Pediatric urinary disorders

Dr. Hamdan Al-Hazmi
Assistant Professor and
Consultant Pediatric Urologist
Deputy Chairman, Department of Surgery
College of medicine,
King Khalid University Hospital

Notes , 427 team , imp!

Surgery team 429

Badra'a Almuharib- Ismail Raslan

Special thanks to Abdullah Alaoqayil

Congenital anomalies of the urogenital system

- Most common of all organ system.
- 10% of population has some type of urogenital anomaly.
- 14:1000 birth has antenatal diagnosis of urogenital anomaly.
- Antenatal ultrasound after 28 weeks gestation. most of congenital anomalies detected by U/s , 24-28 weeks of gestation urinary system start to be a little bit clear !

Congenital anomalies of the urogenital system…

- Antenatal Hydronephrosis
- Anomaly of position, number and rotation
- Cystic abnormalities
- Prune Belly Syndrome
- Hypospadias
- Epispadias
- Bladder Exstrophy

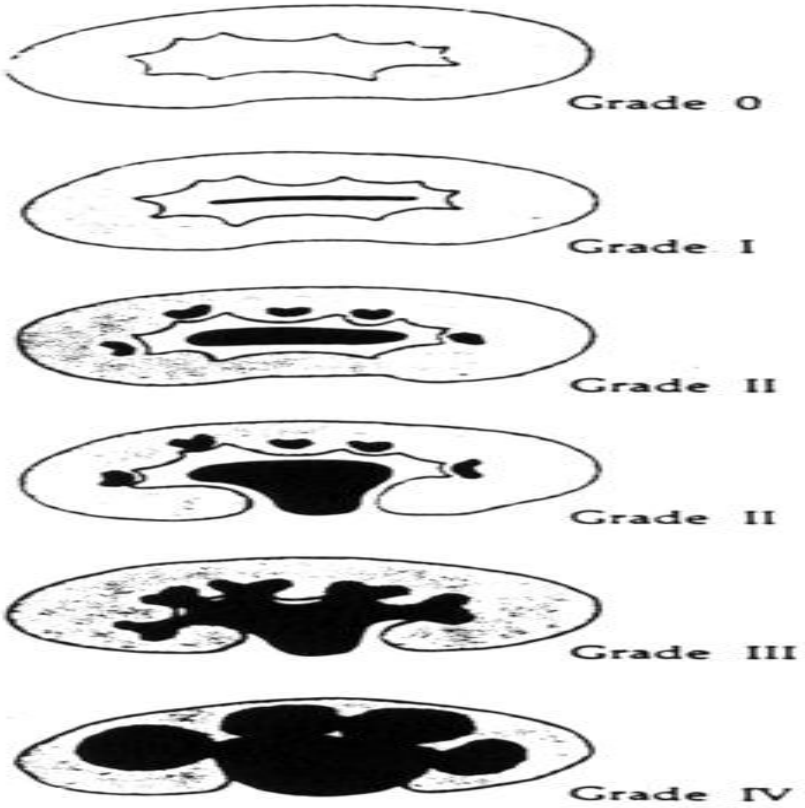
Antenatal Hydronephrosis (ANH)

Not Dx just like syndrome , it's hydronephrosis detected during pregnancy by U/S !!

Causes (DDx) **imp!**:

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

ANH

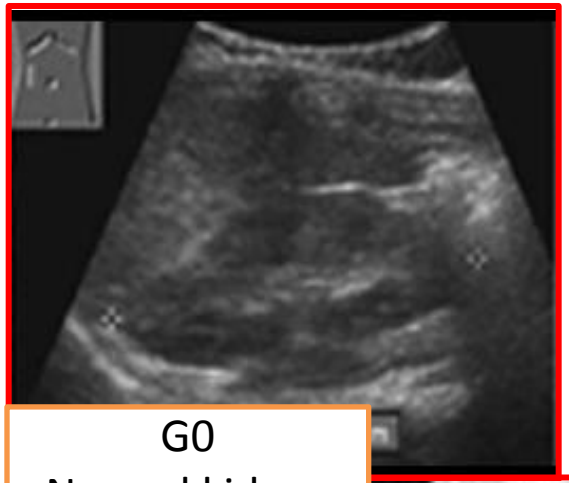


Grades of Antenatal Hydronephrosis

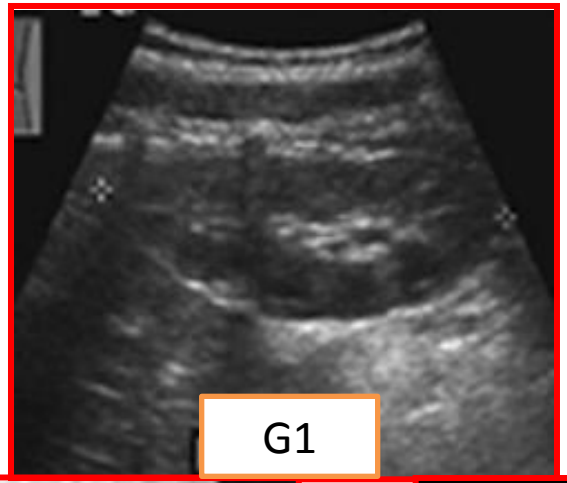
0 > normal Kidney !
from 1 (simplest) to 4 (most severe) depend on the level of the dilatation!

Not imp to know the details of grading just know there is grading system And More sever the grade the more the severity of underlying cause !!

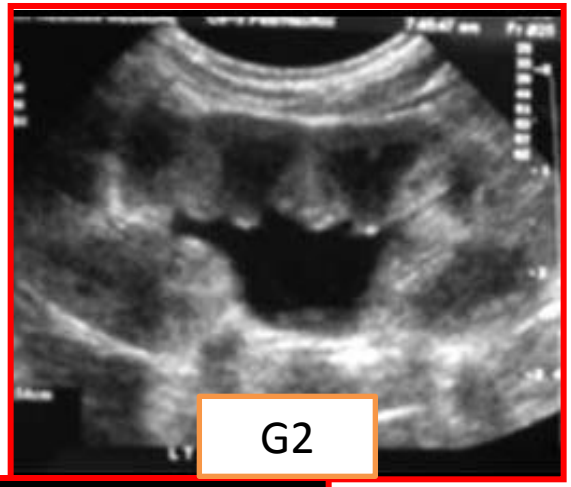
SFU Grading



G0
Normal kidney

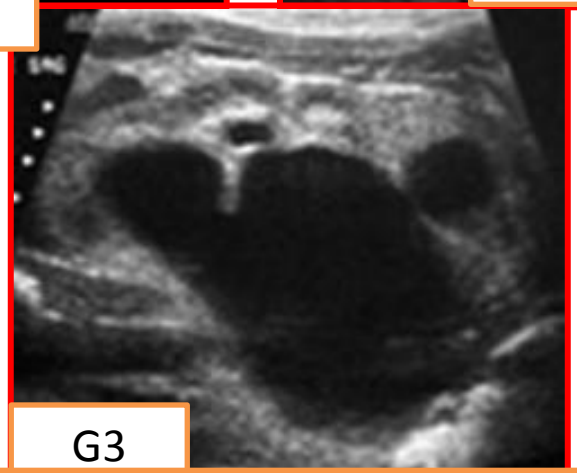


G1

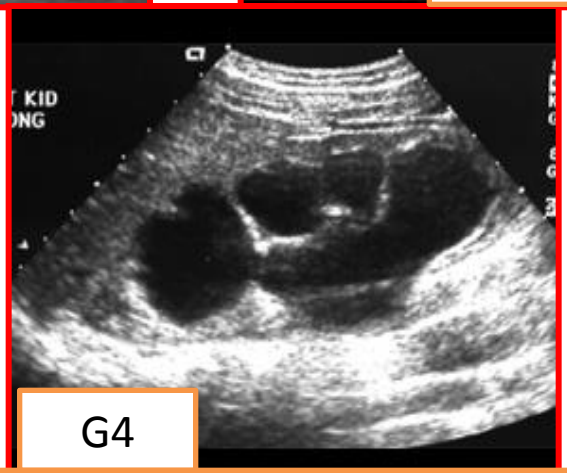


G2

urine in U/S is dark!



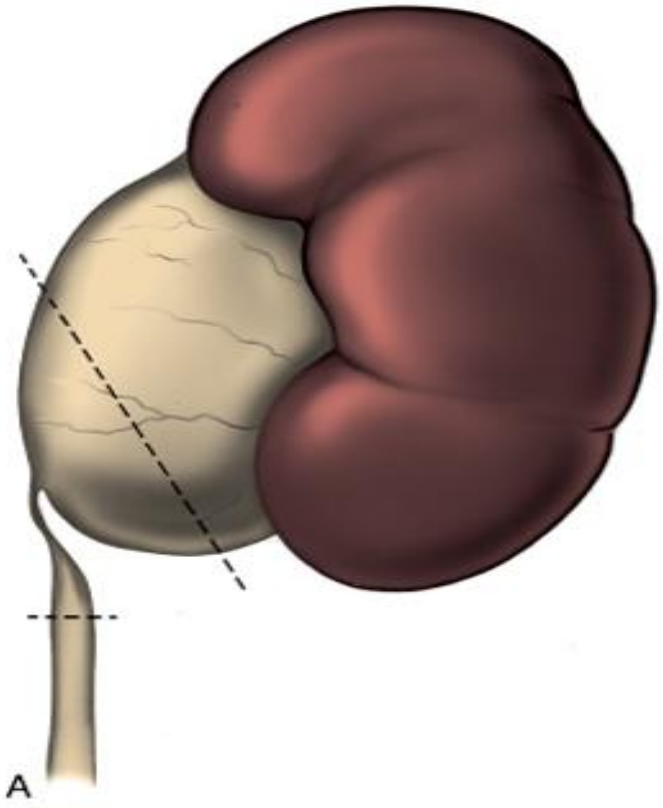
G3

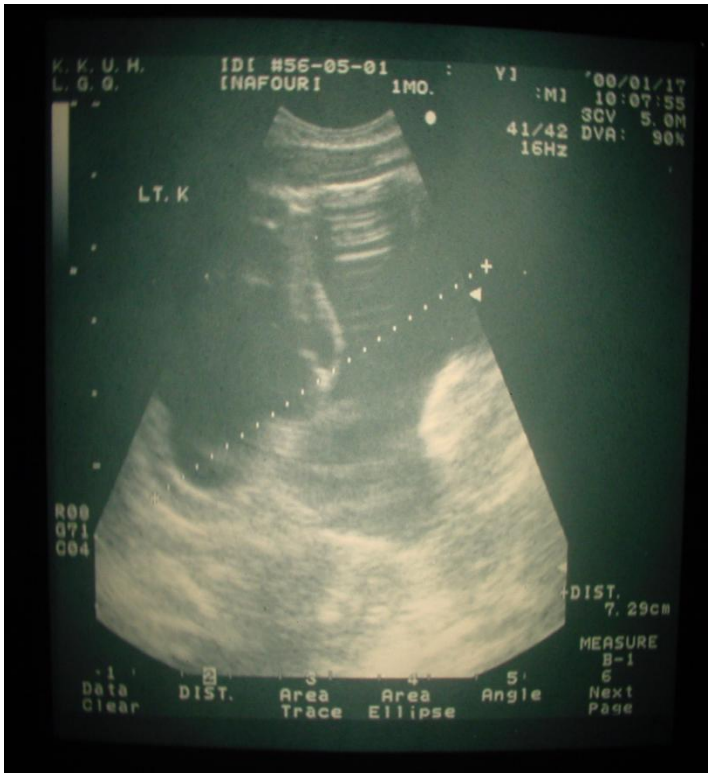


G4

1st Q: What is this anomaly? hydronephrosis , 2nd Q:what is DDx ? !
How to differentiate b/w cyst and hydronephrosis on U/s ? In hydronephrosis there is dilatation of the renal pelvis & renal calyces and there is communication between all But in cyst each cyst is alone and there is no communication b/w cysts !

1st DDx: Pelviureteric junction obstruction(PUJO)





1.It is dilation of renal pelvis due to distal obstruction and the 2.ureter usually is normal

On U/S renal pelvis is dilated but ureter isn't dilated !

Notes !!

- In General what do we mean by hydronephrosis?

- Dilation of pelvicalyceal system

- Ureter : Hydroureter
- Both renal pelvis & ureter:

  Hydroureteronephrosis

PUJO : obstruction of Pelviureteric junction area reflected by dilation of renal pelvis while ureter is normal

IMP: opposite to uretrovesical obstruction : the whole system will be dilated !!

427 notes

- PUJO is the most common cause of (ANH) and children abdominal mass.
- Male: Female ratio 2:1, it occurs in left side more than right.

Etiology (theories) :

- Segmental muscular attenuation
- Angulation
- True Stenosis
- Extrinsic compression
- Crossing vessels ; 20-30%

Associated Findings:

- Reflux, 5-10%
- Contralateral PUJ, 10%
- Contralateral agenesis, 5%

Presentation:

- Incidental in Neonates (by US) or during infancy even b/c during pregnancy we can detect it.
- If it missed during pregnancy the pt will present with all of these symptoms and we do the investigation and we find hydronpehrosis Or detected incidentally during any other investigation for example, pt has trauma and has routine U/S or CT and u find system is dilated !
- Incidental in Children
- the Symptomatic:
  - UTI
  - Pain
  - Mass
  - Hematuria
  - Stone

427 notes

- Investigation:
- U/S
- Renal scan
- VCUG (i.e. voiding cystourethrography)

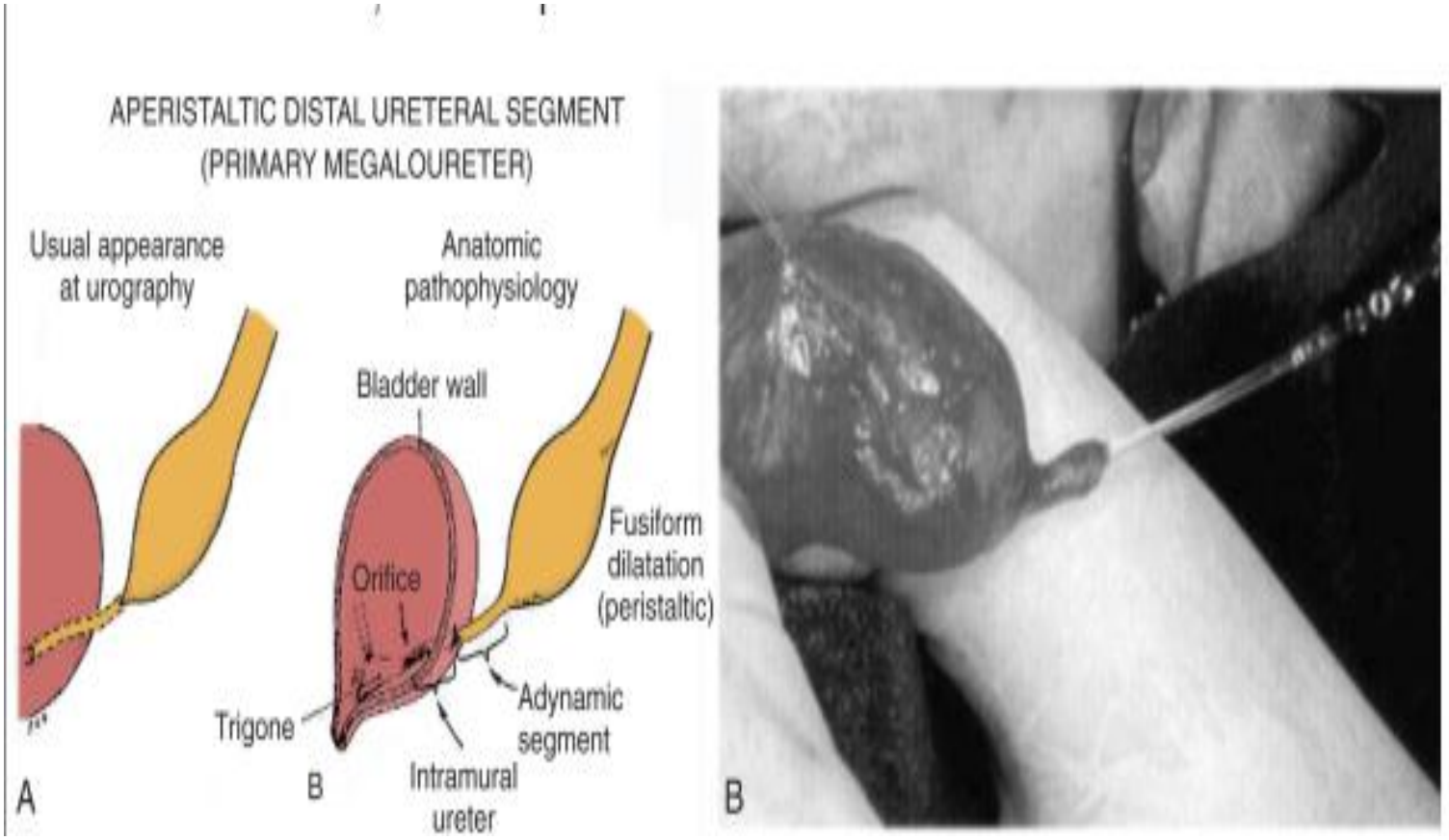
- Indication of surgery:
- Symptomatic pt.
- Incidental finding in neonates:

>worsening hydronephrosis pattern

>Reduced differential renal function

- Incidental finding in children with significant obstruction.

2nd DDx : Ureterovesical junction obstruction



- There is narrow segment cause dilation of whole system involve, Maybe unilateral or bilateral but it's usually Unilateral
- other name is : Megaureter ; Severely dilated Ureter
- What's different b/w Ureterovesical junction obstruction and PUJO ? in Ureterovesical junction obstruction there is dilation of ureter **imp!**

427 notes

- Male : female ratio 3:1
- Left : right ratio 3:1

Types:

- Obstructive not refluxing
- Obstructive refluxing
- Refluxing non obstructive
- Non refluxing non obstructive (adynamic ureter)

Treatment :

- Obstruction: excision and reimplanting of the UVJ
- Reflux: according to the same line of reflux management

3rd DDx: Duplication Anomalies

Duplication Anomalies:

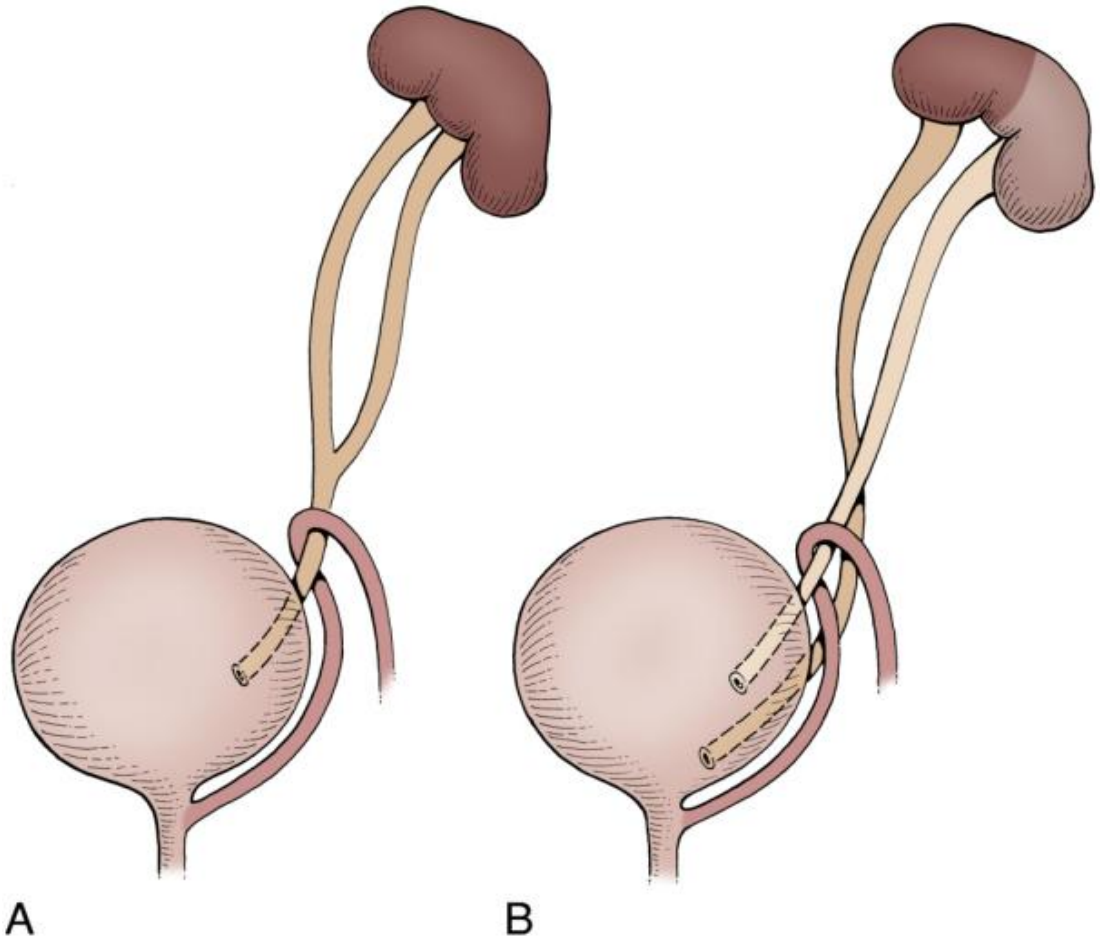
A. Renal and ureteric duplication
B. Complete duplication from upper pole of kidney to lower pole of bladder and vise versa (Weiger-Meyer law)
C. Ureterocele
D. Ectopic ureter

A. Renal and ureteric duplication

- Incidence is 1%, 1.6:1 F:M, 85% unilateral.
- Either two urethral buds meeting the meta-nephros

  or one ureteric bud that bifurcates.
- Associated with: reflux 43%, renal dilatation 29%, ectopic insertion 3%, infections and ureterocele.
- Duplication per se is of no clinical significance, but

  the associated anomalies may require intervention



Incomplete Duplication

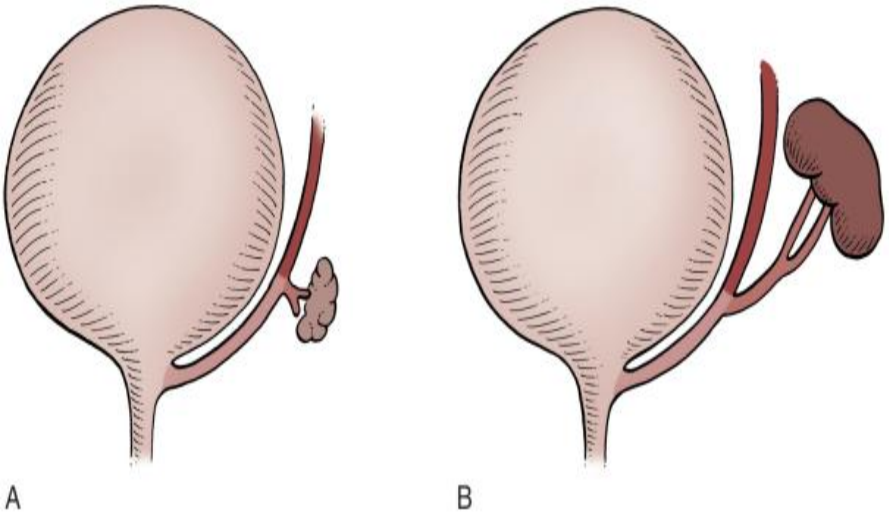
Complete Duplication

-Usually only one ureter comes from each kidney to bladder > single system !

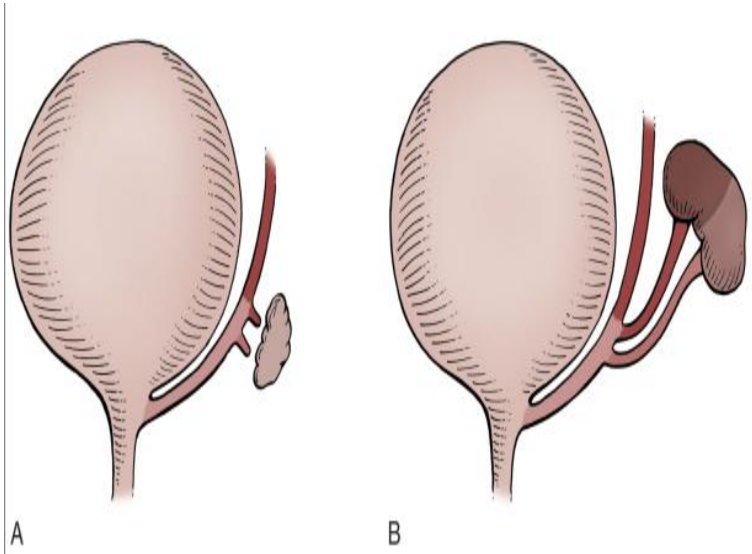
-Duplication Anomalies : 2 ureter coming from each kidney and going to bladder.

-Incomplete Duplication: both ureters meet in their way and one ureter go to bladder.

-Complete Duplication: 2 duplication.



Incomplete ureteral duplication



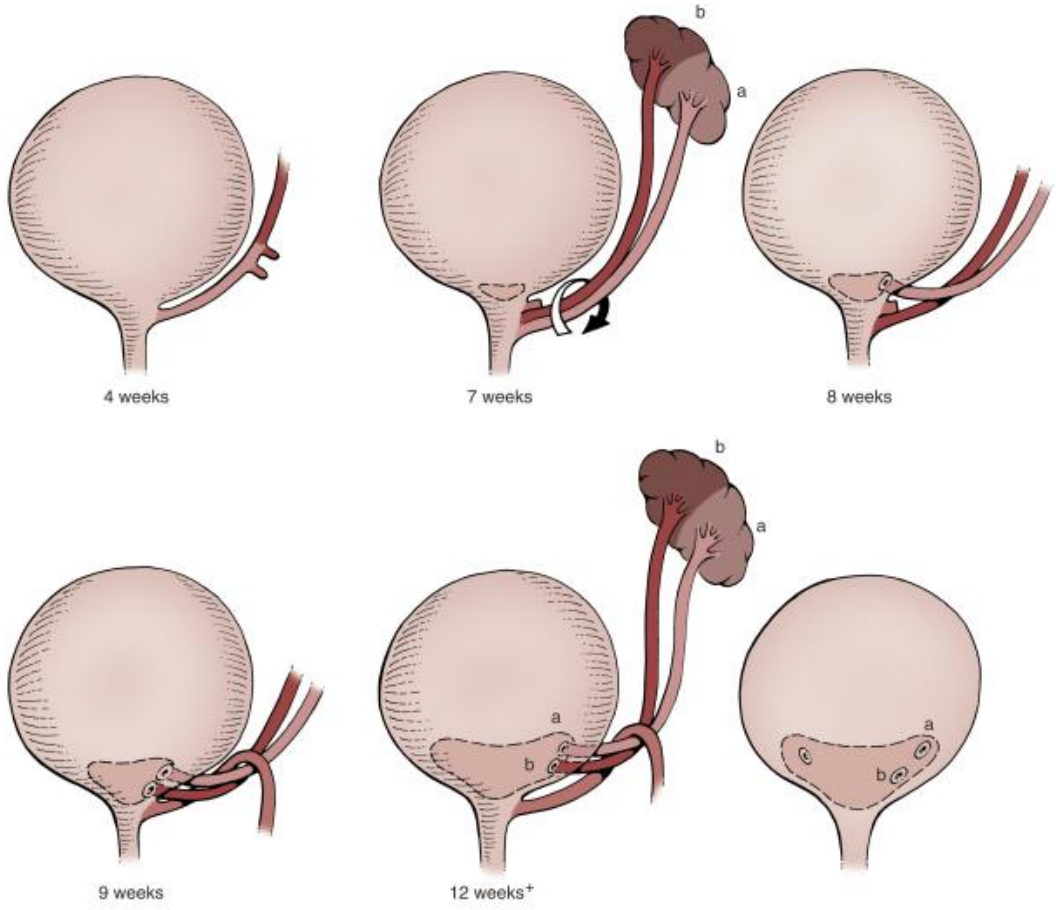
complete ureteral duplication

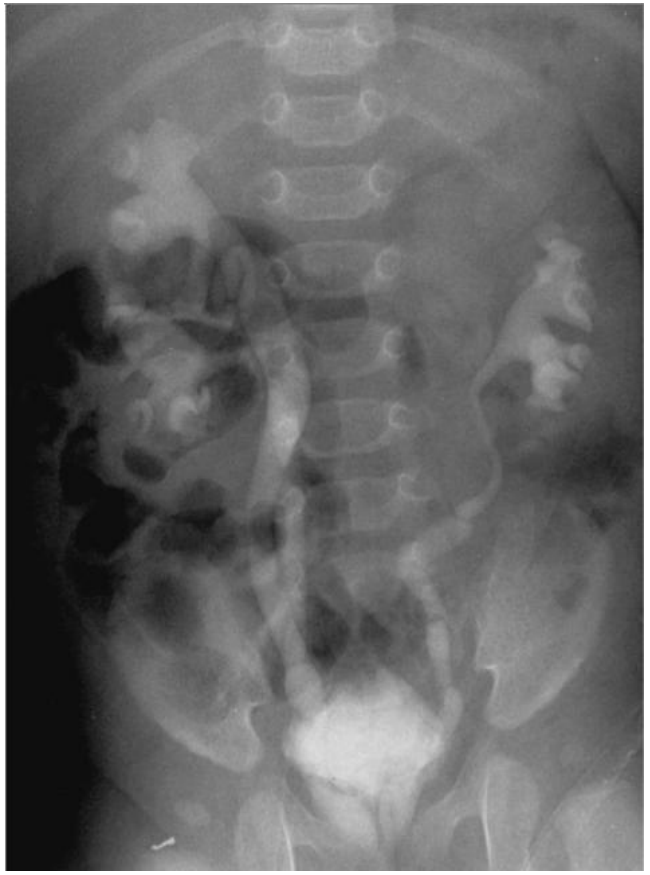
Embryologically :

Normally : One ureteral bed (early precursor of the ureter) meet future kidney.
In Incomplete ureteral duplication: Ureteral bed bifurcate into 2 after the generation >goes to kidney as 2 ureter
In complete ureteral duplication: 2 separate uretral bed (2 future ureter) and come to meet metanephic kidney (future kidney)

If the both ureters coming to kidney and no reflex or obstruction > no harm to kidney but if there is obstruction as in uretrocele or ectopic ureter or reflex > harm to kidney !

B) Weiger - Meyer Law





The upper pole ureter (which drain the upper pole of the kidney) come to lower part of bladder and lower pole ureter coming to upper part of bladder > no need to know explanation

كل جزء من الكلية (العلوي والسفلي) له حوض خاص به لذلك يمكن القول انها كليتين في كليه – لذلك اذا حدث سدد في احد الحالبين فان الجزء من الكليه الذي يصب منه هذا الحالب هو وحده سيتمدد ولكن الجز الاخر سيظل سليم لان الحالب فيه سليم

C) Ureterocele : cystic dilatation of the distal part of the ureter

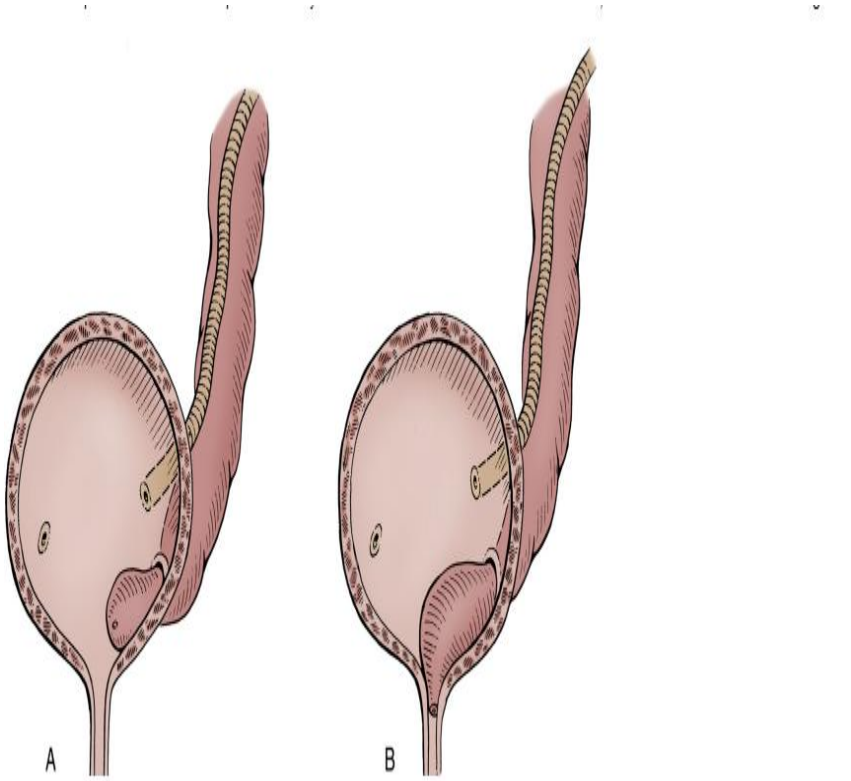
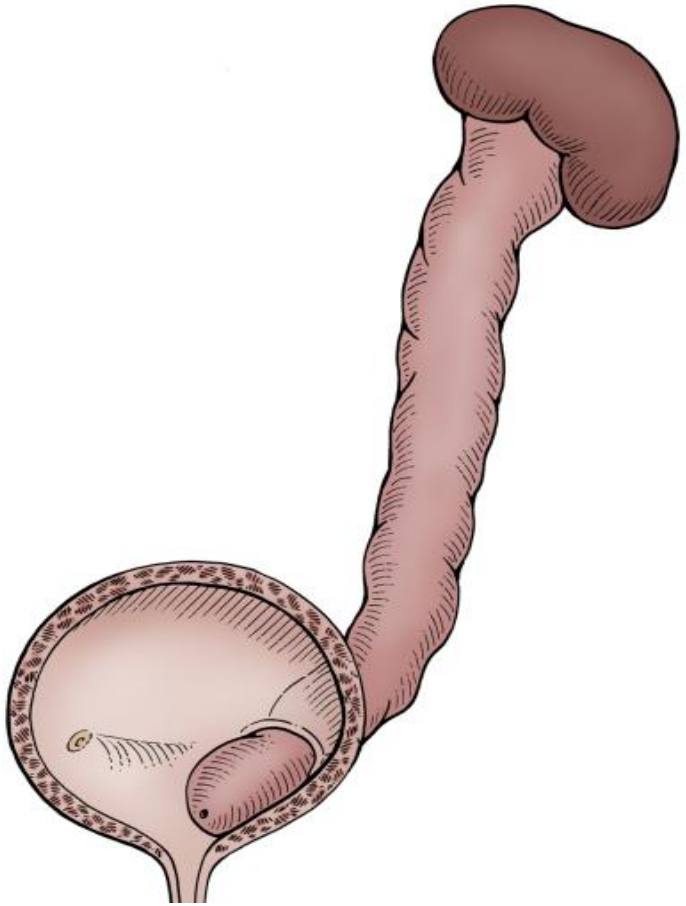


Figure 116-5 A, An intravesical ureterocele located entirely within the bladder. B, The distal portion of an ectopic ureterocele extends outside the bladder and into the urethra.

Ureterocele is associated with Duplication Anomalies



Single system ureterocele

Notes !!

- Ureterocele >cystic dilatation of the distal part of ureter causing obstruction which cause whole ureter is dilated.
- Pt will come with hydrouretronephrosis !
- This other example of Ureterovesical junction obstruction But the difference b/w them is the present of is Cyst in bladder in Ureterocele !

- Sacculation of the terminal portion of the ureter.

Has 2 types :

- 1- Orthotopic = intravesical (inside bladder)=simple=adult type ureterocele.
- 2- Ectopic (start in bladder and extended out side of bladder) = extravesical=duplex system= infant type ureterocele.
- In ectopic ureterocele it involve the upper pole system.
- 7:1 F:M, 10% bilateral, ectopic: orthotopic 4:1
- Commonest cause of urine retention in female infants.

- Presentation:
  - Usually detected Antenatal (U/S) → we use MCUG (micturating cystourethrogram; which is radiological study) to confirm the diagnosis imp!
  - Or pt presented with:
  - Urine retention
  - Infection
  - Calculus formation > pt have infection with stagnation of urine will form stones .
  - interlabial mass in female: one of the DDx for intralabial mass in female is uretrocele!
    - if there is ureterocele and there is vaginatation through urethra.

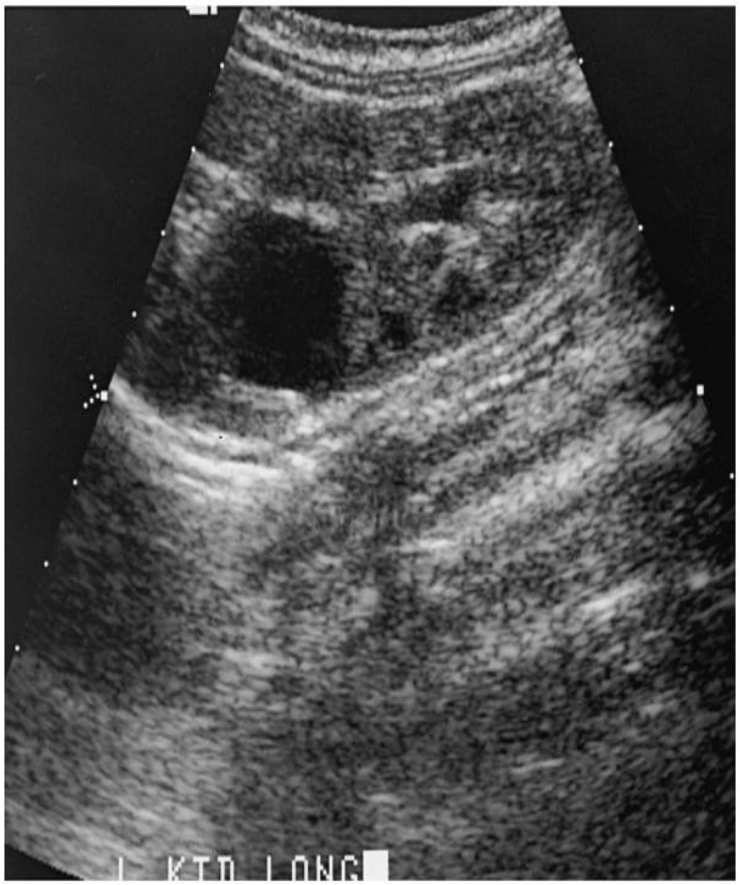
427 notes

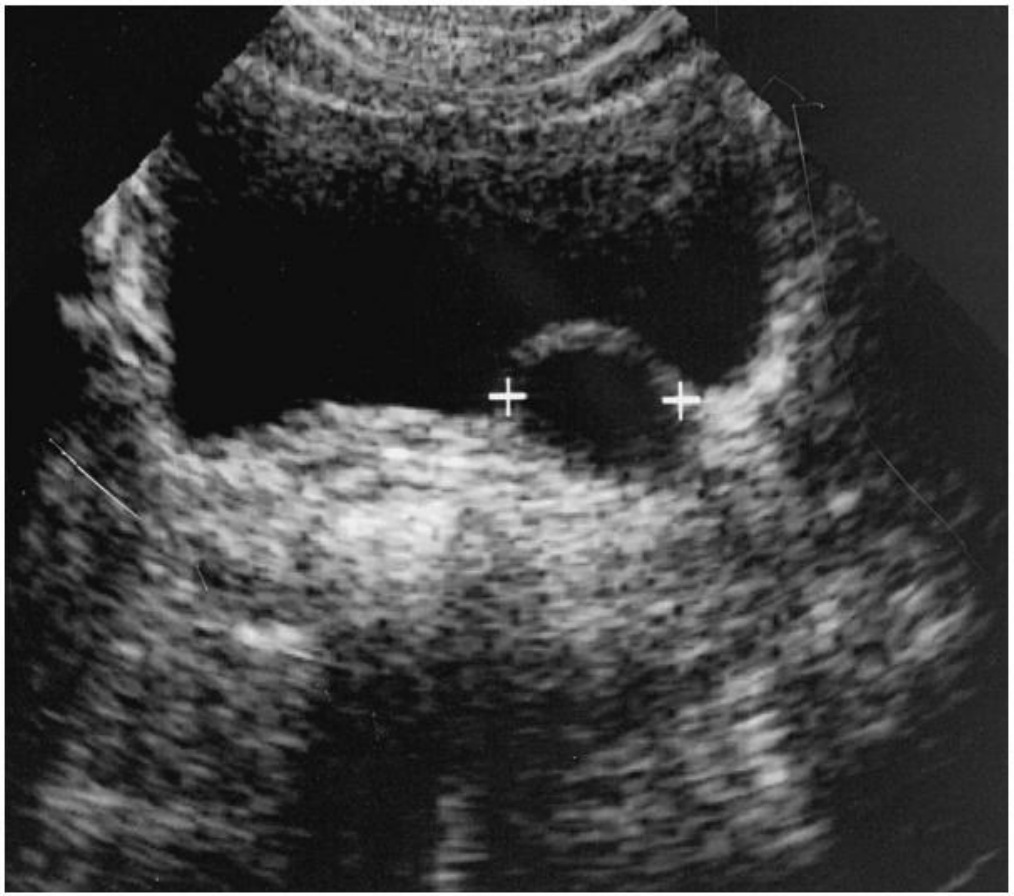
Investigations:

- Antenatal U/S
- IVP
- VCUG
- Renal scan

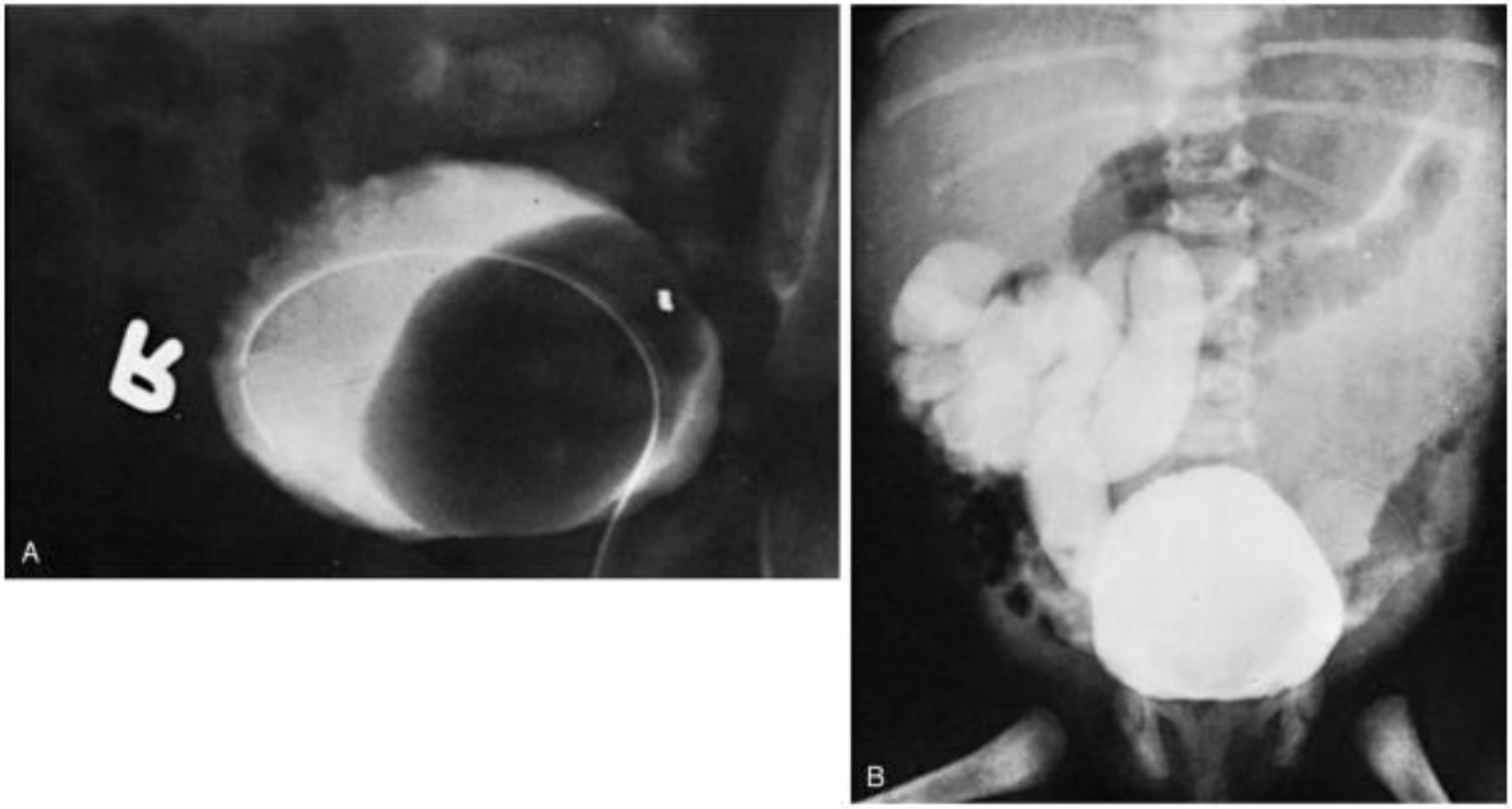
Management:

- Needs urgent intervention
- Incision of ureterocele
- Upper pole heminephroectomy
- Excision of ureterocele and common sheath reimplant.





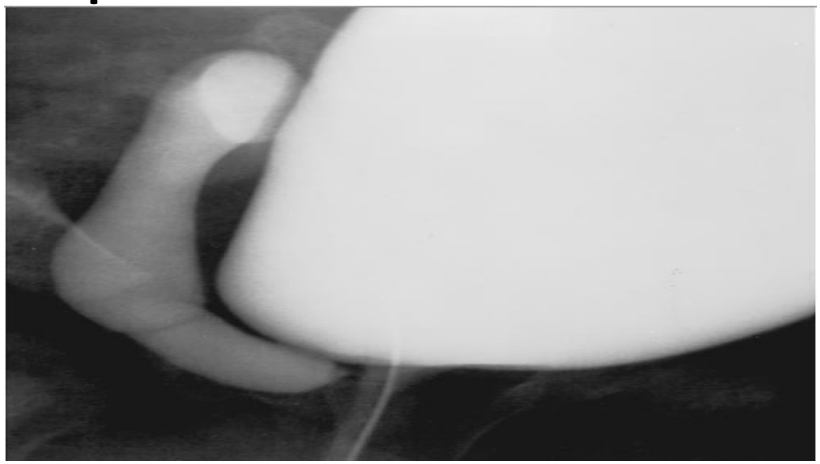
Cystic structure in bladder which is cystic dilation of distal part of ureter



MCUG, put catheter in bladder and use contrast if there is no abnormality whole bladder will be white.
But in ureterocele we will see filling defect imp!

D) Ectopic ureter

- ureter come outside the bladder.
- Simple ectopia any other way in bladder
  - or ureter completely outside bladder(ectopic ureter)
- Most commonly associated with duplex system and with ureterocele.
- Clinical picture depend on: associated anomalies, site and sex of the patient.



427 notes

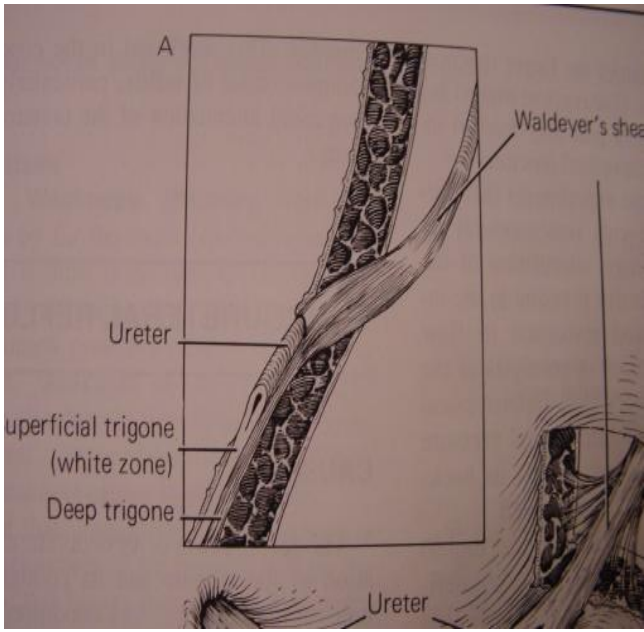
- Investigations include IVP, VCUG, cystoscopy
- Renal scan asses the function of both renal poles in case of duplication.

4th DDx : Vesicoureteric Reflux (VUR)

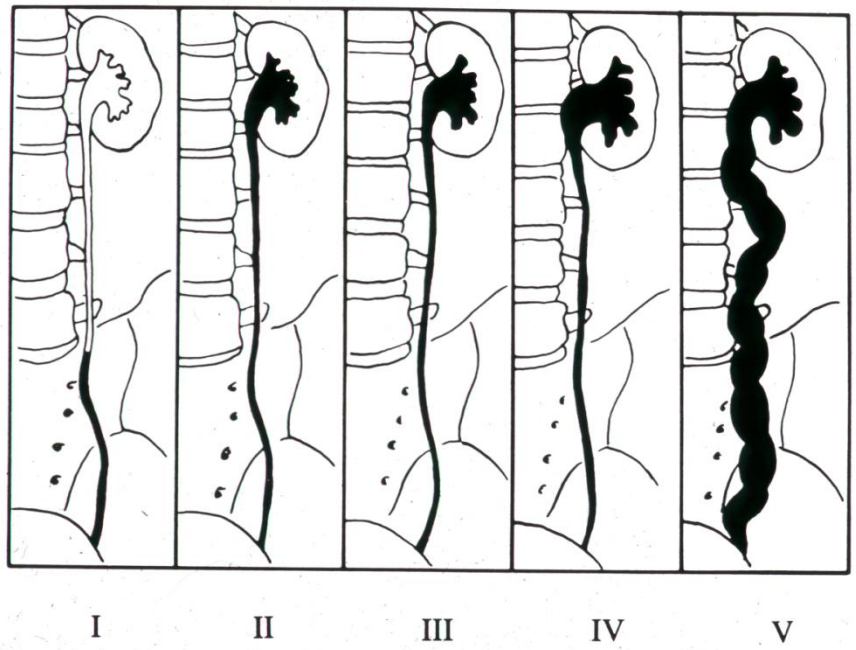
Normal anti-reflux mechanism

"Flap valve" imp!

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.

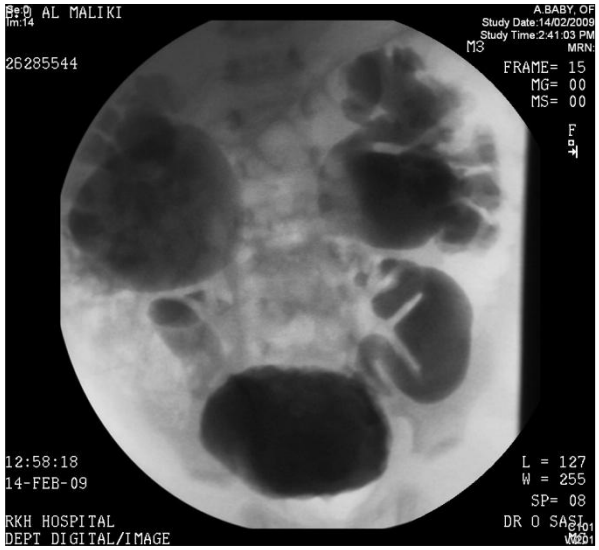


Divided into 5 grads :



1: confined to ureter ; contrast in distal part of ureter!
2: reaches the kidney so the contrast in whole ureter and renal pelvis without any dilation
3: mild ,dilation of renal pelvis and ureter without loss of calyces.
4: moderate , same as 3 but there is loss of calyces.
5: sever , tortuous dilated ureter !

Study to role out reflex is MCUG imp!
If there is no reflux , contrast only in bladder but if there is reflux it will be one of these grades in pic.



Notes !

- Management :

It imp in pt with VUR To keep him in prophylactic antibiotic

Prophylactic dose: (1/3 of therapeutic dose only at night (24h), long term)

427 notes

Resolution of reflux:

87% of Grade 1
63% of Grade 2
53% of Grade 3
33% of Grade 4

Over 3 years period of follow up

Management:

The decision depends on :

- The chance of spontaneous resolution (age and grade at presentation)
- Breakthrough infection
- Renal scarring and renal function
- Compliance with medication

Medical management:

- **Patients with UTI (the most common presentation) and VUR is suspected.**

>continue on prophylactic antibiotics after treatment till the VCUG is done.

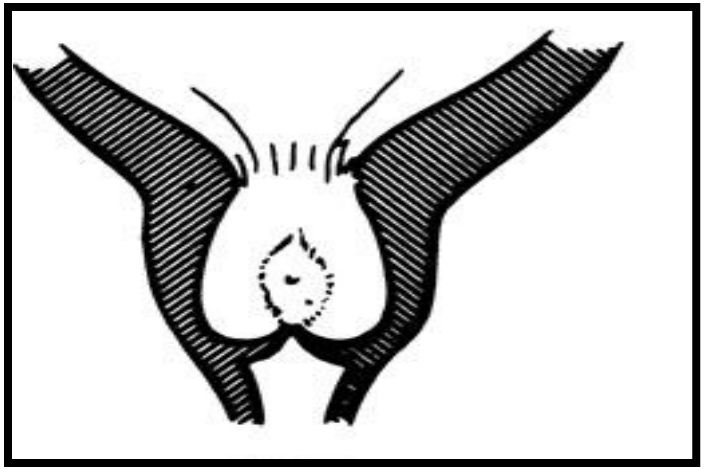
- **Patients for conservative management :**

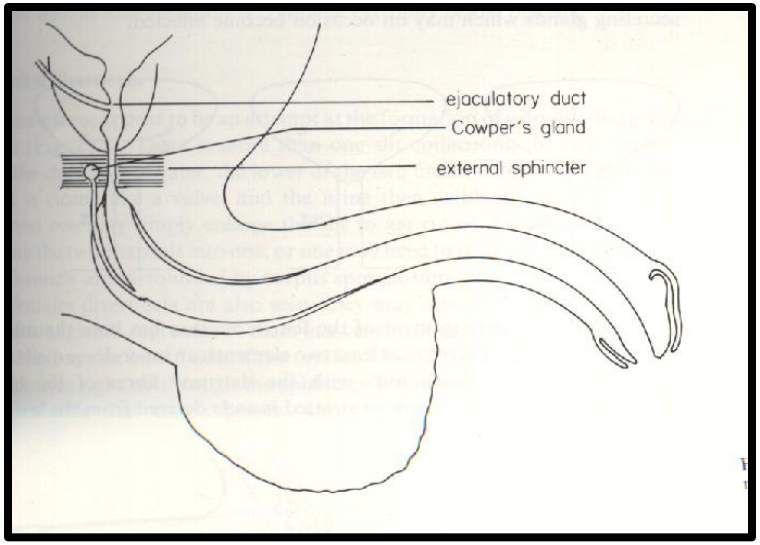
> Continue meticulously on prophylactic antibiotics and surveillance with urine culture and sensitivity, U/S ,and DMSA (dimercaptosuccinic acid) scan

5th DDx: Posterior urethral valves (PUV)

- Incomplete canalization of the posterior urethra
- 1:5000 male infants.
- most common cause of urine retention in male infants.
- 50% have renal impairment.
- The bladder and the kidneys developed under

  high pressure and resistance.
- The more proximal the valve the more sever the condition





Notes!

- Posterior urethra : from bladder neck to membranous urethra.
- anterior urethra :Distal to membranous urethra bulbar, penile urethra .
- b/w anterior & posterior urethra during embryologically there is canalization from distal to proximal and from caudal to cephalic leading to complete tube without any narrowing , But in Posterior urethral valve incomplete canalization of urethra and leave small membrane (posterior urethral valve) which cause obstruction.
- So there is obstruction >bilateral hydroureteronephrosis imp!, the bladder is urogenic imp! b/c during pregnancy the detrusor muscle replaced by collagen so no contraction of muscle .
- Baby start voiding during 24th week of gestation > so in Posterior urethral valve the baby will void against pressure so bladder will be large and trabeculated and urogenic .

- Note : the obstruction is not complete (narrowing or stenosis) b/c if it complete “severe” the pt will die in utero!

Associated findings

1. Oligohydramnios imp! (low amount of Amniotic fluid)

*No output of urine or little > Amniotic fluid -low in U/s b/c there is no secretion but there is absorption .

*Obstruction of esophagus no absorption > Polyhydramnios.

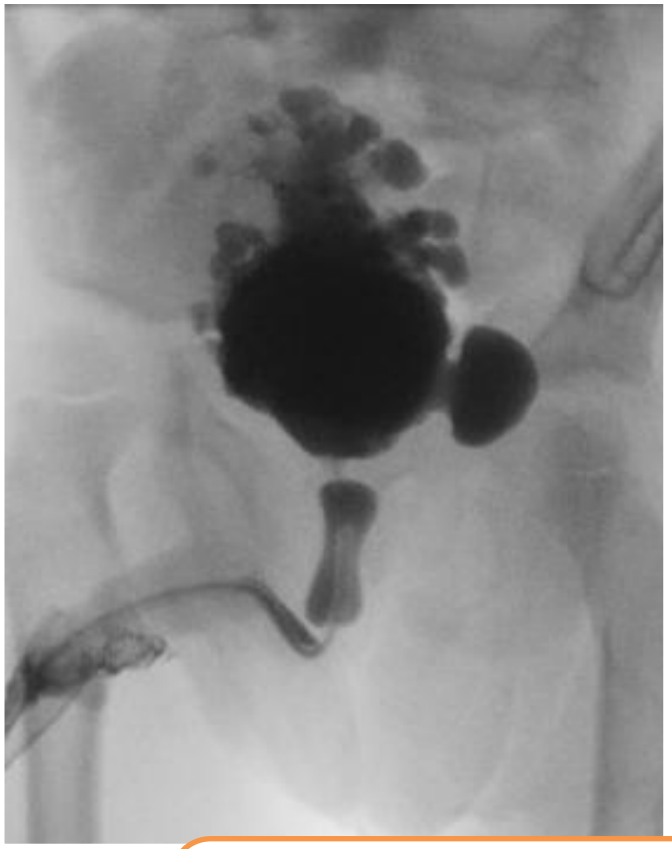
2. Bilateral renal dilatation imp!
3. VUR: 40%
4. Valve bladder > loss of it's Fx and become abnormal bladder
5. Renal impairment in 30-50% , 25% of them will have renal transplantation in future > so very serious and we have to interfere immediately postnatal.

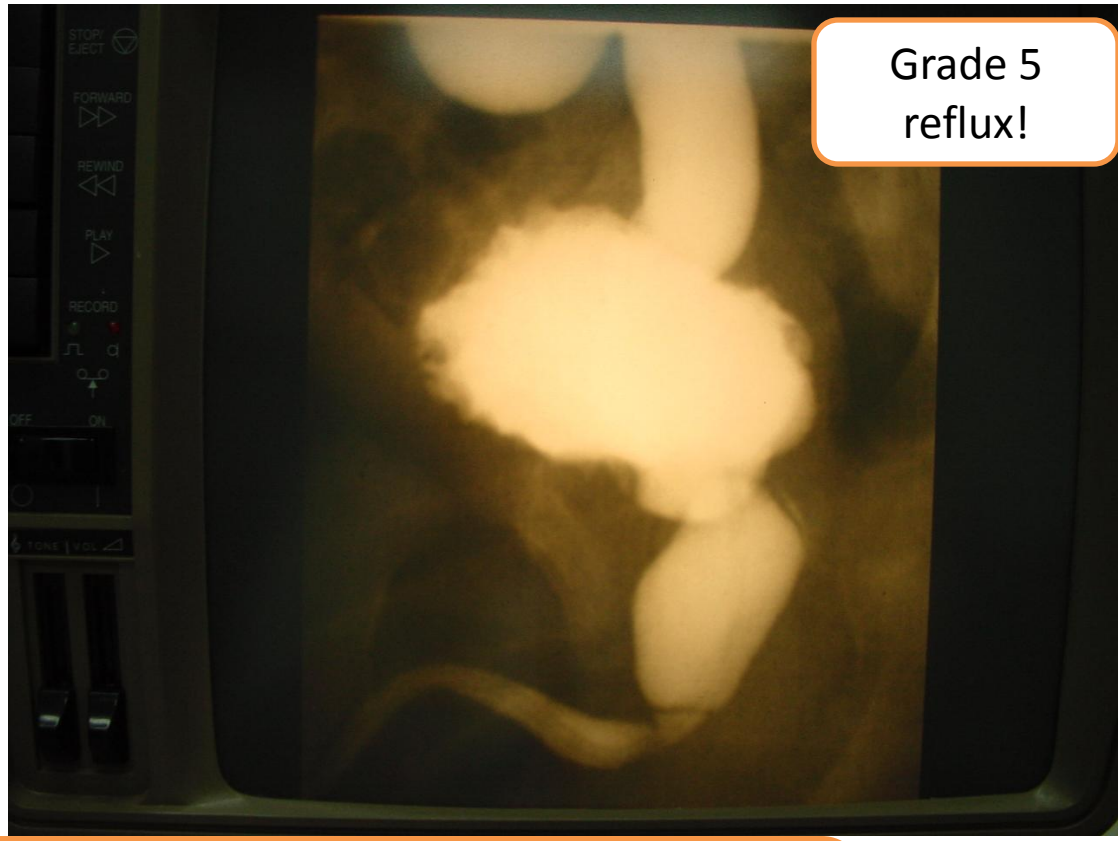
Presentation:

1. Antenatal (hydonephrosis)
2. Urine retention
3. UTI
4. Poor urinary stream
5. CRF; at late stage

Investigations:

Antenatal US , US , VCUG , Renal scan , Renal function studies, Urodynamic study





Grade 5 reflux!

MCUG:

Posterior uethera dilated, normal anterior urthera ,bladder trabeculated and elongated (christmas tree bladder)

Kidney

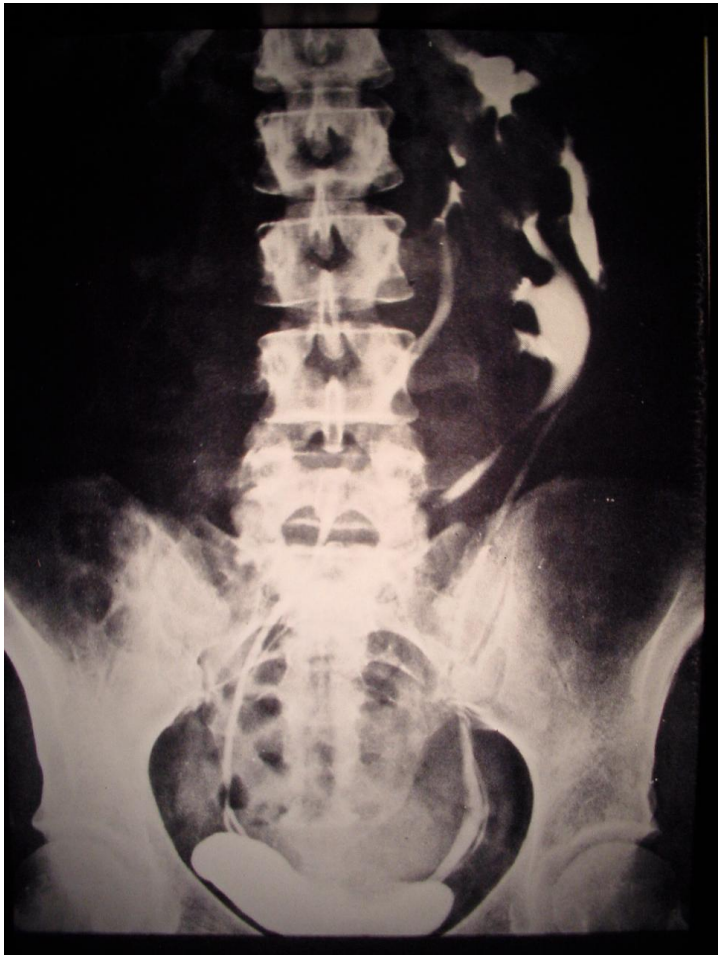
Anomaly of **position**, number and rotation:

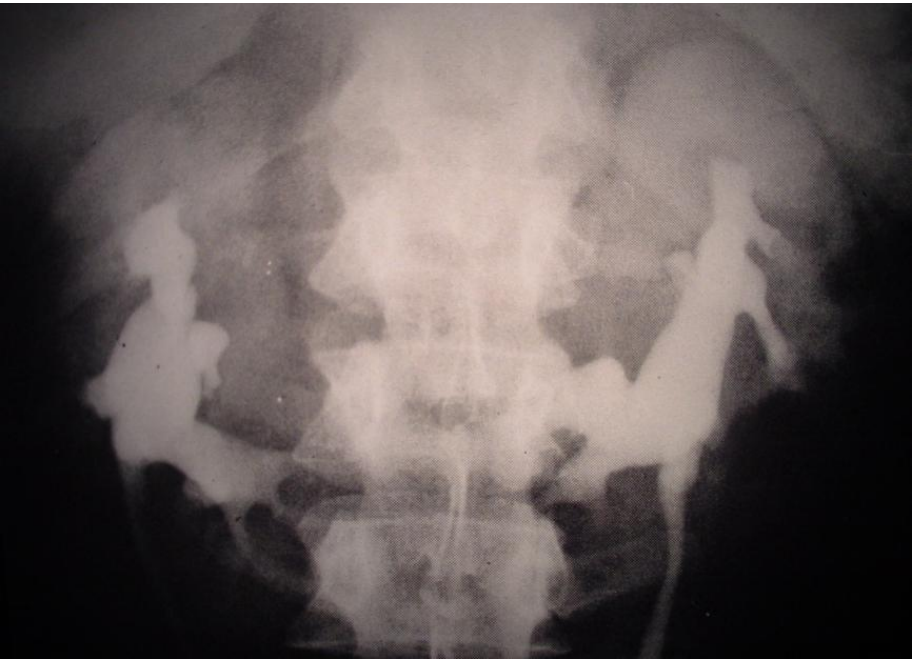
1. Simple ectopia:
   - A kidney that is outside the renal fossa.
   - Pelvic (commonest), lumbar, sacral.
2. Thoracic kidney. kidney in chest
3. Horseshoe kidney (2 kidney fused and connected together : 90% by the lower lobes, 10% upper lobes connected , the connection is either fibrous band or sometimes it's parenchymal tissue)
4. Unilateral renal agenesis.
5. Bilateral renal agenesis.
6. Crossed renal ectopia with no fusion.
7. Crossed renal ectopia with fusion.
8. Malrotated kidney.

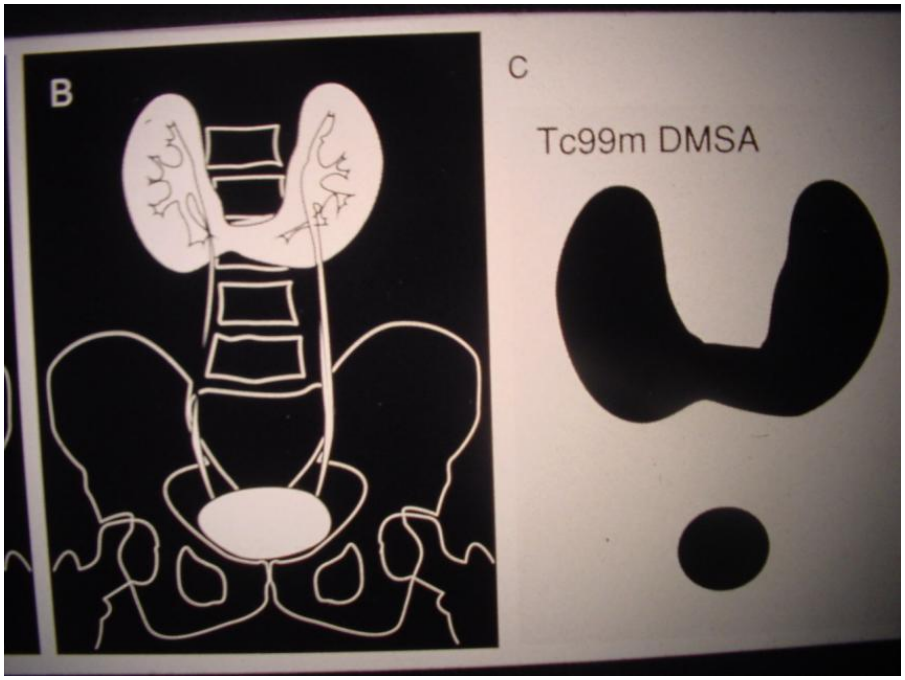
Notes !

- Normal position of kidney : retroperitoneal in flank area. Anywhere except this place > ectopia!
- If it little up or down this called simple ectopia.
- Cross ectopia: kidney goes to other side

Right to left
cross ectopia







Horseshoe kidney

Kidney

Cystic abnormalities:

1. Renal dysplasia هذه الكلمه تعني ان انسجه الكليه تغيرت الى انسجه غير عامله وبالتالي تنتج كليه غير عامله*

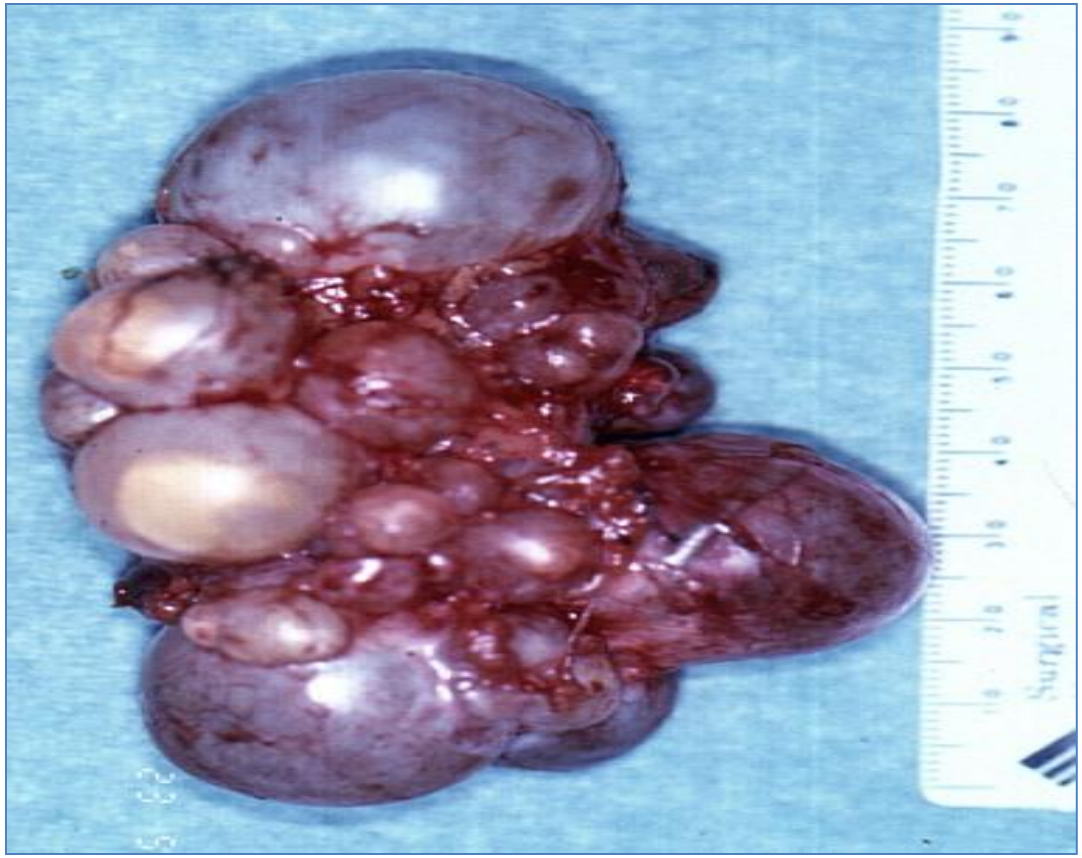
   A) congenital unilateral multicystic kidney.
   B) Segmental and focal renal dysplasia.
   C) Renal dysplasia associated with congenital lower tract obstruction.

2. Congenital polycystic kidney disease: * في هذه الحاله تكون الكليه عامله بشكل سليم

   A) Infantile type
   B) Adult type

3. Simple cyst
4. Calyceal cyst
5. Peripelvic cyst
6. Perinephric cyst

- Just to know the names !



(Multicystic dysplastic kidney)
- One of DDx of ANH
- no fx of kidney
- The whole kidney replaced by cyst so no nephrogenic tissue !
- Multicystic : means multiple cyst , dysplastic : no renal tissue.

But in Polycystic ,
kidney is large usually bilateral
There is no Function of the kidney !

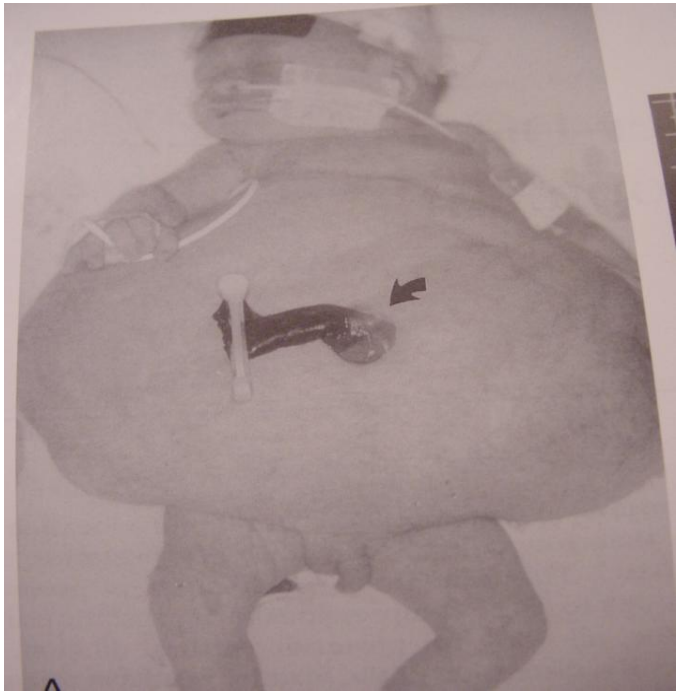
Prune Belly Syndrome (PBS)

- Also called Eagle-Barrett Syndrome, Triad syndrome imp! :

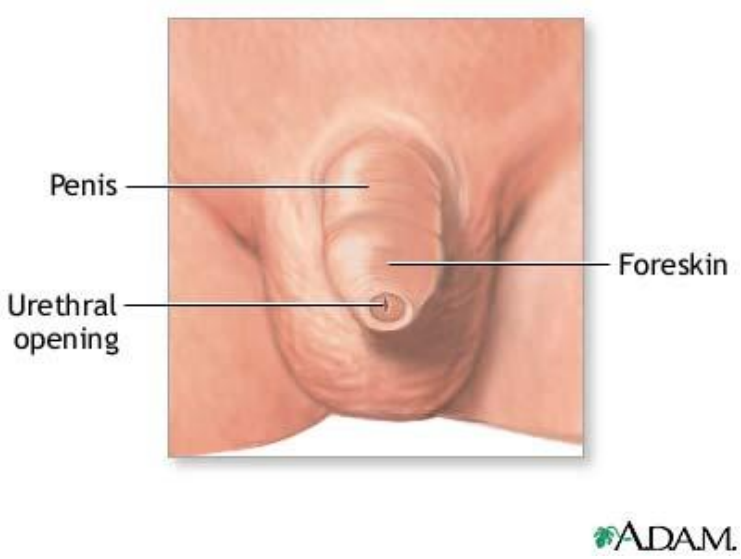
1- Absent abdominal wall muscle(External oblique, Internal oblique, Transverse abdominal muscles), u can feel all the organ and even u can see the bowel movement b/c the muscle either absent or thin layer (hypopalstic).

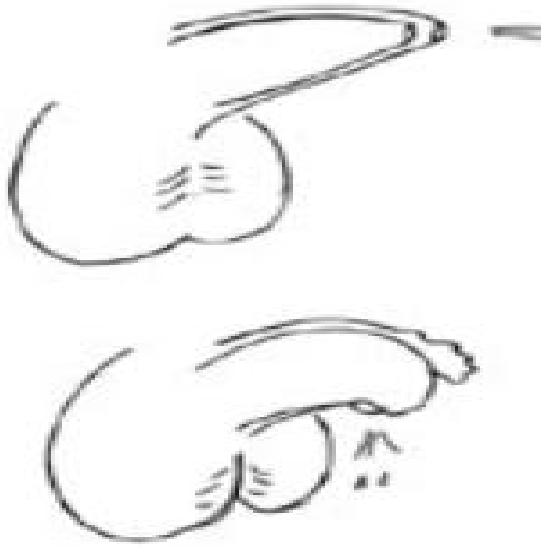
2- Bilateral undescended testis (bilateral intraabdominal testis)

3- obstructive uropathy bilateral hydrouretronephrosis and large bladder !



Hypospadias : abnormally located meatus (which contain the urethral opening)





Meatus open in tip of glans of penis if meatus open anywhere other than that we call it ectopic .

What different b/w Hypospadias ,Epispadias?

Hypospadias: if penis toward the scrotum ventral side

Epispadias :toward abdomen dorsal side

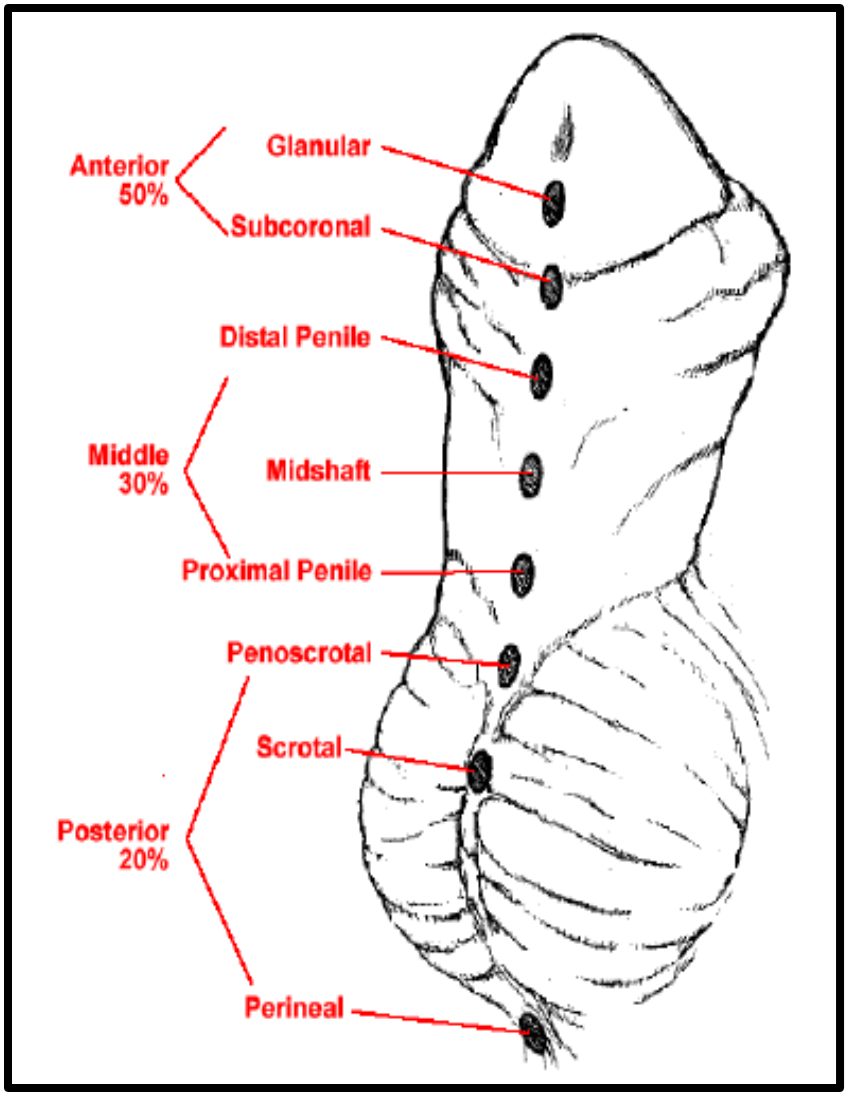
Hypospadias

- Common (2%)
- Abnormal position of the EUM(external urethral meatus):

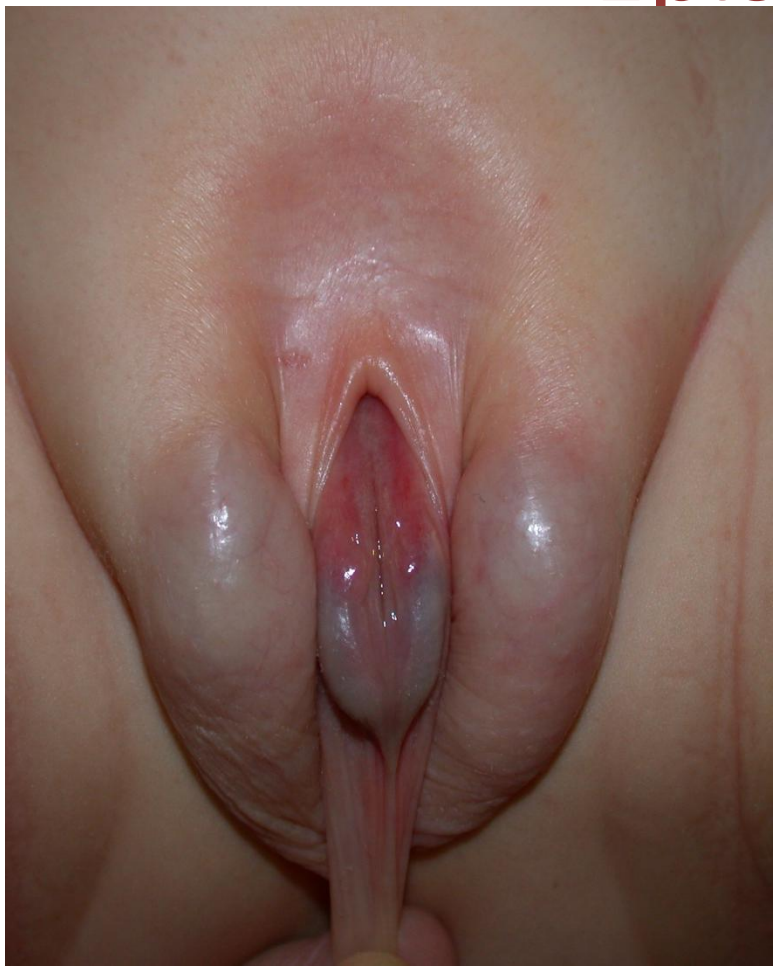
1- Distal hypospadias : (from mid shaft to Glanular)

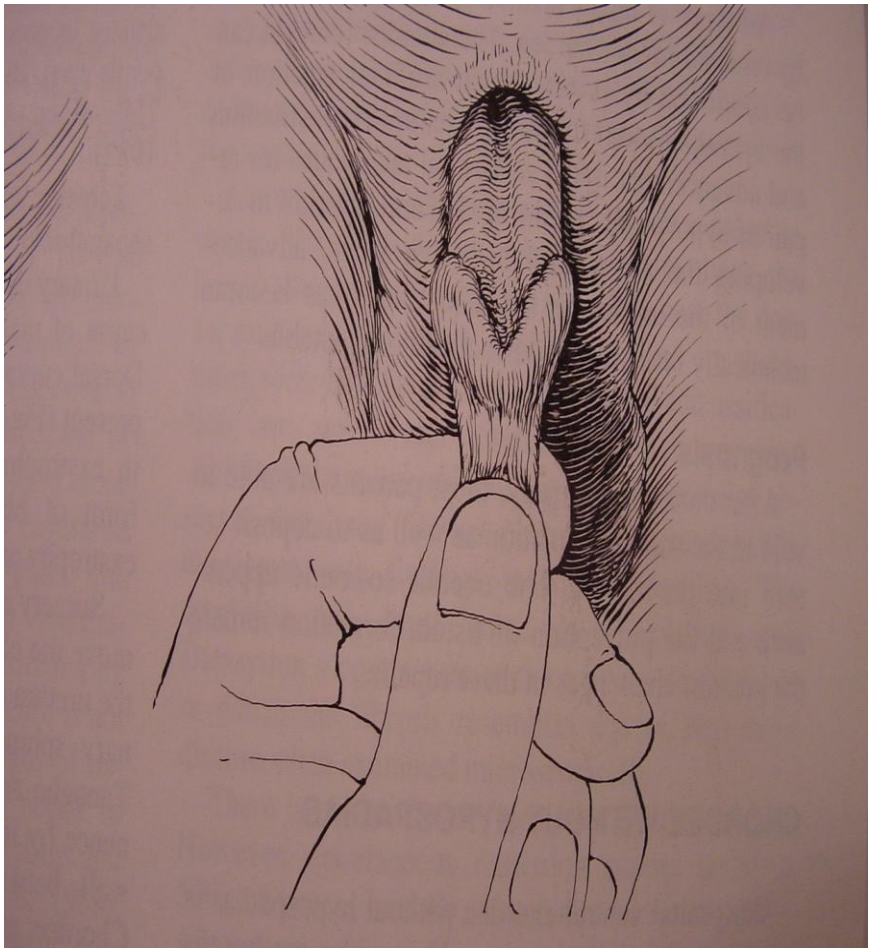
2-Proximal hypospadias (from proximal penile "proximal shaft" to the perineal)

- NO Circumcision imp !! Absolute contraindication " b/c dorsal urethral skin we will need it in repairing especially in proximal hypospadias"
- age to repair :6 to 9 months
- Requires one stage repair


Anterior
50%
Glanular
Subcoronal
Distal Penile
Middle
30%
Midshaft
Proximal Penile
Penoscrotal
Scrotal
Posterior
20%
Perineal

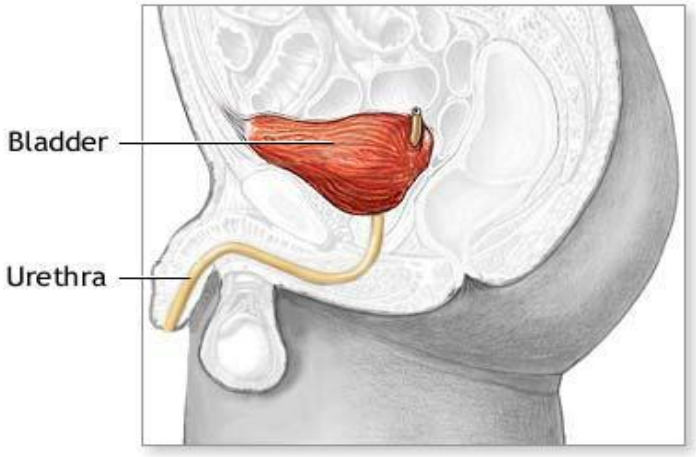
Epispadias

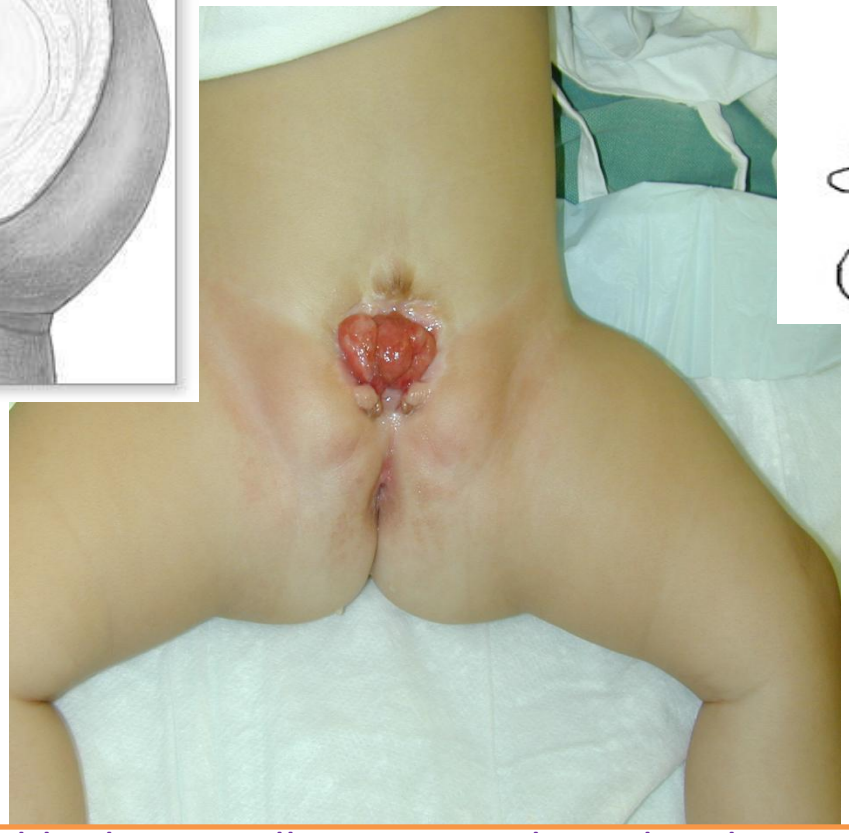


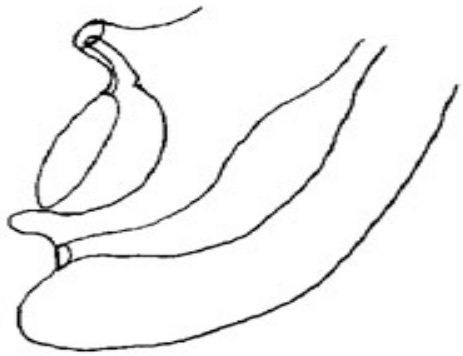


Very rare , Abnormal position of external urethral meatus in dorsal surface of the penis

Bladder Exstrophy







- Bladder has 3 walls : Anterior, lateral and posterior.
- Anterior wall consist of abdominal muscles and skin , in bladder exstrophy the anterior wall is absent (no anterior abdominal wall , no skin) so the lateral wall will be attached to skin to outside. (the bladder is exposed to the outer environment)
- We need to close bladder and to reconstruct abdomen

427 notes

- An exstrophy of bladder
- Rare; 1:30000 live births with a 3:1 male : female ratio
- The results of improper development of anterior abdominal wall, pelvic girdle , and anterior wall of the bladder