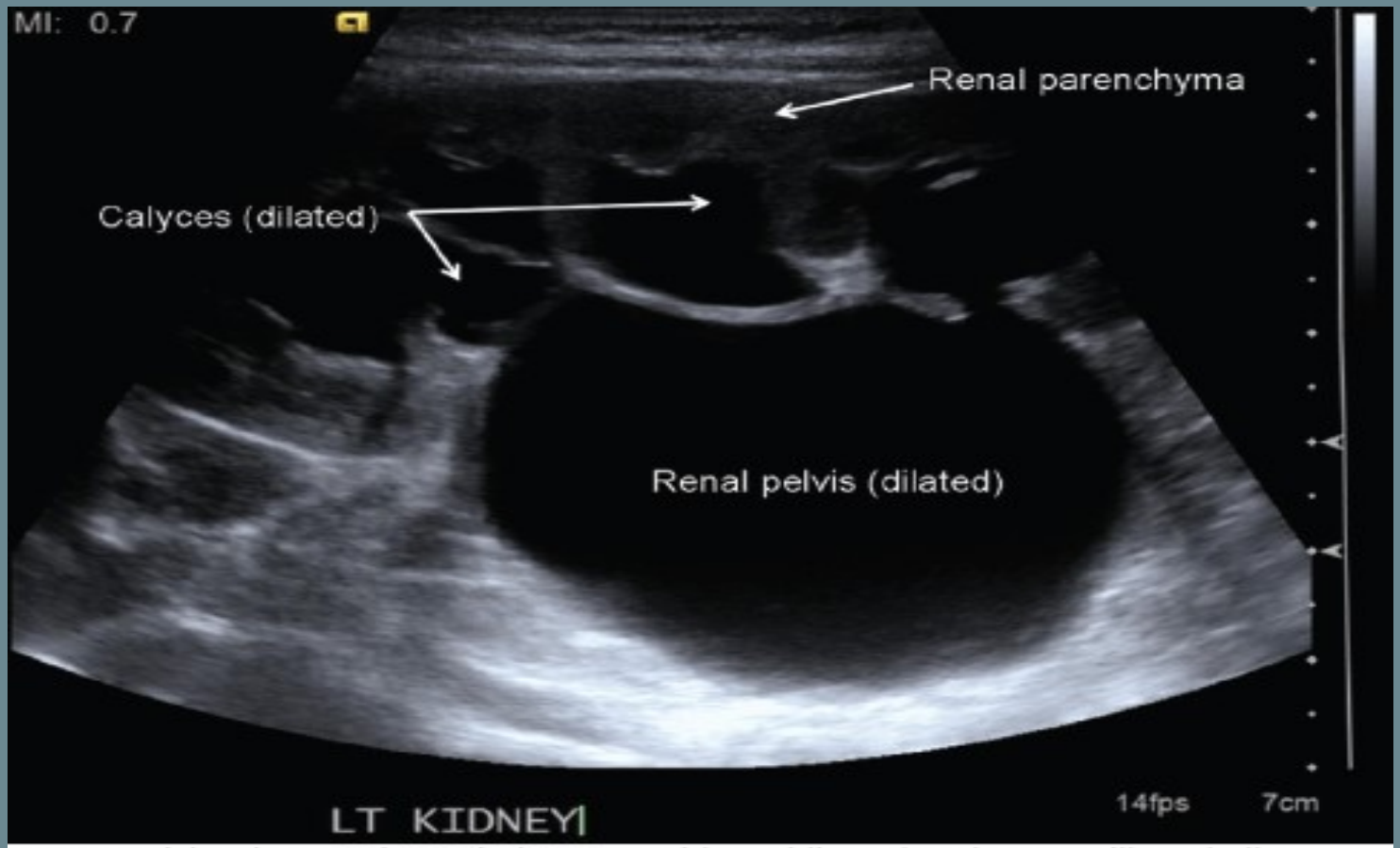
UPJ (CONT'D)

Sonographic Findings:

- Pelvocalyceal dilation without ureteral dilation
- When obstruction pronounced, dilated renal pelvis extends inferiorly and medially
- If vesicoureteral reflux or primary megaureter is present, ureter may be dilated



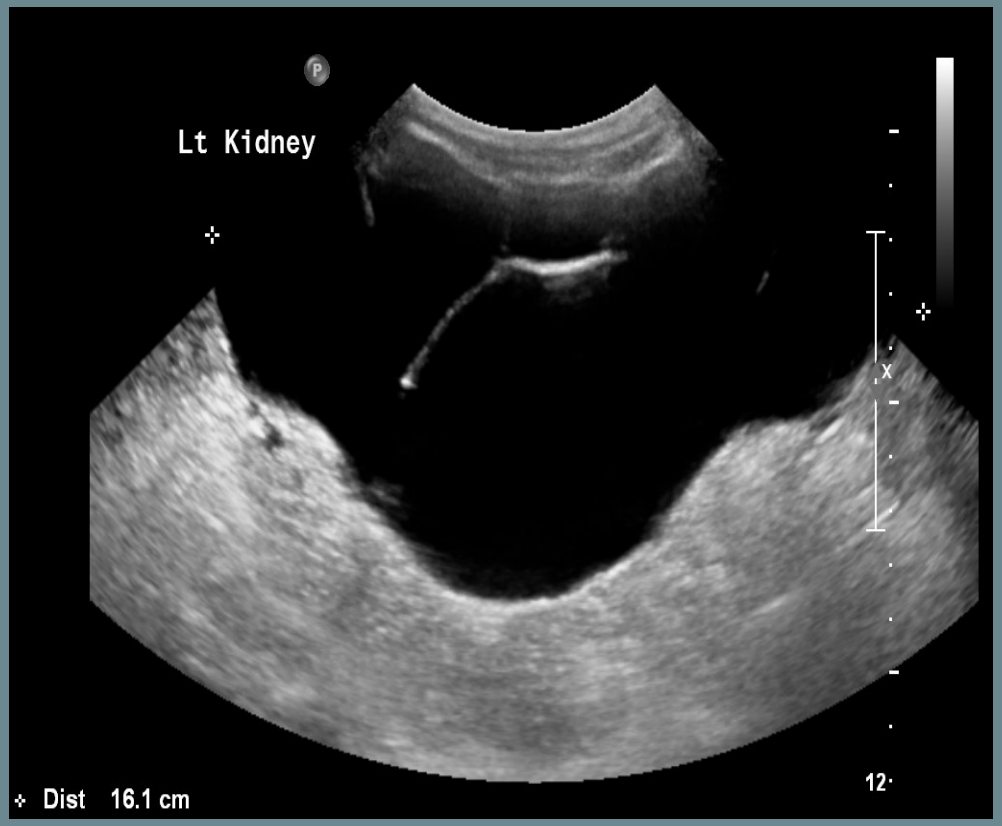
https://www.endourologytraining.in/ureteropelvic-junction-obstruction/



Source: Richard P. Usatine, Mindy Ann Smith, Heidi S. Chumley, Camille Sabella, E.J. Mayeaux, Jr., Elumalai Appachi: *The Color Atlas of Pediatrics*: www.accesspediatrics.com

URETERAL OBSTRUCTION

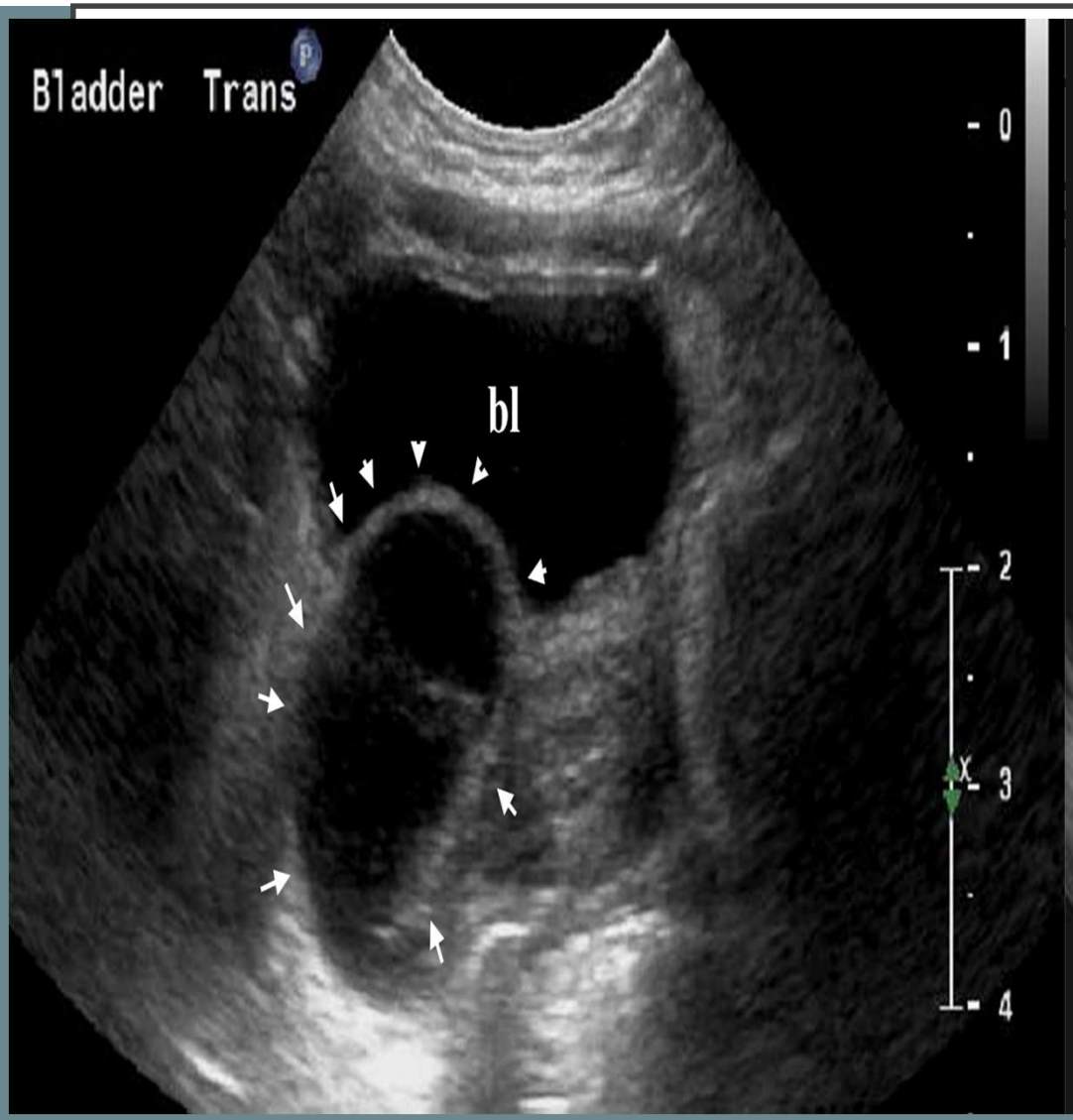
- Ureter may be obstructed anywhere along its course or at ureterovesical junction
- Abscess or lymphoma may cause:
  - Obstruction to ureter
  - Presence of primary megaureter
  - Atresia
  - Ectopic ureter may be cause of obstruction
- Sonographic findings:
  - Hydronephrosis and hydroureter with narrow segment of distal ureter behind bladder
  - Diminished ureteral jet may be seen in the bladder with color Doppler on the side of the obstruction


Lt Kidney
Dist 16.1 cm


RK LONG
URETER

DUPLEX COLLECTING SYSTEM & ECTOPIC URETEROCELE

- Aka. Duplex or double kidney
- Results from an ectopic bladder insertion and cystic dilation of the distal ureter of the upper pole from a completely duplicated renal collecting system
- More commonly in the female and more often on the lef
- Appears as a fluid mass within the urinary bladder
- Sonographic findings:
  - dilated upper pole and ureter to the bladder


Bladder Trans
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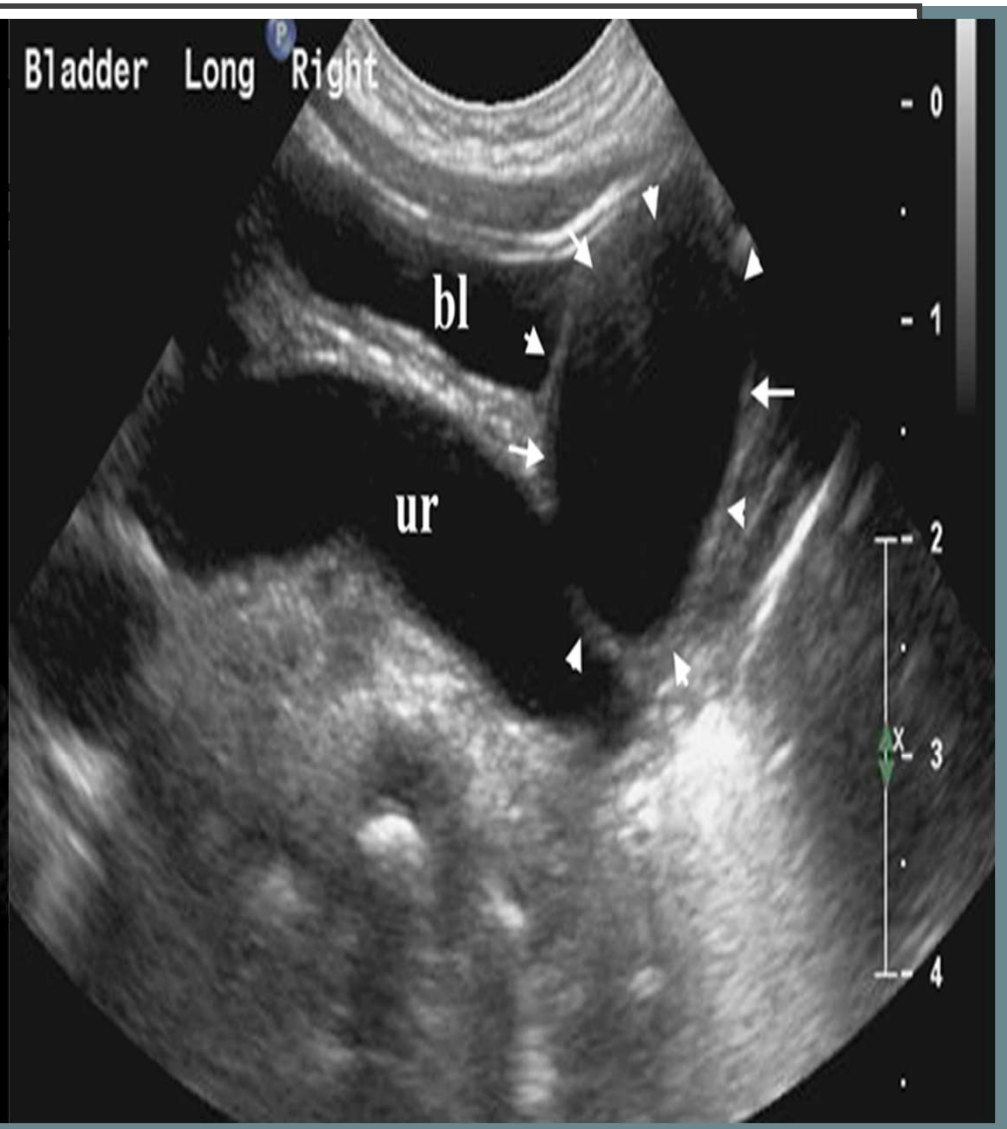
Bladder Long Right
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ur
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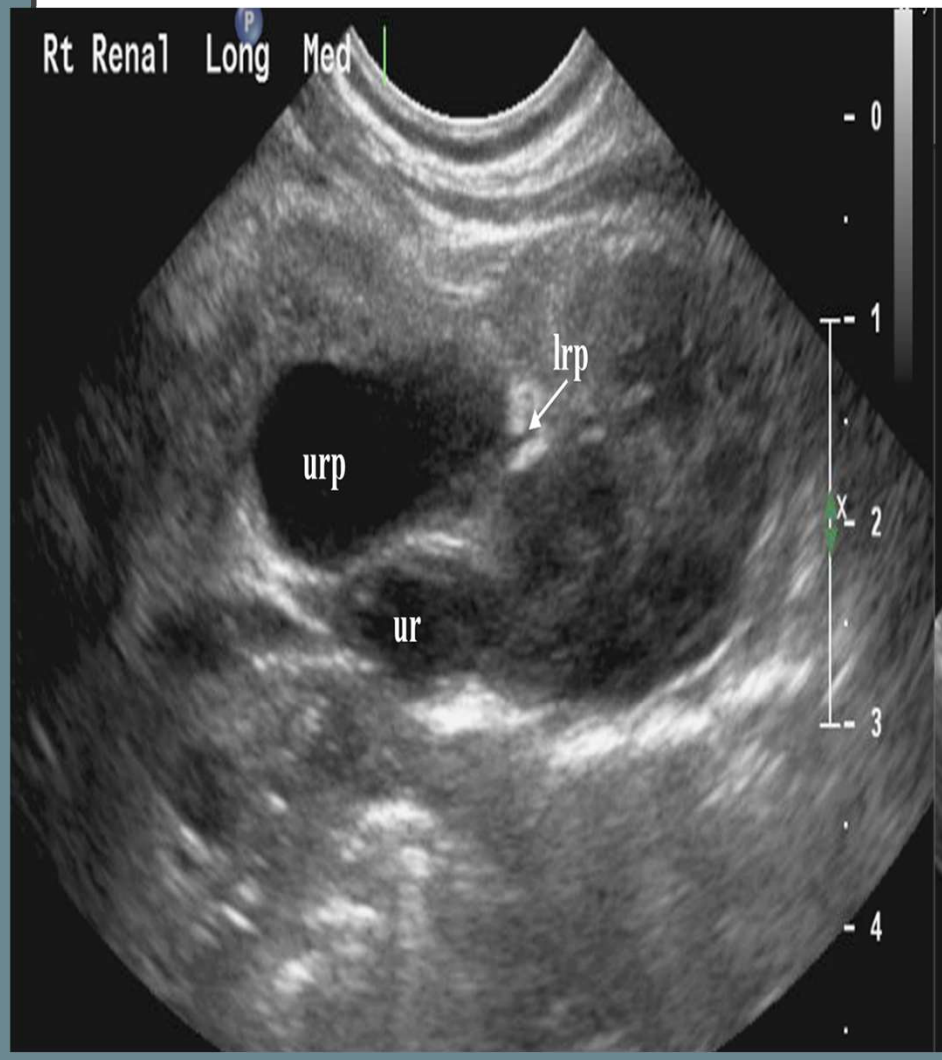
Rt Renal Long Med
lrp
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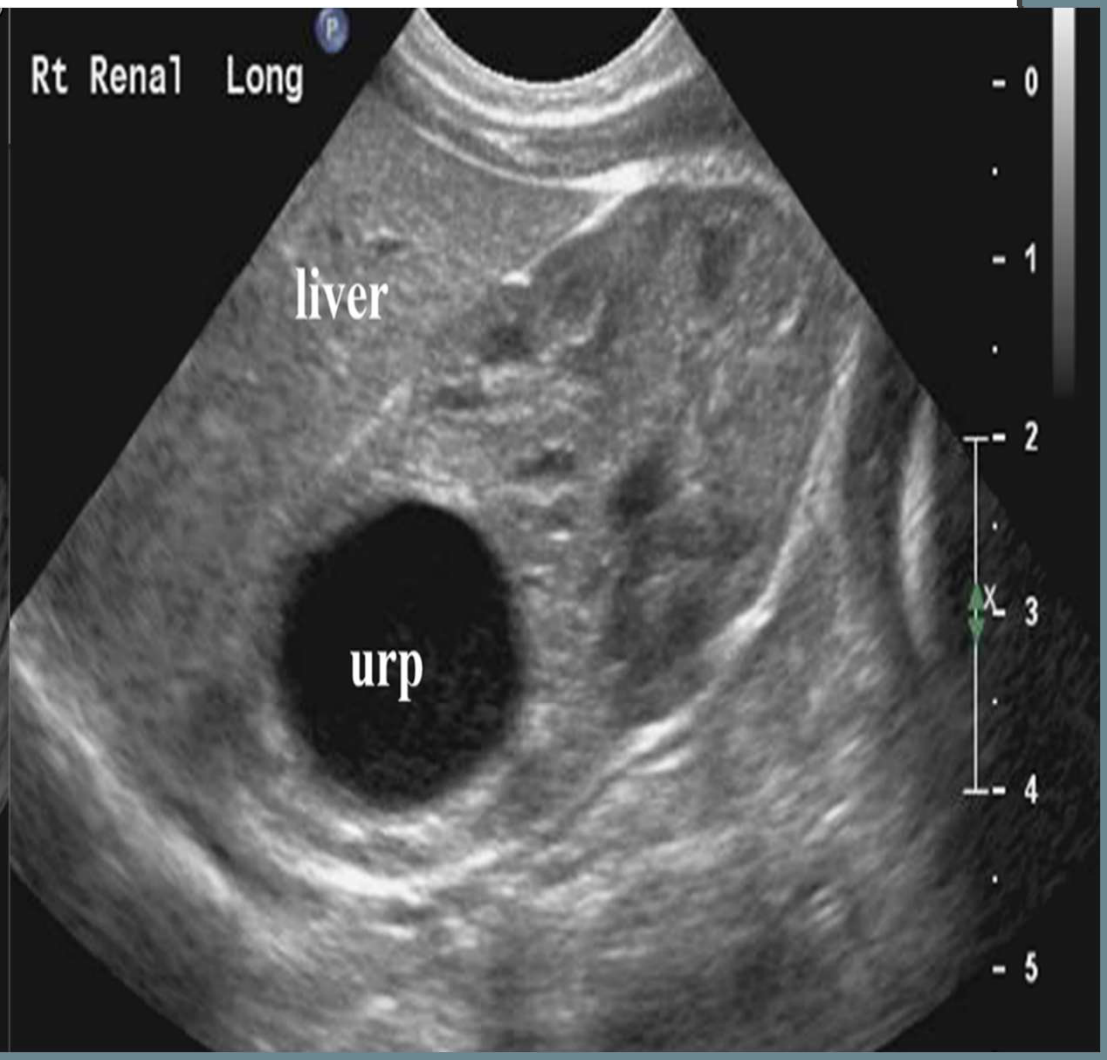

Rt Renal Long
liver
urp

BLADDER OUTLET OBSTRUCTION

- Bilateral hydronephrosis is frequently caused by obstruction at the level of bladder or bladder outlet
- Bladder may be obstructed by neurogenic bladder, pelvic mass, or congenital anomaly such as posterior urethral valves

BLADDER OUTLET OBSTRUCTION (CONT'D)

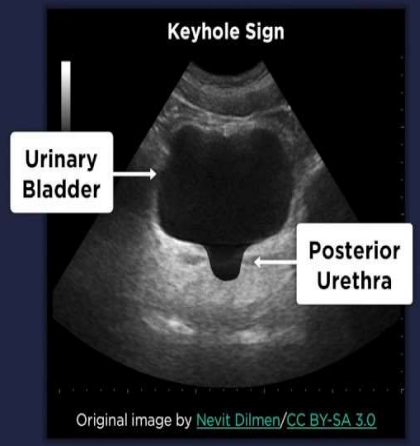
- Sonographic findings:
  - May identify perirenal urinoma- anechoic structure filled with urine
- Other potential causes of perirenal urine extravasation include:
  - trauma
  - UPJ obstruction
  - UVJ obstruction
  - pelvic masses that cause obstruction of the bladder or ureter

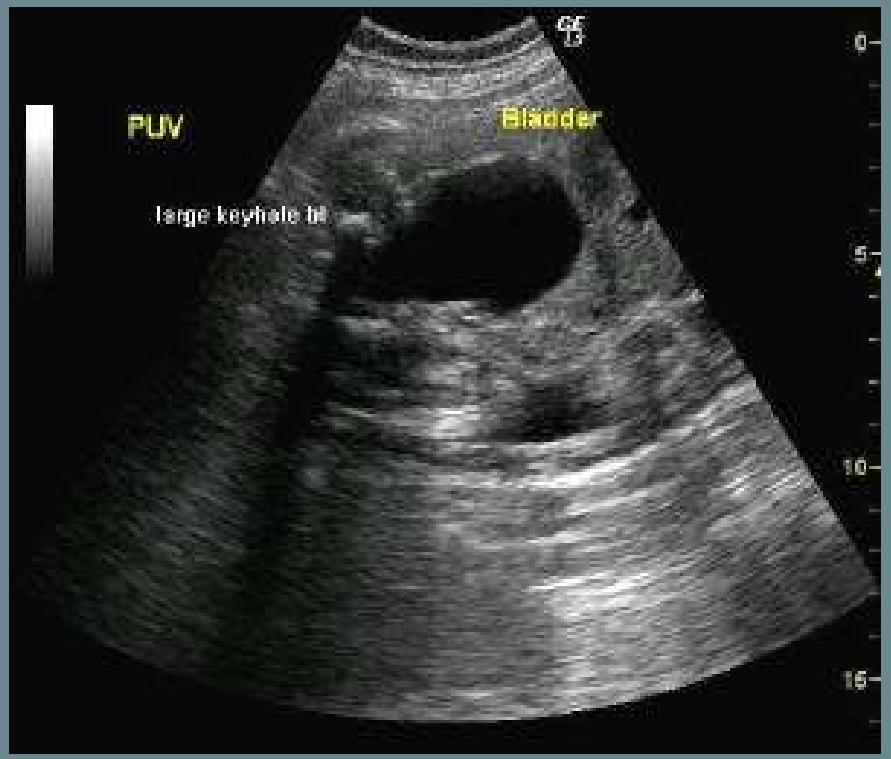
POSTERIOR URETHRAL VALVE OBSTRUCTION (PUV)

- *Most common cause of bladder outlet obstruction in male neonate
- Caused by excessive flaps of tissue in posterior urethra
- Sonographic findings:
  - Keyhole appearance of bladder (neonatal)
  - Wall of bladder appears thickened and trabeculated
  - Midline sagittal imaging with caudal angulation may allow visualization of distended posterior urethra
  - Perineal approach may also be helpful to image posterior urethra
  - Hydronephrosis and hydroureter are usually bilateral
  - May identify perirenal urinoma- anechoic structure filled with urine

PUV

- A large portion of cases are detected on prenatal ultrasound.
  - Ultrasound may reveal a distended and thickened (e.g., lower abdominal mass), hydronephrosis, and/or a dilated posterior urethra (keyhole sign).



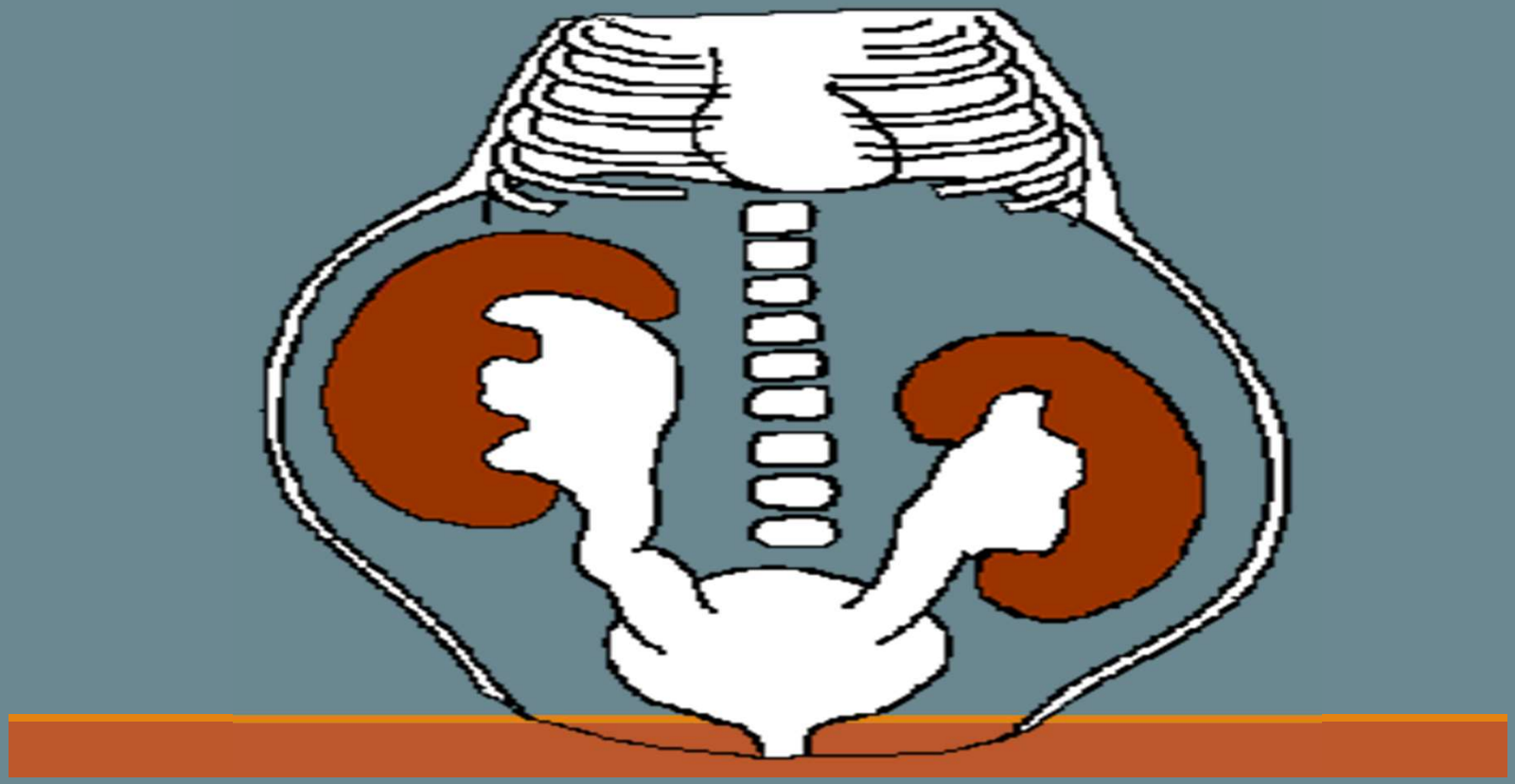


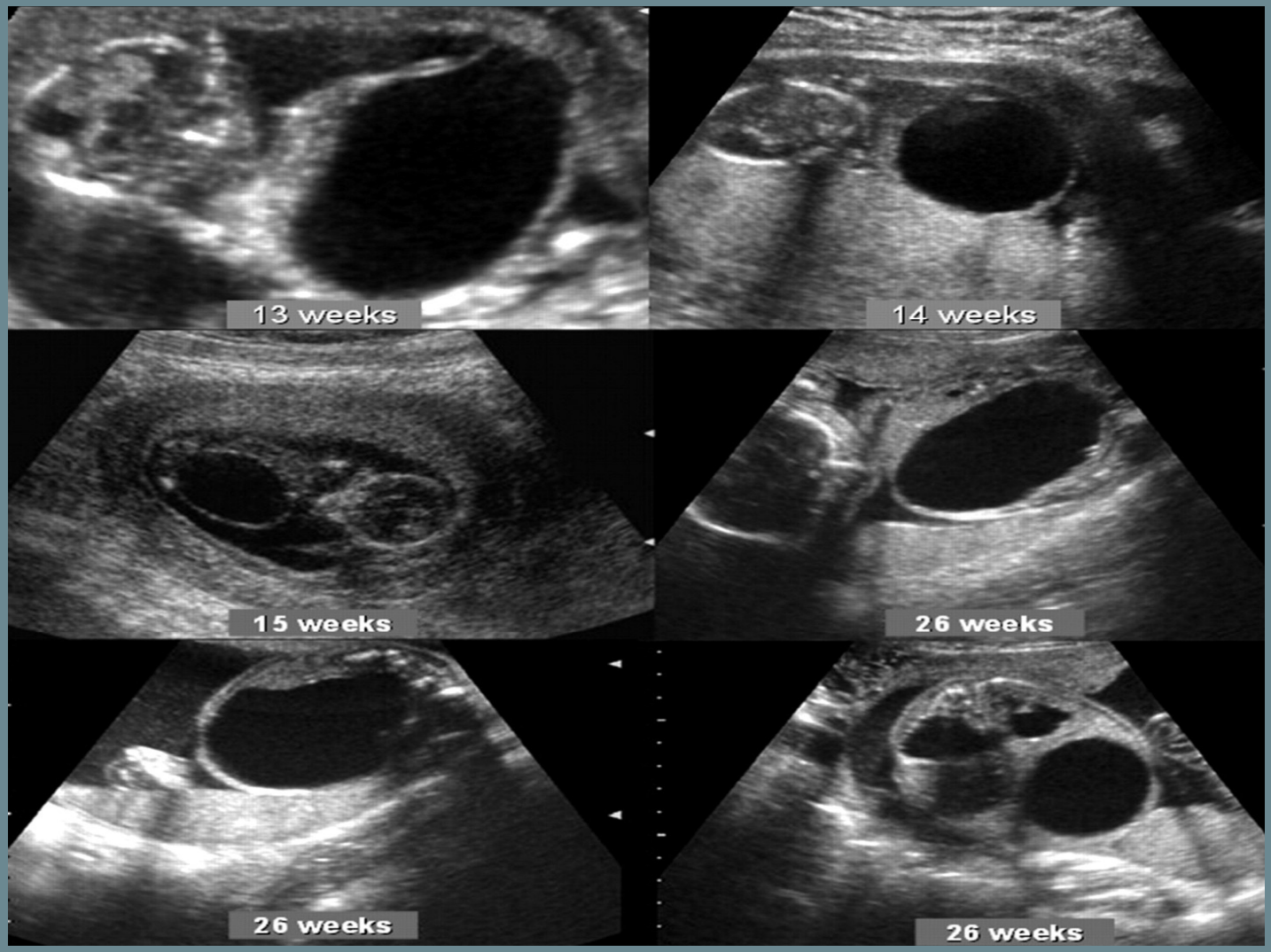
URETHRAL ATRESIA

- Occurs in females
- Similar findings as in PUV
- Less common
- Need to know gender!!!!

PRUNE-BELLY SYNDROME

- Abdominal muscle deficiency syndrome- sever distention of abd cavity
- Aka. Eagle-Barrett Syndrome
- Rare, congenital anomaly of unknown etiology
- Males (96% of cases)
  - severe cases results in early infant death due to pulmonary hypoplasia
- Triad of clinical findings:
  - hypoplasia or deficiency of the abdominal musculature
  - bilat. cryptorchidism
  - urinary tract anomalies- dilated ureters and bladder




13 weeks
14 weeks
15 weeks
26 weeks
26 weeks
26 weeks

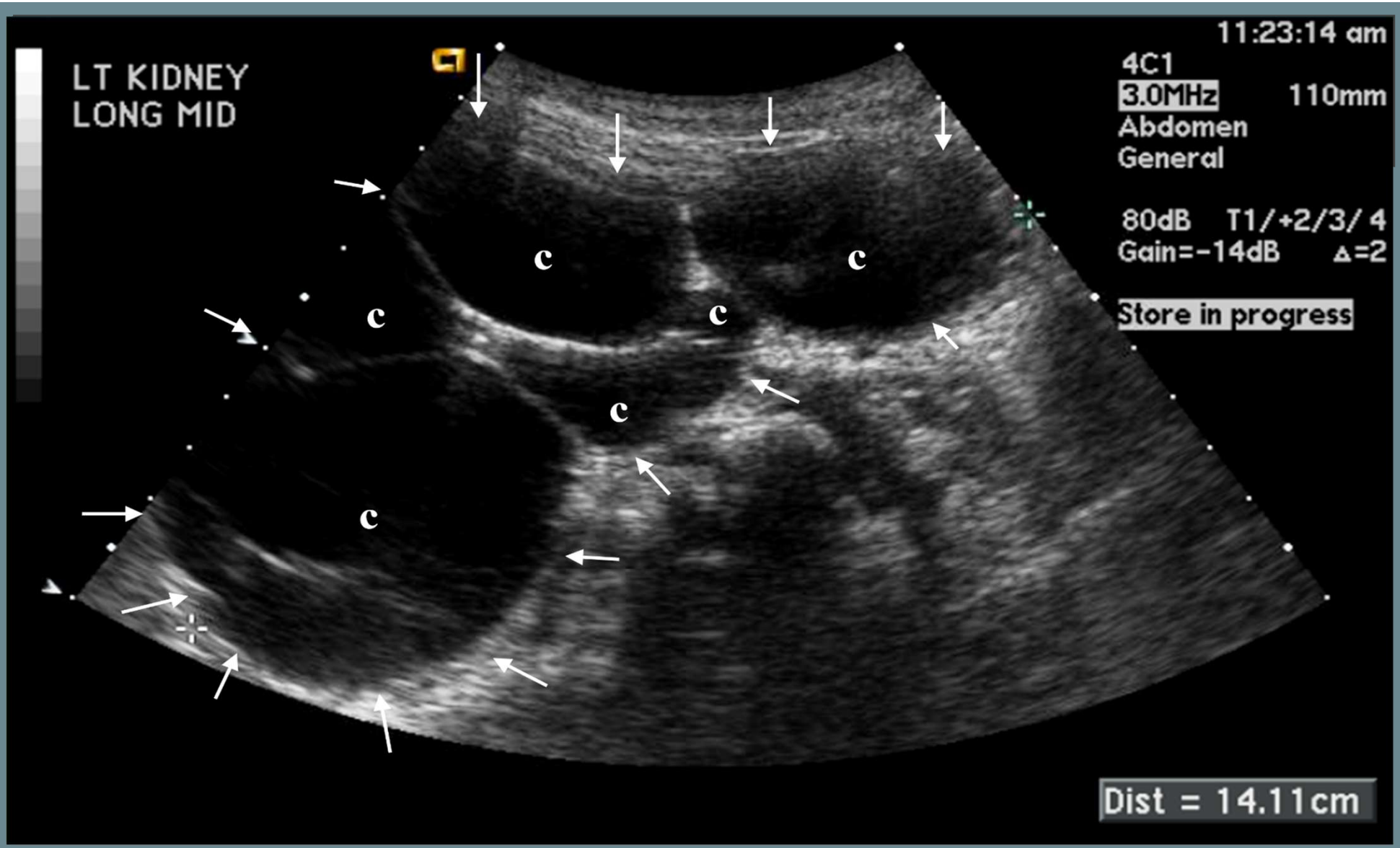
RENAL CYSTIC DISEASE

MULTICYSTIC DYSPLASTIC KIDNEY (MCDK)

- *Most common cause of renal cystic disease in neonate when hydronephrosis is
- excluded
- *Most common cause of abdominal mass in newborn
- Congenital, usually sporadic, renal dysplasia
  - Secondary to severe, generalized interference with ureteral bud function during first trimester

MCKD (CONT'D)

- Collecting tubules enlarge, becoming cystic, grossly distorting the shape of the kidney
- Remaining renal parenchyma becomes virtually nonfunctioning
- Nearly half the cases have contralateral abnormalities
- Sonographic findings:
  - Classic appearance is unilateral mass resembling "cluster of grapes" that represent multiple discrete noncommunicating cysts
- No identifiable renal pelvis


LT KIDNEY
LONG MID
11:23:14 am
4C1
3.0MHz
110mm
Abdomen
General
80dB T1/+2/3/4
Gain=-14dB Δ=2
Store in progress
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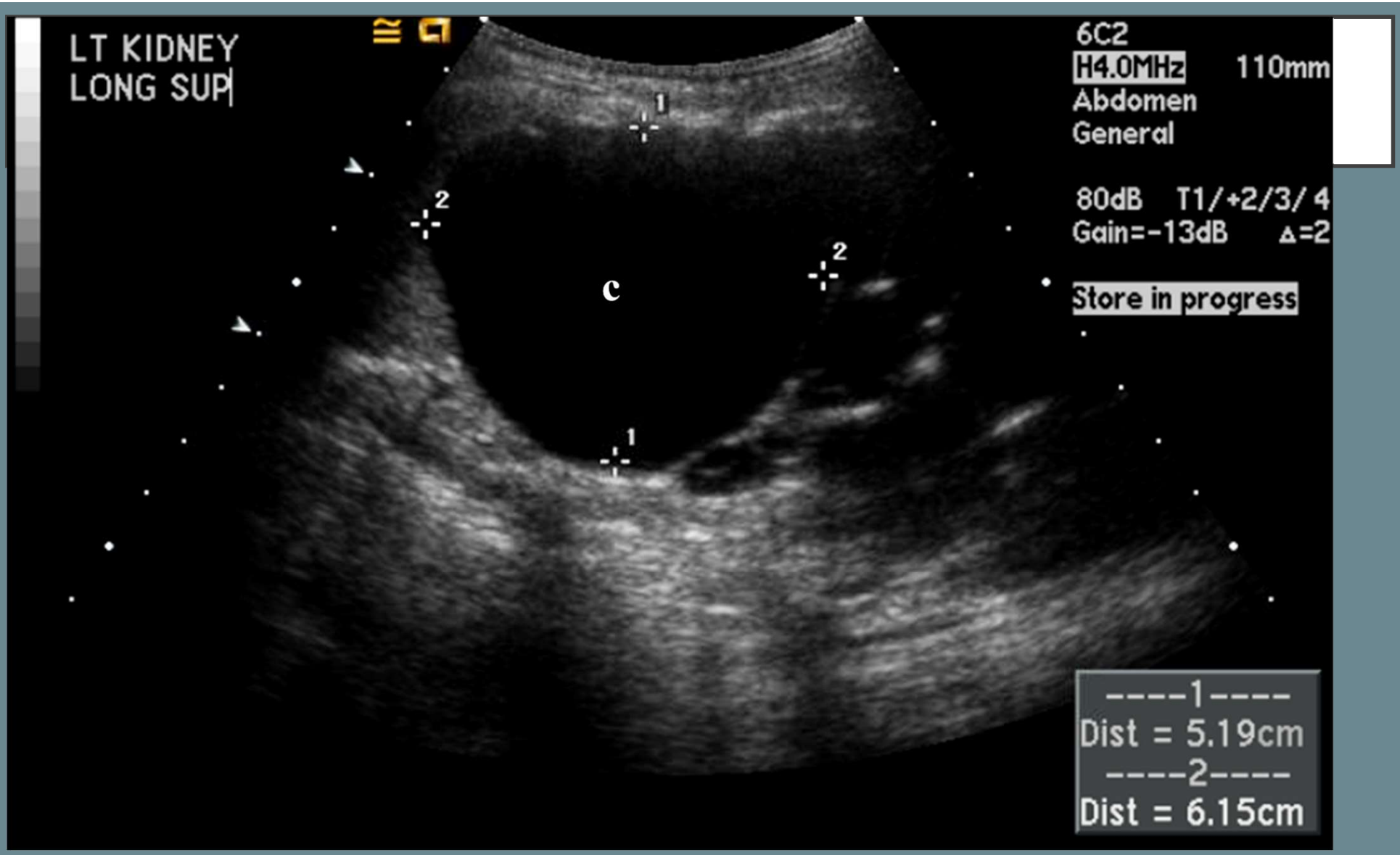
Dist = 14.11cm


LT KIDNEY
LONG SUP
6C2
H4.0MHz
110mm
Abdomen
General
80dB T1/+2/3/4
Gain=-13dB
Store in progress
----1----
Dist = 5.19cm
----2----
Dist = 6.15cm

HEREDITARY RENAL CYSTIC DISEASE

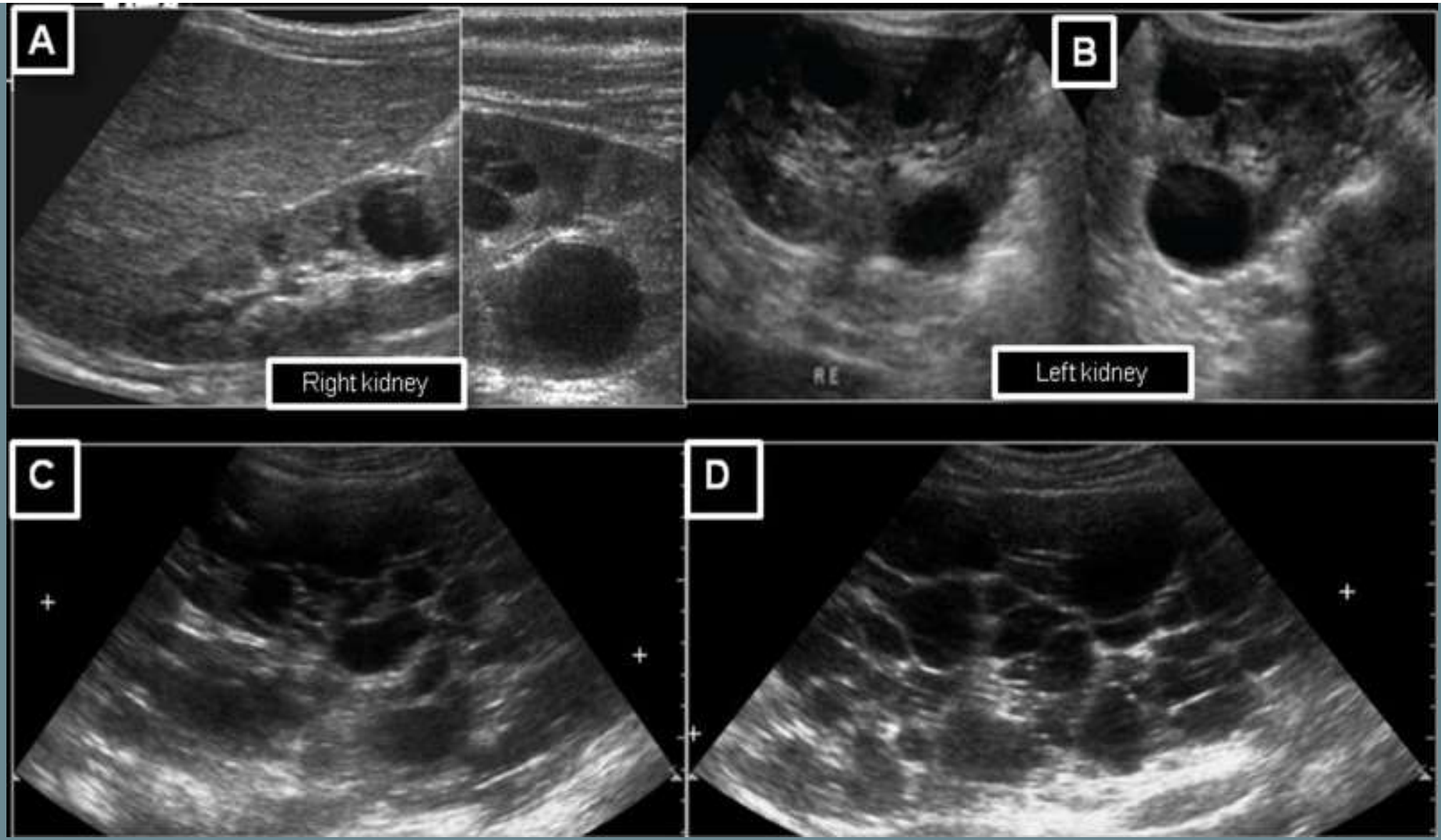
- Hereditary renal cystic disease, which may present in the neonate or pediatric patient include:
  - Autosomal recessive polycystic kidney disease (ARPKD)
  - Autosomal dominant polycystic kidney disease (ADPKD)
  - Tuberous sclerosis
  - Von Hippel-Lindau disease
  - Medullary cystic disease/nephronophthisis

AUTOSOMAL RECESSIVE POLYCYSTIC KIDNEY DISEASE (ARPKD)

- Typical pathologic presentation: diffuse enlargement, sacculations, cystic diverticula of medullary portions of kidneys
- Most severe form seen in neonatal stage- usually fatal within months
- Less severe in infantile to juvenile stage
  - Always bilateral
  - Always congenital hepatic fibrosis
- Mutation of PKHD1 gene on chromosome 6
- Kidneys appear hyperechoic, enlarged with hypoechoic rim representing cortex compressed by expanded pyramids (sponge-like)

AUTOSOMAL RECESSIVE POLYCYSTIC KIDNEY DISEASE (ARPKD) (CONT'D)

- Sonographic findings:
  - Bilateral renal enlargement
  - Diffuse increased echogenicity and loss of definition of renal sinus, medulla, cortex
  - Less severe cases: hepatosplenomegaly and portal hypertension, with normal to echogenic renal parenchyma
  - Pulmonary hypoplasia and Potter fascies may be associated findings

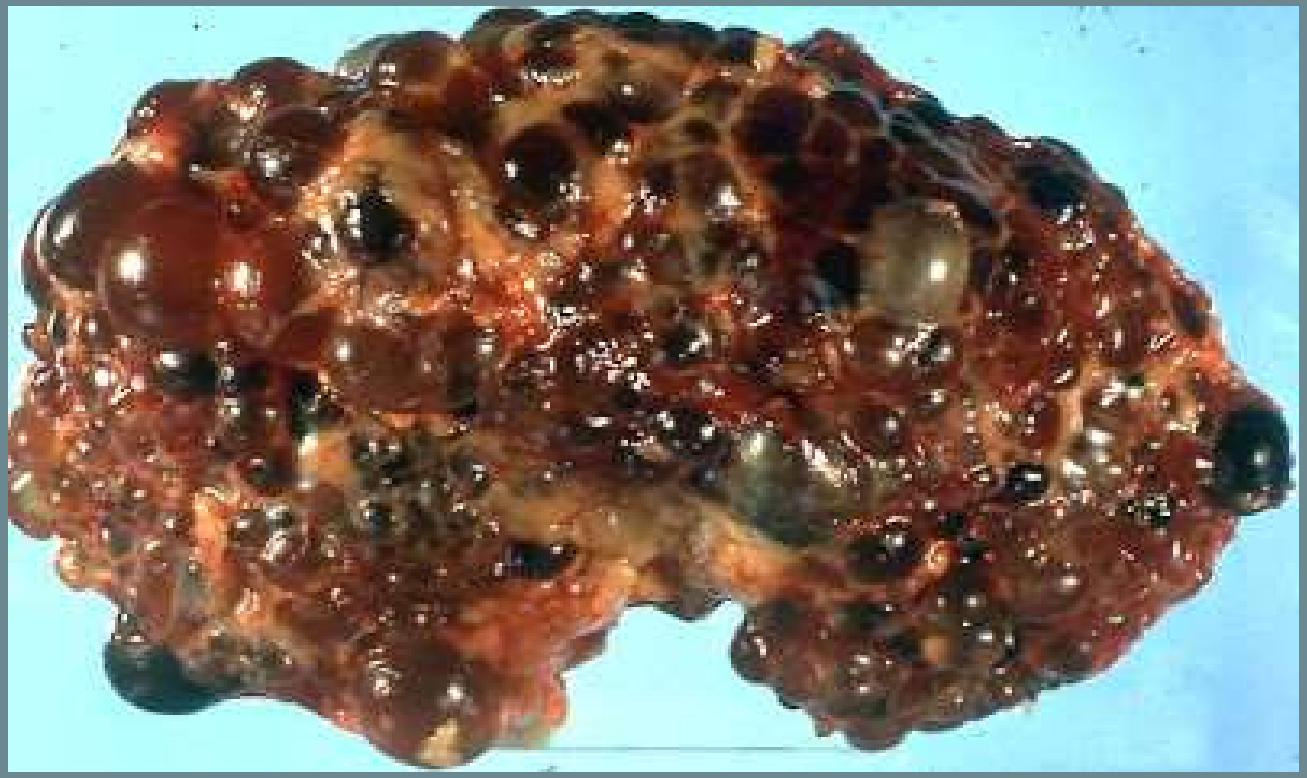

A
Right kidney
B
Left kidney
C
D

AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE (ADPKD)

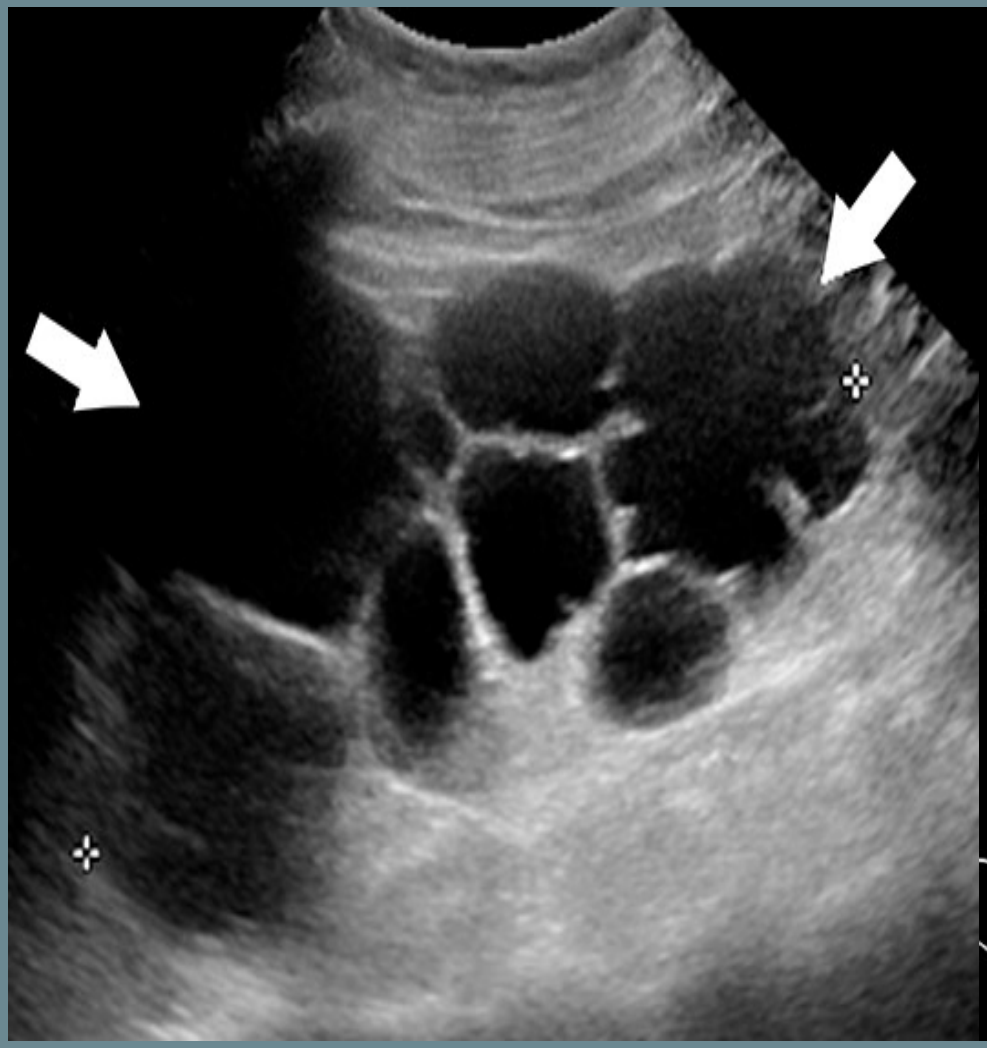
- Adult dominant form of polycystic kidney disease usually appears during middle age (30-50 yrs)
- Rarely reported in young infants
- 90% of cases inherited, 10% are sporadic
- Cysts are macroscopic of varying size; can also form in liver, spleen, pancreas
- Cerebral berry aneurysms known to occur in 10% to 15% of patients with ADPKD
- Increased incidence of RCC

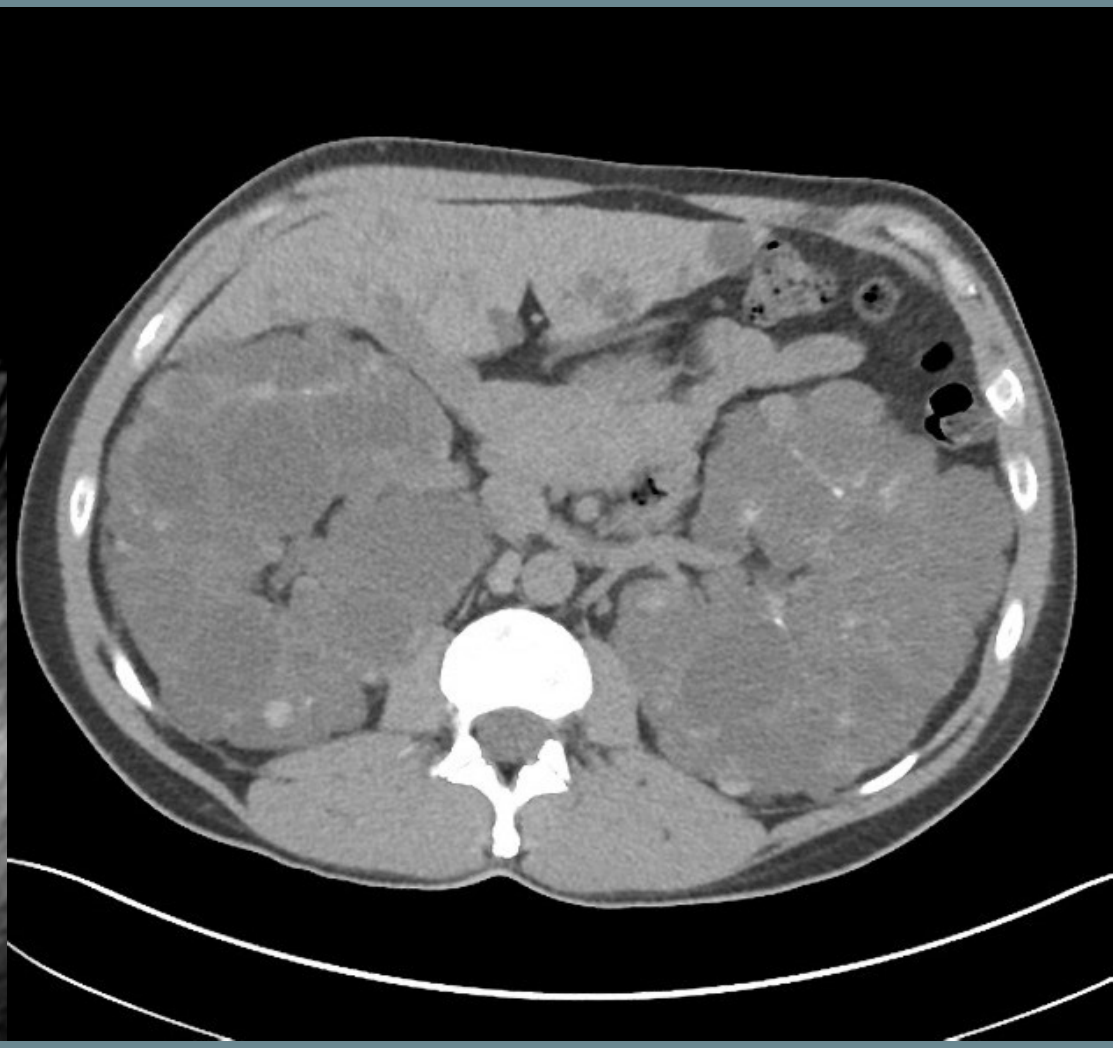
ADPKD (CONT'D)

- Clinical findings:
  - Hypertension
  - Microscopic/gross hematuria
  - Flank pain- stones in 20-30%
- Sonographic findings:
  - Enlarged, often echogenic, multicystic kidneys
  - Often difficult to distinguish borders
  - Cysts in other organs such as liver, spleen, pancreas, ovaries, etc.



https://www.renalfellow.org/2008/04/23/ultrasound-criteria-for-adpkd/

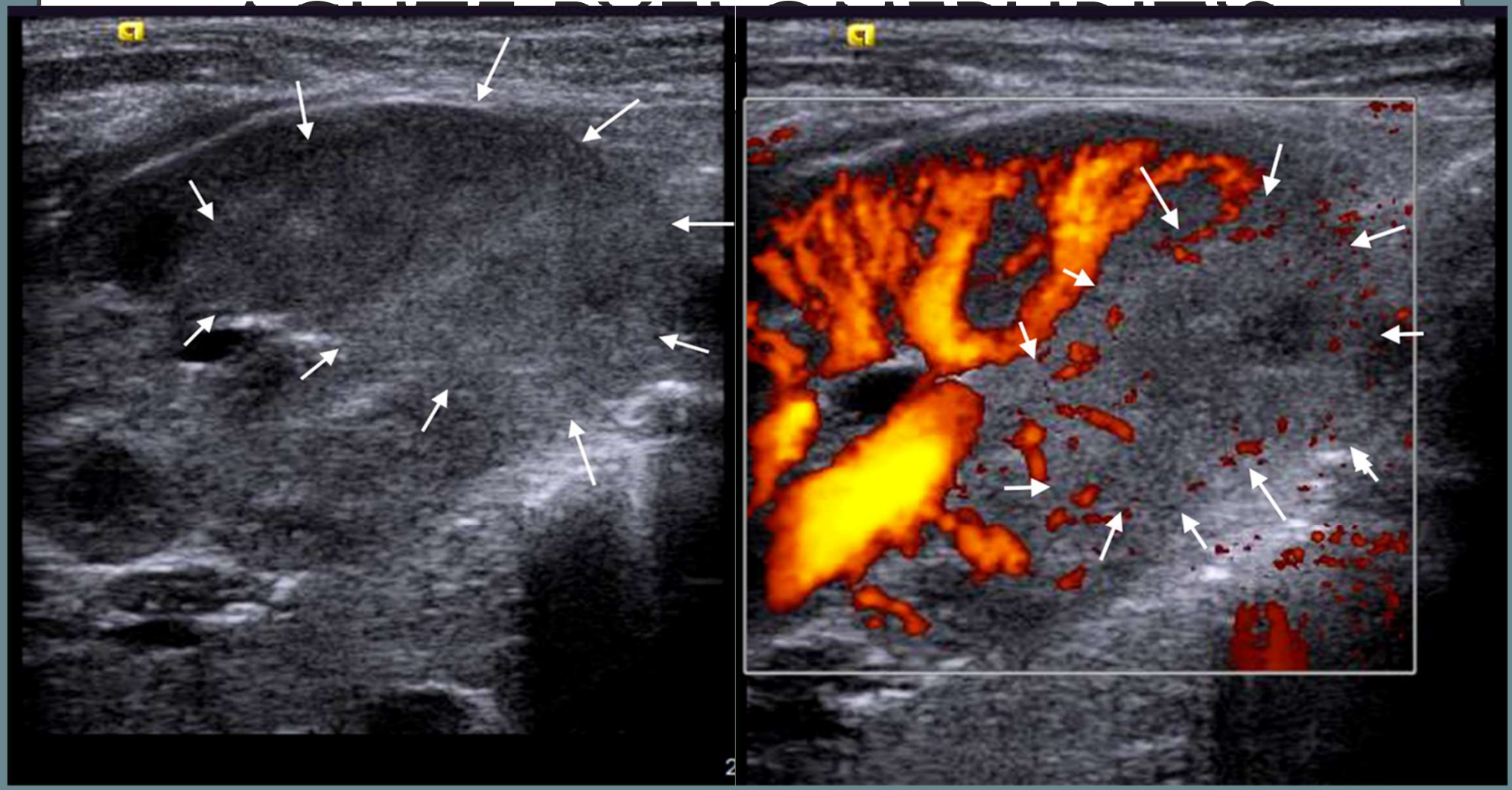




URINARY TRACT INFECTIONS (UTI)

ACUTE PYELONEPHRITIS

- Clinical symptoms include:
  - sudden fever
  - flank pain
  - tenderness
- Infection usually begins in bladder and ascends the ureter into renal pelvis
- Sonographic findings:
  - renal size may be slightly enlarged with altered renal parenchyma echogenicity secondary to edema
  - Color Doppler demonstrates absence of color in affected area of kidney



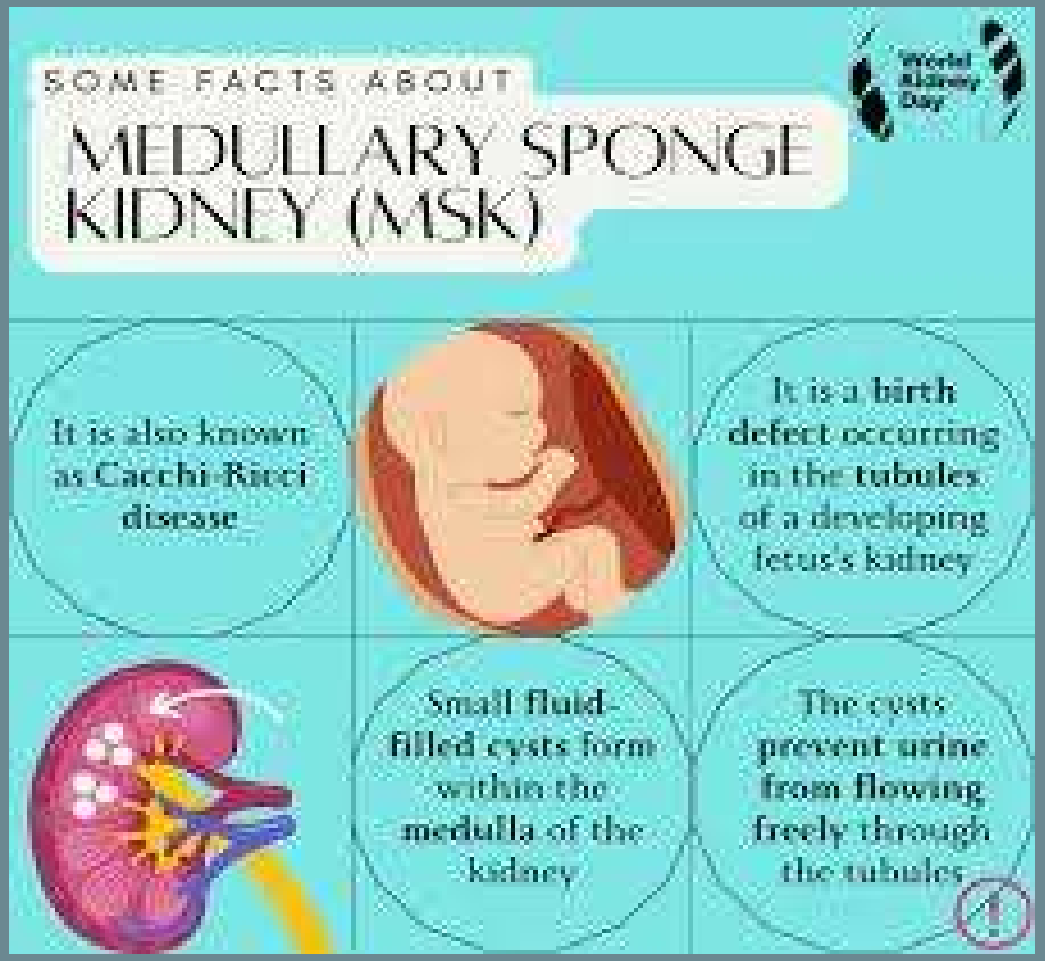
CHRONIC PYELONEPHRITIS

- Repeated episodes of acute pyelonephritis cause the kidney to become scarred and decrease in size
- Outline of the kidney may be irregular from scarring
- Renal cortex becomes more echogenic than liver parenchyma
- Renal pyramids become difficult to separate from the renal parenchyma

ACQUIRED PATHOLOGY

NEPHROCALCINOSIS

- Often in preemies (6-41%)
- Calcification of the renal parenchyma, often in the medulla and rare in the cortex
- Many underlying causes including:
  - Infants receiving long-term treatment for lung or heart disease
  - Medullary sponge kidney (Cacchi-Ricci disease)
  - Bartter syndrome



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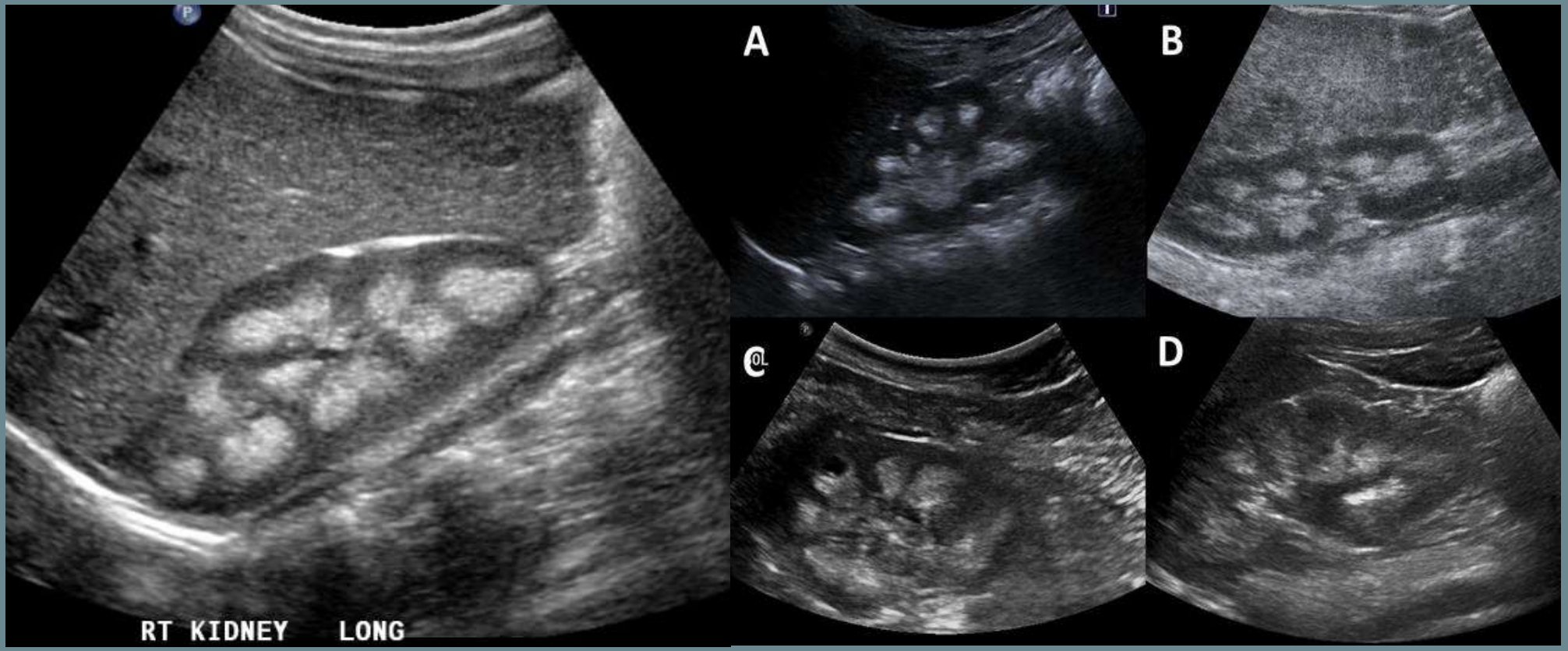


https://www.intechopen.com/chapters/67118

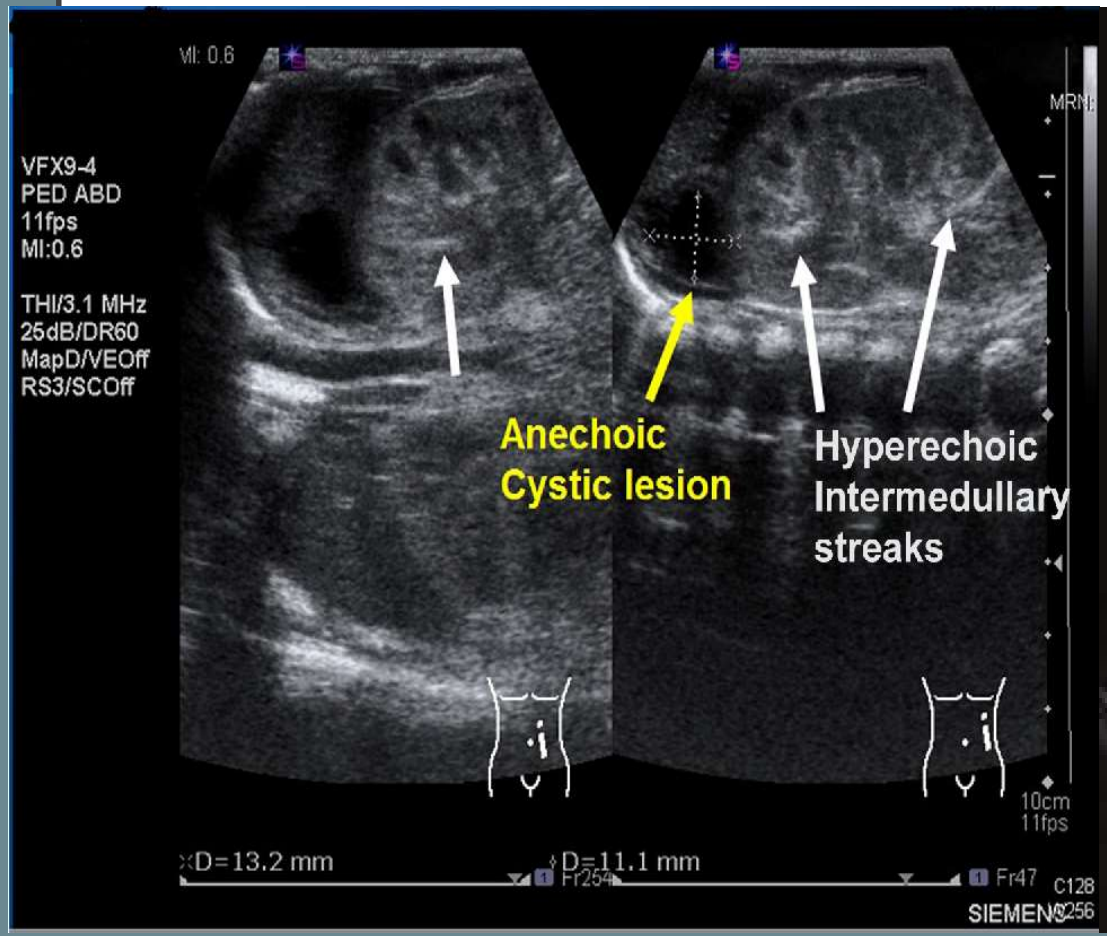
RENAL VEIN THROMBOSIS

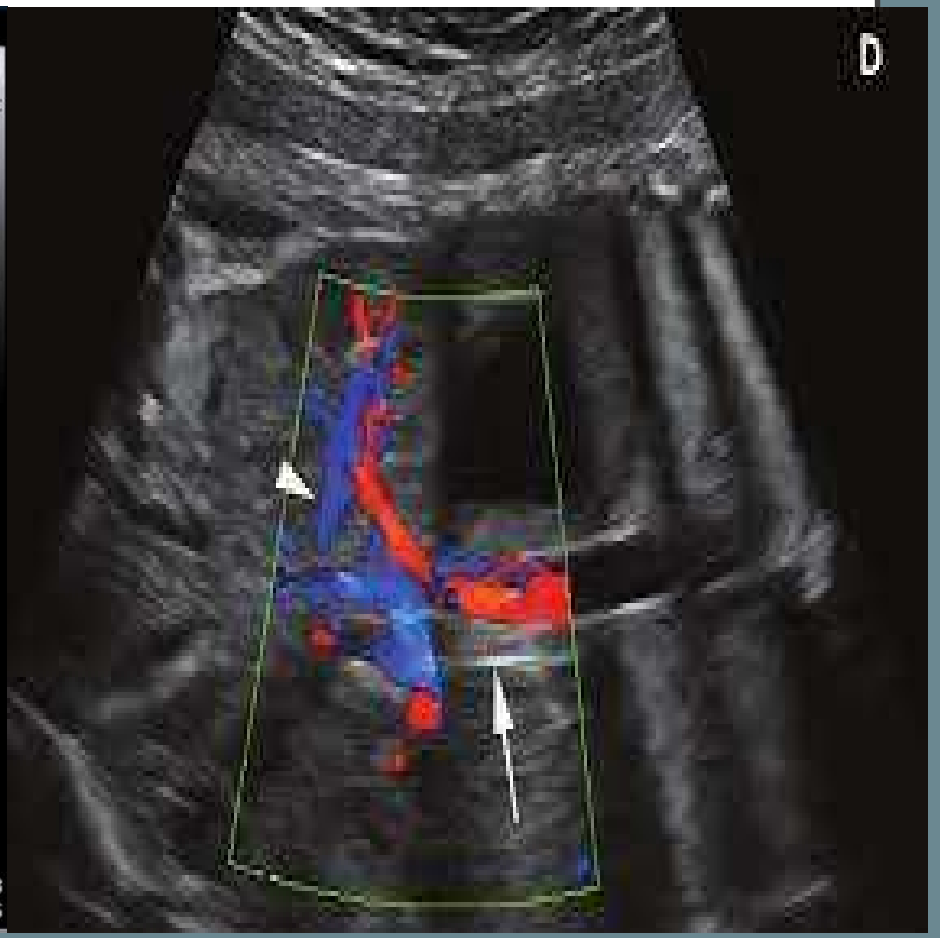
- Renal vein thrombosis is most likely to occur in dehydrated or septic infants
- More prevalent in infants born to diabetic mothers or infants with shock, glomulonephritis ,or nephrotic syndrome
- One or both kidneys may be involved
- If thrombus reaches renal vein or IVC, it may be directly visualized within the vascular lumen

RENAL VEIN THROMBOSIS (CONT'D)

- Clinical findings:
  - Hematuria
  - Low platelet count

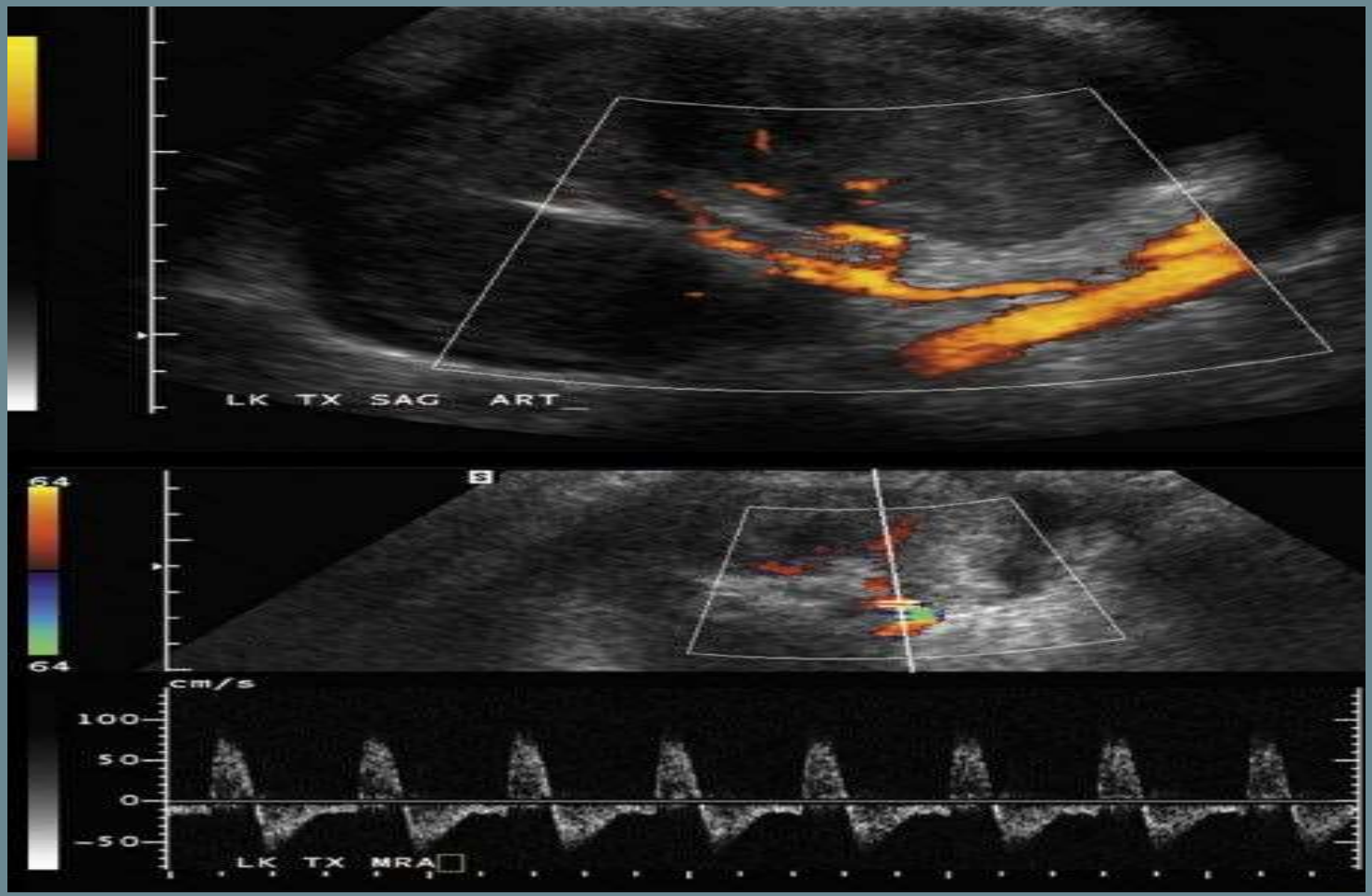
- Sonographic findings:
  - Renal enlargement
  - Intrarenal vascular calcifications
  - Renal parenchymal echogenicity
  - Coexisting adrenal hemorrhage (lt >rt)





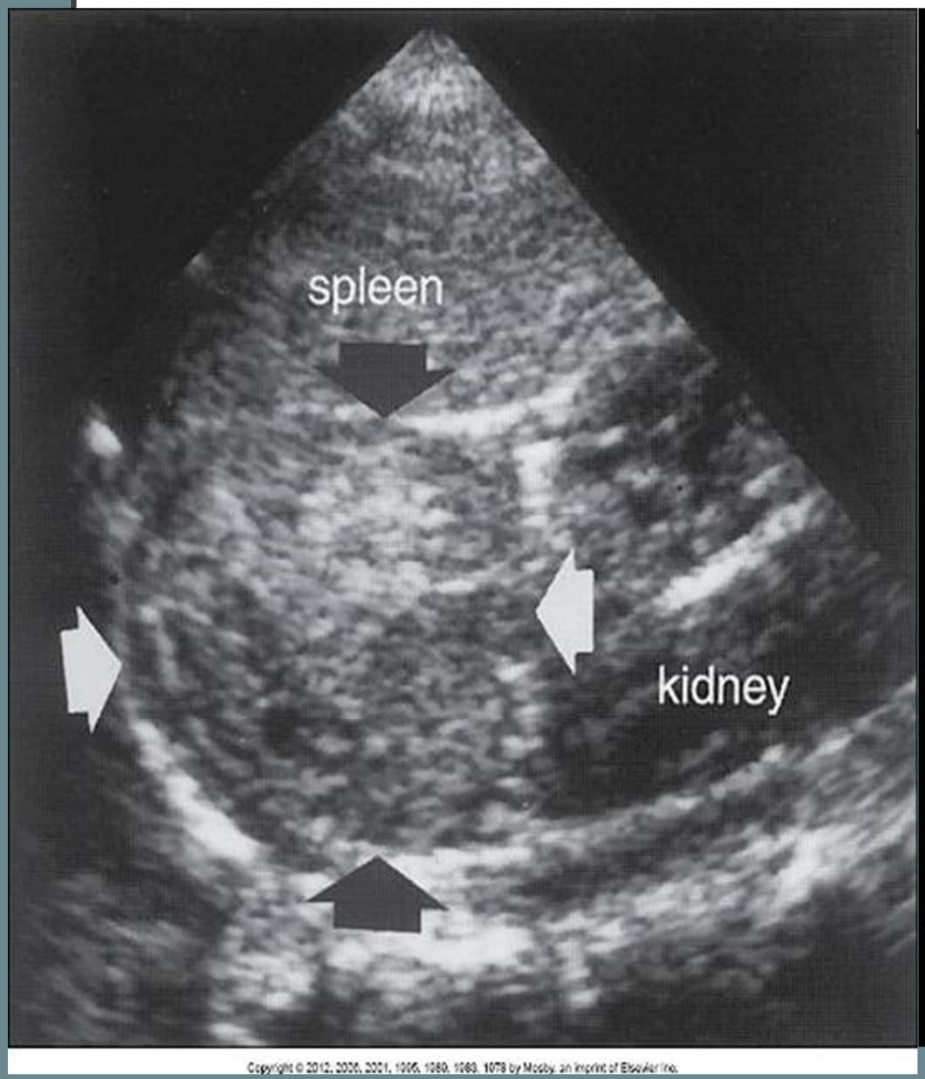
https://www.semanticscholar.org/paper/Unusual-Presentation-of-Renal-Vein-Thrombosis-in-a-Yang-Fu/e08ed0e0b00fa82c78bf6a40a0841711eb169dad

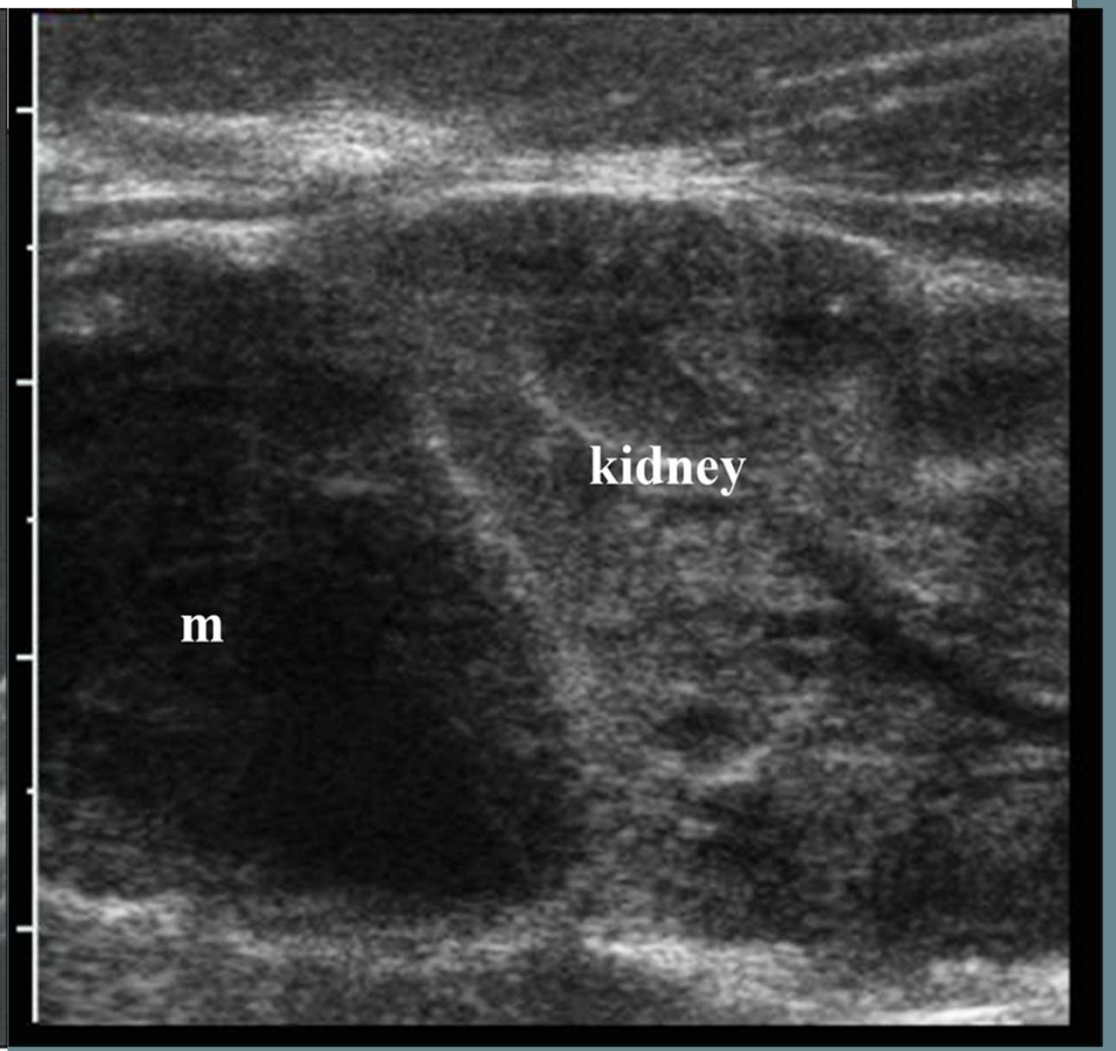
https://onlinelibrary.wiley.com/doi/abs/10.7863/ultra.34.8.1509

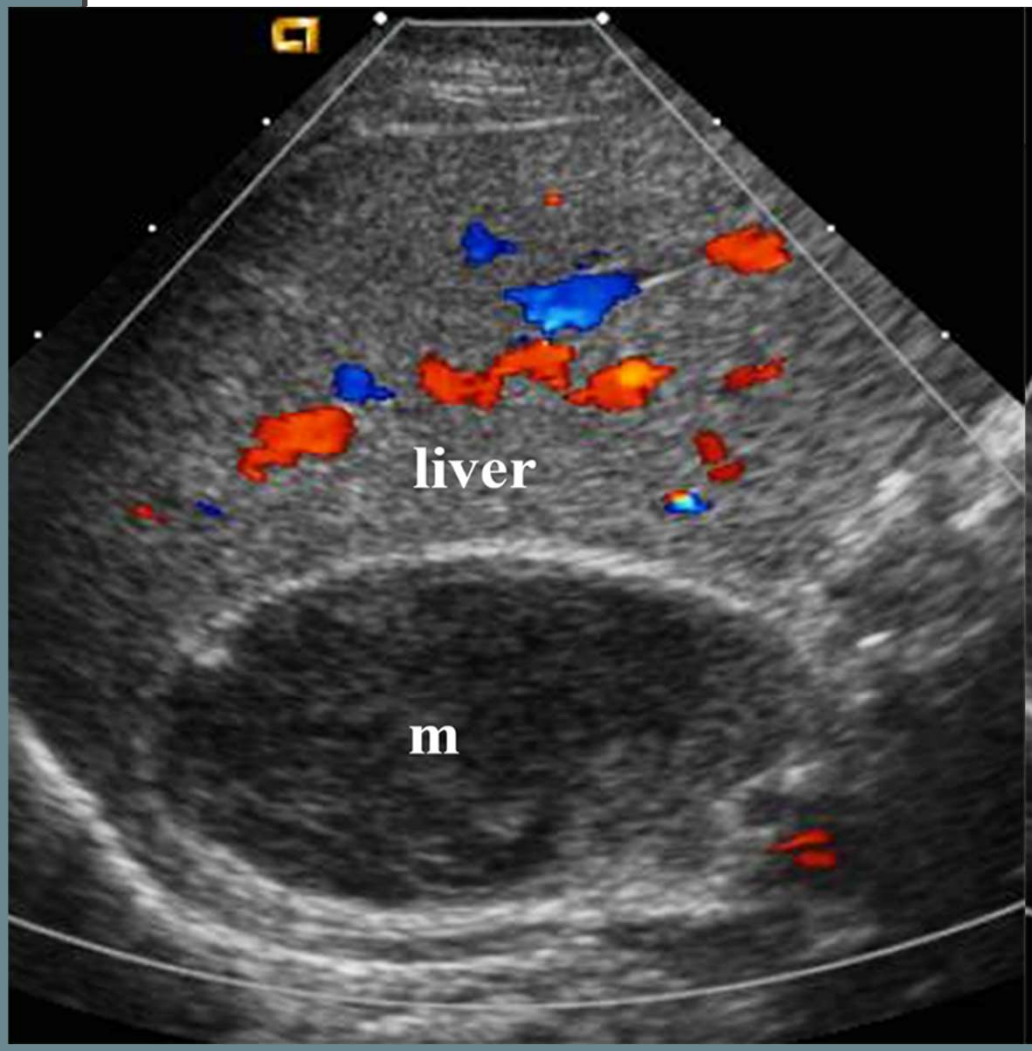

LK TX SAG ART
64
64
cm/s
100
50
0
-50
LK TX MRA

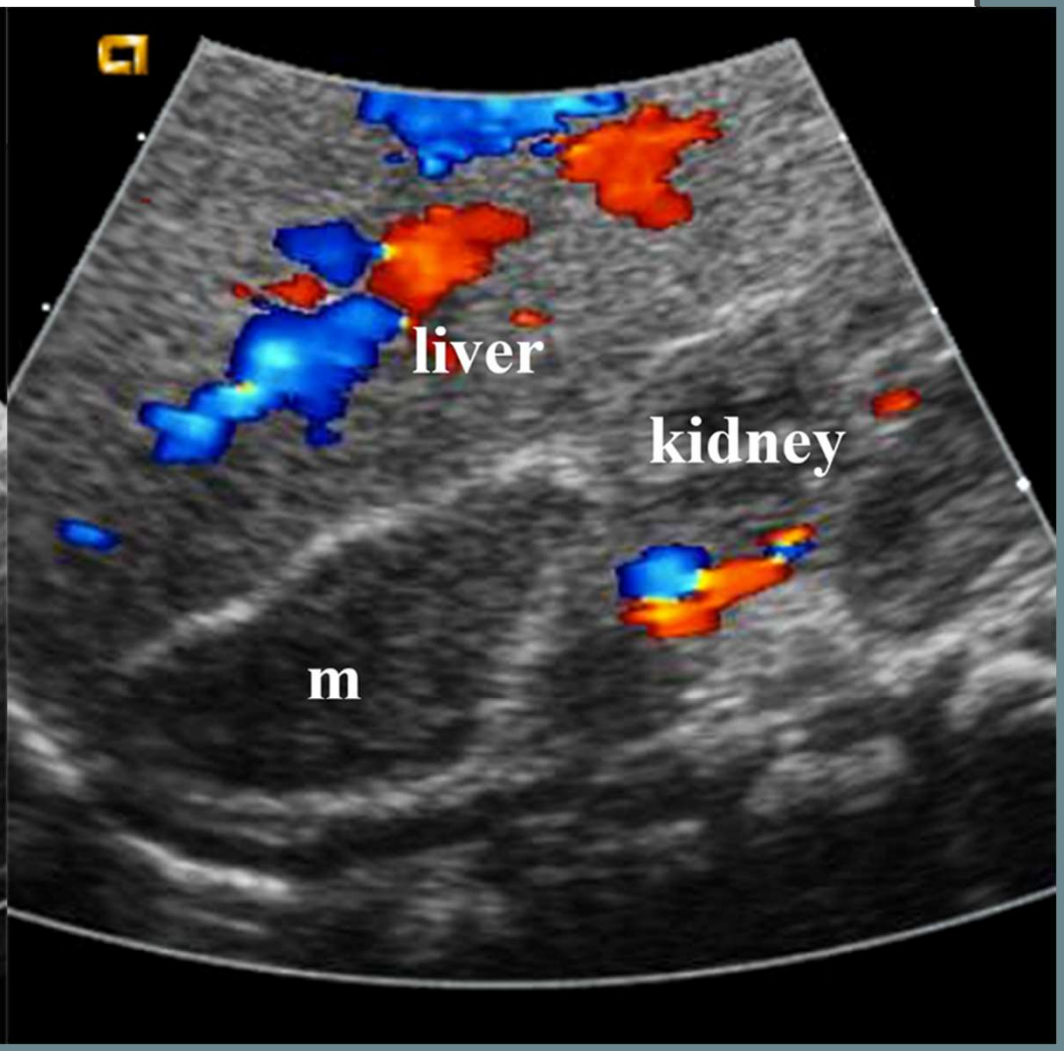
ADRENAL HEMORRHAGE

- Difficult delivery, large size, infants of diabetic mothers, stress, hypoxia at delivery, septicemia, and shock are all predisposing factors
- Sonographic findings:
  - Ovoid enlargement of gland or portion of gland
  - Appearance can range from anechoic to hyperechoic, or may be a mix of echogenicity depending on age and severity of process






liver
m


liver
kidney
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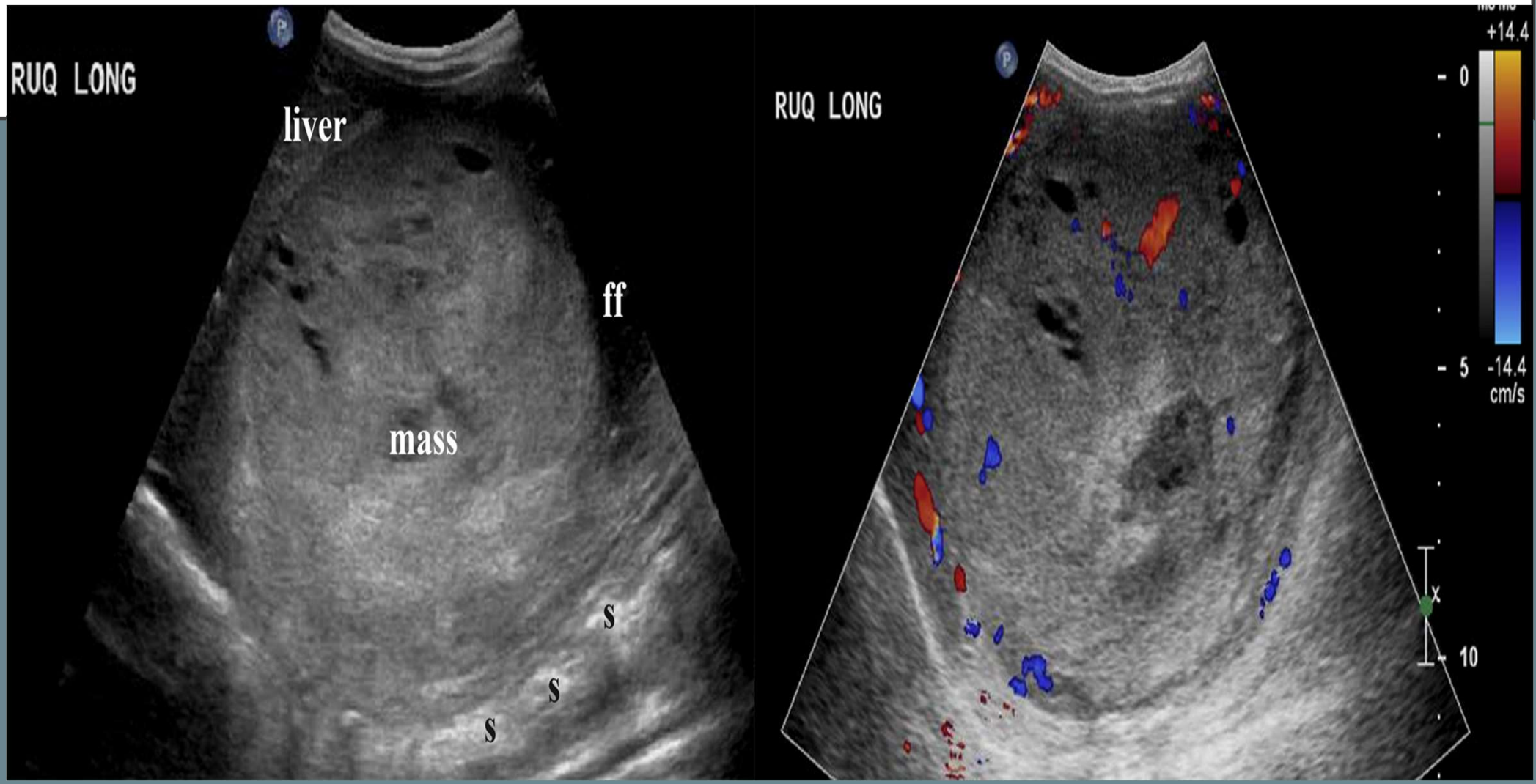
NEPHROBLASTOMA (WILMS TUMOR)

*Most common intraabdominal malignant renal tumor in young children
  - peaks between 2 and 5 years
- Usually unilateral
- Sonography can detect tumoral spread into renal vein, IVC, right atrium, and contralateral kidney
- Spreads through renal sinus and peripelvic soft tissues, lymph nodes in renal hilum, and paraaortic areas

WILMS TUMOR (CONT'D)

- Sonographic findings:
  - Bulky, solid mass that can will distort renal sinus, pyramids, cortex, contour of kidney
  - Hydronephrosis may be present
  - Variable echotexture, areas of echogenicity and calcifications within
  - May demonstrate liquefaction if necrosis and hemorrhage occur


RUQ LONG
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+14.4
0
5
-14.4
cm/s
10

WILMS TUMOR (CONT'D)

- Predisposing factors:
  - hepatoblastoma
  - pancreatoblastoma
  - Beckwith-Wiedemann syndrome
- Wilms tumor may also occur spontaneously
- Differentials include:
  - neuroblastoma
  - congenital mesoblastic nephroma

CONGENITAL MESOBLASTIC NEPHROMA

- Although rare, it's the most common renal tumor of neonate
- Benign, indistinguishable from Wilms
- Sonographic findings:
  - Solid lesion with mixed echogenicity
  - Mass may extend through renal capsule into retroperitoneum
  - Found in children <1 year of age (Wilms occurs in children older than one year)

NEUROBLASTOMA

*Most common malignancy in children under one year

- Malignant tumor arises in adrenal medulla
- May be detected on antenatal sonography or at birth
- Second most common abdominal tumor of childhood, occurring between 2 months and 2 years

NEUROBLASTOMA (CONT'D)

- Sonographic findings:
  - Highly echogenic
  - Calcification may be identified
  - Small tumors are homogeneous, large tumors are more complex
  - Adjacent kidney is displaced inferiorly
  - Use Doppler to distinguish between neuroblastoma and adrenal hemorrhage

OTHER MALIGNANT RETROPERITONEAL TUMORS

- Renal cell carcinoma
- Rhabdomyosarcoma
- Adrenal malignant tumors
  - Pheochromocytoma
  - Adrenocortical carcinomas

GENITALIA MALFORMATIONS:

- Hypospadias
  - abnormal opening of the urethra along the ventral aspect of the penis
- Cryptoorchidism
  - undescended testicle(s)
- Hermaphrodites
  - ambiguous genitalia
  - both ovaries and testicles present
- Guevedoces
  - born male, look like a female, grow penises at puberty
  - lack enzyme that produces dihydrotestosterone
  - occurs in a village in DR (also in Turkey and New Guinea)

- https://www.pbs.org/video/9-months-made-you-jonnys-story/
- https://www.bbc.com/news/magazine-34290981

THINK QUESTIONS…..

1. Discuss the pitfalls a sonographer may encounter in a fetus with renal agenesis.
2. In a fetus with multicystic dysplastic kidney, what should the sonographer look for in the contralateral kidney?
3. If hydronephrosis is noted on the obstetric ultrasound examination, what should be examined by the sonographer?
4. Why is it important to document the fetal gender on an ultrasound examination?