L15

For the Dr notes (Important File!!) click here
(Thanks To Mishal AlSuwayegh & Mohammad AlRashed)

For more info about the lecture
click here

Genitourinary anomalies

Objectives:

(pathophysiology, etiology, clinical manifestations, complications, Guide line of managements)

- 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

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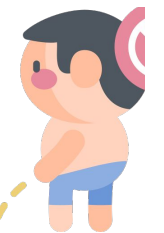
Genitourinary Anomalies

More common

- 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



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

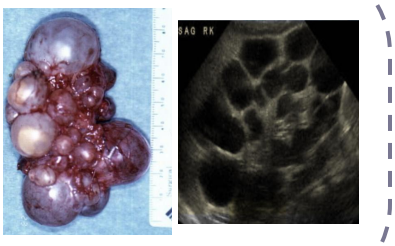
Congenital Anomalies of Kidney

Kidney anomalies

- **Number**
  - Supernumerary — More
  - agenesis — Absence
  - Horseshoe — Fusion
- **Location**
  - Rotation
  - Ectopia — Displacement

Multicystic dysplastic kidney (MCDK) 2nd most common

- There is a difference between MCDK (not working من الأساس)and polycystic kidney disease loss of function over time
- Unilateral, The kidney is completely replaced by cysts, no necrosis but a non-functional kidney or very low function in the kidney (1% -2%)
- There is no nephrons , only disturbed cysts[1]
- 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[2].



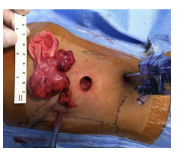
Dark color (fluid-urine)

Diagnosis

- Prenatal US most cases
- Incidental in Neonates/Children
- Symptomatic [3]
  - Mass
  - UTI
  - Pain
- DMSA: nuclear study to look for the function of kidneys
- Little or no uptake of radionuclide [4]
- 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%

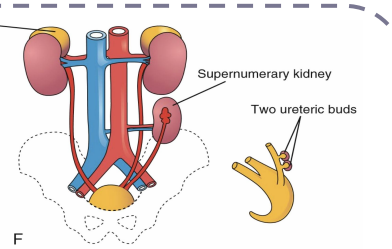
Management

- Observation + follow up (asymptomatic)
  - Cyst fluid disappears
  - Fluid might increase > kidney enlarges > palpable kidney and symptoms start .
- Surgical intervention (nephrectomy)[5]:
  - (renal) Hypertension (medical complications)
  - Pain
  - Pyelonephritis
  - In case the ultrasound report showed worsening of the sizes of the cysts.



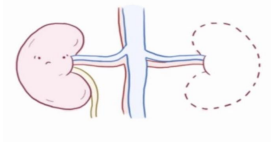
Supernumerary Kidney:

- 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 [6] No intervention until the patient experience symptoms.

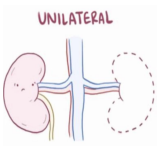


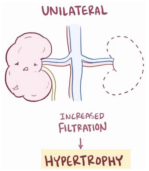
1-There is no renal tissue & there is no communication between them
2-(No Mickey mouse sign unlike in UPJO)
3-same other obstructive uropathy
4-The difference between UPJO and MCDK in US that in MCDK there is NO or very little function (diagnostic).
5-indication
6-detected by US confirmed by radiological like CT

Kidney agenesis:



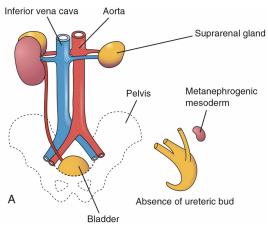
Unilateral Agenesis [2]

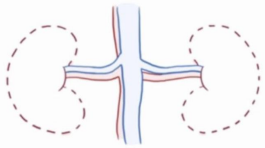




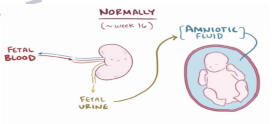
The blood flow normally goes to the each kidney 50%, in case of unilateral renal agenesis the blood flow 100% goes to the only kidney which with time will be exhausted and hypertrophied cause it's doing all the job and shows symptoms: Proteinuria (due to hyper filtration), Pain and Hypertension, we must follow them annually with **urinalysis**, **blood pressure** [1]

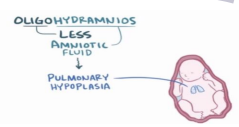
- 1 in 1100 births
- Male: Female of 1.8 : 1
- The left side is absent more frequently
- The ipsilateral ureter is completely absent in 50%



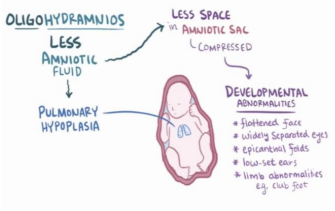


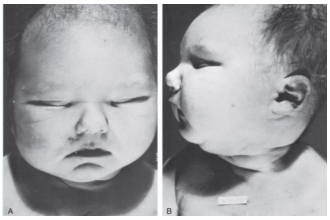
Bilateral Agenesis [3] rare





No kidney = no amniotic fluid[4] = No space for the fetus to develop = leading to Potter's sequence
The most affected organ are the lungs (pulmonary hypoplasia)-this happens due to the high blood pressure which doesn't allow time for the development - 50% die intra- uterine





This is called Potter face (associated with potter syndrome)

Associated anomalies

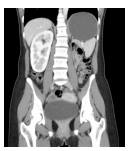
- Anomalies of other organ systems are found frequently in affected individuals: CVS, GIT and MSC
- Müllerian duct abnormalities:
  - 25% to 50% of females
  - 10% to 15% of males
  - Approximately one fourth to one third of women with Mullerian duct anomalies are found to have URA

Associated anomalies

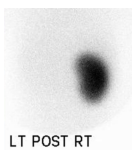
- Ureters are almost always absent.
- Bladder is either absent or hypoplastic.
- Adrenal glands are usually positioned normally.
- Müllerian duct anomalies are commonly observed.

Diagnosis

- Prenatal US
- Incidentally:
  - Abdominal US
  - Abdominal CT
- Confirmed:[5]
  - Nuclear study (DMSA) (100% sensitive)



Abdominal CT shows absent left kidney



DMSA shows if there's renal tissues or not

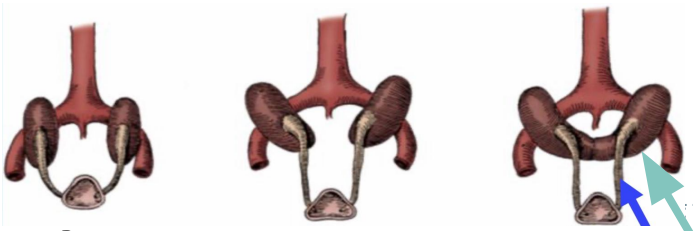
Prognosis

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

1-(checking for renal hypertension)and **ultrasound**
2-if the kidney is absent that doesn't mean that the suprarenal gland will be absent too, because both have different embryological origin
3-After 20 weeks in gestation , the fetal kidney form the amniotic fluid with the urine
4-Oligohydramnios
5-CT alone is not enough since we might miss the diagnosis

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.[1]



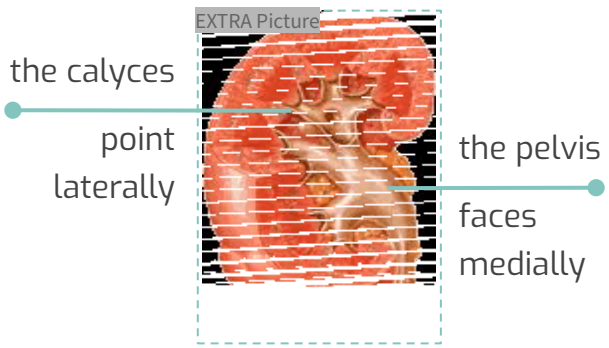
- The calyces [2]
  - normal in number
  - atypical in orientation
  - pelvis remains in the vertical or obliquely lateral plane

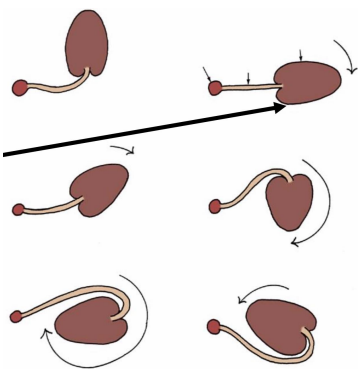
The isthmus is bulky and consists of parenchymatous tissue, it will get stuck under the Inferior mesenteric artery (failure of ascending)

- Horseshoe kidney is frequently found in association with other congenital anomalies.
  e.g. (Trisomies 21 and Turner Syndrome)
- **UPJ** (Ureteropelvic junction) obstruction in one third [3]
- 60 % asymptomatic [4]
- 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





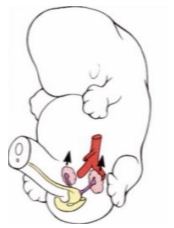
- When this alignment is not exact, the condition is known as malrotation
- Frequently associated with Turner syndrome

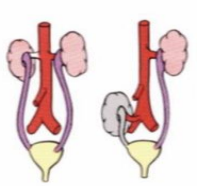
1-Each kidney is drained by its own ureter.
2-by ultrasound or CT
3-due to the acute angle of the ureter
4-The rest 40% of Symptomatic patients is due to **hydronephrosis** (Surgery is required)

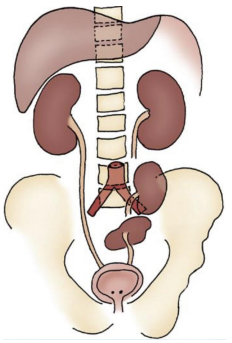
Renal Ectopia:

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

Simple Renal Ectopia [1]



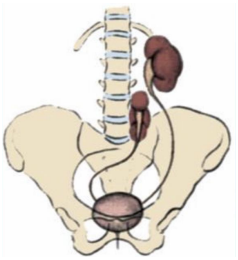




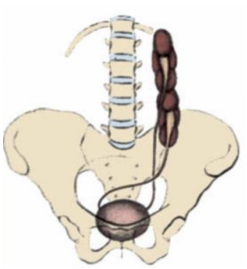
- Left more than the right
- 1 of 2100 to 3000 autopsies.
- Most ectopic kidneys are clinically asymptomatic No need for intervention unless symptoms appear

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.

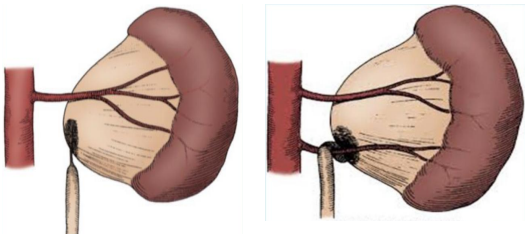
1-The kidney arrested instead of ascending to its normal position in the same original side.

Congenital Anomalies of The Ureter

Ureteropelvic junction obstruction (UPJO) most common

Presentation

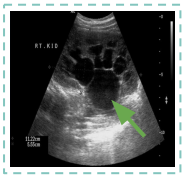
- Prenatal US [1]
- Incidental in Neonates/Children eg : patient comes with abdominal pain,vomiting,crying, they do US and find the obstruction
- By symptoms:
  - UTI:[2]
  - Pain: [3]
  - Mass
  - Hematuria
  - Stone: [4] in renal pelvis



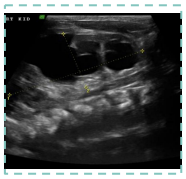
-It's not actually obstruction (if complete obstruction kidney loses its function) , Just narrowing of the ureter
-It's either : intrinsic (due to partial obstruction) or extrinsic (due to the crossing of vessels)

Diagnosis

- First investigation [5] is US [6]
  - Dilated renal pelvis [7]

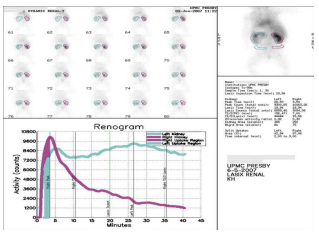


Ultrasound image showing only hydronephrosis (urine inside the kidney appears black). It's called isolated hydronephrosis as it's not associated with other abnormality in the bladder or dilation of the ureter .



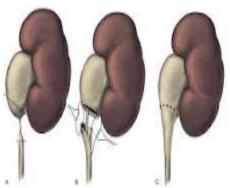
Here longitudinally we see thinning of the renal parenchyma

- **Dynamic** or (diuretic) **renogram** [8] used for conformation and to rule out obstruction [9]
  - Washout curve [10]
  - 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 patient gets older > dilation of pelvis > decrease in resistance (assess by US)
- **Surgical intervention:** (If the patient is symptomatic)
  1. Worsening hydronephrosis
  2. Renal function [11]: less than 40% deteriorating more than 10%
  3. Pyelonephritis [12]:
  4. Stone formation.



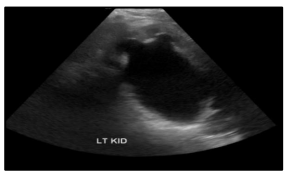
Dismembered Pyeloplasty
we cut the area of narrowing and reattach the ureter, there are a lot of techniques that can be used, but this is the most common one used

1-most cases
2-Pyelonephritis
3-Flank pain
4-Secondary to urine stagnation
5-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
6- and shows Mickey Mouse sign:
7-Dilated major and minor calyces ,The dilatation and distribution of urine are organized unlike in (MCDK)
8-There are 2 studies DTPA and MAG-3 (isotope renography): determines whether the dilatation of the pelvis and calyces is truly obstructive in nature
9-as a cause for hydronephrosis.
10-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
11-in dynamic studies if one kidney functions less than 40% or deteriorating more than 10% after follow up.
12- On top of the pre-existing hydronephrosis

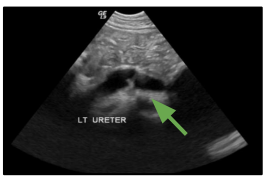
Ureterovesical junction obstruction (UVJO) (Megaureters) 5th most common

Diagnosis

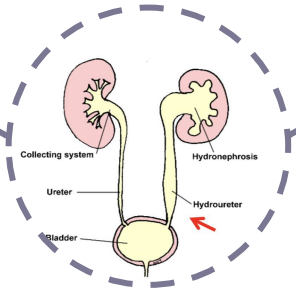
MCUG will be normal



In US dilated kidney



Normally we don't see ureter but here the ureter is dilated Longitudinal black color continuous with the kidney



the obstruction between ureter and bladder dilation of the renal pelvis and ureter [1]

Management [2]

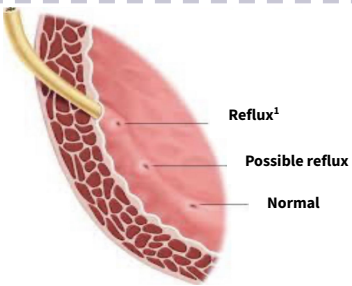
(taking the obstructed part of the urethra out and reattaching the rest)

Vesicoureteral Reflux (VUR) 3rd most common:

Normal anti-reflux mechanism "Flap valve"

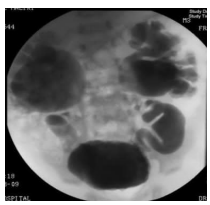
1- Oblique course as it enters the bladder "not angled"
2-Proper muscular attachments to provide fixation
3-Posterior support to enable its occlusion
4- Adequate submucosal length

VUR: means that the urine goes from the bladder back into the UUT



Presentation:

- Asymptomatic
  - Prenatal
  - Fluctuated dilatation
- Febrile UTIs

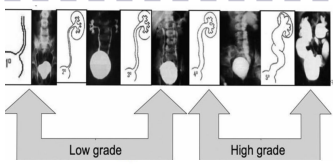


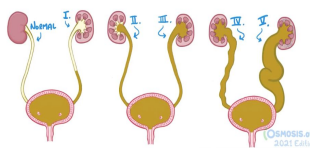
X-Ray with contrast normally the contrast stays in the bladder, If there's any contrast going up it means there's a reflux which is Abnormal

Diagnosis

★ MCUG (VCUG)[3]:

- Done by inserting a feeding tube and injection a contrast per urethra to see the anatomy of LUT- usually the contrast goes to the bladder if it goes to UUT this shows reflux.





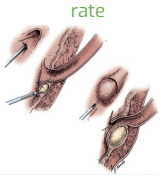
Grade I: urine goes into ureters
Grade II: urine fills entire ureter, renal pelvis
Grade III: urine fills, stretches ureter, renal pelvis
Grade IV: Ureter swollen, curvy and the renal pelvis and calyces become moderately swollen and destroyed.
Grade V: The most severe classification. Urine fills up the ureter, pelvis and calyces causing them swell up completely, and this amount of pressure can lead to kidney failure.

Management

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

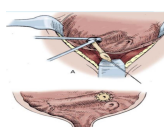
Endoscopic treatment

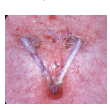
Used with lower graders - 80% success rate

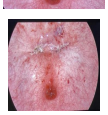


Ureteral reimplantation

higher success rate - laparoscopic or robotic







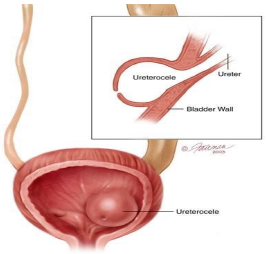
1-(hydrouretronephrosis)
2-We wait there might be spontaneous resolution same as UPJO, the indication for surgery (pain -functional issues)same as UPJO but the procedure here is

Ureteral reimplantation

3-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
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 [1]
- treatment is (Endoscopic incision of ureterocele) [2,3]



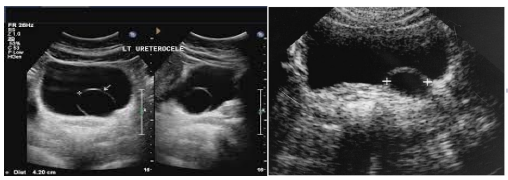
01 **Antenatal (U/S)**

02 **Urine retention**

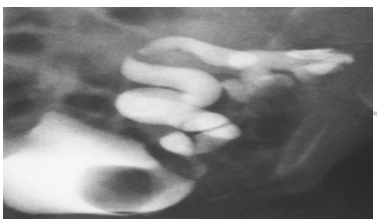
03 **Stone formation**

04 **Infection**

Investigations:



Ultrasound [4]



MCUG-micturating cystourethrogram [5]

Treatment :
- No obstruction ... no treatment
- Ipsilateral obstruction ... postnatal emergency treated by puncture

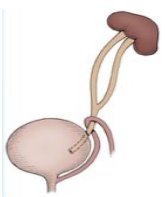
Duplication Anomalies of ureters.

Treatment is indicated only in case of associated complications

- 1% [6]
- 1.6:1 female to male ratio
- 85% unilateral

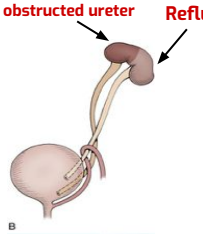
- Associated with:
  - Reflux 43 %
  - Renal dilatation 29%
  - Ectopic insertion 3%
  - Ureterocele

Incomplete duplications



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

Complete duplications

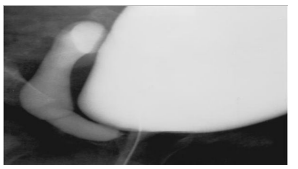


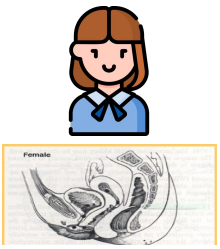
- There are 2 moieties (upper & lower) of renal pelvises & ureter.
- Usually, upper moiety of (ureter & Kidney) is Associated with obstruction
- Lower moiety Associated with reflux "bcs the ureter is short"

1-Ureterocele cause obstruction and the ureter will be dilated
2-Initial treatment
3-Not indicated if there's no back pressure.
4-The best modality , and 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
5-Used for confirmation, Picture Showing filling defect of bladder.
6- of all births

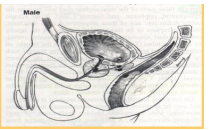
Ectopic Ureter

- An ectopic ureter is any ureter, single or duplex, that doesn't enter the trigonal area of the bladder.[1]
  - Investigation : US , if you suspect it do anatomical study





- In females the ectopic ureter may enter anywhere from the bladder neck to the perineum and into the vagina, uterus,and even rectum.
  - The ureter opens in an area with no sphincter near it .
- One of the classic symptoms is continuous wetting

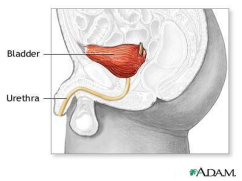


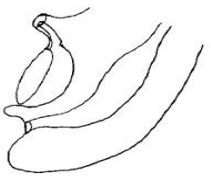
- In males the ectopic ureter always enters the urogenital system above the external sphincter or pelvic floor, and usually into the wolffian structures including vas deferens, seminal vesicles, or ejaculatory duct [2].
  - The ureter opens in an area with a sphincter near it .
  - Symptoms occurs as recurrent "epididymitis" or epididymo orchitis

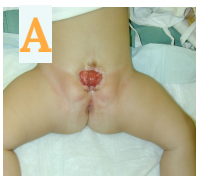
- Dribbling incontinence in a child should raise the suspicion of an ectopic ureter, in which the ureter from the upper pole opens outside the control of the urethral mechanism. The abnormal ureter must be reimplanted in the bladder.

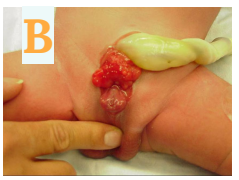
Congenital Anomalies of the Bladder

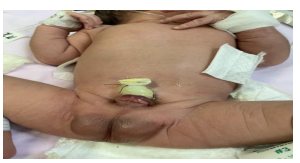
Bladder exstrophy

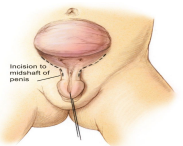












Bladder Exstrophy [3]

- 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 you can see the urine coming out
  - Usually presents as complex with epispadias.
  - Recommendation: delay the treatment till 6 month old

Bladder Exstrophy- Epispadias complex

Pictures are very important

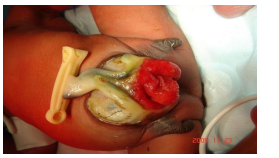
1-The ectopic ureter may open into abnormal site in the urinary bladder, or outside the urinary bladder (most severe) (99% are found out the bladder, most cases open into urethra
2-Symptoms occurs as recurrent infections of "all the pathway".
3-The bladder is normally covered by skin, subcutaneous tissue & 3 muscle layers. In this anomaly these coverings will be absent

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

Congenital Anomalies of the Bladder

Cloacal Exstrophy [1]

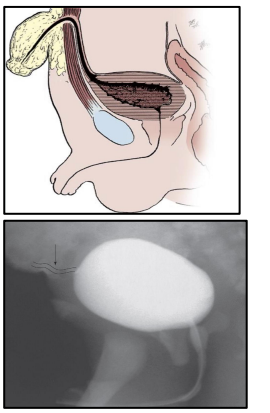
- 1 per 200,000 live births [2]
- Cloaca is the embryologic origin that divides into bladder and rectum.
- Associated anomalies:
  - Omphalocele
  - Gastrointestinal anomalies: Malrotation, duplication, duodenal atresia and Meckel diverticulum
  - Genitourinary anomalies: Separate bladder halves and bifid genitalia



Boy with exstrophy of the bladder and part of GIT and a complete separation of the penis

Urachal Abnormalities

- Usually detected postnatally due to umbilical drainage [3]
- The urachus runs from the apex of the bladder to the umbilicus. It is normally obliterated at birth - closed at its end- it opens because of bladder pathology [4] but may give rise to cysts, a urinary fistula, or a discharging umbilical sinus if parts of it remain patent. Symptomatic remnants require excision.
- 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. remnants are Initially treated with drainage and antibiotics, followed by surgical excision.

03

Non-resolved urachal remnants should be excised the tract or if it reaches bladder we do partial cystectomy due to the increased risk of later adenocarcinoma formation

1- 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.

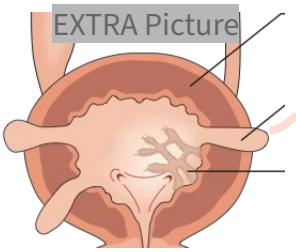
2-rare but serious
3-mother complain of wet umbilicus
4-The only normal part in this urethra is the distal urethra

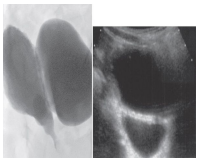
Bladder Diverticulum

Expansion of the mucosa outside of the bladder .

- **Primary Diverticula:** Arises as a localized herniation of **bladder mucosa** at the ureteral hiatus & are most likely caused by a **congenitally** deficient bladder wall [1]
  - Usually it is one sac

EXTRA Picture



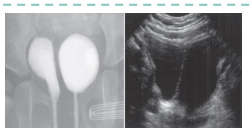


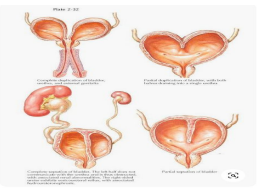
- **Secondary Paraureteral Diverticula** treat the underlying cause : Are acquired and develop due to existing infravesical obstruction. Usually due to chronic retention.
  - Multiple diverticula.

Diagnosis and Management

- Bladder diverticula can be detected on **prenatal ultrasound**.
- The gold standard remains VCUG (Voiding cystourethrogram), which will reveal possible accompanying VUR
- Symptomatic diverticula [2], Especially in conjunction with VUR 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
  - Treat by : connecting 2 bladders together and selecting one good urethra and closing the other one by removing it .

Treatment

01

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

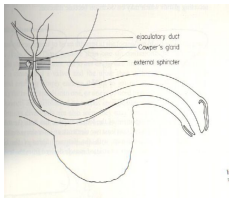
02

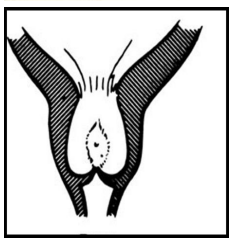
- **Long term goal:**
  - Achieving continence and reconstructing the internal and external genitalia
  - Due to the rarity of the disease & the large variety of presentation, the surgeries must be individualized

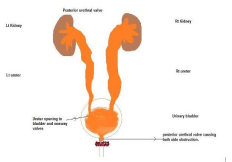
1-"weak wall = forming a sac".
2-We don't treat unless it's Symptomatic or very large diverticula

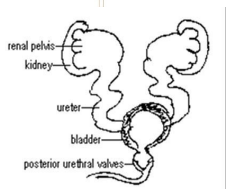
Urethra and external genitalia.

Posterior Urethral Valves (PUV) 4th most common [1,2,3]









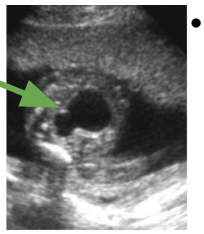
- The main urethra has 2 parts :Anterior urethra (extends from the Fossa Navicularis to the membranes urethra) ; and the Posterior (extends from membranous urethra to the bladder neck ... due to the junction
- extra tissue in posterior part "narrow, not completely close" the opening of bladder
- Disease of boys (universal obstruction of urinary tract)
- 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. 30% have end stage renal failure .

Associated Findings

- Presentation usually is **antenatally**[4]
- Urine retention, UTI, Poor urinary stream, Urinary incontinence, CRF (ESRD)
- Usually the patient needs NICU due to consequence like pulmonary hyperplasia
- 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 [5]
  So it's associated with: complications
  - Oligohydramnios[6] pulmonary hypoplasia same as bilateral agenesis
  - Bilateral renal Dilatation
  - VUR: 40%
  - Valve bladder
  - Renal impairment
  - Respiratory distress [7]

01 PUV Investigations

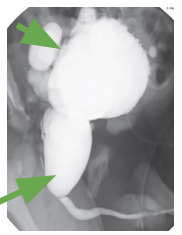
Keyhole sign



- bilateral hydronephrosis & thickened bladder wall.

Sometimes there is reflux

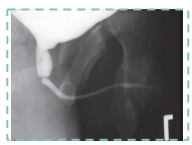
Dilated part of urethra "between extra valve and bladder"



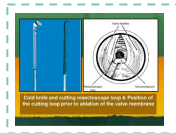
- MCUG is the best choice for diagnosis
- The only normal part in this urethra is the distal urethra
- Trabeculated and elongated bladder (called Christmas tree)

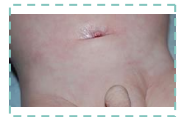
02 PUV Treatment

- **Initial treatment** [8]
  - Feeding tube insertion for temporary recompression of urinary tract
  - Start antibiotic prophylactic
  - MCUG & U/S
- Surgical treatment: (treatment have long term consequences)
  - **Endoscopic valve ablation**[9]
  - **Cutaneous Vesicostomy**[10]: After 1 year, we do endoscopic valve ablation.



Anterior urethra is normal but posterior urethra is dilated





1-It's an extra tissue that grow in posterior part of urethra
2-it's not common but serious, why? B/c the obstruction will affect all urinary tract
3-There's another anomaly called (Anterior UV), but it's very rare.
4-by US
5-(Kidney start to produce urine at age of 20 weeks.)
6-Detected by US prenatally
7-Due to lung hypoplasia. Why lung hypoplasia? Because amniotic fluid normally gets breathed into the developing lungs, which helps to expand the airways and also provides the amino acid proline and both of these things are critical to normal lung development therefore with less amniotic fluid, the lungs may not develop completely.
8-(whatever patient condition, we start by these to stabilize patient:
9-Classic treatment, simply to cut the extra tissue by a knife "only cutting, not removing"
10-In patients with renal failure or low birth weight,

Hypospadias 6th most common & Epispadias

Hypospadias

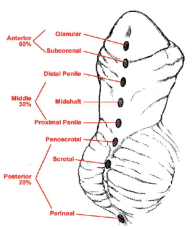
- Very Common
- Abnormal (ectopic) position of the EUM (external urethral meatus) on the ventral (towards the scrotum) surface

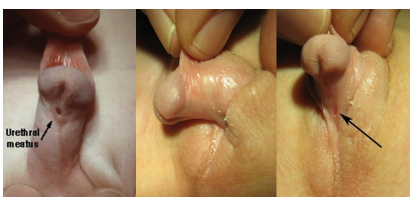
If associated with undescended testes > investigate for gender through chromosomal analysis , might be female with hormonal abnormalities , this is called disorder of sexual differentiation

- Types:
  - The closer to original meatus called: **Distal hypospadias** (less curvature of the penis)
  - If the opening closer to the scrotum, it called: **Proximal hypospadias** (more curvature of the penis)
- 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) used as graft!

- 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





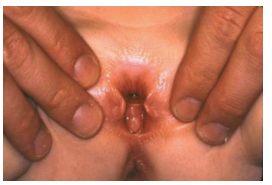
Epispadias [1,2]

- Ectopic opening of the external urethral meatus in the dorsal side. towards the abdomen

For male we have 3 types :

- Penopubic (most severe and associated with urinary incontinence)
- Penile (continence)
- Granular

- 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

1-Very rare
2-Females present with continuous urinary incontinence

GU congenital anomalies: Others

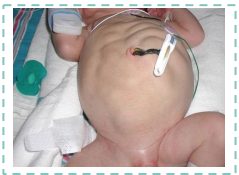
Prune-Belly syndrome

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

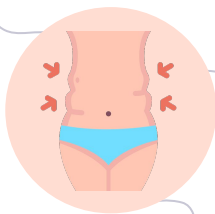
- Others names:
  - Triad syndrome
  - Eagle-Barrett syndrome
  - Abdominal musculation syndrome

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) Only skin and subcutaneous tissues " no muscle "

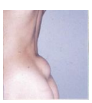


02 Bilateral intra-abdominal testes
no intra abdominal pressure to descend the testes

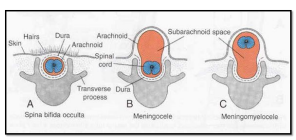
03 Anomalous urinary tract
bilateral hydroureteronephrosis and an enlarged bladder same as PUV

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

Quiz!

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

1. Ultrasound
2. Isotope renography
3. Micturating cystourethrogram

Q2: The most common cause of antenatal hydronephrosis?

1. Pelviureteric junction obstruction
2. Vesicoureteric reflux
3. 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?

1. Dynamic renogram
2. Voiding cystourethrography
3. Cystoscopy

Q4: An infant presented to you with a discharge from umbilicus. What is the most likely diagnosis?

1. Ureterocele
2. Prune belly syndrome
3. Patent urachus

Q1) **B** / *Q2)* **A** / *Q3)* **B** / *Q4)* **C**

القادة

رزان المهنا　　في الدوسري　　محمد الغامدي

نوف الضلعان　　وعد أبو نخاع

الأعضاء

عزام العتيبي　　محمد الزير

شكر خاص لتيم الجراحة دفعة ٤٣٩

حسبي الله لا إله إلا هو عليه توكلت وهو رب العرش العظيم.
اللهم إني أستودعك ما قرأت وما حفظت وما تعلمت فرده لي عند حاجتي إليه إنك على كل شيء قدير.

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