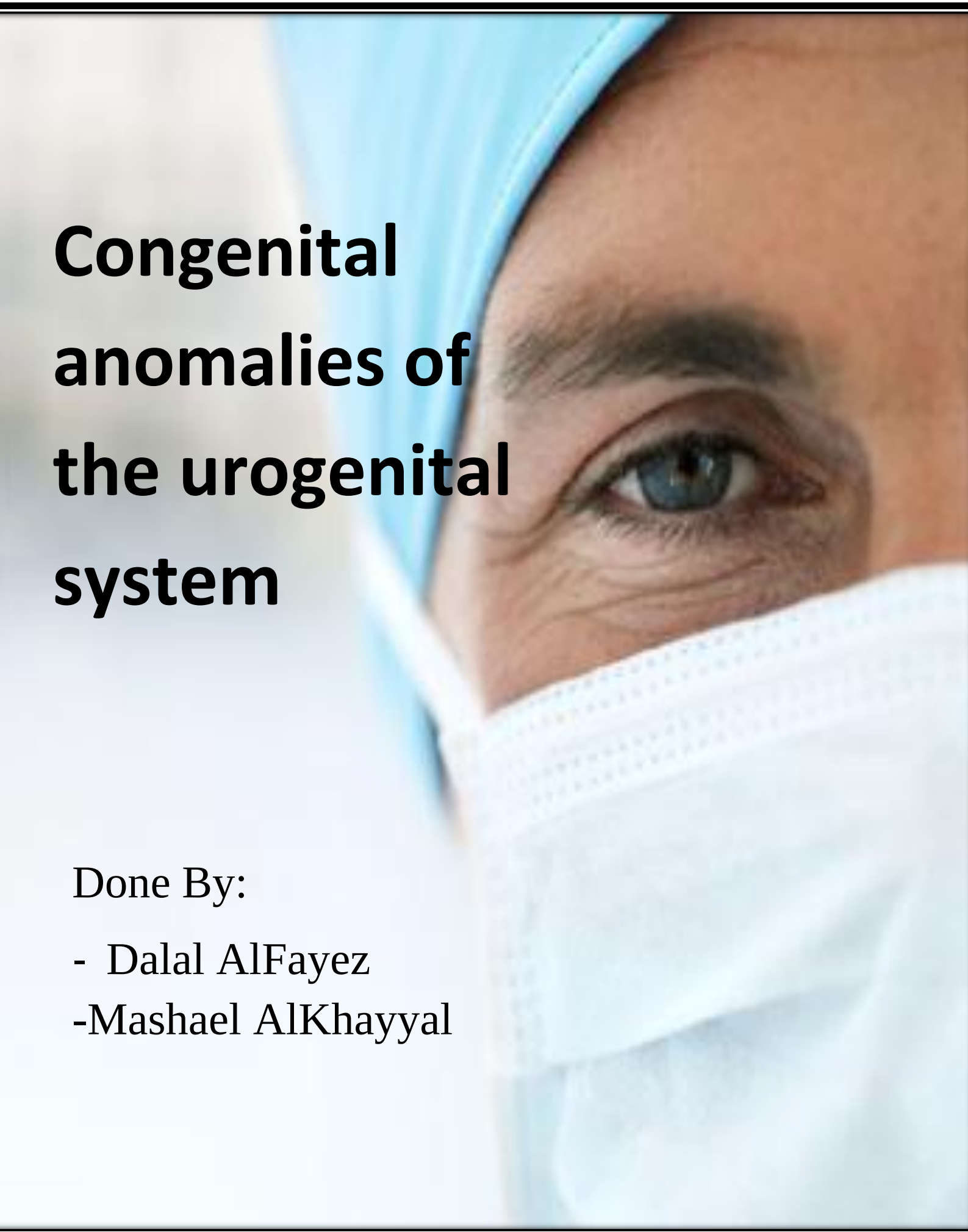
430 Surgery Team



Congenital anomalies of the urogenital system

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Congenital genitourinary anomalies are the most common of all organ system anomalies.

- 10% of population has some type of urogenital anomaly.
- 14:1000 births have antenatal diagnosis of urogenital anomaly. (UGS anomalies could be detected during pregnancy=antenatal)
- Antenatal ultrasound after 28 weeks gestation.

Congenital anomalies of UGS are:
-Antenatal Hydronephrosis
-Anomaly of position, number and rotation
-Cystic abnormalities
-Prune Belly Syndrome
-Hypospadias
-Epispadias
-Bladder e Exstrophy

A-External anomalies:

1-**Hypospadias**:

Abnormal opening of the external urethral meatus on the ventral side. (Normal EUM opens at the tip of the glans) (N.B. Ventral side 'downward', dorsal side of the penis is towards the abdomen 'upward')

-EUM might open Distally (from mid-shaft to glandular) or Proximally (from proximal penile to perineal).

-Circumcision is absolutely contraindicated after childbirth. (The extra skin of foreskin removed by circumcision is needed in the repair surgery)

-Treatment: Surgery at the age of 6-9 months.

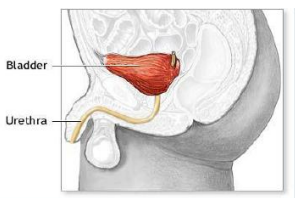
-**Epispadias**: abnormal opening of the external urethral meatus on the dorsal side.

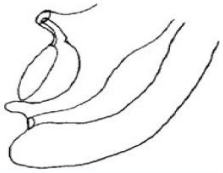
-Circumcision is contraindicated as well.

-Surgical repair of epispadias needs more skin --> skin graft.

2-**Bladder Exstrophy**: absence of the anterior abdominal wall (exstrophy: the turning inside out of an organ).

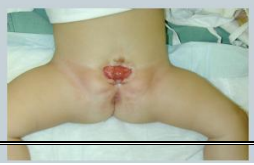
-Often associated with Indirect Inguinal hernia.





-Treatment: Surgery immediately after childbirth. (A neonatal pediatric emergency)

-Immediate surgery is performed to prevent

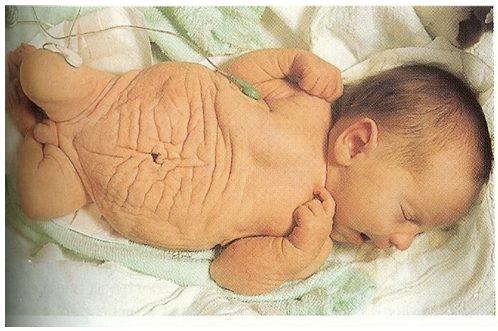


complications of exposed bladder (e.g. infections)

3-Prune Belly Syndrome or Triad Syndrome (in males) or Incomplete Prune Belly Syndrome (in females)

-Includes:

1. Absence of abdominal muscles or presence of a very thin layer. (Bowel movement is visible)
2. Bilateral undescended testis: not palpable testis.
3. Obstructive uropathy.



B-Internal anomalies:

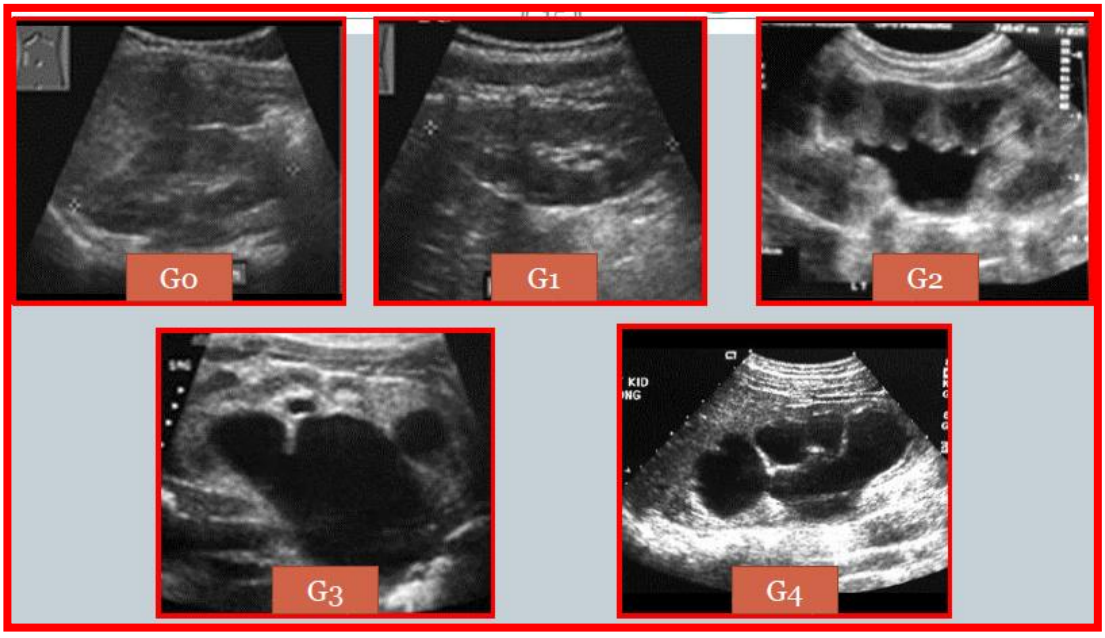
1-Hydronephrosis: dilation of the renal pelvis. (hydronephrosis is a finding not a diagnosis!)

-Hydroureterosis: dilation of the ureter.

-Hydroureteronephrosis: dilation of both ureter and pelvis.

-Investigation: detected by ultrasound: - (**Antenatal Hydronephrosis**: detected during pregnancy)

-SFU grading system is used:



-SFU grade 0: No splitting of renal sinuses (normal kidney)

-SFU grade 1: Urine in pelvis barely splits sinus

-SFU grade 2: Urine fills intrarenal pelvis + Urine fills extra renal pelvis major calyces dilated

-SFU grade 3: SFU Gr 2 and minor calyces uniformly dilated and parenchyma preserved

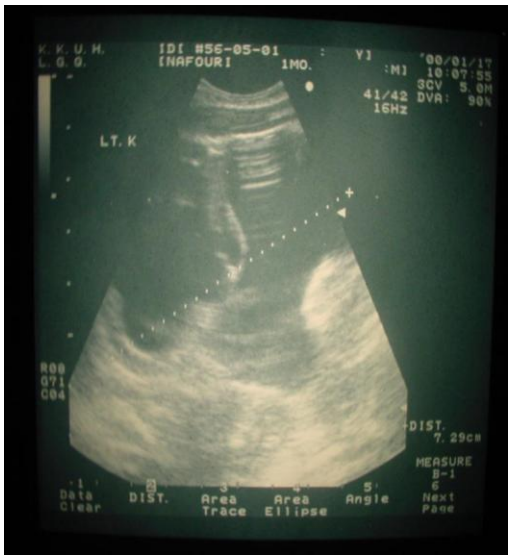
-SFU grade 4: SFU Gr 3 and parenchyma thin (severe dilation of renal pelvis)

*Urine normally appears black in US

*To differentiate between hydronephrosis and renal cyst (In hydronephrosis there will be communication between the renal pelvis and calyces, but in renal cyst there will be no communication between cysts)

-Causes of Antenatal Hydronephrosis:

A- Pelviureteric junction obstruction (41%): obstruction of the junction between renal pelvis of the kidney and the ureter = dilated pelvis+normal ureter



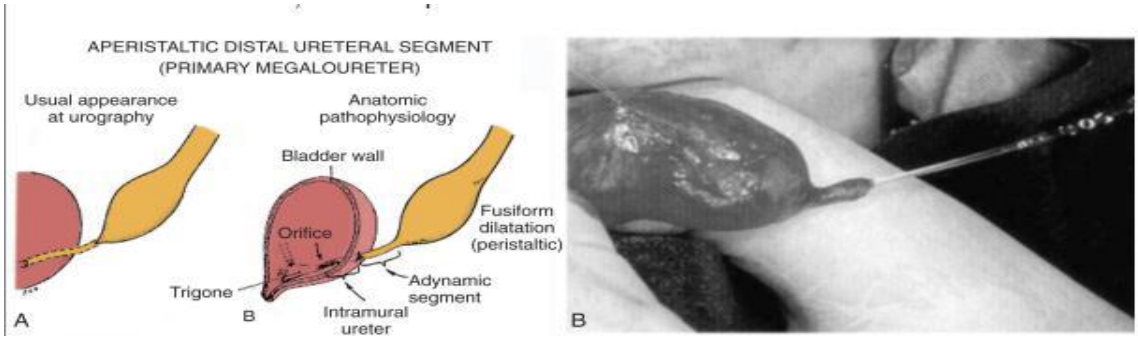
Presentation: Incidental in children and neonates\Symptomatic (UTI,pain,hematuria,stone,mass)

Treatment: Surgery; Pyeloplasty with or without stent

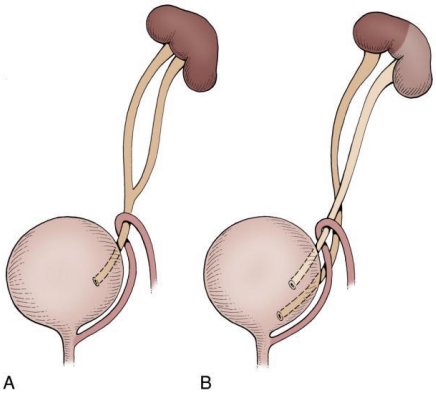
B- Ureterovesical junction obstruction (23%): obstruction of the junction between ureter and bladder = dilated pelvis+dilated ureter (Megaureter)

Types: Obstructive not refluxing, Obstructive refluxing, Non obstructing refluxing, Non obstructive non refluxing

Treatment: Surgery; Uretral Reimplantation (cut and re-connect ureter and bladder)



C- Duplication anomalies



(13%): renal and ureteric duplication = reflux 43%, renal dilatation 29%, ectopic insertion 3%, infections and ureterocele.

Incidence: 1%, 1.6:1 F:M, 85% unilateral.

Embryology: Either two urethral buds meeting the meta-nephros or one ureteric bud that bifurcates. (Not important)

Types: Incomplete: in which one common ureter enters the bladder, Complete: in which 2 ureters ipsilaterally enter the bladder

*Weiger-meyer law: in cases of double ureter, the ureter from the upper pole of the kidney usually opens below and medial to the one from the lower pole.

Presentation: Asymptomatic\Symptomatic: if there is an associated anomaly.

Duplication per se is of no clinical significance, but the associated anomalies may require intervention.

3-**Ureterocele**:

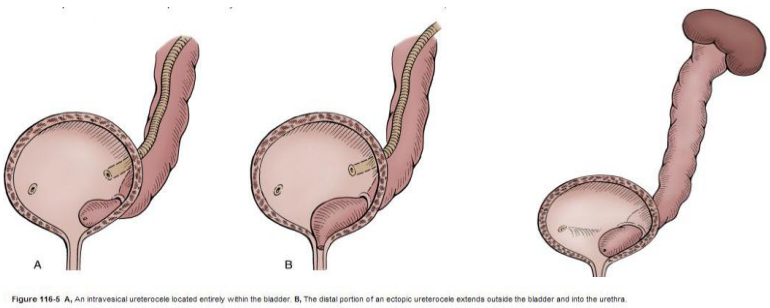


Figure 116-5 A, An intravesical ureterocele located entirely within the bladder. B, The distal portion of an ectopic ureterocele extends outside the bladder and into the urethra.

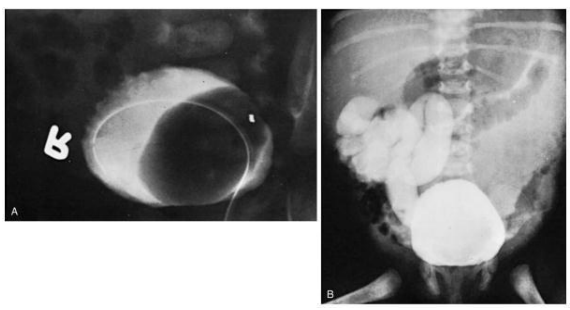
Sacculation of the terminal portion of the ureter (Cystic dilation of the terminal part of ureter due to stenosis of the uretral orifice)

- Can occur in children with single or double ureters (<double)

Incidence: 7:1 F:M, 10% bilateral, ectopic: orthotopic 4:1

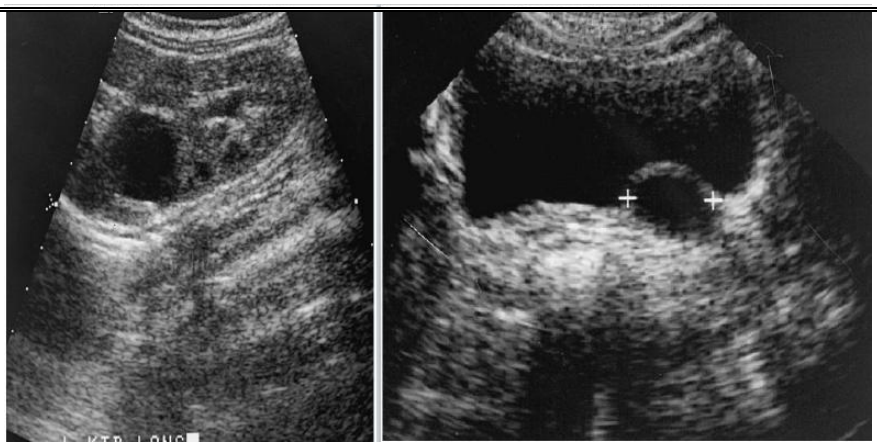
Types: 1- Orthotopic = intravesical=simple=adult type ureterocele.

2- Ectopic = extravesical=duplex system= infant type ureterocele. (Involves the upper pole system)



- Commonest cause of urine retention in female infants.
- Presentation: Incidental (antenatal US), urine retention, calculus formation, infection.

Treatment: Puncture



4-**Ectopic ureter**:

Is an abnormally located terminal portion of the ureter. Instead of the ureter opening in the bladder, it opens in the urethra, vagina, or uterus.

-Most commonly associated with duplex system and with ureterocele.

-Clinical picture depend on: associated anomalies, site and sex of the patient.

-Treatment: ureteric reimplantation if the kidney still works

D-Vesicoureteric reflux (7%):

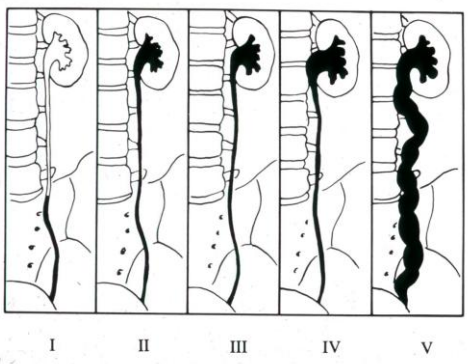
Normal anti-reflux mechanism "Flap valve"

1. Oblique course as it enters the bladder.
2. Proper muscular attachments to provide fixation.
3. Posterior support to enable its occlusion.
4. Adequate submucosal length.

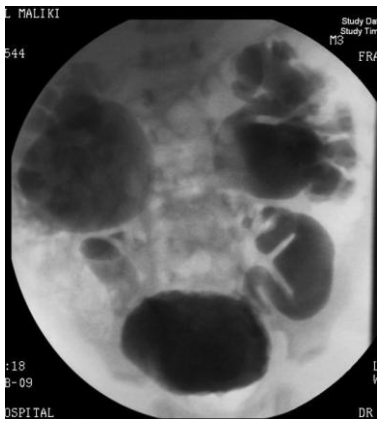
-Detected by MCUG (Micturating Cysto Urethrogram): -

-5 grades:

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



-Patient is given prophylactic antibiotics even before investigations to prevent UTI and protect the kidney. Prophylactic dose: (1/3 of therapeutic dose only at night (24h))

-Grade 1,2 and most of 3 > spontaneous resolution (but greater grades need surgery either ureteric implantation or endoscopic correction of reflux)

E-Posterior urethral valves (10 %):

Incomplete canalization between anterior and posterior urethra (Urethral atrasia: no canalization occurred > patient will die)= Oligohydramnios (low amount of Amniotic fluid), Bilateral renal dilatation, VUR: 40%, valve bladder, and renal impairment. (Will cause universal destruction)

Incidence: 1:5000 male infants.

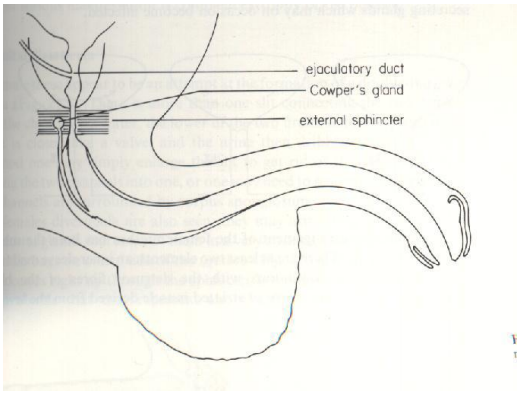
*Most common cause of urine retention in male infants.

50% have renal impairment.

*The bladder and the kidneys developed under high pressure and resistance.

Presentation: Incidental: Antenatal US, Urine retention, UTI, Poor urinary stream, CRF (chronic renal failure)

Treatment: ablation by cystoscopy



F-MCDK (B\M) Multicystic Dysplastic Kidney

G-Others (6%)

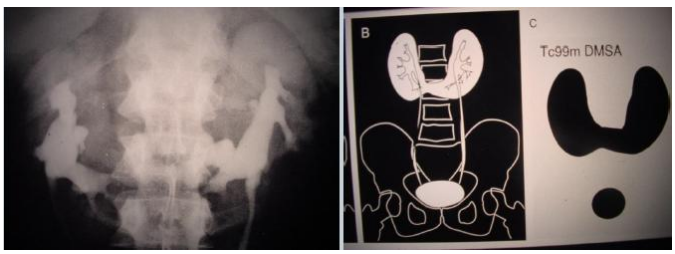
Kidney.

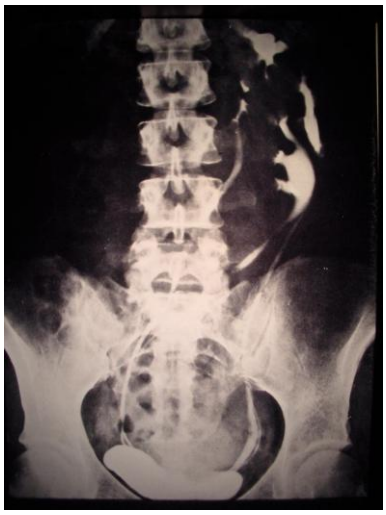
A-Anomalies of position, number and rotation:-

1. Simple ectopia: Normal position of kidney: retroperitoneal in flank area. Anywhere except this place= ectopia

- A kidney that is outside the renal fossa.
- Pelvic (commonest), lumbar, sacral.

2. Thoracic kidney.





3. Horseshoe kidney. (The 2 kidneys fuse together mostly in the lower lobes by fibrous tissue)

4. Unilateral renal agenesis.

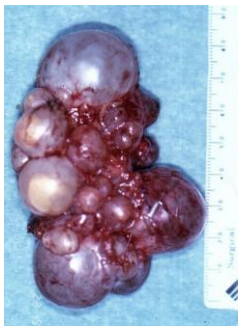
5. Bilateral renal agenesis.

6. Crossed renal ectopia with no fusion. (Kidney goes to other side)

7. Crossed renal ectopia with fusion.

8. Malrotated kidney.

B-Cystic abnormalities



1. Renal dysplasia (dysplasia=no renal tissue)

-Congenital unilateral multicystic kidney (a failed kidney with gross multiple cysts, treatment is nephroectomy if it gets larger)

-Segmental and focal renal dysplasia.

-Renal dysplasia associated with congenital lower tract obstruction.

2. Congenital polycystic kidney disease (polycystic=renal tissue is present)

-Infantile type

-Adult type

3. Simple cyst

4. Calyceal cyst

5.Peripelvic cyst

6. Perinephric cyst