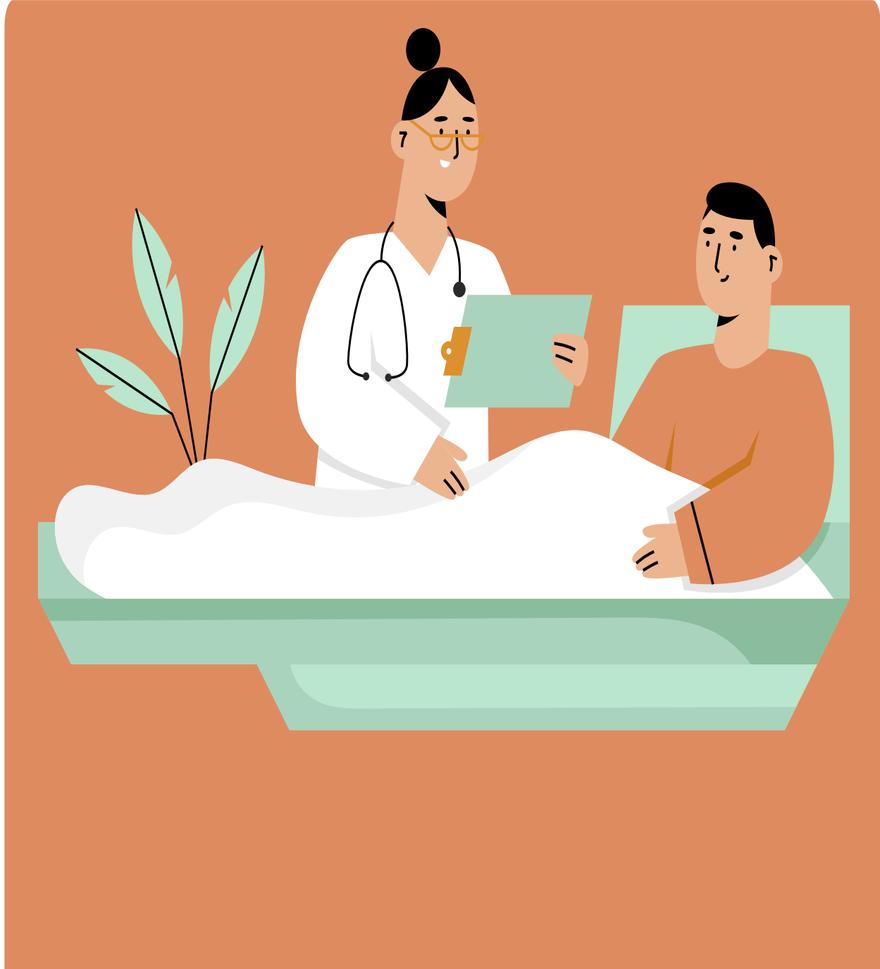
MED437
KING SAUD UNIVERSITY

SURGERY TEAM 437



Pediatric Urinary Disorders

Objectives:

1. *Identify* the common congenital anomalies.
2. How to detect this anomaly on radiological investigations.
3. îlmportant steps in management.

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Color Index:

- Important
- Doctor's Notes
- Extra
- Davidson's

Editing File / Feedback

Congenital Urinary Disorders

Anomalies of:

Anomalies of the Upper Urinary Tract

Kidney anomalies

Most of the congenital kidney abnormalities are asymptomatic.

- Number
  - Renal Agenesis
    - Unilateral
    - Bilateral
  - Supernumerary Kidney
- Ascent
  - Simple renal ectopia
  - Cephalad renal ectopia
  - Thoracic kidney
- Form and Fusion
  - Crossed renal ectopia
    - With fusion
    - Without fusion
  - Horseshoe kidney
  - Multicystic dysplastic kidney
- Rotation

Ureter anomalies

- UreteroPelvic Junction (UPJ) Obstruction
- UreteroVesical Junction (UVJ) Obstruction
- Megaureters
- Ectopic ureter
- Ureterocele
- Vesicoureteral reflux (VUR)

Anomalies of the Lower Urinary Tract

Urinary bladder anomalies

- Urachal abnormalities
- Bladder diverticulum
- Bladder duplication
- Classic bladder exstrophy

Urethral anomalies

- Posterior urethral valves
- Anterior urethral valves
- Urethral duplication
- Congenital urethral stricture
- Urethral polyps

Congenital anomalies of the external genitalia

- Hypospadias
- Epispadias
- Micropenis
- Cloacal exstrophy

You should know the anomaly, how it presents, how to diagnose it and the outline of treatment.
In the investigations focus on **the pictures** because they might come with a case scenario

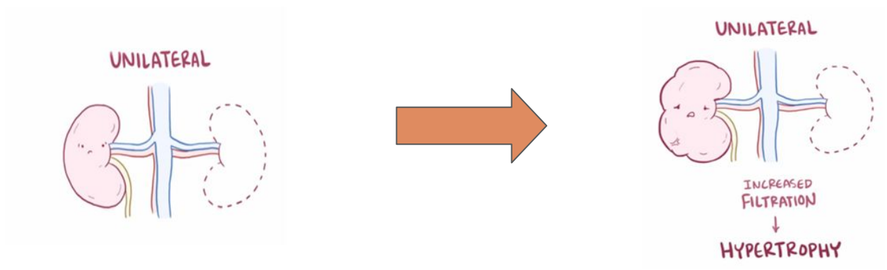
Upper Urinary Tract Anomalies: Kidney

A. Anomalies of Number

Renal Agenesis

(RA) is the complete absence of development of one or both kidneys.

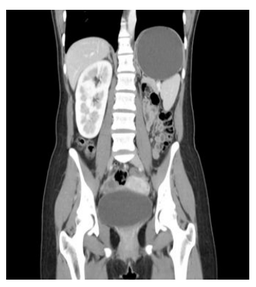
Unilateral Renal Agenesis (URA)



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

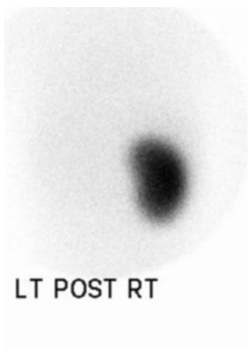
Presentation

- **Prenatal US**
- **Incidentally**: Abdominal US - Abdominal CT Some may present with abdominal pain, you do them an ultrasound and you will find a single kidney.



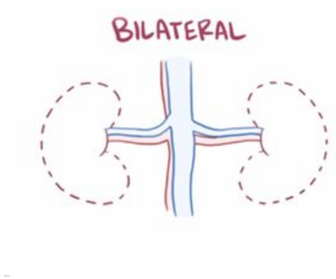
Diagnosis

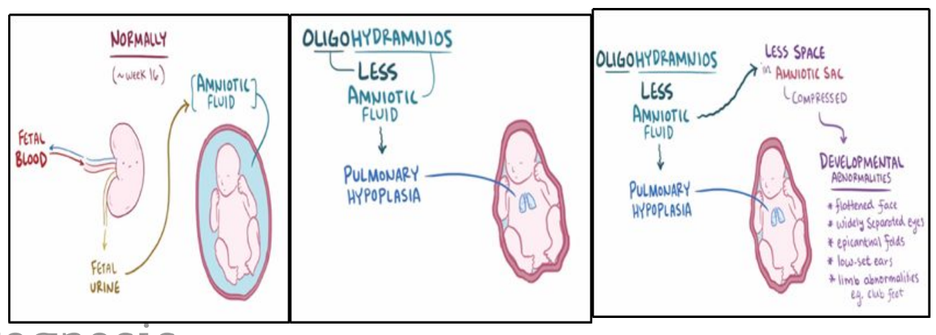
- CT abdomen
  Tell you the position of the kidney
  And if its ectopic or not.
- Nuclear study (DMSA) :
  (**confirmatory**)
  (**DMSA**)= radionuclide scan
  that uses dimercaptosuccinic acid



The best test to confirm the the absence of the kidney is DMSA ***DMSA will show you that the kidney is not uptaking the radionuclide

Bilateral Renal Agenesis

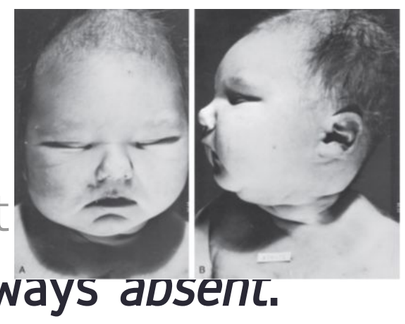




Prognosis:

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

The characteristic:



- Potter's syndrome.
- Oligohydramnios
  (deficiency of amniot
- **Ureters are almost always *absent*.**
- **Bladder is either absent or *hypoplastic*.**
- **Adrenal glands are usually positioned *normally*(**Why? because the kidneys and adrenal glands don't share the same embryonic origin"**.**
- Müllerian duct anomalies are commonly observed.

Since they have anhydramnios the infants will be squished inside resulting in what we know as potter's face. Flat face, nose, and their forehead will be inside. They may also have limb deformities as well.

Management of renal agenesis:

We don't do anything for those patients but we inform them to do a regular routine urinalysis +BP measurement

Renal agenesis happens as result of failing of interaction between ureteric bud and metanephric mass.

Presentation of patients with unilateral kidney agenesis:

- These patients will usually have a normal life. It is often detected incidentally by an ultrasound. Because they are asymptomatic!! Some may present with abdominal pain, you do them an ultrasound and you will find a single kidney.

- **Investigations done to patients with unilateral kidney agenesis:**

- If you do ultrasound or a CAT scan and you don't find the kidneys in the kidney area you can't say immediately it's absent. The kidney can be in another location. i.e ectopic kidney. DMSA however can tell you if there is another kidney somewhere else.

- If the kidney is absent on one side, the vas will mostly be absent on that side as well because they share the same origin i.e mesonephric duct "wolffian duct" Female organs come from the mullerian duct.

- If a child is born with an absent kidney, the contralateral kidney **grows more and increases its filtration rate to compensate. This could be demonstrated by an ultrasound in which you could see the present kidney is not in its usual site.**

Why do those with Bilateral kidney Agenesis present with Oligohydramnios??

- After the 18th week of gestation, the kidneys of the fetus start functioning and start producing urine which makes the amniotic fluid. normally. The fetus will be voiding and drinking his urine but if the kidneys are not there, then no urine and thus no amniotic fluid will be produced resulting in Oligohydramnios or anhydramnios which affects the surfactant production resulting in severe **pulmonary hypoplasia** which could be the cause of death in those kids if you are not prepared to save them onspot. .

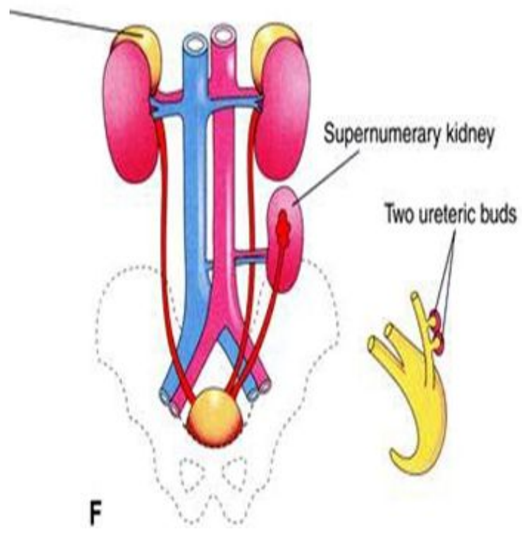
Upper Urinary Tract Anomalies: Kidney

A. Anomalies of Number: *continued*

Supernumerary Kidney (The Dr did not focus on it)

An extra kidney is very rare. It's like having multiple nipples in the breast. He could have pain, you do ultrasound, boom you find an extra kidney.

- 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. (meaning that the 2 ureters on the same side can either be connected to each other or seperated)

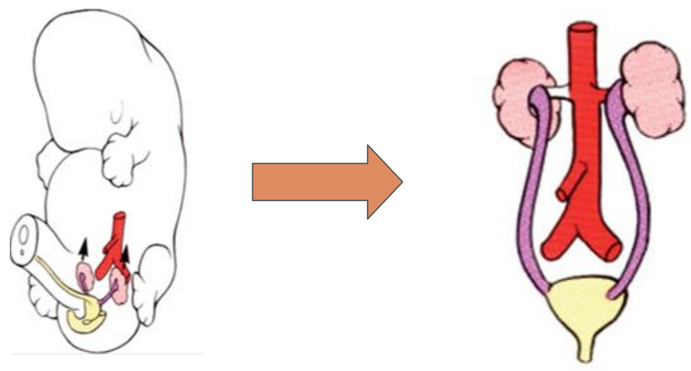


B. Anomalies of Ascent: *Embryologically the kidneys are in the pelvis then they ascend to their normal position in the abdomen*.

1-Simple Renal Ectopia Most common location is pelvic kidney

Ectopia means outside normal position in the abdomen (it may remain in the pelvis or go up to the thorax). if it arrests in the same side it is simple renal ectopia, if it crossed the other side then it is called crossed renal ectopia.

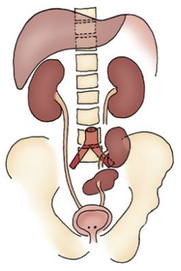
Ectopic kidneys could have normal function but they are more prone to have congenital anomalies.



- The left is more than the right.
- Pelvic ectopia has been estimated to occur in 1 of 2100 to 3000 autopsies.

Associated Anomalies:

- 50% have a hydronephrosis related to:
  - ❑ Obstruction :UPJO (ureteropelvic junction obstruction) and UVJO (ureterovesical junction obstruction) (both will be discussed later)
  - ❑ Reflux (VUR) :grade III or greater
  - ❑ Malrotation (may result in stasis which might prone to stones and infections)
- VUR is found in 30%
- The incidence of genital anomalies in the patient with ectopia is about 15%.
- Most ectopic kidneys are clinically asymptomatic



2-Cephalad Renal Ectopia

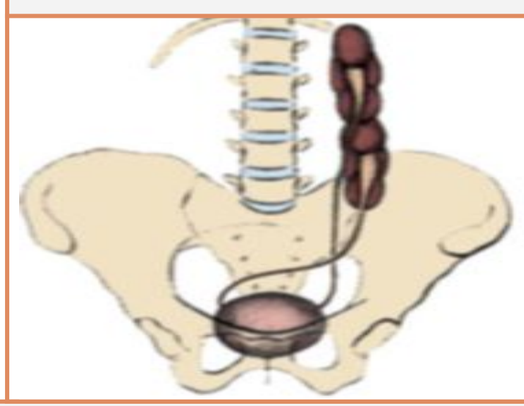
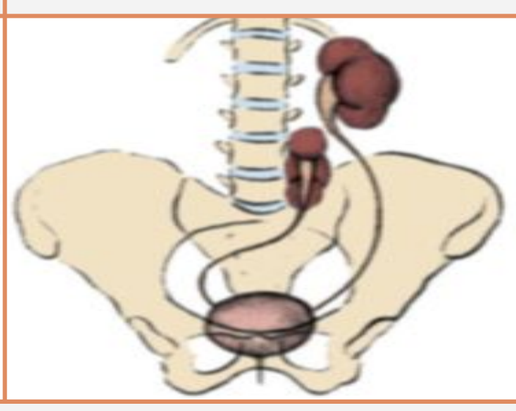
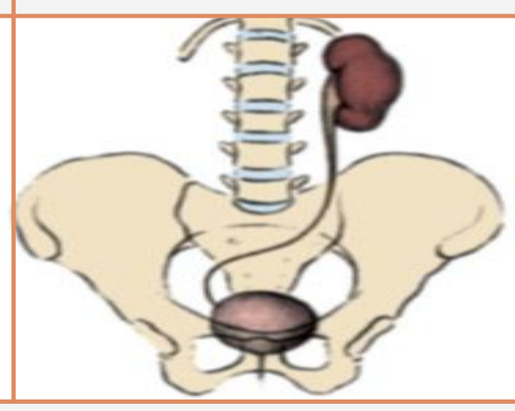
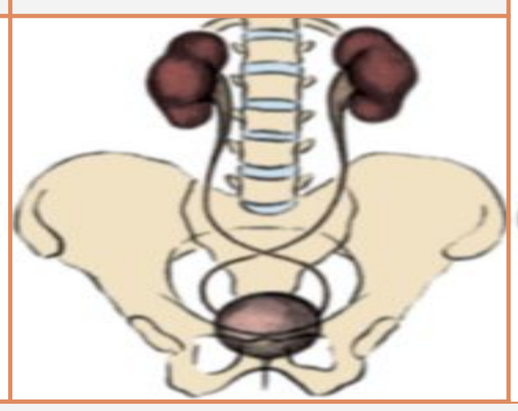
3-Thoracic Kidney

Upper Urinary Tract Anomalies: Kidney

C. Anomalies of Form and Fusion

1-Crossed Renal Ectopia (with and without Fusion)

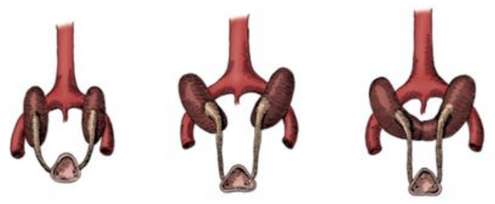
- Crossed ectopia: kidney is located on the side opposite from that in which its ureter inserts into the bladder.
- The ureter from each kidney is usually orthotopic (meaning that if the kidney crossed to the right side, the ureter will still be in the left side).
- 90% are fused with their mate.
- The superior pole of the ectopic kidney usually joins with the inferior aspect of the normal kidney. Types (it can be with or without fusion. Also one kidney can cross or both may cross).

Crossed renal ectopia with fusion (most common one)	Crossed renal ectopia without fusion	Solitary crossed renal ectopia	Bilaterally crossed renal ectopia
			

2-Horseshoe Kidney

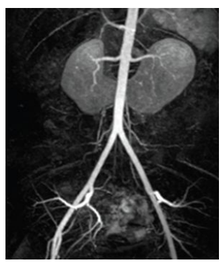
The kidneys are never at their right location. Their ascent stops at the level of inferior mesenteric artery.

Occurs 1 in 400 persons.



Morphology:

- 90% of the kidneys is attached at the lower border, and 10% in the upper border.
- The isthmus is bulky and consists of parenchymatous tissue or fibrous tissue
- The calyces:
  - ❏ Normal in number
  - ❏ Atypical in orientation.
  - ❏ Pelvis remains in the vertical or obliquely lateral plane
- **The blood supply can be quite variable** (for surgical manner)

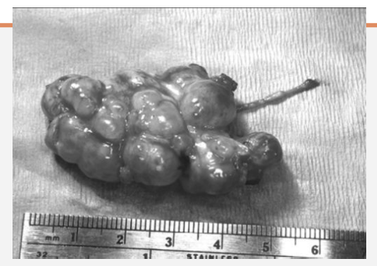


Presentation:

- Horseshoe kidney is frequently found in association with other congenital anomalies.
- UPJ obstruction in one third.
- **60 % asymptomatic**(**some will have symptoms related to stasis like recurrent infections, kidney stones. Also, they could have narrowing of the UPJ and UVJ or reflux (VUR)**
- They are totally separate kidneys only connected by isthmus (**we don't intervene for surgery**)
- Best way to confirm it is by doing DMSA.
- They are more prone to stasis thus UTIs and stones are very common among those patients.

3-Multicystic Dysplastic Kidney (MCDK)

Renal cortex is replaced by numerous cysts of multiple sizes.

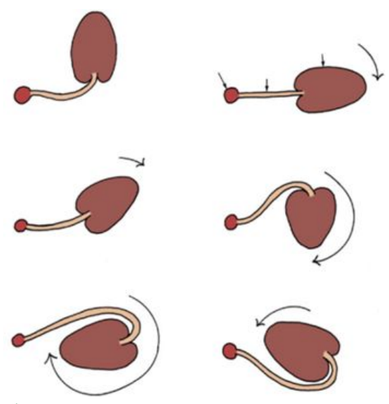


Upper Urinary Tract Anomalies: Kidney

D. Anomalies of Rotation:

Anomalies of Rotation

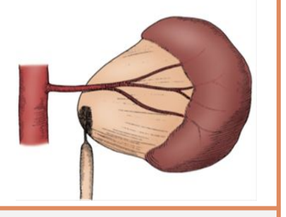
- The kidney and renal pelvis normally rotate 90 degrees ventromedially during ascent:
  - ❑ the calyces point **laterally**
  - ❑ the pelvis faces **medially**
- When this alignment is not exact, the condition is known as malrotation.
- Frequently associated with **Turner syndrome**

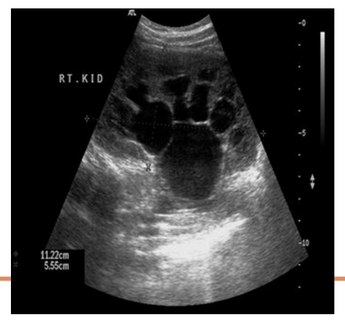
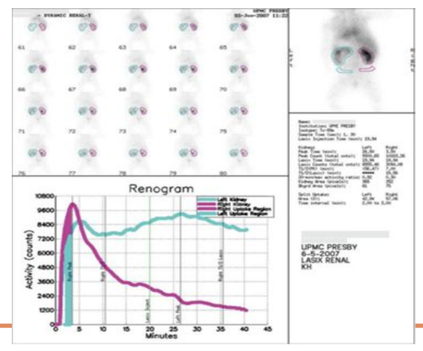
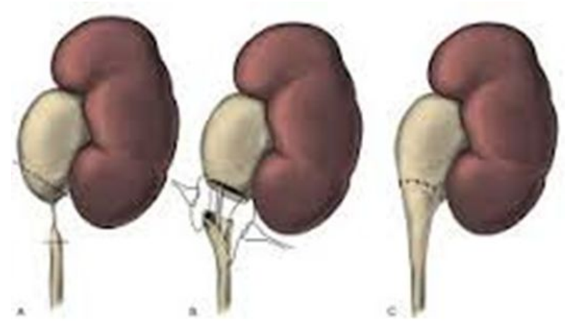


Malrotated kidney can cause stasis hence leads to infections and stone formation

Upper Urinary Tract Anomalies: Ureter

1. UreteroPelvic Junction (UPJ) Obstruction



Presentation	Diagnosis	Management
• Incidental in Neonates • **Incidental in Children** • Symptomatic: ❑ UTI From stasis ❑ Flank Pain ❑ Mass ❑ Hematuria ❑ Stone(due to stasis and infections)	• Ultrasound: Shows isolated hydronephrosis (meaning only the pelvis is dilated and the ureters are normal no hydroureter). • Dynamic renogram=Diuretic renal scan (Nuclear)*  	Dismembered Pyeloplasty (we cut the area of narrowing and reattach the ureter) 

***IMPORTANT:** How to confirm that this patient has obstruction and needs surgery?
By nuclear study called dynamic renogram There are 2 types of nuclear study:

- Static renogram: If you want to see if there is renal tissue or function issue (Is this kidney functioning or not?) like in pt. with unilateral renal agenesis.
- **Dynamic renogram: If you want to see if there is renal obstruction. (ex: GFR)**

- Usually each kidney take 50% and the summation for both is 100%, if the function decreased (less than 40%) this is indication to do surgical intervention in presence of hydronephrosis and obstruction.

Summary: decreased function + hydronephrosis ⟶ diagnosis: UPJO ⟶ intervention will be pyeloplasty!

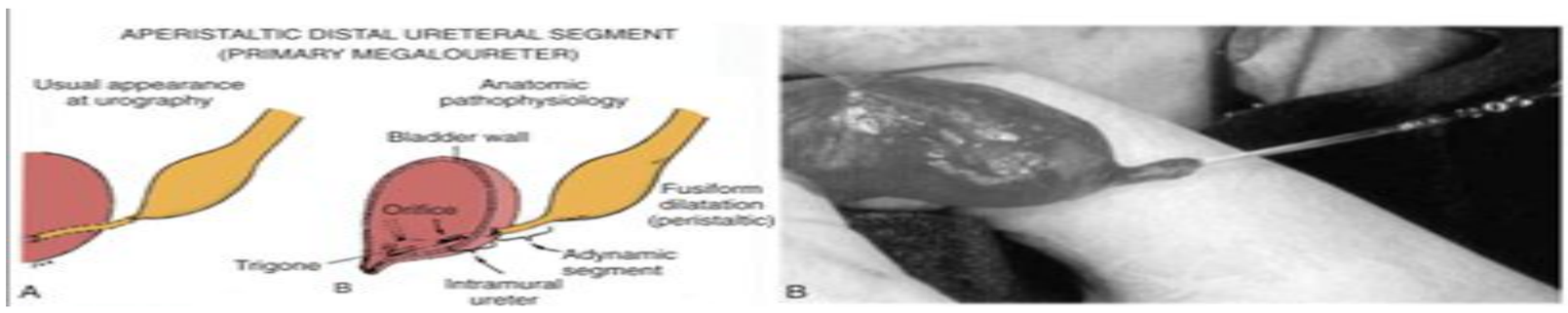
Upper Urinary Tract Anomalies: Ureter

2-Ureterovesical junction (UVJ) obstruction

And Megaureters*:

Same investigations and management as UPJO.

- On US: UPJ shows isolated hydronephrosis, because only renal pelvis is dilated. But here both renal pelvis & ureter are dilated so we call it **hydroureteronephrosis.**
- Nuclear study: decreased function with obstruction, so this is indication to do surgery; we excise the abnormal area then we implant the ureter → we call this ureteral reimplantation. ("large ureter" when a ureter is more than 7 mm)

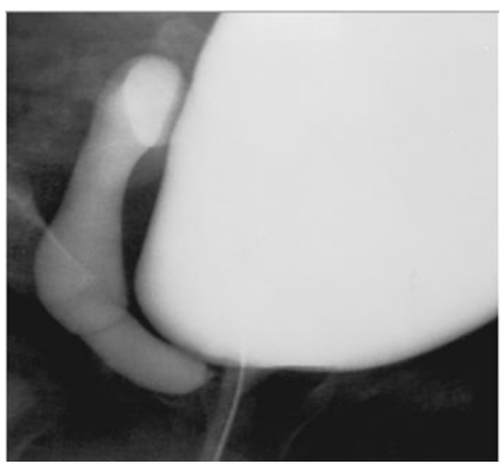


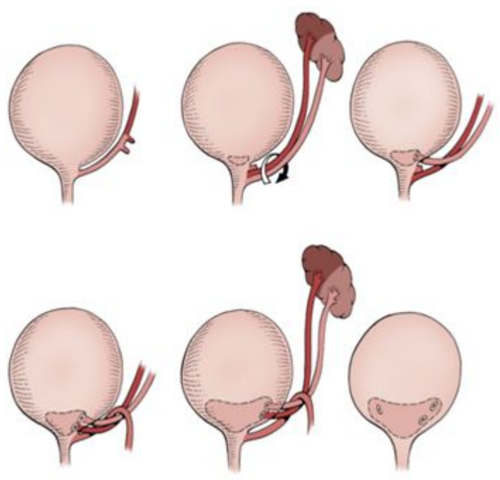
3-Ectopic ureter

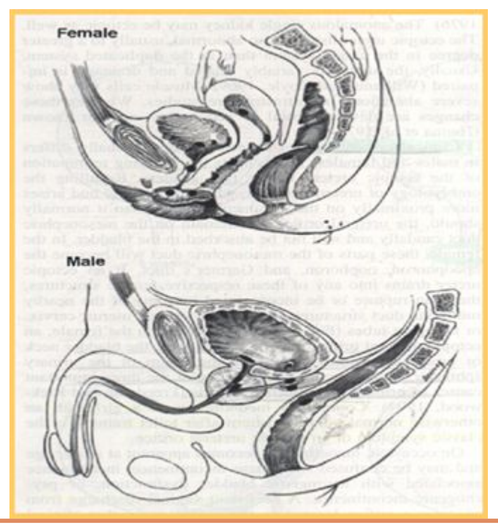
- An ectopic ureter is any ureter, single or duplex, **that doesn't enter the trigonal area of the bladder**
- In a *duplex* system the ectopic ureter is inevitably the **upper pole ureter** due to its budding from the mesonephric duct later (more cephalad) than the lower pole ureteral bud.

Presentation **(Symptoms depend on where the ureter opens or is found)**

- 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 (because it will open in the vagina which has no sphincter).**
- 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.





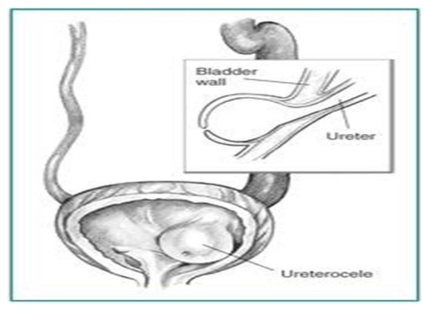


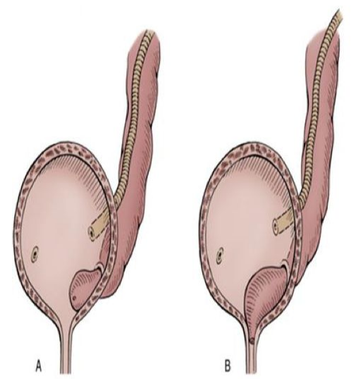
Females can present with incontinence because their urethra is short. However, men are usually dry unless it occurs distal to the sphincter or they may be presenting as with recurrent epididymitis, **epididymo-orchitis. No wetting\dribbling.**

Management: reimplantation (if kidney function is normal!) if its not functioning → nephrectomy.

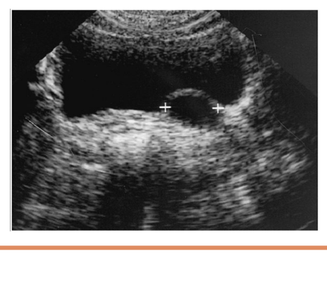
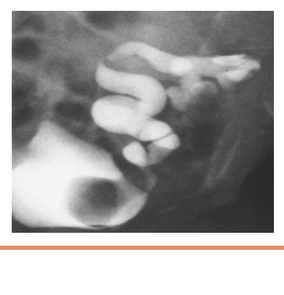
Ureteral Anomalies

5. Ureterocele (A cystic dilation of the distal aspect of the ureter).

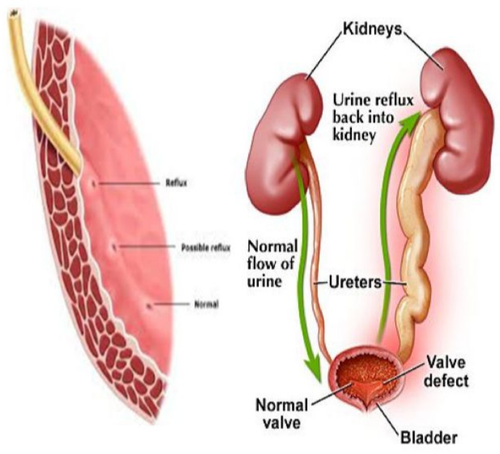




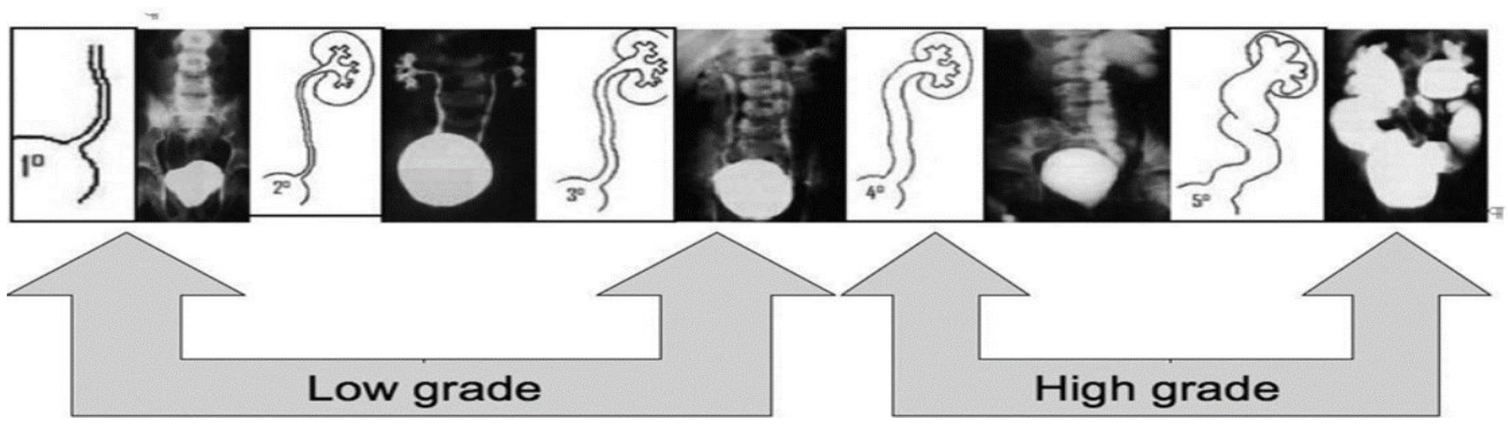
- Located either within the bladder or spanning the bladder neck and urethra.
- They usually have a megaureter. There will be cystic dilatation and a narrow opening at the end causing back pressure, ultimately they will develop hydronephrosis
- **Types:**
  - Intravesical ⟶ Orthotopic , Simple , Adult type
  - Extravesical ⟶ Ectopic , Duplex system , Infant type

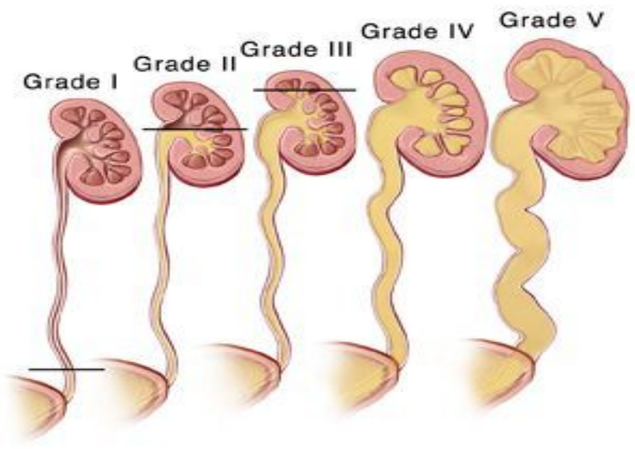
Presentation	Diagnosis	Management
• Antenatal (U/S) • If the cyst is too big, it can extend to the neck of the urinary bladder causing **Urine retention** • Infection(recurrent UTI) • Calculus formation secondary to stasis	• Ultrasound • MCUGIVP(voiding cystourethrogram; contrast study to confirm • There is a filling defect in the voiding cystogram.+ a rflux  	We go by cystoscope, incise it causing to drain (relieve the obstruction). If this does not help, then we excise it and reconnect the ureter to the bladder again(Reimplantation)

6. Vesicoureteral Reflux (VUR) Video(06:34)



- **Normal anti-reflux mechanism: "Flap valve"**
  a. Oblique course as it enters the bladder.
  b. Proper muscular attachments to provide fixation.
  c. Posterior support to enable its occlusion.
  d. Adequate submucosal length. If shorter the chance will be higher
  (Simply, he function of the flap valve is to prevent the back the flow of urine, once it's disturbed then VUR develops).
  e. Reflux per se is not a problem, we are concerned more with complications that could develop from it such as cystitis or urinary tract infections.. Here we have an open highway to the kidneys so patients can have pyelonephritis and can become severely sick and may develop scarring of the kidneys. We don't usually treat males who do not have recurrent infections or not symptomatic.
  f. Lower grade means higher chances to resolve and vice versa

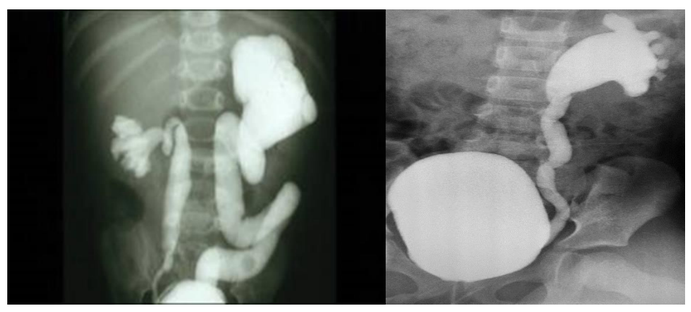
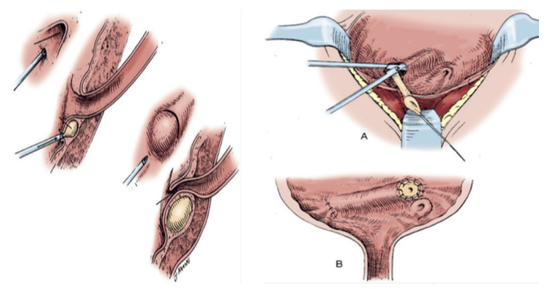




Ureteral Anomalies

6. Vesicoureteral Reflux (VUR):

Presentation	Diagnosis	Management

• Asymptomatic
  ○ Prenatal
  ○ Fluctuated dilatation
• Febrile UTIs (febrile = with fever)
• On US: it will show **hydronephrosis (not specific) so we do MCUG to confirm**
	• MCUG modality of choice helps you to confirm presence of a reflux and to know the size of the bladder.
	-If the contrast goes up=its a Reflux,Normally it will be in the bladder only
	 
		• Prophylactic antibiotic
		• Surgical treatment:
		  ○ Endoscopic (cystoscopy) treatment
		  ○ Ureteral reimplantation
		 

Surgical treatment: The classic solution is disconnecting the ureter from the bladder, create a tunnel, bring another opening then reconnect properly.

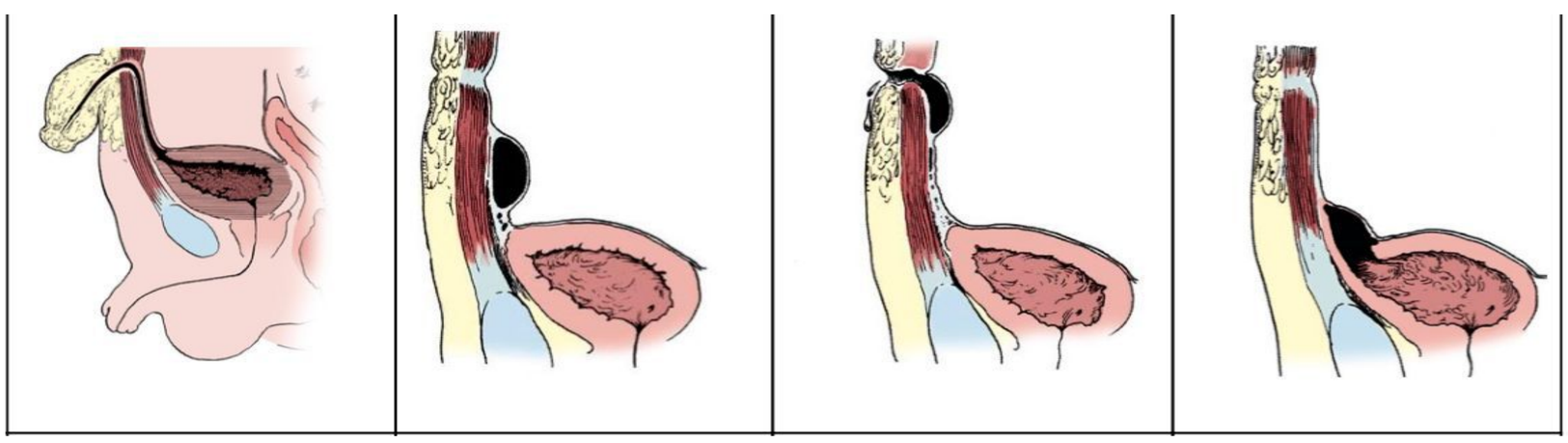
Giving them prophylactic antibiotics reduces the risk of infections but not a 100%.

We can give them injectable blocking agents like what we use for the fillers. These agents work by narrowing the dilated segments; works well when we have low grade reflux.

Bladder Anomalies

1. Urachal Abnormalities (Embryologically the bladder is connected to the umbilicus by the urachus(which is Urachus is the canal that drains the urinary bladder of the fetus that joins and runs within the umbilical cord) but normally it will degrade. If it is patent you will have these anomalies).

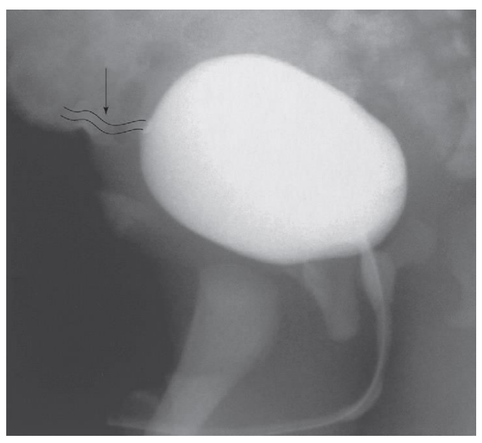
- Urachal Anomalies are usually detected postnatally due to umbilical drainage.



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

Urachus should be obliterated by the time the baby is born. If not then patent urachus develops Which causes urine to leak from the umbilicus.

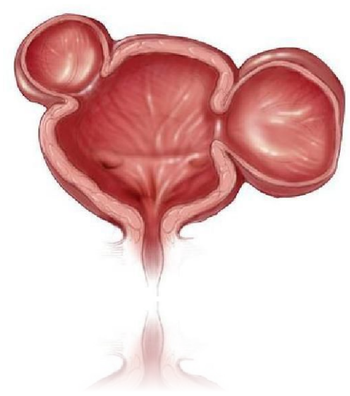
Bladder Anomalies

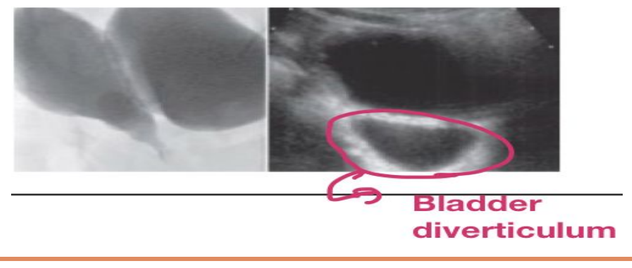
Diagnosis	Management
• Imaging possibilities include Ultrasound, CT & VCUG (**MCUG**). • There is a continuation between the bladder & the umbilicus and the contrast going inside 	• **Conservative** Treatment with observation: Justified in **asymptomatic** cases due to possible spontaneous resolution. • **Infected urachal remnants: Initially are treated with drainage and antibiotics, followed by surgical excision.** • **Nonresolved** urachal remnants should be **excised** due to the increased risk of adenocarcinoma formation later

If a child is born with Urachus anomalies, where the the urachus remains patent or there are some remnants, we will interfere surgically and excise it. This a potential area for Adenocarcinoma. They may develop recurrent infections which is another reason to remove.

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



Presentation	Diagnosis	Management
Urinary retention. Post void urinary residual Recurrent UTI's and stones due to stasis or they can be asymptomatic	• Bladder diverticula can be detected on a prenatal ultrasound. • The **gold standard** is VCUG(voiding cystourethrogram) which will reveal possible accompanying VUR. 	• Asymptomatic: Not Treated • Symptomatic: Especially in conjunction with VUR, should be treated surgically

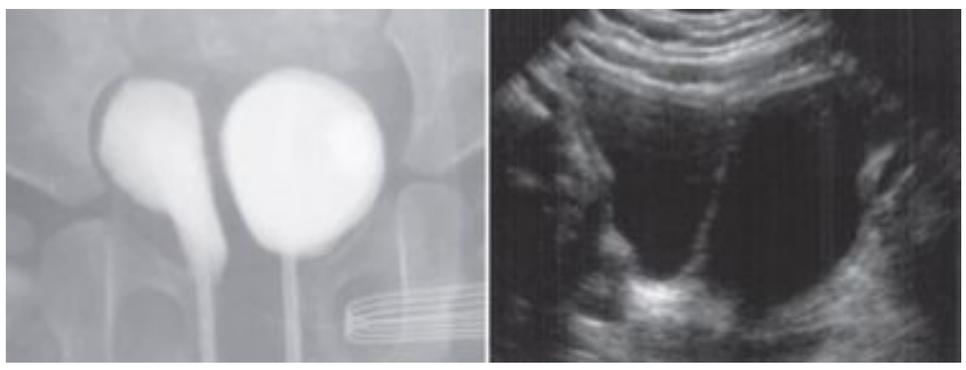
Bladder Anomalies

3. Bladder Duplication

Just know we can have two bladders and two urethras; very rare. This all of what we have to know about it.

- Very rare,
- Bladder duplication is often associated with duplication anomalies of the external genitalia & lower GIT.

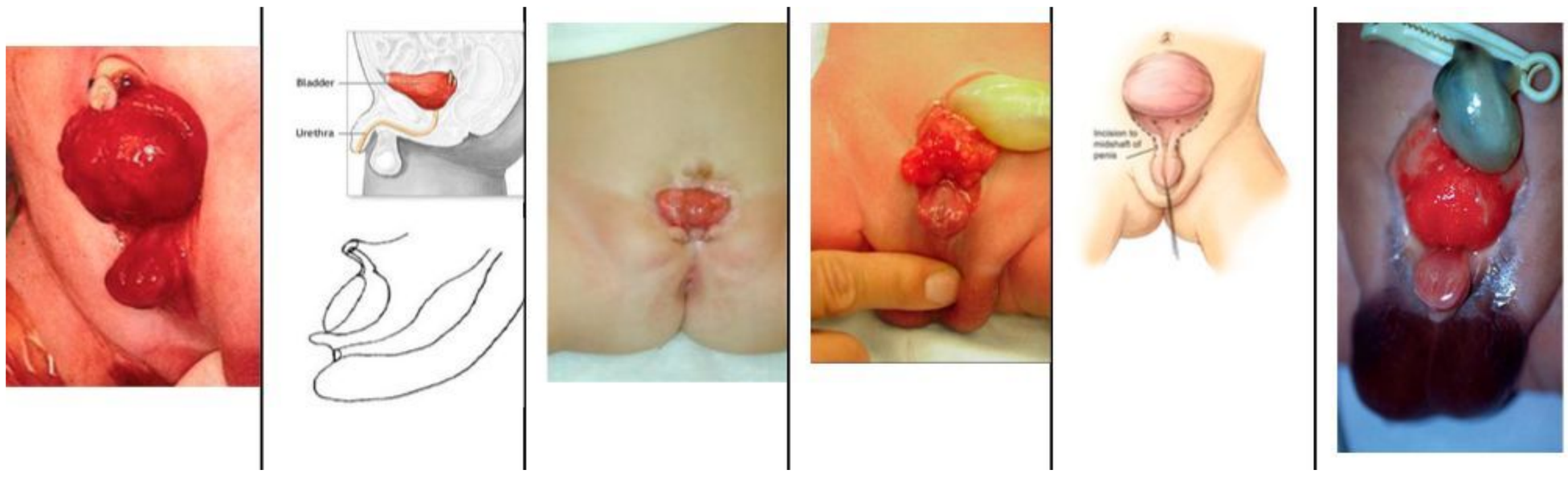
Diagnosis	Management


MCUG to confirm the diagnosis
	• **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 Video (04:30)

Bladder exstrophy is when we have deficient abdominal muscle and the bladder wall will be open to the skin, you can see the trigone of the bladder in the pictures

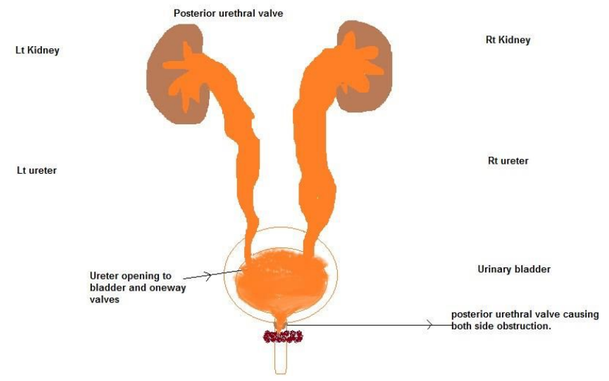
- The incidence of bladder exstrophy has been estimated as between 1 in 10,000 and 1 in 50,000.



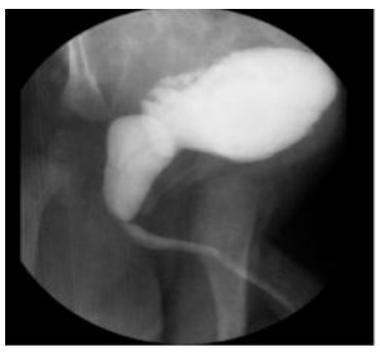
What you see is the inside of the bladder and the ureteric orifices. Males will usually have a short penis. Has to be fixed by surgery after birth. Usually done during the first week of life.

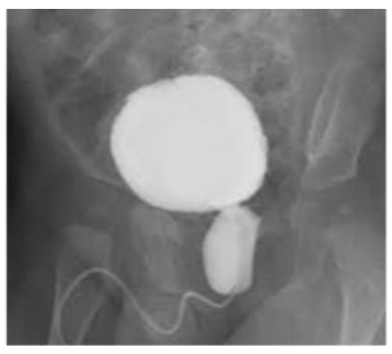
Urethral Anomalies

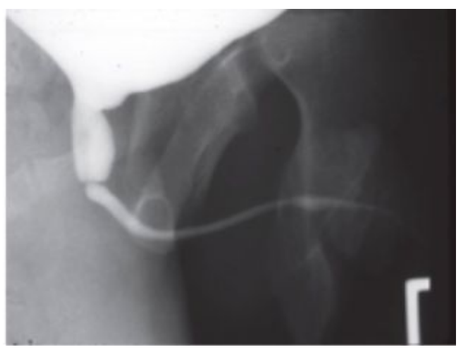
1. Posterior Urethral Valves (PUV)



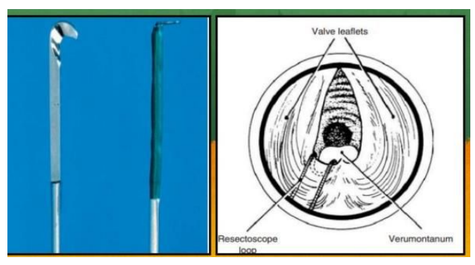
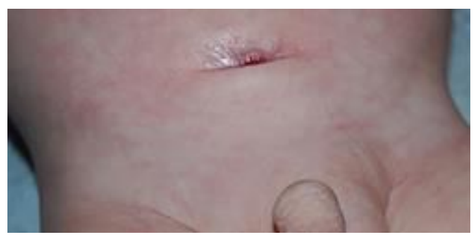
- 1 in 8000 to 25,000 live births
- Makes up 10% of urinary obstruction diagnosed in utero.
- **Most common cause of urine retention in male infants**.
- 50% have renal impairment
- the bladder & the kidneys developed under high pressure & resistance. Which can be severe or mild
- With time the bladder wall muscles will become thick which leads to neurogenic bladder, also will develop reflux
- **Associated findings:**
  - **Oligohydramnios**
  - **Bilateral renal dilatation**
  - **VUR in 40%**
  - Valve bladder
  - Renal impairment(In the end)







Presentation	Diagnosis	Management

• Antenatal
• Urine retention
• UTI
• Poor urinary stream
• Urinary incontinence
• CRF (ESRD)
 	
Causes obstruction and difficulties with urination which leads to severe bilateral hydronephrosis
 	
Usually a baby boy with severe bilateral hydronephrosis and Oligohydramnios will have posterior Urethral reflux until proven otherwise. Those kids will also have pulmonary hypoplasia.
	• US suspected Findings:
	  ○ Dilated posterior urethra
	  ○ Thick wall bladder
	  ○ Dilated kidney and ureters.
	• MCUG
	
	An ultrasound is done first to confirm the presence of hydronephrosis then we do MCUG to look for PUV
		• Initial treatment:
		  ○ Feeding tube insertion
		  ○ Ultrasound
		  ○ Start antibiotic prophylactic
		  ○ MCUG.
		• Surgical treatment:
		  ○ Endoscopic valve ablation
		  ○ Cutaneous vesicostomy
		
		 
		
		 

whenever the baby is stable we will go by a cystoscope and incise the valve and urethra to make voiding easier. Vesicostomy can be done when the baby is too small or premature. We bring the bladder into the anterior abdominal wall as a temporary diversion.

Other urethral anomalies include:

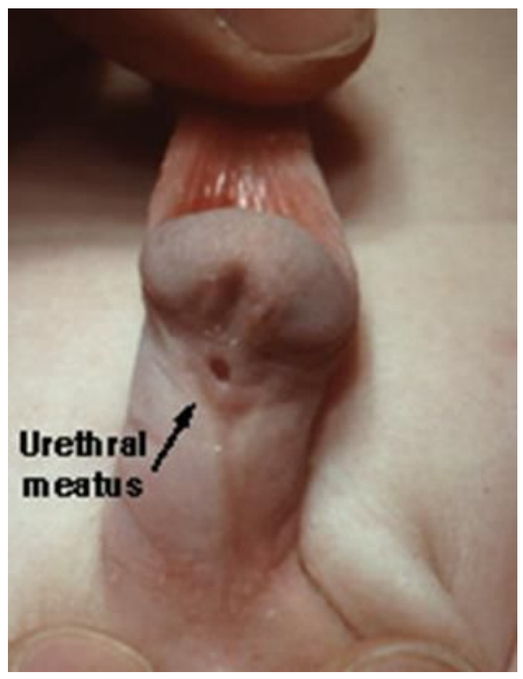
2. Anterior Urethral Valve **3. Urethral Duplication**

4. Congenital urethral stricture **5. Urethral polyps**

III. Congenital Anomalies of the External Genitalia

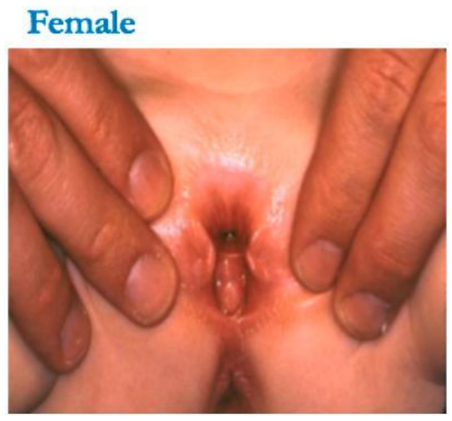
1. Hypospadias

- Abnormal position of the external urethral meatus (EUM) on the **ventral** surface.
- Types: Distal hypospadias, Proximal hypospadias
- **NO Circumcision absolute contraindication why?because we need every part of the skin in the surgery**
- 6 to 9 months repair.(because of the risk of anesthesia).
- Its triad of:
- 1-:incomplete foreskin, the ventral part is missing.
- 2- the opening is lower than where it should be.
- 3-In some cases there will be penile curvature.
- We do surgery to fix it for cosmetic purposes



2. Epispadias

- Abnormal position of the external urethral meatus on **the dorsum surface.**

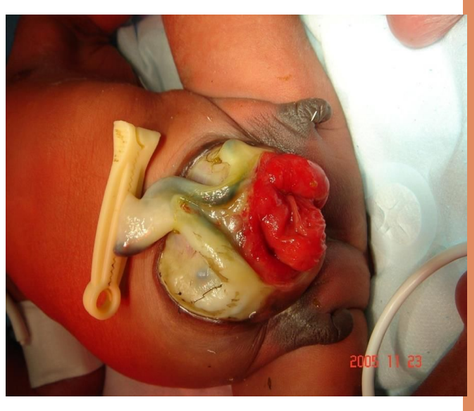


The opening could be lower just like Hypospadias, could be midshaft or penupubic. In the latter the sphincter mechanism is gone and they could have incontinence if you don't fix it surgically.

3. Micropenis

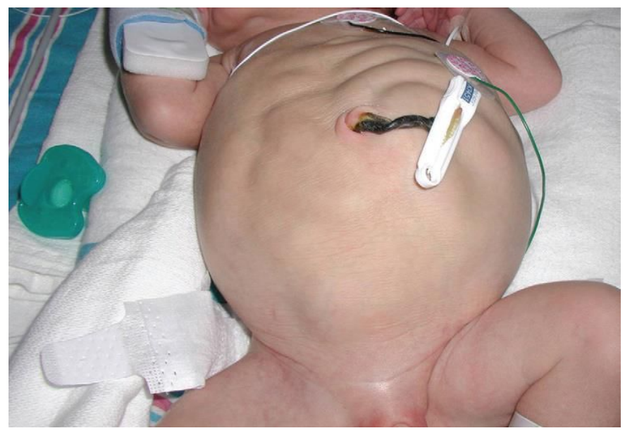
4. Cloacal Exstrophy

More severe form of bladder exstrophy
+here you can see the bowel not the Bladder only unlike bladder exstrophy.
-They have poor quality of life, usually terminate it during pregnancy



IV. Other Congenital Anomalies

1. Prune-Belly Syndrome They could have cardiac or limb anomalies.

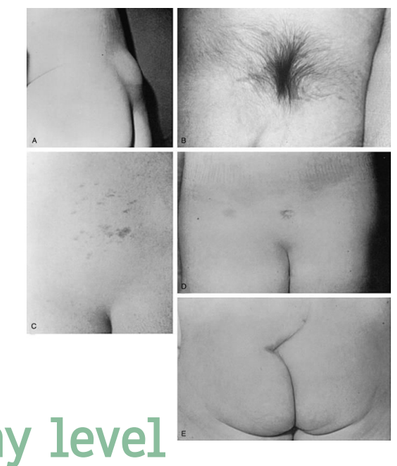


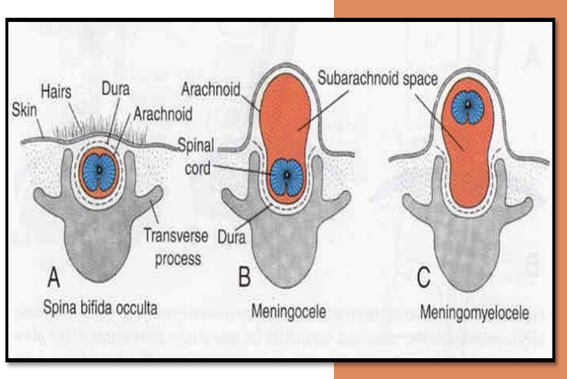
- The incidence: 1:29,000 to 1 in 40,000 live birth.
- **3 major findings:**
  - Deficiency of the abdominal musculature
  - Bilateral intra- abdominal testes **should be corrected surgically.**
  - Anomalous urinary tract bilateral hydronephrosis
- Other names::
  - **Triad Syndrome**
  - Eagle-Barrett Syndrome
  - Abdominal Musculation Syndrome
  - Problems with these kids are: they don't have enough muscles thus they can't generate enough pressure to defecate nor to urinate. They may have recurrent infections and pneumonia because they can't cough the secretions.. They can develop recurrent UTI's because they will have residual urine in their bladder.

IV. Other Congenital Anomalies

2. Neuro Spinal Dysraphisms AKA spina bifida

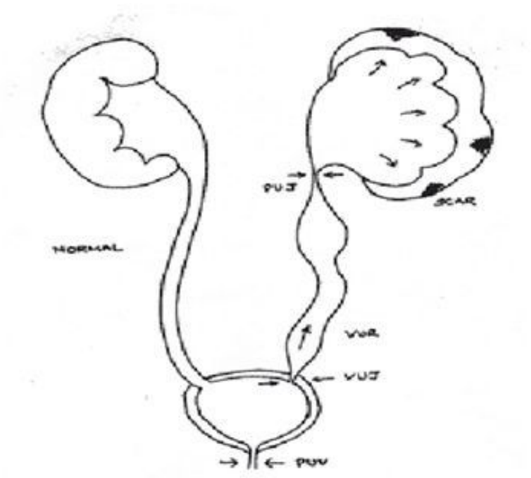
- **The most common cause of neurogenic bladder dysfunction** in children is abnormal development of the spinal canal and internecine spinal cord.
- Cutaneous lesions occur in 90% of children with various occult dysraphic states.
- **These lesions vary from:** that's why they check babies back in the clinic
  - Small lipomeningocele
  - Hair patch
  - Dermal vascular malformation
  - Sacral dimple
  - Abnormal gluteal cleft
  - It happens as as result of defect in transverse process
  - The commonest site is usually L5-S1. Can happen at any level
  - Spina bifida commonest cause of paraplegia in kids.50% of the kids that are wheelchair dependent have Spina bifida.





3. Antenatal Hydronephrosis (Hydronephrosis that developed in pregnancy)

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



Surgical 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)

Summary

Upper Urinary Tract Disorders

Kidney Anomalies

Anomalies of number	Anomalies of ascent	Anomaly of fusion
1. **URA** ○ More common in male ○ Usually left side URA ○ DMSA (best confirmatory test) 2. **BRA** ○ **P**ulmonary Hypoplasia ○ **P**otter's syndrome ○ Best confirmatory test > DMSA ○ Bladder is usually hypoplastic	1. **SRE** ○ Pelvic kidney is the most common ○ Most are asymptomatic ○ Hydronephrosis ○ VUR is common	1. **Horseshoe kidney** ○ Most are asymptomatic ○ UPJ is common ○ Variable blood supply

Ureteral Abnormalities

	Presentation	Diagnosis	Management
UPJO	-	• U/S: Hydronephrosis w/o hydroureter • Confirm Dx: Dynamic renogram=Diuretic renal scan	Dismembered Pyeloplasty
VUR	Febrile UTI	• MCUGIVP	• Prophylactic antibiotic • Cystoscopy • Ureteral reimplantation
Ureterocele	Urinary retention > UTI + Stones	• MCUGIVP	

Summary

Lower Urinary Tract Disorders

Bladder		Urethral
Urachal	Diverticulum	PUV
1. **Patent urachus:** ○ Asymptomatic: conservative Rx ○ Infected: Drain + Abx then excision ○ Nonresolved: Excision bc risk of adenocarcinoma	1. **Primary:** ○ congenital ○ Herniation of bladder wall 2. **Secondary:** ○ Acquired: ○ due to infravesical obstruction **I**f symptomatic, especially w/ VUR: surgery	○ Most common cause of urinary retention in male infants ○ Bilateral hydronephrosis ○ Renal impairment (CRF) ○ Dx: MCUG

External Genitalia Disorders

Hypospadias	Prune-Belly Syndrome	Neurospinal Dysraphisms
○ EUM on the ventral surface ○ Distal hypospadias is more common ○ NO circumcision	○ Three Major findings: ○ Deficiency of abdominal musccles ○ Bilateral intraabdominal testes ○ Abnormal urinary tract	○ Most common cause of neurogenic bladder in children ○ Cutaneous lesions: ○ Hair patch ○ Sacral dimple ○ Abnormal gluteal cleft ○ Lipomeningocele

Quiz

1. Which one of the following is the most common cause of urinary retention in male newborns?
   a. Posterior Urethral Valve
   b. Bladder Duplication
   c. Vesicoureteral Reflux
   d. Anterior Urethral Valve
2. In which of the following is circumcision an absolute contraindication?
   a. Hypospadias
   b. Epispadias
   c. Triad Syndrome
   d. Spina Bifida
3. Which one of the following diseases require management with prophylactic antibiotics?
   a. Horseshoe kidney
   b. Ureteropelvic junction obstruction
   c. Vesicoureteral reflux
   d. Crossed renal ectopia
4. An infant presents to you with urinary discharge from the umbilicus, what is the most likely diagnosis?
   a. Ureterocele
   b. Duplication anomalies
   c. Micropenis
   d. Patent urachus
5. What is the best confirmatory test for unilateral renal agenesis?
   a. Ultrasound
   b. CT Abdomen
   c. DMSA
   d. Micturating cystourethrogram
6. What is the best test for diagnosing vesicouretral reflux?
   a. Ultrasound
   b. CT abdomen
   c. DMSA
   d. Micturating cystourethrogram
7. Potter's syndrome is characterstic of:
   a. Bilateral renal agenesis
   b. Unilateral renal ageneisi
   c. Supernumerary kidney
   d. Simple renal ectopia
8. All of the following can show dilated ureter on ultrasound except:
   a. Uretrovesical obtruction
   b. Vesicoureteral reflux
   c. Ureterocele
   d. Ureteropelvic obstruction

Answers:

1. A
2. A
3. C
4. D
5. C
6. D
7. A
8. D