

Congenital Pediatric Urinary Disorders

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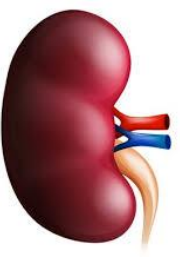
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Learning Objectives:

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

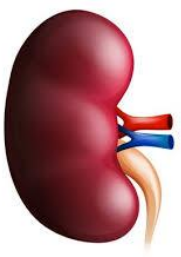
Congenital Urinary Disorders

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



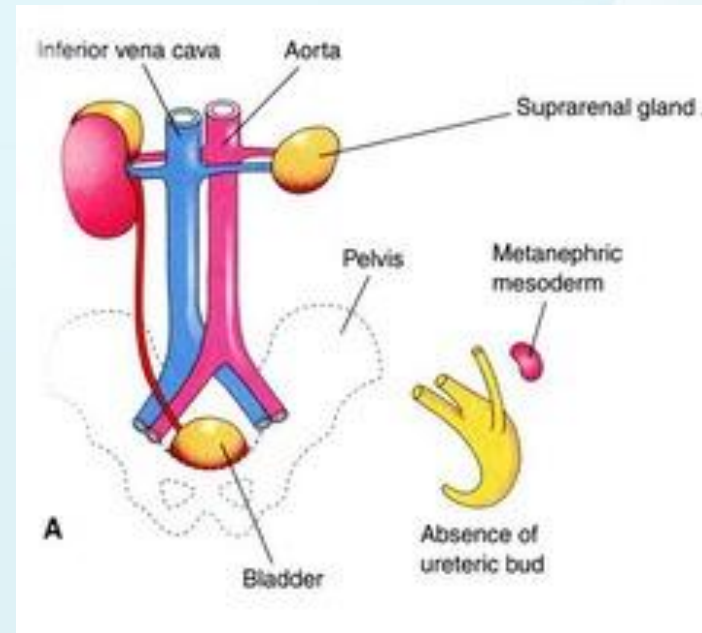
Anomalies of the kidney

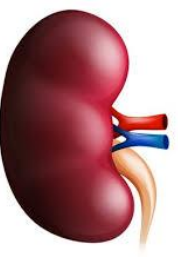
- **Anomalies of:**
 - Number
 - Ascent
 - Form and Fusion
 - Rotation



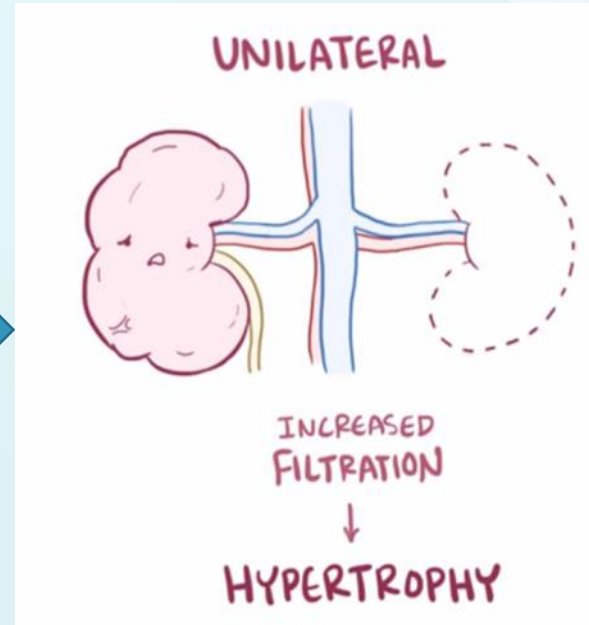
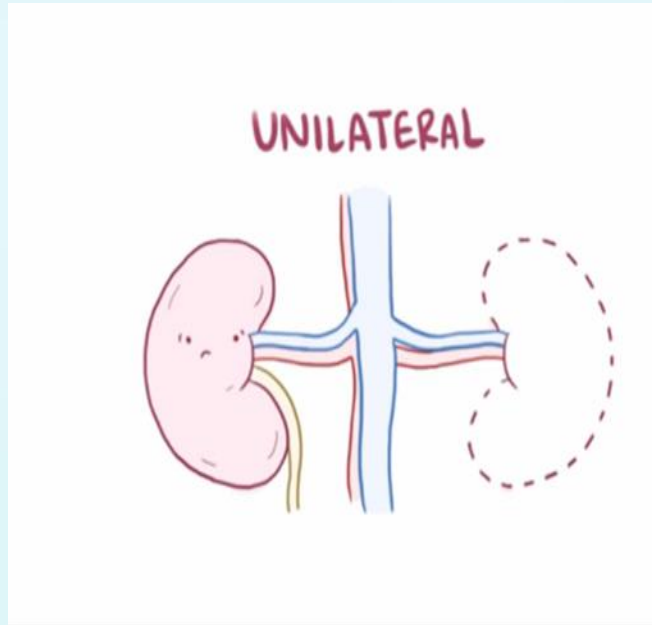
1- Unilateral Renal Agenesis

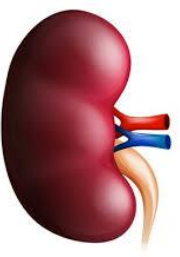
- ▶ 1 in 1100 births.
- ▶ Male: Female of 1.8 : 1
- ▶ The left side is absent more frequently .
- ▶ The ipsilateral ureter is completely absent in 50%.





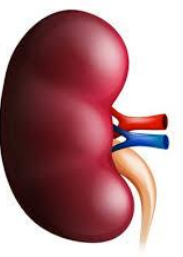
Unilateral Renal Agenesis...





Unilateral Renal Agenesis....

- ▶ Associated anomalies:
 - ▶ Anomalies of other organ systems are found frequently in affected individuals
CVS,GIT,MSC
 - ▶ Müllerian duct abnormalities
 - ▶ 25% to 50% of females
 - ▶ 10% to 15% of males
 - ▶ Approximately one fourth to one third of women with Mullerian duct anomalies are found to have URA.

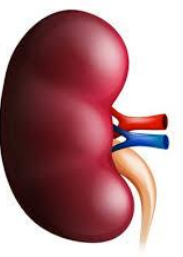


Unilateral Renal Agenesis...

➤ Presentation

- Prenatal US
- Incidentally
 - Abdominal US
 - Abdominal CT



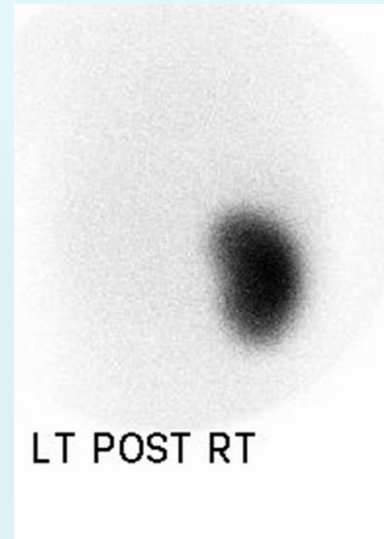


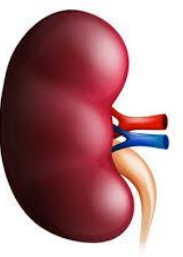
Unilateral Renal Agenesis...

‣ **Diagnosis**

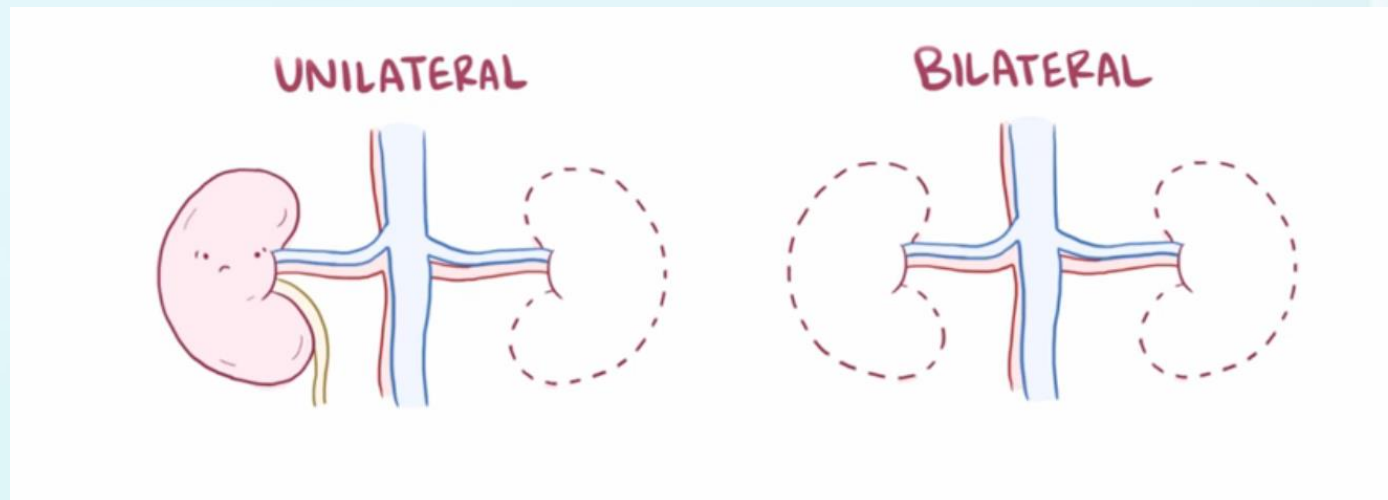
‣ *Confirmed*

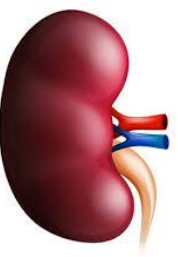
‣ *Nuclear study (DMSA)*



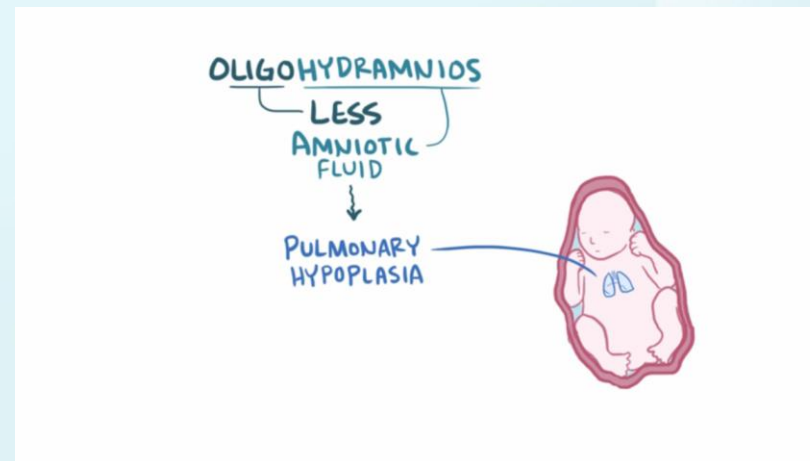
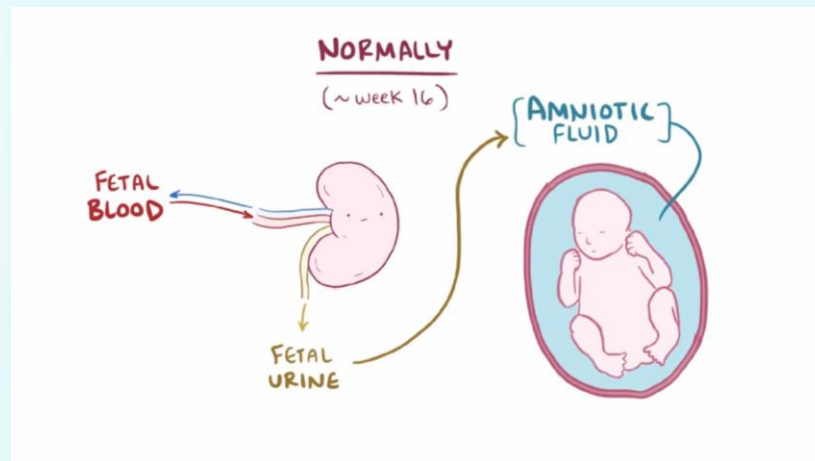


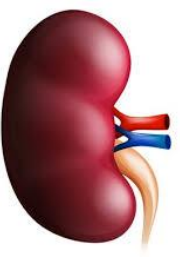
2- Bilateral Renal Agenesis



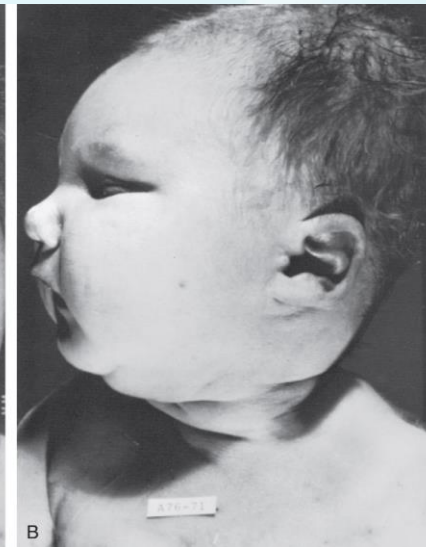
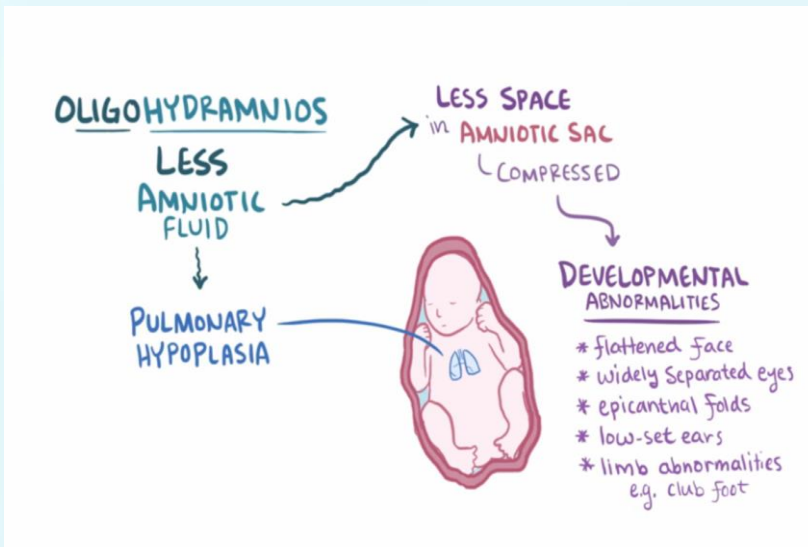


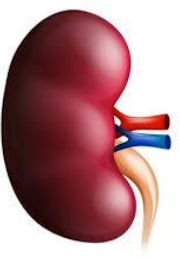
Bilateral Renal Agenesis...





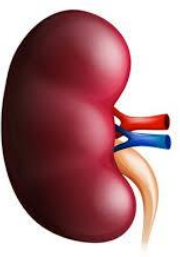
Bilateral Renal Agenesis...





Bilateral Renal Agenesis

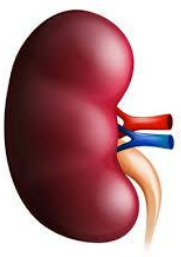
- Ureters are almost always *absent*.
- Bladder is either absent or *hypoplastic*.
- Adrenal glands are usually positioned *normally*.
- Müllerian duct anomalies are commonly observed.



Bilateral Renal Agenesis...

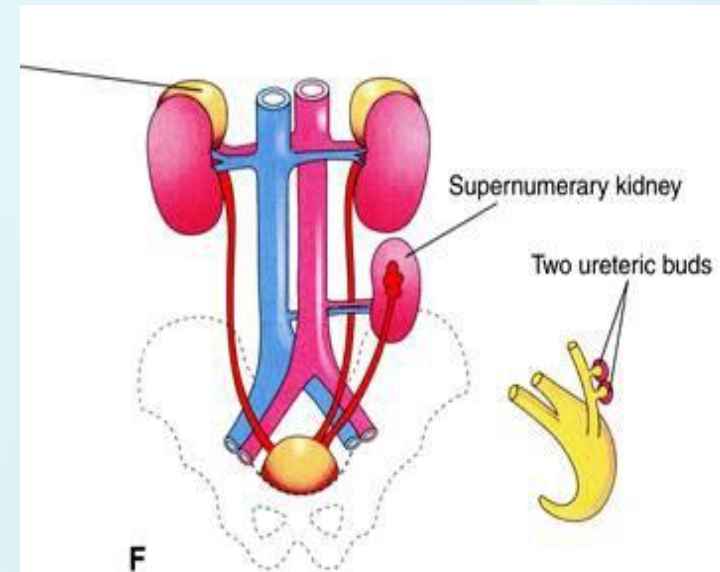
‣ Prognosis

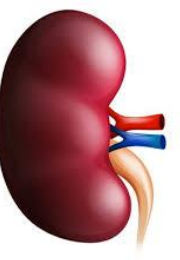
- 40% are stillborn.
- Do not survive beyond 48
 - hours due to respiratory distress associated with pulmonary hypoplasia.



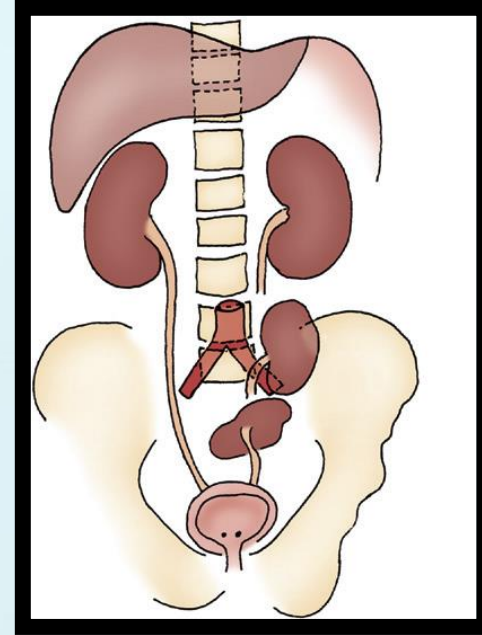
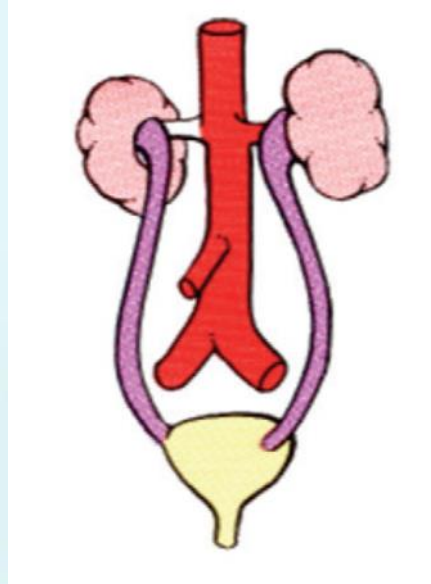
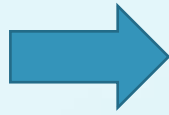
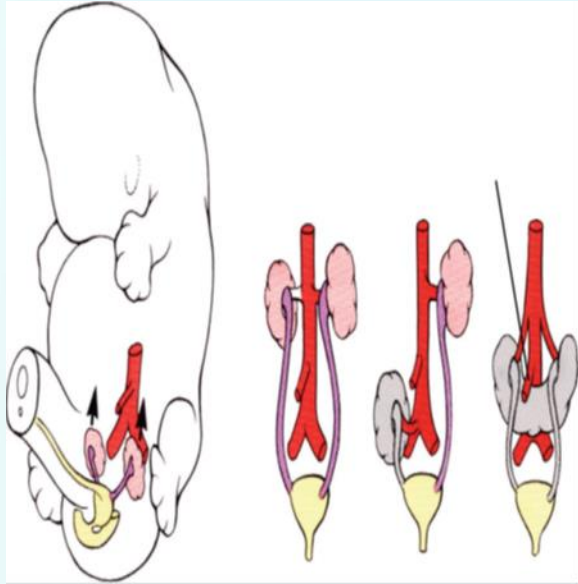
3- Supernumerary Kidney

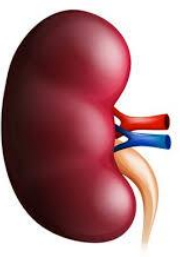
- Definitive **accessory** organ with its own collecting system, blood supply, and distinct encapsulated parenchyma.
- Either completely separate or loosely attached to the kidney on the ipsilateral side.
- The ureteral inter-relationships on the side of the supernumerary kidney can be variable.





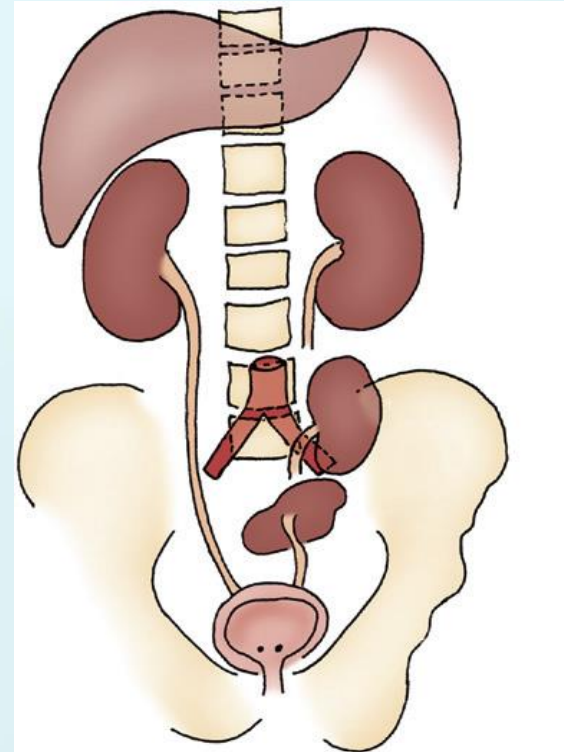
4- Simple Renal Ectopia

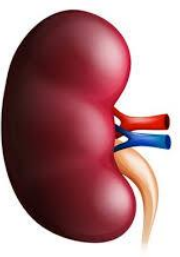




4- Simple Renal Ectopia

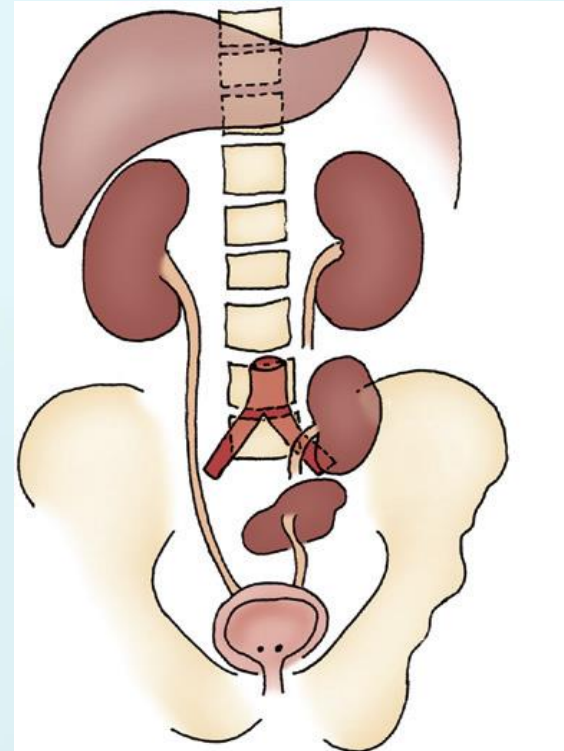
- **Left** more than the right.
- 1 of 2100 to 3000 autopsies.
- Most ectopic kidneys are clinically asymptomatic.

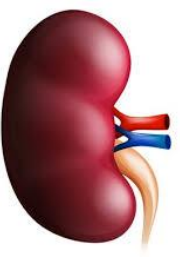




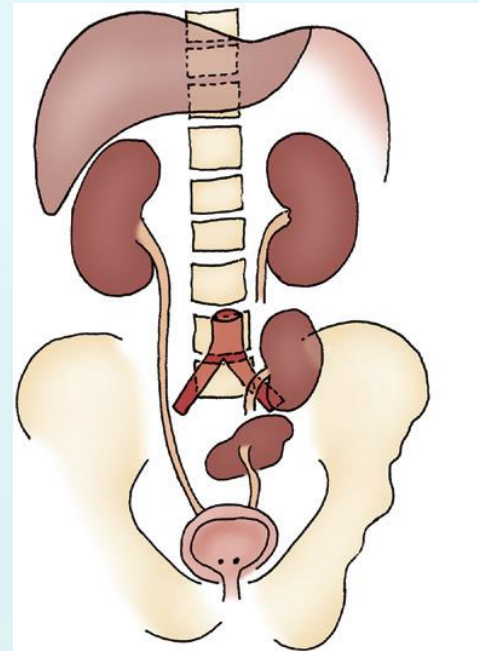
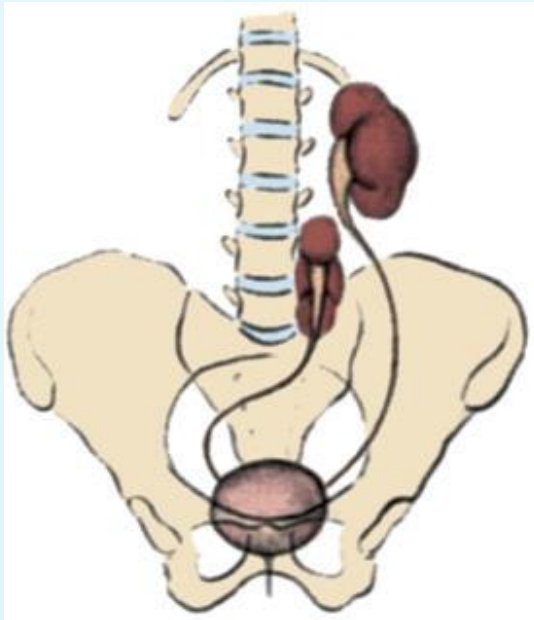
4- Simple Renal Ectopia...

- ▶ Associated Anomalies:
 - ▶ 50% have a **hydronephrosis**:
 - ✓ Obstruction: UPJO and UVJO
 - ✓ Reflux (VUR): grade III or greater
 - ✓ Malrotation
 - ▶ Genital anomalies in the patient with ectopia is about 15%.

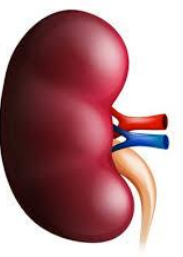




5- Crossed Renal Ectopia

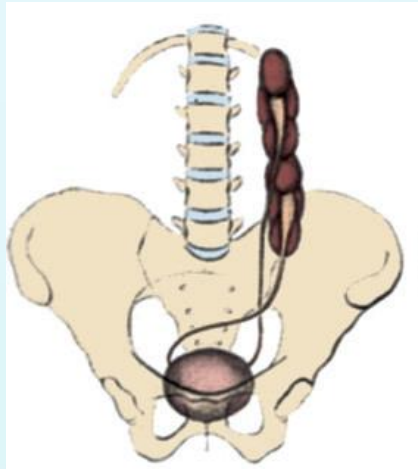


Crossed ectopia: kidney is located on the side opposite from that in which its ureter inserts into the bladder. The ureter from each kidney is usually orthotopic.

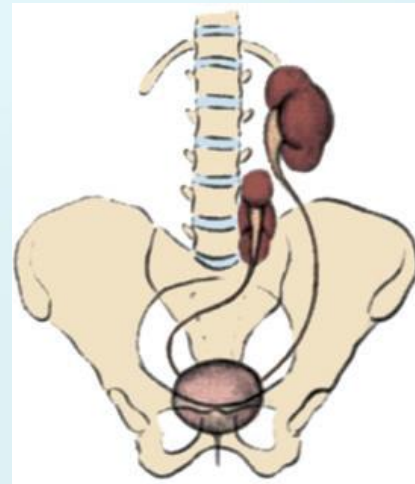


Crossed Renal Ectopia...

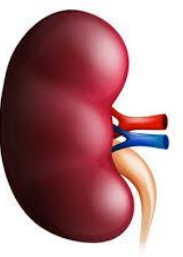
with Fusion



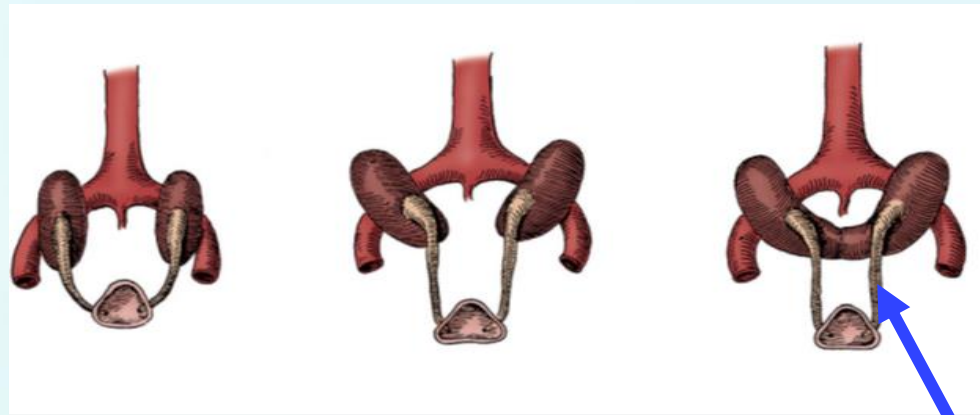
without Fusion



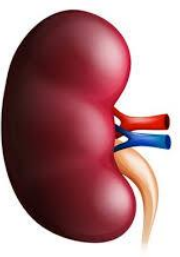
90% are fused
the superior pole of the ectopic kidney usually joins with the inferior aspect of the normal kidney.



6- Horseshoe Kidney



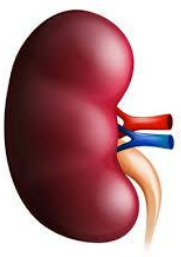
The **isthmus** is bulky and consists of parenchymatous tissue.



Horseshoe Kidney

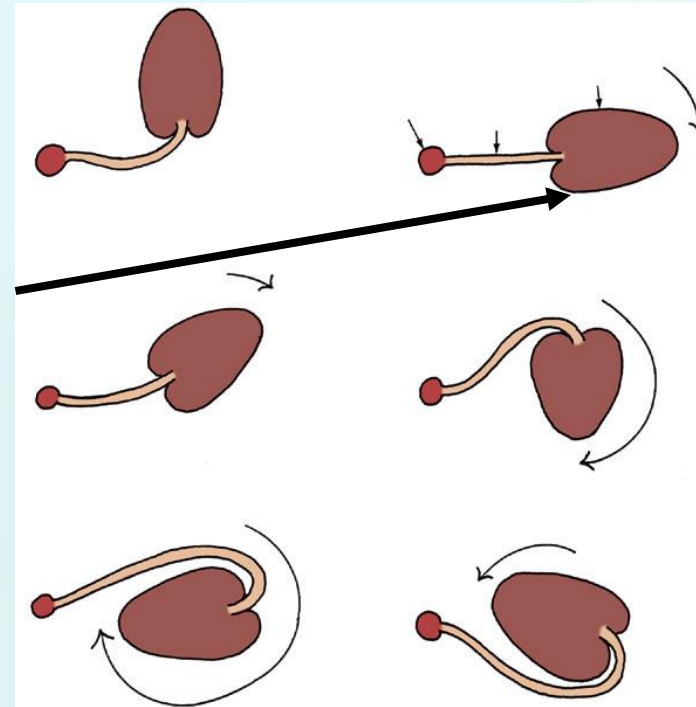
- Occurs 1 in 400 persons.
- The calyces:
 - ✓ normal in number
 - ✓ atypical in orientation.
 - ✓ pelvis remains in the vertical or obliquely lateral plane
- Horseshoe kidney is frequently found in association with other congenital anomalies.
 - UPJ obstruction in one third.
- 60 % asymptomatic.



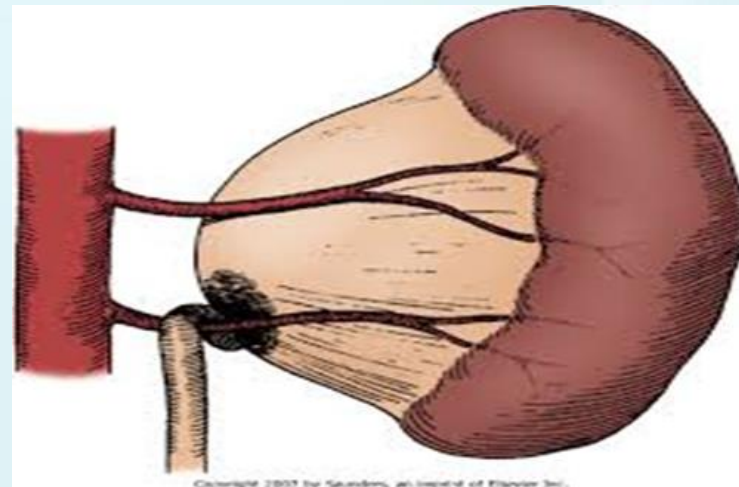
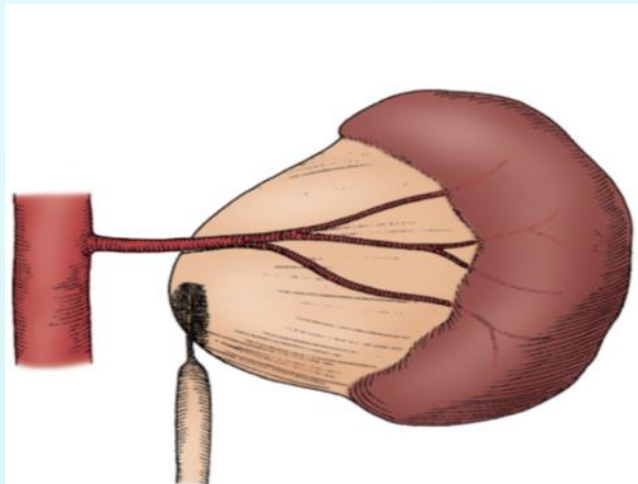


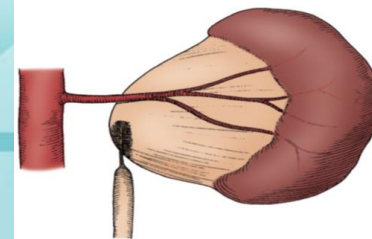
7- Anomalies of Rotation

- ▶ 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.



8-Ureteropelvic junction (UPJ) obstruction

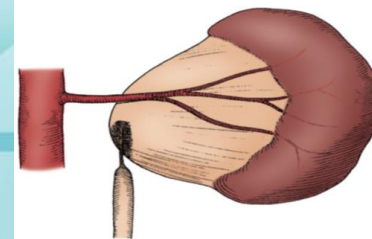




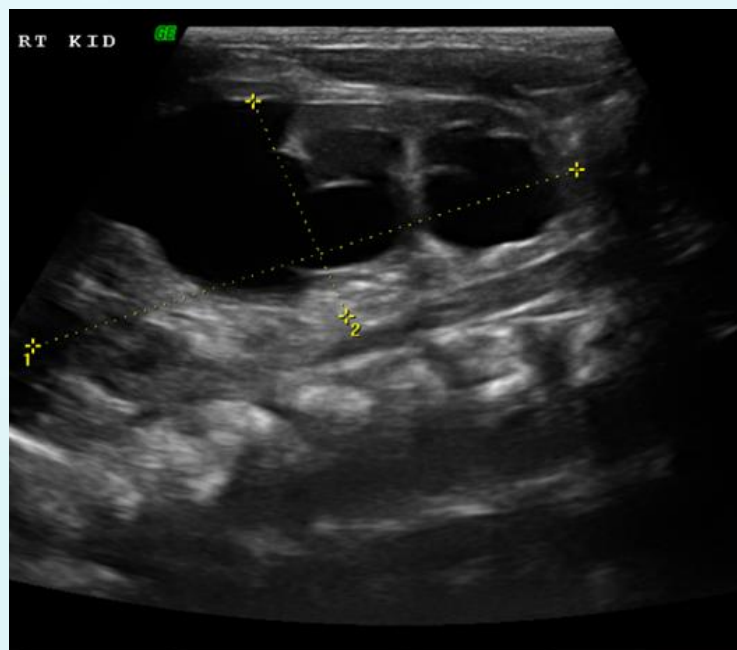
UPJ...

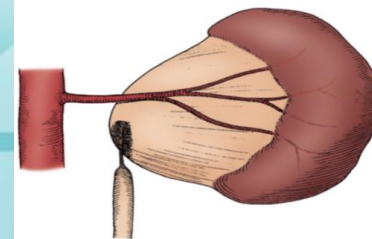
Presentation:

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



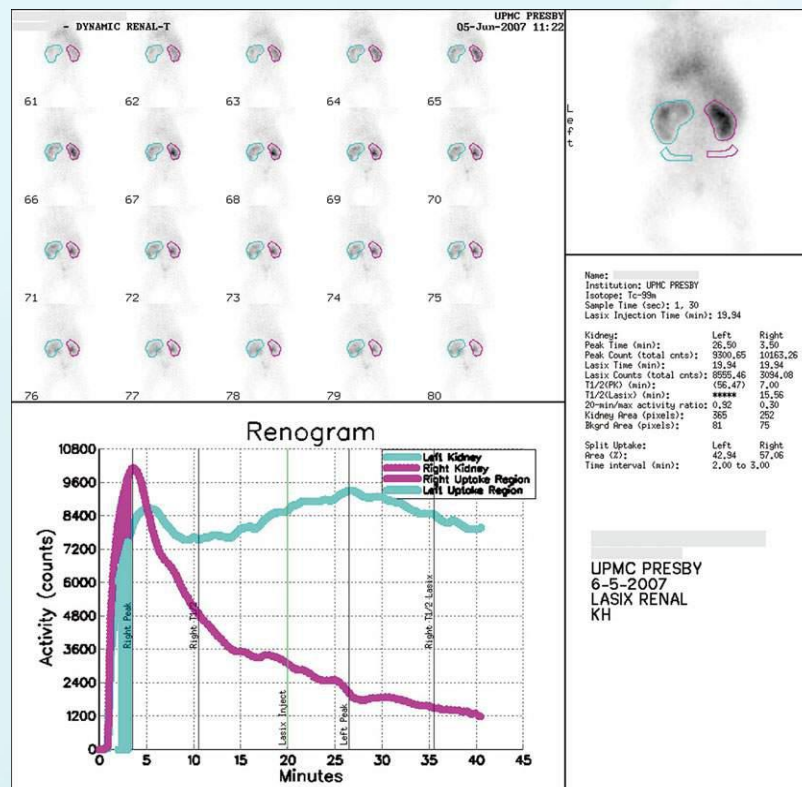
UPJO...

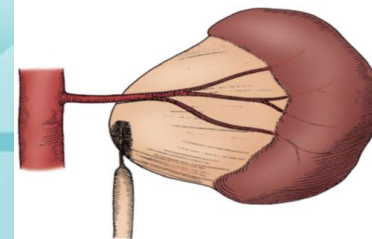




UPJO...

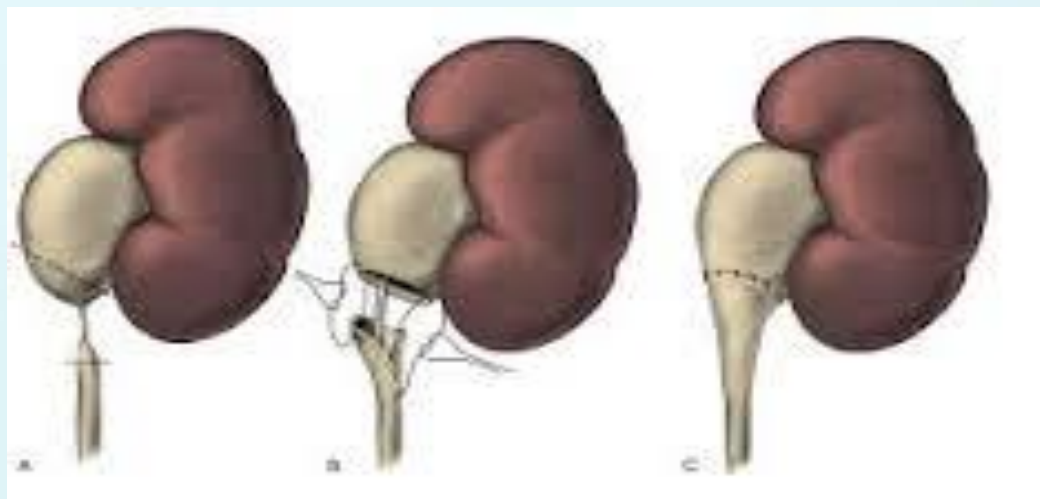
➤ Dynamic renogram



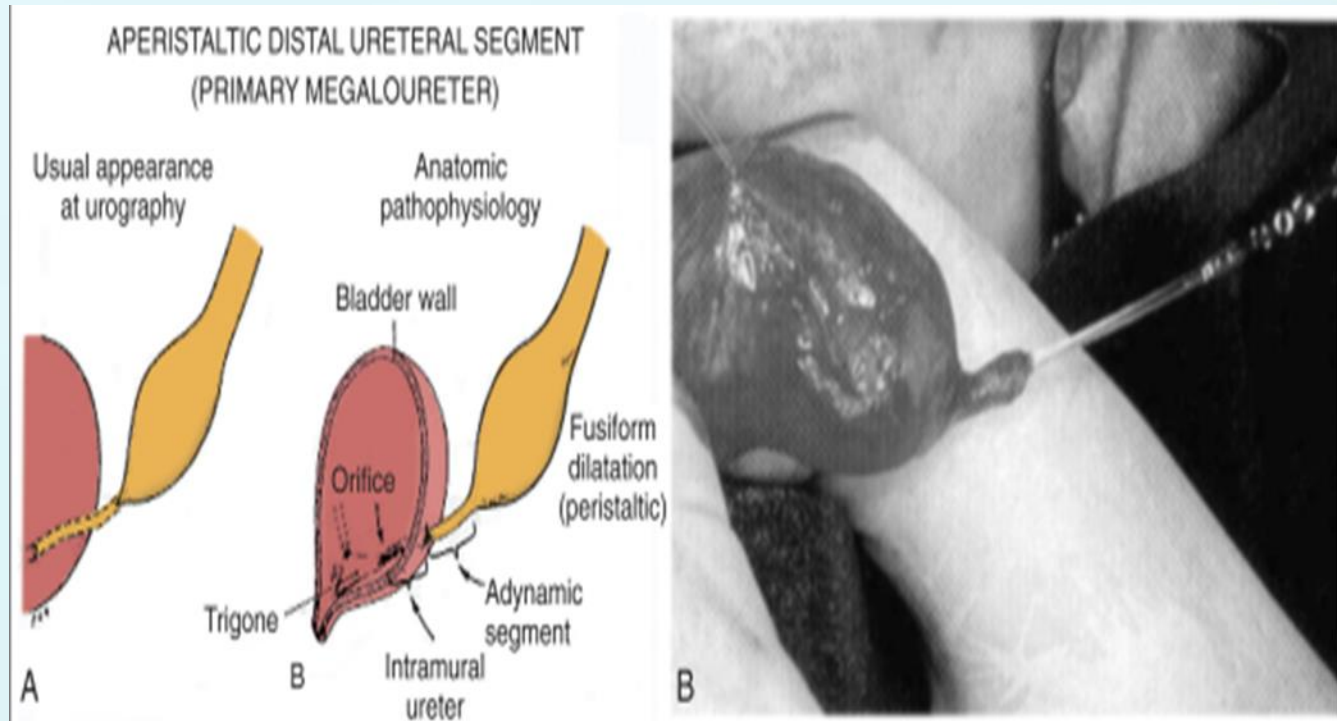


UPJO...

- ▶ **Dismembered Pyeloplasty**



9-Ureterovesical junction (UVJ) obstruction (Megaureters)



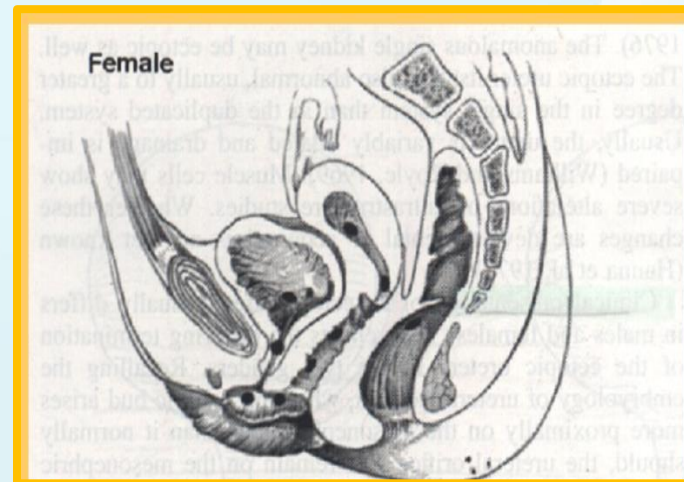
11- Ectopic Ureter

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



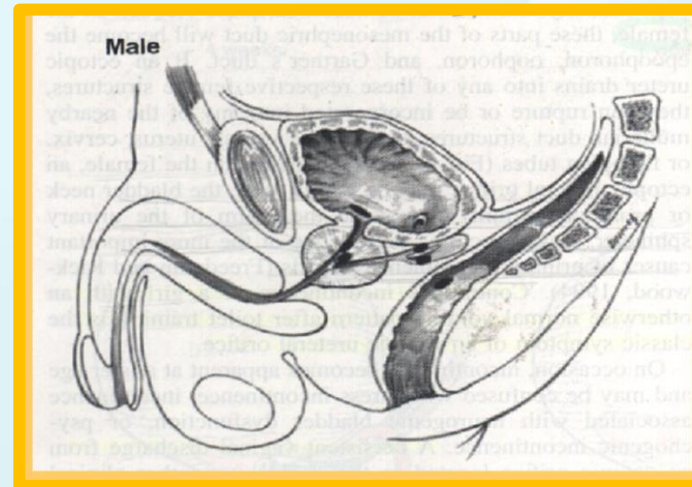
Ectopic Ureter...

- ▶ 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**.



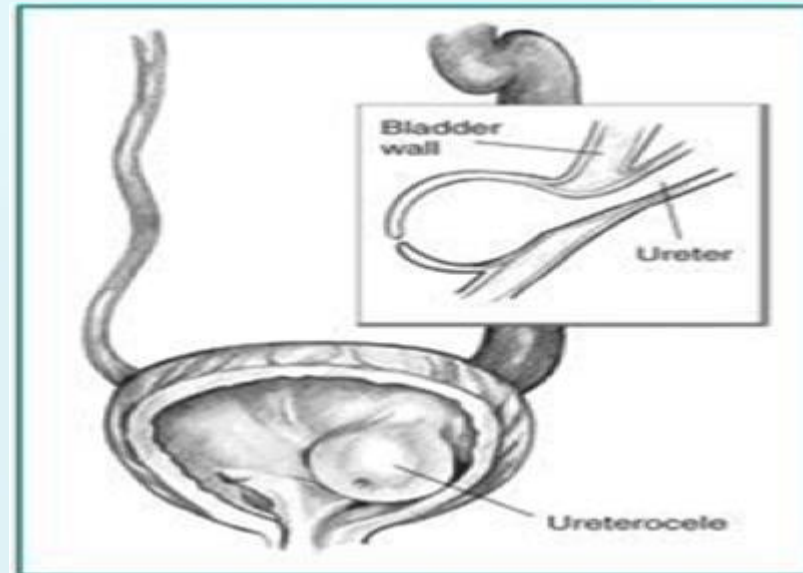
Ectopic Ureter

- ▶ 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.

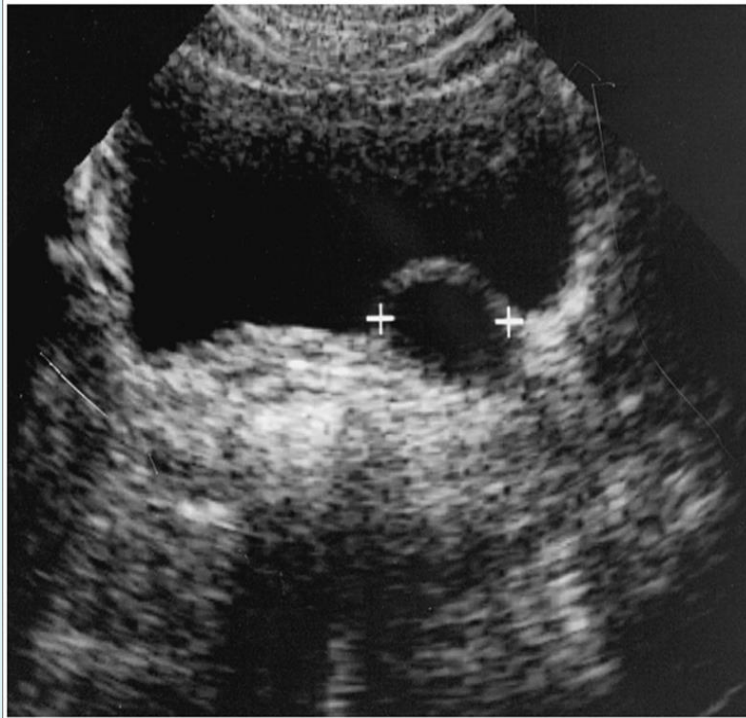


12- Ureterocele

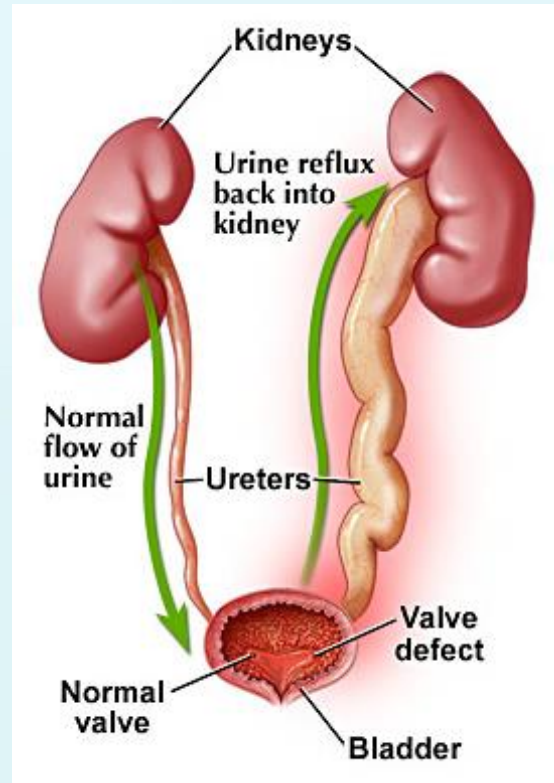
- A cystic dilation of the distal aspect of the ureter
- Located either within the bladder or spanning the bladder neck and urethra.
- **Presentation:**
 - Antenatal (U/S)
 - Urine retention
 - Infection
 - Calculus formation



Ureterocele...



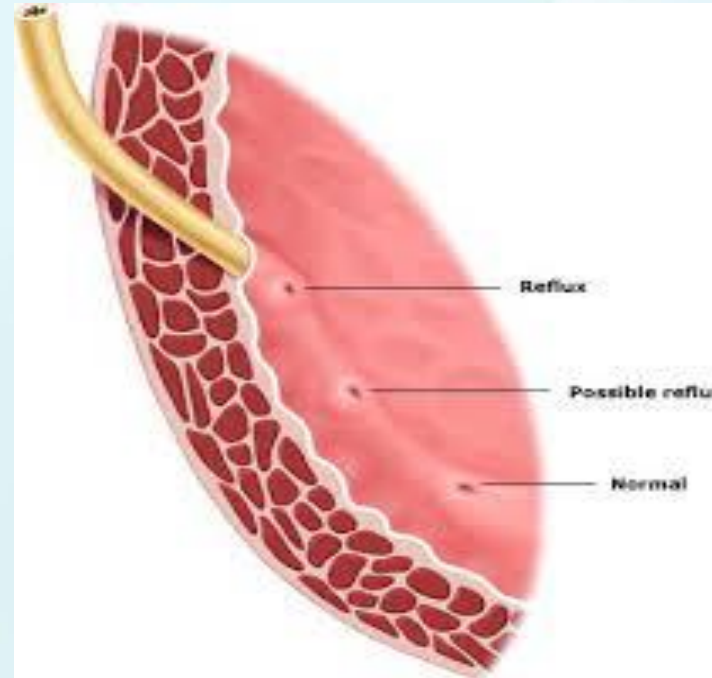
13-Vesicoureteral Reflex (VUR)



Vesicoureteral Reflux (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.



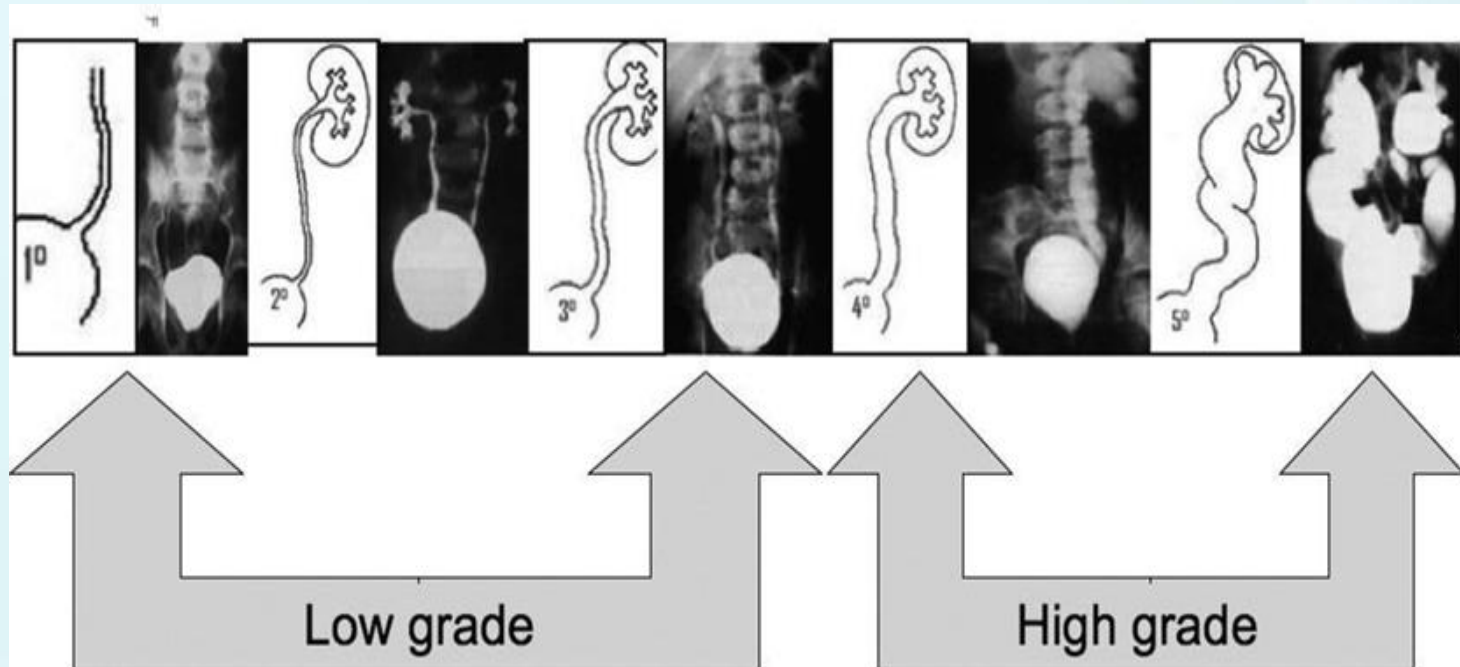
VUR...

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

VUR...

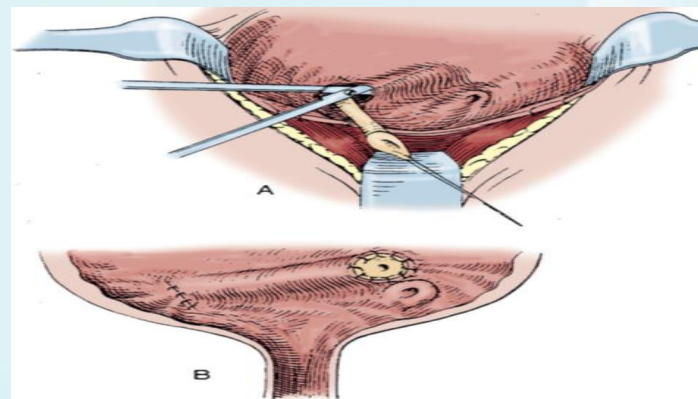
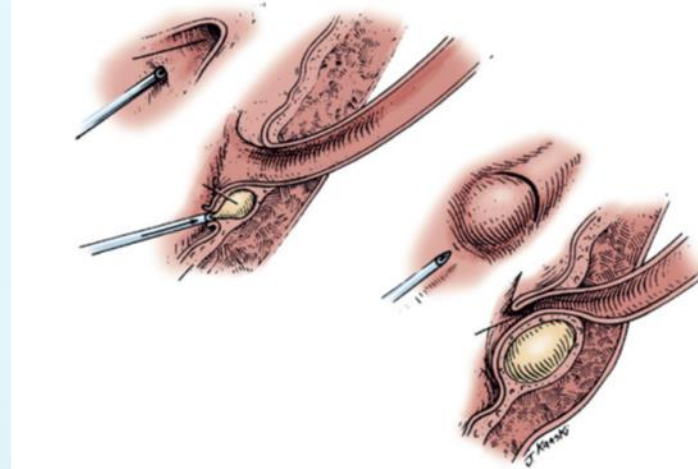
▷ Diagnosis:

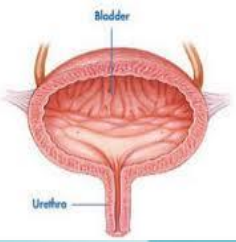
▷ MCUG (VCUG)



VUR...

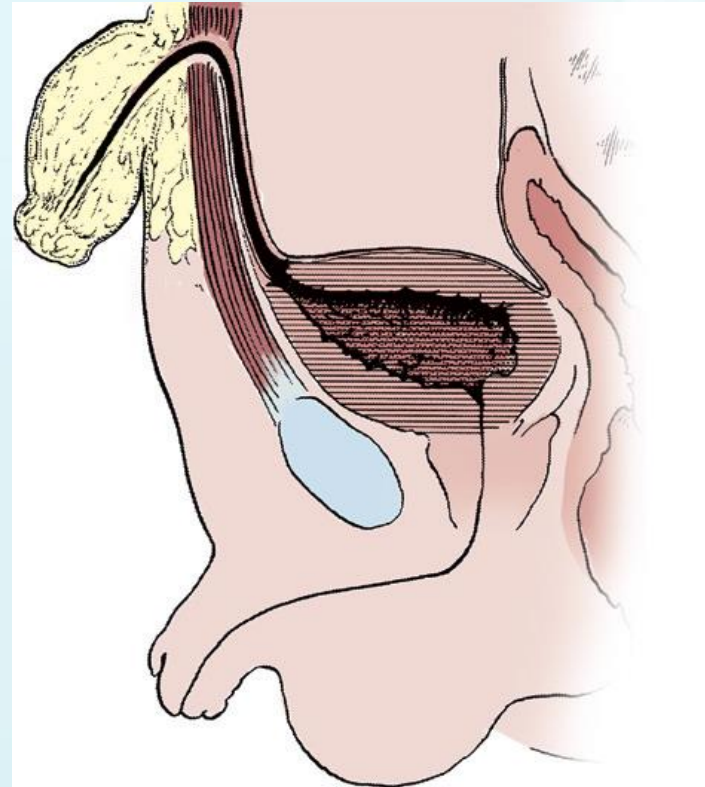
- **Management:**
- Prophylactic antibiotic
- Surgical treatment
 - Endoscopic treatment
 - Ureteral reimplantation

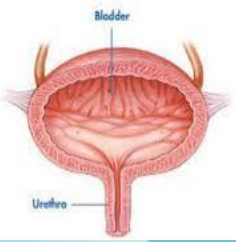




14-Urachal abnormalities

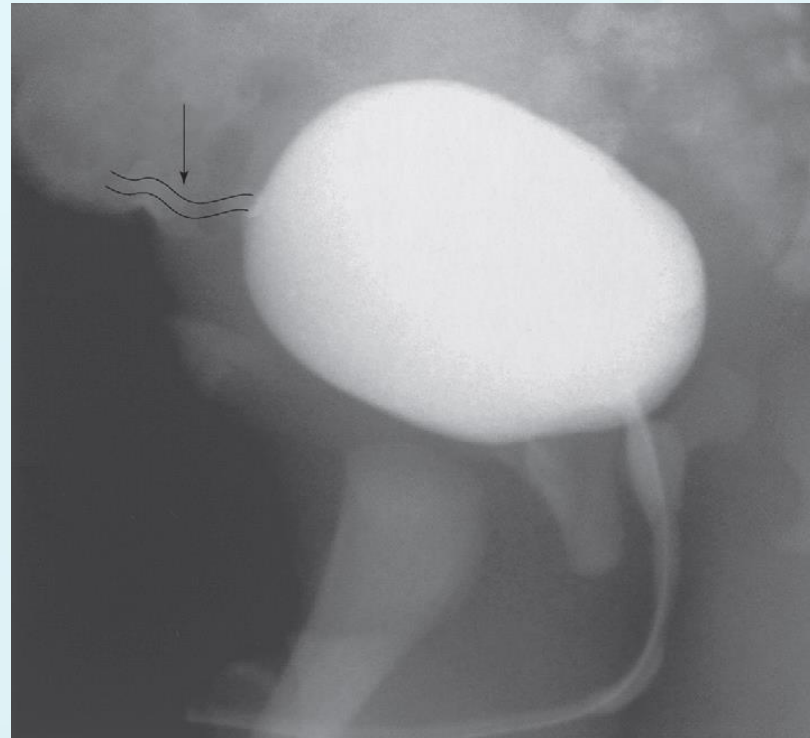
- ▶ Urachal anomalies are usually detected postnatally due to umbilical drainage.
- ▶ Imaging possibilities include ultrasound, CT, and VCUG.

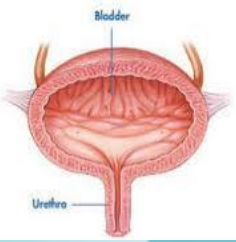




14-Urachal abnormalities...

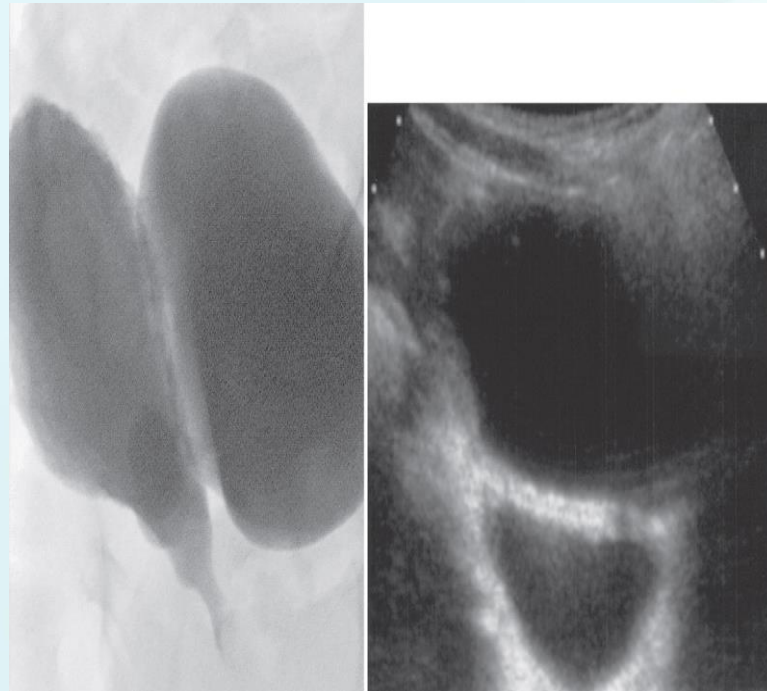
- ▶ **Conservative** treatment with observation is justified in asymptomatic cases due to possible spontaneous resolution
- ▶ **Infected urachal** remnants are initially treated with drainage and antibiotics, followed by surgical excision.
- ▶ Nonresolved urachal remnants should be excised due to the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

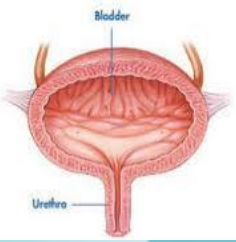




15-Bladder Diverticulum

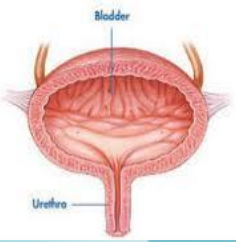
- ▶ Bladder diverticula can be detected on prenatal ultrasound, but the gold standard remains VCUG, which will reveal possible accompanying VUR.





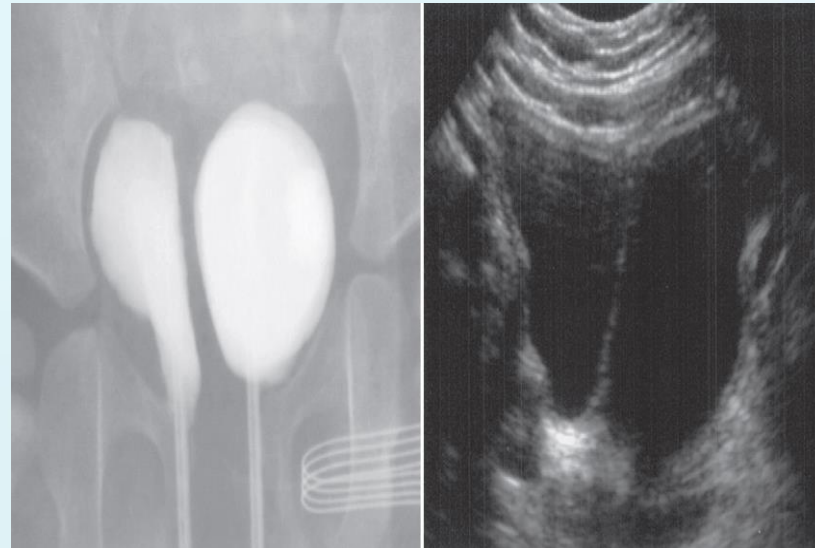
Bladder Diverticulum...

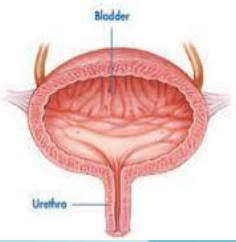
- Types:
 - Primary diverticula
 - arise as a localized herniation of **bladder mucosa** at the ureteral hiatus and are most likely caused by a **congenitally** deficient bladder wall.
 - Secondary para-ureteral diverticula
 - are acquired and develop due to existing infra- vesical obstruction.
- Symptomatic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



16- Bladder Duplication

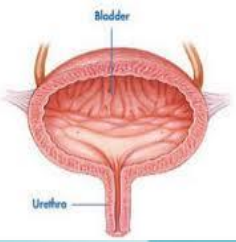
- ▶ Often associated with duplication anomalies of the external genitalia and lower gastrointestinal tract.
- ▶ Initial treatment is directed toward
 - ▶ renal preservation.
 - ▶ prevention of infections.



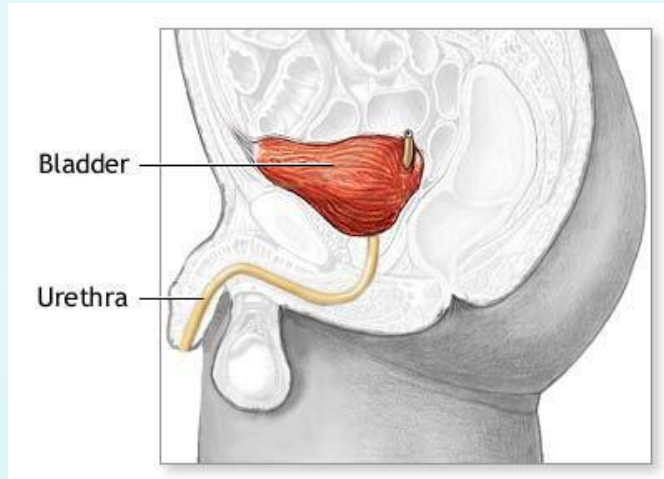


Bladder Duplication...

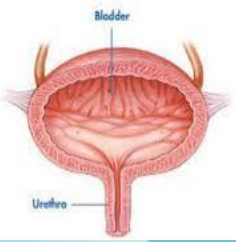
- ▶ Long-term goals include achieving continence and reconstructing the internal and external genitalia.
- ▶ Due to the rarity of the disease and the large variety of presentations, the surgeries must be individualized



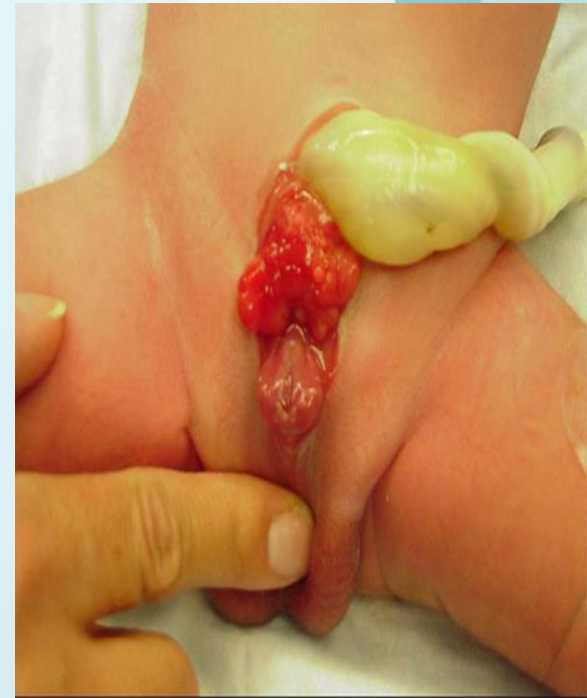
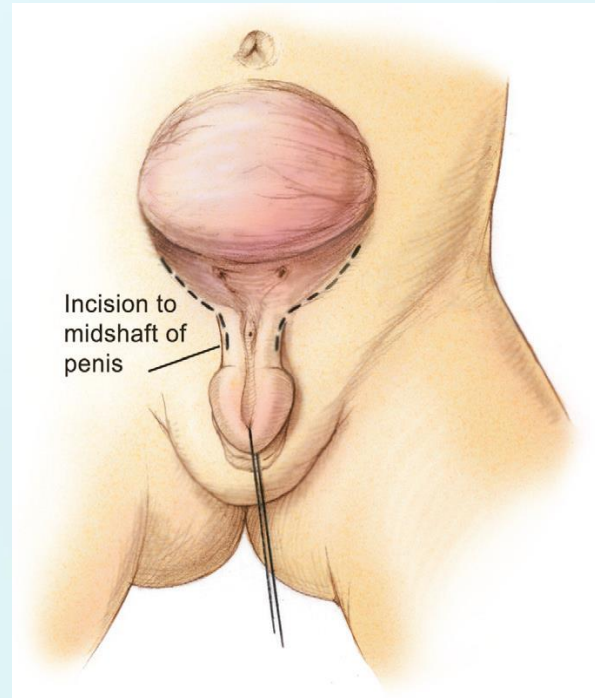
17- Bladder Exstrophy



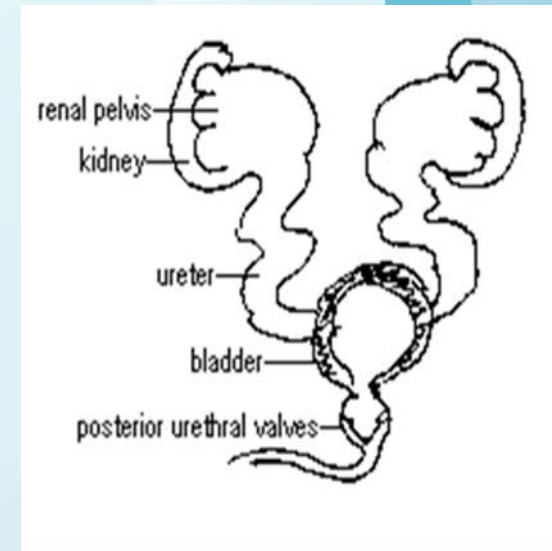
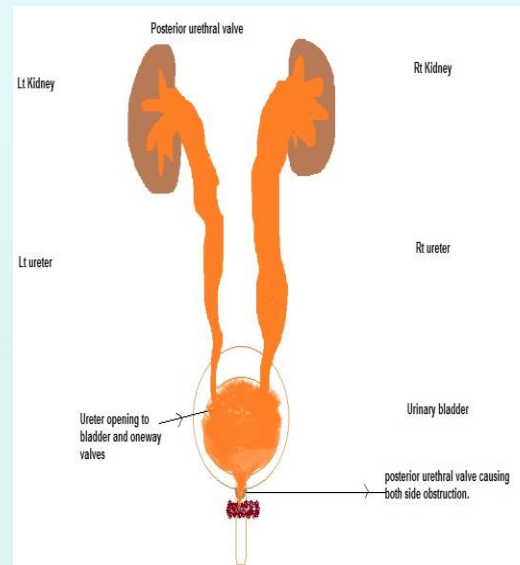
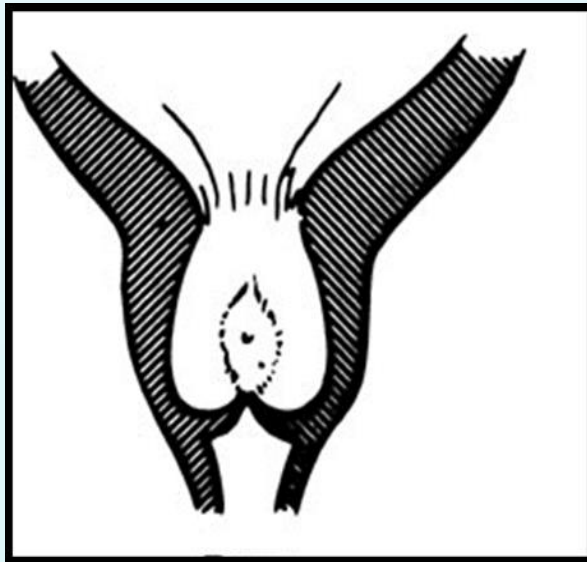
The incidence of bladder exstrophy has been estimated as between 1 in 10,000 and 1 in 50,000.



Bladder Exstrophy...



18-Posterior Urethral Valves (PUV)



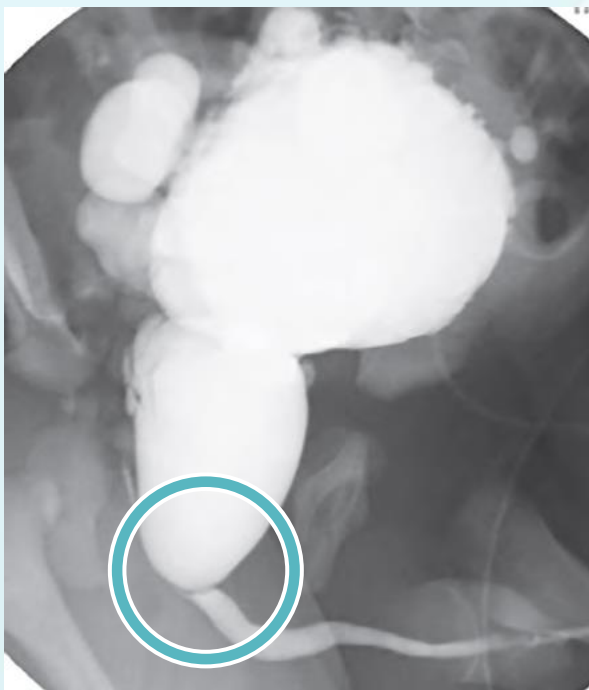
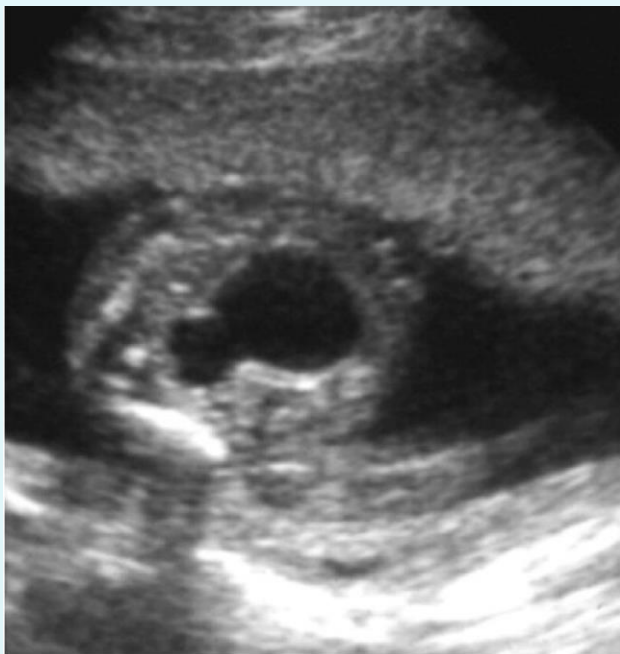
The bladder and the kidneys developed under high pressure and resistance.

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...



PUV...

➤ **Presentation:**

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

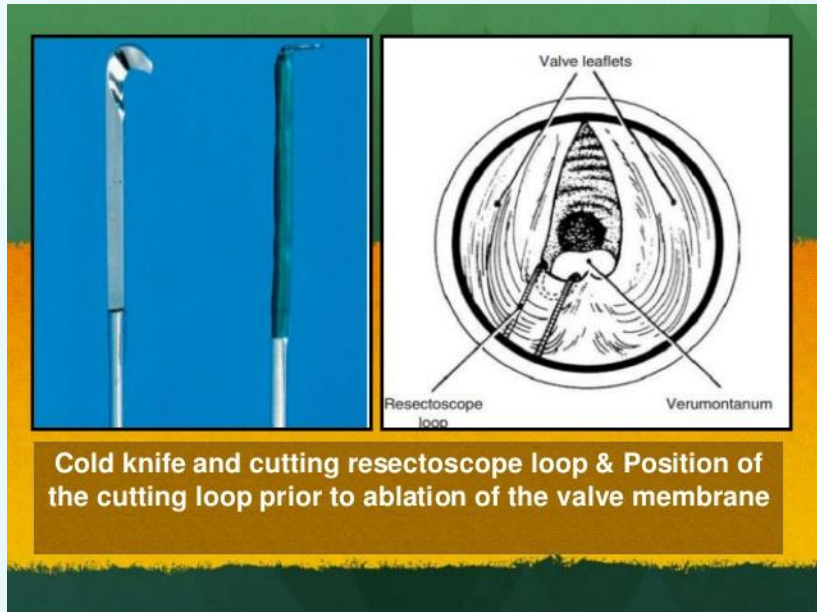
• **Associated findings:**

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

PUV...

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

PUV...



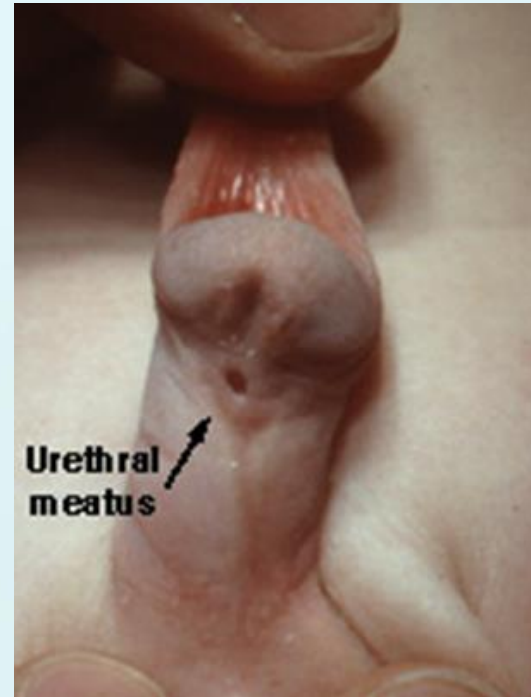
Endoscopic valve ablation



Cutaneous vesicostomy

19-Hypospadias

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



20- Epispadias

Male



Female



21-Cloacal Exstrophy



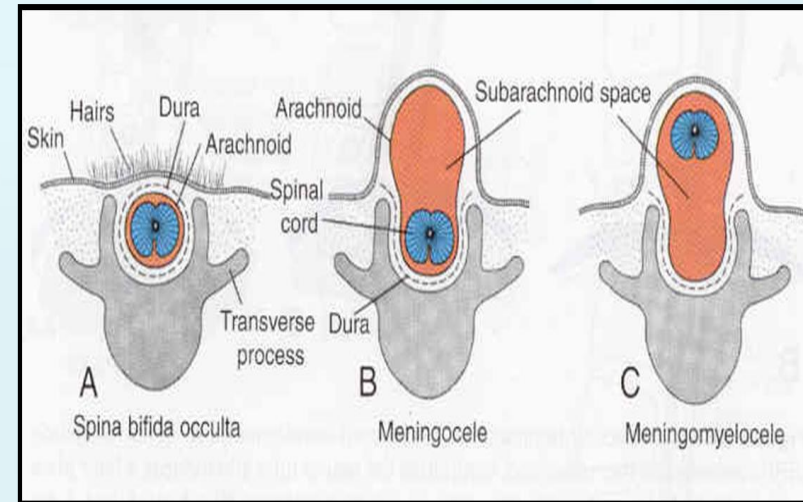
22- Prune-Belly Syndrome

- 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



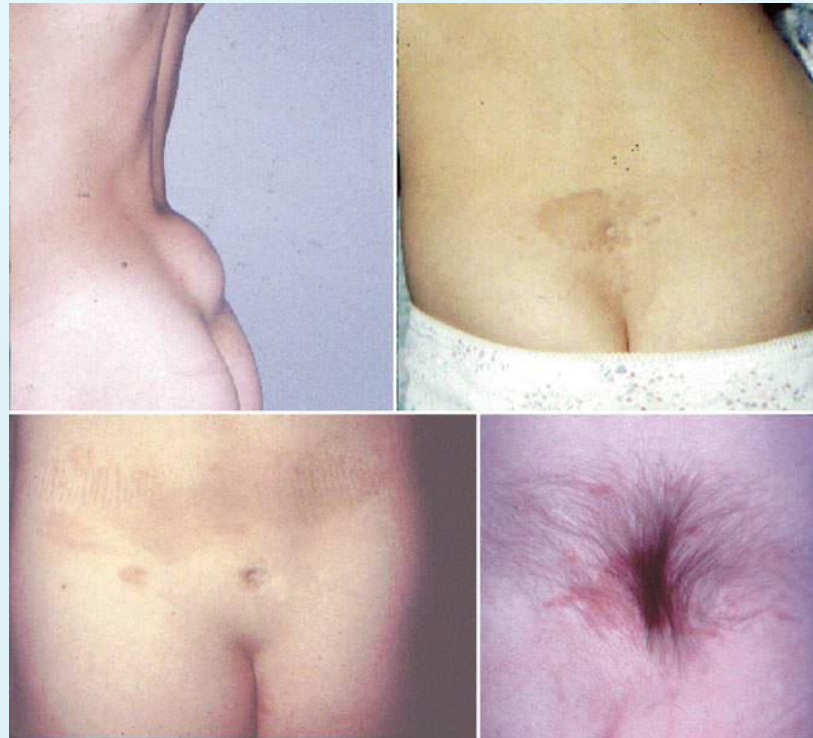
23-NEUROSPINAL DYSRAPHISMS

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



NEUROSPINAL DYSRAPHISMS...

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

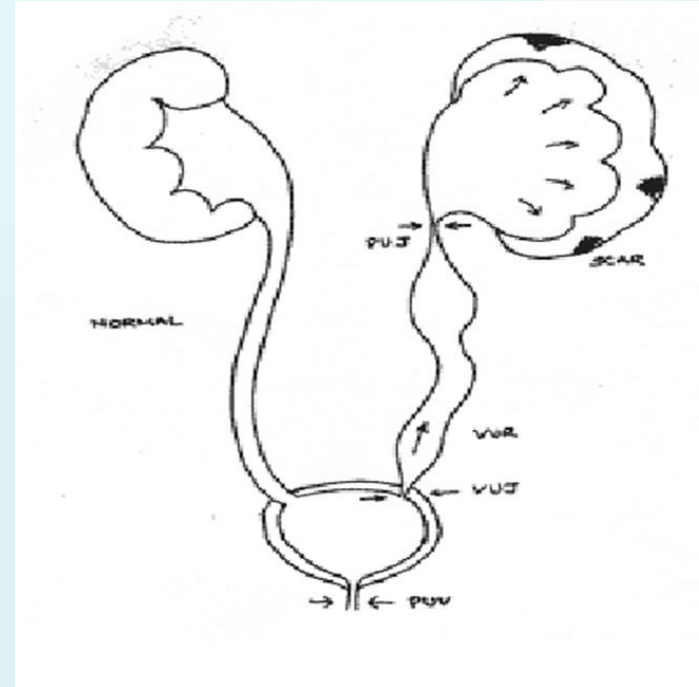


Antenatal Hydronephrosis(ANH)

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%)



THANK
YOU!

