GENITOURINARY ANOMALIES

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Lecture Objectives

- 13. GU anomalies (pathophysiology, etiology, clinical manifestations, complications, Guideline of managements)

  - 13.1. Discuss congenital anomalies of **kidney** (K/C)
  - 13.2. Discuss congenital anomalies of **ureter** (K/C)
  - 13.3. Discuss congenital anomalies of **bladder** (K/C)
  - 13.4. Discuss congenital anomalies of **urethra** (K/C)
  - 13.5. Recognize congenital anomalies of **genitalia** (K/C)

K- Knowledge C- Cognitive
P-Psychomotor Co-Communication

More common

1. Ureteropelvic junction obstructiion (UPJO)
2. Multicystic dysplatic kidney (MCDK)
3. Vesicoureteral Reflux (VUR)
4. Posterior Urethral Valve (PUV)
5. Ureterovesical Junction Obstruction (UVJO)
6. Hypospadias

Less common

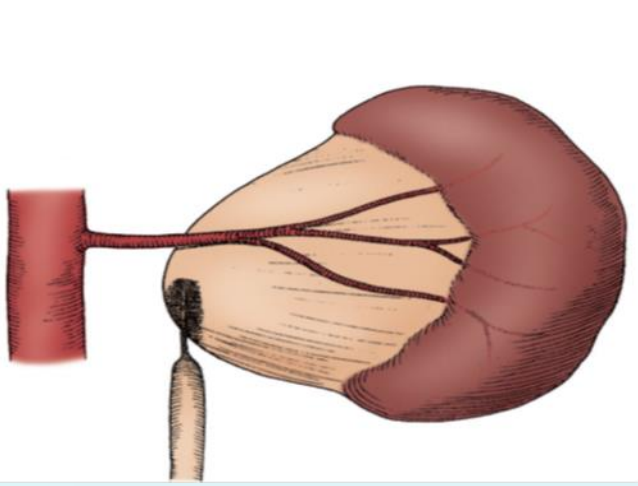
1. Unilateral Renal Agenesis
2. Ureterocele
3. Duplication Anomalies
4. Ectopic ureter
5. Horseshoe Kidney
6. Crossed Renal Ectopia
7. Bladder Diverticulum
8. Prune -Belly Syndrome
9. Epispadias
10. Bladder Exstrophy

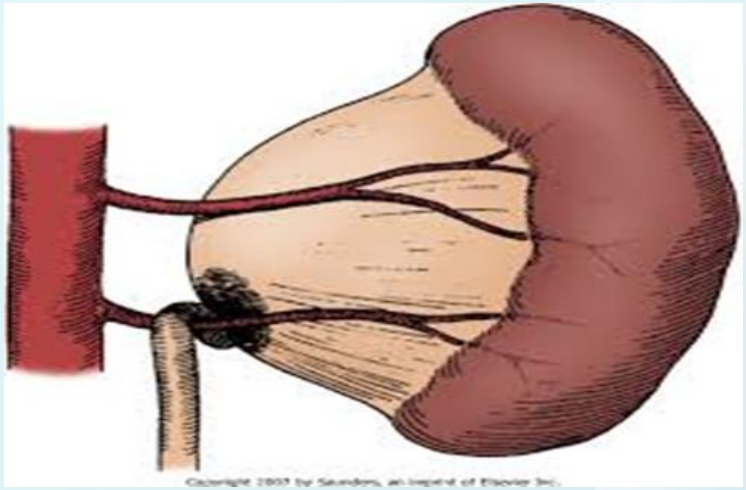
Uncommon

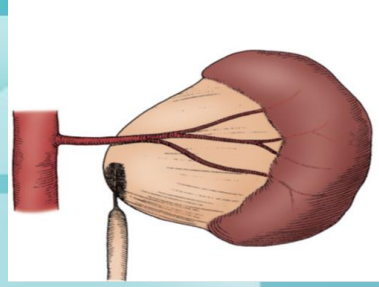
1. Bilateral Renal Agenesis
2. Supernumerary Kidney
3. Anomalies of rotation
4. Bladder duplication
5. Cloacal exstrophy
6. Urachal abnormalities
7. Neurospinal dysraphism

UPJO

Ureteropelvic junction (UPJ) obstruction



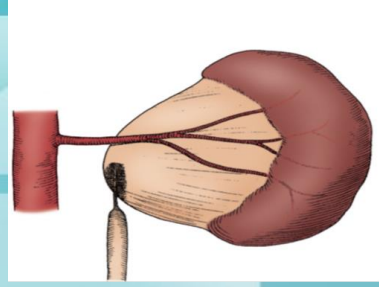




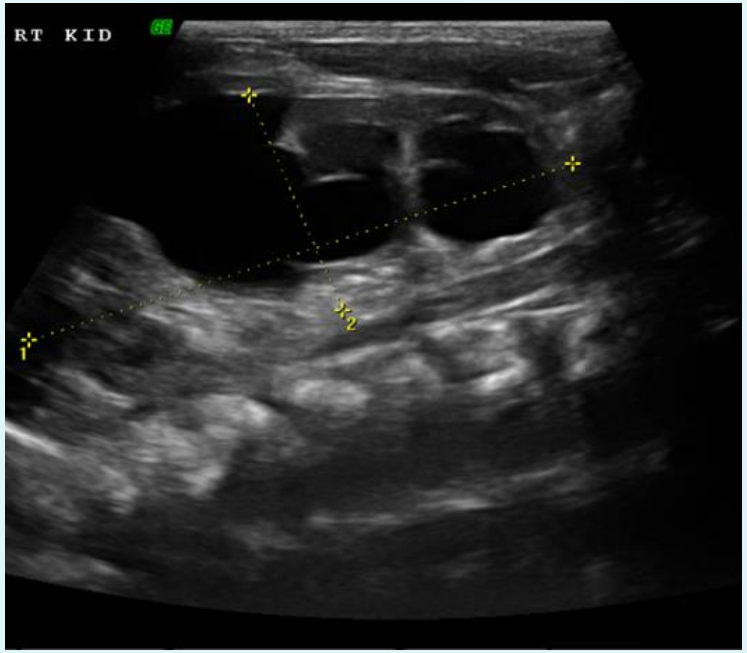
UPJO…

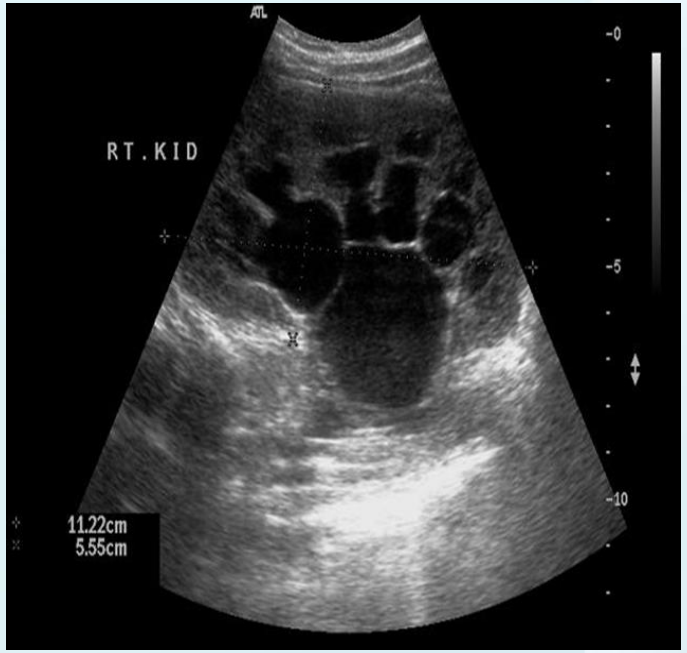
Presentation:

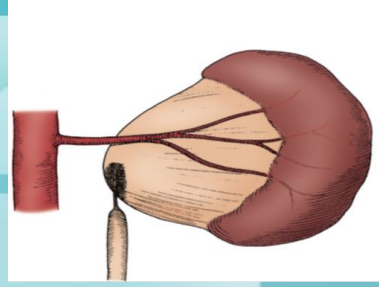
- Prenatal US
- Incidental in Neonates/Children
- Symptomatic:
  - ✓ UTI
  - ✓ Pain
  - ✓ Mass
  - ✓ Hematuria
  - ✓ Stone



UPJO…

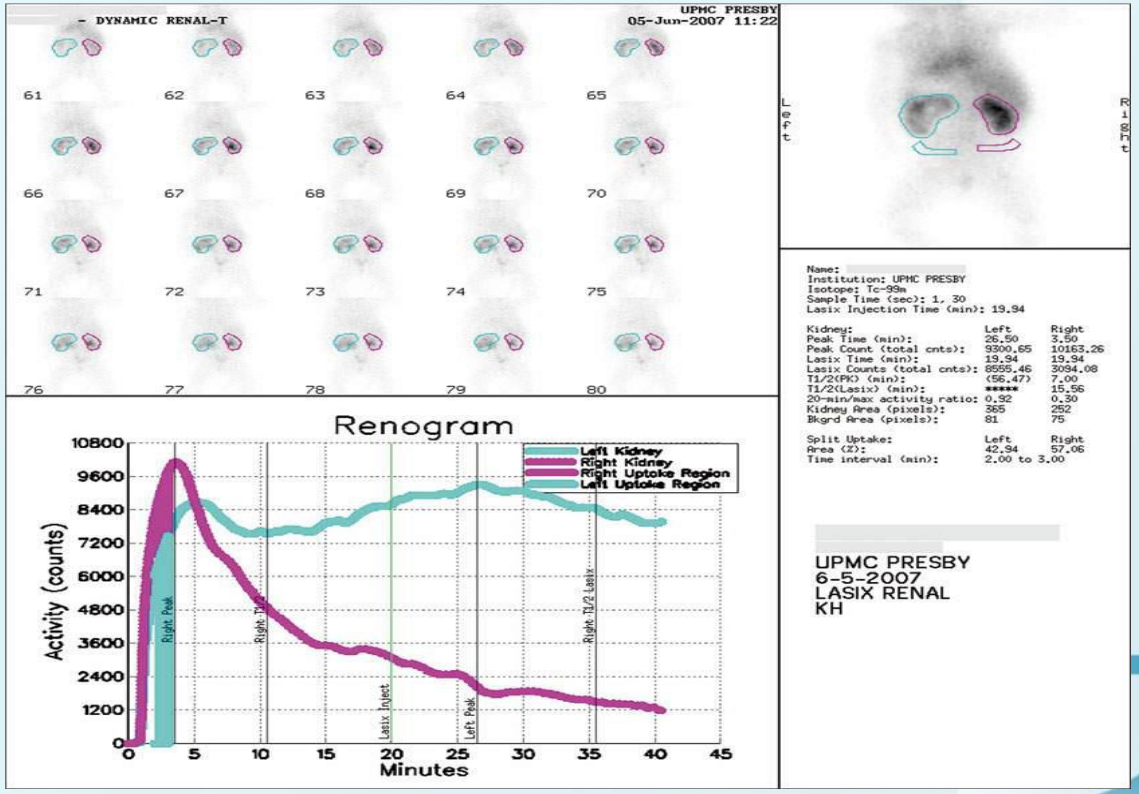






UPJO…

- **Dynamic renogram**
  - Washout curve
  - Function



Management:

A. Observation
   - ❑ Spontaneous resolution

B. Surgerical intervention
   1. Worsoning hydronephrosis
   2. Renal function
      - ❑ less than 40%
      - ❑ deterioration more than 10%
   3. Pyelonephritis
   4. Stone formation

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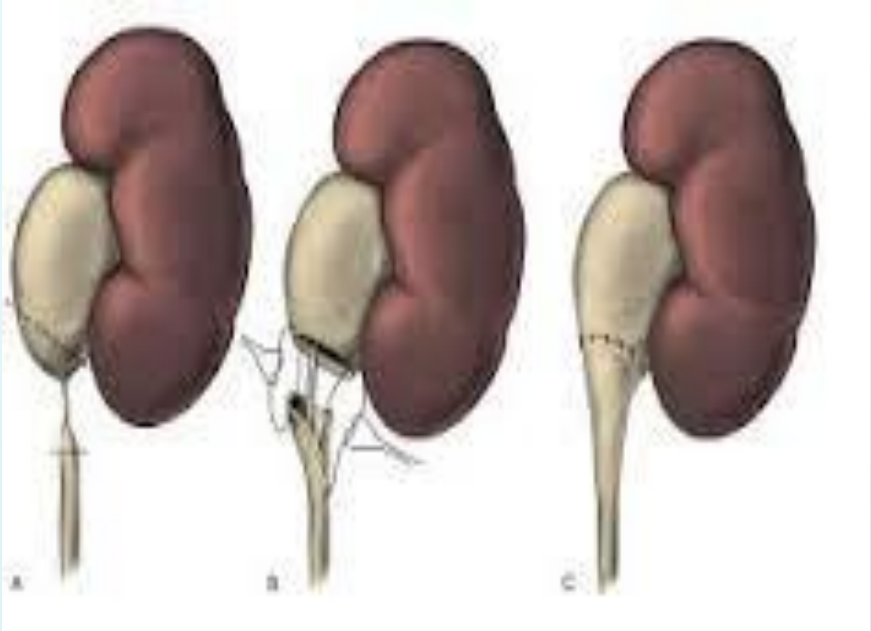
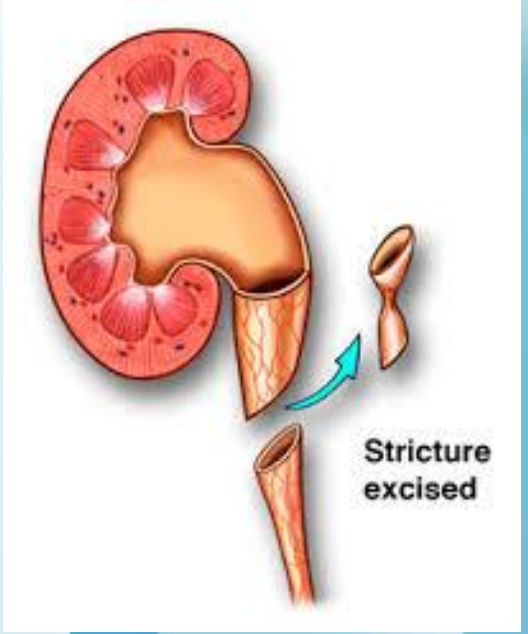
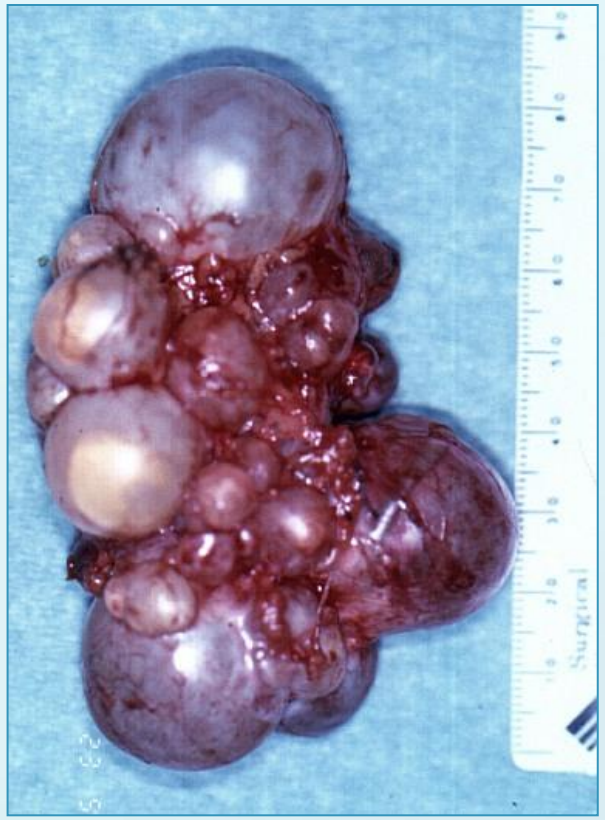


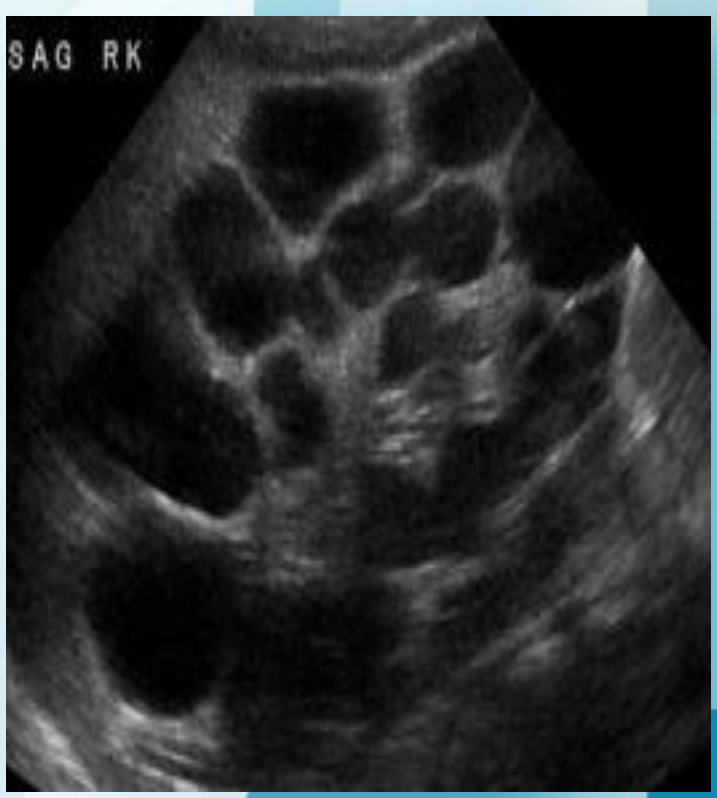


Dismembered Pyeloplasty

Multicystic dysplatic kidney (MCDK)

- Unilateral
- The kidney non functioning
- Ultrasound:
  - Very thin and abnormal renal parenchyma, surrounded by multiple cysts of various sizes that do not connect, nor they connect to the renal pelvis.





MCDK

Diagnosis:

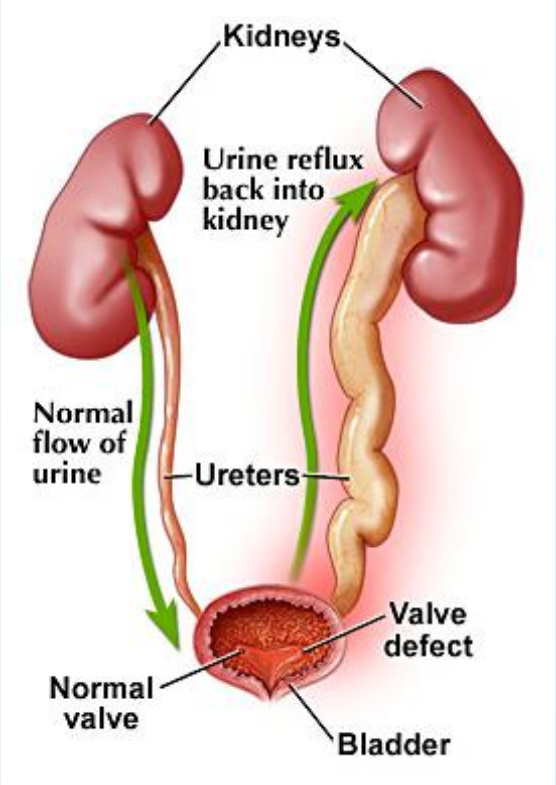
- Prenatal US
- Incidental in Neonates/Children
- Symptomatic:
  - Mass
  - UTI
  - Pain
- DMSA
  - Little or no uptake of radionuclide
- MCUG/ VCUG
  - Contralateral VUR
  - 18%-43%

Management:

A. Observation
   - ❑ Cyst fluid disappears
B. Surgerical intervention
   1. Hypertension
   2. Pain
   3. Pyelonephritis

Nephrectomy

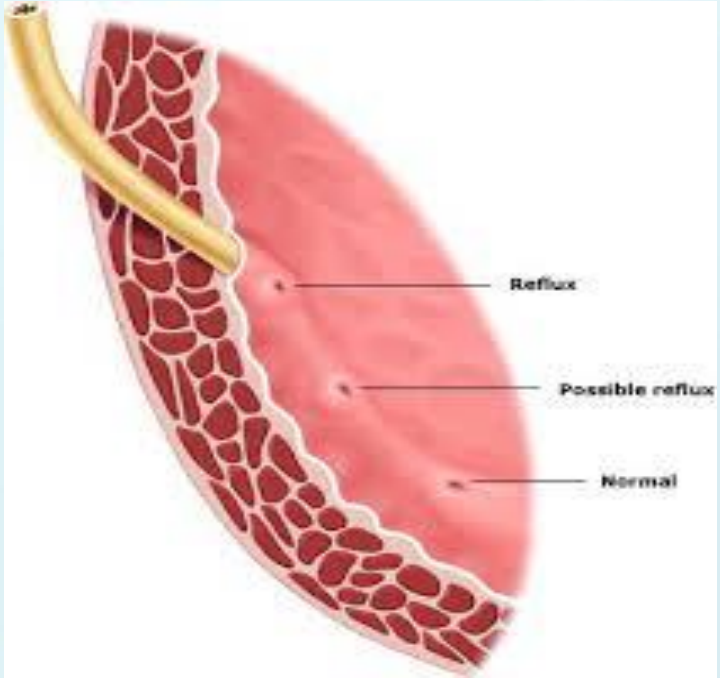
Vesicoureteral Reflux (VUR)



Vesicoureteral Reflux (VUR)…

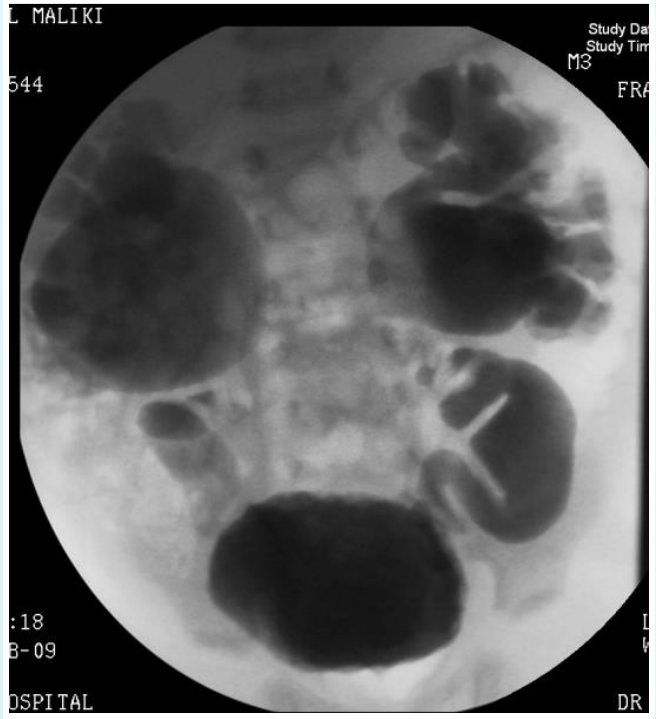
Normal anti-reflux mechanism
"Flap valve"

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.



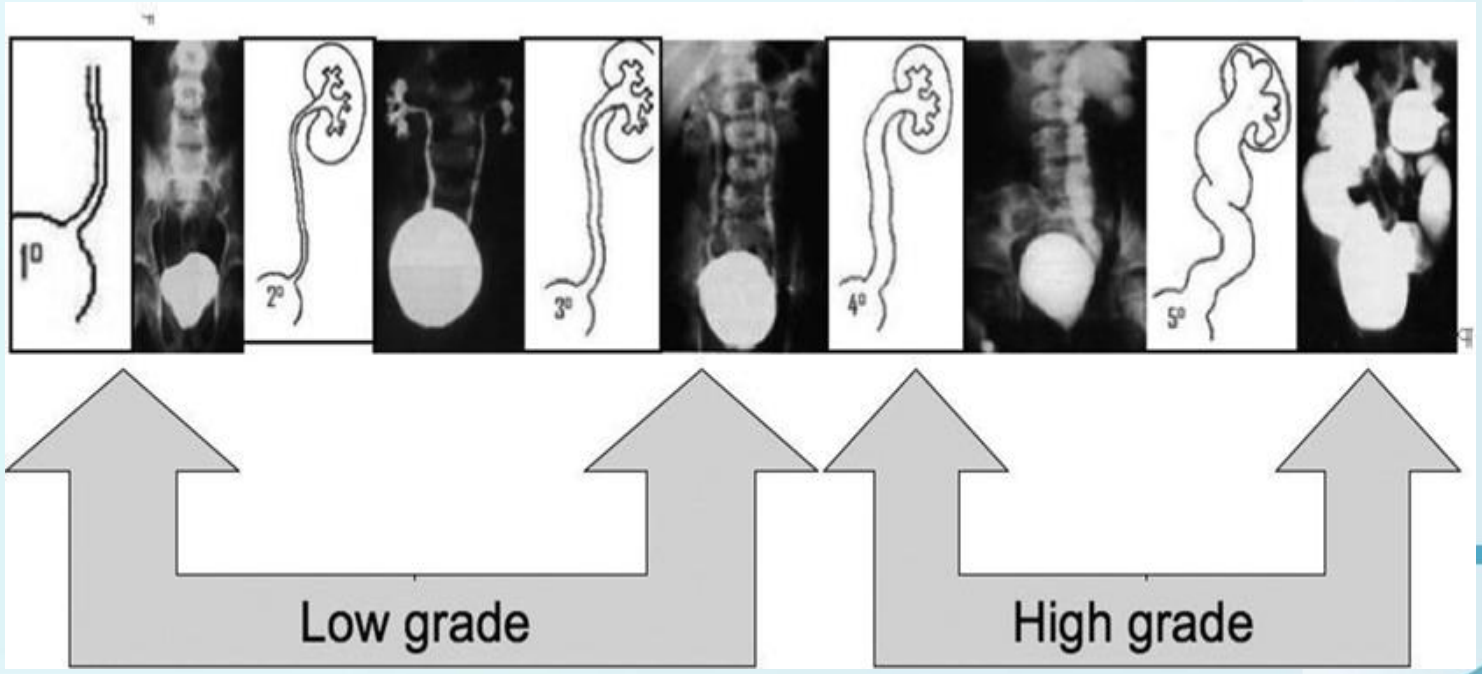
VUR…

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



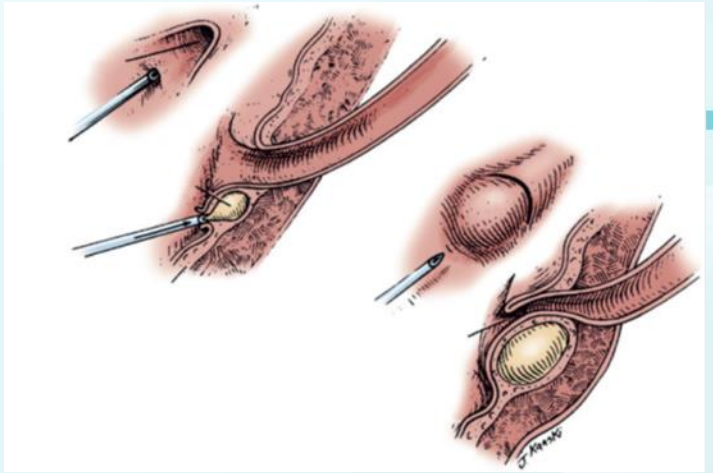
VUR…

- **Diagnosis:**
  - **MCUG (VCUG)**

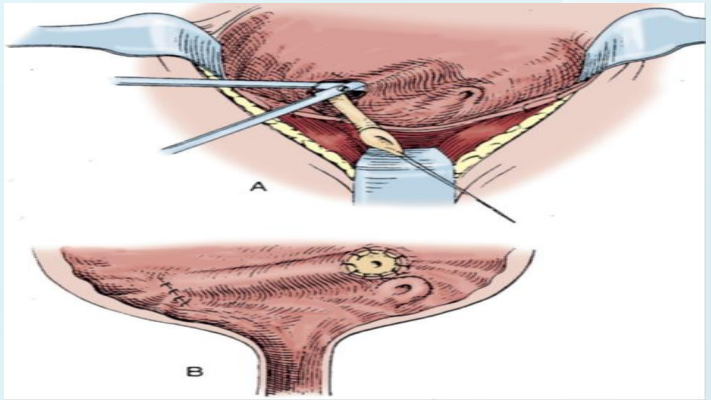


VUR…

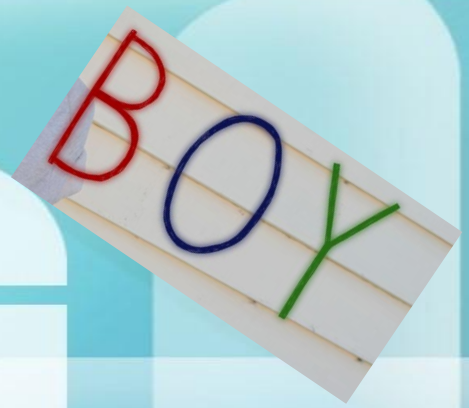
- Management:
- Prophylactic antibiotic
  - Spontaneous resolution
- Surgical treatment
  1. Recurrent pyelonephritis on antibiotic prophylaxis (outbreak infection)
  2. Noncompliant with medical treatment
  3. Persistence of reflux (high grade)



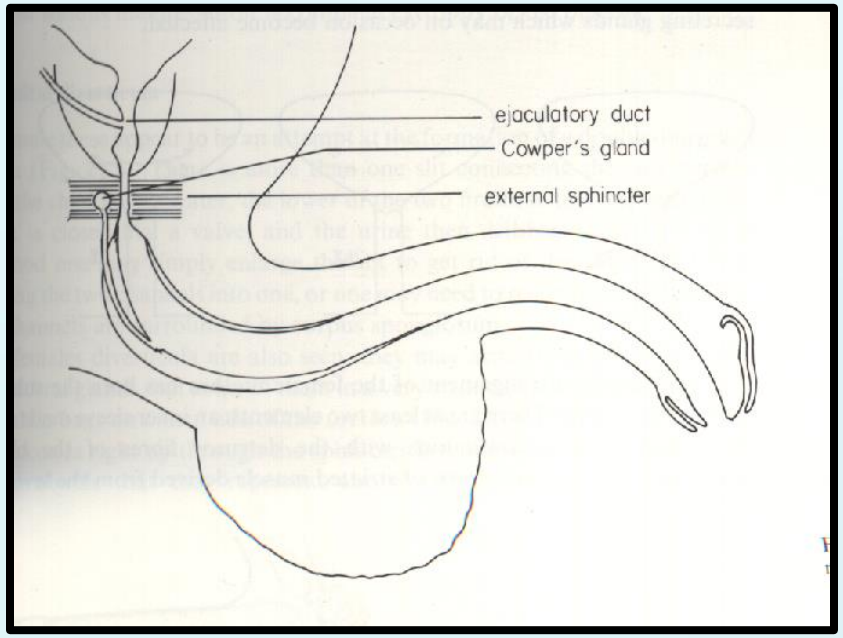
Endoscopic treatment

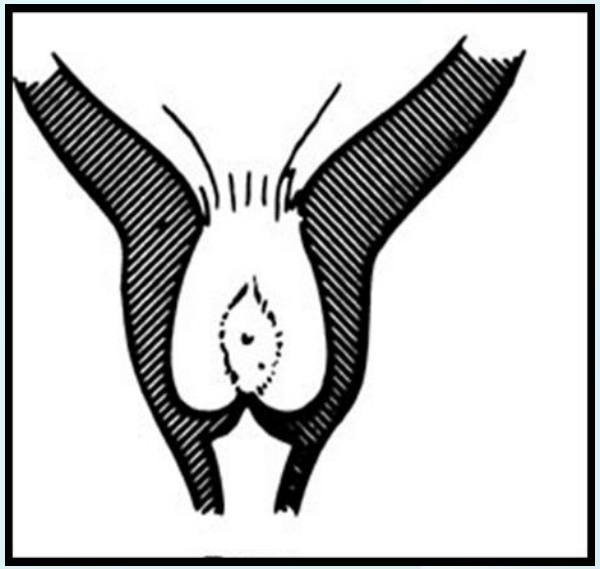


Ureteral reimplantation

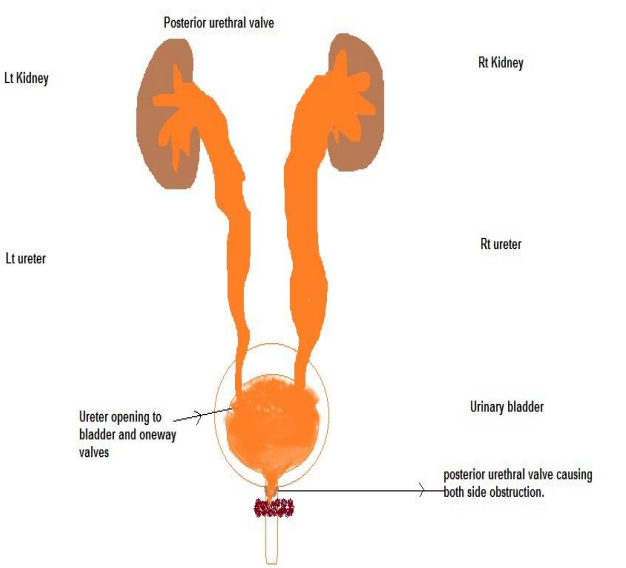


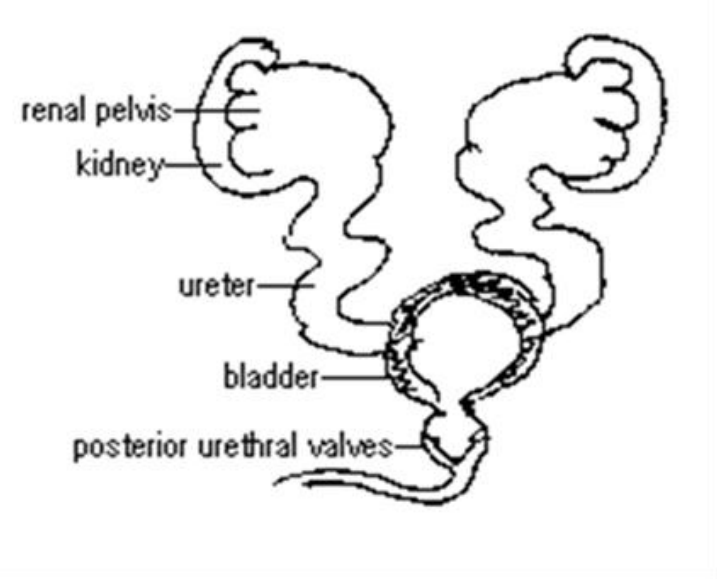
Posterior Urethral Valves (PUV)





Posterior Urethral Valves (PUV)





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

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

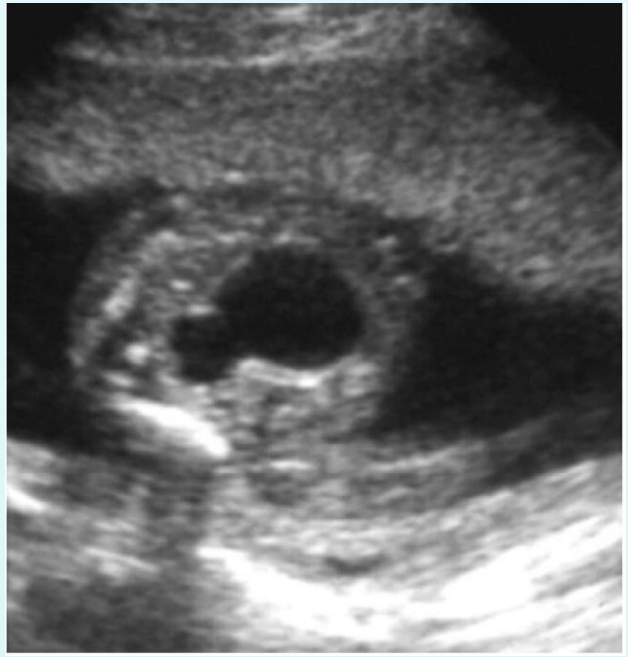
PUV…

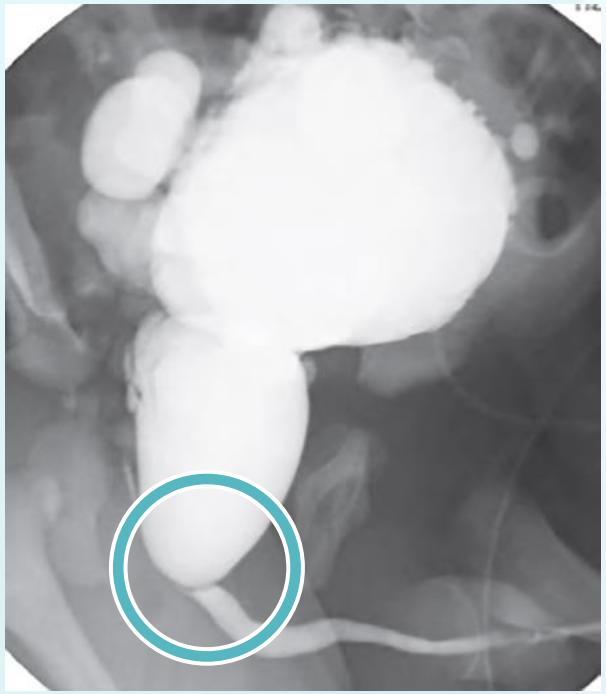
- 1 in 8000 to 25,000 live births.
- Make up 10% of urinary obstructions diagnosed in utero.
- Most common cause of urine retention in male infants.
- 50% have renal impairment.

PUV…

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

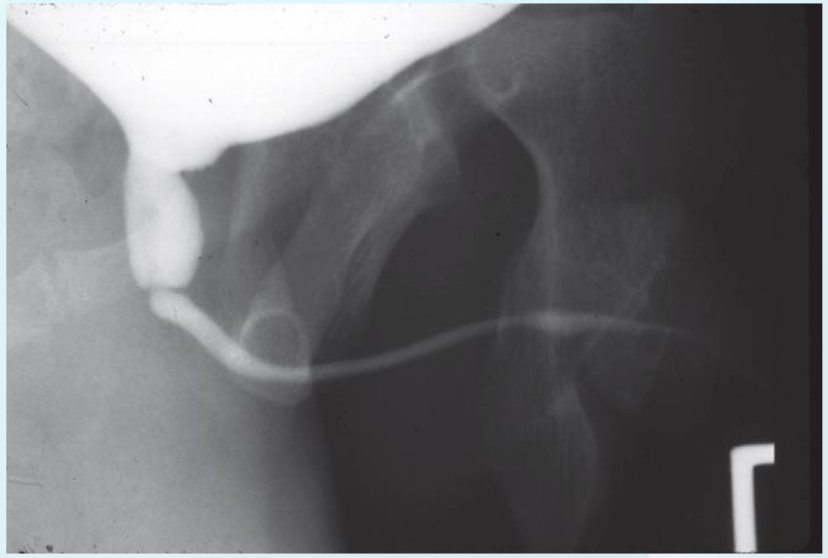
PUV…



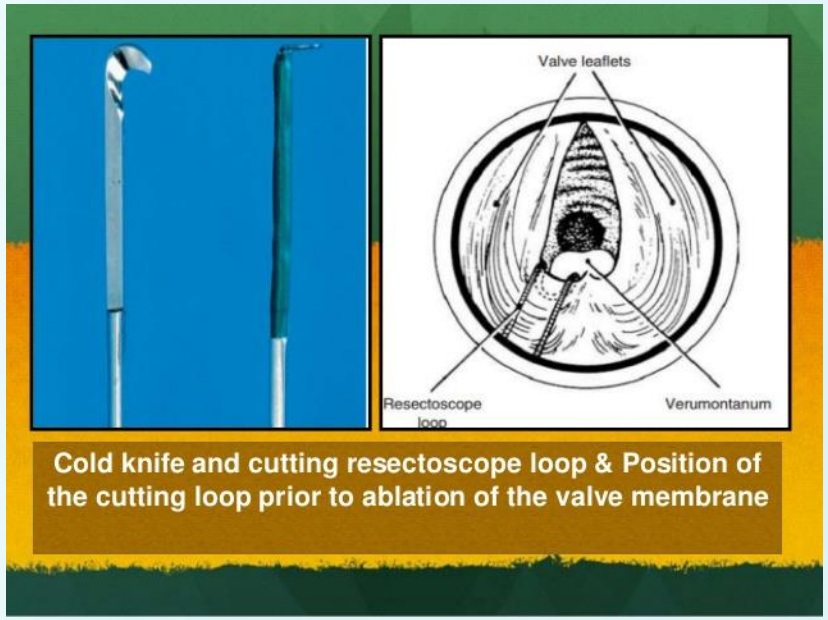


PUV…

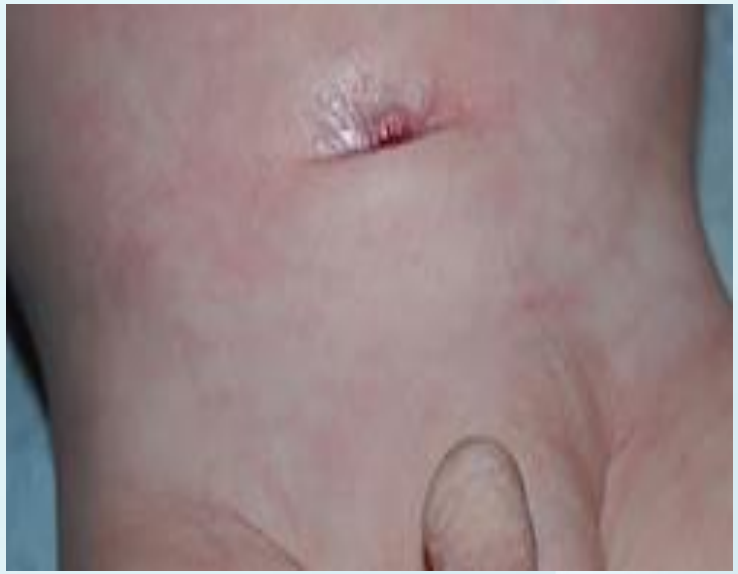
- Initial urological l treatment
  - Feeding tube insertion
  - Start antibiotic prophylactic
  - Ultrasound
  - MCUG



PUV…

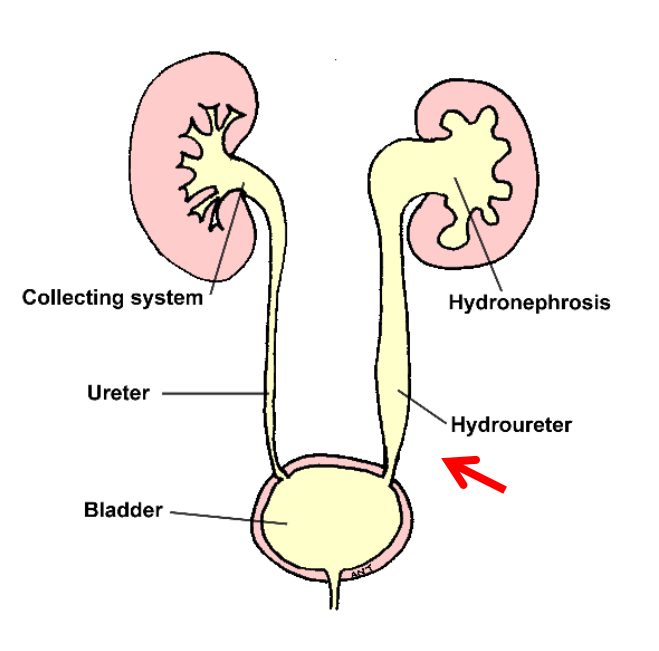


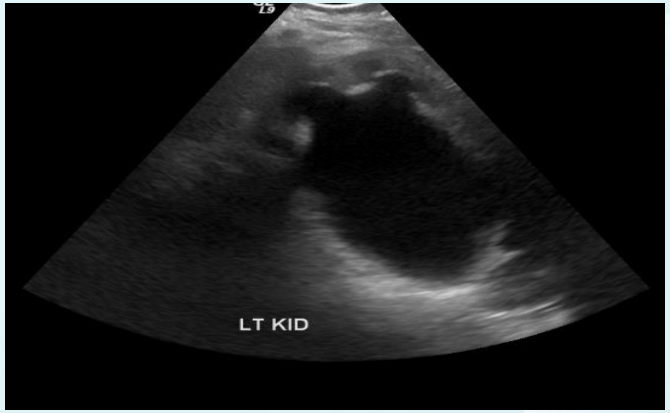
Endoscopic valve ablation

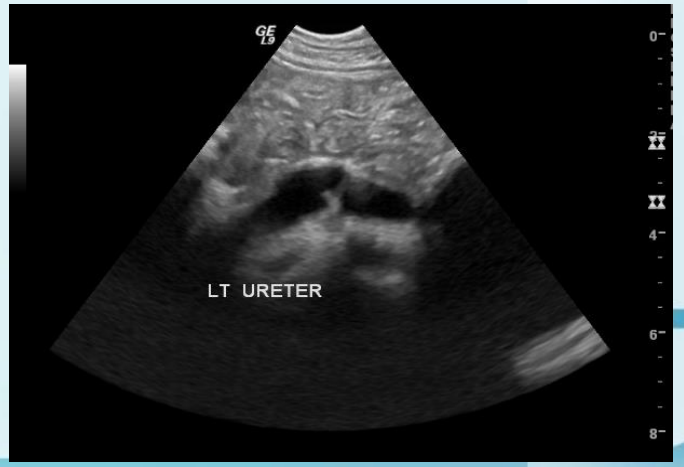


Cutaneous vesicostomy

Ureterovesical junction obstruction (UVJO) (Megaureters)



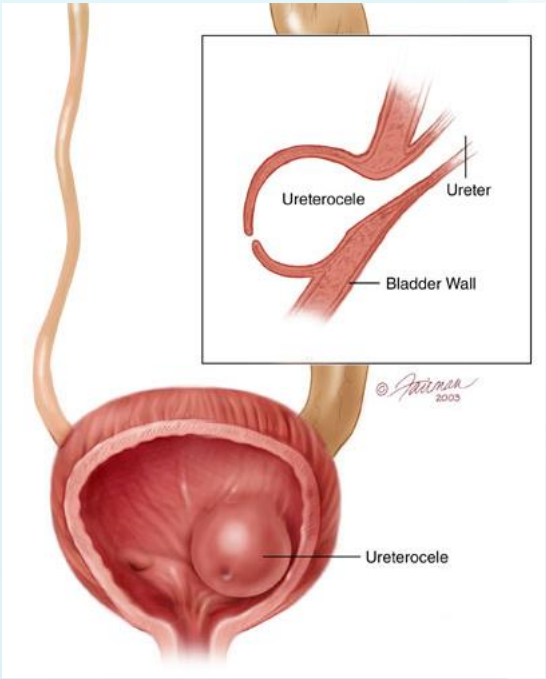




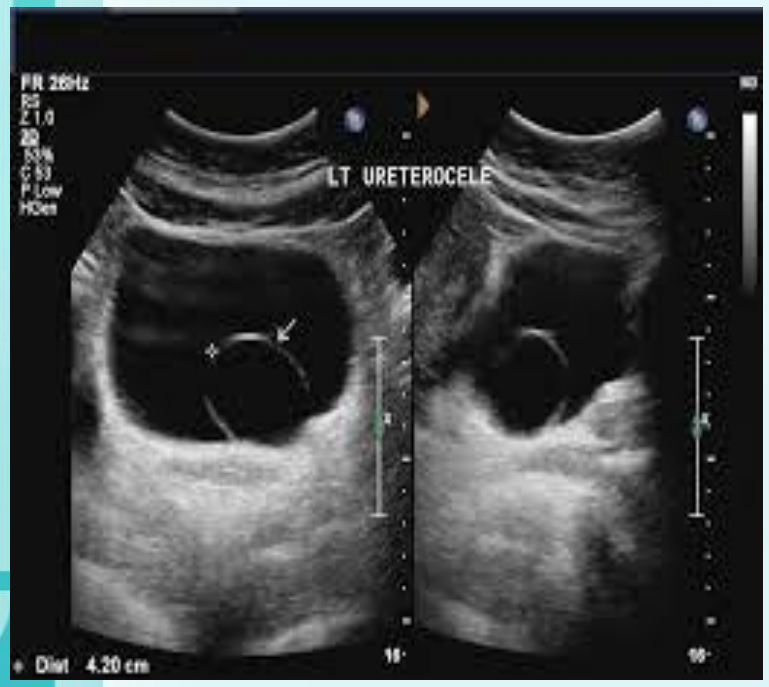
Ureteral reimplantation

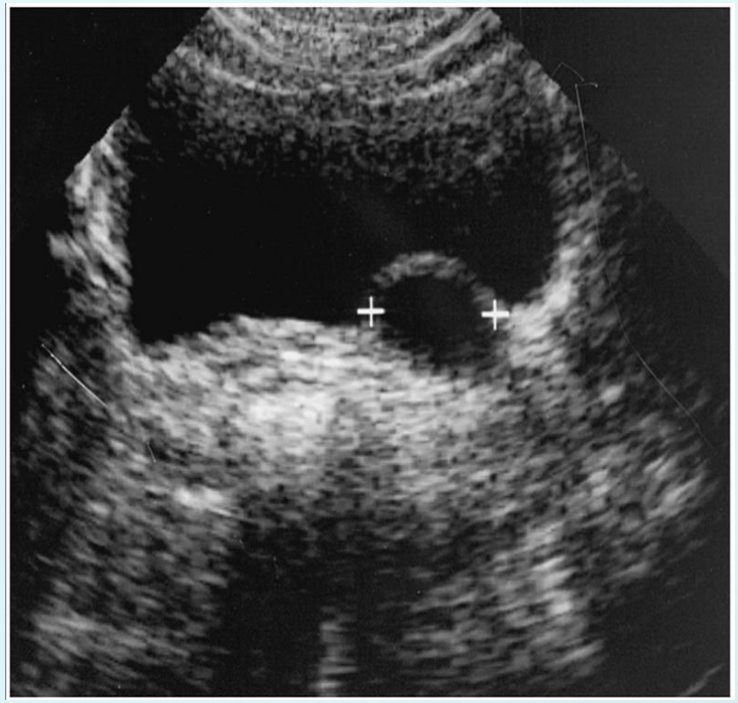
Ureterocele

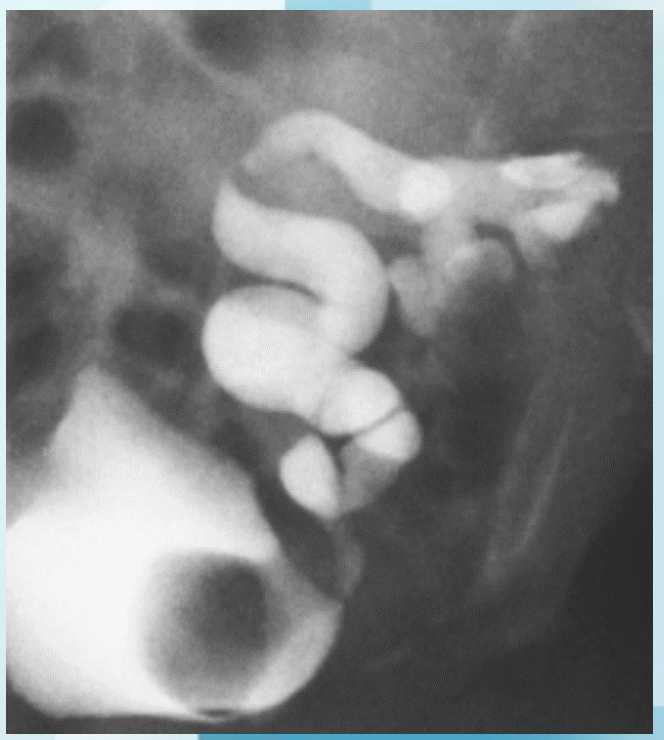
- A cystic dilation of the distal aspect of the ureter
- Located either within the bladder or spanning the bladder neck and urethra.
- **Presentation:**
  - Antenatal (U/S)
  - Urine retention
  - Infection
  - Stone formation



Ureterocele…



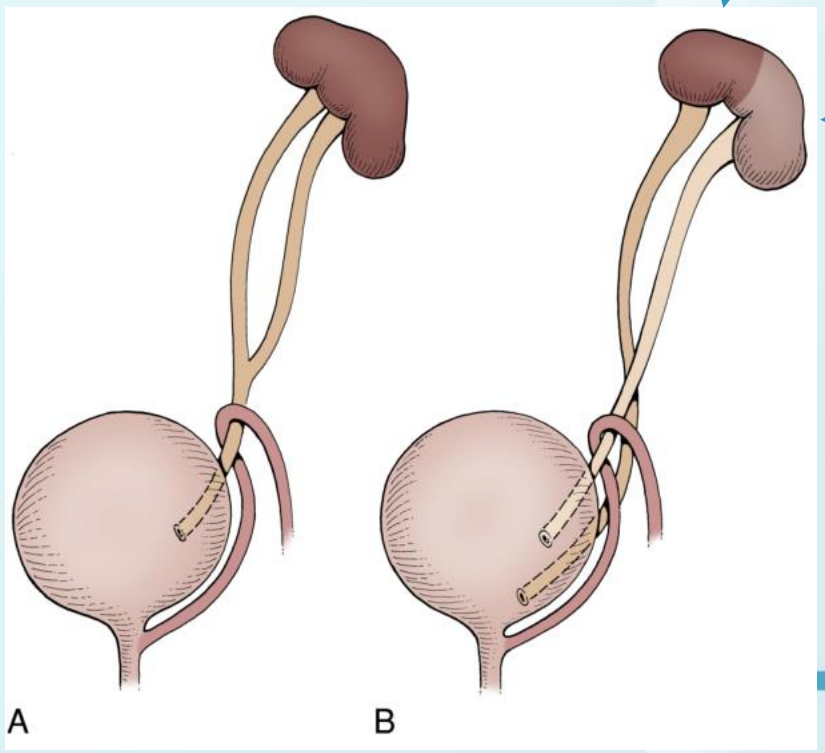




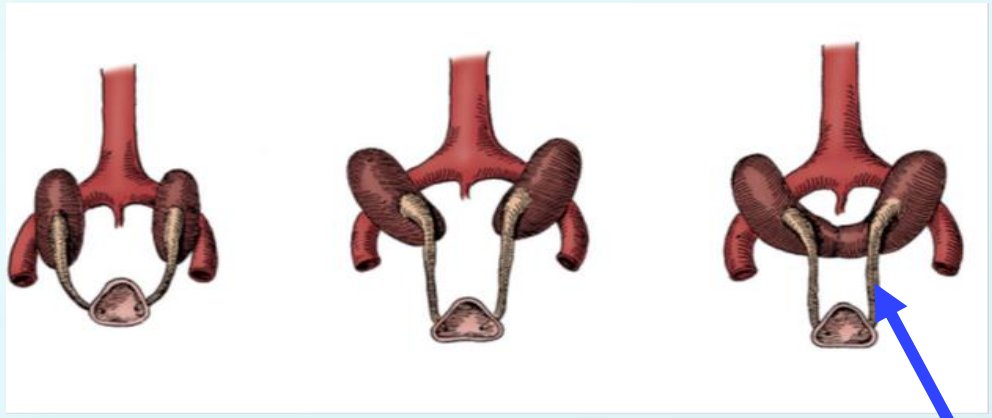
Endoscopic incision of ureterocele

Duplication Anomalies

- 1%
- 1.6:1 F:M
- 85% unilateral.
- Complete or incomplete
- Associated
  - Reflux 43%
  - Renal dilatation 29%
  - Ectopic insertion 3%
  - Ureterocele.



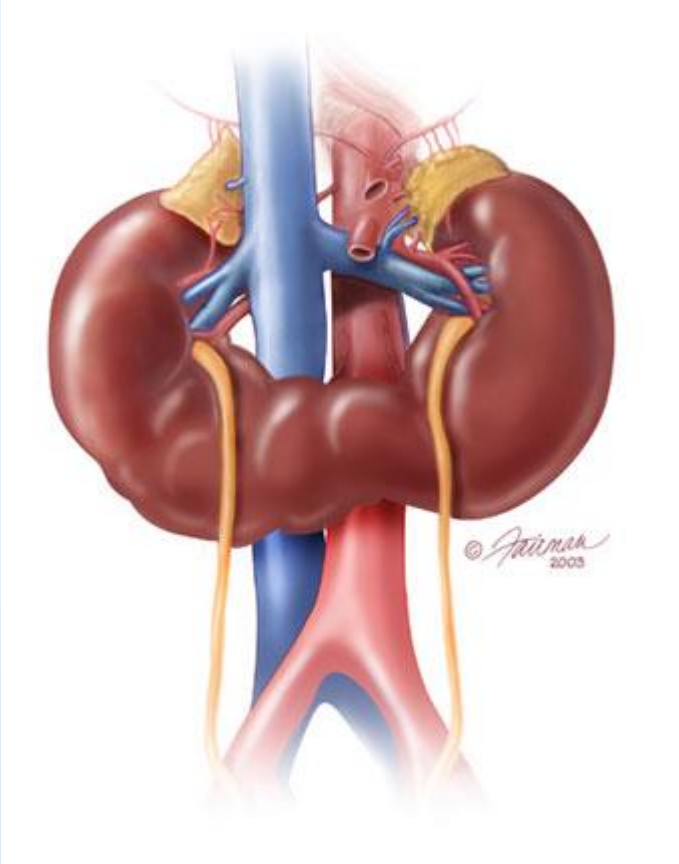
Horseshoe Kidney



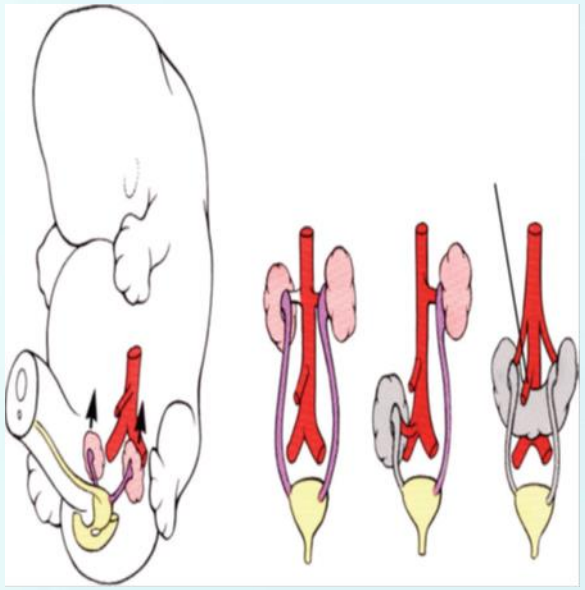
The isthmus is bulky and consists of parenchymatous tissue.

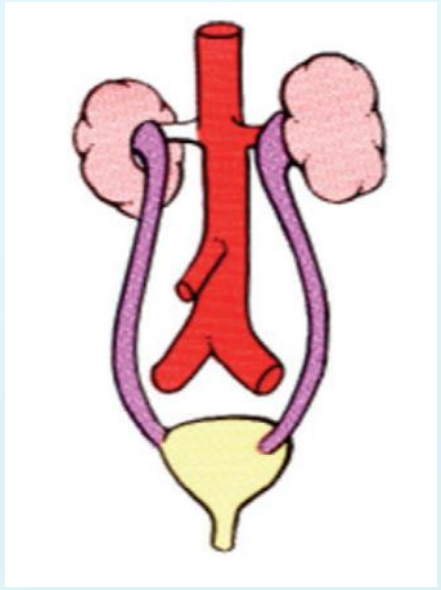
Horseshoe Kidney

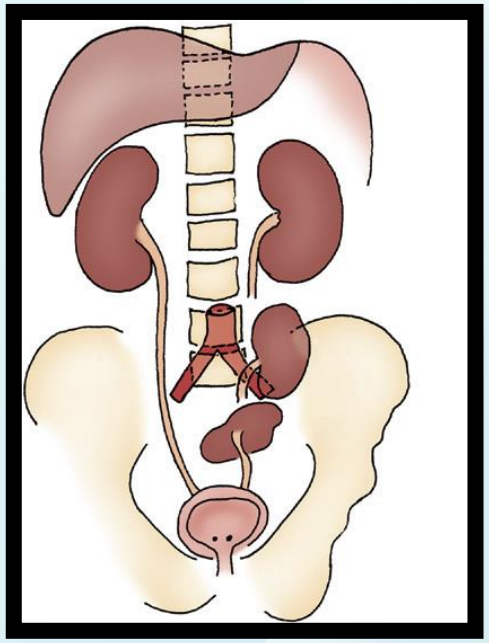
- Occurs 1 in 400 persons.
- The calyces:
  - ✓ normal in number
  - ✓ atypical in orientation.
  - ✓ pelvis remains in the vertical or obliquely lateral plane
- Horseshoe kidney is frequently found in association with other congenital anomalies.
- UPJ obstruction in one third.
- 60 % asymptomatic.



Simple Renal Ectopia

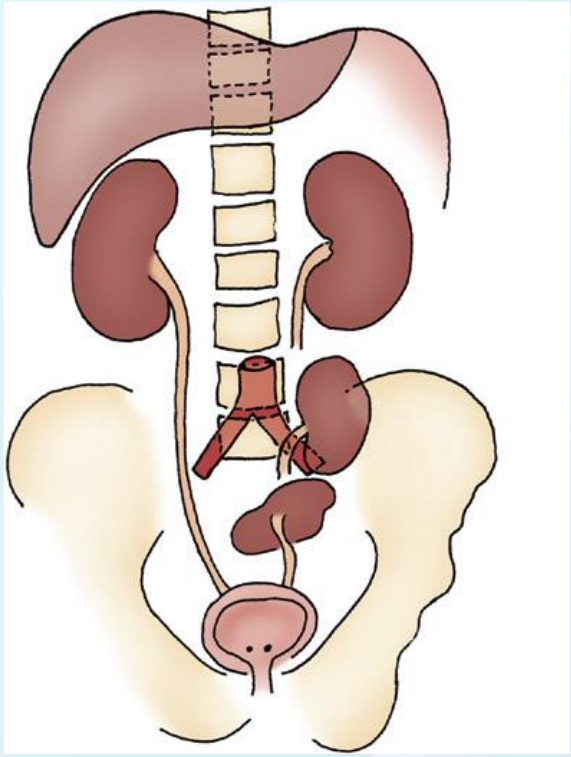






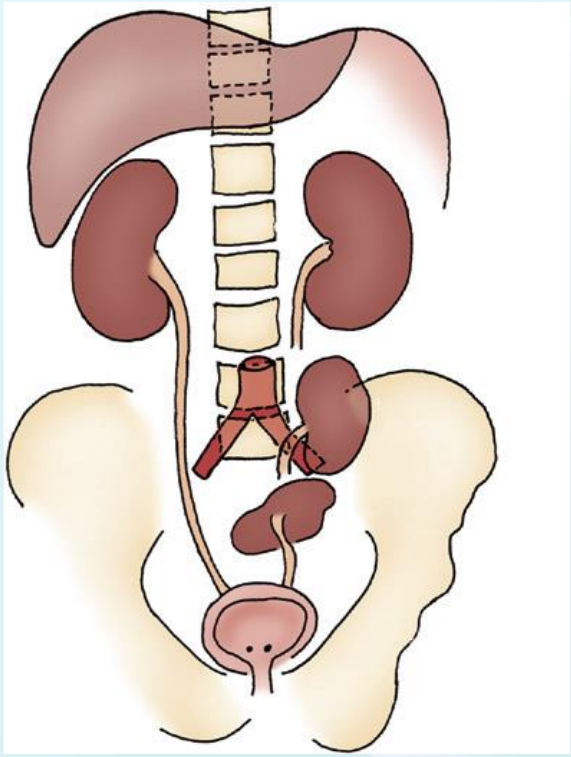
Simple Renal Ectopia

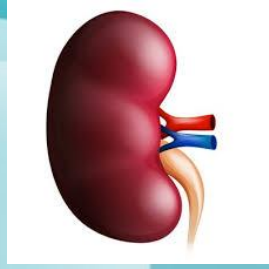
- Left more than the right.
- 1 of 2100 to 3000 autopsies.
- Most ectopic kidneys are clinically asymptomatic.



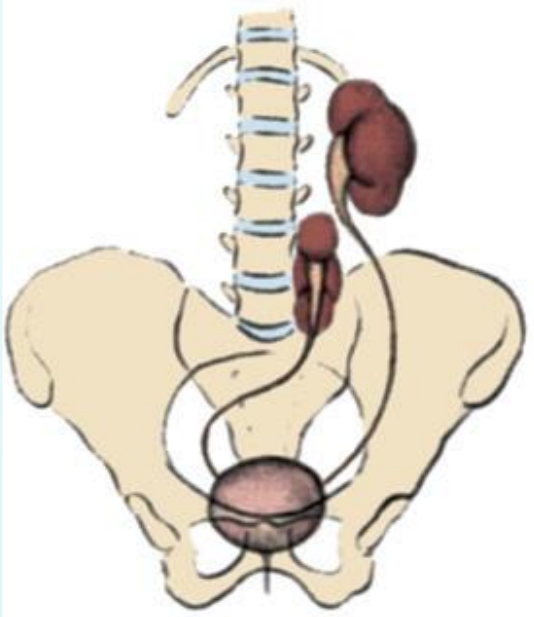
Simple Renal Ectopia…

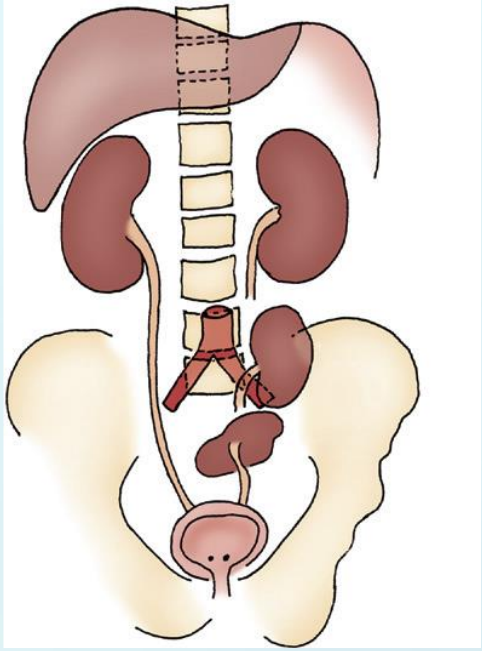
- Associated Anomalies:
  - 50% have a hydronephrosis:
    - ✓ Obstruction: UPJO and UVJO
    - ✓ Reflux (VUR): grade III or greater
    - ✓ Malrotation
  - Genital anomalies in the patient with ectopia is about 15%.



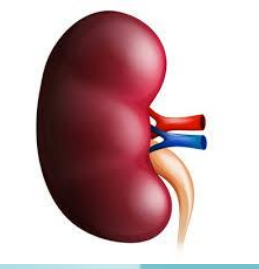


Crossed Renal Ectopia



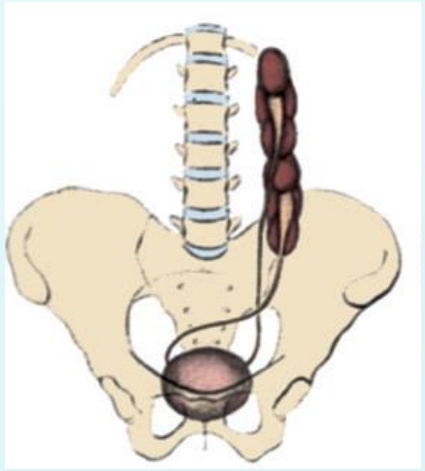


Crossed ectopia: kidney is located on the side opposite from that in which its ureter inserts into the bladder.
The ureter from each kidney is usually orthotopic.

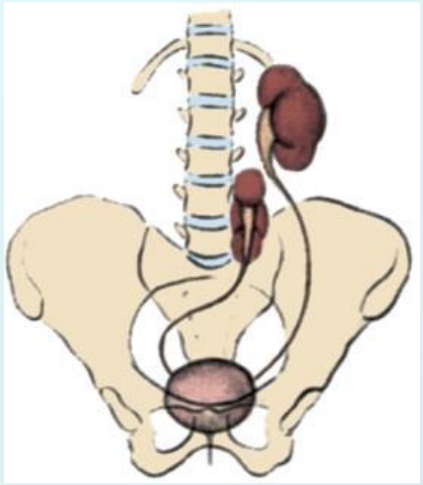


Crossed Renal Ectopia…

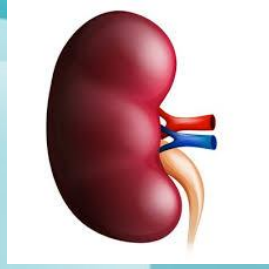
with Fusion



without Fusion

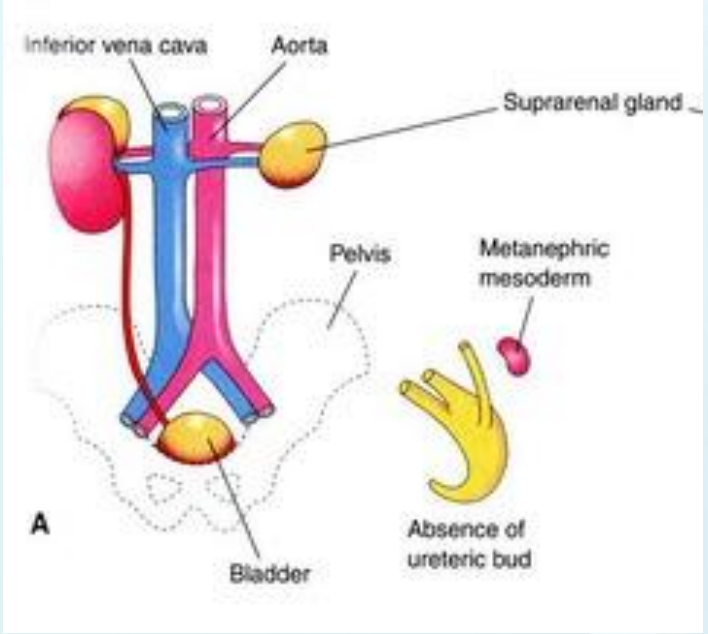


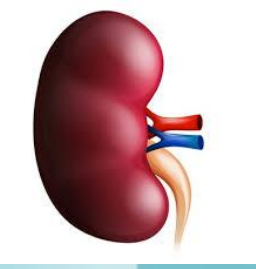
90% are fused
the superior pole of the ectopic kidney usually joins with the inferior aspect of the normal kidney.



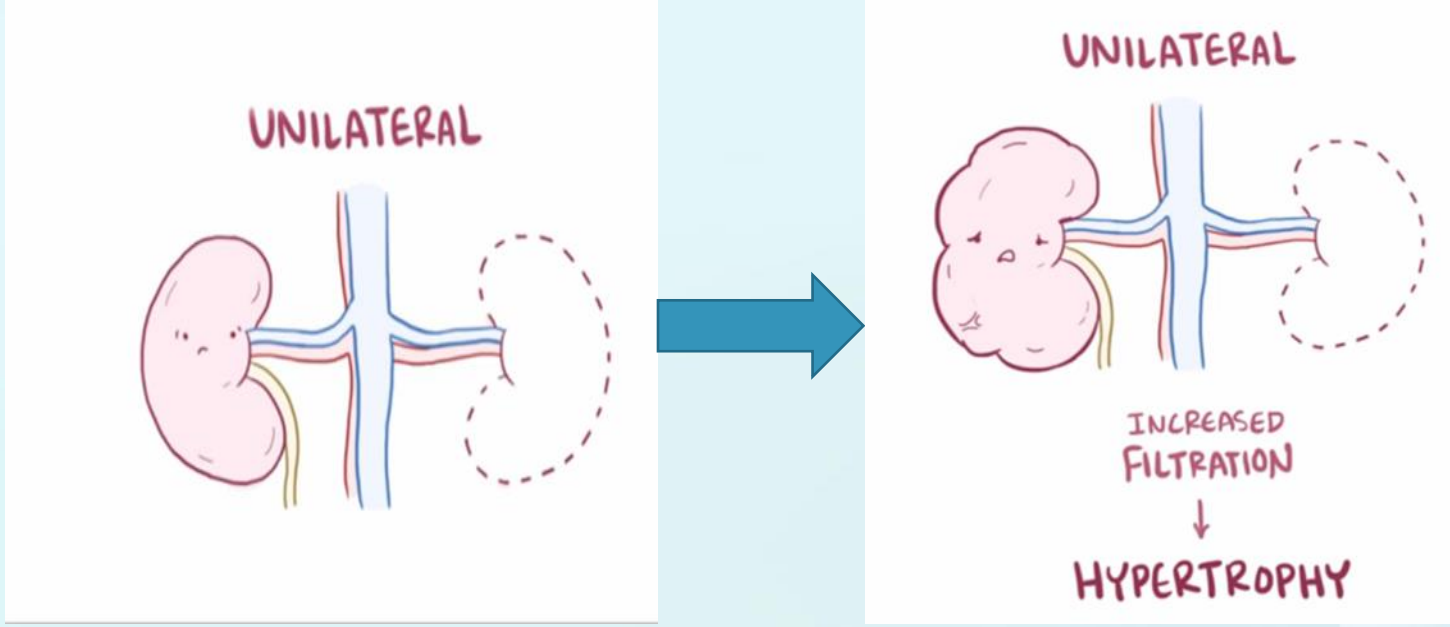
Unilateral Renal Agenesis

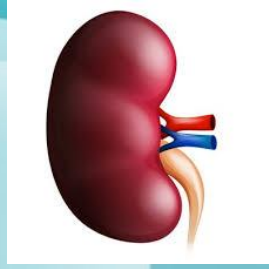
- 1 in 1100 births.
- Male: Female of 1.8 : 1
- The left side is absent more frequently .
- The ipsilateral ureter is completely absent in 50%.





Unilateral Renal Agenesis…

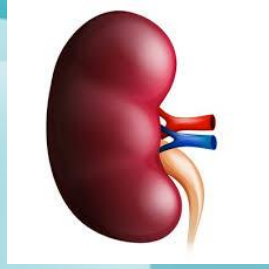




Unilateral Renal Agenesis….

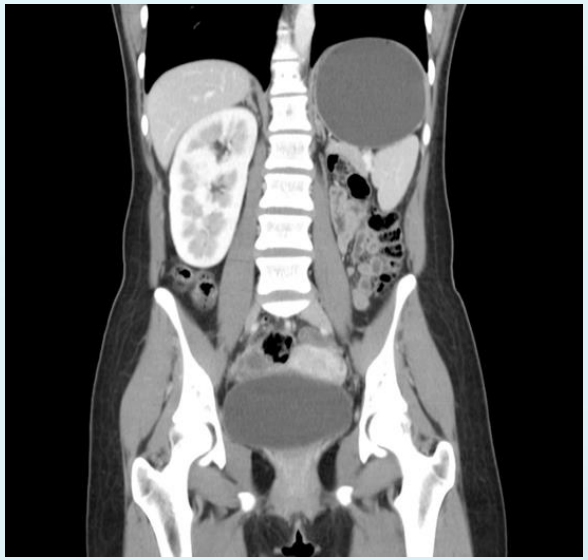
- Associated anomalies:
  - Anomalies of other organ systems are found frequently in affected individuals

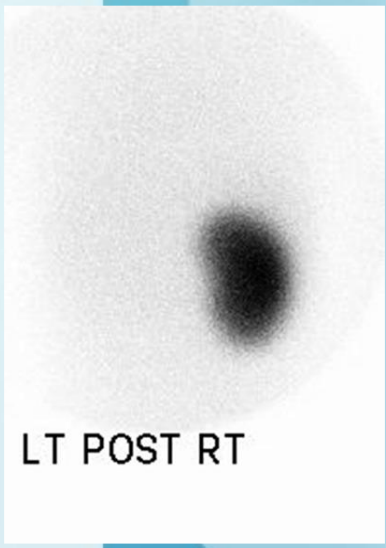
    CVS,GIT,MSC
  - Müllerian duct abnormalities
    - 25% to 50% of females
    - 10% to 15% of males
    - Approximately one fourth to one third of women with Mullerian duct anomalies are found to have URA.



Unilateral Renal Agenesis…

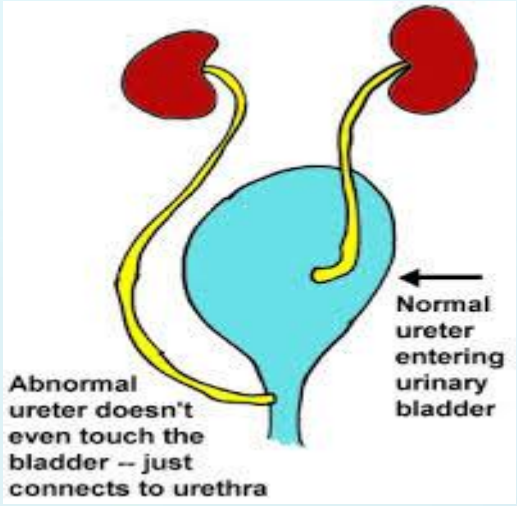
- **Diagnosis:**
  - Prenatal US
  - Incidentally
    - Abdominal US
    - Abdominal CT
  - *Confirmed*
    - *Nuclear study (DMSA)*

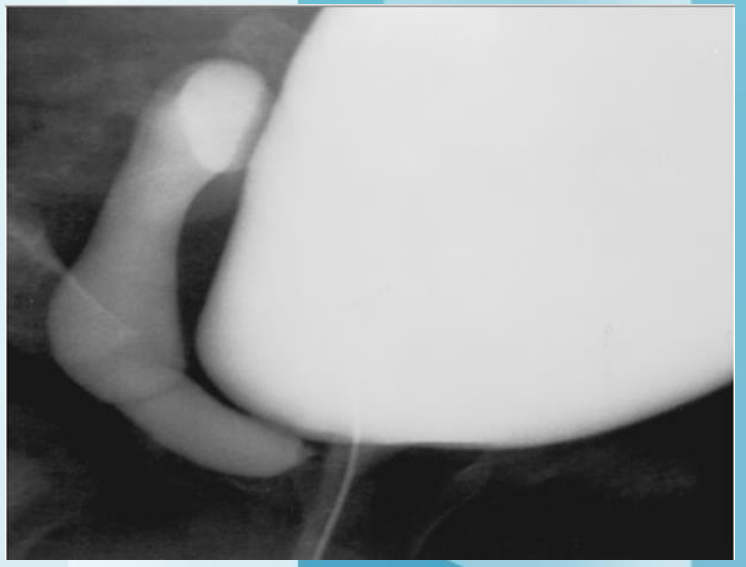




Ectopic Ureter

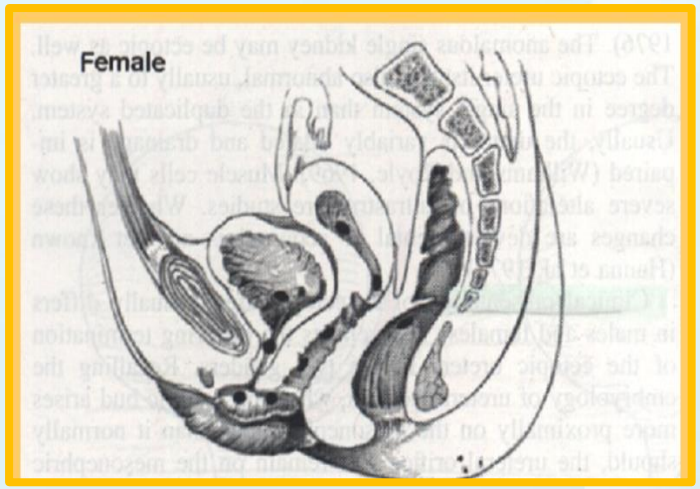
- An ectopic ureter is any ureter, single or duplex, that doesn't enter the trigonal area of the bladder.





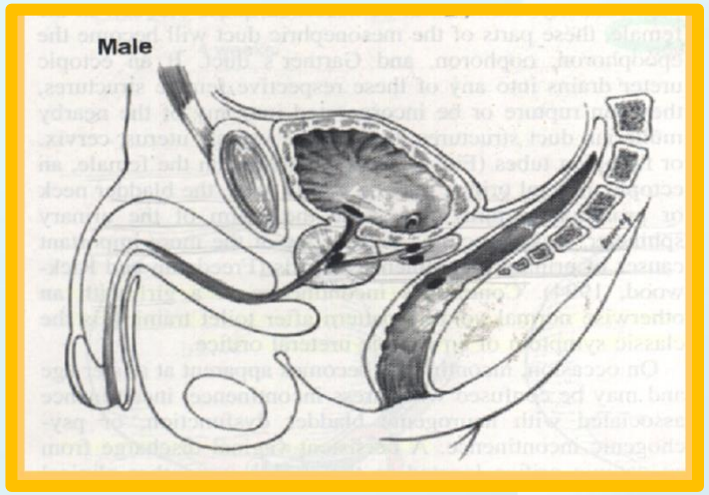
Ectopic Ureter…

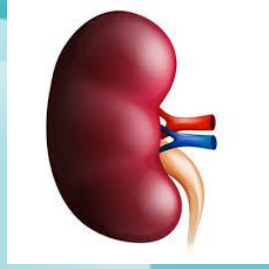
- In females the ectopic ureter may enter anywhere from the bladder neck to the perineum and into the vagina, uterus,and even rectum.
- One of the classic symptoms is **continuous wetting.**



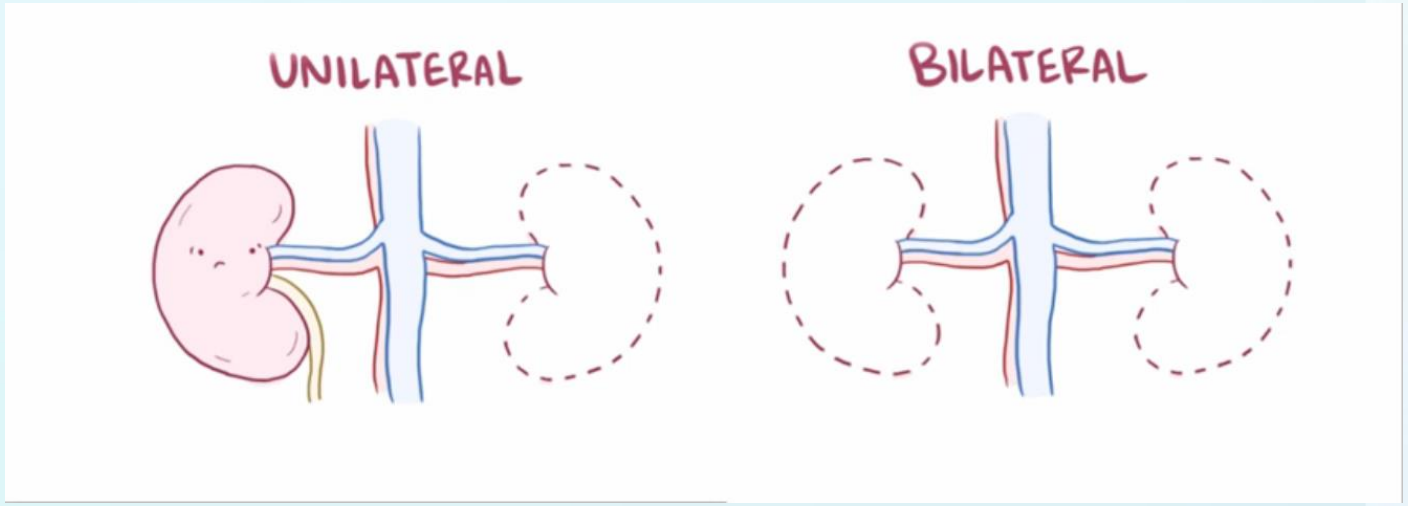
Ectopic Ureter

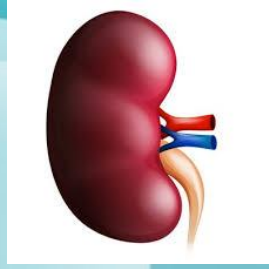
- In males the ectopic ureter always enters the urogenital system above the external sphincter or pelvic floor, and usually into the wolffian structures including vas deferens, seminal vesicles, or ejaculatory duct.



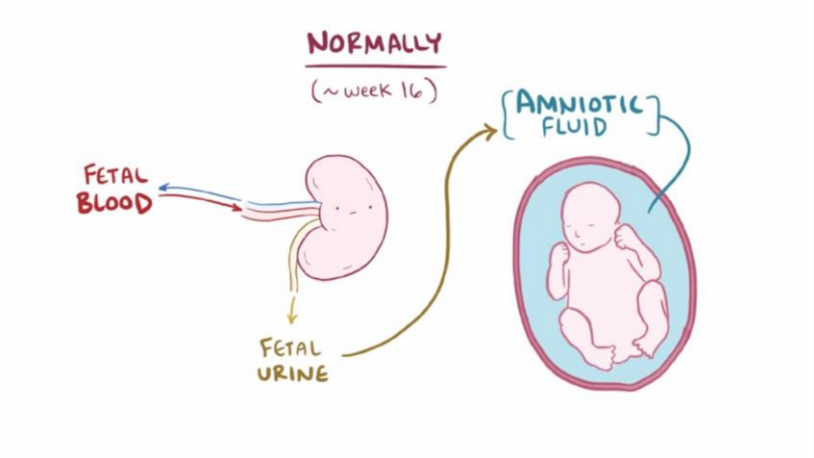


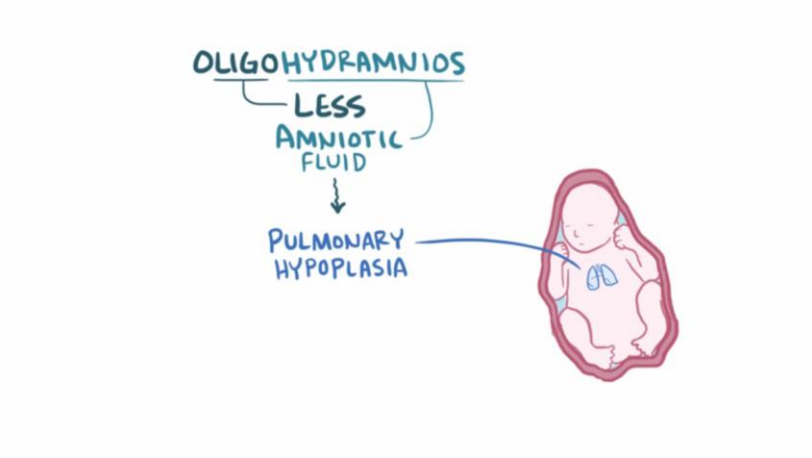
Bilateral Renal Agenesis

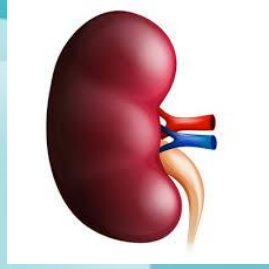




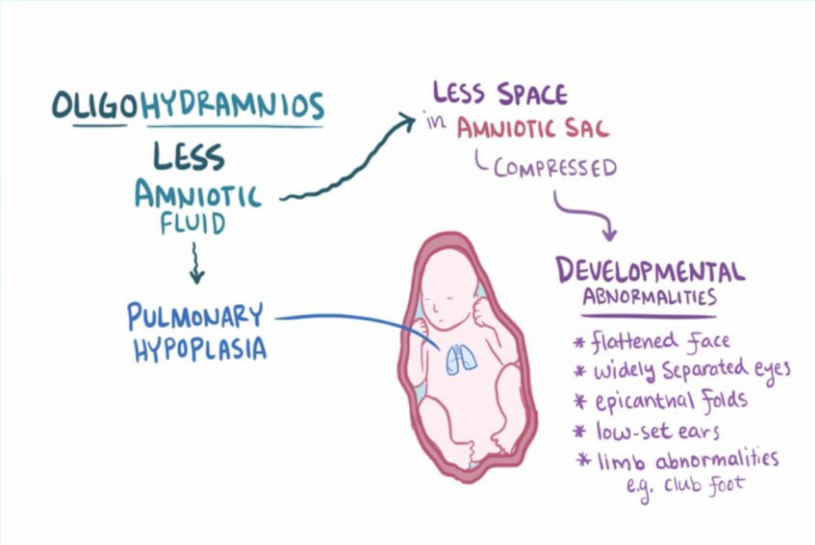
Bilateral Renal Agenesis…

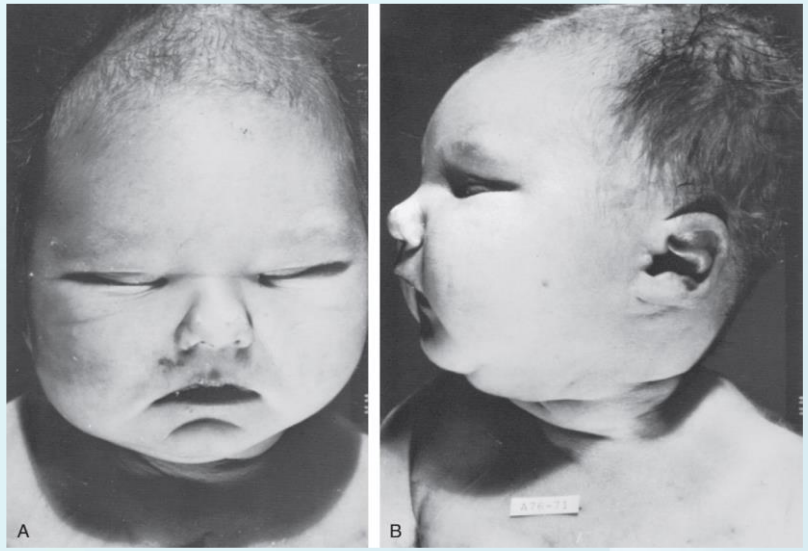


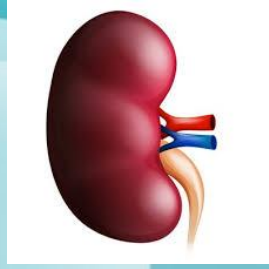




Bilateral Renal Agenesis…

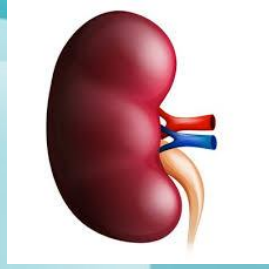






Bilateral Renal Agenesis

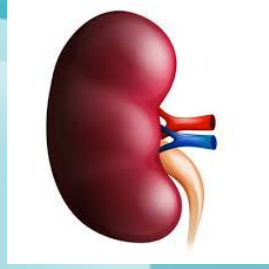
- Ureters are almost always *absent*.
- Bladder is either absent or *hypoplastic*.
- Adrenal glands are usually positioned *normally*.
- Müllerian duct anomalies are commonly observed.



Bilateral Renal Agenesis…

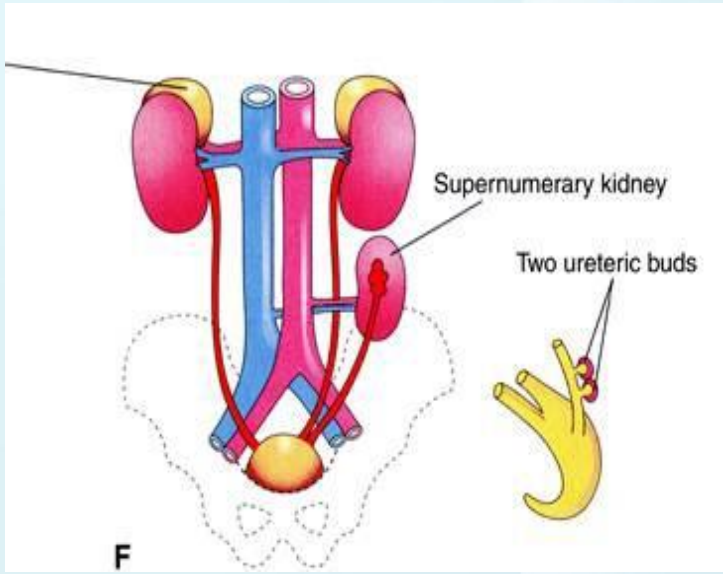
Prognosis

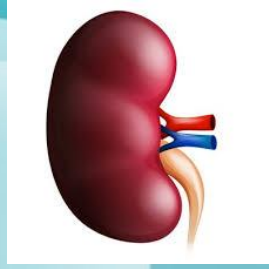
- 40% are stillborn.
- Do not survive beyond 48 hours due to respiratory distress associated with pulmonary hypoplasia.



Supernumerary Kidney

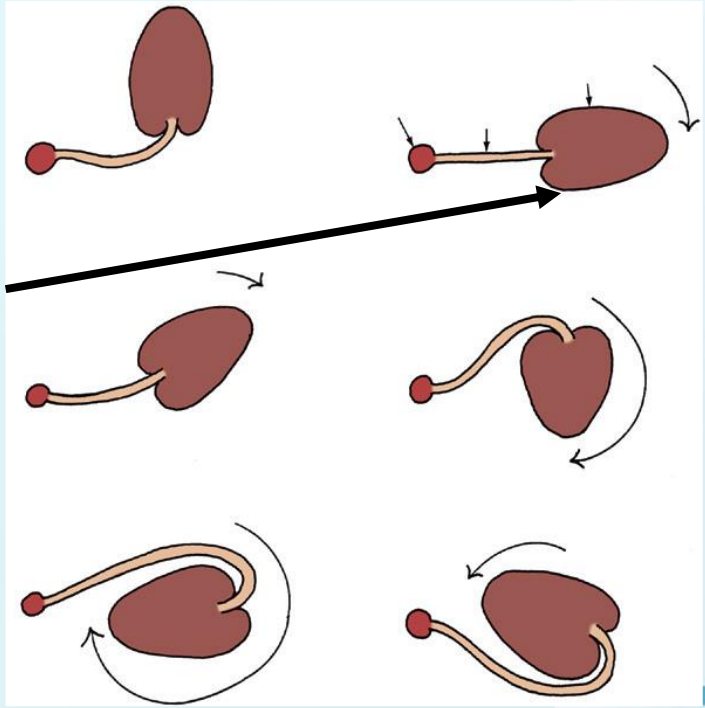
- Definitive **accessory** organ with its own collecting system, blood supply, and distinct encapsulated parenchyma.
- Either completely separate or loosely attached to the kidney on the ipsilateral side.
- The ureteral inter-relationships on the side of the supernumerary kidney can be variable.





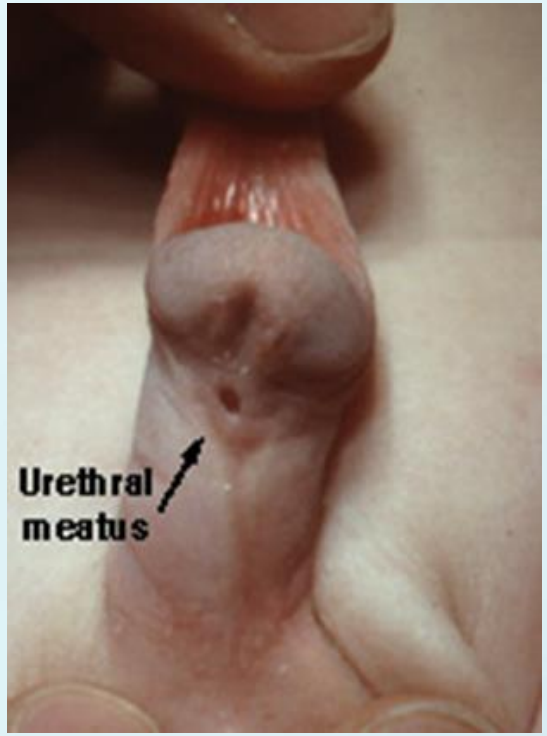
Anomalies of Rotation

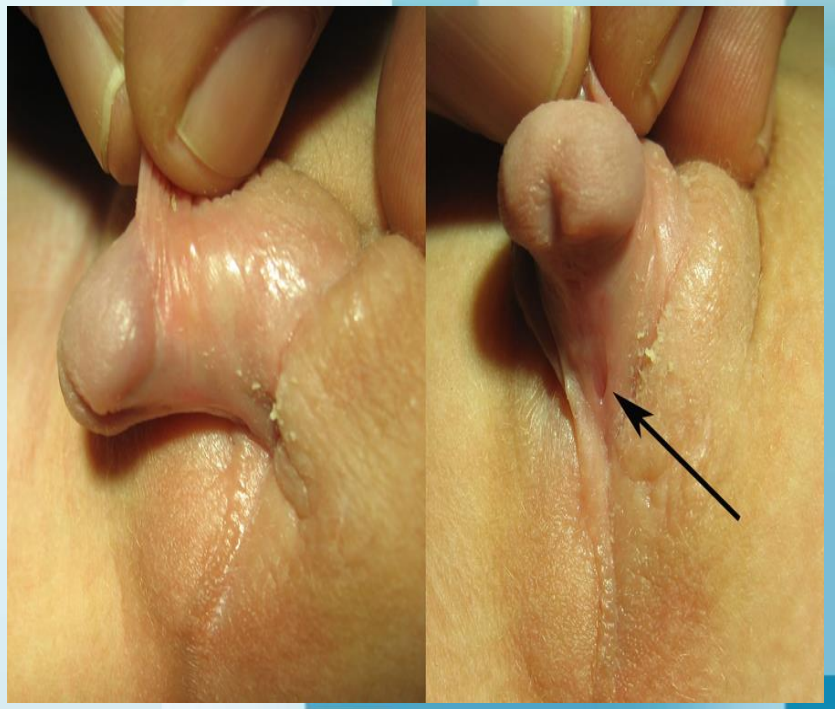
- The kidney and renal pelvis normally rotate 90 degrees ventromedially during ascent
  - ✓ the calyces point laterally.
  - ✓ the pelvis faces medially.
- When this alignment is not exact, the condition is known as malrotation.
- Frequently associated with Turner syndrome.



Hypospadias

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

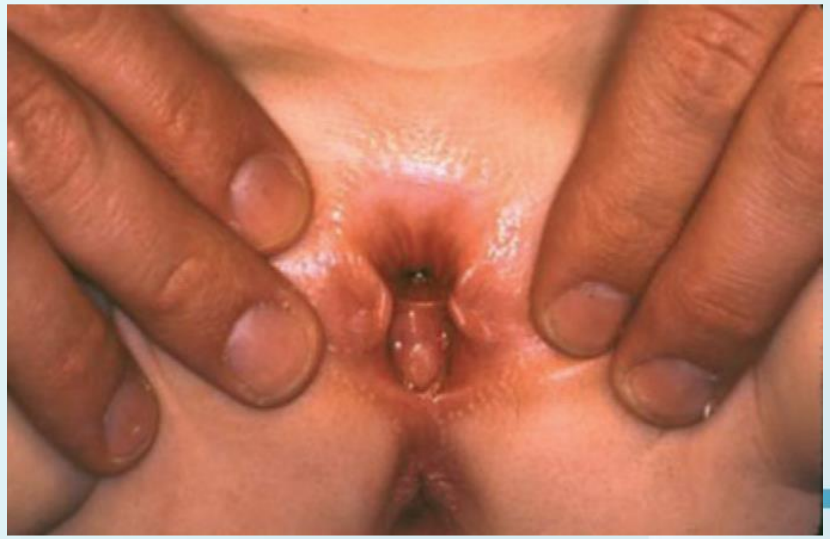




Epispadias

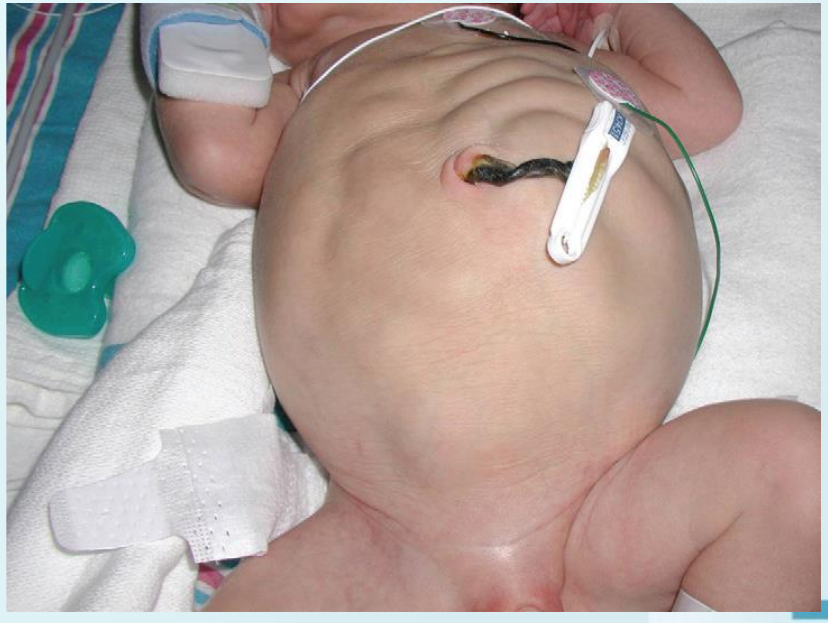
Male

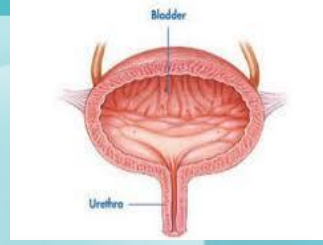
Female



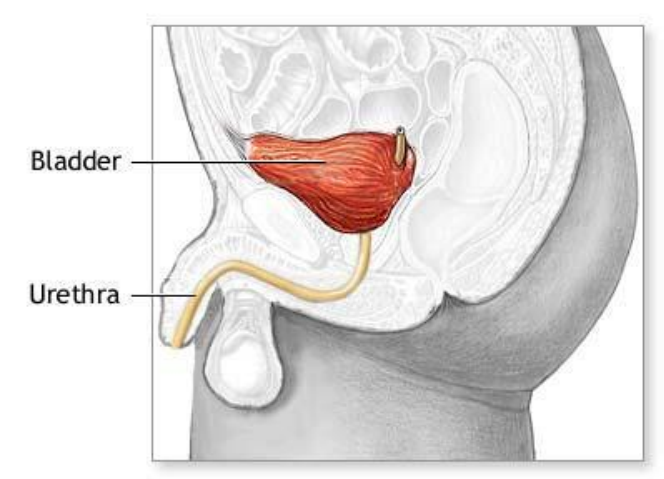
Prune-Belly Syndrome

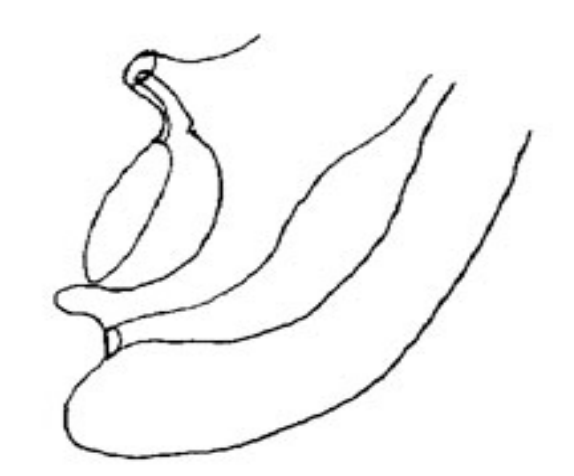
- The incidence :1 in29,000 to 1 in 40,000 live births
- The three major findings are
  - deficiency of the abdominal musculature,
  - bilateral intra-abdominal testes,
  - anomalous urinary tract
- Other names
  - Triad syndrome
  - Eagle-Barrett syndrome
  - abdominal musculation syndrome



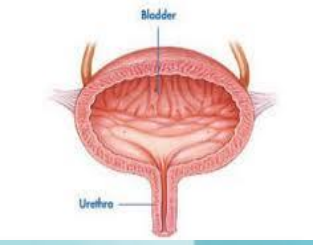


Bladder Exstrophy

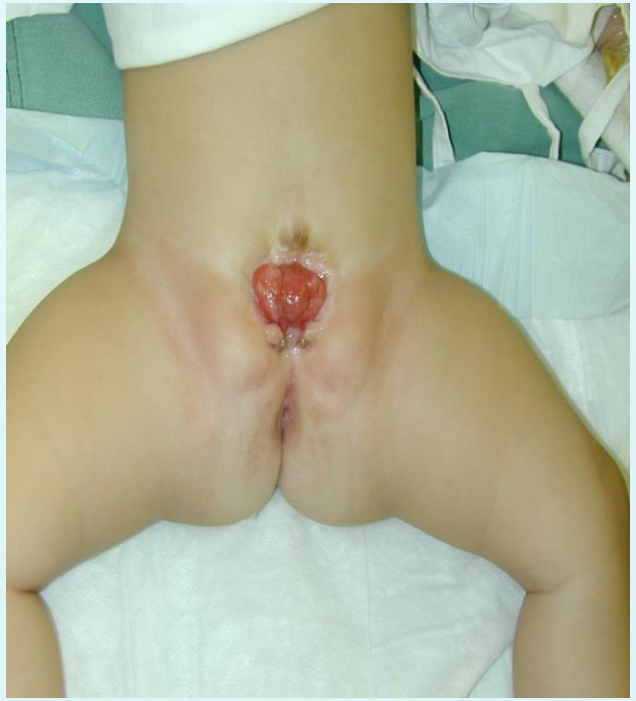


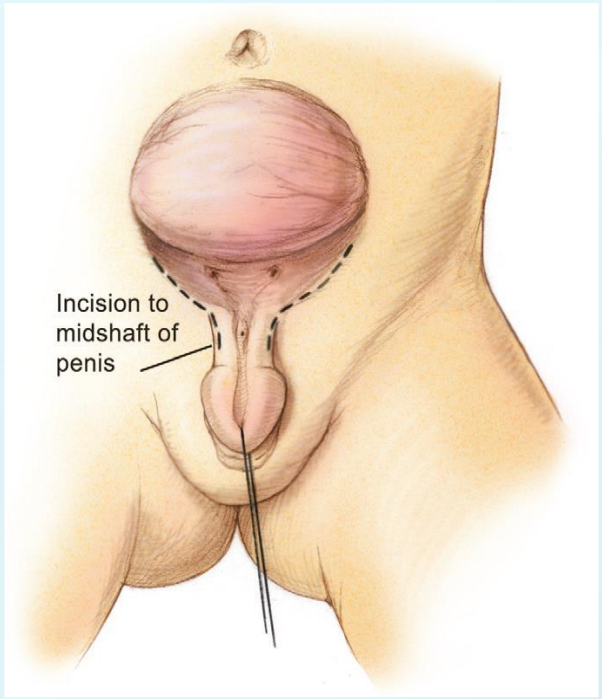


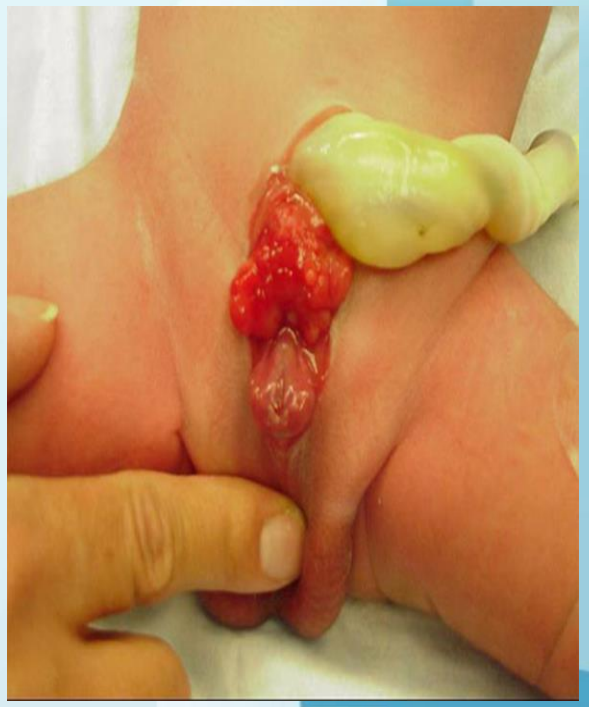
The incidence of bladder exstrophy has been estimated as between 1 in 10,000 and 1 in 50,000.



Bladder Exstrophy-Epispadias Complex

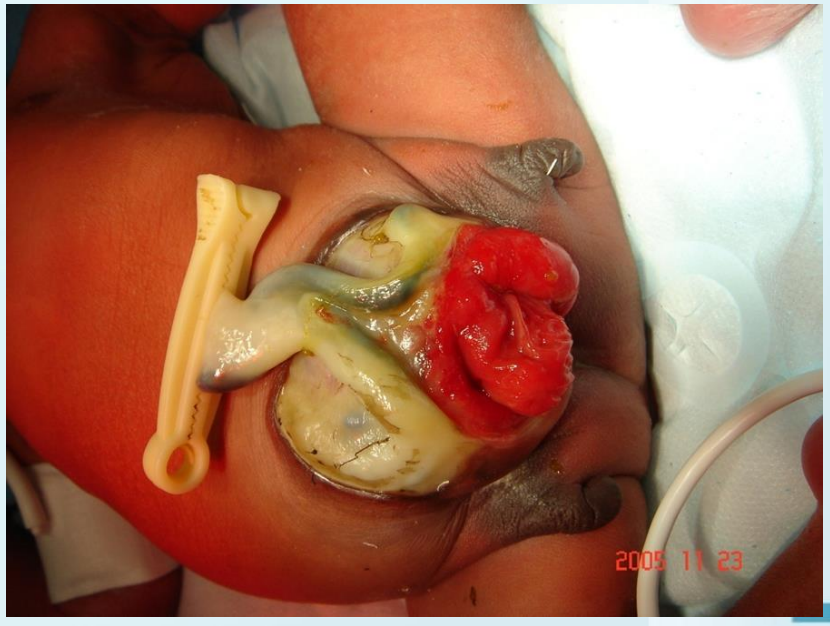


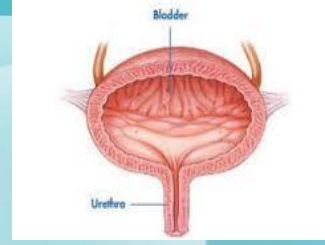




Cloacal Exstrophy

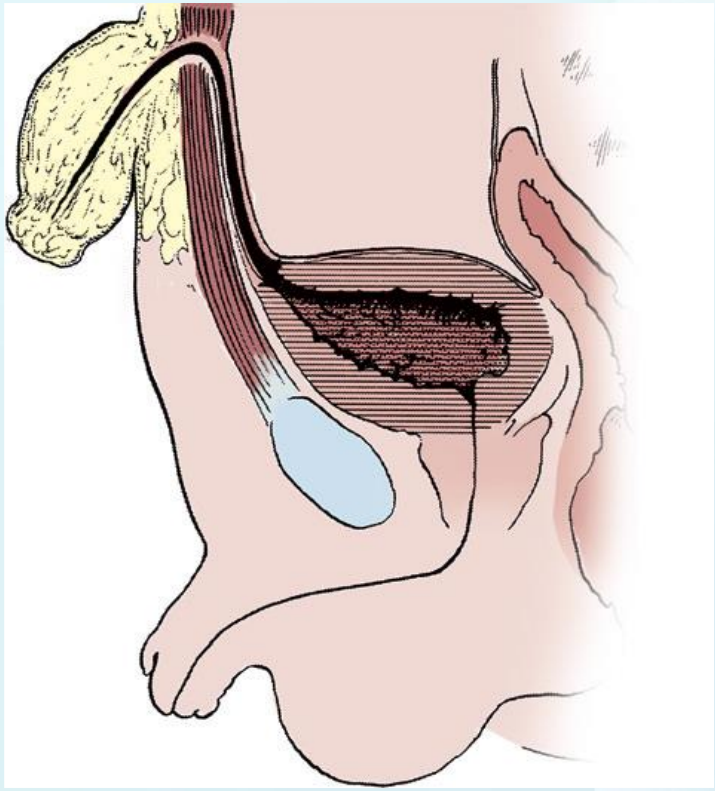
- 1 per 200,000 live births
- Associated anomalies:
  - Omphalocele
  - Gastrointestinal anomalies
    - Malrotation, duplication,duodenal atresia, Meckel diverticulum
  - Genitourinary anomalies
    - Separate bladder halves,bifid genitalia

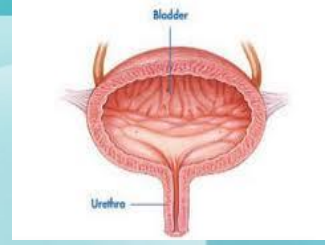




Urachal abnormalities

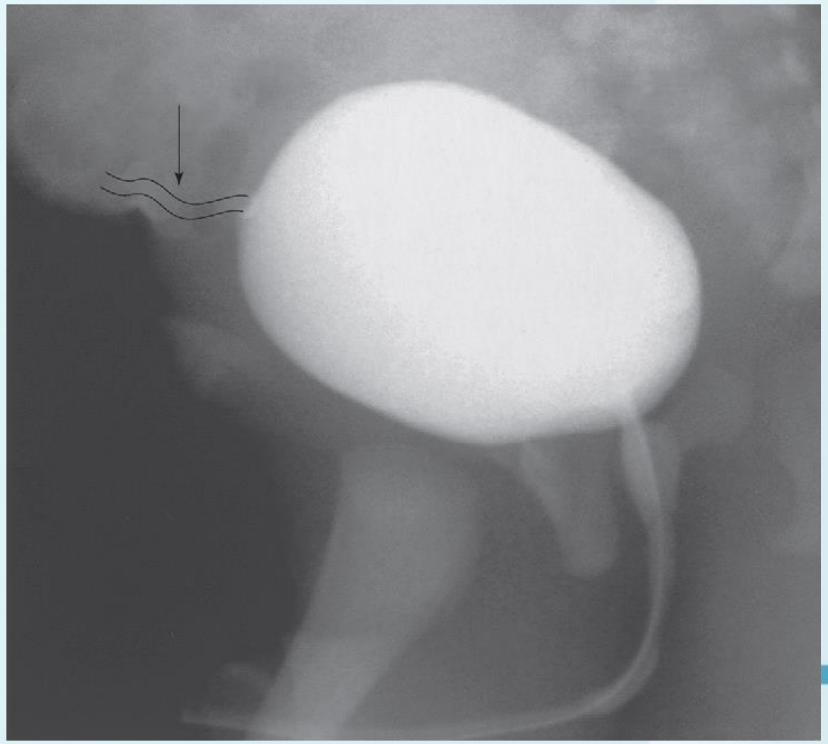
- Urachal anomalies are usually detected postnatally due to umbilical drainage.
- Imaging possibilities include ultrasound, CT, and VCUG.

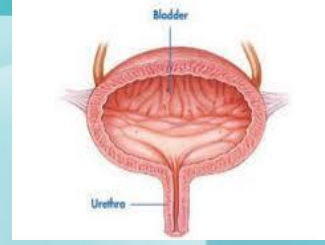




Urachal abnormalities…

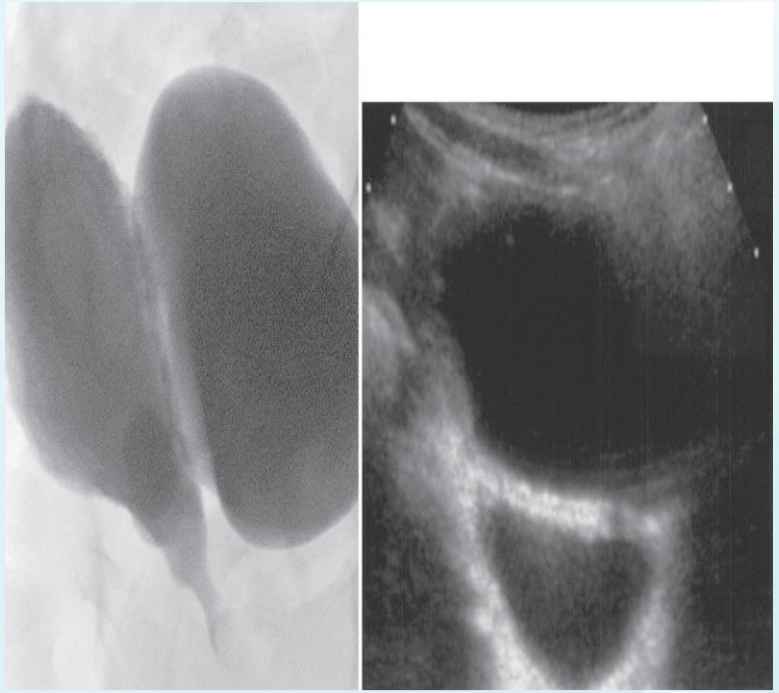
- Conservative treatment with observation is justified in asymptomatic cases due to possible spontaneous resolution
- Infected urachal remnants are initially treated with drainage and antibiotics, followed by surgical excision.
- Nonresolved urachal remnants should be excised due to theincreased risk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

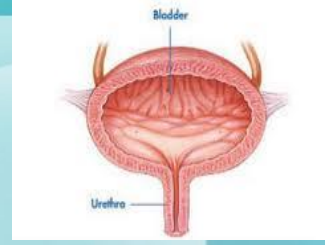




Bladder Diverticulum

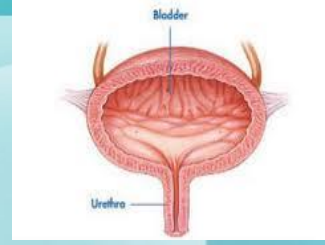
- Bladder diverticula can be detected on prenatal ultrasound, but the gold standard remains VCUG, which will reveal possible accompanying VUR.





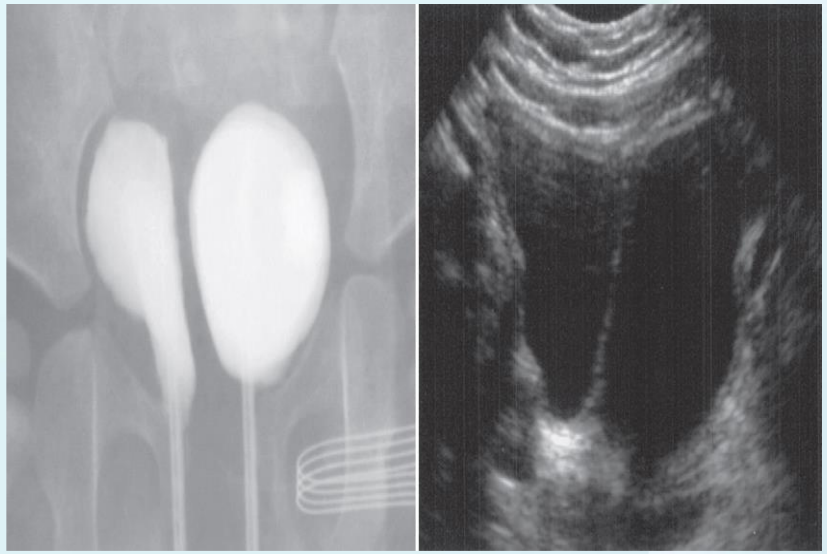
Bladder Diverticulum…

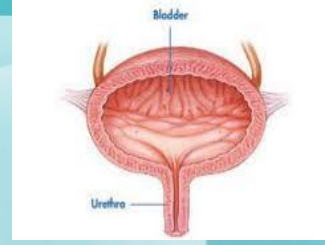
- Types:
  - Primary diverticula
    - arise as a localized herniation of bladder mucosa at the ureteral hiatus and are most likely caused by a congenitally deficient bladder wall.
  - Secondary para-ureteral diverticula
    - are acquired and develop due to existing infra- vesical obstruction.
- Symptomatic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



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.



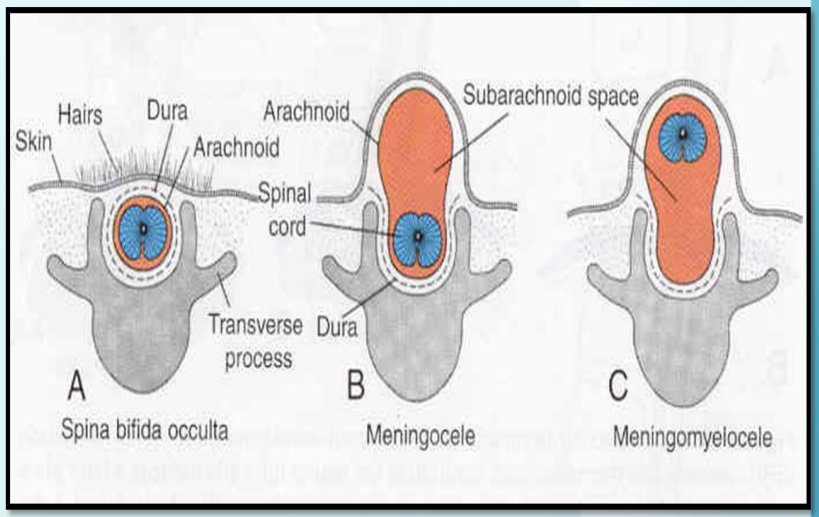


Bladder Duplication…

- 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

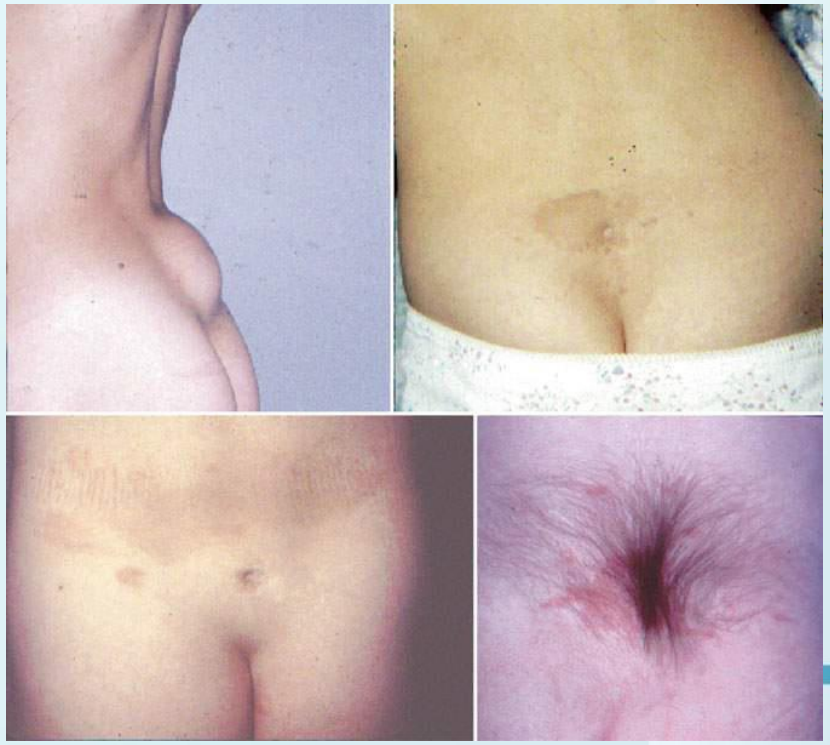
NEUROSPINAL DYSRAPHISMS

- 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!