THE UNIVERSITY OF JORDAN

CYSTIC DISEASES of THE KIDNEY

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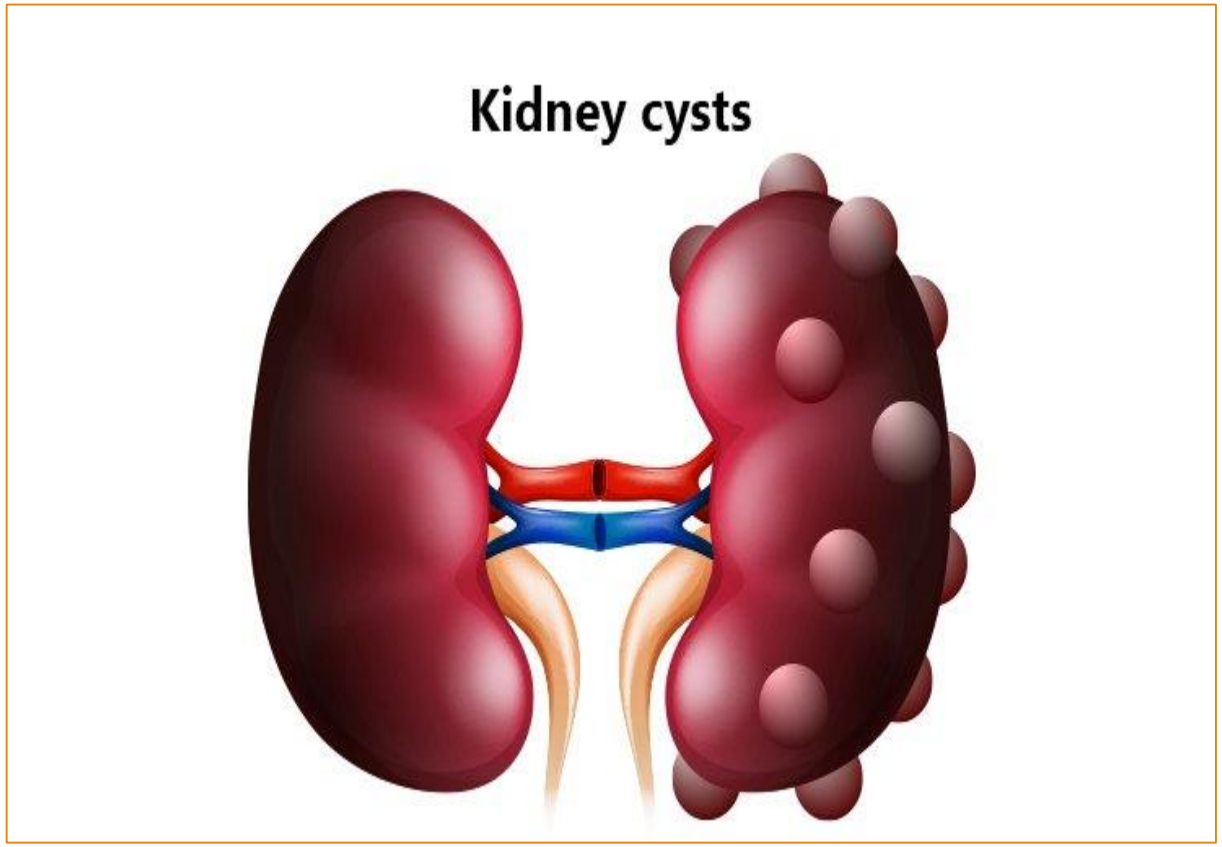


Types of cysts

1-Simple Cysts

2-Dialysis-associated acquired cysts

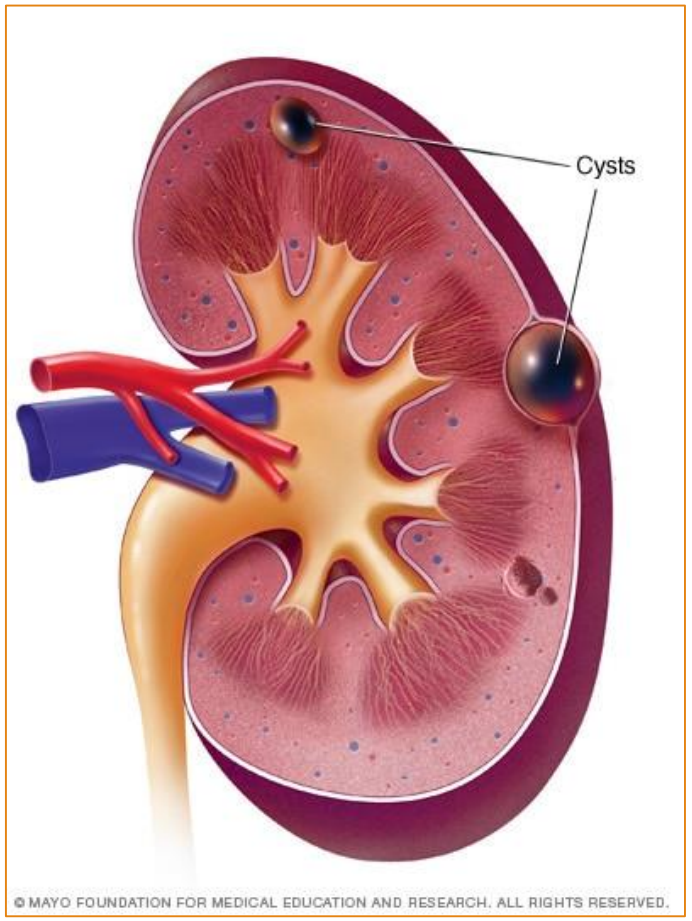
3-Autosomal Dominant (Adult) Polycystic Kidney Disease

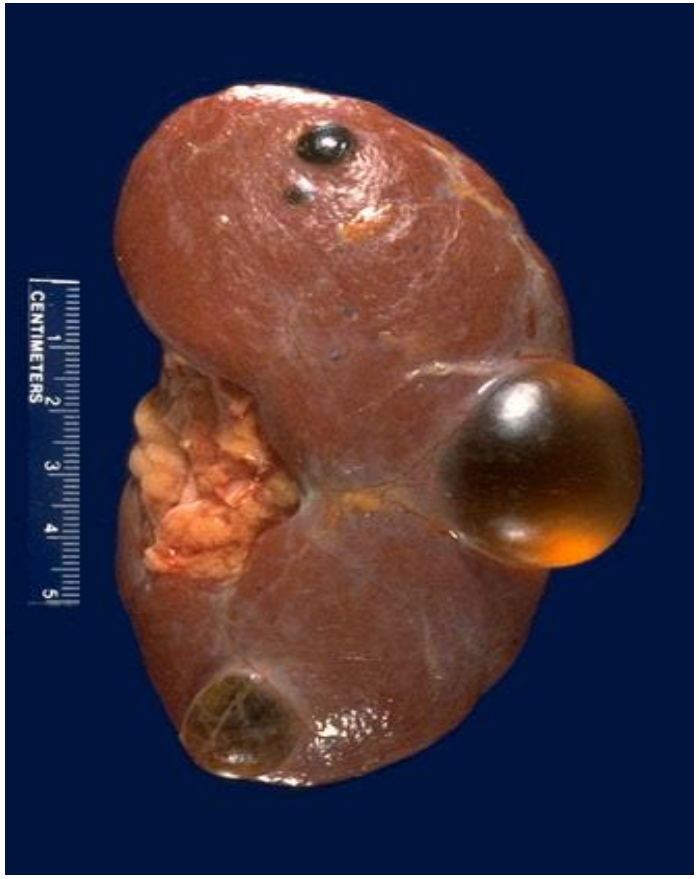
4-Autosomal Recessive (Childhood) Polycystic Kidney Disease

5-Medullary Cystic Disease

1- Simple Renal Cysts

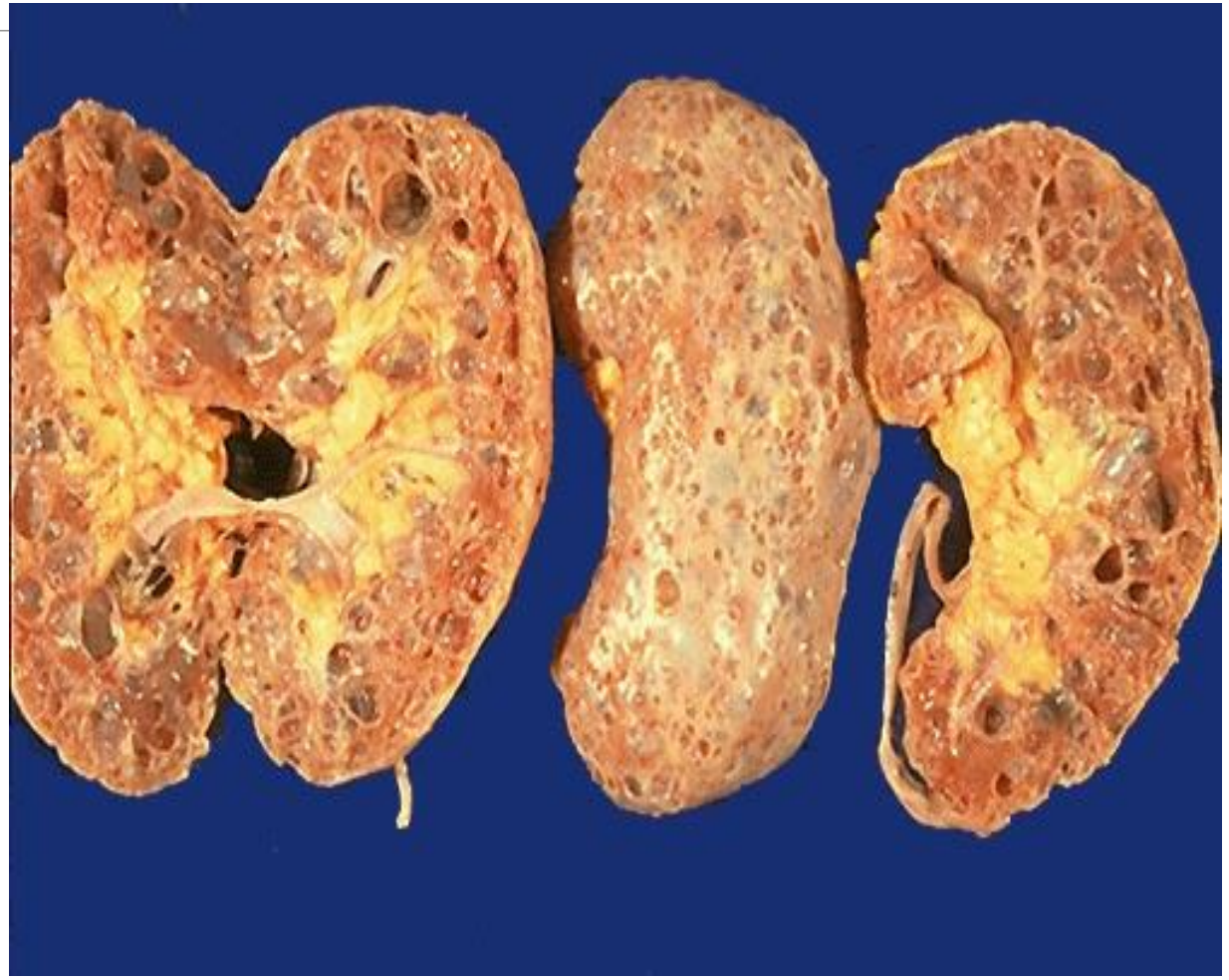
- Multiple or single
- 1-5 cm in diameter
- filled with clear fluid.
- confined to the cortex.
- no clinical significance.
- Usually discovered incidentally or because of hemorrhage and pain
- Importance: to differentiate from kidney tumors





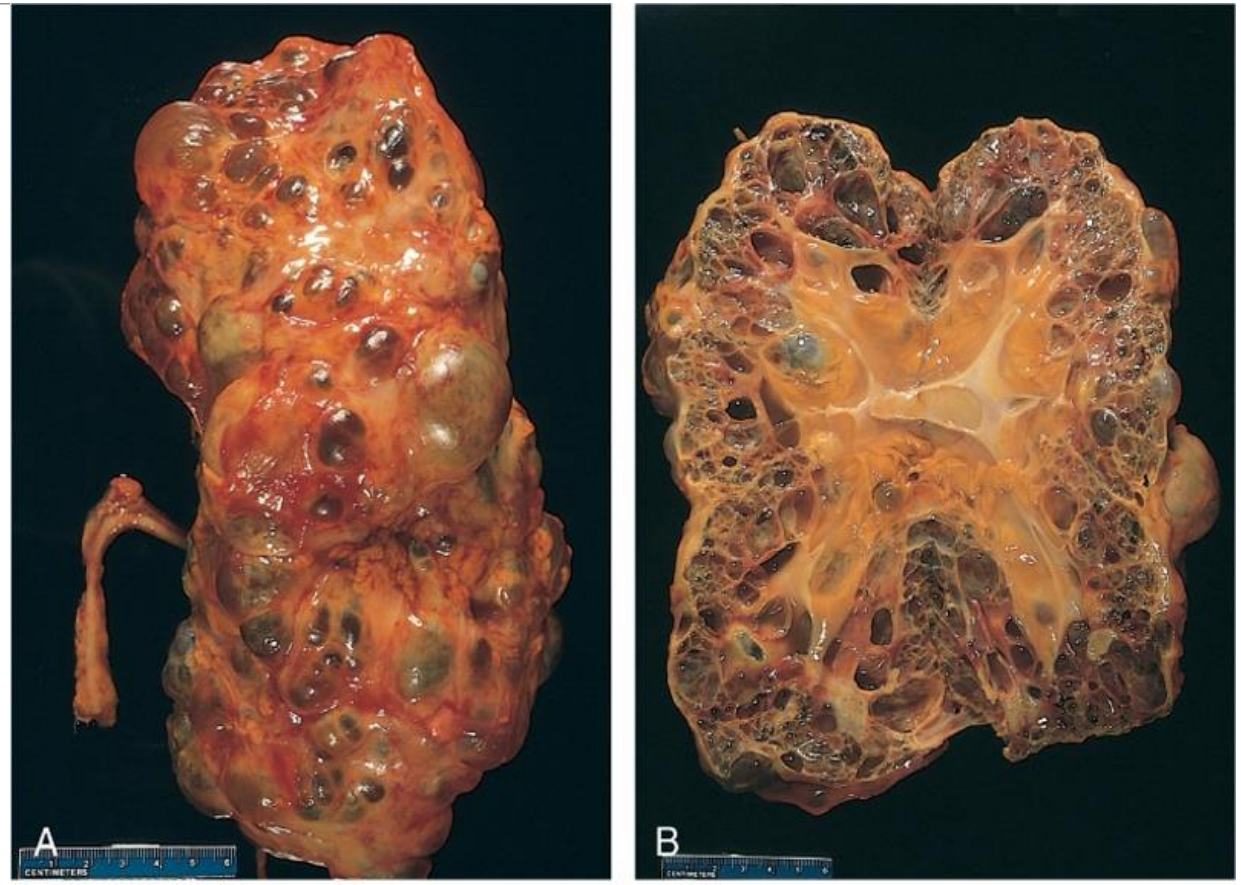
2- Cysts Associated With Chronic Dialysis

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

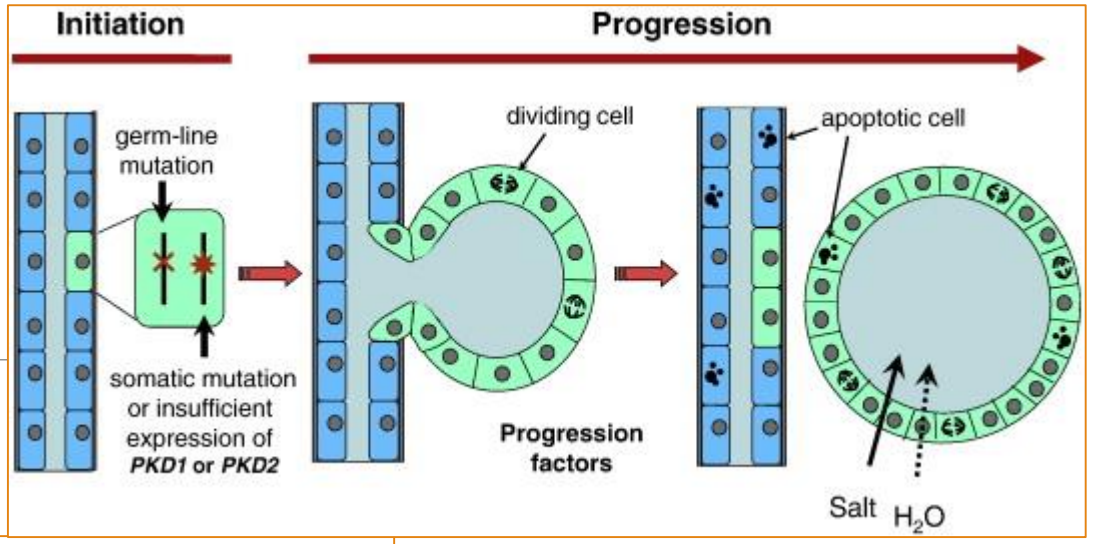


3- Autosomal Dominant (Adult) Polycystic Kidney Disease

- ❑multiple bilateral cysts
- ❑eventually destroy the renal parenchyma.
- ❑Incidence (1: 500-2000) 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).***



(Adult) Polycystic Kidney Disease



Clinical presentation :

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

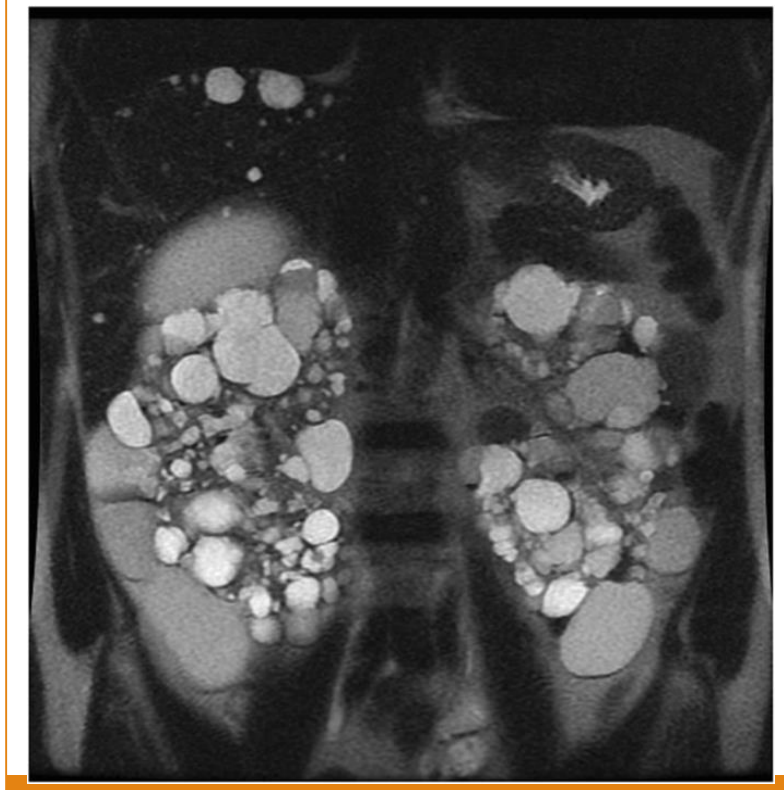
Complications

1- *hypertension* (75%)*

2- urinary infection

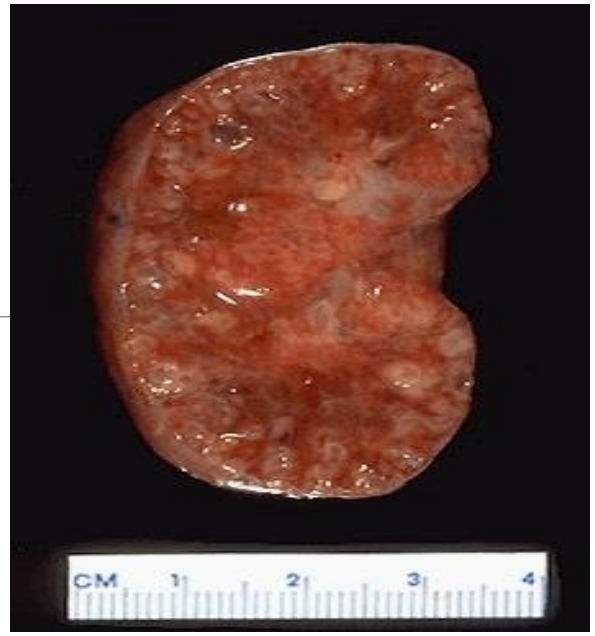
3- vascular aneurysms of circle of Willis* (10% -30%)→ (subarachnoid hemorrhage).

4- renal failure at age 50

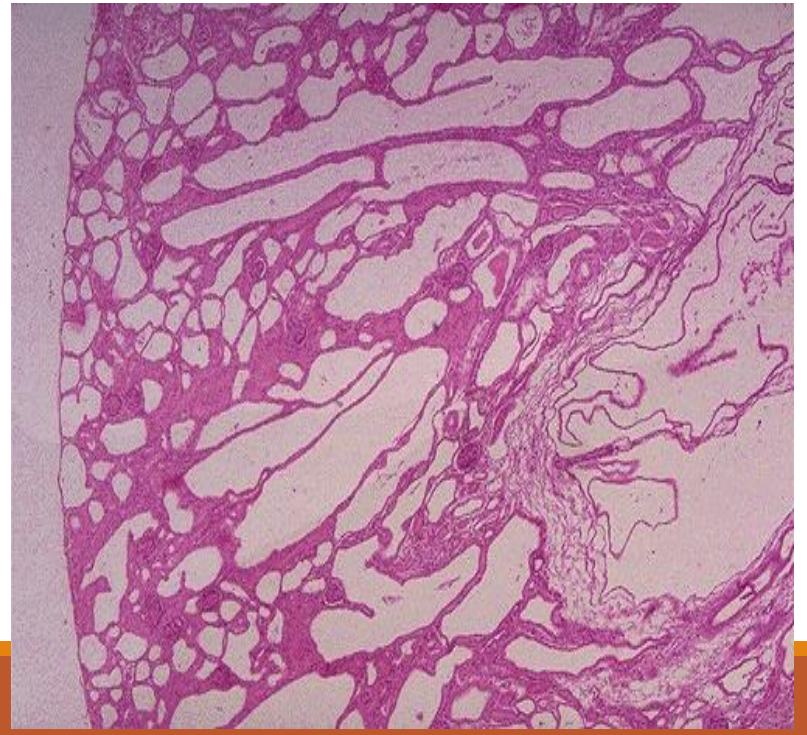


Coronal T2-weighted image shows symmetric enlargement of the kidneys, which contain multiple cysts with variable size

4-Autosomal Recessive (Childhood) Polycystic Kidney Disease

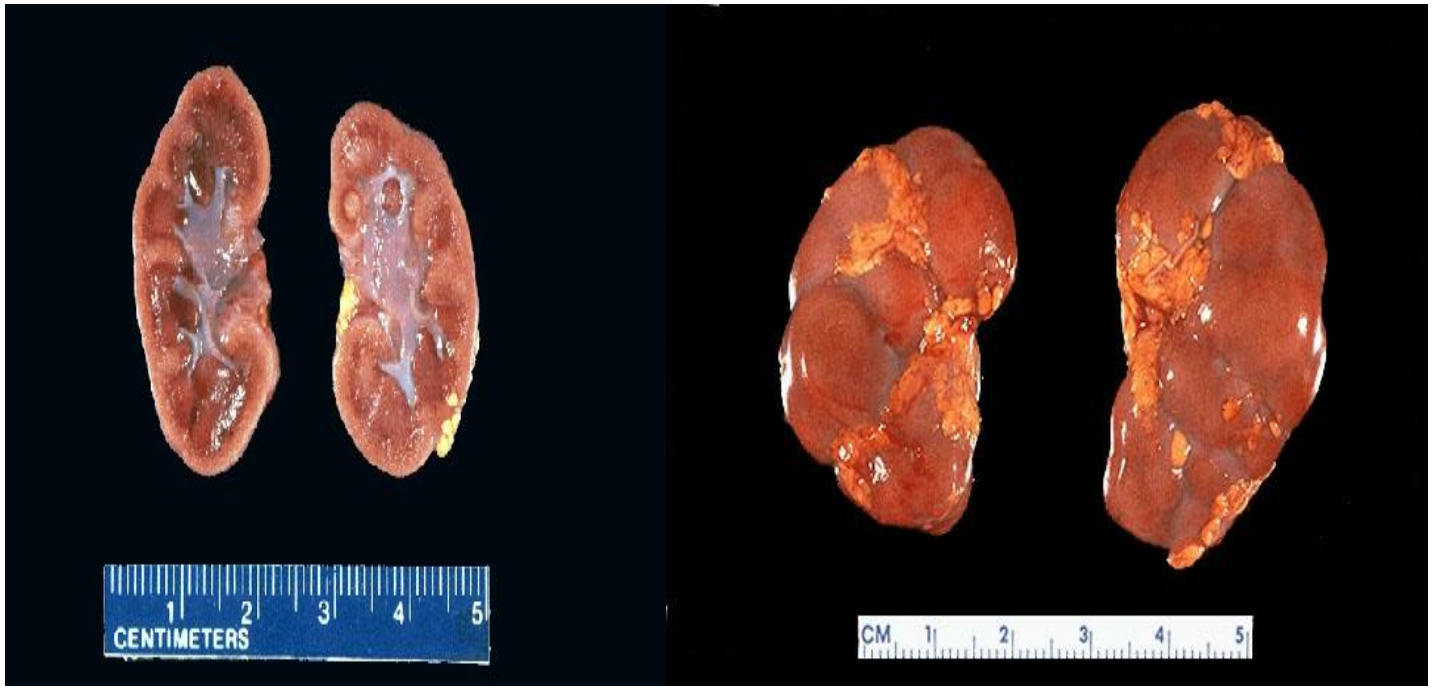


- autosomal recessive
- 1:20,000 live births.
- Types: perinatal, neonatal, infantile, and juvenile.
- Presents early in life
- Associated with liver cysts
- Mutations in ***PKHD1*** gene coding for ***fibrocystin***.
- Fibrocystin may be involved in the function of cilia in tubular epithelial cells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

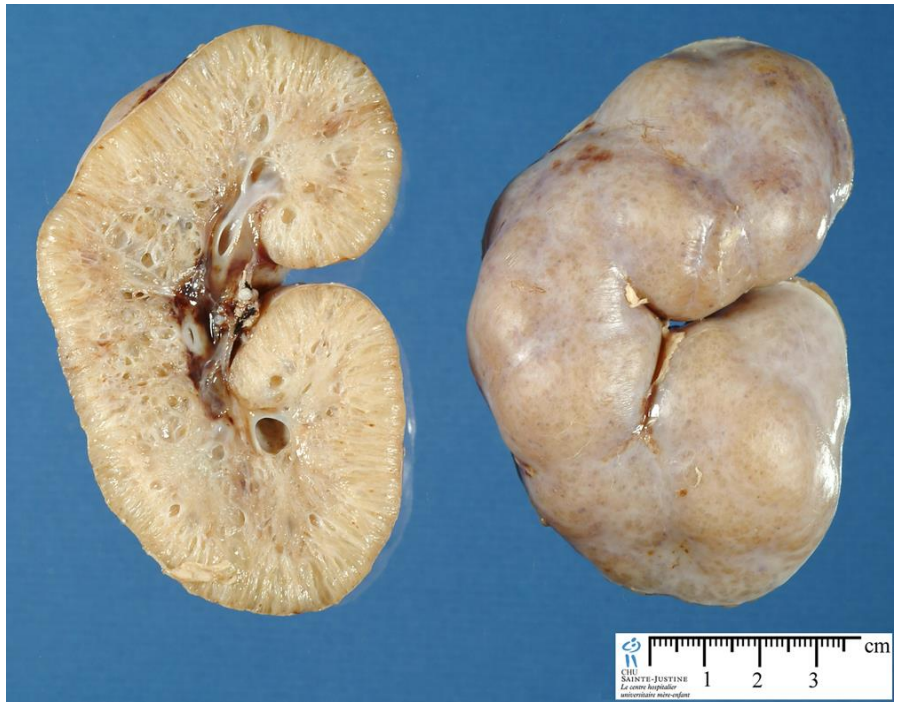


Normal vs childhood polycystic kidneys

NORMAL TERM INFANT KIDNEYS



CHILDHOOD POLYCYSTIC KIDNEYS



Adult vs childhood polycystic kidney disease

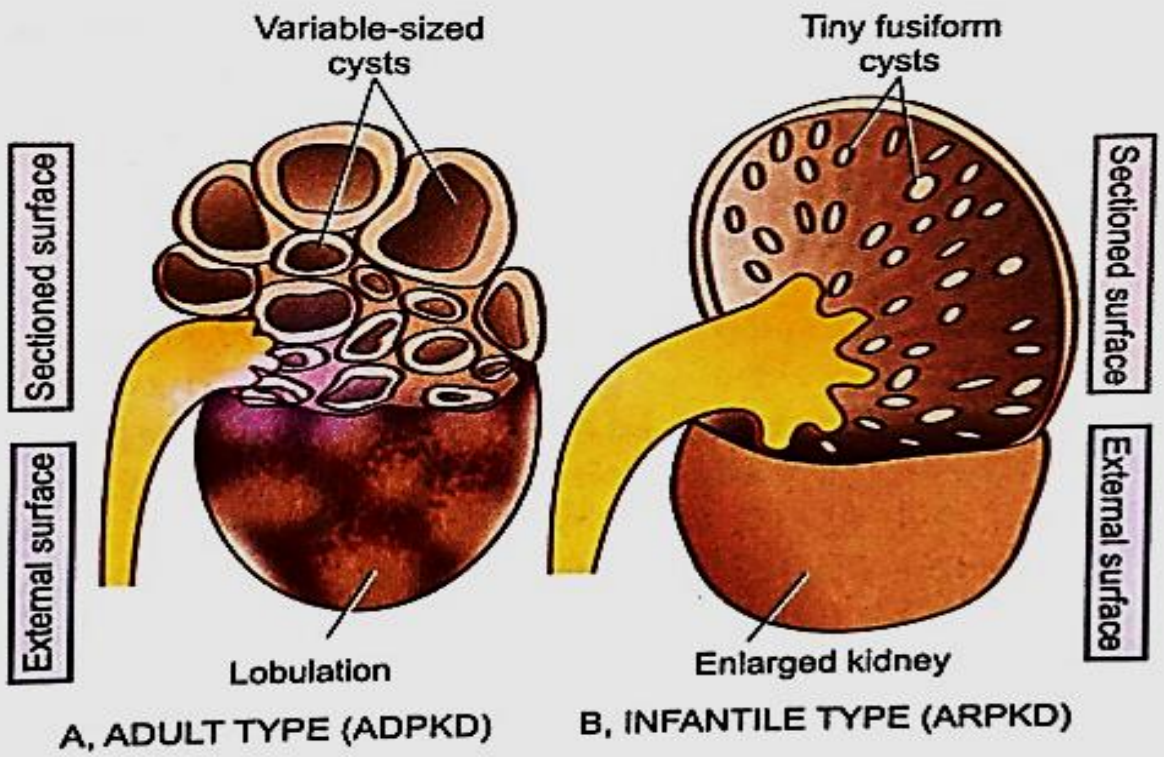


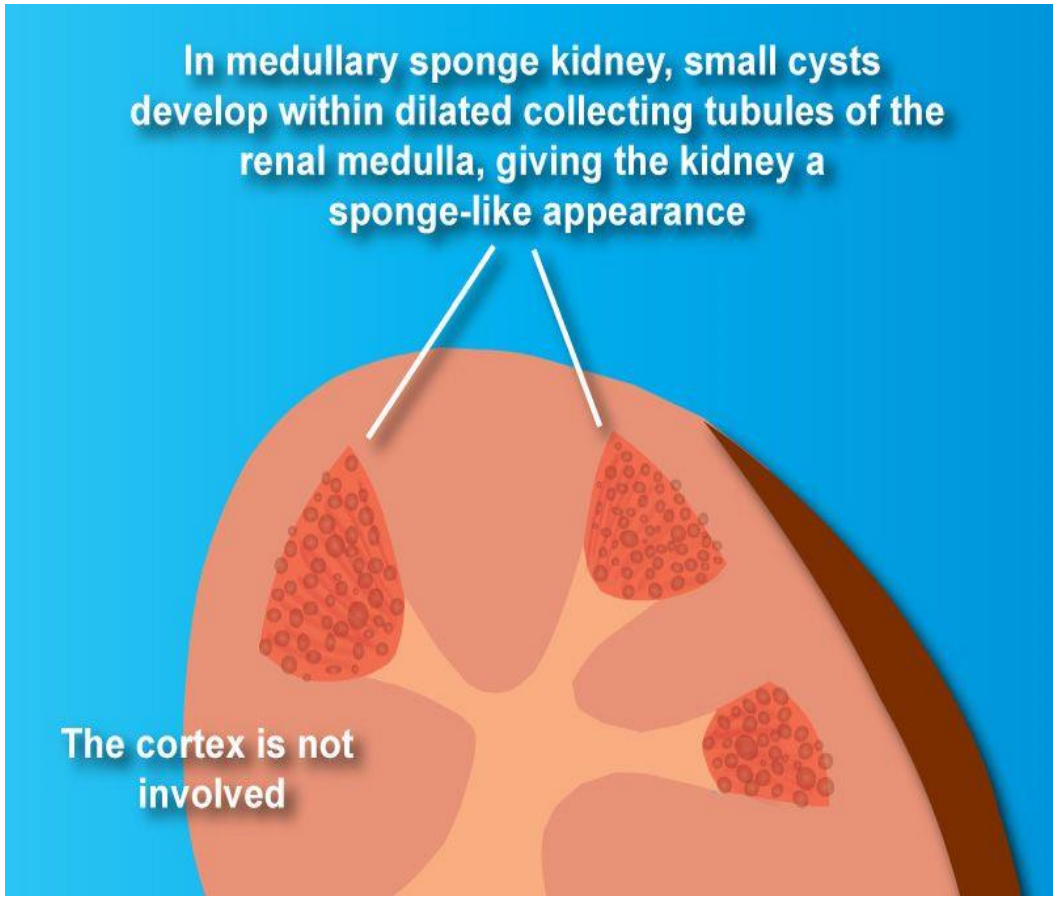
Figure 22.8 Polycystic kidney disease. Diagrammatic representation of comparison of gross appearance of the two main forms.

5- Medullary Cystic Disease

- **2 major types:**

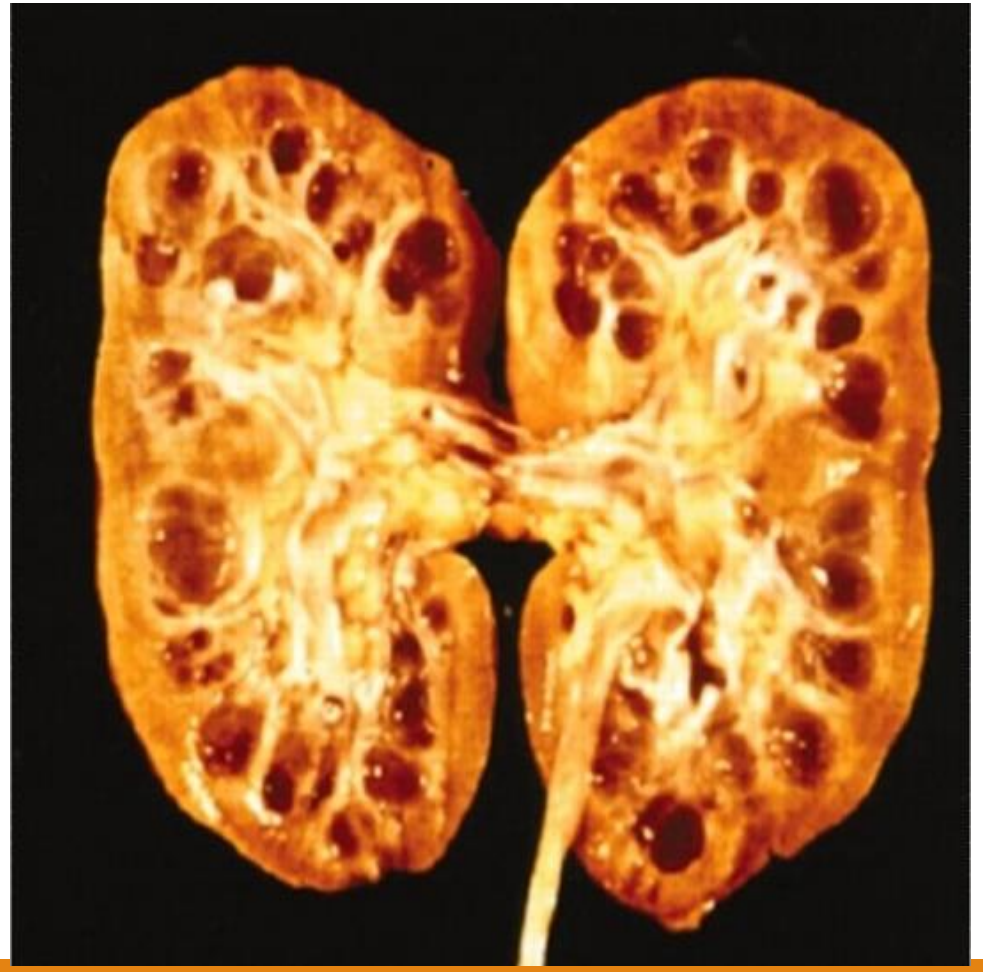
1-Medullary sponge kidney

- **common and nonthreatening condition.**
- **Rare, developmental abnormality characterized by ectatic (dilatory) or cystic malformations in the medullary collecting ducts of the kidney resulting in medullary cysts.**
- **Most patients are asymptomatic and the condition may be diagnosed based on incidental findings following radiologic investigation for other reasons.**



2-N*ephronophthisis-medullary cystic disease complex*

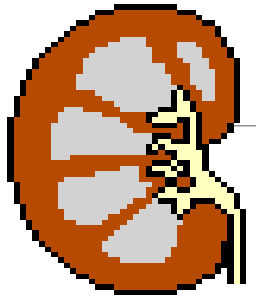
- **- almost always associated with renal dysfunction.**
- **- usually begins in childhood.**
- **- Cysts are at cortico-medullary junction**
- **More than 9 gene mutations are described**
- **All share in common renal histologic triad of tubular basement membrane disintegration, tubular atrophy with cyst development, and interstitial cell infiltration with fibrosis**



Nephronophthisis-medullary cystic disease complex (medullary- uremic type)

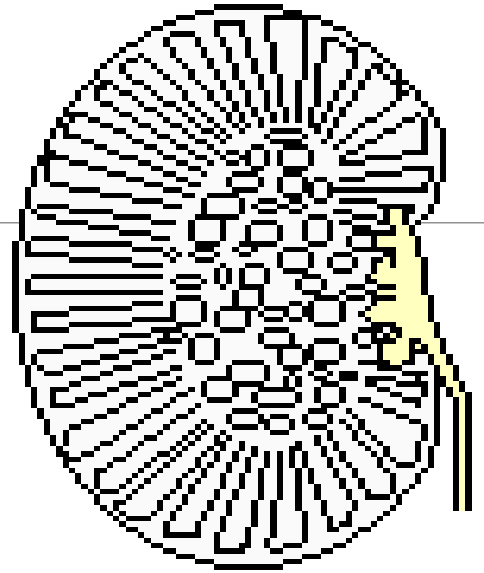
- **Clinical features:**
- **polyuria and polydipsia (↓tubular function).**
- **renal failure over 5-10-year**
- **A positive family history and unexplained chronic renal failure in young patients should lead to suspicion of medullary cystic disease.**

Kidney Cysts

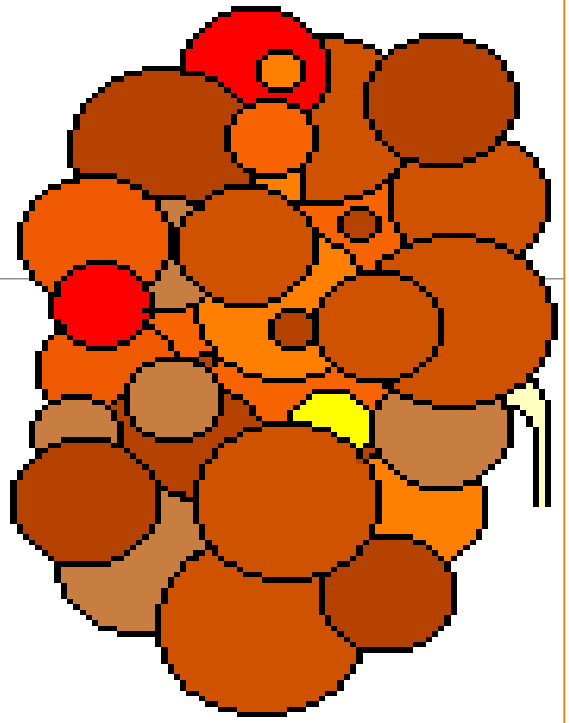


No cysts

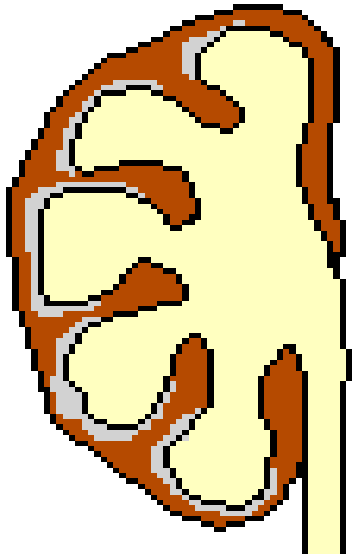
Simple cysts



Recessive polycystic



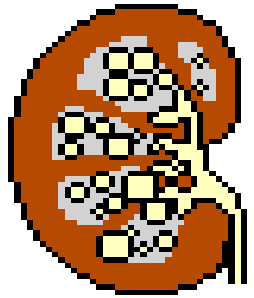
Dominant polycystic



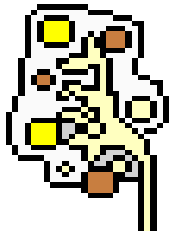
Hydronephrosis is not cysts

"Dysplasia"

Medullary sponge



Medullary uremic



Dialysis cystic