432 Surgery Team

15 Pediatric urinary disorders

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COLOR GUIDE: • Females' Notes • Males' Notes • Important • Additional

Objectives

Were not given

INTRODUCTION:

- Most common anomalies of all organ systems.
- 10% of the population have some type of urogenital anomaly
- 14:1000 births have an antenatal diagnosis of urogenital anomaly.
- The antenatal diagnosis is done by ultrasound after 28 weeks of gestation because at 24-28 weeks the urinary system starts to be clear on ultrasound

ANOMALIES OF THE KIDNEY:

1. ANOMALIES OF POSITION, NUMBER AND ROTATION:

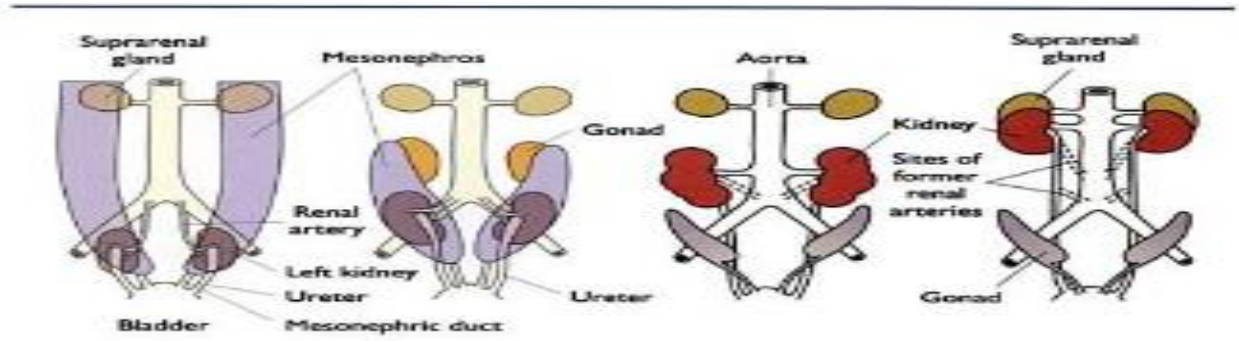
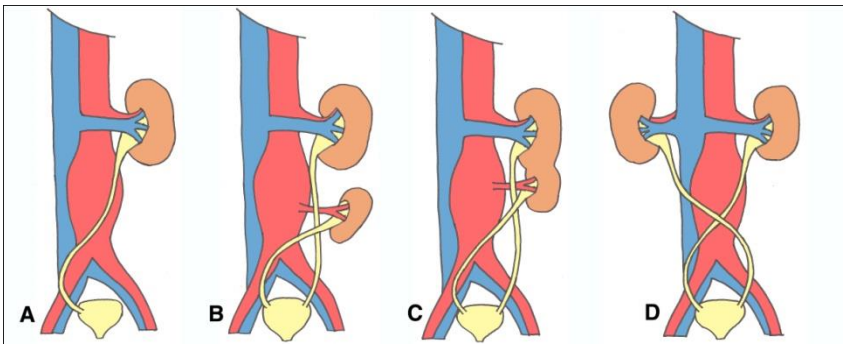


Figure 5.1 Normal ascend and rotation of the kidney. The kidneys develop in the fetal pelvis after the 5th week and ascend into their retroperitoneal position. The kidney also rotates antero-medially. Variations of ureteric and collecting ducts, ascend and arteries are common.

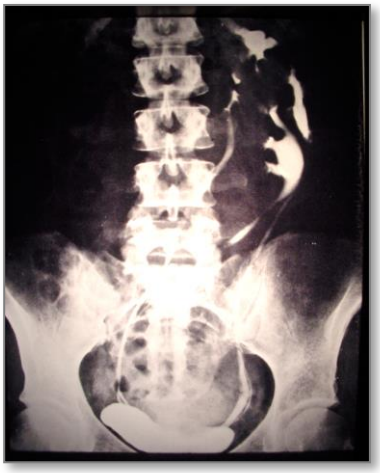
When the kidneys ascend from the pelvis to their permanent location in the upper lumbar region, they come into apposition with the adrenal glands, which develop in situ. During ascent, the kidneys rotate medially so that the hilum, which initially faced anteriorly, (the pelvis is posterior and the calyces are anterior), now faces medially, (the pelvis is medial and the calyces are lateral). The segmental vessels supplying the kidney are added cranially and lost caudally during ascent.

Normal position of the kidney: retroperitoneal in flank area. Anywhere except this place is called ectopia

1. Simple Ectopia:
   - A kidney that is outside the renal fossa. but in the same side of the ureter.
   - Pelvic (commonest), lumbar, sacral.
2. Crossed renal Ectopia with no fusion. Cross ectopia = kidney goes to other side but the ureter stays in the same side.
3. Crossed renal ectopia with fusion.



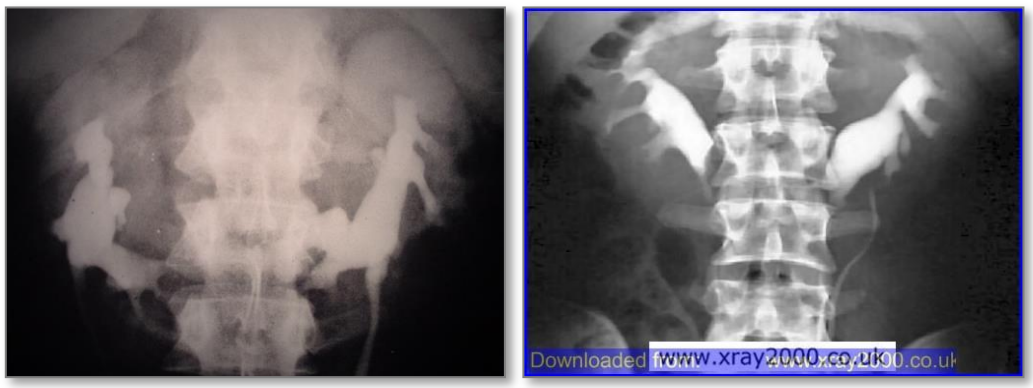
Crossed renal ectopia. **A**, Solitary crossed kidney**. B**, Crossed kidney without or **C**. with fusion**. D.** Bilaterally crossed kidney



Right to left ectopia(cross)

4. Thoracic kidney:kidney in thoracic cavity.
5. Horseshoe kidney = 2 kidneys fused and connected together

- The connection is either fibrous band or Sometimes its parenchymal tissue. (the area where they connect is called the isthmus)
- 90% by the lower lobes, 10% upper lobes connected.



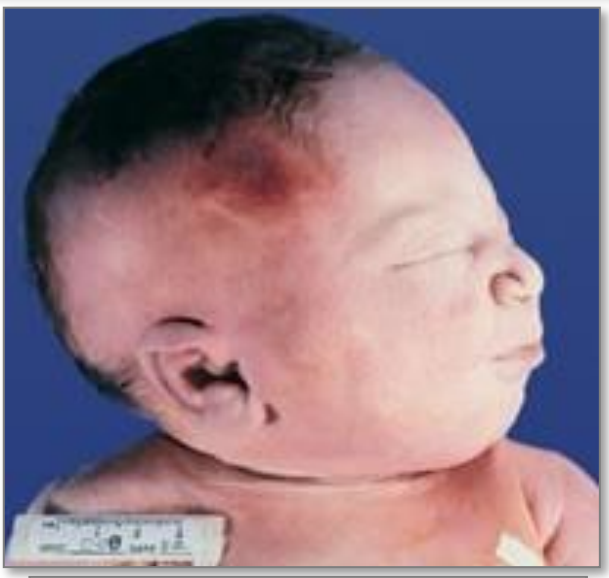
Horseshoe kidney

6. Mal-rotated kidney.(any position other than the calyces laterally and the pelvis medially is diagnostic)
7. A supernumerary kidney(Have its own collecting system)
8. Unilateral renal agenesis.
9. Bilateral renal agenesis. (The fetus will die in utero or in the 1st 24 hrs. because during development most of the amniotic fluid is formed from urine, if absent, there will be no amniotic fluid which will cause compression on the whole infant "flat face, nose, and ears". There is also a compression on lungs, so the infant will die 2ry to pulmonary insufficiency in the 1st 24 hrs.)

Note(s):

- Remember that abnormalities of renal ascend, rotation and vasculature mal-development often co-exist.
- Hourseshoe kidney and simple ectopia are Usually undetected (asymptomatic), diagnosis is incidentally: common symptoms is due to obstruction, hydronephrosis or reflux.

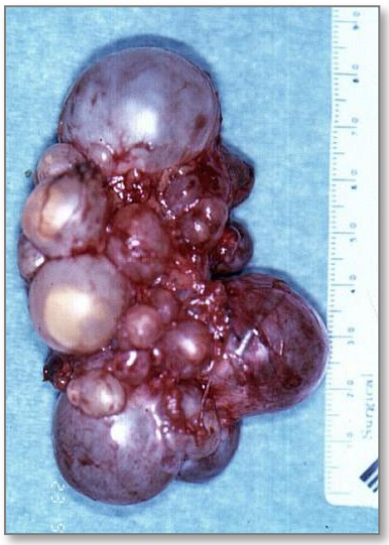
*- A hallmark of renal agenesis, hypoplasia, or dysfunction in utero is **oligohydramnios** (low amniotic fluid volume) since the amniotic fluid is produced by the kidneys. Reduced amniotic fluid volume causes increased pressure on the developing fetus, resulting in a **sloped forehead**, **"parrot beak" nose**, **shortened fingers**, and **hypoplasia of internal organs**, particularly the **gut** and **lungs**. Collectively, this sequence of anomalies is known as the **Potter sequence.***



Sloped forehead, "parrot beak" nose

2. CYSTIC ABNORMALITIES:

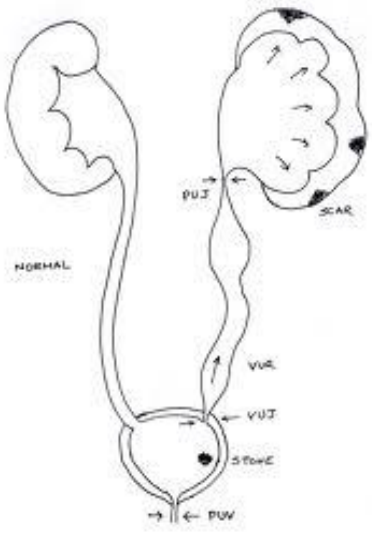
1- Renal dysplasia
   - ✓ Congenital unilateral multicystic kidney.
   - ✓ Segmental and focal renal dysplasia.
2- Renal dysplasia associated with congenital lower tract obstruction .Congenital polycystic kidney disease:
   - ✓ Infantile type
   - ✓ Adult type
3- Simple cyst
4- Calyceal cyst
5- Peripelvic cyst
6- Perinephric cyst



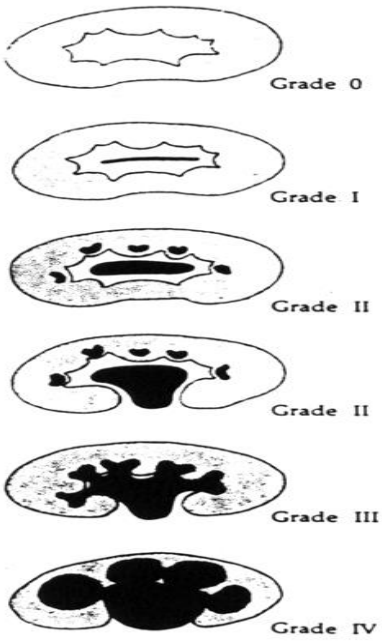
Multicystic dysplastic kidney:

- One of the DDx of ANH
- Kidney is non functional
- The whole kidney replaced by cysts so no nephrogenic tissues.
- Multicystic: means multiple cysts
- Dysplastic: no renal tissue.
- Polycystic: kidney is large usually bilateral

Antenatal Hydronephrosis(ANH):



- This is hydronephrosis detected during pregnancy by ultrasound.
- It is a condition which has a differential diagnosis and causes. It is not a diagnosis on its own!
- **Causes**:
  - ✓ Pelviureteric junction obstruction(PUJ) (41%).
  - ✓ Ureterovesical junction obstruction (UVJ)(23%)
  - ✓ Vesicoureteric reflux (7%)
  - ✓ Duplication anomalies (13%)
  - ✓ Posterior urethral valves (10 %)
  - ✓ MCDK (Multicystic dysplastic kidney)
  - ✓ Others (6%)
- **Grades of ANH:**



Specific details of grading are not important but you should know:

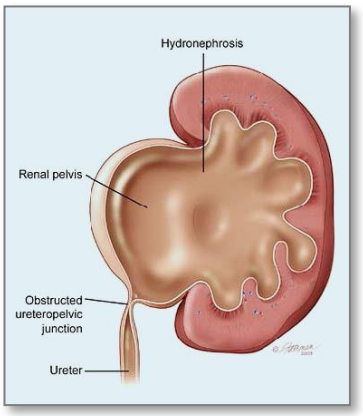
- ✓ Urine appears <u>black</u> on ultrasound
- ✓ From I-IV: 1 is the simplest and 4 is the worst
- ✓ Grading depends on <u>level of dilatation</u> by ultrasound

- So if you see hydronephrosis in an antenatal ultrasound the
  - ✓ Diagnosis is: antenatal hydronephrosis.
  - ✓ the differential diagnosis is: (DDx Below)

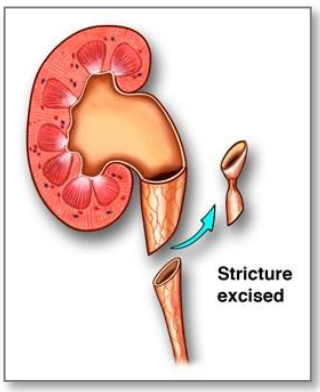
Note(s):DDx

*- **Hydronephrosis** is dilation of the pelvicalyceal system.*
*- **Hydroureter**: dilatation of the ureter*
*-**mhydroureteronephrosis** : Both renal pelvis and ureter*

1. PELVIURETERIC JUNCTION OBSTRUCTION (PUJO)



- Dilation of the renal pelvis due to obstruction at the junction between the pelvis and ureter. Because the obstruction is before the Ureter in these cases is usually not affected. So on Ultrasound the renal pelvis is dilated and the ureter is normal "isolated hydronephrosis"
- It is the most common cause of ANH
- **Etiology**: (theories): Segmental muscular attenuation, Angulation, True. Stenosis, Extrinsic compression, Crossing vessels; 20-30%.
- **Associated findings:**
  - ✓ Reflux in 5-10%
  - ✓ Contralateral PUJO in 10%
  - ✓ Contralateral agenesis in 5%
- **Presentation**:
  - ✓ More in males and occurs in the left side more than the right.
  - ✓ Incidental in neonates by US or in infancy
  - ✓ If the diagnosis is missed during pregnancy or early infancy the child could come with symptoms like: UTI, pain, mass, hematuria, and stones.
- **Investigations include**: ultrasound, renal scans and VCUG (voiding Cystourethrography)
- **Indications of surgery**: surgical modality of treatment is Pyeloplasty (to cut the narrow area and anastomose the normal ureter to the renal pelvis).
  - ✓ Symptomatic patients
  - ✓ If the finding is incidental:
  - ✓ Neonates: worsening pattern or reduced renal function
  - ✓ Children: significant obstruction

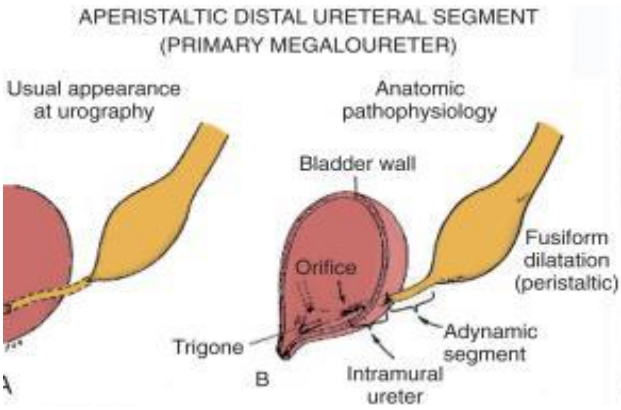


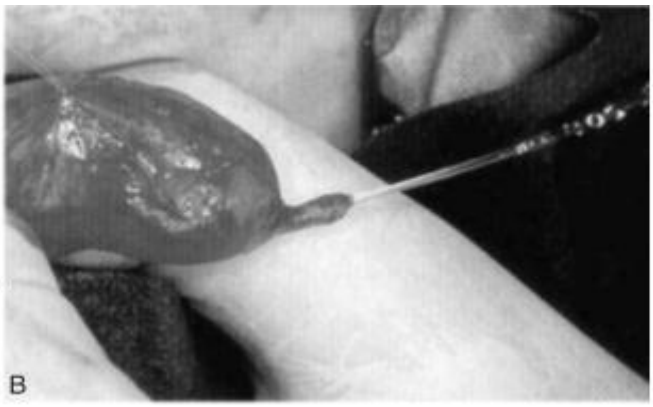
Pyeloplasty

Note(s):

*- **voiding cystourethrogram (VCUG),** also micturating cystourethrogram (MCUG), is a technique for visualizing a person's urethra and urinary bladder while the person urinates (voids). The technique consists of catheterizing the person in order to fill the bladder with a radiocontrast agent, typically cystografin. Under fluoroscopy (real time x-rays) the radiologist watches the contrast enter the bladder and looks at the anatomy of the patient. If the contrast moves into the ureters and back into the kidneys, the radiologist makes the diagnosis of vesicoureteral reflux.*

2. URETEROVESICAL JUNCTION OBSTRUCTION



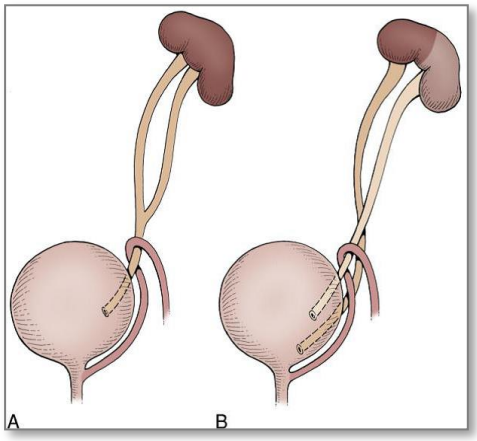


- Also called Mega-ureter or severely dilated ureter.
- There is a narrow segment that causes the dilatation of the whole renal system
- Could be bilateral or unilateral but it is mostly unilateral
- It is different from PUJO in that the ureterovesical junction obstruction has a dilated ureter (in US, hydrouretronephrosis).
- **Presentation:**
  - ✓ Male 3:1 female
  - ✓ Left 3:1 Right
- **Types**:
  - ✓ Obstructive non refluxing
  - ✓ Obstructive refluxing
  - ✓ Refluxing non obstructive
  - ✓ Non refluxing non obstructive (adynamic ureter)
- **Treatment**:
  - ✓ Obstruction: (ureteral reimplantation): excision of the abnormal segment and reimplanting of the UVJ.
  - ✓ Reflux: according to the same line of reflux management

3. Duplication Anomalies:

3.1 RENAL AND URETERIC DUPLICATION (not mentioned by the DR)

- 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
- **Embryological view**:
  - ✓ Normally: One ureteral bud (early precursor of the ureter) meet future kidney.
  - ✓ In Incomplete ureteral duplication: Ureteral bud bifurcate into 2 after the generation they go to kidney as 2 ureters
  - ✓ In complete ureteral duplication: 2 separate ureteral buds 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 that will be harmful



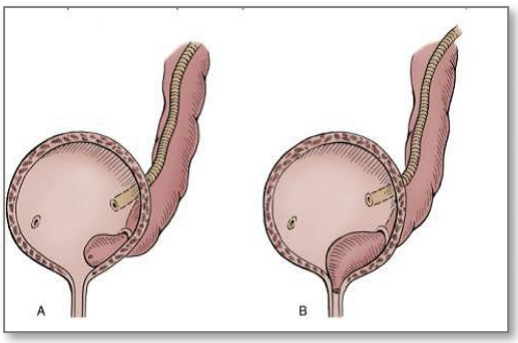
A: incomplete duplication
B: complete duplication
- Usually only one ureter comes from each kidney to bladder: single system.
- **Duplication Anomalies:** 2 ureters coming from each kidney and going to bladder.
- **Incomplete Duplication**: both ureters meet in their way and one ureter go to bladder.
- **Complete Duplication**: 2 duplications.

3.2 COMPLETE DUPLICATION FROM UPPER POLE OF KIDNEY TO LOWER POLE OF BLADDER (not mentioned by the DR)

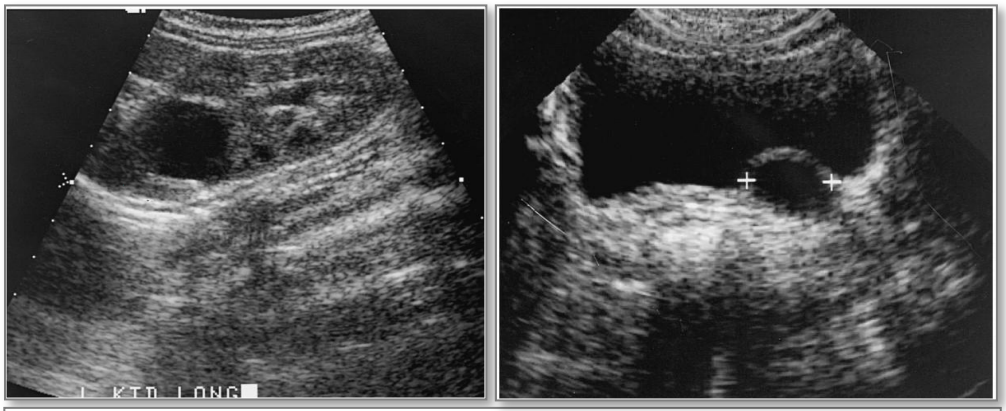
Here a principle called the Weiger-Meyer Law takes place which states: The upper pole ureter (which drain the upper pole of the kidney) comes to lower part of bladder and lower pole ureter coming to upper part of the bladder

3.3 URETROCELE

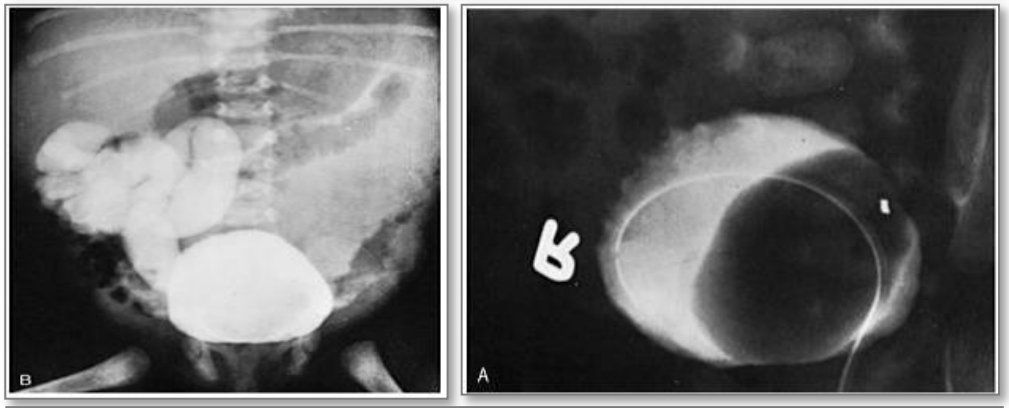
- Commonest cause of urine retention in female infants
- Cystic dilatation of the distal part of the ureter → This will lead to obstruction and the whole ureter will get dilated
- **Associated with**: duplication anomalies
- This can be confused with Uretrovesical junction obstruction but the difference here is there is cystic dilatation which can be present in the bladder
- Sacculation of the terminal portion of the ureter has **2 types**:



  - ✓ Orthotopic
    - Intravesical (inside bladder)
    - Simple OR adult type ureterocele.
  - ✓ Ectopic
    - Start in bladder and extended outside the bladder
    - Extravesical=duplex system OR infant type ureterocele.
    - In ectopic ureterocele it involve the upper pole system.
- **Presentation**:
  - ✓ Hydrouretronephrosis
  - ✓ 7:1 F:M, 10% bilateral, ectopic : orthotopic 4:1
  - ✓ Symptoms include: urine retention, infection and presence of stones
  - ✓ Intralabial mass: One of the differential diagnosis for an Intralabial mass in females is a ureterocele
- **Investigations**:
  - ✓ Usually detected by Antenatal (U/S) "seen as a thin-wall intravesical structure with the same consistency of the urine"
  - ✓ we use MCUG (micturating cystourethrogram " here it will show a filing defect. In MCUG, you put a catheter in the bladder with a contrast and look for the shape of the bladder, and see if the urine goes up) to confirm the diagnosis.
- **Management**:
  - ✓ Needs urgent intervention
  - ✓ Incision of ureterocele (Endoscopic puncture of ureterocele)
  - ✓ Upper pole heminephroectomy
  - ✓ Excision of ureterocele and common sheath reimplant.



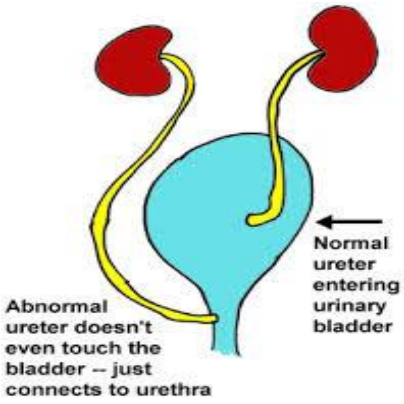
Cystic structure in bladder which is cystic dilation of distal part of ureter



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

3.4 ECTOPIC URETER

- Ureter does not implant in the bladder and stays outside
- Most commonly associated with duplex system and with ureterocele.(Normal ureter open in the trigone)
- Clinical picture depend on: associated anomalies, site and sex of the patient. (in case of a female: continuous wetting , but a male will not present with that, he may present with epididym-orchitis if the ureter opens in the vas deference)
- Can be:
  - ✓ Simple ectopia: implanted in abnormal position
  - ✓ Ectopic ureter: it is completely outside the bladder

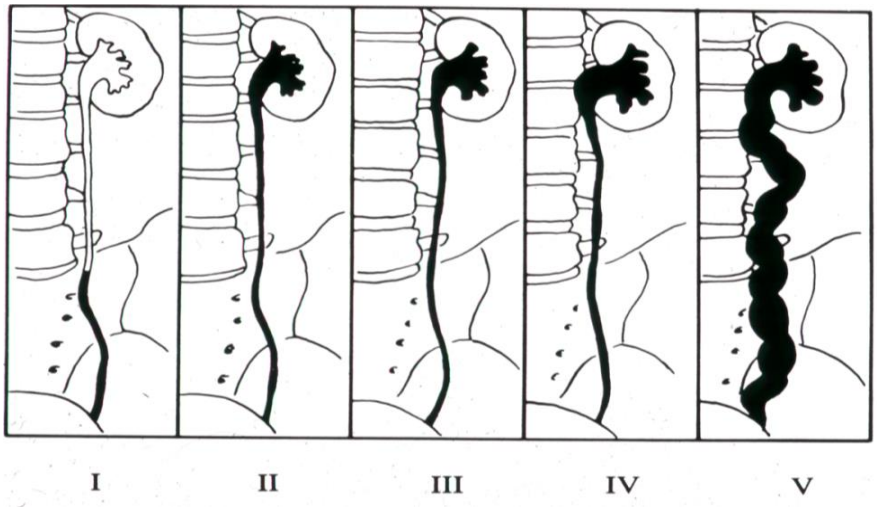


- **Investigations** include IVP, VCUG, cystoscopy
- Renal scan asses the function of both renal poles in case of duplication

4.VESICOURETERIC REFLUX

- There is a normal anti-reflux mechanism between the bladder and the ureter a "Flap Valve" which depends on:
  - ✓ Ureter has an Oblique course as it enters the bladder.
  - ✓ Proper muscular attachments to provide fixation.
  - ✓ Posterior support to enable its occlusion.
  - ✓ Adequate submucosal length.

- The study to rule out reflux is MCUG and it is also used for grading:
  Normal: contrast in bladder
  - ✓ Grade I: confined to ureter, contrast is in the distal part of the ureter
  - ✓ Grade II: contrast reaches the kidney but there is no dilation
  - ✓ Grade III: Mild dilation of the renal pelvis and ureter without loss of
  - ✓ calyces
  - ✓ Grade IV: moderate dilation but there is loss of calyces
  - ✓ Grade V: severe dilation and tortuous dilated ureter"



- Resolution of reflux: Over 3 year follow up:
  - ✓ 87% of Grade 1
  - ✓ 63% of Grade 2
  - ✓ 53% of Grade 3
  - ✓ 33% of Grade 4

- **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

  - ✓ Surgical management**:** (for grade 4 and 5 and if medical treatment failed "febrile infection") prevent the reflux by:
    - Ureteral reimplantation (surgical modality of choice)
    - Endoscopic correction: cystoscopic Deflux injection for correction

Note(s):

Prophylactic dose:

- 1l3 of therapeutic dose
- every (24h)
- if for urinary prophylaxis it will be excreted by kidneys

4. POSTERIOR URETHRAL VALVE (PUV):

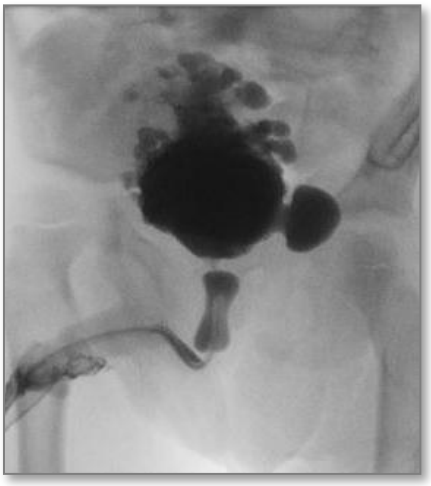
- Incomplete canalization of the posterior urethra
- 1:5000 male infants.
- Most common cause of urine retention in male infants.
- 50% have renal impairment.
- The bladder and the kidneys developed under high pressure and resistance.
- The more proximal the valve the more sever the condition
- Associated findings:
  - ✓ Oligohydramnios :
    - low amount of Amniotic fluid
    - No output of urine or little → Amniotic fluid
    - Low in Ultrasound "because there is no secretion but there is absorption".
    - Obstruction of esophagus → no absorption → Polyhydramnios.
  - ✓ Bilateral renal dilatation :
    - VUR: 40%
    - Valve bladder → loss of its Function and become abnormal bladder
    - The bladder is urogenic, because during pregnancy the detrusor muscle is replaced by collagen so no contraction of muscle occurs.
    - 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
    - "Posterior urethral valve is the universal destruction of urinary tract because it affects kidney, bladder, ureters, bladder, and urethra
    - Renal impairment: in 30-50%, 25% of them will have renal transplantation in the future
    - The obstruction is not complete (narrowing or stenosis) because if it is complete "severe" the patient will die in utero.
- **Presentation**:
  - ✓ Antenatal (hydonephrosis)
  - ✓ Urine retention
  - ✓ UTI
  - ✓ Poor urinary stream

Note(s):

- Between 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

- ✓ CRF; at late stage

- **Investigations**:
  - ✓ Antenatal US, US, VCUG, Renal scan, renal function studies, Urodynamic studies.
  - ✓ MCUG: Posterior urethra dilated, normal anterior urethra ,bladder trabeculated and elongated (Christmas tree bladder)
- **Treatment** :
  - ✓ Endoscopic primary valve ablation



Other ANOMALIES:

1. PATENT URACHUS:

- the urachus is a remnant of a channel between the bladder and the umbilicus (where urine initially drains in the fetus during the 1st trimester of pregnancy). The channel of the urachus usually obliterates around the 12th week of gestation.
- **Presentation**: varying amounts of clear urine to leak at the umbilicus.
- **Investigations** In MCUG : you will see contrast in abdomen.
- **Treatment :**
  - ✓ Usually asymptomatic and it will close by itself eventually with time.
  - ✓ If the urachal disorder presents with an infection, the infection is treated first. And occasional surgical drainage of any infected cyst or poorly draining cavity.
  - ✓ Once the infection is under control, excision of the urachus is usually performed. This can usually be done laparoscopically or with a small incision on the lower abdomen.

2. BLADDER DIVERTICULUM:

- Out pouching in bladder.
- Primary → congenital, solitary.
- Secondary → acquired, multiple.

  Acquired bladder diverticula are typically the result of obstruction of the bladder outlet (e.g. an enlarged prostate or area of scarred urethra, posterior urethral valve), bladder dysfunction from nerve injury or, rarely, as a result of prior bladder surgery.
- **Treatment**:

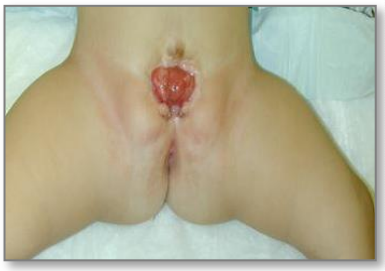
  Follow up, do not always require treatment. If becomes very large → excise it. (Because there will be urine stasis→ stones).

3. BLADDER DUPLICATION:

- Complete: each one has its own urethra and urethral opening.
- Incomplete : two bladders opens in one urethra

4. BLADDER EXSTROPHY (only bladder)

- 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
- if you see part of bowel also → cloacal extrophy
- 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

5. EPISPADIAS

- Very rare, Abnormal position of external urethral meatus in dorsal surface of the penis
- If in a female, commonest presentation is incontinence

6. Hypospadias :

- Abnormally located meatus which contains the urethral opening
- The meatus usually opens in the tip of the glans penis and if it opens anywhere else it is considered an ectopic opening.
  - ✓ In hypospadias the opening is towards the scrotum or ventral side
  - ✓ In Epispadias the opening is towards the abdomen or dorsal side
- Common (2%)
- Abnormal position of the EUM(external urethral meatus):
  - ✓ Distal hypospadias: (from mid shaft to Glans)
  - ✓ Proximal hypospadias (from
- proximal penile “proximal shaft” to the perineal)
- **Treatment:**
  - ✓ NO Circumcision
  - ✓ Absolute contraindication because dorsal urethral skin we will be needed in repairing later on especially in proximal hypospadias
  - ✓ Age to repair:6 to 9 months
  - ✓ Requires one stage repair

7. PRUNE BELLY SYNDROME (Eagle-Barrett Syndrome):

- Consists of a triad of:

1- Absent abdominal wall muscles

- External oblique, Internal oblique, Transverse abdominal muscles
- You can feel all the organs (kidneys and liver) and you can even see the bowel movement because the muscle layer is either absent or thin (hypopalstic).

2- Bilateral undescended testis (intra abdominal)

3- obstructive uropathy

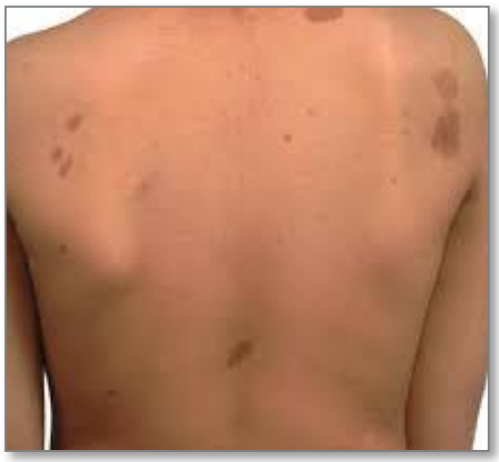
- bilateral hydrouretronephrosis and large bladder (dilation of whole urinary system because no peristalsis "abnormality in smooth muscles" → urine stasis)

- In a female → pseudo prune belly syndrome (because classically it is a triad and in a male)

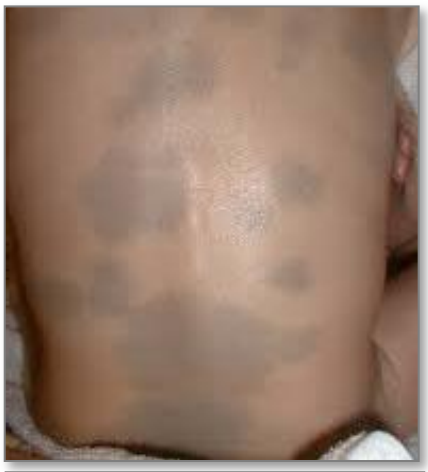
8. Myelo-meningocele

- (1) The baby may have a closed swelling containing spinal cord and/or spinal nerves. (2) More often, the spinal canal is open and leaks CSF, with his flattened cord forming a plaque on its surface→ Causes neurogenic bladder .
- Always examine from the back in a baby with urological symptoms.
- presentation :
  - ✓ Loss of bladder or bowel control
  - ✓ Partial or complete lack of sensation
  - ✓ Partial or complete paralysis of the legs
  - ✓ Weakness of the hips, legs, or feet of a newborn
  - ✓ Skin lesions :
    - café-au-lait = coffee with milk, hyper-pigmented lesions that may vary in color from light brown to dark brown. The borders may be smooth or irregular.
    - Hypertrichosis (abnormal amount of hair growth over the body)
    - Dermal Sinus Tracts

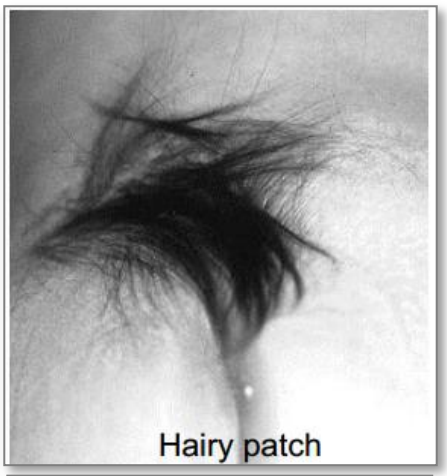
- Mongolian spots → green spots are normal finding. (Flat, gray-blue in color (almost looking like a bruise), are very common in any part of the body of dark-skinned babies).



café-au-lait



Mongolian spots



Hypertrichosis

SUMMARY

1. Bilateral renal agenesis *in utero is* **oligohydramnios** → *increased pressure on the developing fetus* → flat face, nose, and ears".
2. If you can see posterior bladder wall only → bladder extrophy , if you can see part of bowel also → cloacal extrophy
3. if in a female with epispadias, commonest presentation is incontinence.
4. circumcision is contraindicated with any abnormality in the penis
5. prune belly syndrome is classically in a male and a triad of absent abdominal wall muscles- bilateral undescended testis - obstructive uropathy
6. always examine from the back in a baby with urological symptoms → maybe myelo-meningocele

PUJO	**US: isolated hydronephrosis**	**Pyeloplasty.**	**most common cause of antenatal hydronephrosis**
URETEROVESICAL JUNCTION OBSTRUCTION	US: hydrouretronephrosis	ureteral reimplantation	
URETROCELE	▪ MCUG: filing defect. ▪ US: thin-wall cyst in bladder	Endoscopic puncture of ureterocele	Commonest cause of urine retention in female infants
ECTOPIC URETER	IVP, VCUG, cystoscopy	-	In a female → continuous wetting , but a male will not present with that
VESICOURETERIC REFLUX	MCUG	Medical management: UTI → prophylaxis Surgical management: Ureteral reimplantation	
POSTERIOR URETHRAL VALVE	▪ MCUG : Posterior urethra dilated , Christmas tree bladder ▪ US : Bilateral renal dilatation, Oligohydramnios	Endoscopic primary valve ablation	

Questions

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

a. epispadias
b. uretrocele.
c. prune belly syndrome
d. patent urachus

2) what is the best modality to rule out urine reflux?

a. MCUG
b. US
c. IVP
d. ERCP

3) A female child presented with Intralabial mass what is the most likely diagnosis?

a. prune belly syndrome
b. epispadias
c. ureterocele
a. myelo-meningocele

4) Which of the following congenital anomalies is an absolute contraindication for Circumcision:

a) Bilateral undescended testis.
b) Hypospedias.
c) posterior urethral valve.
d) Vesioureteral Refulx.

5) 5 Years old female presented with history of recurrent febrile UTI with incontinence, whats the best modality in diagnosing VUR?

d. Intravenous ureterogram(IVP)
e. nuclear study
f. Voiding Cysturethrogram (VCUG)
g. Renal and bladder US

Answers:

1st Questions: d

2nd Questions: a

3rd Questions: c

4th question : B

5th Question :c