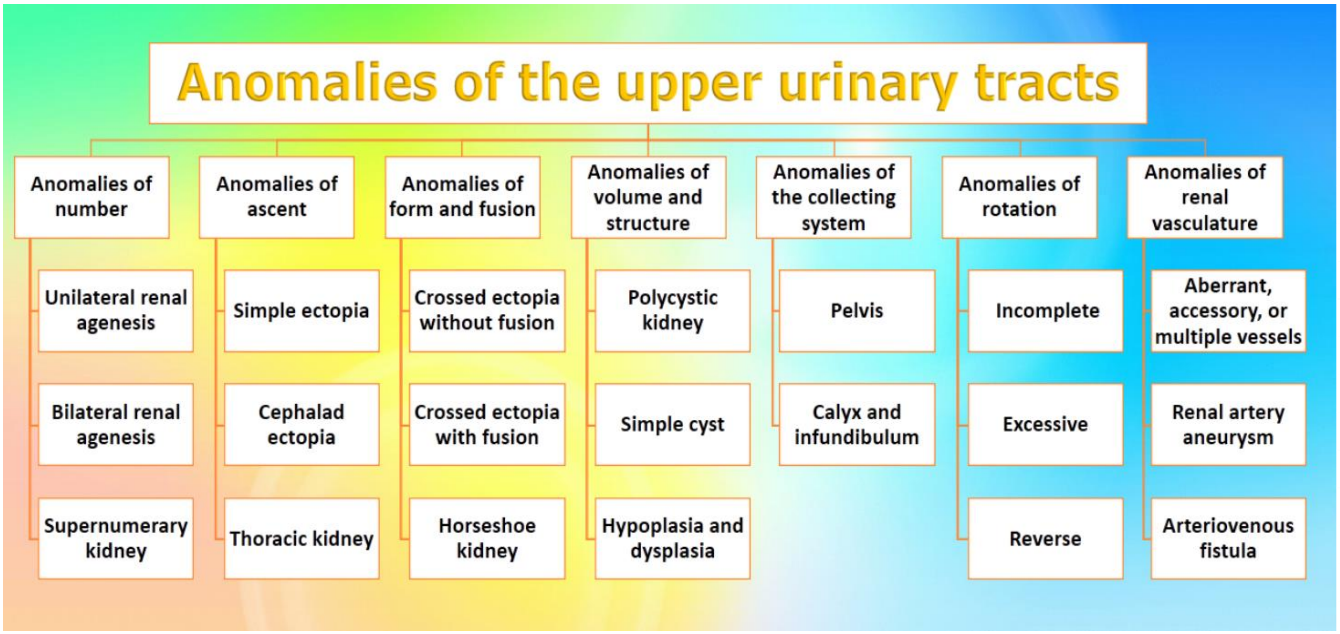
Lec:13 **Congenital Anomalies of The Upper Urinary Tract** المرحلة الخامسة

د. وليد خالد العبيدي

- Congenital anomalies of the upper urinary tract comprise a group of abnormalities, ranging from complete absence to aberrant location, orientation, and shape of the kidney as well as aberrations of the collecting system and blood supply.

Surgical anatomy

- The parenchyma of each kidney usually drains into seven calyces, three upper, two middle and two lower calyces. Each of the three segments represents an anatomically distinct unit with its own blood supply.
- The kidney and renal pelvis normally rotate 90 degrees ventromedially (toward midline) as they leave the true pelvis during beginning of ascent at 6th week of gestation so that the calyces point laterally and the pelvis faces medially. When this alignment is not exact, the condition is known as malrotation.



Unilateral Renal Agenesis (URA)

Incidence: 1: 1400 births

- Found accidentally, more frequently on the left side.
- Ipsilateral adrenal agenesis is rarely encountered with URA
- Other Genital anomalies are much more frequently observed.

Symptoms: Asymptomatic

Diagnosis: U/S, CT scan: absent kidney on that side + compensatory hypertrophy of the contralateral kidney

Treatment: no specific treatment.

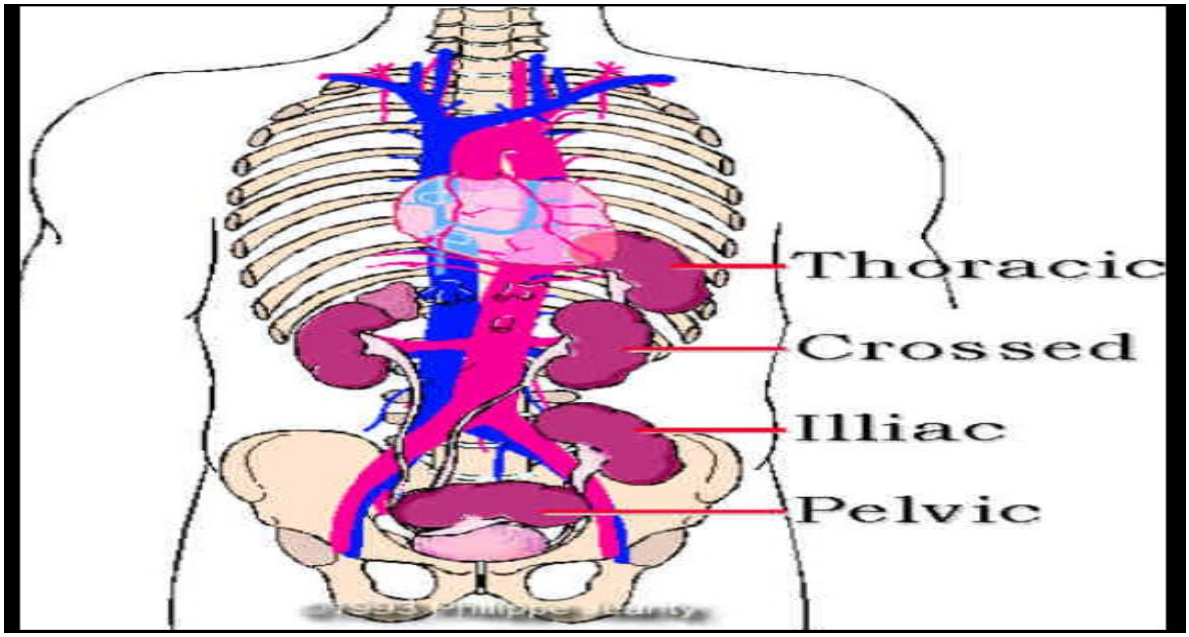
Prognosis: no evidence that they have an increased susceptibility to other diseases

Bilateral agenesis: rare, incompatible with life

ANOMALIES OF ASCENT

1. Simple Renal Ectopia

- When the mature kidney fails to reach its normal location in the "renal" fossa, the condition is known as renal ectopia. The term is derived from the Greek words ek ("out") and topos ("place") and literally means "out of place."
- Incidence The incidence is 1 in 1000
- An ectopic kidney can be found in one of the following positions: pelvic, iliac, abdominal, thoracic, and crossed.
- The renal pelvis is usually anterior (instead of medial) to the parenchyma, because the kidney has incompletely rotated. As a result, some of ectopic kidneys have a hydronephrotic collecting system due to obstruction of the ureteropelvic or the ureterovesical junction.



Associated Anomalies:

1. Contralateral agenesis: The incidence of appears high.
2. Hydronephrosis secondary to obstruction or reflux may be seen in the contralateral kidney

Clinical features: Most ectopic kidneys are asymptomatic

Diagnosis: U/S, CT scan.

Prognosis: The ectopic kidney is no more susceptible to disease than the normally positioned kidney except for the development of hydronephrosis or urinary calculus formation or the presence of ectopic ureter**.**

ANOMALIES OF FORM AND FUSION

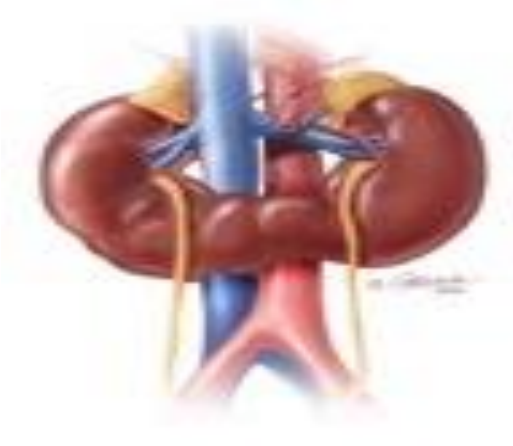
Crossed Renal Ectopia With and Without Fusion

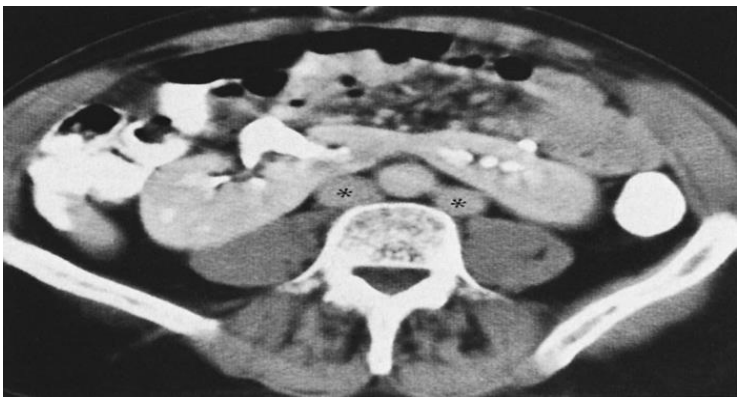
- When a kidney is located on the side opposite from that in which its ureter inserts into the bladder, the condition is known as crossed ectopia.

Horseshoe Kidney

- Probably the most common of all renal fusion anomalies.
- Incidence: 1: 1000 births
- The anomaly consists of two distinct renal masses lying vertically on either side of the midline and connected at their respective lower poles by a parenchymatous or fibrous isthmus that crosses the midplane of the body.
- Fusion of the renal masses early in embryonic life, so its ascent will be impeded by inferior mesenteric artery.
- The kidneys are low located at the level of the 4th lumbar vertebrae, malrotated and pelvis lie anteriorly.

Diagnosis: ultrasound, CT scan.





Symptoms: When present, they are related to complications like hydronephrosis, infection, or calculus formation due to ureteric angulation or obstruction with impaired urine drainage

Treatment:

Medical: pain relief and antibiotics to control infection

Surgical: if present, stone removal, Pelviureteric junction obstruction correction.

Prognosis: usually they have normal life.

Cystic disease of the kidneys

Polycystic kidney disease:

- The kidney is one of the most common sites in the body for cysts
- Two types:

1. AUTOSOMAL RECESSIVE ("INFANTILE") POLYCYSTIC KIDNEY DISEASE
2. AUTOSOMAL DOMINANT ("ADULT") POLYCYSTIC KIDNEY DISEASE

Autosomal dominant polycystic kidney disease

- Autosomal dominant, transmitted by either parent.
- 50% of offspring affected.
- Both kidneys replaced by large no. of cysts of variable size which make the kidney of large size.
- 15% associated with cystic disease of liver, lung, pancreas or spleen.

Aetiology & Pathogenesis

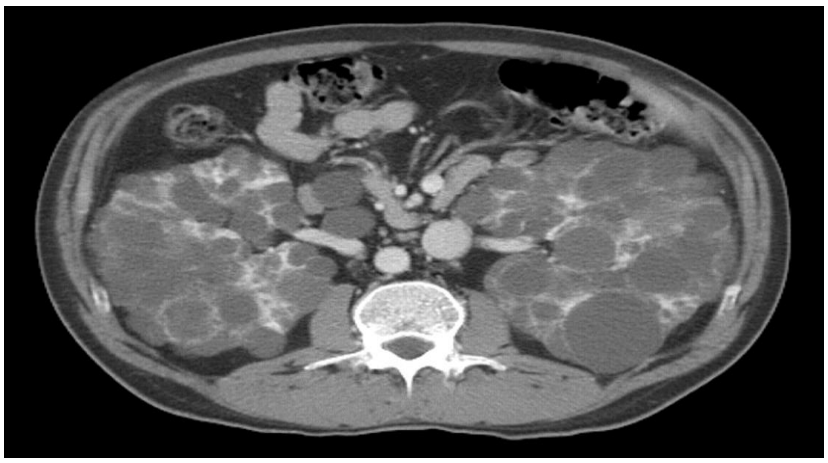
- The cysts occur because of defects in the development of the collecting and uriniferous tubules and in the mechanism of their joining.
- Blind secretory tubules that are connected to functioning glomeruli become cystic.

Clinical presentation:

- Rarely gives clinical manifestation before 4o years

1. Asymptomatic: diagnosed accidentally.
2. Pain: due to pedicle stretching, stone, bleeding inside cyst or infection.
3. Hematuria
4. Infection
5. Hypertension: in 70%, unknown cause.
6. Renal impairment
7. Renal enlargement

Diagnosis: Family history of polycystic disease. U/S, CT scan, MRI.



Treatment:

Medical:

1. To control infection, hypertension, pain and anemia.
2. Renal impairment: by low protein diet and dialysis.

Surgical:

1. Rovsing's operation (deroofing) for large cysts causing symptoms or obstruction.
2. Stone removal.
3. Renal failure: Renal transplantation.

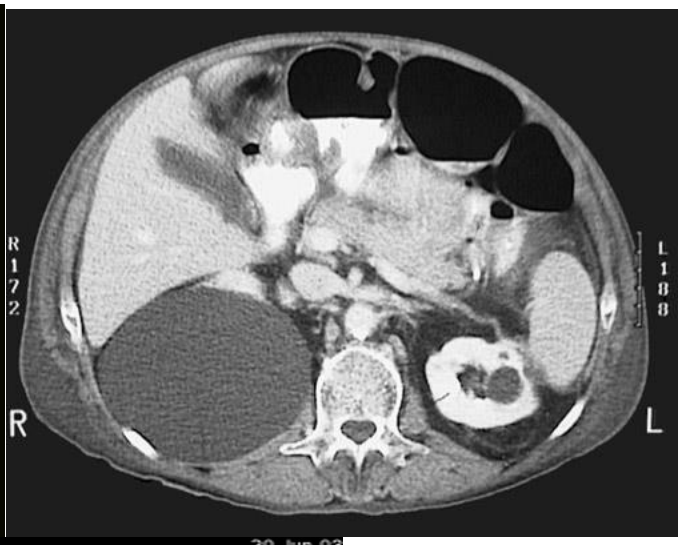
Autosomal recessive polycystic kidney disease

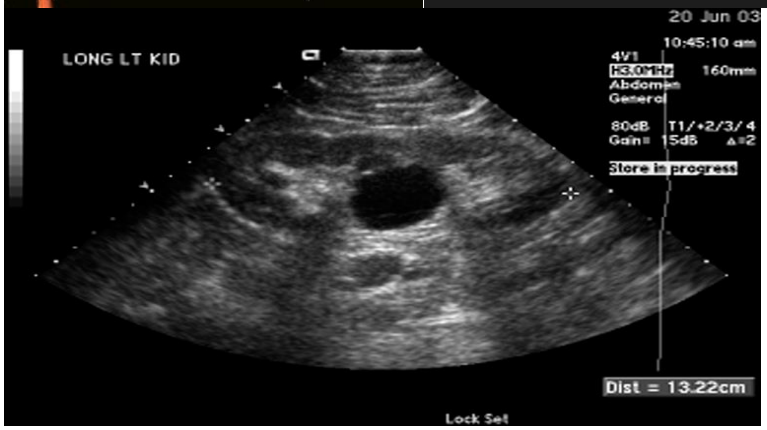
- Rare autosomal recessive, incompatible with life. 50% die at birth.
- Both kidneys are large in size and replaced by large number of cysts which may obstruct labor. Associated with hepatic fibrosis

Simple (solitary) renal cyst

⇒ Common condition.
⇒ Single or multiple.
⇒ unilateral or bilateral.
⇒ Congenital or acquired.
⇒ Usually asymptomatic. In 10% symptomatic: pain,heaviness, infection, bleeding inside the cyst or pressure effect on the ureter causing hydronephrosis.

Diagnosis: U/S, CT scan &MRI





Treatment: usually no treatment needed
Symptomatic patients:

1. Aspiration and injection of sclerosing agent.
2. Rovsing's operation (deroofing).

⇒ N.B. Malignant cyst: radical nephrectomy.

Congenital Anomalies of Renal pelvis & Ureter

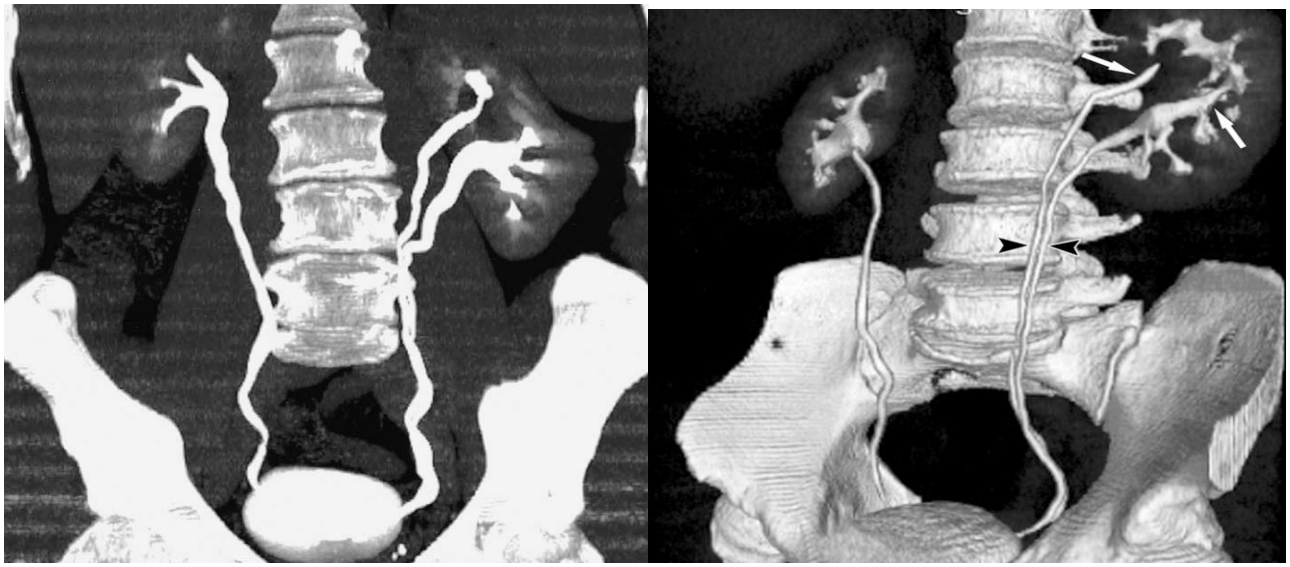
Duplication of Renal Pelvis: More common on left side.

Duplication of the ureter:

- Usually the ureters fuse & have common orifice in the bladder although they may open independently in the bladder.
- ***Ureteral duplication: partial and complete***

1. Partial duplication: is more common. Two ureters draining single kidney for variable length, then unite together before entering the bladder in one ureteric orifice. Rarely the lower part is duplicated as inverted Y ureter.
2. *Complete duplication:* Less frequent, the whole ureter is duplicated, and each one opens in separate orifice in the bladder. The ureter draining the upper part opens more distally in the bladder.

Clinical features: usually asymptomatic



Ectopic Ureters

- ✓ Ectopic ureter is the ureter that does not enter the trigonal area of the bladder.
- ✓ In the male, the posterior urethra is the most common site of termination, also to seminal vesicle
- ✓ In the female, the urethra and vestibule are the most common sites

Clinical features: According to the site of orifice

- ✓ In females: continuous dribbling.
- ✓ In males: urinary tract infection.

Diagnosis: IVU, U/S, CT scan, cystoscopy

Treatment:

1. Ureteric reimplantation
2. Partial nephrectomy (or nephrectomy): Ectopic ureters may drain renal moieties (either an upper pole or a single system kidney) that have minimal function.

Ureteroceles

- ✓ Is due to congenital atresia of the ureteric orifice which causes a cystic dilatation of the intramural portion of the ureter
- ✓ Women > men
- ✓ Sometimes involves with ectopic ureter
- ✓ More prone to stone disease & UTIs

Clinical Features: asymptomatic, Repeated UTIs, Hematuria.
Diagnosis: IVU ('cobra head sign'), cystoscopy, cystogram.
Treatment

1. Asymptomatic: no treatment
2. Cystoscopy with diathermy incision of the ureterocele.
3. In complicated cases, ureteral reimplantation.
4. or Nephrectomy in non-functioning kidney.

Ureteropelvic Junction (UPJ) (PUJ) Obstruction (stenosis)

- ⇒ The most common cause of significant dilation of the collecting system in the fetal kidney
- ⇒ Boys > Girls
- ⇒ Left-sided lesions predominate
- ⇒ Could be bilateral

ETIOLOGY

1. Intrinsic (intramural): interruption in the development of the circular musculature of the UPJ or mucosal fold that causes valve like effect.
2. Extrinsic: An aberrant, accessory, or early-branching lower-pole renal artery.

SYMPTOMS / PRESENTATION

- ⇒ Most infants are asymptomatic.
- ⇒ most children are discovered because of their symptoms

1. Episodic flank or upper abdominal pain associated with nausea and vomiting.
2. Recurrent infections.
3. failure to thrive, diarrhea.
4. loin mass.

DIAGNOSIS

1. U/S, IVU, CT scan, Magnetic Resonance Imaging.
2. Radionuclide Renography: MAG3 to see the split function of each kidney.

Treatment:
Medical:

1. control infection and pain.
2. Suppressive antibiotics

Surgical:

Indications for surgery:

1. Progressive hydronephrosis.
2. UTI despite antibiotic cover, and symptomatic patients.
3. Severe hydronephrotic nonfunctioning kidney.
4. Deterioration of renal function

A. **SURGICAL REPAIR:**

✓ Open & laparoscopic surgical techniques

1. Anderson-Hynes dismembered pyeloplasty: excision of the pathologic UPJ & appropriate reanastamosis.
2. Flap technique or flap operation

✓ Endoscopic Approaches:

1. Balloon dilatation
2. Antegrade endopyelotomy

B. Nephrectomy : for nonfunctioning kidney.

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