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Genitourinary Anomalies

Objectives

- Discuss congenital anomalies of kidney
- Discuss congenital anomalies of ureter
- Discuss congenital anomalies of bladder
- Discuss congenital anomalies of urethra
- Recognize congenital anomalies of genitalia

Colour Index

- Main Text
- Males slides
- Females slides
- Doctor notes
- Textbook
- Important
- Golden notes
- Extra

Editing File

Genitourinary Anomalies

- Ureteropelvic junction obstruction (UPJO)
- Multicystic dysplastic kidney (MCDK)
- Vesicoureteral Reflux (VUR)
- Posterior Urethral Valve (PUV)
- Ureterovesical Junction Obstruction (UVJO)
- Hypospadias

U Must Void Properly
Unless you Have an anomaly

More common

Less common

- Unilateral Renal Agenesis
- Ureterocele
- Duplication Anomalies
- Ectopic ureter
- Horseshoe Kidney
- Crossed Renal Ectopia
- Bladder Diverticulum
- Prune -Belly Syndrome
- Epispadias
- Bladder Exstrophy

Uncommon

- Bilateral Renal Agenesis
- Supernumerary Kidney
- Anomalies of rotation
- Bladder duplication
- Cloacal exstrophy
- Urachal abnormalities
- Neurospinal dysraphism

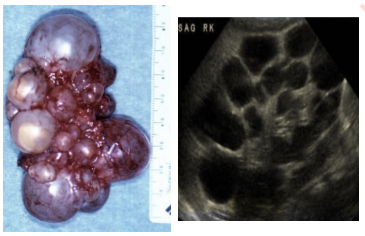
Congenital Anomalies of Kidney

Kidney anomalies

- **Number**
  - Supernumerary — More
  - agenesis — Less
  - Horseshoe — Joined
- **Location**
  - Rotation
  - Ectopia

Multicystic dysplastic kidney (MCDK) 2nd most common

- There is a difference between MCDK and polycystic kidney disease
- Unilateral
- There is no renal tissue, only disturbed cysts & there is no communication between them
- Ultrasound: Very thin and abnormal renal parenchyma, surrounded by multiple cysts of various sizes that do not connect, nor they connect to the renal pelvis. Urine differ from UPJO case by bring in-organized in distribution (No Mickey mouse sign unlike in UPJO)



Diagnosis

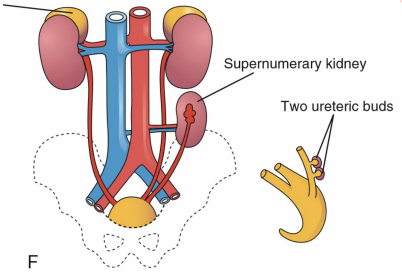
- Prenatal US
- Incidental in Neonates/Children
- Symptomatic same other obstructive uropathy
  - Mass
  - UTI
  - Pain
- DMSA: nuclear study to look for the function of kidneys
  - Little or no uptake of radionuclide
  - The difference between UPJO and MCDK in US that in MCDK there is NO or very little function (diagnostic).
- MCUG/ VCUG (X-ray + contrast)
  - To detect if there is reflux or not, but now days not used routinely unless the patient symptomatic
  - Contralateral VUR
  - 18%-43%

Management

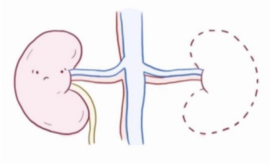
- Observation + follow up (asymptomatic)
  - Cyst fluid disappears
- Surgical intervention (nephrectomy) indication:
  - Hypertension
  - Pain
  - Pyelonephritis

Supernumerary Kidney:

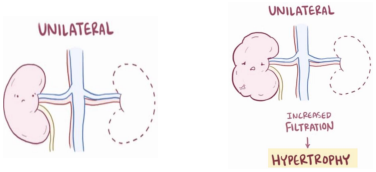
- Definitive accessory organ with its own collecting system, blood supply, and distinct encapsulated parenchyma.
- Either completely separate or loosely attached to the kidney on the ipsilateral side.
- The ureteral inter-relationships on the side of the supernumerary kidney can be variable.
- detected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



Kidney agenesis:

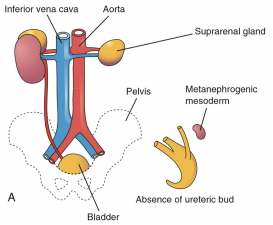


Unilateral Agenesis

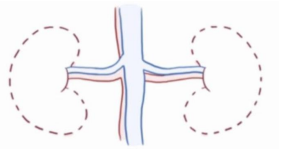


The blood flow normally goes to the each kidney 50%, in case of unilateral renal agenesis the blood flow 100% goes to the only kidney which with time will be exhausted and shows symptoms: Proteinuria, Pain and Hypertension, we must follow them annually with **urinalysis**, **blood pressure** and **ultrasound**

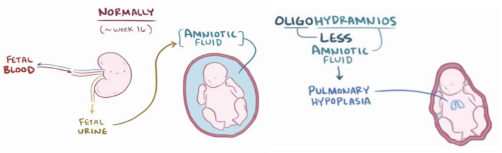
- 1 in 1100 births
- Male: Female of 1.8 : 1
- The left side is absent more frequently
- The ipsilateral ureter is completely absent in 50%



- if the kidney is absent that doesn't mean that the suprarenal gland will be absent too, because both have different embryological origin

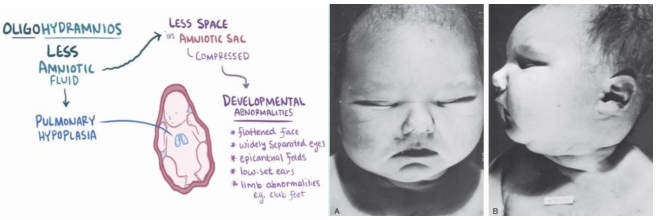


Bilateral Agenesis rare



kidney form the amniotic fluid with urine
No kidney = no amniotic fluid = No space for the fetus to develop = Oligohydramnios leading to Potter's sequence

the most affected organ are the lungs (pulmonary hypoplasia)



This is called Potter face (associated with potter syndrome)

Associated anomalies

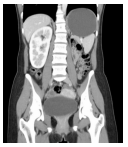
- Anomalies of other organ systems are found frequently in affected individuals: CVS, GIT and MSC
- Müllerian duct abnormalities:
  - 25% to 50% of females
  - 10% to 15% of males
  - Approximately one fourth to one third of women with Mullerian duct anomalies are found to have URA

Associated anomalies

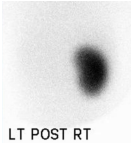
- Ureters are almost always absent.
- Bladder is either absent or hypoplastic.
- Adrenal glands are usually positioned normally.
- Müllerian duct anomalies are commonly observed.

Diagnosis

- Prenatal US
- Incidentally:
  - Abdominal US
  - Abdominal CT
- ★ Confirmed: CT alone is not enough since we might miss the diagnosis
  - Nuclear study (DMSA)



Abdominal CT shows absent left kidney



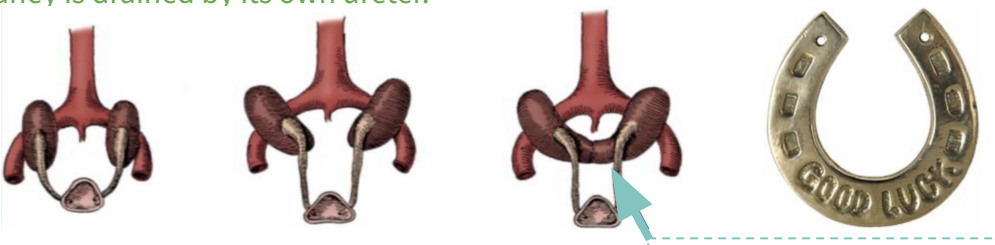
DMSA shows if there's renal tissues or not

Prognosis

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

Horseshoe Kidney:

- Occurs 1 in 400 persons
- Normally the kidneys in the embryological life is inter-pelvic structure facing anteriorly, & with time it will ascend to the normal position.
- sometimes for unknown reason, the 2 kidneys fuses together and connected at lower poles (90%) or upper poles (10%) forming the classical horseshoe kidney.
- Each kidney is drained by its own ureter.

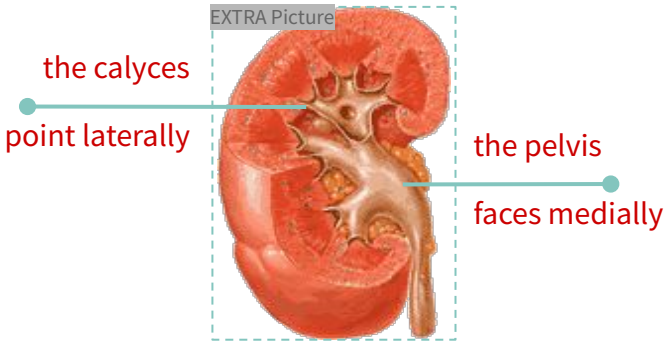


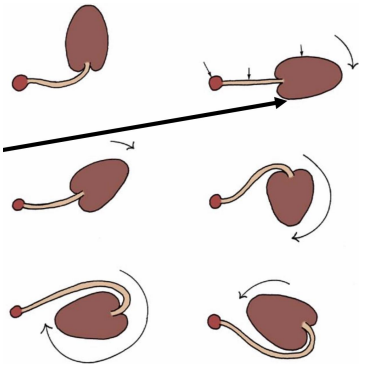
The isthmus is bulky and consists of parenchymatous tissue, it will get stuck under the Inferior mesenteric artery (failure of ascending)

- The calyces by ultrasound or CT:
  - 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** (Ureteropelvic junction) obstruction in one third due to the acute angle of the ureter
- 60 % asymptomatic The rest 40% of Symptomatic patients is due to **hydronephrosis** (Surgery is required)
- There is no surgery to separate the the 2 kidneys, we just treat the symptoms if it occurs (treat the associated pathology not Horseshoe kidney).

Anomalies of rotation:

- The kidney and renal pelvis normally rotate 90 degrees ventromedially during ascent





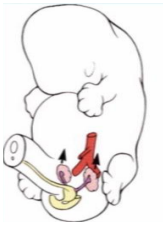
- When this alignment is not exact, the condition is known as malrotation
- Frequently associated with Turner syndrome

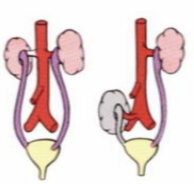
Renal Ectopia:

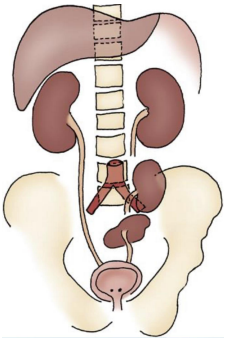
- There is no need to do surgery, we just treat the symptoms if exist

Simple Renal Ectopia

- The kidney arrested instead of ascending to its normal position in the same original side.
- Left more than the right
- 1 of 2100 to 3000 autopsies.
- Most ectopic kidneys are clinically asymptomatic

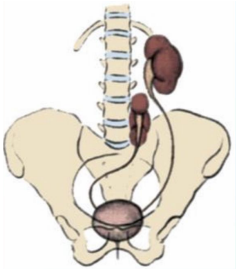




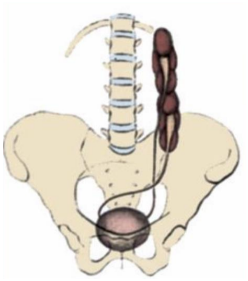


Associated Anomalies

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



Without fusion



With fusion

Crossed Renal Ectopia

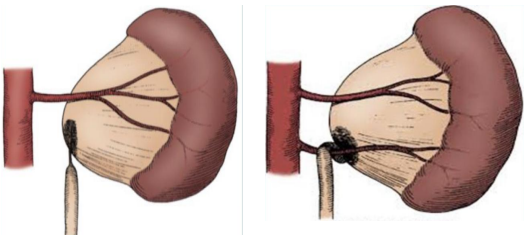
- Crossed ectopia: kidney is located on the side opposite from that in which its ureter inserts into the bladder unlike simple ectopia
- The ureter from each kidney is usually orthotopic
- 90% are fused: the superior pole of the ectopic kidney usually joins with the inferior aspect of the normal kidney.

Congenital Anomalies of The Ureter

Ureteropelvic junction obstruction (UPJO) most common

Presentation

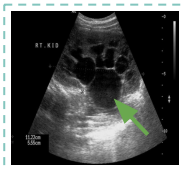
- Prenatal US most cases
- Incidental in Neonates/Children
- By symptoms:
  - UTI: Pyelonephritis
  - Pain: Flank pain
  - Mass
  - Hematuria
  - Stone: Secondary to urine stagnation



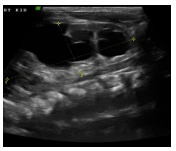
It's not actually obstruction (if complete obstruction kidney loses its function) , Just narrowing of the ureter

Diagnosis

- First investigation[1] is US and shows Mickey Mouse sign:
  - Dilated renal pelvis
  - Dilated major and minor calyces
  - The dilatation and distribution of urine are organized unlike in (MCDK)

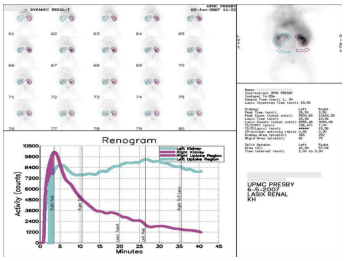


Ultrasound image showing only hydronephrosis (urine inside the kidney appears black)



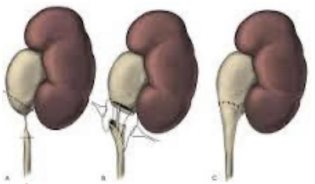
Here longitudinally we see thinning of the renal parenchyma

- **Dynamic renogram**[2] used for conformation and to rule out obstruction as a cause for hydronephrosis. It shows:
  - Washout curve the right kidney curve reaches the top and then drop immediately means there is excretion and draining. The left doesn't drop so there is an obstruction
  - Function: we measure the uptake of isotopes by the kidneys, the summation of both kidneys uptake should be 100% (e.g if it is 30% for 1 kidney then it'll be 70% for the other). Normally each kidney takes 50%



Management

- **Observation:** We wait first and observe there may be spontaneous resolution of the hydronephrosis
- **Surgical intervention:**
  1. Worsening hydronephrosis
  2. Renal function: in dynamic studies if one kidney functions less than 40% or deteriorating more than 10% after follow up.
  3. Pyelonephritis: On top of the pre-existing hydronephrosis
  4. Stone formation



Dismembered Pyeloplasty

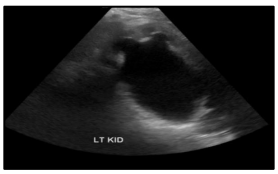
we cut the area of narrowing and reattach the ureter, there are a lot of techniques that can be used, but this is the most common one used

1. When hydronephrosis observed the next step is to rule out vesicoureteral reflux by MCUG since VUR is a more common cause of hydronephrosis than ureteric obstruction
2. There are 2 studies DTPA and MAG-3 (isotope renography): determines whether the dilatation of the pelvis and calyces is truly obstructive in nature

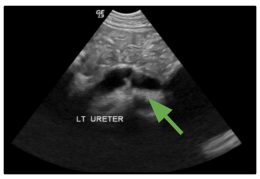
Ureterovesical junction obstruction (UVJO) (Megaureters) 5th most common

Diagnosis

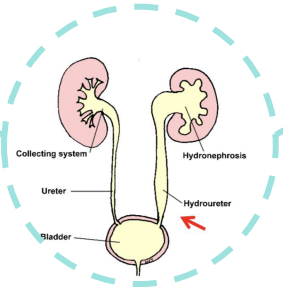
MCUG will be normal



In US dilated kidney



Normally we don't see ureter but here the ureter is dilated



the obstruction between ureter and bladder dilation of the renal pelvis and ureter (hydrouretronephrosis)

Management

We wait there might be spontaneous resolution, the indication for surgery same as UPJO but the procedure here is **Ureteral reimplantation**

Vesicoureteral Reflux (VUR) 3rd most common:

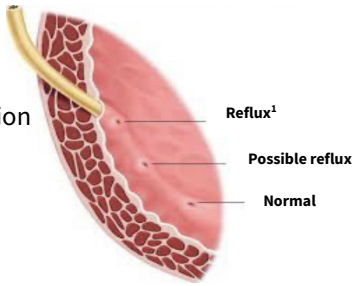
Normal anti-reflux mechanism "Flap valve"

01 Oblique course as it enters the bladder

02 Proper muscular attachments to provide fixation

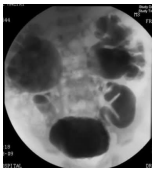
03 Posterior support to enable its occlusion

04 Adequate submucosal length



Presentation:

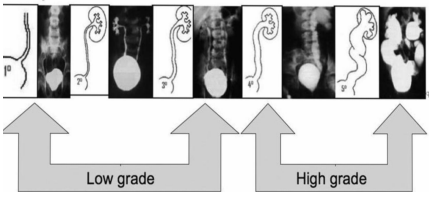
- Asymptomatic
  - Prenatal
  - Fluctuated dilatation
- Febrile UTIs



X-Ray with contrast normally the contrast stays in the bladder, If there's any contrast going up it means there's a reflux which is Abnormal

Diagnosis

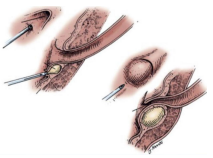
★ MCUG (VCUG): we stage it for treatment: low grade 1,2,3 no need to treat surgically (Spontaneous resolution of reflux) give patients prophylactic antibiotics to prevent UTIs unlike high grade 4,5 needs surgery



Management

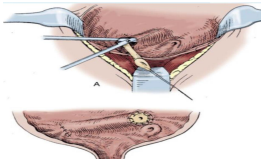
- **Prophylactic antibiotic:** Spontaneous resolution
- **Surgical treatment:**
  1. Recurrent pyelonephritis on antibiotic prophylaxis (outbreak infection)
  2. Noncompliant with medical treatment
  3. Persistence of reflux (high grade)

Endoscopic treatment



Ureteral reimplantation

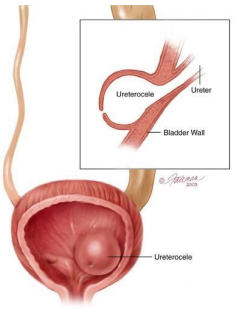
higher success rate



1. Short ureter length entering the bladder result in **primary reflux:** in children may correct on its own if it is not associated with any other abnormality. **Secondary reflux:** If the abnormality in the bladder or urethra or in adults needs definitive surgical management in the form of vesicoureteroplasty

Ureterocele

- A cystic dilation of the distal aspect of the ureter
  - Either within the bladder
  - or spanning the bladder neck and urethra
  - Ureterocele cause obstruction and the ureter will be dilated
- Initial treatment is (Endoscopic incision of ureterocele)
  - Not indicated if there's no back pressure.



Presentation:

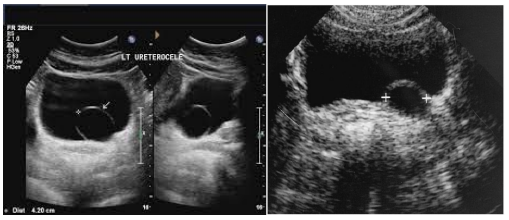
01 **Antenatal (U/S).**

02 **Urine retention.**

03 **Calculus formation.**

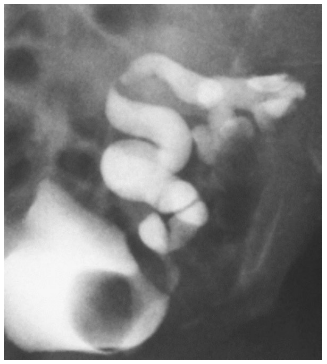
04 **Infection.**

Investigations:



Ultrasound

- The best modality.
- You can see the bulging distal ureter, which is stenosed and prevent urine to reach Urinary Bladder -the stenosis severity may vary from mild to complete occlusion-



MCUG-micturating cystourethrogram

- Used for confirmation.
- Picture Showing filling defect of bladder.

Duplication Anomalies of ureters.

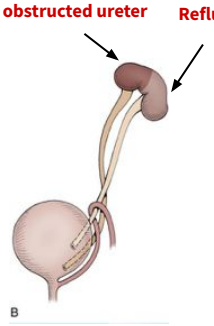
- 1% of all births
- 1.6:1 female to male ratio
- 85% unilateral

- Associated with:
  - Reflux 43 %
  - Renal dilatation 29%
  - Ectopic insertion 3%
  - Ureterocele

Incomplete duplications

- 2 renal pelvises drain into 2 separate ureters that join together distally and form a single ureter

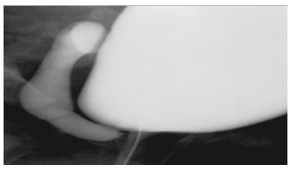
Complete duplications

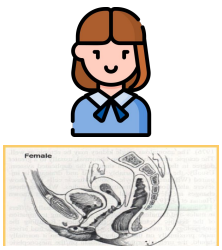


- There are 2 moieties (upper & lower) of renal pelvises & ureter.
- Usually, upper moiety of (ureter & Kidney) is Associated with obstruction
- Lower moiety Associated with reflux “bcs the ureter is short”

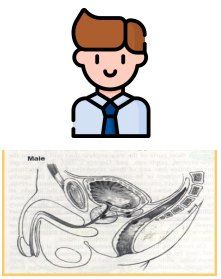
Ectopic Ureter

- An ectopic ureter is any ureter, single or duplex, that doesn't enter the trigonal area of the bladder.
- The ectopic ureter may open into abnormal site in the urinary bladder, or outside the urinary bladder (most severe) (99% are found out the bladder, most cases open into urethra)





- **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**

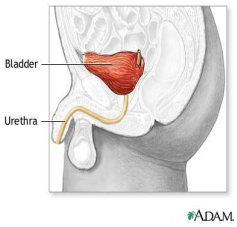


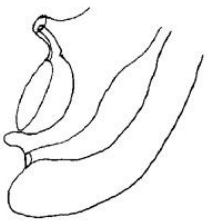
- In males the ectopic ureter always enters the urogenital system above the external sphincter or pelvic floor, and usually into the wolffian structures including vas deferens, seminal vesicles, or ejaculatory duct.
- Symptoms occurs as recurrent infections of “all the pathway, E,g : **epididymitis**”

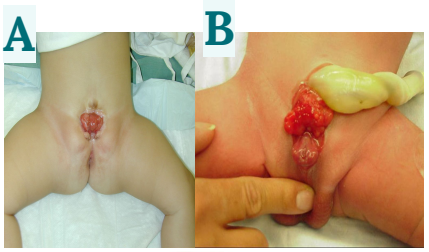
- Dribbling incontinence in a child should raise the suspicion of an ectopic ureter, in which the ureter from the upper pole opens outside the control of the urethral mechanism. The abnormal ureter must be **reimplanted in the bladder**.

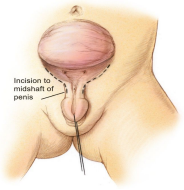
Congenital Anomalies of the Bladder

Bladder exstrophy









Bladder Exstrophy

- The incidence of bladder exstrophy has been estimated as between 1 in 10,000 and 1 in 50,000.
- Exstrophy means “inside-out”, the inside of the bladder here is exposed to the outer
- It is a urological emergency
- The bladder is normally covered by skin, subcutaneous tissue & 3 muscle layers. In this anomaly these coverings will be absent
- Usually presents as complex with epispadias.

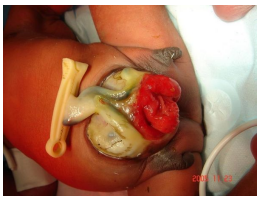
Bladder Exstrophy-Epispadias complex

Pictures are very important

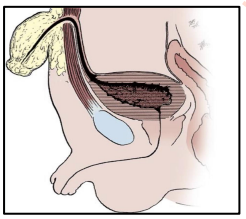
- Picture A : Female
- Picture B: Male, there’s testes

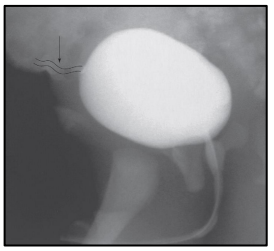
Cloacal Exstrophy

- 1 per 200,000 live births (Rare, but serious)
- Cloaca is the embryologic origin that divides into bladder and rectum.
- It is a urological emergency
- GI tract is involved here.
- 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.
- Associated anomalies:
  - Omphalocele
  - Gastrointestinal anomalies: Malrotation, duplication, duodenal atresia and Meckel diverticulum
  - Genitourinary anomalies: Separate bladder halves and bifid genitalia



Urachal Abnormalities





- Usually detected postnatally due to **umbilical drainage** mother complain of wet umbilicus
- The urachus runs from the apex of the bladder to the umbilicus. It is normally obliterated at birth - closed at its end- but may give rise to **cysts**, a urinary **fistula**, or a discharging umbilical **sinus** if parts of it remain patent. Symptomatic remnants require excision.
- The only normal part in this urethra is the distal urethra
- Imaging possibilities include ultrasound, CT, and VCUG.

01

Asymptomatic patients

Conservative treatment with observation due to possible spontaneous resolution.

02

Infected urachal remnant

Usually in urachal sinus. Initially are treated with drainage and antibiotics, followed by surgical excision.

03

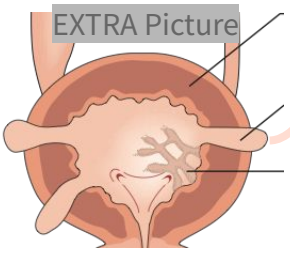
Non-resolved urachal remnants

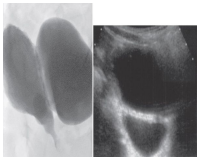
should be excised due to the increased risk of later **adenocarcinoma** formation

Bladder Diverticulum

- **Primary Diverticula:** Arises as a localized herniation of bladder mucosa at the ureteral hiatus & are most likely caused by a **congenitally** deficient bladder wall "weak wall = forming a sac".

EXTRA Picture



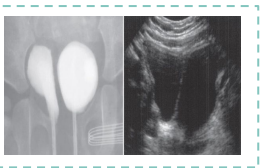


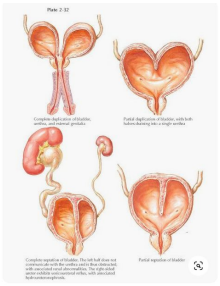
- **Secondary Paraureteral Diverticula:** Are acquired and develop due to existing infravesical obstruction. Usually due to chronic retention.

Diagnosis and Management

- Bladder diverticula can be detected on **prenatal ultrasound.**
- The gold standard remains VCUG (Voiding cystourethrogram), which will reveal possible accompanying VUR
- We don't treat unless it's Symptomatic diverticula, Especially in conjunction with VUR or if it's very large should be **treated surgically**

Bladder Duplication





- Bladder duplication is often associated with duplication anomalies of the external genitalia & lower GIT
- Presentation vary depending on the abnormality. It may presents with separated urethra or with common urethra, one genitalia or more than one

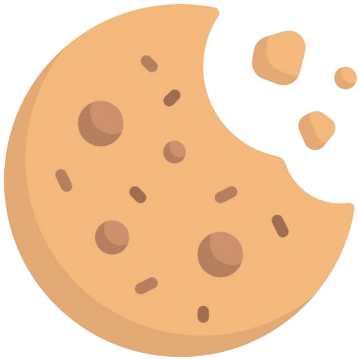
We connect the two bladders and make one external urethral meatus

01

- Initial treatment:
  - Directed towards renal preservation.
  - Prevention of infections

02

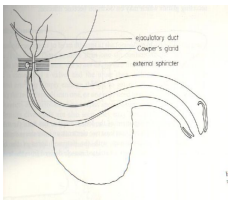
- Long term goal:
  - Achieving continence and reconstructing the internal and external genitalia
  - Due to the rarity of the disease & the large variety of presentation, the surgeries must be individualized

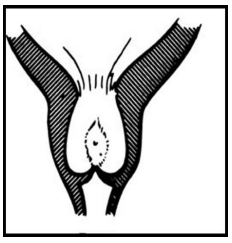


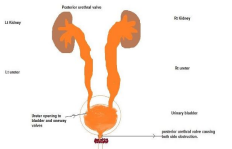
You deserve a break!

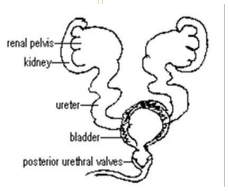
Urethra and external genitalia.

Posterior Urethral Valves (PUV) 4th most common









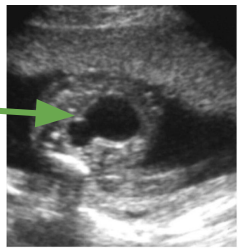
- It's an extra tissue that grow in posterior part of urethra and "narrow, not completely close" the opening of bladder
- Disease of boys, it's not common but serious, why? B/c the obstruction will affect all urinary tract (universal obstruction of urinary tract)
- There's another anomaly called (Anterior UV), but it's very rare.
- 1 in 8000 to 25,000 live births
- Make up 10% of urinary obstructions diagnosed in utero
- Most common cause of urine retention in male infants
- 50% have renal impairment.

Associated Findings

- Presentation usually is **antenatally** by US
- Urine retention, UTI, Poor urinary stream, Urinary incontinence, CRF (ESRD)
- **Features of classic case presentation**
  - Boy, bilateral hydronephrosis and has bilateral hydroureter with thickened bladder, Keyhole sign on Ultrasound
- The in utero bladder and the kidneys developed under high pressure and resistance (Kidney start to produce urine at age of 20 weeks.) So it's associated with:
  - Oligohydramnios Detected by US prenatally
  - Bilateral renal Dilatation
  - VUR: 40%
  - Valve bladder
  - Renal impairment

01 PUV Investigations

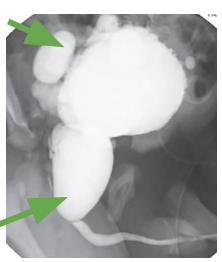
Keyhole sign



- In U/S you can see **bilateral hydronephrosis** & thickened bladder wall.

Sometimes there is reflux

Dilated part of urethra "between extra valve and bladder"

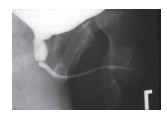


★ MCUG is the **best choice** for diagnosis

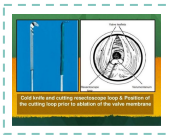
- The only normal part in this urethra is the distal urethra

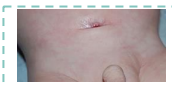
02 PUV Treatment

- **Initial treatment** (whatever patient condition, we start by these to stabilize patient:
  - Feeding tube insertion
  - Start antibiotic prophylactic
  - MCUG & U/S
- Surgical treatment:
  - **Endoscopic valve ablation:** Classic treatment, simply to cut the extra tissue by a knife "only cutting, not removing"
  - **Cutaneous Vesicostomy:** In patients with renal failure or low birth weight, After 1 year, we do endoscopic valve ablation.

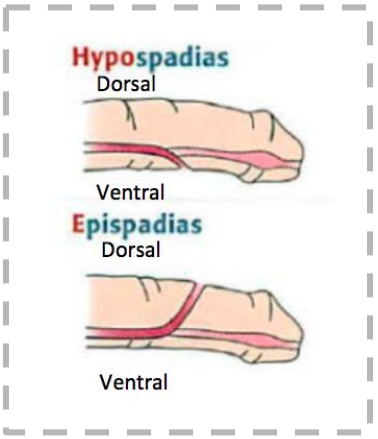


Anterior urethra is normal but posterior urethra is dilated



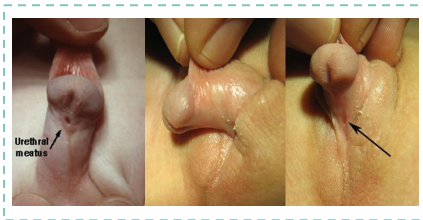


Hypospadias 6th most common & Epispadias



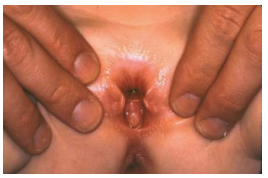
Hypospadias

- Very Common
- Abnormal position of the EUM (external urethral meatus) on the **ventral** (towards the scrotum) surface
- Types:
  - The closer to original meatus called: **Distal hypospadias**
  - If the opening closer to the scrotum, it called: **Proximal hypospadias**
  - The more proximal the opening, the more severe it is.
- The corpus spongiosum may be scarred and fibrosed, leading to a ventral curvature or chordee of the penis
- ★ **NO Circumcision** (Absolute contraindication, because we need this skin for reconstruction of the meatus in the future)
- 6 to 9 months repair (Surgery before 6 months associated with poor outcome).
- The aim of treatment is to correct the chordee by excising the fibrosis, and then to construct a new urethral opening in the normal position on the glans. This procedure should be ideally completed before the boy goes to school



Epispadias

- Very rare
- Ectopic opening of the external urethral meatus in the dorsal side.
- Females present with continuous urinary incontinence
- Other associated abnormalities include separation of the symphysis pubis and rectal prolapse.
- Reconstruction of these deformities is not always successful, and urinary incontinence may remain a major problem and require urinary diversion.



Female

Male

GU congenital anomalies: Others

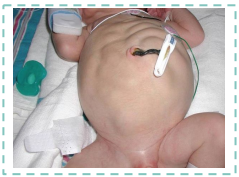
Prune-Belly syndrome

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

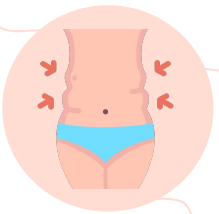
- Others names:
  - Triad syndrome
  - Eagle-Barrett syndrome
  - Abdominal musculation syndrome

The 3 major findings (triad):

Three systems are affected: musculoskeletal, urinary and genital



01 **Deficiency of the abdominal musculature**
(You can see the bowel movement very clearly)



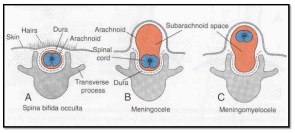
02 **Bilateral intra-abdominal testes**
no intra abdominal pressure to descend the testes

03 **Anomalous urinary tract**
bilateral hydroureteronephrosis and an enlarged bladder same as PUV

Neuro-Spinal Dysraphisms (Dysraphisms means incomplete fusion)

- 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 (Any mass or abnormality that compress or compromise the spinal cord may affect the bladder function. So, **don't forget to examine the back** of any patient who is presenting with urological symptoms)
- Lesions vary like:

01 small lipomeningocele. Needs immediate surgical intervention



02 Abnormal Gluteal cleft

03 Sacral Dimples

04 Hair Patches (Spina bifida occulta)

This patient have cafe au lait spots

05 Dermal Vascular malformation

Summary

	Presentation	Association	Diagnosis	Management

MCDK	• Symptomatic
	  ○ Mass
	  ○ UTI
	  ○ Pain
		• Contralateral VUR 18%-43%	• Prenatal US
			• Incidental in Neonates/Children
			• DMSA: Little or no uptake of radionuclide
			• MCUG/ VCUG: now days not used routinely unless the patient symptomatic
				• Observation
				• Surgical intervention: :
				  ○ Hypertension
				  ○ Pain
				  ○ Pyelonephritis

Super numerary kidney	-	-	• By US	-

Unilateral renal agenesis	• If there's symptoms: Proteinuria, Pain and Hypertension	• Anomalies of other organ systems: CVS, GIT and MSC
		• Müllerian duct abnormalities
			• Prenatal US
			• Incidentally:
			  ○ Abdominal US
			  ○ Abdominal CT
			• DMSA: confirm
				• If symptomatic follow them annually with **urinalysis**, **blood pressure** and **ultrasound**

Bilateral renal agenesis	• Do not survive beyond 48 hours due to respiratory distress associated with pulmonary hypoplasia	• Ureters absent • Bladder absent or hypoplastic • Adrenal glands normal • Müllerian duct abnormalities	-	Do not survive beyond 48 hours due to respiratory distress associated with pulmonary hypoplasia

Horseshoe kidney	-	• UPJO	• The calyces by ultrasound or CT:
			  ○ normal in number
			  ○ atypical in orientation
			  ○ pelvis remains in the vertical or obliquely lateral plane
				• There is <u>no surgery</u> to separate the the 2 kidneys, we just treat the symptoms

Simple renal ectopia	• Mostly asymptotic	• 50% have a hydronephrosis:
		  ○ Obstruction: UPJO and UVJO
		  ○ Reflux (VUR): grade III or greater
		  ○ Malrotation
		• 15% Genital anomalies
			-	• There is no need to do surgery, we just treat the symptoms if exist

Summary

	Presentation	Association	Diagnosis	Management
Crossed renal ectopia	-	• 90% are fused	-	• There is no need to do surgery, we just treat the symptoms if exist

UPJO	• UTI
	• Pain
	• Mass
	• Hematuria
	• Stone
		• only hydronephrosis	• Prenatal US: only hydronephrosis
			• Incidental
			• Dynamic renogram used for conformation
				• Observation
				• Surgical intervention (**Dismembered Pyeloplasty**):
				  ○ Worsening hydronephrosi
				  ○ deteriorating Renal function
				  ○ Pyelonephritis
				  ○ Stone formation

UVJO	-	• Hydrouretronephrosis	• US: dilated kidney	We wait there might be spontaneous resolution, the indication for surgery same as UPJO but the procedure here is **Ureteral reimplantation**

VUR	• Asymptomatic
	• Febrile UTIs
		-	• MCUG (VCUG	• **Prophylactic antibiotic:** Spontaneous resolution
				• **Surgical indication:**
				  1. Recurrent pyelonephritis on antibiotic prophylaxis (outbreak infection)
				  2. Noncompliant with medical treatment
				  3. Persistence of reflux (high grade)
				• Procedure:
				  ○ **Ureteral reimplantation**
				  ○ **Endoscopic treatment**

Ureterocele	• Antenatal US • Urine retention • Calculus formation • Infection	-	• US: the best modality • MCUG: confirmation	• Endoscopic incision of ureterocele: not indicated if there's no back pressure

Summary

	Presentation	Association	Diagnosis	Management

Ureter duplication	-	• Associated with:
		  ○ Reflux 43 %
		  ○ Renal dilatation 29%
		  ○ Ectopic insertion 3%
		  ○ Ureterocele
			-	-

Ectopic ureter	• Recurrent infections of "all the pathway, • classic symptom in female is **continuous wetting**	-	-	• **reimplantation in the bladder**
Bladder exstrophy	• Emergency • coverings above bladder will be absent	• Usually presents as complex with epispadias.	-	-
Cloacal exstrophy	• Emergency • GI is involved	• Omphalocele • Gastrointestinal anomalies • Genitourinary anomalies	-	-
Urachal abnormalities	• Usually detected postnatally due to **umbilical drainage** mother complain of wet umbilicus	-	• ultrasound • CT • VCUG	• Asymptomatic patients: **conservative** • Infected urachal remnant: **drainage and antibiotics**, followed by **surgical excision** • Non-resolved urachal remnants: should be **excised** due to the increased risk of later **adenocarcinoma**
Bladder diverticulum	-	• conjunction • VUR	• prenatal ultrasound • VCUG: gold standard	• We don't treat unless it's Symptomatic diverticula, • in conjunction with VUR or if it's very large should be **treated surgically**

Summary

	Presentation	Association	Diagnosis	Management

Bladder duplication	• separated urethra or with common urethra
	• one genitalia or more than one
		• duplication anomalies of the external genitalia & lower GIT	-	• **Initial treatment:** renal preservation and Prevention of infections
				• **Long term goal:**
				  ○ 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

PUV	• Urine retention
	• UTI
	• Poor urinary stream
	• Urinary incontinence
	• CRF (ESRD)
		• Oligohydramnios
		• Bilateral renal Dilatation
		• VUR: 40%
		• Valve bladder
		• Renal impairment
			• U/S: bilateral hydronephrosis & thickened bladder wall and keyhole sign
			• MCUG is the **best choice** for diagnosis
				• **Initial treatment**:
				  ○ Feeding tube insertion
				  ○ Start antibiotic prophylactic
				• **Surgical**:
				  ○ Endoscopic valve ablation
				  ○ Cutaneous Vesicostomy

Hypospadias	-	• ventral curvature or chordee of the penis	-	• NO Circumcision • 6 to 9 months repair
Epispadias	• continuous urinary incontinence in female	• separation of the symphysis pubis • rectal prolapse	-	• Reconstruction of these deformities is not always successful • incontinence require urinary diversion.
Prune-belly syndrome	• Three systems are affected: musculoskeletal, urinary and genital	• Deficiency of the abdominal musculature • Bilateral intra-abdominal testes • Anomalous urinary tract	-	-
Neuro-spinal dysraphisms	• Cutaneous lesions occur in 90% of children with various occult dysraphic states	• Small lipomeningocele • Abnormal Gluteal cleft • Sacral Dimples • Hair Patches • Dermal Vascular malformation	• examination of the back	• Refer to neurosurgeon

Quiz

MCQ

Q1: what is the best test to establish that dilatation is caused by obstruction?

A) Ultrasound
B) Isotope renography
C) Micturating cystourethrogram

Q2: The most common cause of antenatal hydronephrosis?

A) Pelviureteric junction obstruction
B) Vesicoureteric reflux
C) Ureterovesical junction obstruction

Q3: A 1-year-old girl is brought to the physician because of fever and crying while passing urine for 2 days, she was treated for a urinary tract infection with oral cefixime. Renal ultrasonography shows hydronephrosis of the left kidney. Empirical antimicrobial therapy is initiated. Which of the following is the most appropriate next step in diagnosis?

A) Dynamic renogram
B) Voiding cystourethrography
C) Cystoscopy

Q4: 1 day-old boy with a history of bilateral antenatal hydronephrosis and hydroureter with thick walled bladder and mild oligohydramnios. After confirmation by MCUG and stabilizing the patient what is the best procedure to do?

A) Ureteral reimplantation
B) Cutaneous vesicostomy
C) endoscopic valve ablation

Q5: An infant presented to you with a discharge from umbilicus. What is the most likely diagnosis?

A) Ureterocele
B) Prune belly syndrome
C) Patent urachus

Q6: surgical treatment for vesicoureteral reflux is indicated in which of the following?

A) Stone formation
B) Pyelonephritis
C) non-compliant medical treatment

Answers

Q1	B	Q4	C
Q2	A	Q5	C
Q3	B	Q6	C

Extra Questions

Good Luck!

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Feedback