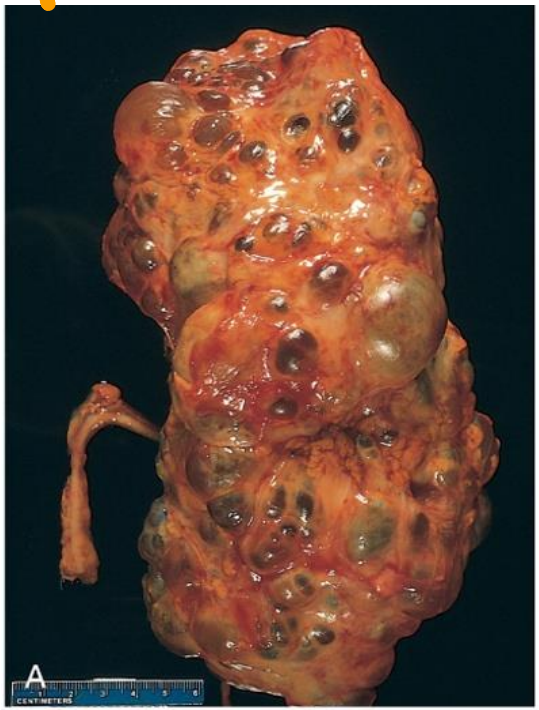
Modified by: Jaysal Massad

THE UNIVERSITY OF JORDAN

CYSTIC DISEASES of THE KIDNEY

Dr. Nisreen Abu Shahin



cysts are dilated tubules usually due ↑ pressure or due to other causes

Types of cysts

1-Simple Cysts

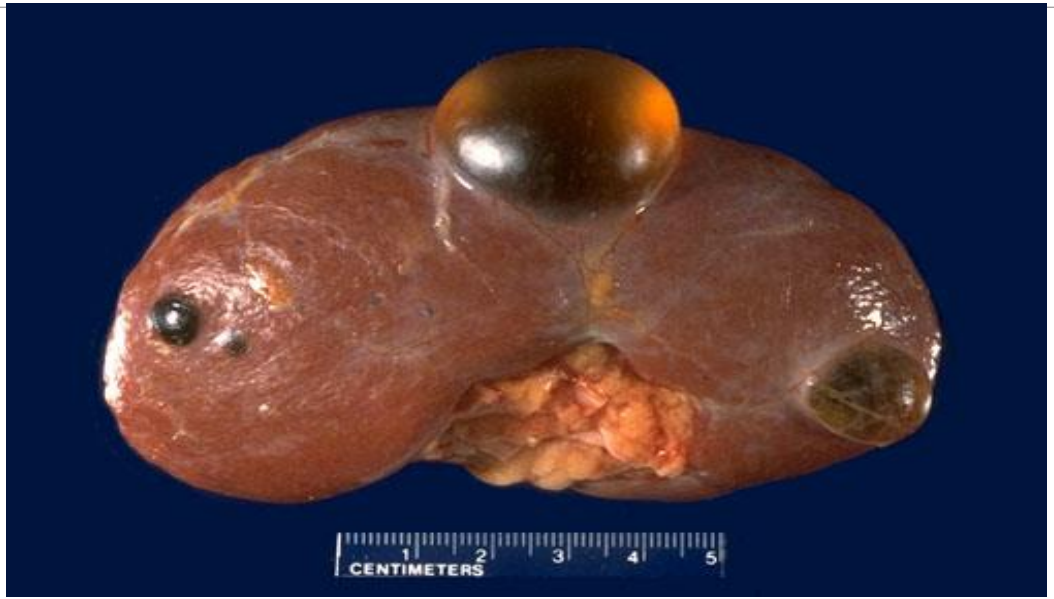
2-Dialysis-associated acquired cysts

3-Autosomal Dominant (Adult) Polycystic Kidney Disease

4-Autosomal Recessive (Childhood) Polycystic Kidney Disease

5-Medullary Cystic Disease

Simple renal Cysts



1-Simple Cysts

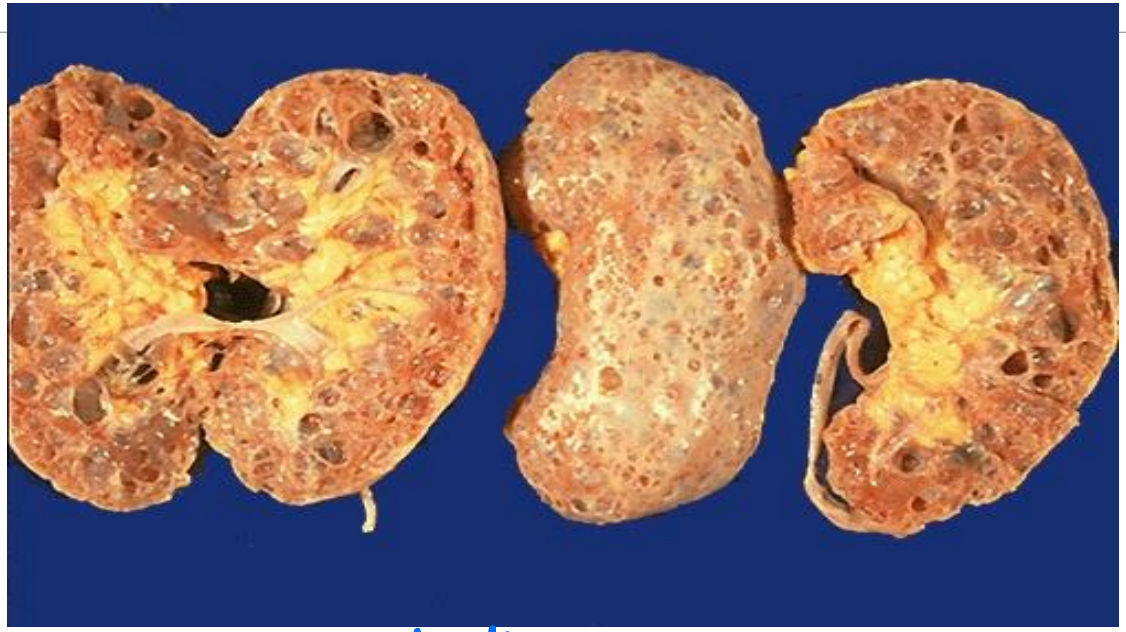
- Multiple or single
- 1-5 cm in diameter
- filled with clear fluid.
- confined to the cortex.
- no clinical significance.
- Usually discovered incidentally or because of hemorrhage and pain
- Importance: to differentiate from kidney tumors

Follow up by imaging techniques

Not very Serious ⇒ mild Complications

Patients with CRF ⇐ مرضى

Cysts associated with chronic dialysis



Possible Complications:

1) ↑ risk of infection
2) . rupture of cyst → hemorrhage
3) stone formation
4) malignancy inside cyst.

2-Dialysis-associated acquired cysts

⇒ * Those cysts are significant *

- in patients with renal failure who have prolonged dialysis.
- both cortex and medulla multiple cells / variable in size
- Complications: hematuria; pain . may be numerous
- Increased risk of renal carcinomas (100 times greater * than in the general population)

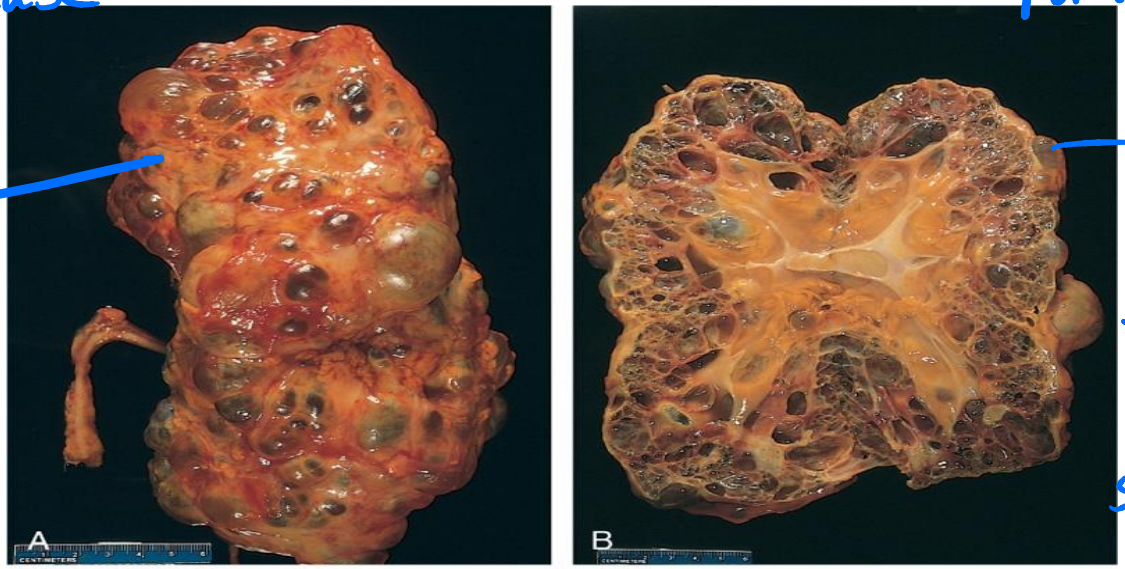
abnormal function / inflammation ⇒ ↑ Growth factor secretion ⇒ tubular epithelial proliferation

* kidney transplantation is the optimum solution *

Autosomal Dominant (Adult) Polycystic Kidney Disease

Genetic Cause

* onset of disease * manifestations



© Elsevier. Kumar et al: Robbins Basic Pathology 8e - www.studentconsult.com

* weight = 1.5 kg

normal 150 g

→ cortex is lost & parenchyma lost so function ↓ so CRF & urine impairment

* # hence causing abdominal masses

3- Autosomal Dominant (Adult) Polycystic Kidney Disease

- multiple bilateral cysts
- eventually destroy the renal parenchyma.
- Incidence (1: 500-1000) persons
- 10% of chronic renal failure.
- inheritance of one of 2 autosomal dominant genes:
- (1)- *PKD1:* 85-90% (encodes polycystin-1)
- (2)- *PKD2* :10-15% (encodes *polycystin- 2).*

i.e → disease was always found.

during adulthood

3-Autosomal Dominant (Adult) Polycystic Kidney Disease – cont.

Clinical presentation :

- ***asymptomatic until the 4th decade*** due to slow progression
- **Symptoms: *flank pain* , heavy dragging sensation, abdominal mass, hemorrhage, obstruction, *Intermittent gross hematuria***

Complications

1- ***hypertension*** ***(75%)***

2- ***urinary infection*** → small % of Patients

3- **vascular aneurysms of circle of Willis (10% -30%)→ (subarachnoid hemorrhage).**

↳ Berry ??

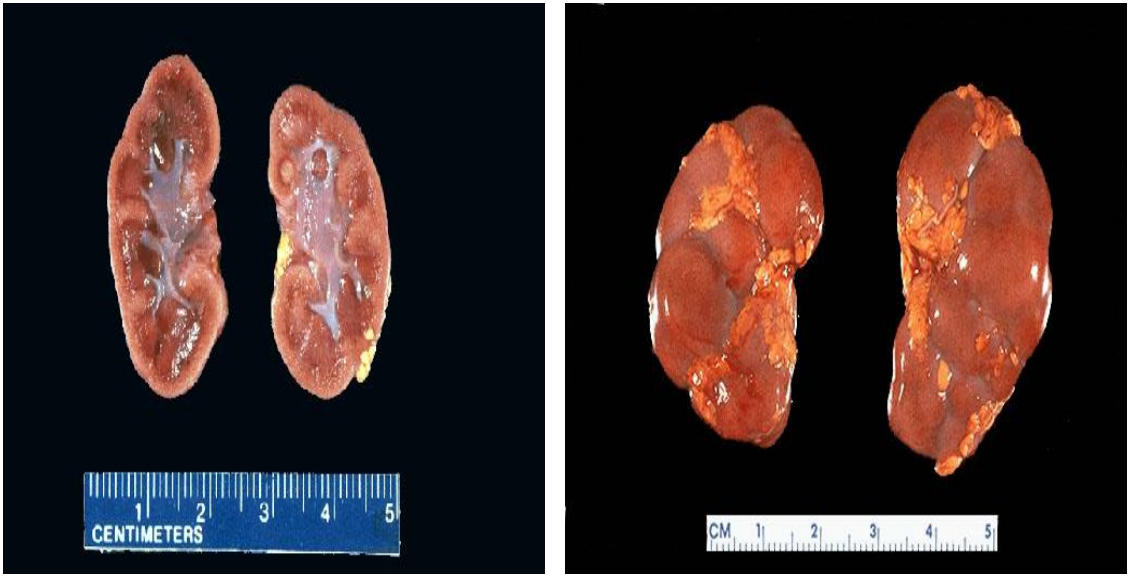
4- **renal failure at age 50**

Transplantation is needed.

4-Autosomal Recessive (Childhood) Polycystic Kidney Disease

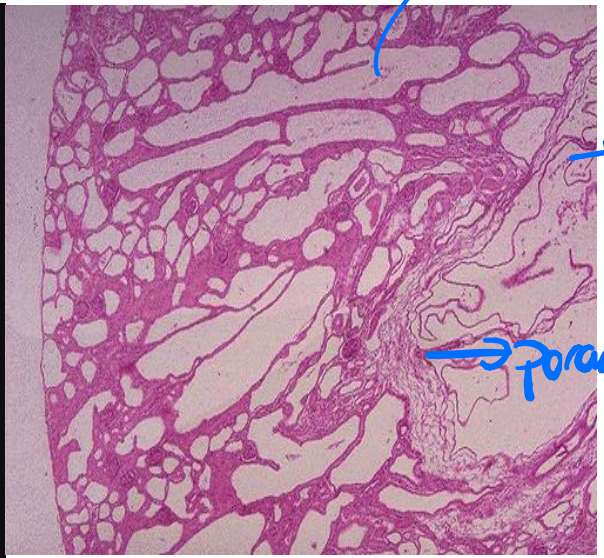
- autosomal recessive
- 1:20,000 live births.
- Types: perinatal, neonatal, infantile, and juvenile.
- Associated with liver cysts
- Mutations in ***PKHD1*** gene coding for ***fibrocystin***.
- **Fibrocystin** may be involved in the function of cilia in tubular epithelial cells .

Normal term infant kidneys



Autosomal Recessive (Childhood) Polycystic Kidney Disease:

Treatment is also transplantation



space of cyst

→ cortex & medulla

→ parenchymal loss

Cysts only in medulla.

5- Medullary Cystic Disease

- 2 major types:

usually without dangerous Clinical effects

1-medullary sponge kidney

- common and innocent condition.

2-nephronophthisis-medullary cystic disease complex → Genetically determined by many genes

kidney wasting

- almost always associated with renal dysfunction.
- usually begins in childhood.
- Cysts are at cortico-medullary junction

Tubules & interstitial abnormalities

5- Medullary Cystic Disease

- **Clinical features:**
- polyuria and polydipsia (↓tubular function). ⇒ diluted urine
- renal failure over 5-10-year
- A positive family history and unexplained chronic renal failure in young patients should lead to suspicion of medullary cystic disease.

tubular Atrophy and interstitial fibrosis leading to Renal failure

URINARY OUTFLOW OBSTRUCTION

↳ an obstruction anywhere in the outflow track

The most important.

•Renal Stones (*Urolithiasis*)

- stone formation at any level in the urinary collecting system.
- Most common in kidney.
- (1%) of all autopsies. → Can be asymptomatic
- Symptomatic more common in men
- Familial tendency toward stone formation
- unilateral in 80%
- Variable sizes

Stone = inorganic salt (98%) + organic matrix (2%)

→ minerals ex. Ca^{++}

← from cells or bacterial colonies

❖ Types are according to inorganic salt:

1- calcium oxalate/ calcium oxalate+ calcium phosphate-- (80%) .

2- Struvite (magnesium ammonium phosphate) (<10%)

3- uric acid (6-7%)

4- cystine stones (2%) ← least Common

Causes of Renal Stones

1-increased urine concentration of stone's constituents exceeds solubility in urine (supersaturation).

- 50% of *calcium stones* pts have hypercalciuria with no hypercalcemia.
- 5% to 10% → hypercalcemia and hypercalciuria.

2-The presence of a nidus

- Urates provide a nidus for calcium deposition.
- Desquamated epithelial cells
- Bacterial colonies

3-urine pH

- *Magnesium ammonium phosphate (struvite) stones* occur with alkaline urine due to UTIs.
- Uric acid stones form in acidic urine (under pH 5.5).

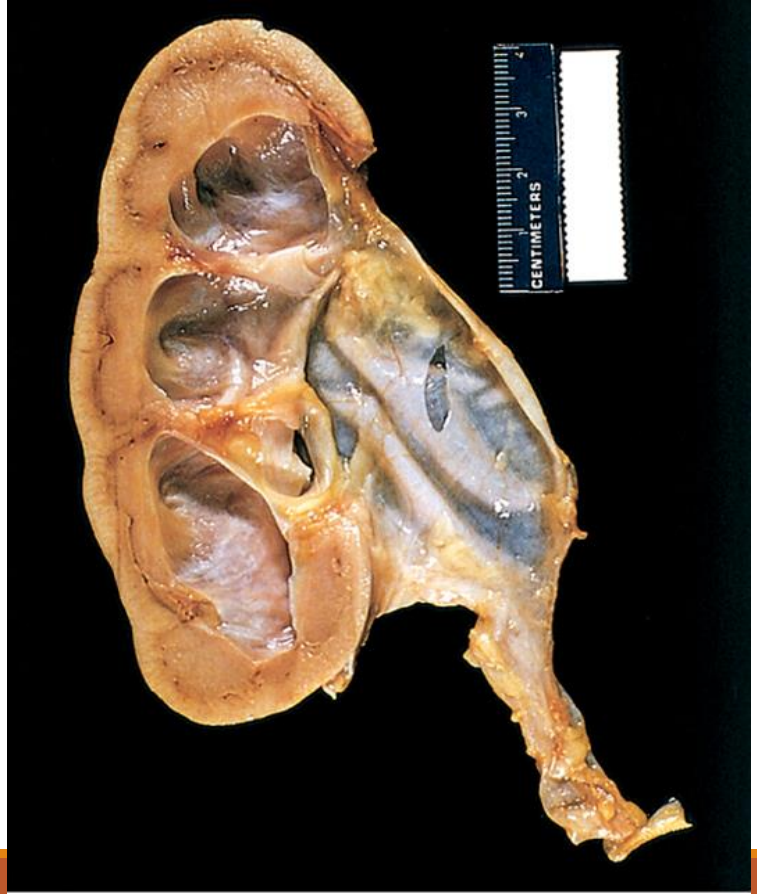
4-infections

urea-splitting bacteria (*Proteus vulgaris* and staph).

Hydronephrosis

Hydronephrosis

- dilation of the renal pelvis and calyces due to obstruction, with accompanying atrophy of kidney parenchyma.
- sudden or insidious
- Obstruction at any level from the urethra to the renal pelvis.
- The most common causes are :



Hydronephrosis of the kidney, with marked dilation of the pelvis and calyces and thinning of renal parenchyma.

1-Congenital:

examples

- Atresia of urethra
- Valve formations in ureter or urethra
- Aberrant renal artery compressing ureter
- Renal ptosis with torsion or kinking of ureter

2-Acquired:

- ***Examples:***

- **Foreign bodies**: Calculi, necrotic apillae · Stones
- **Tumors**: prostatic hyperplasia, prostate cancer, bladder tumors, cervix or uterus cancer.
- **Inflammation**: Prostatitis, ureteritis, urethritis,
- **Neurogenic**: Spinal cord damage
- Normal **pregnancy**: rare, mild and reversible