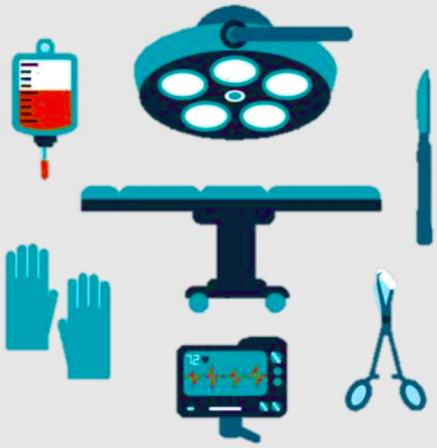

Pediatric Urinary Disorders

Objectives:

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

Resources:

- 436 doctors slides.
- 435's teamwork.
- Surgical Recall.

Done by: Ruba Barnawi, Sarah Alshamrani, Nawaf Aldarwish
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Revised by: Maha AlGhamdi

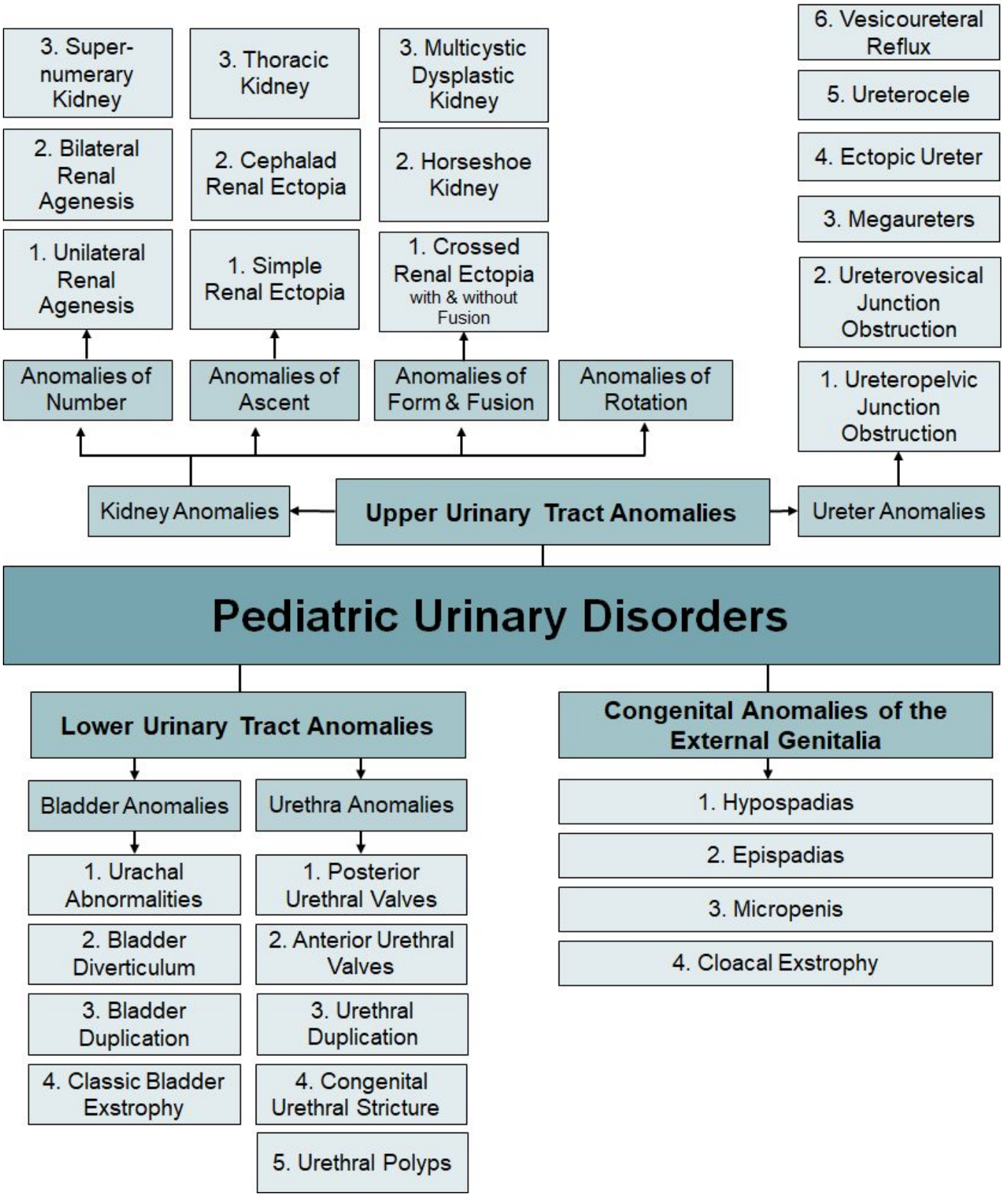
Color index:

Notes , Important , Extra , Davidson's

Editing file

Feedback

Outline



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.

I. Upper Urinary Tract Anomalies (Kidney + Ureter)

Kidney Anomalies

For simplicity we divide them into: number, ascent, form & fusion, and rotation. (they may occur together)

(Most of the congenital kidney anomalies present asymptomatic unless associated with other anomalies causing obstructive uropathy or reflux)

A. Anomalies of Number:

Renal Agenesis[1] Video(04:52) highly recommended!

Unilateral Renal Agenesis (URA)

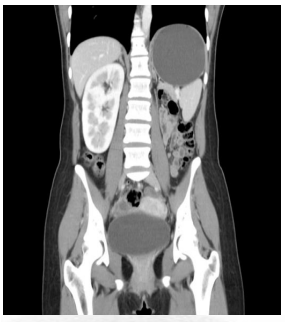
- 1 in 1100 births.
- **Male** : Female of 1.8 : 1
- The **left** side is absent more frequently.
- The ipsilateral **ureter** is completely absent in 50%.
- Anomalies of other organ systems are found frequently in affected individuals CVS,GIT,MSC
- Müllerian duct abnormalities: 25% to 50% of females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

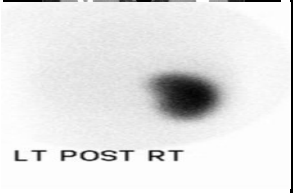
-Normally, renal blood flow distributed 50% to each kidney.In this case, the whole blood flow will go to that one kidney (either left or right) => Increase work of this kidney => hypertrophy (but not clinically important).

- **Asymptomatic**, diagnosed incidentally

Presentation:

- Prenatal US
- Incidentally:
  - Abdominal US (will say kidney was not visualized, to be documented by other study.) Which study? **Nuclear study.**
  - Abdominal CT (not sensitive) kidney may not be visualized in CT either bc it was obscured by the bowel gases, difficult on the examiner or different position "ectopic kidney".





Diagnosis:

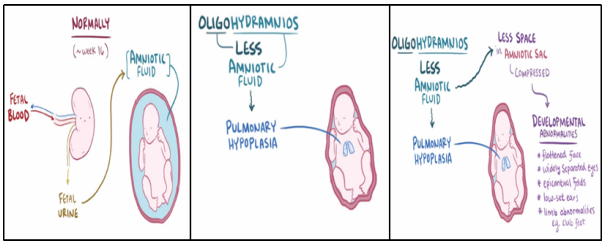
- Nuclear study **(DMSA[2]):** confirmed

Management:

- It is not clinically significant, we do nothing about it: we inform the patient and do regular routine urinalysis (bc they may develop proteinuria in the future) + BP measurements.

Bilateral Renal Agenesis

- More serious.
- No kidney = no amniotic fluid! (which protects child from contractions of uterus)

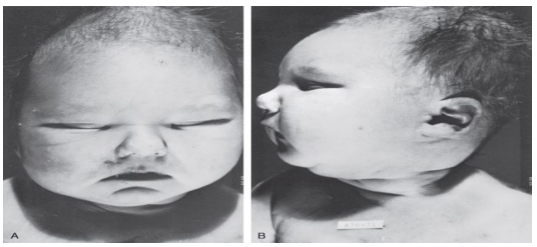


Prognosis:

- 40% are stillborn (die at birth).
- If they didn't die at birth they do not survive beyond 48 hours due to respiratory distress associated with **pulmonary hypoplasia** (because no fluid is reabsorbed) + renal failure.

Characteristics:

- Potter's syndrome.
- Oligohydramnios (deficiency of amniotic fluid)
- Ureters are almost always absent.
- Bladder is either absent or hypoplastic. (bc no kidney => no urine => no distention)
- Adrenal glands are usually positioned normally (if the kidney is absent it doesn't mean the adrenal will be absent, since different embryological origin).
- Müllerian duct anomalies are commonly observed.

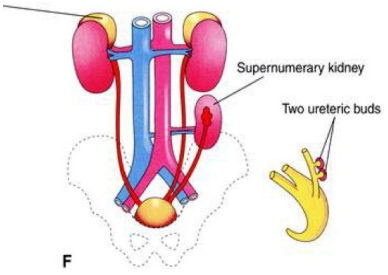


[1]Renal agenesis (RA) is the complete absence of development of one or both kidneys.

[2] It's a radionuclide scan that uses dimercaptosuccinic acid.

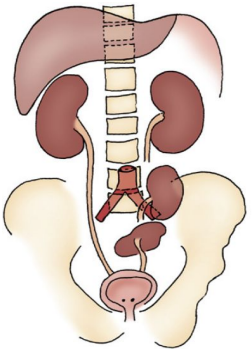
Supernumerary Kidney very rare

- Definitive accessory organ (one or two or three extra kidneys) with its own collecting system (own ureter draining into bladder) , blood supply, and distinct encapsulated parenchyma.
- Both the accessory & original kidneys are working
- 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[3]

- 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 and UVJO (will be discussed later)
  - Reflux (VUR): grade III or greater
  - Malrotation
- The incidence of genital anomalies in the patient with ectopia is about 15%.
- Most ectopic kidneys are clinically **asymptomatic** unless it was associated with reflux or obstruction).

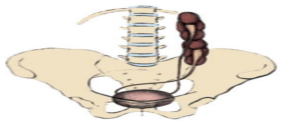
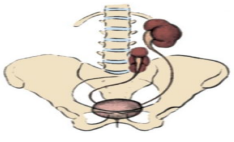
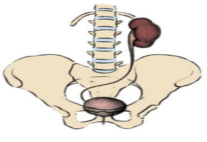
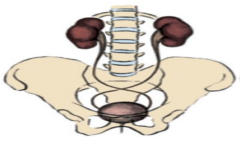
2. Cephalad Renal Ectopia

3. Thoracic Kidney

C. Anomalies of Form and Fusion:

1. Crossed Renal Ectopia (with and without Fusion): Anomaly on top of an anomaly.

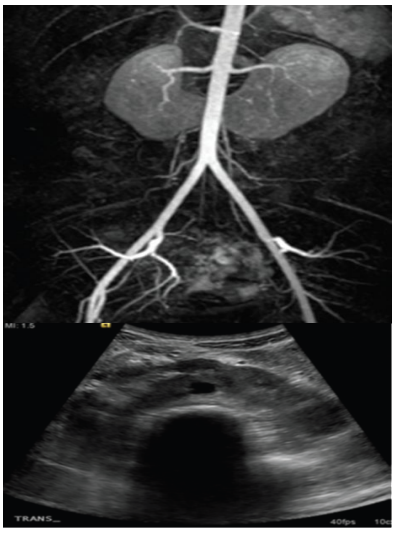
- Crossed ectopia: kidney is located on the side opposite from that in which its ureter inserts into the bladder.
- The ureter from each kidney is usually orthotopic (meaning that if the kidney crossed to the right side, the ureter will still be in the left side). (How can we differentiate if this kidney is crossed or not? If the ureter crossed the midline or not)
- 90% are fused with their mate. (both kidneys are attached together)
- 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	Crossed renal ectopia without fusion	Solitary crossed renal ectopia	Bilaterally crossed renal ectopia
			

[3] 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.

2. Horseshoe Kidney [Video(03:28)]

- Occurs 1 in 400 persons.



Morphology:

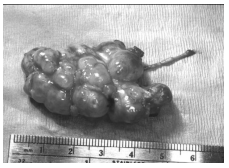
- 90% of the kidneys is attached at the lower border, and 10% in the upper border.
- The isthmus is bulky and consists of parenchymatous tissue .
- The calyces:
  - normal in number
  - atypical in orientation.
- pelvis remains in the vertical or obliquely lateral plane

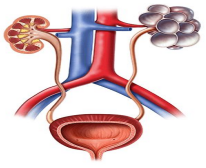
Presentation:

- Horseshoe kidney is frequently found in association with other congenital anomalies.
- UPJ obstruction in one third.
- 60 % **asymptomatic**. (diagnosed incidentally)
- The blood supply can be quite variable (for surgical manner)
- They are totally separate kidneys only connected by isthmus[4].
- We don't separate them!

3. Multicystic Dysplastic Kidney (MCDK)

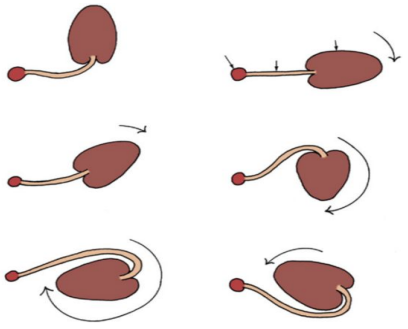
- Renal cortex is replaced by numerous cysts of multiple sizes.





D. Anomalies of Rotation (Malrotation):

- 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 ventromedially during ascent:
  - the calyces point **laterally**, and the pelvis faces **medially**.
- When this alignment is not exact, the condition is known as malrotation.
- Frequently associated with **Turner syndrome**.



Ureteral Anomalies

(Very important anomaly that you should know)

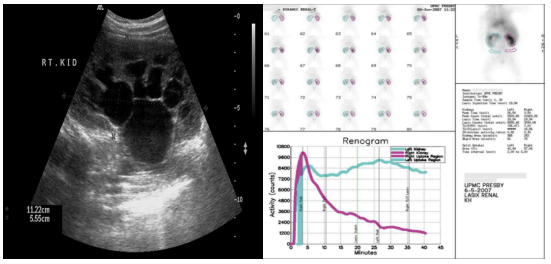
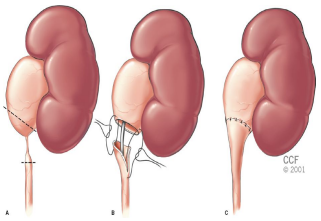
We have **3 anatomical narrowing points** of the ureters: **1.** ureteropelvic junction (UPJ), **2.** the point where the ureter crosses anterior to the iliac vessels and **3.** the ureterovesical junction (UVJ). (click here to see a picture)

1. UreteroPelvic Junction (UPJ) Obstruction (or stenosis → narrowing not complete obstruction)

(Why is it important? Bc usually it is the most common congenital anomaly **detected incidentally during pregnancy with US**, when detected, we need to immediately **correct it** before it affect the function of the kidney.)

[4] Isthmus is where the two kidneys are fused, 90 % renal tissue 10 % fibrous tissue

Presentation	Diagnosis	Management

• Usually detected:
  ○ Prenatal US
  ○ Incidental in Neonates/Children
• If missed pt will become symptomatic:
  ○ UTI
  ○ Pain
  ○ Palpable mass
  ○ Hematuria
  ○ Stone
	• Ultrasound: Shows isolated **hydronephrosis** (meaning only the pelvis is dilated and the ureters are normal **no** hydroureter).
	• **Dynamic renogram***: Shows obstruction
	 
	
	(fluid appears black → Looks like mickey mouse)
		• **Dismembered Pyeloplasty**
		(we cut the area of narrowing and reattach the ureter, there are many types of pyeloplasty; you don't have to know how the procedure is done but you need to know the **name**)
		 

*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:

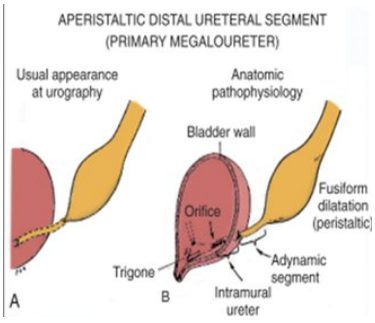
- **Static renogram**: If you want to see if there is renal tissue or function issue (Is this kidney functioning or not?) like in pt. with unilateral renal agenesis.
- **Dynamic renogram**: If you want to see if there is renal obstruction. (ex: GFR)

- 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!

2. UreteroVesical Junction (UVJ) Obstruction "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 hydro**ureter**onephrosis.
- 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

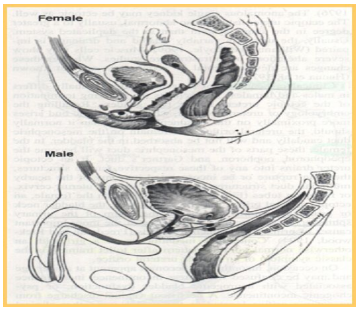
- An ectopic ureter is any ureter, single or duplex, that doesn't enter the **trigonal** area of the bladder .

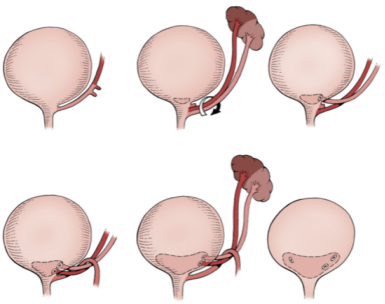
Presentation:

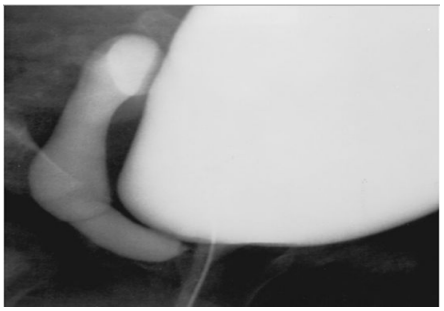
- In **females** the ectopic ureter may enter anywhere from the bladder neck to the perineum and into the vagina, uterus,and even rectum.
- One of the classic symptoms is **continuous wetting\dribbling** (Incontinence). (Since the ureter opens in an area (like the **vagina**) where there is no sphincter 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 (area proximal to sphincter).
- In **males** it will be presented with **recurrent** epididymitis, **epididymo-orchitis**. No wetting\dribbling because epididymis has sphincter unlike the vagina.

Investigation: MCUG

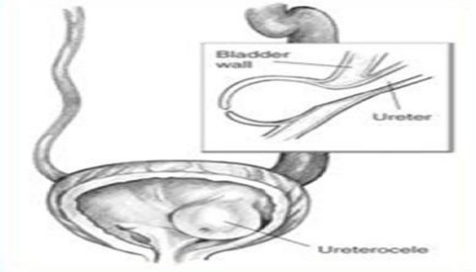
Treatment: reimplantation (**if** kidney function is normal!) if not functioning → nephrectomy.



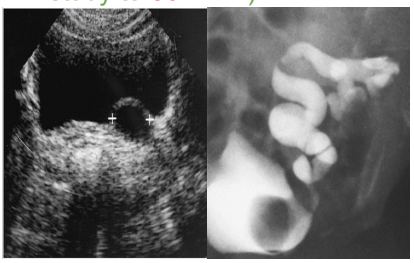




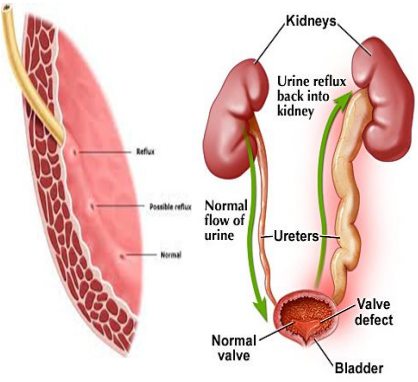
5. Ureterocele



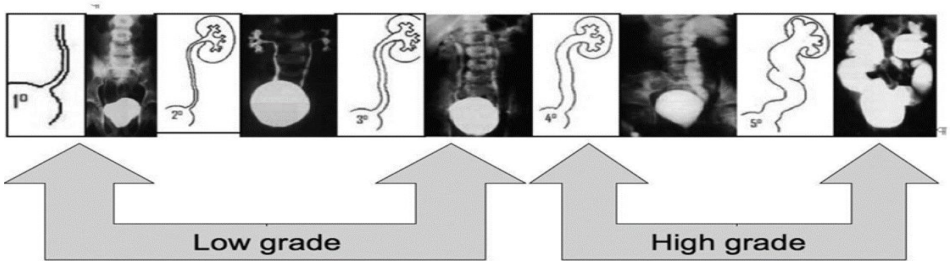
- A cystic dilation (Fluids) of the distal aspect of the ureter. (a sac or cyst at the end of the ureter that opens inside the bladder)
- Located either within the bladder or spanning the bladder neck and urethra.
- **Types**:
  A. Intravesical → Orthotopic , Simple , Adult type
  B. Extravesical → Ectopic , Duplex system , Infant type

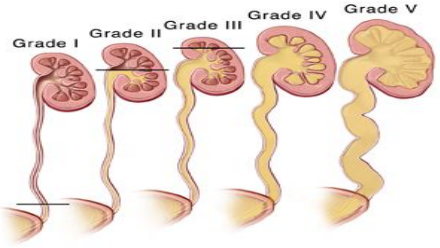
Presentation	Diagnosis	Management
• Antenatal detected during pregnancy (U/S) • Urine retention • Infection • Calculus formation Always keep this in mind for anomalies: Either detected during pregnancy (antenatal) or if missed: Found incidentally or is symptomatic (most common symptoms obstructive uropathy: pain, infection or stone formation)	• Ultrasound • MCUGIVP (voiding cystourethrogram; contrast study to **confirm**) 	• Initially (immediate) management: Incision/**puncture** the ureterocele to release obstruction. • Then later on treat accordingly (**definitive** treatment): ○ if there is obstruction → excise the area and reimplant ○ If there is reflux → reimplantation

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

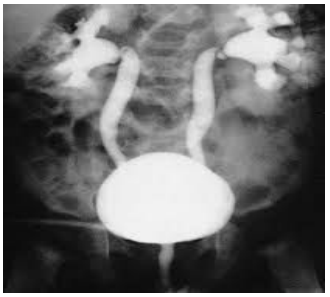
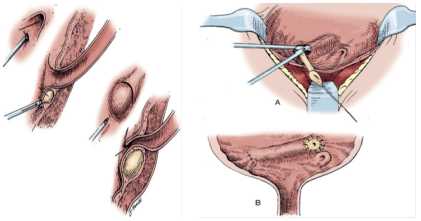


- Normal anti-reflux mechanism: "Flap valve "
  1. Oblique course as it enters the bladder.
  2. Proper muscular attachments to provide fixation.
  3. Posterior support to enable its occlusion.
  4. Adequate submucosal length. If shorter the chance will be higher
- In the **primary** reflux, the entrance of the ureter to the bladder is abnormal (either course is short or not entering obliquely), but in the **secondary** reflux there's anomaly in the bladder (ex: neurogenic bladder).
- It's important to differentiate between them because of treatment:
  - In primary they are born with it, while in secondary if we treat the underlying cause it will go away.
- **Staging:** we stage it for treatment: low grade 1,2,3 no need to treat surgically unlike high grade 4,5 needs surgery (you don't have to know it)





Presentation	Diagnosis	Management

• Asymptomatic
  ○ Prenatal
  ○ Fluctuated dilatation
Antenatally they present with special finding: **alternating hydronephrosis***
 	
• **Febrile UTIs** (febrile = with fever)
• Could lead to pyelonephritis
	• **MCUG**** (micturating cystourethrogram) **modality of choice**.
	 
	On US it will show hydronephrosis (not specific) so we do MCUG to confirm
		• **Prophylactic antibiotic^**
		• Surgical treatment^^.
		  ○ Endoscopic (cystoscopy) treatment (80% success)
		  ○ Ureteral reimplantation (98% success).
		 

All this is extra explanation by the doctor for your understanding. Skip it if you want.

***** It means if I do the US is at 25 weeks there will be hydronephrosis then when repeated at 30 weeks there is no hydronephrosis then there will be hydronephrosis. **Why does this happen?** It depends on what time the US is taken during voiding. If bladder is empty (fetus already voided) there is no hydronephrosis, but if **during** voiding there will be hydronephrosis. So protocol now is to do US during pregnancy then repeat after 15 mins (to rule out false -ve / +ve)
If missed antenatally, they will present symptomatically with infection, pyelonephritis, **rarely** may cause renal failure.

******MCUG/VCUG[5] : It is an X-ray, we put contrast in the bladder => If there is no reflux the contrast will stay in the bladder. If it is going to upper tract this mean that this pt. has reflux.

^What is the difference between prophylactic antibiotics vs. therapeutic antibiotics?

- Prophylactic antibiotics is broad spectrum, while therapeutic is targeted to a specific organism.
- The dose is different: in prophylactic we give one third (⅓) of the therapeutic dose.
- Different frequency, ex: we give a pt. a therapeutic antibiotic 3 or 2 times/day, while prophylactic antibiotic will usually be given once/day.

For all the patient with reflux we will start with the prophylactic antibiotics (especially high grade). **Why**? Bc low grade reflux will go by itself, we call it **spontaneous resolution** of reflux. For example, grade in grade one 80%-90% will go by itself without anything. As the grade increase, the spontaneous resolution of reflux will decrease. Once you reach to grade 5 spontaneous resolution of reflux will be 3%. That's mean as we go high in the grade we need more surgical intervention.

^^What type of surgery intervention? Our aim is to prevent the urine in the bladder from going up to the kidney.

- Either we do it **endoscopic** by cystoscopy where we inject material to narrow the opening to allow urine to come down but not go up. It is a one-day surgery (patients leave the same day) but it has a success rate of 80%.
- Or we can do reimplantation (open surgery), pt will stay at least 3 days in the hospital, there is comorbidities from the incision and he will have hematuria after the surgery (but it has high success rate = 98%).
- We discuss all of this with the family, and let them decide.

What is the difference between the reimplantation here and the reimplantation in the ureterovesical junction (UVJ) obstruction?
In UVJ obstruction we **excise the abnormal area** then we implant ureter, but here in the reflux we **just reimplant** ureter bc there is no abnormal segment to be excise.

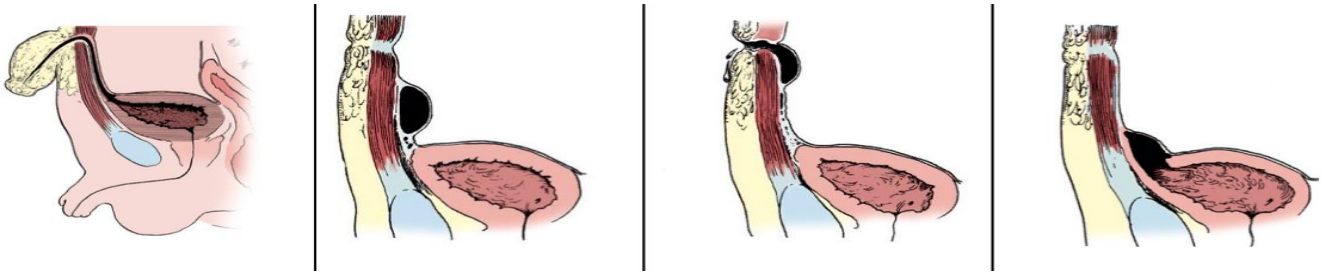
[5] VCUG: Voiding CystoUrethroGram

II. Lower Urinary Tract Anomalies (Bladder + Urethra)

Bladder Anomalies

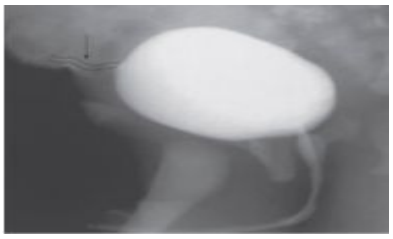
1. Urachal Abnormalities

- Embryologically the bladder is connected to the umbilicus by the urachus, but normally it degrades. If it is patent you will have these anomalies.
- Urachal Anomalies are usually detected postnatally due to **umbilical drainage**.

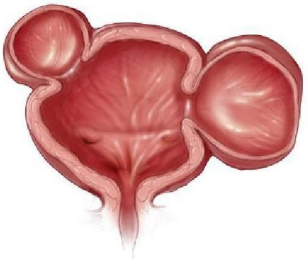


1. Patent Urachus 2. Urachal Cyst 3. Umbilical- Urachal Sinus 4. Vesicourachal Diverticulum

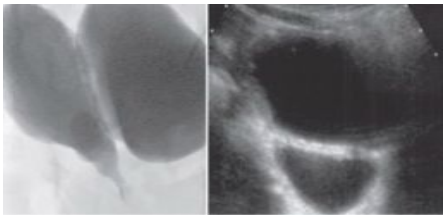
(patent urachus: dripping through the umbilicus → presentation: mother will tell you the umbilicus is usually wet. Also 2 way stream during voiding (periurethra and from umbilicus). What is the importance? This area will be precancerous, need to excise tract later)

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. (may take 1 week or 1 year but usually close spontaneously due to secondary effects of fibrosis) • **Infected urachal remnants**: Initially treated with drainage and antibiotics, followed by **surgical excision** (definitive treatment). • **Nonresolved** urachal remnants should be **excised** due to the **increased risk of adenocarcinoma** formation later

2. Bladder Diverticulum



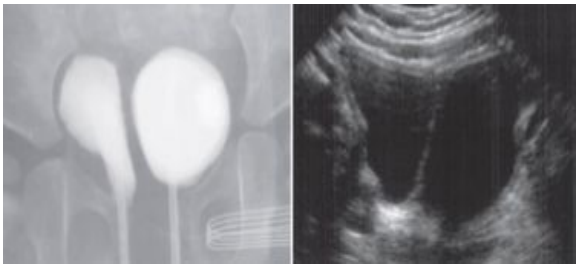
- Weak bladder muscle, not formed well leading to outpouching of mucosa (Out pushing from bladder to form another sac). As pocket fills with urine, it may cause infection or urinary retention.
- **Types:**
  - **Primary Diverticula**: Arises as a localized herniation of bladder mucosa at the ureteral hiatus & are most likely caused by a congenitally deficient bladder wall. Usually there is one **single** diverticula (90%) unless pt has muscular dystrophy.
  - **Secondary Paraureteral Diverticula**: Are acquired and develop due to existing infravesical obstruction. It occurs usually multiple, (usually there is normal musculature but high pressure: pressure → sacculation → diverticulum. Like pts with secondary VUR)

Presentation	Diagnosis	Management
Symptoms: • Symptoms of obstructive uropathy like stone (due to stagnation) • **Retention** (more specific to diverticulum)	• Bladder diverticula can be detected on a prenatal ultrasound. • The **gold standard**, is **VCUG** which will reveal possible accompanying VUR. 	• **Asymptomatic**: Not Treated • **Symptomatic**: Especially in conjunction with VUR, should be treated surgically (excise it: bladder diverticulectomy)

3. Bladder Duplication

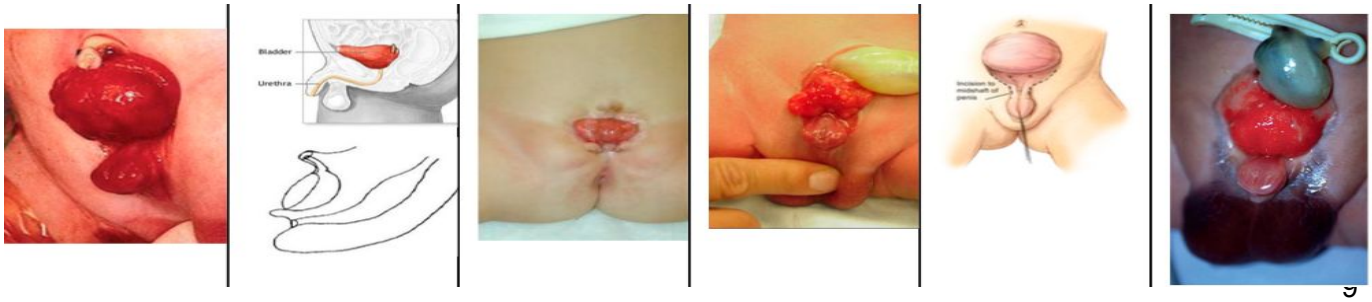
- Two bladders and each ureter opens in one. They either have a common urethra or a separate one (two urethras).
- **Very rare**, seen in conjoined twins.
- Bladder duplication is often associated with duplication anomalies of the external genitalia & lower GIT.

Diagnosis	Management

• **MCUG** to confirm.
 
	• **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)

- The incidence of bladder exstrophy has been estimated as between 1 in 10,000 and 1 in 50,000.
- In males, it's associated with exstrophy-epispadias complex "Epispadias open in the dorsal side".
- 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 (Neonatal **emergency**, we need to correct it immediately after birth) , you will see the posterior wall, ureters & urine come out.
  [A couple of years ago they got a **picture of bladder exstrophy** and were asked to give diagnosis.]

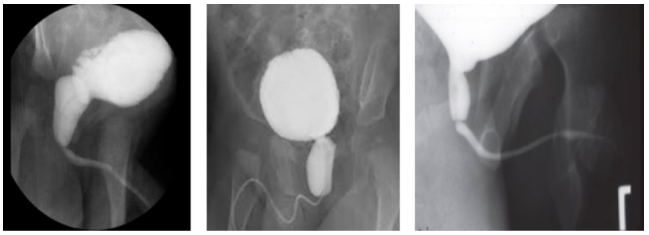


Urethral Anomalies

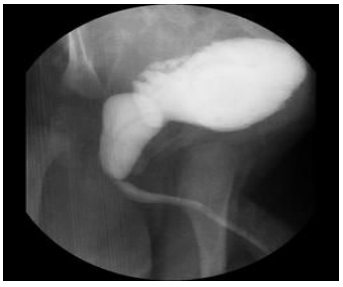
(another important anomaly that you should know)

1. Posterior Urethral Valves (PUV) (Disease of **boys**)

- 1 in 8000 to 25,000 live births (rare but serious).
- Makes up 10% of urinary obstruction diagnosed in utero.
- **Most common** cause of urine retention in male infants.
- 50% have renal impairment and may need transplant.
- The bladder & the kidneys developed under high pressure & resistance.
- **Associated findings:**
  - Oligohydramnios
  - Bilateral renal dilatation
  - VUR in 40% (will have secondary VUR)
  - Valve bladder
  - Renal impairment



Presentation	Diagnosis	Management

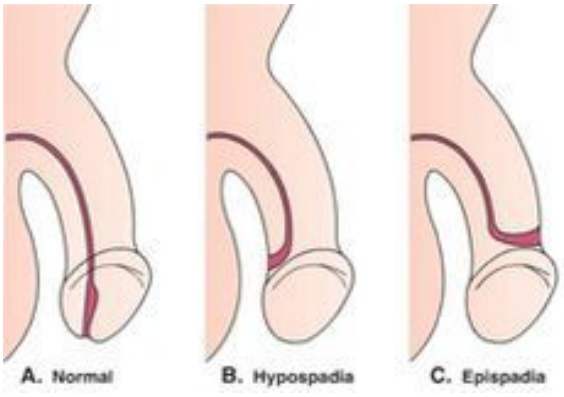
• Antenatal US
• Urine retention
• UTI
• Poor urinary stream (mother will tell you not voiding complete stream only dripping)
• Urinary Incontinence
• CRF (ESRD*)
Very serious disease, we have to interfere quickly or baby may come back with renal failure if missed
 	
*40% will end up with ESRD whatever you do.
	• **US** suspected Findings:
	  ○ Dilated posterior urethra
	  ○ Thick wall bladder
	  ○ Dilated kidney and ureters.
	Bilateral hydroureteronephrosis + thick bladder wall (hypertrophy) = **keyhole sign**. US not enough to diagnose because these findings have other differentials.
	
	• MCUG (Confirmatory) **Radiology of choice** (You will see normal ant. Urethra, filling defect in the post. Urethra and dilated posterior urethra).
	 
		(Neonatal emergency)
		• **Initial treatment:**
		  ○ **Feeding tube insertion**
		  ○ Ultrasound
		  ○ **Start antibiotic prophylactic**
		  ○ MCUG.
		• **Surgical treatment:**
		  ○ Endoscopic valve ablation (We go by cystoscope, and open/relieve the area of obstruction. allowing the obstruction to go)
		  ○ Cutaneous vesicostomy (we do it when the patient is too small. We open the bladder into the skin to drain the urine and bypass the obstruction, then once the patient is 1 year old we close it and do endoscopic ablation)

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

Absolute Contraindication to do Circumcision*

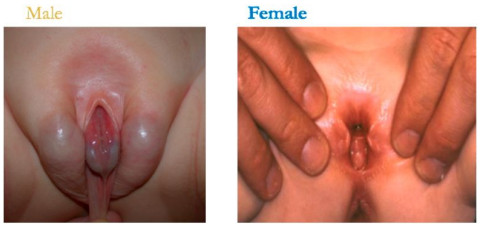


1. Hypospadias

- Only know the meaning + contraindication.
- Abnormal position of the EUM[6] on the **ventral** surface (toward the scrotum)
- NO Circumcision absolute contraindication (because we use the skin for repair "reconstruction in the future")
- Types: distal and proximal.
- 6-9 months repair (because of the risk of anesthesia)

2. Epispadias

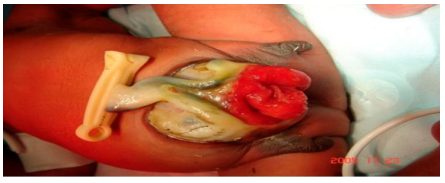
- Only know the meaning.
- Abnormal position of the EUM on the **dorsum** surface.
- If it opens on the bladder neck pt will present with incontinence.
- Commonest type: **penopubic**.



3. Micropenis

4. Cloacal Exstrophy

- Only know the meaning.
- Remember in the bladder exstrophy, the bladder is opened to the outside but the GI is intact and there is an anal opening.
- But here, there is **no anal opening** and all systems are open together (See bowel in the pic)
- One examination to differentiate between cloacal exstrophy and bladder exstrophy: do **rectal exam**

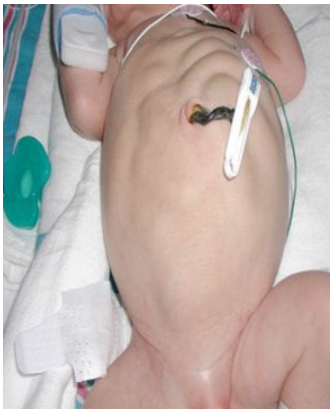


*Any congenital anomaly of male external genitalia it is **absolute contraindicated** to do circumcision because we need the skin to reconstruct, so we have to explain it to the family and refer the patient to a pediatric urologist.

[6] EUM = external urethral meatus

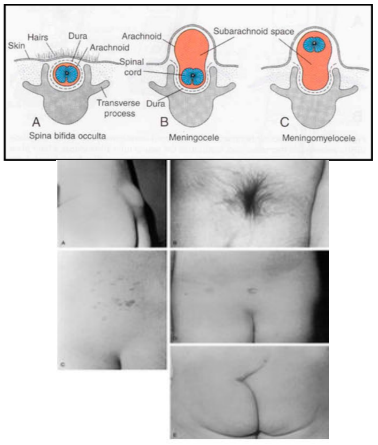
IV. Other Congenital Anomalies

1. Prune-Belly Syndrome



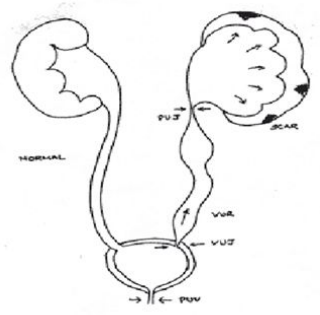
- The incidence: 1:29,000 to 1 in 40,000 live birth.
- 3 major findings: Affects 3 systems.
  - Deficiency of the abdominal musculature (**1. MSK**), on examination you can feel kidneys, liver and you can see the bowel movement clearly.
  - Bilateral intra- abdominal testes (**2. Genital tract**)
  - Anomalous urinary tract **bilateral hydronephrosis** (**3. Urinary tract**)
- Other names:
  - **Triad Syndrome** (bc it affects MSK, Urinary tract and Genital tract)
  - **Eagle-Barrett Syndrome**
  - Abdominal Musculation Syndrome
- Why did we mention it here? Because it is one of the causes of bilateral hydronephrosis.

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. (Don't forget to **examine the pt back**!!)
- Cutaneous lesions occur in 90% of children with various occult dysraphic states.
- These lesions vary from:
  - Small lipomeningocele
  - Hair patch
  - Dermal vascular malformation
  - Sacral dimple
  - Abnormal gluteal cleft
- Why did we mention it here? Any pt evaluated for any voiding dysfunction you have to examine the back because if you find spina bifida we keep in mind pt will have some form of neurogenic bladder.

3. Antenatal Hydronephrosis (Hydronephrosis detected by US during pregnancy)



- Causes: (we covered most of them in the lecture this is like a recap)
  - **Pelviureteric junction obstruction (41%)**
  - Ureterovesical junction obstruction (23%)
  - Vesicoureteric reflux(7%)
  - Duplication anomalies (13%)
  - Posterior urethral valves (10 %)
  - MCDK
  - Others (6%)

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 Anomalies

Kidney Anomalies

Anomalies of Number:	Anomalies of Ascent	Anomalies of Form and Fusion:	Anomalies of Rotation
1- Unilateral Renal Agenesis -CT Abdomen (not sensitive) -**DMSA** (Nuclear study) **2- Bilateral Renal Agenesis** - Do not survive beyond 48 hours due to respiratory distress associated with pulmonary hypoplasia. **3-Supernumerary Kidney** **-asymptomatic** & found incidentally	**1. Simple Renal Ectopia** - 50% have a hydronephrosis due: Obstruction Reflux Malrotation - Most are **asymptomatic** **2. Cephalad Renal Ectopia** **3. Thoracic Kidney**	**1. Crossed Renal Ectopia with and without Fusion:** - Kidney is located on the side opposite from that in which its ureter inserts into the bladder. **2. Horseshoe Kidney** -They are totally separate kidneys only connected by isthmus - 60% **asymptomatic** **3. Multicystic Dysplastic Kidney**	- The most important thing to know is if the kidneys in the normal position or mal-positioned. -Frequently associated with Turner syndrome

Upper Urinary Tract Anomalies

Ureteral Anomalies

Ureteropelvic junction (UPJ) obstruction	Ureterovesical junction (UVJ) obstruction	Ectopic Ureter	Ureterocele	Vesicoureteral Reflux
Presentation: Incidental or if symptomatic: -UTI -Pain -Palpable mass -Hematuria -Stone **Diagnosis:** -Ultrasound **(isolated hydronephrosis)** -Dynamic renogram (obstruction) **Management:** -Dismembered Pyeloplasty	**Diagnosis:** - US: **hydroureteronephrosis**, - Nuclear study: decreased function with obstruction, so this is indication to do surgery. **Management:** - Pyeloplasty (ureteral reimplantation).	-Is any ureter that doesn't enter the trigonal area of the bladder. **Presentation:** -in females classic symptoms is **continuous wetting\dribbling.** - In males it will be presented with recurrent **epididymitis,** epididymo-orchitis. **Diagnosis:** -MCUG	**Presentation:** -Antenatal (U/S) -Urine retention -Infection -Calculus formation **Diagnosis:** -Ultrasound -MCUGIVP **Management:** Initially Incision\puncture the ureterocele.	**Presentation:** **Asymptomatic** -Prenatal -Fluctuated dilatation **Febrile UTIs** **Diagnosis:** Best study for diagnose is **MCUG** **Management:** -Prophylactic antibiotic -Surgical treatment. 1-Endoscopic (cystoscopy) 2-Ureteral reimplantation.

Lower Urinary Tract Anomalies

Bladder Anomalies

Urachal Abnormalities

Diagnosis:
Ultrasound, CT & VCUG (MCUG).
Management:
-Conservative Treatment with observation if asymptomatic.
-Infected urachal remnants need surgical excision to avoid adenocarcinoma

Bladder Diverticulum

Diagnosis:
prenatal ultrasound, The gold standard, is VCUG
Management:
Asymptomatic: Not Treated
Symptomatic: Especially in conjunction with VUR, should be treated surgically

Bladder Duplication

Management:
Initial treatment:
-Directed towards, renal preservation
-Prevention of infections
Long term goals
-Achieving continence and reconstructing the internal and external genitalia

Classic Bladder Exstrophy

-You can find it in neonate delivery, you will see the posterior wall, ureters & urine come **out.**

Management:
Neonatal emergency, we need to correct it immediately after birth

urethral Anomalies

Posterior Urethral Valves

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

diagnosis
US (keyhole sign)
& MCUG

Treatment:

- **Initial treatment:**
  -Feeding tube insertion
  -Ultrasound
  -Start antibiotic prophylactic
  -MCUG
- **Surgical treatment:**
  -Endoscopic valve ablation
  -Cutaneous vesicostomy

Congenital Anomalies of the External Genitalia

Hypospadias

Abnormal position of the EUM on the ventral surface
ABSOLUTE contraindication for circumcision

Epispadias

Abnormal position of the EUM on the dorsum surface

Micropenis

cloacal Exstrophy

Other Congenital Anomalies

Prune-Belly Syndrome

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

Neuro Spinal Dysraphisms:

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

Antenatal Hydronephrosis

Weigert-Meyer Rule Exstrophy

Questions

Q1. Parents came to the emergency with their baby, what is absolute contraindication for circumcision?
A. Hypospadias
B. Posterior urethral valve
C. Undescended testis
D. Vesicoureteric reflux

Q2. A newborn girl evaluated for a unilateral hernia. Postnatal US revealed a normal right kidney, left hydronephrosis, no hydroureter and normal urinary bladder. What is the most likely diagnosis?
A. Vesicoureteric reflux
B. Pelviureteric junction obstruction
C. Ureterovesical junction obstruction
D. Posterior urethral valves

Q3. 1 day-old boy with a history of bilateral antenatal hydronephrosis and hydroureter with thick walled bladder and mild oligohydramnios. What is the initial test to do?
A. Insert tube in the bladder
B. Cystourethroscope
C. Voiding (micturating) cystourethrogram
D. Nuclear study

Q4. 5-year-old female presented with history of recurrent febrile urinary tract infection with incontinence. what is the best study to diagnose the vesicoureteric reflux?
A. Intravenous uretrograme (IVP)
B. Nuclear study
C. Renal and bladder ultrasound
D. Voiding (micturating) cystourethrograme (VCUG)

Q5. Which one of the following is true about hypospadias?
A. circumcision is recommended
B. it's a two-stage surgery
C. repair is best during 6 to 9 months
D. repair is best during 6 to 9 years

Q6. The most common cause of antenatal hydronephrosis?
A. Pelviureteric junction obstruction
B. Vesicoureteric reflux
C. Ureterovesical junction obstruction
D. Duplication anomalies

Q7. An infant presented to you with a discharge from umbilicus. What is the most likely diagnosis?
A. Epispadias
B. Uretrocele.
C. Prune belly syndrome
D. Patent urachus

Answers: 1-A 2-B 3-C 4-D 5-C 6-A 7-D