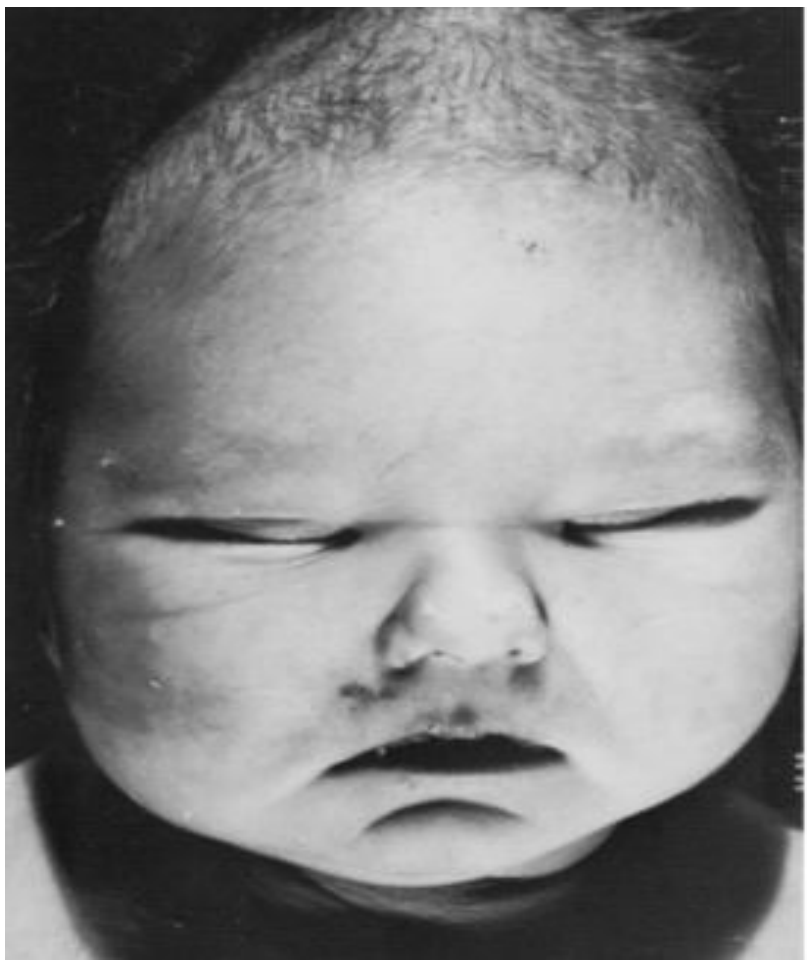
Kidney and Ureters : Agenesis, Horseshoe kidneys, congenital cysts, Megaureter, Ectopic ureter, Ureterocele

Dept Of Surgery

Bilateral Renal Agenesis

- Bilateral Renal Agenesis was first recognized in 1671 by Wolfstrigel
- Can occur secondary to a defect of the wolffian duct, ureteric bud, or metanephric blastema
- Bilateral agenesis occurs in 1 of every 4000 births
- Male predominance

- 40% of the affected infants are stillborn
- Children who are born alive do not survive beyond 48 hours because of respiratory distress associated with pulmonary hypoplasia
- The adrenal glands are usually normally positioned
- Characteristic Potter facies and presence of oligohydramnios are pathognomonic
- Complete ureteral atresia is observed in slightly more than 50% of affected individuals



Potter's facial appearance

Diagnosis

- The characteristic Potter facies and the presence of oligohydramnios are pathognomonic and should alert for this severe urinary malformation.

- Amnion nodosum—small white, keratinized nodules found on the surface of the amniotic sac—may also suggest this anomaly

- Anuria after the first 24 hours without distention of the bladder should suggest renal agenesis

Diagnosis

- BRA has been detected in higher proportion in cryptophthalmos or Frazer's syndrome, Klinefelter's syndrome , Kallmann's syndrome, esophageal atresia.

- Renal ultrasonography confirm the presence or absence of urine within these structures.

- Absence of uptake of the radionuclide in the renal fossa above background activity confirms the diagnosis of BRA.

- Umbilical artery catheterization and an aortogram defines the absence of renal arteries and kidneys.

Unilateral Renal Agenesis

- There are no tell tale signs (as with BRA) that suggest an absent kidney .

- Diagnosis not suspected unless careful examination of the external and internal genitalia uncovers an abnormality that is associated with renal agenesis or an imaging study is done.

- Unilateral agenesis occurs once in 1100 births

- Males predominate in a ratio of 1.8:1

- More frequent on the left side

- Ipsilateral ureter is completely absent in about half of the patients

- Structures derived from the müllerian or wolffian duct are most often anomalous

- Anomalies of other organ systems involve the cardiovascular (30%), gastrointestinal (25%), and musculoskeletal (14%) systems

Unilateral renal agenesis to be associated with other urologic abnormalities in 48% of patients

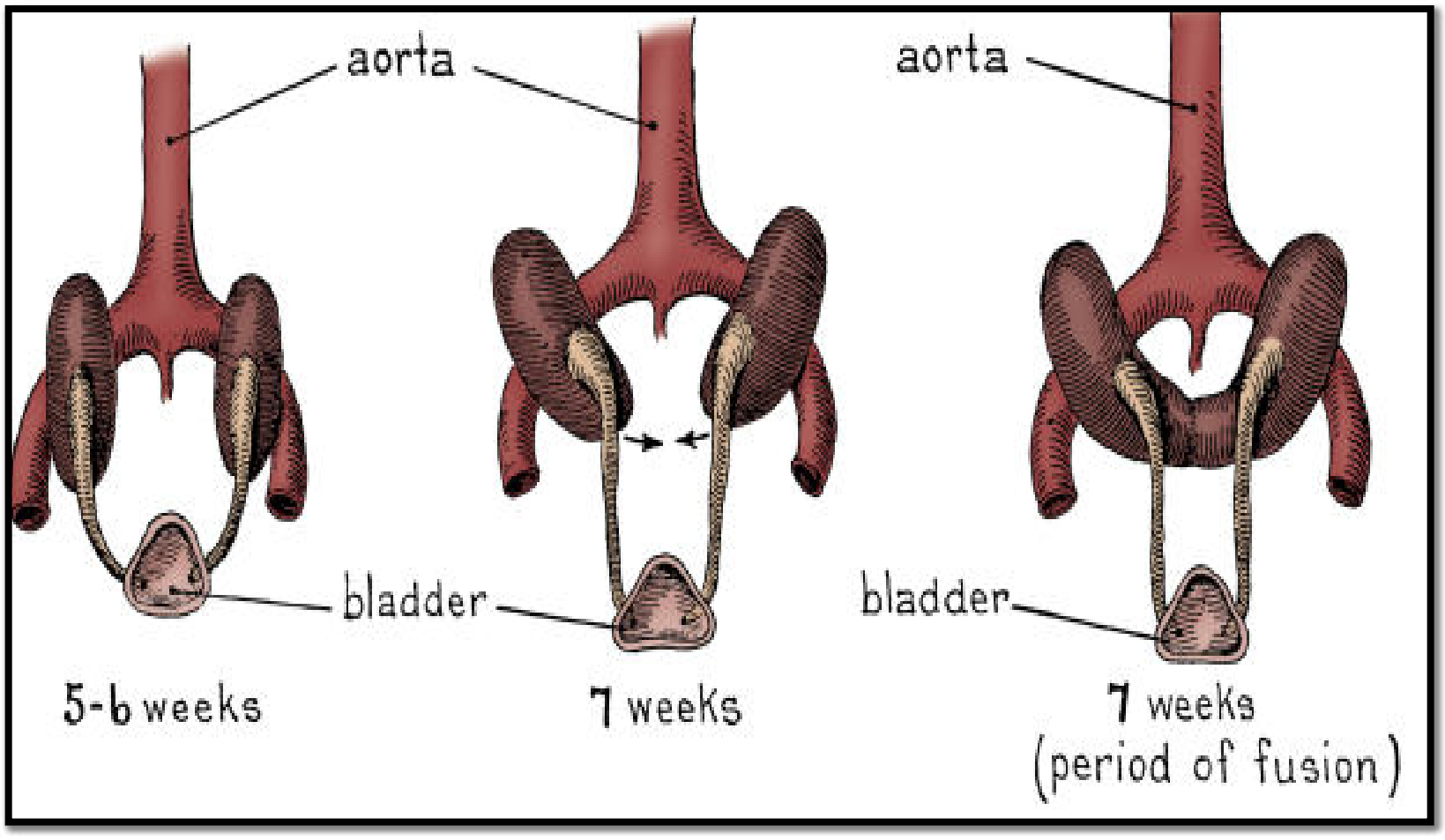
- Primary vesicoureteral reflux (28%)
- Obstructive megaureter (11%)
- Ureteropelvic junction obstruction (3%)

Diagnosis

- No specific symptoms heralding an absent kidney
- The diagnosis should be suspected during a physical examination when the vas deferens or body and tail of the epididymis is missing or hypoplastic vagina is associated with a unicornuate or bicornuate uterus
- Radionuclide imaging
- No clear-cut evidence that patients with a solitary kidney have an increased susceptibility to other diseases

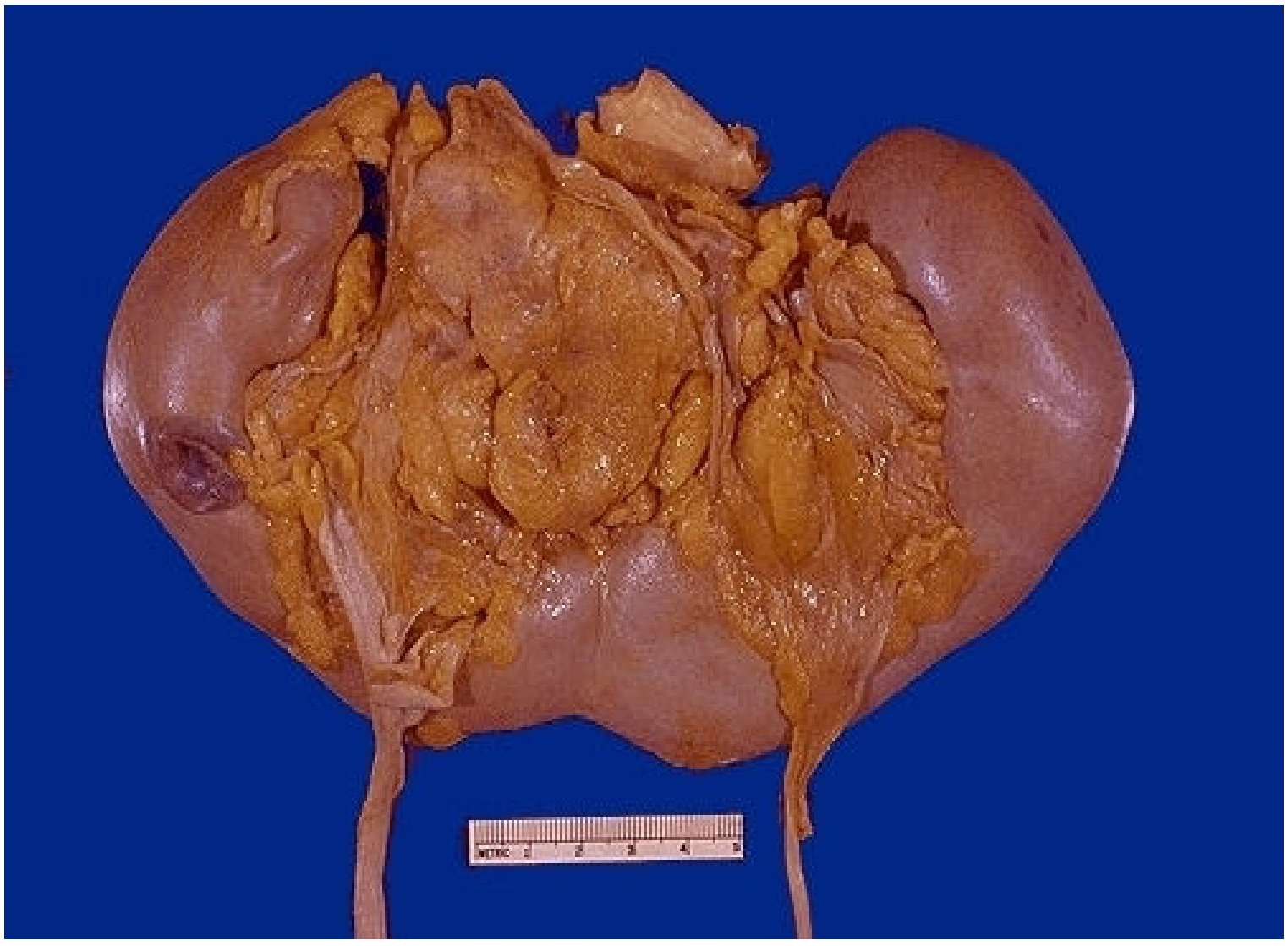
Horseshoe kidneys

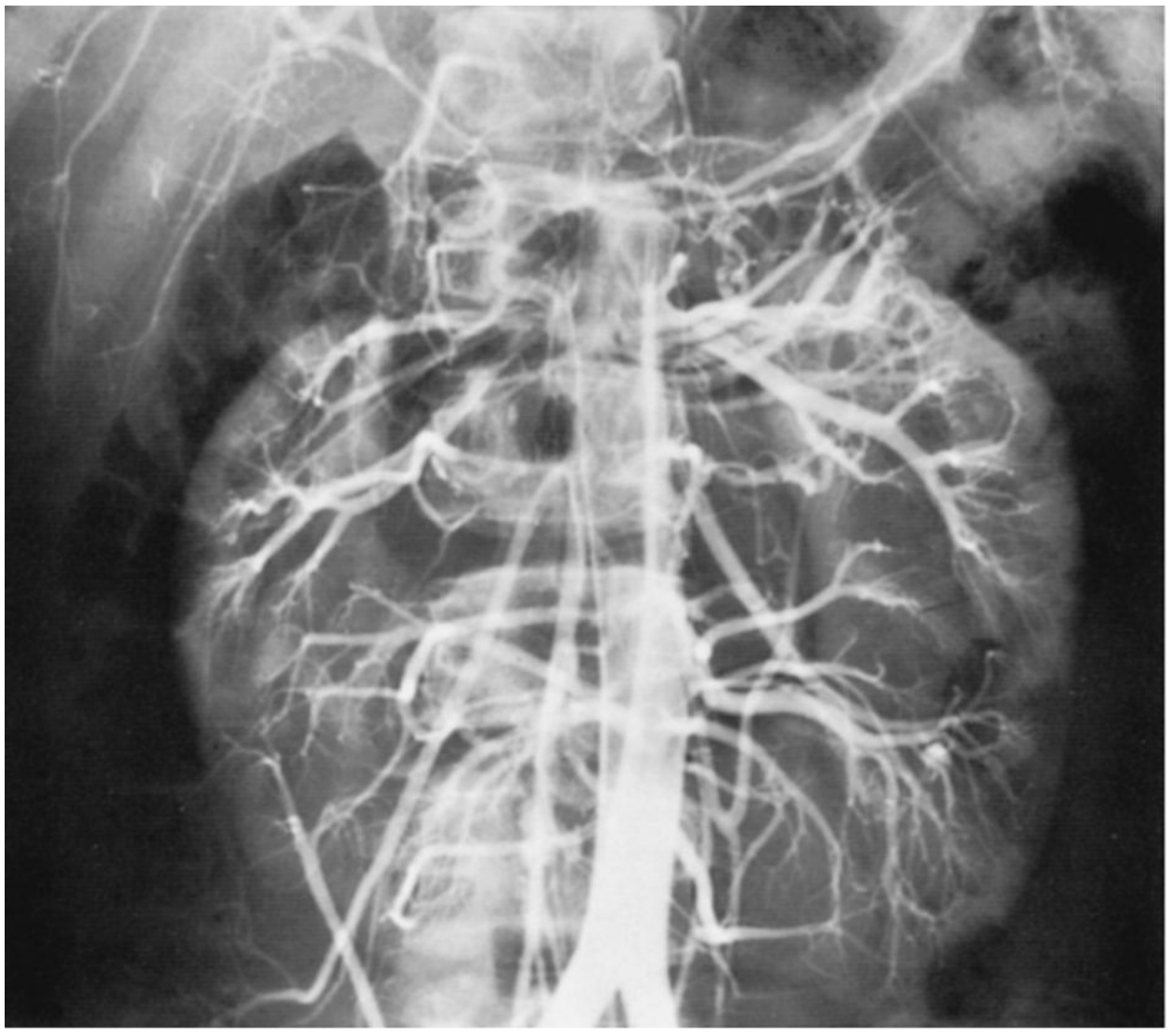
- Most common of all renal fusion anomalies
- Occurs in 0.25% of the population
- fusion occurs before the kidneys have rotated on their long axis



The lower poles of the two kidneys touch and fuse as they cross the iliac arteries

- In 95% of patients, the kidneys join at the lower pole; in a small number, an isthmus connects both upper poles instead
- Calyces normal in number, are atypical in orientation
- Ureter may insert high on the renal pelvis and lie laterally
- Blood supply to the horseshoe kidney can be quite variable





Arteriogram showing a multiplicity of arteries supplying kidney arising from aorta and common iliac arteries

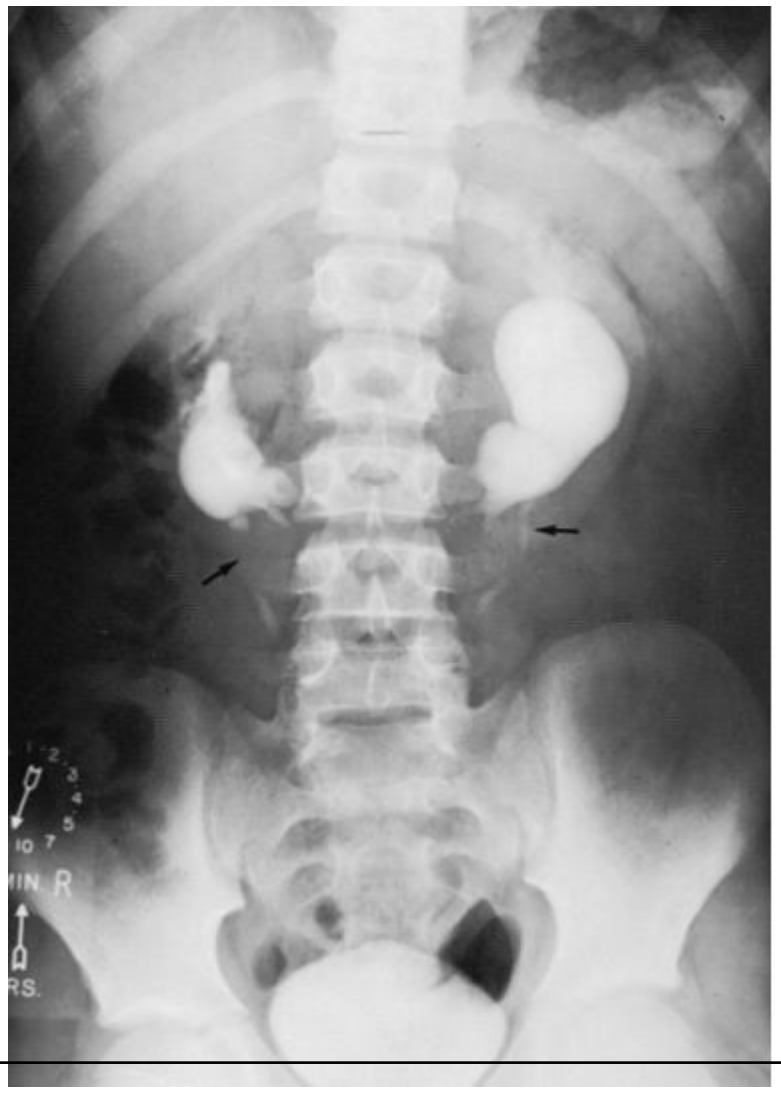
- UPJ obstruction, causing hydronephrosis, occurs in one third of individuals
- 60% patients remain asymptomatic for aprox. 10 years

Associated Anomalies

Genital Anomalies	
Hypospadias	4%
Undescended testes	4%
Bicornuate uterus	7%
Septate vagina	7%
Urinary Collecting System	
Ureteral duplication	10%
Ureteropelvic junction obstruction	20%
Vesicoureteral reflux	50%
Renal Parenchymal Abnormalities	
Multicystic dysplasia	1%
Autosomal recessive polycystic kidney	1%
Metabolic Derangements in Patients with Stones	
Hypercalciuria, hyperoxaluria, hypocitraturia, hypouricuria	50%

Diagnosis

- **Excretory urogram**

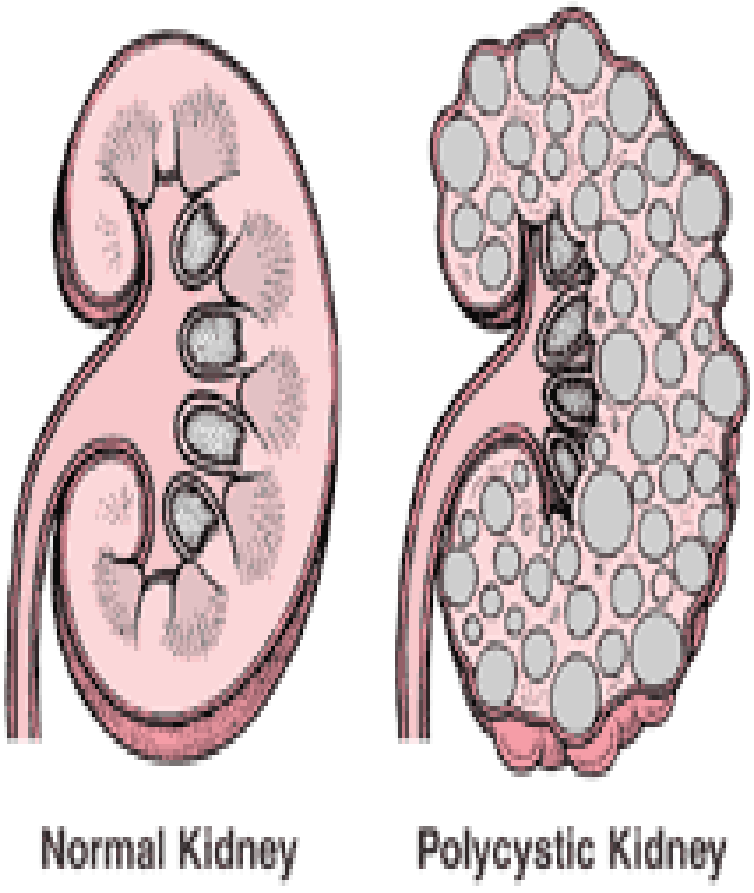


Prognosis

- 13% have persistent urinary infection or pain
- 17% develop recurrent calculi
- Renal carcinoma has been reported within a horseshoe kidney in 123 patients
- Incidence of Wilms' tumor in horseshoe kidneys is more than twice

Congenital cysts

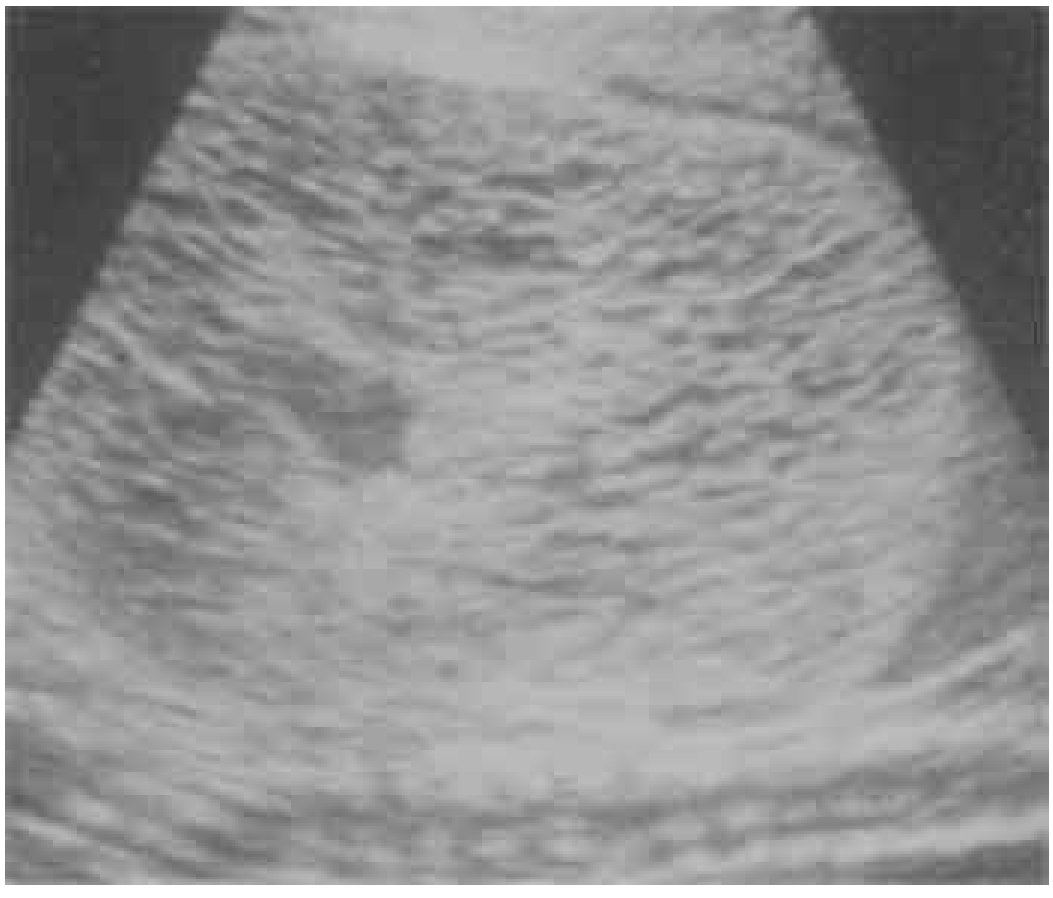
- Kidney is one of the MC sites in body for cysts
- Arise from the nephrons and collecting ducts after they have formed



Genetic
Autosomal recessive (infantile) polycystic kidney disease
Autosomal dominant (adult) polycystic kidney disease
Juvenile nephronophthisis/medullary cystic disease complex
Juvenile nephronophthisis (autosomal recessive)
Medullary cystic disease (autosomal dominant)
Congenital nephrosis (familial nephrotic syndrome) (autosomal recessive)
Familial hypoplastic glomerulocystic disease (autosomal dominant)
Multiple malformation syndromes with renal cysts (e.g., tuberous sclerosis, von Hippel-Lindau disease)
Nongenetic
Multicystic kidney (multicystic dysplastic kidney)
Benign multilocular cyst (cystic nephroma)
Simple cysts
Medullary sponge kidney
Sporadic glomerulocystic kidney disease
Acquired renal cystic disease
Calyceal diverticulum (pyelogenic cyst)

Cystic Diseases of the Kidney

- Multicystic refers to a dysplastic entity
- Polycystic most inherited, all without dysplasia and all with nephrons throughout the kidney
- Many of the polycystic kidney disease entities progress to renal failure



‘Snowstorm’ appearance of **infantile** polycystic disease

Ectopic Kidney

- Kidney not located in usual position
- 1 in 1,000 births, but only about one in 10 of these are ever diagnosed; up to 10% bilateral

<u>Most common:</u>

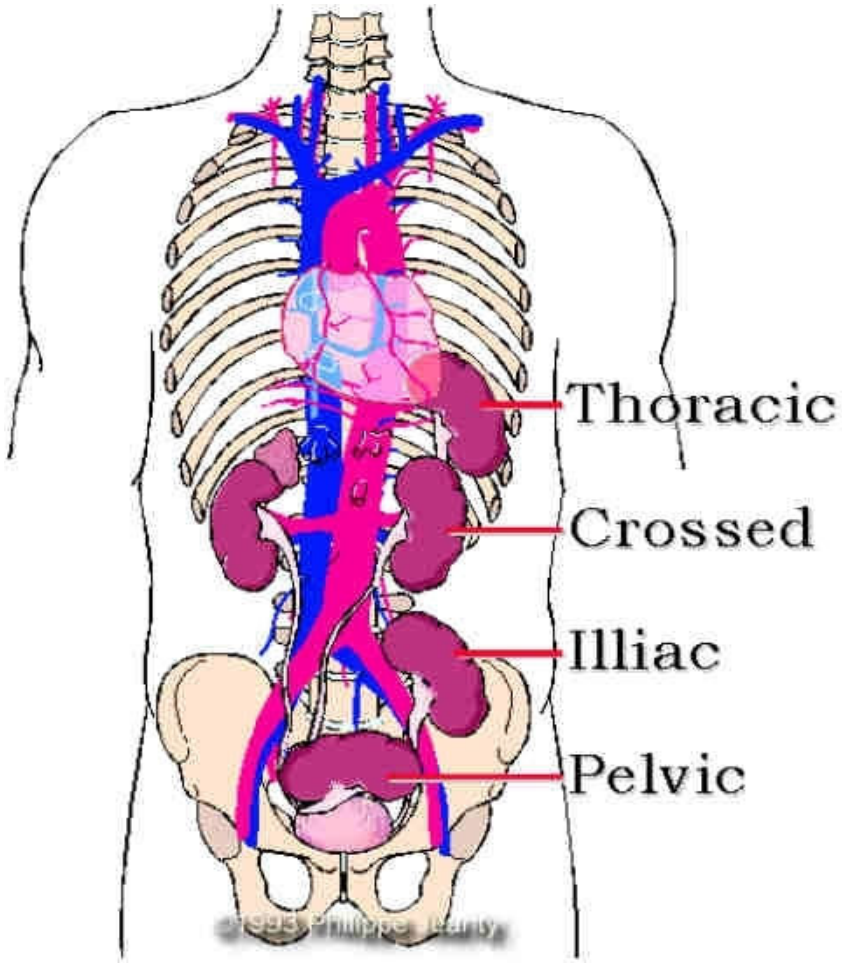
- Horseshoe Kidney
- Unilateral renal agenesis
- Pelvic kidney

~~(Left kidney more likely to be abnormal)~~

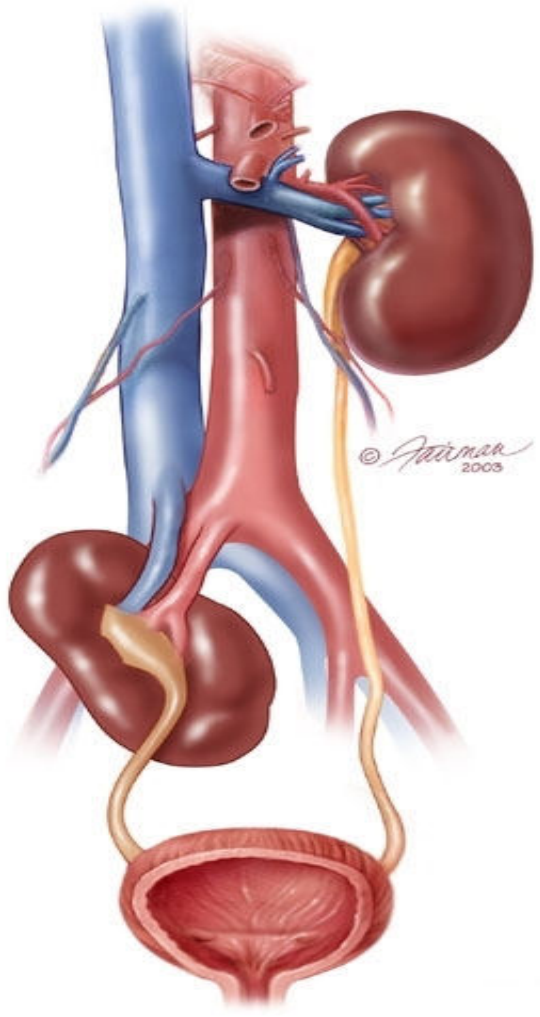
Ectopic Kidney

- Function is generally normal initially
- Abnormal position leads to obstruction in 50% of ectopic kidneys
- Increased risk UTI, kidney stones, VUR
- Frequently associated with abnormalities of other organ systems (uterine, cardiac, skeletal)

Ectopic Kidney Locations

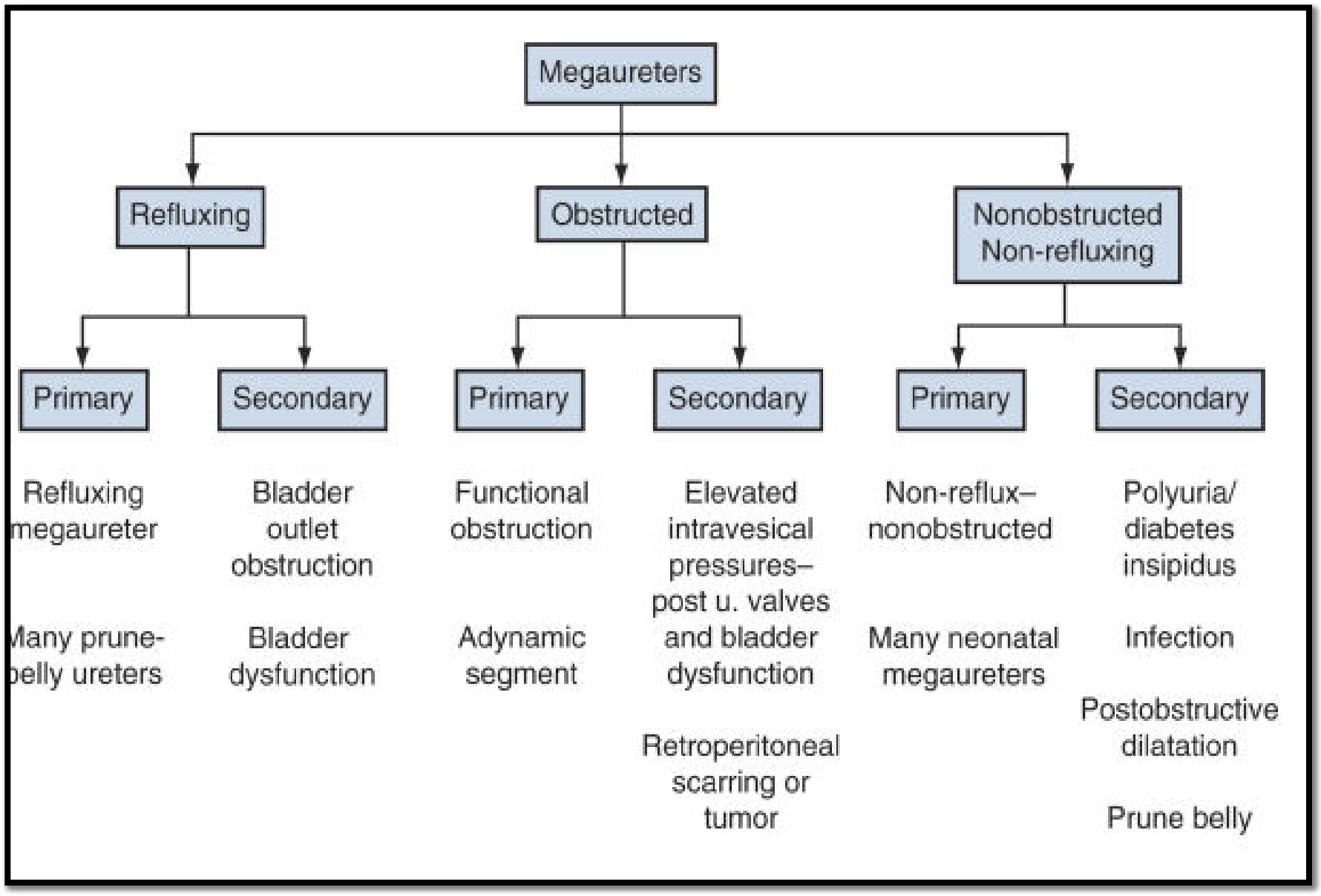


Ectopic Kidney
(simple renal ectopia)



Mega ureter

- Ureters wider than 7 to 8 mm
- Normal ureteral diameter is rarely greater than 5 mm
- Primary MGU is 2-4 times more common in boys than girls
- Slight predilection (1.6 to 4.5 times) for the left side
- Bilateral in approximately 25% of patients
- In 10% to 15% of children contralateral kidney may be absent or dysplastic



Three major classifications of megaureter based on primary and secondary causes

Pathophysiology

- Distal end of the ureter, as it becomes intramural and subsequently submucosal, rearranges the muscular layers in its wall.
- All layers become longitudinally oriented
- Ureteral adventitia fuses to the bladder trigone by attachment to Waldeyer's sheath
- Sympathetic and parasympathetic innervation to the distal ureter and UVJ area is believed to modulate primarily ureteral peristalsis

Diagnosis

Ultrasound

- Distinguishes MGU from UPJ obstruction based on the presence or absence of a dilated ureter

VCUG

- to rule out reflux

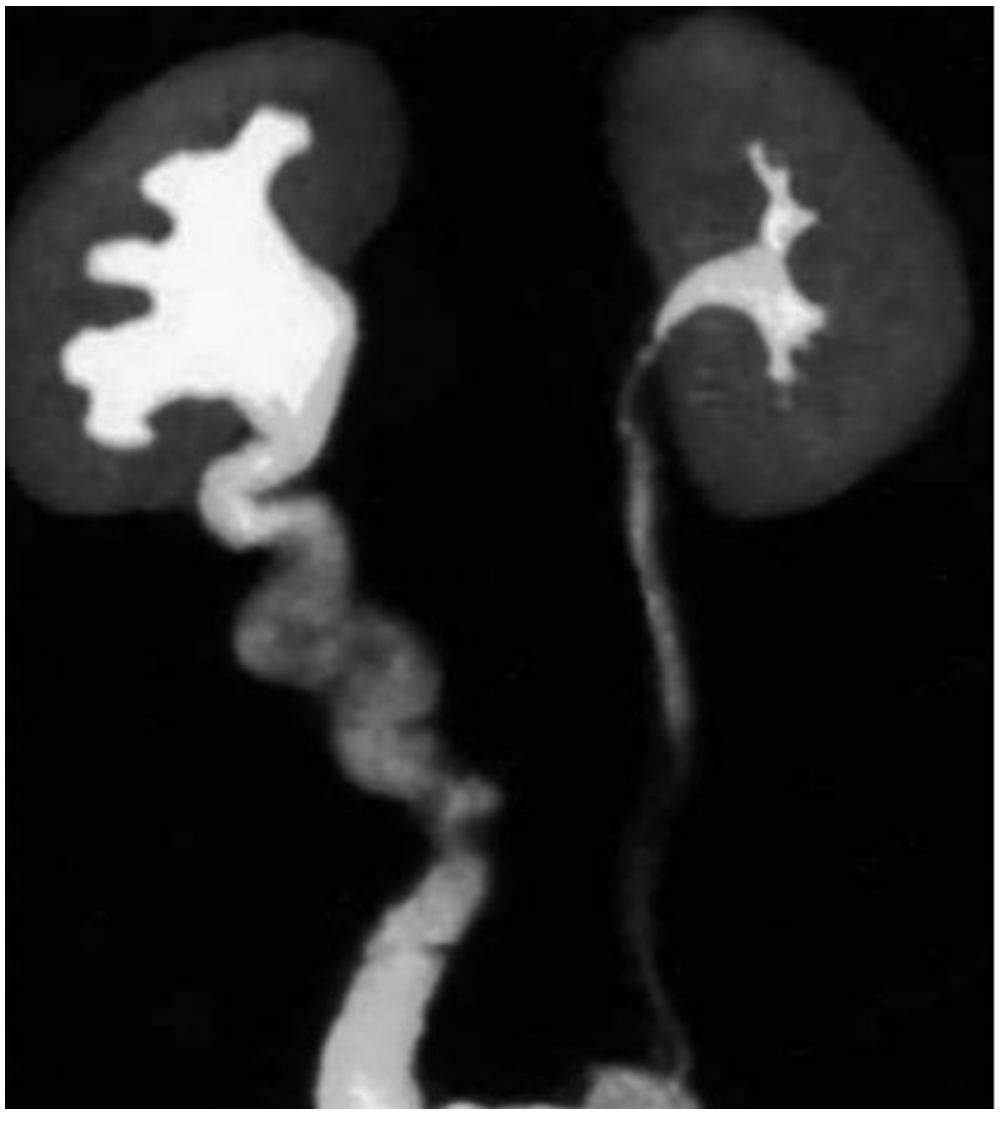
Renal scintigraphy

- Provides objective, reproducible parameters of function and obstruction

Whitaker's perfusion test & ureteral opening pressure

- To evaluate obstruction, but their invasiveness and requirement for anaesthesia are drawbacks in children

Magnetic resonance urography



Magnetic resonance urogram showing obstruction at the right ureterovesical junction

Management

Primary Refluxing Megaureter

- Medical management is often the initial approach

- **Surgery**
  - Endoscopic subureteric injection, is recommended for persistent high-grade reflux in older children

- **Reconstructive surgery of a dilated ureter**
  - distal ureterostomy for unilateral reflux
  - vesicostomy for bilateral disease

- **Secondary Refluxing or Obstructive Megaureter**
  - Management of secondary MGUs is initially directed at their root cause

- **Primary "Dilated" Nonrefluxing Megaureter: Nonobstructive versus Obstructive**
  - Expectant management is preferred
  - Antibiotic suppression & radiologic surveillance is appropriate in most cases
  - Surgical correction

- **Surgical Options**
  - Plication or infolding for moderately dilated ureter

Complications

Persistent reflux and obstruction

Postoperative VUR

Ectopic ureter

- Ureter whose orifice terminates anywhere other than the normal trigonal position
- **Lateral ectopia** : an orifice more cranial and lateral than normal
- **Caudal ectopia** : orifice is more medial and distal than the normal position

- 80% are associated with a duplicated collecting system
- Females :
  - More than 80% are duplicated
  - Urethra and vestibule are the most common sites
- Males:
  - most ectopic ureters drain single systems
  - posterior urethra is the most common site
- Drainage into the genital tract involves the seminal vesicle three times more often than the ejaculatory duct and vas deferens combined

Ureterocele

(outpouching of ureter as it enters bladder)