Congenital Anomalies of The Upper Urinary Tract

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Congenital anomalies of the upper urinary tract comprise a group of abnormalities, ranging from complete absence to aberrant location, orientation, and shape of the kidney as well as aberrations of the collecting system and blood supply.

Surgical anatomy

The parenchyma of each kidney usually drains into seven calyces, three upper, two middle and two lower calyces. Each of the three segments represents an anatomically distinct unit with its own blood supply.

The kidney and renal pelvis normally rotate 90 degrees ventromedially (toward midline) as they leave the true pelvis during beginning of ascent at 6^{th} week of gestation so that the calyces point laterally and the pelvis faces medially. When this alignment is not exact, the condition is known as malrotation

Anomalies of the upper urinary tracts

- Anomalies of number
  - Unilateral renal agenesis
  - Bilateral renal agenesis
  - Supernumerary kidney
- Anomalies of ascent
  - Simple ectopia
  - Cephalad ectopia
  - Thoracic kidney
- Anomalies of form and fusion
  - Crossed ectopia without fusion
  - Crossed ectopia with fusion
  - Horseshoe kidney
- Anomalies of volume and structure
  - Polycystic kidney
  - Simple cyst
  - Hypoplasia and dysplasia
- Anomalies of the collecting system
  - Pelvis
  - Calyx and infundibulum
- Anomalies of rotation
  - Incomplete
  - Excessive
  - Reverse
- Anomalies of renal vasculature
  - Aberrant, accessory, or multiple vessels
  - Renal artery aneurysm
  - Arteriovenous fistula

Unilateral Renal Agenesis (URA)

Found accidentally, more frequently on the left side.
Ipsilateral adrenal agenesis is rarely encountered with URA

Symptoms: Asymptomatic
Diagnosis: U/S or IVU,CT scan: absent kidney on that side + compensatory hypertrophy of the contralateral kidney
Treatment: no specific treatment

Bilateral agenesis: rare, incompatible with life

Renal Agenesis

Unilateral renal agenesis
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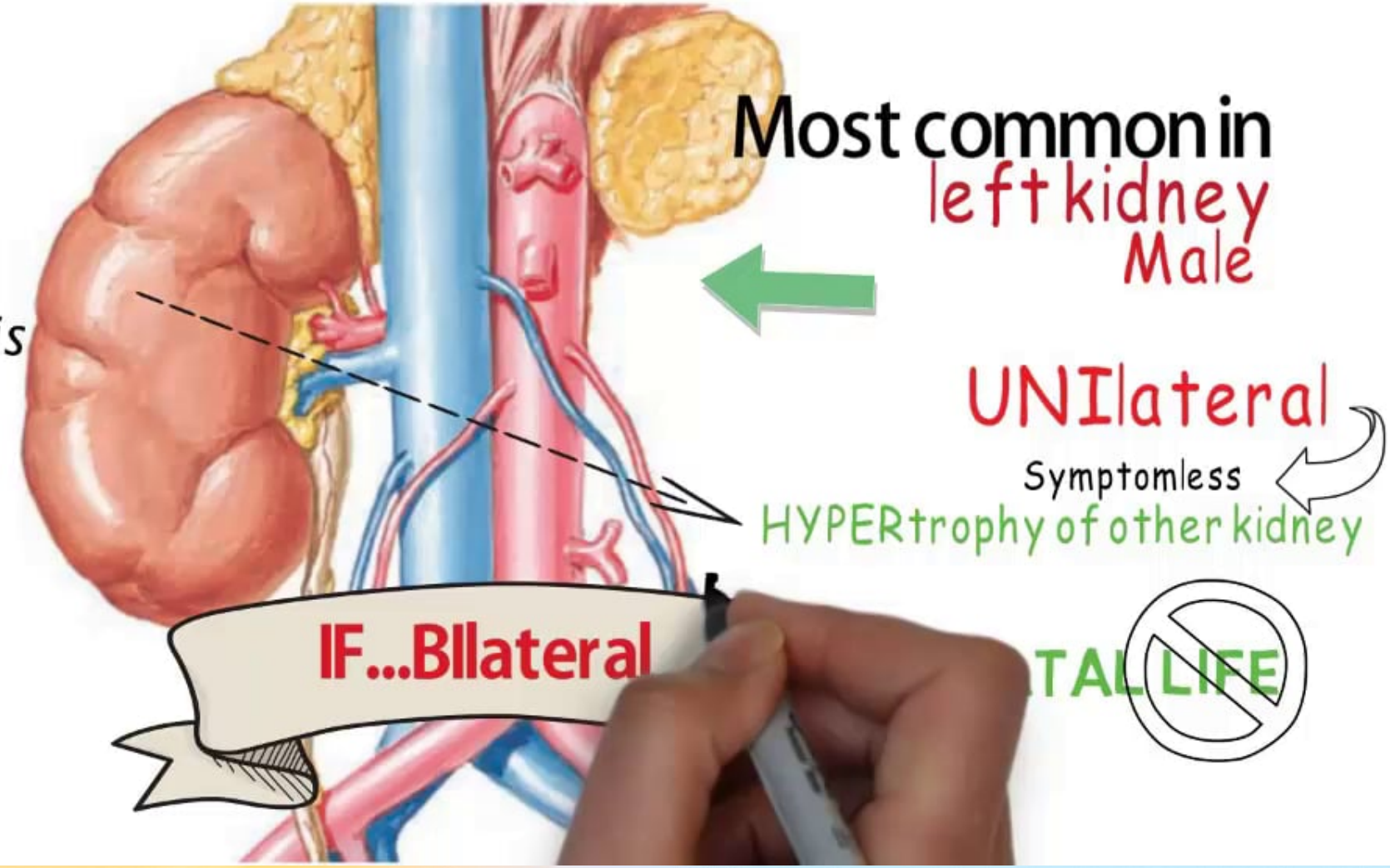
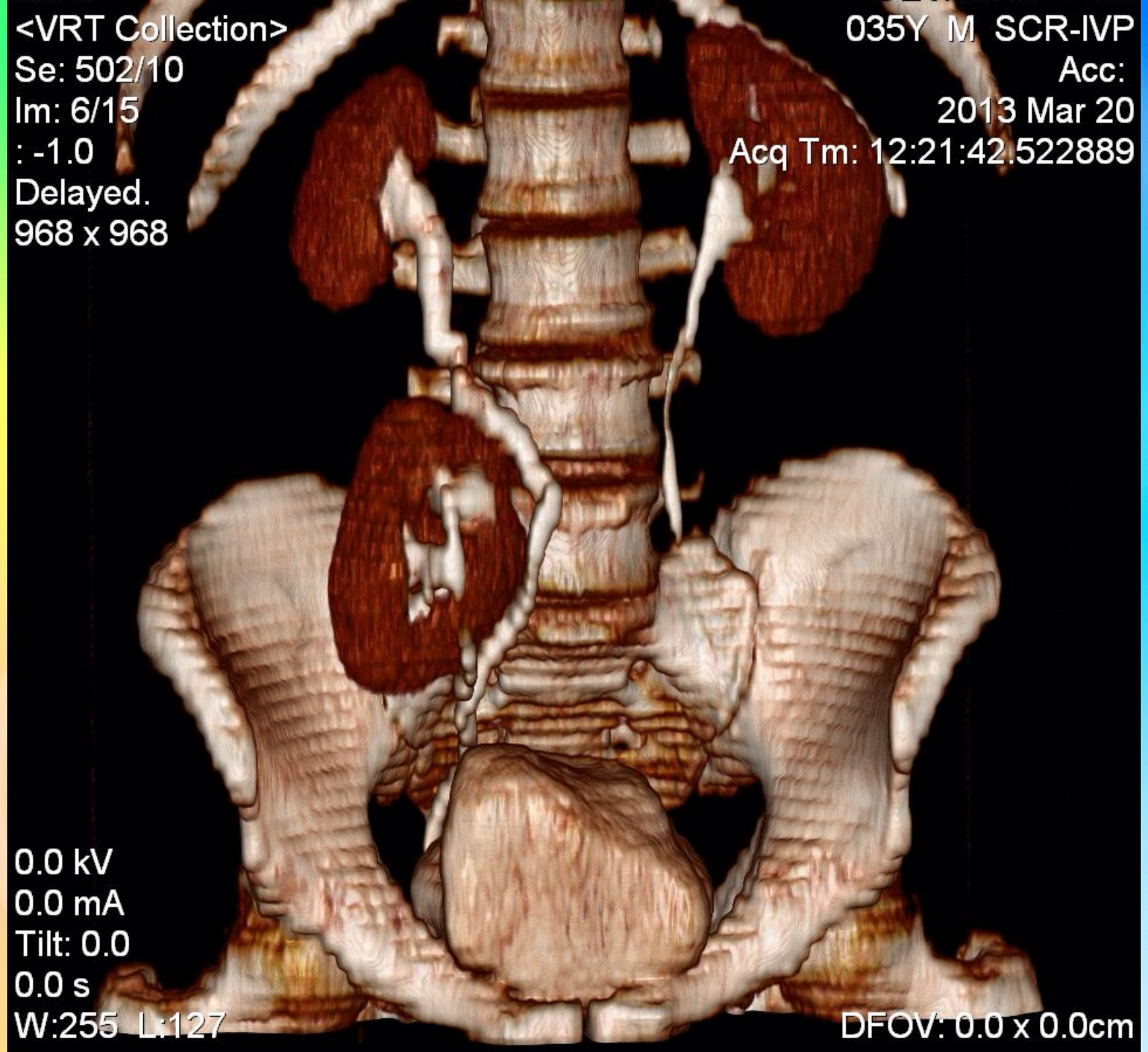


Supernumerary Kidney

The supernumerary kidney is a distinct mass of renal parenchyma that may be either completely separate or only loosely attached to the major kidney on the ipsilateral side.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ANOMALIES OF ASCENT

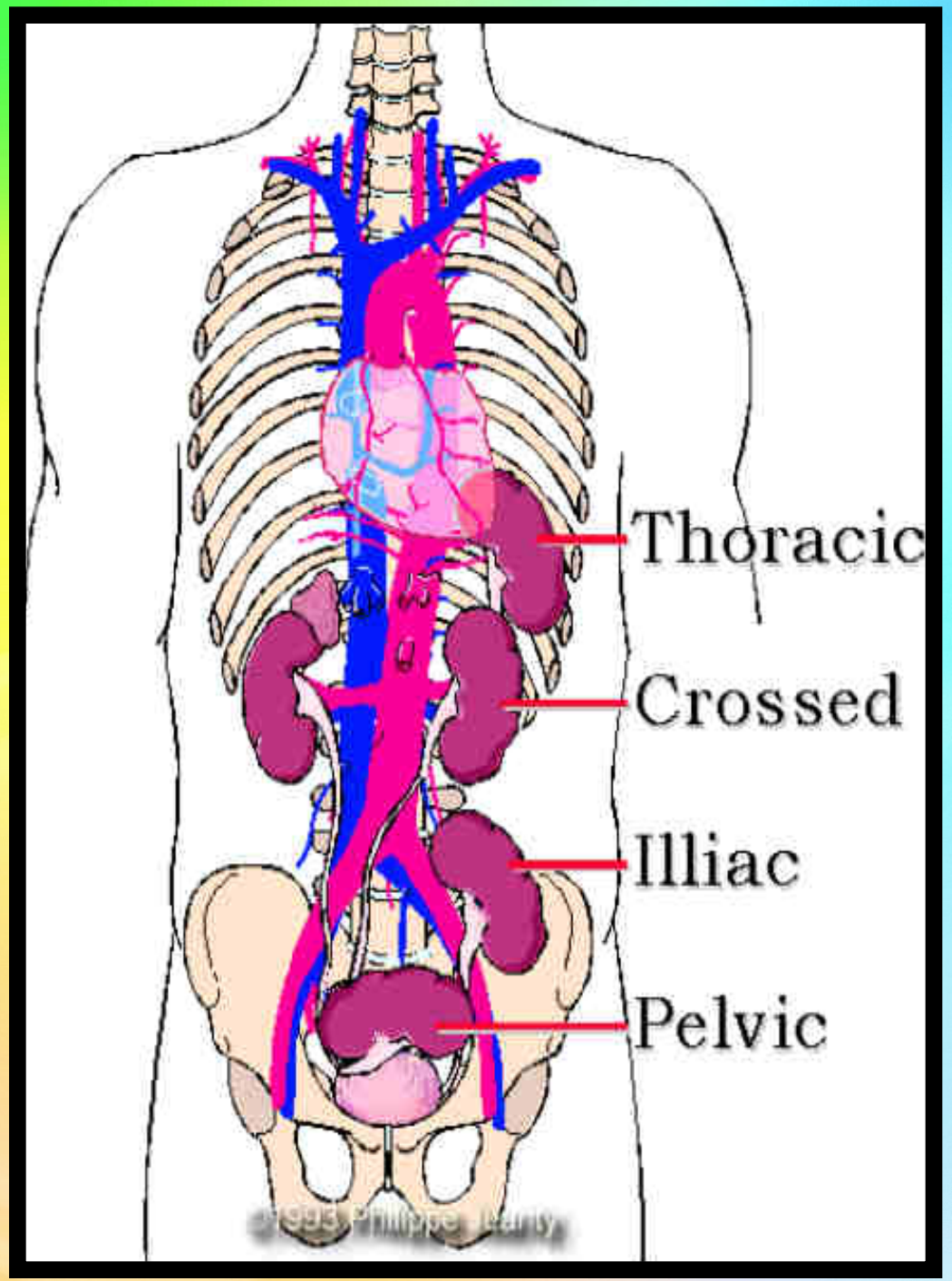
1. Simple Renal Ectopia

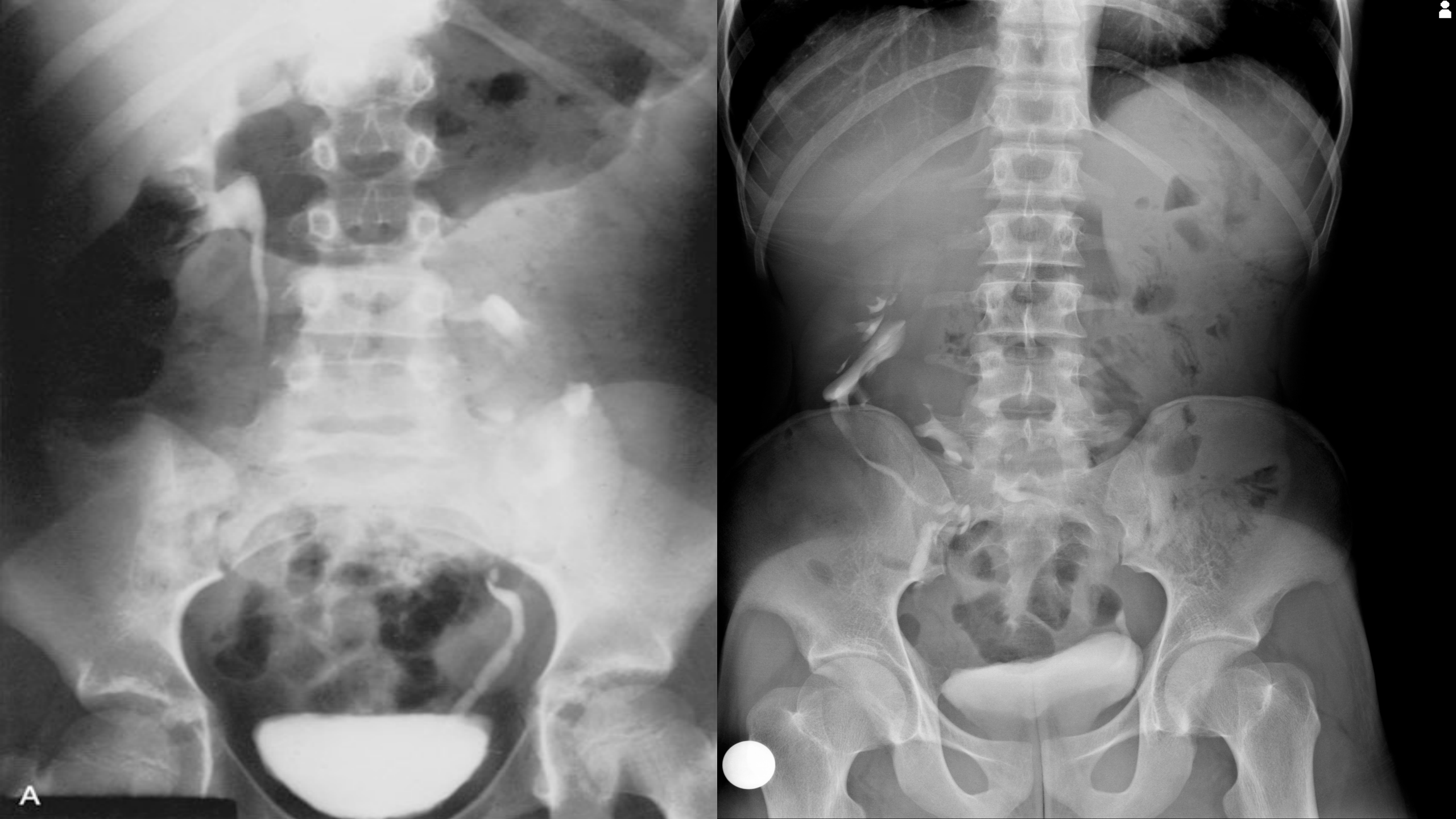
When the mature kidney fails to reach its normal location in the “renal” fossa, the condition is known as renal ectopia. The term is derived from the Greek words ek (“out”) and topos (“place”) and literally means “**out of place**.”

An **ectopic kidney** can be found in one of the following positions:

pelvic, iliac, abdominal, thoracic, and crossed.

The renal pelvis is usually anterior (instead of medial) to the parenchyma, because the kidney has incompletely rotated. As a result, some of ectopic kidneys have a hydronephrotic collecting system due to obstruction of the ureteropelvic or the ureterovesical junction.


Thoracic
Crossed
Illiac
Pelvic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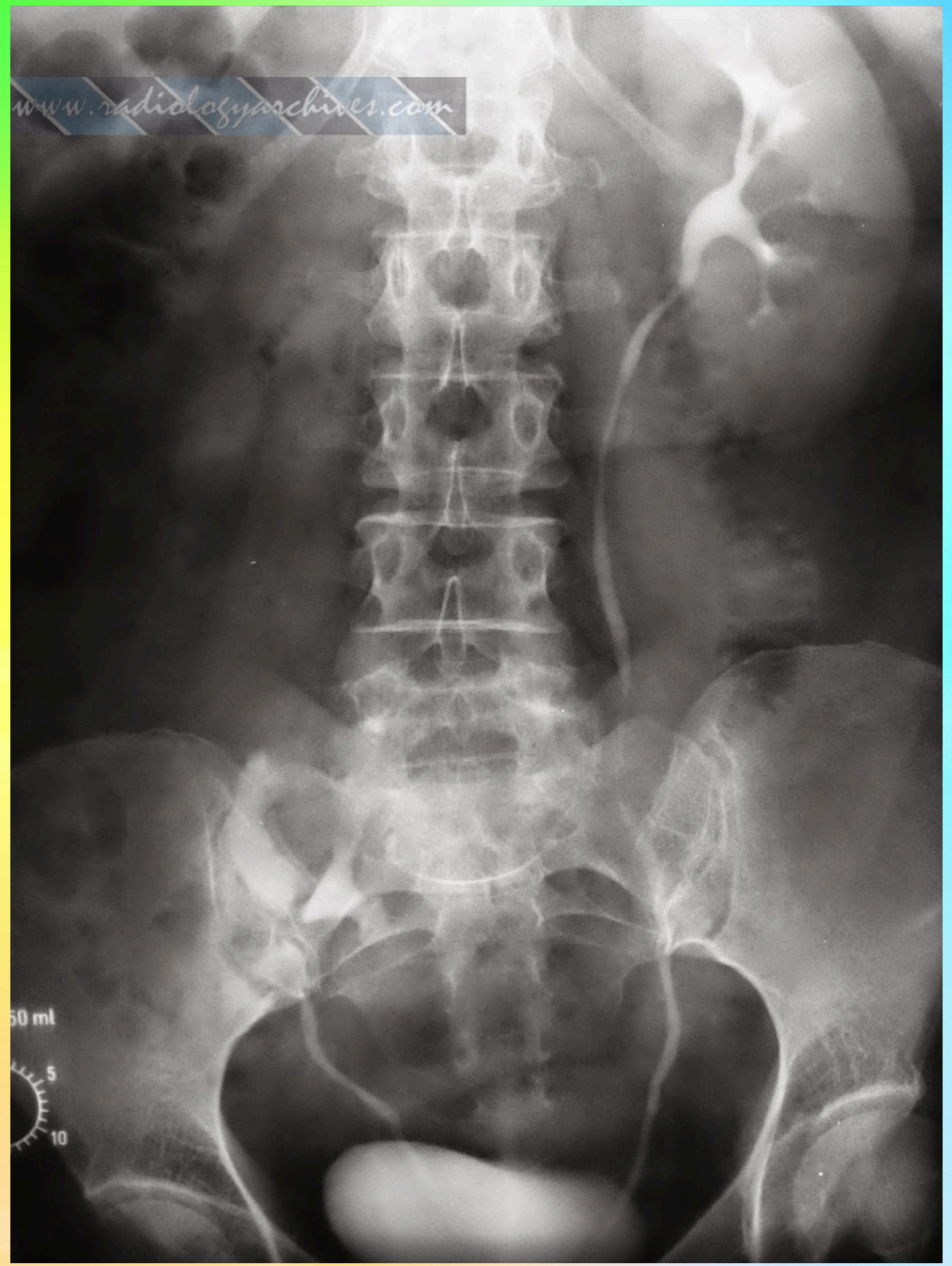
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Associated Anomalies: The incidence of contralateral agenesis appears to be rather high. Hydronephrosis secondary to obstruction or reflux may be seen in the contralateral kidney

Clinical features: Most ectopic kidneys are asymptomatic

Diagnosis: U/S, IVU, CT scan

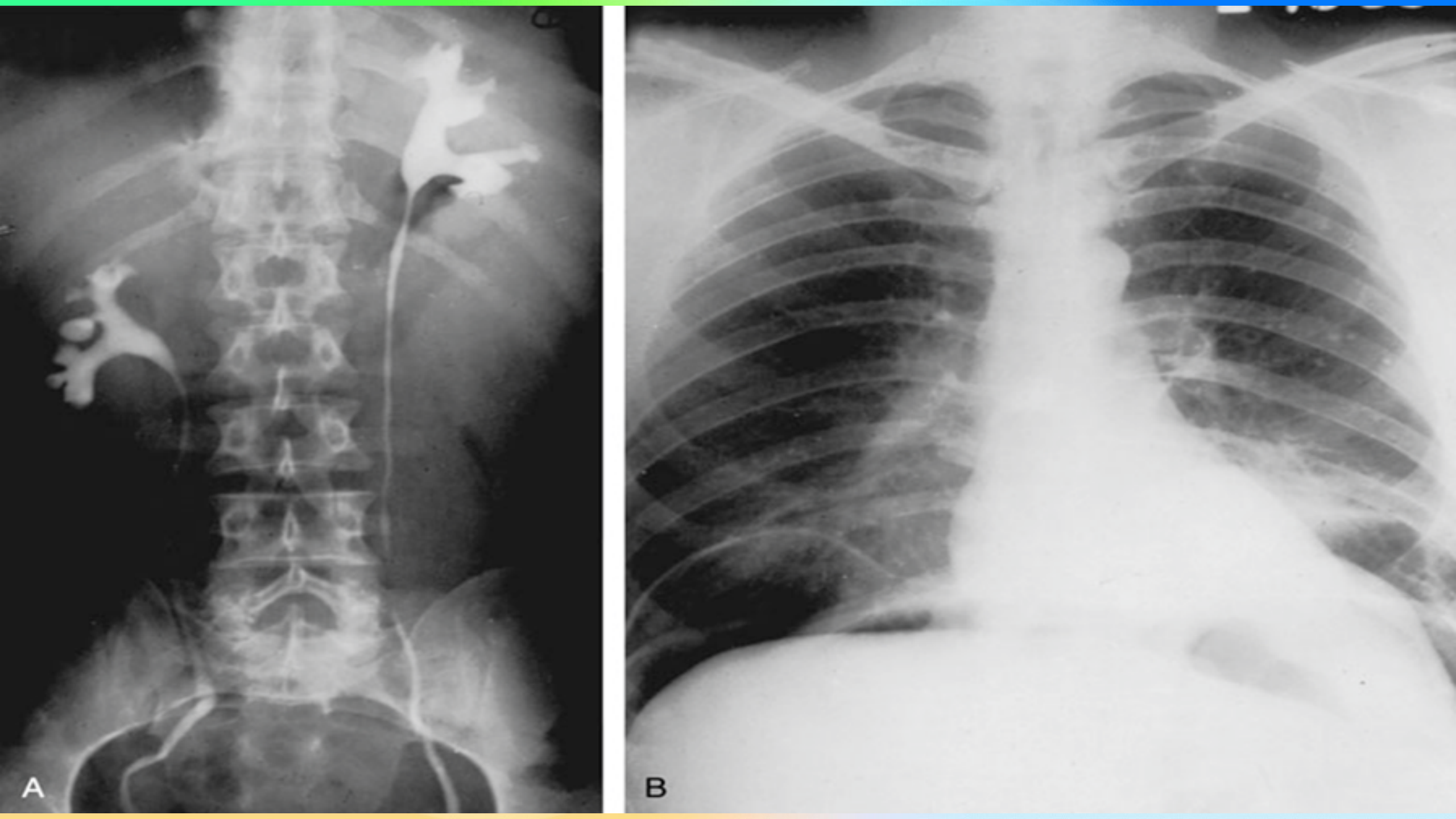
Prognosis: The ectopic kidney is no more susceptible to disease than the normally positioned kidney except for the development of hydronephrosis or urinary calculus formation or the presence of ectopic ureter.

2. Cephalad Renal Ectopia

The kidney may be positioned more cranial than normal.

3. Thoracic Kidney

Intrathoracic ectopia denotes either partial or a complete protrusion of the kidney above the level of the diaphragm into the posterior mediastinum


A
B

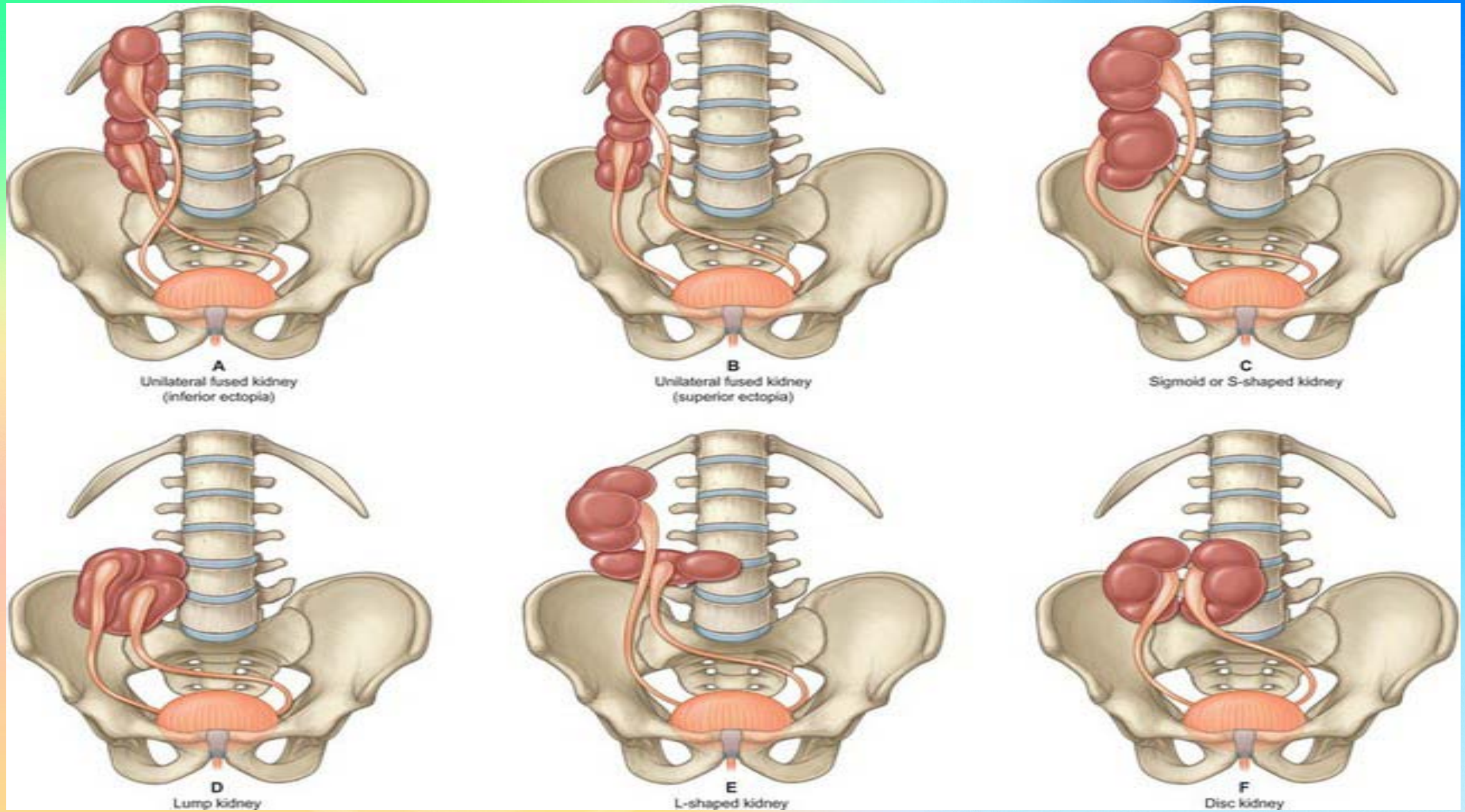
ANOMALIES OF FORM AND FUSION

Crossed Renal Ectopia With and Without Fusion

When a kidney is located on the side opposite from that in which its ureter inserts into the bladder, the condition is known as crossed ectopia.

Types of fused ectopia

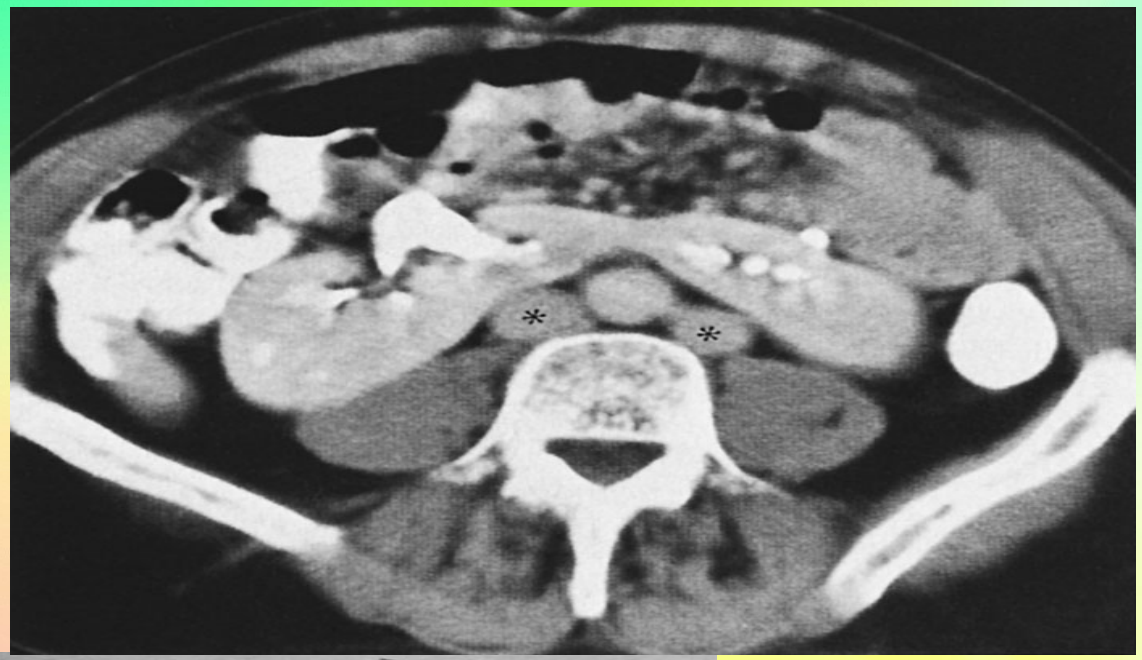
A. Inferior Ectopic Kidney: The upper pole of the crossed kidney is attached to the inferior aspect of the normally positioned mate.
B. Superior Ectopic Kidney: crossed ectopic kidney that lies superior to the normal kidney.
C. Sigmoid, or S-Shaped, Kidney: they face in opposite directions from one another
D. Cake or Lump Kidney: fusion has taken place over a wide margin
E. L-Shaped Kidney: crossed kidney assumes a transverse position.
F. Disc Kidney: joined at the medial borders of each pole

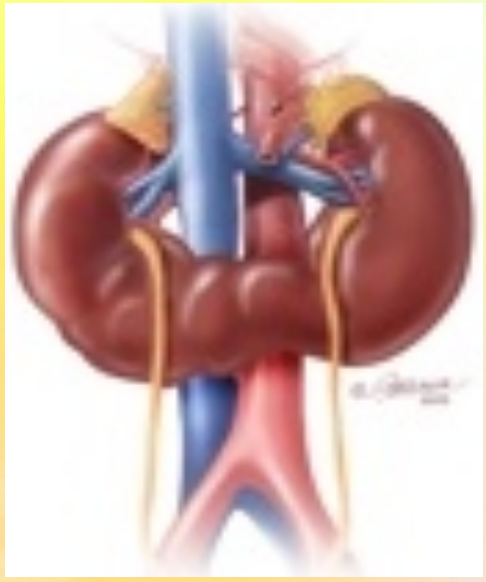

A
Unilateral fused kidney
(inferior ectopia)
B
Unilateral fused kidney
(superior ectopia)
C
Sigmoid or S-shaped kidney
D
Lump kidney
E
L-shaped kidney
F
Disc kidney

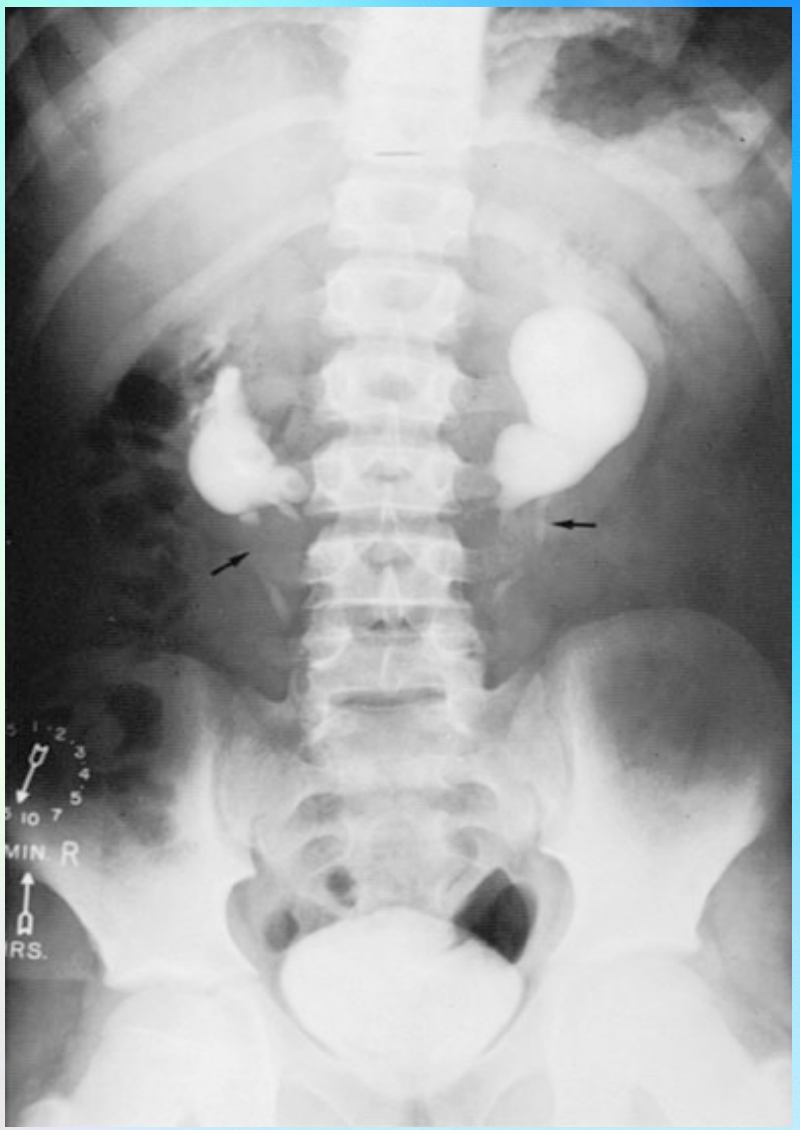
Horseshoe Kidney

- probably the most common of all renal fusion anomalies.
- The anomaly consists of two distinct renal masses lying vertically on either side of the midline and connected at their respective lower poles by a parenchymatous or fibrous isthmus that crosses the midplane of the body.
- Fusion of the renal masses early in embryonic life, so its ascent will be impeded by inferior mesenteric artery.
- The kidneys are low located at the level of the 4^{th} lumbar vertebrae, malrotated and pelves lie anteriorly

Diagnosis: ultrasound, IVU, CT scan







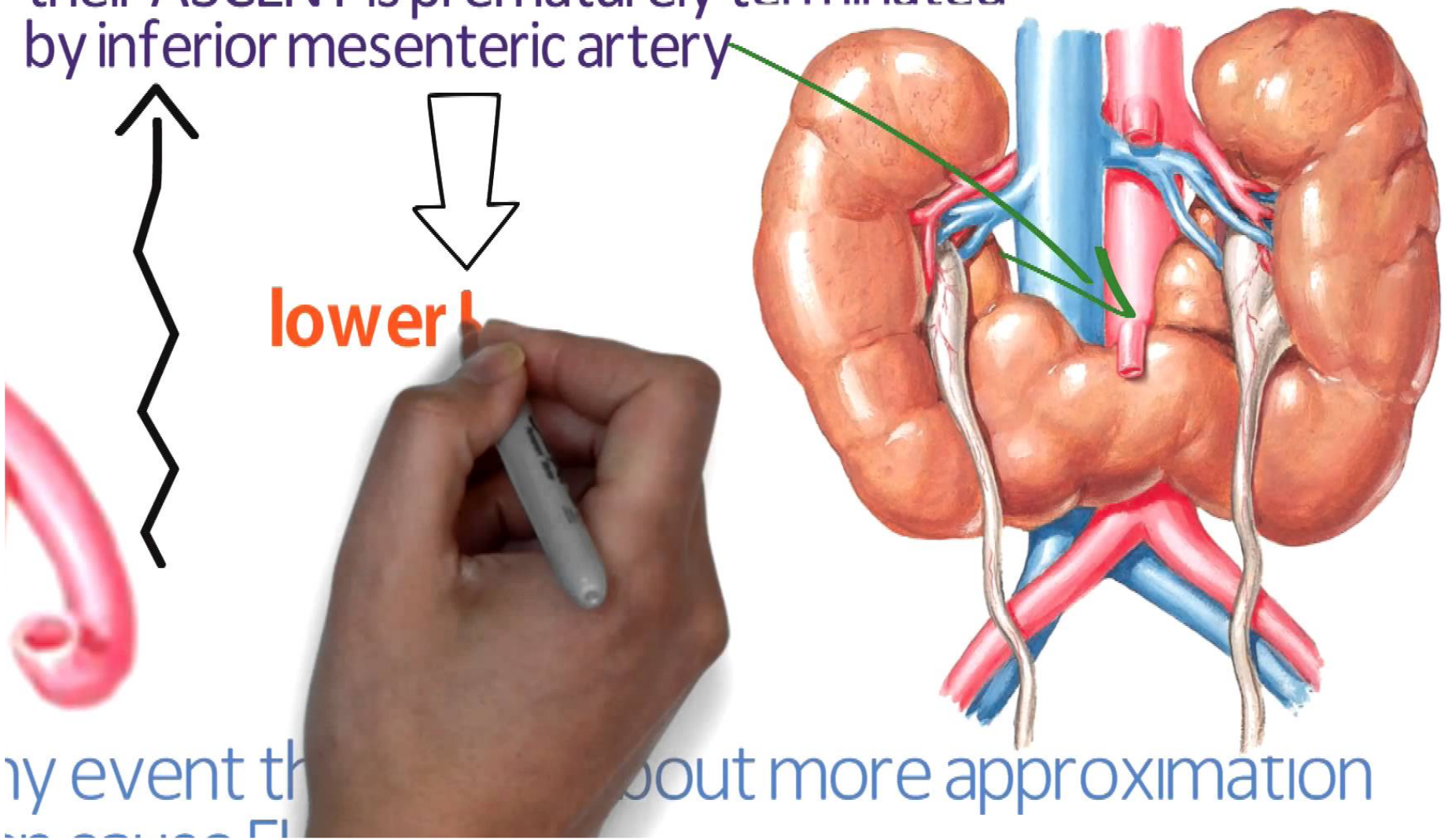
Symptoms: When present, they are related to complications like hydronephrosis, infection, or calculus formation due to ureteric angulation or obstruction with impaired urine drainage

Treatment:

Medical: pain relief and antibiotics to control infection

Surgical: if present, stone removal, Pelviureteric junction obstruction correction.

their ASCENT is prematurely terminated
by inferior mesenteric artery



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Cystic disease of the kidneys

Polycystic kidney disease :

The kidney is one of the most common sites in the body for cysts

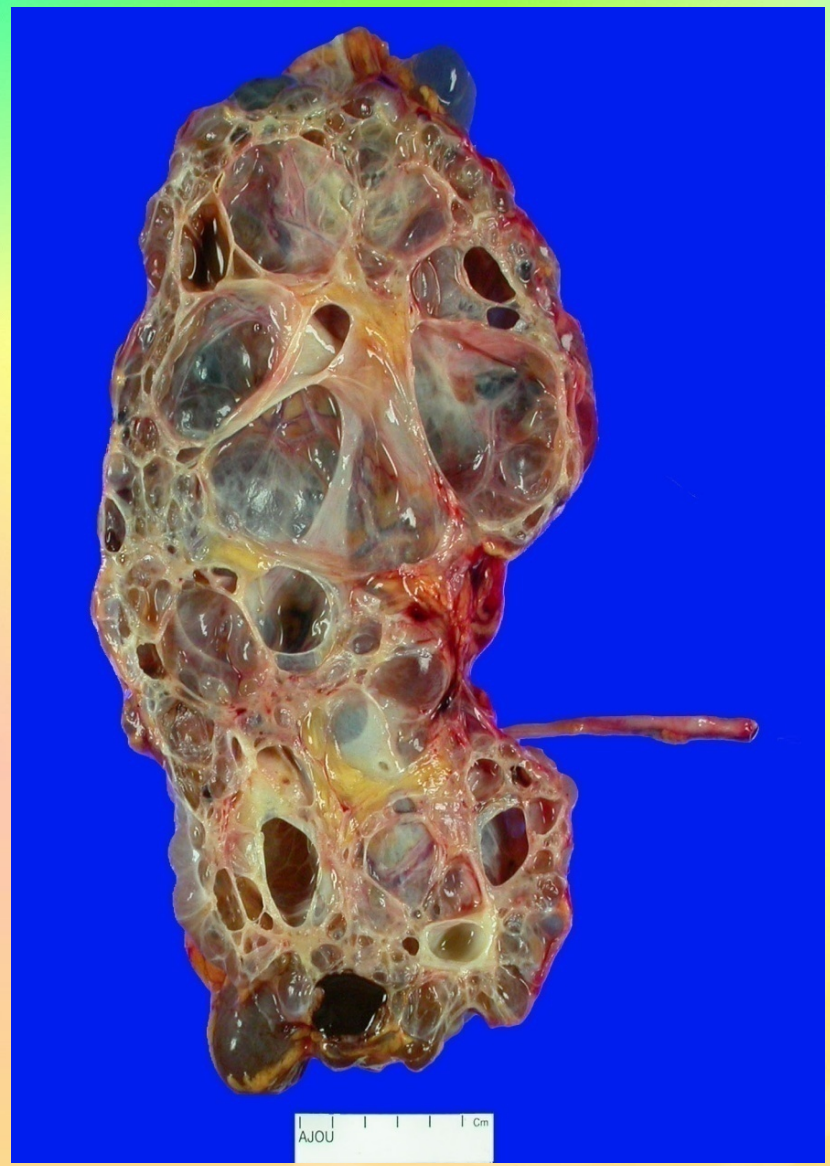
Two types:

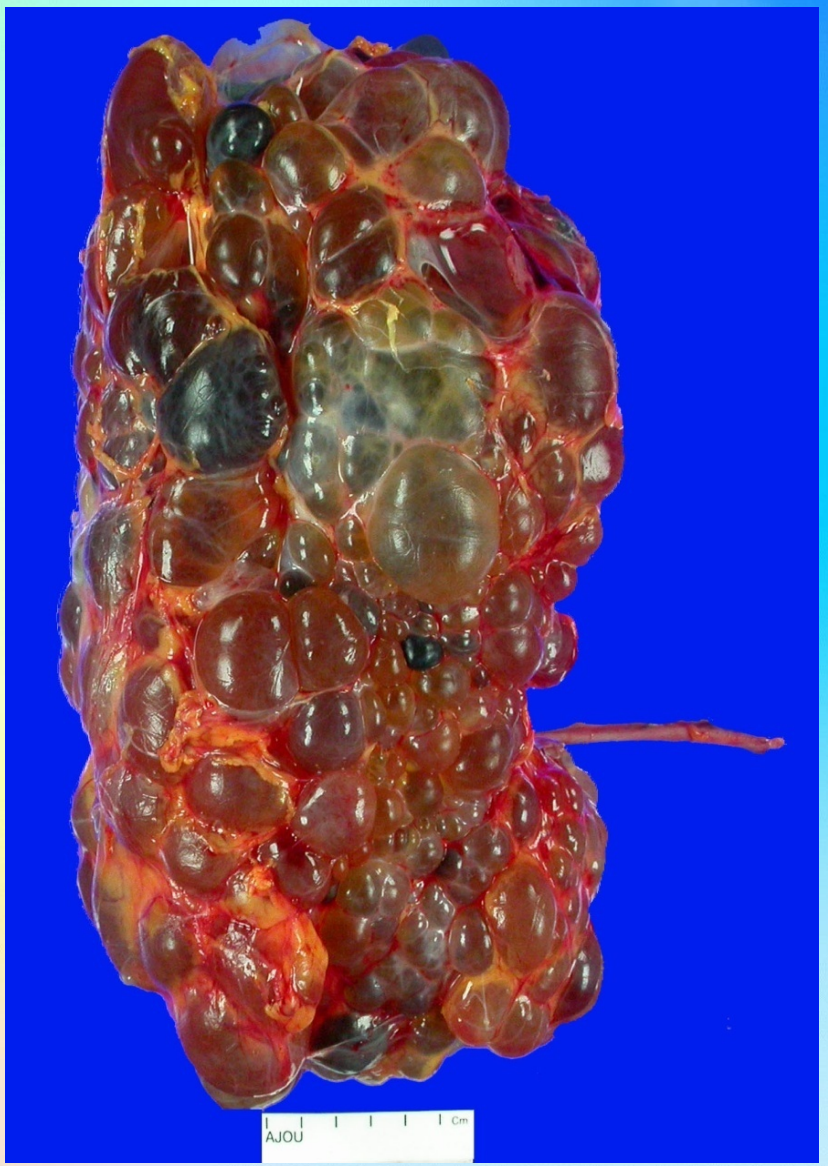
- AUTOSOMAL RECESSIVE ("INFANTILE") POLYCYSTIC KIDNEY DISEASE
- AUTOSOMAL DOMINANT ("ADULT") POLYCYSTIC KIDNEY DISEASE

Autosomal dominant polycystic kidney disease

- Autosomal dominant, transmitted by either parents, 50% of offspring affected.
- Both kidneys replaced by large no. of cysts of variable size which make the kidney of large size.
- 15% associated with cystic disease of liver, lung, pancreas or spleen.

Adult polycystic kidney disease



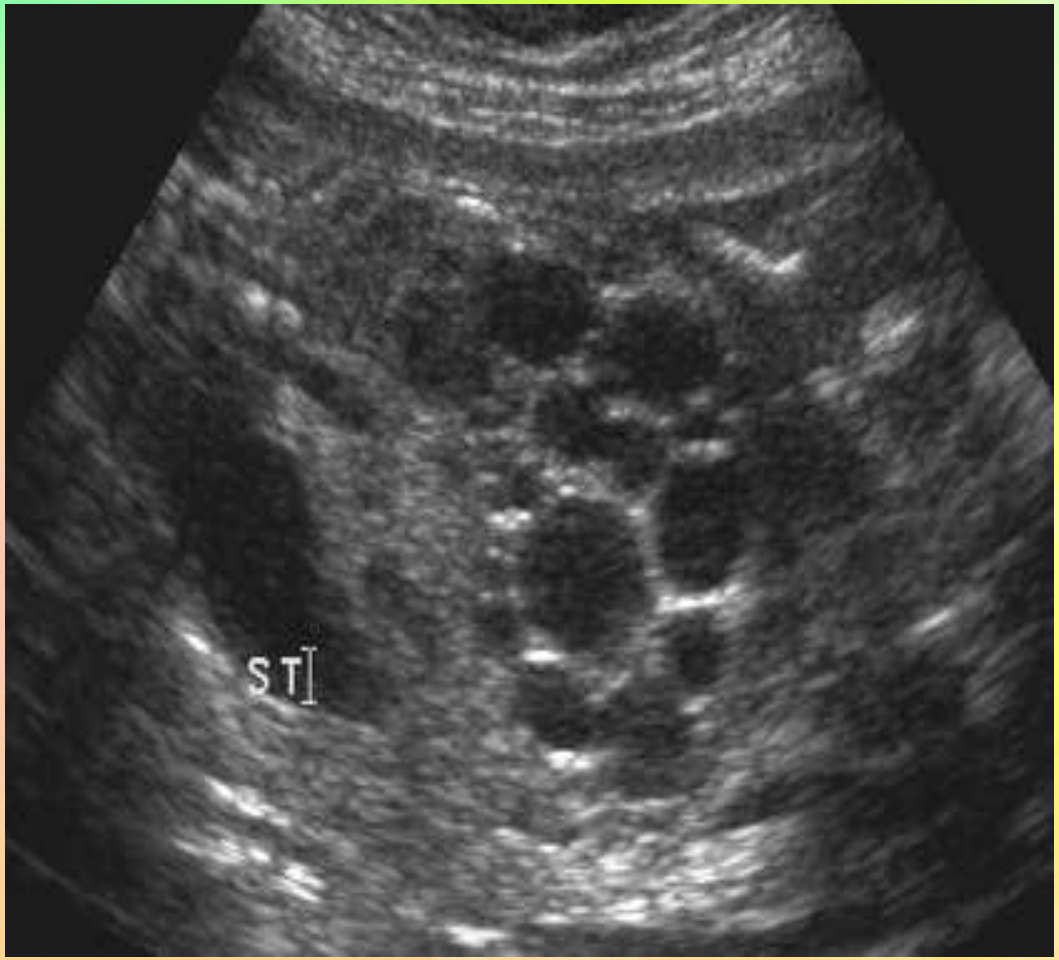


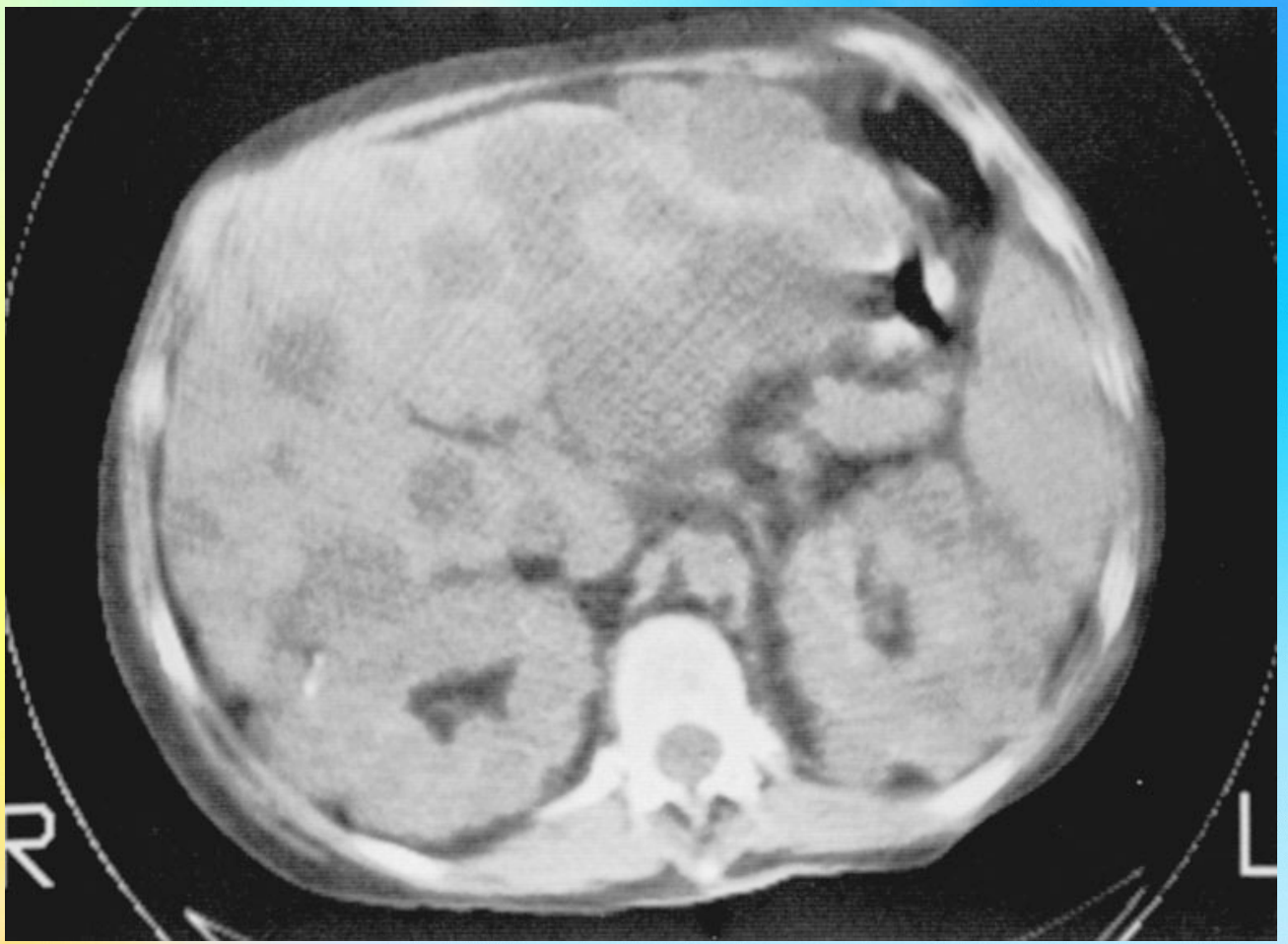
Clinical presentation:

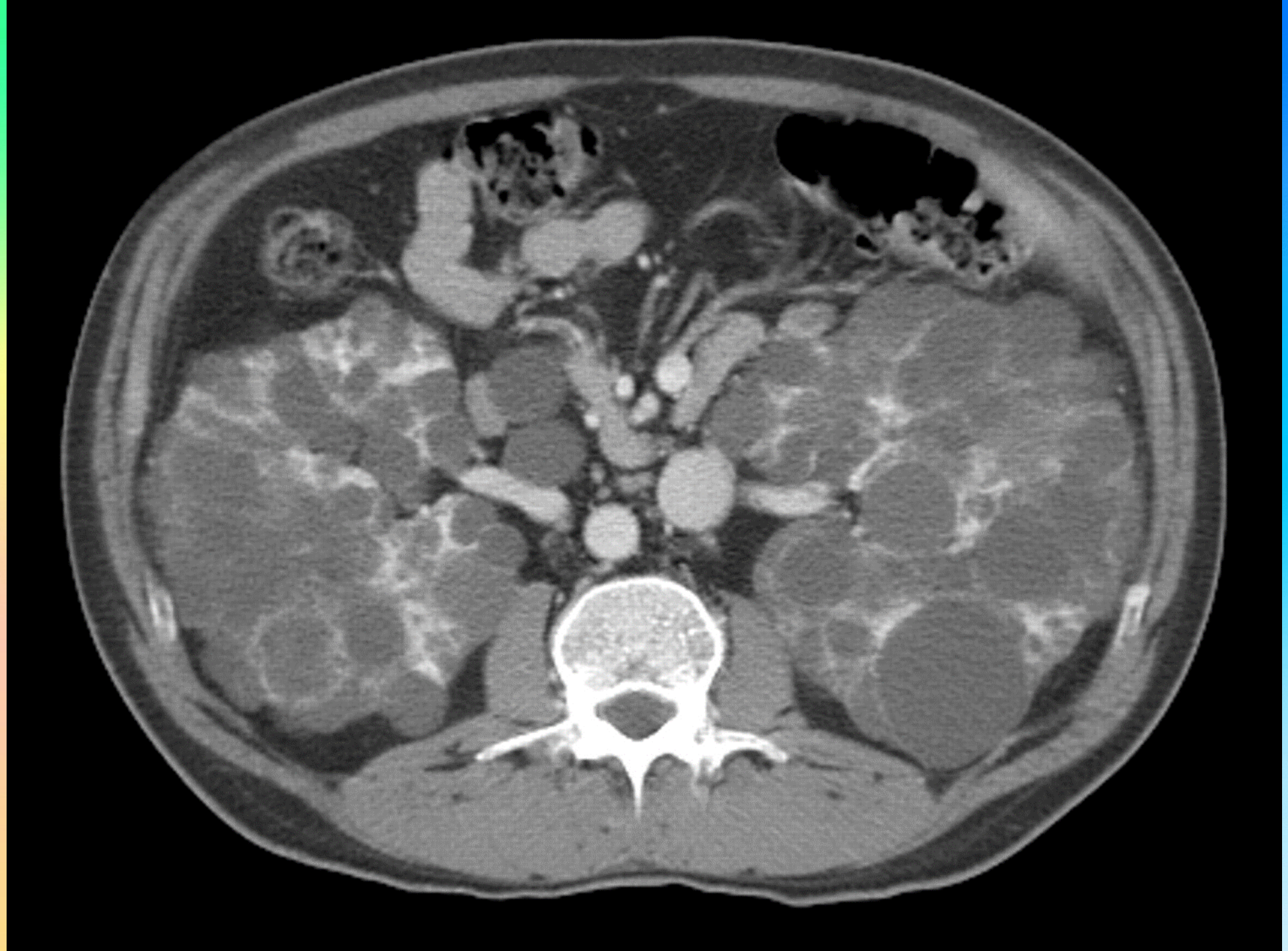
Rarely gives clinical manifestation before 4o years

Asymptomatic
Pain
Hematuria
Infection
Hypertension
Renal impairment
Renal enlargement

Diagnosis: Family history of polycystic disease. U/S, IVU, CT scan, MRI.







Treatment:

Medical:

To control infection, hypertension, pain and anemia.
Renal impairment: by low protein diet and dialysis.

Surgical:

Rovsing's operation (deroofing) for large cysts causing symptoms or obstruction.
Stone removal.
Renal failure: Renal transplantation.

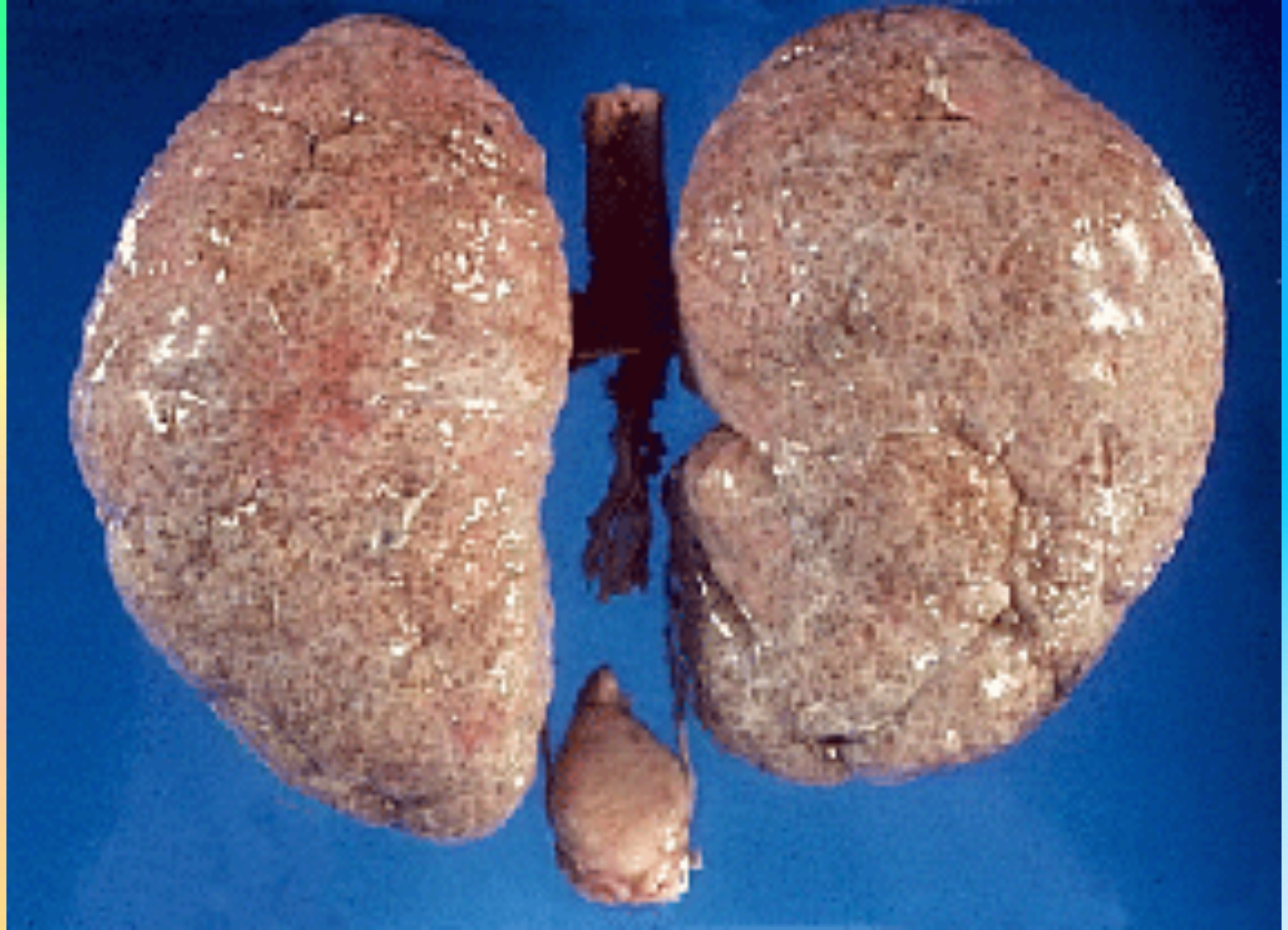
Autosomal recessive polycystic kidney disease

Rare autosomal recessive, incompatible with life. 50% die at birth.

Both kidneys are large in size and replaced by large number of cysts which may obstruct labor. Associated with hepatic fibrosis

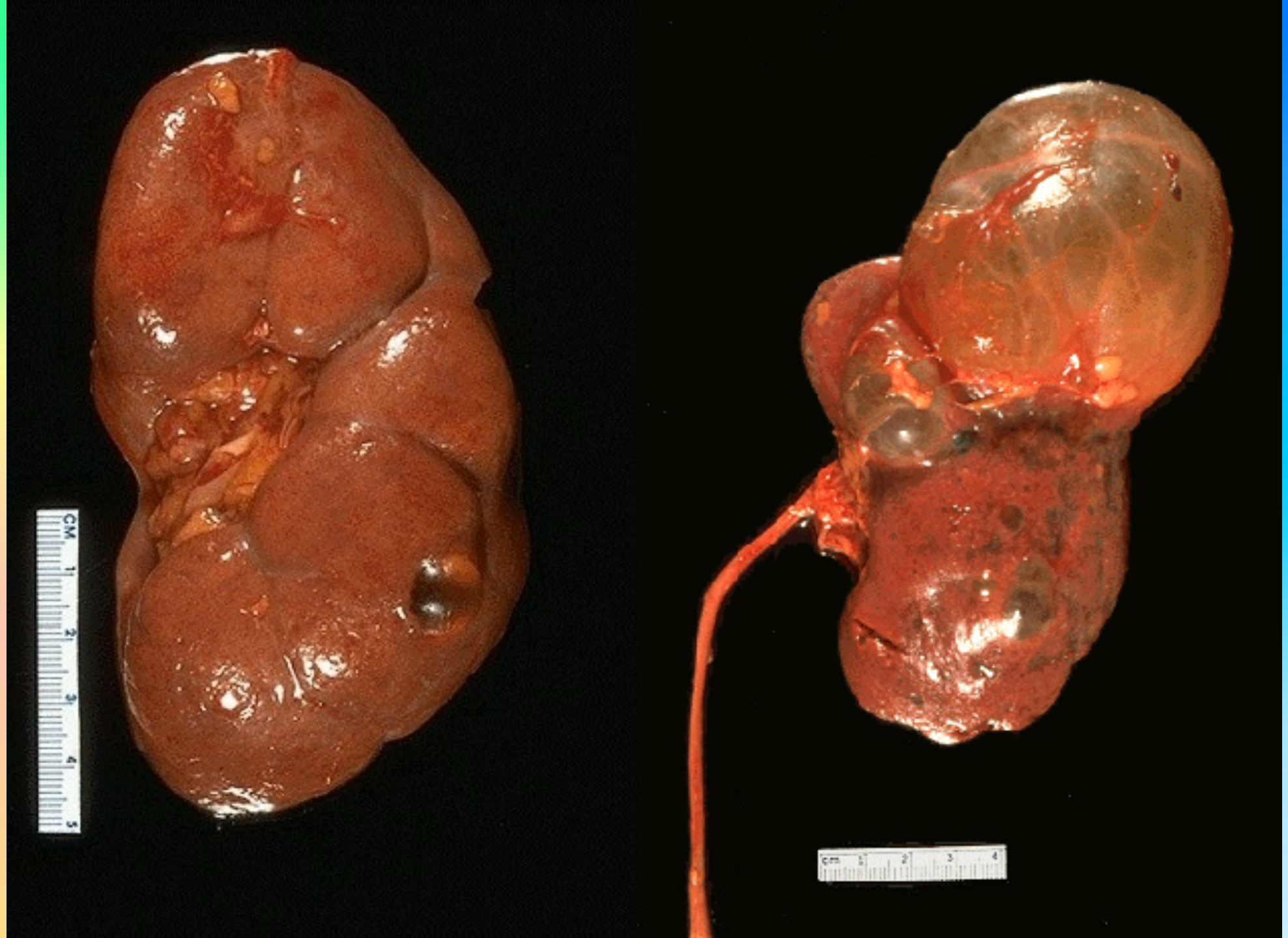
Clinical features: oligohydramnios, respiratory distress, uremia, hypertension,

Treatment: according to presentation. treat hypertension, treat hepatic failure, transplant.



Simple (solitary) renal cyst

Common condition.
Single or multiple.
uni or bilateral.
Congenital or acquired.
Usually asymptomatic. In 10% symptomatic: pain, heaviness, infection, bleeding inside the cyst or pressure effect on the ureter causing hydronephrosis.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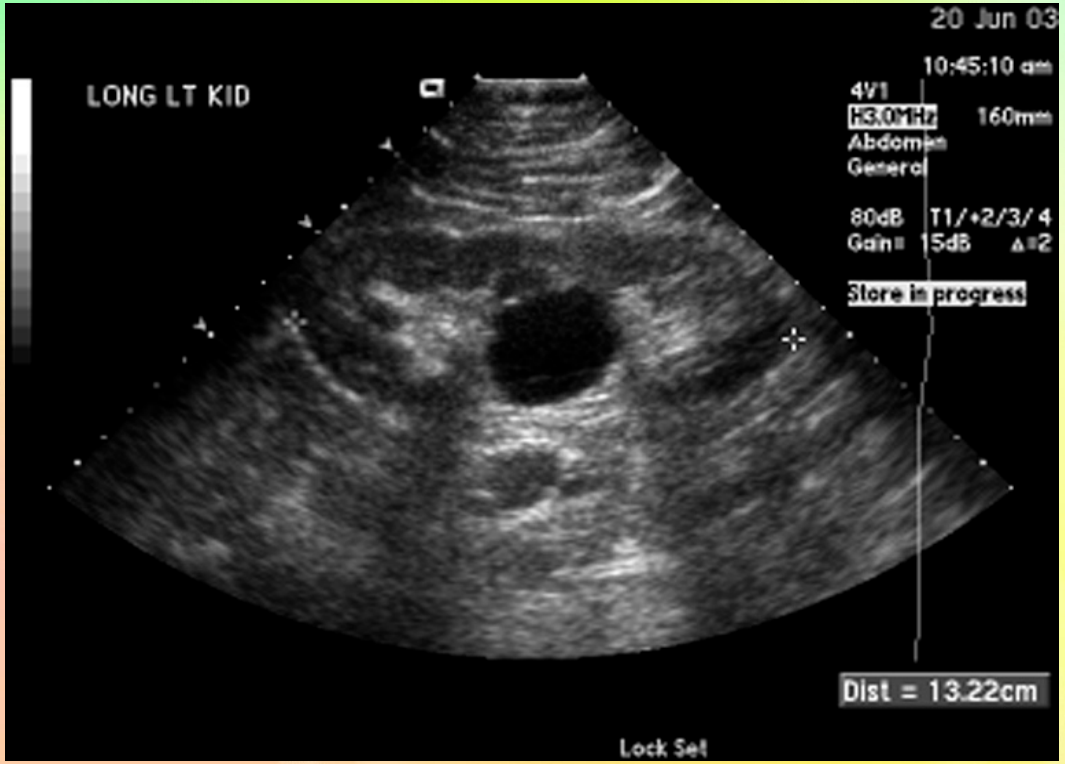
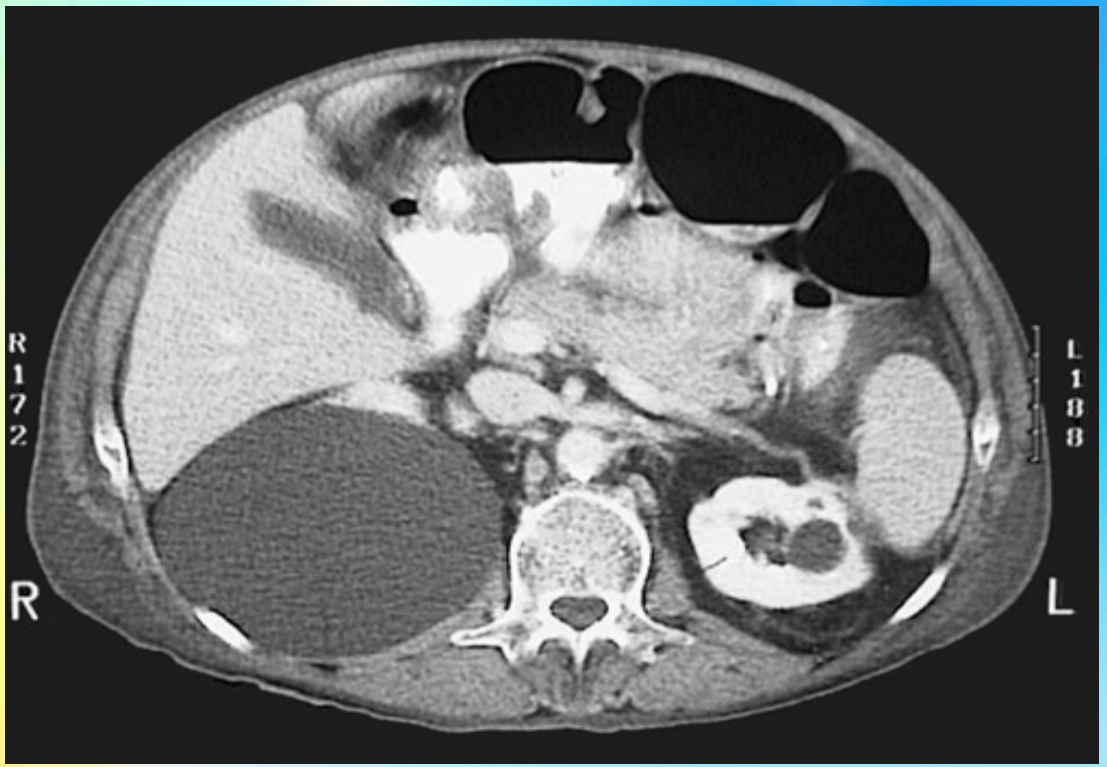
Diagnosis

U/S, KUB, IVU, CT scan &MRI





Treatment: usually no treatment needed

Symptomatic patients:

- Aspiration and injection of sclerosing agent.
- Rovsing’s operation (deroofing).
- Partial or total nephrectomy in destructed kidney.

Congenital Anomalies of Renal pelvis & Ureter

Duplication of Renal Pelvis: More common on left side. Renorenal reflux may occur from one pelvis to the other.

Duplication of the ureter: Usually the ureters fuse & have common orifice in the bladder although they may open independently in the bladder.

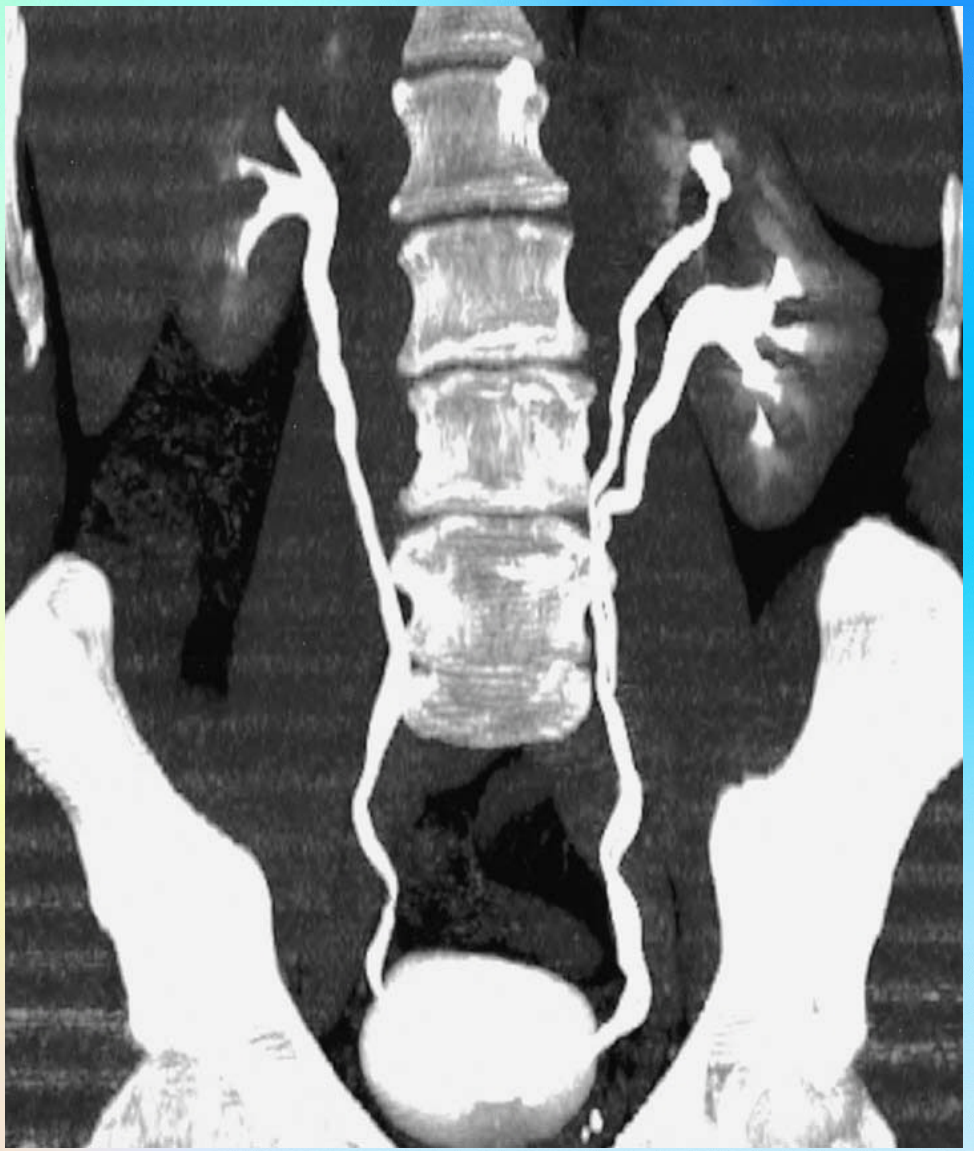
Clinical features : usually asymptomatic

More prone to infections, calculus disease & hydronephrosis

Treatment: expectant

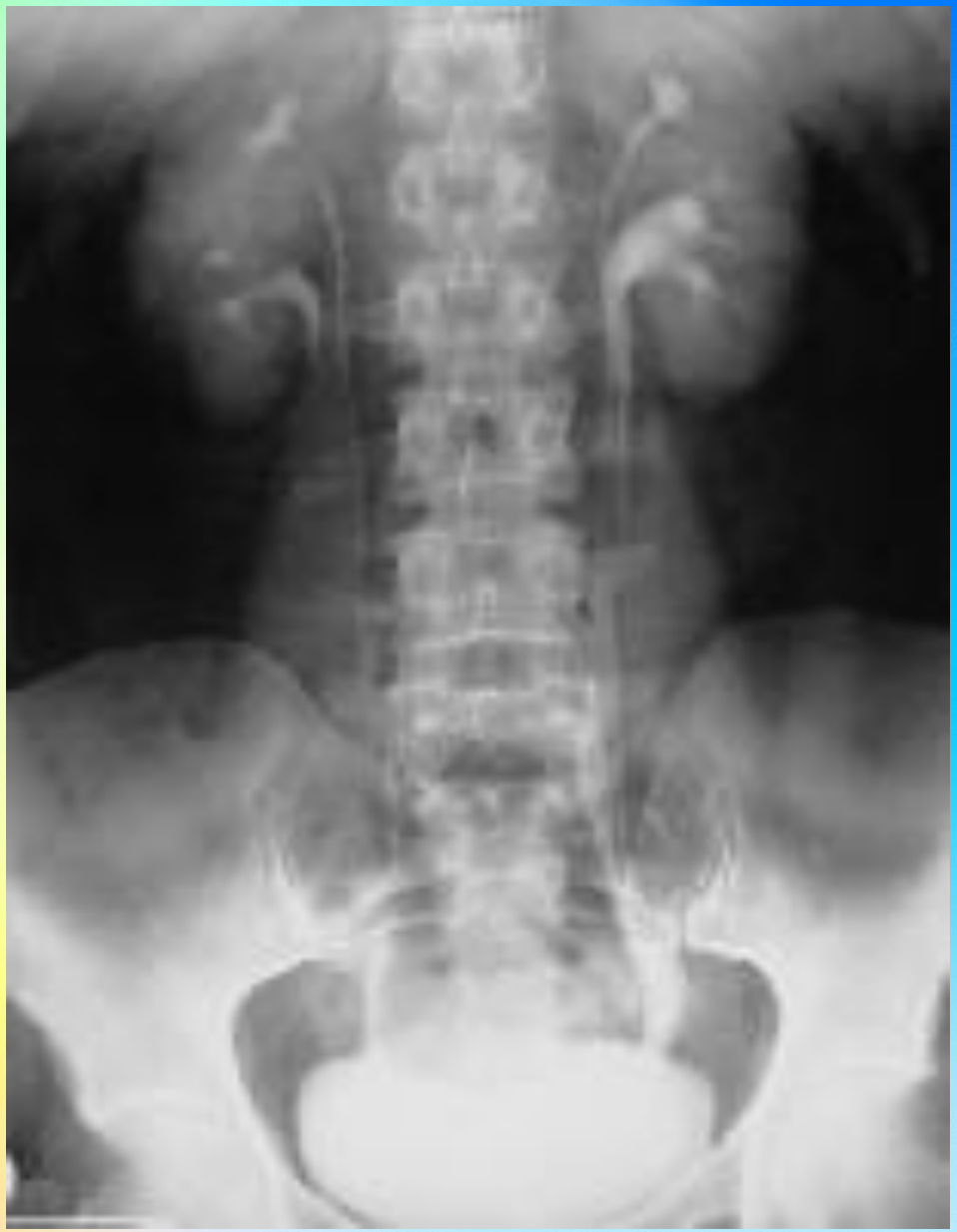
Ureteral duplication: partial and complete

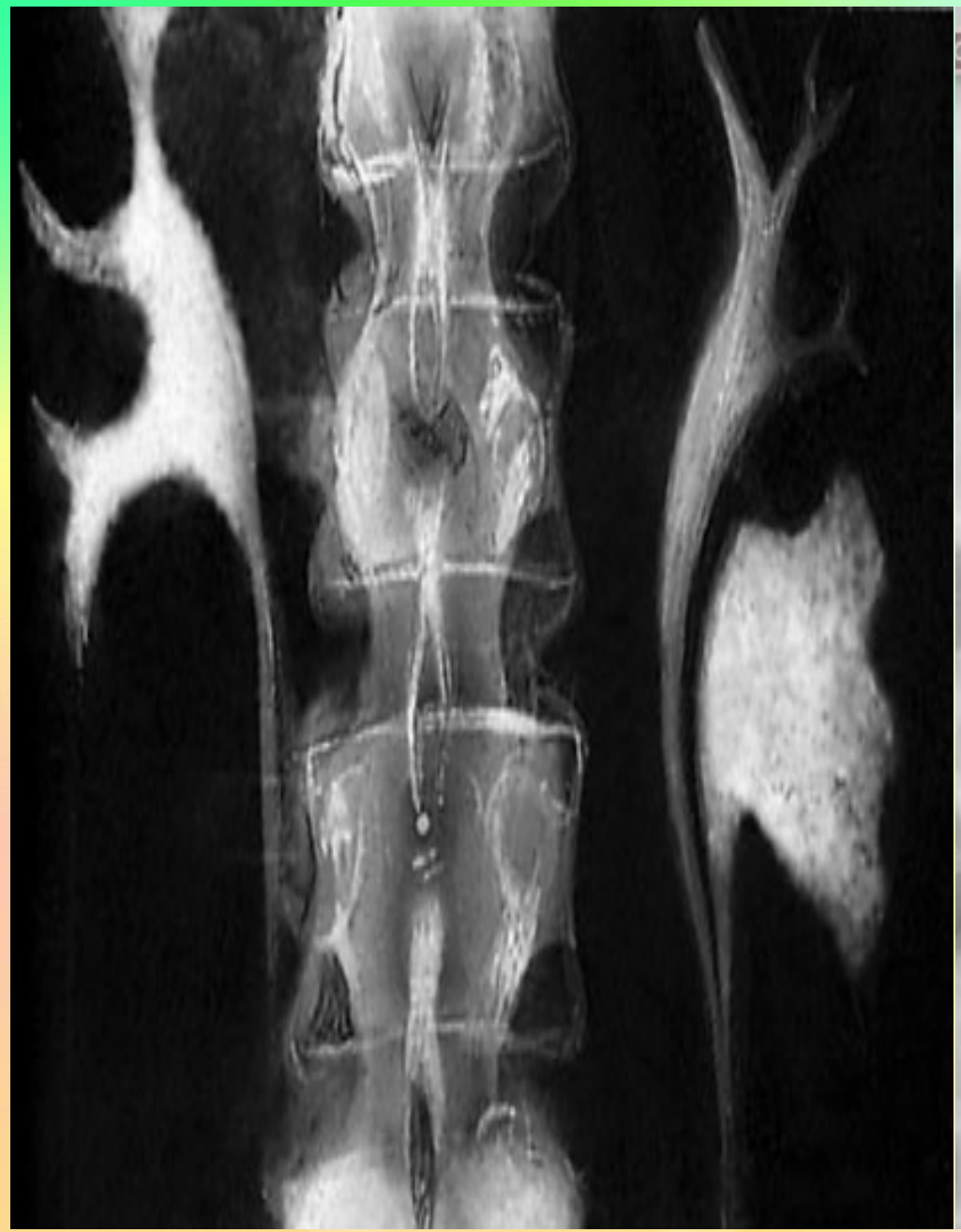
- Partial duplication: is more common. Two ureters draining single kidney for variable length, then unite together before entering the bladder in one ureteric orifice. Rarely the lower part is duplicated as inverted Y ureter.



Complete duplication:

Less frequent, the whole ureter is duplicated, and each one opens in separate orifice in the bladder. The ureter draining the upper part opens more distally in the bladder.



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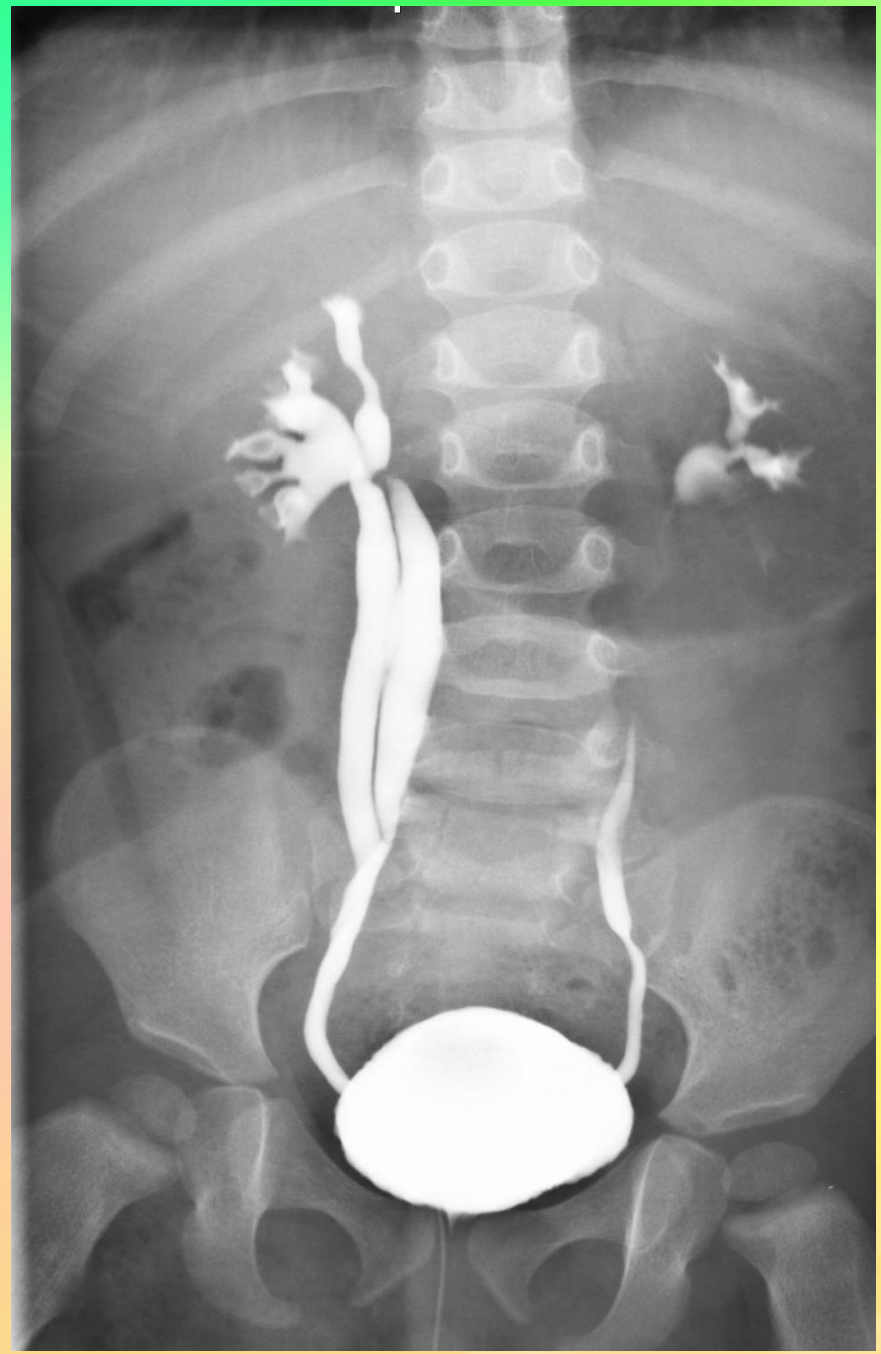
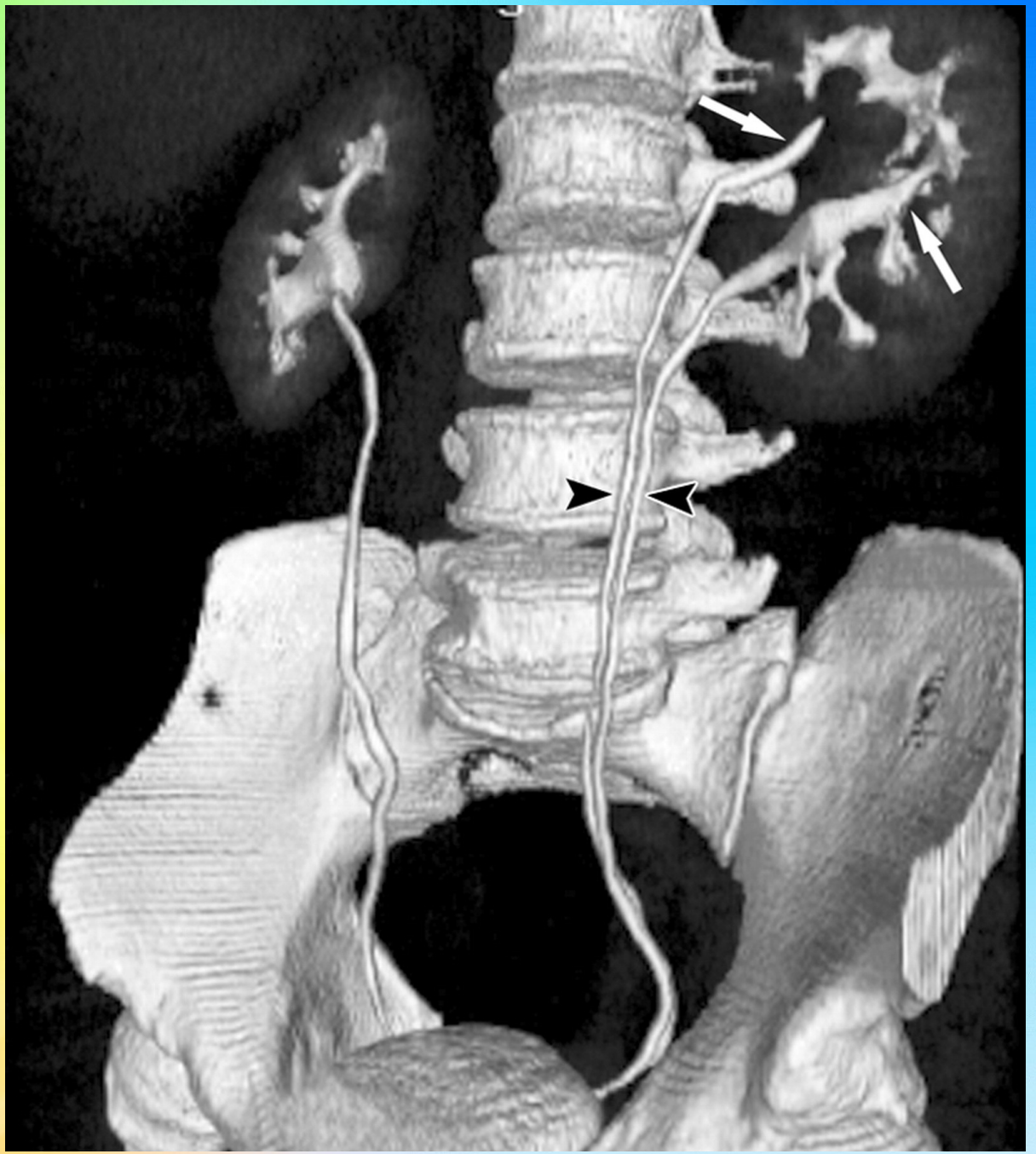
Bifid renal pelvis





Ectopic Ureters

Ectopic ureter is the ureter that does not enter the trigonal area of the bladder.

In the male, the posterior urethra is the most common site of termination, also to semenal vesicle

In the female, the urethra and vestibule are the most common sites

Clinical features: According to the site of orifice

In females: continuous dribbling

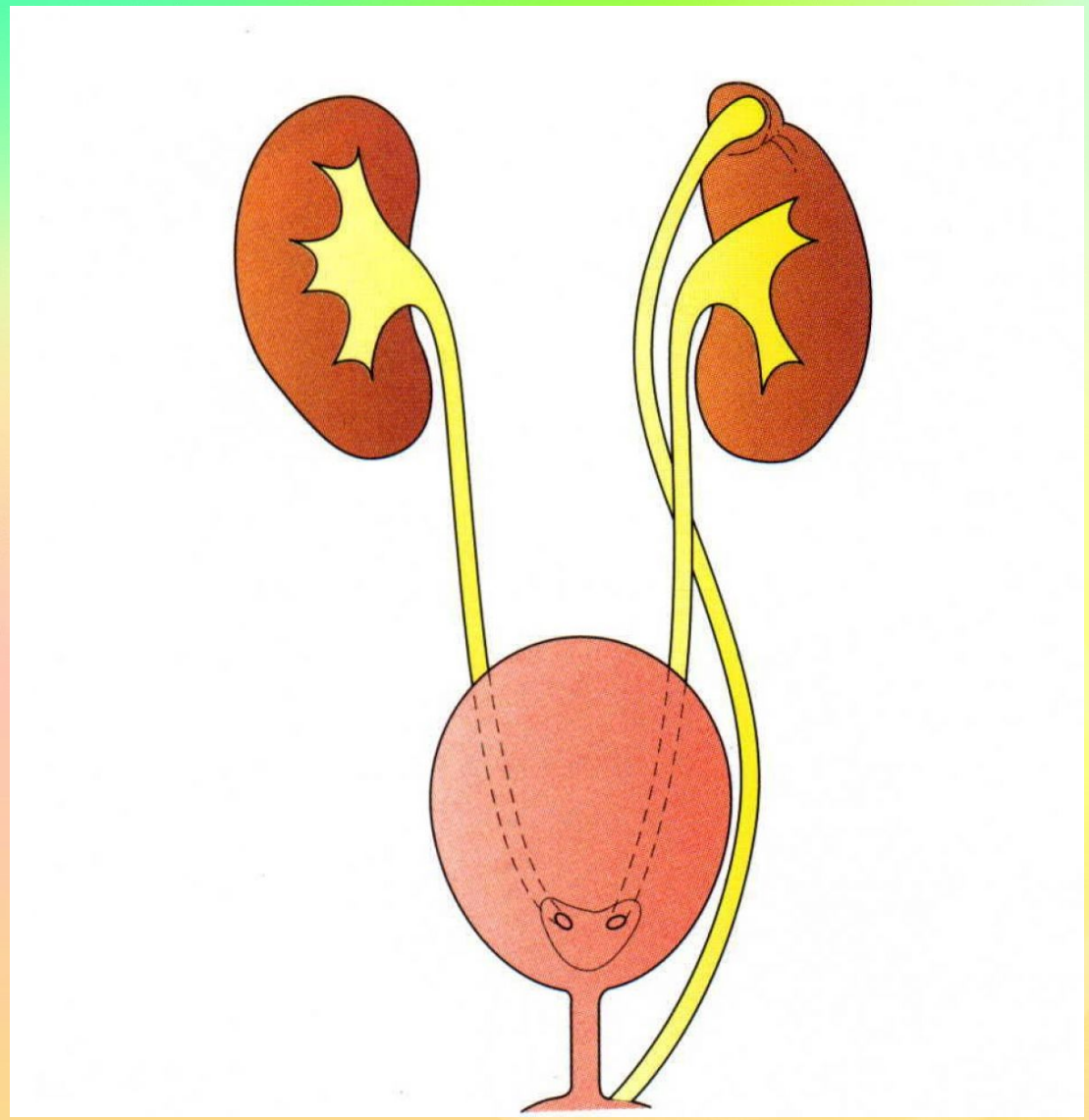
In males: urinary tract infection

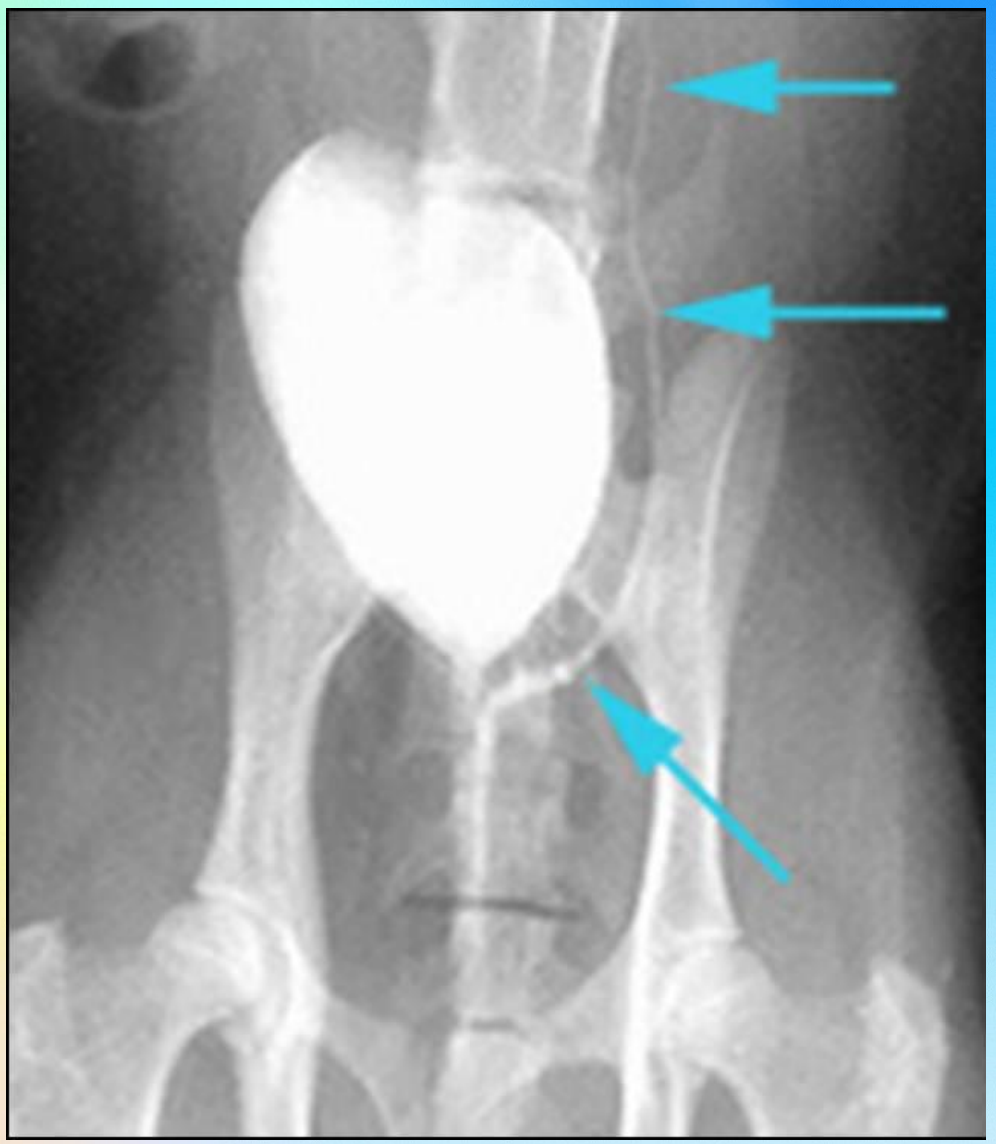
Diagnosis: IVU, U/S, CT scan, cystoscopy

Treatment: Ureteric reimplantation

Ectopic ureters may drain renal moieties (either an upper pole or a single-system kidney) that have minimal function. Therefore, upper pole partial nephrectomy (or nephrectomy of single system) is sometimes recommended

Complete ureteral duplication and ectopic ureteric orifice.





Ureteroceles

Is due to congenital atresia of the ureteric orifice which causes a cystic dilatation of the intramural portion of the ureter

Women > men

Sometimes involves with ectopic ureter

More prone to stone disease & UTIs

Clinical Features : asymptomatic, Repeated UTIs, Hematuria

Diagnosis

IVU, cystoscopy, cystogram

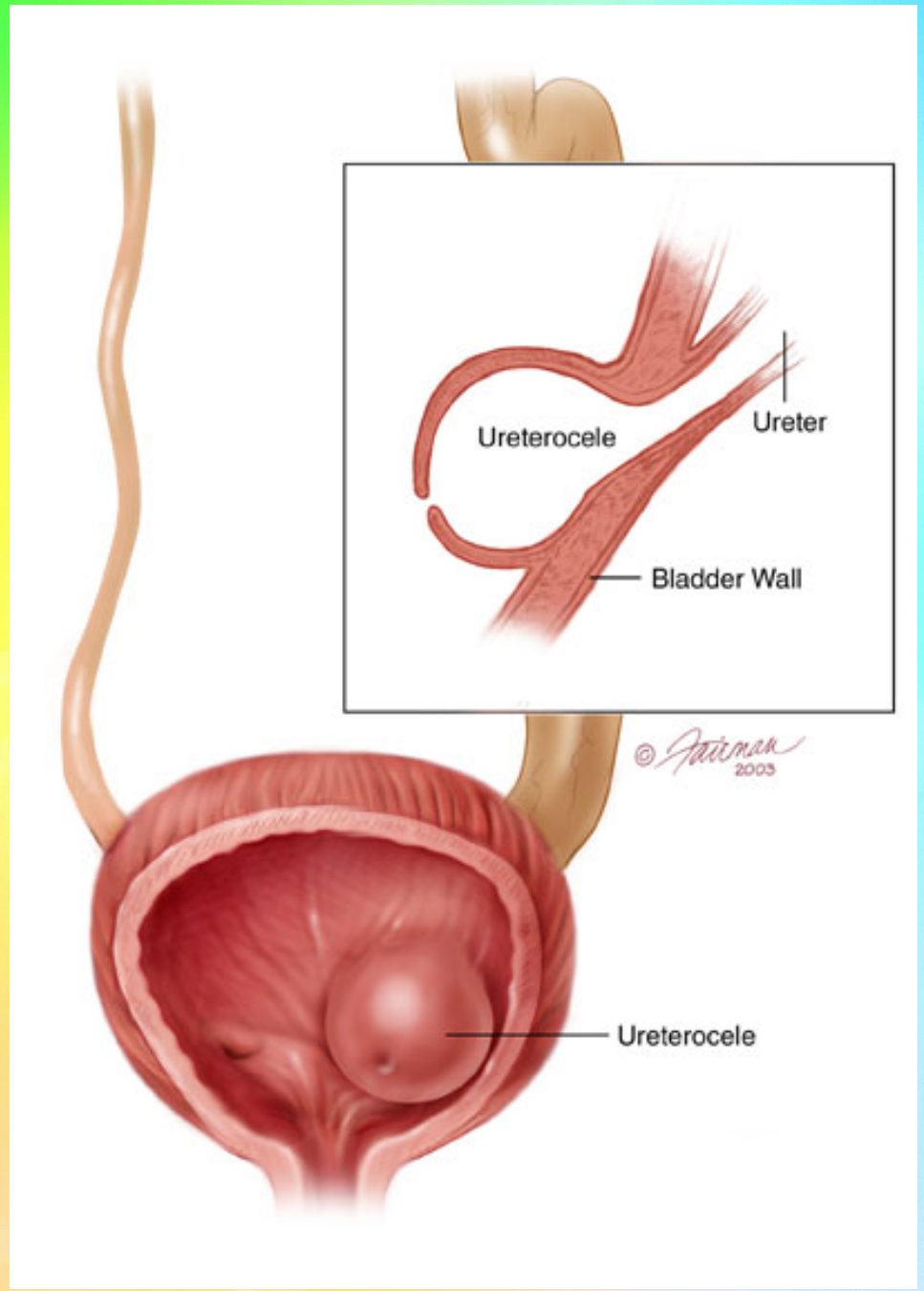
The 'cobra head sign' on excretory urography is typical.

Treatment

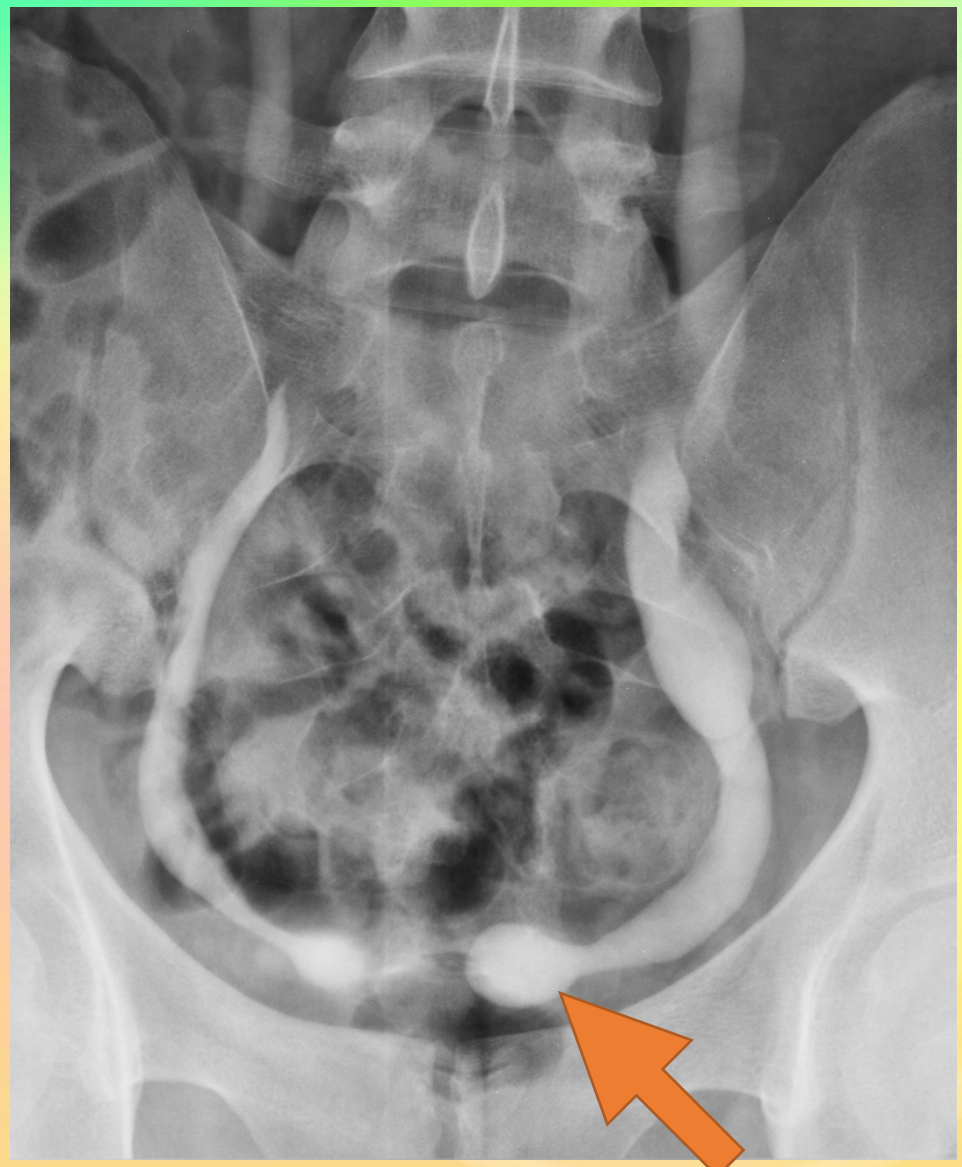
Asymptomatic : no treatment

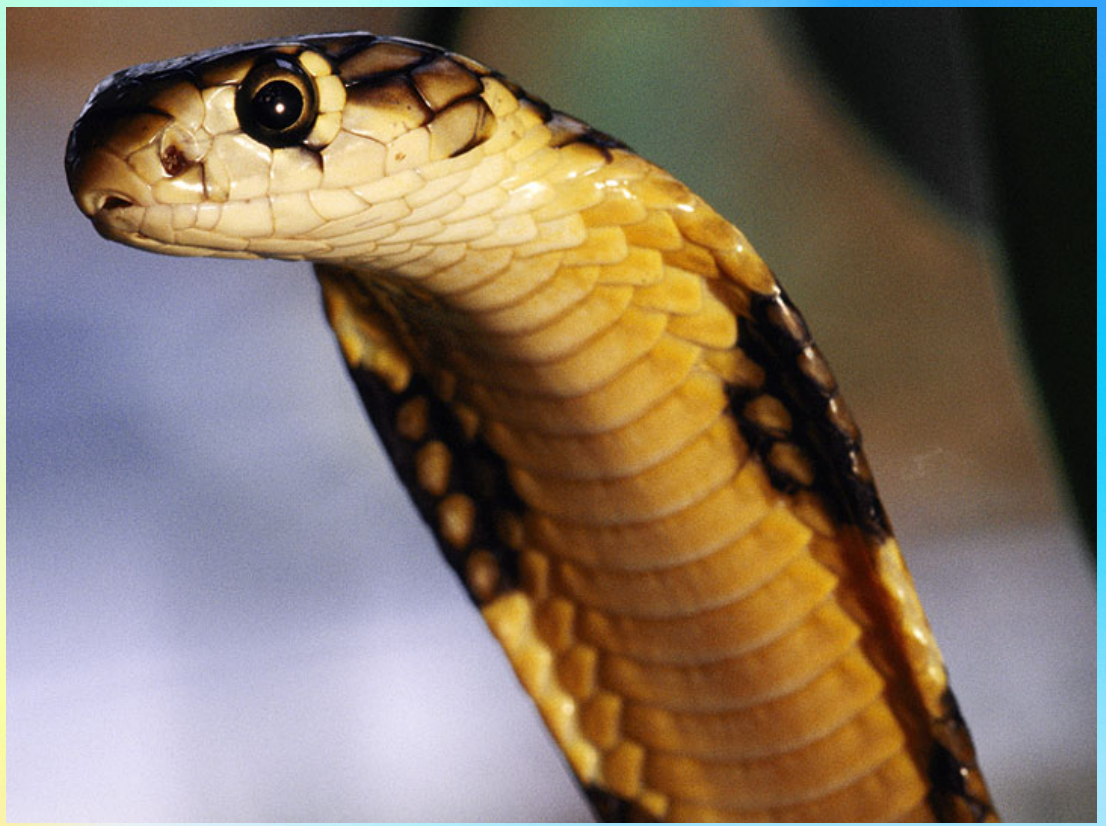
Cystoscopy with diathermy incision of the ureterocele or Nephrectomy in non functioning kidney

In complicated cases, ureteral reimplantation.


Ureterocele
Ureter
Bladder Wall
© Tortora 2003
Ureterocele

Cobra (Adder) head appearance of ureterocele





Ureteropelvic Junction (UPJ) (PUJ) Obstruction (stenosis)

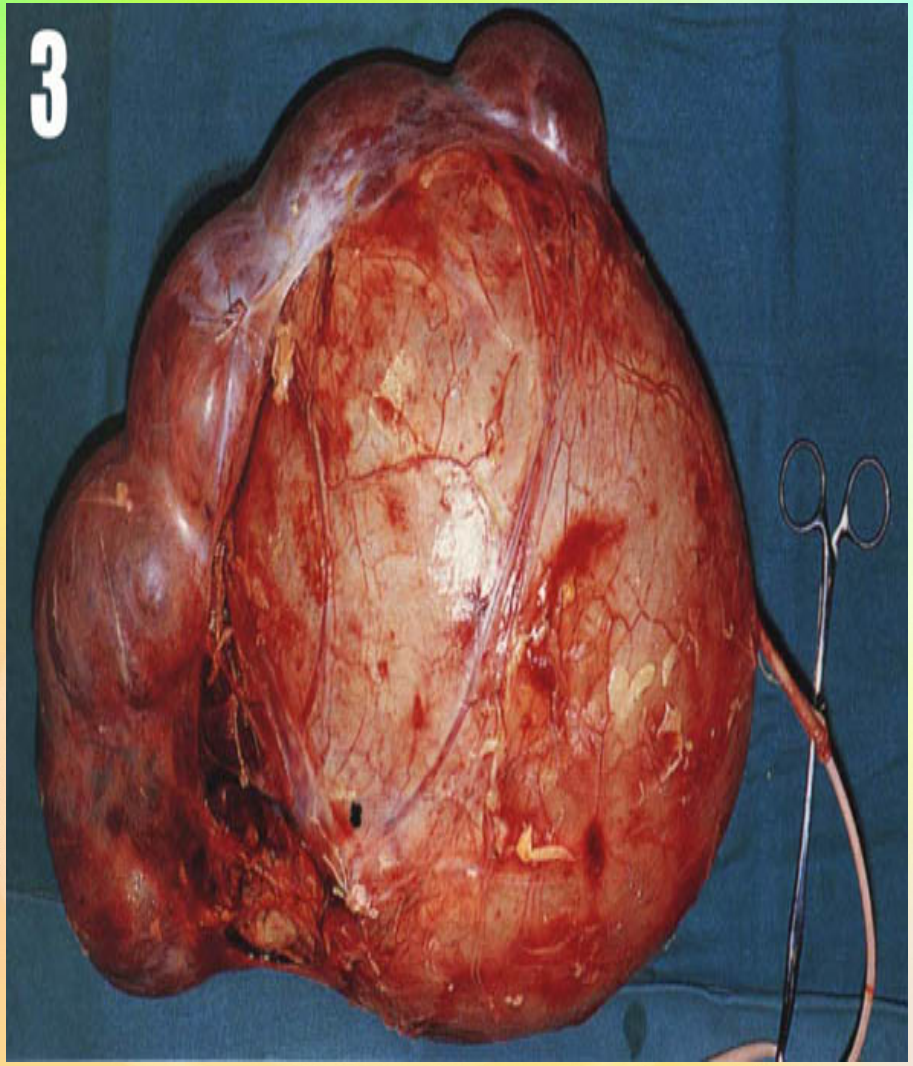
The most common cause of significant dilation of the collecting system in the fetal kidney
Boys > Girls
Left-sided lesions predominate
Could be bilateral

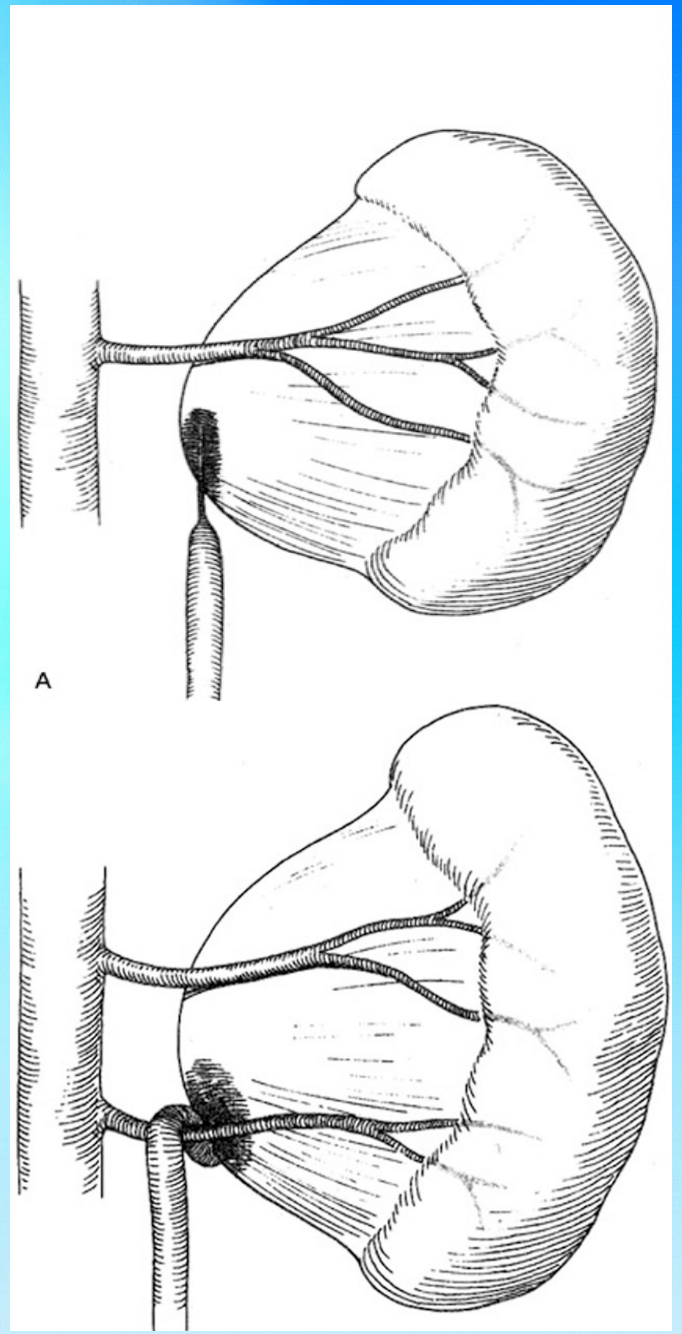
ETIOLOGY

Intrinsic (intramural): interruption in the development of the circular musculature of the UPJ or mucosal fold that causes valve like effect.

Extrinsic: An aberrant, accessory, or early-branching lower-pole renal artery

PUJ Obstruction – gross pathology





SYMPTOMS / PRESENTATION

Most infants are asymptomatic and most children are discovered because of their symptoms
Episodic flank or upper abdominal pain with recurrent infections, sometimes associated with nausea and vomiting, failure to thrive, diarrhea, and loin mass.

DIAGNOSIS

U/S, IVU, CT scan, Magnetic Resonance Imaging, Radionuclide Renography: to see the split function of each kidney, Pressure-Flow Studies and Whitaker test

Treatment:

Medical:

control infection and pain.
Suppressive antibiotics

Surgical:

Indications for surgery:
1-progressive hydronephrosis.
2- UTI despite antibiotic cover, and symptomatic patients.
3- Severe hydronephrotic non functioning kidney.
4- deterioration of renal function

SURGICAL REPAIR: including open surgical techniques, laparoscopic, & endoscopic approaches

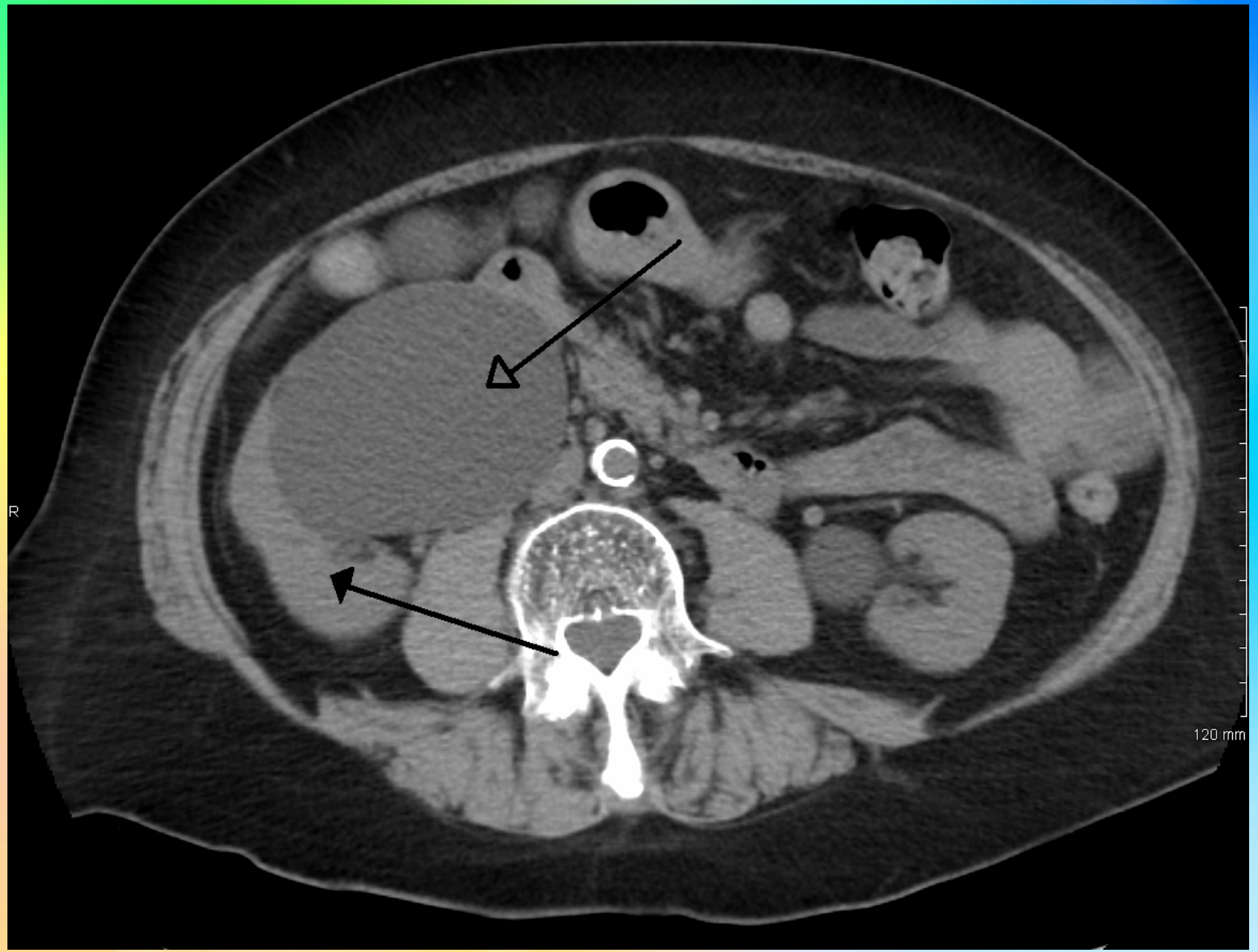
Open & laparoscopic surgical techniques

Anderson-Hynes dismembered pyeloplasty: excision of the pathologic UPJ & appropriate reanastamosis.

Flap technique or flap operation

Endoscopic Approaches:

- Balloon dilatation
- Antegrade endopyelotomy
- Nephrectomy for non functioning kidney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