THE UNIVERSITY OF JORDAN

# CYSTIC DISEASES of THE KIDNEY

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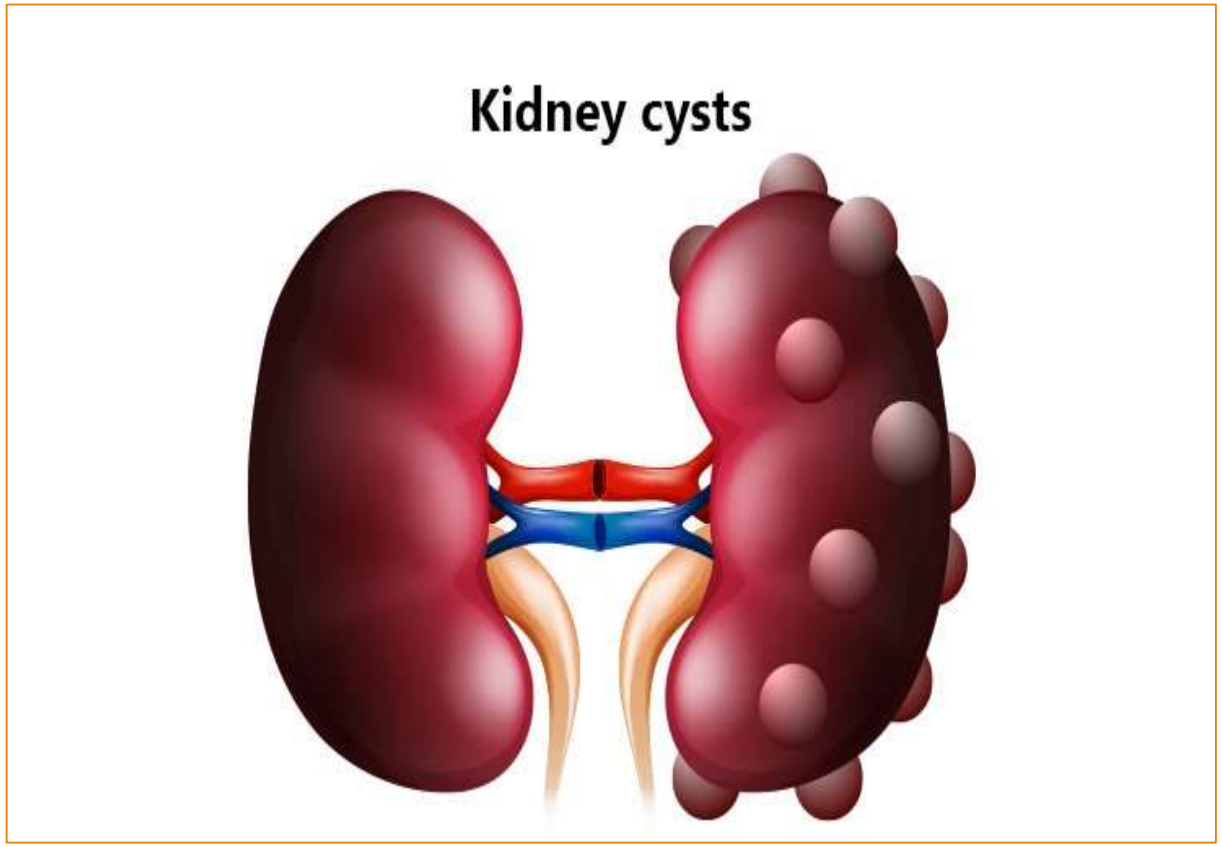


# Types of cysts

1-Simple Cysts

2-Dialysis-associated acquired cysts

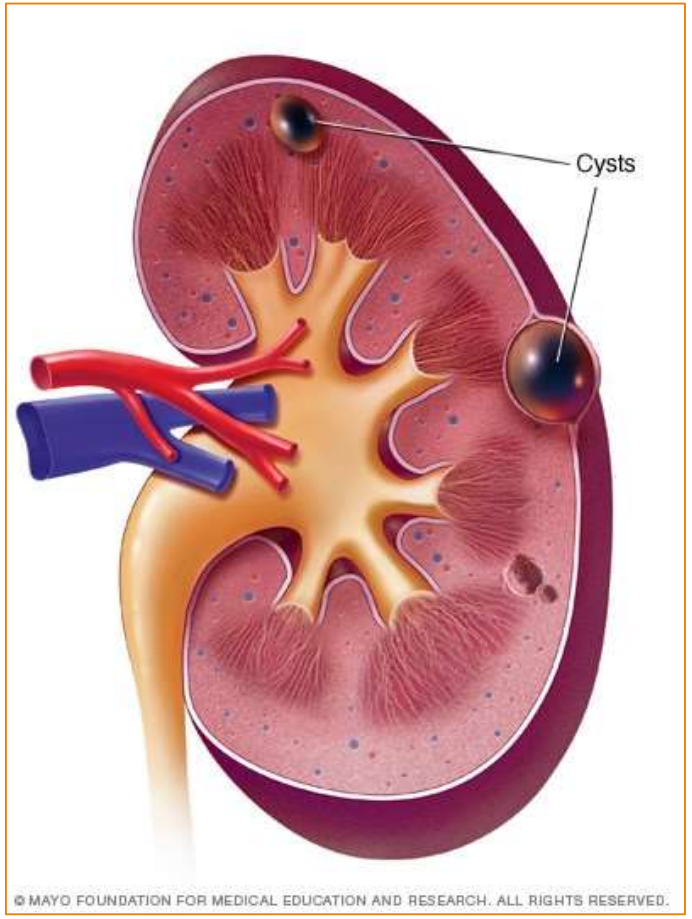
3-Autosomal Dominant (Adult) Polycystic Kidney Disease

4-Autosomal Recessive (Childhood) Polycystic Kidney Disease

5-Medullary Cystic Disease

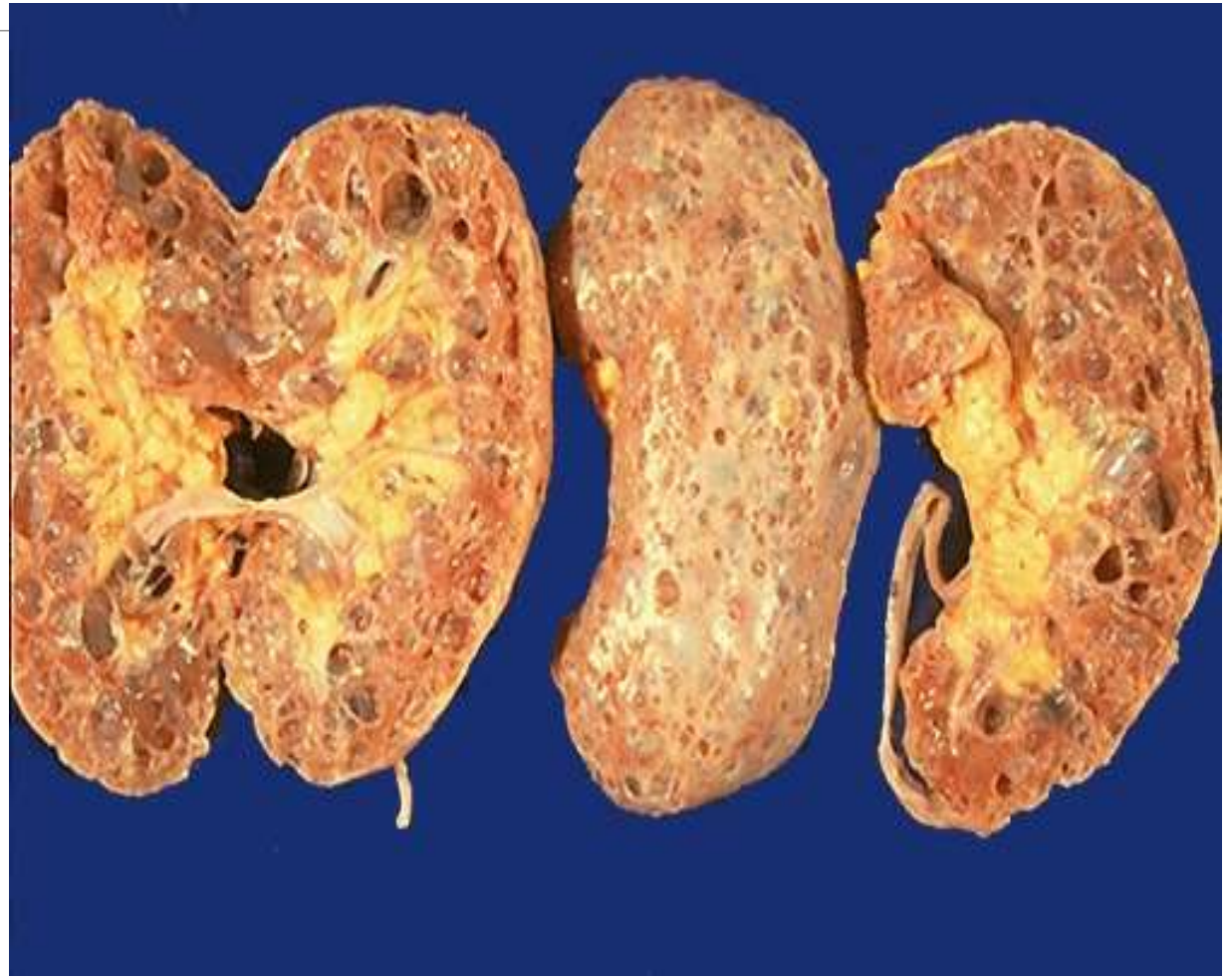
# 1- Simple Renal Cysts

- 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



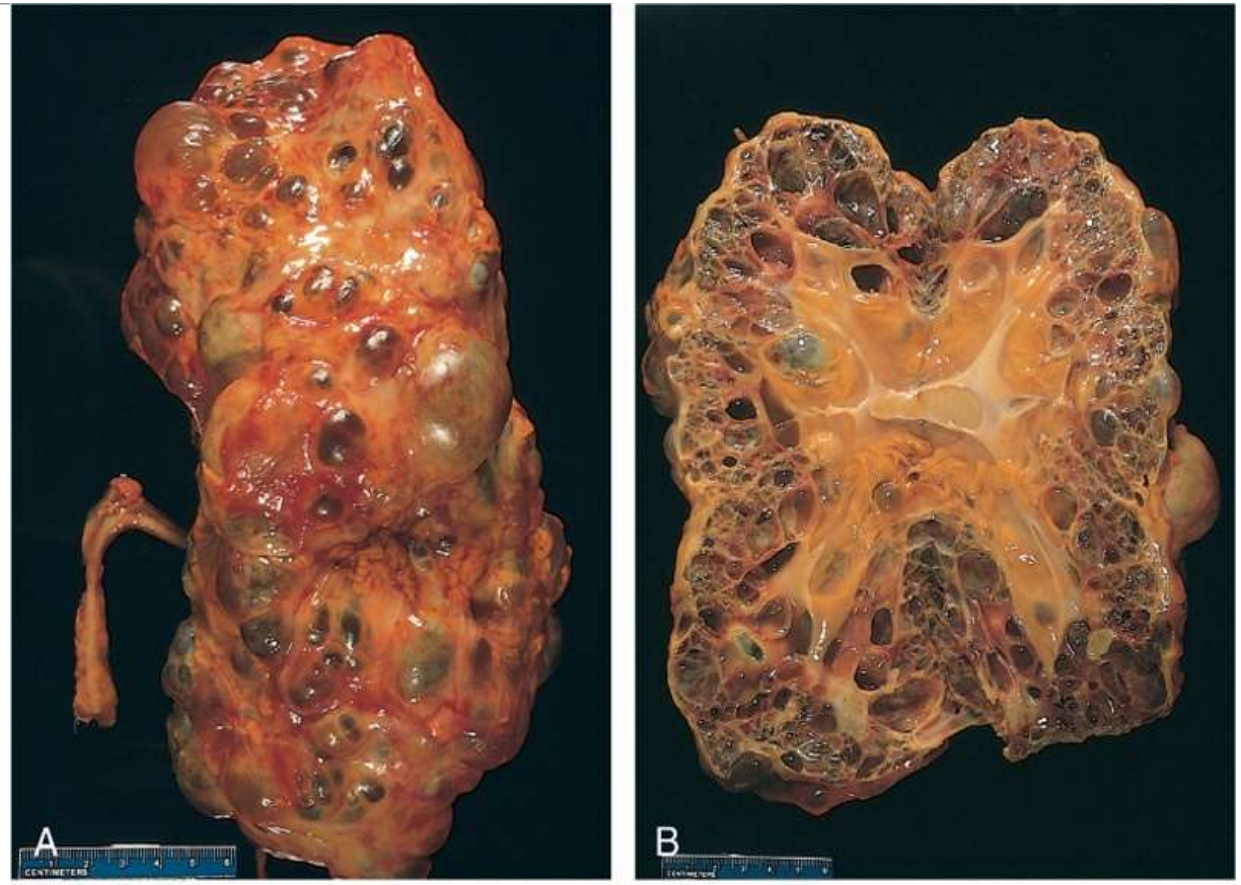

# 2- Cysts Associated With Chronic Dialysis

- **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)**

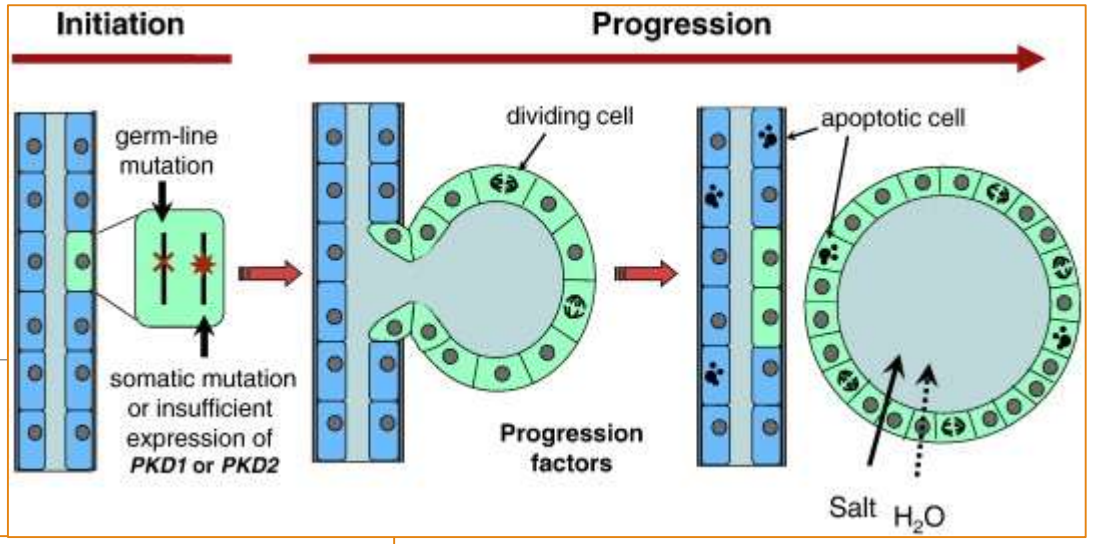


# 3- Autosomal Dominant (Adult) Polycystic Kidney Disease

- ❑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***

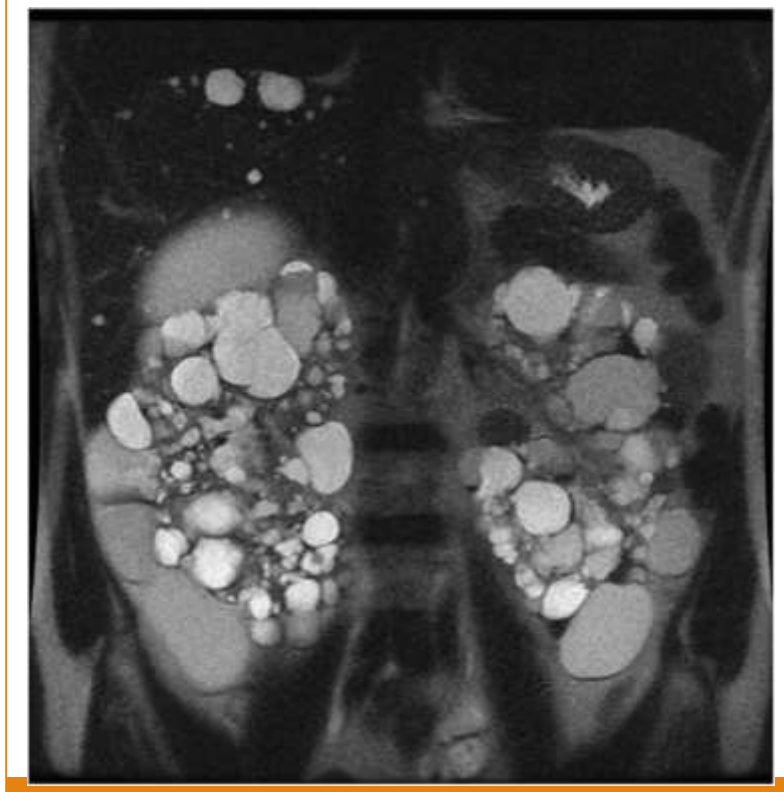
**Complications**

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

***2- urinary tract infections***

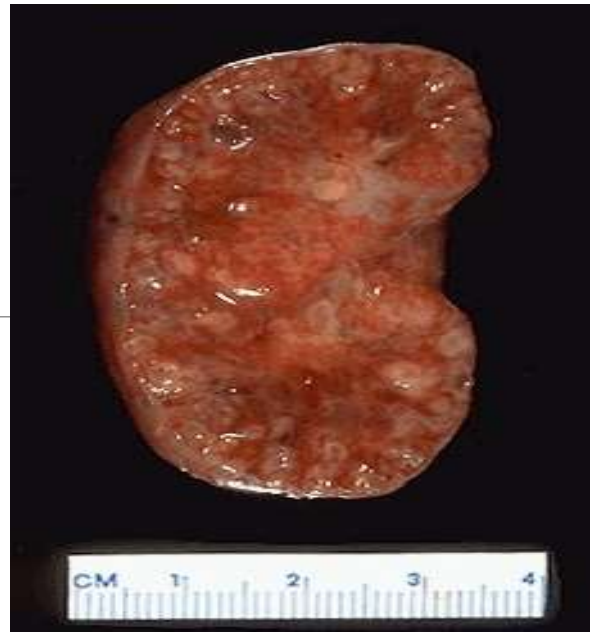
**3- vascular aneurysms of circle of Willis* (10% -30%)→ (subarachnoid hemorrhage ).**

**4- renal failure at age 50 (25%; risk increases with age)**

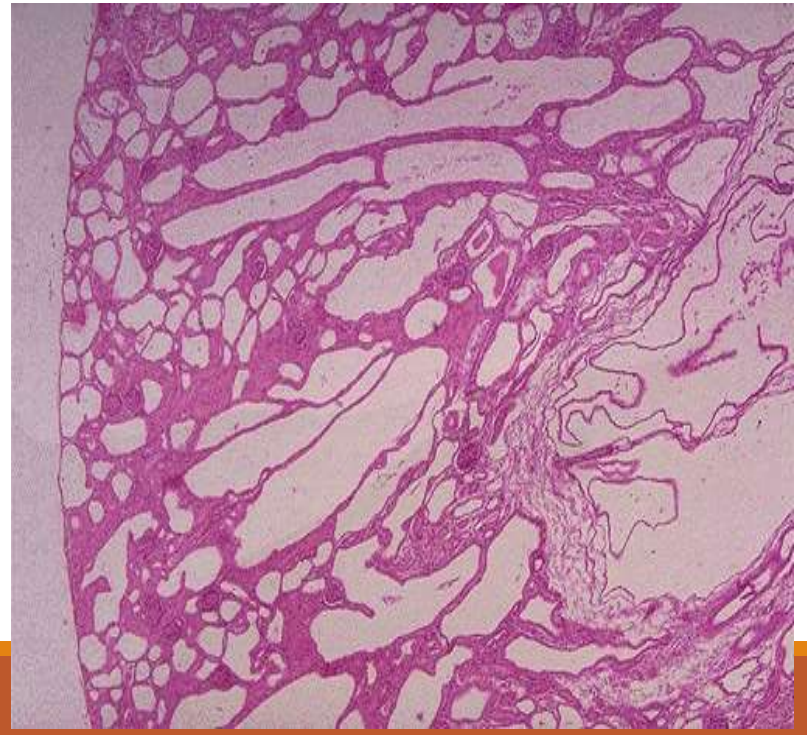


Coronal T2-weighted image shows symmetric enlargement of the kidneys, which contain multiple cysts with variable size

# 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 <u>liver</u> cysts** + hepatic fibrosis
- **Mutations in ***<u>PKHD1</u>* **gene coding for ***<u>fibrocystin</u>*.
- **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

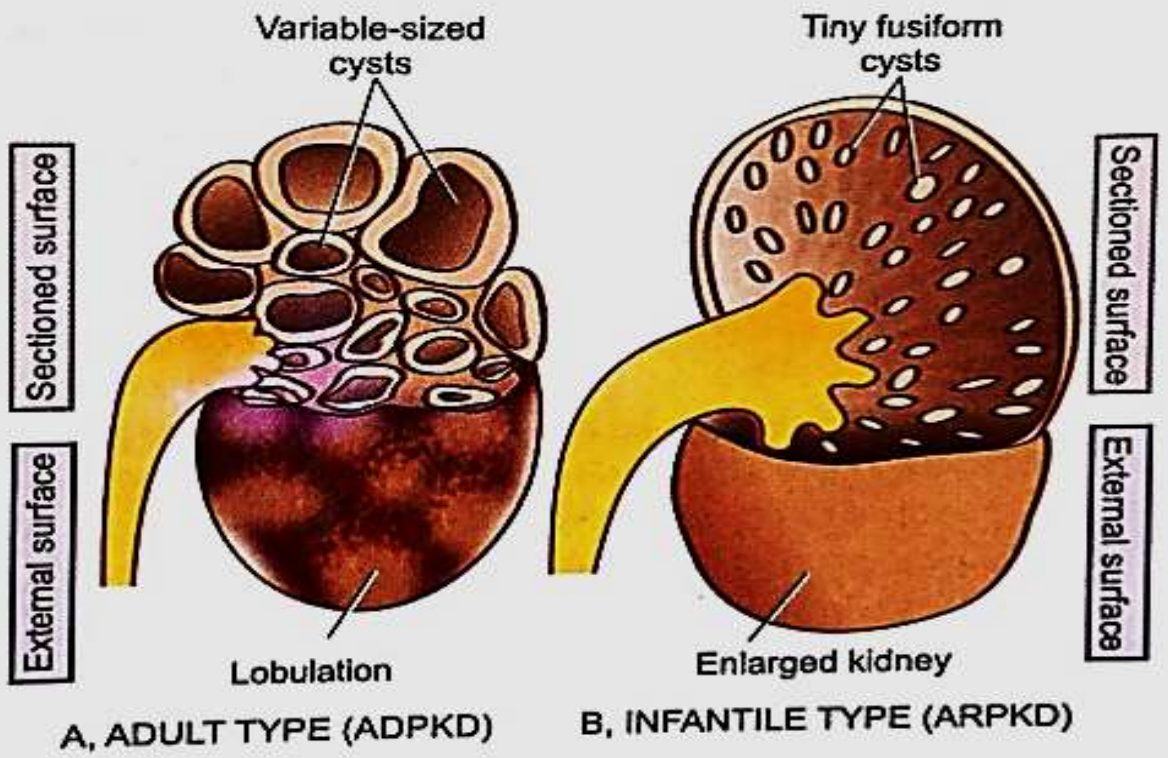


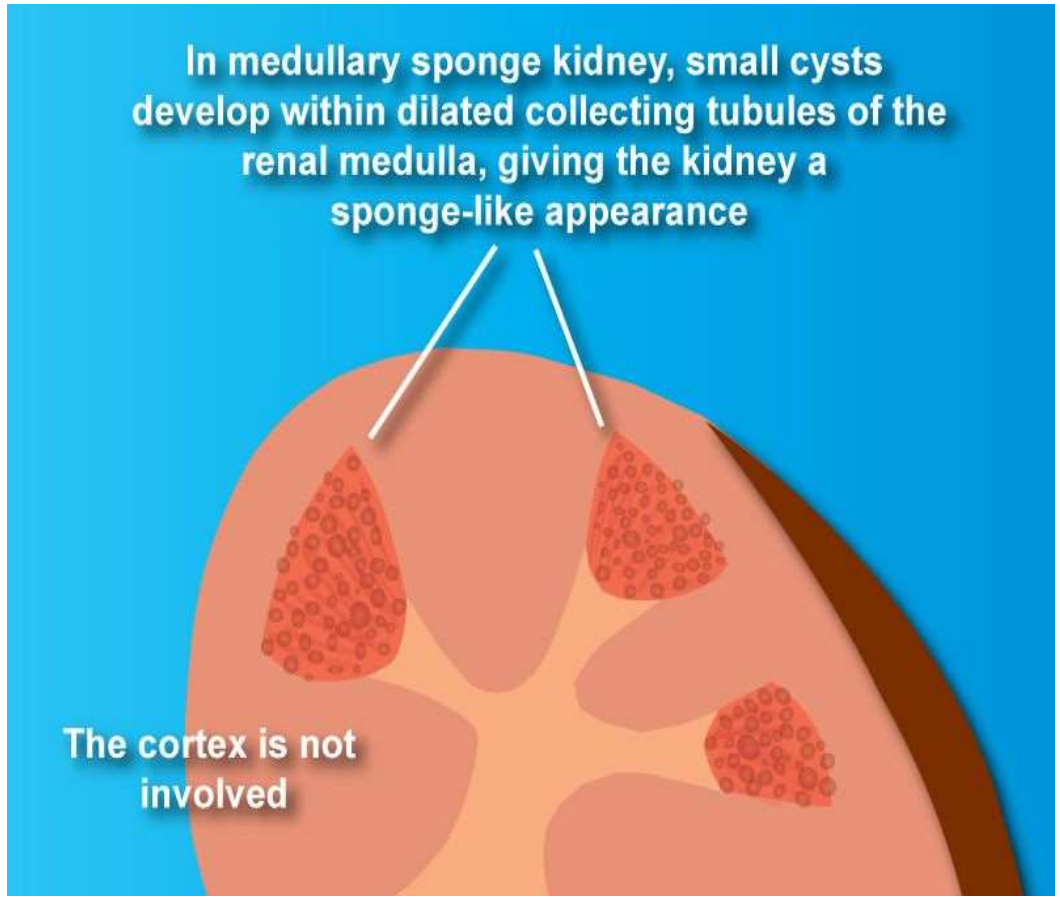

**Figure 22.8** Polycystic kidney disease. Diagrammatic representation of comparison of gross appearance of the two main forms.

# 5- Medullary Cystic Disease

- **2 major types:**

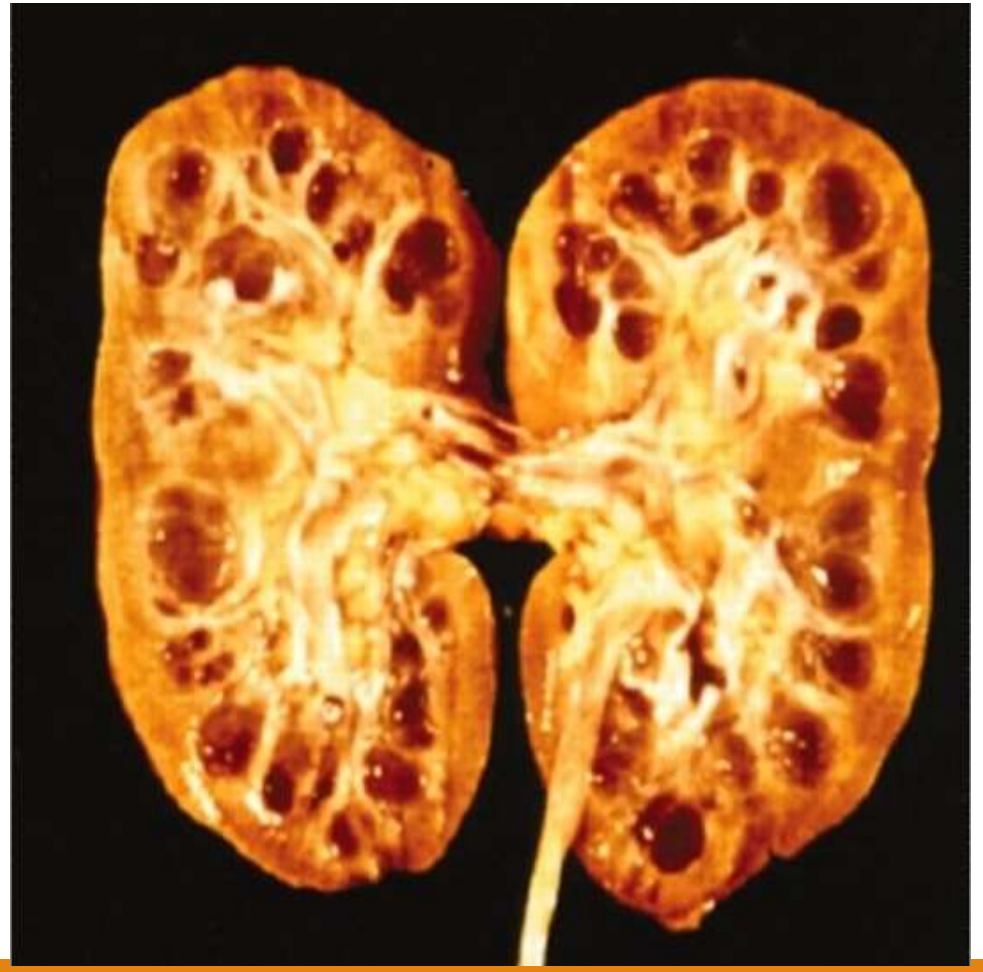
***1-Medullary sponge kidney***

- **A rare nonthreatening condition.**
- **developmental abnormality characterized by ectatic (dilated) ducts or cystic malformations in the medullary collecting ducts of 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*

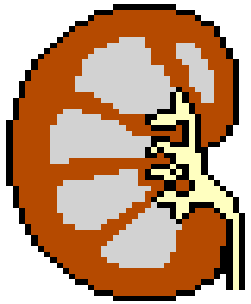
- **- almost always associated with renal dysfunction.**
- **- usually begins in childhood.**
- **- Cysts are at cortico-medullary junction**
- **More than 9 gene mutations are described**
- **All share in common renal histologic triad of tubular basement membrane disintegration, tubular atrophy/ cyst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



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

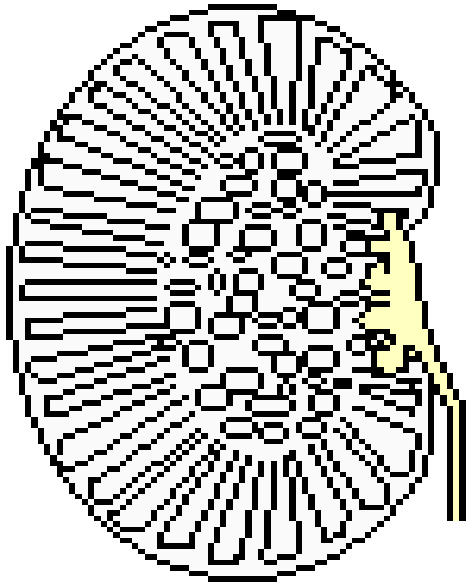
- **Clinical features:**
- **polyuria and polydipsia (↓tubular function).**
- **renal failure over 5-10-year**
- **A positive family history and unexplained chronic renal failure in young patients should lead to suspicion of medullary cystic disease.**

# Kidney Cysts

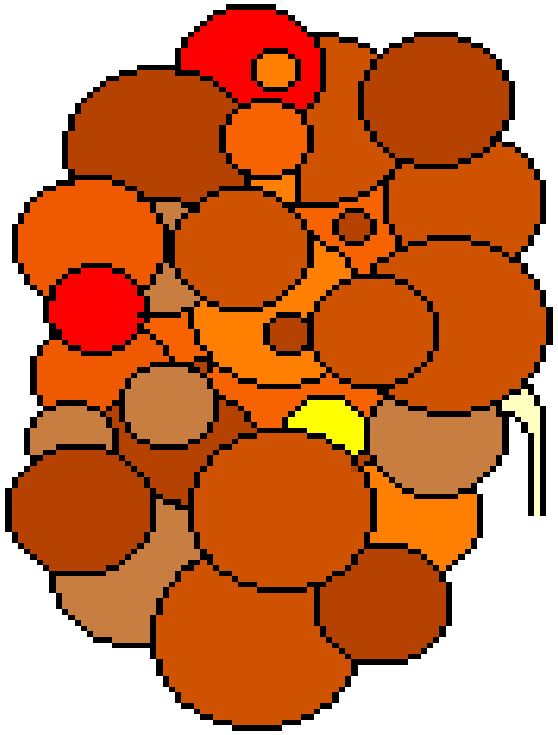


No cysts

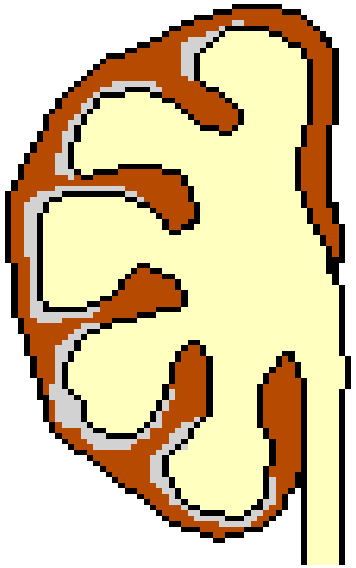
Simple cysts



Recessive polycystic



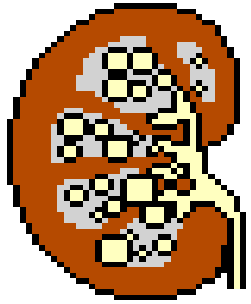
Dominant polycystic



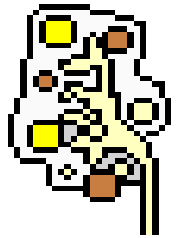
Hydronephrosis is not cystsis not cysts

"Dysplasia"

Medullary sponge



Medullary uremic



Dialysis cystic