جراحة
SURGERY

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((Congenital anomalies of the upper urinary tract))

Anomalies of number:

-Agenesis : Unilateral , Bilateral .

-Supernumerary kidney .

Anomalies of volume and structure:

-Hypoplasia

-Multicystic kidney

-Polycystic kidney : Infantile , Adult , Other cystic disease , Medullary cystic disease

Anomalies of ascent:

-Simple ectopia

-Cephalad ectopia

-Thoracic kidney

Anomalies of form and fusion:

Crossed ectopia with and without fusion

- Unilateral fused kidney (inferior ectopia)
- Sigmoid or S-shaped kidney
- Lump kidney
- L-shaped kidney
- Disc kidney
- Unilateral fused kidney (superior ectopia)
- Horseshoe kidney

Anomalies of rotation:

-Incomplete

-Excessive

-Reverse

Anomalies of the collecting system:

1. Calyx and infundibulum

- Calyceal diverticulum
- Hydrocalyx
- Megacalycosis
- Unipapillary kidney
- Extrarenal calyces

- Anomalous calyx (pseudotumor of the kidney)
- Infundibulopelvic dysgenesis

2.Pelvis

- Extrarenal pelvis
- Bifid pelvis

Anomalies of renal vasculature:

Aberrant, accessory, or multiple vessels

Renal artery aneurysm

Arteriovenous fistula

Congenital anomalies of the upper urinary tract

In summery comprise a diversity of abnormalities, ranging from: complete absent kidney, supernumerary Kidney , aberrant location, orientation, and shape of the kidney ,aberrations of the collecting system &blood supply.

ANOMALIES OF NUMBER :

- ***Unilateral Renal Agenesis (URA):***

Incidence : **1: 1400 births**

Found accidentally, more frequently on the left side.

Embryology : Complete absence of a ureteric bud or aborted ureteral development prevents maturation of the metanephric blastema into adult kidney tissue.

*Ipsilateral adrenal agenesis is rarely encountered with URA

*Other Genital anomalies are much more frequently observed

Asymptomatic

Diagnosis : U/S or IVU,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.
- ***Supernumerary Kidney:*** truly an accessory organ

Incidence: very rare

Symptoms: It may not produce symptoms until early adulthood, if at all.
Diagnosis: accidentally by IVU or abdominal U/S
Treatment: no treatment

ANOMALIES OF ASCENT :

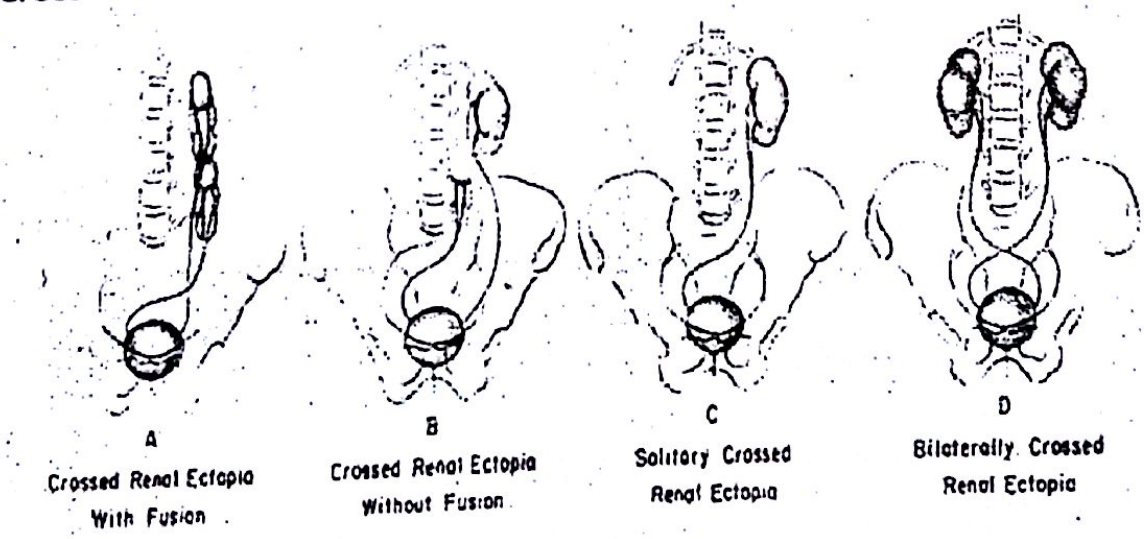
- **Simple Renal Ectopia :** When the mature kidney fails to reach its normal location in the "renal fossa "

Incidence: **The incidence is 1 in 1000**
Associated Anomalies: The incidence of contralateral agenesis appears to be rather high , Hydronephrosis secondary to obstruction or reflux may be seen in as many as 25% of none contralateral kidneys
Clinical features: Most ectopic kidneys are asymptomatic
Diagnosis: **U/S, IVU, 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

- **Cephalad Renal Ectopia**
- **Thoracic Kidney**

ANOMALIES OF FORM AND FUSION :

Crossed Renal Ectopia With and Without Fusion



Horseshoe Kidney

found in 1:1000 necropsies and is more common in men.

probably the most common of all renal fusion anomalies

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 occurs early in embryonic life, so its ascent will be impeded by inferior mesenteric artery.

The kidneys are low located, mal rotated and pelves lie anteriorly

Symptoms:

When present, they are related to complications like hydronephrosis, infection, or calculus formation

diagnosis: US , IVU , CT scan.

Treatment:

Medical: *pain relief and to control infection*

Surgical: *stone removal, PUJ stenosis correction and isthmus division in cases of operations on the aorta*

Prognosis: usually they have normal life.

CYSTIC DISEASES OF THE KIDNEY :

- **Polycystic kidney disease :**

The kidney is one of the most common sites in the body for cysts

Two types:

a. AUTOSOMAL RECESSIVE ("INFANTILE") POLYCYSTIC KIDNEY DISEASE

b. AUTOSOMAL DOMINANT ("ADULT") POLYCYSTIC KIDNEY DISEASE .

1. *Congenital cystic kidney (polycystic kidney) (Adult cystic renal disease)*

Autosomal dominant, transmitted by either parents, 50% of offspring affected.

Both kidneys replaced by large no. of cysts of variable size which make the kidney of large size.

The cysts contain clear fluid but sometimes blood.

The cysts progressively increase in size causing pressure atrophy of the renal parenchyma and pressing the ureter.

15% associated with cystic disease of liver, lung, pancreas or spleen.

Etiology & 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 pictures : Rarely gives clinical manifestation before 4o years

1. ***Asymptomatic:*** diagnosed accidentally.
2. ***Pain:*** due to pedicle stretching, stone, ureteric obstruction, bleeding inside cyst or infection.
3. ***Hematuria:*** cyst distention and rupture to the collecting system.
4. ***Infection:*** renal or cyst infection causes fever, rigor and loin pain.
5. ***Hypertension:*** in 70%, Unknown cause.
6. ***Renal impairment:*** anorexia, headache, nausea, vomiting, , drowsiness and coma.
7. **Renal enlargement:** large knobby palpable kidney.

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

Treatment:

Medical: (Expectant)

To control infection, hypertension, pain and anemia.

Renal impairment: by low protein diet and dialysis.

Surgical:

Rovsing's operation (deroofing) for large cysts causing symptoms or obstruction.

Stone removal.

Renal failure: Renal transplantation.

2. Infantile polycystic disease of the kidney

Rare autosomal recessive, incompatible with life.

Both kidneys are large in size and replaced by large number of cysts which may obstruct labor.

The condition is due to failure of ureteric bud to fuse with metanephrose.

3. ***Simple (solitary) renal cyst***

Common condition . single or multiple . uni 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

Examination: usually –ve, big cyst cause painless loin mass, & painful if complicated by bleeding or infection

U/S: echo free area (cystic lesion).

KUB: soft tissue shadow.

IVU: stretched calyx, filling defect or hydronephrosis.

CT scan &MRI: are diagnostic.

Treatment: usually no treatment needed

Symptomatic cases:

Aspiration and injection of sclerosing agent.

Rovsing's operation (deroofing).

Partial or total nephrectomy in destructed kidney.

N.B. Malignant cyst: radical nephrectomy.

N.B. Hydatid cyst aspiration is contraindicated because of anaphylaxis and dissemination.

CONGENITAL ANOMALIES OF RENAL PELVIS & URETER :

- **Duplication of Renal Pelvis**

Incidence: 4 % , More common on left side Renorenal reflux may occur from one pelvis to the other

- **Duplication of the ureter**

Incidence: 3 % , Usually the ureters fuse & have common orifice in the bladder although they may open independently in which case the ureters cross each other so that the ureter that drain the upper pelvis open below (more distally) in the bladder & vise versa.

Clinical features : usually asymptomatic

More prone to infections, calculus disease & hydronephrosis

treatment : expectant

Ureteral duplication: partial and complete

- **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.

- ***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.

- **Ectopic Ureters**

80% are associated with a duplicated collecting system

In the male, the posterior urethra is the most common site of termination, also to semenal 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: Ureteric reimplantation or implantation of one ureter to the other ureter is used

Ectopic ureters may drain renal moieties (either an upper pole or a single-system kidney) that have minimal function. Therefore, upper pole partial nephrectomy (or nephrectomy of single system) is sometimes recommended .

Complete ureteral duplication and ectopic ureteric orifice.

- **Congenital Megaureter:**

Grossly dilated ureter

Unilateral or bilateral

More common in male

Clinical features:

Asymptomatic, pain, repeated UTIs

Lower ureter might be obstructed

Sometimes associated with

vesicoureteral reflux

Diagnosis : IVU

Treatment :

Infection should be controlled

Excision of the lower stenotic segment (if present)

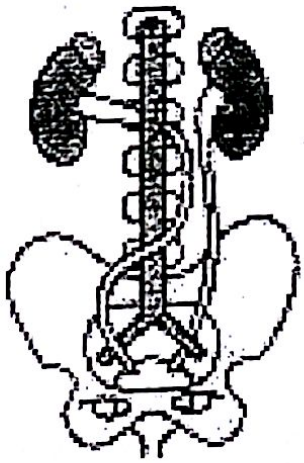
Ureteric tapering & reimplantation into the bladder

Nephroureterectomy for non functioning kidney

Postcaval (*Retrocaval)* ureter (*Preureteral Vena Cava*)

The right ureter pass behind the inferior vena cava

This might causes obstruction



- **Vascular abnormality**

Incidence: about 1 in 1500 , Although it is congenital,most patients present at 3rd or 4th decade.

Diagnosis: IVU

Treatment: surgical correction involves ureteral division, with relocation and ureteroureteral or ureteropelvic reanastomosis, usually with excision or bypass of the retrocaval segment, which can be aperistaltic

- **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, cystoscopy, cystogram

The 'adder head' on excretory urography is typical.

Treatment:

Asymptomatic : no treatment

Cystoscopy with diathermy cauterization of the hole

Nephrectomy in non functioning kidney

In complicated cases, ureteral reimplantation and vesical reconstruction

- **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 , *15% bilateral*

Etiology:

Intraluminal : mucosal fold that causes valve like effect.

Intrinsic (intramural) : interruption in the development of the circular musculature of the UPJ

Extrinsic : An aberrant, accessory , or early-branching lower-pole renal artery

Symptomsl/presentation:

Most infants are asymptomatic and most children are discovered because of their symptoms , Episodic flank or upper abdominal pain, sometimes associated with nausea and vomiting.

Diagnosis:

1. U/S: hydronephrosis.

2. IVU: diagnostic , hydronephrosis with fixed stenotic segment or complete obstruction.

3. CT scan: hydronephrosis that ends abruptly.

4. Magnetic Resonance Imaging.
5. Radionuclide Renography: to see the split function of each kidney.
6. Pressure-Flow Studies :Whitaker test.

Treatment:

Medical : control infection and pain.

Surgical : *Indications for surgery:*

1-progressive hydronephrosis.

2- UTI, and symptomatic patients.

3- Severe hydronephrotic non functioning kidney.

Treatment

SURGICAL REPAIR including open surgical techniques, laparoscopic & endoscopic approaches.

Open & laparoscopic surgical techniques :

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

Endoscopic Approaches **:** balloon dilatation , Antegrade endopyelotomy

Nephrectomy : for non functioning kidney