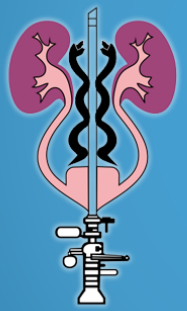
Urogenital system anomalies



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Common congenital anomalies

Common Renal anomalies

- Abnormal number: agenesis
- Abnormal form or position: horseshoe kid.

Common Bladder anomalies

- Bladder Extrophy.

Common Urethral & penile anomalies

- Hypospadias.
- Epispadias.

Common ureteral & renal pelvis anomalies

- UPJ obstruction.
- Vesico-uretral reflux.
- Duplication.
- Uretrocele.
- Ectopic ureter.

Common genital anomalies

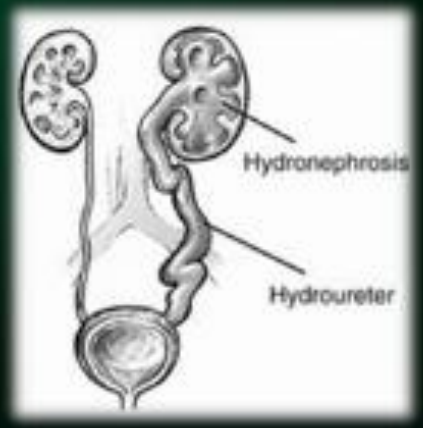
- Undescended testis.
- Intersex (disorders of sexual differentiation).

congenital diseases of the kidney

- Hydronephrosis
  - Obstructive(UPJ,UVJ obstruction, duplication)
- Renal agenesia
  - Multicystic dysplastic kidney
- Polycystic kidney
- Horshoe kidney
- Renal dysplasia
- Renal hypoplasia
- Vascular abnormalities of the kidney

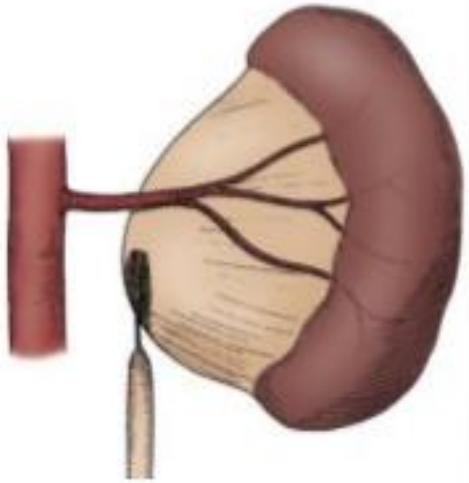
a) Hydronephrosis

It is the dilatation of the renal pelvis which may be found as unilateral or bilateral. It may be due to obstruction of urine flow in the distal urinary tract or reflux of urine up the ipsilateral ureter or due to bladder neck obstruction or urethral obstruction.



Pelviureteric junction obstruction

Obstruction of the junction between the renal pelvis & ureter.

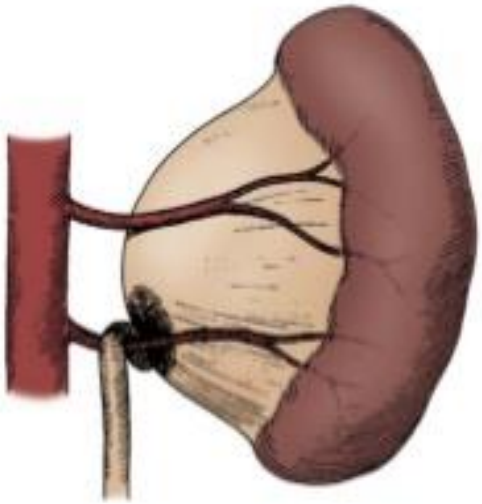


Aetiology

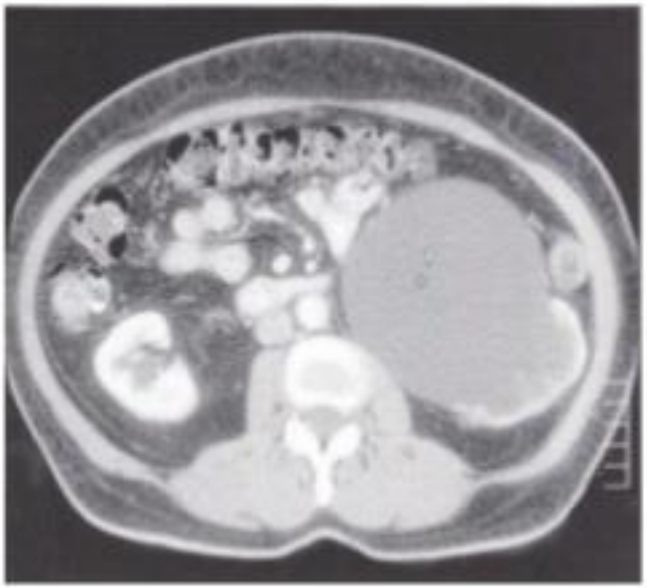
- aperistaltic segment of ureter due to absent muscles.

or

- crossing vessels over UPJ.



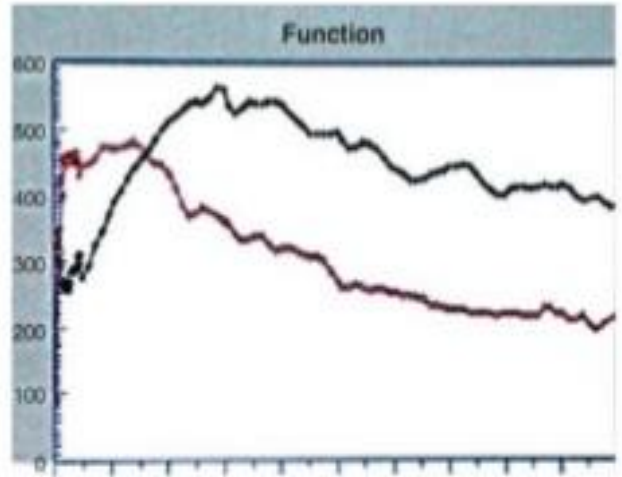
Pelviureteric junction obstruction- *Presentation*



- may present at any time (before birth, in childhood, or in adulthood) by:
- abdominal mass.
- abdominal pain.
- Haematuria after fairly minor abdominal trauma.

Pelviureteric junction obstruction- *Evaluation*

- ***IVU*** shows delay in appearance of contrast and dilated renal pelvis and calices.
- ***Renal scan*** shows differential renal function and confirms obstruction.



Pelviureteric junction obstruction- *Treatment*

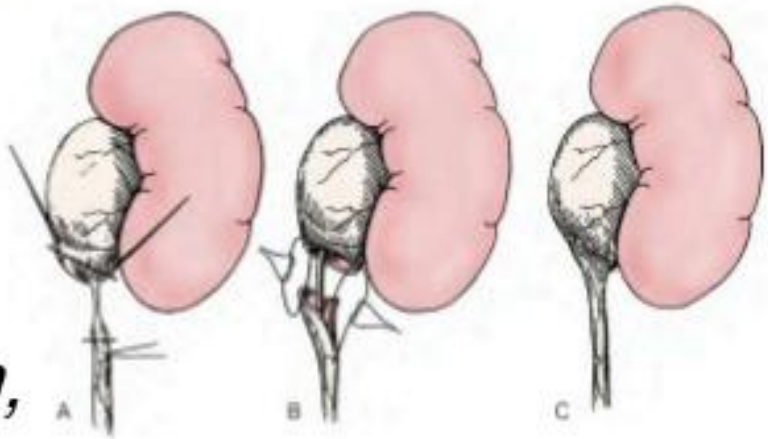
- *Surgery is indicated for:*

  1. obstructive symptoms,

  2. stone formation,

  3. recurrent urinary infection,

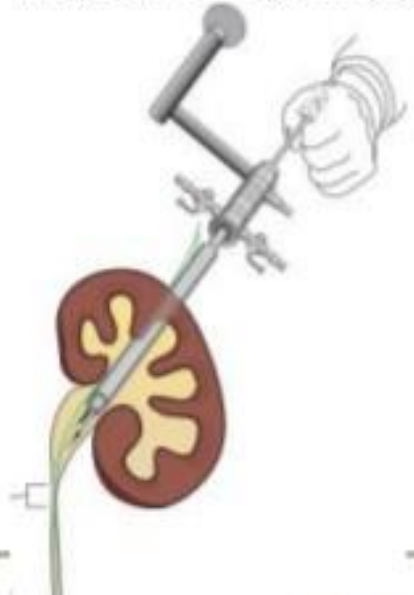
  4. progressive renal impairment.



- **Pyeloplasty** is the treatment of choice
- **Nephrectomy** is performed if the affected kidney is <10% of total renal function.

Pelviureteric junction obstruction

- Minimally invasive alternative techniques:
  1. ***antegrade endopyelotomy.***
  2. ***Laparoscopic pyeloplasty is becoming more used now.***

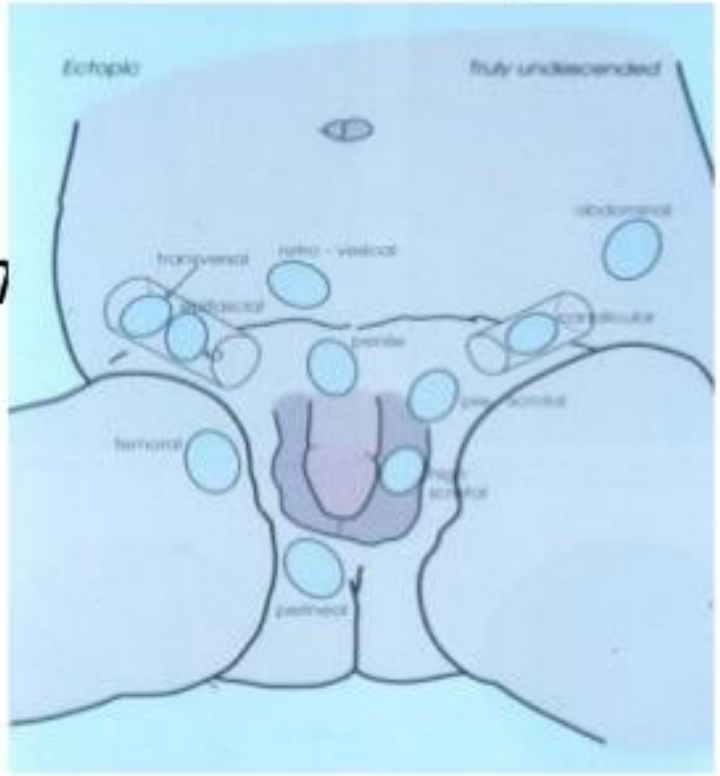


Undescended testis

- The incidence ranges from 3.4% to 5.8% in full term boys
- It decreases to 0.8% in one year old boys.
- Cause is unclear, but androgens may have an important role.

Undescended testis vs Ectopic

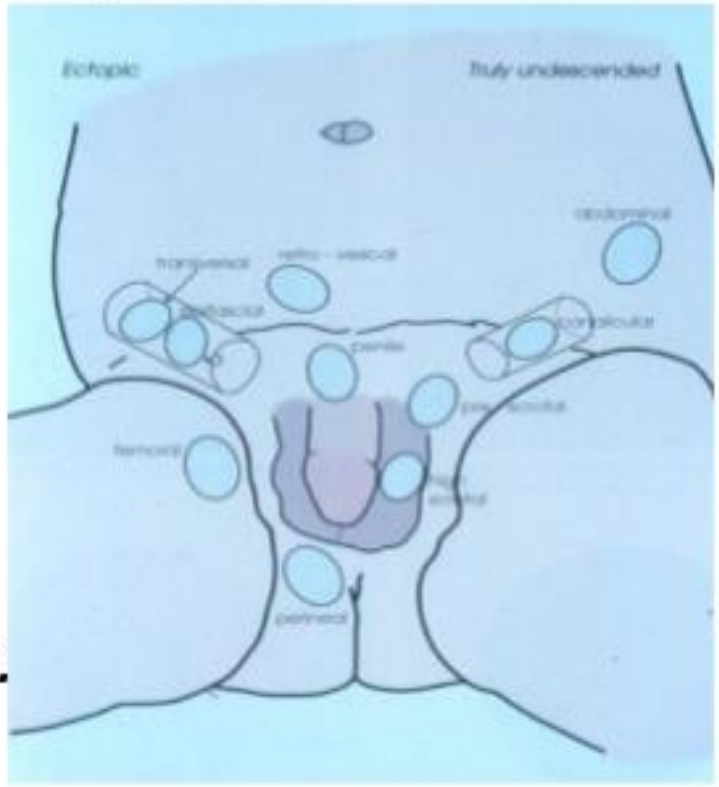
- ***Undescended testis*** can be classified by its ***location***:
  1. *upper scrotum,*
  2. *superficial inguinal pouch*
  3. *inguinal canal,*
  4. *abdomen.*



- In 80% of cases, the undescended testis will be palpable in the inguinal canal.

Undescended testis vs Ectopic

- ***Ectopic testis*** *(testis outside path of normal descent)* may be:
  1. *penile,*
  2. *femoral,*
  3. *perineal,*
  4. *retrovesical.*
  5. *Superficial inguinal pouch.*



Undescended testis-
Complications

- Patients with undescended testes have two major concerns:

  1. increased incidence of testicular cancer: up to 10 times higher than in a normal testis
  2. subfertility.

- Spontaneous descent of undescended testis is rare after the age of one year.

Undescended testis-

- ***Examination*** (esp, under anesthesia) is more accurate than imaging.
- ***Laparoscopy*** is the **investigation of choice** for non-palpable testes to differentiate intra-abdominal from vanished from inguinal testis.
- ***Imaging:*** *Ultrasound, CT, MRI* (inaccurate).
- *chromosomal and endocrine evaluation* if bilateral non-palpable testes.

Undescended testis- *Management*

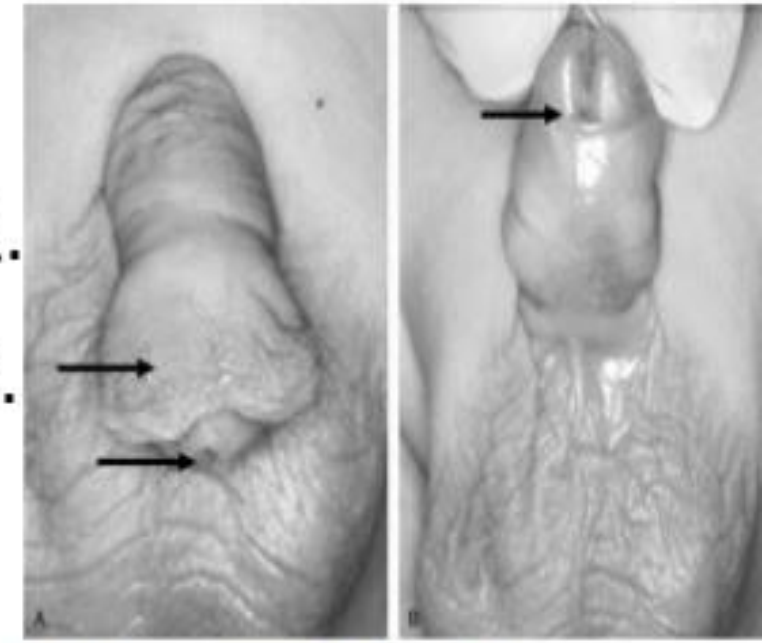
- ***Orchidopexy*** if testis is felt in inguinal canal or below, it should be performed as soon as possible.
- ***Orchidectomy*** If an atrophic intra-abdominal testis is detected especially after puberty, as the testis is incapable of spermatogenesis and the risk of malignancy.

Retractile testis

- Commonly confused with undescended testis.
- the testis can be delivered into bottom of scrotum.
- low undescended testis will immediately pop back to its undescended position after being released.

Hypospadias

- congenital condition results in underdevelopment of urethra.
- affects 3 per 1000 male infants.
- Consists of 3 anomalies:



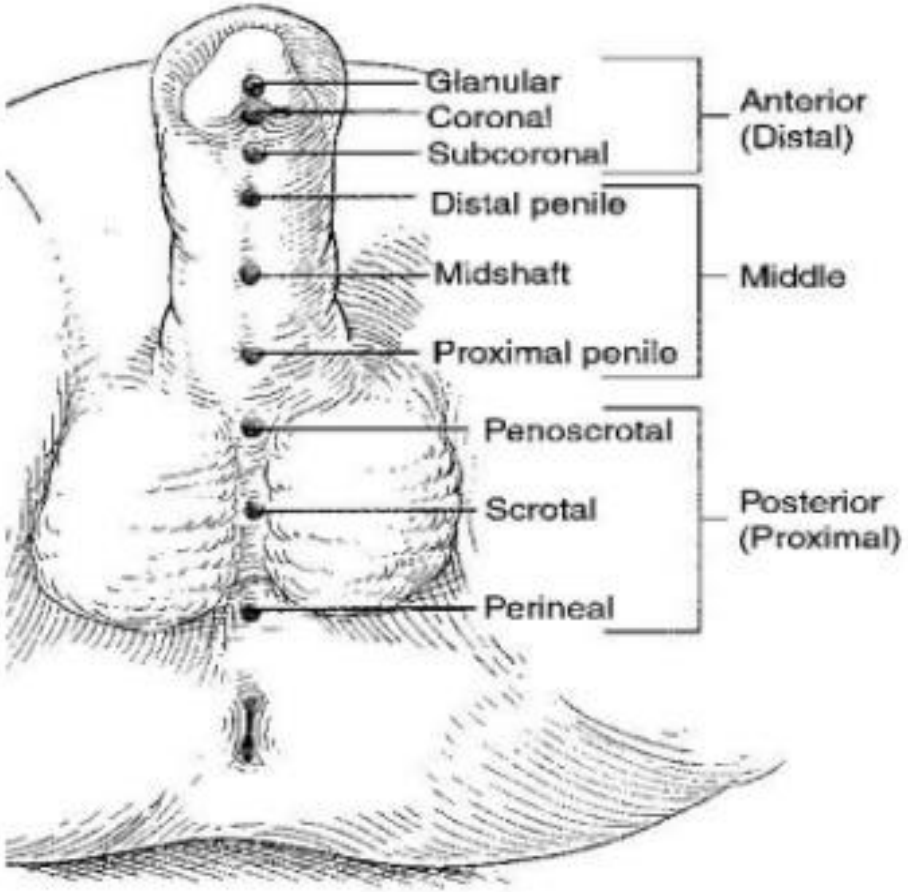
(1) Abnormal ventral opening of the urethral meatus.

(2) Ventral curvature of the penis (chordee).

(3) Deficient prepuce ventrally.

Hypospadias- *Evaluation*

- Site Of the meatus.
- Circumcised or not.
- Associated anomalies:
  meatal stenosis, or
  undecended testis.
- Penile curvature.



Hypospadias- *Examples*

Hypospadias- *Treatment*

- The child should be referred for urological assessment and surgical treatment.
- The ideal age for surgery is 6–12 months.

RENAL AGENESIS

- Bilateral: it is very rare(<400 cases)
  - Not survivable
  - Olygohydramnios
  - Pulmoner hypoplasia
  - Facial deformities(Potter face)

UNILATERAL RENAL AGENESIS

- Ureteric bud did not develop or contact with metanephric blastema
- Ureter does not exist on the ipsilateral side in 50% of the cases.
- Blind ending ureter may persist
- Usually asymptomatic and may be associated with the other anomalies such as cardiac, vertebral column, long bones, hands, genital and anal anomalies,

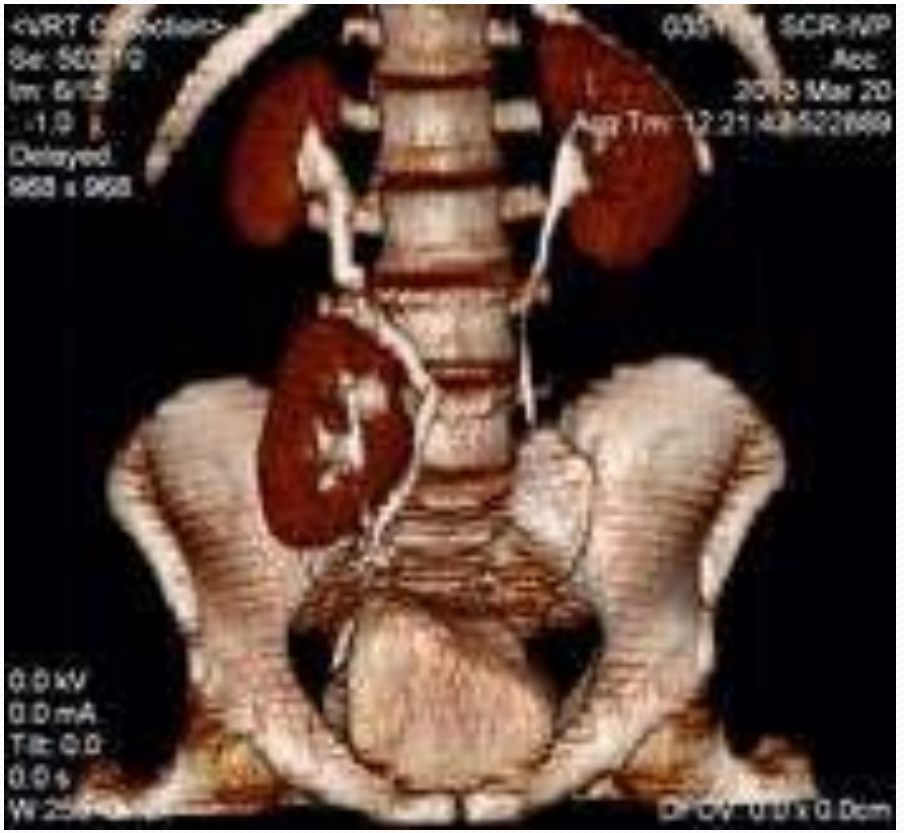
Fig. 3.3 Renal agenesis. Note the aborted right renal artery (arrow).

RENAL HYPOPLASIA

- Congenitally small kidney
- Dysplasia may coexist
- Incidence may increase in case of antenatal exposure to cocaine and alcohol
- Differential diagnosis includes renal artery stenosis or pyelonephritic atrophic kidney
- Renal artery has a small diameter which may cause hypertension(nephrectomy is the treatment of choice)

Supernumerary kidneys

- It is a very rare congenital anomaly
- Fourth kidney has been reported only once
- Complete duplication is a different entitiy

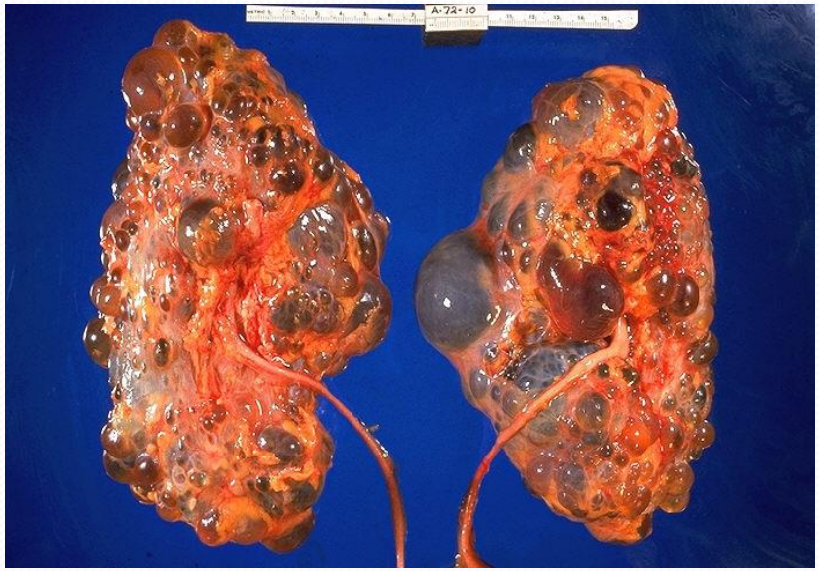


DYSPLASIA AND MULTICYSTIC KIDNEY

- MULTICYSTIC DYSPLASIA
  - Usually one sided
  - Not inherited
  - Characterized by an irregular cystic mass
  - Ureter is absent or atresic
  - Collector tubes and nephron are not united
  - Treatment is usually not necessary but follow-up is required because of malignity potential
  - Contralateral renal and ureteral anomalies may coexist(i.e., UPJ obstruction)

ADULT POLYCYSTIC KIDNEY

- Autosomal dominant inheritance, 95% bilateral
- Symptoms appear after 40 years of age
- Liver, spleen and pancreas cysts may coexist
- Collector and distal tubules are not connected. Cysts are developed because of continuing glomerular secretion
- The fluid inside the cysts is amber coloured



DIAGNOSIS

- SYMPTOMS AND SIGNS
  - Pain(tension on the vascular pedicle, obstruction, infection, distension of cyst walls due to bleeding
  - Hematuria
    - Microscopic or macroscopic
  - Renal colic
  - Abdominal mass
  - Tenderness of costovertebral angle
  - Hypertension
  - Proteinuria
  - Chills, fever and flank pain(infection!!)
  - Lower urinary tract symptoms
  - Signs and symptoms of renal failure
    - Fatigue, nausea, vomiting, headache, weigt loss, anemia, hyperazotemia, proteinuria, edema, ascites, pericardial effusion, congestive heart failure, hypertension, electrolyte disturbances, hypertensive retinopathy, uremic encephalopathy

LABORATORY FINDINGS

- Anemia(chronic blood loss or bone marrow toxicity due to uremia)
- Proteinuria
- Microscopic or macroscopic hematuria
- Pyuria
- Bacteriuria
- Low urine density(due to the decrease in concentration ability)
- Creatinine clearance is decreased
- Patients already have renal failure when first presented

RADIOLOGICAL FINDINGS

- Renal contours are enlarged on KUB
- Spider deformity on IVU due to cyst pressure on calyces
- Enlarged kidneys with multiple cysts on USG
- Computed tomography better delineates stones, tumors
- No uptake of radionuclide by the cysts on scintigraphy “cold spots”

COMPLICATIONS

- Pyelonefritis(may be asymptomatic)
- Infection (pain, fever, chills)
- Life threatening macroscopic hematuria
- Renal colic
- Hypertensive retinopathy, encephalopathy
- Anemia
- Chronic renal failure

TREATMENT

- Treatment is usually supportive and conservative
- General measures:
  - Diet (protein restriction 0.5-0.75 g/kg/day)
  - Forced diuresis(3000 ml/day)
  - Restriction of strenuous exercise
  - Control of blood pressure
  - Hemodialysis in the presence of chronic renal failure
- Surgery: cyst resection, renal transplantation
- Treatment of complications: pyelonephritis, drainage of infected cyst, blood transfusion, embolization, stone treatment

SIMPLE CYST

- usually solitary and one sided
- It is not clear whether it is congenital or acquired
- It is common in the patients on chronic hemodialysis
- It may cause obstruction, infection and deterioration of renal function
- It is usually located at the lower pole and becomes symptomatic after 10 cm
- Cyst fluid is amber coloured. Hemoragic cyst is found in 5% and half of them harbour papillary tumors
- They have no connections with the collecting system(contrary to the calyceal diverticula)
- Multilocular cyst may be misdiagnosed as tumors
- USG is helpful for differential diagnosis

CLINICAL FINDINGS

- Symptoms:
  - Pain
  - Gastrointestinal upset
  - Mass
  - Fever, fatigue and pain if it is infected
- Signs:
  - mass
  - CVA tenderness

LABORATORY FINDINGS

- Urinalysis is usually normal
- Microscopic hematuria is not common
- Renal functions are normal

- X-Ray findings:

  - Bulging in renal contour on KUB, a change in renal axis, calcification in the cyst wall
  - IVU: space occupying lesion, distorsion of renal pelvis and calyces, extrinsic obstruction of ureter,
  - CT: the best method to differentiate the tumor and cyst. Density of the cyst fluid is the same with water
- USG, Scintigraphy, cystography and aspiration help for differential diagnosis

DIFFERENTIAL DIAGNOSIS

- Tumor
- Polycystic kidney
- Renal carbuncle
  - History of skin infection a couple of weeks ago, pain and CVA tenderness)
- Hydronephrosis
- Extrarenal tumor
- Hydatic cyst(usually calcified)

ANALYSIS OF CYST FLUID

- Cytology
- Lipid, protein content
- LDH<250 mIU
  - I f the cyst fluid is hemorrhagic and rich of protein, lipids with elevated LDH, tumor should be suspected
- Microbiological investigation

COMPLICATIONS

- Cyst becomes infected
  - Bleeding inside the cyst
  - Obstruction
  - Hydronephrosis
  - Pyelonephritis
  - Hypertension
- Treatment:

Specific measures:

Cyst puncture and sclerotherapy(iophendylate , ethanol 95% injection)

Cystectomy

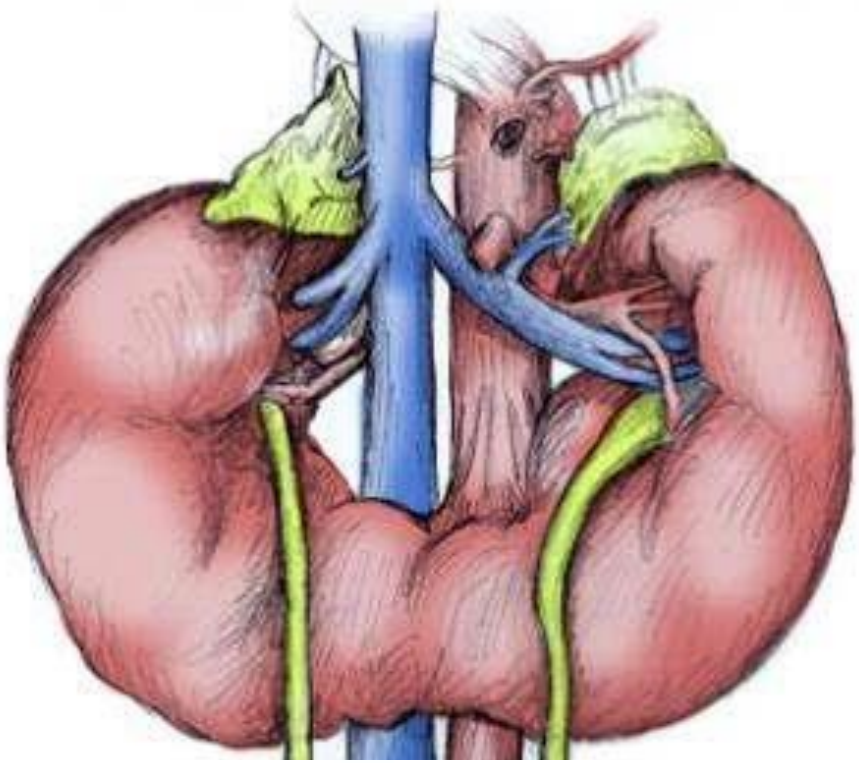
Laparoscopic

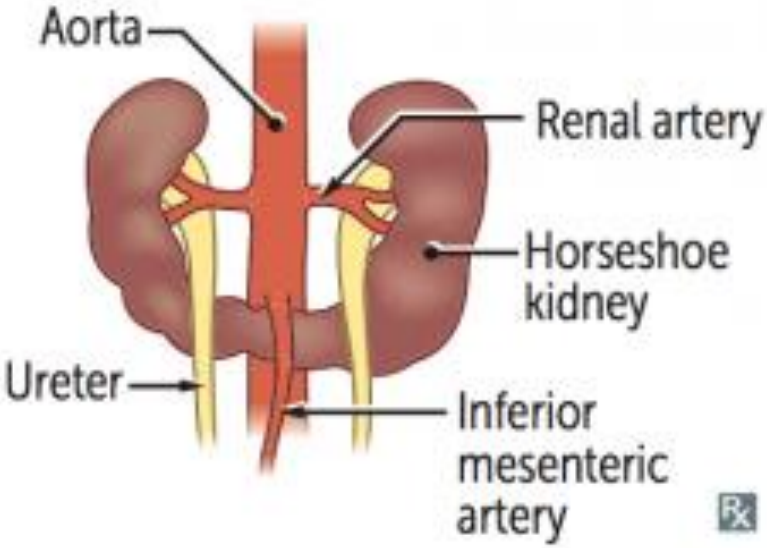
Exploration

Treatment of complications

RENAL FUSION

- Renal fusion anomalies is seen 1/1000 and most of them are horseshoe kidneys
- Even if the fused kidneys are located on the same side, ureteral openings are normal
- Kidneys are located below and have variable vasculature
- In the patients who have both fusion and ectopy, extraurinary and the other urinary abnormalities are seen 78% and 65% respectively




Aorta
Renal artery
Horseshoe kidney
Ureter
Inferior mesenteric artery

PATHOLOGY

- The kidneys are not normally rotated
- Pelvis located anteriorly and ureters pass over isthmus
- Aberrant vasculature and course of ureters may result in obstruction and hydronephrosis
- Stone and infection may supervene
- Vesicoureteral reflux is more common
- The kidneys are usually fused at the lower pole, the axis is vertical
- More advanced form of fusion is a cake kidney

CLINICAL FINDINGS

- Symptoms
  - Usually asymptomatic
  - Pain, fever, chills if there is infection or stone
  - Gastrointestinal upset and renodigestive reflex
- Signs:
  - Palpable mass
  - Rovsing sign is also used in patients with horseshoe kidney, consisting of abdominal pain, nausea, and vomiting with hyperextension of the spine.

LABORATORY FINDINGS

- Urinalysis is usually normal unless infection is present
- Renal functions are usually normal
- X-Ray: renal axes are parallel to the spine
  - IVU : the pelvises are anteriorly located, lower calyces may across the other side with the ureters superimposed.
    - Cross ectopy : kidneys are located on the same side as the renal pelvises and ureters, however, one of the ureters crosses over to the other side
    - Cake kidney may exert pressure on the bladder when it is located in the pelvis
  - CT: reveals more detailed information

COMPLICATIONS

- Ureteral obstruction
- Hydronephrosis
- Stone development
- Infection

TREATMENT is directed towards complications

ECTOPIC KIDNEY

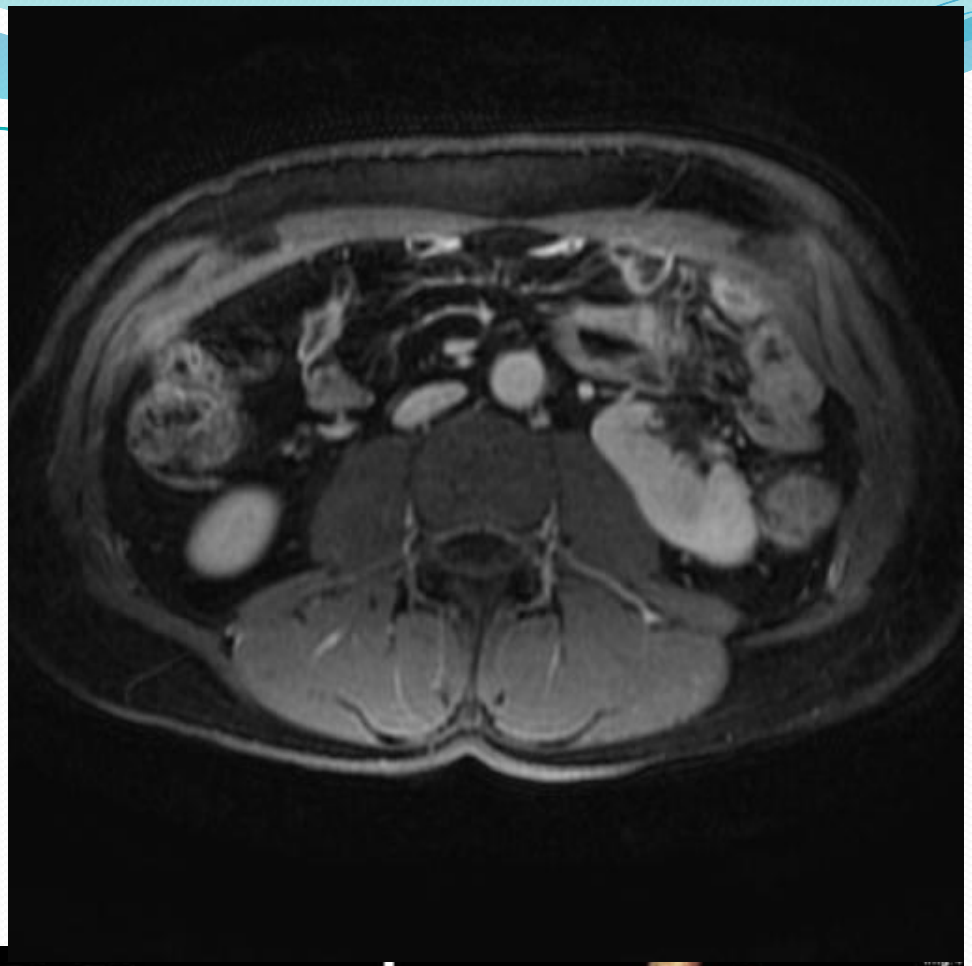
- asymptomatic unless obstruction or infection supervene

- 1-Simple ectopia: kidney is on the same side but at a lower level because of incomplete ascend.
- It differs from nephroptosis or acquired ectopia by the length of its ureter correlated with the kidney's location
-
- 2- Cross ectopia without fusion : both kidneys are on the same side

Fig. 3.15 Pelvic kidney, **a** KUB, **b** tomography, **c** retrograde pyelogram.

ROTATION ANOMALIES OF THE KIDNEY

- Malrotation rarely causes problems



MEDULLARY SPONGE KIDNEY

- Cystic dilatation of renal collector tubules.
- Autosomal recessive
- Often bilateral, occasionally unilateral, and/or involving one papilla
- Cystic dilatation is found in tubules, prone to infection and stone development
- Symptoms are related to the infection or stones
- Diagnosis is made by IVU, renal pelvis and calyces are normal. Dilated tubules may be filled up with contrast material.
- Treatment is directed towards complications which are rare.

ABNORMALITIES OF THE RENAL VESSELS

- Solitary renal artery is seen in 75-85%.
- Abberant arteries may cause infundibular or ureteropelvic junction obstruction
- Aberrant veins are less common