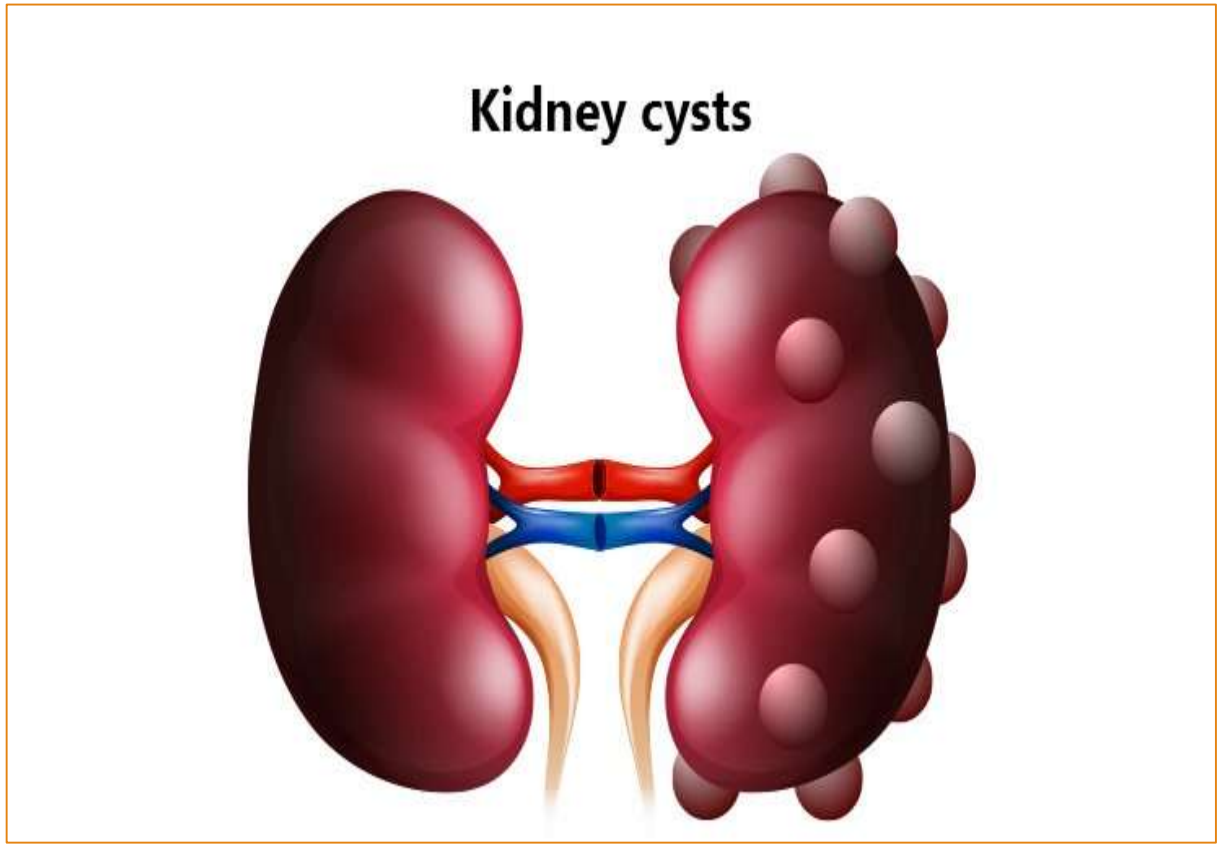
THE UNIVERSITY OF JORDAN

CYSTIC DISEASES of THE KIDNEY

Dr. Nisreen Abu Shahin



Types of cysts

1-Simple Cysts

2-Dialysis-associated acquired cysts

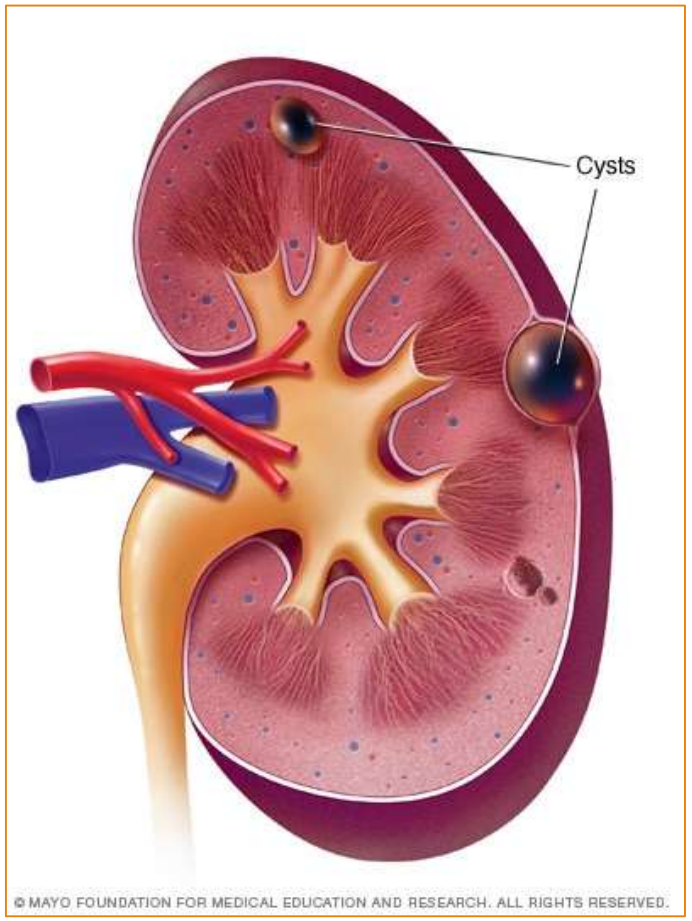
3-Autosomal Dominant (Adult) Polycystic Kidney Disease

4-Autosomal Recessive (Childhood) Polycystic Kidney Disease

5-Medullary Cystic Disease

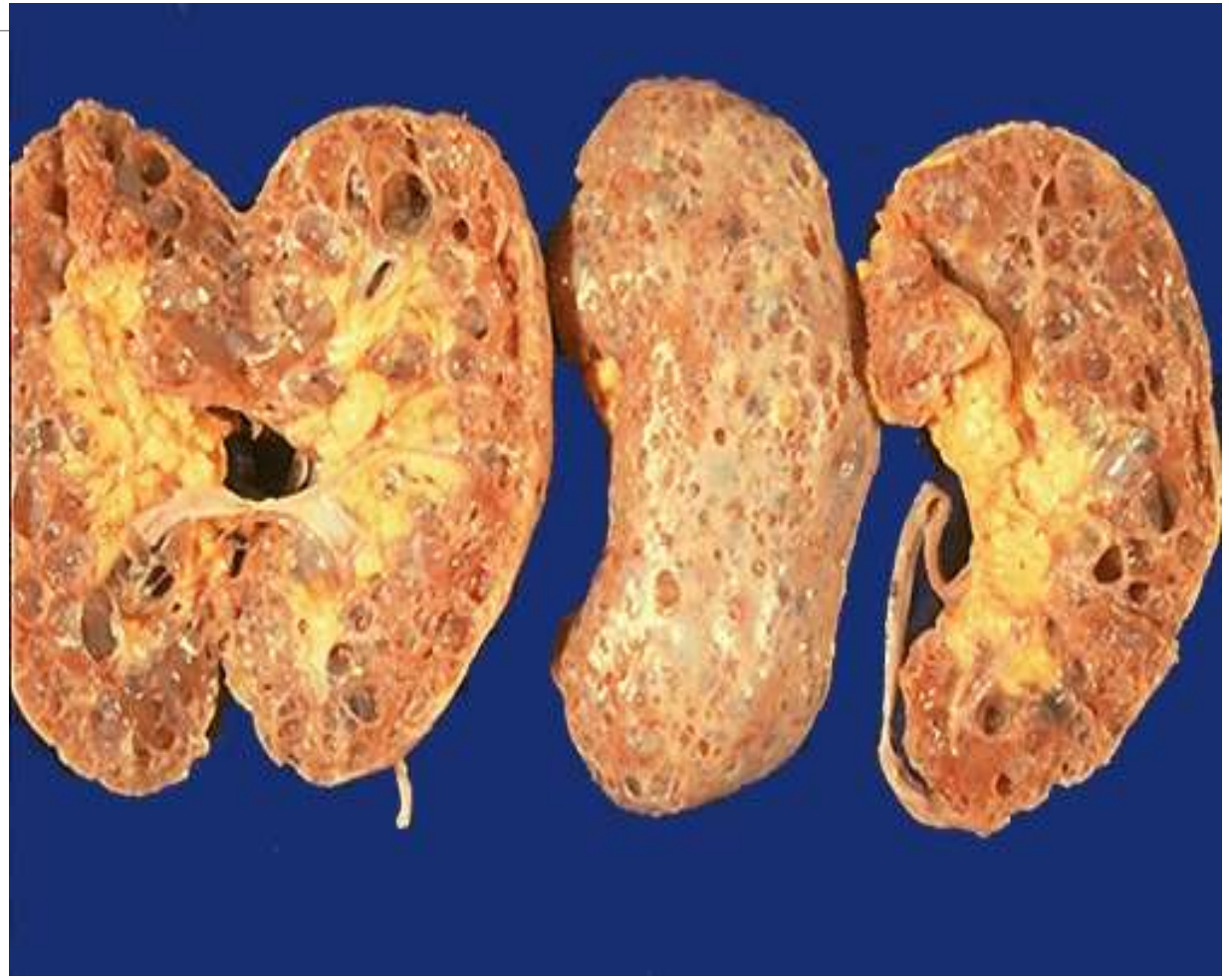
1- Simple Renal Cysts

- 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



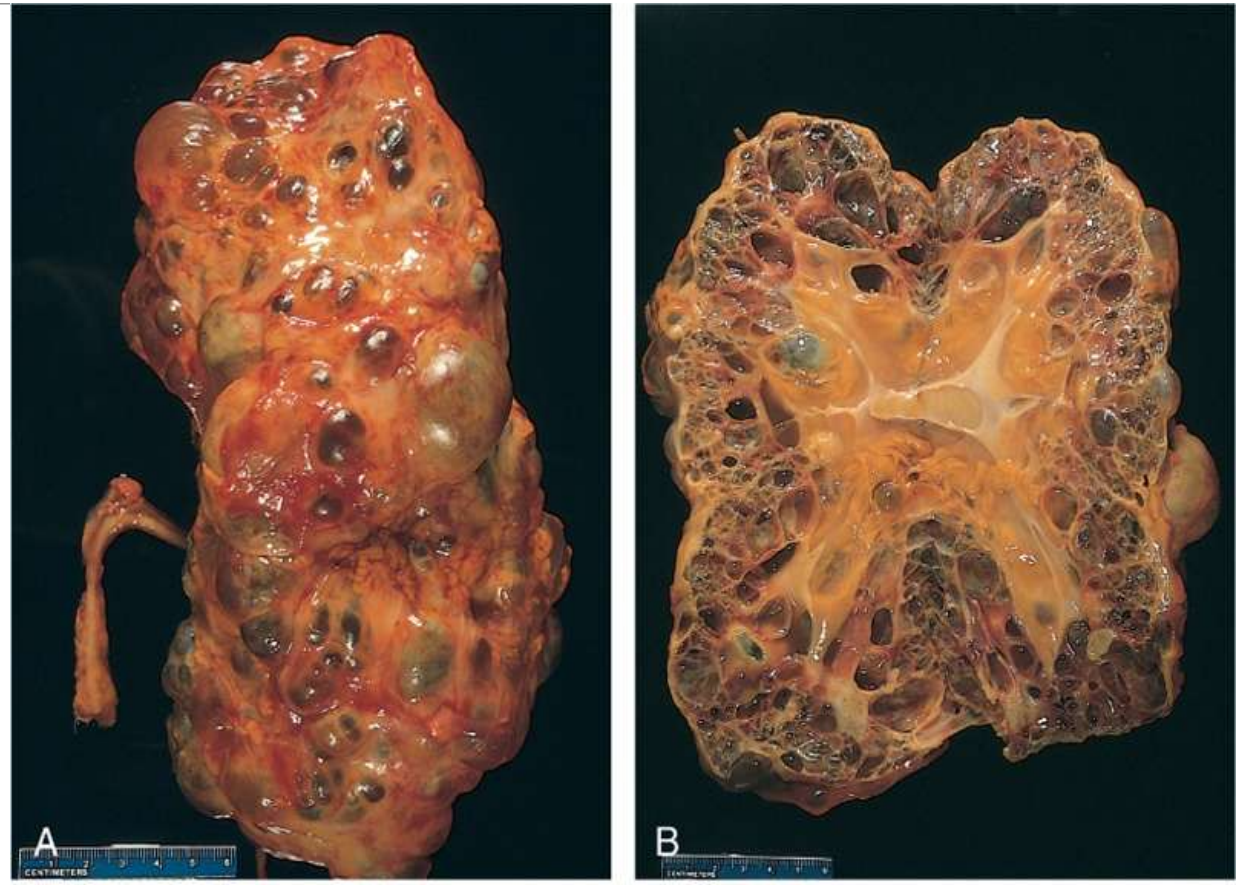
2- Cysts Associated With Chronic Dialysis

- **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)

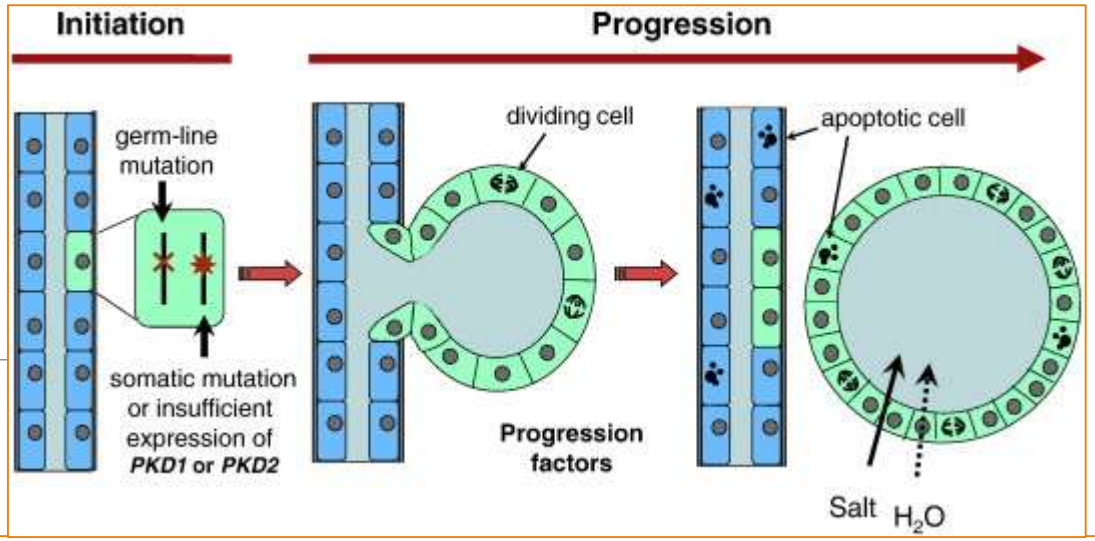


3- Autosomal Dominant (Adult) Polycystic Kidney Disease

- ❑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

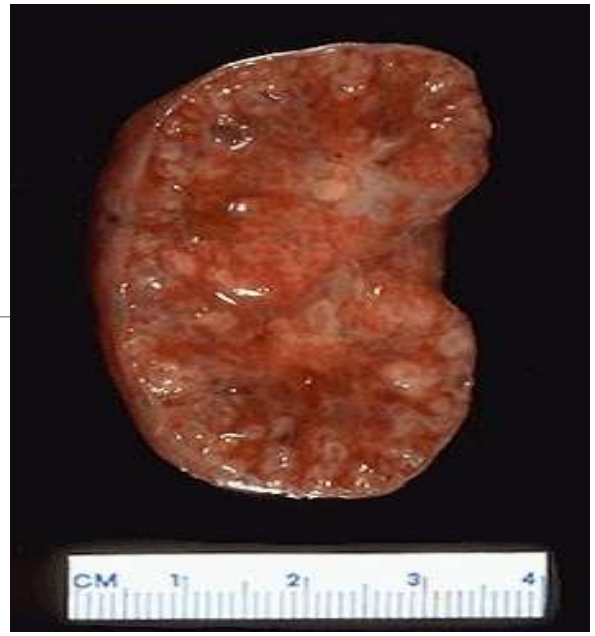
1- *hypertension* (75%)

2- urinary tract infection

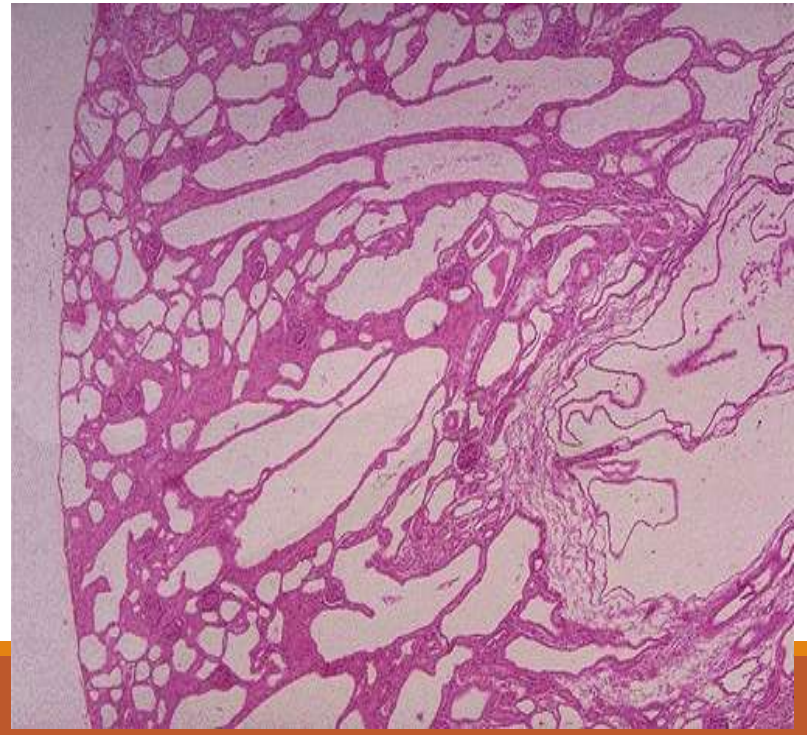
3- vascular aneurysms of circle of Willis (10% -30%)→→ (subarachnoid hemorrhage)

4- chronic renal failure at age 50 (≈25% ; % increases with age)

4-Autosomal Recessive (Childhood) Polycystic Kidney Disease

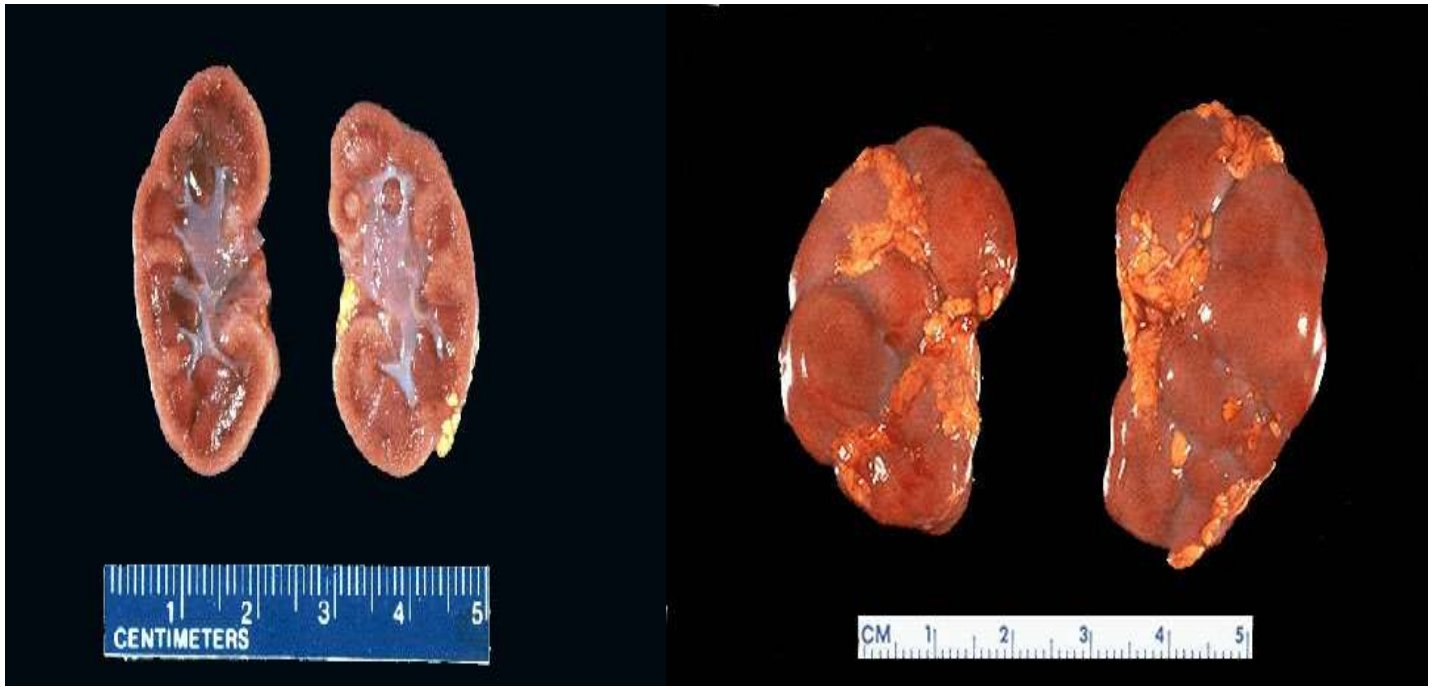


- **autosomal recessive**
- **1:20,000 live births.**
- **Types: perinatal, neonatal, infantile, and juvenile.**
- **Presents early in life**
- **Associated with liver cysts and fibrosis**
- **Mutations in *PKHD1* gene coding for *fibrocystin*.**
- **Fibrocystin may be involved in the function of cilia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



Normal vs childhood polycystic kidneys

NORMAL TERM INFANT KIDNEYS



CHILDHOOD POLYCYSTIC KIDNEYS

Adult vs childhood polycystic kidney disease

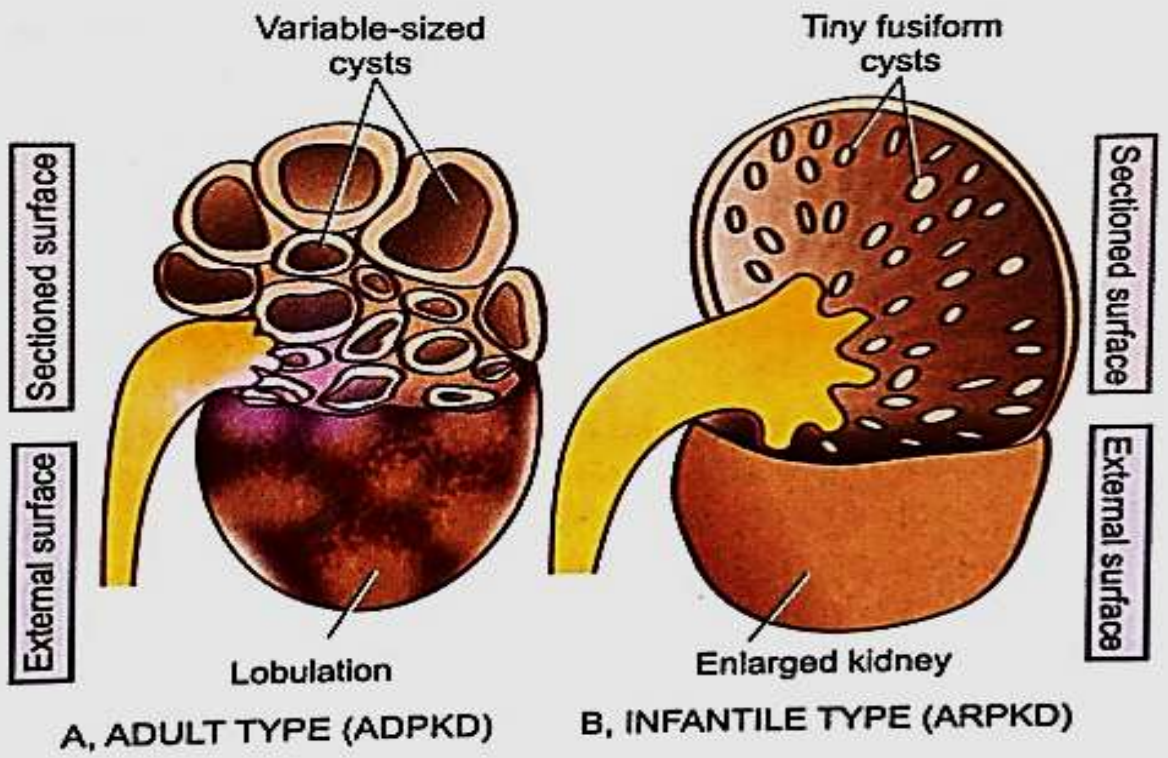


Figure 22.8 Polycystic kidney disease. Diagrammatic representation of comparison of gross appearance of the two main forms.

5- Medullary Cystic Disease

- **2 major types:**

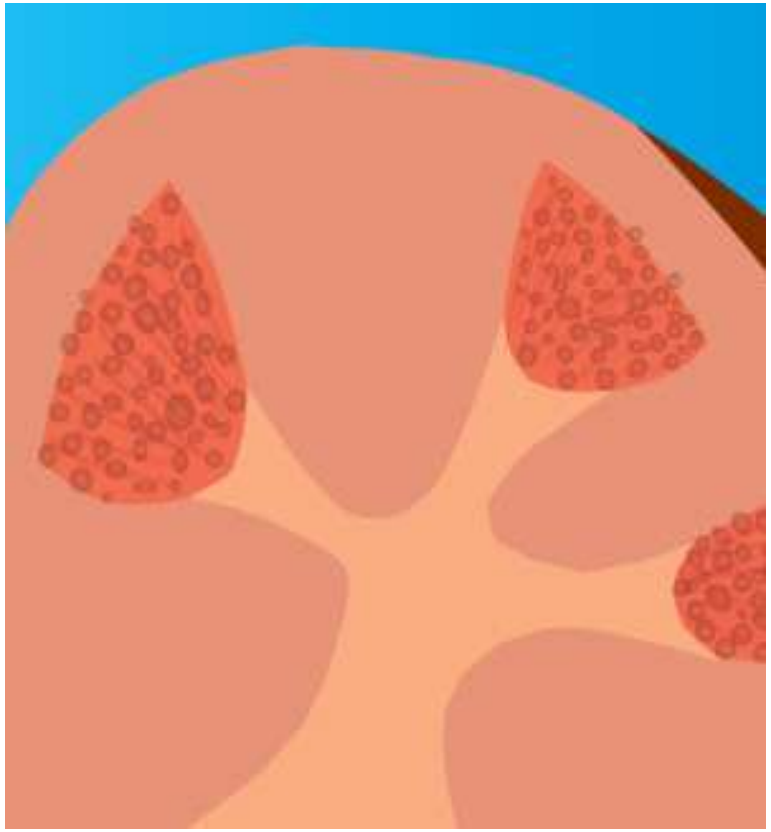
1-medullary sponge kidney

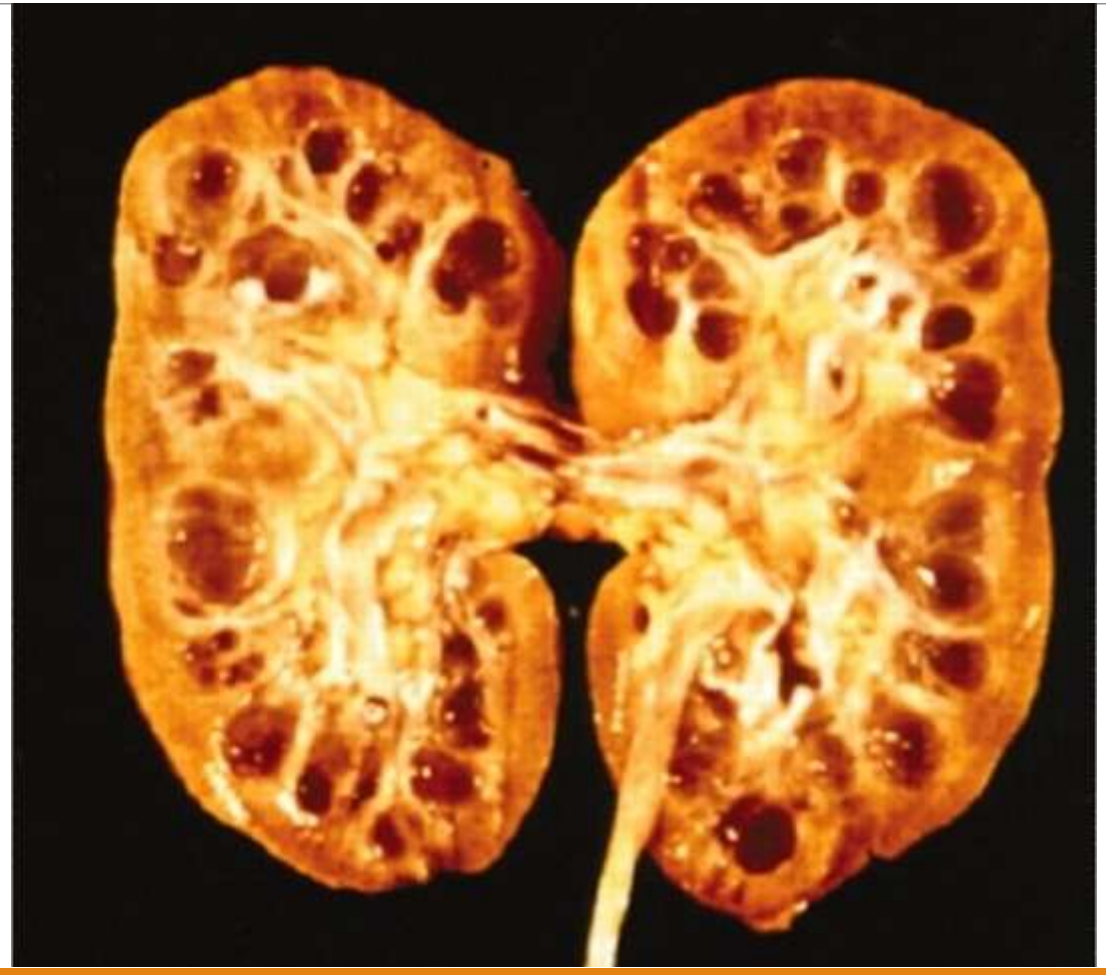
- **common and innocent condition**

2-*nephronophthisis-medullary cystic disease complex*

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

Medullary Sponge Kidney



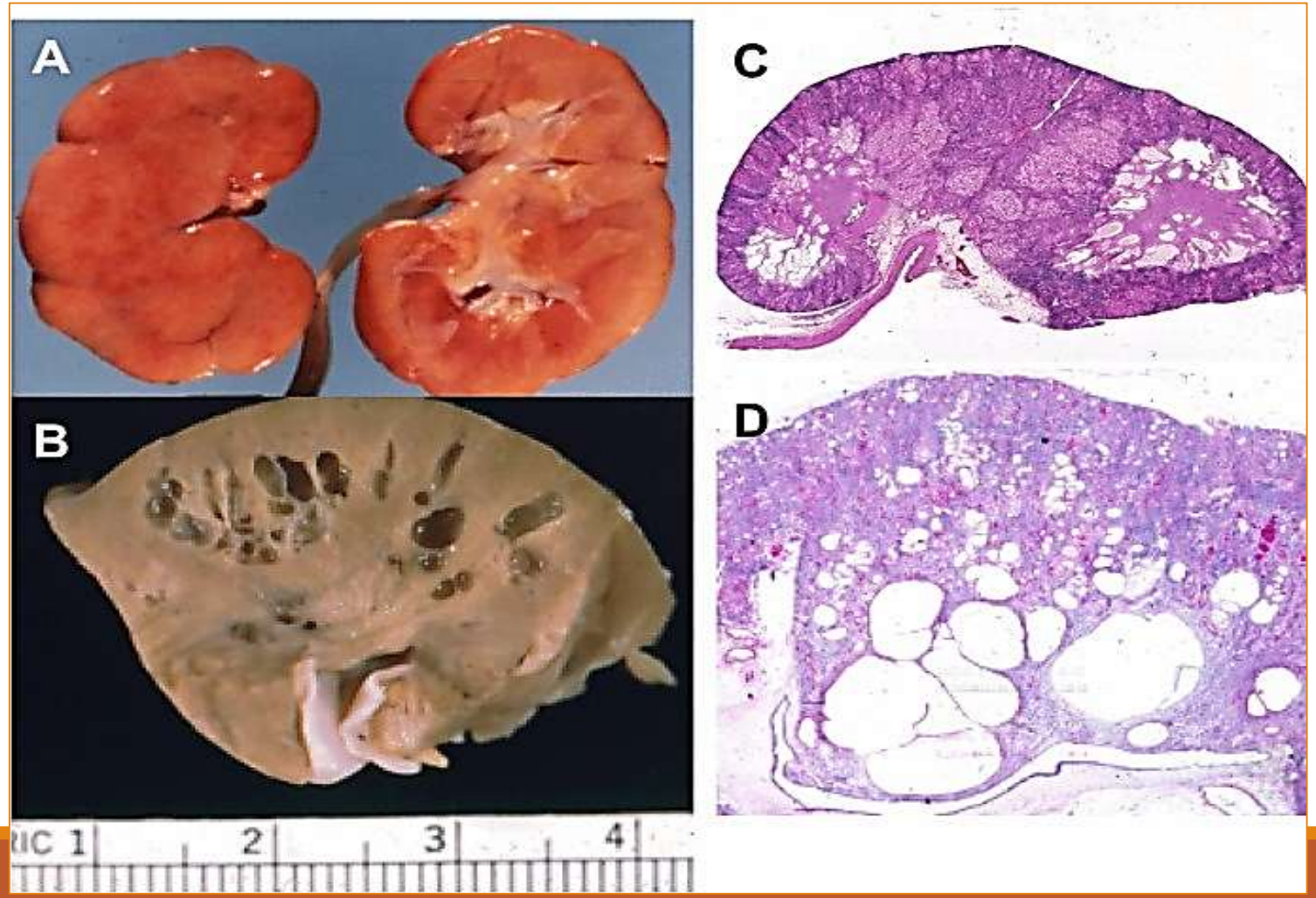


5- Medullary Cystic Disease

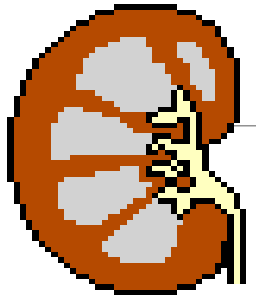
2- Nephronophthisis-medullary cystic disease complex (medullary- uremic type)

- **A worse disease with progressive renal impairment**
- **polyuria and polydipsia (↓tubular function)**
- **renal failure over 5-10 years**
- **A positive family history and unexplained chronic renal failure in young patients should lead to suspicion of medullary cystic disease.**

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

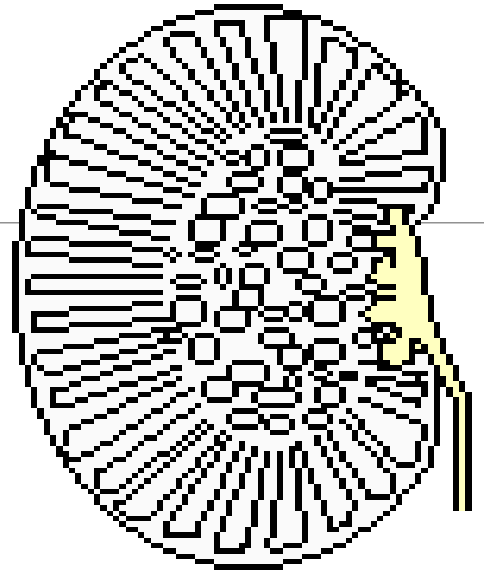


Kidney Cysts

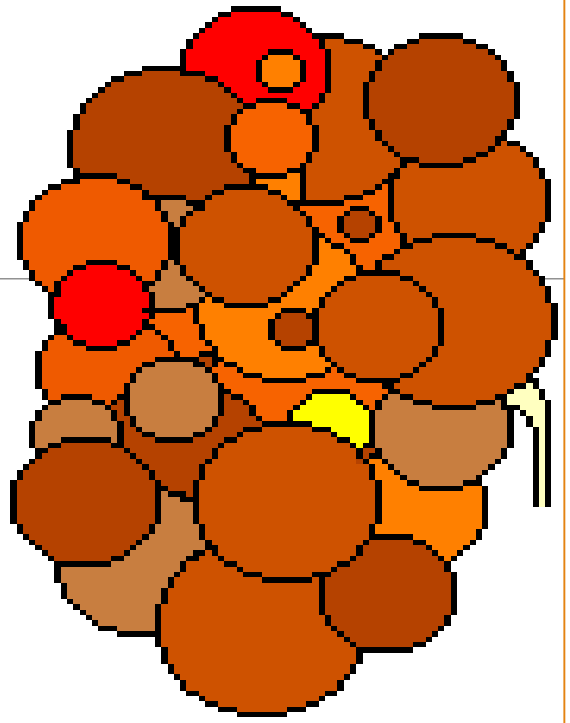


No cysts

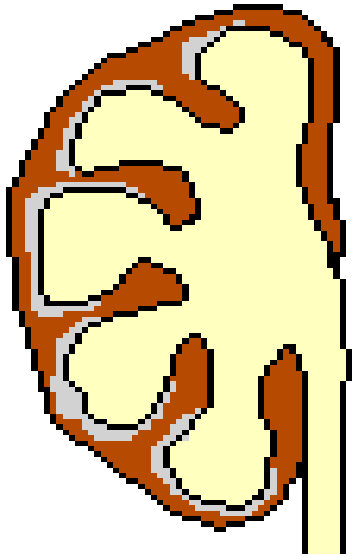
Simple cysts



Recessive polycystic



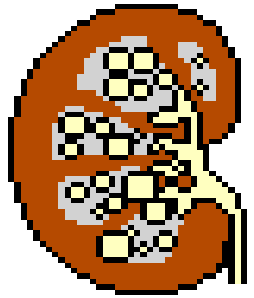
Dominant polycystic



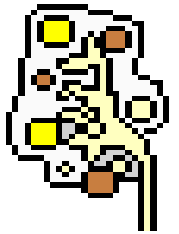
Hydronephrosis is not cysts

"Dysplasia"

Medullary sponge



Medullary uremic



Dialysis cystic