Pediatric urinary disorders

Objectives:

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

Resources:

- **Davidson's.**
- **Slides**
- **Surgical recall.**
- **Raslan's notes.**

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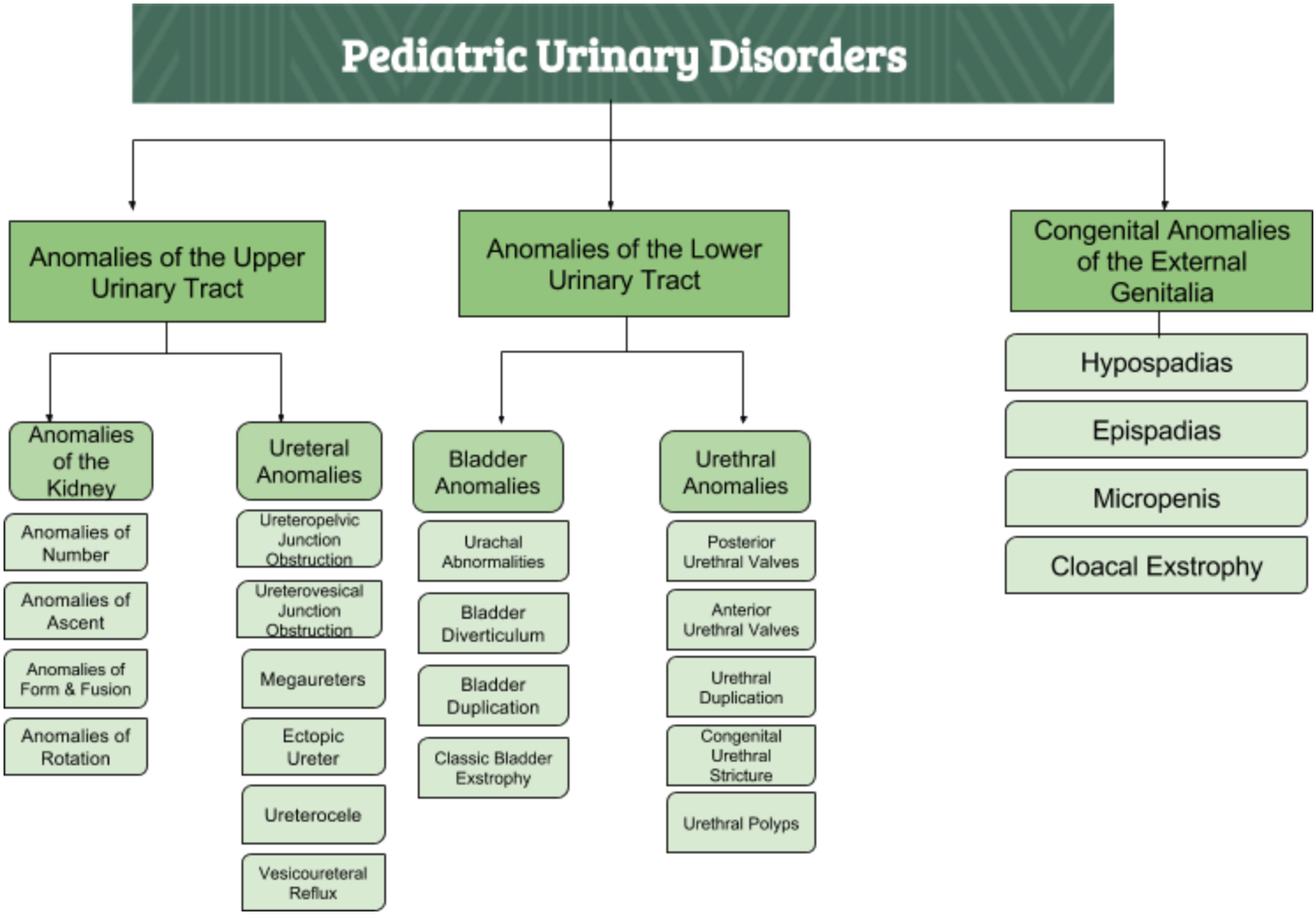
Once you stop learning
you start dying.


Pediatric Urinary Disorders
Anomalies of the Upper Urinary Tract
Anomalies of the Lower Urinary Tract
Congenital Anomalies of the External Genitalia
Anomalies of the Kidney
Anomalies of Number
Anomalies of Ascent
Anomalies of Form & Fusion
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Ureteral Anomalies
Ureteropelvic Junction Obstruction
Ureterovesical Junction Obstruction
Megaureters
Ectopic Ureter
Ureterocele
Vesicoureteral Reflux
Bladder Anomalies
Urachal Abnormalities
Bladder Diverticulum
Bladder Duplication
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Urethral Anomalies
Posterior Urethral Valves
Anterior Urethral Valves
Urethral Duplication
Congenital Urethral Stricture
Urethral Polyps
Hypospadias
Epispadias
Micropenis
Cloacal Exstrophy

Upper Urinary Tract Anomalies

The normal position of the kidneys in the fetus is in the pelvis, and the orientation is anterior.
But at the age of delivery which is about about 32 weeks, the position of the kidneys are in the flank area, and the orientation is directed medially.

Kidney Anomalies:

❖ Anomalies of Number

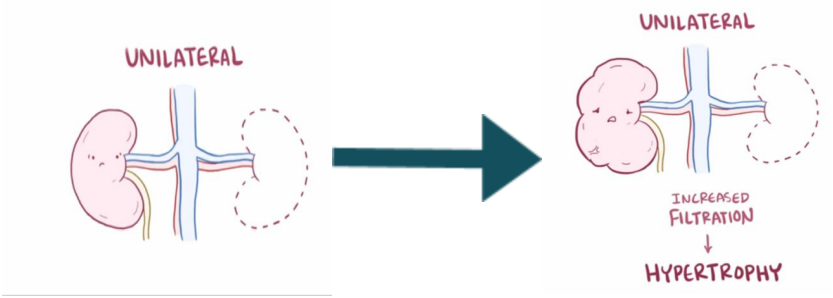
1/Renal Agenesis:

A/Unilateral Renal Agenesis (URA)

Renal agenesis

This occurs when one kidney is absent.
If the normal function of the kidney is preserved and the person is asymptomatic, this condition cause no harm.
If the person doesn't have a left kidney for example, he will still have the left suprarenal gland cause it's of different embryological origin.
The kidney will have to work double time to compensate for the missing one, this will lead to hypertrophy due to increased filtration.

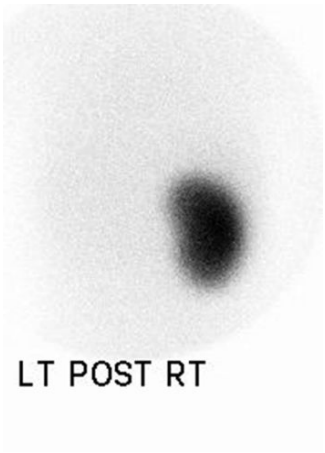
- 1 in 1100 births.
- Male : Female of 1.8 : 1
- The left side is absent more frequently .
- The ipsilateral ureter is completely absent in 50%.
- Anomalies of other organ systems are found frequently in affected individuals CVS,GIT,MSC
- Müllerian duct abnormalities occur in 25% to 50% of cases of females with URA compare with wolffian duct anomalies in 10% to 15% of males with URA.
- Approximately one fourth to one third of women with müllerian duct anomalies are found to have URA.

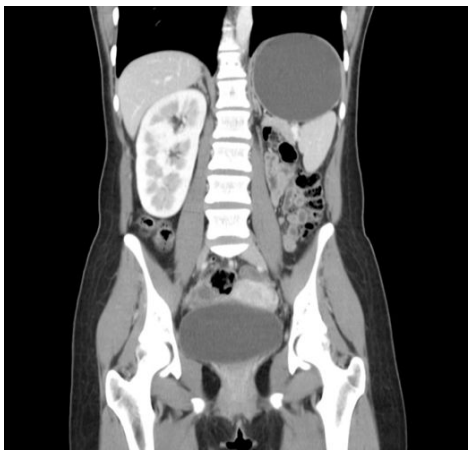


Diagnosis:

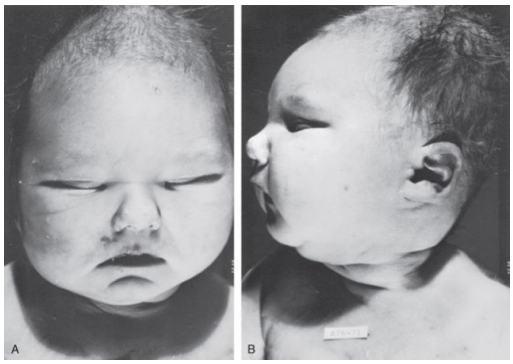
- CT Abdomen
- DMSA

If we want to know the presence of tissue we use STATIC nuclear medicine, and it will tell us if the kidney is present or absent. So it is diagnostic of URA. Ultrasound is not sensitive in this case, even if the kidney is not visualized it could be ectopic.





B/Bilateral Renal Agenesis:

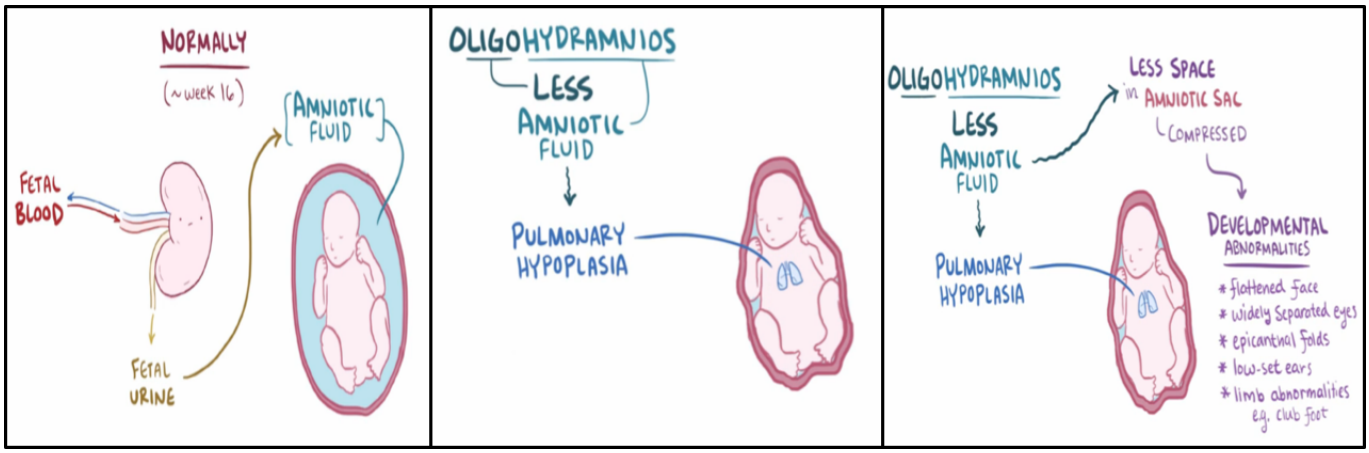


This case is clinically significant, because there is no compensation for the lost function of the kidneys.
The **amniotic fluid** is the protective liquid contained by the amniotic sac of a gravid Amniote. This fluid serves as a cushion for the growing fetus, but also serves to facilitate the exchange of nutrients, water, and biochemical products between mother and fetus. Amniotic fluid is generated from maternal plasma, and passes through the fetal membranes by osmotic and hydrostatic forces. When fetal kidneys begin to function in about week 16, fetal urine also contributes to the fluid. The amniotic fluid in the 20th week of gestation is formed from the urine.
If there's no kidneys → no urine → no amniotic fluid. So the fetus will mostly die. The cause of death is respiratory distress. Also the amniotic fluid protect the fetus from the contractions of the uterus, so if the amniotic fluid is absent this could affect the fetal development, which will result in the classical potter appearance.

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

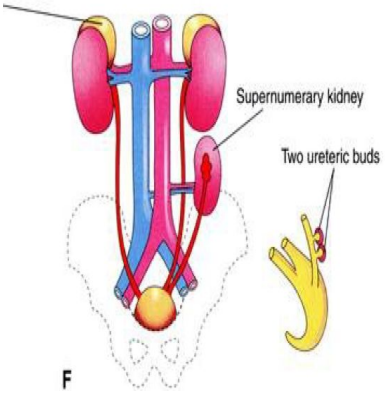
Characteristics:

- ○ Potter's syndrome.
- ○ Oligohydramnios
- Ureters are almost always absent.
- Bladder is either absent or hypoplastic.
- Adrenal glands are usually positioned normally.
- Müllerian duct anomalies are commonly observed.



2/Supernumerary Kidney:

This condition occurs when the patient has more than two kidneys, but it is extremely rare to have more than 3 kidneys.
All the kidneys are functional, and it's discovered incidentally because usually they are asymptomatic.

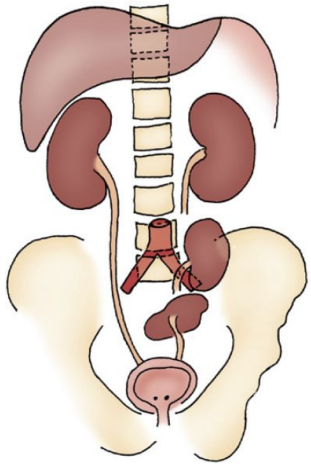


- Definitive accessory organ with its own collecting system, blood supply, and distinct encapsulated parenchyma.
- Usually asymptomatic and found incidentally.
- Either completely separate or loosely attached to the kidney on the ipsilateral side.
- The ureteral inter-relationships on the side of the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

❖ Anomalies of Ascent

1/Simple Renal Ectopia

It's abnormal position of the kidney but in the same side.The ureter is in the same side.



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

We don't intervene unless it's symptomatic.We don't need to correct the position of the kidney because it has already taken the blood supply from that area.

2/Cephalad Renal Ectopia

3/Thoracic Kidney

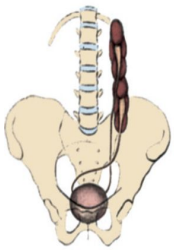
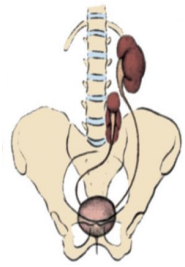
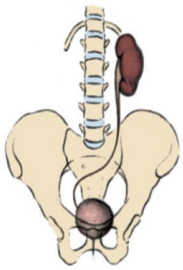
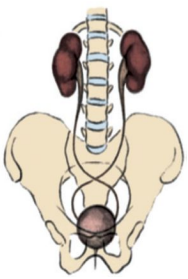
❖ Anomalies of Form and Fusion

1/Crossed Renal Ectopia with and without Fusion

It's abnormal position of the kidney but in the opposite side. The ureter is not crossed to the opposite side.

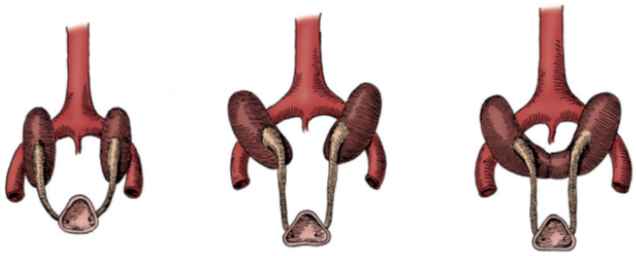
- Crossed ectopia: kidney is located on the side opposite from that in which its ureter inserts into the bladder.
- 90% are fused with their mate
- the superior pole of the ectopic kidney usually joins with the inferior aspect of the normal kidney.
- The ureter from each kidney is usually orthotopic(meaning that if the kidney crossed to the right side, the ureter will still be in the left side).

Knowing the different types is not important, what's important is knowing how to diagnose it, and to differentiate between simple and crossed.

Types of Crossed Renal Ectopia			
			
Crossed renal ectopia with fusion	Crossed renal ectopia without fusion	Solitary crossed renal ectopia	Bilaterally crossed renal ectopia

2/Horseshoe Kidney

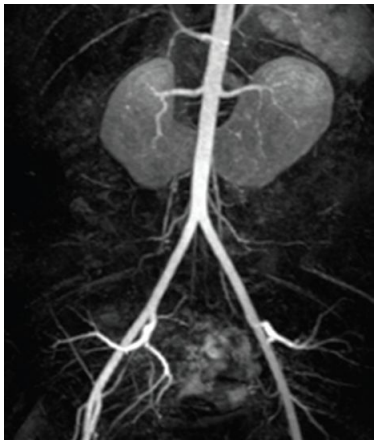
If it's asymptomatic we don't intervene, unless it's associated with other anomalies like hydronephrosis.



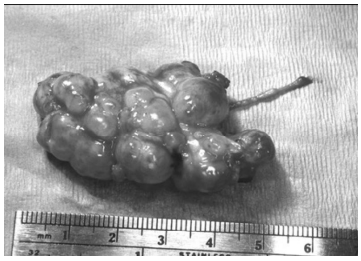
- Occurs 1 in 400 persons.

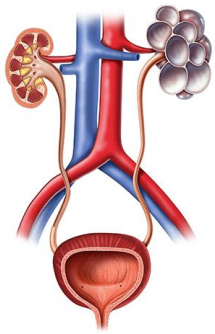
90% of the kidneys is attached at the lower border, and 10% in the upper border.

- 80% the isthmus is bulky and consists of parenchymatous tissue .
- The calyces:
  - normal in number
  - atypical in orientation.
  - pelvis remains in the vertical or obliquely lateral plane
- Horseshoe kidney is frequently found in association with other congenital anomalies.
- UPJ obstruction in one third.
- 60 % asymptomatic.
- The blood supply can be quite variable (for surgical manner)
- They are totally separate kidneys only connected by isthmus



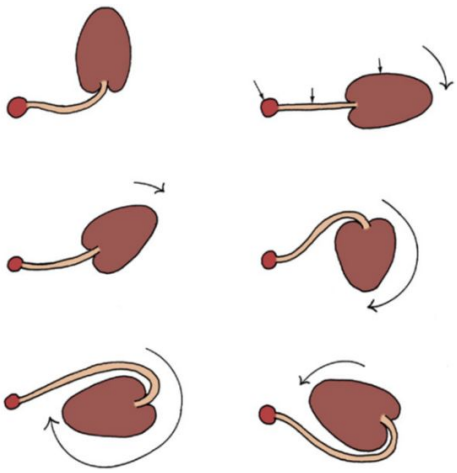
3/Multicystic Dysplastic Kidney (MCDK)





❖ Anomalies of Rotation

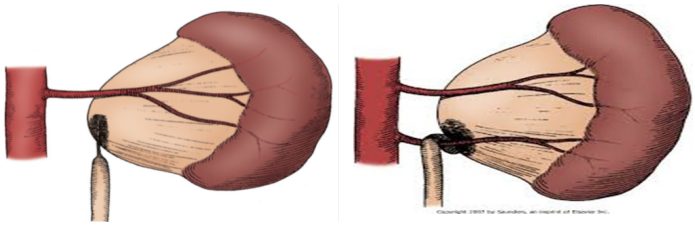
The most important thing to know is if the kidneys in the normal position or malpositioned.



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

Ureteral Anomalies:

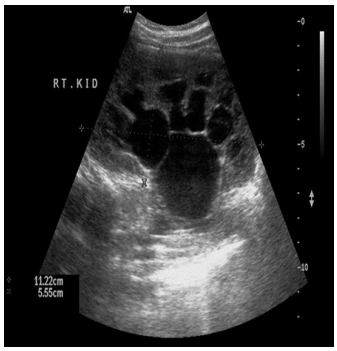
1/Ureteropelvic junction (UPJ) obstruction

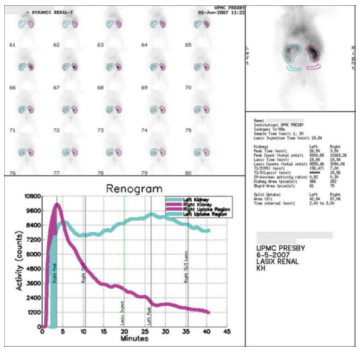


Also known as infra-uretero junction

Presentation:

- Incidental in Neonates
- Incidental in Children
- Symptomatic: (may come with all urinary congenital disorders)
  - UTI
  - Pain
  - Mass
  - Hematuria
  - Stone



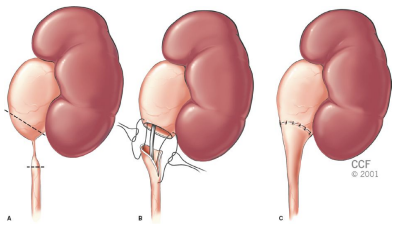


Diagnosis:

- Ultrasound
- Dynamic renogram (MAG3)(To confirm diagnosis)

Management:

- Dismembered Pyeloplasty[1]



2/Ureterovesical junction (UVJ) obstruction

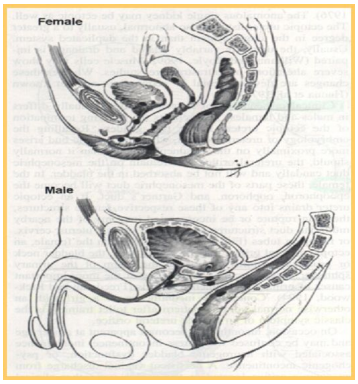
3/Megaureters

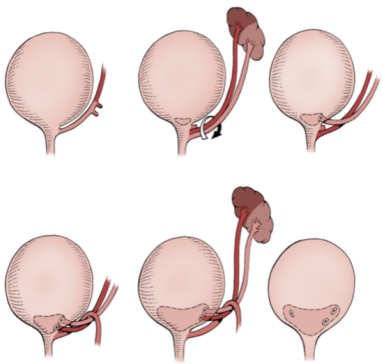
4/Ectopic Ureter

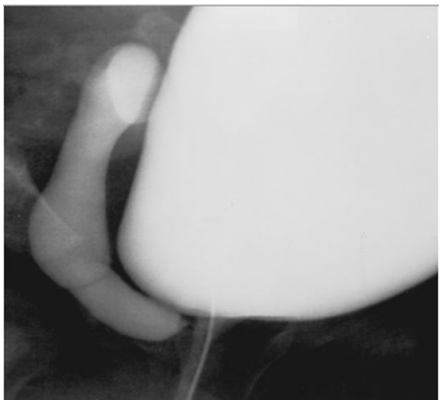
- An ectopic ureter is any ureter, single or duplex, that doesn't enter the trigonal area of the bladder .
- In a duplex system the ectopic ureter is inevitably the upper pole ureter due to its budding from the mesonephric duct later (more cephalad) than the lower pole ureteral bud.
- In females the 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. (Due to the ureter open in an area there is no sphincter distal to it)

- 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.
In males it will be presented with recurrent epididymitis, epididymo-orchitis.

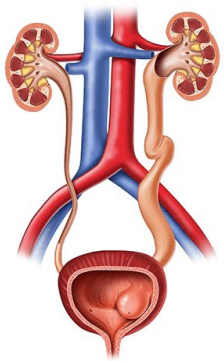






[1] It's a procedure where we excise the area of stenosis, then attach the normal ureter to the pelvis

5/Ureterocele



By itself it's not needed to be treated, unless it's causing obstruction or reflux.

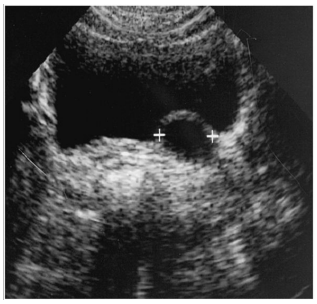
- A cystic dilation of the distal aspect of the ureter
- Located either within the bladder or spanning the bladder neck and urethra.

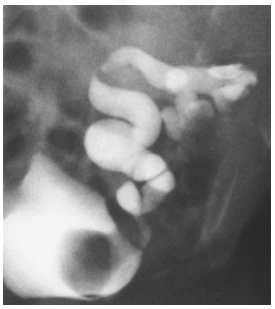
Presentation:

- Antenatal (U/S)
- Urine retention
- Infection
- Calculus formation

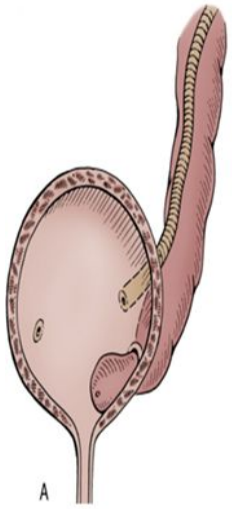
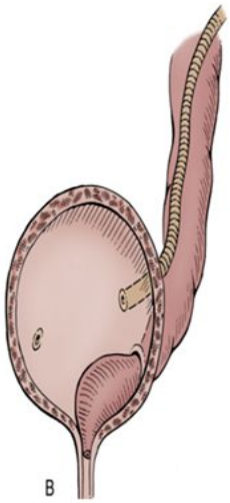
Diagnosis:

- Ultrasound (In the ultrasound the ureterocele will present as cystic like structures inside the bladder)
- MCUGIVP





Types:

Intravesical	Extravesical
	
▪ Orthotopic ▪ Simple ▪ Adult type	▪ Ectopic ▪ Duplex system ▪ Infant type

Treatment:

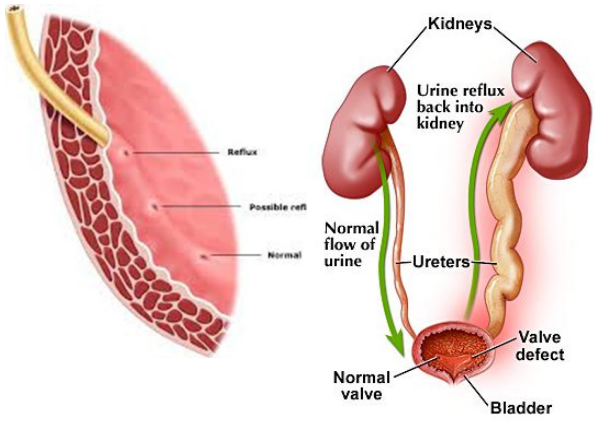
If we have a patient with ureterocele present with obstruction we do cystoscopy
And we make punctures to the ureterocele to drain the urine.Used in emergency.

6/Vesicoureteral Reflux VUR

Normal anti-reflux mechanism “Flap valve ”

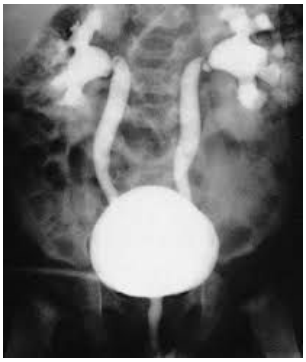
1. Oblique course as it enters the bladder.
2. Proper muscular attachments to provide fixation.
3. Posterior support to enable its occlusion.
4. Adequate submucosal length. If shorter the chance will be higher

In the primary reflux, the entrance of the ureter to the bladder is abnormal, but in the secondary reflux there's anomaly in the bladder. It’s important to differentiate between them.



Presentation

- Asymptomatic
  - Prenatal
  - Fluctuated dilatation
- Febrile UTIs

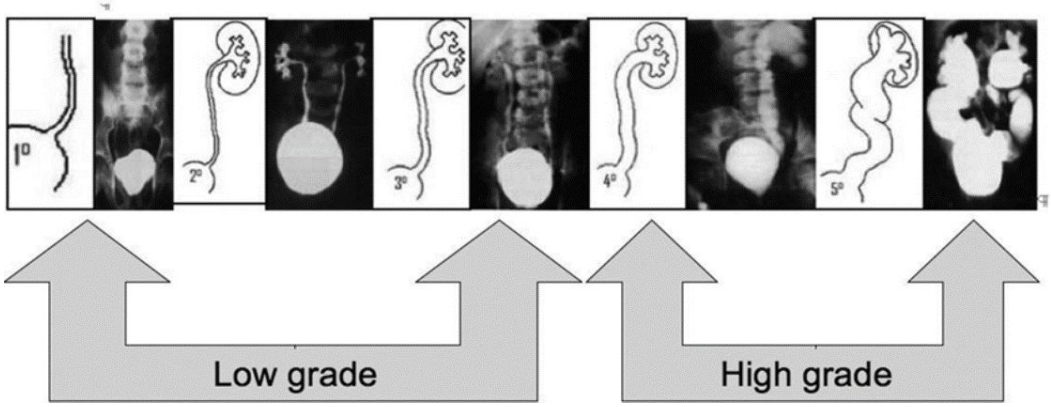


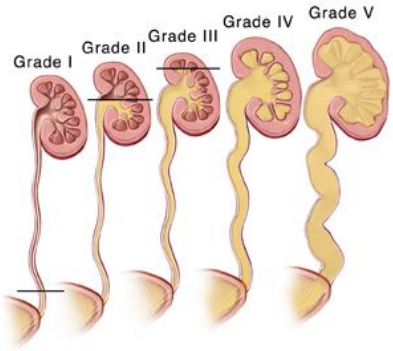
Diagnosis:

- **MCUG**[2]

When we do ultrasound and there’s dilatation, doing it again after 30 min there’ll be no dilatation, this is the pathognomonic finding in the ultrasound, so If we do the US while the fetus is voiding, the urine will go up to the kidney and it will be dilated, but if we do it while the bladder is empty, the kidney won’t be dilated, this is called alternating hydronephrosis.

Staging: not important



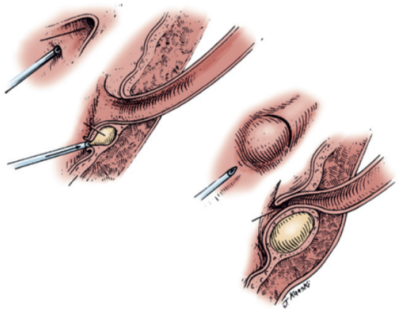


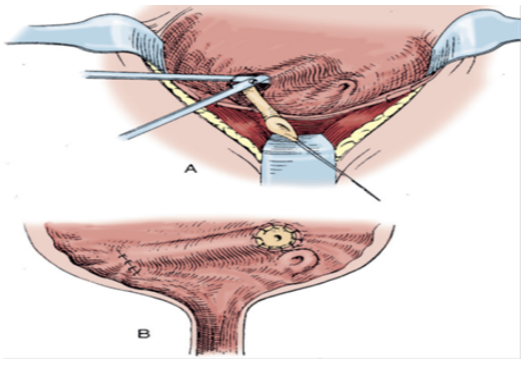
Management:

- Prophylactic antibiotic.It’s ⅓ of the normal dose, and the frequency is once daily.

For example if the patient is presented with reflux immediately after birth we give amoxicillin, but we don’t give Trimethoprim/sulfamethoxazole because of it’s hepatotoxic effect on newborns from 1-3 months old.

- Surgical treatment.
  - Endoscopic (cystoscopy) treatment (80%)
  - Ureteral reimplantation.





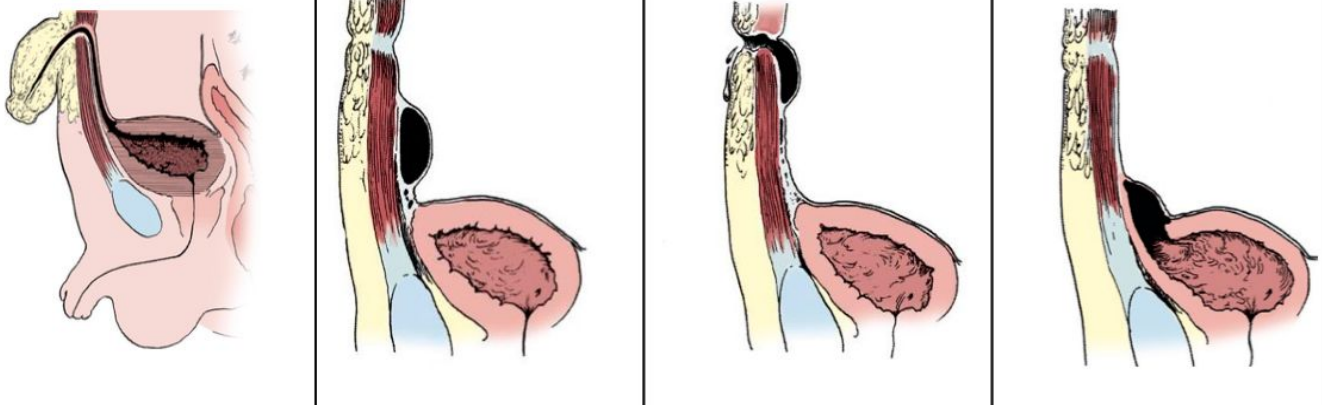
[2] micturating cystourethrogram

Lower Urinary Tract Anomalies

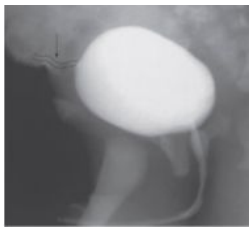
Bladder Anomalies:

1/ Urachal Abnormalities.

- Urachal Anomalies are usually detected postnatally due to umbilical drainage.



1. Patent Urachus (dripping through the umbilicus)
2. Urachal Cyst
3. Umbilical- Urachal Sinus
4. Vesicourachal Diverticulum



Diagnosis:

- Imaging possibilities include Ultrasound, CT & VCUG (MCUG).
- *There is a continuation between the bladder & the umbilicus and the contrast going inside*

Management:

Conservative Treatment with observation	Justified in asymptomatic cases due to possible spontaneous resolution.
Infected urachal remnants	Initially treated with drainage and antibiotics, followed by surgical excision.
Nonresolved urachal remnants	Should be excused due to the increased risk of adenocarcinoma formation later

2/ Bladder Diverticulum.

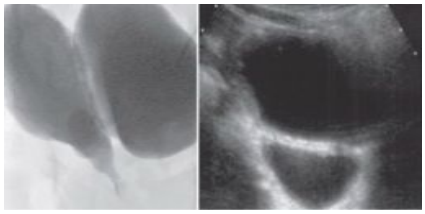
- Weak bladder muscle, not formed well leading to outpouching of mucosa. As pocket fills with urine, it may cause *infection* or *urinary retention*.

Types:

Primary Diverticula	Secondary Paraureteral Diverticula
Arises as a localized herniation of *bladder mucosa* at the ureteral hiatus & are most likely caused by a *congenitally* deficient bladder wall.	Are acquired and develop due to existing infravesical obstruction. It occurs usually multiple

Diagnosis:

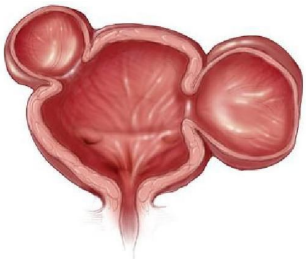
Bladder diverticula can be detected on a prenatal ultrasound.
The gold standard, is VCUG which will reveal possible accompanying VUR



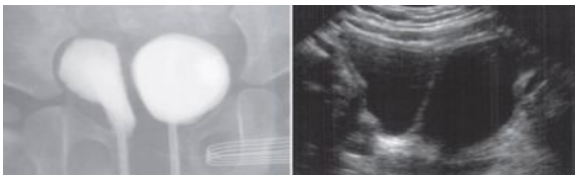
Treatment:

Asymptomatic	Not Treated
Symptomatic	Especially in conjunction with VUR, should be treated *surgically.*

3/ Bladder Duplication. rare



- Two bladders and each ureter opens in one. They either have a common urethra or a separate one.
- Very rare.
- Bladder duplication is often associated with duplication anomalies of the external genitalia & lower GIT.

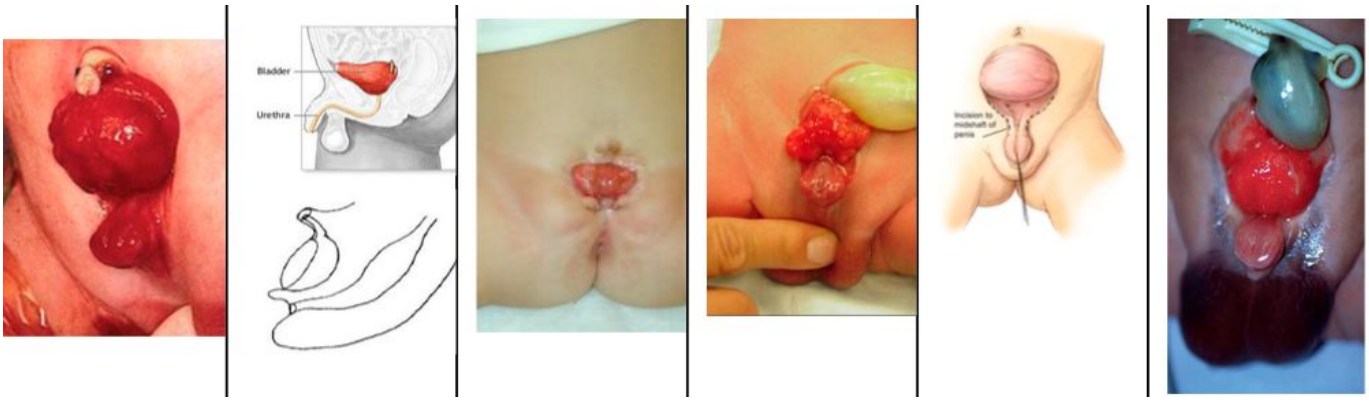


Management:

Initial treatment	- Directed towards, renal preservation - Prevention of infections
Long term goals	- Achieving continence and reconstructing the internal and external genitalia - Due to the rarity of the disease & the large variety of presentation, the surgeries must be individualized.

4/ Classic Bladder Exstrophy.

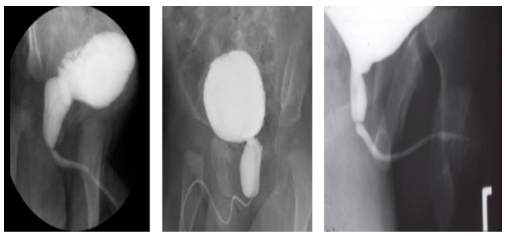
- The incidence of bladder exstrophy has been estimated as between 1 in 10,000 and 1 in 50,000.
- The bladder is usually covered by skin, subcutaneous tissue & 3 muscle layers. If you cut those coverings and anterior wall of the bladder you will get it.
- You can also find it in neonate delivery, you will see the posterior wall, ureters & urine come out.



Urethral Anomalies:

1. Posterior urethral Valves 2.Anterior Urethral Valves 3. Urethral Duplication 4.Congenital Urethral Stricture 5. Urethral Polyps

1/ Posterior Urethral Valves (PUV).



- 1 in 8000 to 25,000 live births.
- Makes up 10% of urinary obstruction diagnosed in utero.
- *Most common* cause of urine retention in male infants.
- *50%* have renal impairment.
- The bladder & the kidneys developed under high pressure & resistance.

Presentation:

- Antenatal
- Urine retention
- UTI
- Poor urinary stream
- Urinary Incontinence
- CRF (ESRD)

Associated findings:

- Oligohydramnios
- Bilateral renal dilatation
- VUR in 40%
- Valve bladder
- Renal impairment

Diagnosis:

- US suspected **Findings:**
  - Dilated posterior urethra
  - Thick wall bladder
  - Dilated kidney and ureters.
- MCUG (Confirmatory) "christmas tree bladder (trabeculation), & mismatch of urethral caliber"

How do we know the exact site for the valve? By filling defect.

Management:

Initial Treatment	- Feeding tube insertion - Start antibiotic prophylactic - Ultrasound - MCUG
Surgical Treatment	- Endoscopic valve ablation - Cutaneous vesicostomy (temporary bc pt is infant)

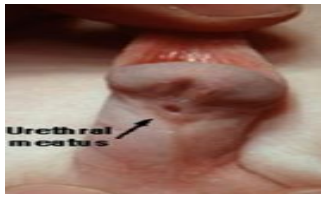
Congenital Anomalies of the External Genitalia

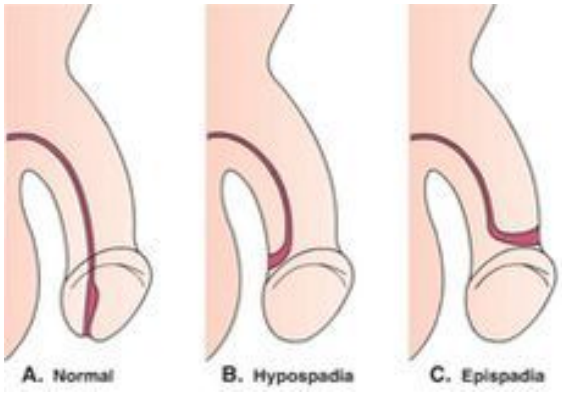
- **Absolute Contraindication to do Circumcision!**

1/ Hypospadias.

2 Types: I. Distal hypospadias II. Proximal Hypospadias

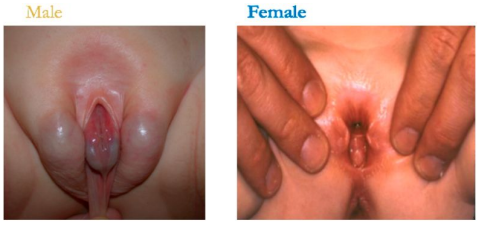
- Abnormal position of the EUM on the ventral surface
- NO Circumcision *(because we use the skin for repair)*
- 6-9 months repair *(because of the risk of anesthesia)*





2/ Epispadias.

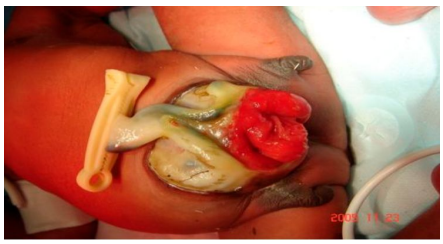
- Abnormal position of the EUM on the dorsum surface.



3/ Micropenis.

4/Cloacal Exstrophy. pt has no anus compare to bladder exstrophy

A child with cloacal exstrophy is born with the bladder and a part of the intestines open to the outside.The bony pelvis is also split open like a book.
In males the penis is either flat and short or sometimes split. In females the clitoris is split and there may be two vaginal openings. The intestine may be short and the anus may not open.



1.Prune-Belly Syndrome

The incidence: 1:29,000 to 1 in 40,000 live birth.

3 major findings:

- Deficiency of the abdominal musculature
- Bilateral intra- abdominal testes
- Anomalous urinary tract

Other names:

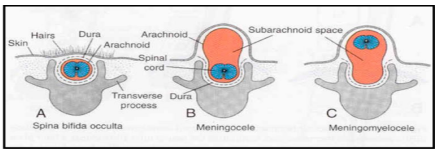
- Triad Syndrome
- Eagle-Barrett Syndrome
- Abdominal Musculation Syndrome

- US is the same as PUV you can differentiate it **by examination:**
- Impalpable testes because of undescended testes.
- Anterior abdominal muscle either absent or hypoplastic and becomes one layer.

Check bowel movement.

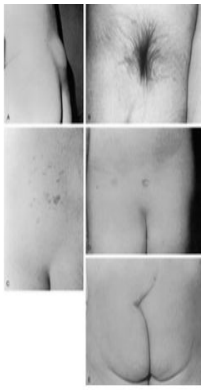
2.Neuro Spinal Dysraphisms

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



- Cutaneous lesions occur in 90% of children with various occult dysraphic states[3].
- These lesions vary from:

- Small lipomeningocele
- Hair patch
- Dermal vascular malformation
- Sacral dimple
- Abnormal gluteal cleft



3.Antenatal Hydronephrosis

Causes:

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

4.Weigert-Meyer Rule

Applies to duplex collecting systems, when complete, duplicated ureters insert separately into the bladder. The **Weigert-Meyer rule** states that. the upper pole ureter is the ectopic ureter and its orifice inserts inferomedially in the bladder in relationship to the lower pole normal ureter.

Recall:

What are the contraindications to circumcision?
Hypospadias etc. because the foreskin might be needed for future repair of the abnormality.
What is Eagle-Barrett's syndrome?
A.k.a. Prune belly; congenital inadequate abdominal musculature (very lax and thin)

[3] Differ than Congenital Dermal Melanocytosis (Mongolian Spot) which is found naturally as blue-gray pigmentation usually on the sacral area of healthy infants.