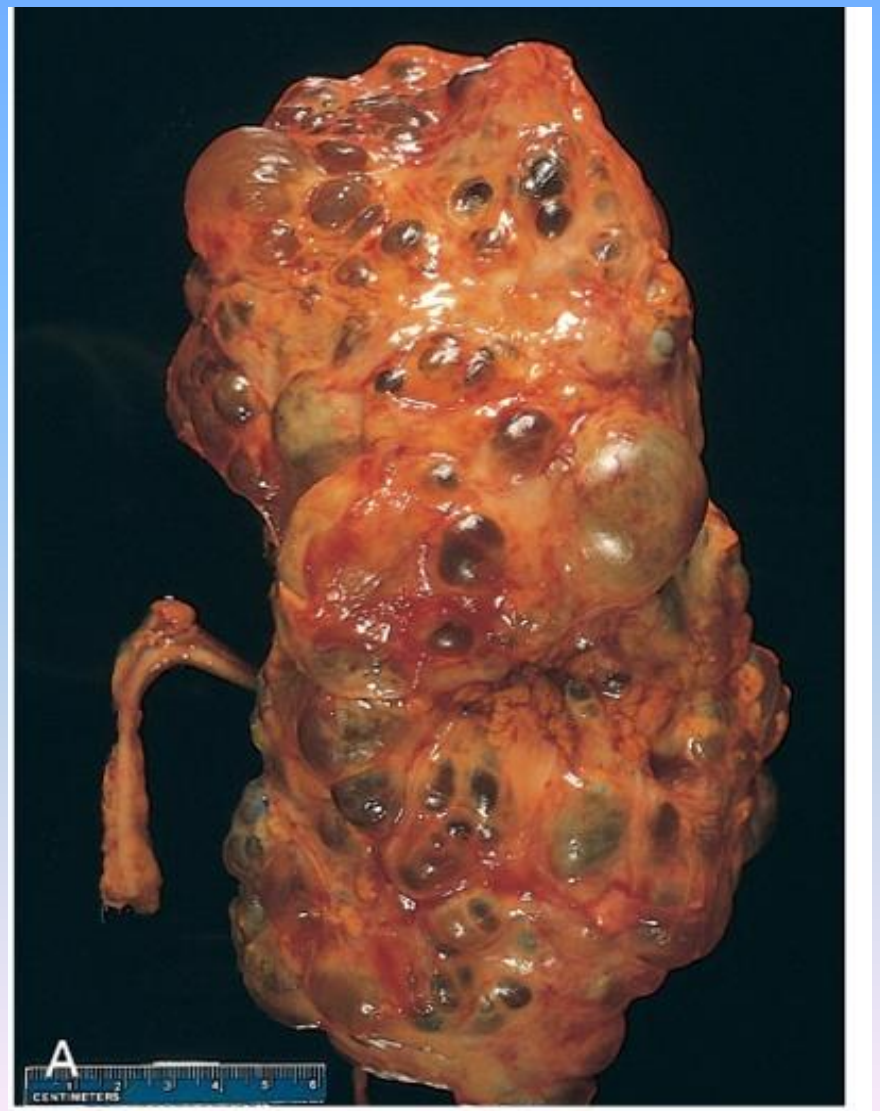
CYSTIC DISEASES OF THE KIDNEY



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- **Types of cysts**

1-Simple Cysts

2-Dialysis-associated acquired cysts

3-Autosomal Dominant (Adult) Polycystic Kidney Disease

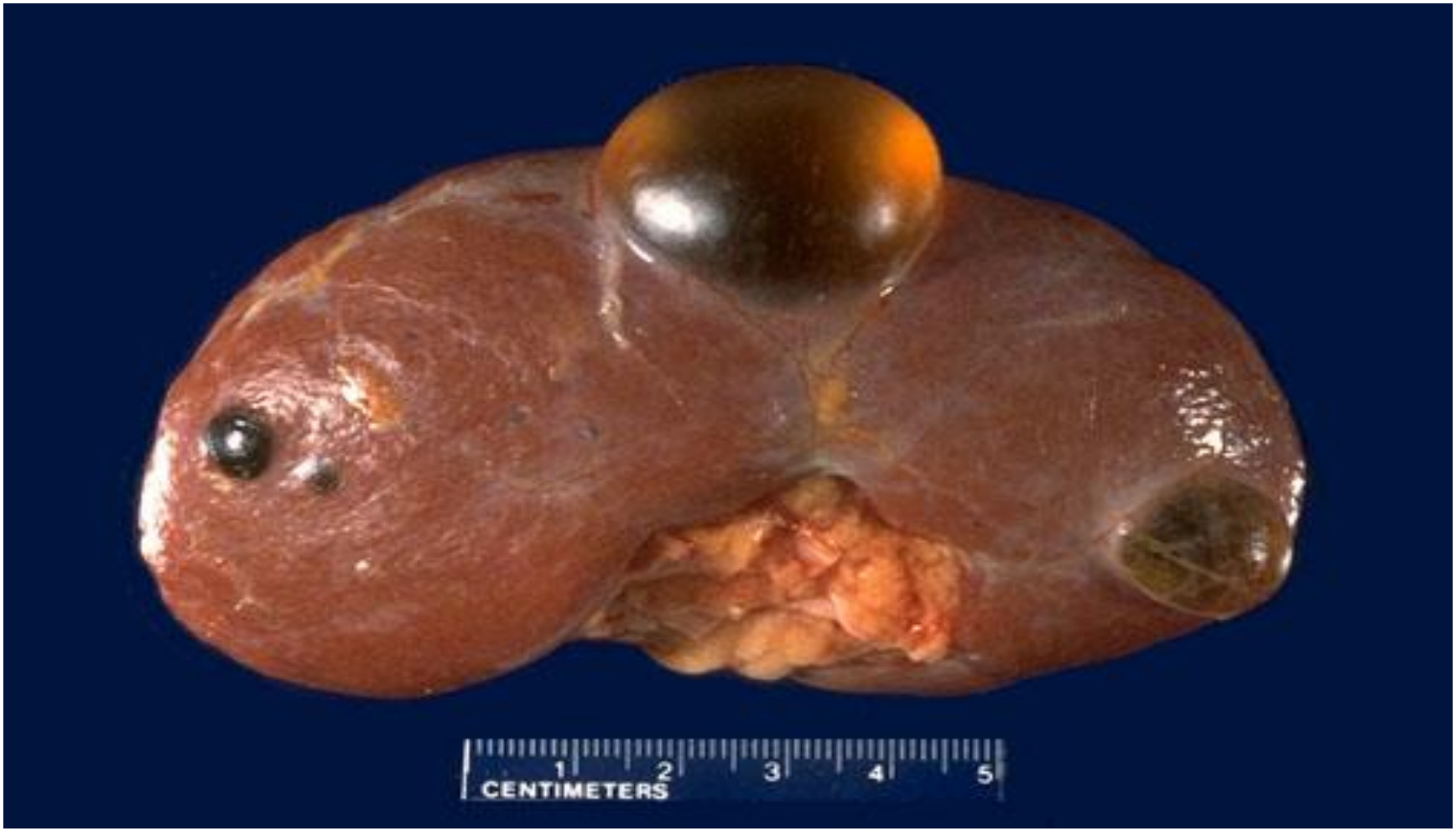
4-Autosomal Recessive (Childhood) Polycystic Kidney Disease

5-Medullary Cystic Disease

1-Simple 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**

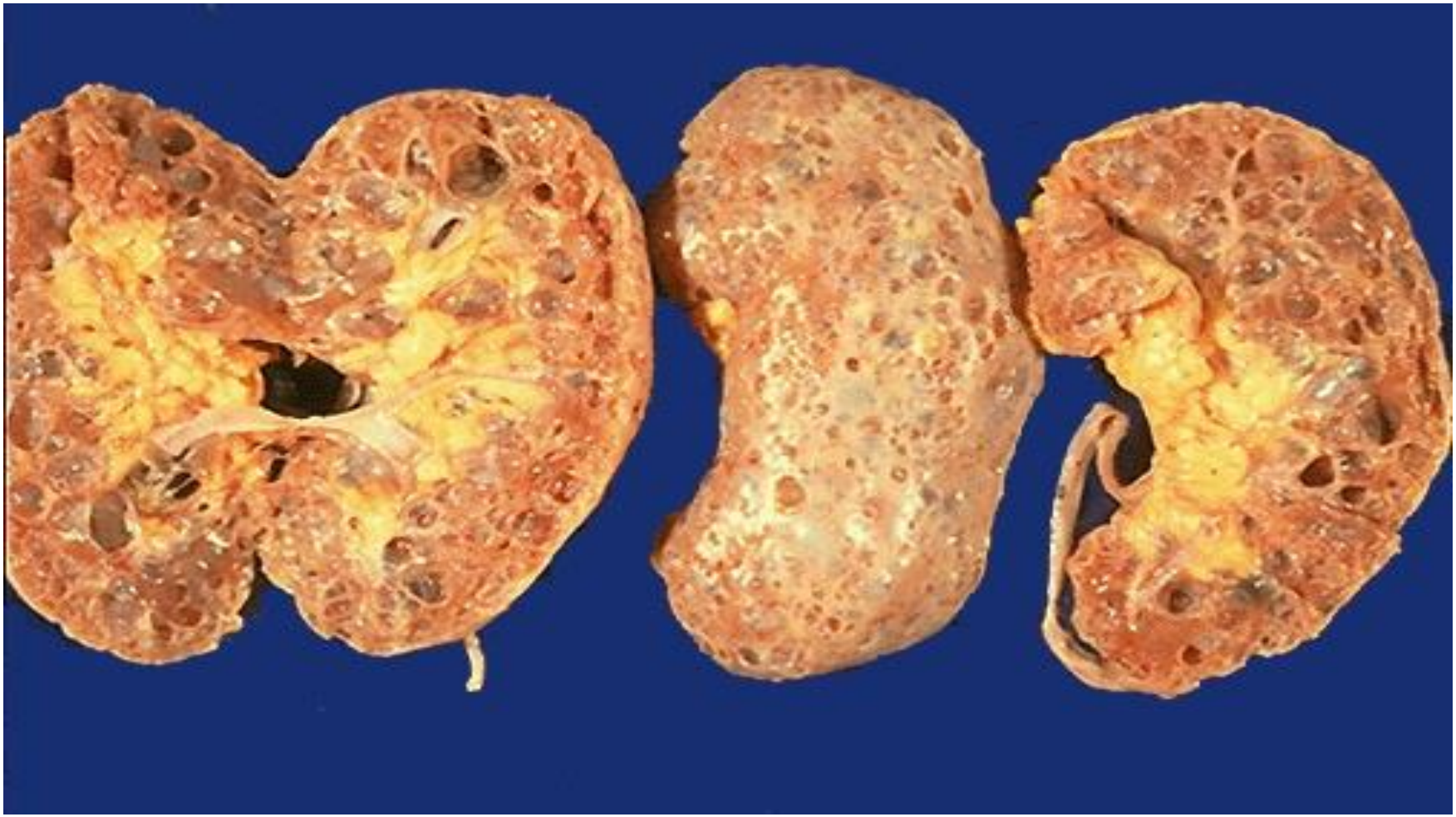
Simple renal Cysts



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)

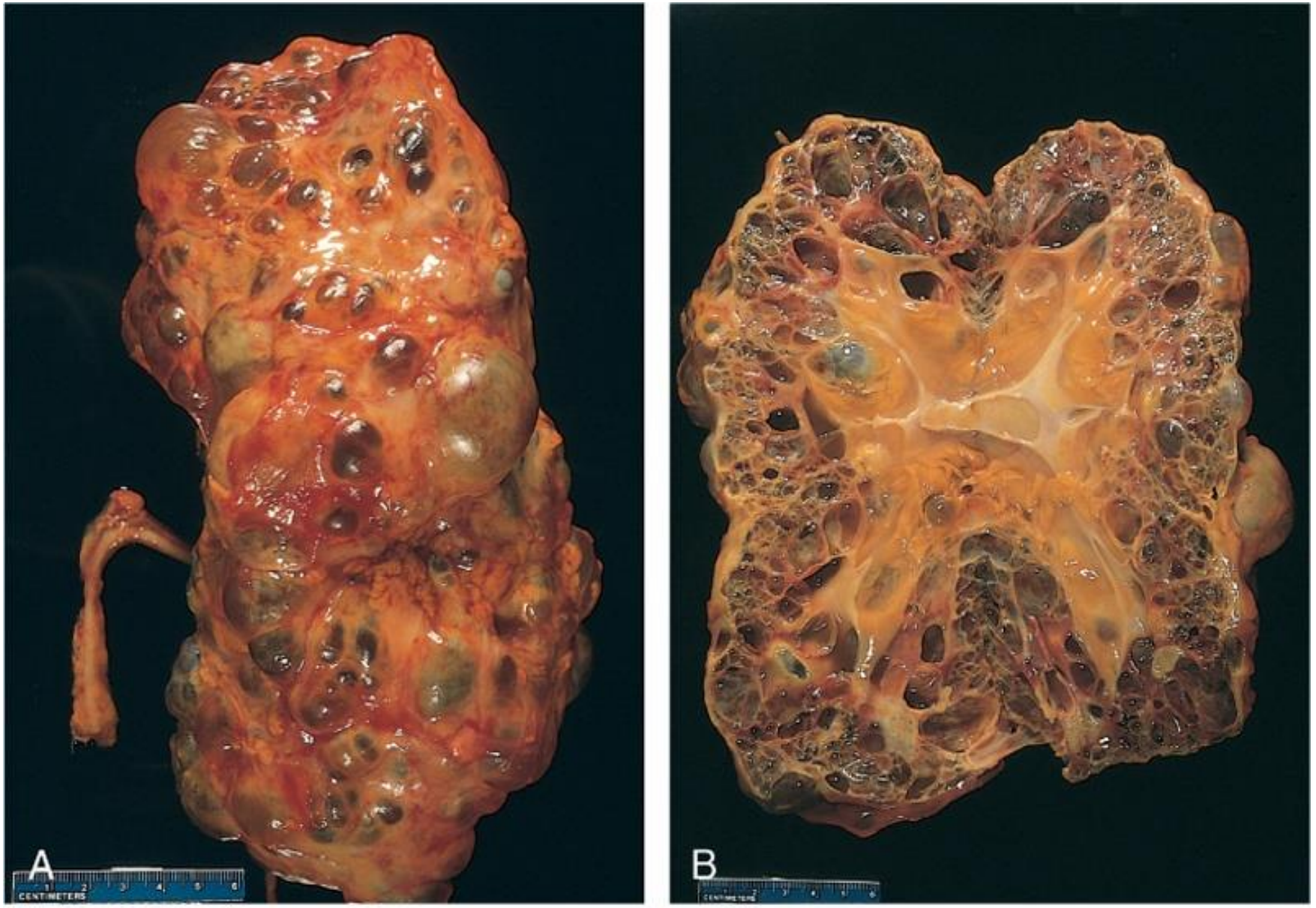
Cysts associated with chronic dialysis.



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).*



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

- **Clinical presentation**
- **small cysts start to develop in adolescence.**
- ***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.

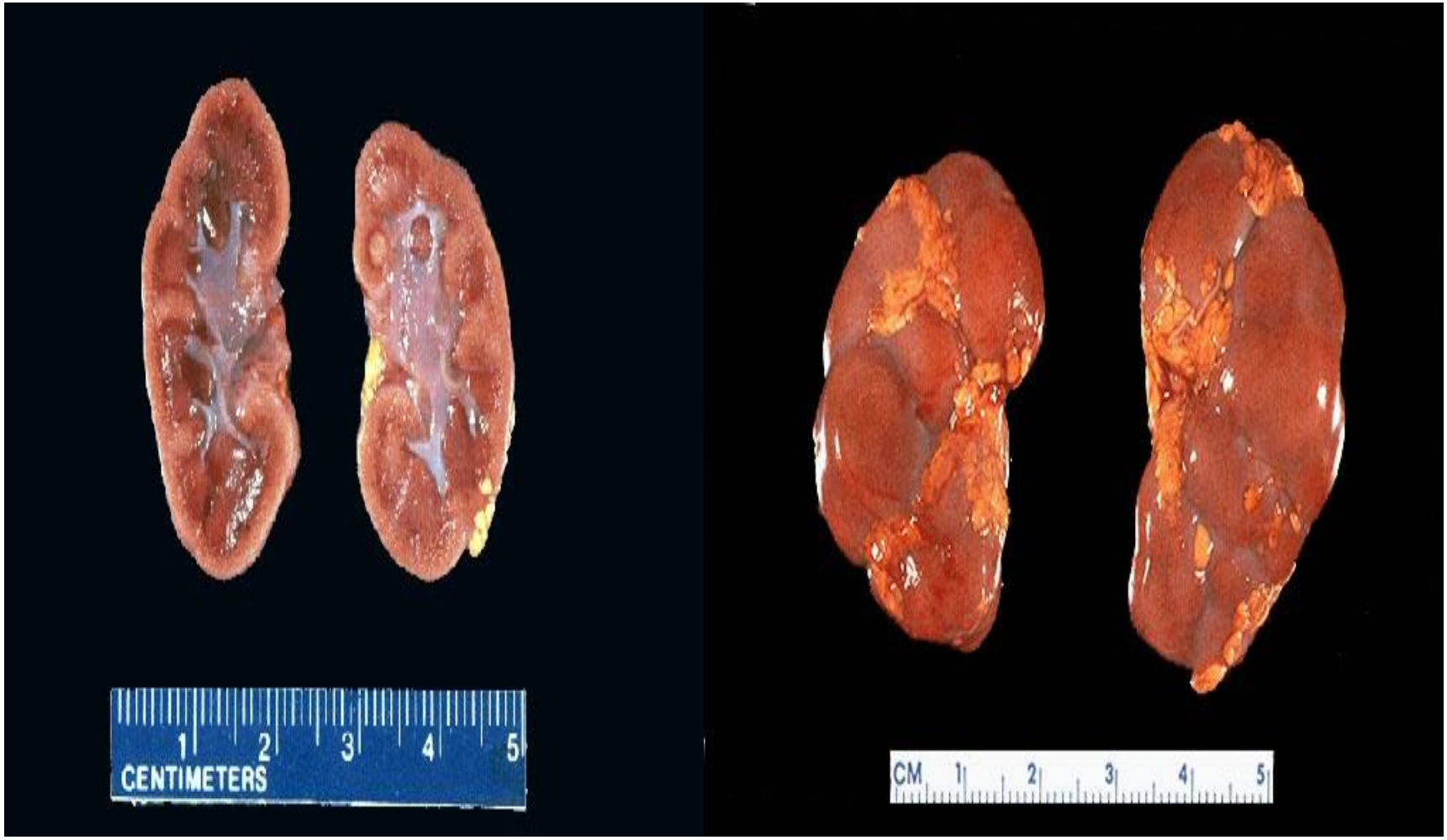
3- vascular aneurysms of circle of Willis (10% -30%)→ (subarachnoid hemorrhage).

4- renal failure at age 50

4-Autosomal Recessive (Childhood) Polycystic Kidney Disease

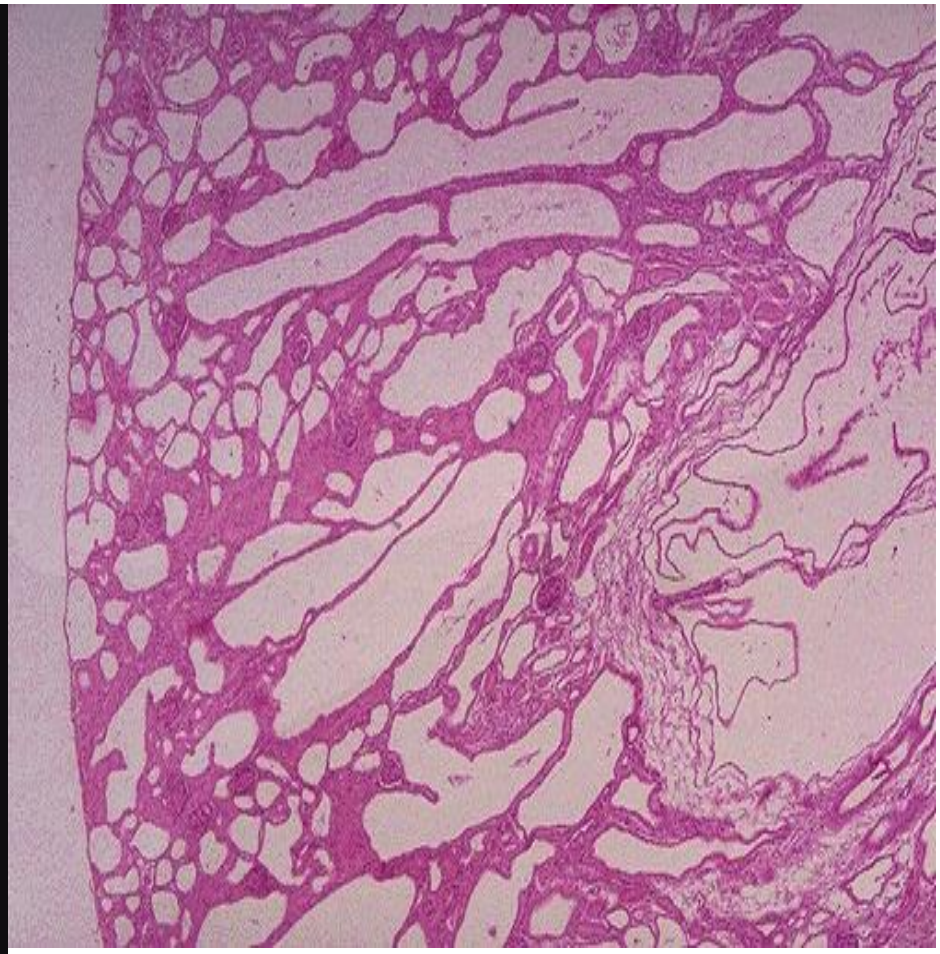
- **autosomal recessive**
- **1:20,000 live births.**
- **Types: perinatal, neonatal, infantile, and juvenile.**
- **Associated with liver cysts**
- **Mutations in *PKHD1* gene coding for *fibrocystin*.**
- **Fibrocystin may be involved in the function of cilia in tubular epithelial cells .**

Normal term infant kidneys



Cysts are fairly small but uniformly distributed throughout the parenchyma so that the disease is usually symmetrical in appearance

with both kidneys markedly enlarged.



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 :**

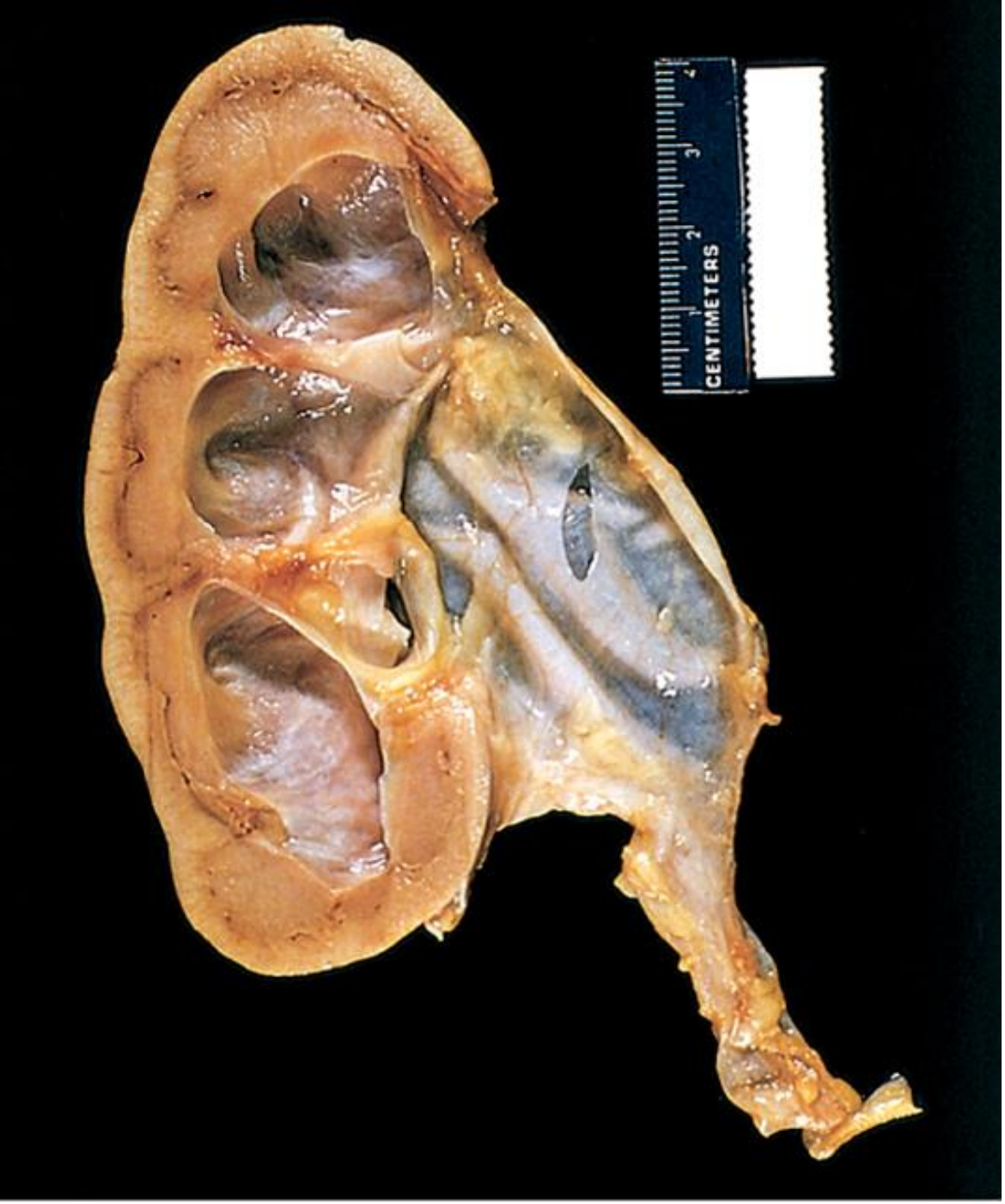
- ***1-Congenital:***

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: causes***

- **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



Hydronephrosis of the kidney, with marked dilation of the pelvis and calyces and thinning of renal parenchyma.

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DISEASES AFFECTING TUBULES AND INTERSTITIUM

Tubulointerstitial Nephritis

- Causes :
- 1- bacterial infection.
- 2- drugs.
- 3- metabolic disorders
- 4- physical injury (irradiation).
- 5- immune reactions.
- **divided into :**
- **1-acute**
- **2-chronic**

Urinary tract infections

1- lower UT (cystitis, prostatitis, urethritis).

2- upper UT(pyelonephritis).

Infectious : Acute Pyelonephritis

- inflammation of kidney and renal pelvis, u**sually due to bacterial infection**.
- **Most commonly:**

1- Escherichia coli

Others:

2- *Proteus.*

3- Klebsiella.

4- Enterobacter.

5- *Pseudomonas.*

6- Staphylococci and *Streptococcus faecalis* (uncommon).

Drug-Induced Interstitial Nephritis

- **Two forms:**
- **1-Acute Drug-Induced Interstitial Nephritis**
- **2- chronic (Analgesic) Nephropathy**

- **Acute TIN**
- **Most common: synthetic penicillins (methicillin, ampicillin)**
- **Others: synthetic antibiotics; diuretics; NSAIDs; other drugs**

- **Pathogenesis**
- **immune mechanism.**
- **? type I hypersensitivity.**
- **? T cell-mediated (type IV) hypersensitivity reaction.**

- **Morphology**

- **interstitium : infiltration by lymphocytes and macrophages, eosinophils and neutrophils**

- **glomeruli are normal**

Clinical course

- **2-40 days after exposure to drug.**
- ***fever*, *eosinophilia* & *rash* (25%)**
- ***renal abnormalities:* hematuria, minimal or no proteinuria, and leukocyturia**
- **withdrawal of the offending drug is followed by recovery**

Analgesic Nephropathy: chronic drug-induced

- Consumption of large quantities of analgesics over long periods may cause **chronic interstitial nephritis** *often with* ***renal papillary necrosis***.
- **Aspirin and acetaminophen are common**
- **Pathogenesis** not entirely clear.
  - ***covalent binding*** **and** ***oxidative damage***
  - **inhibition of prostaglandin synthesis**

Clinical Course

- Chronic renal failure, hypertension, and anemia (results from damage to red cells)

- **A complication of analgesic abuse is: increased incidence of *transitional-cell carcinoma* of the renal pelvis**

Acute Tubular Necrosis (ATN)

- characterized morphologically by damaged tubular epithelial cells and clinically by acute suppression of renal function.
- ***It is the most common cause of acute renal failure.***
- **ATN is a reversible condition if treated properly and quickly.**

- **Clinical manifestations: electrolyte abnormalities, acidosis, uremia, signs of fluid overload, often oliguria.**
- Proximal tubular epithelial cells are particularly sensitive to hypoxemia and toxins

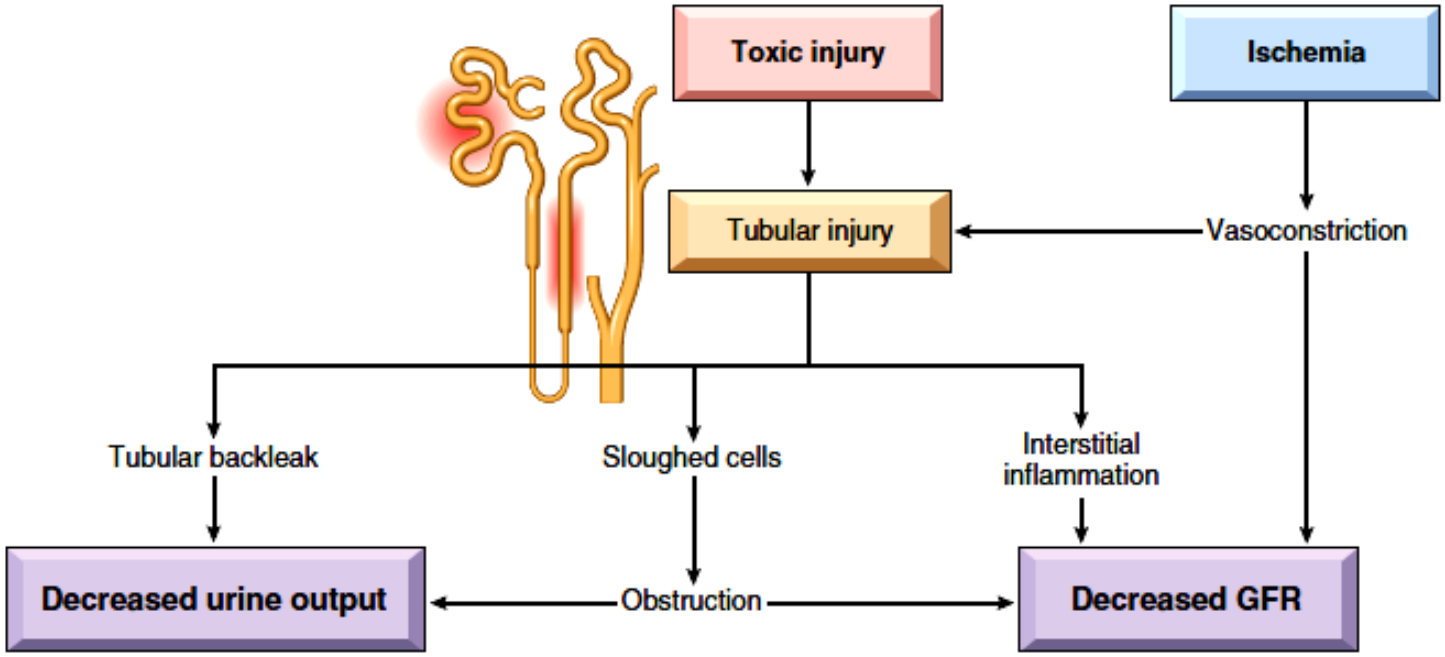


Fig. 14.16 Postulated sequence in ischemic or toxic tubular injury.

- ***Types:***

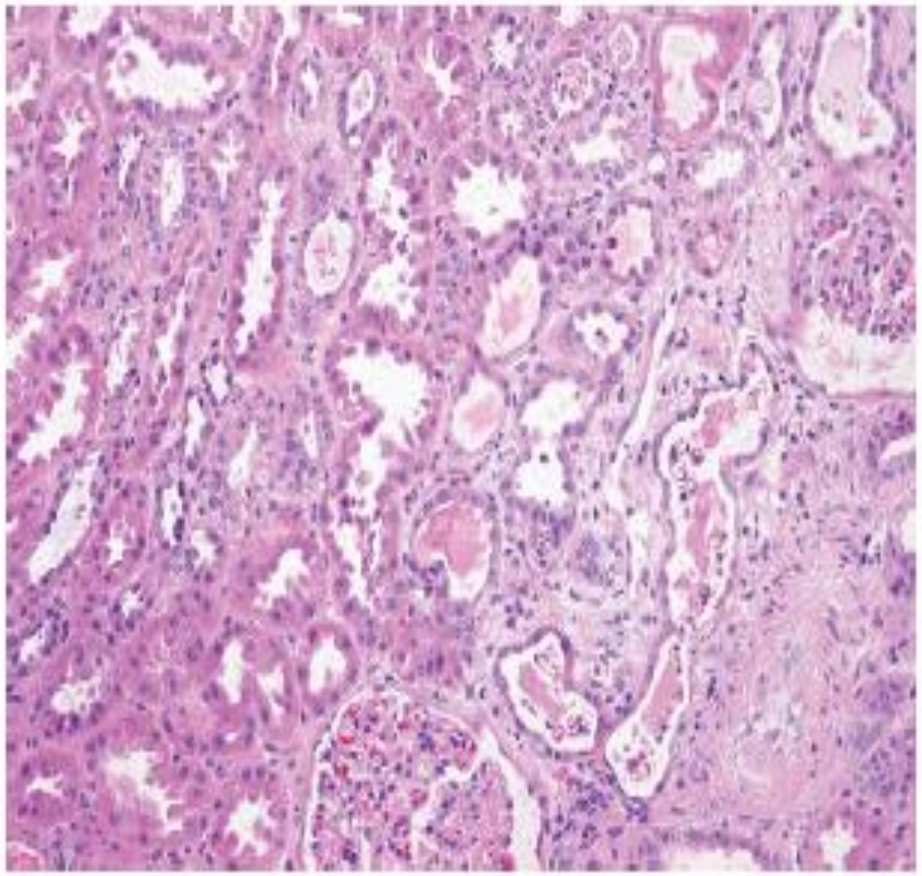
- ***1- ischemic ATN :***

- most common

- associated with shock (e.g. **severe trauma; acute pancreatitis; septicemia; mismatched blood transfusions,hemolytic crises, myoglobinuria, etc)**

- ***2- nephrotoxic ATN***
- **poisons including heavy metals (e.g., mercury)**
- **organic solvents (e.g., carbon tetrachloride)**
- **drugs such as gentamicin and other antibiotics, and radiographic contrast agents.**

Acute tubular epithelial cell injury with blebbing at the luminal pole, detachment of tubular cells from their underlying basement membranes,
and granular casts



ATI

- repair and tubular regeneration lead to gradual clinical improvement
- With supportive care, patients who survive have a good chance of recovering renal function
- those with preexisting chronic kidney disease, complete recovery is less frequent