CYSTIC DISEASES of THE KIDNEY

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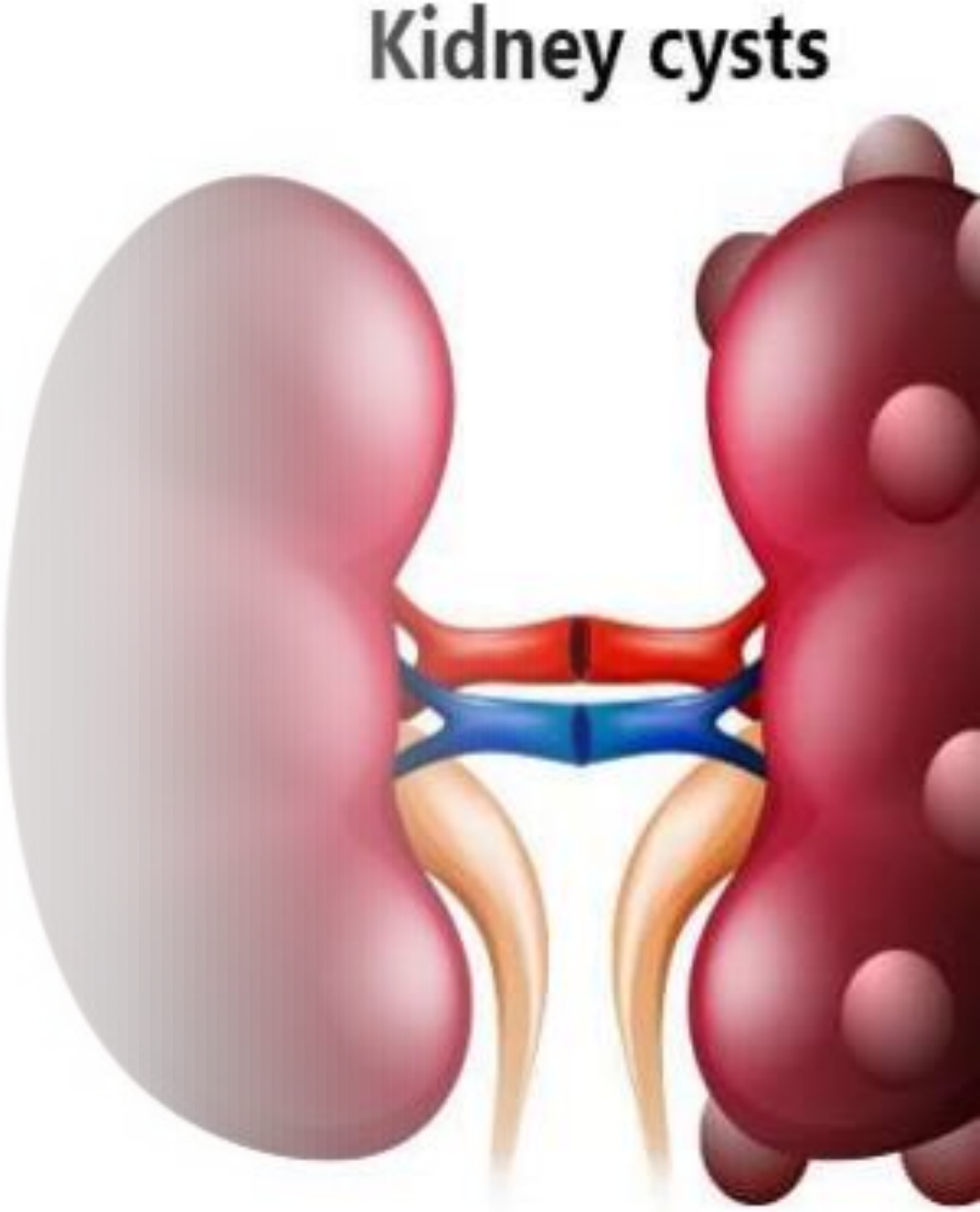


Cystic diseases of kidney

A heterogeneous group that could be:

1. Hereditary.

2. Developmental.

3. Acquired disorders.

Types of cysts:

1-Simple Cysts

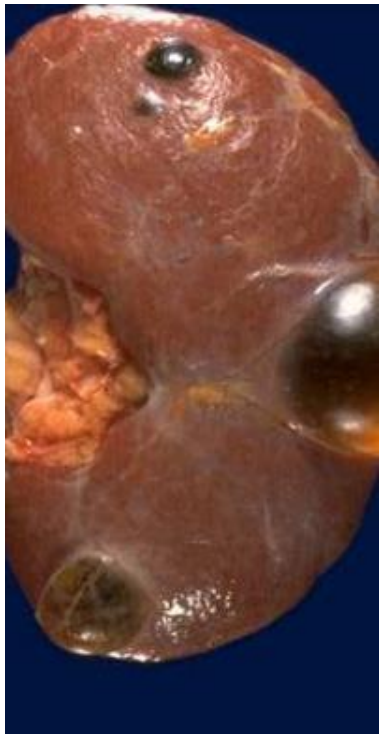
2-Dialysis-associated acquired cysts

3-Autosomal Dominant (Adult) Polycystic Kidney Disease

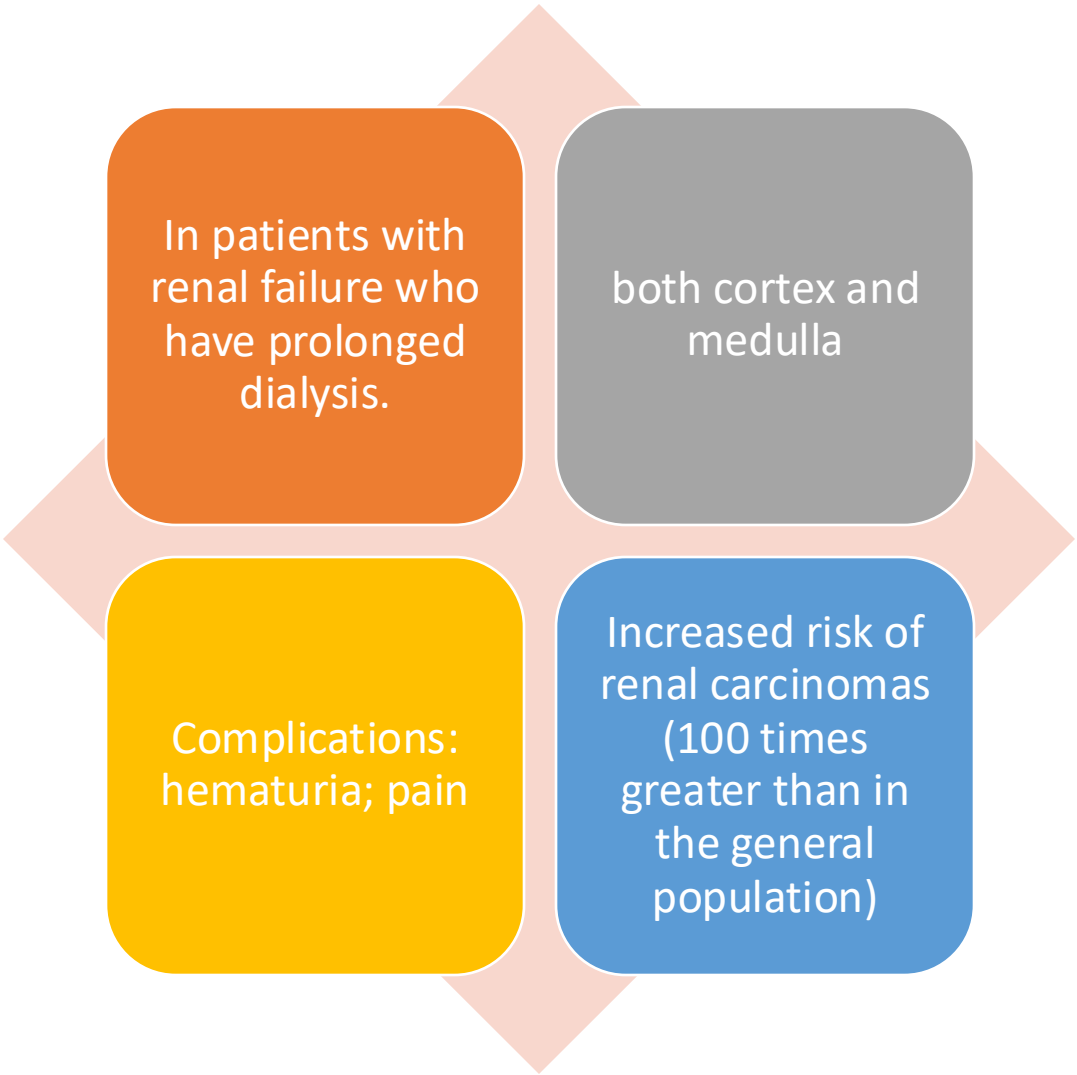
4-Autosomal Recessive (Childhood) Polycystic Kidney Disease

5-Medullary Cystic Disease

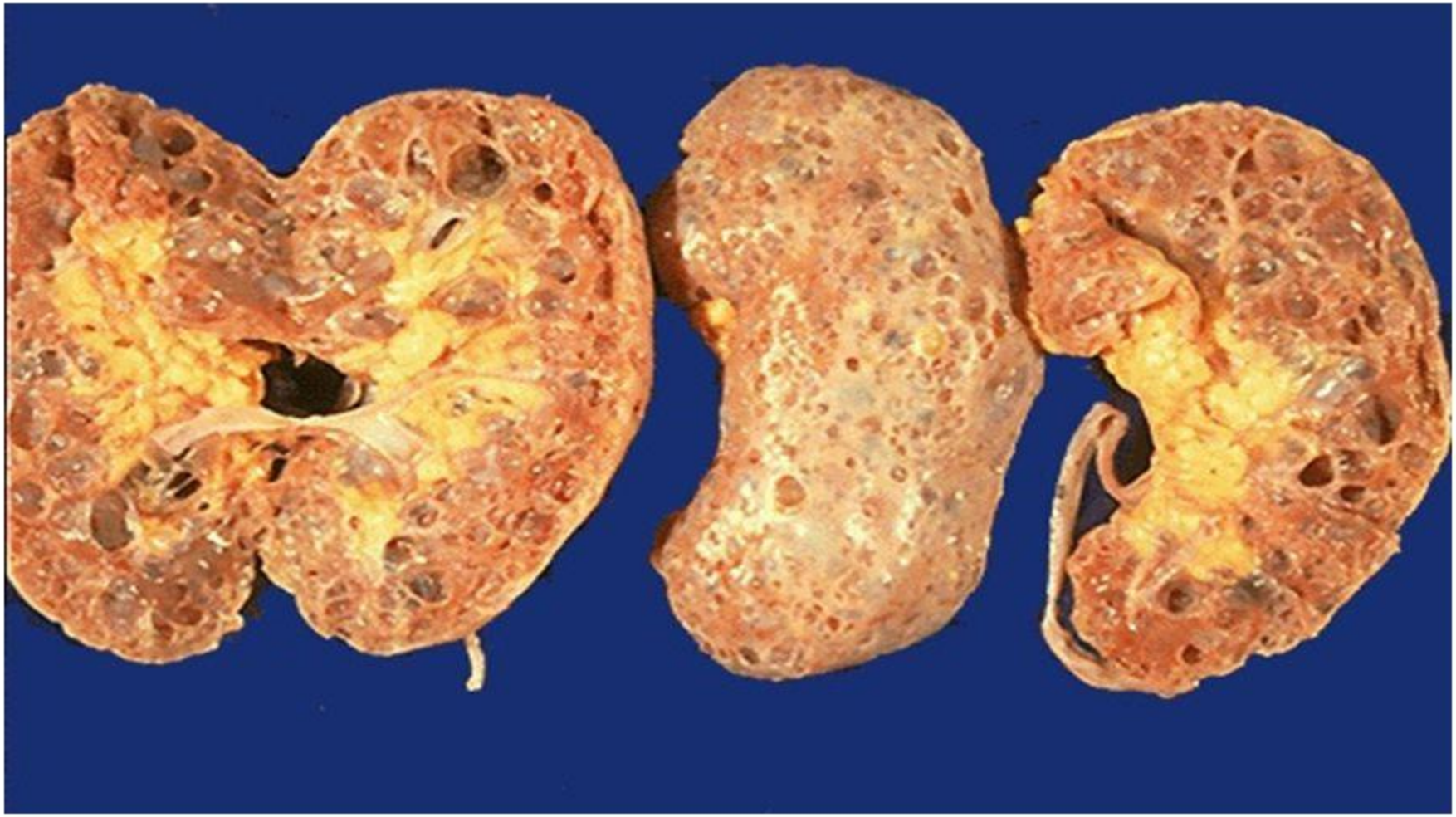
1- Simple Renal Cysts

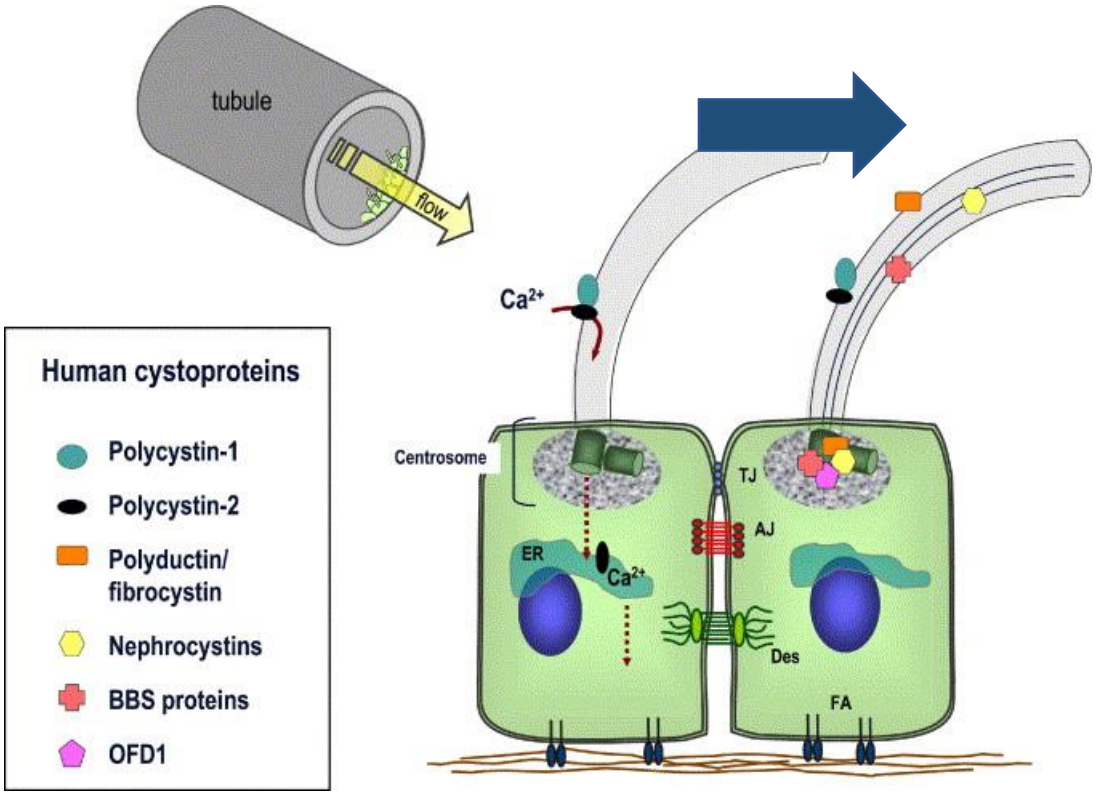


- **1-Simple Cysts :**
- **Multiple or single**
- **1-5 cm in diameter**
- **Filled with clear fluid.**
- **Confined to the cortex.**
- **No clinical significance.**
- **Usually discovered incidentally or because of hemorrhage and pain**
- **Importance: to differentiate from** kidney tumors


In 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)

Cystic change associated with chronic renal dialysis.





Hereditary cystic diseases:

Defect is in cilia–centrosome complex of tubular epithelial cells, interfere with fluid absorption & cellular maturation

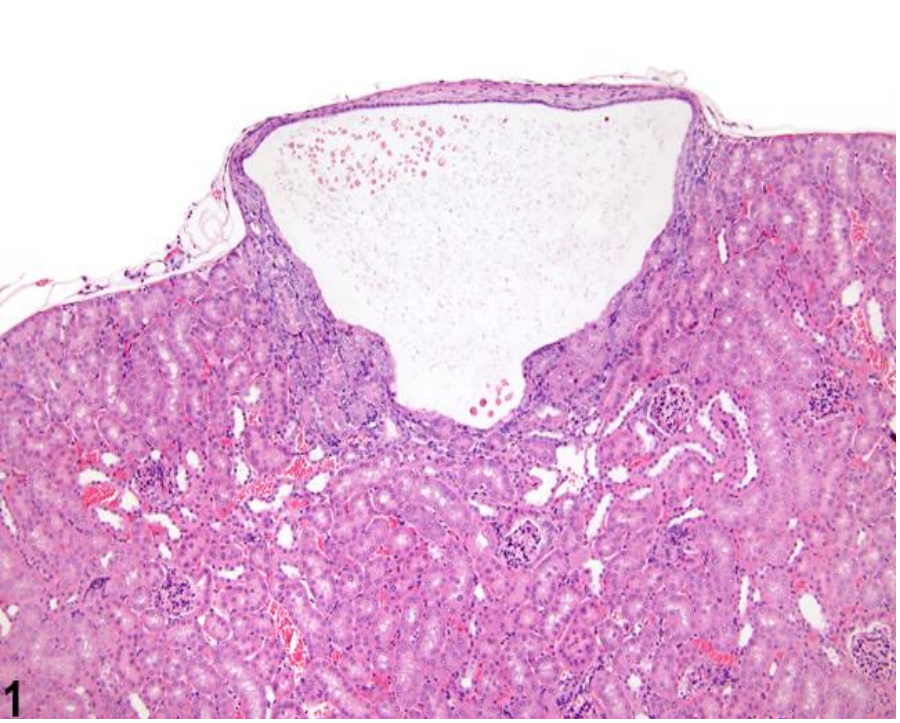
Cyst formation.

Most common form:

1. Simple cysts.
2. Polycystic kidney disease.

Microscopic examination:
Membranes are composed of single layer of cuboidal or flattened epithelium or completely atrophied.

Radiographic studies:
In contrast with renal tumors, renal cysts:
1. Have smooth contours.
2. Always avascular.
3. Produce fluid rather than solid tissue signals on ultrasonography.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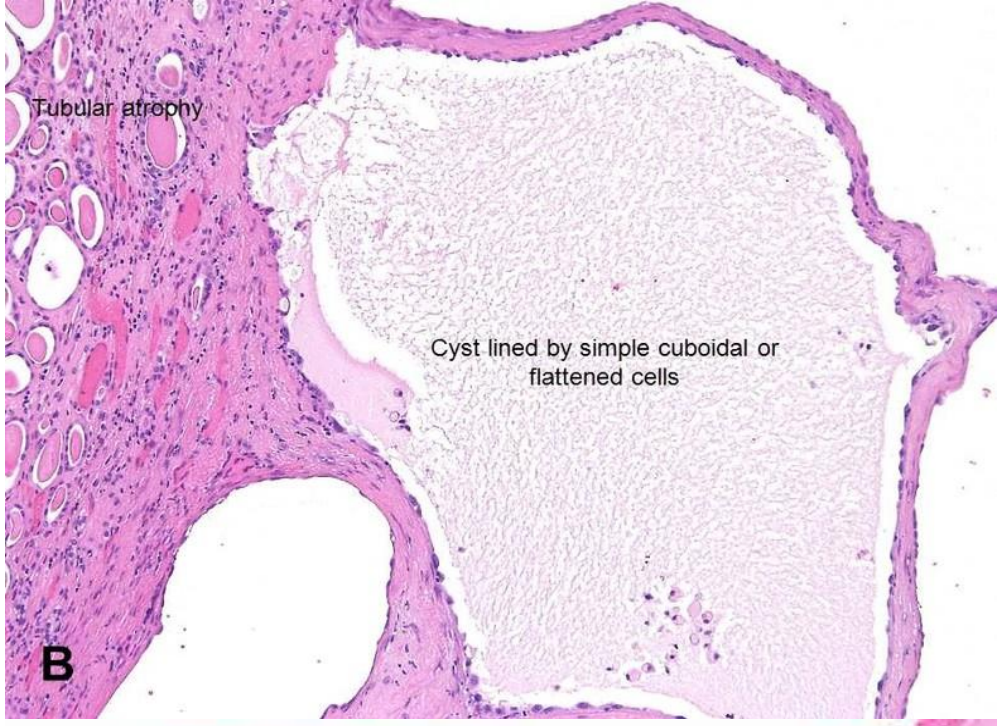
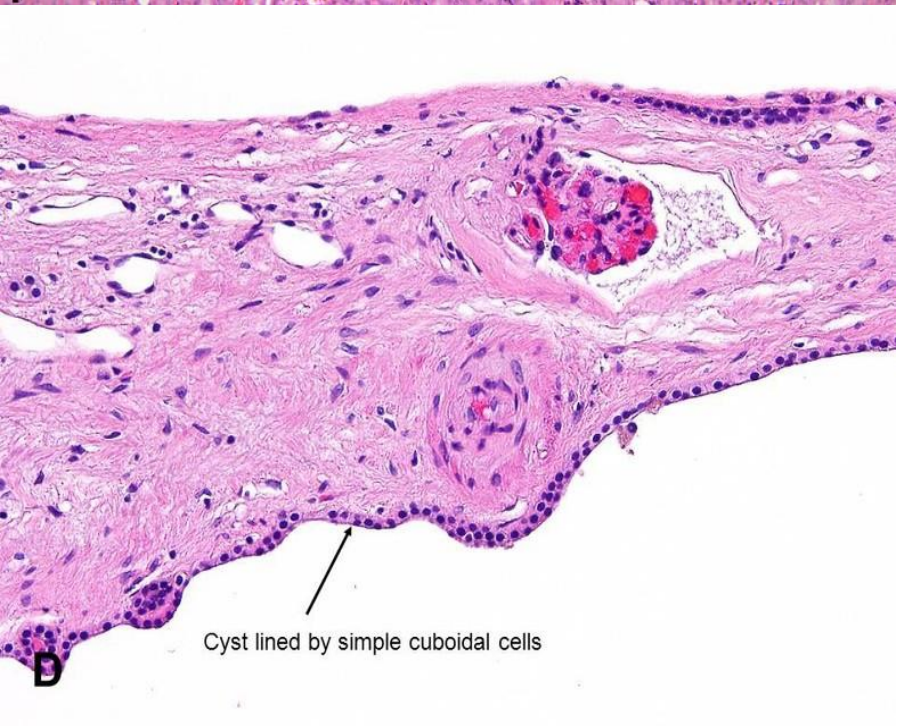
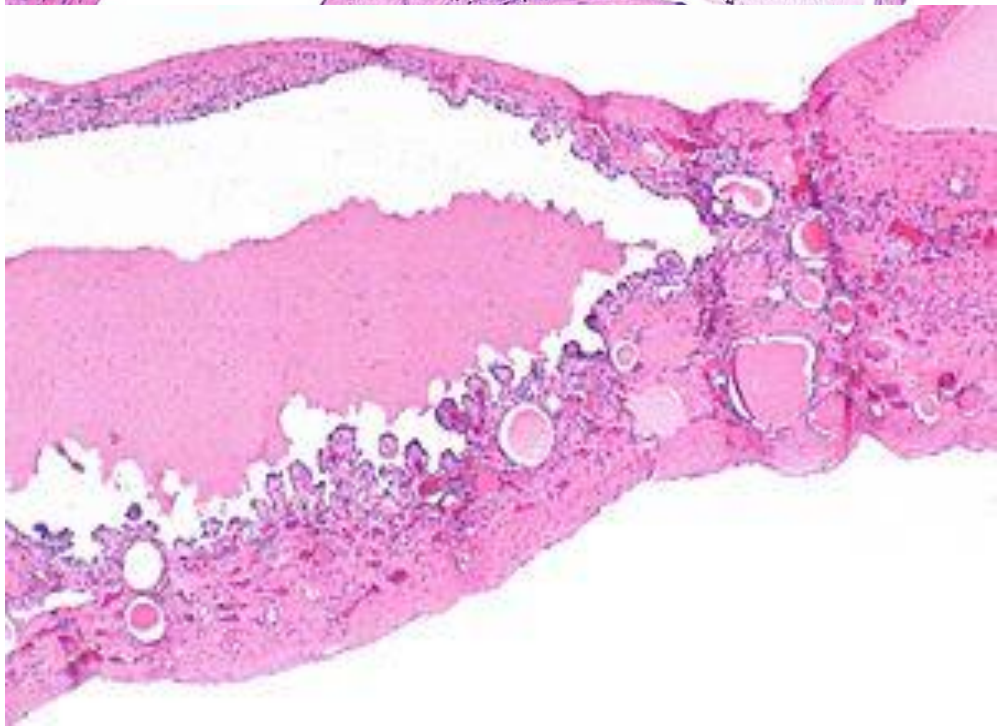
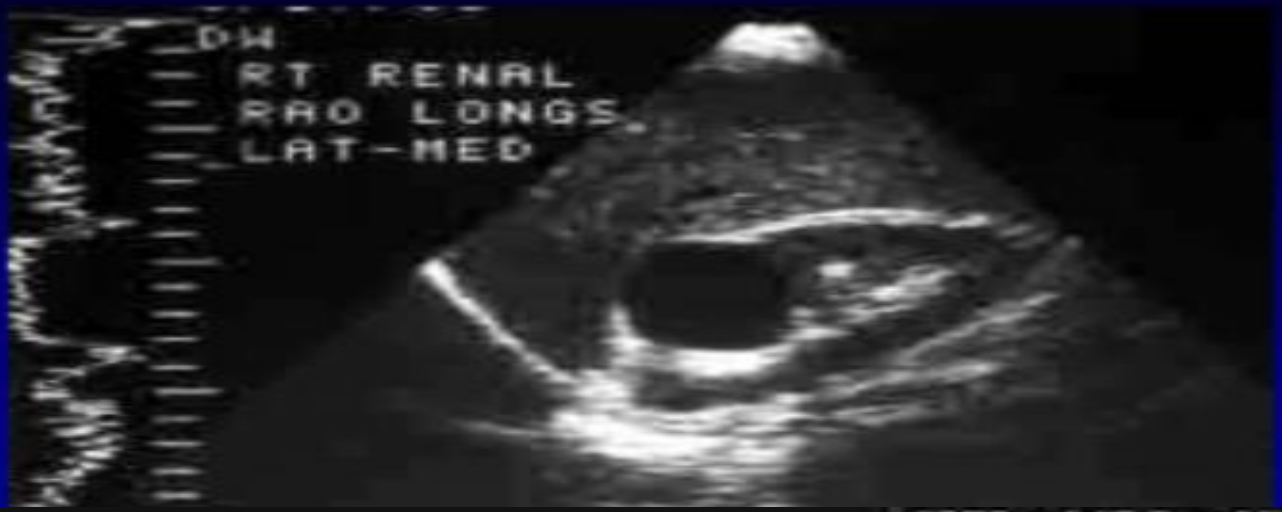
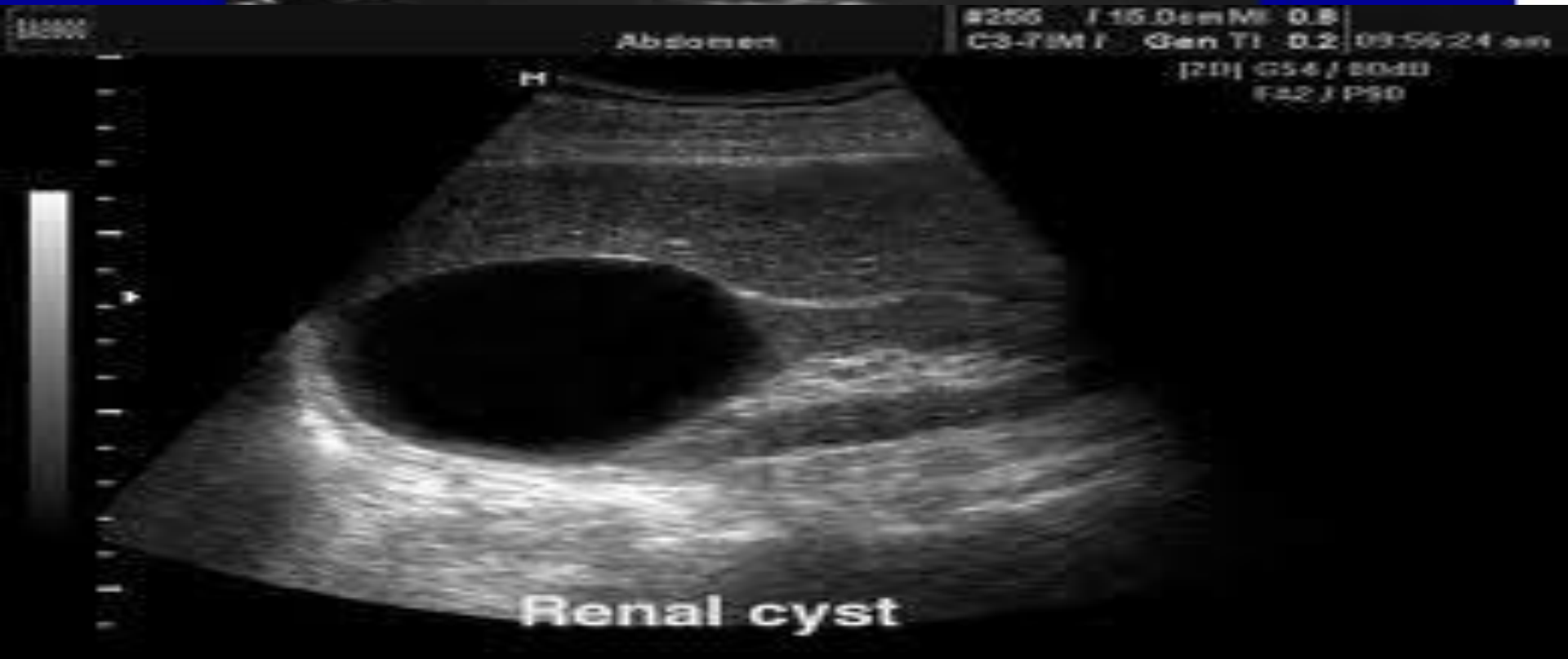

Tubular atrophy
Cyst lined by simple cuboidal or flattened cells
B


Cyst lined by simple cuboidal cells
D

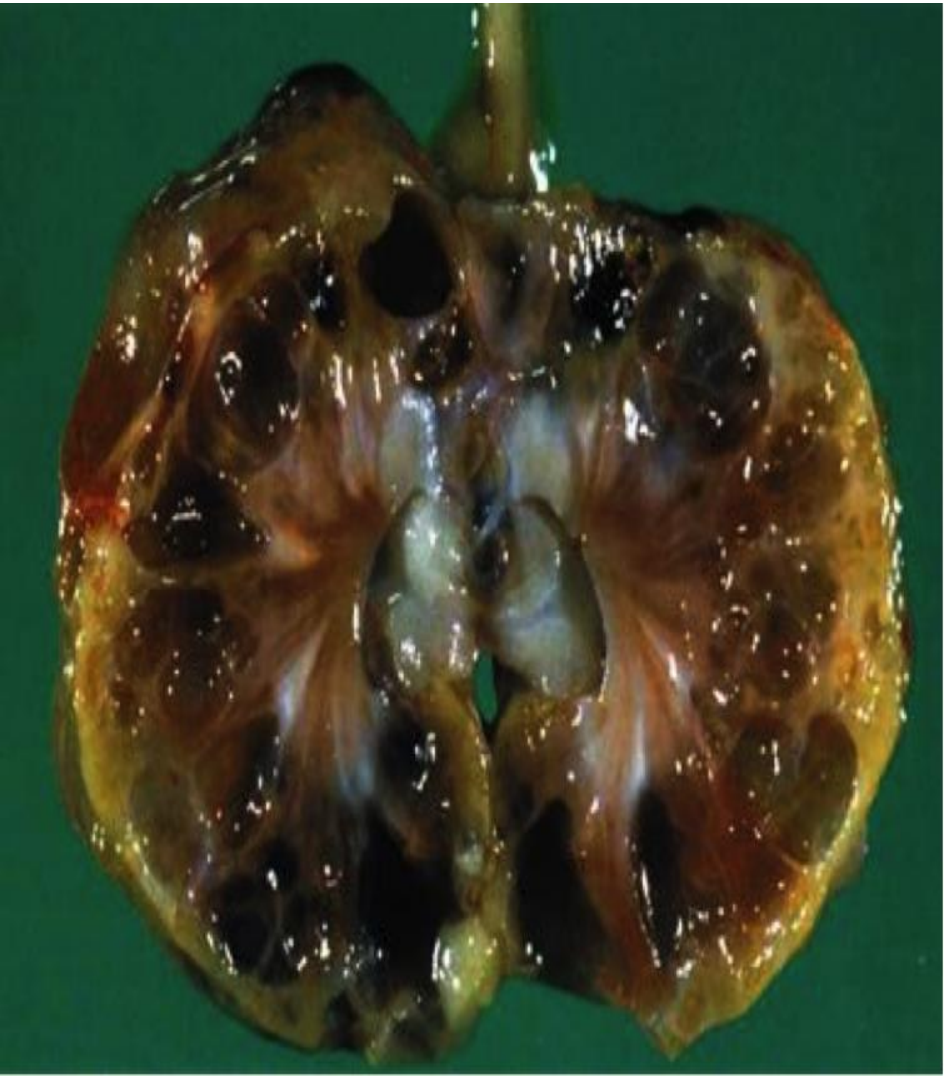


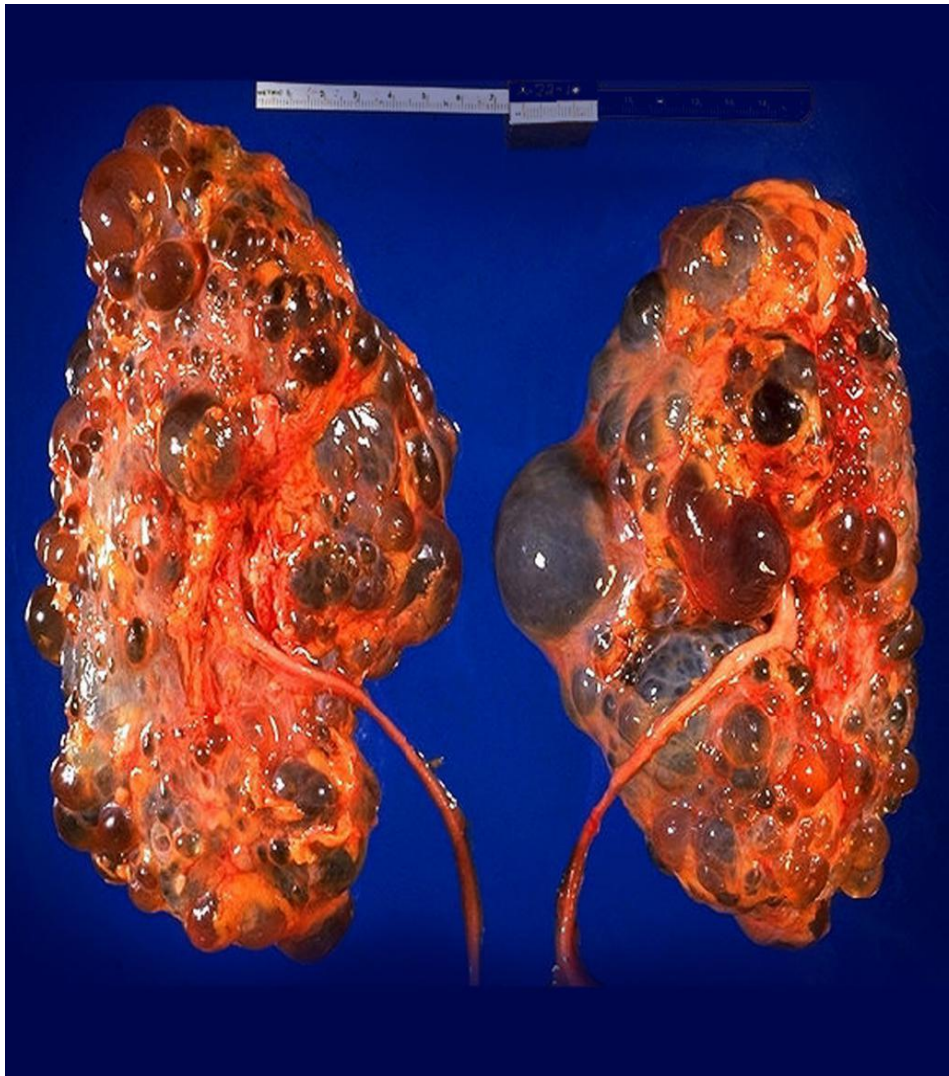
SIMPLE RENAL CYSTS – U/S





3- Autosomal Dominant (Adult) Polycystic Kidney Disease





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).

*Clinical presentation :

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

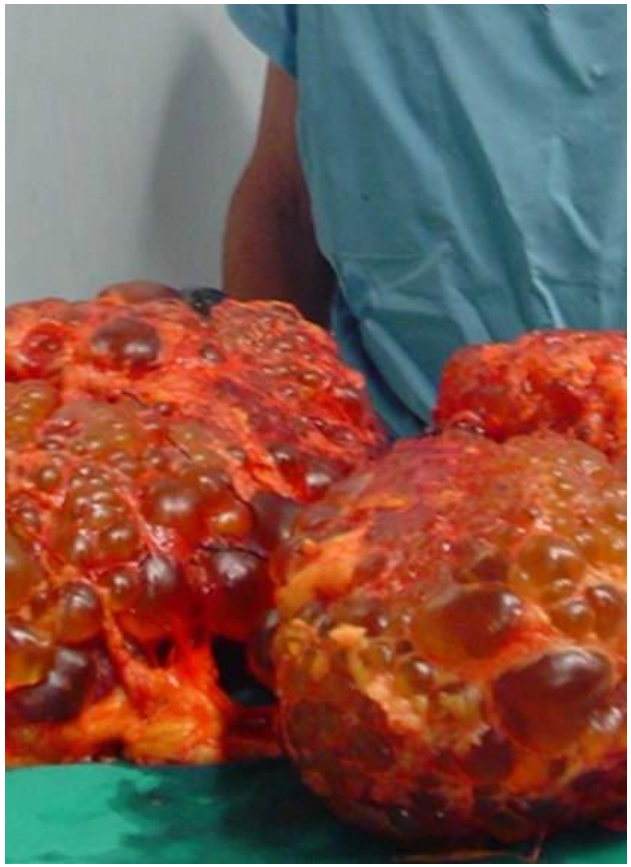
*Complications

- 1- hypertension (75%)
- - urinary infection
- 3- vascular aneurysms of circle of Willis (10% -30%) (subarachnoid hemorrhage).
- 4- renal failure at age 50

Morphology

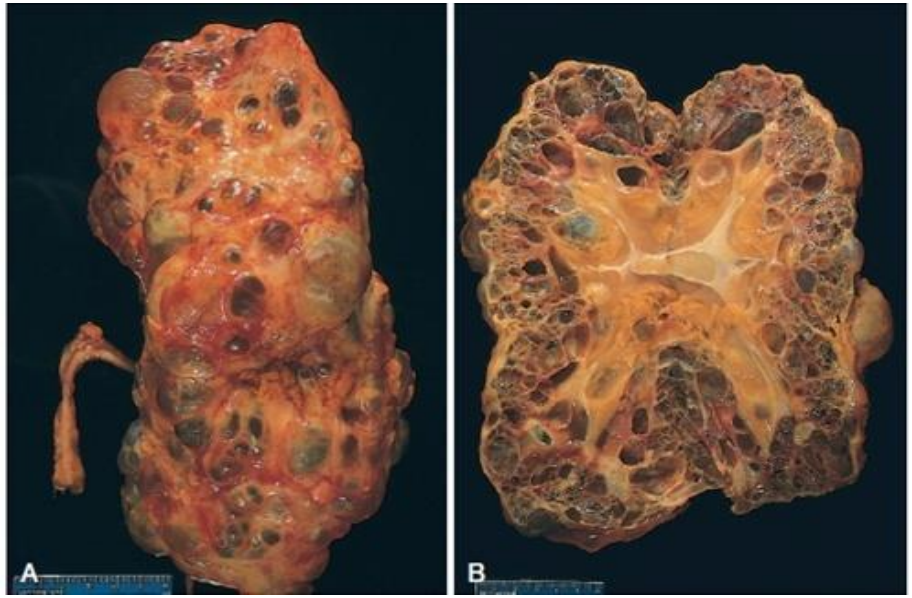
- Kidney reach very large size & weights as 4kg. (Normal: 125-170 mg)
- Large kidneys are palpable abdominally as masses extending into the pelvis.

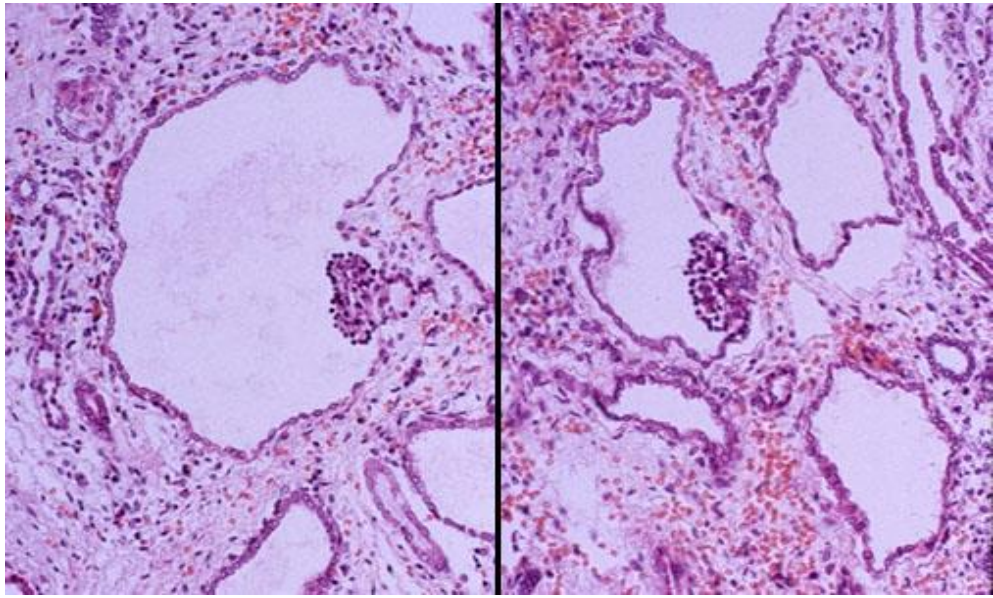
- Gross examination:
- Kidney is composed of mass of cysts of various sizes up to 3 or 4 cm in diameter.
- No intervening parenchyma.
- Cysts are filled with fluid:
- Clear, turbid, or hemorrhagic.

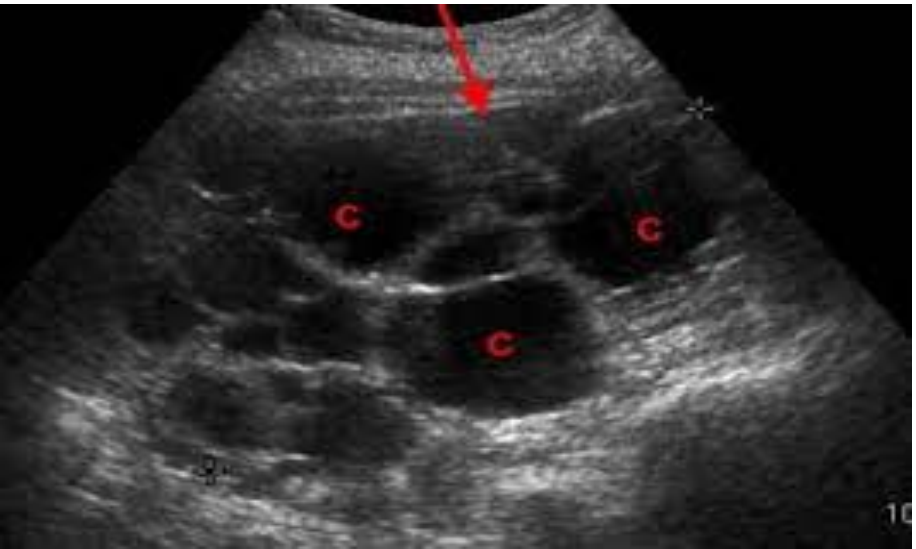


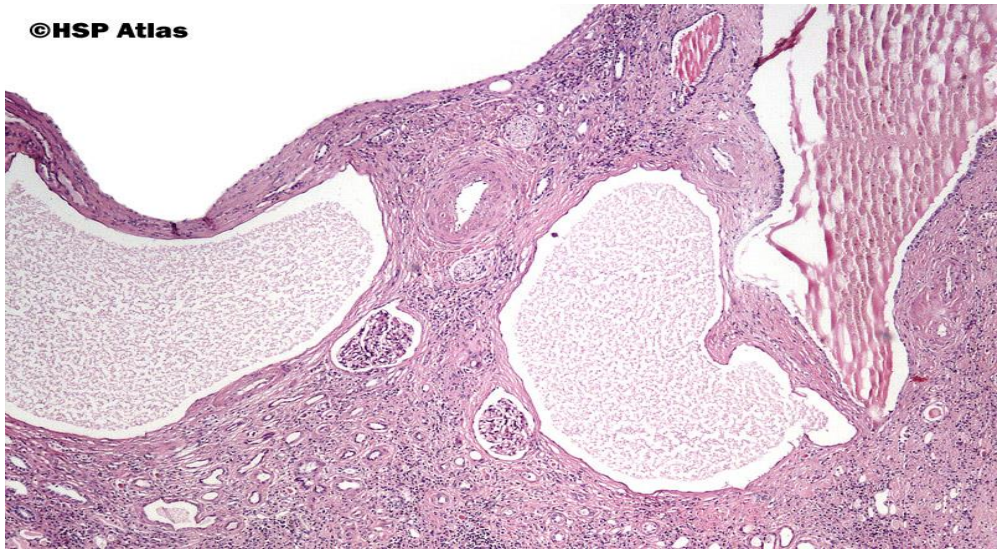
Autosomal dominant adult polycystic kidney:

Kidney is enlarged, with numerous dilated cysts.









4-Autosomal Recessive (Childhood) Polycystic Kidney Disease

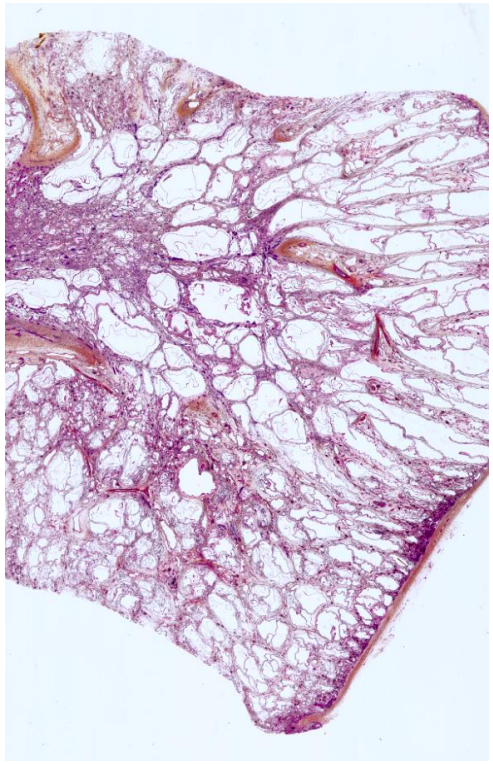
- Autosomal recessive
- 1:20,000 live births.
- Types: perinatal, neonatal, infantile, and juvenile.
- Associated with liver cysts
- Mutations in PKHD1 gene coding for fibrocystin.
- Fibrocystin may be involved in the function of cilia in tubular epithelial cells .

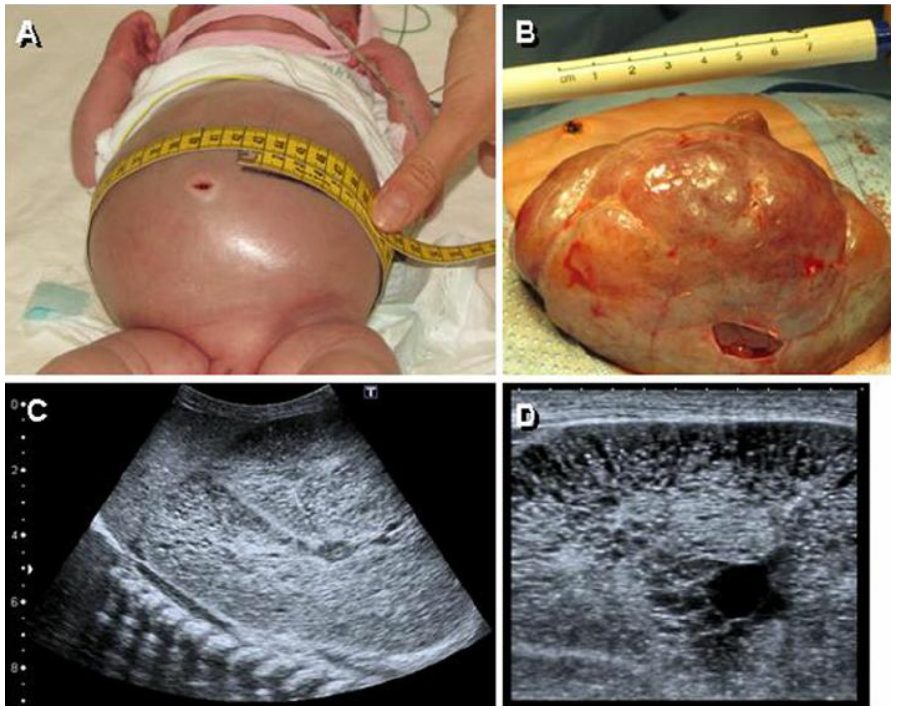
Autosomal recessive (childhood) polycystic Kidney disease

- **Perinatal: Period immediately before & after birth**
- **Neonatal: Newborn.**
- **Infantile: babies or very young children.**
- **Juvenile: Youth or young person.**

Baby with autosomal recessive polycystic kidney disease (ARPKD).
A. Distended abdomen due to
large kidneys.

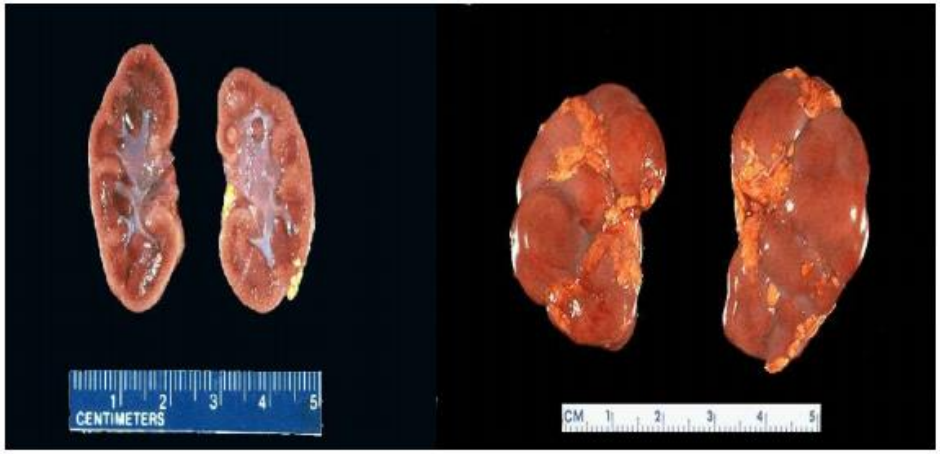
B. Nephrectomized kidney.





Normal vs childhood polycystic kidneys

NORMAL TERM INFANT KIDNEYS



CHILDHOOD) POLYCYSTIC KIDNEYS

Adult vs childhood polycystic kidney disease

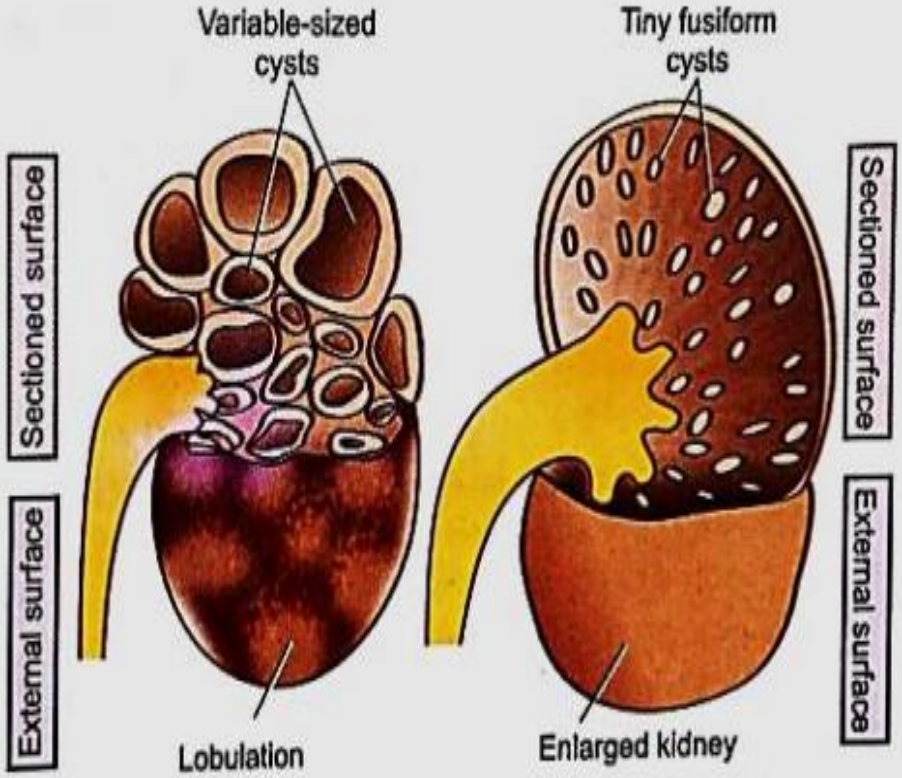
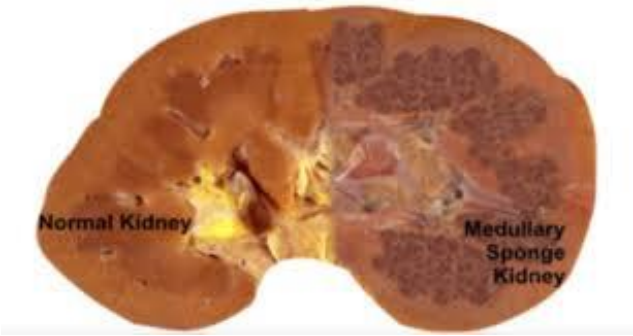
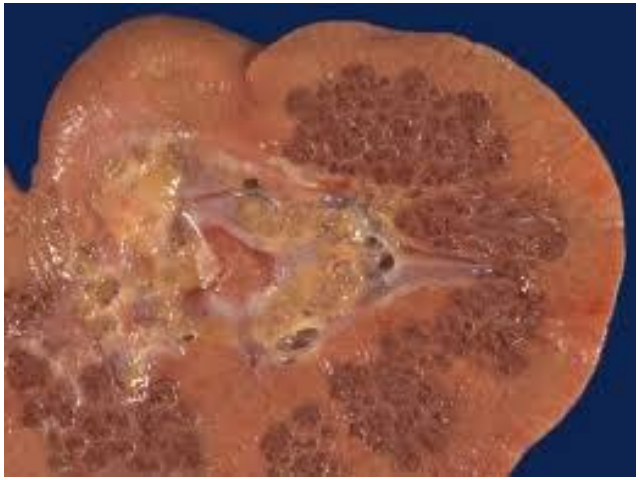


Figure 22.8 ◆ Polycystic kidney disease. Diagrammatic representation of comparison of gross appearance of the two main forms.





5-Medullary Cystic Diseases

Two major types:

1-Medullary sponge kidney

2-Nephronophthisis-medullary cystic disease complex

- almost always associated with renal dysfunction.

- usually begins in childhood.

- Cysts are at cortico-medullary junction.

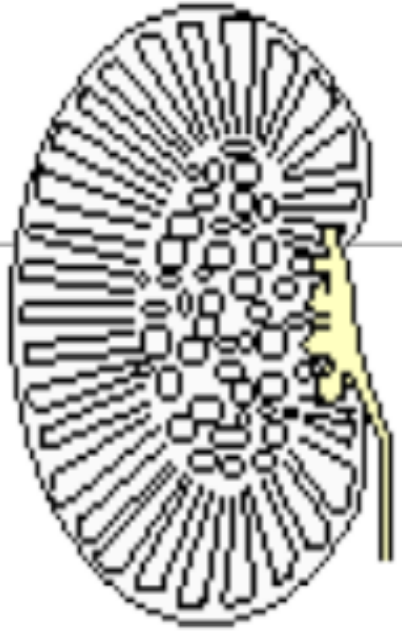
Clinical Course

- 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

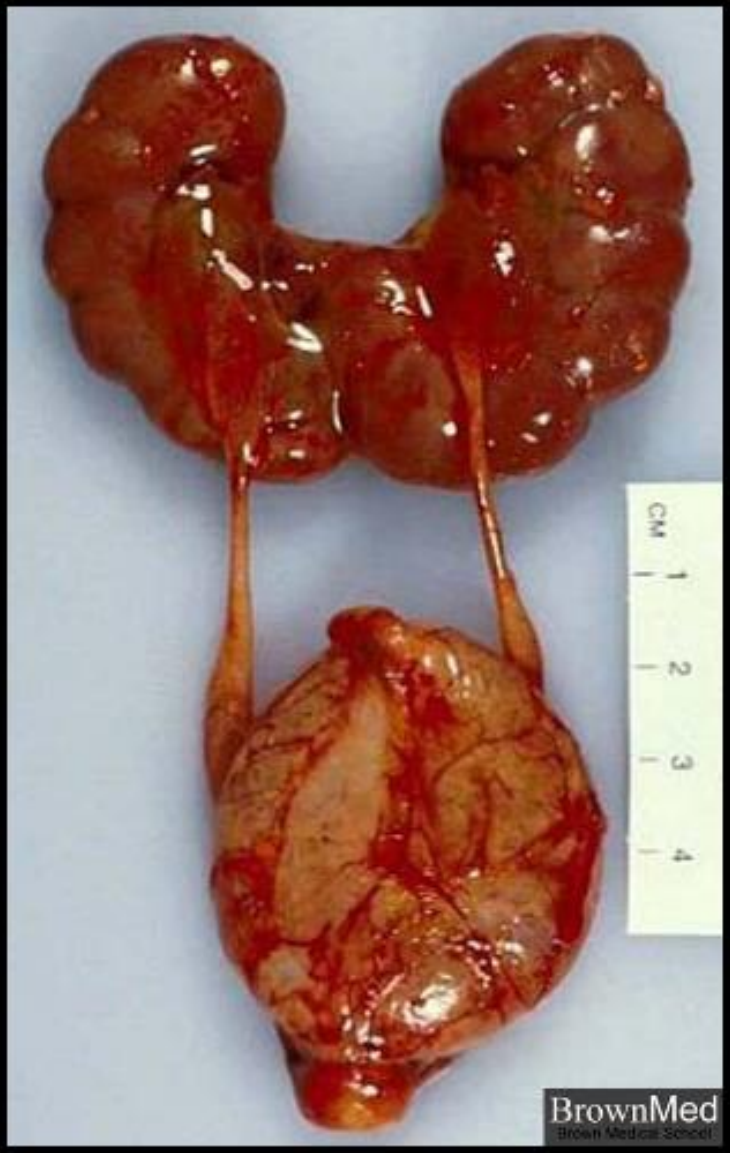
Congenital anomalies

- **Agenesis** :Complete absence of renal tissue; unilateral or bilateral.
- **Bilateral agenesis:** incompatible with life; associated with large adrenal glands; leads to Potter (oligohydramnios) sequence.
- **Unilateral agenesis:** not fatal
- **Duplication of ureters** : < 1% of individuals
- **Ectopic (displaced) kidneys** :Usually at pelvic brim
- **Horseshoe kidney** :Most common congenital kidney anomaly, 0.15 - 0.25% of all newborns , 90% are fused at lower pole. Associated with obstruction, anomalous superior vena cava . Complete fusion of the kidneys produces a formless mass in the pelvis (pancake kidney)
- **Hypoplasia** : Rare; failure of kidney to develop to normal size without scarring.

CONGENITAL ANOMALIES OF THE KIDNEY

- Congenital cystic diseases
- *Multicystic dysplasia* is the **most common** form of renal cystic disease in childhood.
- The term *dysplasia* refers to a developmental rather than a preneoplastic lesion.
- Often associated with obstruction in the lower urinary tract, increased hydrostatic pressure in the developing kidney is thought to play a role in its development.
- The kidneys are usually grossly distorted; the cysts range from microscopic to several centimeters in diameter.

Fused kidney (horseshoe)




Thank you
Good luck