The Fetal Urogenital System

Chapter 64

Embryology

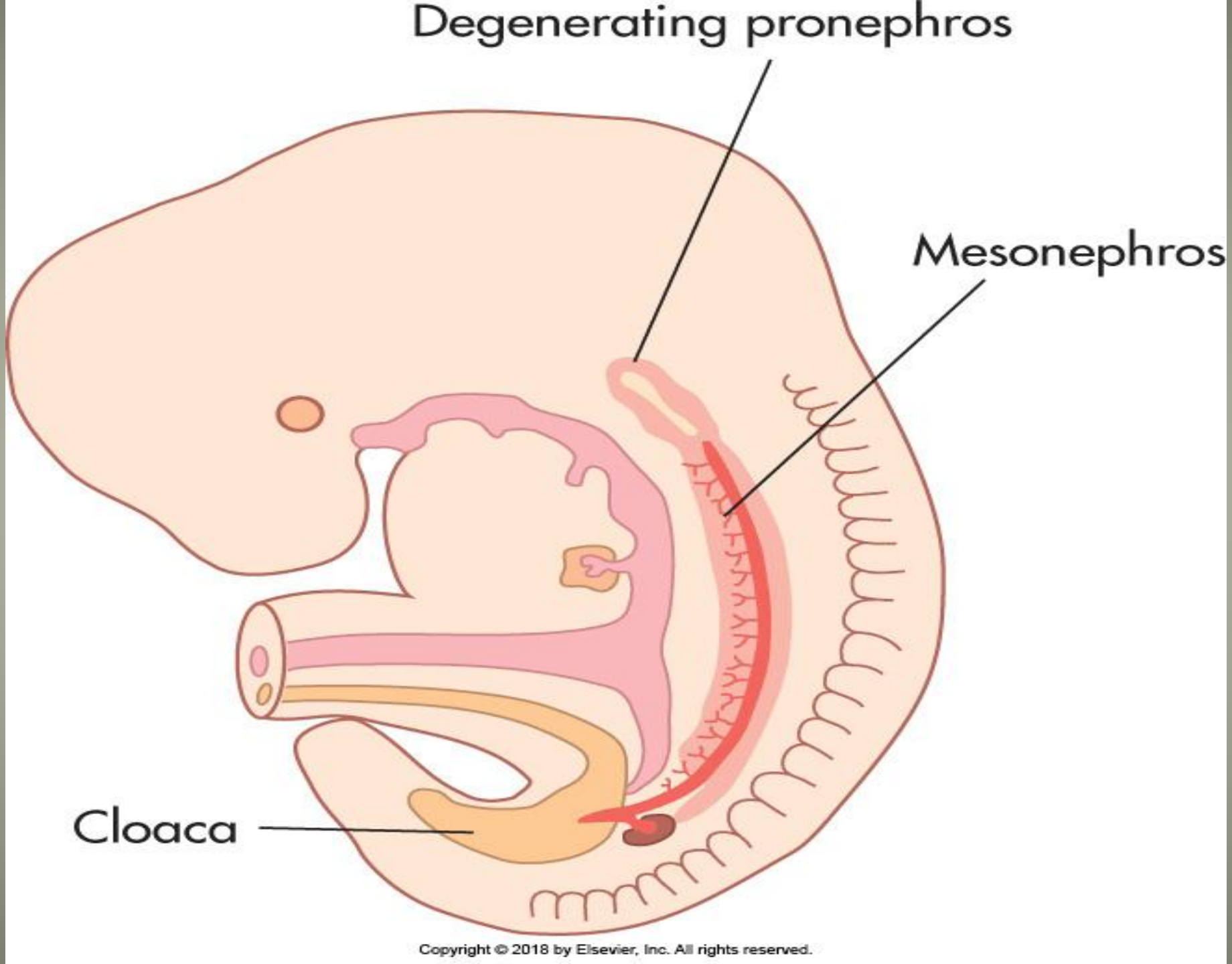
- Systems developed from intermediate mesoderm
  - Urinary
  - Genital
- Overlapping of the two systems well seen in male system
- Mesonephric duct
  - First serves as urinary duct
  - Later is transformed into main genital duct
    - *Vas deferens*

Embryology

- Intermediate mesoderm forms longitudinal mass on both sides of the aorta
  - "Urogenital ridge"
- Both urinary/genital systems develop in these ridges
- Urinary system is the first to develop

Kidney Development

- Three sets of excretory organs develop in the embryo
  - Third set remains as the “permanent kidneys”
- Permanent kidneys (metanephros)
  - Begins to develop in the fifth week
  - Begins to function and produce urine around the 11th week



Kidney Development

- Urine formation continues to fill amniotic cavity throughout fetal life
- Kidneys do not eliminate waste in utero
  - Placenta eliminates waste from fetal blood
- Permanent kidneys develop from two different sources:
  - 1. Metanephric diverticulum or ureteric bud
    - Gives rise to
      - Ureter, renal pelvis, calyces, and collecting tubules
  - 2. Metanephric mesoderm

Kidney Development

- Kidneys initially lie very close together in pelvis
- Gradually they migrate into the abdomen
  - Become separated from one another
  - Normally complete migration by ninth week
- In some cases
  - One kidney remain in the pelvic cavity
    - Other migrates into posterior flank

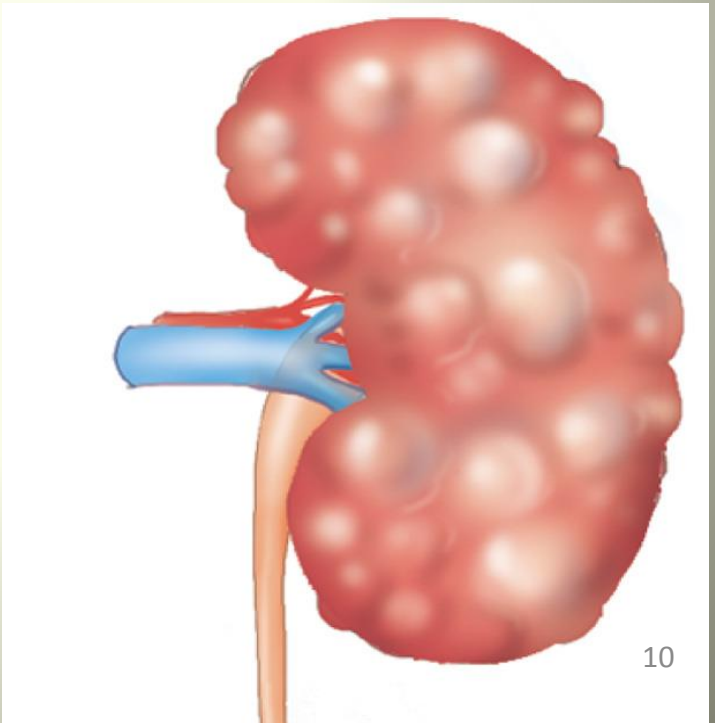
Congenital Malformations of the Kidneys

Renal Agenesis

- Complete absence of the kidneys
- Occurs when
  - Ureteric buds fail to develop
  - Kidneys degenerate before they can induce the metanephric mesoderm to form nephrons

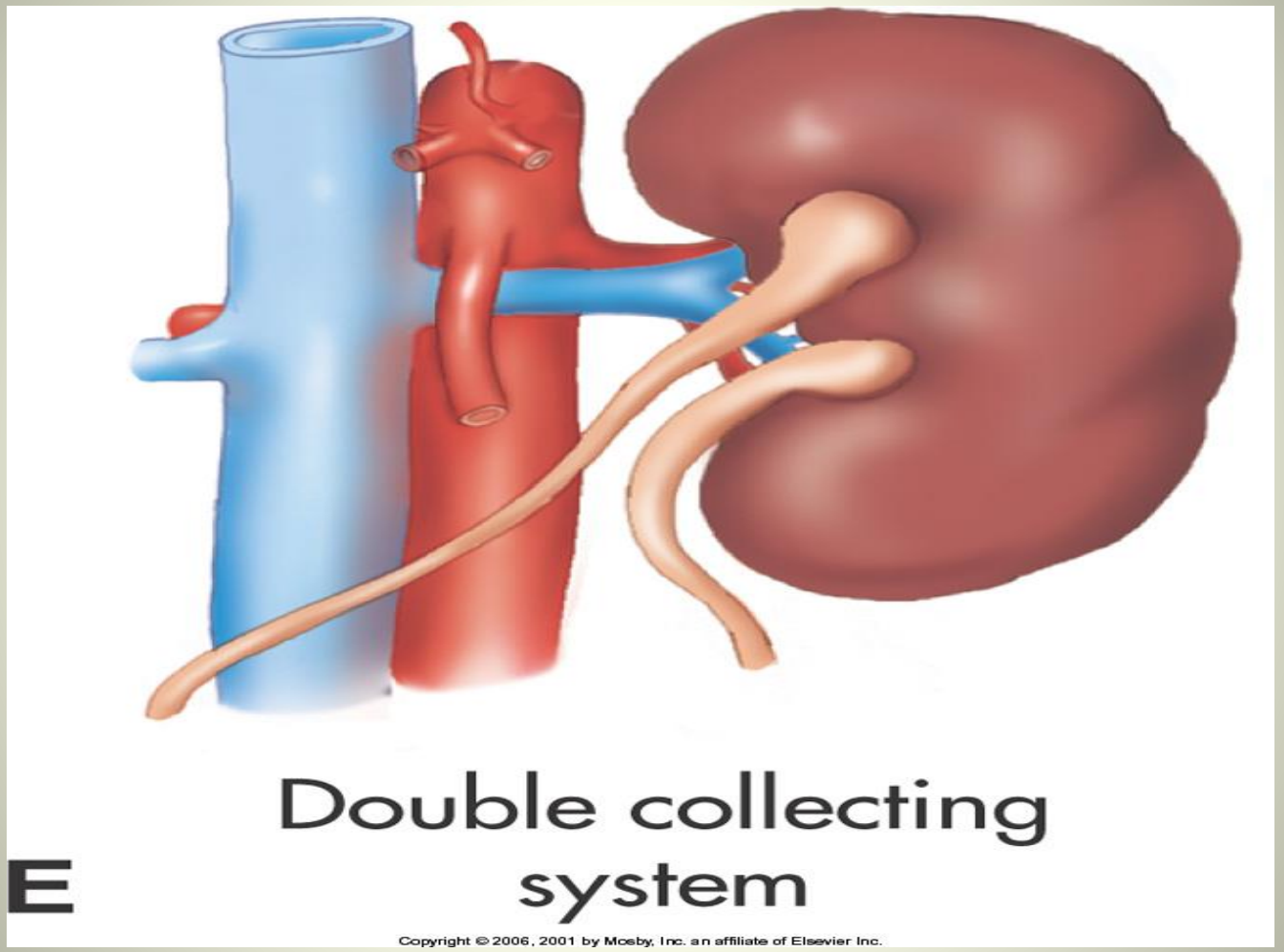
Congenital Polycystic Disease

- Uriniferous tubules develop from two different sources
- Failure of the tubules to join
- Each uriniferous tubule consists of two parts:
  - Nephron
  - Collecting tubule



Double or Divided Kidney

- Results from division of the ureteric bud at an early stage
  - Ureteral bud closest to the sinus drains the lower renal pole and enters the bladder at the trigone
  - Ureter that drains the upper pole enters the bladder more medial and caudal (ectopic ureter)
- Lower pole more prone to reflux
- Upper pole more prone to obstruction



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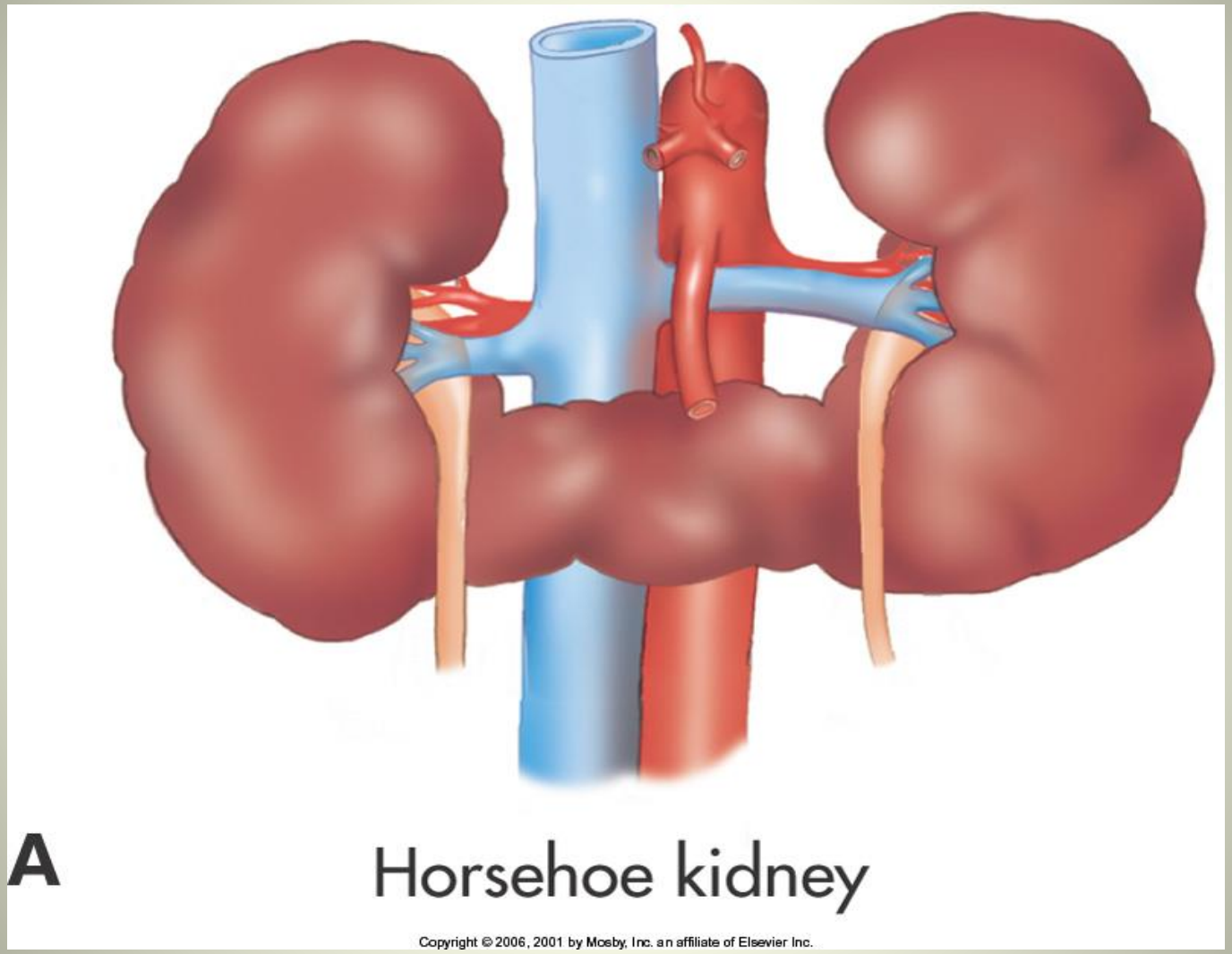
Double collecting system

Horseshoe Kidney

- Forms when the inferior poles of the kidney fuse while they are in the pelvis
- May occur in 1 to 4 per 1000 births
- More common in males



A

Horsehoe kidney

Crossed Renal Ectopia
Abnormal Renal Position

- Kidney located on the opposite side of its ureteral orifice
- Usually affects the left kidney
  - –Kidney goes towards the right
- Positioned inferior to the normal kidney

Bladder Development

- Derived from the hindgut derivative
  - Known as the urogenital sinus
- As the kidneys migrate upward
  - Orifices of the ureters also move cranially
  - Ejaculatory ducts (derived from the mesonephric ducts) move toward one another and enter the prostatic part of the urethra

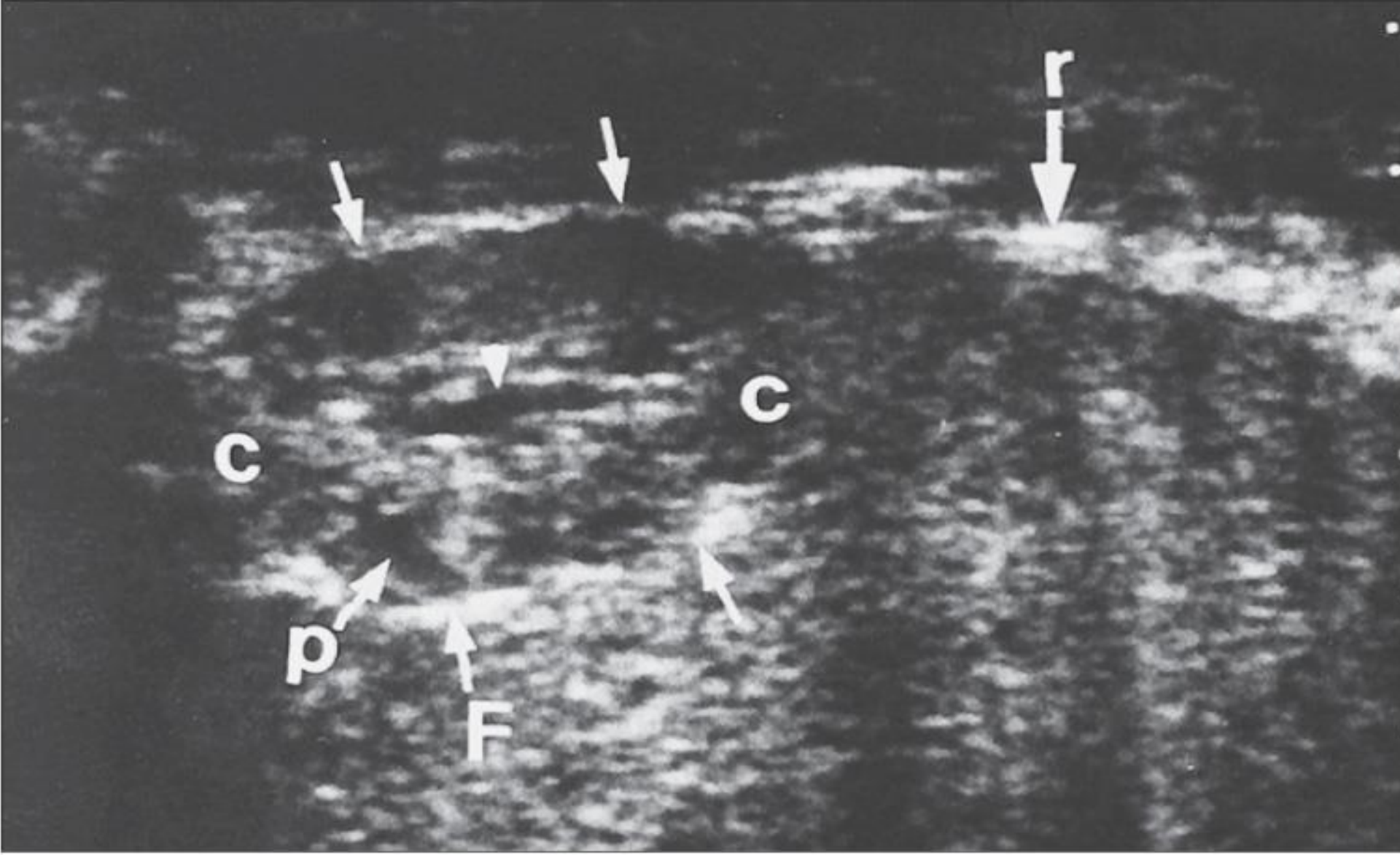
Bladder Exstrophy

- Occurs primarily in males
  - 1 in 30,000 births
- Characterized by protrusion of the posterior wall of the bladder
  - (Contains the trigone of the bladder and ureteric orifices)
- Caused by defective closure of the inferior part of the anterior abdominal wall during the fourth week
- As a result
  - No muscle or connective tissue forms in the anterior abdominal wall to cover the urinary bladder
  - Bladder is formed external to the abdominal wall

Sonographic Evaluation of the Urinary Tract

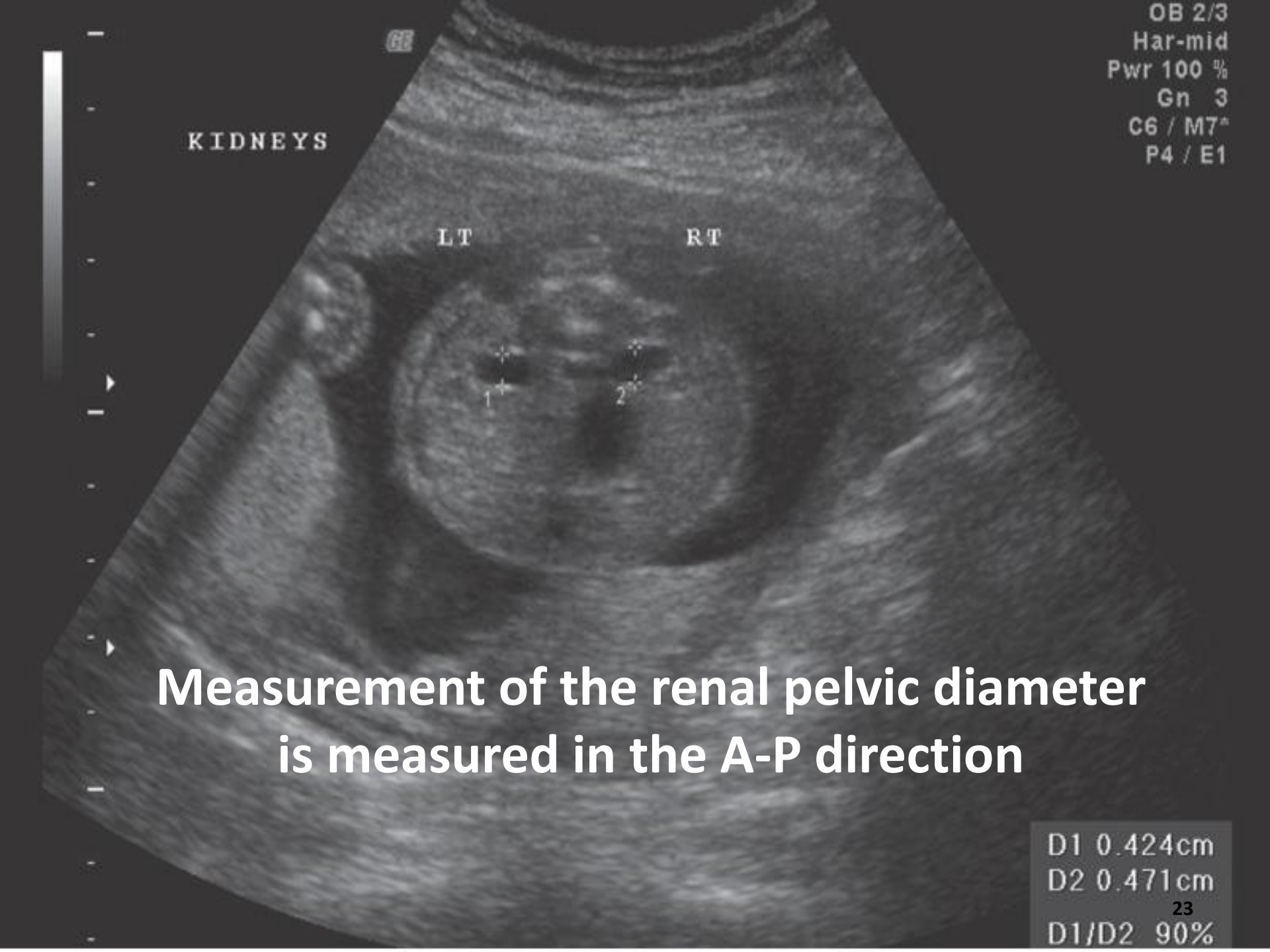
- **Endovaginal**
  - **9 weeks**
    - **Kidneys have been documented**
- **Transabdominal**
  - **13 weeks**
    - **Kidneys and bladder may be seen**
      - **Kidneys appear as bilateral hyperechoic structures in the paravertebral regions**
      - **Renal pelvises can be seen as sonolucent areas within the central kidney**
      - **Bladder imaged as rounded echo-free area centrally in the pelvis**

- **25 weeks**
  - **Possible to:**
    - **Distinguish the renal cortex from the medulla**
    - **Outline the renal capsule clearly**
    - **See central echogenic area in the renal sinus region**
- **Kidneys should be evaluated by assessing their:**
  - **Anatomy**
  - **Texture**
  - **Size**
- **Normal anatomic structures include:**
  - **Relatively homogeneous renal cortex and parenchyma**
  - **Echogenic pyramids and calyces**
  - **Anechoic renal pelvis**
- **Kidney texture that appears significantly echogenic should be a cause for concern**



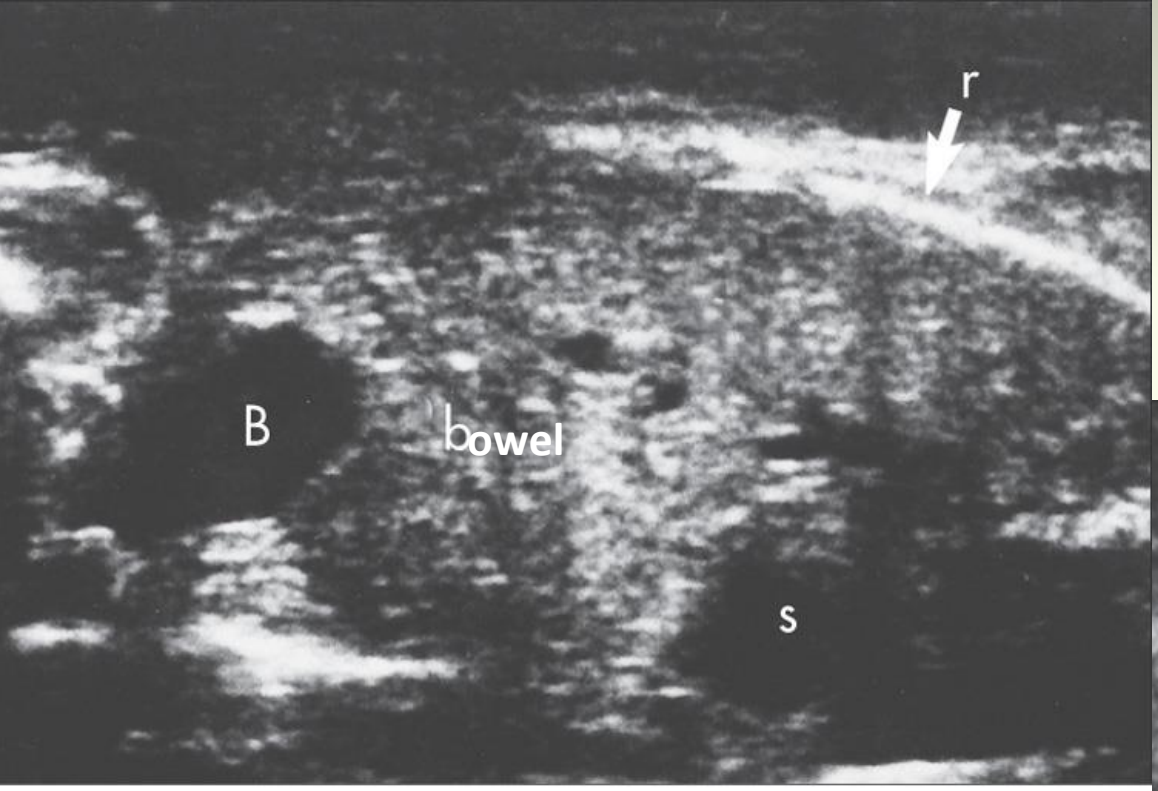
Cortex *(c),* pelvis *(arrowhead)*, and pyramids *(p)*,capsule of the kidney *(arrows)*, perirenal fat *(F). r,* Rib

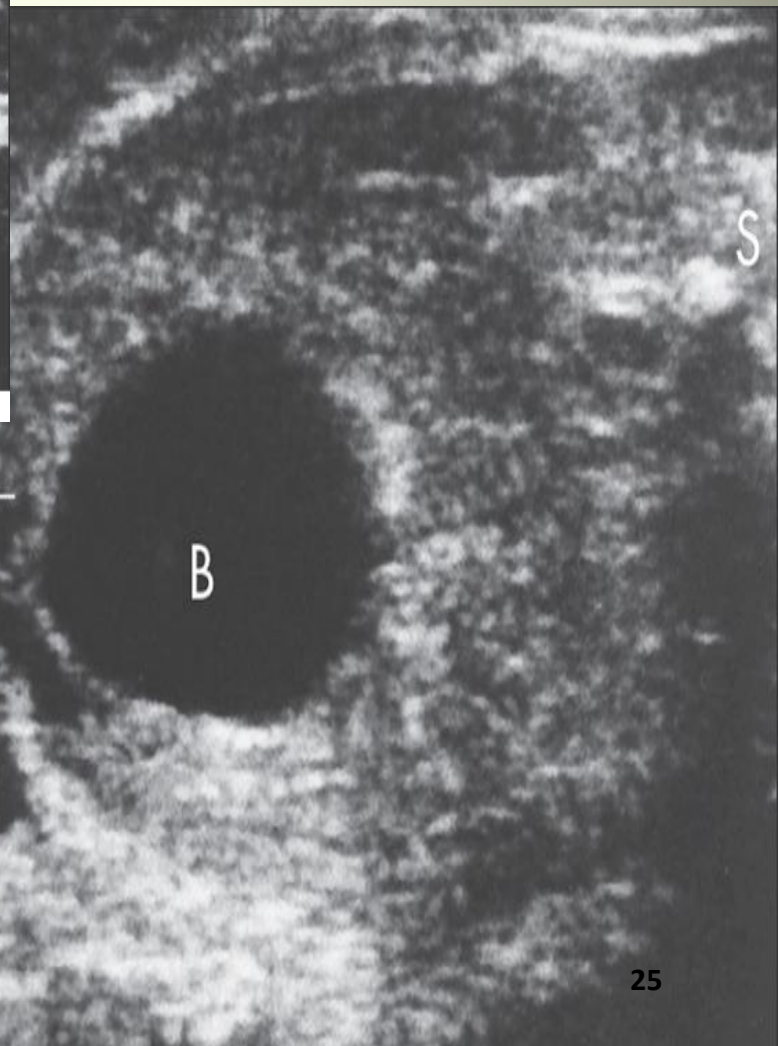
- **Small amount of urine may be seen in the renal pelvis in the normal fetus**
  - **AP diameter <4 mm before third trimester**
  - **AP diameter <7 mm from the third trimester until term**
  - **Should not be misinterpreted as an abnormal collection of urine within the renal pelvis**
    - ***Pelviectasis* (5–9 mm)**
    - ***Caliectasis* (> 10 mm)**
      - **Rounded calyces with renal pelvis dilation**
      - **May lead to severe hydronephrosis**
- **AP Renal pelvis diameter**
  - **>10 to 15 mm is considered abnormal**



Measurement of the renal pelvic diameter is measured in the A-P direction

- Bladder normally visualized in all fetuses
- If bladder appears too large:
  - Evaluated again at the end of the study to see if normal emptying has occurred
    - (Assuming examination takes at least 45–60 minutes)
- Failure to observe bladder may indicate severe renal abnormality
  - When oligohydramnios is present
- Bladder wall should be thin in a normal fetus
- Bladder wall becomes hypertrophied
  - When obstruction occurs at the level of the urethra
    - Ureteral jets may rule out obstruction





C

Abnormalities of the Urinary Tract

Renal Malformations

- Divided into two categories:
    1. Congenital malformation
    2. Resulting from obstructive process
- Consequences of renal malformations vary
  - Depends on
    - Type of lesion
    - Extent of obstruction

Amniotic Fluid

- Critical marker in assessment of renal function
- Kidneys excrete urine after the 11th week
  - Not a major contributor of fetal urine until the 14 - 16 weeks
    - Amniotic fluid volume does not increase until 14-16 weeks
  - Normal amniotic fluid volume before this time does not exclude the possibility of renal agenesis

Amniotic Fluid

- With severe renal disease
  - Amniotic fluid is reduced
- Most severe malformations
  - Absent
- When severe oligohydramnios is seen
  - Usually both kidneys or ureters and the urethra are malformed
- With unilateral obstructions
  - Normal amount of amniotic fluid may be seen
    - Contralateral kidney produces urine

Fetal Bladder

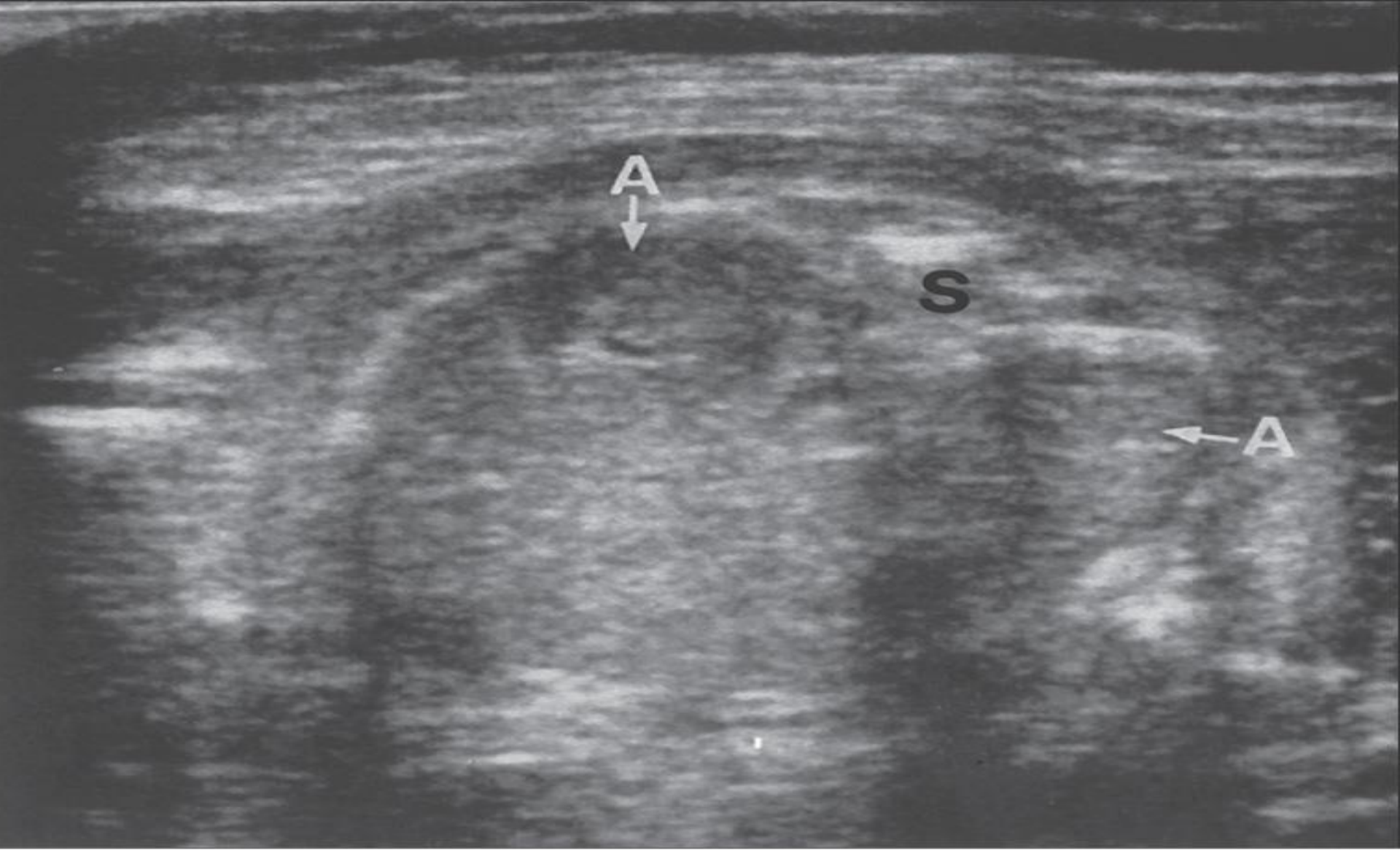
- Important to identify early in the exam to make sure adequate fluid is present
- Bladder not seen:
  - Reevaluate the bladder at the end of the examination
  - Usually takes at least 30 minutes to fill and empty the fetal bladder

Abnormalities of Kidney Development

- **Incompatible with life**
  - **Renal agenesis**
  - **Infantile polycystic kidney disease**
- **Immediate neonatal death**
  - **Bilateral Multicystic dysplastic kidneys**

Renal Agenesis

- Absence of the kidneys
  - Bilateral agenesis occurs in 1 in 3,000 to 1 in 10,000 births
  - Kidneys and bladder are not visualized sonographically
  - Amniotic fluid is absent or severely decreased
    - Urine is not being produced
- Important to remember in early stages
  - Fluid may be visible because it is produced from other fetal sources
- Adrenal glands may be large and may mimic the kidneys



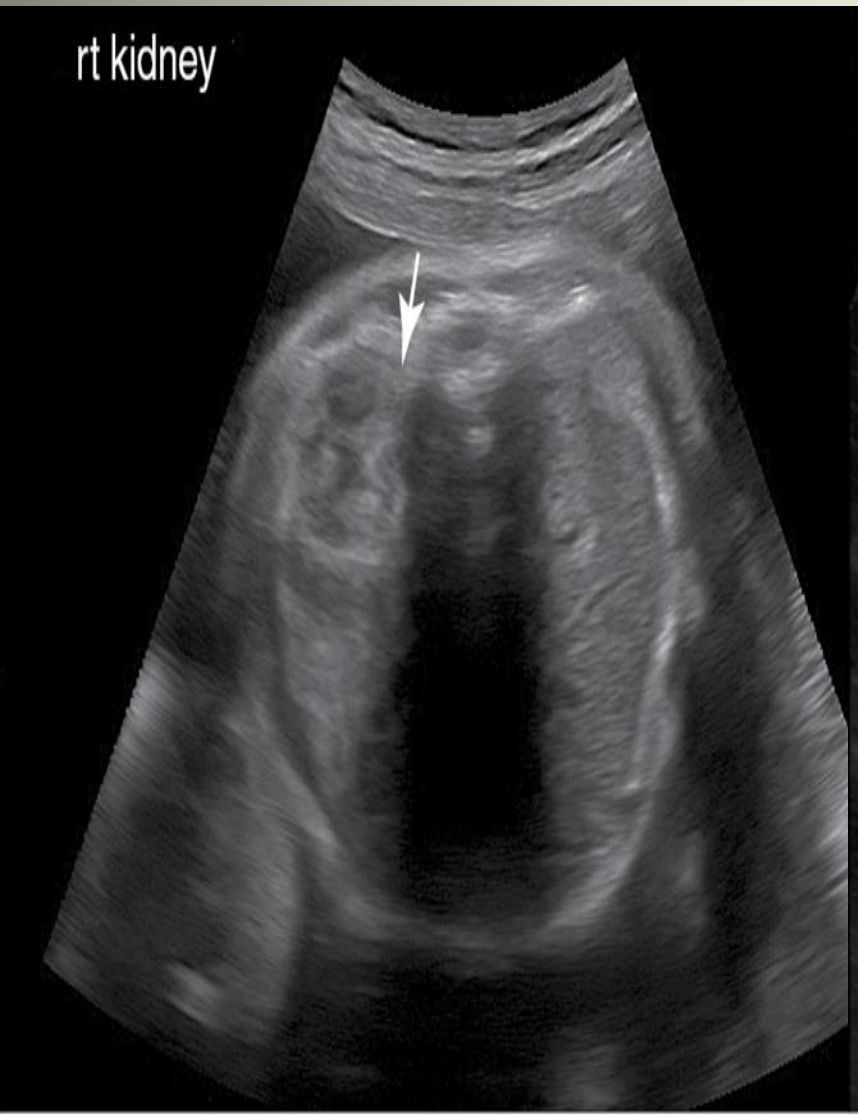
Enlarged adrenal glands *(A)* occupying the renal spaces

Sonographic Findings

- Severe oligohydramnios after 13 to 15 weeks' menstrual age
- Persistent absence of urine in fetal bladder
- Failure to visualize kidneys
  - (use color flow to outline renal arteries)
- Abnormally small thorax
- Fetal pelvis should be carefully scanned when kidneys cannot be found in their normal locations

Unilateral Renal Agenesis

- Occurs in 1 in 600 to 1 in 1000 births more commonly than bilateral disease
- May be associated with
  - Uterine anomalies in females
  - Testicular hypoplasia
  - Hypospadias
- Contralateral kidney may be hypertrophied
  - Compensate for the absent kidney
  - Adequate amounts of amniotic fluid are produced and chances for survival are excellent



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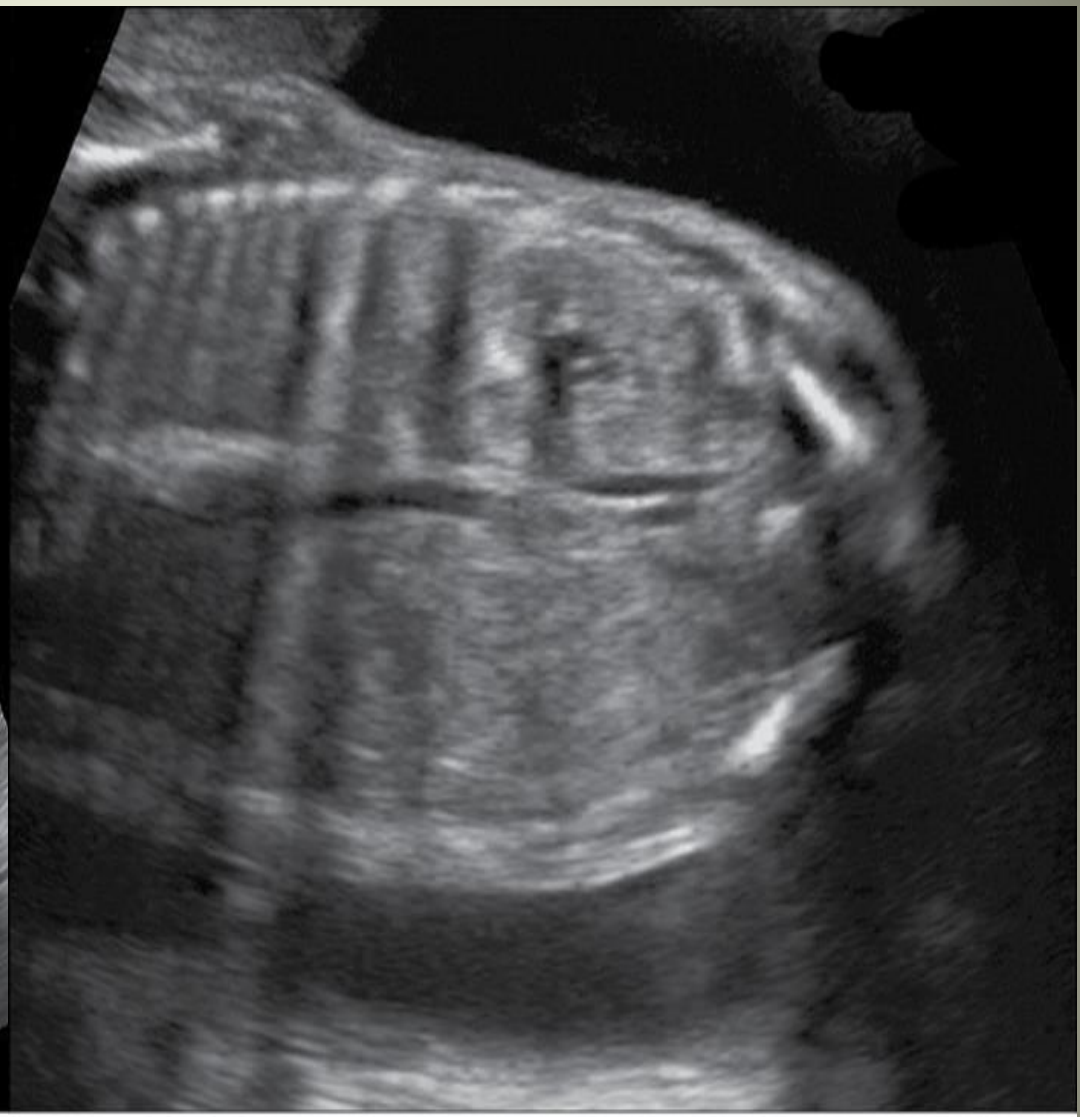


B

Horseshoe Kidney

- Fusion of lower poles of both kidneys
- Transverse images of fetal abdomen demonstrate abnormal lie of the kidney
- If spine is down
  - Connecting isthmus may be seen anterior to the aorta

Renal Ectopia

- Kidney lies outside normal position
  - Usually in the pelvis
- Occasionally, crossed fused ectopia occurs
  - Both kidneys found on the same side
    - Right side most common
- Adrenal gland may fill the renal fossa
- Evaluate the pelvis
  - Kidney lying superior to the bladder

Potter's Sequence

- Characterized by
  - Renal agenesis
  - Oligohydramnios
  - Pulmonary hypoplasia
  - Malformed hands and feet
- Can be unilateral or bilateral

Classifications of Cystic Renal Anomalies

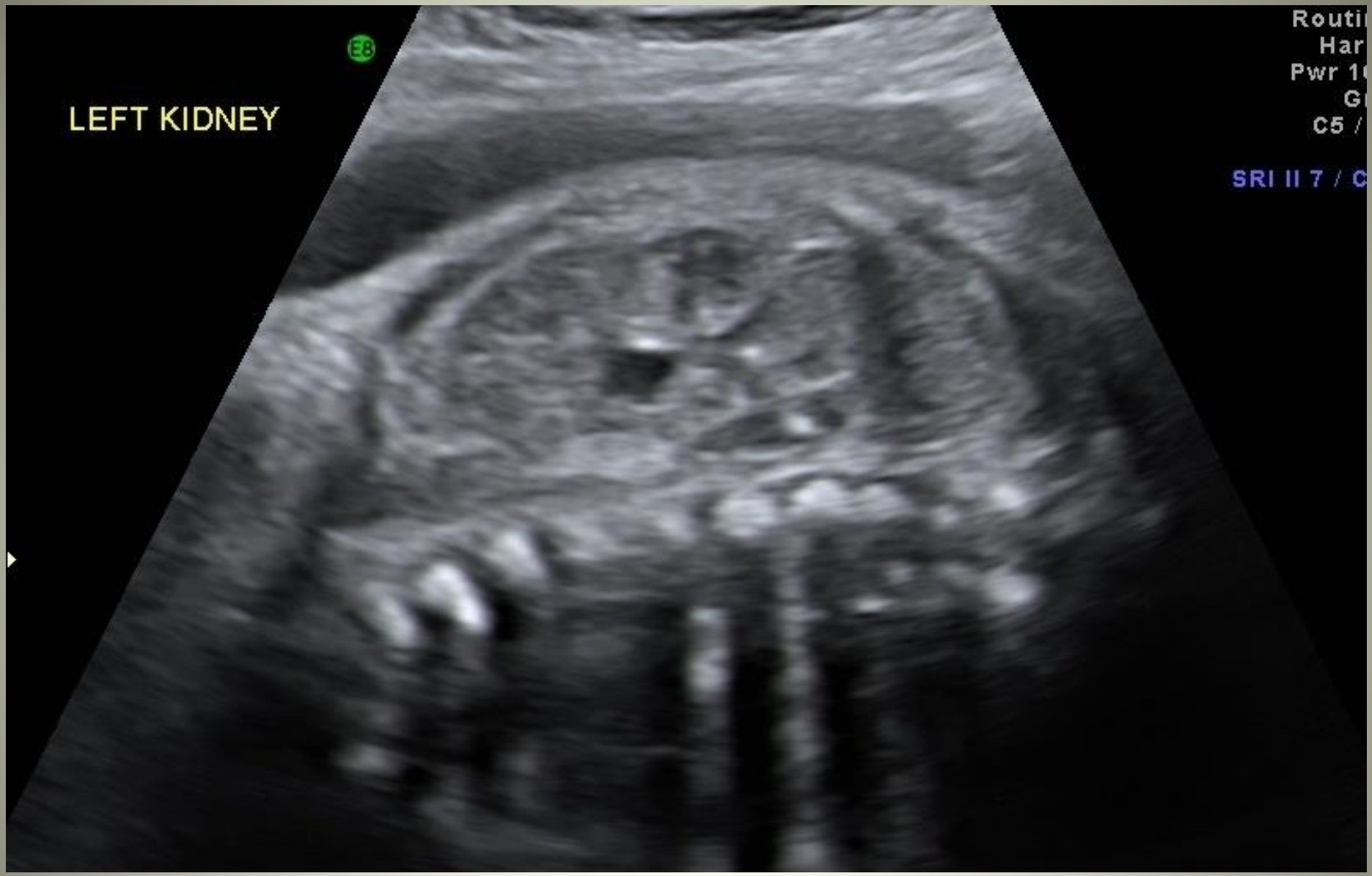
- **Potter described:**
- **Type 1: (AR) infantile polycystic kidney disease**
- **Type 2: Renal agenesis**
  Multicystic kidneys
  Renal dysplasia
- **Type 3: (AD) polycystic kidney disease**
- **Type 4: Renal dysplasia**
  obstructive kidney disease

Type I

- Infantile polycystic kidney disease (IPKD)
  - Autosomal-recessive disorder
    - (25% chance of recurrence)
  - Affects the fetal kidneys and liver
- Most severe forms of IPKD are those found prenatally
- Individual cysts are not identified sonographically
  - Kidneys are enlarged because of dilated tubules
  - Enlargement may not occur until the 24th week

Type I
Sonographic Findings

- Progressive renal enlargement
- Echogenic renal parenchyma
- Empty bladder and oligohydramnios
- Some cases - kidneys are so massive they fill the entire abdomen



IPKD

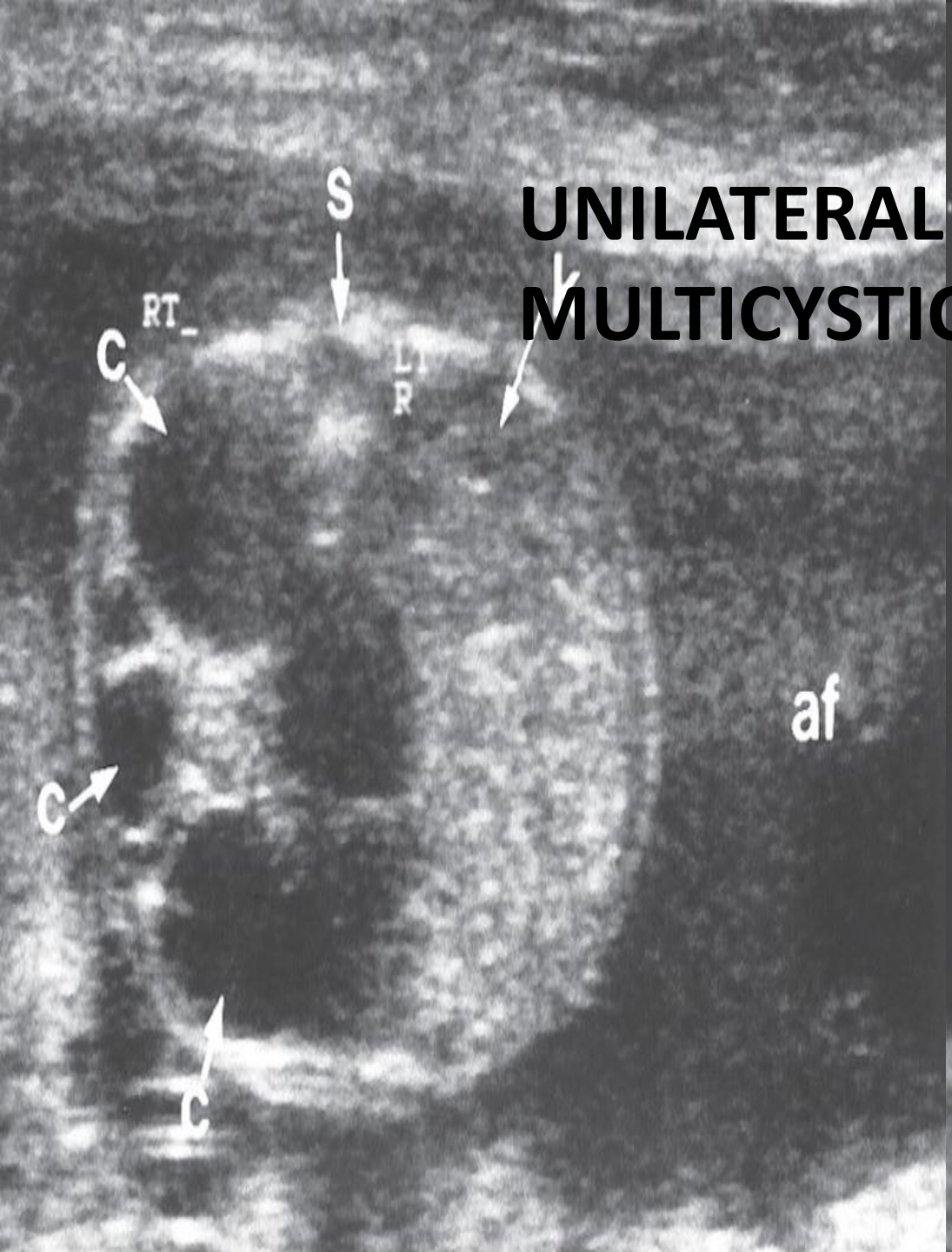
Type II

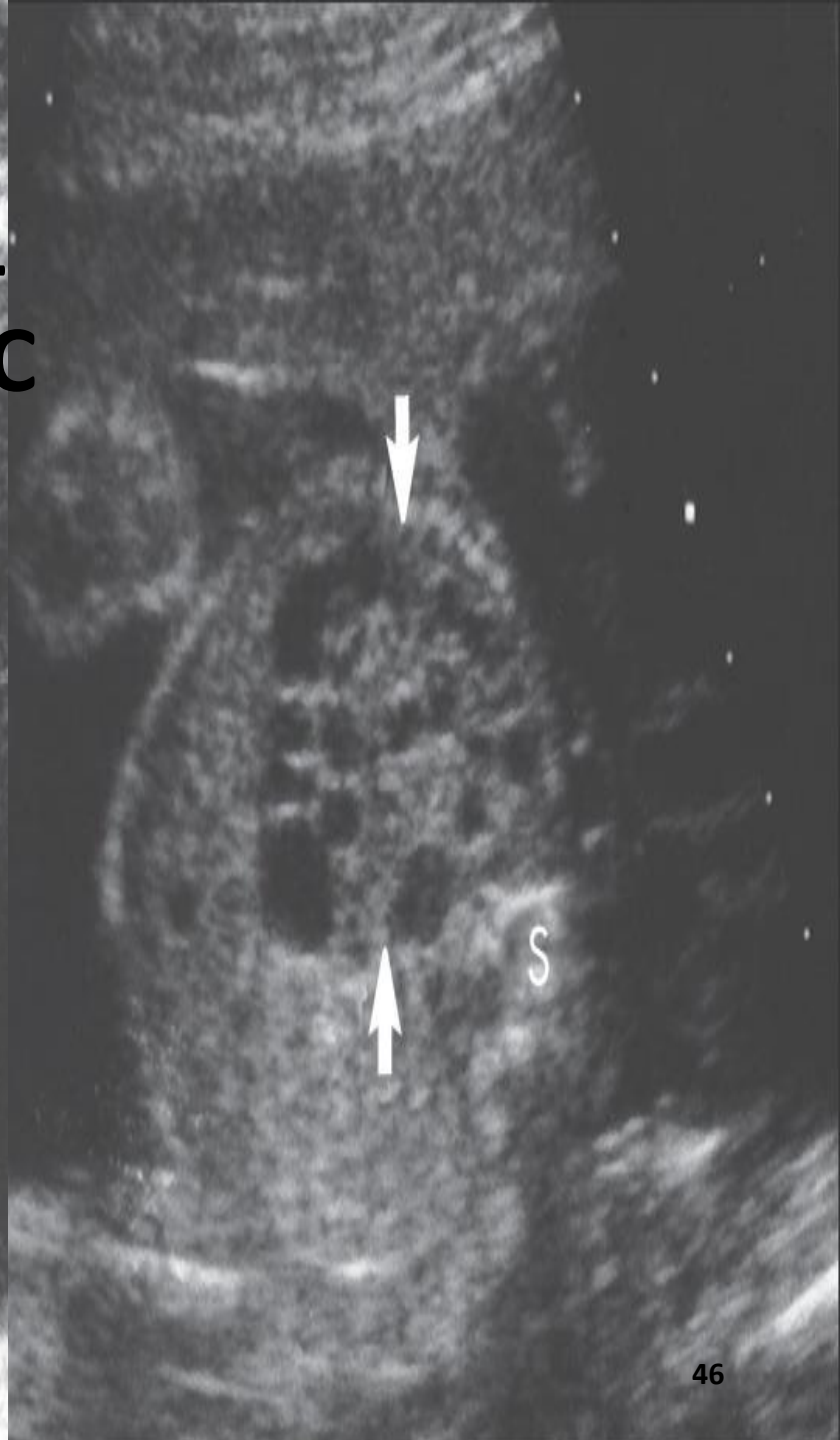
- Multicystic Dysplastic Kidney Disease
- Renal tissue is replaced by cysts of varying sizes found throughout the kidney
- Borders difficult to define because of the distorted renal outline
- Affected kidney is nonfunctional
- When multicystic kidney identified
  - Search for anomalies of opposite kidney
- Lethal condition for the neonate

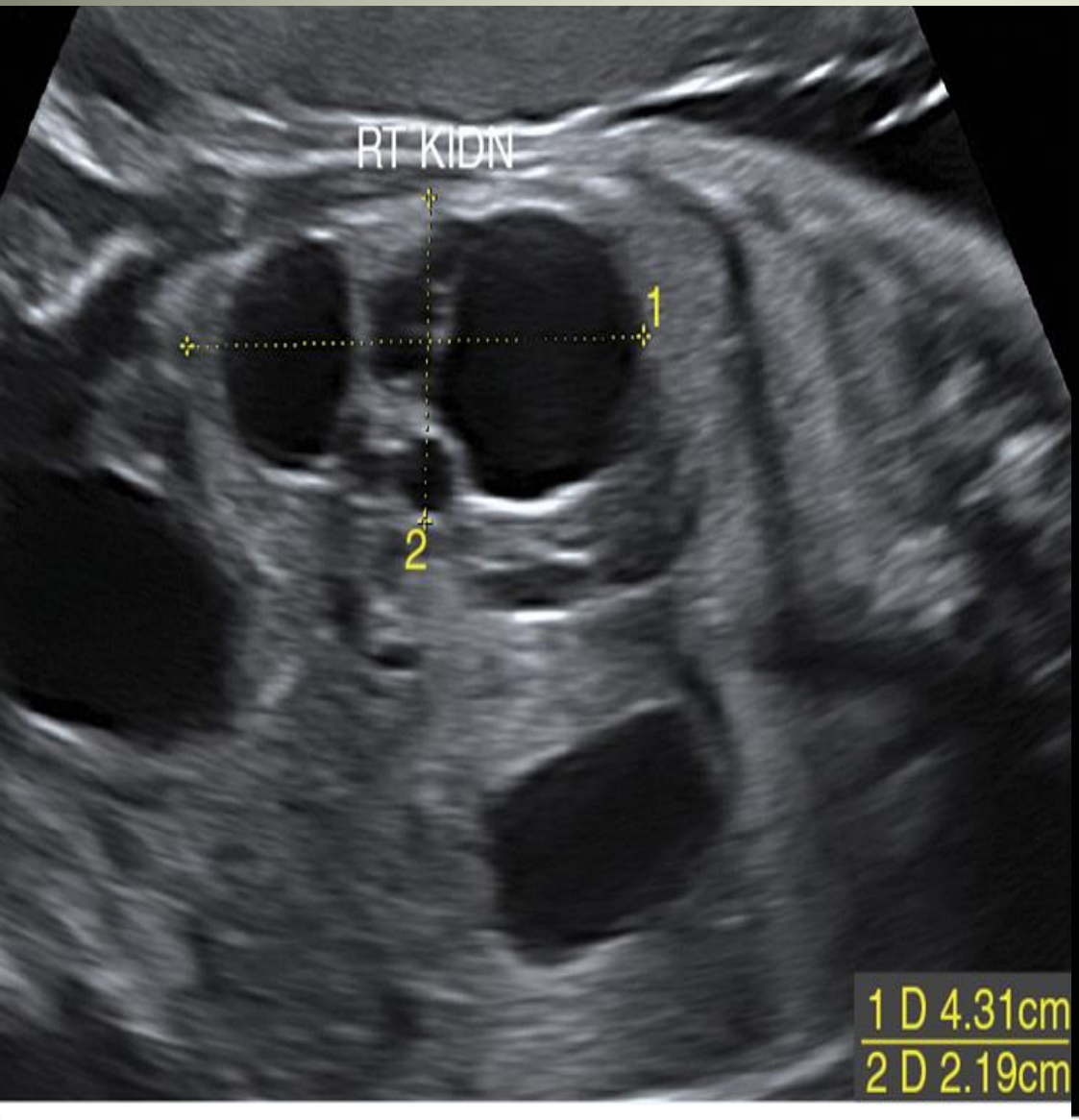
Type II Sonographic Findings

- Multiple cysts of variable size
- No distinct renal pelvis
- No distinct renal parenchyma
- Renal size may be normal, hypoplastic, or enlarged
- Severe oligohydramnios if bilateral
- A multicystic kidney needs to be distinguished from hydronephrosis and calyceal dilation
- When bilateral
  - Oligohydramnios and an absent bladder are expected

UNILATERAL MULTICYSTIC







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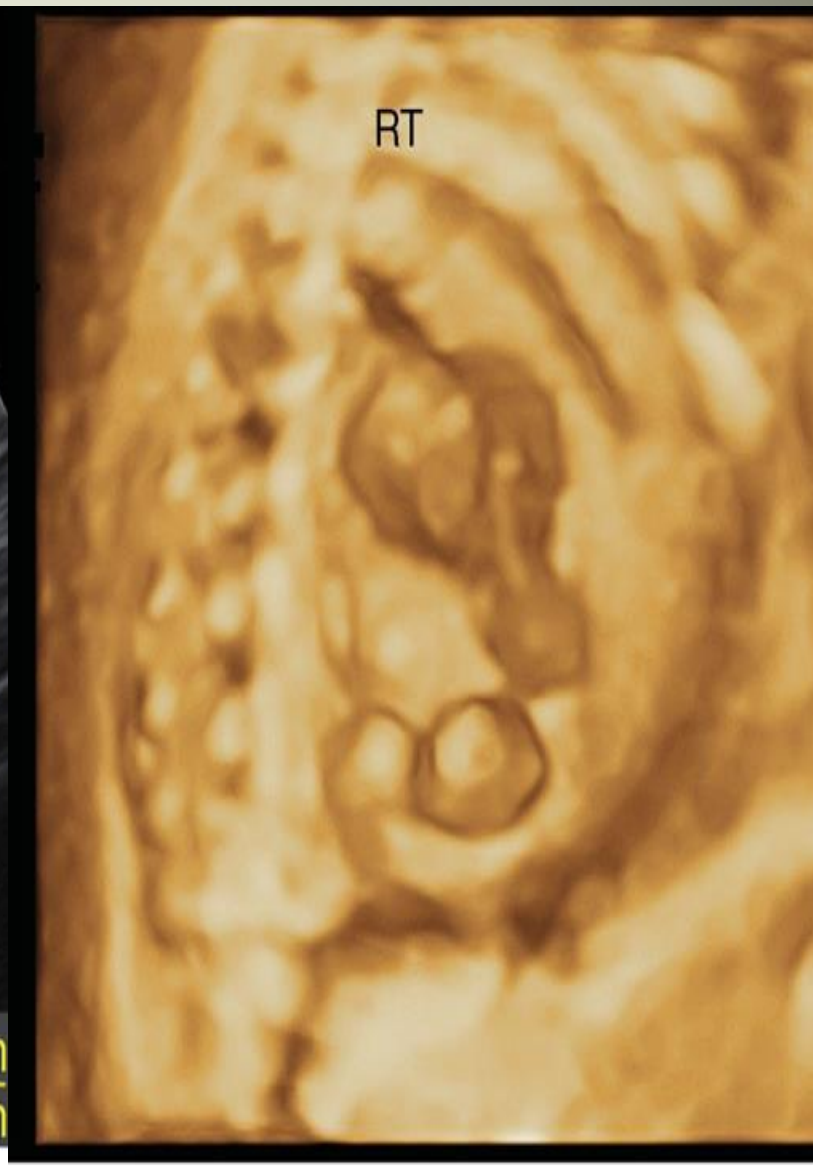

B

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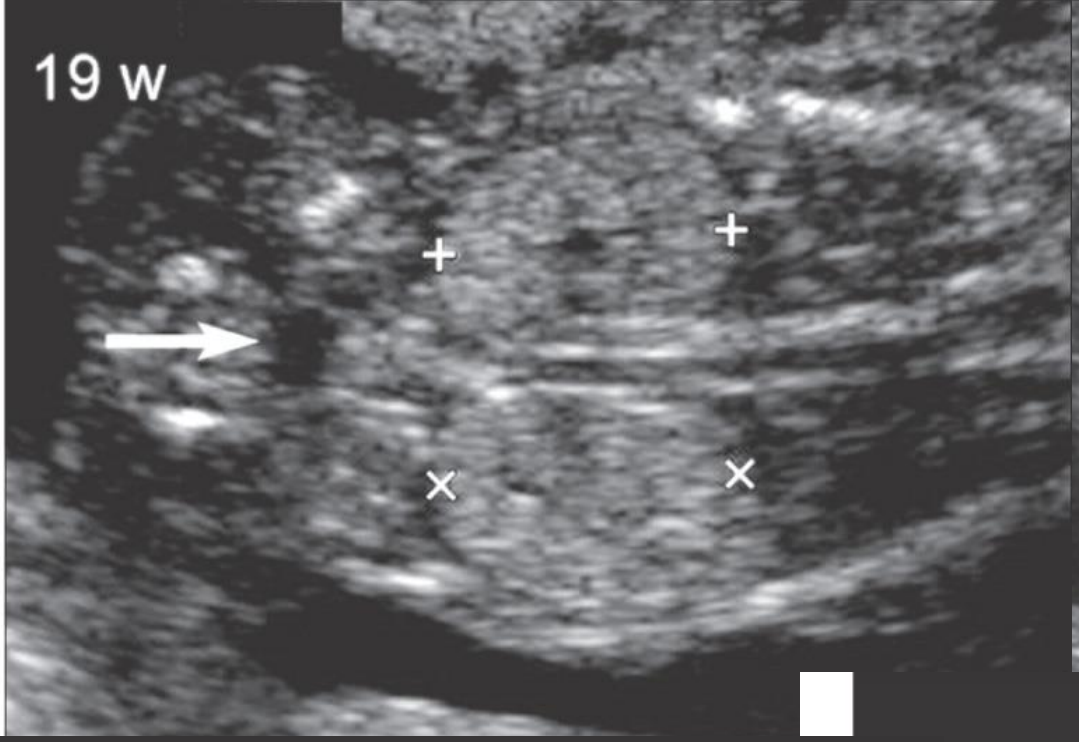
Type III

- Adult Dominant Polycystic Kidney Disease (APKD)
  - Autosomal-dominant
- May be differentiated in a fetus when there is a family history of polycystic kidneys, liver, or both
- Kidneys appear large and echogenic
  - Rarely are cysts observed prenatally
- Bilateral enlargement of kidneys may prompt a renal and liver work-up in the parents to exclude this disorder

Type III Sonographic Findings

- Large kidneys with hyperechoic parenchyma
- Size may be asymmetric
- Amniotic fluid volume is normal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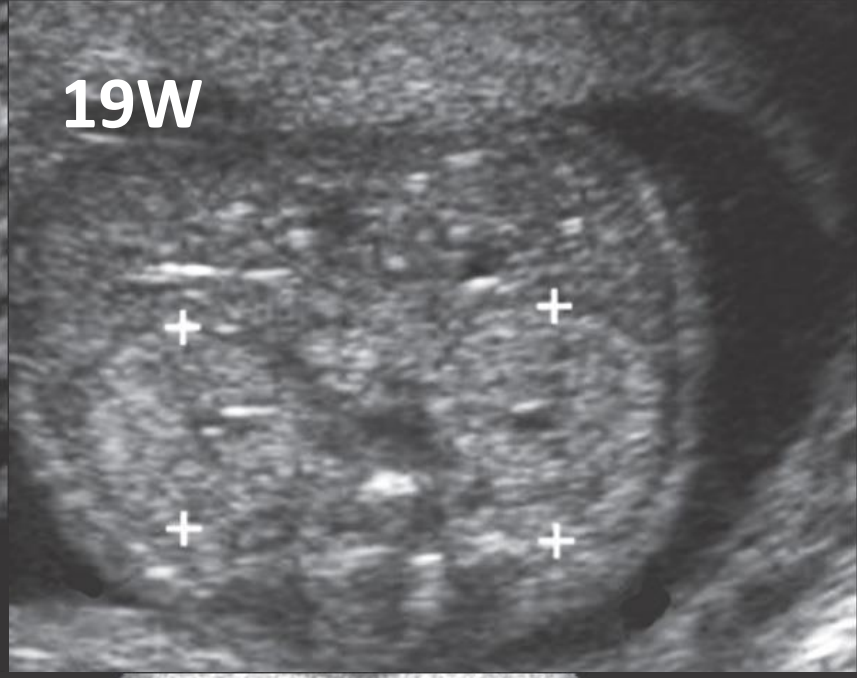
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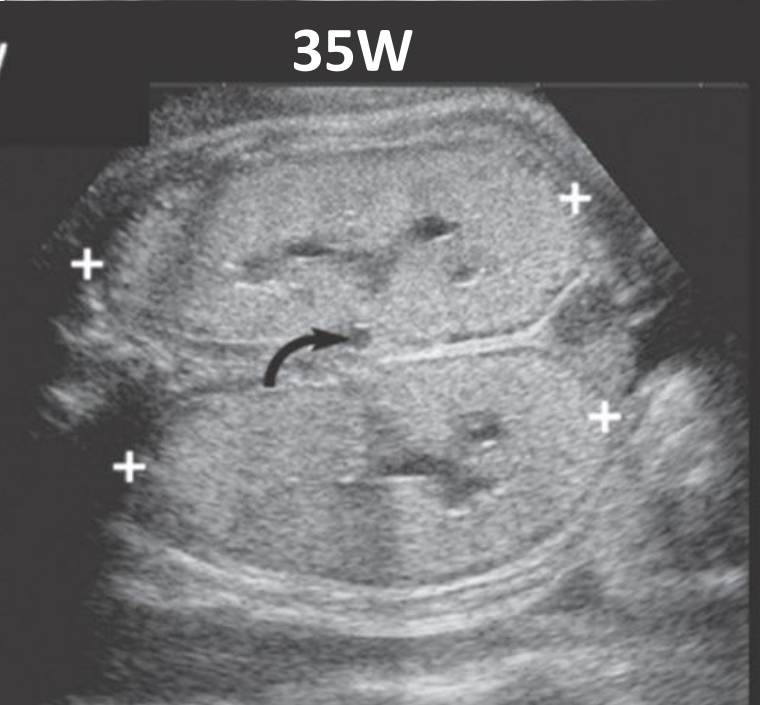
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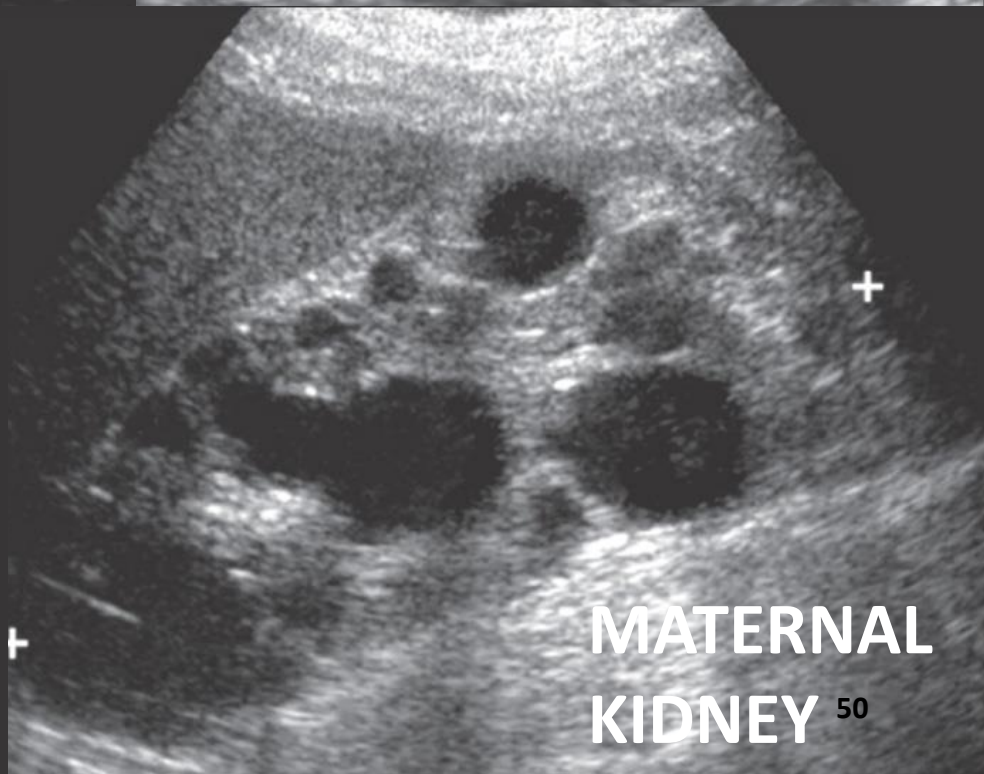
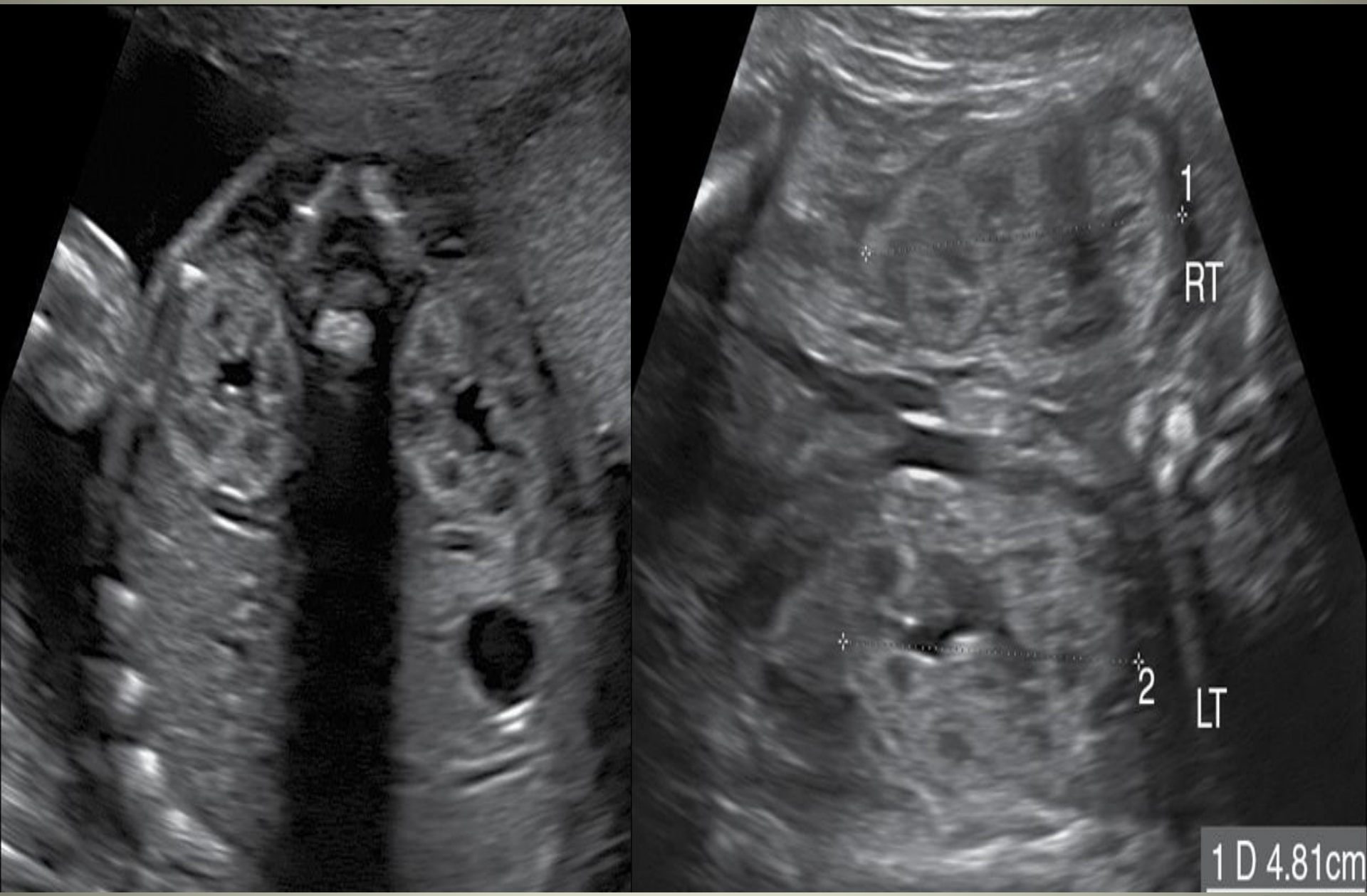

MATERNAL
KIDNEY


1
RT
2
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1 D 4.81cm

Type IV

- Obstructive cystic dysplasia
  - Renal dysplasia occurs secondary to obstruction in the first or early second trimester
- Caused by early renal obstruction
- Unilateral disease can be caused by:
  - Pelviureteral junction obstruction
  - Vesicoureteric junction obstruction
- Bilateral obstructive dysplasia caused by
  - Severe bladder outlet obstruction
  - Usually urethral atresia or posterior urethral valves

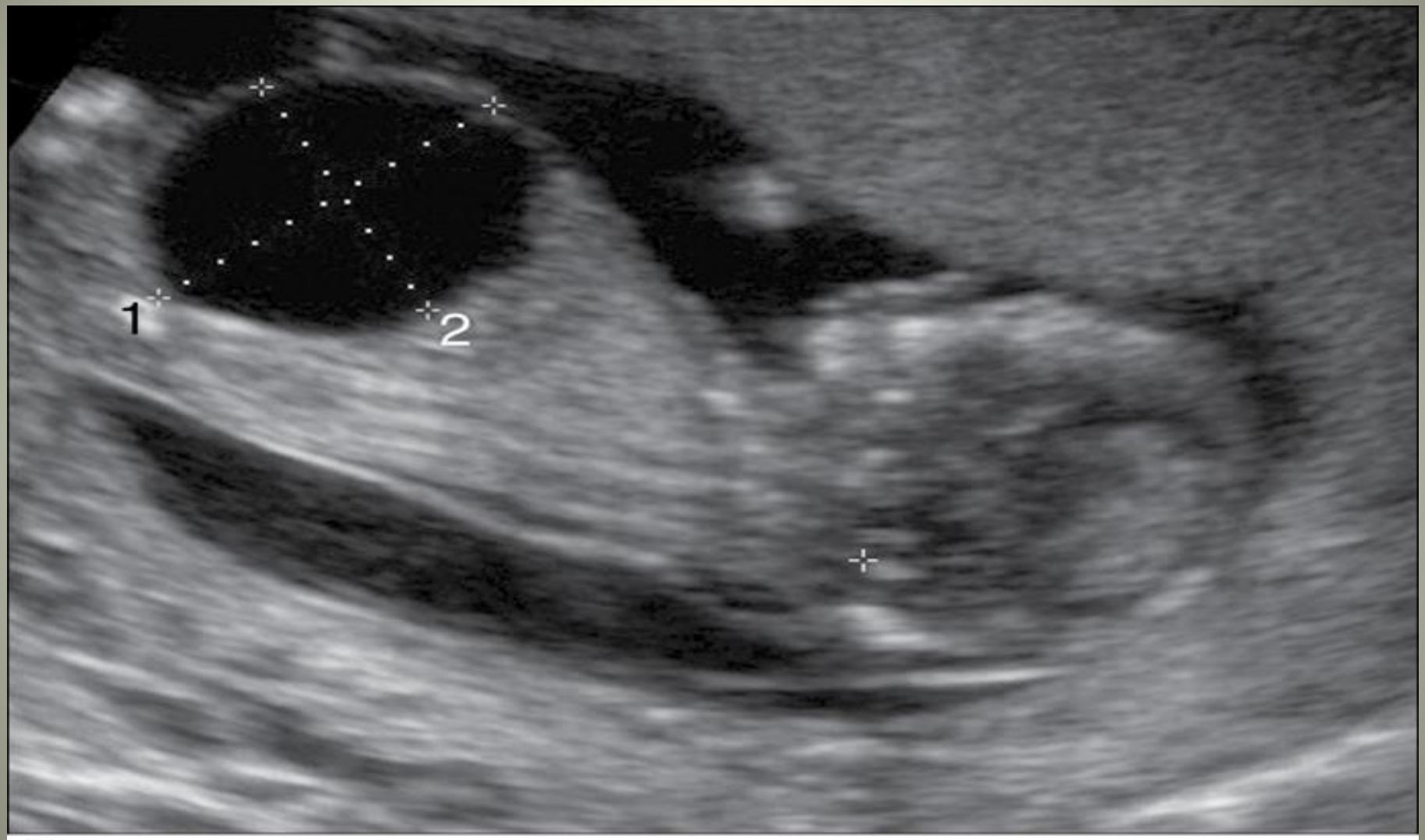
Type IV Sonographic Findings

- Kidneys appear small and echogenic with cortical peripheral cysts
- If bilateral look for:
  - Early bladder outlet obstruction
  - Bilateral hydronephrosis
  - Thick-walled bladder
  - Severe oligohydramnios
- Cortex is dysplastic and replaced with the multiple cortical cysts

Obstructive Urinary Tract Abnormalities

- May be obstructed
  - Ureteropelvic junction
    - Junction of the ureter entering the renal pelvis
  - Ureterovesical junction
    - Junction of the ureter as it enters the bladder
  - Megacystis
    - At the level of the urethra

Obstructive Cystic Dysplasia



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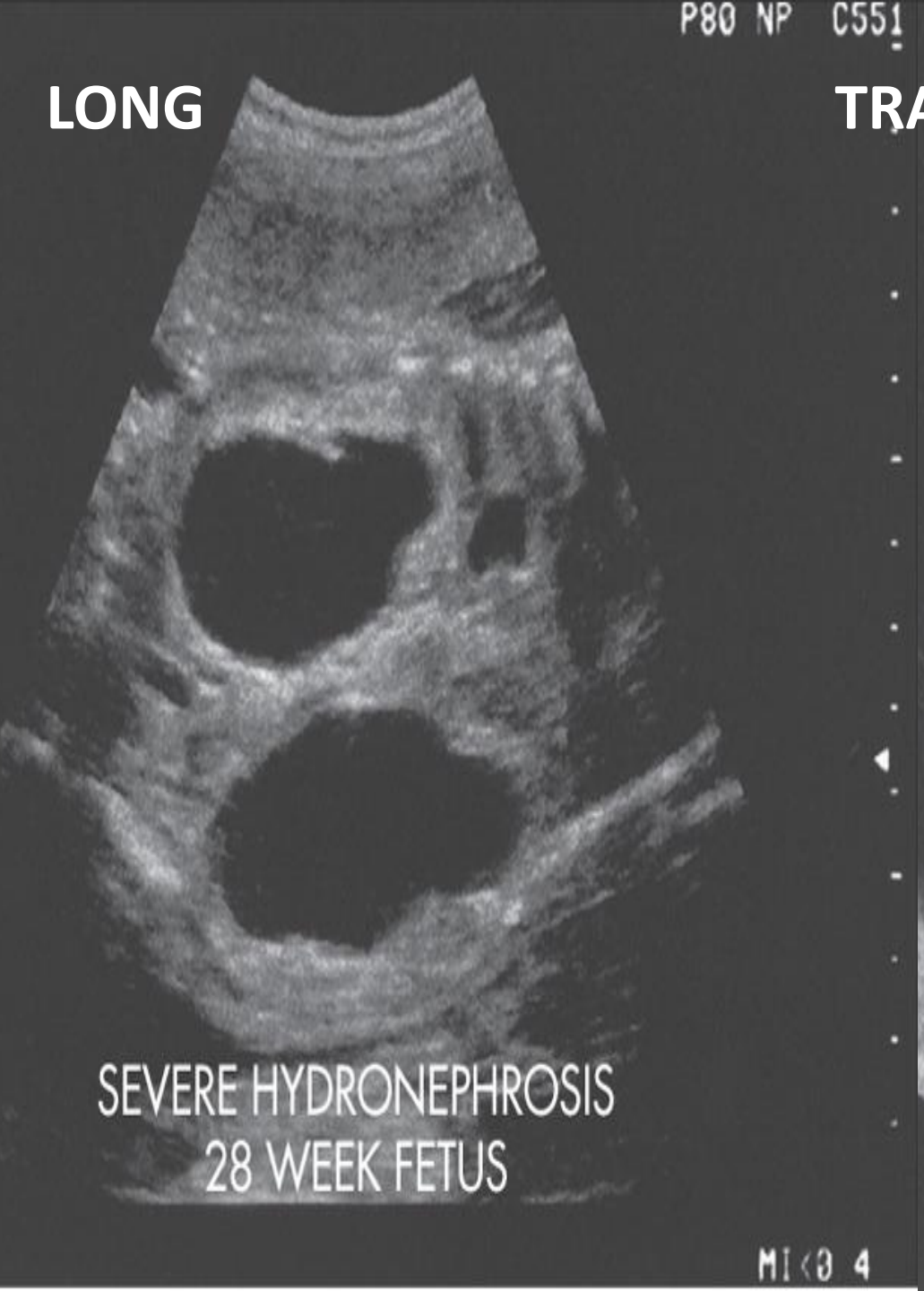
Hydronephrosis

- Most common fetal anomaly
- Sonographic appearance of urinary tract obstruction varies, depending on the site and extent of blockage
- Dilation of the renal pelvis occurs as a response to a blockage of urine at some junction
- Commonly occurs when obstruction is in the
  - Ureter, bladder, or urethra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SEVERE HYDRONEPHROSIS
28 WEEK FETUS
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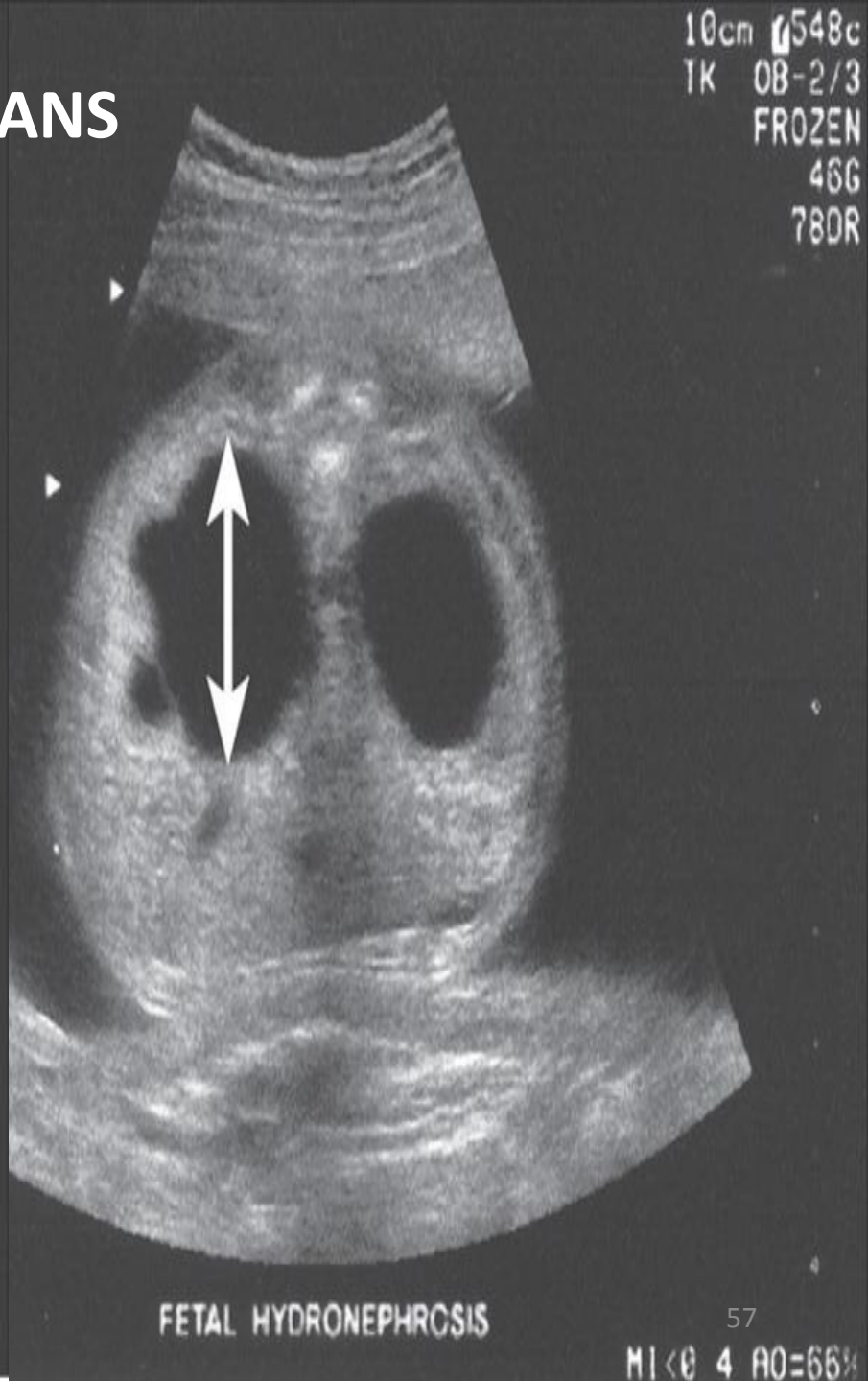
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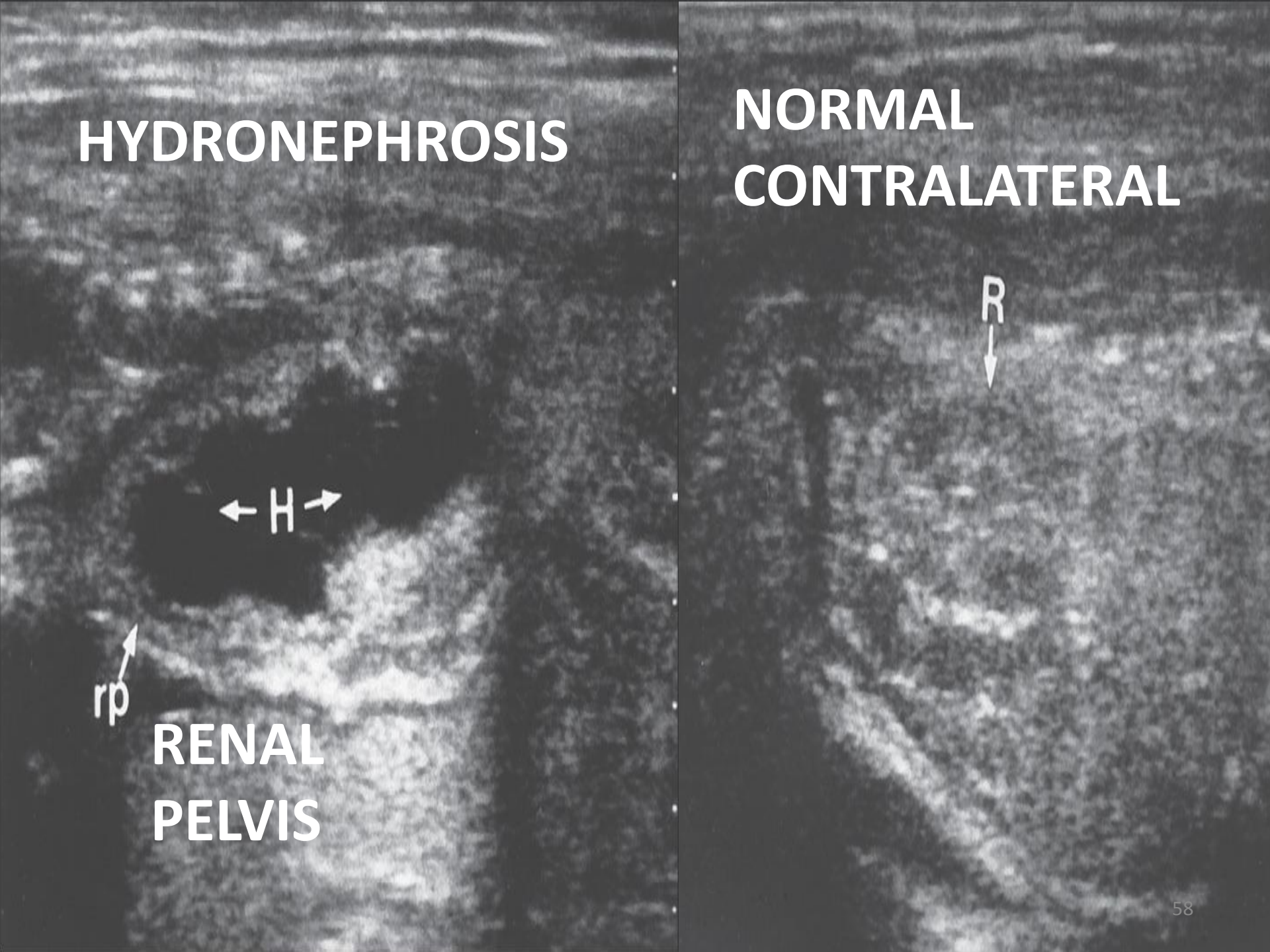

HYDRONEPHROSIS
H
rp
RENAL PELVIS
NORMAL CONTRALATERAL
R

Sonographic Findings

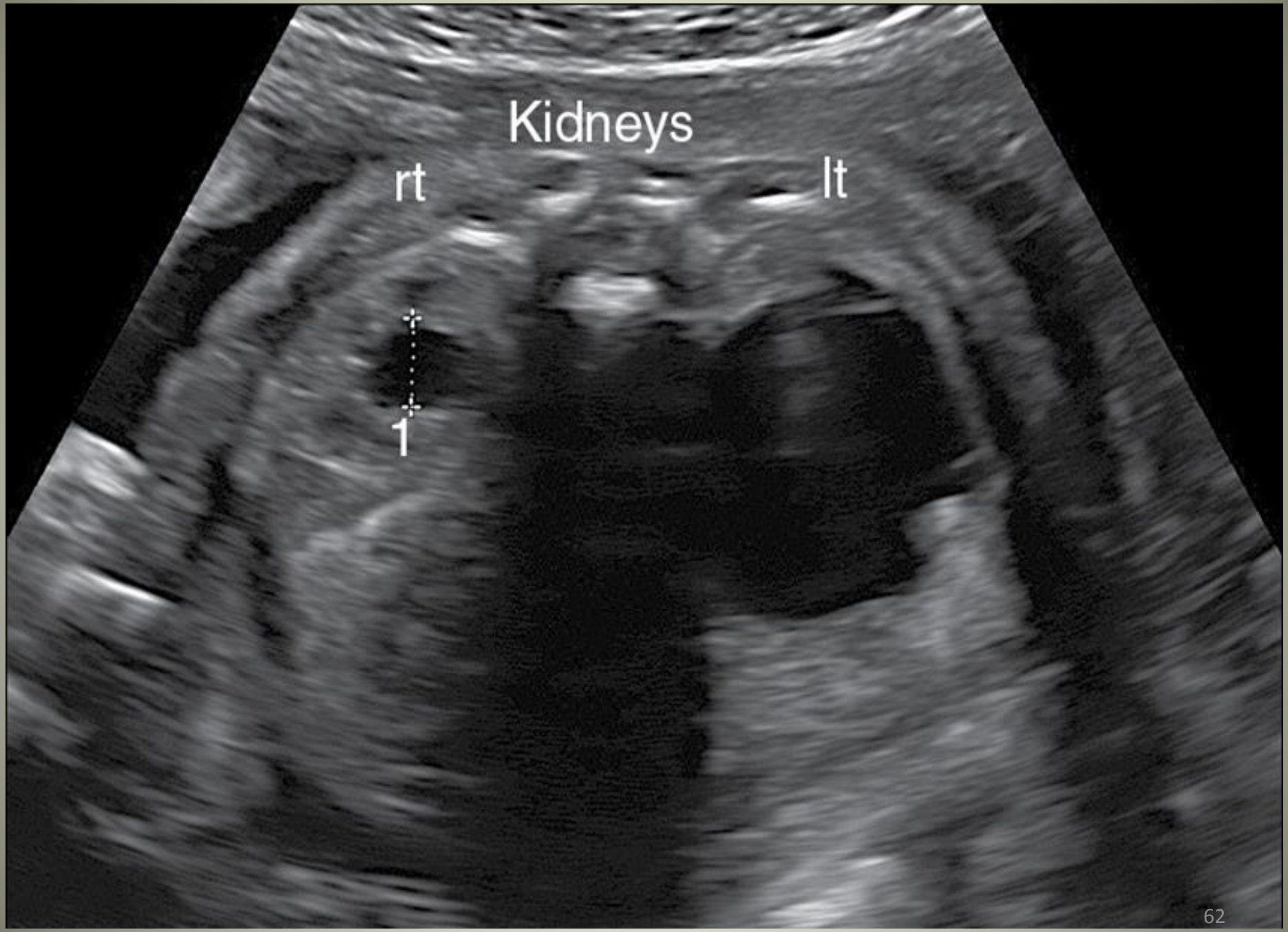
- AP renal pelvic diameter:
  - > 7 mm considered mild hydronephrosis
  - 7-15 mm considered moderate hydronephrosis
  - > 15 mm considered severe hydronephrosis
- Rim of renal parenchyma preserved
- Calyceal distention with central pelvis communication

Sonographic Findings

- Renal dysplasia often occurs and represents cystic changes within the renal tissue.
- May occur unilateral or bilateral
  - Unilateral commonly results from obstruction at the ureteropelvic junction (UPJ)
    - Junction of the renal pelvis and the ureter

Hydronephrosis

- Prognosis depends on severity and cause
- When considering severity without relating to cause or dysplasia, mild hydronephrosis has been found to resolve antenatally or postnatally without surgical intervention
- Requires image of true cross-sectional plane through mid renal pelvis
  - Accurate AP pelvis measurement
  - Optimally, spine is located anteriorly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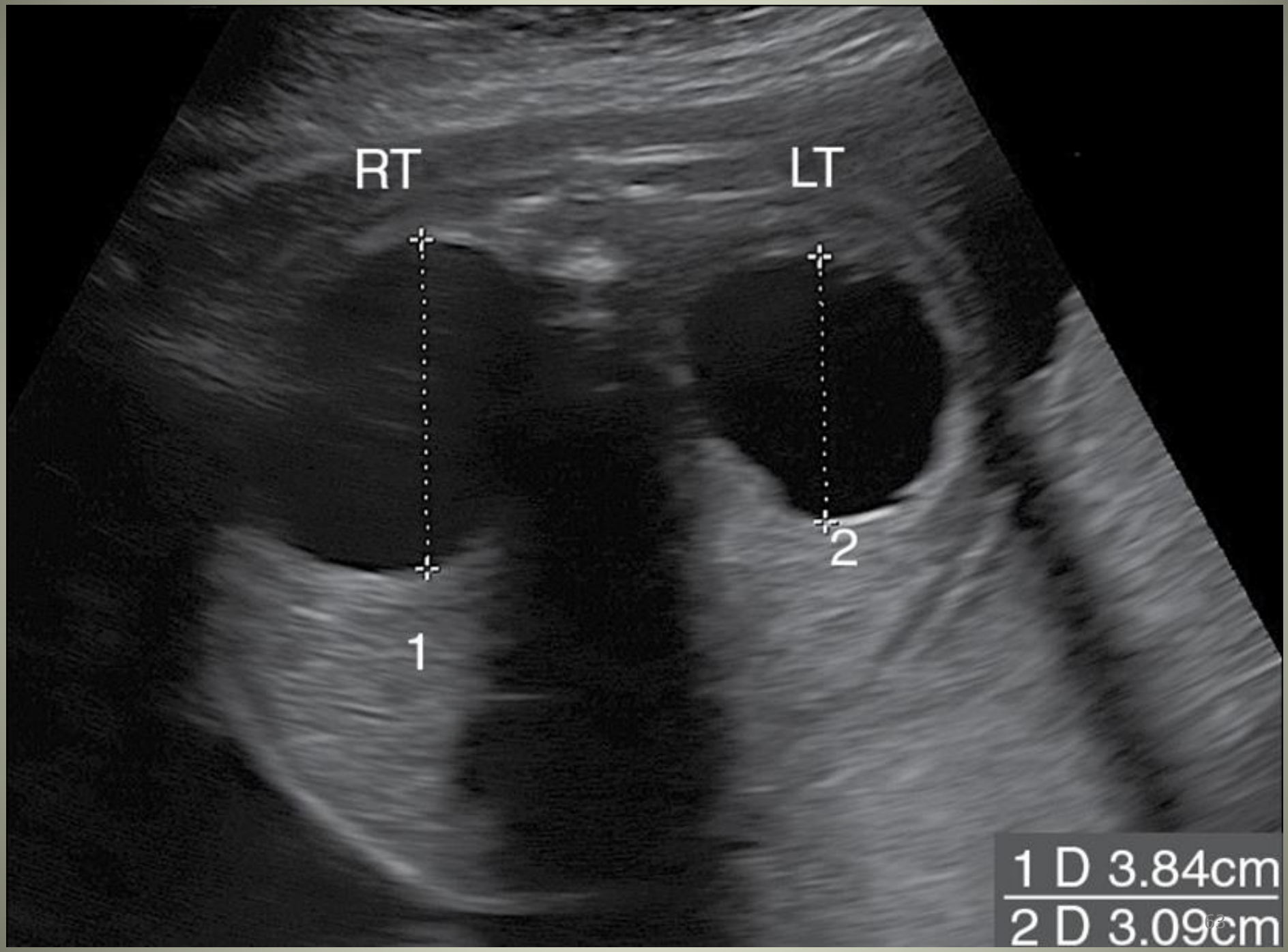
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1 D 3.84cm
2 D 3.09cm

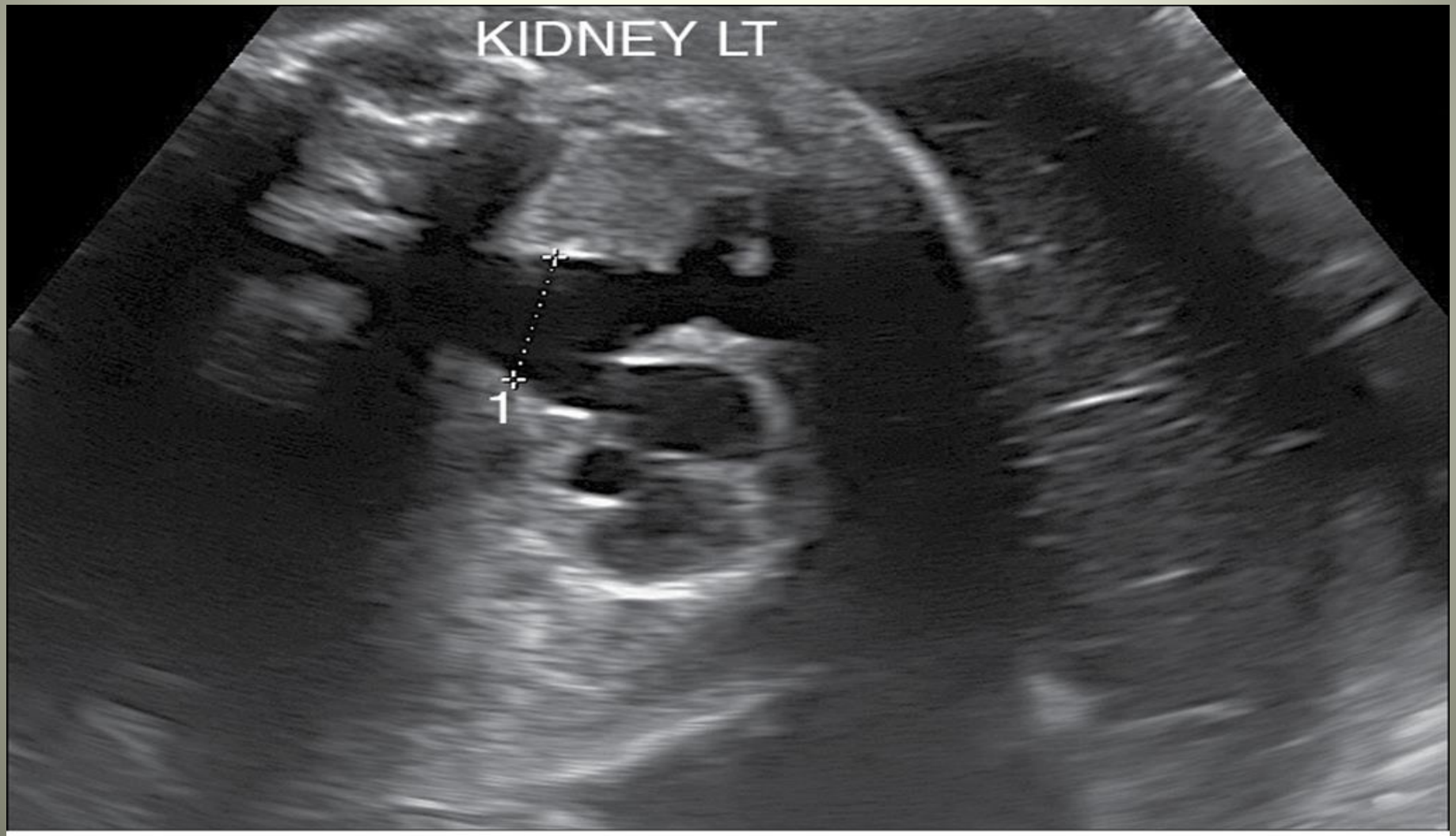
Ureteropelvic Junction Obstruction

- UPJ is the most common reason for hydronephrosis in neonate
- Male prevalence
- Early prenatal detection may improve long-term renal function

Ureteropelvic Junction Obstruction

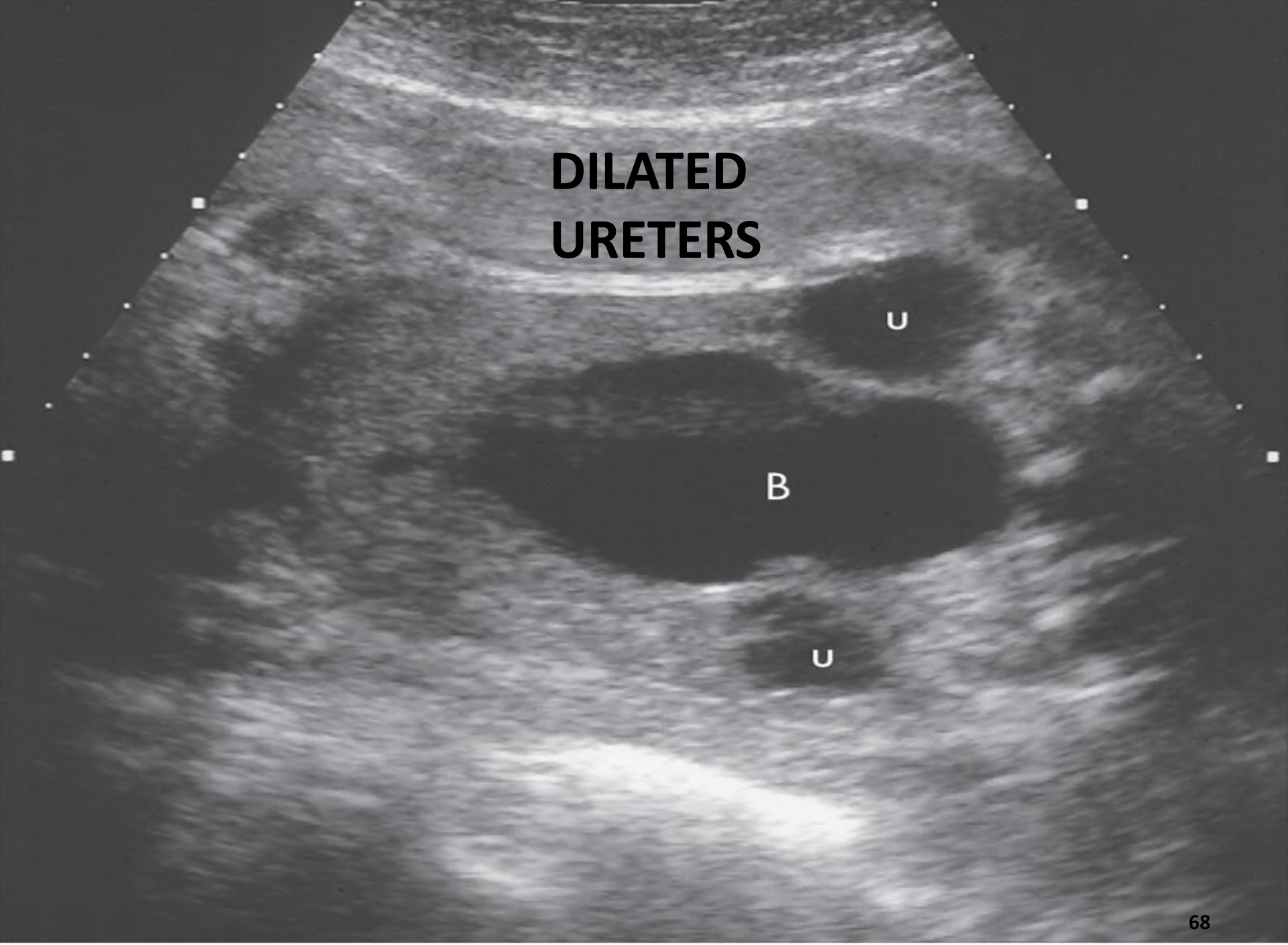
- **Sonographically:**
  - **Collection of anechoic urine located medially within renal pelvis that communicates with calyces**
  - **Renal cortex, ureter, bladder, amniotic fluid usually normal in cases of unilateral involvement**

Ureteropelvic Junction Obstruction



Ureterovesical Junction Obstruction

- Commonly presents with dilation of ureter (Megaureter)
  - May result from
    - Primary ureteral defect
      - Stenotic ureteral valves or fibrosis
    - Secondary to obstruction at another level
      - Causing reflux or backward flow of urine



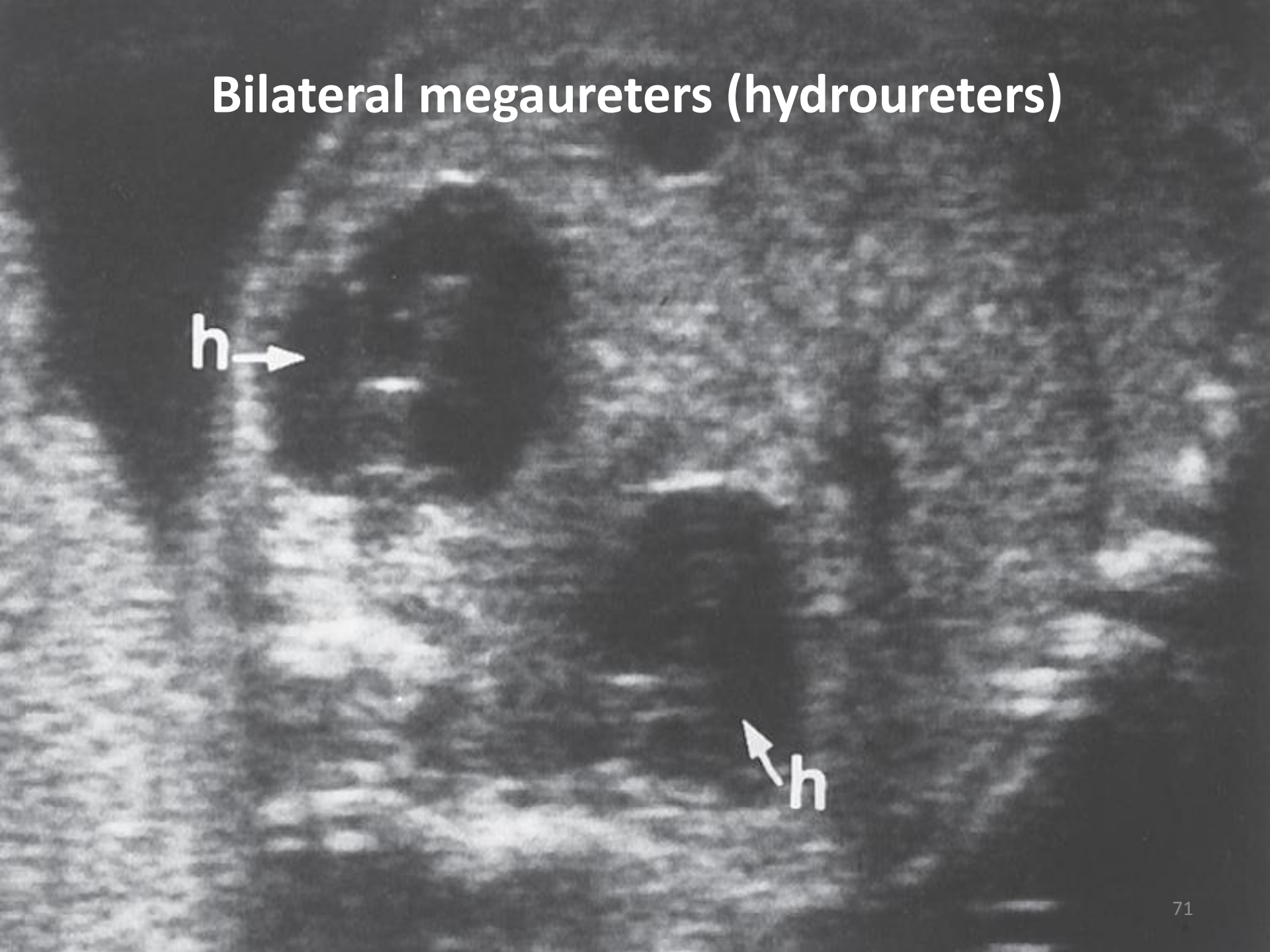
Secondary Obstruction to Ureterocele and Ectopic Ureter

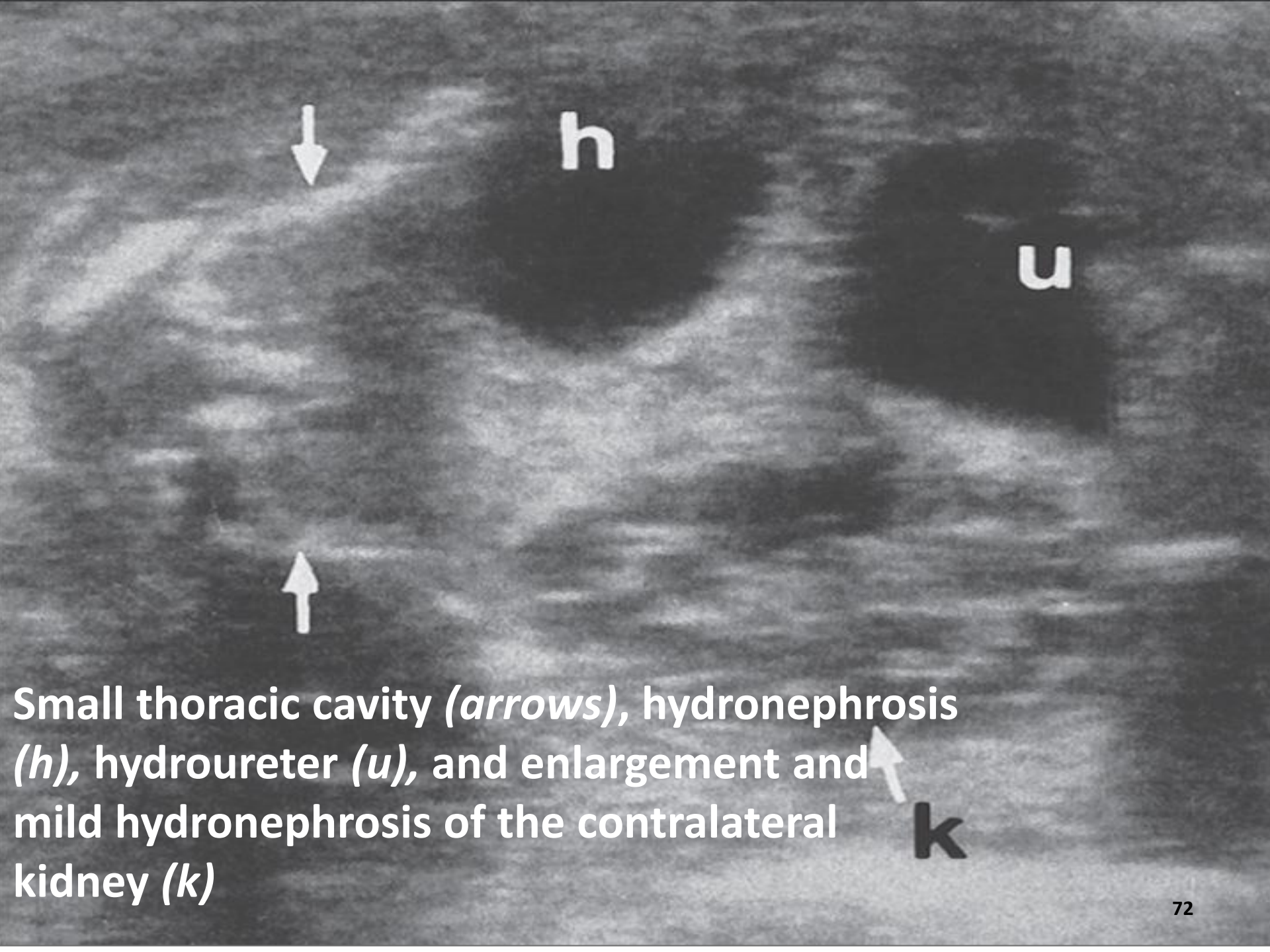
- Ureterocele - cystic dilation of intravesical (bladder) segment of distal ureter
- Ectopic ureter - does not insert near posterolateral angle of trigone of bladder
  - In females it may insert in the vagina, vestibule, or uterus
  - In males, it may insert in the seminal vesicle, vas deferens, or ejaculatory ducts
- May also insert into bladder in an ectopic location
- Unless hydronephrosis is present
  - Condition is difficult to diagnose in utero

Posterior Urethral Valve Obstruction

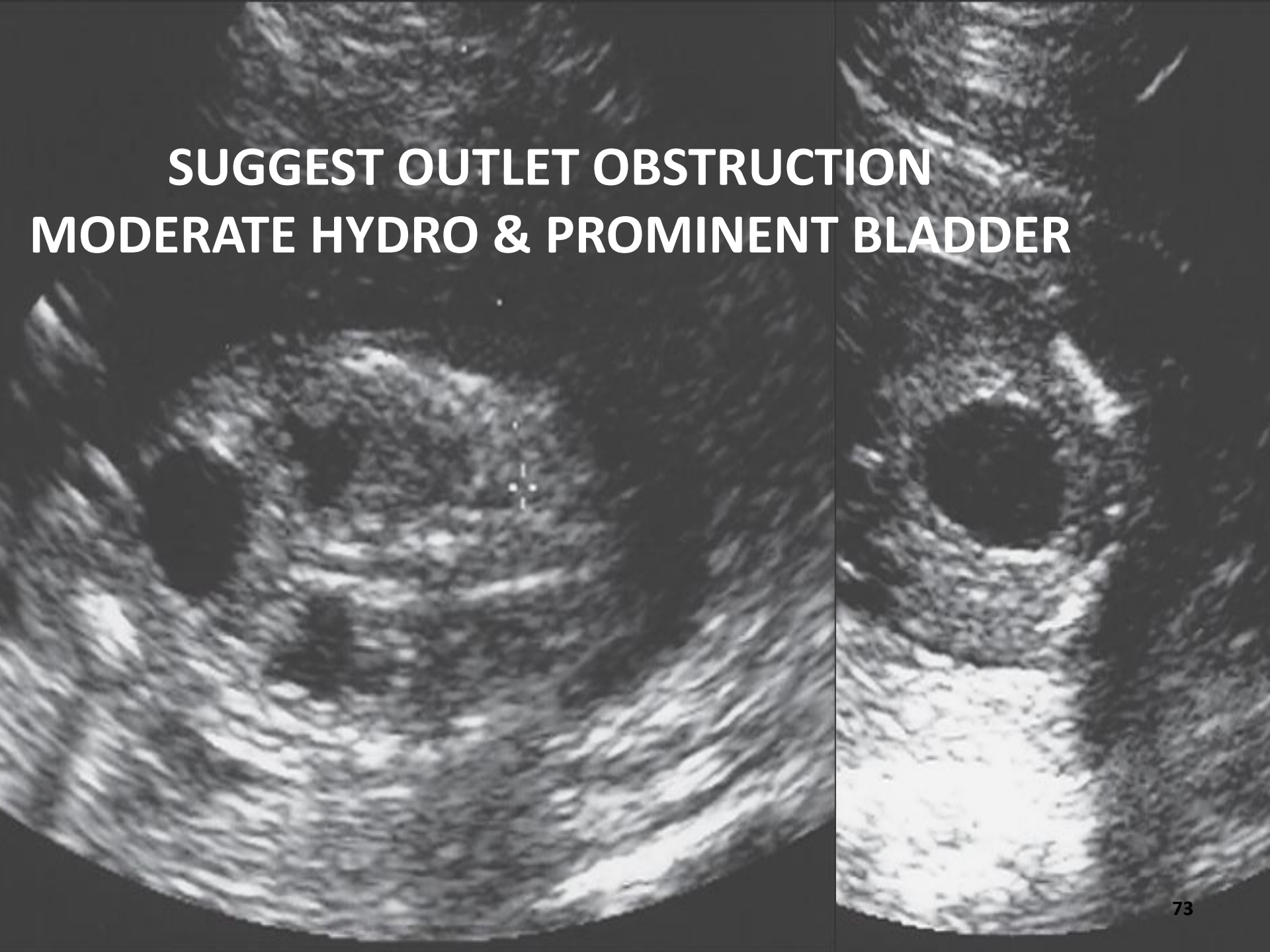
- Results in
  - Hydronephrosis
  - Hydroureters
  - Dilation of the bladder and posterior urethra
- Occurs only in males
  - Manifested by the presence of a valve(s) in the posterior urethra
- Urine is unable to pass through the urethra
- Causes a back-up in the bladder, ureter, and, in the most severe cases, the kidneys

Bilateral megaureters (hydroureters)



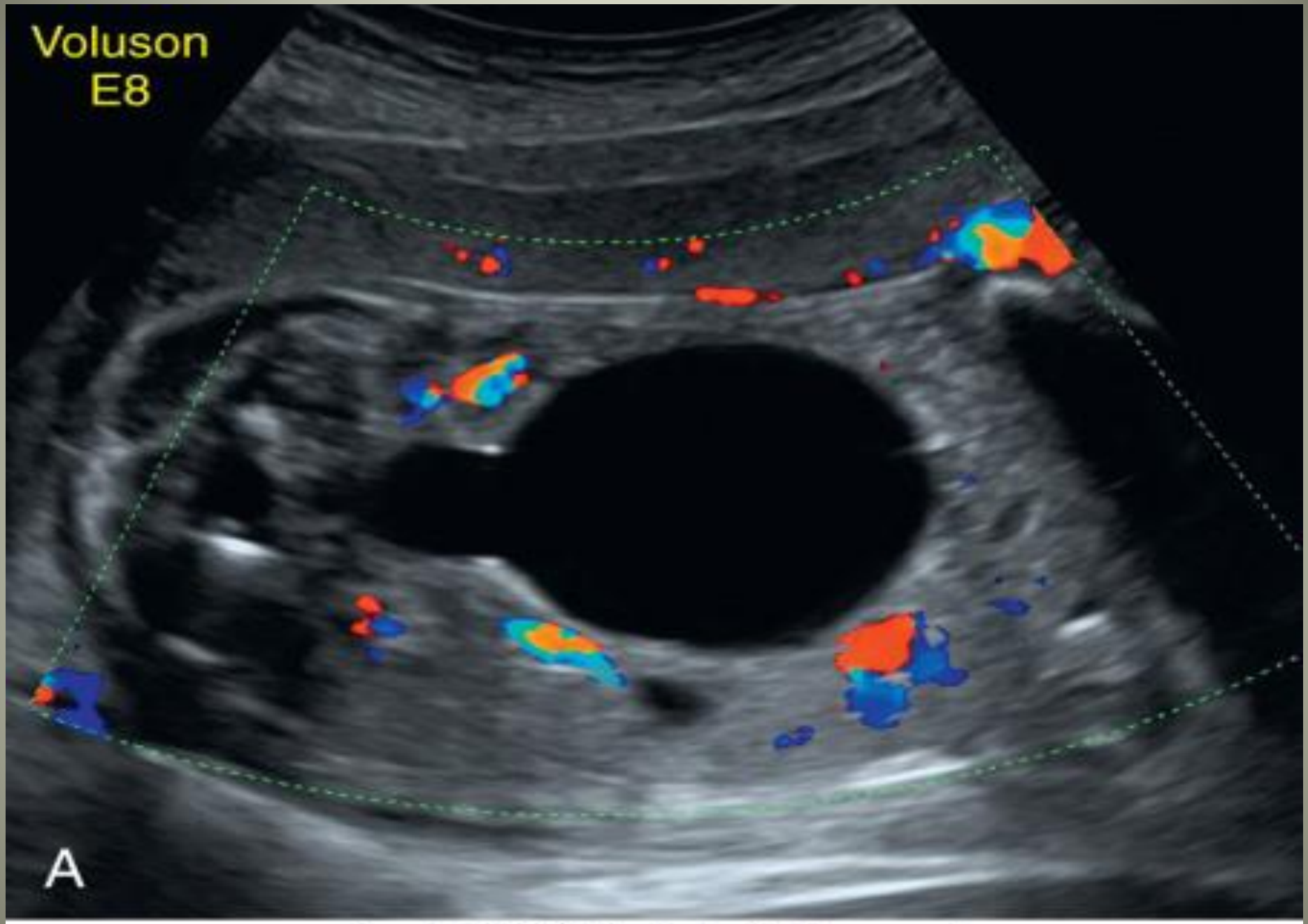


Small thoracic cavity *(arrows)*, hydronephrosis *(h),* hydroureter *(u),* and enlargement and mild hydronephrosis of the contralateral kidney *(k)*


SUGGEST OUTLET OBSTRUCTION
MODERATE HYDRO & PROMINENT BLADDER

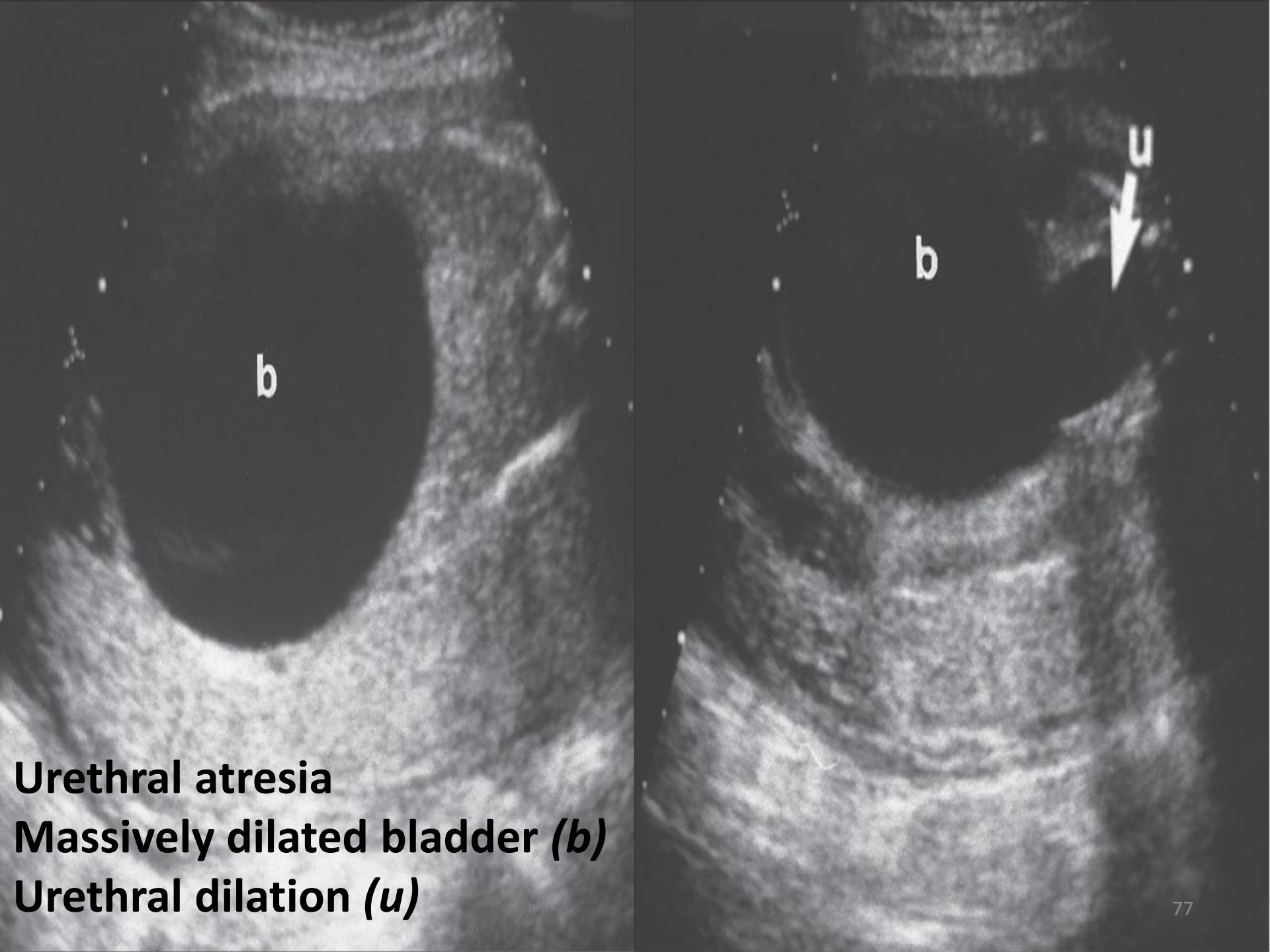
Sonographic Findings

- Dilated bladder (thickening of the bladder wall)
- Dilated posterior urethra "KEYHOLE SIGN"
- Oligohydramnios
- Hydroureters
- Hydronephrosis and dysplasia
- Fetal ascites (some cases)
- Distention of fetal abdomen (urethral obstruction malformation complex prune-belly syndrome)
- Male fetus

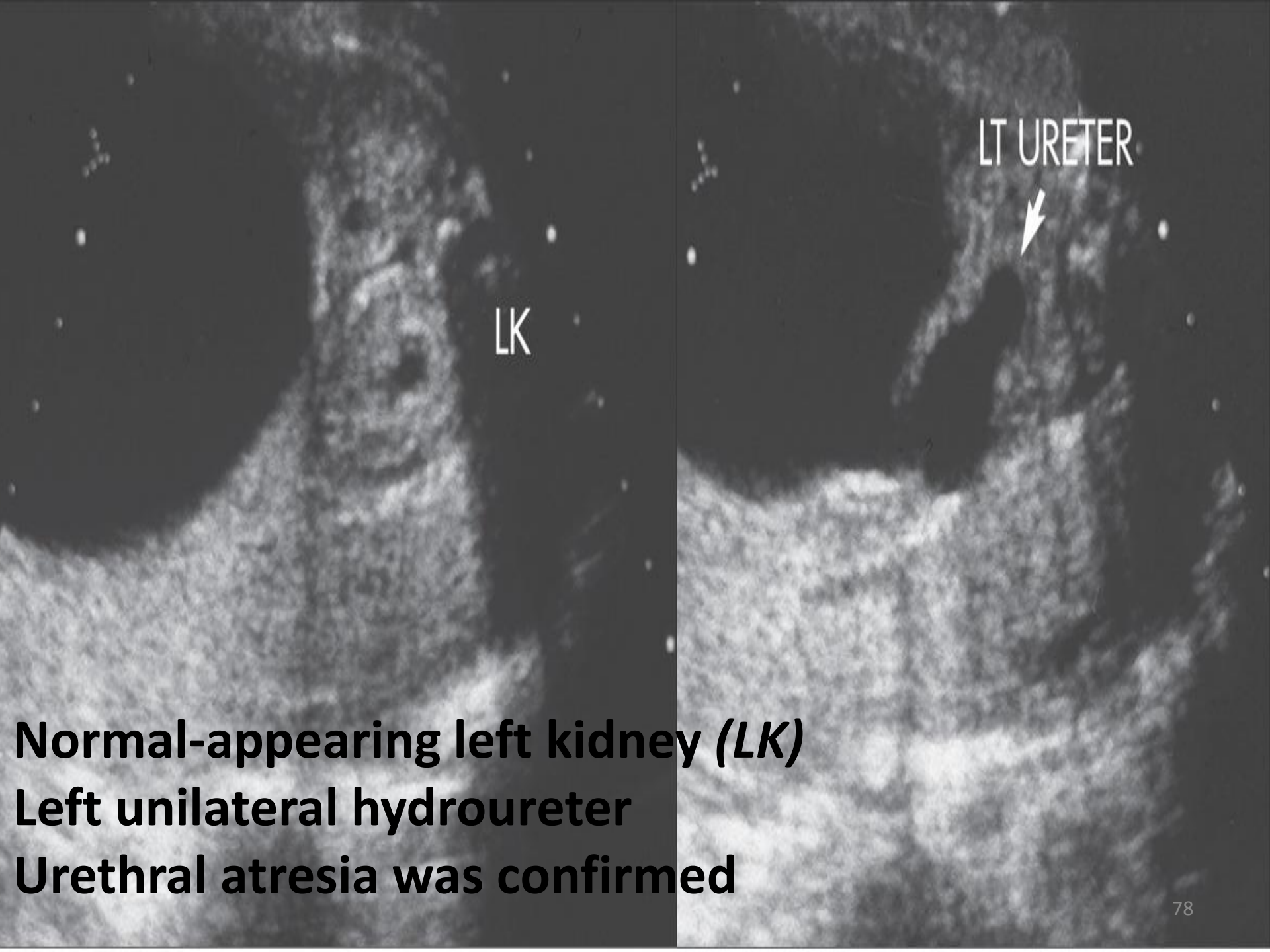


Prune-Belly Syndrome

- May be called “urethral obstruction malformation complex” or Eagle Barrett syndrome
- Consists of:
  - Cryptorchidism
  - Agenesis of abdominal wall muscle
  - Megaureters
  - Bladder outlet obstruction
    - Caused by
      - Urethral anomalies
        - Atresia
        - Stenosis valves
        - Diverticulum



Urethral atresia
Massively dilated bladder *(b)*
Urethral dilation *(u)*



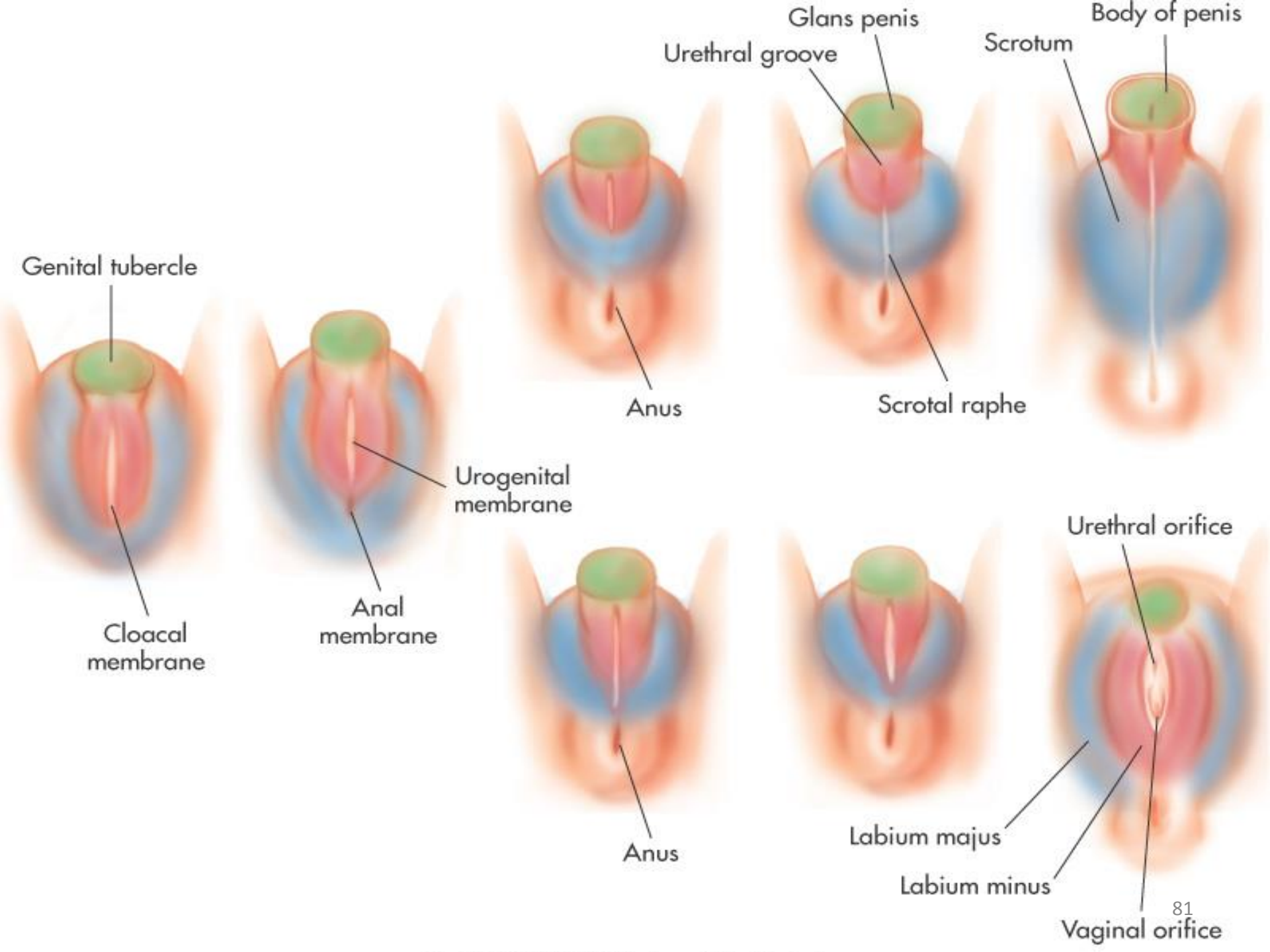
Normal-appearing left kidney *(LK)*
Left unilateral hydroureter
Urethral atresia was confirmed

Sonographic Findings

- Oligohydramnios
- Mild to severe bilateral hydronephrosis
- Fetal ascites
- Hypoplastic lungs
- Abdomen extremely distended compared with small thoracic cavity
- Dilated ureters and bladder appear as numerous cystic lesions within the distended abdominal cavity

Development of the External Genitalia

- Early development of external genitalia is similar for both sexes
  - Distinguishing sexual characteristic begins during the ninth week
  - External genital organs are fully differentiated by the 12th week

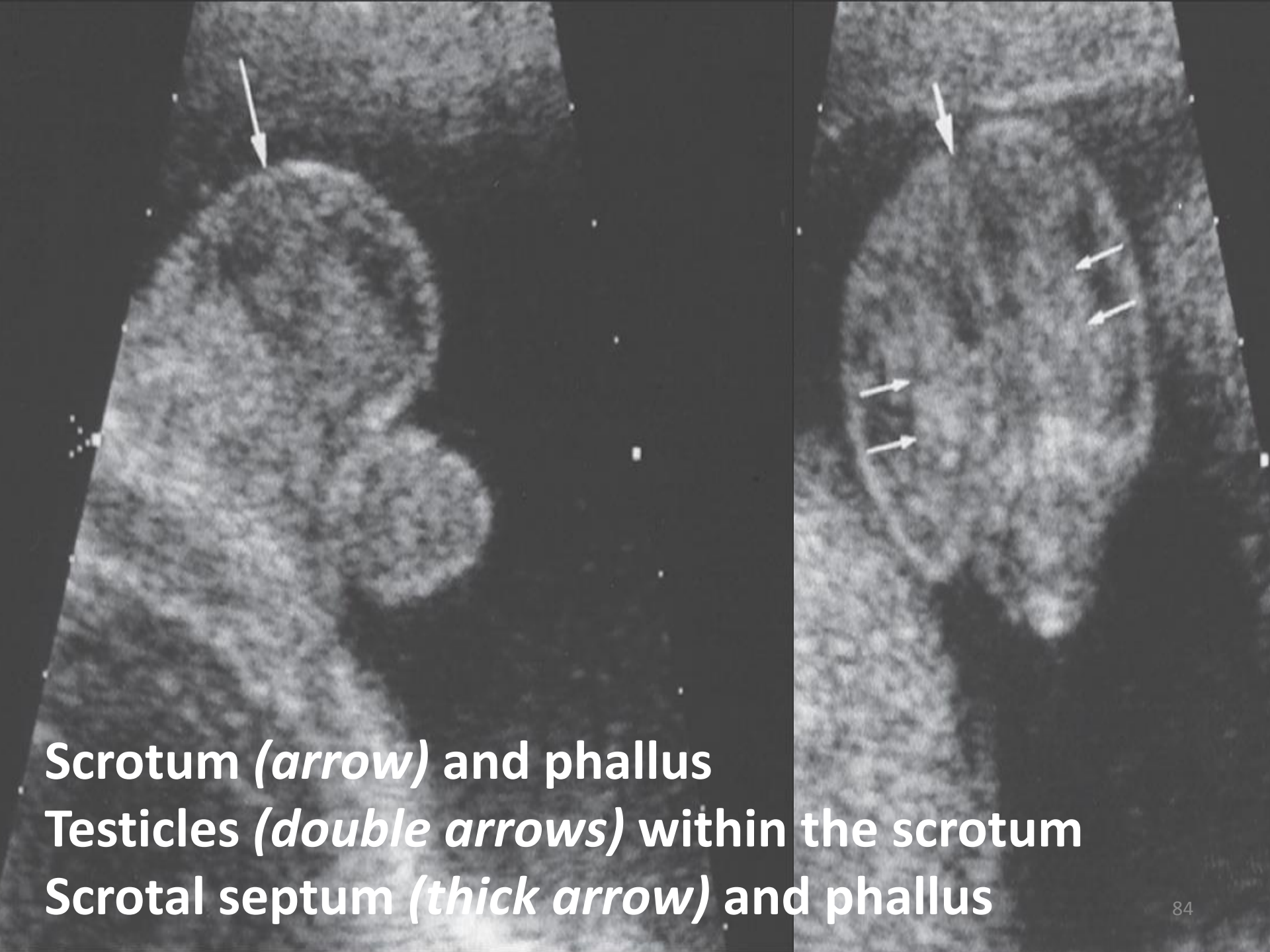

Glans penis
Urethral groove
Scrotum
Body of penis
Genital tubercle
Anus
Scrotal raphe
Urogenital membrane
Cloacal membrane
Anal membrane
Urethral orifice
Anus
Labium majus
Labium minus
Vaginal orifice

Development of the External Genitalia

- 4th week
  - Genital tubercle develops at the cranial end of the cloacal membrane
  - Labioscrotal swellings and urogenital folds develop off either side of this membrane
  - Genital tubercle elongates to form a phallus
    - Similar in both sexes

Development of Male External Genitalia

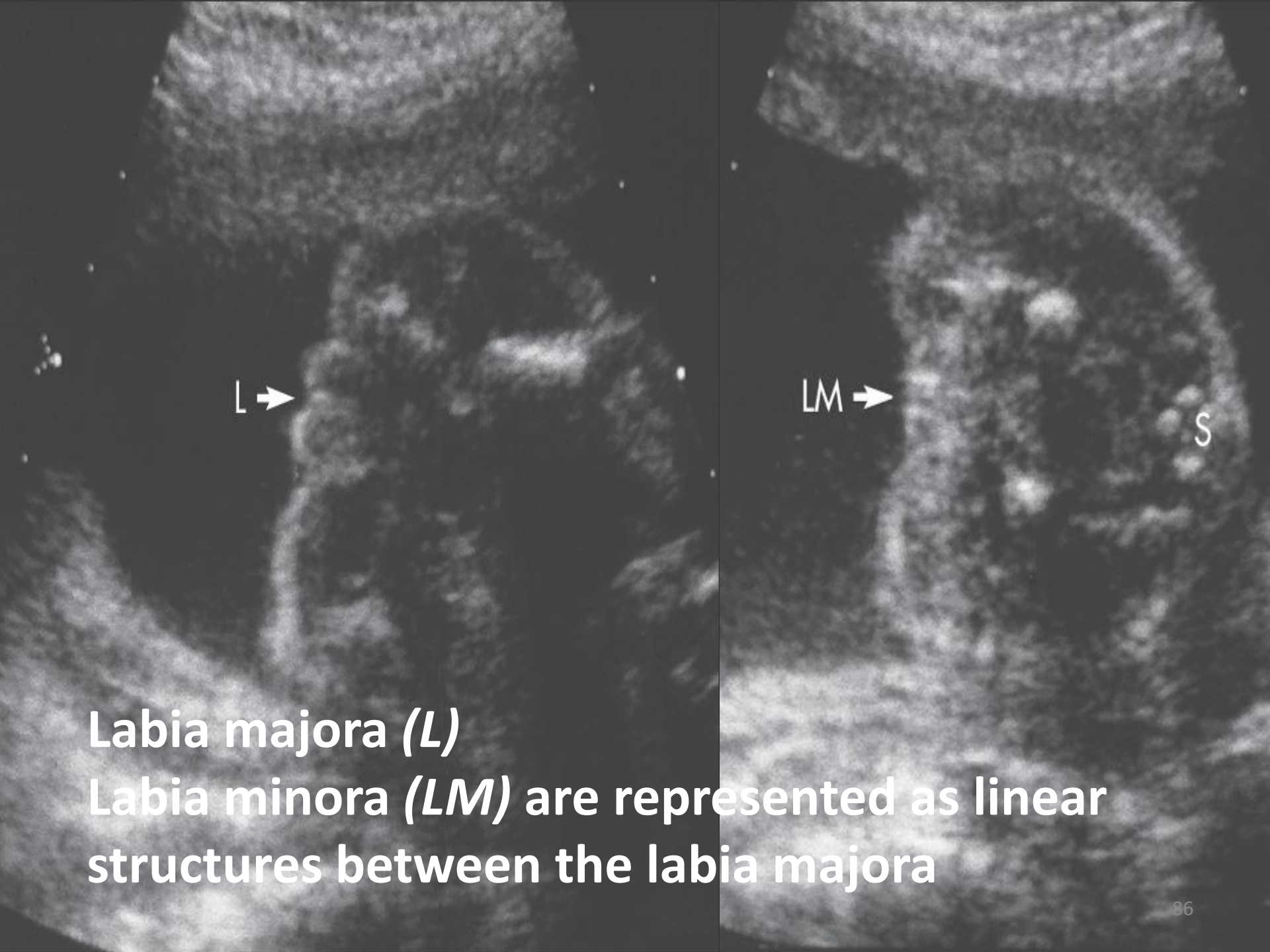
- Fetal testes produce androgens that cause the masculinization of the external genitalia
- Phallus elongates to form the *penis*
- Urogenital folds fuse on ventral surface of penis to form *spongy urethra*
- Labioscrotal swellings grow toward the median plane and fuse to form the *scrotum*
- Line of fusion of the labioscrotal folds
  - Called the *scrotal raphe*



Scrotum *(arrow)* and phallus
Testicles *(double arrows)* within the scrotum
Scrotal septum *(thick arrow)* and phallus

Development of the Female External Genitalia

- Both urethra and vagina open into the urogenital sinus
  - "vestibule of the vagina"
- Urogenital folds become the labia minora
- Labioscrotal swellings become the labia majora
- Phallus becomes the clitoris



Labia majora *(L)*
Labia minora *(LM)* are represented as linear structures between the labia majora

Congenital Malformations of the Genital System

- Rare
- May be requested to determine the gender
  - When a gender-linked disorder is considered
    - Hemophilia
    - Aqueductal stenosis
  - These conditions usually occur in male fetuses
- Abnormal fetal genitalia may be indicative of syndromes of the endocrine and genital systems

Malformations of the Uterus and Vagina

- With incomplete fusion of two müllerian ducts
  - Various forms of duplication of the uterus and/or vagina may occur
- Complete failure of the fusion
  - Duplication of the entire female genital tract
    - Uterus didelphys (double uterus and double vagina)
- Duplication of uterus with one vagina
  - Bicornuate uterus

Hydrocele

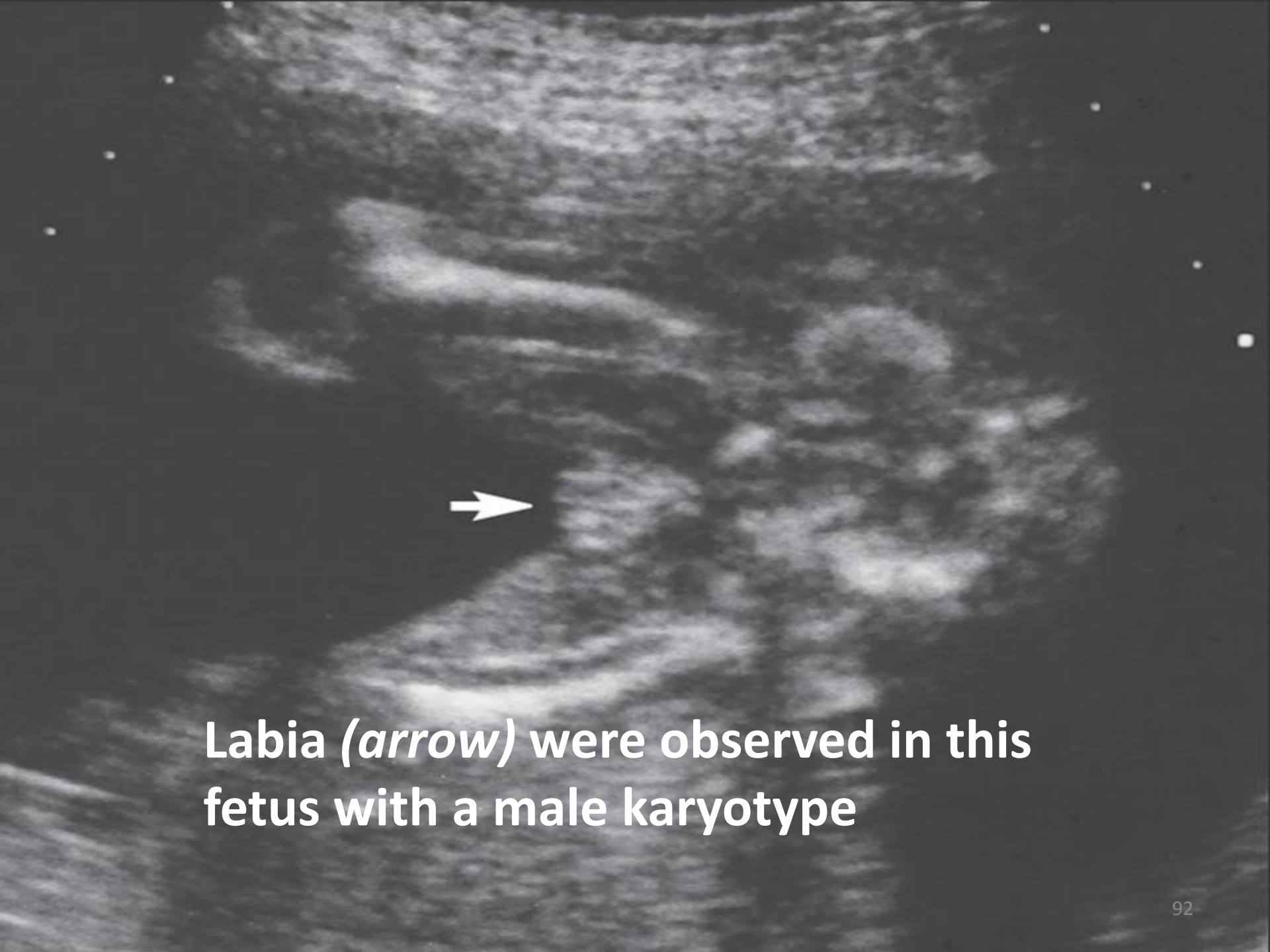
- Accumulation of serous fluid surrounding the testicle
  - Results from a communication with peritoneal cavity
- May occur
  - unilateral
  - bilateral lesions
- Generally benign

Descent of the Gonads

- Eventually descend from the abdomen to the pelvis
- Testes remain near the deep inguinal area until the 28th week
- Descend through the inguinal canals and enter the scrotum before birth
- Cryptorchidism - (undescended testes)
  - Failure to descend

Ambiguous Genitalia

- **Hermaphroditism**
  - **Occurs when errors take place in determining male or female sexuality**
- **True hermaphrodism**
  - **Rare condition in which both ovarian and testicular tissues are present**
  - **Internal and external genitalia are variable**
- **Most fetuses have a normal karyotype, but some are mosaics**
  - **(46,XX/46,XY)**
- **Must be careful in making the differential of ambiguous genitalia because penile and clitoral size may vary in normal fetus**



Labia *(arrow)* were observed in this fetus with a male karyotype

Female Pseudohermaphrodites

- Fetus has a 46,XX karyotype
- Most common cause
  - Congenital virilizing adrenal hyperplasia
    - Causes masculinization of the external genitalia
      - Enlarged clitoris
      - Abnormalities of urogenital sinus
      - Partial fusion of labia majora

Male Pseudohermaphrodites

- Fetus has testes and a 46,XY karyotype
- Variable external and internal genitalia depending on the development of the penis and genital ducts

Other Pelvic Masses

Hydrometrocolpos

- Collection of fluid in the vagina and uterus
- Appears as a hypoechoic “cystlike” mass posterior to the bladder (area of the uterus)
- Predominantly cystic
  - May contain
    - midlevel echoes
    - Fluid–debris levels

Ovarian Cyst

- Often appears as multiseptated and bilateral
- Results from maternal hormonal stimulation
  - usually benign
- May twist on itself and lead to
  - Torsion
  - Rupture
  - Intestinal obstruction
- Differential diagnosis
  - Mesenteric cyst
  - Urachal cyst
  - Enteric duplication