

Congenital anomalies of kidney and ureter

Department of Urology

- Anomalies of the Upper Urinary Tract
- Congenital cyst
- Ectopic ureter
- Ureterocoele

Anomalies of the Upper Urinary Tract

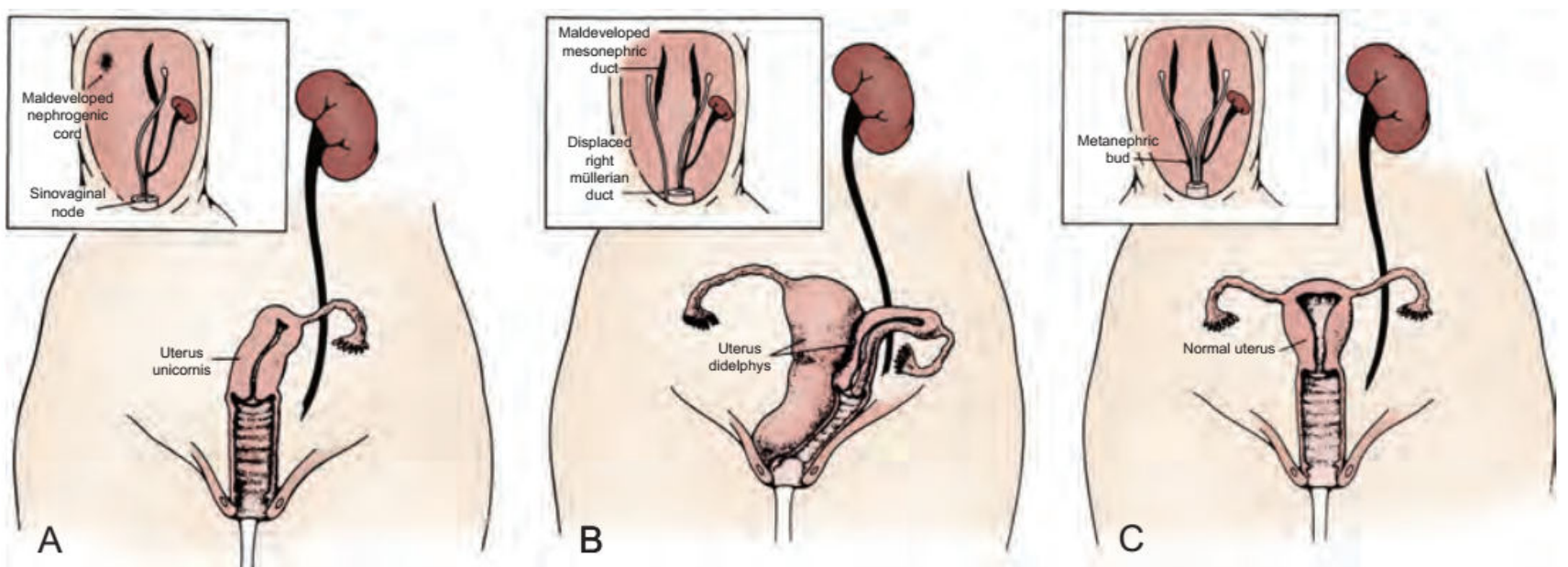
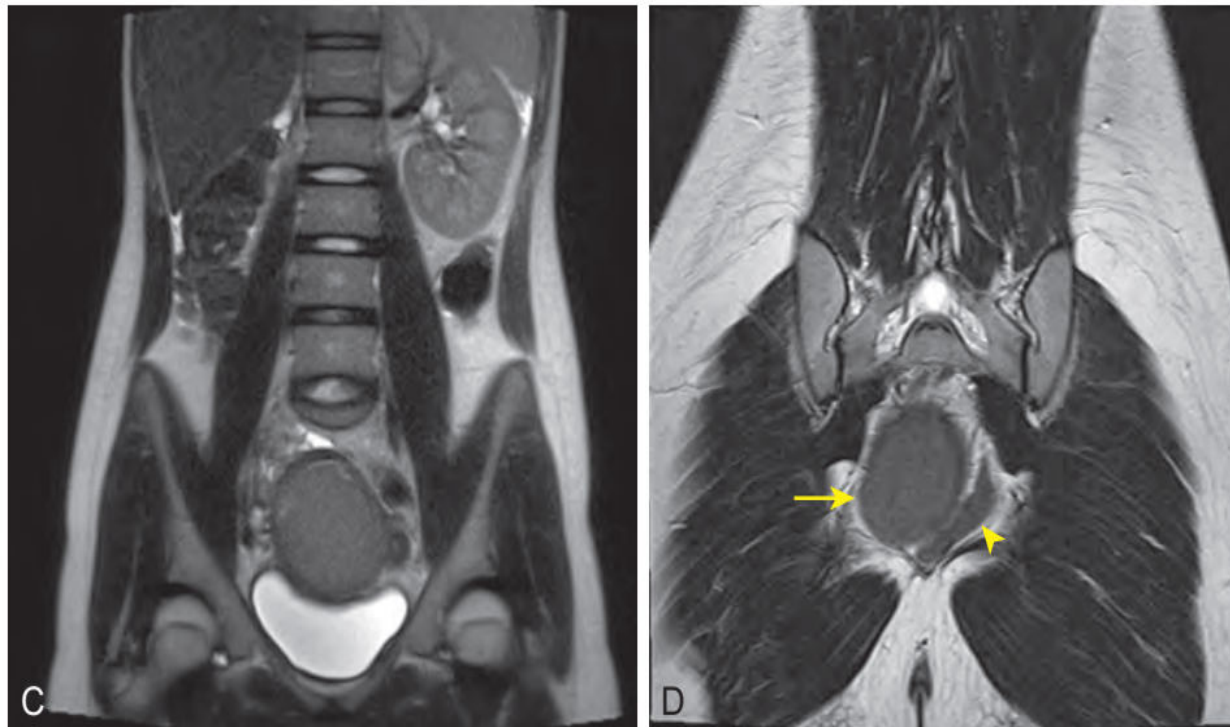
- ANOMALY OF **NUMBER**
 - Bilateral Renal Agenesis
 - *Unilateral Renal Agenesis*
 - *Supernumerary Kidney*
 - ANOMALIES OF **ASCENT**
 - Simple Renal Ectopia
 - Cephalad Renal Ectopia
 - Thoracic Kidney
 - ANOMALIES OF **FORM AND FUSION**
 - Crossed Renal Ectopia with and without Fusion
 - Horseshoe Kidney
 - ANOMALIES OF **ROTATION**
 - ANOMALIES OF **RENAL VASCULATURE**
 - Aberrant, Accessory, or Multiple Vessels
 - Renal Artery Aneurysm
 - Renal Arteriovenous Fistula
 - ANOMALIES OF THE **COLLECTING SYSTEM**
 - Calyceal Diverticulum
 - Hydrocalycosis
 - Megacalycosis
 - Infundibulopelvic Stenosis
 - Bifid Pelvis
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- ANOMALY OF **NUMBER**
 - Bilateral Renal Agenesis
 - *Unilateral Renal Agenesis*
 - *Supernumerary Kidney*



- prominent fold and skin crease beneath each eye, blunted nose, depression between lower lip and chin

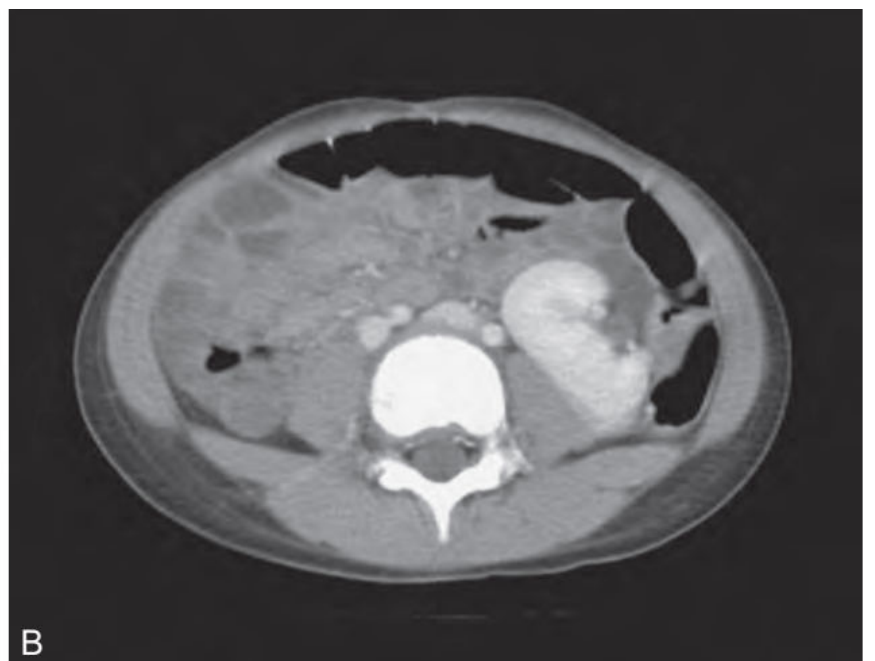
Unilateral Renal Agenesis

- 1 in 1100 births
- Kallmann syndrome, Turner syndrome, Poland syndrome, Fraser syndrome, branchio-oto-renal (BOR) syndrome, DiGeorge syndrome
- Anomaly of other organ



Supernumerary Kidney

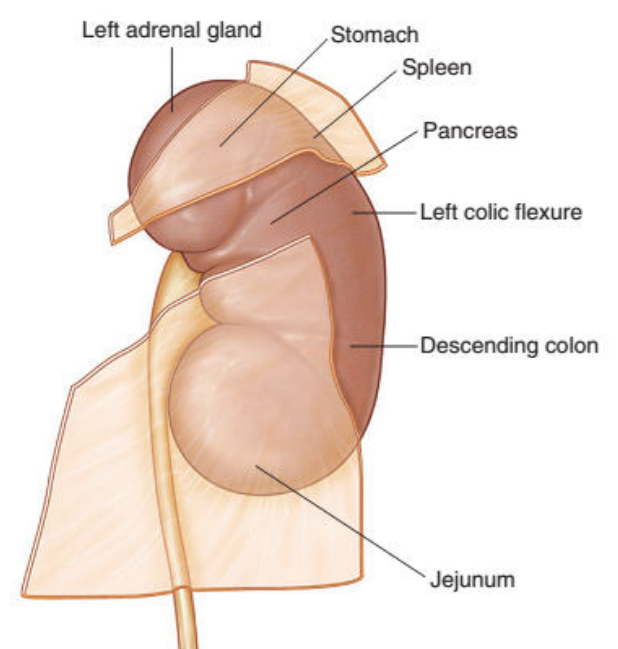
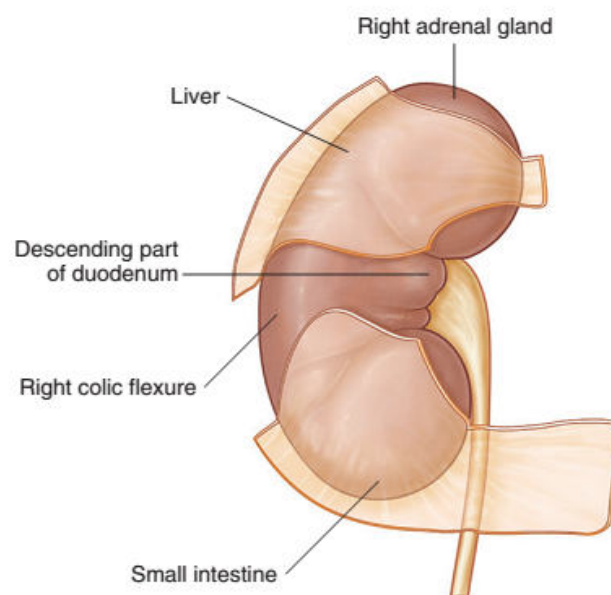
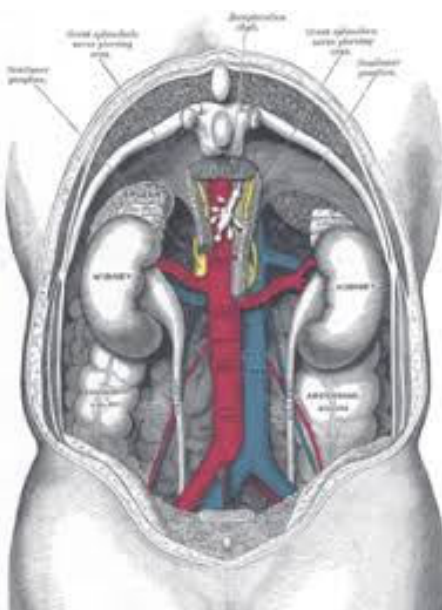
- distinct encapsulated parenchymal mass with its own collecting system, blood supply
- Very rare
- Asymptomatic/symptomatic
- T/t- if symptom

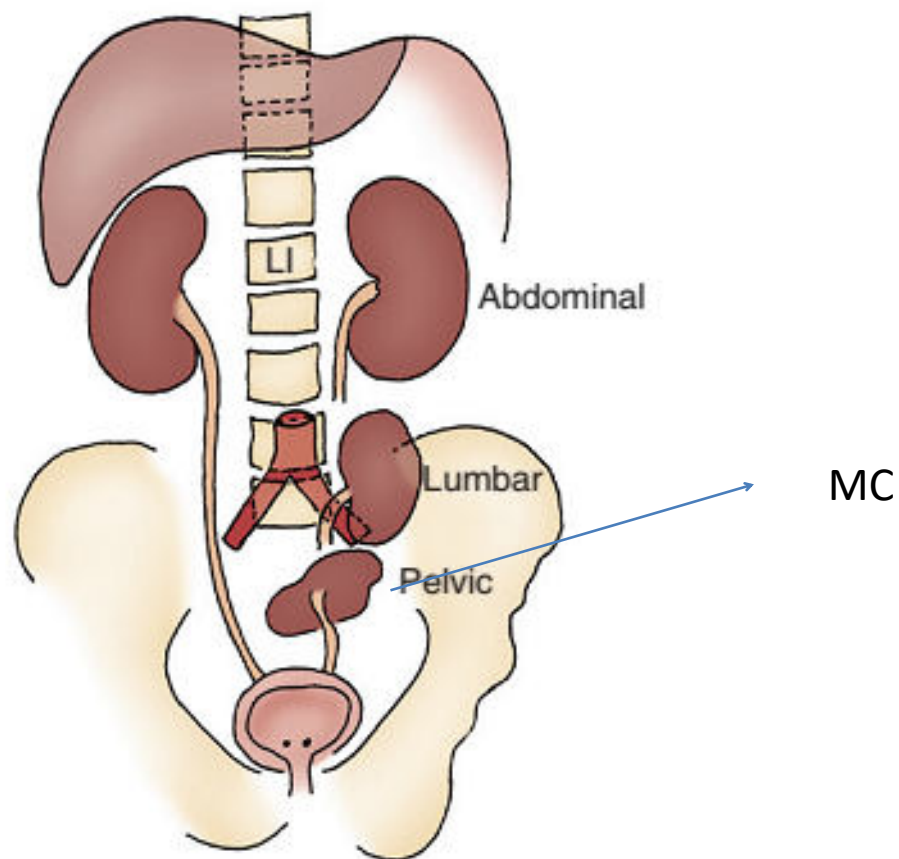


ANOMALIES OF ASCENT

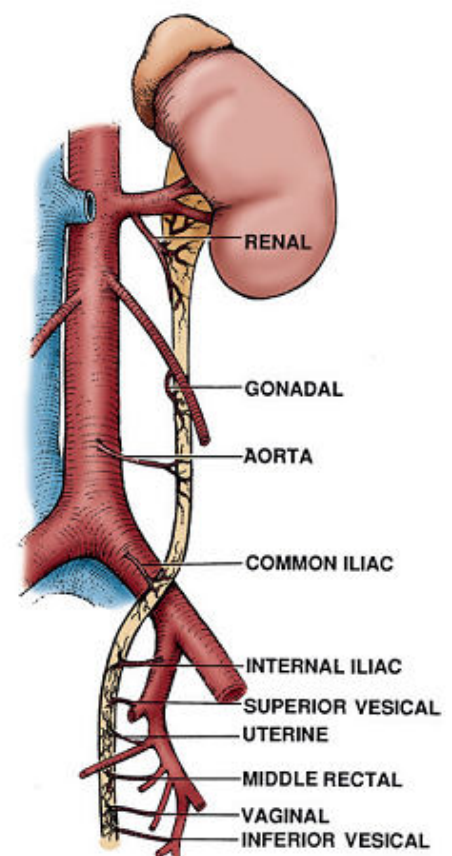
- Simple Renal Ectopia
- Cephalad Renal Ectopia
- Thoracic Kidney

ek (“out”) and *topos* (“place”)

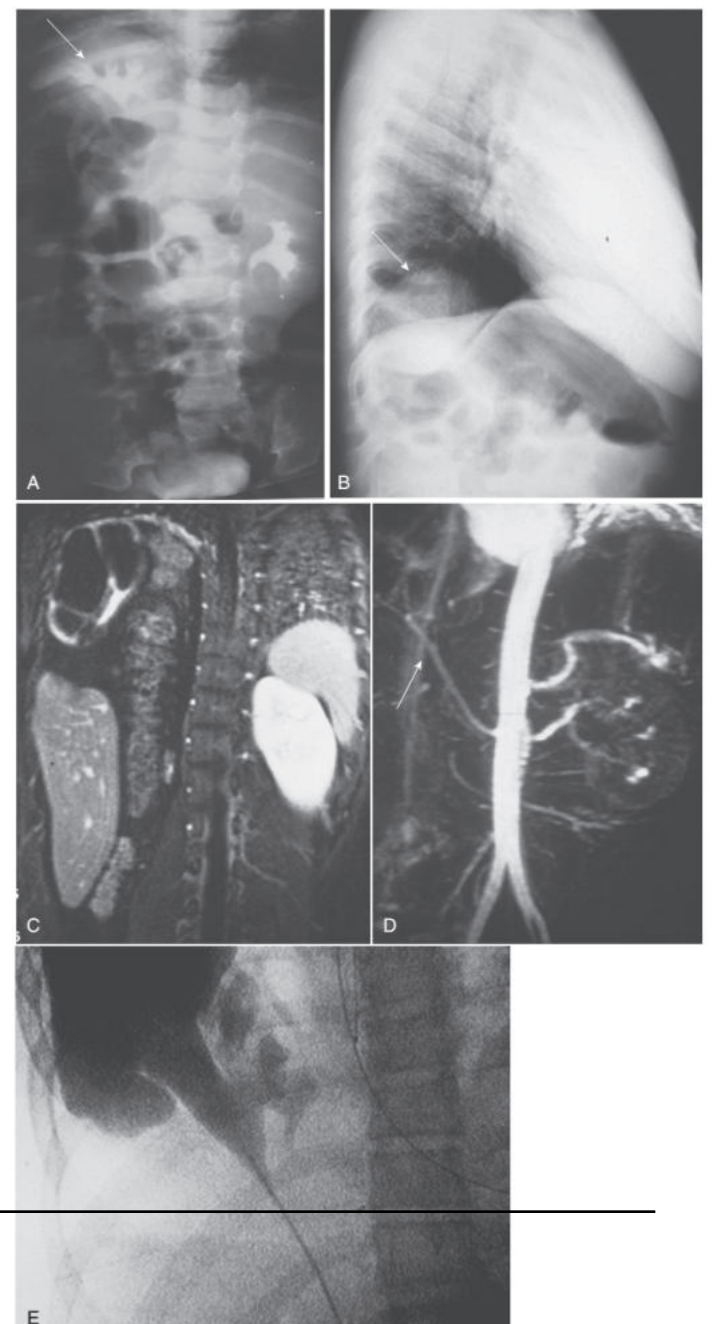
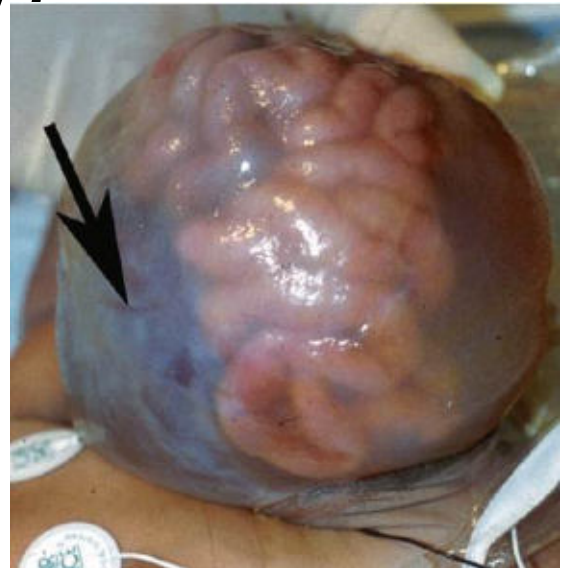




- Mostly asymptomatic
- **renal pelvis is usually anterior (instead of medial)**
- **Hydronephrotic**
 - PUJO
 - VUJO
 - Reflux
 - Malrotation
- **Ureter- not redundant**
- **Vascular supply-**
 - Not from abdominal aorta

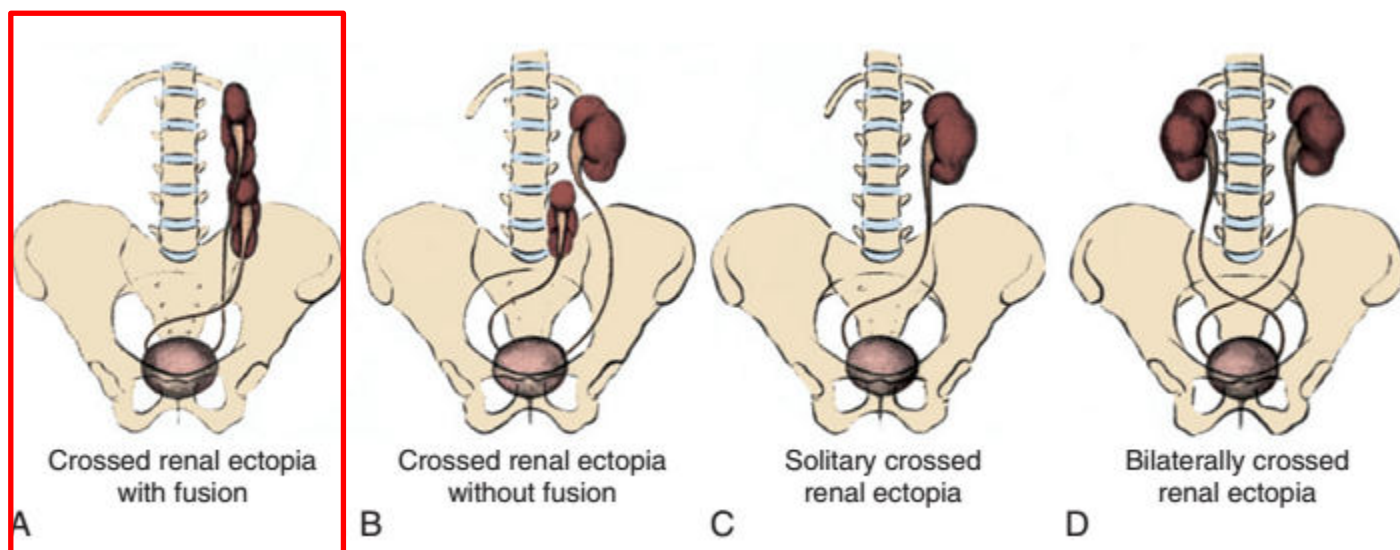


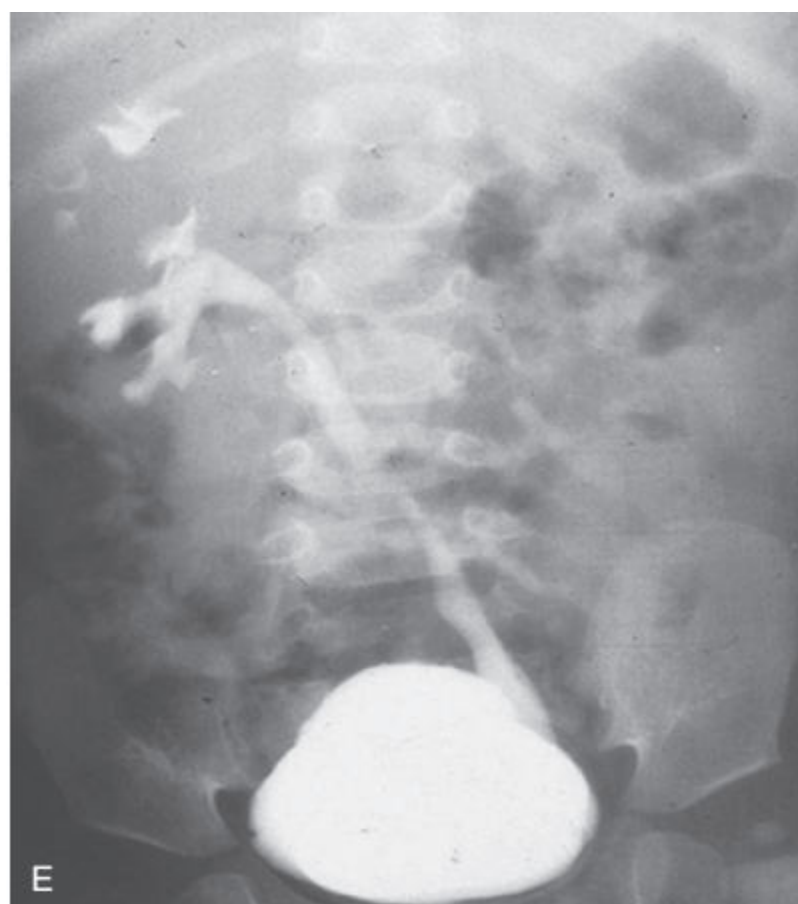
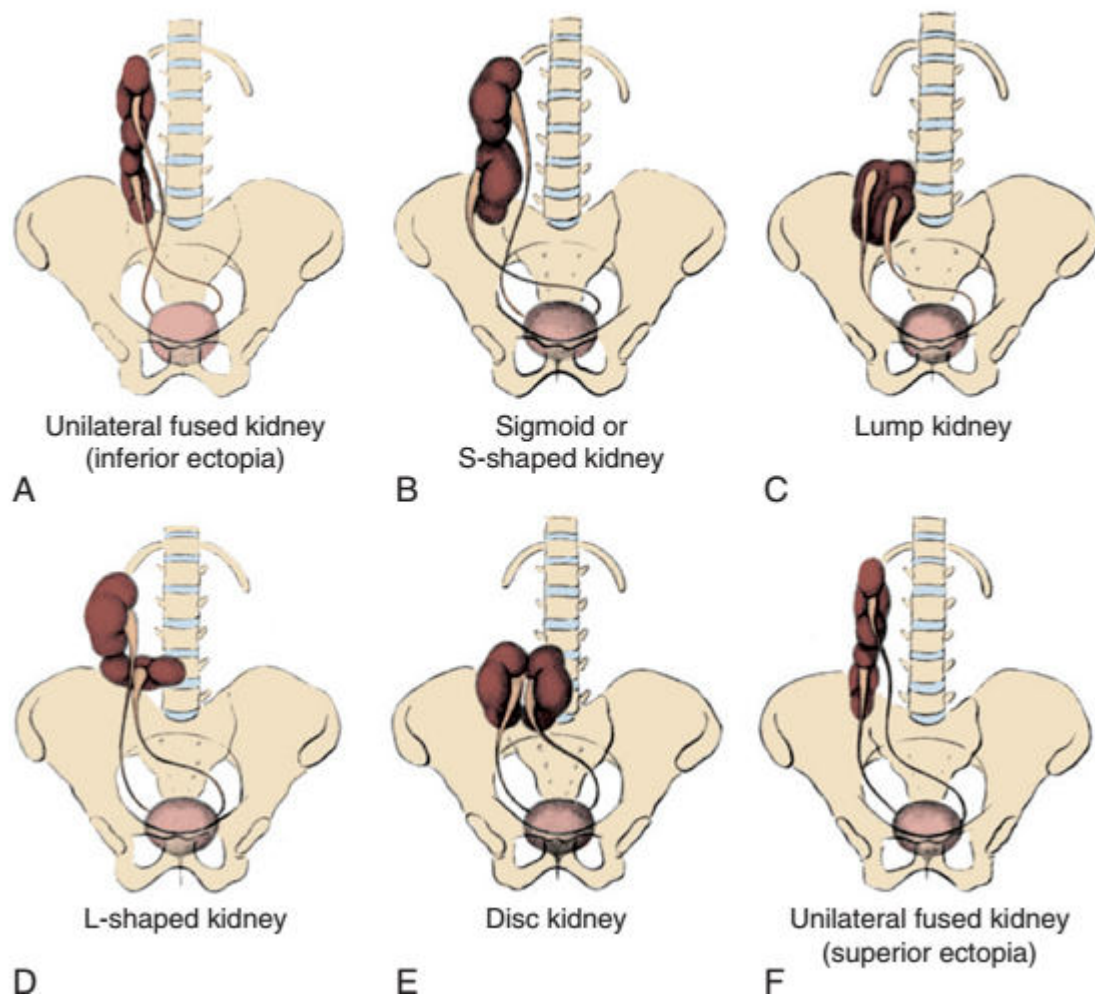
- Dx- Incidental
- USG/CT/MRI/Nuclear scan
- Clinical implication-
 - Calculus(Obstruction-malrotation, High insertion, crossing vessel)
 - RVH
- No difficult labour

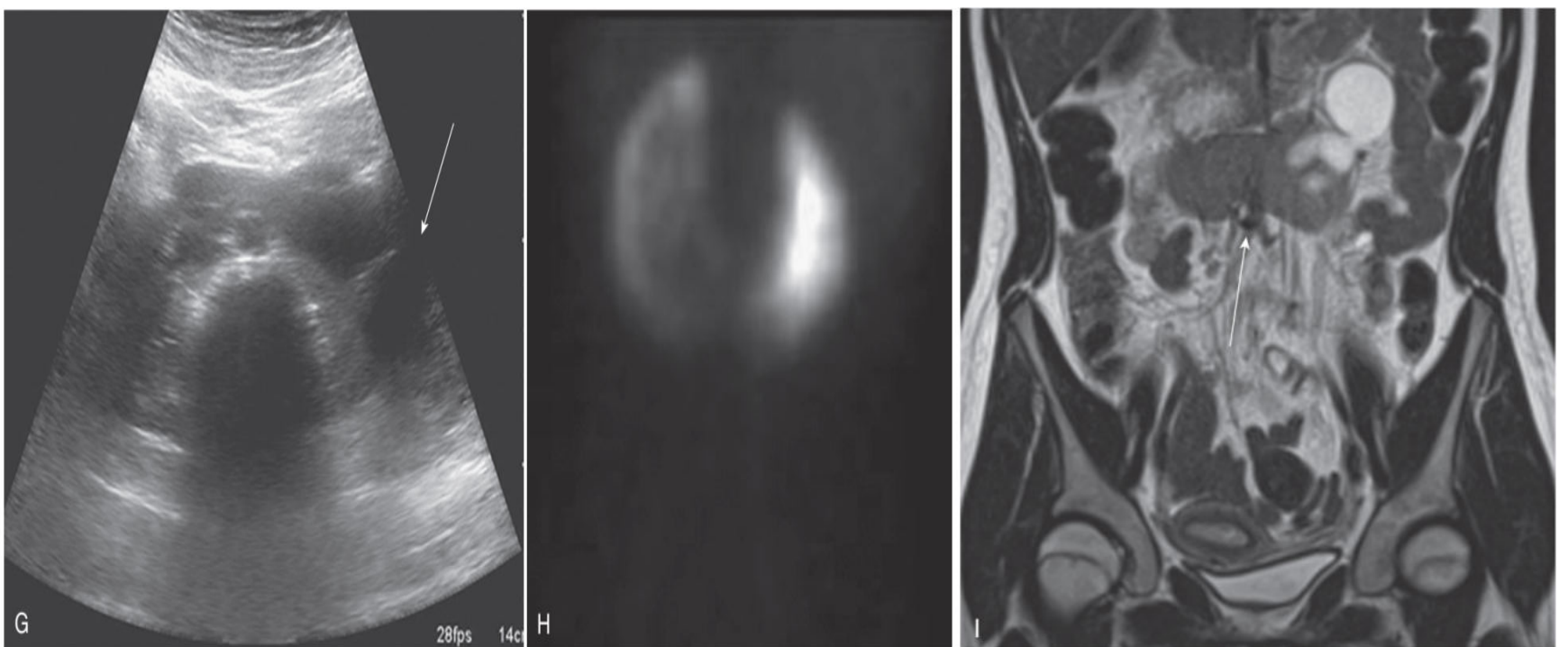
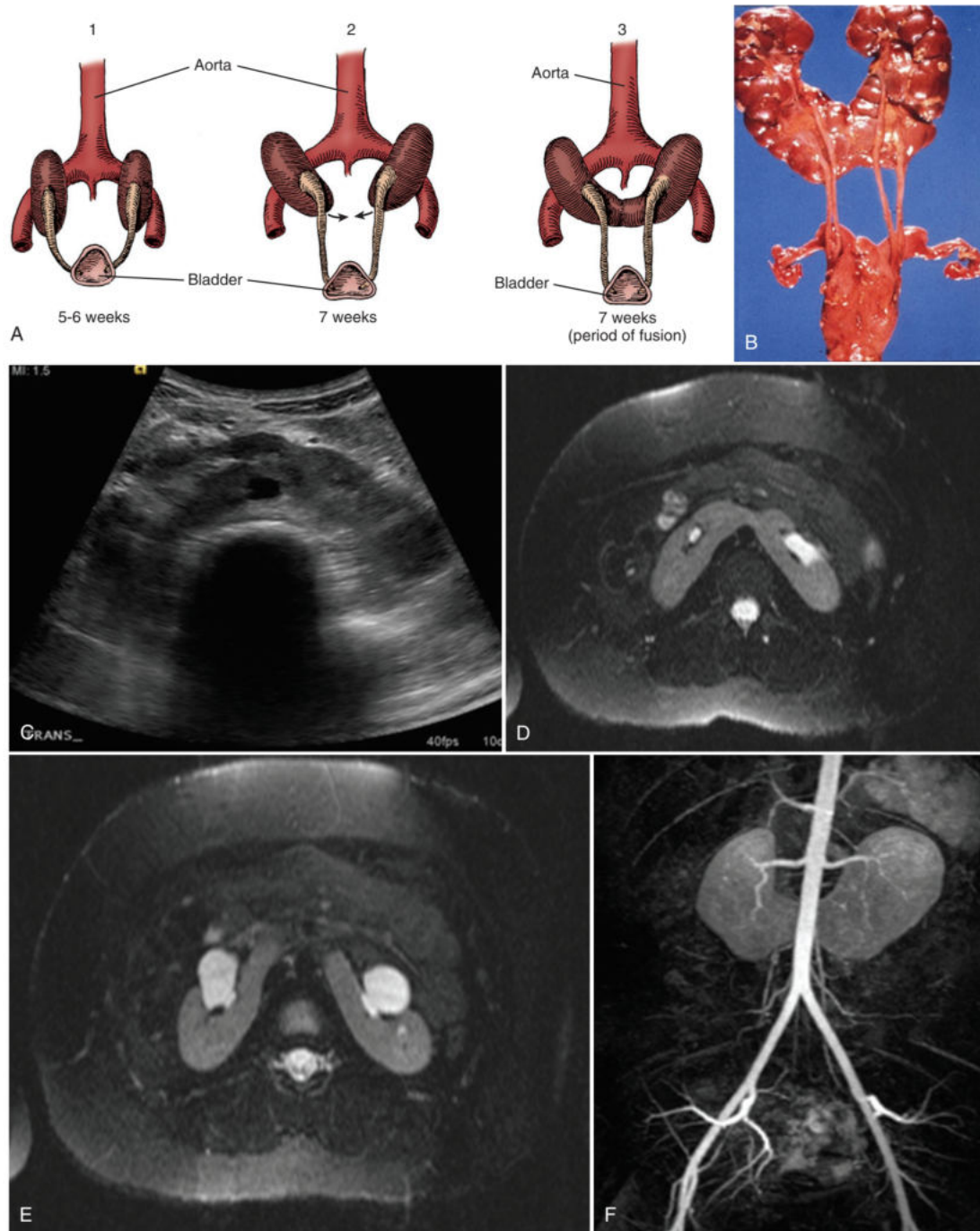


- **ANOMALIES OF FORM AND FUSION**
 - Crossed Renal Ectopia with and without Fusion
 - Horseshoe Kidney

Crossed Renal Ectopia with and without Fusion



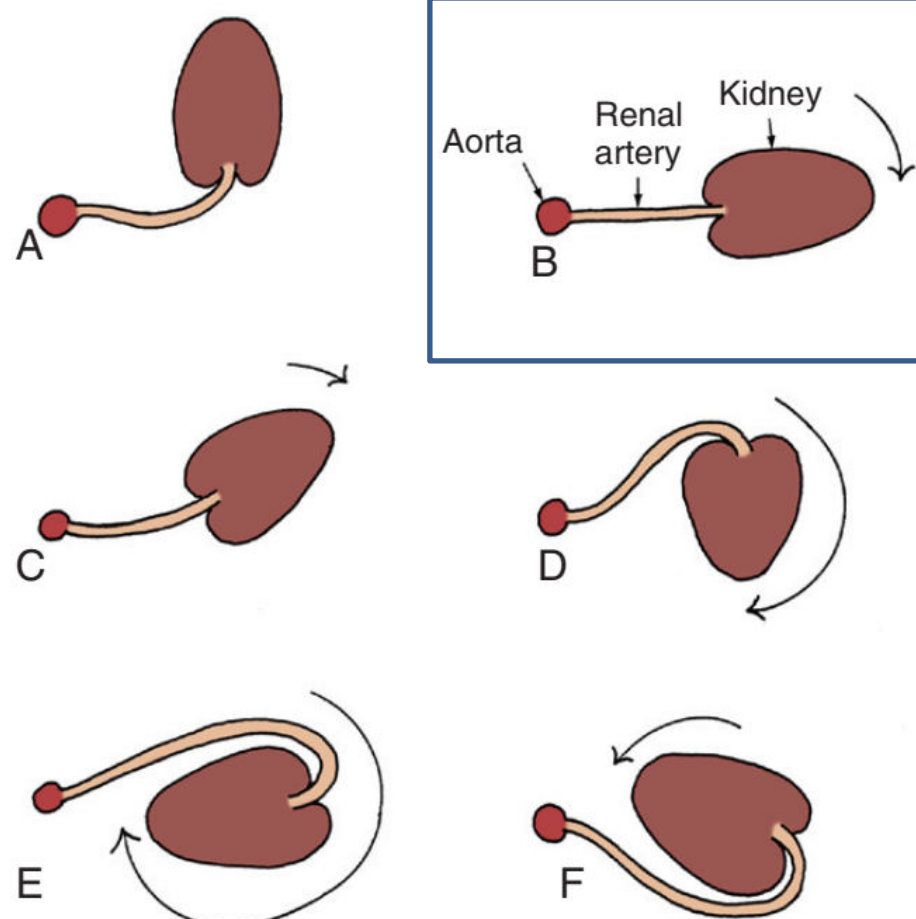




ANOMALIES OF ROTATION

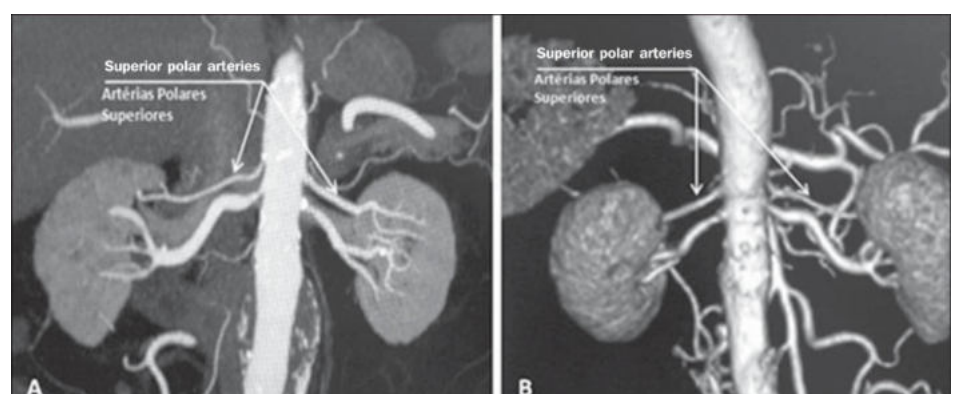
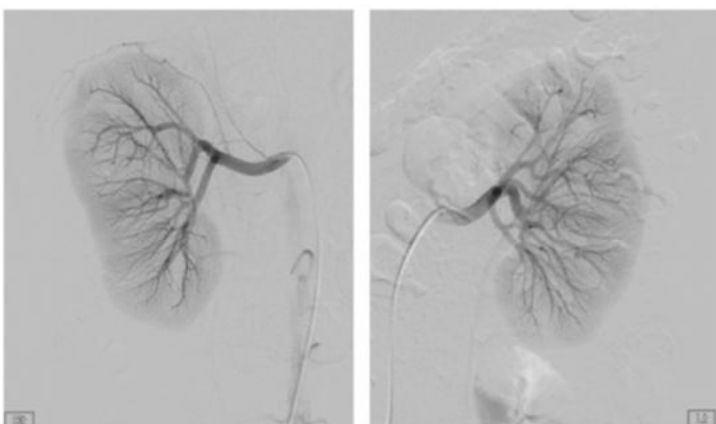
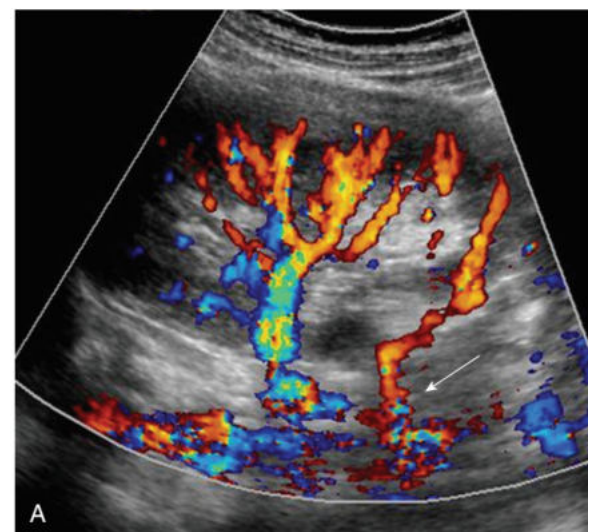
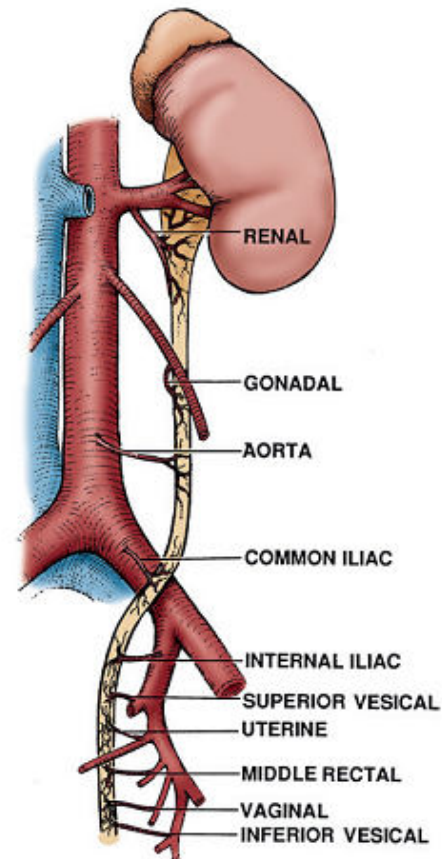
The kidney and renal pelvis normally rotate 90 degrees ventromedially during ascent so that the calyces point laterally and the pelvis faces medially.

When this alignment is not exact, the condition is known as **malrotation**



ANOMALIES OF RENAL VASCULATURE

- **Aberrant**- not arising from aorta (iliac/gonadal/mesenteric)
- **Accessory**-more than one artery supplying same segment
- **Multiple Vessels**-more than one artery supplying different segment



- Nephrectomy
- Transplantation

Renal Artery Aneurysm

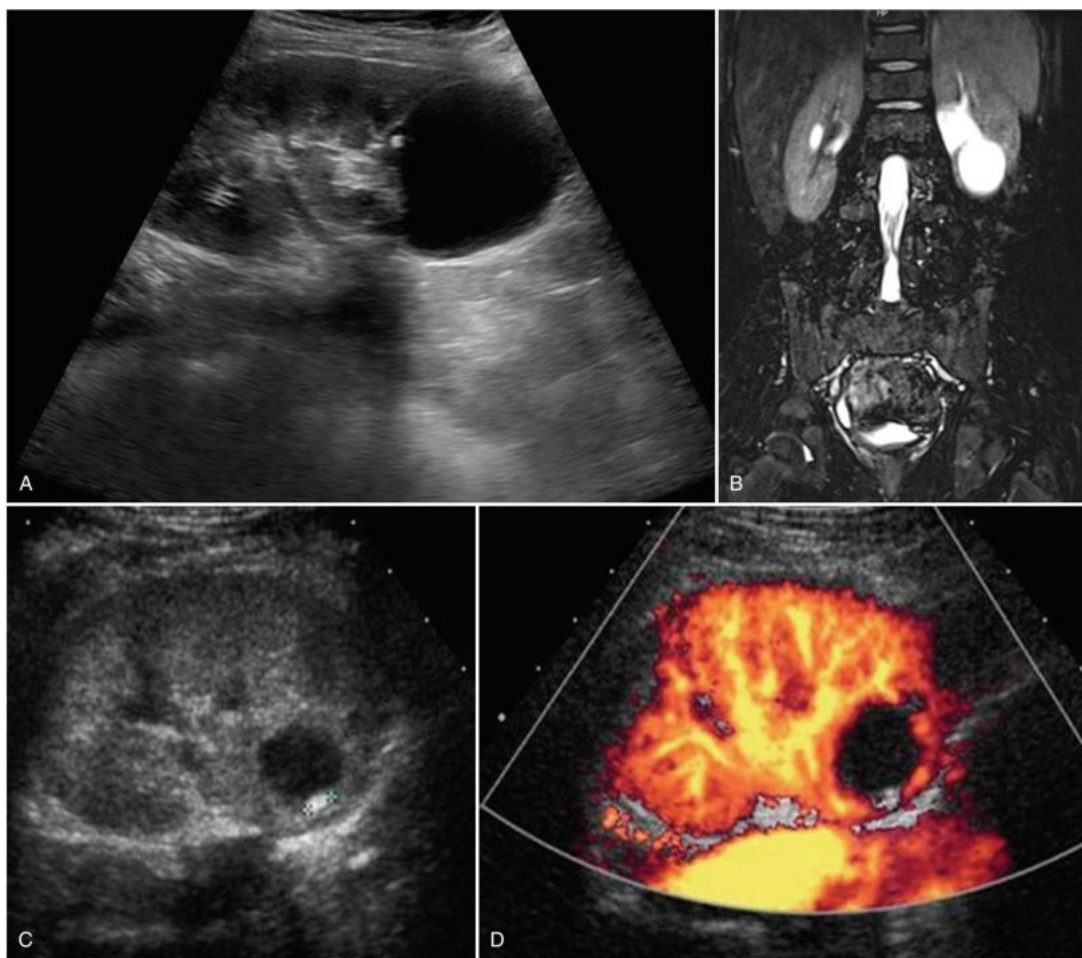
- Incidence-0.1% - 0.3%.
Most -silent,
- diagnosed during an evaluation of hypertension.
- pulsatile mass in the
region of the renal hilum or
- abdominal bruit
- calcification in the area of the renal
artery or its branches (30%) is highly suggestive

Renal Arteriovenous Fistula

- congenital
- acquired
 - trauma, inflammation, renal surgery, or percutaneous needle biopsy)
- Hypertension, Cardiac failure
- CD/MRA/CTA/DSA
- Angioembolisation/Surgical ligation

ANOMALIES OF THE COLLECTING SYSTEM

- **Calyceal Diverticulum**
 - a cystic cavity within the kidney lined by transitional epithelium communicates with a calyx or, less commonly, with the renal pelvis through a narrow isthmus.
 - Dx**-CT or MR urography.
 - T/t**-marsupialization of the diverticulum, fulguration of the epithelial lining,



- **Hydrocalycosis** –obstruction/VUR
- **Megacalycosis**-Nonobstructive
- **Extrarenal Calyces**
- **Infundibulopelvic Stenosis**
- **Extrarenal Pelvis**
- **Bifid pelvis**

Congenital cyst

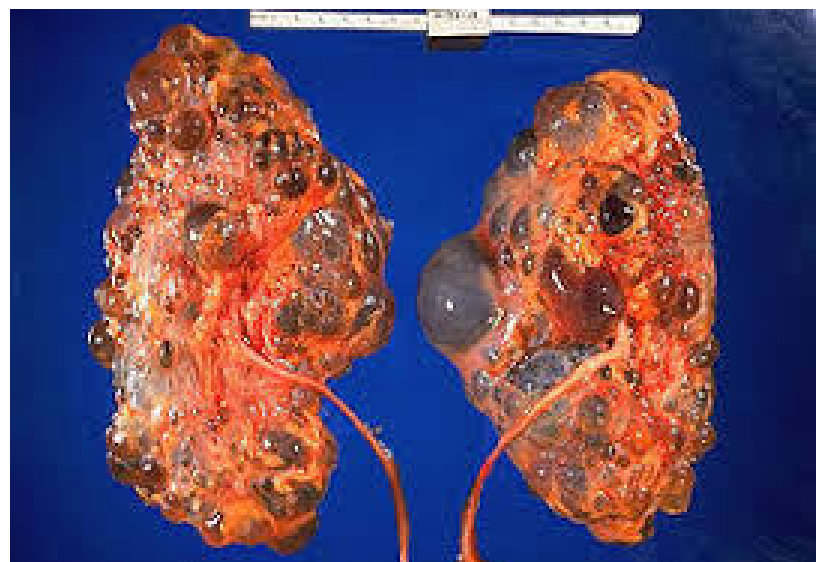
Congenital/Acquired

- Autosomal Recessive (Infantile) Polycystic Kidney Disease
- Autosomal Dominant (Adult) Polycystic Kidney Disease
- Juvenile Nephronophthisis and Medullary Cystic Disease Complex
- Other Inheritable Renal Cystic Diseases (Congenital Nephrosis)
- Familial Hypoplastic Glomerulocystic Kidney Disease (Cortical Microcystic Disease)
- Multiple Malformation Syndromes with Renal Cysts
- 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)
- Parapelvic and Renal Sinus Cysts

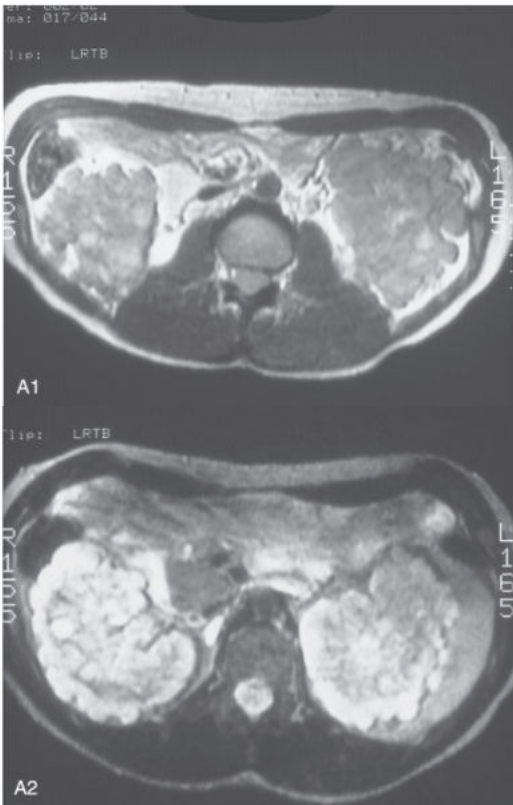
ARPKD



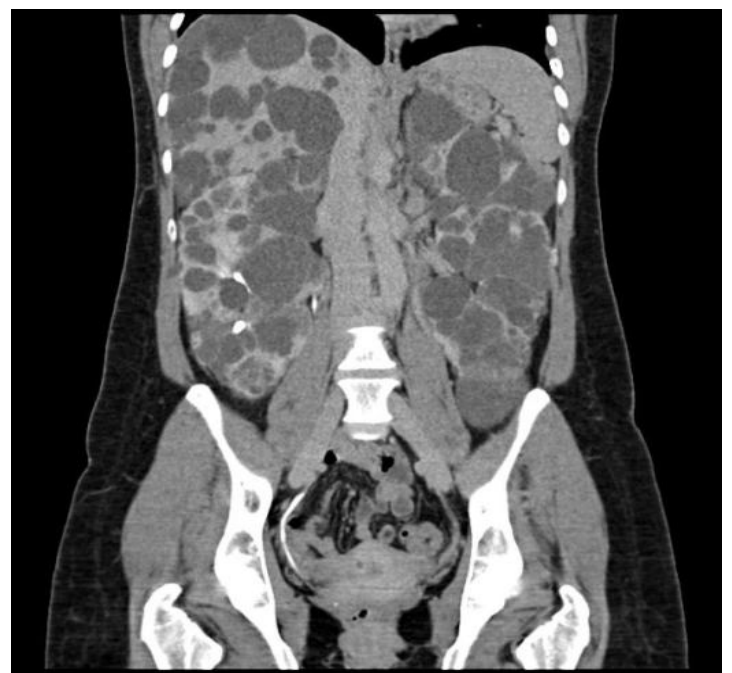
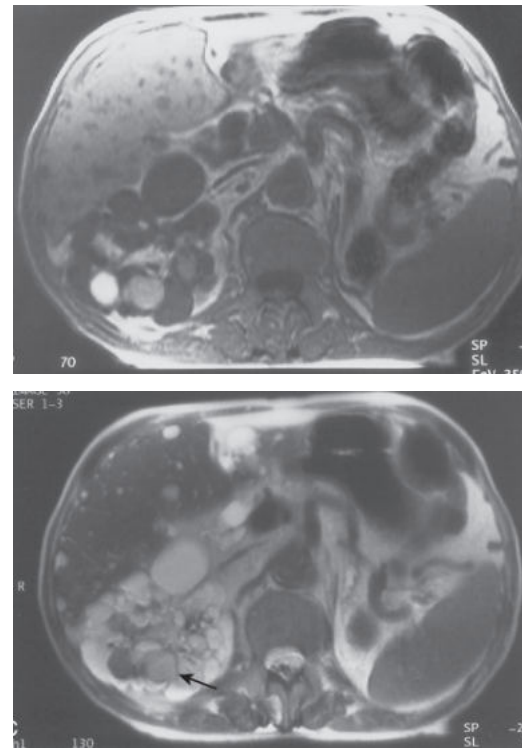
ADPKD



ARPKD



ADPKD



Chromosome 6
1: 5000-40,000
Perinatal
Symmetrically enlarged, homogeneous,
hyperechogenic
Collecting duct ectasia; cysts derived
principally from collecting duct
Always with congenital hepatic fibrosis
but of varying severity
None

Chromosome 4 and 16
1: 500-1100
Third to fifth decades
Large, cystic kidneys, sometimes asymmetrical

Microcysts and macrocysts derived from the
entire nephron
Cysts, mostly in adults

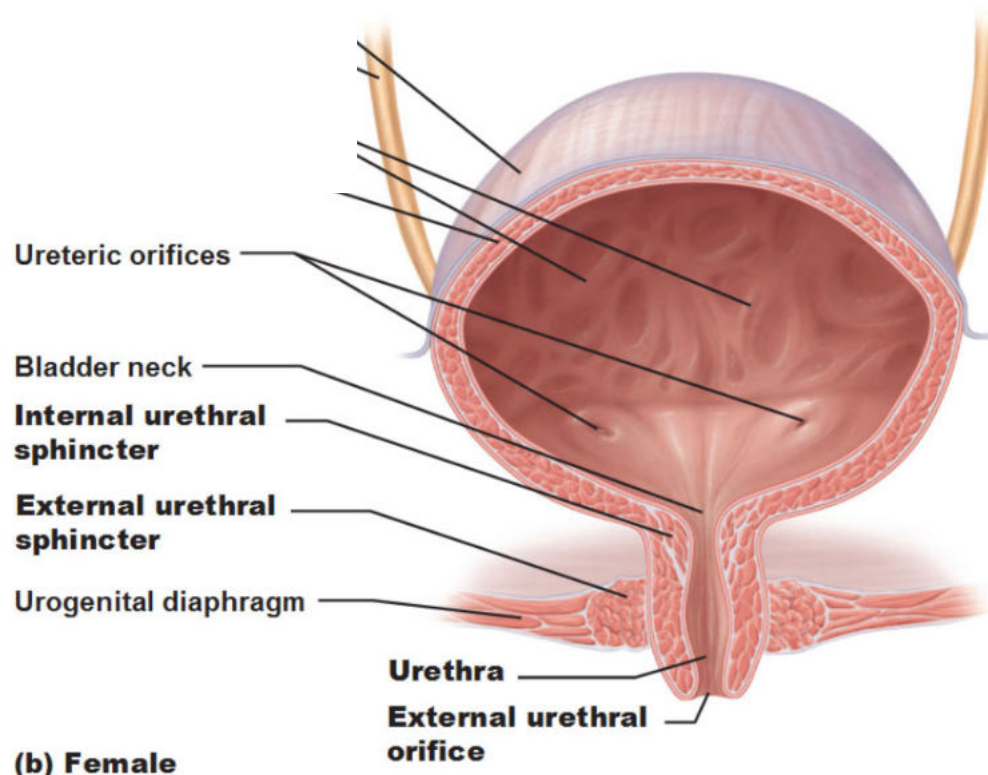
Intracranial (berry) aneurysms, colonic
diverticuli; mitral valve prolapse; cysts of
other organs (seminal vesicle, arachnoid
membrane, pancreas)

- PKDH1 gene
- Firocystin (polyductin)
- Renal/ hepatic failure and portal hypertension
- Most die early

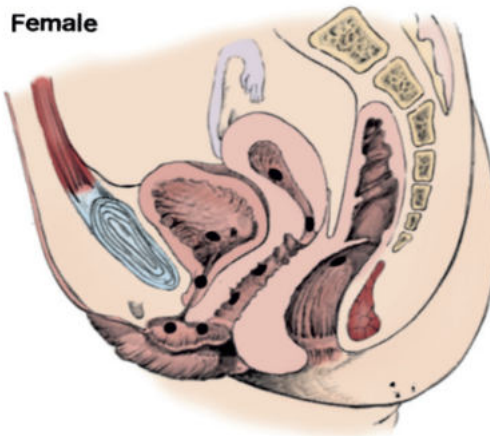
PKD 1 and PKD 2 gene
Hypertension, ESRD
Anti hypertensive(ACE inhibitors)
Renoprotective measures
Aspiration/ Deroofing / Nephrectomy

Ectopic ureter and Ureterocoele

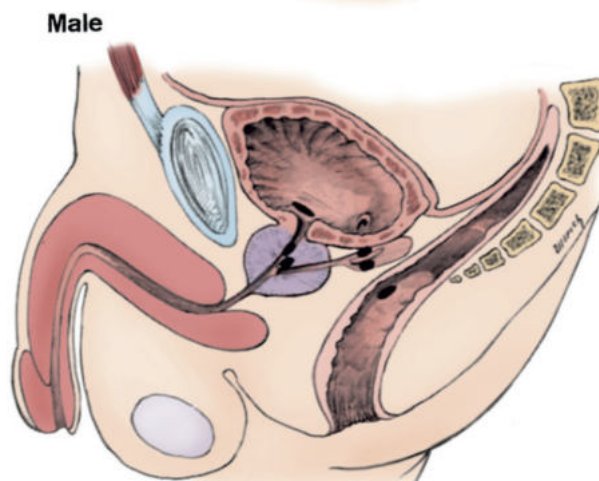
Urinary Bladder and Urethra – Female



Ectopic Ureter does not enter the trigonal area



vagina, uterus,
Perineum,
urethra, rectum

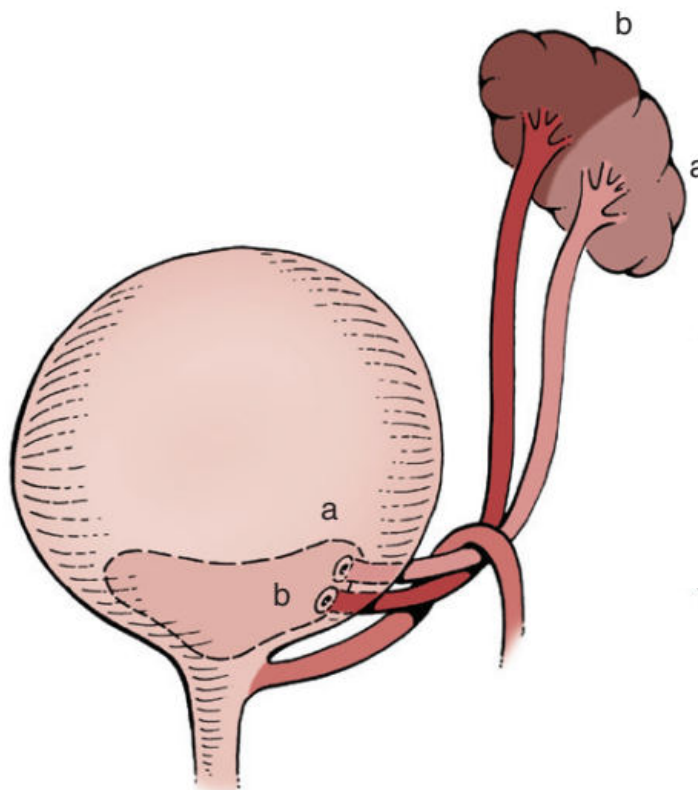


Seminal vesicle, vas
deferens, prostatic
urethra,

- **Ureterocele** –
- cystic dilation of the distal ureter
- Located either within the bladder or spanning the bladder neck and urethra.

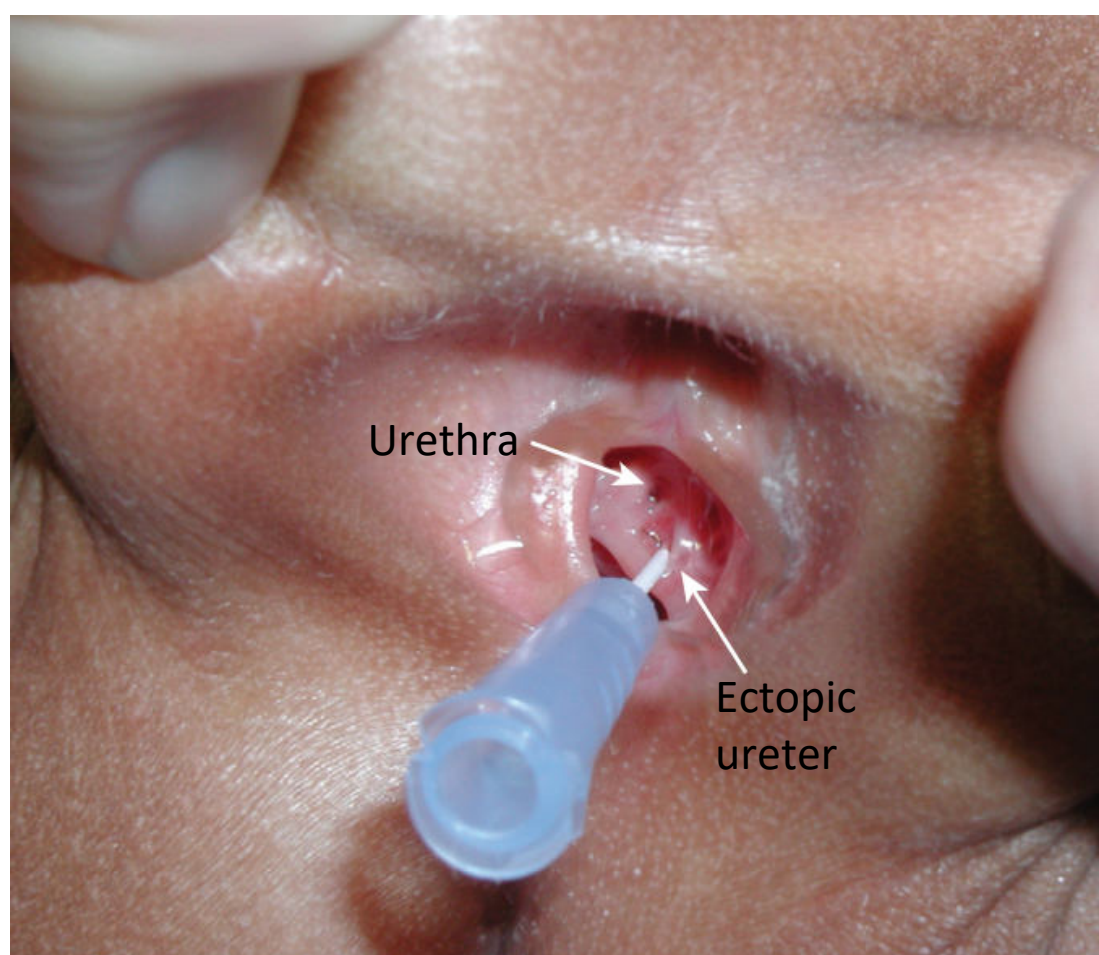


- Single / duplex system
- Unilaterally/ bilaterally
- MC duplex system unilaterally



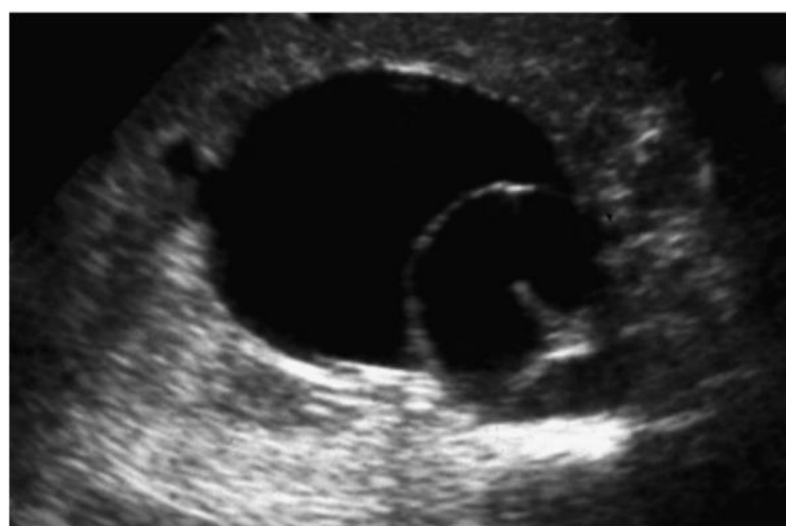
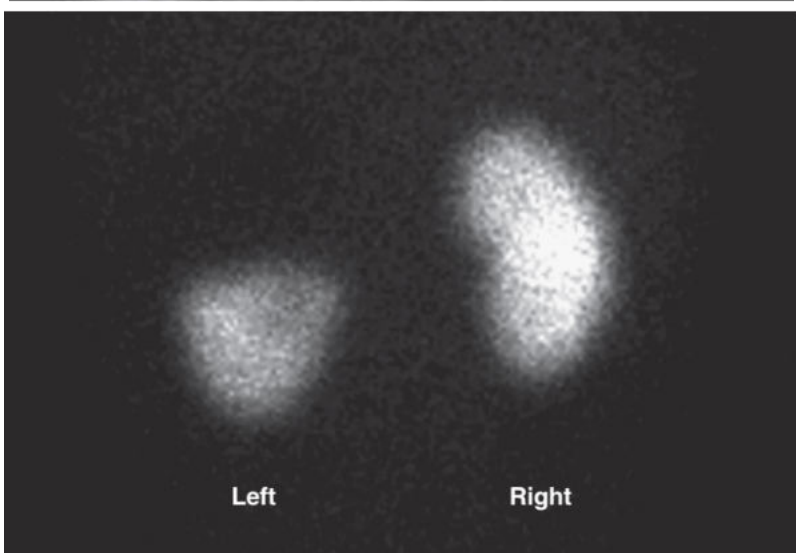
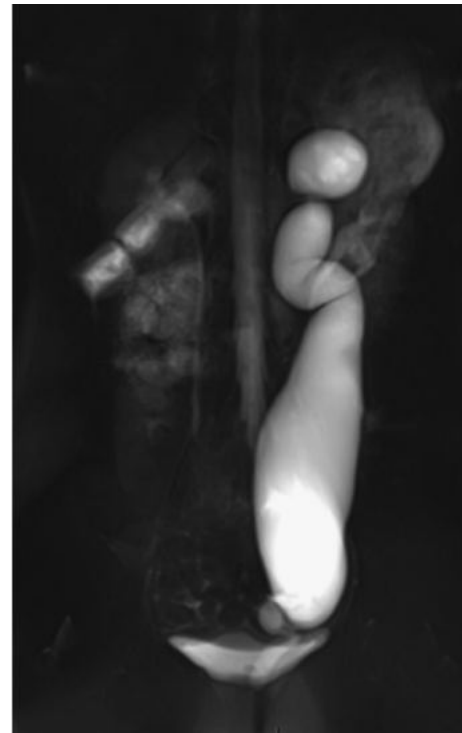
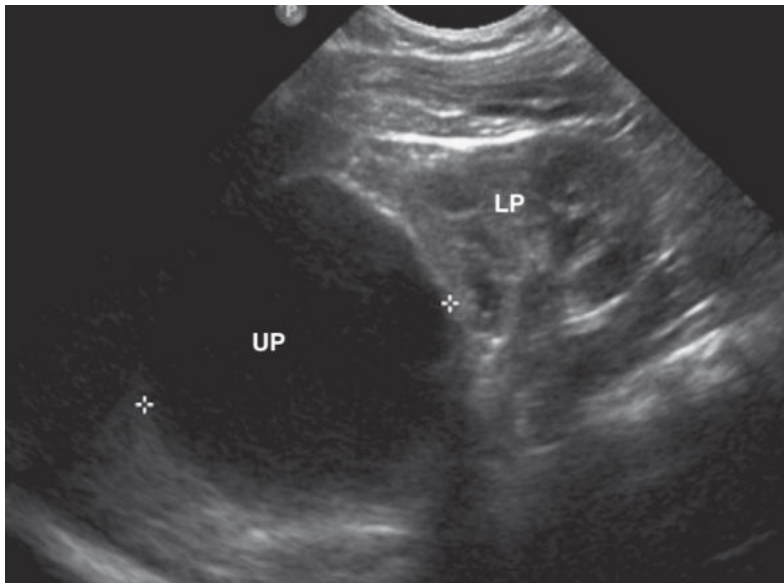
In a duplex system the ectopic ureter is inevitably the upper pole ureter due to its budding from the mesonephric duct later (more cephalad) than the lower pole ureteral bud.

- C/F-
- Incidental
- Vague abdominal discomfort
- Recurrent UTI
- Incontinence
- Prolapsed ureterocoele

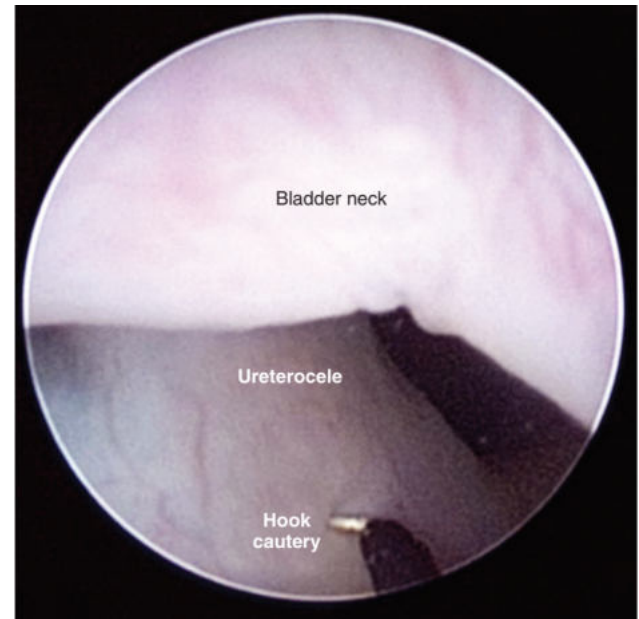
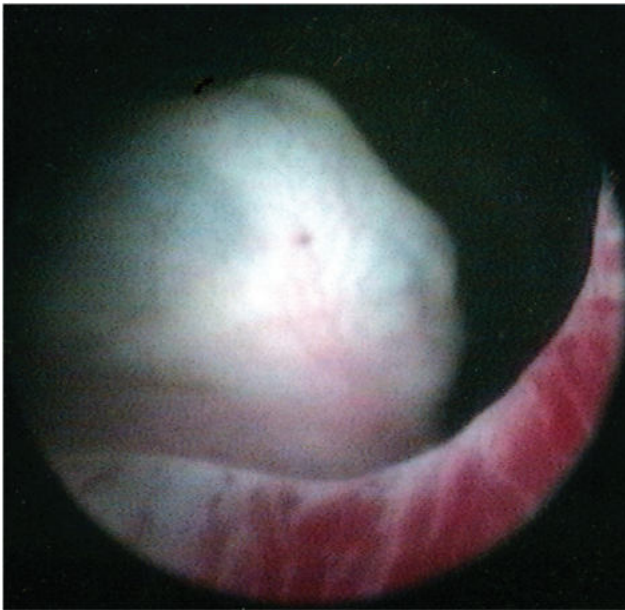




- Diagnosis:
 - USG
 - IVP
 - CT Urography
 - MRU
 - Endoscopy



Ureterocoele



- Transurethral incision
- Ureterocele excision
- Upper pole nephrectomy
- Ureteric reimplantation

- Thank you

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