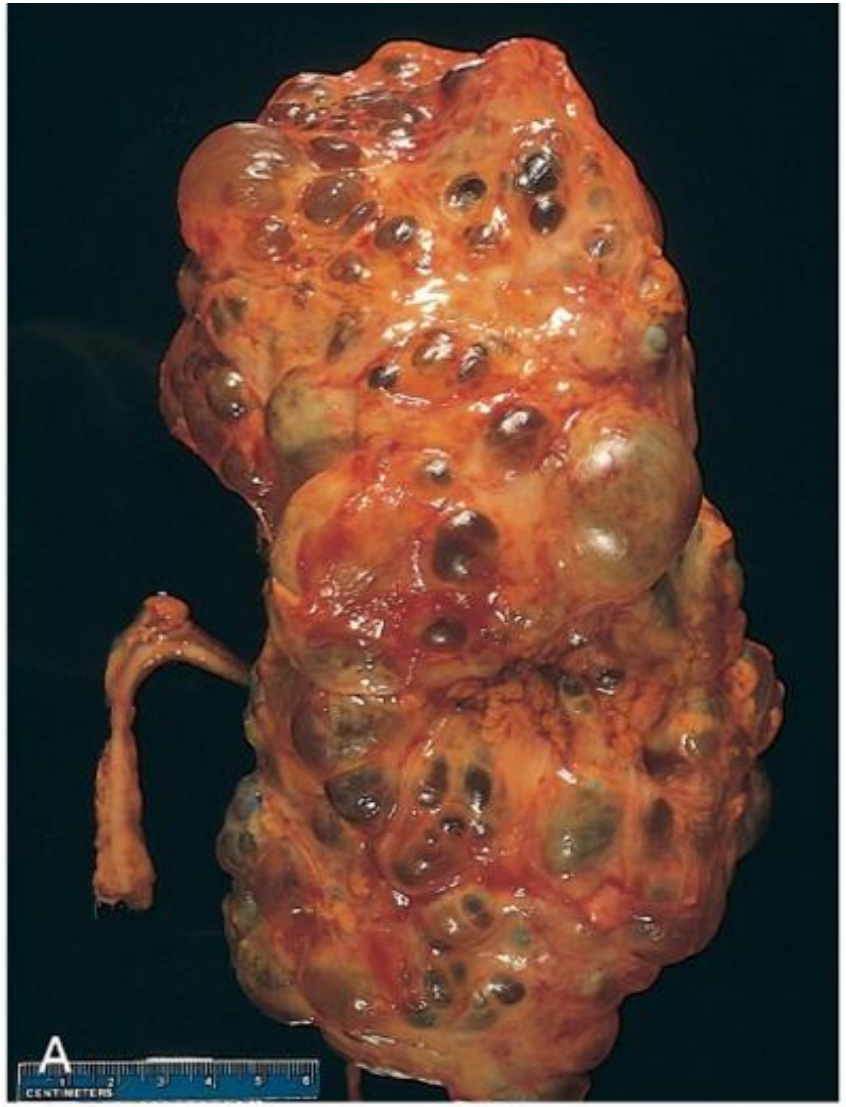
THE UNIVERSITY OF JORDAN

CYSTIC DISEASES of THE KIDNEY

Dr. Nisreen Abu Shahin



Types of cysts

1-Simple Cysts

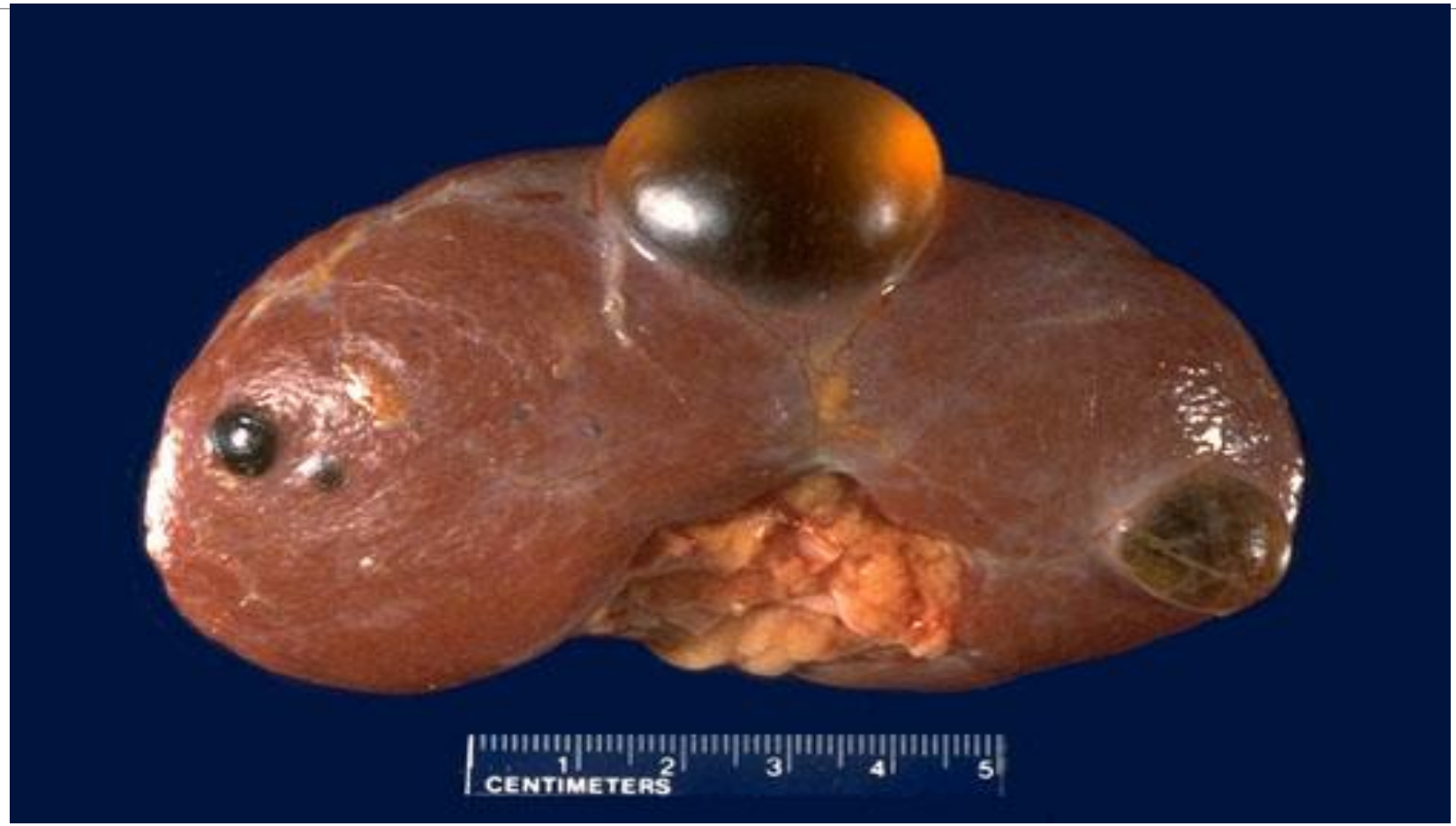
2-Dialysis-associated acquired cysts

3-Autosomal Dominant (Adult) Polycystic Kidney Disease

4-Autosomal Recessive (Childhood) Polycystic Kidney Disease

5-Medullary Cystic Disease

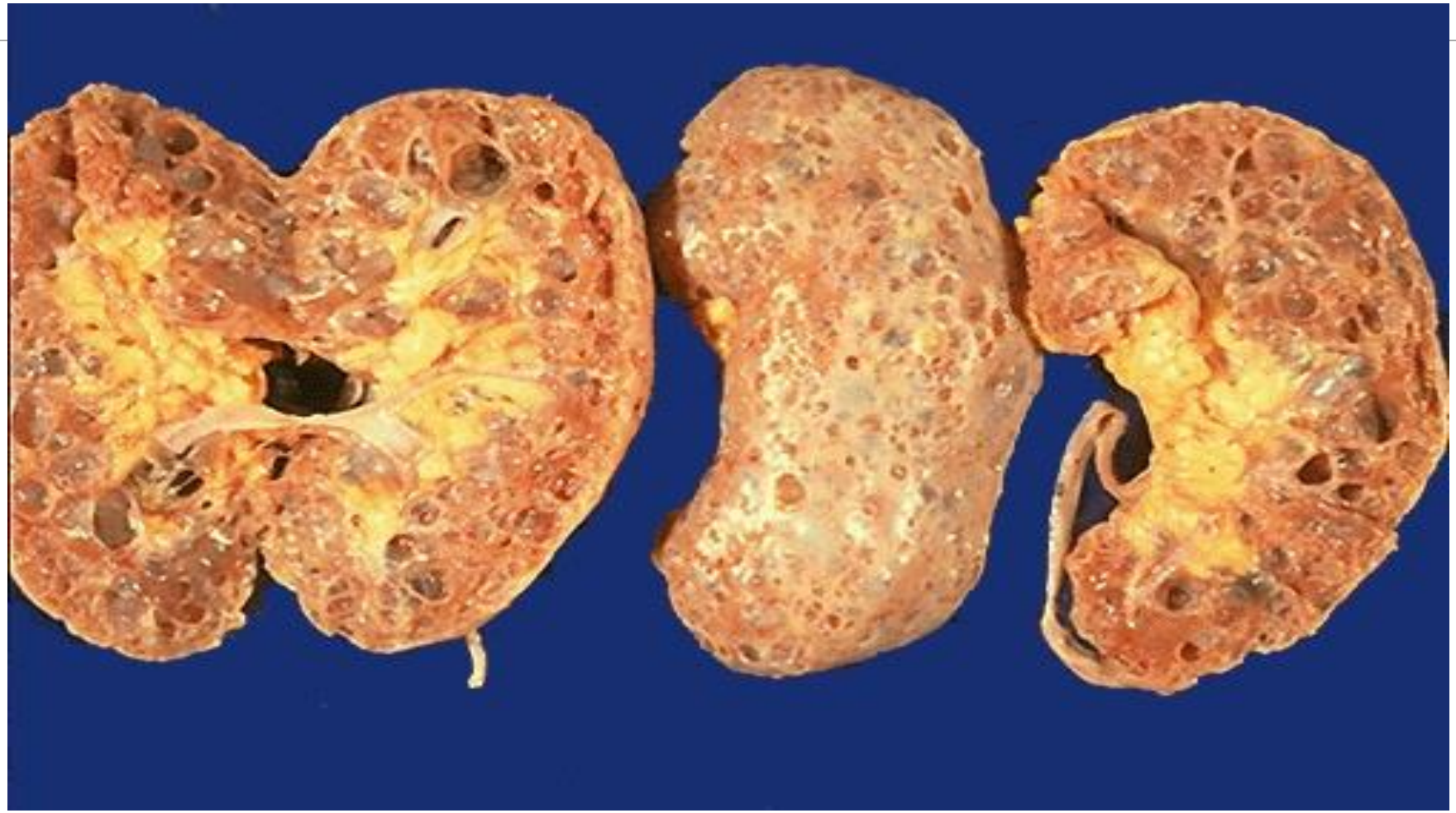
Simple renal Cysts



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

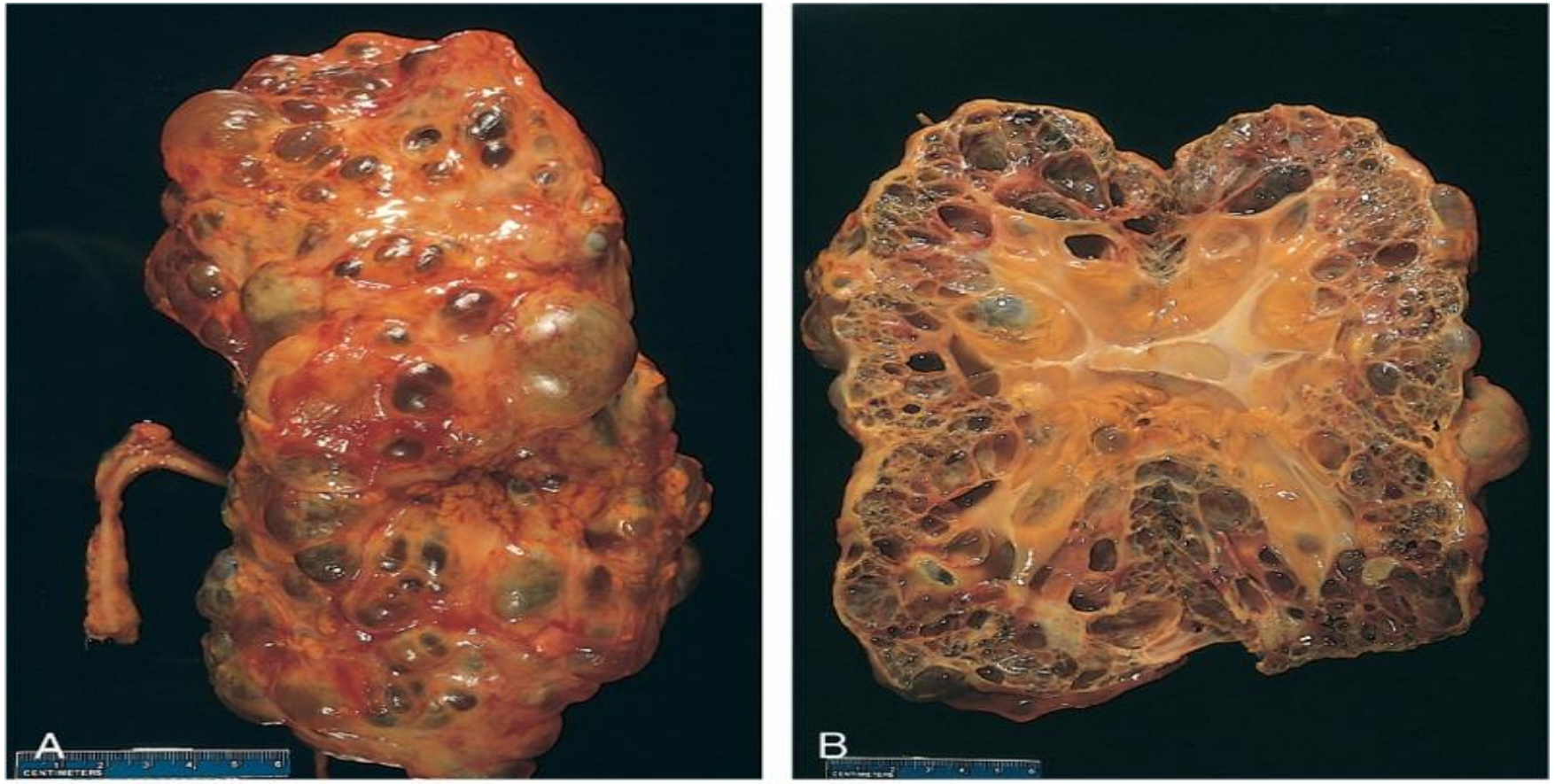
Cysts associated with chronic dialysis



2-Dialysis-associated acquired cysts

- **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)

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).***

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

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*

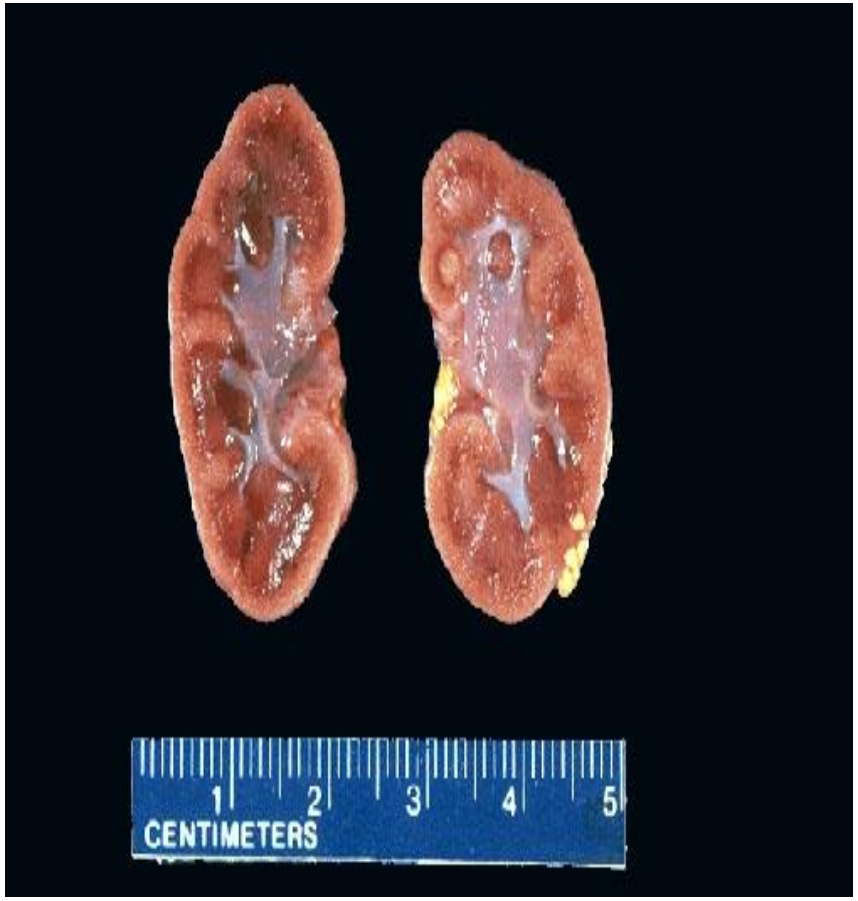
3- vascular aneurysms of circle of Willis (10% -30%)→ (subarachnoid hemorrhage).

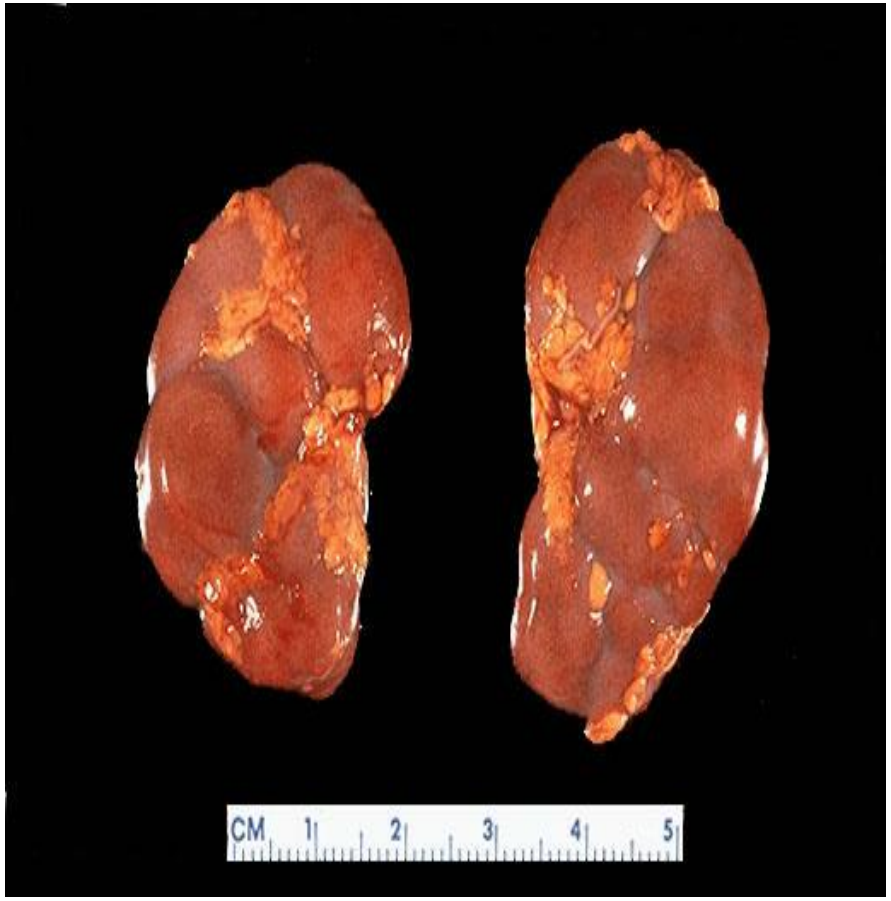
4- renal failure at age 50

4-Autosomal Recessive (Childhood) Polycystic Kidney Disease

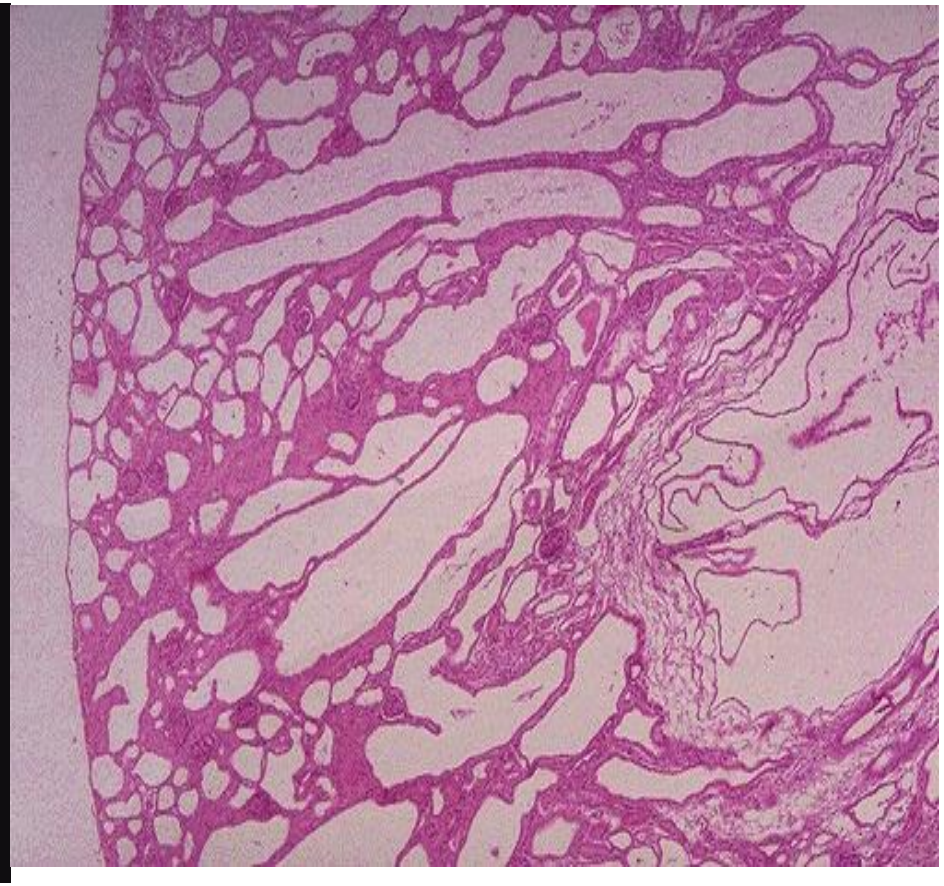
- **autosomal recessive**
- **1:20,000 live births.**
- **Types: perinatal, neonatal, infantile, and juvenile.**
- **Associated with liver cysts**
- **Mutations in *PKHD1* gene coding for *fibrocystin*.**
- **Fibrocystin may be involved in the function of cilia in tubular epithelial cells .**

Normal term infant kidneys





Autosomal Recessive (Childhood) Polycystic Kidney Disease:



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**

5- Medullary Cystic Disease

- **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.

URINARY OUTFLOW OBSTRUCTION

- **Renal Stones (*Urolithiasis)***

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

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

❖ **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%)

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).

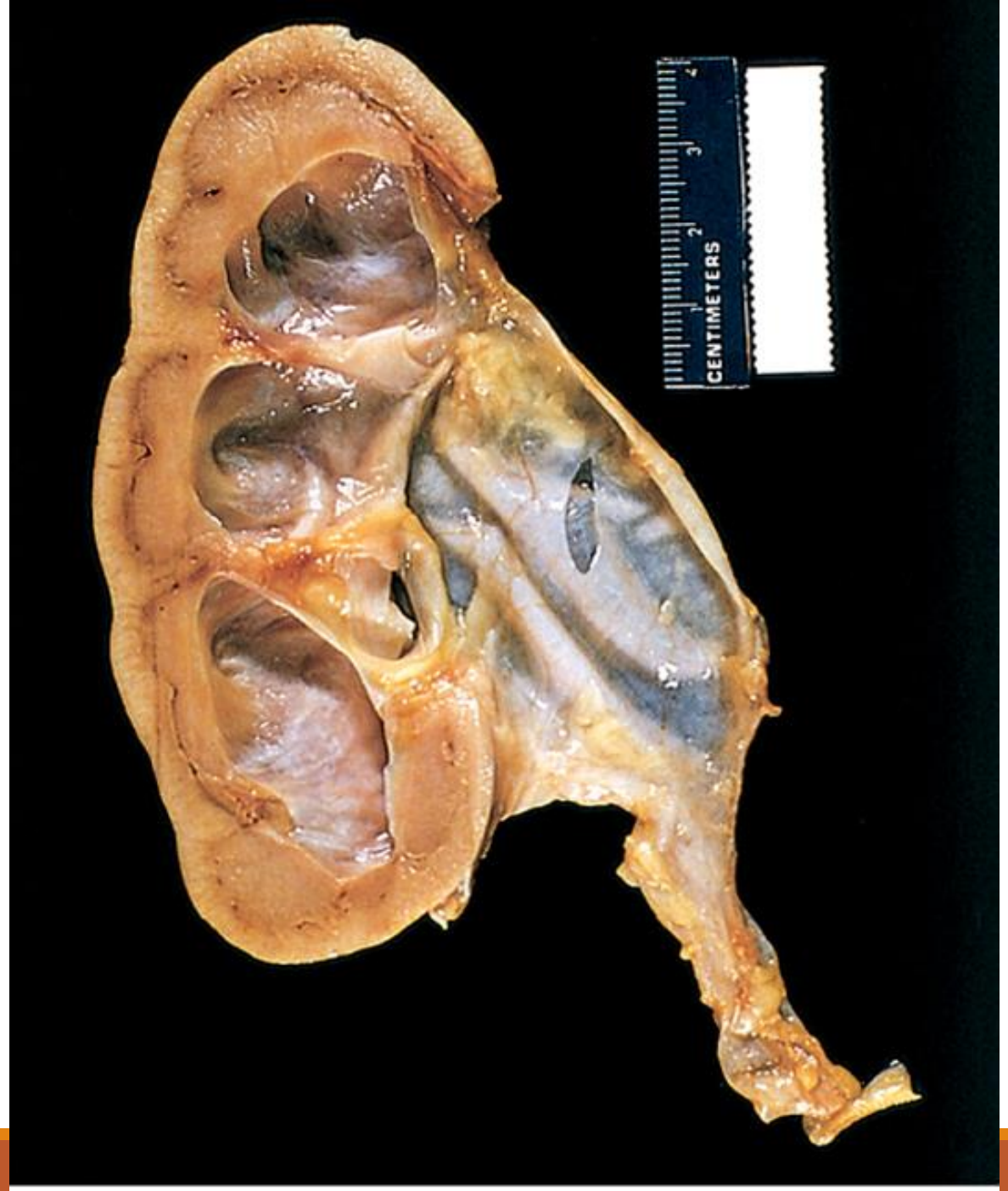
4-infections

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
- **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