Congenital Pediatric Urinary Disorders

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Learning Objectives:

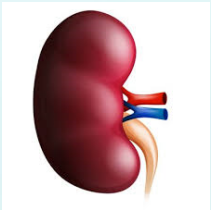
- Identify the common congenital anomalies.
- How to detect this anomaly on radiological investigations.
- Important steps in management.

Congenital Urinary Disorders

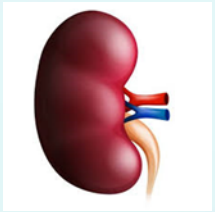
- Anomalies of the Upper Urinary Tract
  - Kidney
  - Ureter
- Anomalies of the Lower Urinary Tract
  - Urinary Bladder
  - Urethra

Anomalies of the Upper Urinary Tract

kidney



Anomalies of the kidney



- **Anomalies of:**
  - Number
  - Ascent
  - Form and Fusion
  - Rotation

Anomalies of the kidney

- **Anomalies of:**
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Anomalies of Number

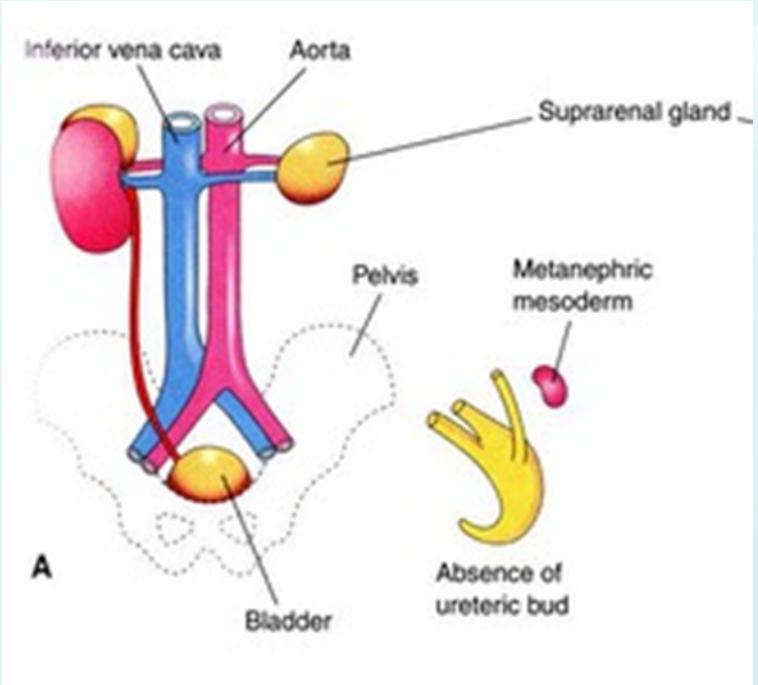
- Renal Agenesis:
  - Unilateral
  - Bilateral

- Supernumerary Kidney

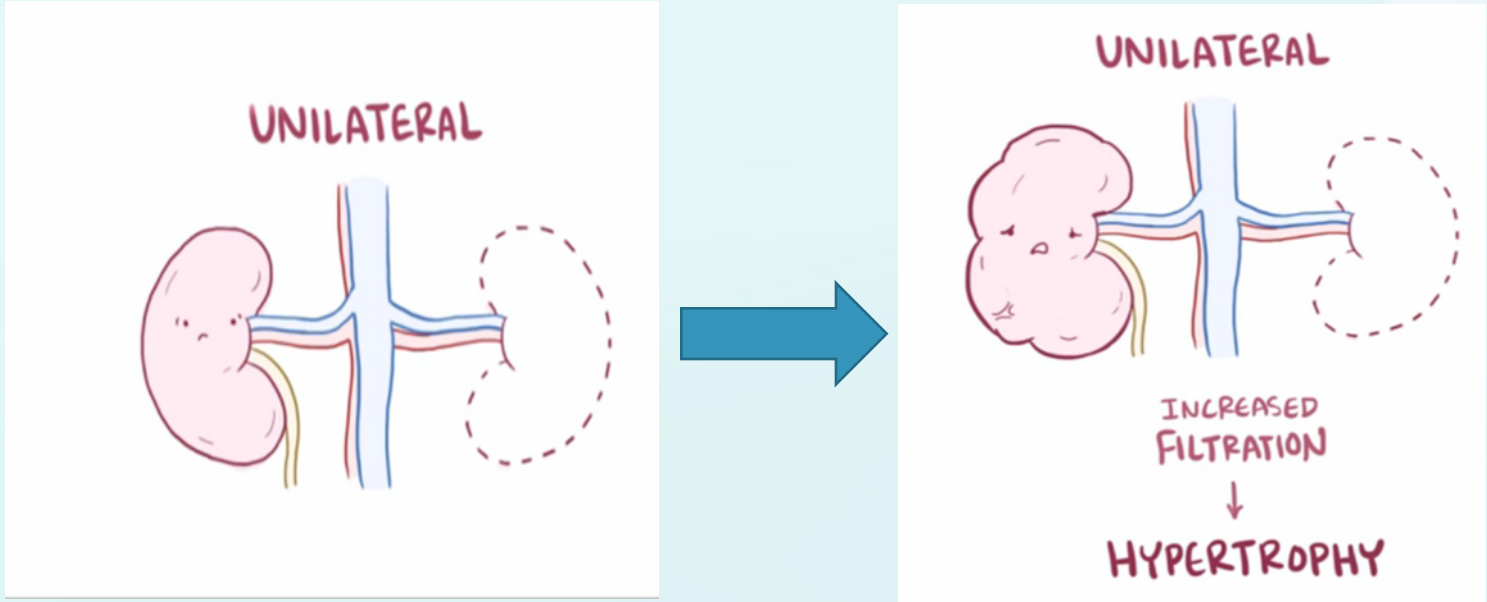
Unilateral Renal Agenesis (URA)

- 1 in 1100 births.
- Male: Female of 1.8 : 1
- The left side is absent more frequently .
- The ipsilateral ureter is completely absent in 50%.
- Anomalies of other organ systems are found frequently in affected individuals

  CVS,GIT,MSC



URA



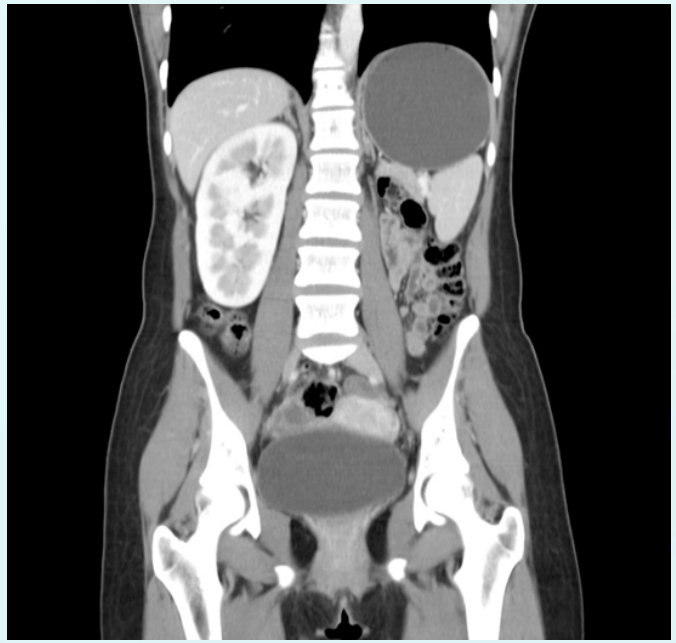
URA…

- Müllerian duct abnormalities occur in 25% to 50% of cases of females with URA compared with wolffian duct anomalies in 10% to 15% of males with URA.

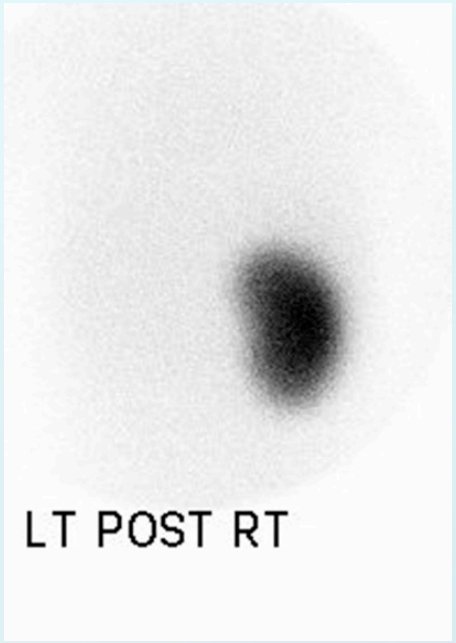
- Approximately one fourth to one third of women with müllerian duct anomalies are found to have URA.

Diagnosis

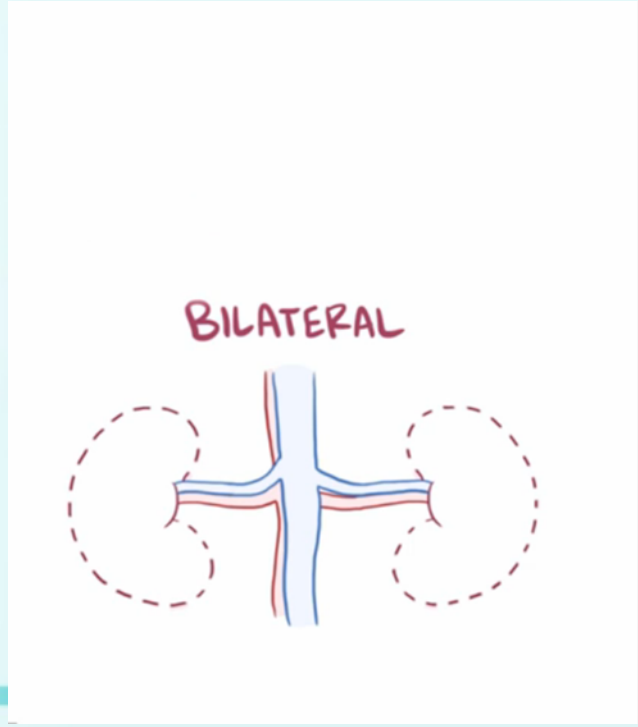
CT Abdomen



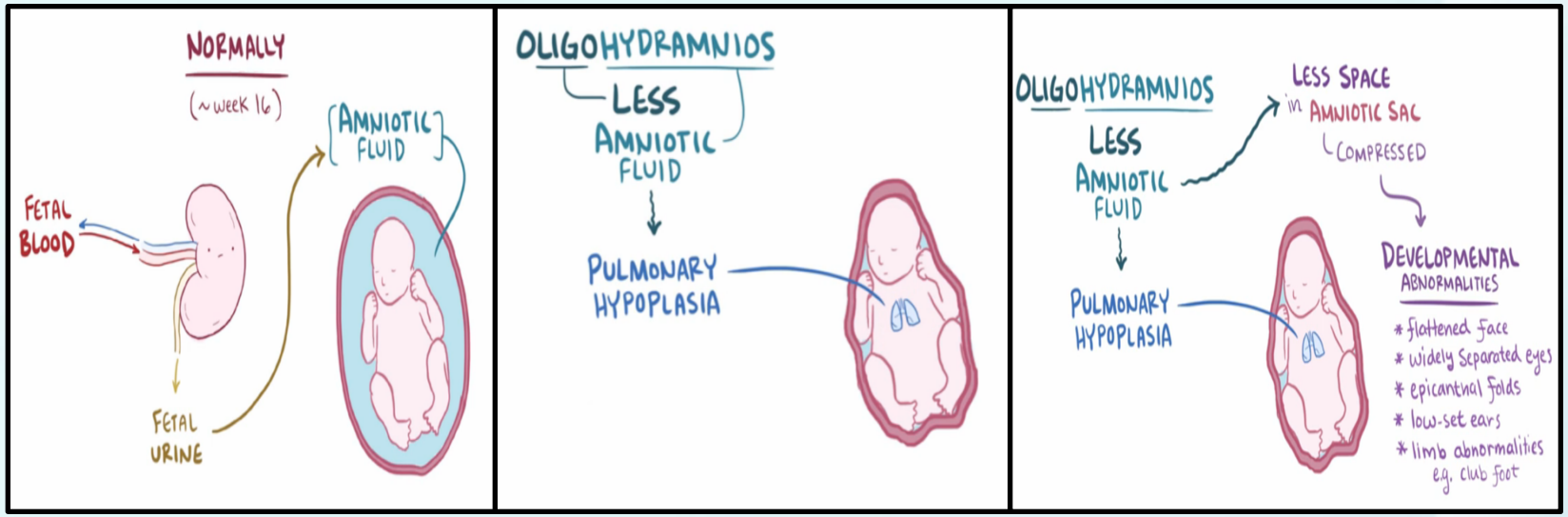
DMSA



Bilateral Renal Agenesis

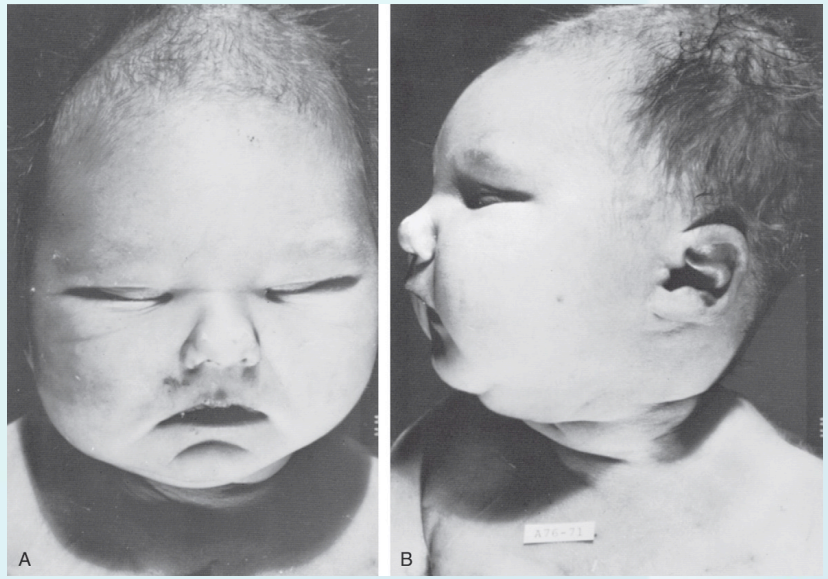


Bilateral Renal Agenesis…



Bilateral Renal Agenesis…

- 40% are stillborn.
- Do not survive beyond 48 hours due to respiratory distress associated with pulmonary hypoplasia.
- The characteristic
  - Potter's syndrome.
  - Oligohydramnios

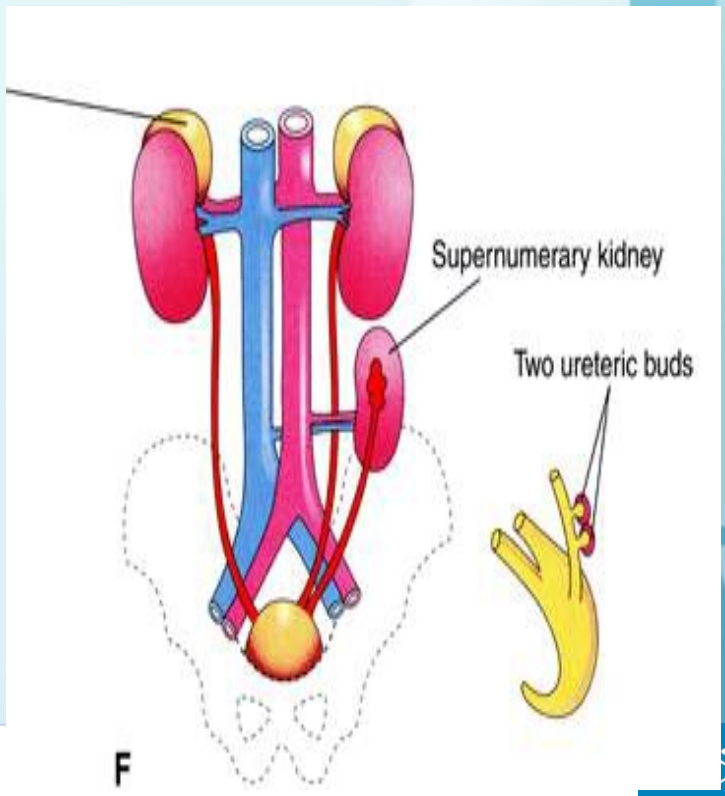


Bilateral Renal Agenesis

- Ureters are almost always *absent*.
- Bladder is either absent or *hypoplastic*.
- Adrenal glands are usually positioned *normally*.
- Müllerian duct anomalies are commonly observed.

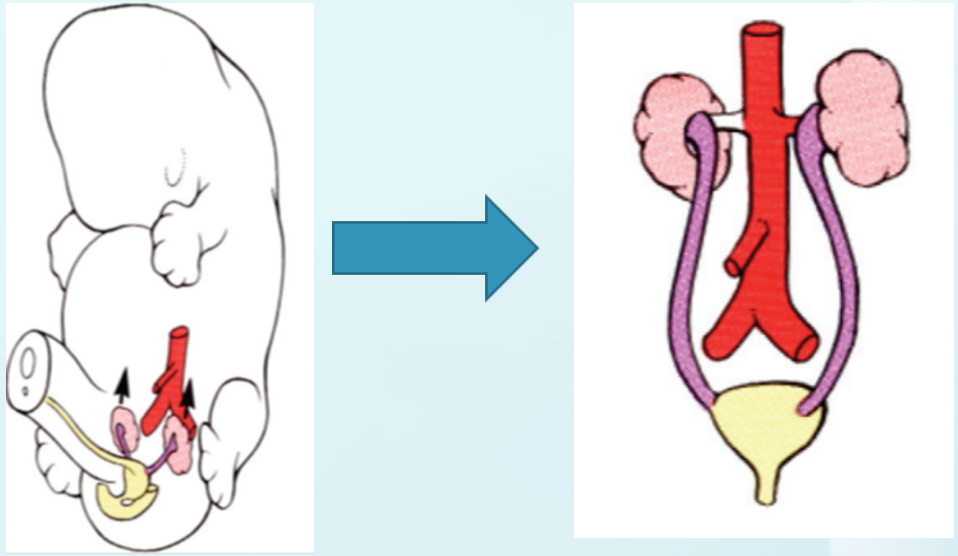
Supernumerary Kidney

- Definitive **accessory** organ with its own collecting system, blood supply, and distinct encapsulated parenchyma.
- Either completely separate or loosely attached to the kidney on the ipsilateral side.
- The ureteral inter-relationships on the side of the supernumerary kidney can be variable.



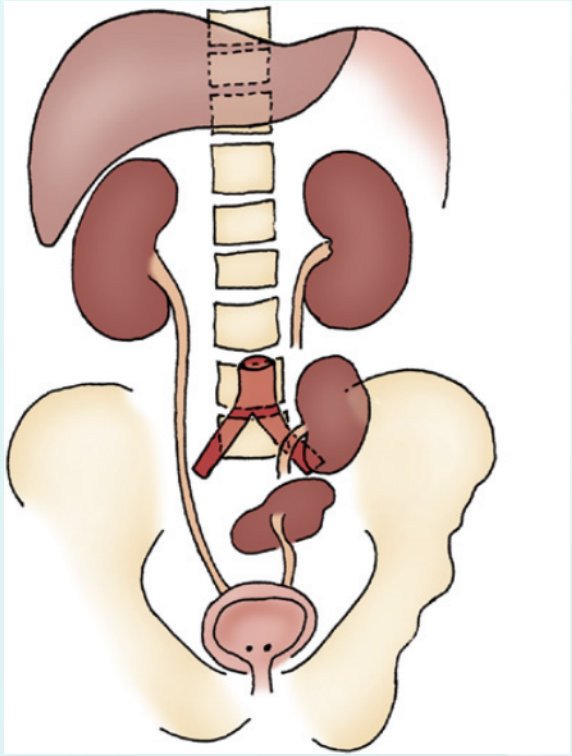
Anomalies of the kidney

- Anomalies of:
  - Number
  - **Ascent**
  - Form and Fusion
  - Rotation



Simple Renal Ectopia

- The left is more than the right.
- Pelvic ectopia has been estimated to occur in 1 of 2100 to 3000 autopsies.
- 50% have a hydronephrosis:
  - ✓ Obstruction: UPJO and UVJO
  - ✓ Reflux: grade III or greater
  - ✓ Malrotation
- VUR is found in 30%
- The incidence of genital anomalies in the patient with ectopia is about 15%.
- Most ectopic kidneys are clinically asymptomatic



Anomalies of Ascent

- Simple Renal Ectopia
- Cephalad Renal Ectopia
- Thoracic Kidney

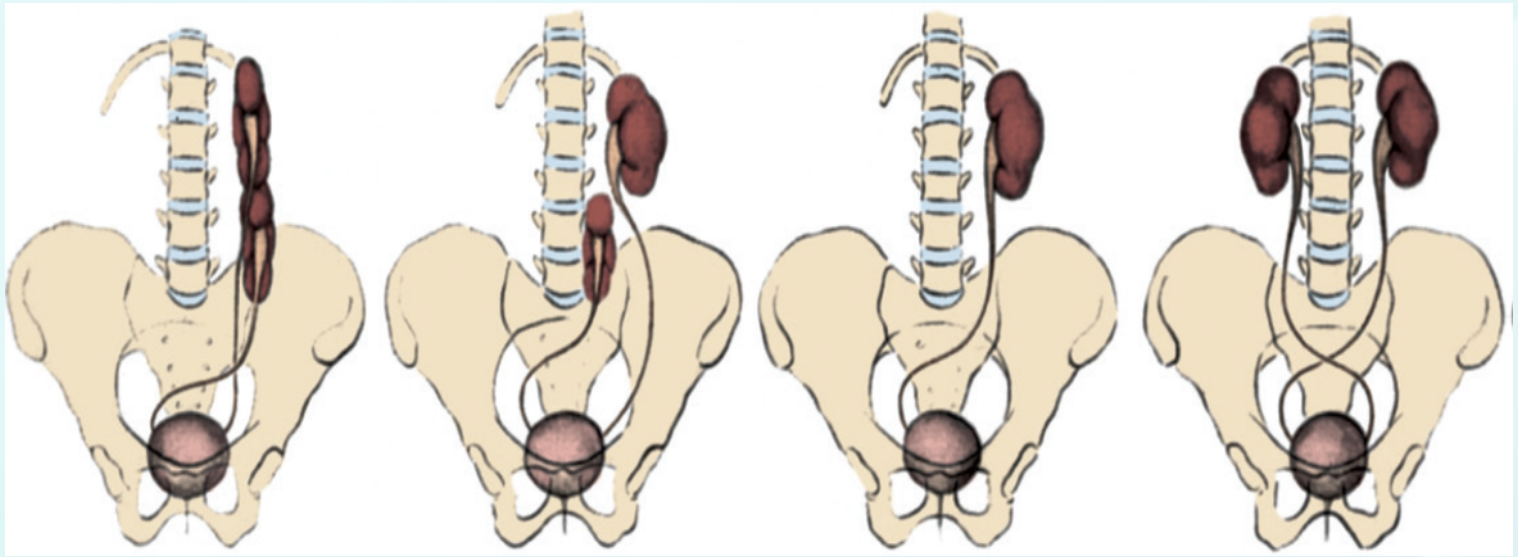
Anomalies of the kidney

- Anomalies of:
  - Number
  - Ascent
  - **Form and Fusion**
  - Rotation

Anomalies of Form and Fusion

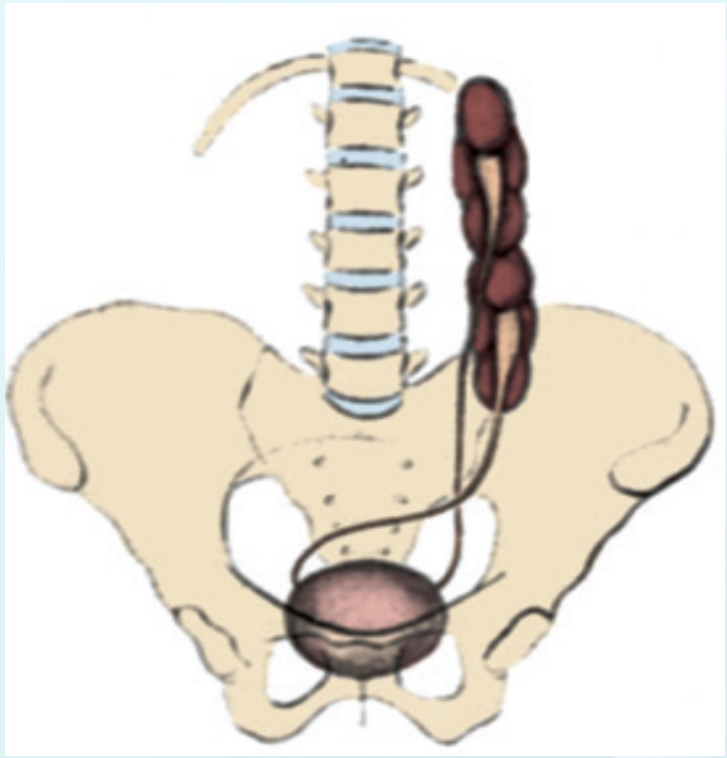
- Crossed Renal Ectopia:
  - with Fusion
  - without Fusion
- Horseshoe Kidney

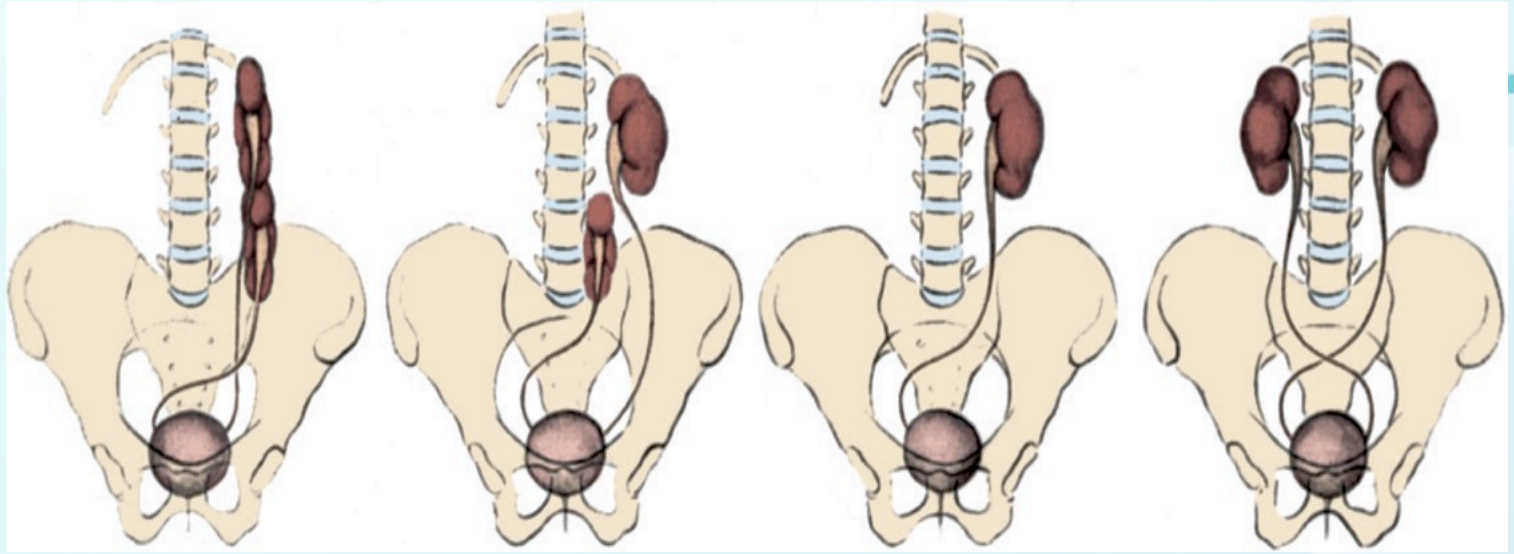
Crossed Renal Ectopia with and without Fusion



Crossed Renal Ectopia with and without Fusion

- **Crossed ectopia**: kidney is located on the side opposite from that in which its ureter inserts into the bladder.
- 90% are fused with their mate
- the superior pole of the ectopic kidney usually joins with the inferior aspect of the normal kidney.
- The ureter from each kidney is usually orthotopic.





A)
Crossed renal ectopia with fusion

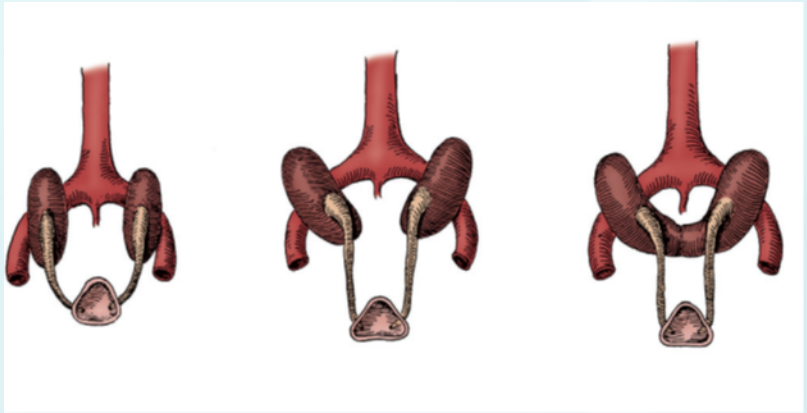
B)
Crossed renal ectopia without fusion

C)
Solitary crossed renal ectopia

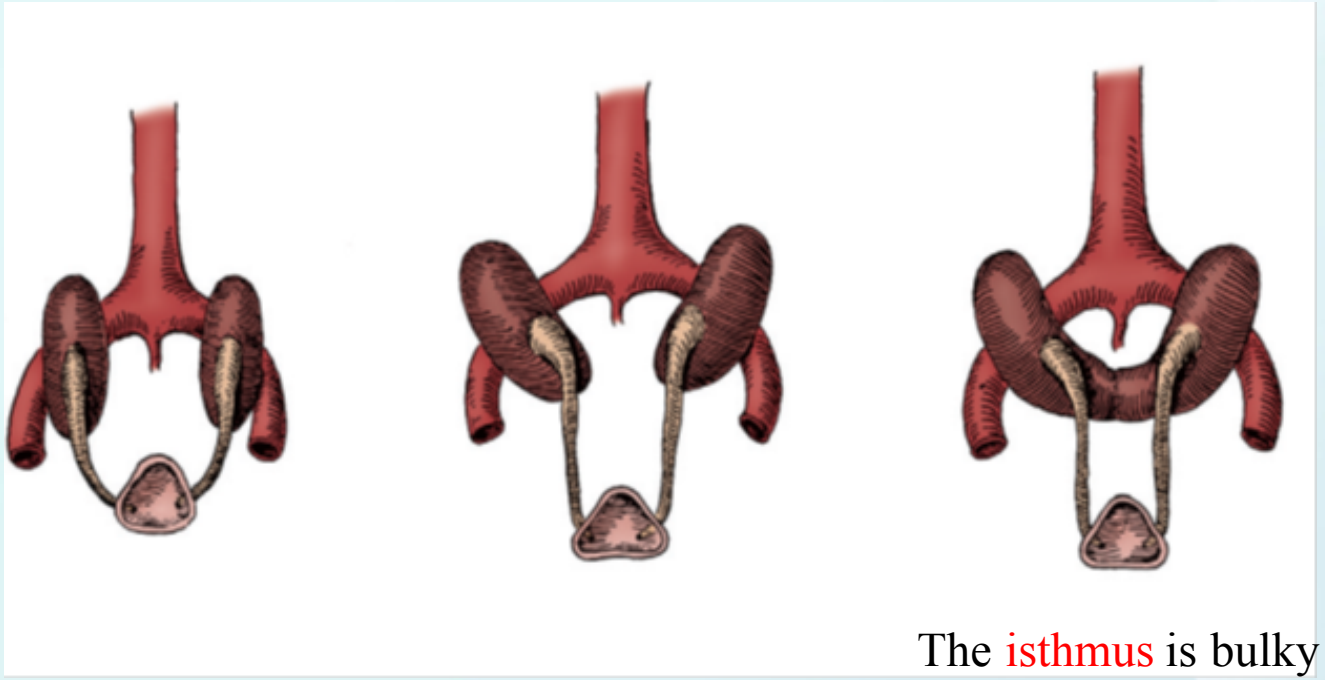
D)
Bilaterally crossed renal ectopia

Horseshoe Kidney

- Occurs 1 in 400 persons.
- The isthmus is bulky and consists of parenchymatous tissue.
- The calyces:
  - ✓ normal in number
  - ✓ atypical in orientation.
  - ✓ pelvis remains in the vertical or obliquely lateral plane
- The blood supply can be quite variable.
- Horseshoe kidney is frequently found in association with other congenital anomalies.
- UPJ obstruction in one third.
- 60 % asymptomatic.



Horseshoe Kidney



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Horseshoe Kidney

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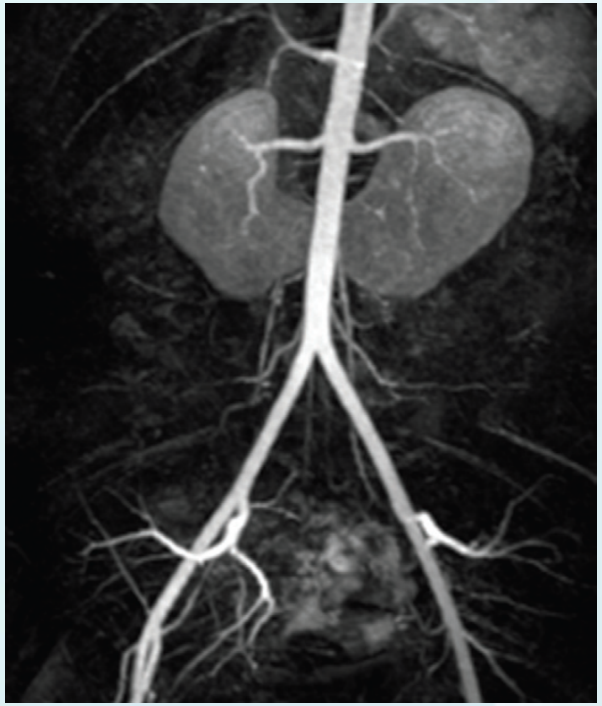

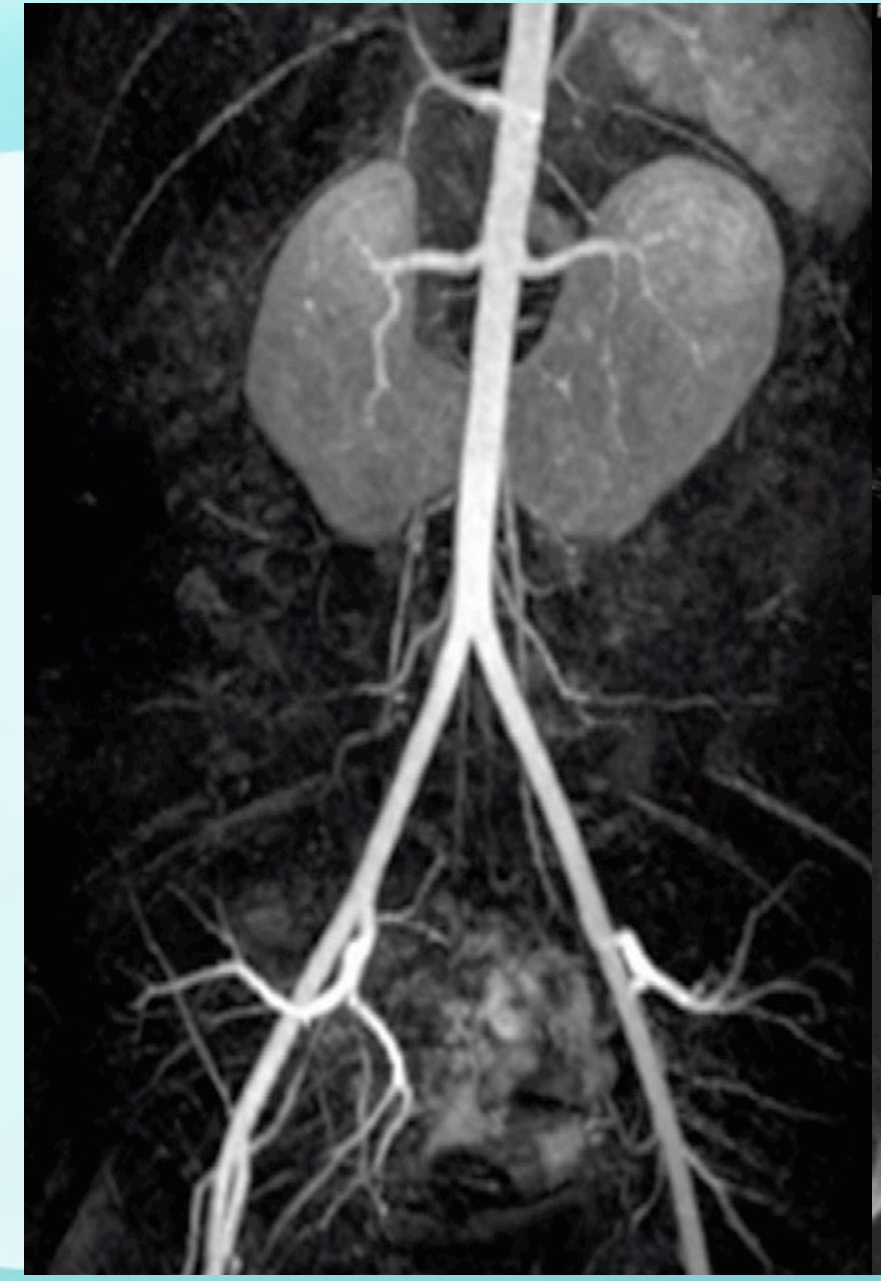

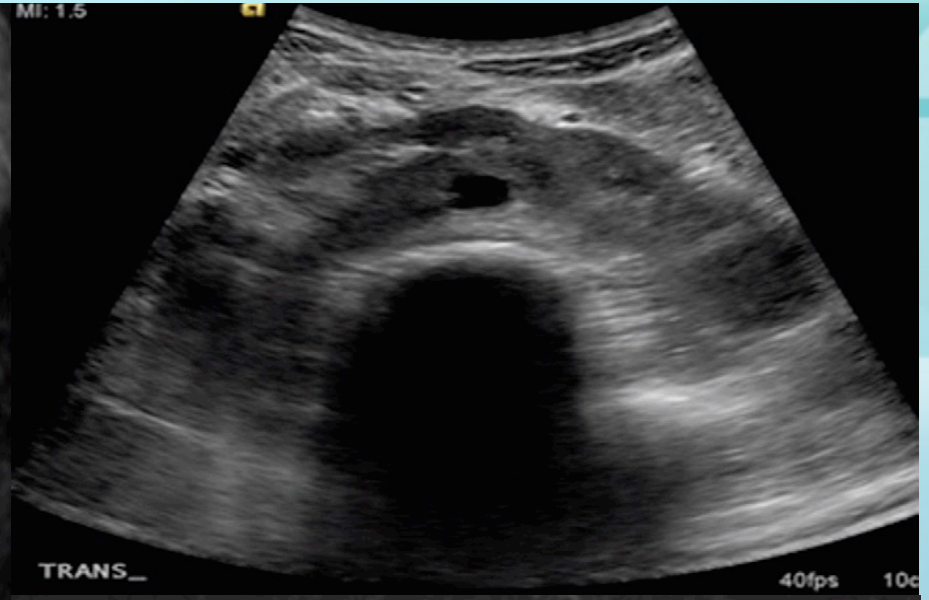
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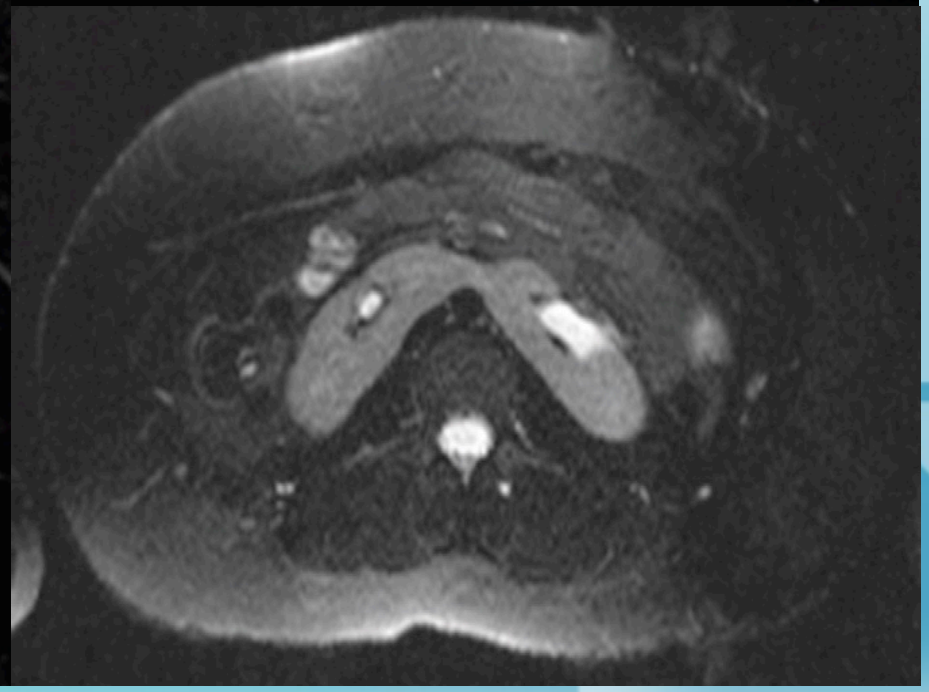
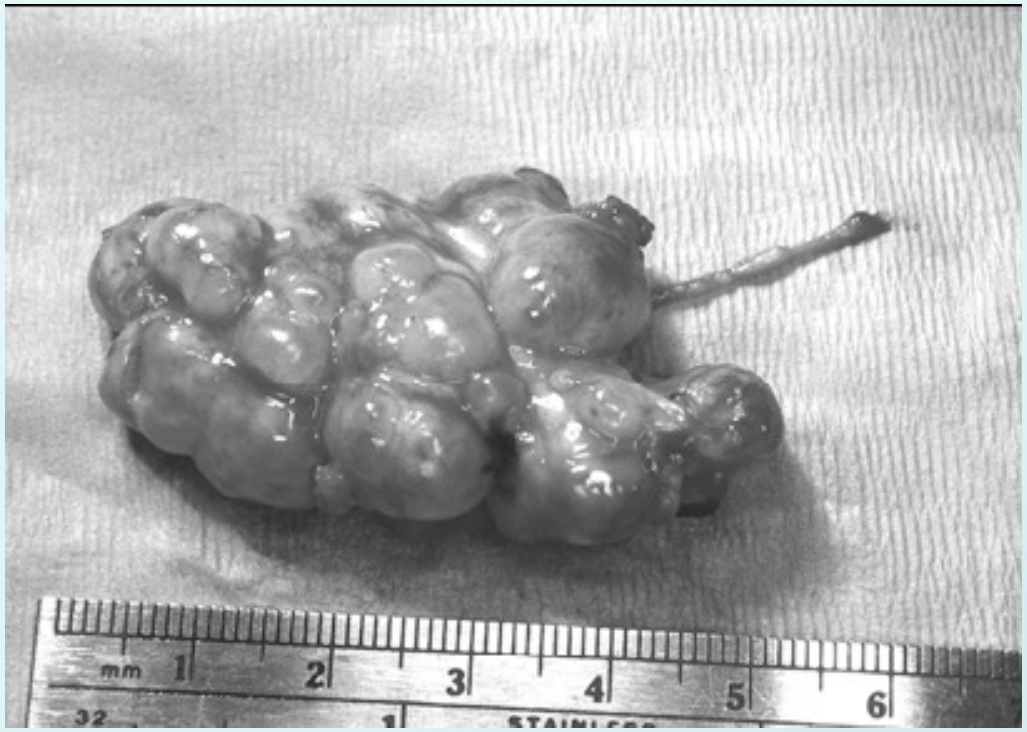


Multicystic Dysplastic Kidney(MCDK)



Anomalies of the kidney

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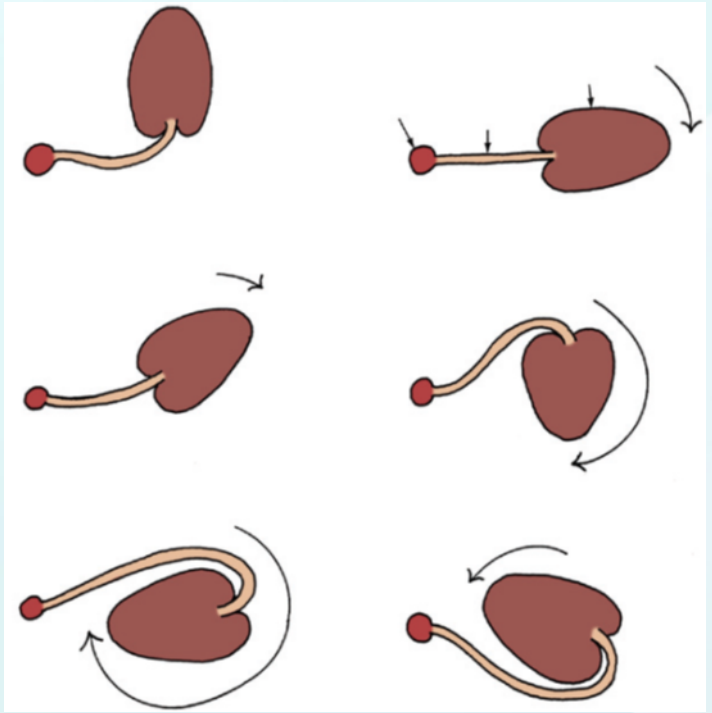
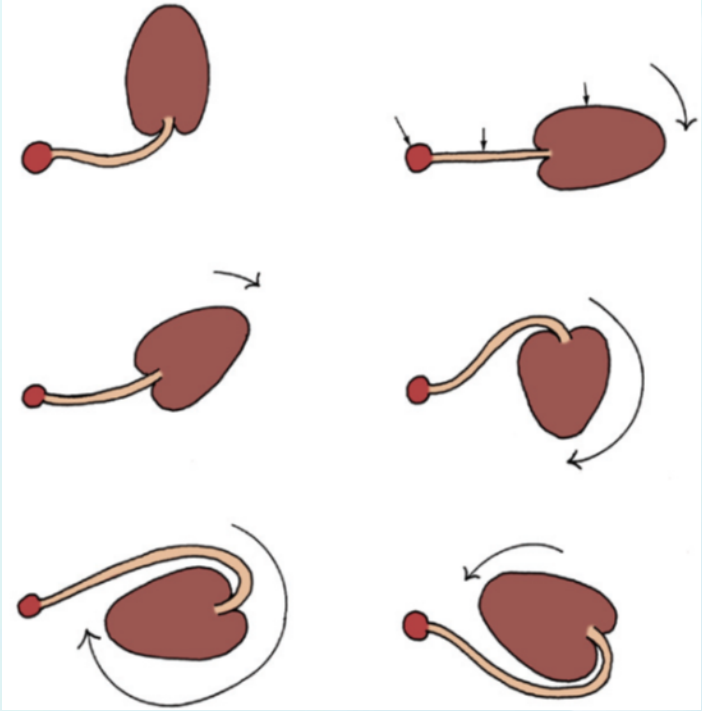
  - Number
  - Ascent
  - Form and Fusion
  - **Rotation**



- The kidney and renal pelvis normally rotate 90 degrees ventromedially during ascent
  - ✓ the calyces point laterally.
  - ✓ the pelvis faces medially.
- When this alignment is not exact, the condition is known as malrotation.
- Frequently associated with Turner syndrome.



Ureteral Anomalies

- Ureteropelvic junction (UPJ) obstruction
- Ureterovesical junction (UVJ) obstruction
- Megaureters
- Ectopic Ureter
- Ureterocele
- Vesicoureteral Reflux (VUR)

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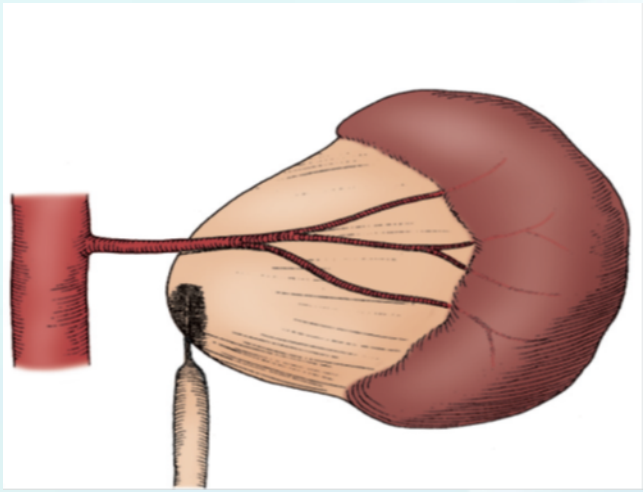
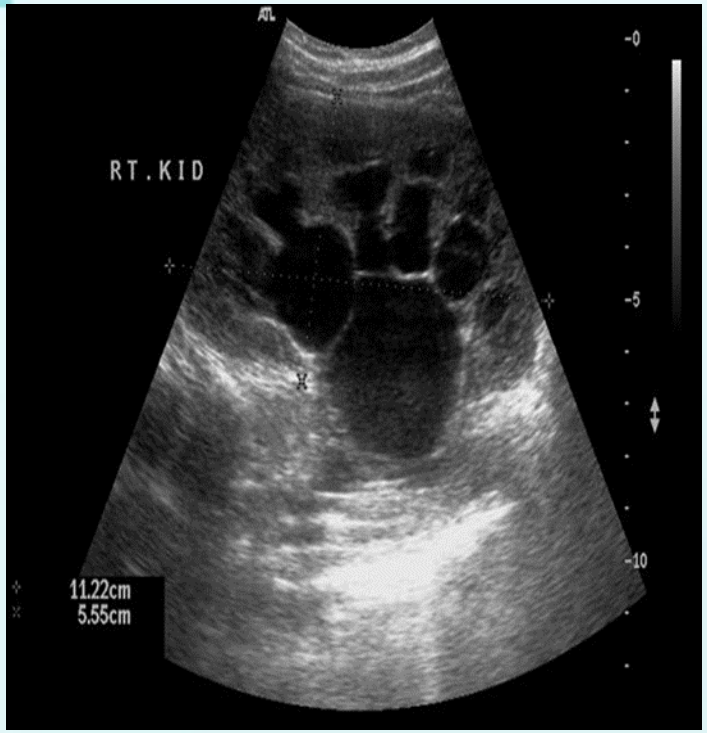
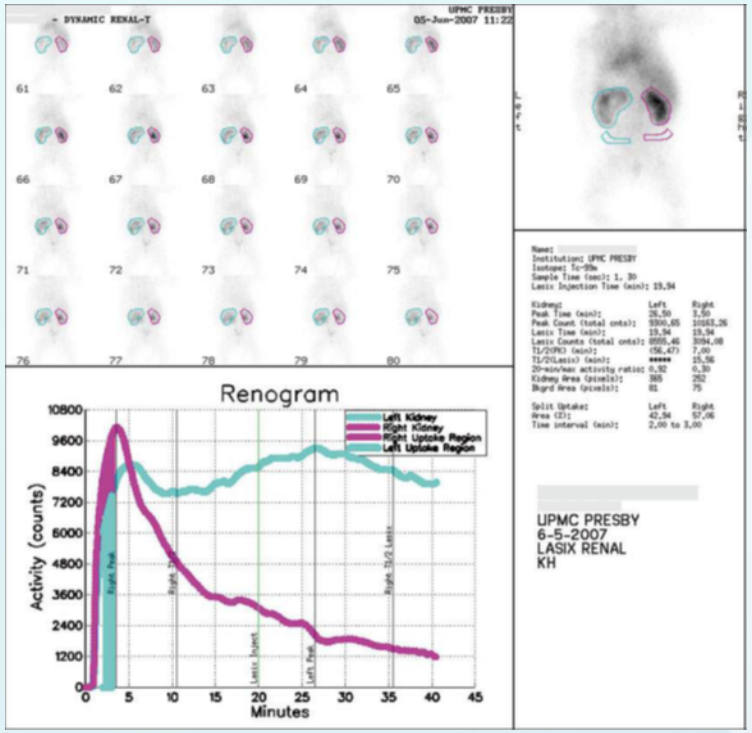


UPJ…

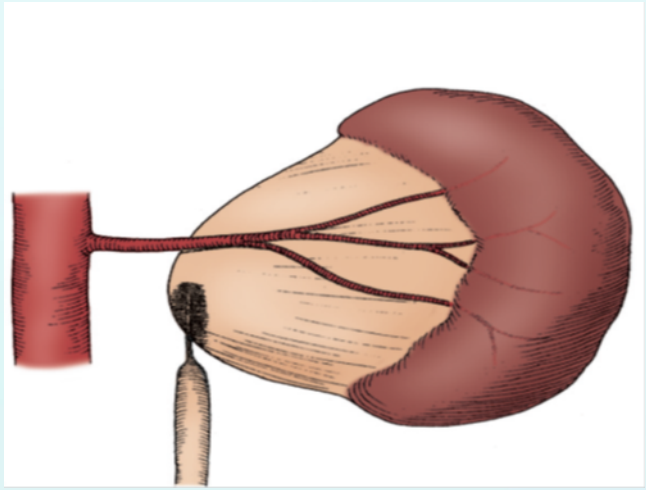
Presentation:

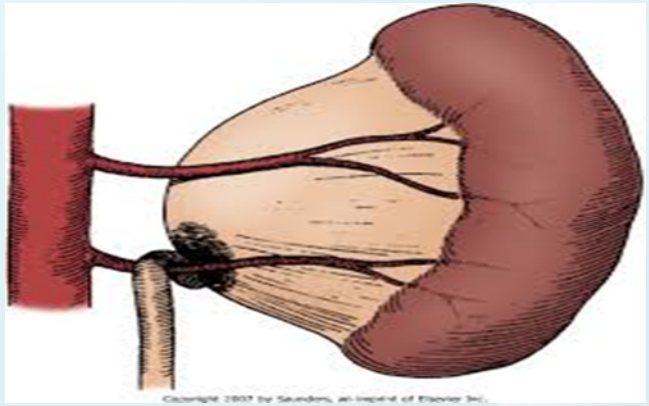
- Incidental in Neonates
- Incidental in Children
- Symptomatic:
  - ✓ UTI
  - ✓ Pain
  - ✓ Mass
  - ✓ Hematuria
  - ✓ Stone



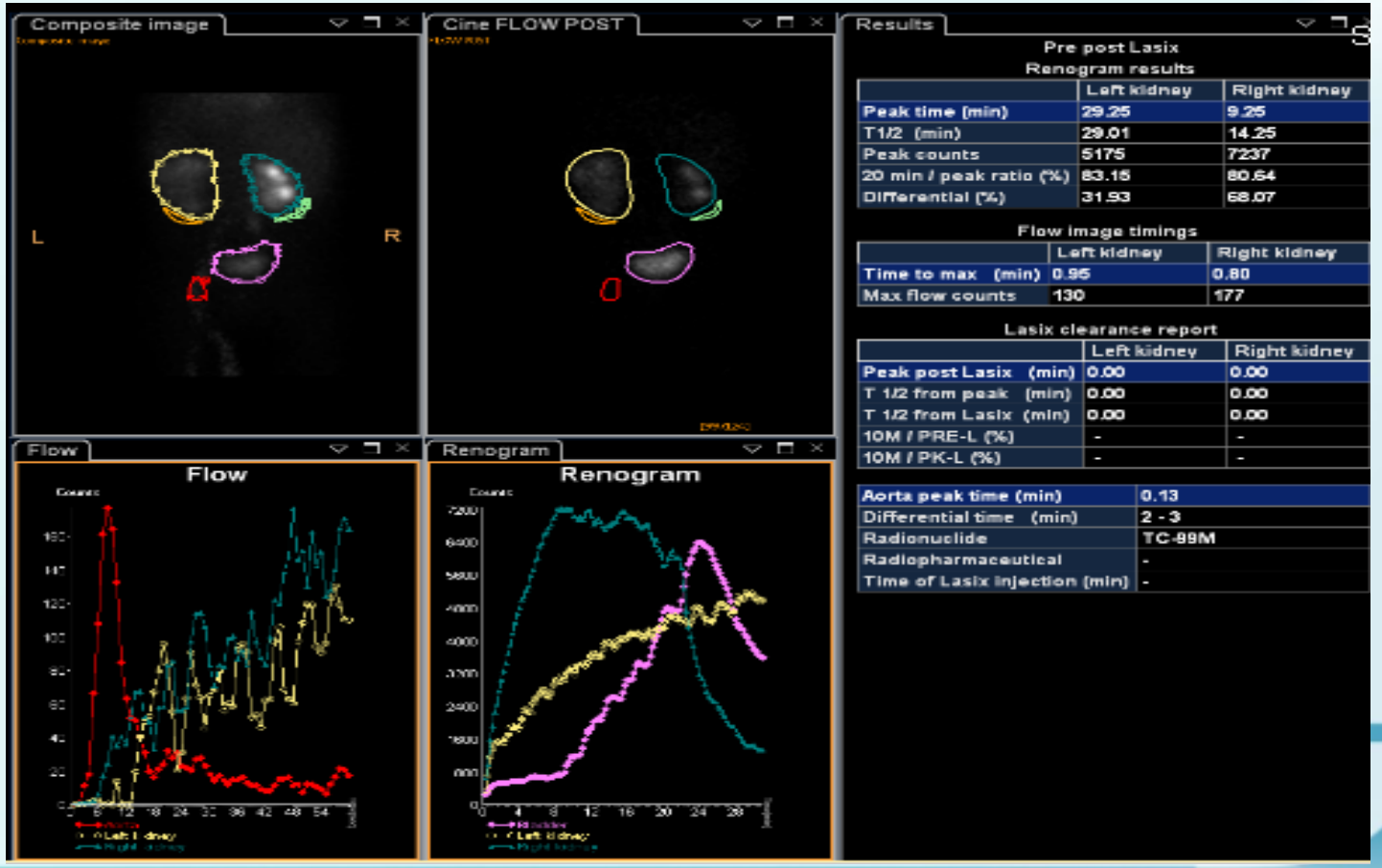


Ureteropelvic junction (UPJ) obstruction



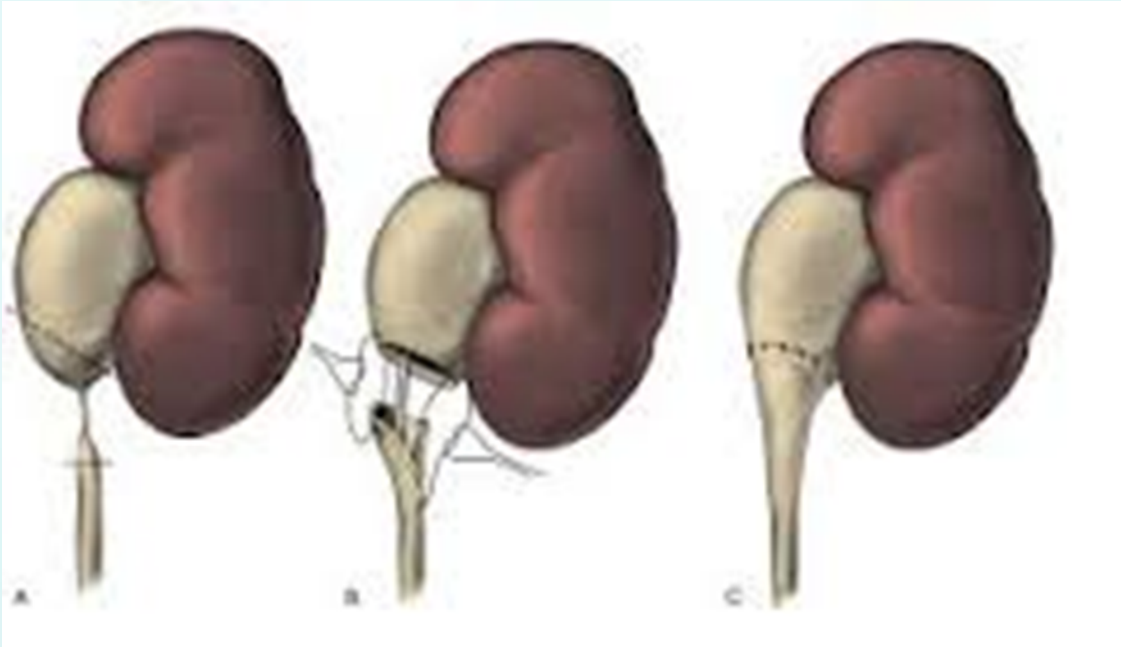


PUJO…
Dynamic renogram



PUJO…

Dismembered Pyeloplasty



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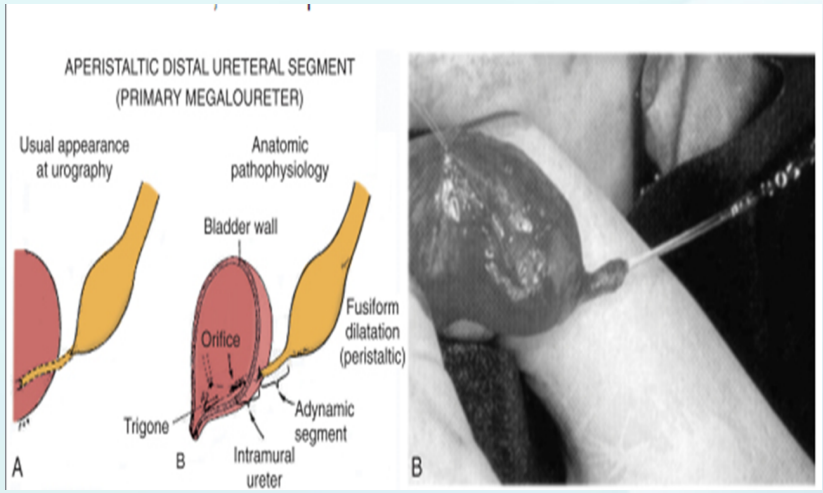
- Ureteropelvic junction (UPJ) obstruction
- **Ureterovesical junction (UVJ) obstruction**
- **Megaureters**
- Ectopic Ureter
- Ureterocele
- Vesicoureteral Reflux (VUR)



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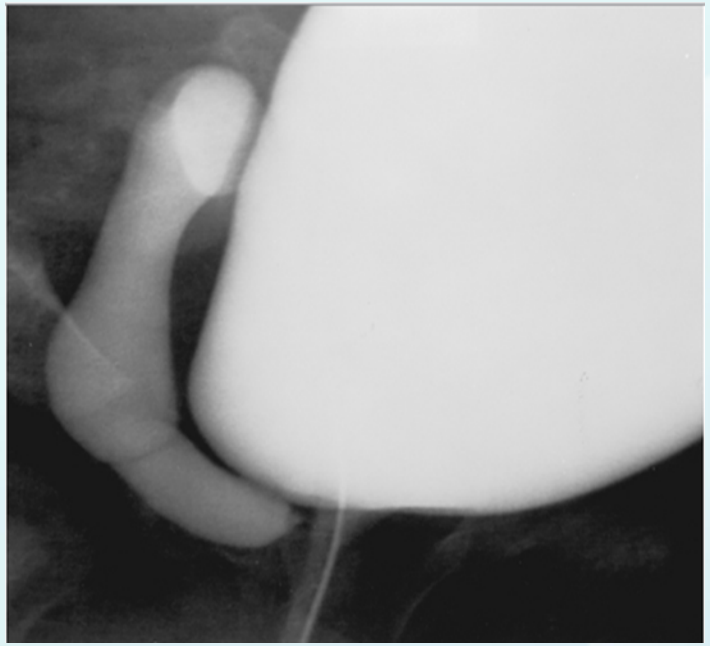
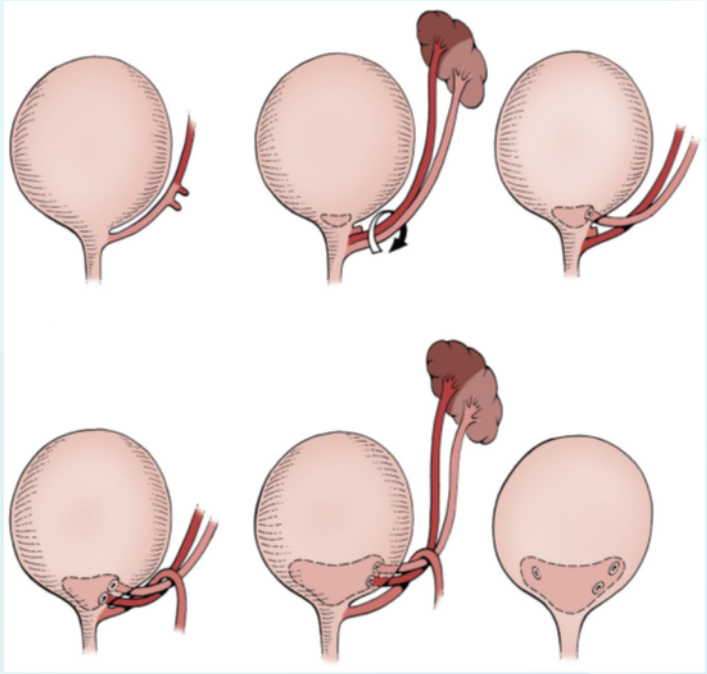


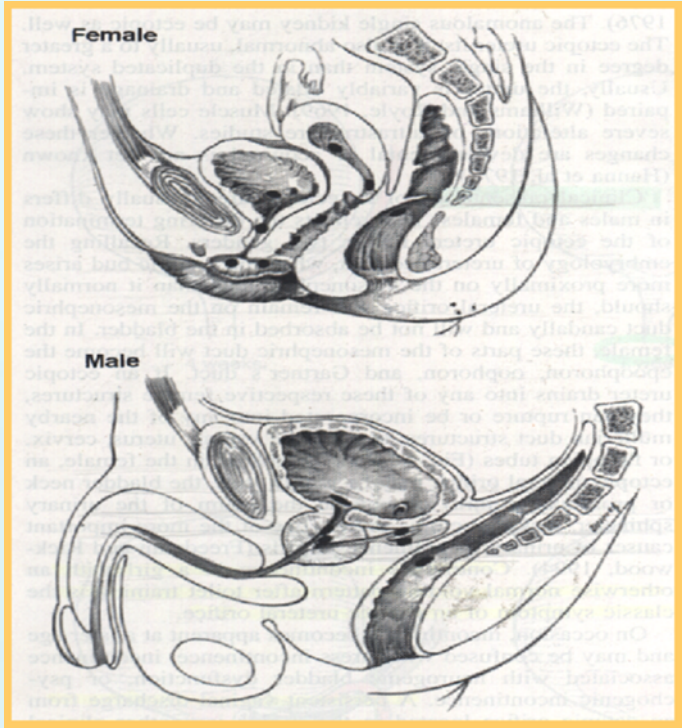
Ectopic Ureter

- An ectopic ureter is any ureter, single or duplex, that doesn't enter the trigonal area of the bladder.
- 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.



Ectopic Ureter

- In females the ectopic ureter may enter anywhere from the bladder neck to the perineum and into the vagina, uterus,and even rectum.
- One of the classic symptoms is **continuous wetting.**
- In males the ectopic ureter always enters the urogenital system above the external sphincter or pelvic floor, and usually into the wolffian structures including vas deferens, seminal vesicles, or ejaculatory duct.



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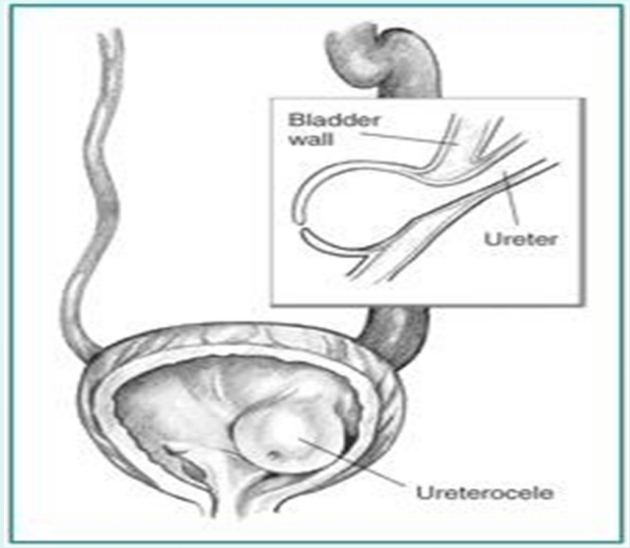
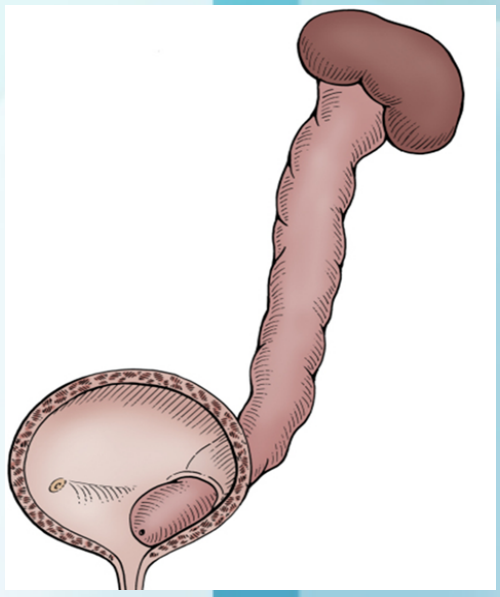
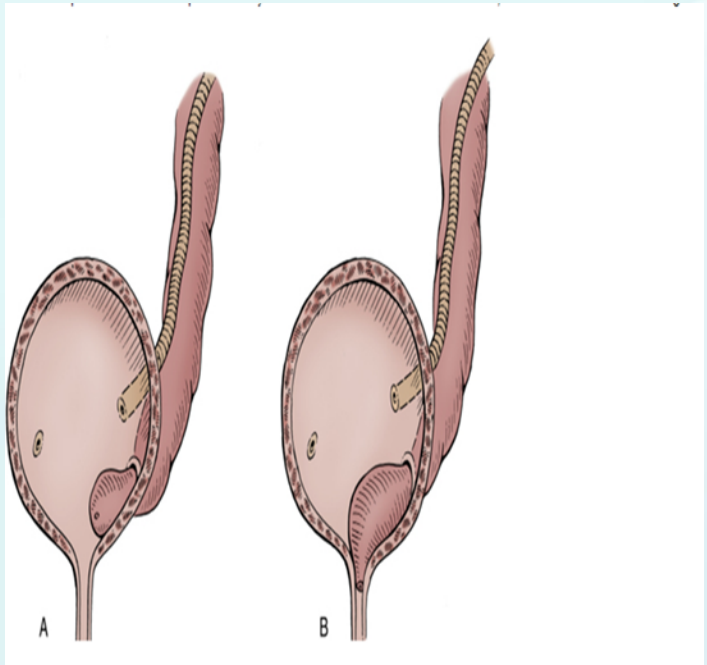
Ureterocele

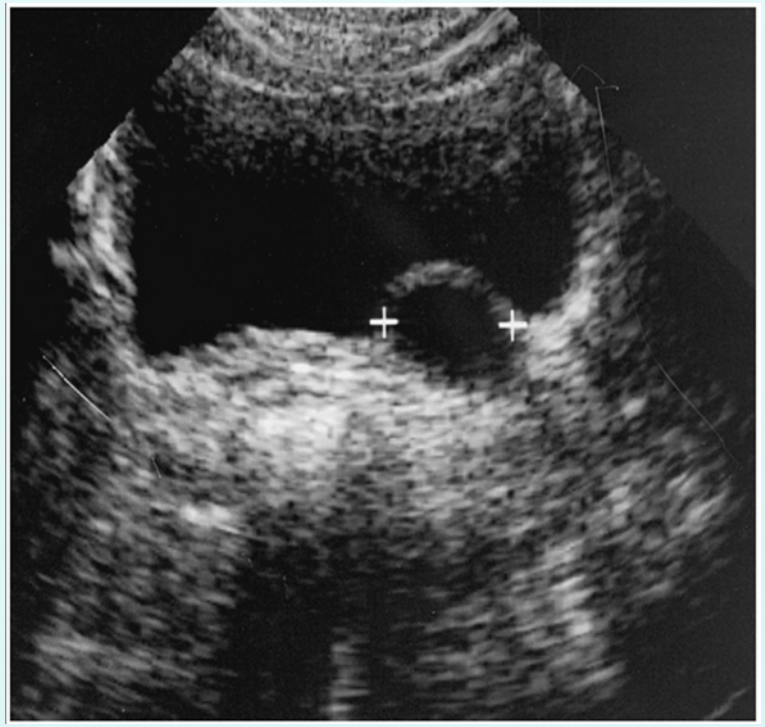
- A cystic dilation of the distal aspect of the ureter
- Located either within the bladder or spanning the bladder neck and urethra.
- **Presentation:**
  - Antenatal (U/S)
  - Urine retention
  - Infection
  - Calculus formation

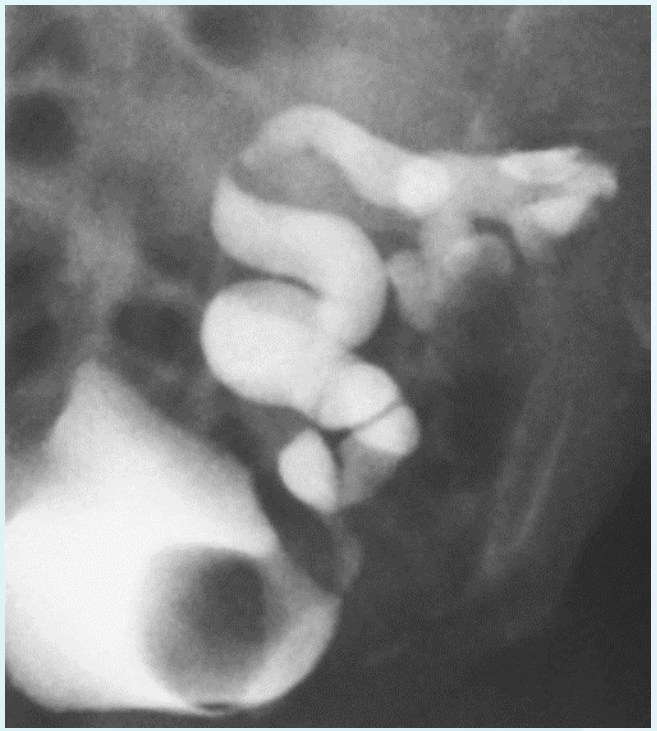




- **Types**:
  - ✓ Intravesical: Orthotopic, simple, adult type.
  - ✓ Extravesical: Ectopic, duplex system, infant type.







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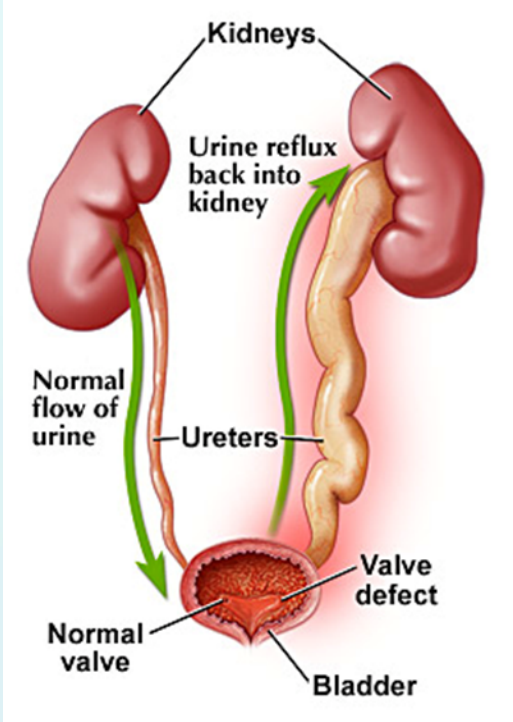
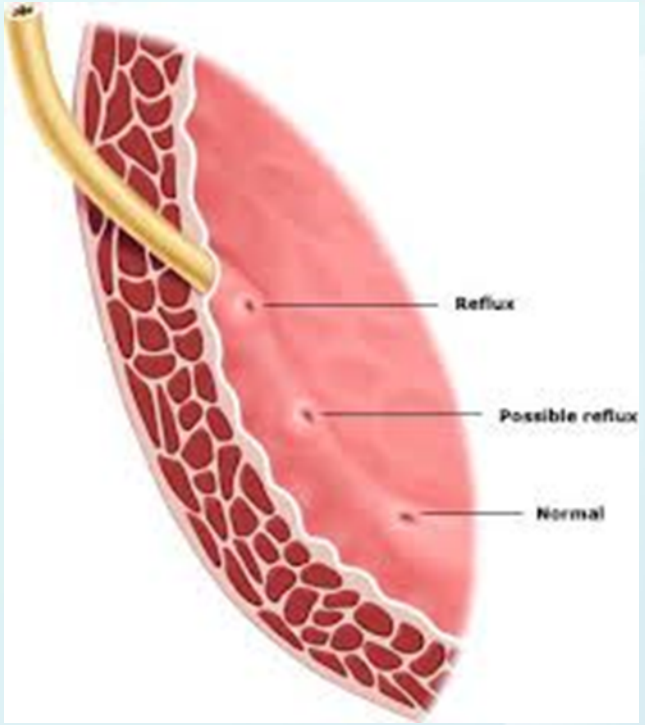


VUR

Normal anti-reflux mechanism
"Flap valve"

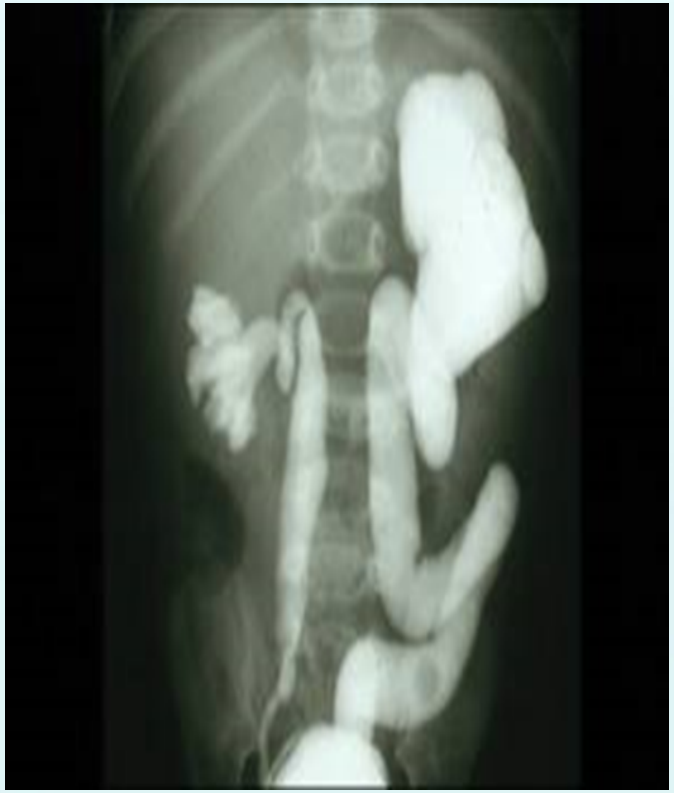
1. Oblique course as it enters the bladder.
2. Proper muscular attachments to provide fixation.
3. Posterior support to enable its occlusion.
4. Adequate submucosal length.

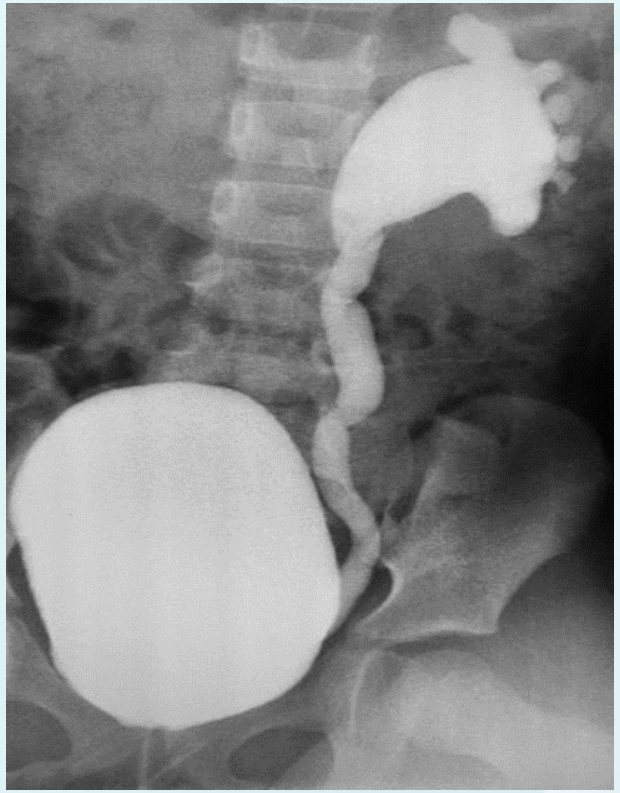


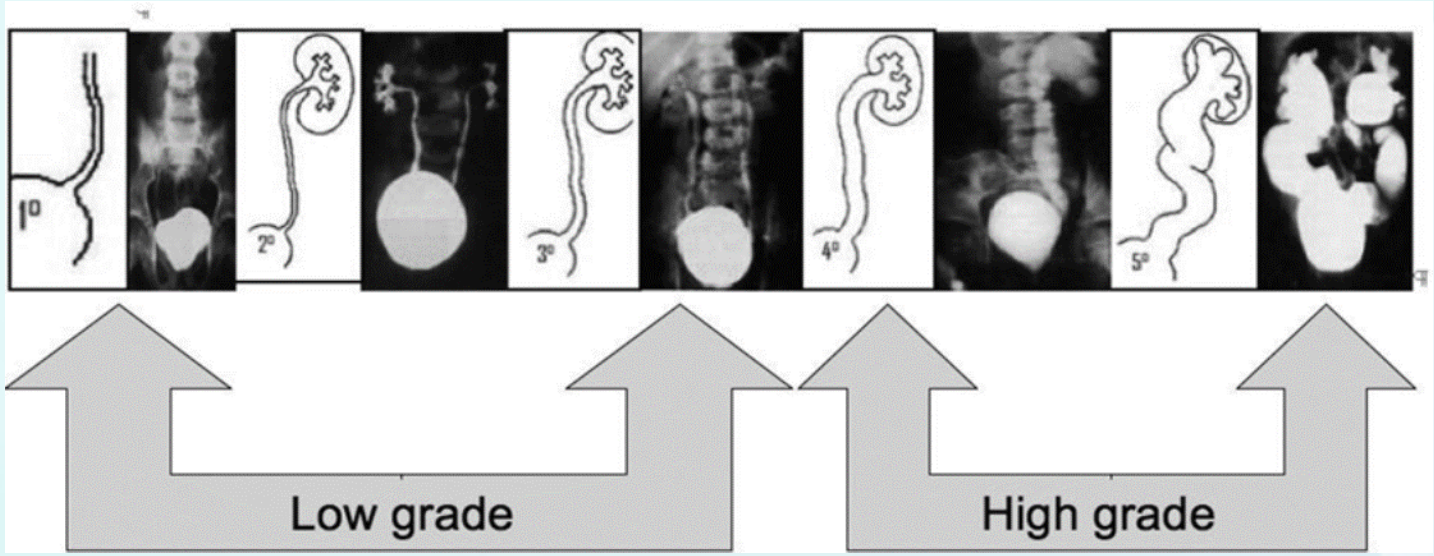
VUR

- Presentation
  - Asymptomatic
    - Prenatal
    - Fluctuated dilatation
  - Febrile UTIs

MCUG

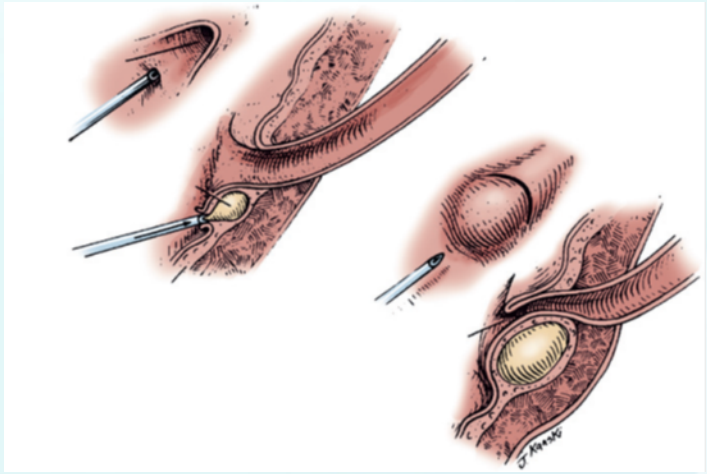


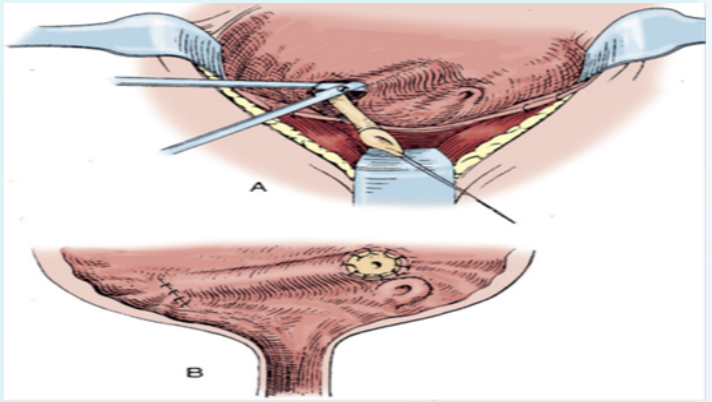



1°
2°
3°
4°
5°
Low grade
High grade

VUR: Management

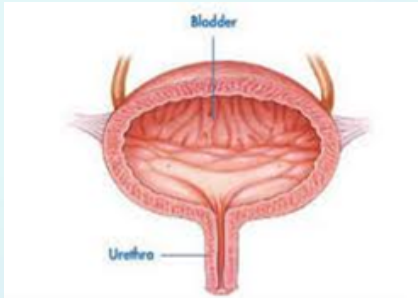
- Prophylactic antibiotic
- Surgical treatment
  - Endoscopic treatment
  - Ureteral reimplantation





Bladder Anomalies

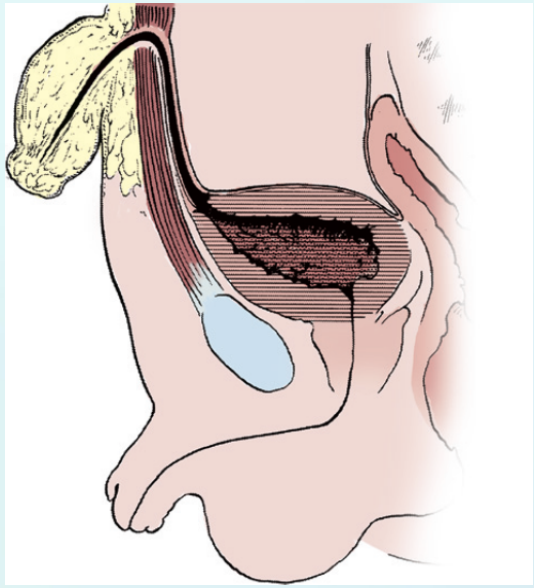
- Urachal abnormalities
- Bladder diverticulum
- Bladder duplication
- Bladder exstrophy



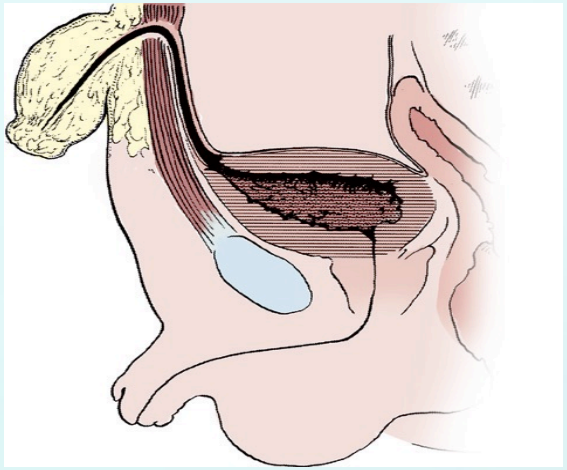
Bladder Anomalies

Urachal abnormalities

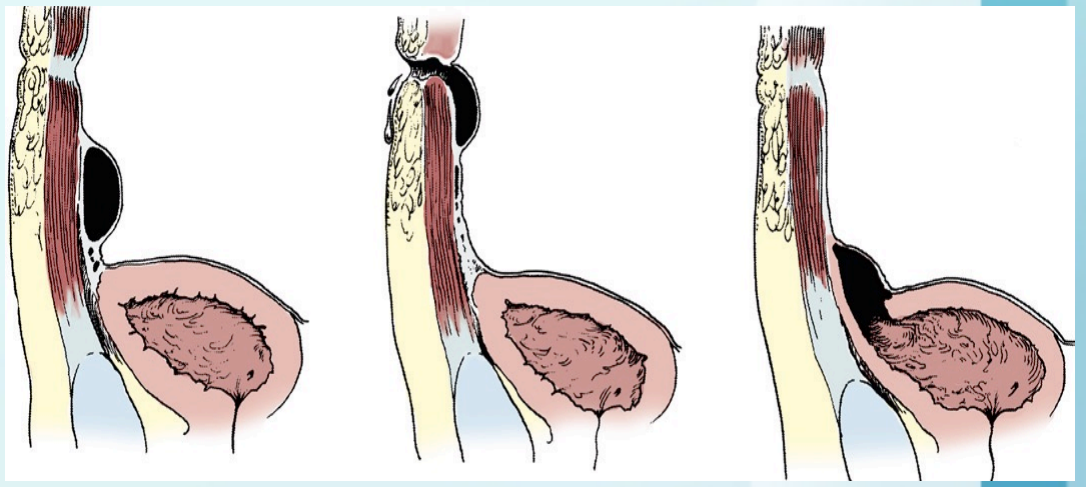
- Urachal anomalies are usually detected postnatally due to umbilical drainage.
- Imaging possibilities include ultrasound, CT, and VCUG



Urachal abnormalities



Patent urachus



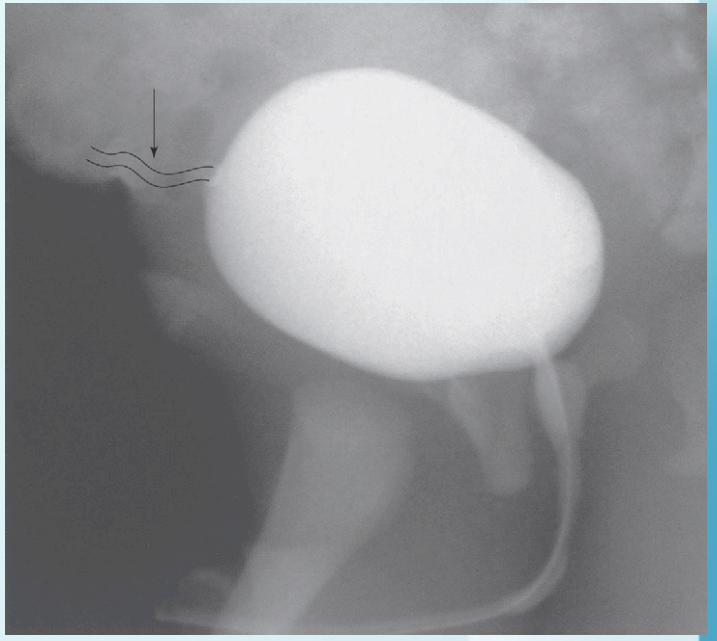
Urachal cyst

Umbilical-urachus sinus

Vesicourachal diverticulum

Urachal abnormalities

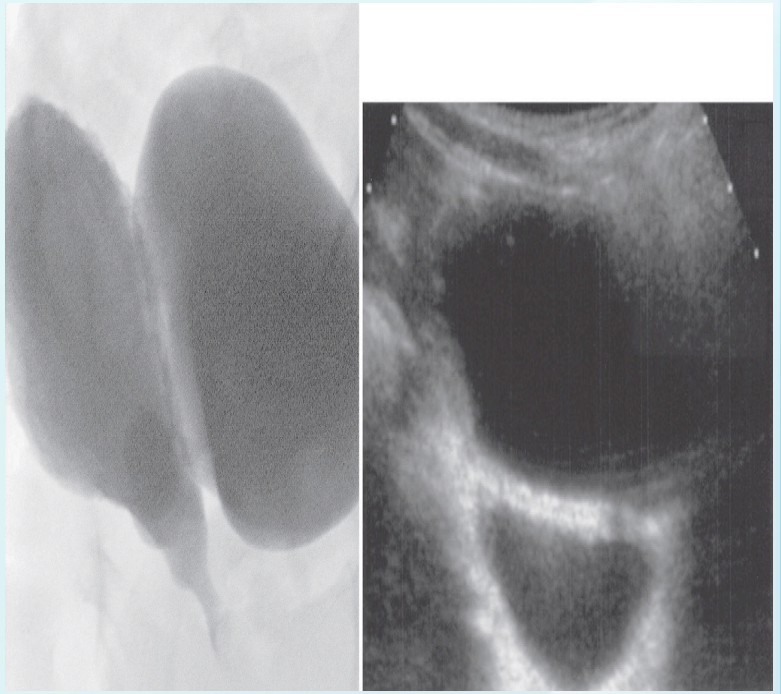
- Conservative treatment with observation is justified in asymptomatic cases due to possible spontaneous resolution
- Infected urachal remnants are initially treated with drainage and antibiotics, followed by surgical excision.
- Nonresolved urachal remnants should be excised due to theincreased risk of later adenocarcinoma formation



Bladder Anomalies

Bladder Diverticulum

- Bladder diverticula can be detected on prenatal ultrasound, but the gold standard remains VCUG, which will reveal possible accompanying VUR.

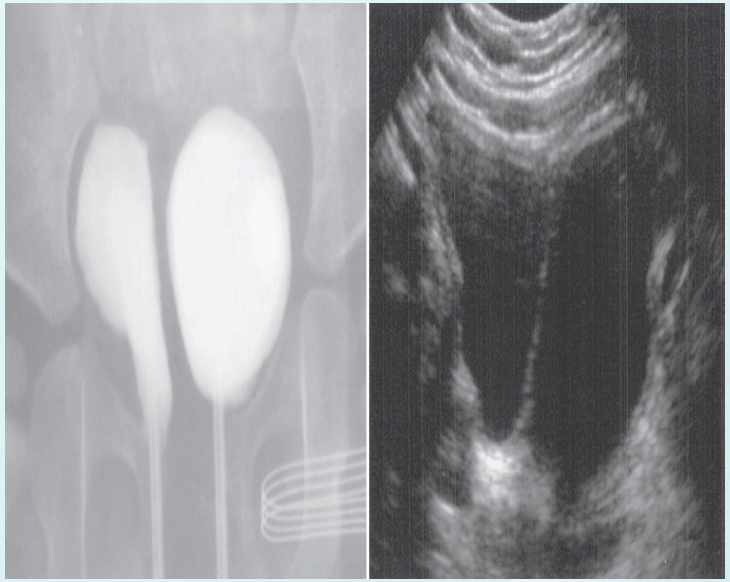


Bladder Diverticulum

- Primary diverticula
  - arise as a localized herniation of bladder mucosa at the ureteral hiatus and are most likely caused by a congenitally deficient bladder wall.
- Secondary para-ureteral diverticula
  - are acquired and develop due to existing infra- vesical obstruction.
- Symptomatic diverticula, especially in conjunction with VUR, should be treated surgically.

Bladder Duplication

- Bladder duplication is often associated with duplication anomalies of the external genitalia and lower gastrointestinal tract.
- Initial treatment is directed toward
  - renal preservation.
  - prevention of infections.

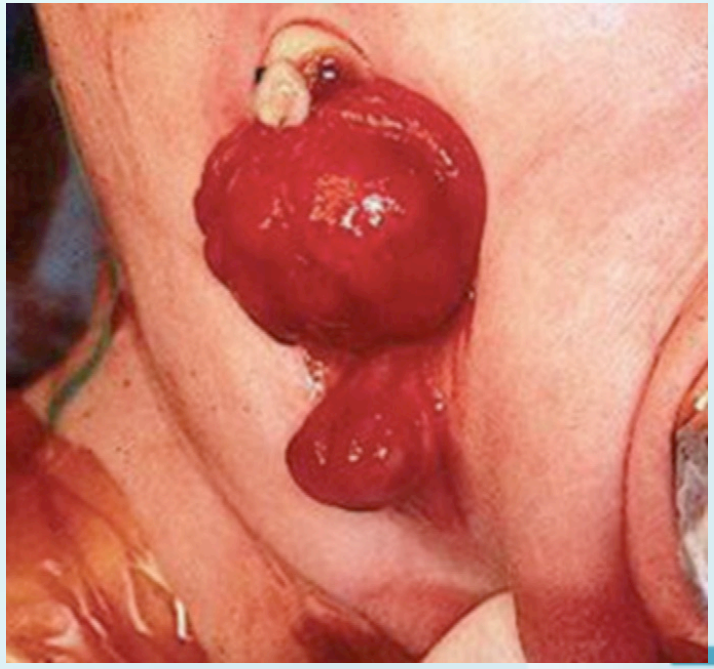


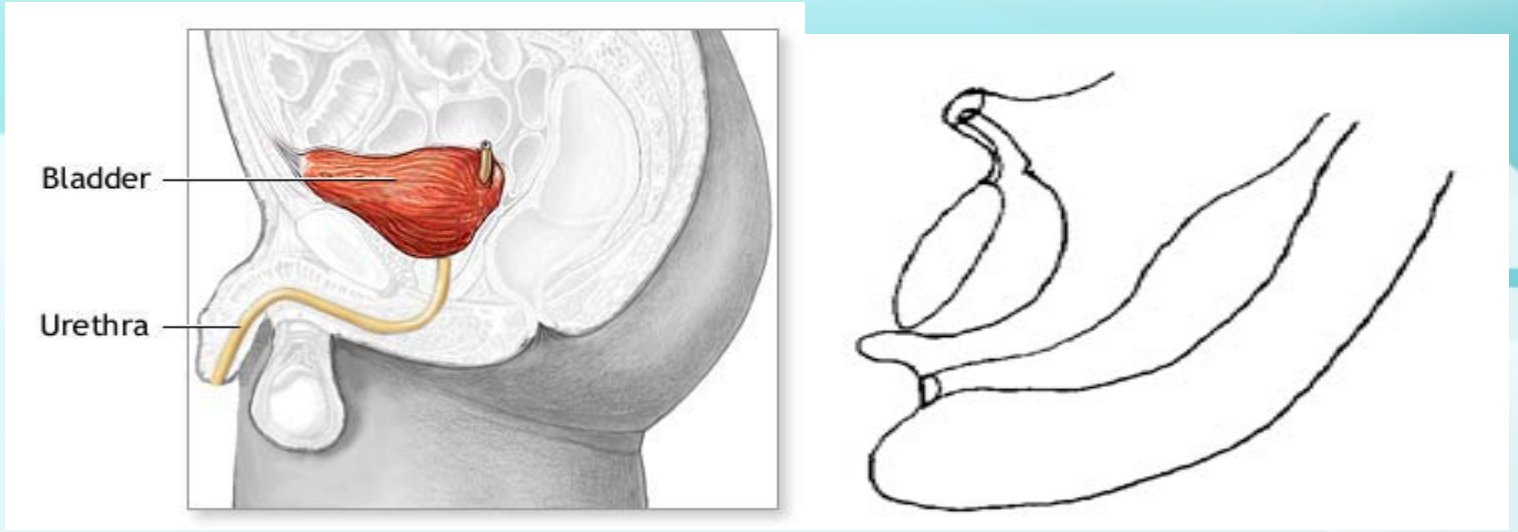
Bladder Duplication

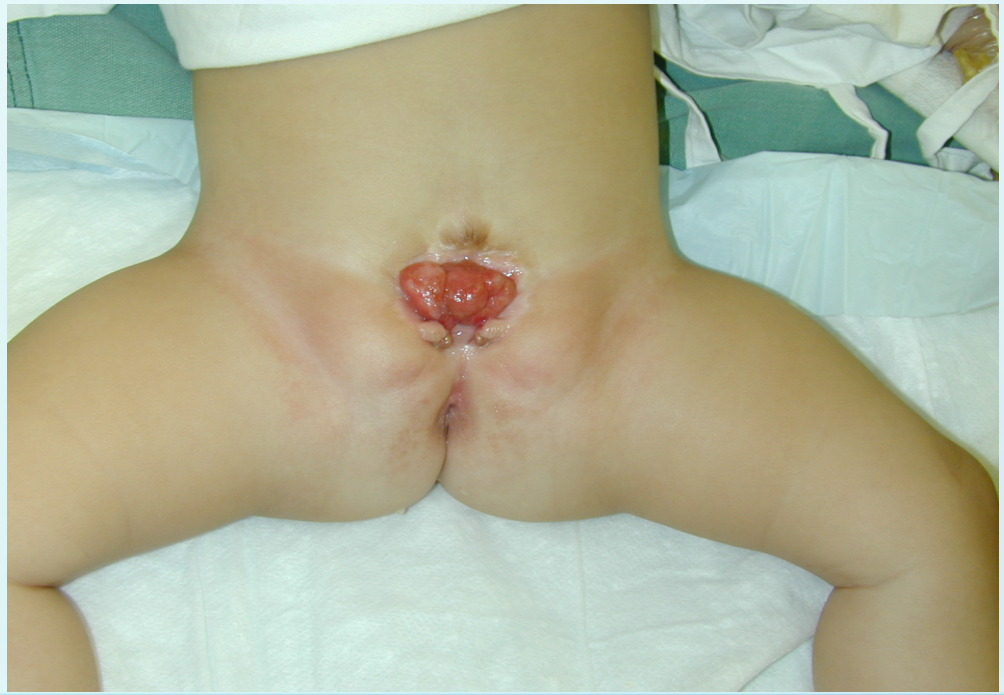
- Long-term goals include achieving continence and reconstructing the internal and external genitalia.
- Due to the rarity of the disease and the large variety of presentations, the surgeries must be individualized

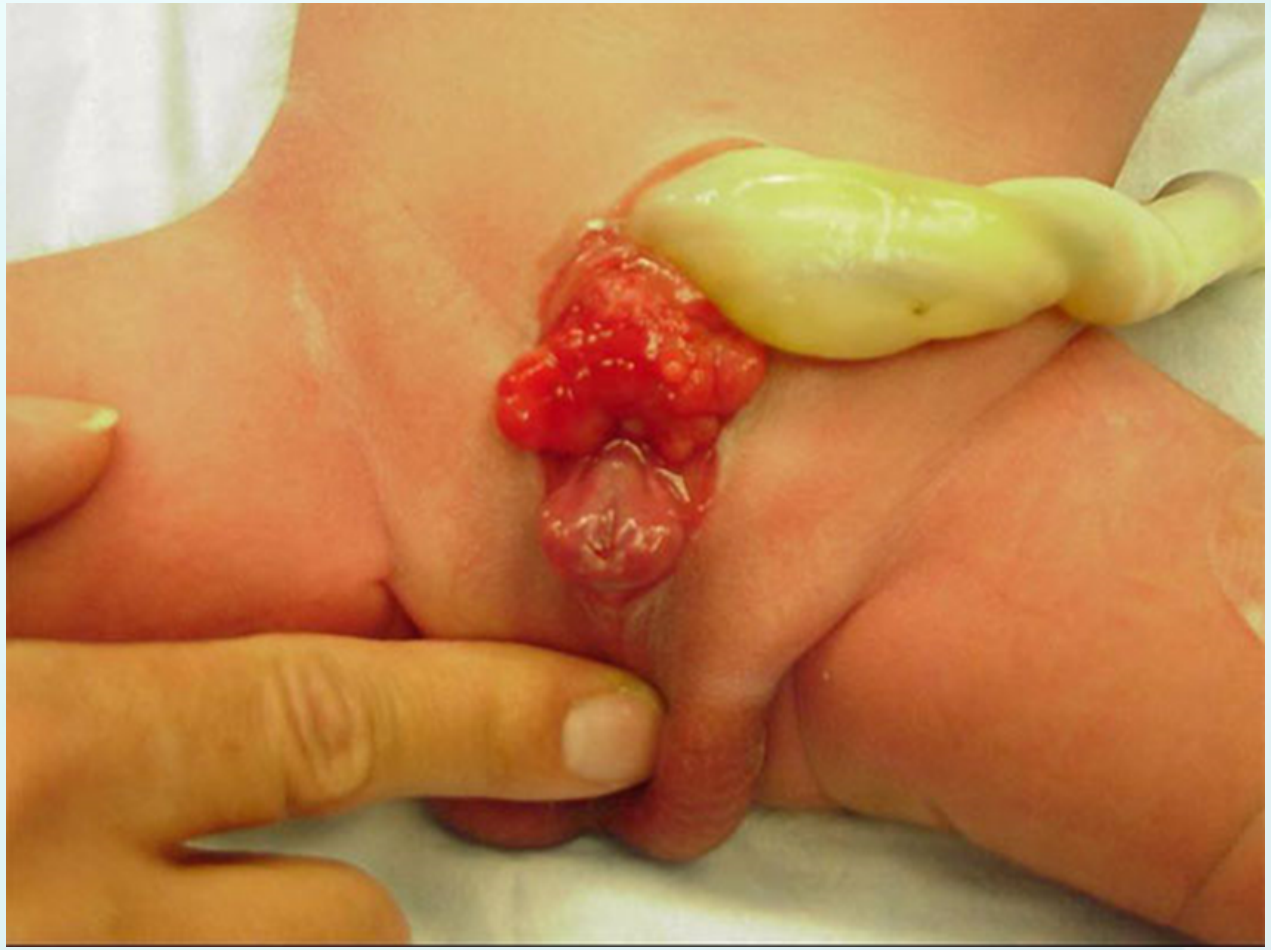
Classic Bladder Exstrophy

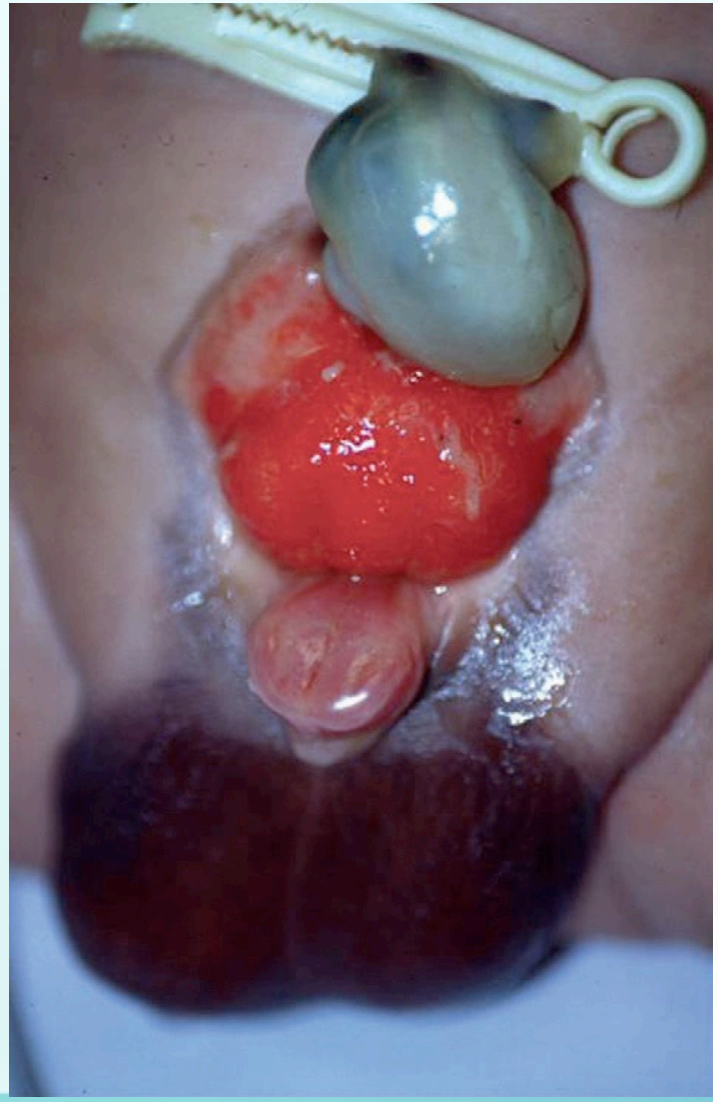
- The incidence of bladder exstrophy has been estimated as between 1 in 10,000 and 1 in 50,000.

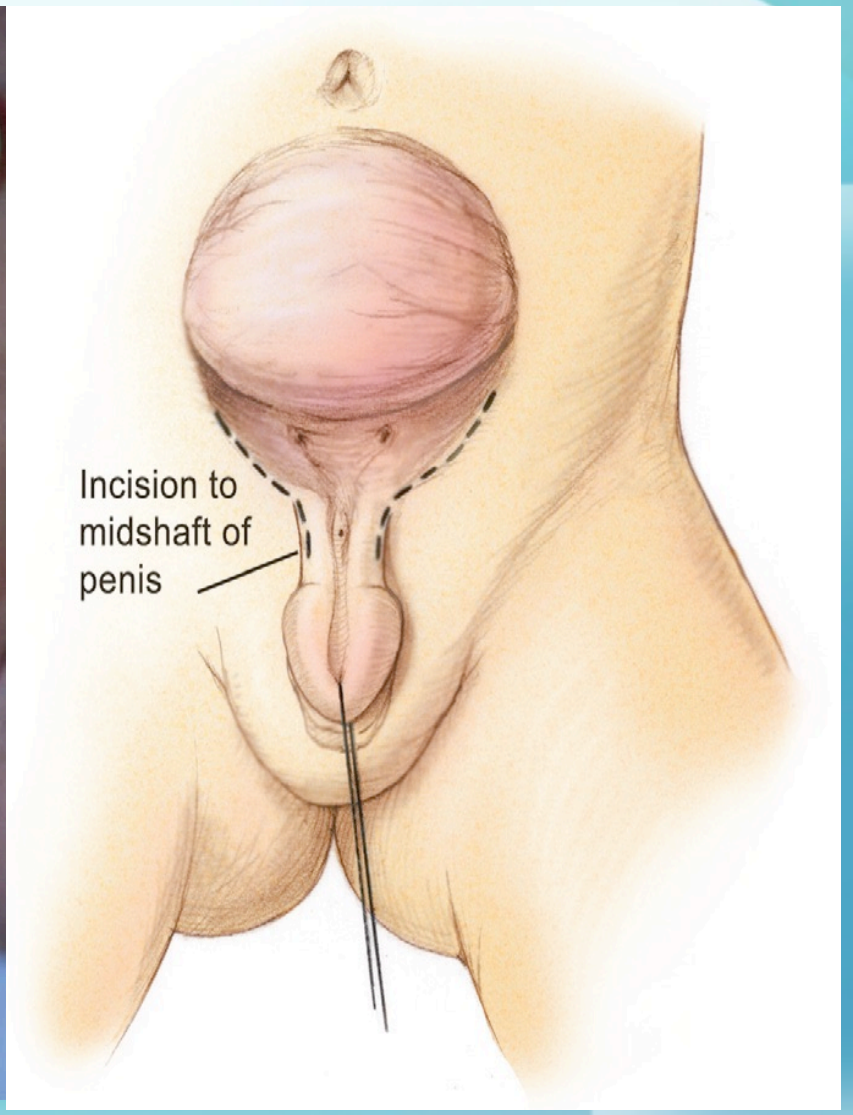



Bladder
Urethra








Incision to
midshaft of
penis

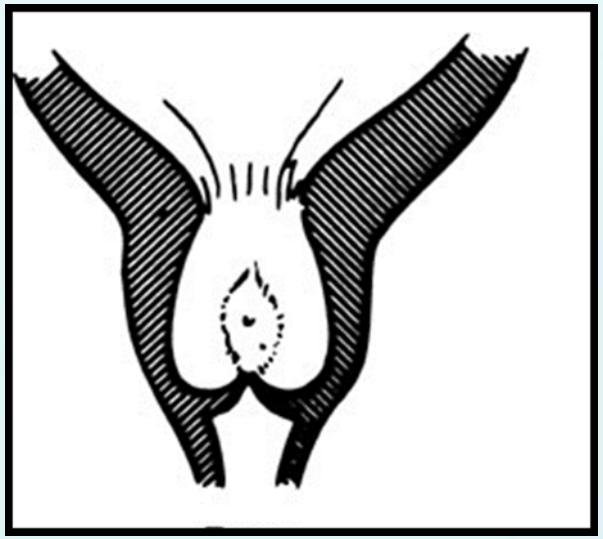
Urethral anomalies

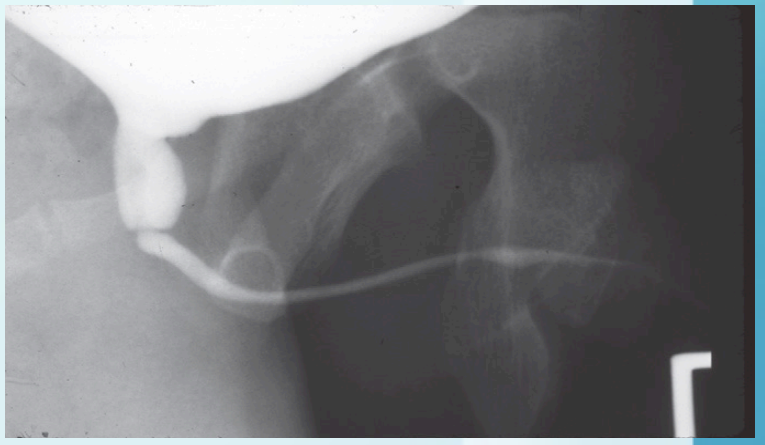
- Posterior Urethral Valves
- Anterior Urethral Valves
- Urethral Duplication
- Congenital Urethral Stricture
- Urethral Polyps

Urethral anomalies

- **Posterior Urethral Valves**
- Anterior Urethral Valves
- Urethral Duplication
- Congenital Urethral Stricture
- Urethral Polyps

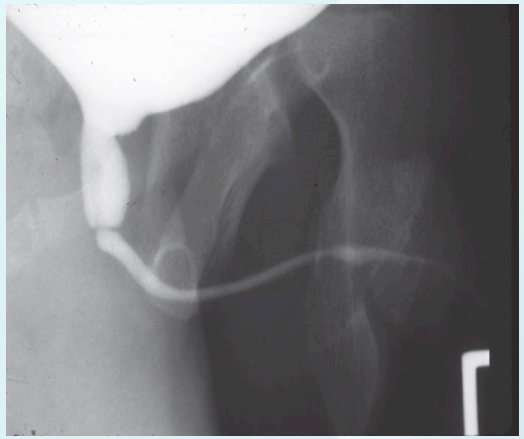
Posterior Urethral Valves (PUV)





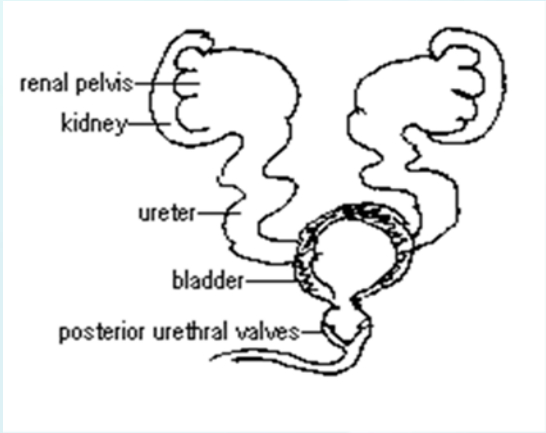
PUV

- 1 in 8000 to 25,000 live births.
- Make up 10% of urinary obstructions diagnosed in utero.
- Most common cause of urine retention in male infants.
- 50% have renal impairment.



PUV

- The bladder and the kidneys developed under high pressure and resistance.

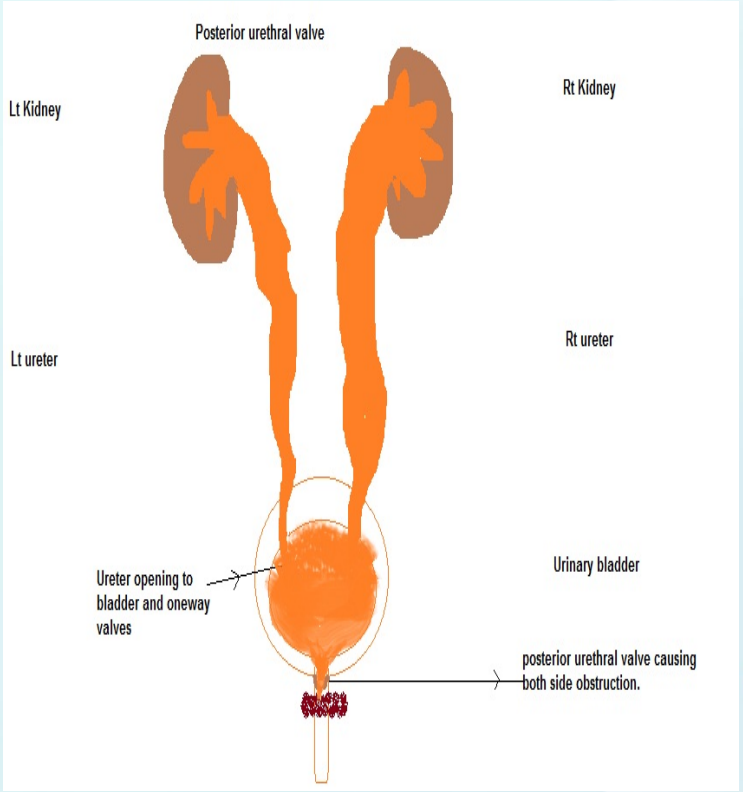


PUV

- Presentation:
  1. Antenatal
  2. Urine retention
  3. UTI
  4. Poor urinary stream
  5. Urinary incontinence
  6. CRF (ESRD)

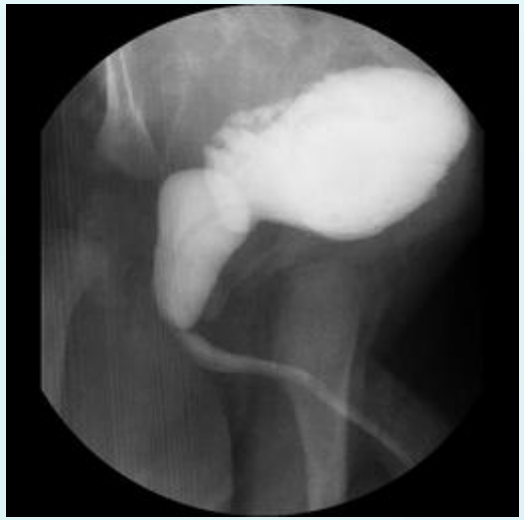
PUV

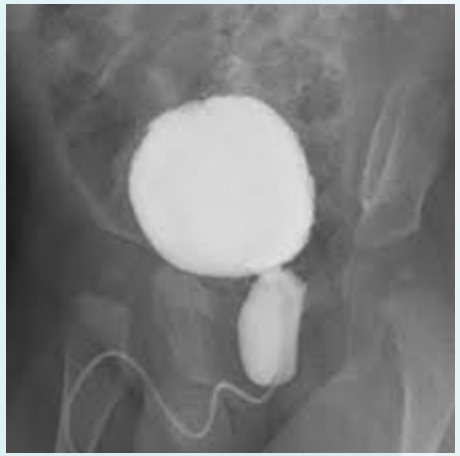
- **Associated findings:**
  1. Oligohydramnios
  2. Bilateral renal dilatation
  3. VUR: 40%
  4. Valve bladder
  5. Renal impairment



PUV…

MCUG





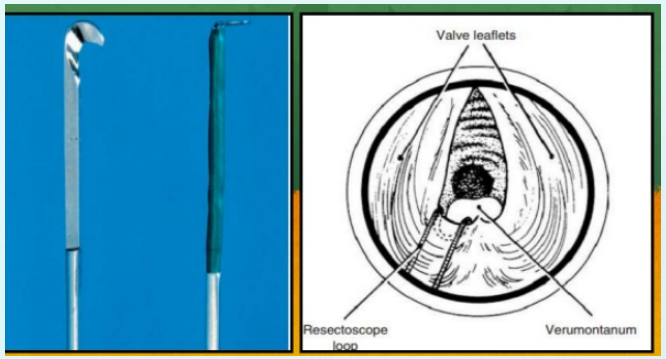
PUV…

Management

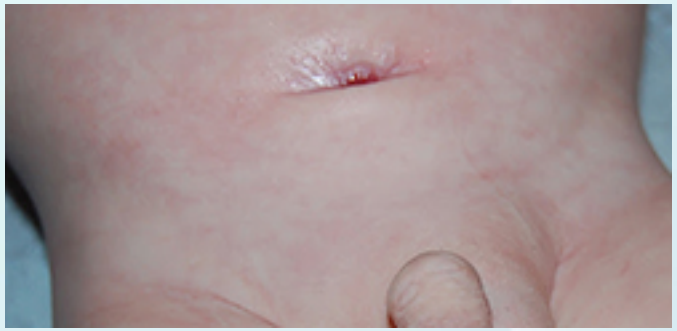
- Initial treatment
  - Feeding tube insertion
  - Start antibiotic prophylactic
  - Ultrasound
  - MCUG

PUV…
Surgical treatment

Endoscopic valve ablation



Cutaneous vesicostomy



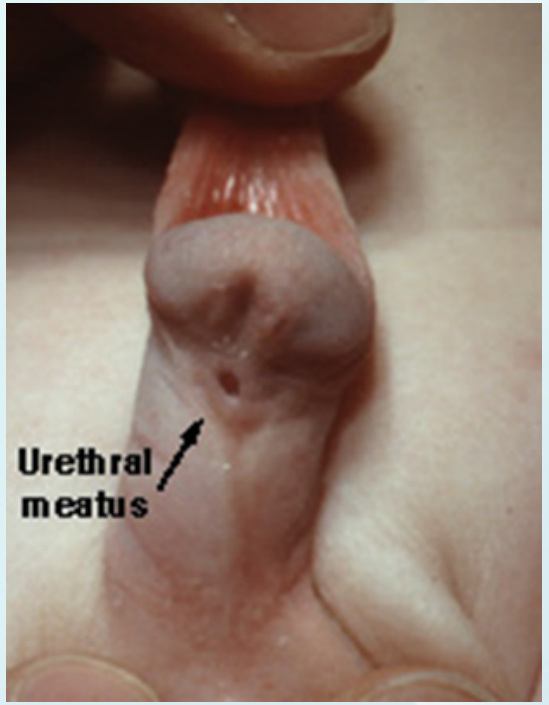
Congenital Anomalies of the external Genitalia

Congenital Genital disordered

- Hypospadias
- Epispadias
- Micropenis
- Bladder Exstrophy
- Cloacal Exstrophy

Hypospadias

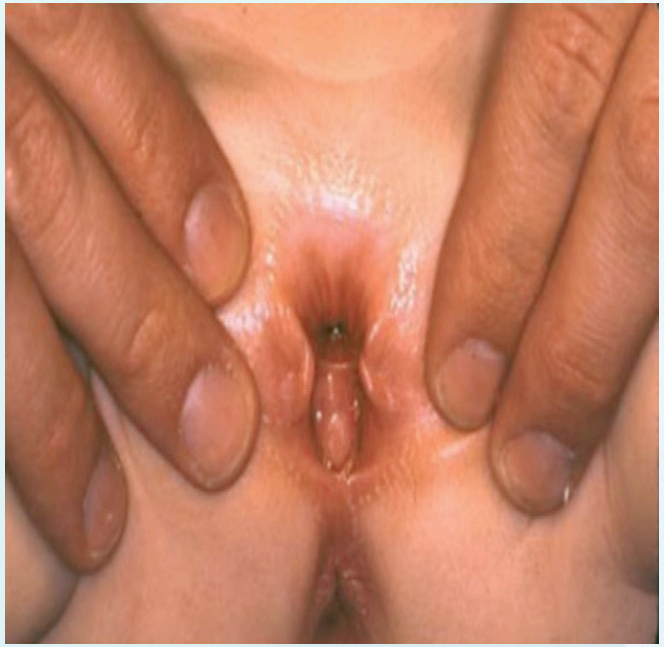
- Abnormal position of the EUM on the ventral surface.
- Types:
  - ✓ Distal hypospadias.
  - ✓ Proximal hypospadias.
- NO Circumcision
- 6 to 9 months repair.



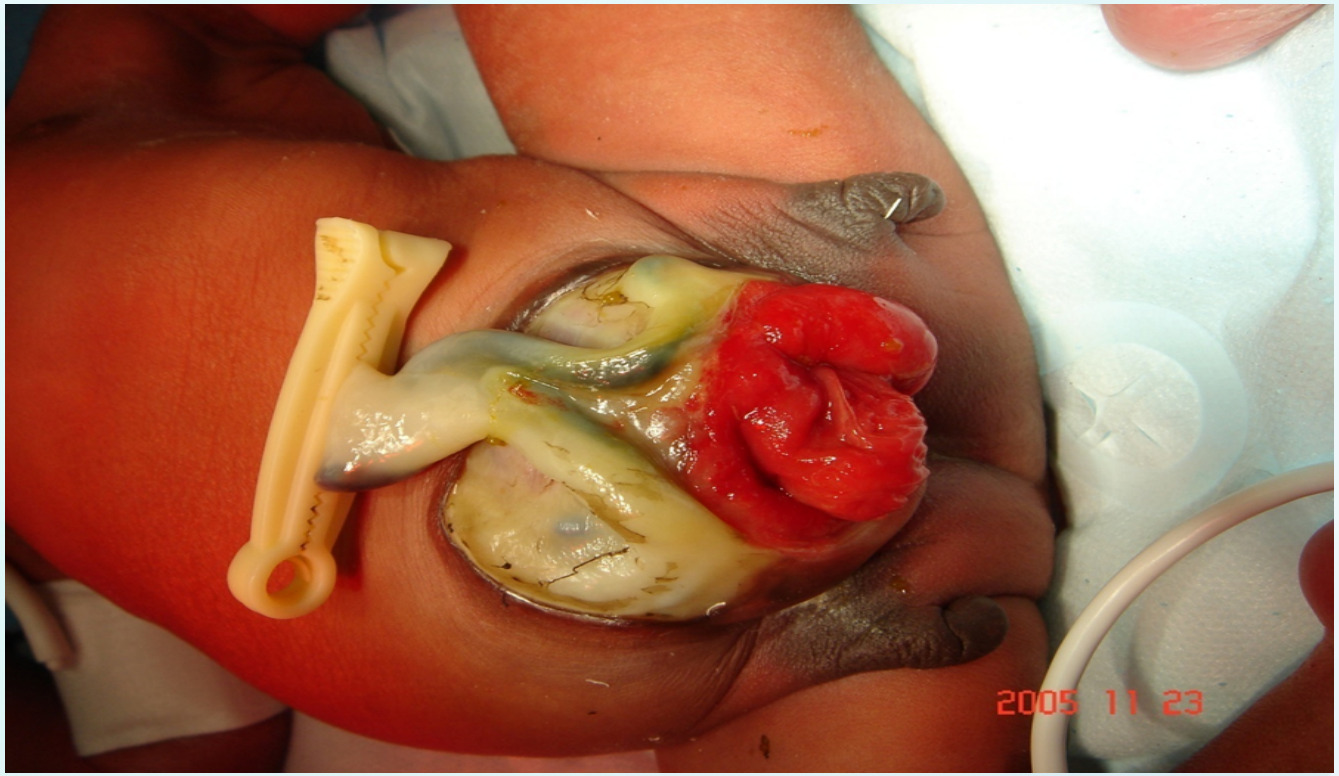
Epispadias

Male

Female

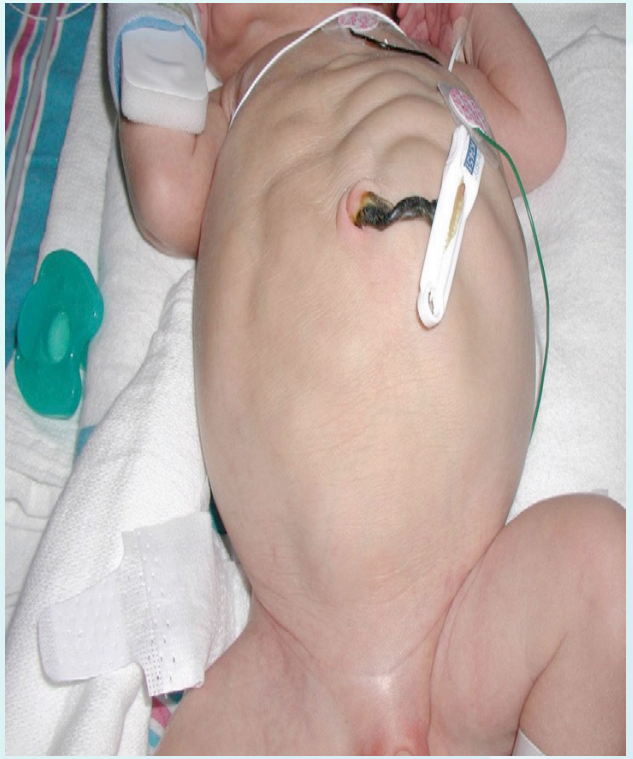


Cloacal Exstrophy



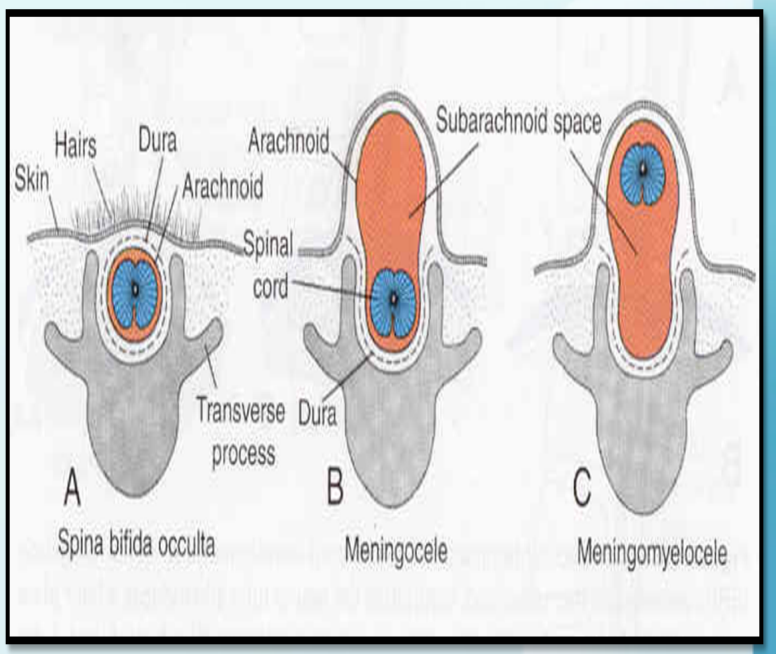
Prune-Belly Syndrome

- The incidence :1 in29,000 to 1 in 40,000 live births
- The three major findings are
  - deficiency of the abdominal musculature,
  - bilateral intra-abdominal testes,
  - anomalous urinary tract
- Other names
  - Triad syndrome
  - Eagle-Barrett syndrome
  - abdominal musculation syndrome



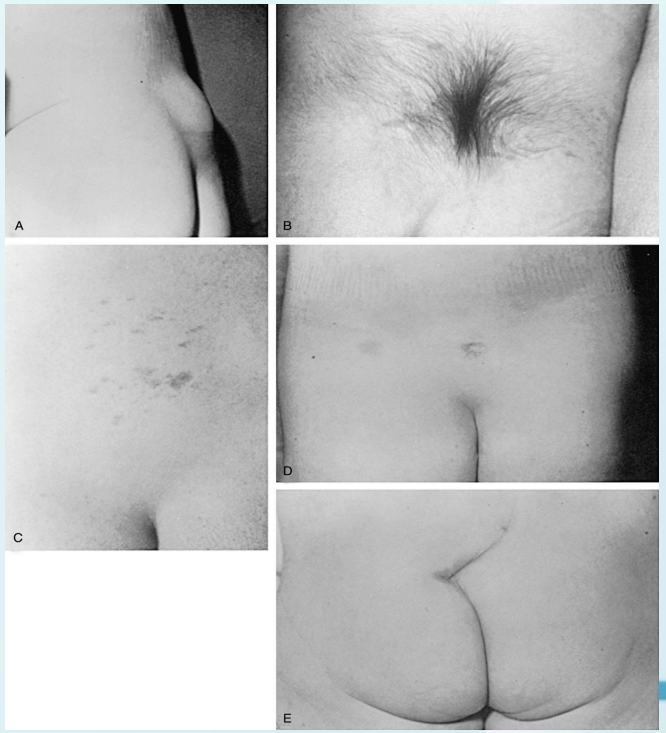
NEUROSPINAL DYSRAPHISMS

- The most common cause of neurogenic bladder dysfunction in children is abnormal development of the spinal canal and internecine spinal cord.



NEUROSPINAL DYSRAPHISMS

- Cutaneous lesions occur in 90% of children with various occult dysraphicstates.
- These lesions vary from
  - small lipomeningocele
  - hair patch
  - dermal vascular malformation
  - sacral dimple
  - abnormal gluteal cleft.



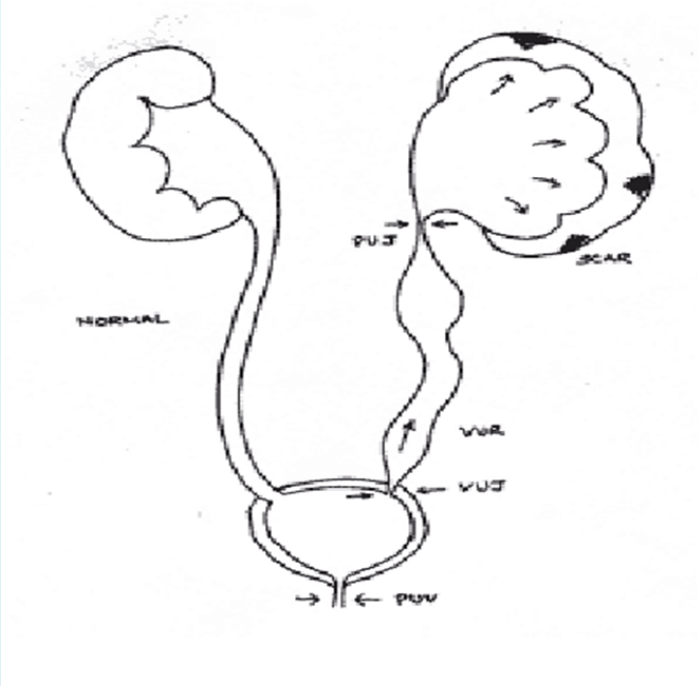
THANK YOU!

Antenatal Hydronephrosis(ANH)

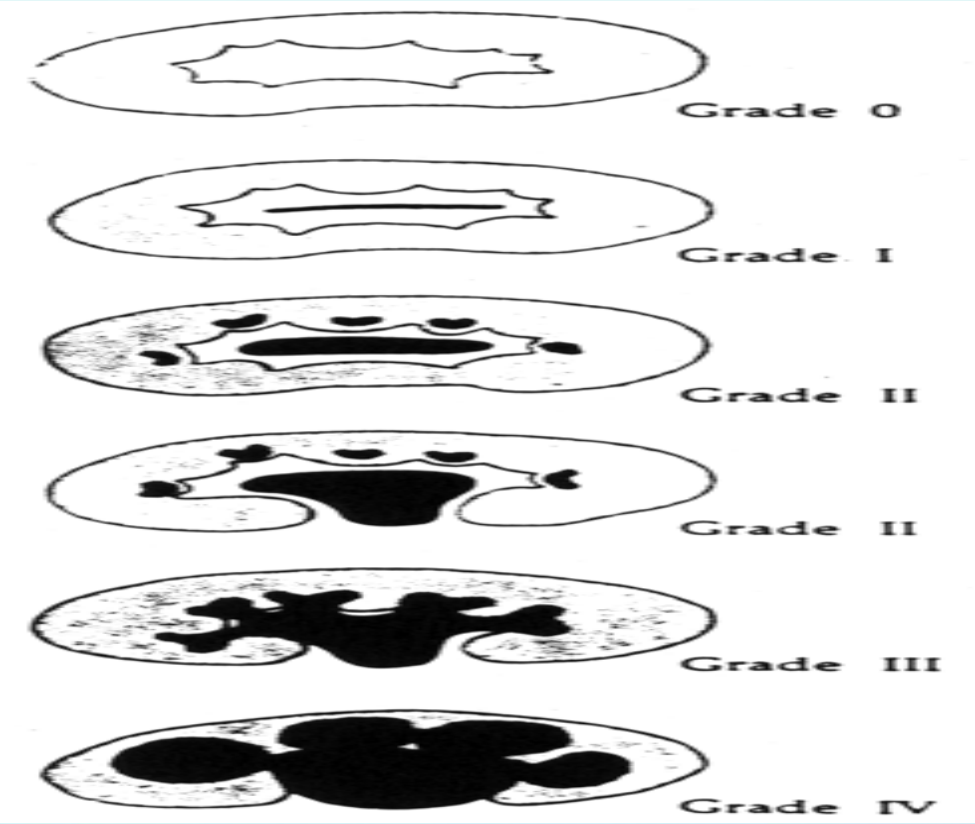
Antenatal Hydronephrosis(ANH)

Causes:

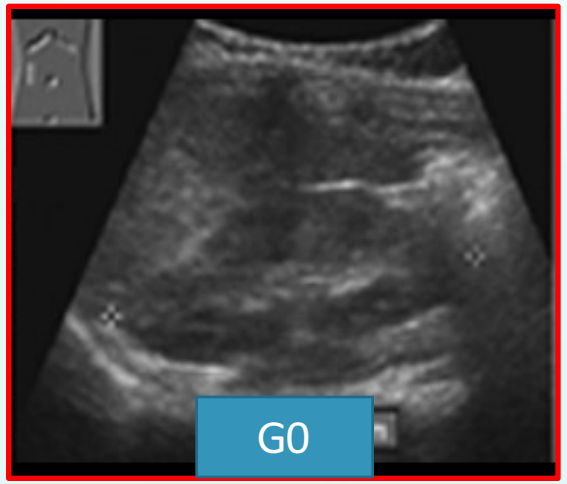
- Pelviureteric junction obstruction (41%)
- Ureterovesical junction obstruction (23%)
- Vesicoureteric reflux(7%)
- Duplication anomalies (13%)
- Posterior urethral valves (10 %)
- MCDK
- Others (6%)

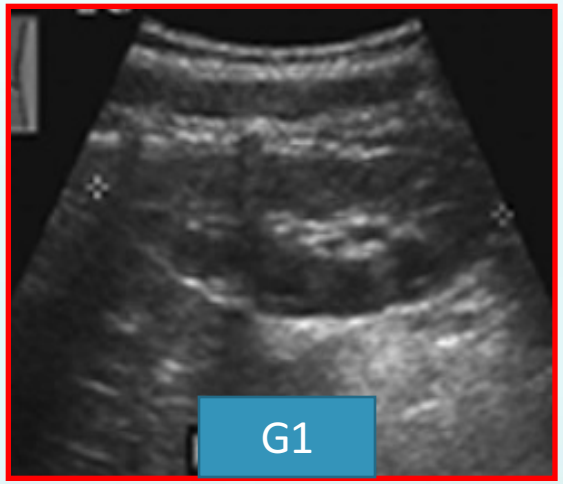


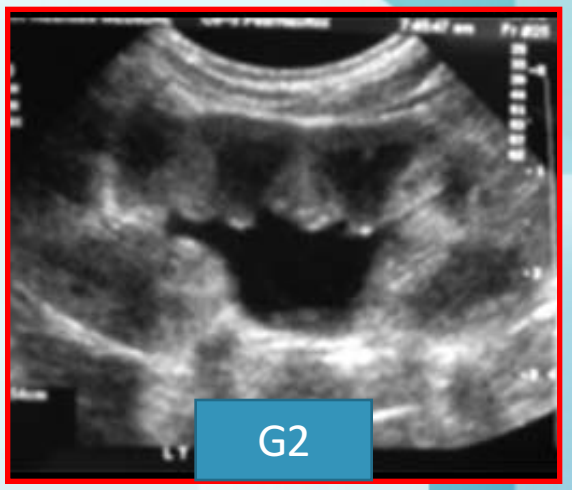
ANH

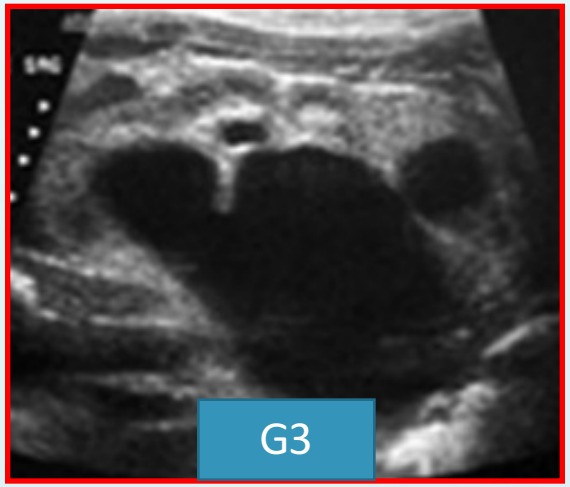


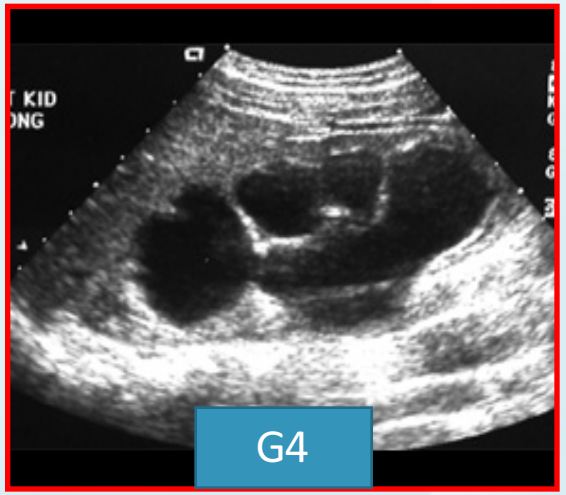
SFU Grading



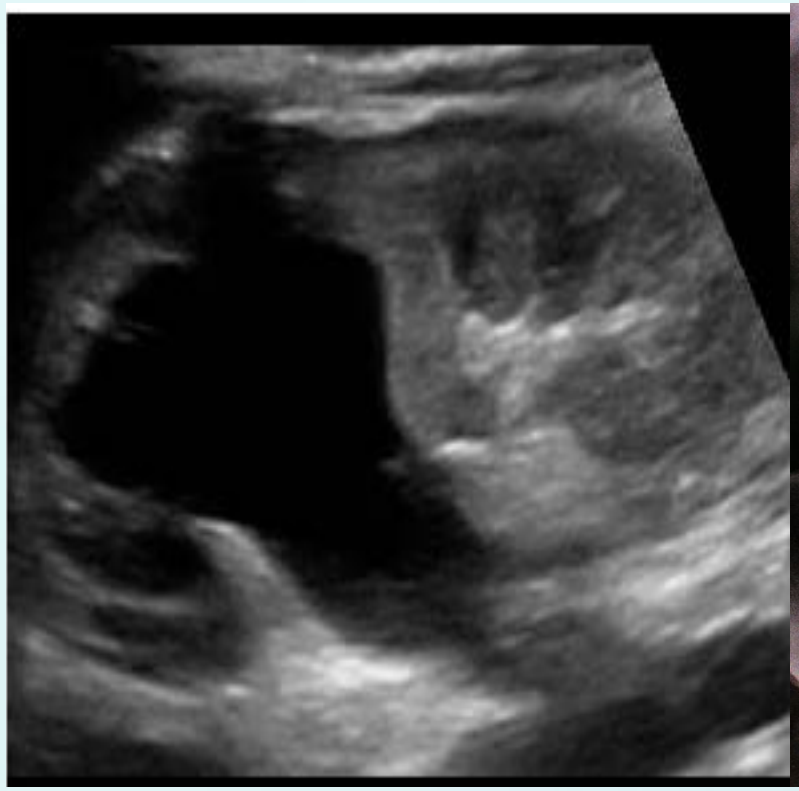


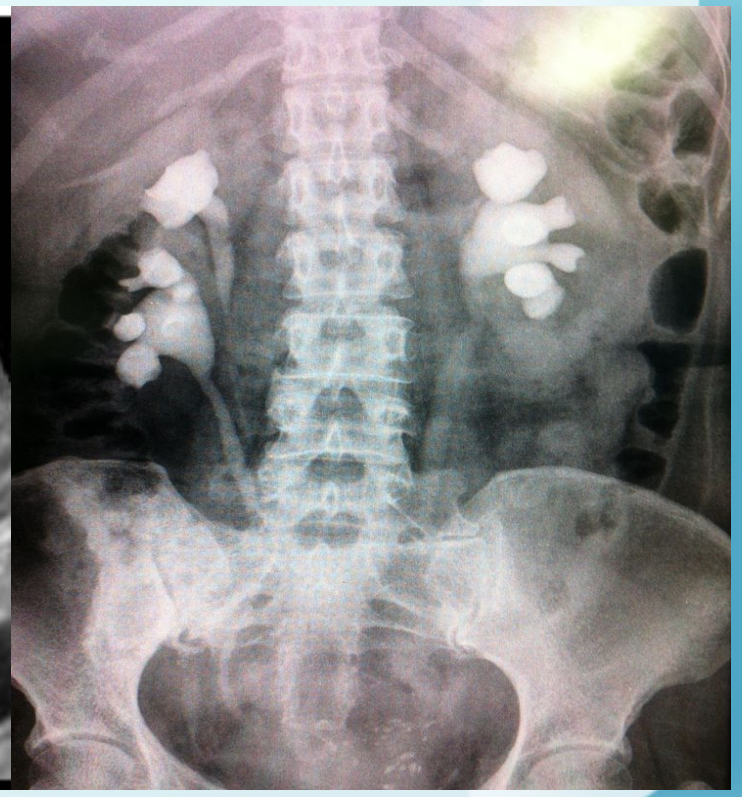


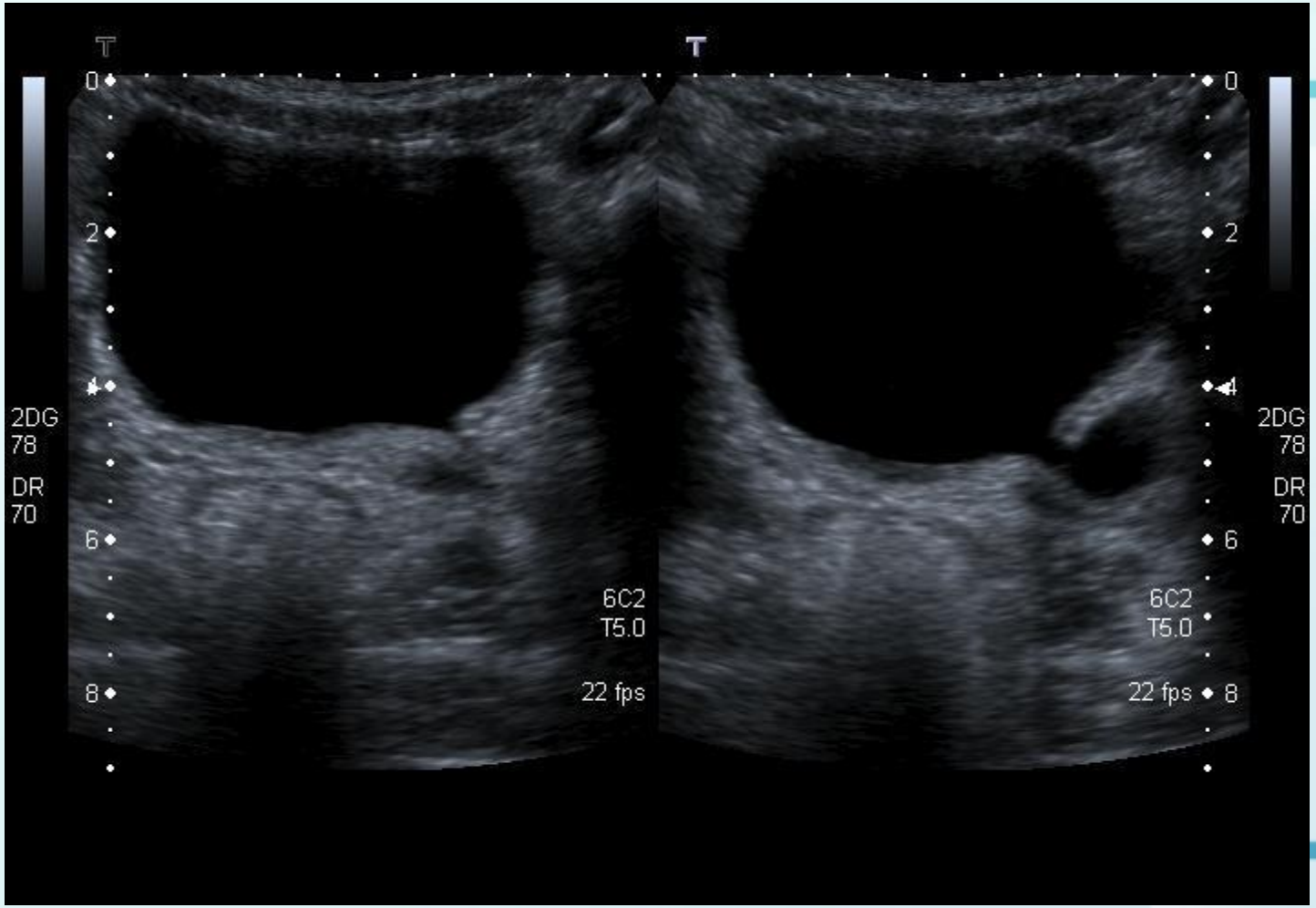




Weigert- Meyer Rule



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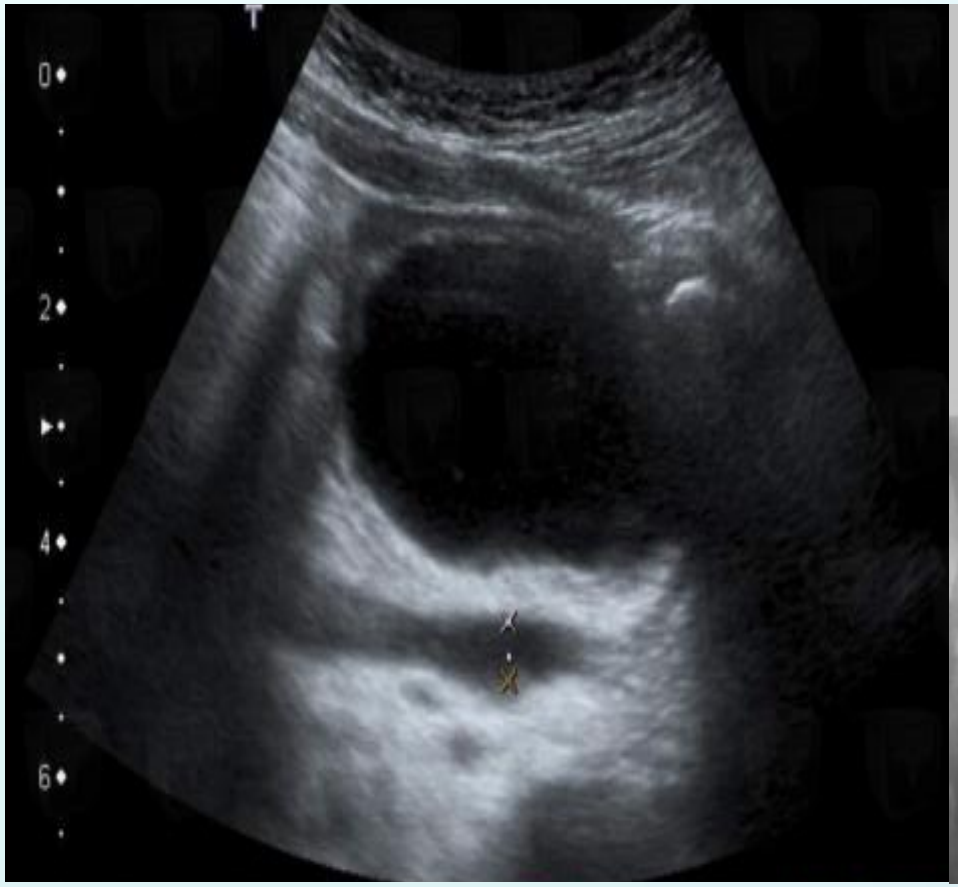
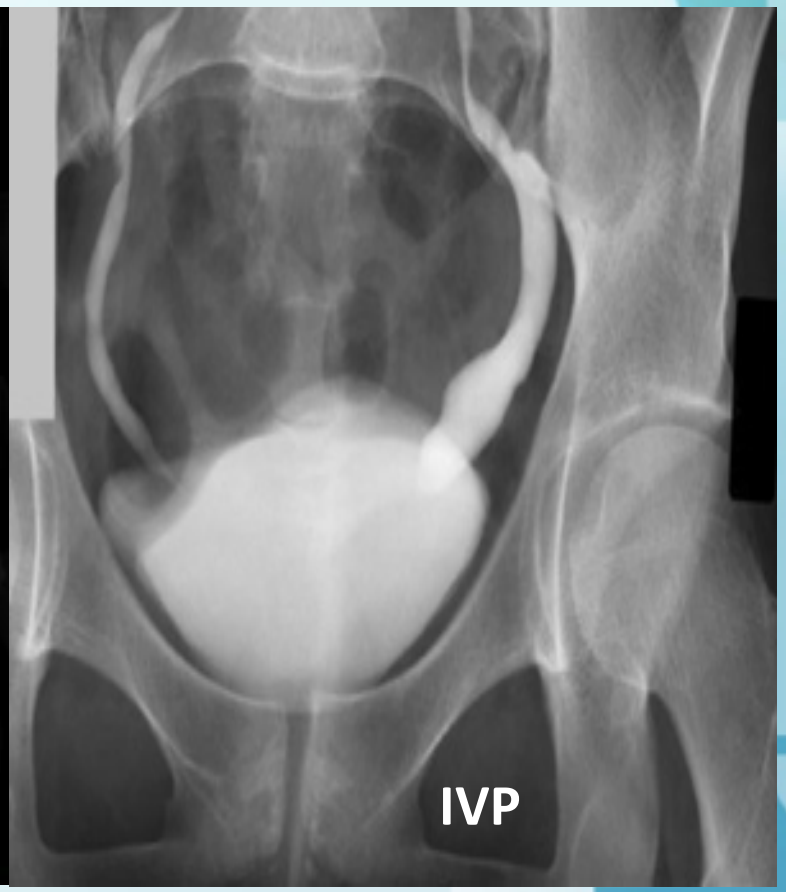
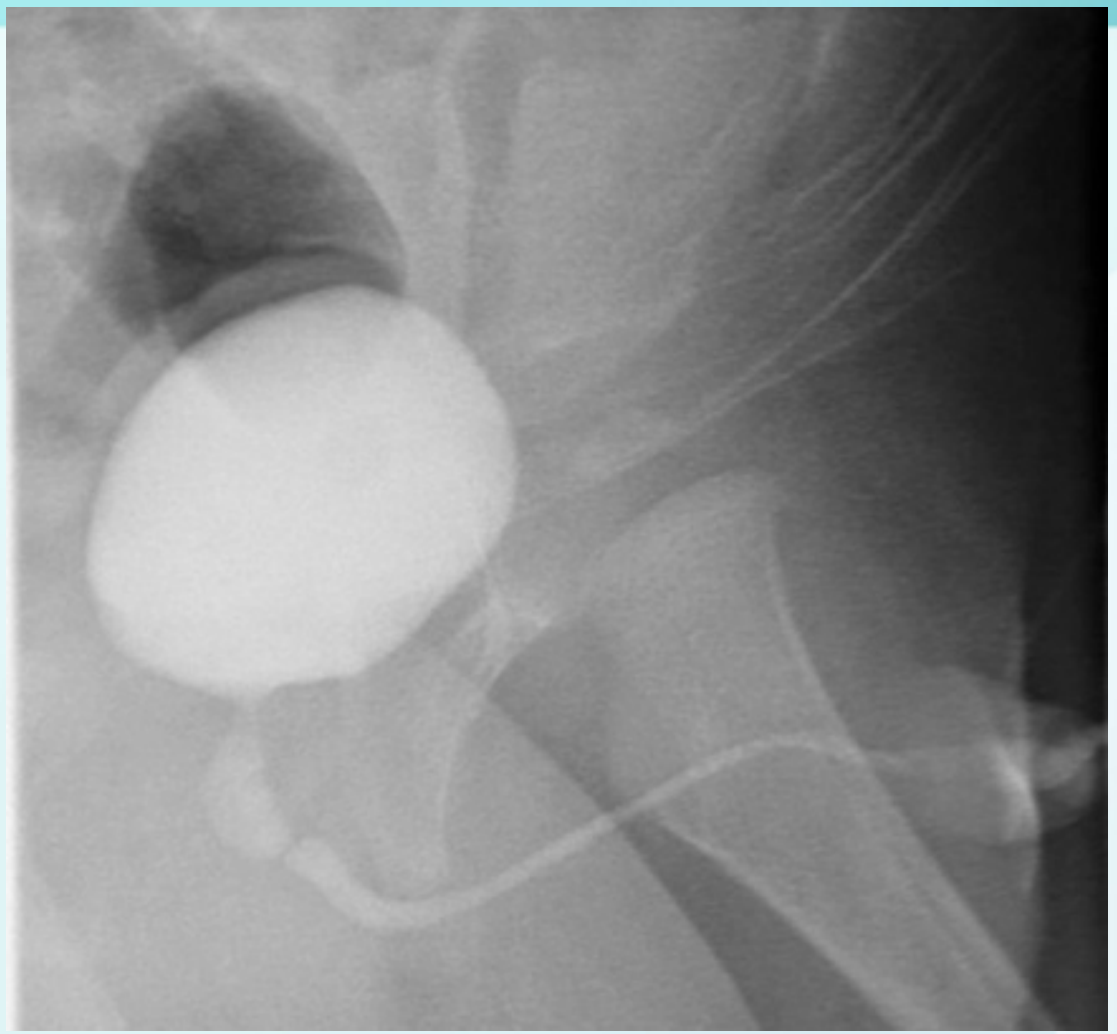
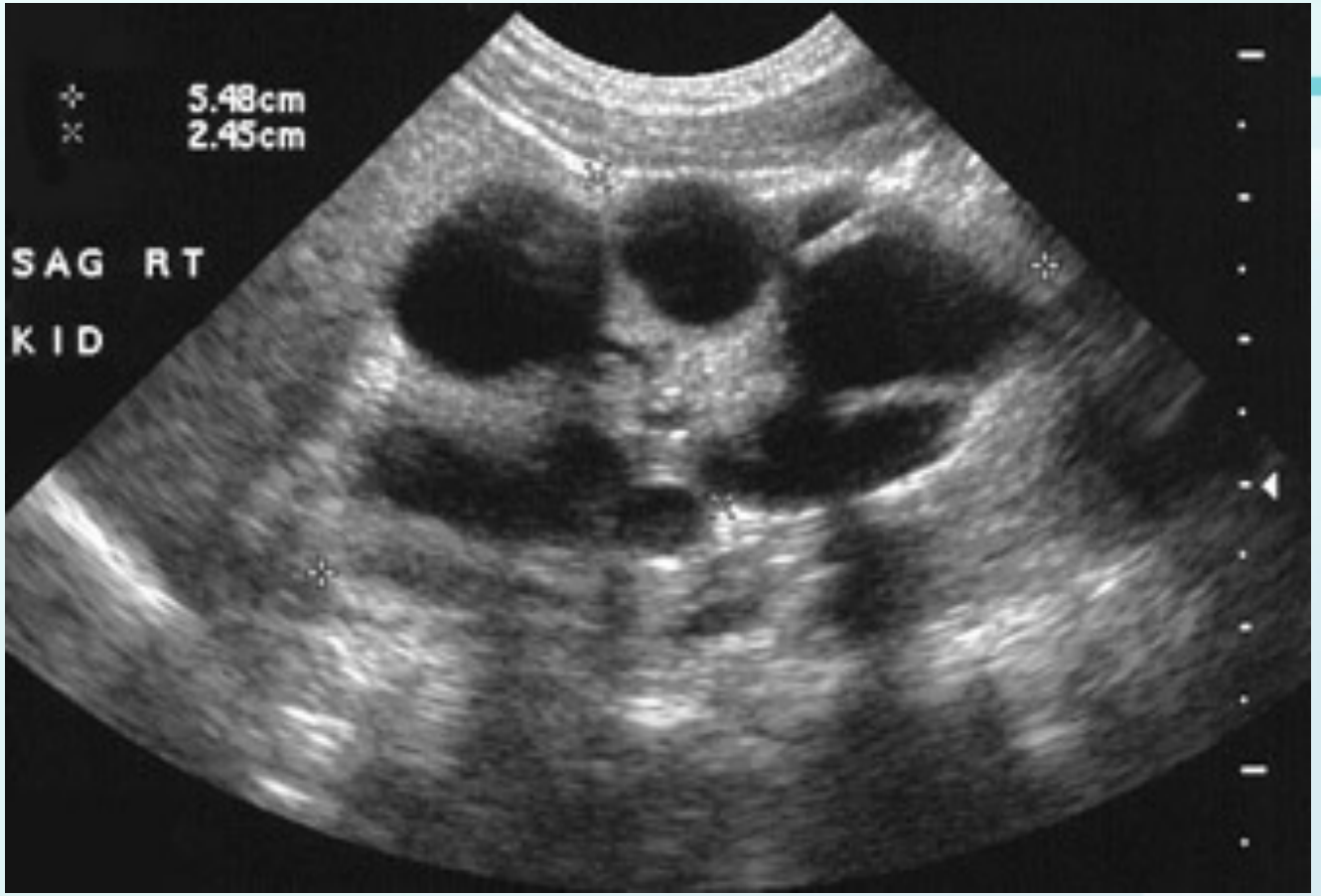



IVP

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KID