

# **Congenital Pediatric Urinary Disorders**

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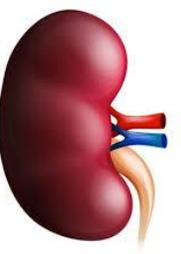
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## **Learning Objectives:**

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

# **Congenital Urinary Disorders**

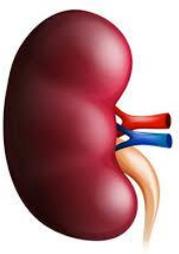
- **Anomalies of the Upper Urinary Tract**
  - **Kidney**
  - **Ureter**
- **Anomalies of the Lower Urinary Tract**
  - **Urinary Bladder**
  - **Urethra**



## **Anomalies of the kidney**

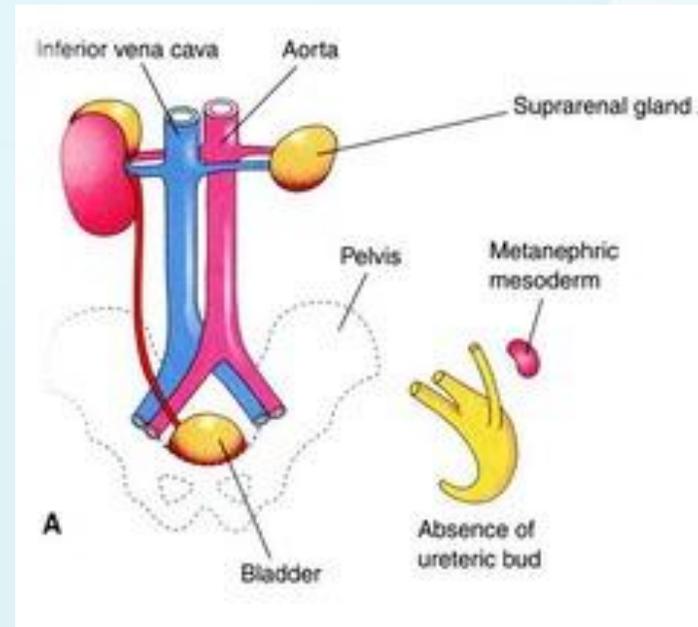
### ‣ **Anomalies of:**

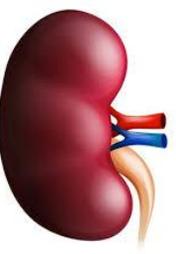
- Number
- Ascent
- Form and Fusion
- Rotation



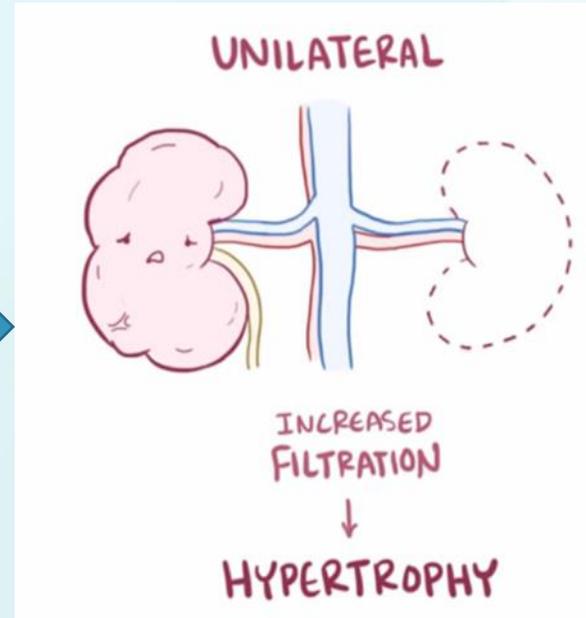
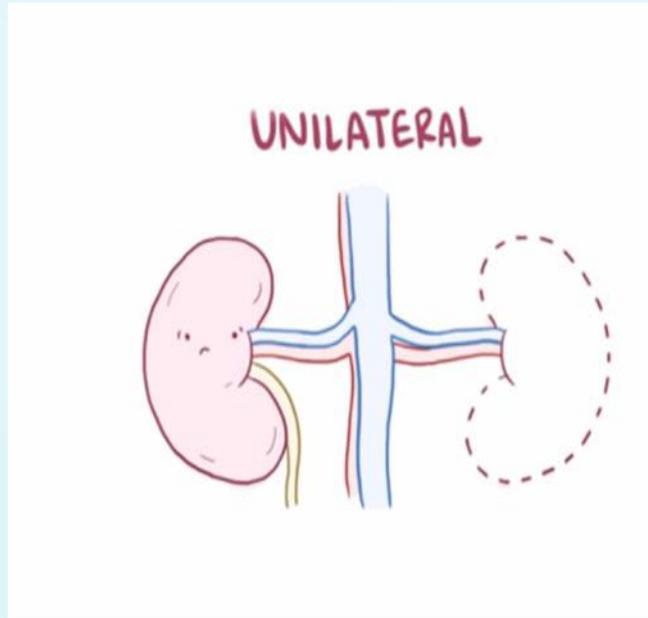
## 1- Unilateral Renal Agenesis

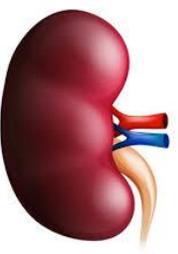
- ▶ 1 in 1100 births.
- ▶ Male: Female of 1.8 : 1
- ▶ The left side is absent more frequently .
- ▶ The ipsilateral ureter is completely absent in 50%.





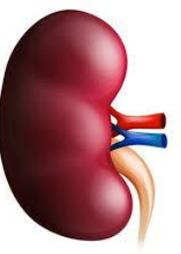
## Unilateral Renal Agenesis...





## 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.

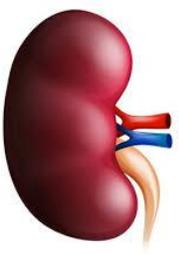


## Unilateral Renal Agenesis...

### ➤ Presentation

- Prenatal US
- Incidentally
  - Abdominal US
  - Abdominal CT



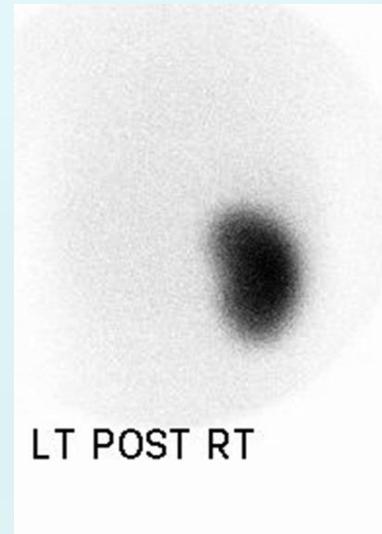


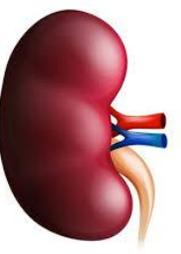
## Unilateral Renal Agenesis...

### ‣ **Diagnosis**

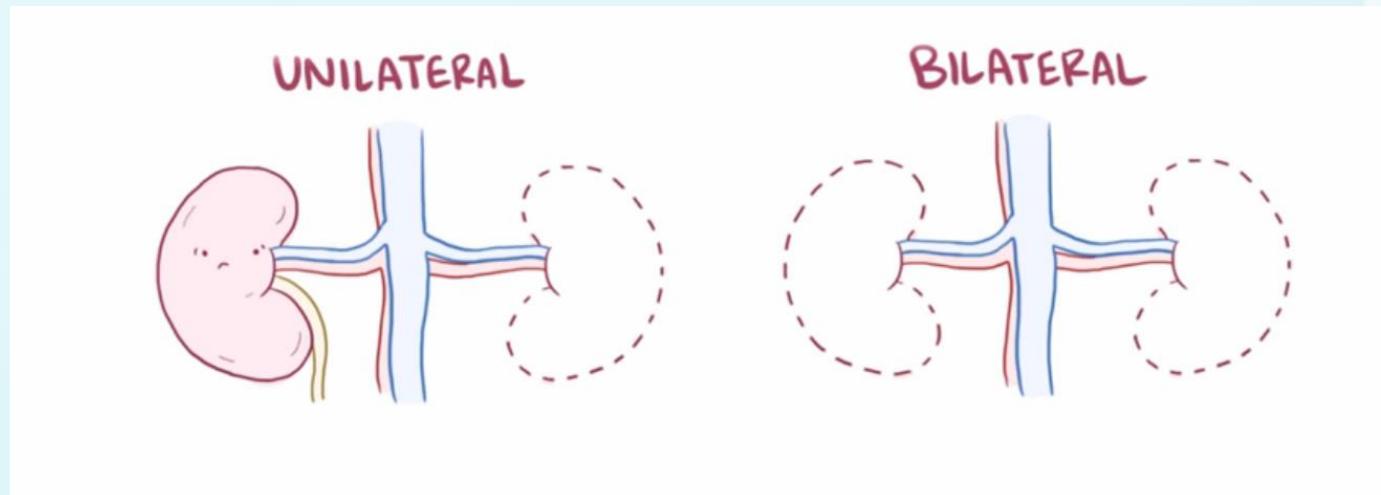
‣ *Confirmed*

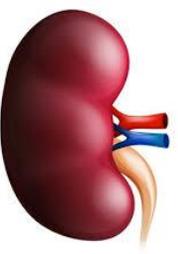
‣ *Nuclear study (DMSA)*



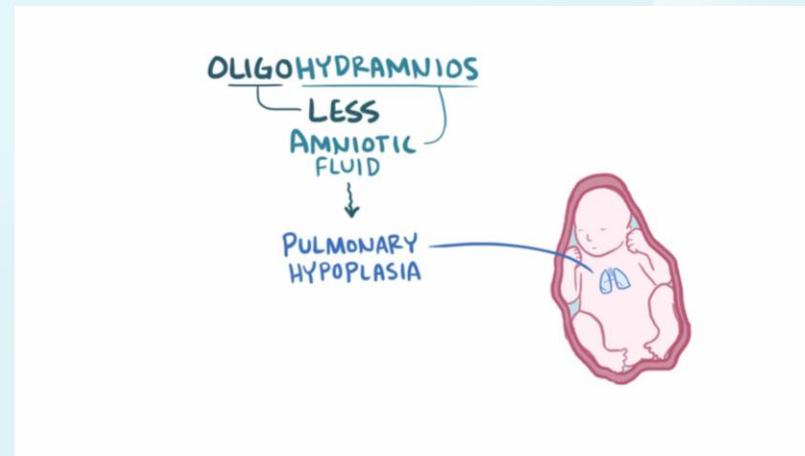
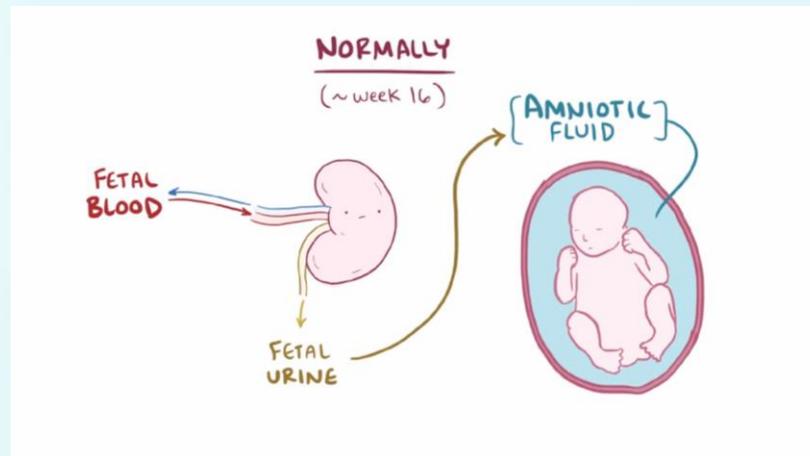


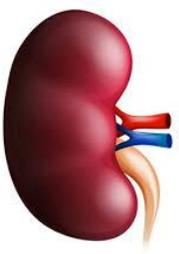
## 2- Bilateral Renal Agenesis



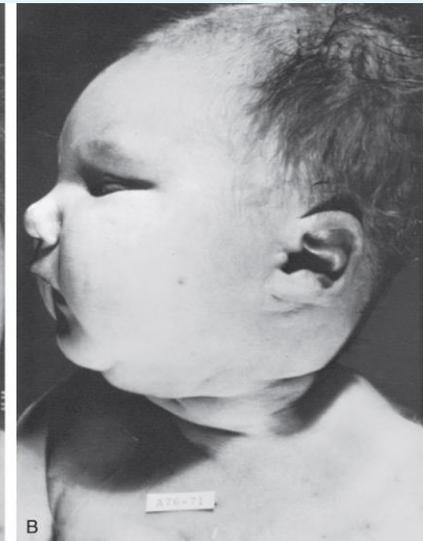
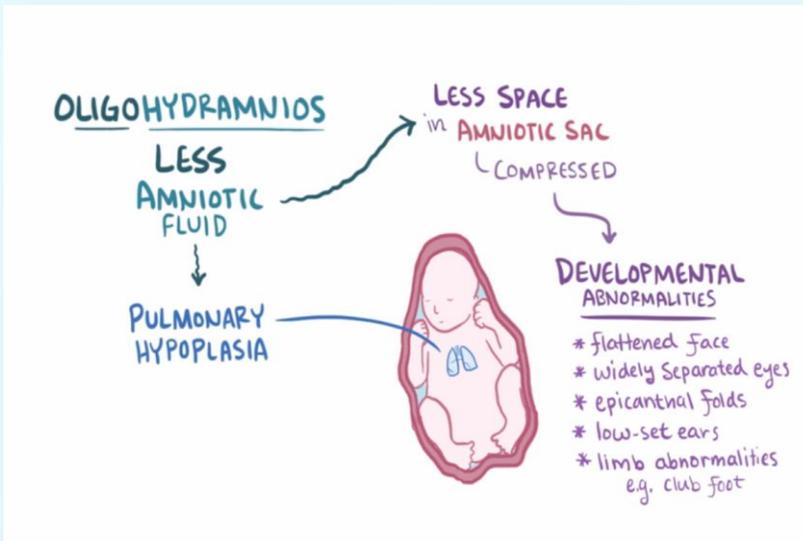


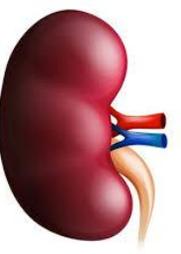
## Bilateral Renal Agenesis...





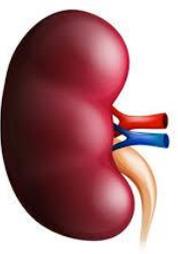
## Bilateral Renal Agenesis...





## Bilateral Renal Agenesis

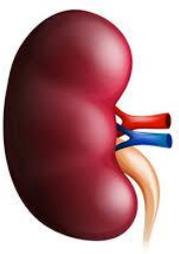
- Ureters are almost always *absent*.
- Bladder is either absent or *hypoplastic*.
- Adrenal glands are usually positioned *normally*.
- Müllerian duct anomalies are commonly observed.



## Bilateral Renal Agenesis...

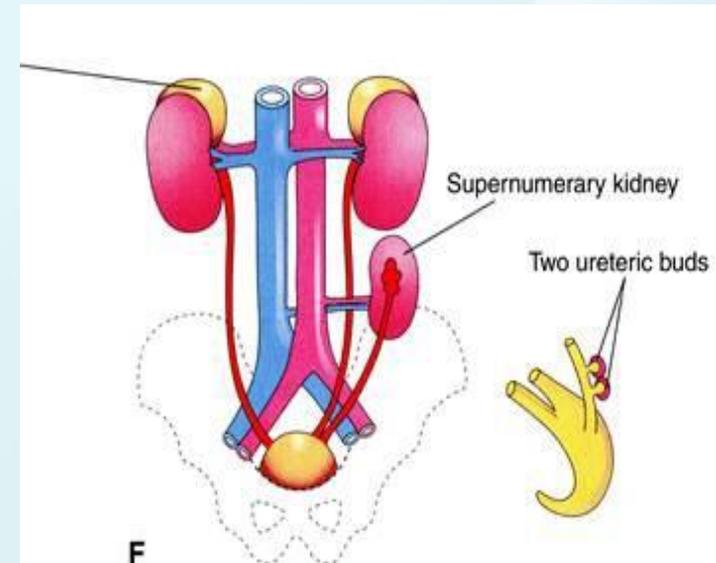
### ‣ Prognosis

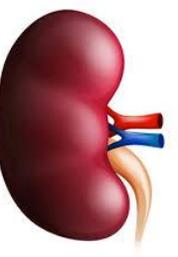
- 40% are stillborn.
- Do not survive beyond 48
  - hours due to respiratory distress associated with pulmonary hypoplasia.



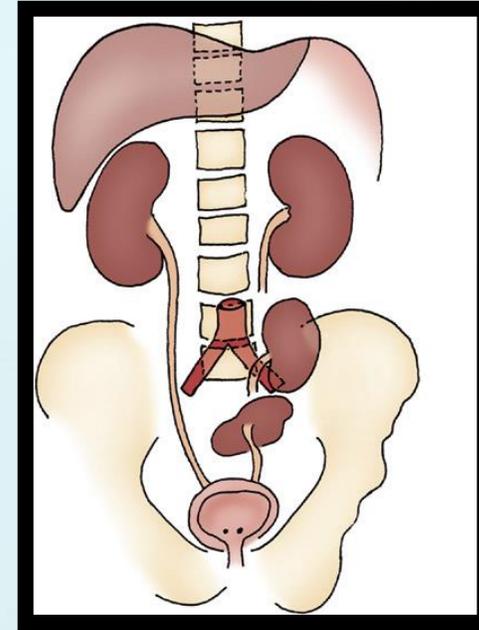
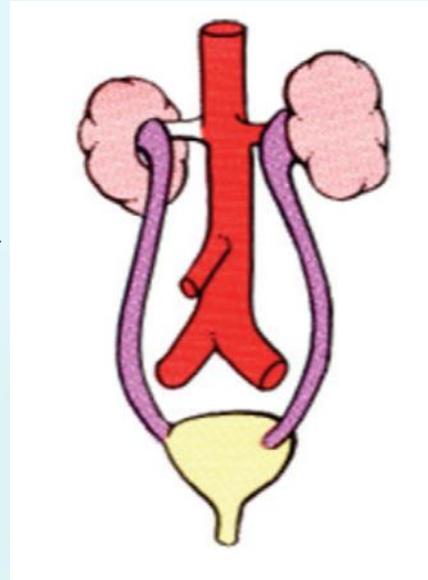
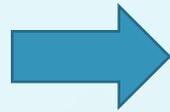
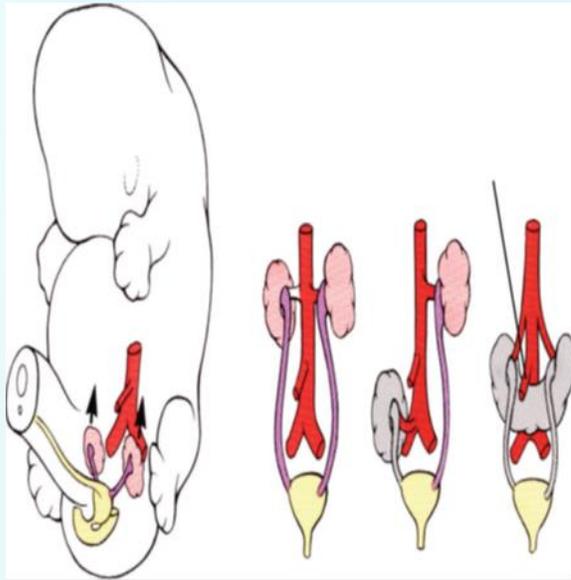
### 3- Supernumerary Kidney

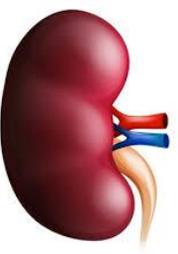
- Definitive **accessory** organ with its own collecting system, blood supply, and distinct encapsulated parenchyma.
- Either completely separate or loosely attached to the kidney on the ipsilateral side.
- The ureteral inter-relationships on the side of the supernumerary kidney can be variable.





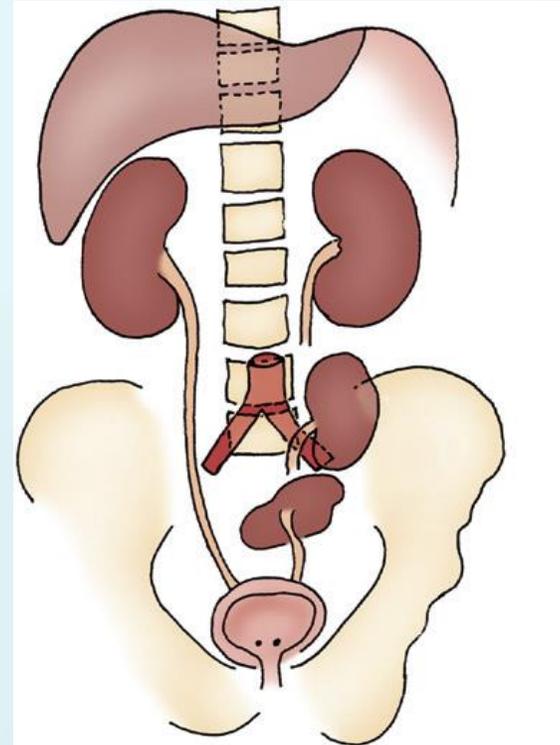
## 4- Simple Renal Ectopia

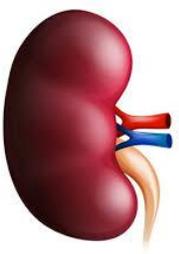




## 4- Simple Renal Ectopia

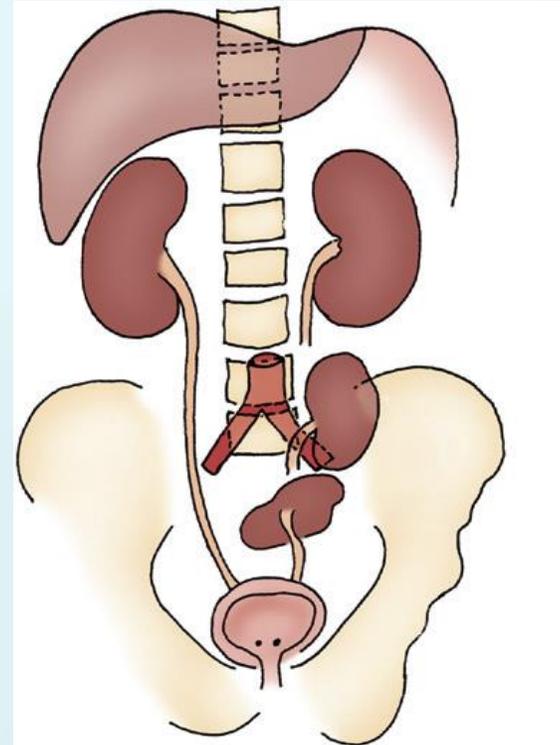
- **Left** more than the right.
- 1 of 2100 to 3000 autopsies.
- Most ectopic kidneys are clinically asymptomatic.

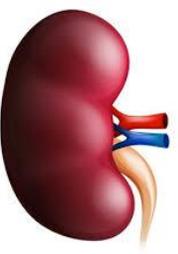




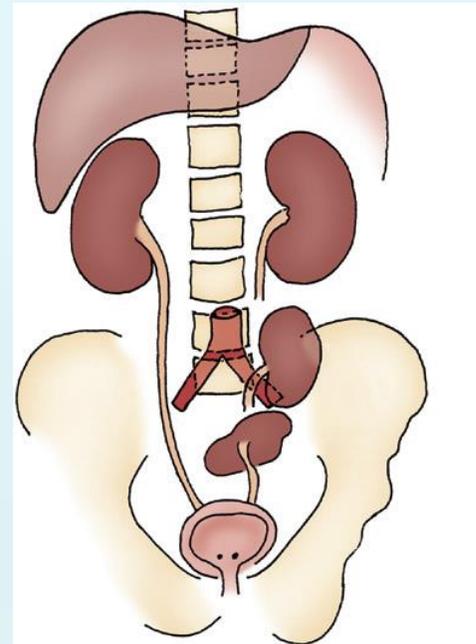
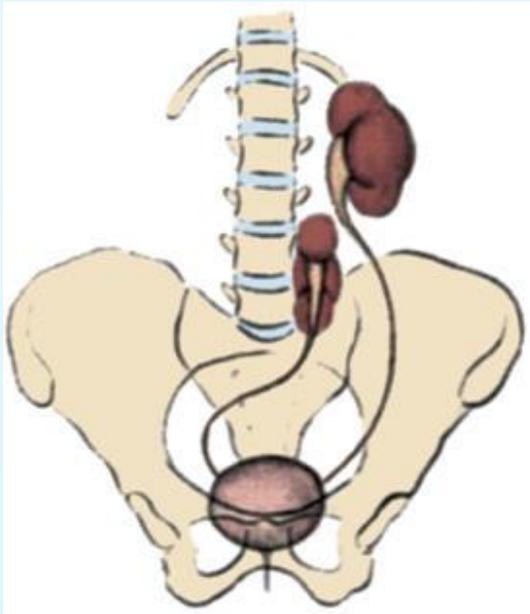
## 4- Simple Renal Ectopia...

- ▶ 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%.

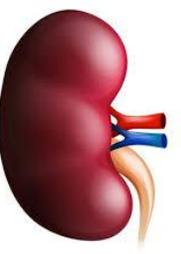




## 5- Crossed Renal Ectopia

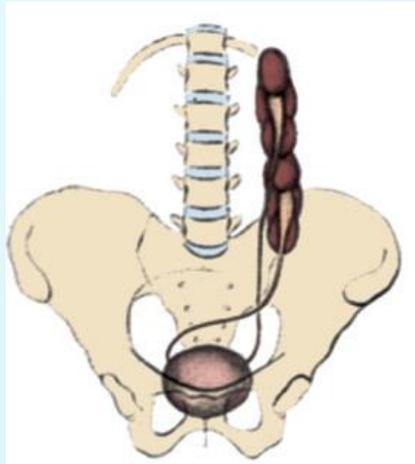


**Crossed ectopia:** kidney is located on the side opposite from that in which its ureter inserts into the bladder. The ureter from each kidney is usually orthotopic.

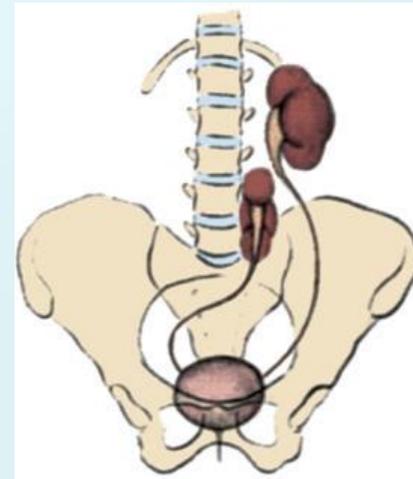


## Crossed Renal Ectopia...

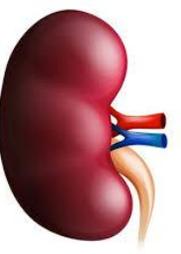
**with Fusion**



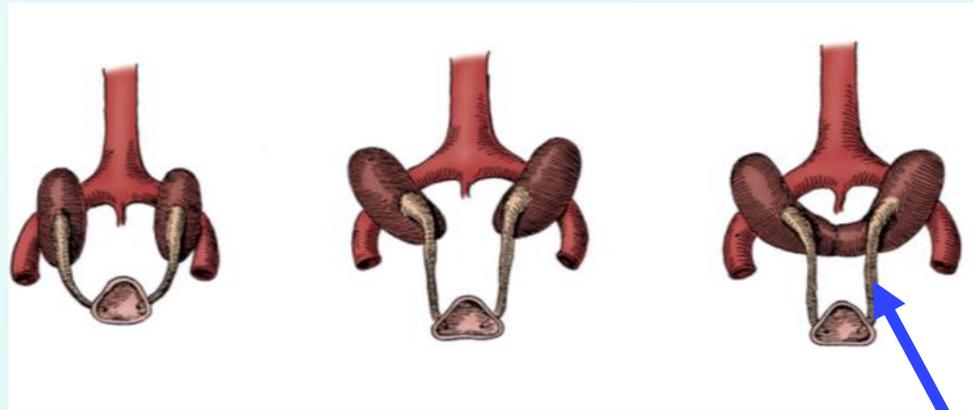
**without Fusion**



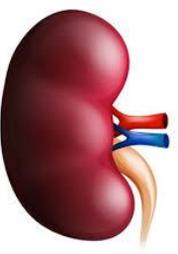
90% are fused  
the superior pole of the ectopic kidney usually joins with the inferior aspect of the normal kidney.



## 6- Horseshoe Kidney



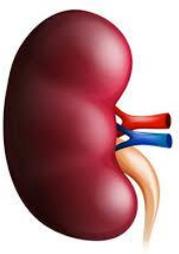
The **isthmus** is bulky and consists of parenchymatous tissue.



## Horseshoe Kidney

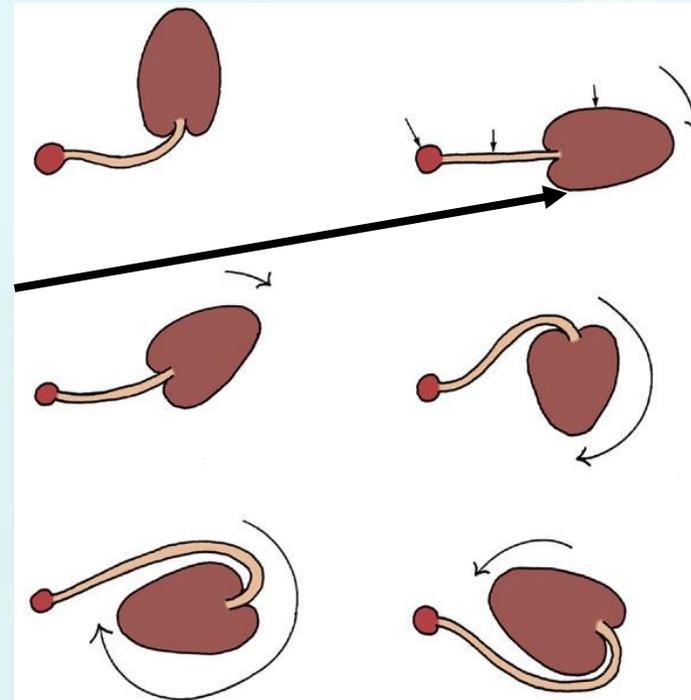
- Occurs 1 in 400 persons.
- The calyces:
  - ✓ normal in number
  - ✓ atypical in orientation.
  - ✓ pelvis remains in the vertical or obliquely lateral plane
- Horseshoe kidney is frequently found in association with other congenital anomalies.
  - UPJ obstruction in one third.
- 60 % asymptomatic.



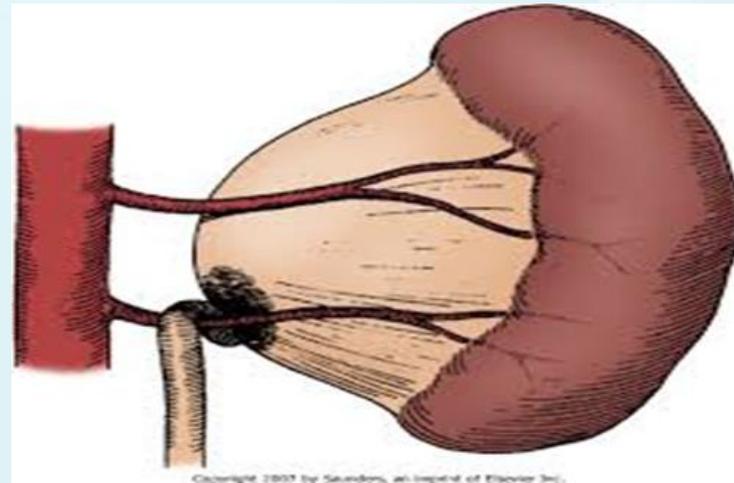
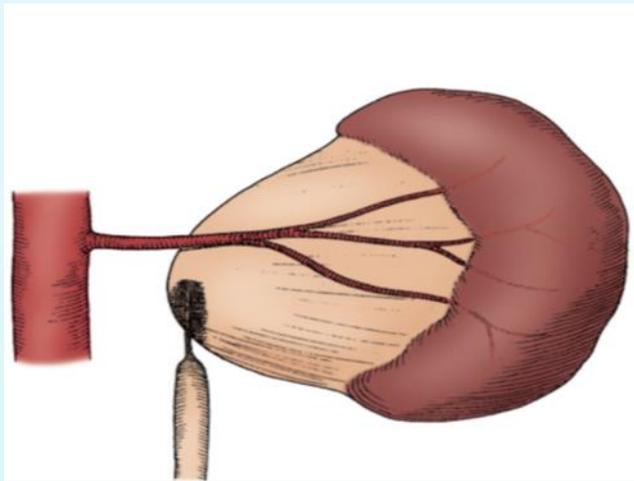


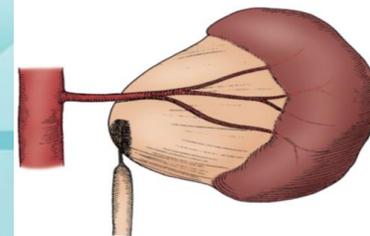
## 7- Anomalies of Rotation

- ▶ 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

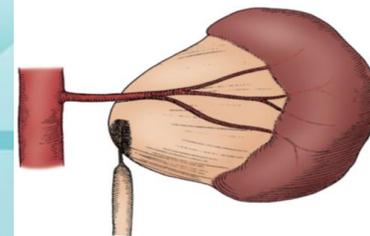




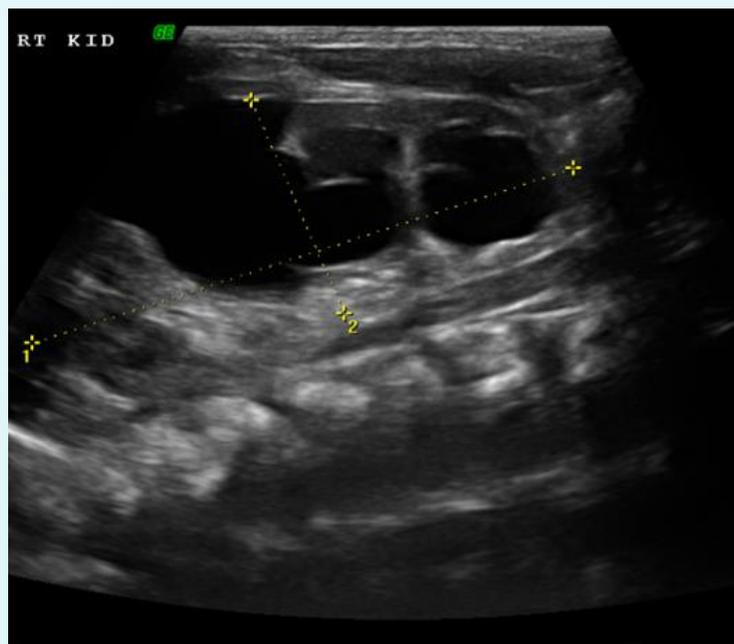
## UPJ...

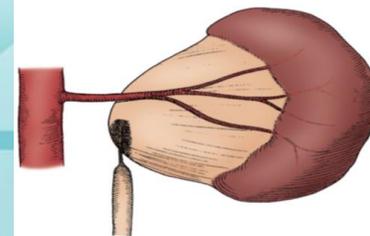
### Presentation:

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



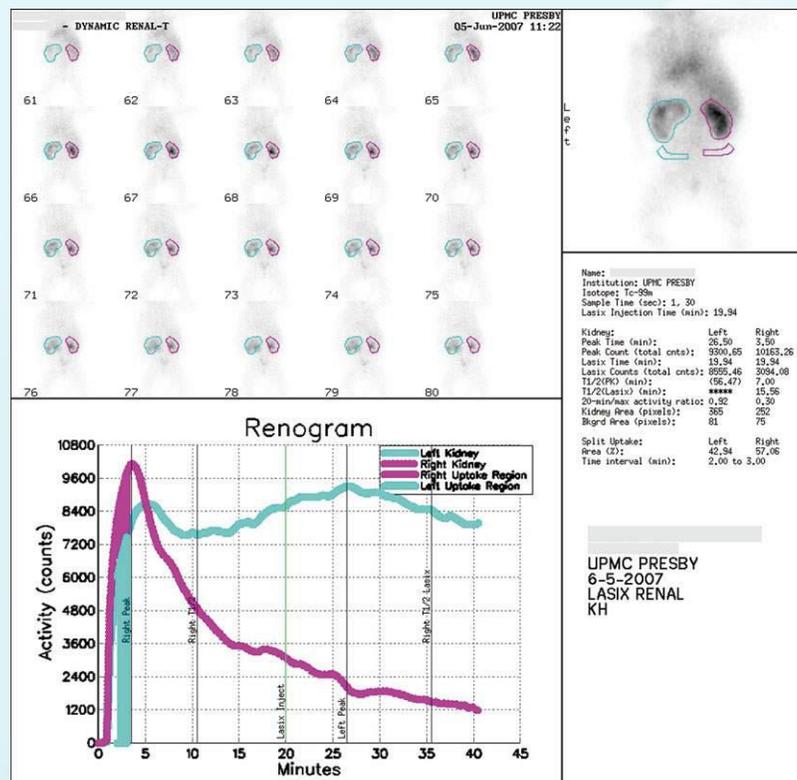
## UPJO...

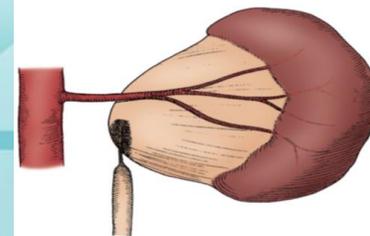




# UPJO...

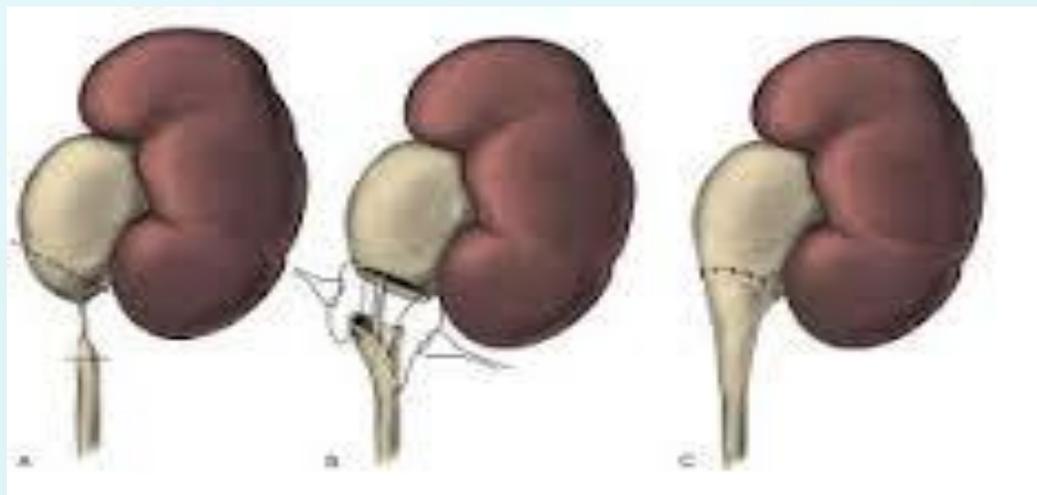
## ➤ Dynamic renogram



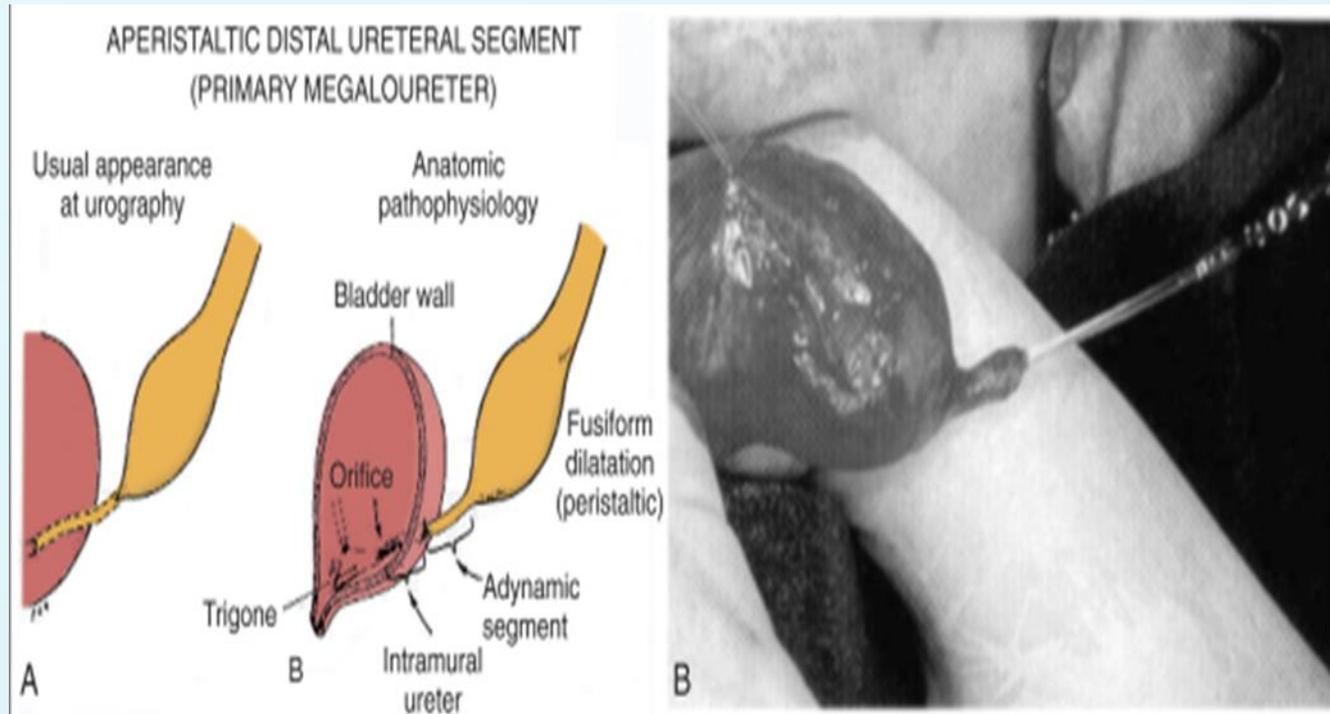


## UPJO...

- ▶ **Dismembered Pyeloplasty**



## 9-Ureterovesical junction (UVJ) obstruction (Megaureters)



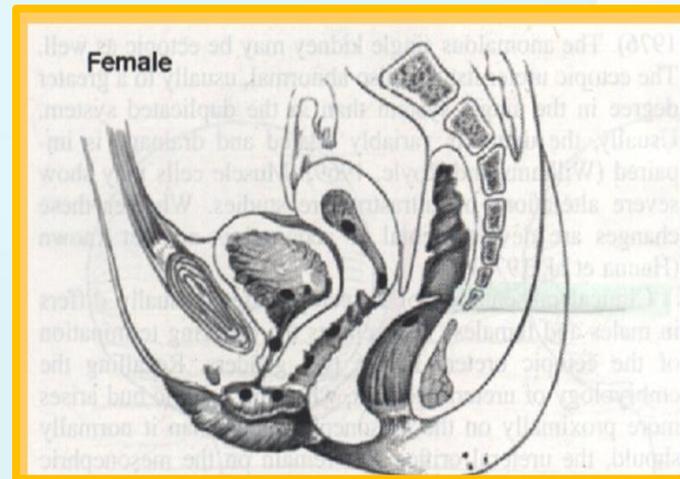
## 11- Ectopic Ureter

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



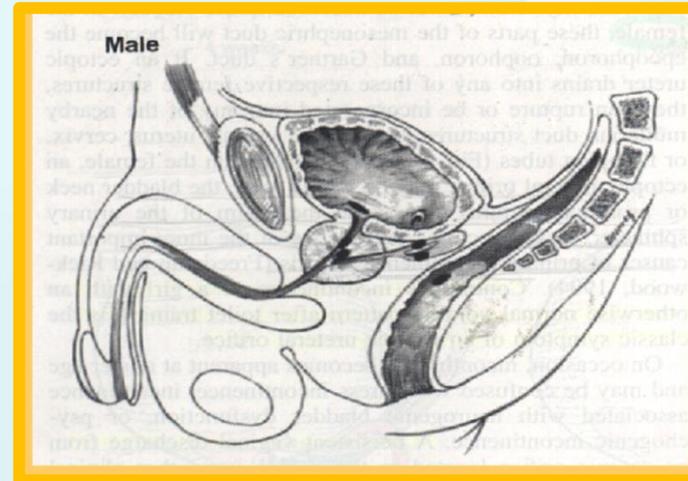
## Ectopic Ureter...

- ▶ 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**.



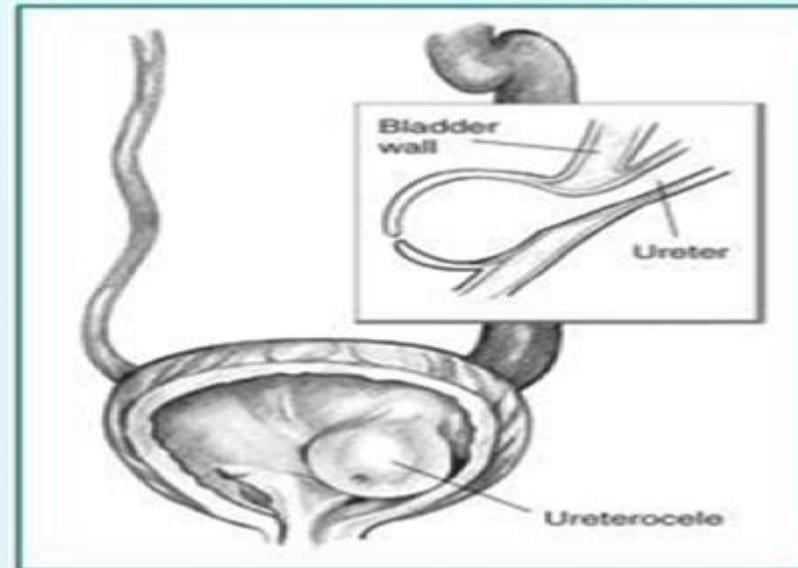
## Ectopic Ureter

- ▶ 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.

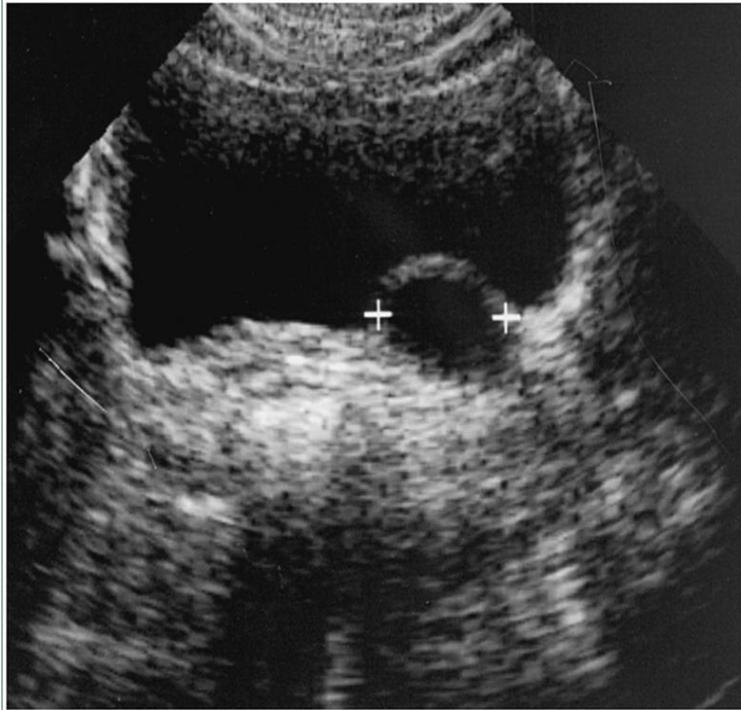


## 12- Ureterocele

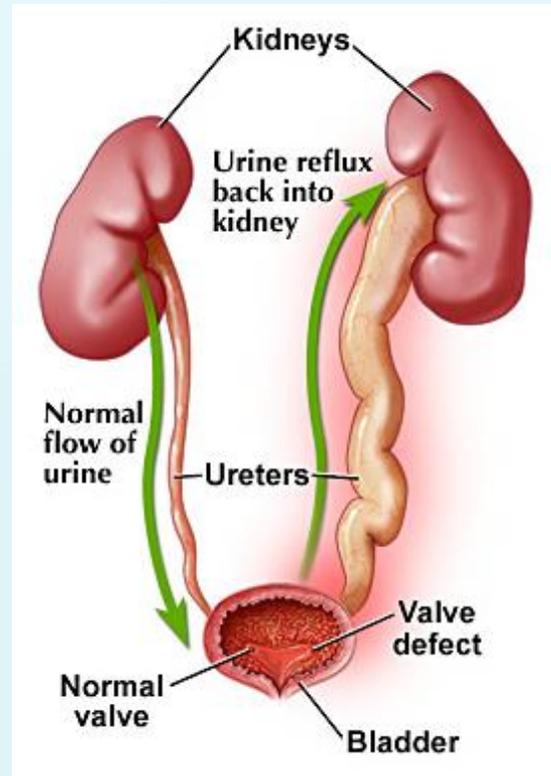
- A cystic dilation of the distal aspect of the ureter
- Located either within the bladder or spanning the bladder neck and urethra.
- **Presentation:**
  - Antenatal (U/S)
  - Urine retention
  - Infection
  - Calculus formation



## Ureterocele...



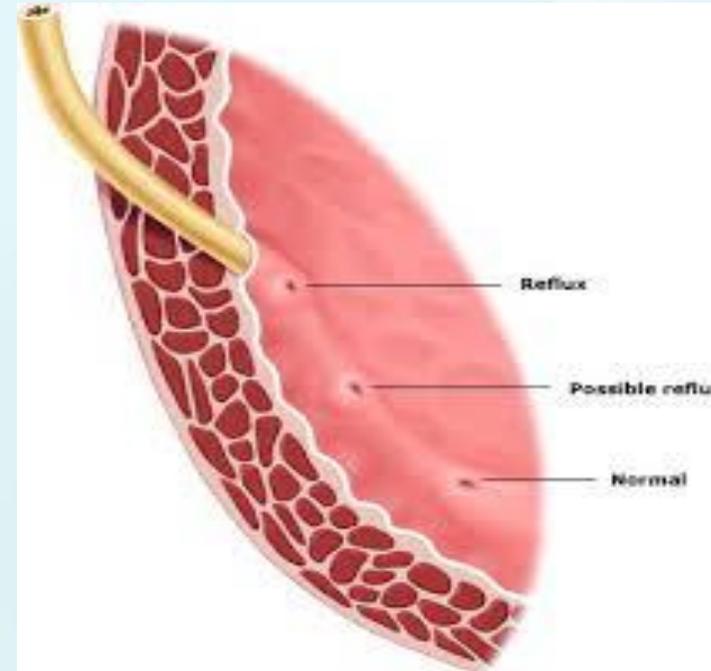
## 13-Vesicoureteral Reflex (VUR)



## 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.



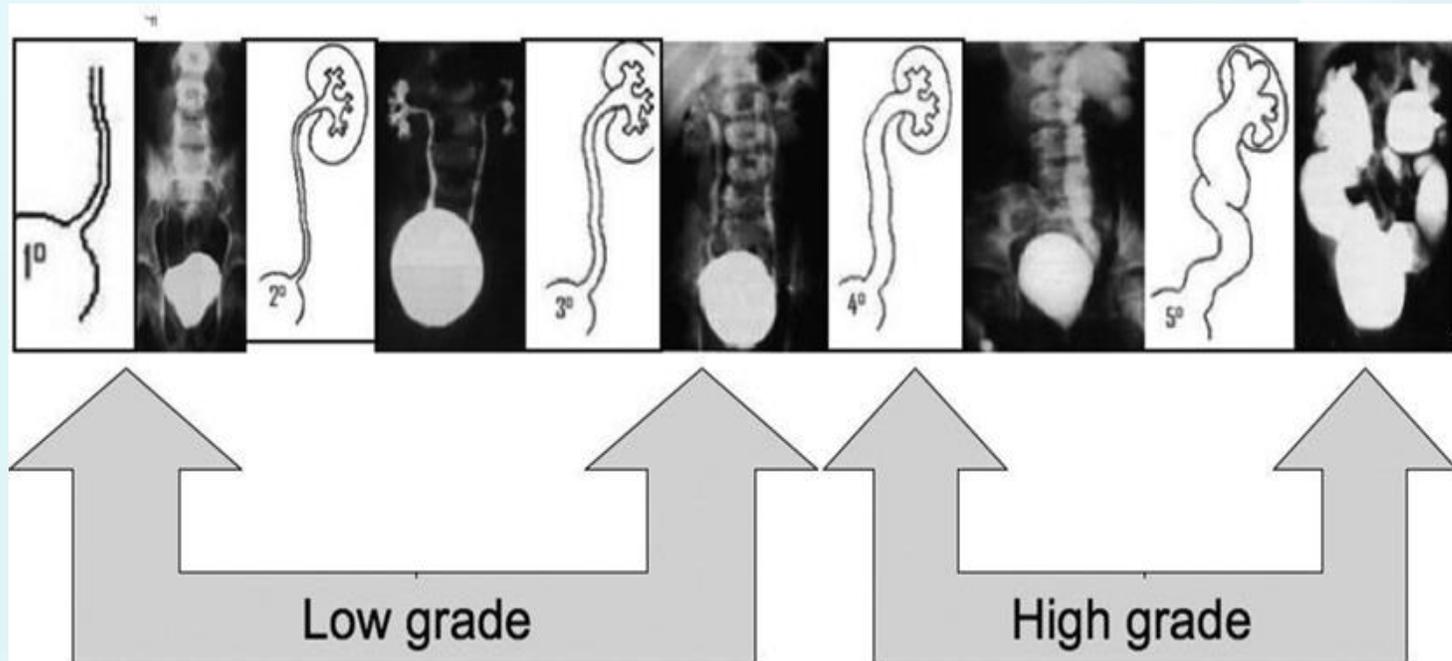
## **VUR...**

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

## VUR...

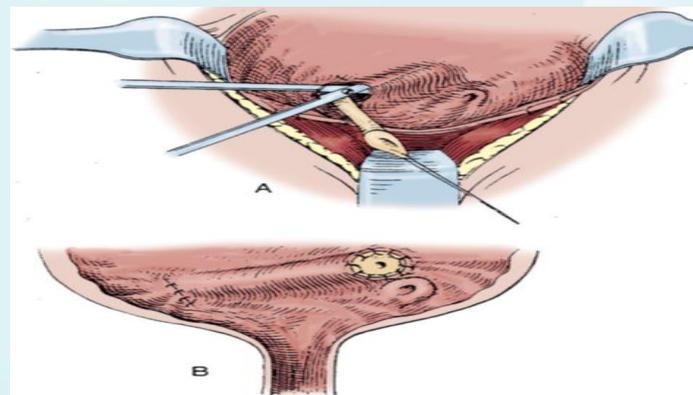
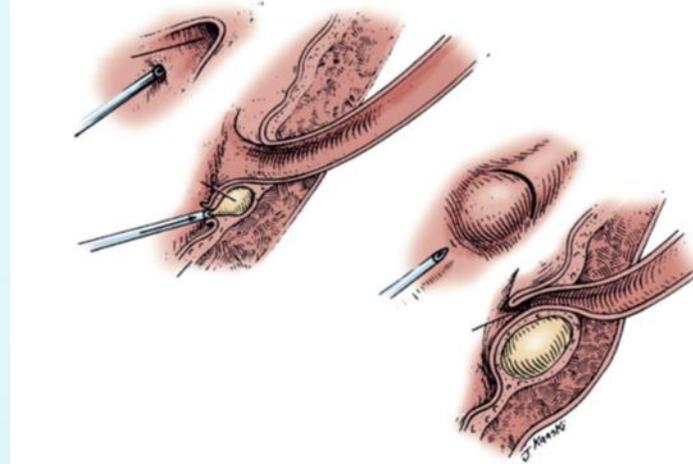
### ▷ Diagnosis:

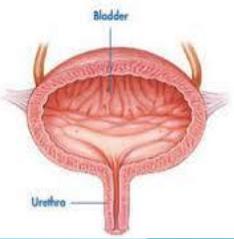
#### ▷ MCUG (VCUG)



## VUR...

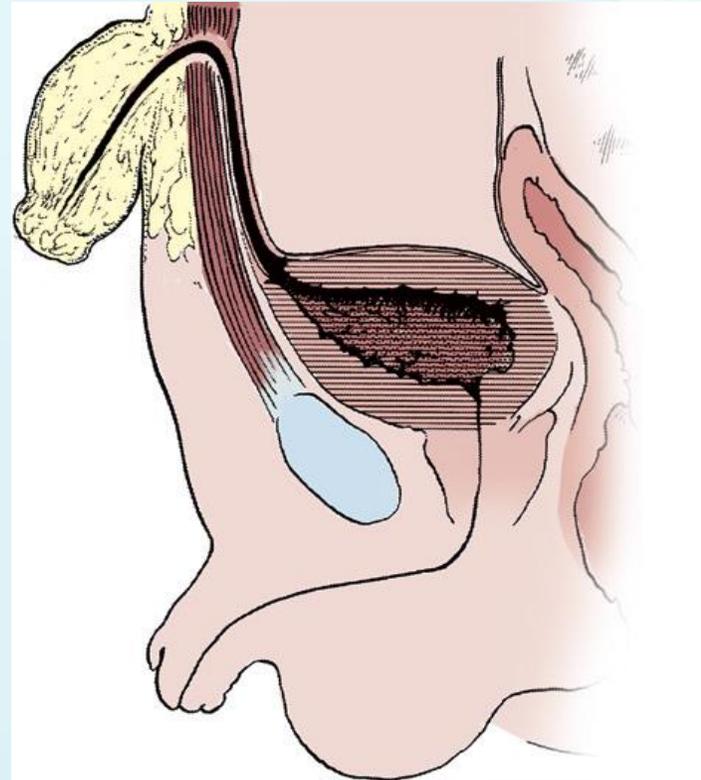
- **Management:**
- Prophylactic antibiotic
- Surgical treatment
  - Endoscopic treatment
  - Ureteral reimplantation

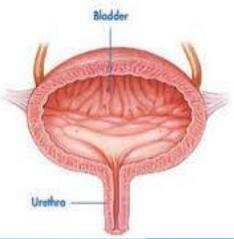




## 14-Urachal abnormalities

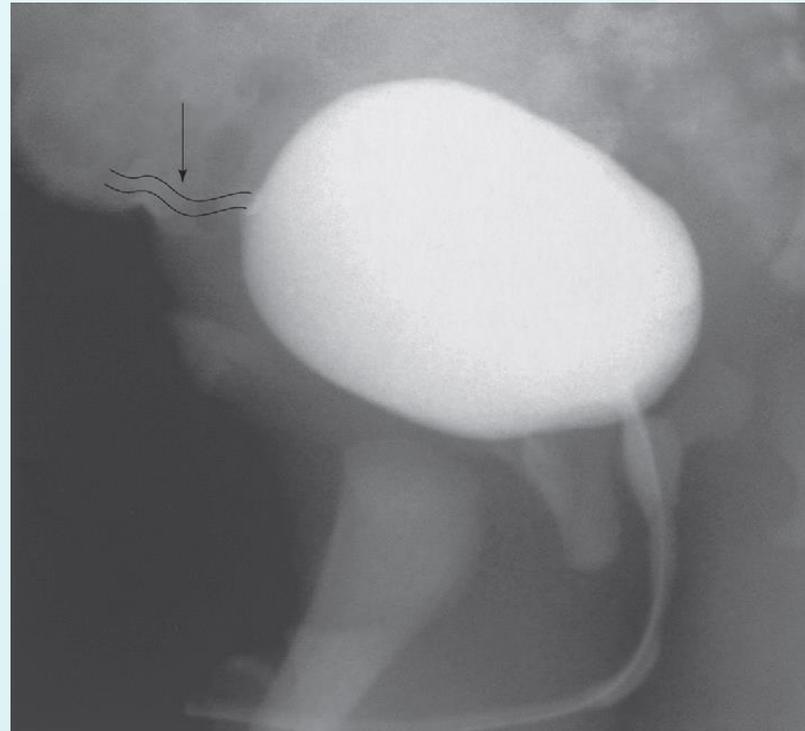
- ▶ Urachal anomalies are usually detected postnatally due to umbilical drainage.
- ▶ Imaging possibilities include ultrasound, CT, and VCUG.

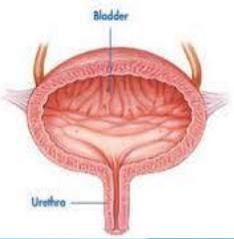




## 14-Urachal abnormalities...

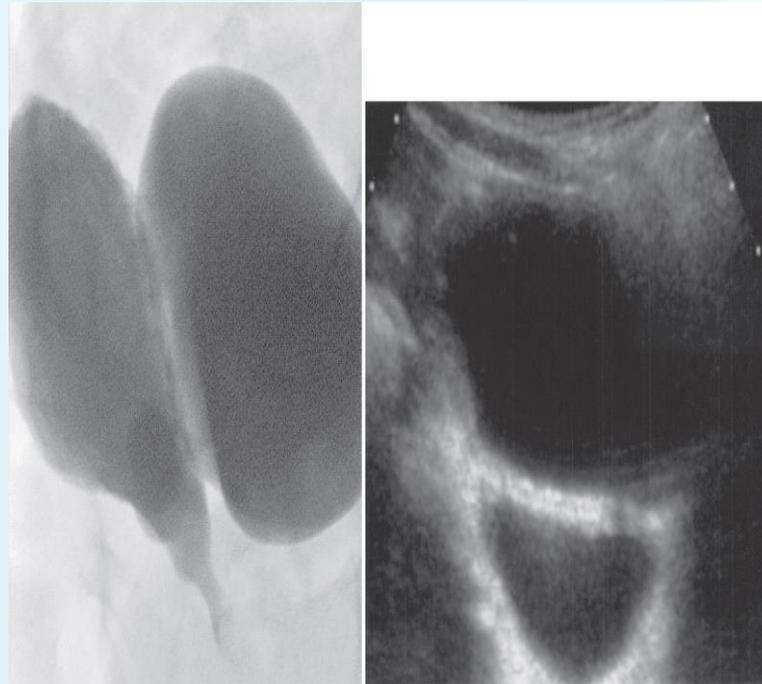
- ▶ **Conservative** treatment with observation is justified in asymptomatic cases due to possible spontaneous resolution
- ▶ **Infected urachal** remnants are initially treated with drainage and antibiotics, followed by surgical excision.
- ▶ Nonresolved urachal remnants should be excised due to the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

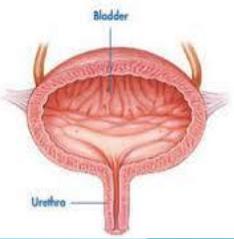




## 15-Bladder Diverticulum

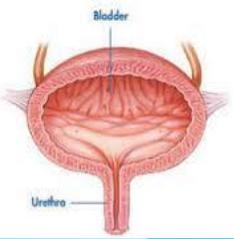
- ▶ Bladder diverticula can be detected on prenatal ultrasound, but the gold standard remains VCUG, which will reveal possible accompanying VUR.





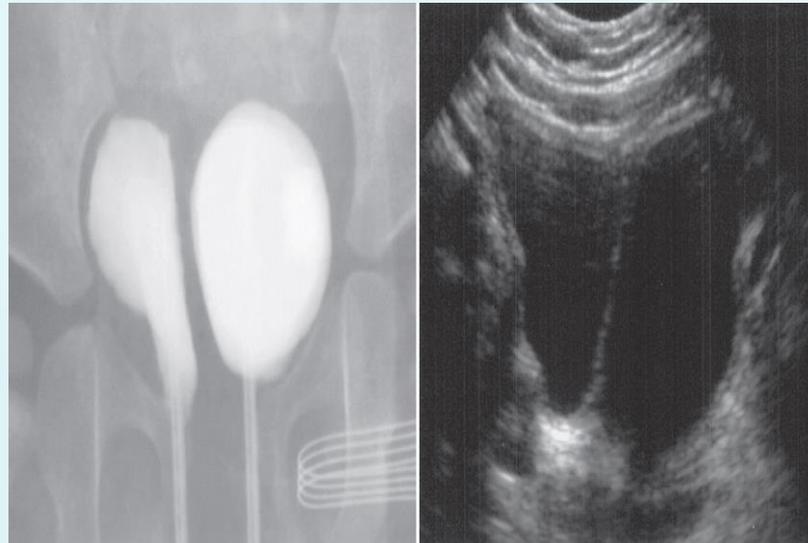
## Bladder Diverticulum...

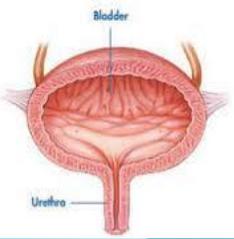
- Types:
  - Primary diverticula
    - arise as a localized herniation of **bladder mucosa** at the ureteral hiatus and are most likely caused by a **congenitally** deficient bladder wall.
  - Secondary para-ureteral diverticula
    - are acquired and develop due to existing infra- vesical obstruction.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



## 16- Bladder Duplication

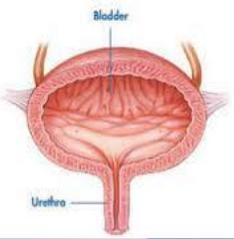
- ▶ Often associated with duplication anomalies of the external genitalia and lower gastrointestinal tract.
- ▶ Initial treatment is directed toward
  - ▶ renal preservation.
  - ▶ prevention of infections.



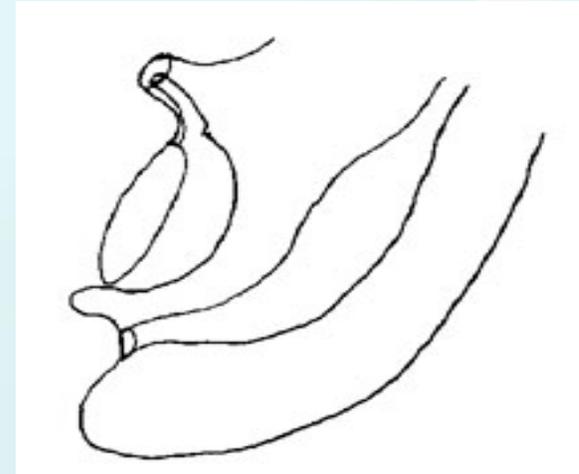
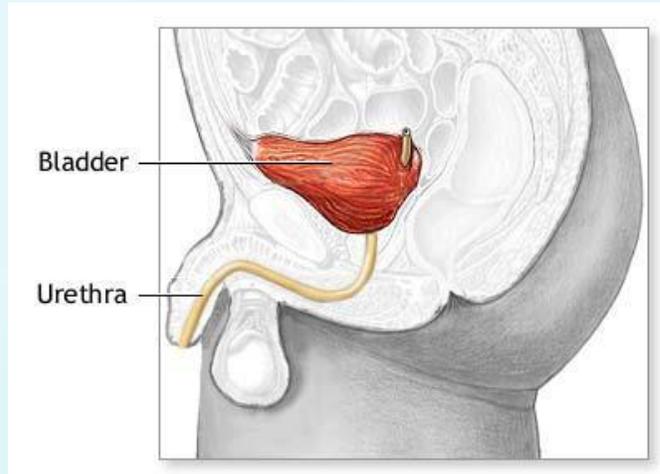


## Bladder Duplication...

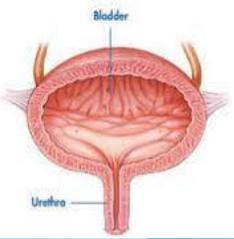
- ▶ Long-term goals include achieving continence and reconstructing the internal and external genitalia.
- ▶ Due to the rarity of the disease and the large variety of presentations, the surgeries must be individualized



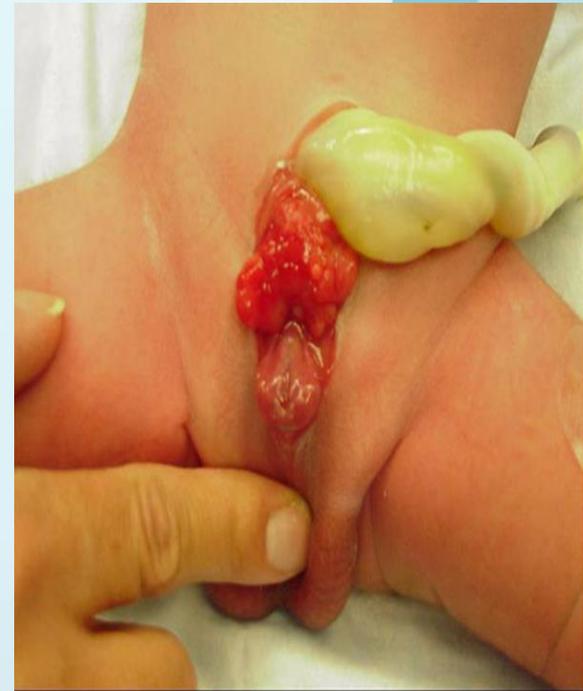
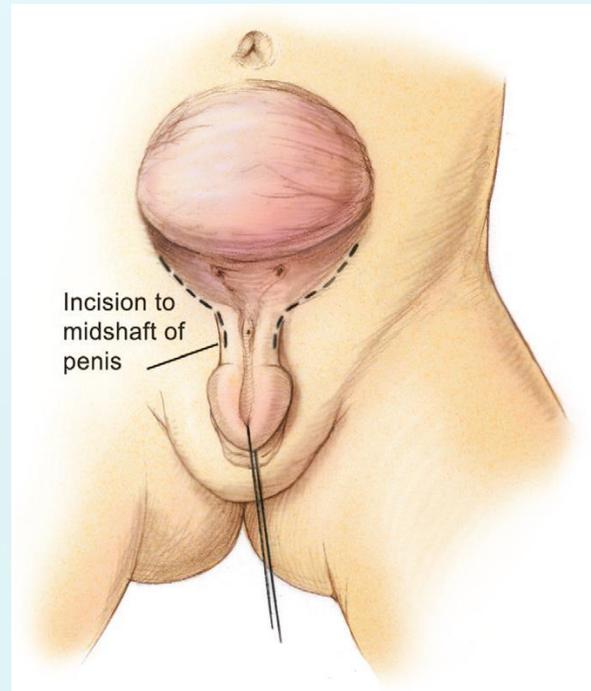
## 17- Bladder Exstrophy



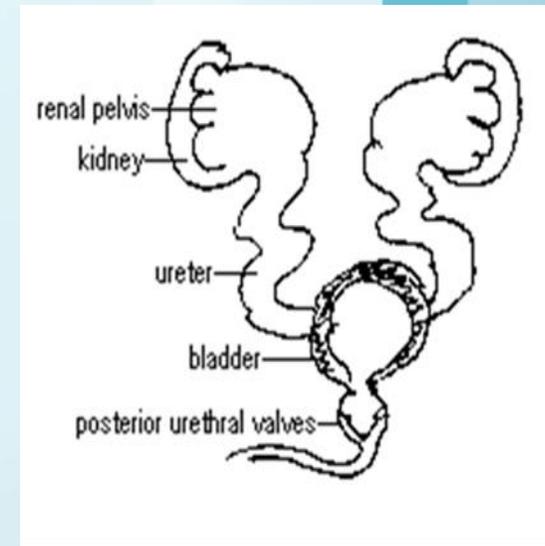
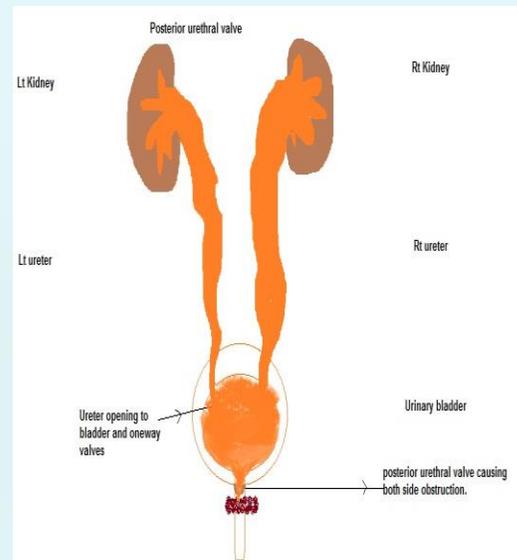
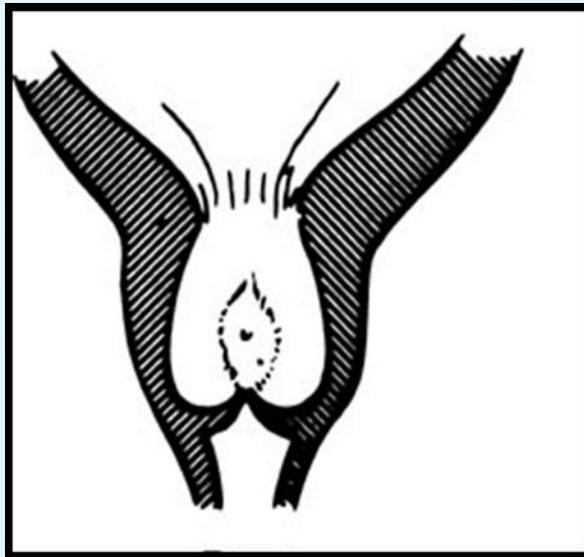
The incidence of bladder exstrophy has been estimated as between 1 in 10,000 and 1 in 50,000.



## Bladder Exstrophy...



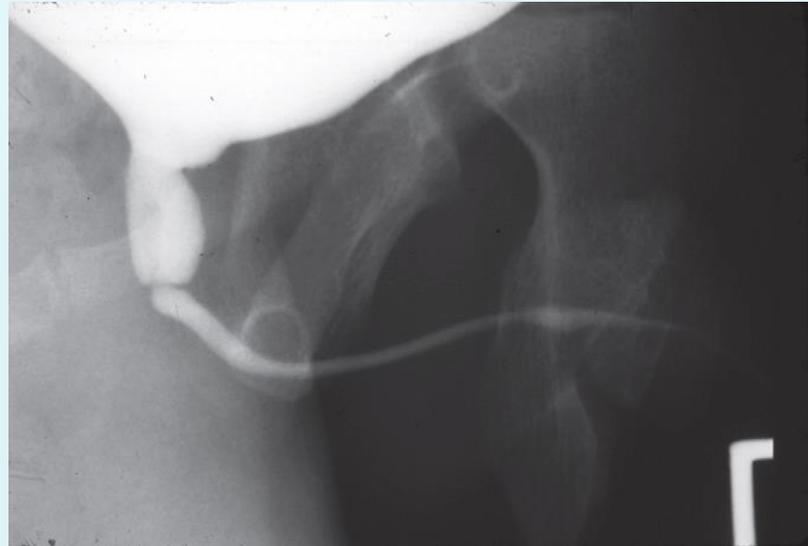
## 18-Posterior Urethral Valves (PUV)



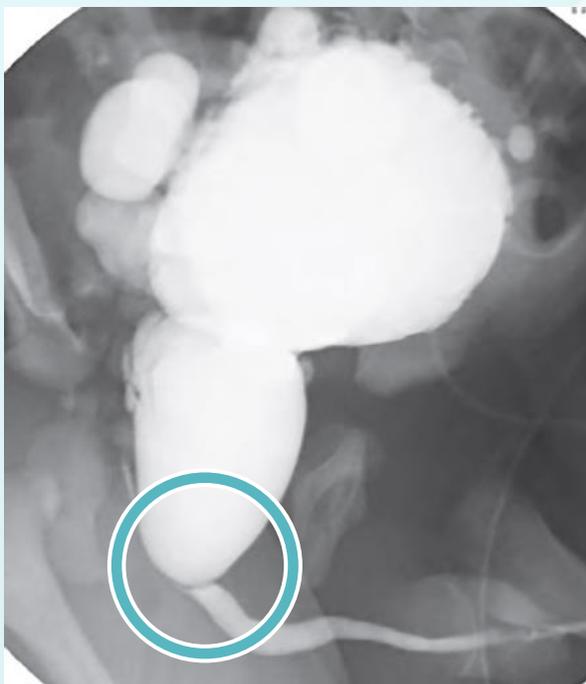
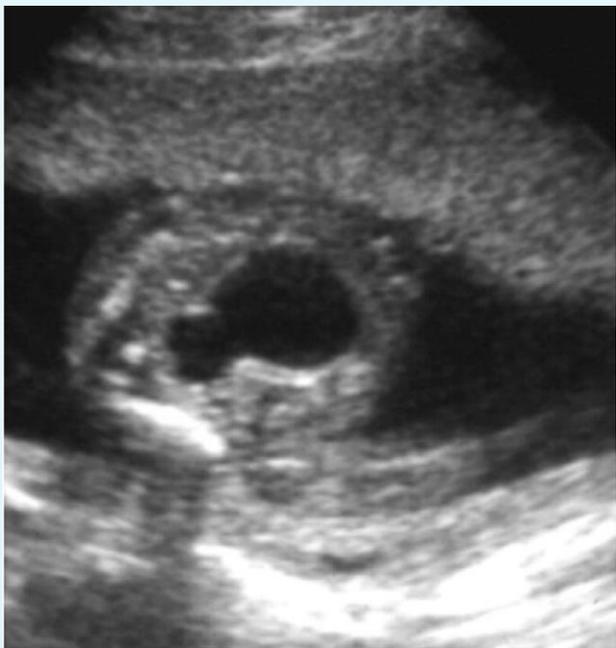
The bladder and the kidneys developed under high pressure and resistance.

## 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)

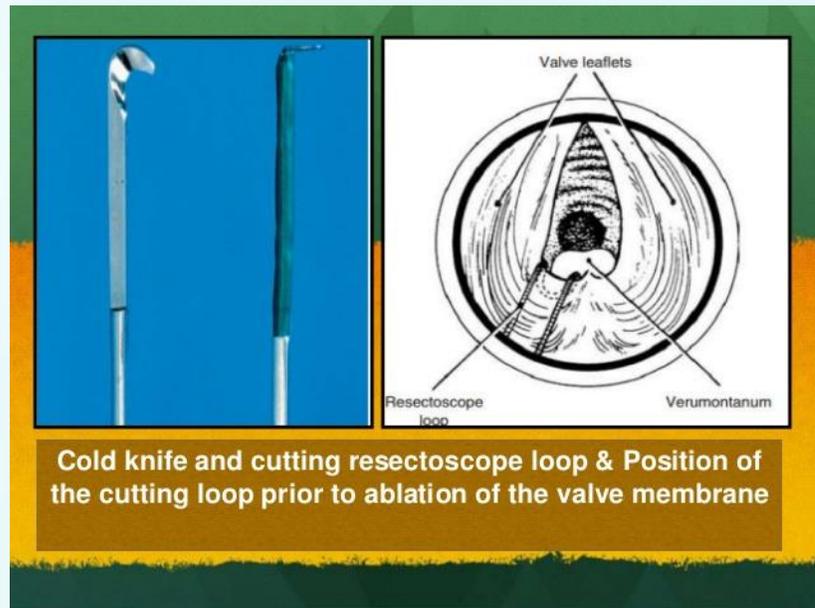
### • **Associated findings:**

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...



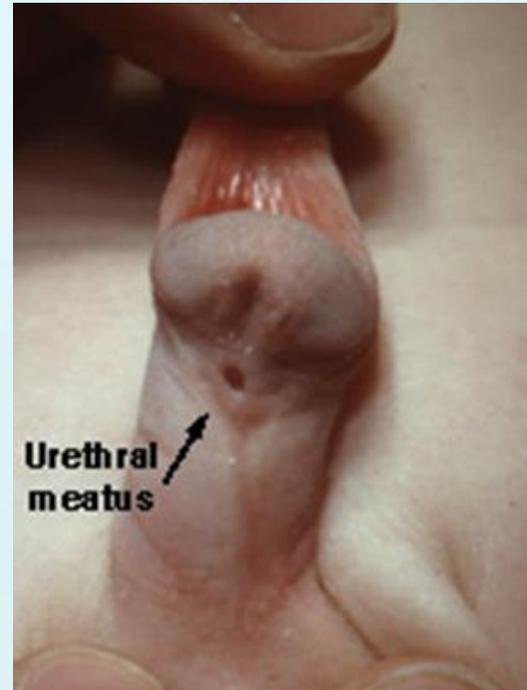
Endoscopic valve ablation



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



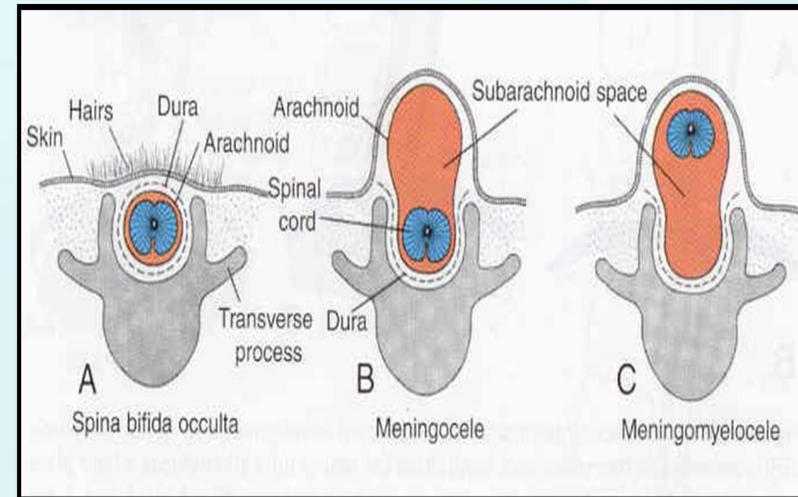
## 22- Prune-Belly Syndrome

- 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.

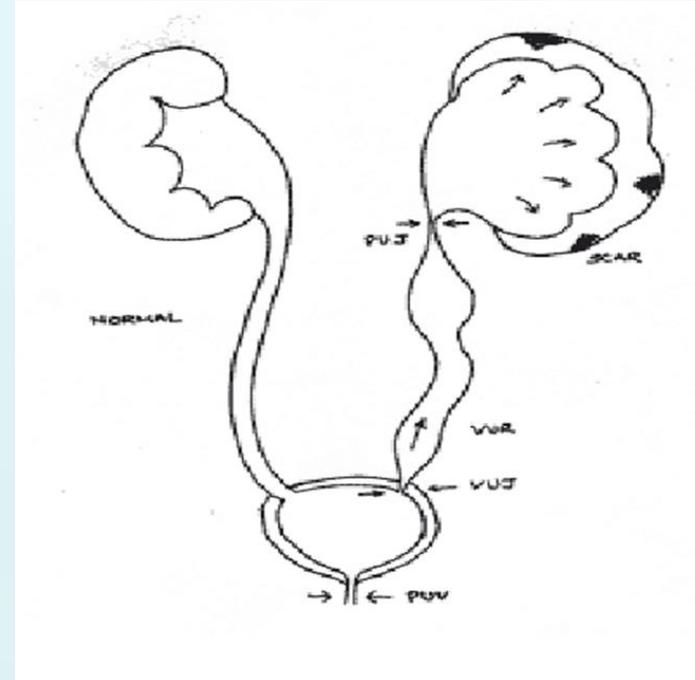


## **Antenatal Hydronephrosis(ANH)**

# 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!

