

RENAL MALFORMATIONS, RENAL VASCULAR DISEASES, POLYCYSTIC KIDNEY

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DEPT OF PATHOLOGY**

CONGENITAL MALFORMATIONS

- AGENESIS
- HYPOPLASIA
- ECTOPIC
- HORSESHOE

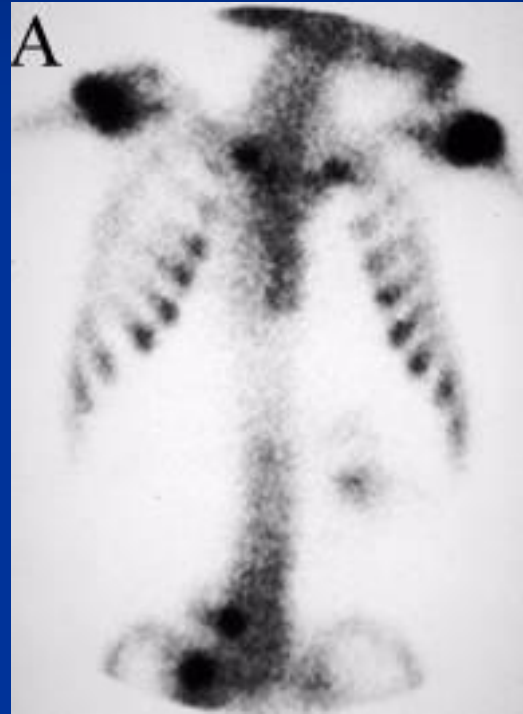
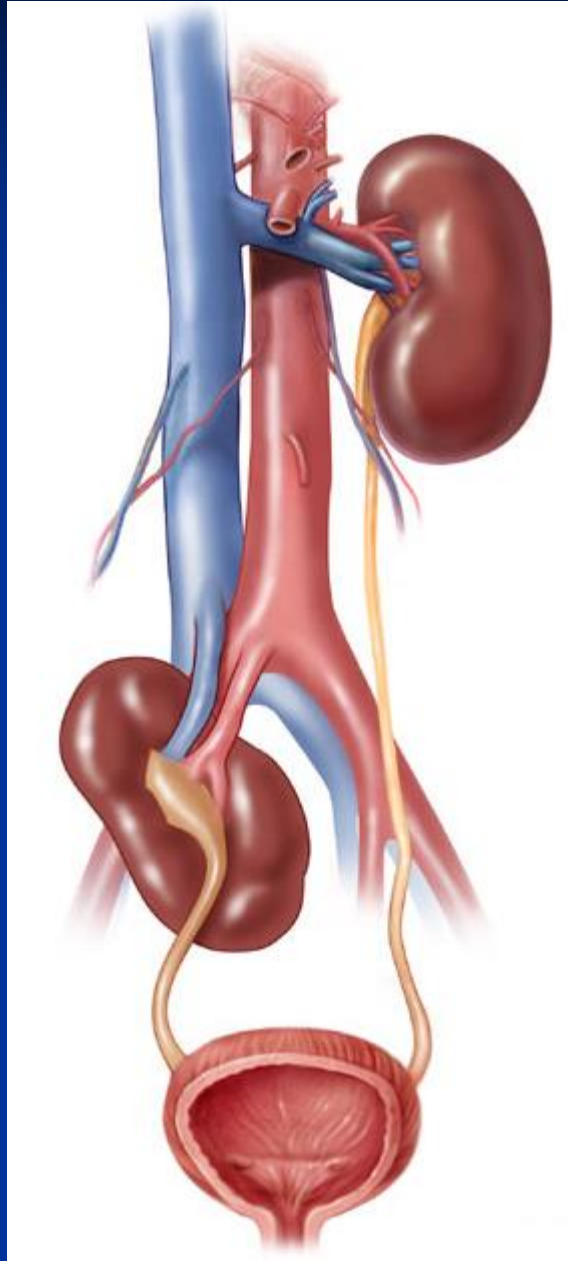
AGENESIS



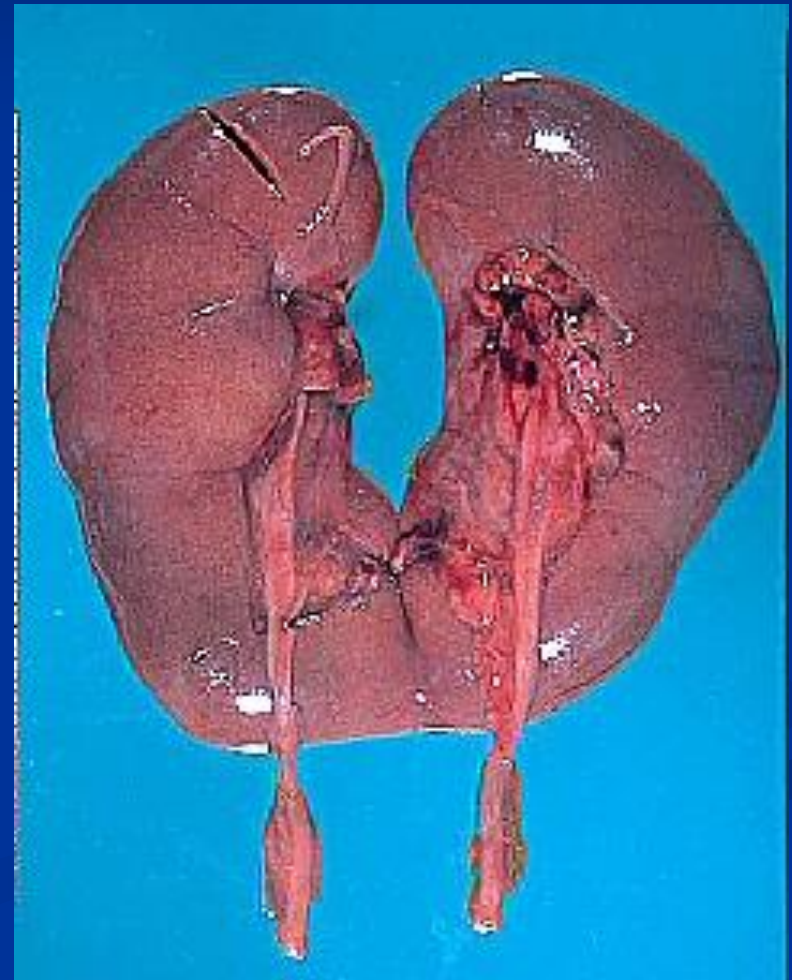
HYPOPLASIA



ECTOPIC (usually PELVIC)



HORSESHOE

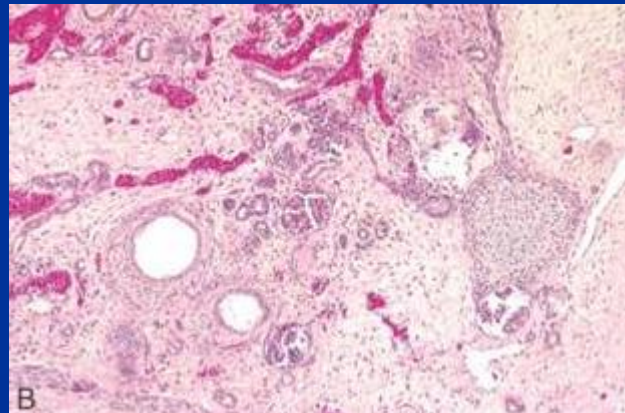
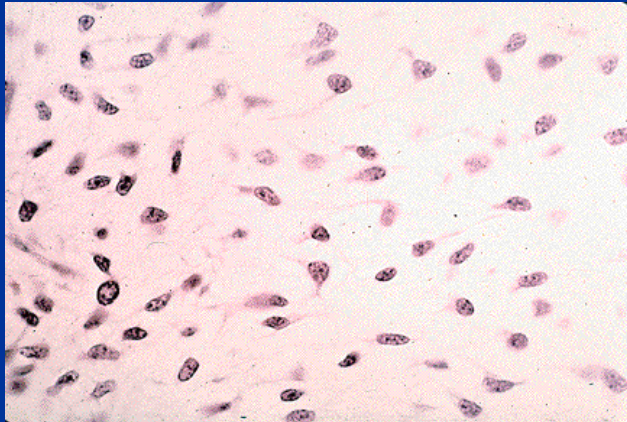


CYSTIC DISEASES

- CYSTIC RENAL “DYSPLASIA”
- Autosomal **D**OMINANT (**AD**-ULTS)
- Autosomal RECESSIVE (CHILDREN)
- MEDULLARY
 - Medullary Sponge Kidney (MSK)
 - Nephronopththisis-Medullary
- ACQUIRED
- SIMPLE

CYSTIC RENAL “DYSPLASIA”

- ENLARGED
- UNILATERAL or BILATERAL
- CYSTIC
- Have “MESENCHYME”
- NEWBORNS
- VIRAL, GENETIC (rare)



“SIMPLE” CYSTS

- Cortical
- Also called “retention” cysts
- Also “acquired”
- Incidental, asymptomatic
- **VERY** very very common



- Here is a normal adult kidney.
- The capsule has been removed and a pattern of fetal lobulations still persists, as it sometimes does.
- The hilum at the mid left contains some adipose tissue.
- At the lower right is a smooth-surfaced, small, clear fluid-filled simple renal cyst.
- Such cysts occur either singly or scattered around the renal parenchyma and are not uncommon in adults.

- In cross section, this normal adult kidney demonstrates the lighter outer cortex and darker medulla with central pelvis.





- Here is a much larger simple renal cyst of the upper pole.
- Other smaller cysts are also scattered around the kidney.
- The ureter exits south on the left. Such a large renal cyst would be seen on a radiographic imaging procedure, but could probably be distinguished from a neoplasm by its fluid density.

Renal Vascular Diseases

Renal vascular disease affects the blood flow into and out of the kidneys. It may cause kidney damage, kidney failure, and high blood pressure.

WHO AT RISK?

- Older age >50 y/o
- Female
- Atherosclerosis
- High blood pressure
- Smoking
- High cholesterol
- Diabetes
- Family hx of peripheral arterial disease
- Family hx of renal artery disease

WHAT CAUSE IT?

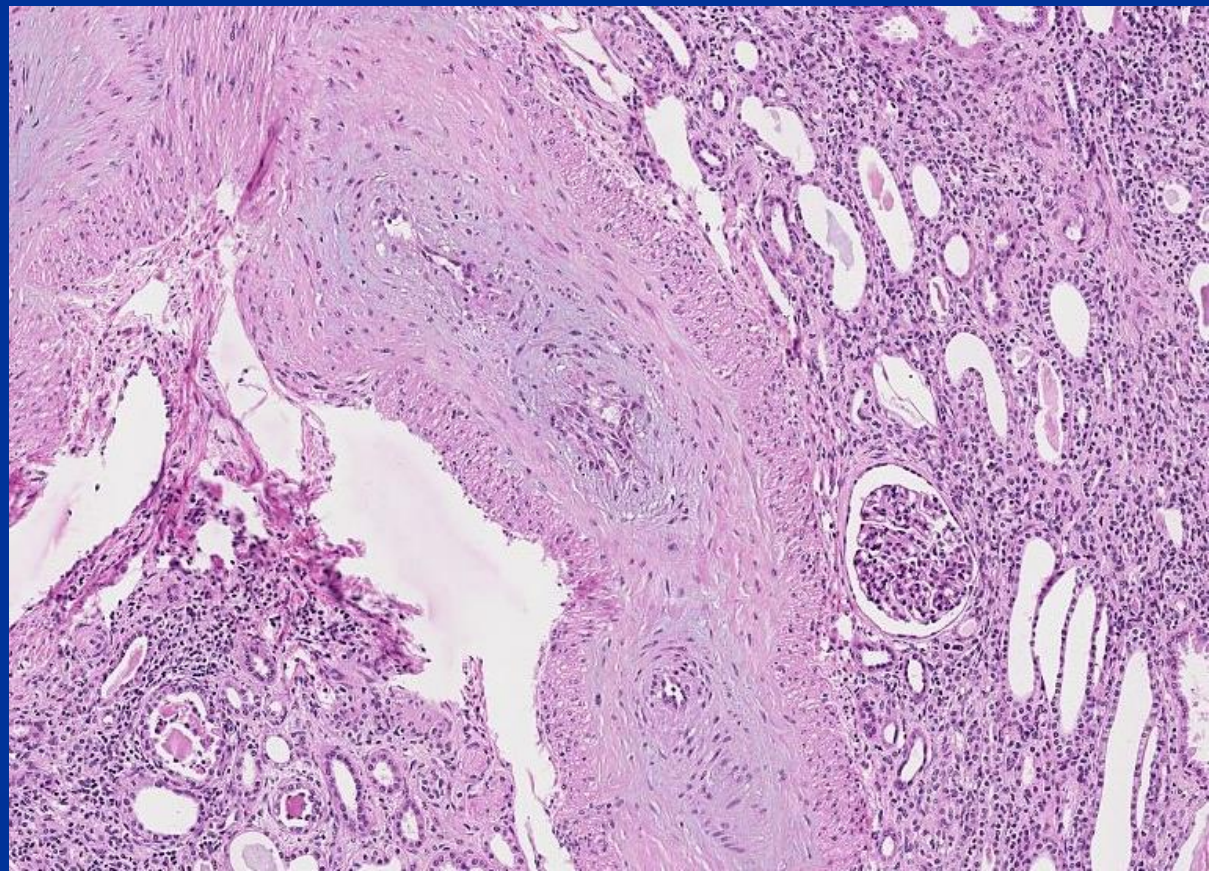
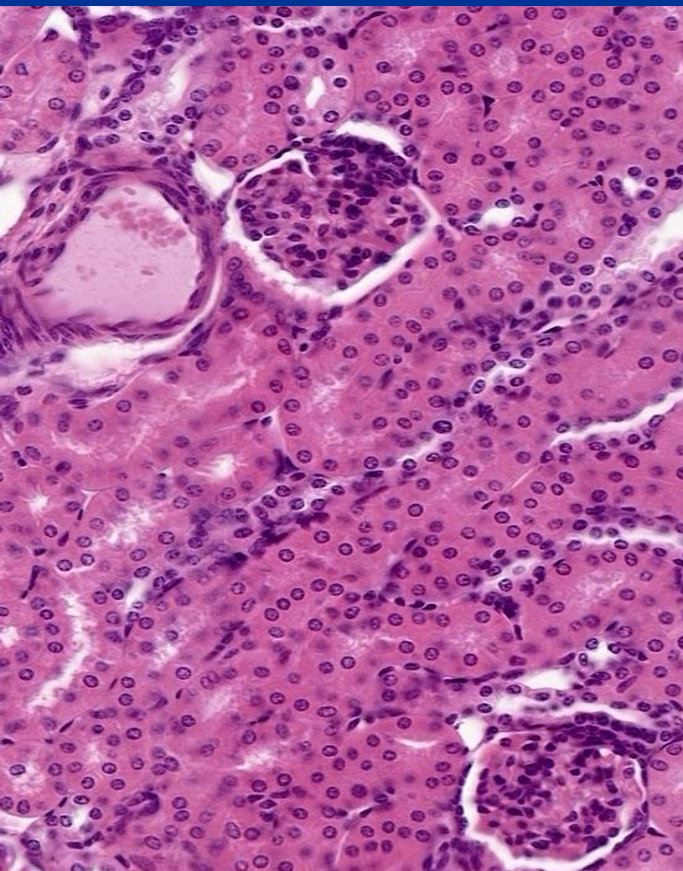
- Atherosclerosis
- Injury
- Infection
- Inflammatory or other underlying disease
- Surgery
- Tumour
- Aneurysm
- Pregnancy
- Birth defect
- Medication

VASCULAR DISEASES

- BENIGN NEPHROSCLEROSIS
- MALIGNANT NEPHROSCLEROSIS (i.e., malignant hypertension)
- RENAL ARTERY STENOSIS
- THROMBOTIC MICROANGIOPATHIES
 - Hemolytic-Uremic Syndromes, Child, Adult, TTP
- THROMBI, EMBOLI, INFARCTS
 - SICKLE CELL
 - DIFFUSE CORTICAL NECROSIS

BENIGN NEPHROSCLEROSIS

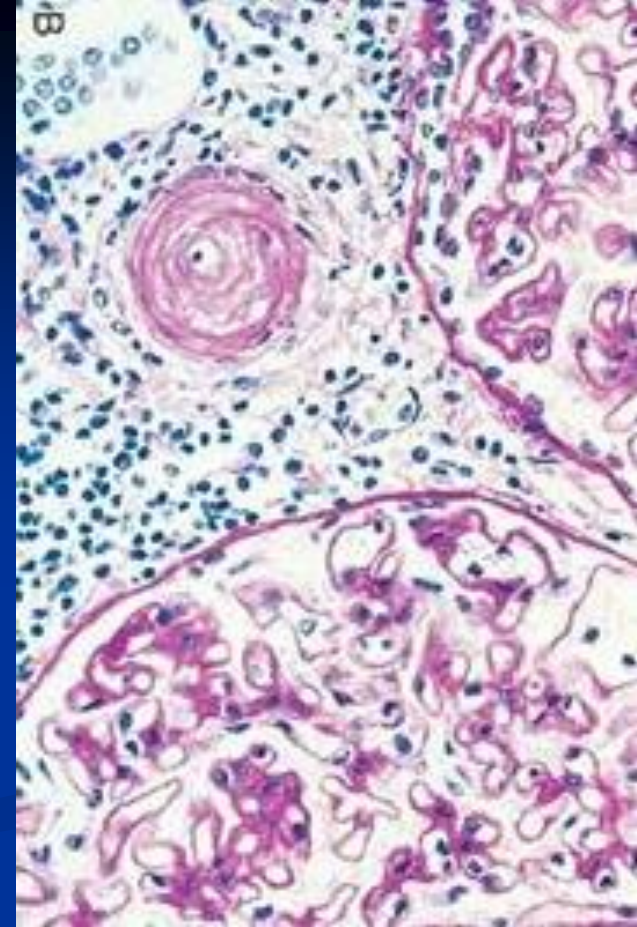
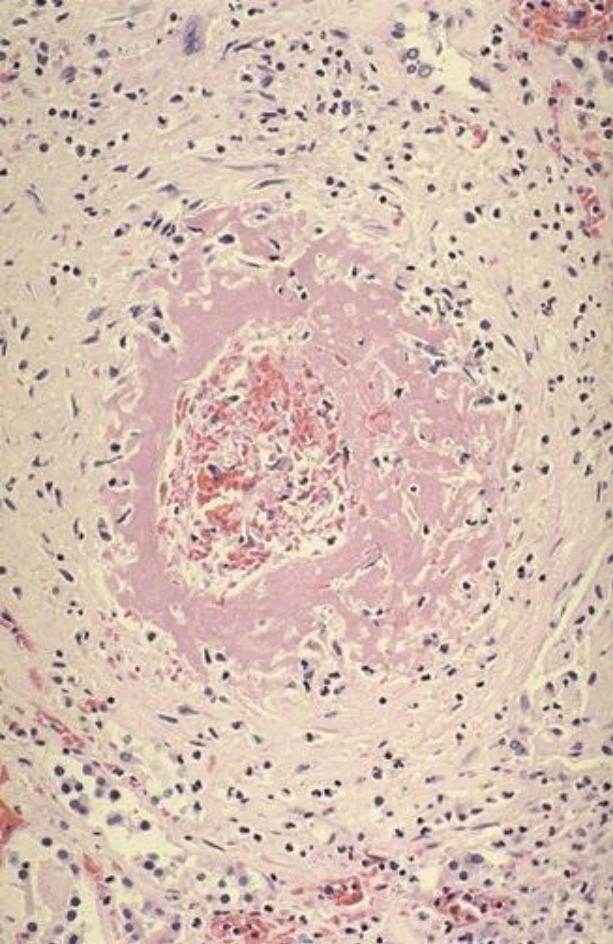
- Sclerosis, i.e., “hyalinization” of arterioles and small arteries, i.e., **arterio-, arteriolo-**
- Is this part of “routine” atherosclerosis????
- **VERY VERY VERY** common



MALIGNANT NEPHROSCLEROSIS

(i.e., malignant hypertension)

- NOT a part of “routine” atherosclerosis
- By definition, associated with rapidly progressive hypertension (1-2% of HTN)
- VASCULAR DAMAGE
- FIBRINOID NECROSIS
- “ONION SKINNING”
- SIGNIFICANT LUMINAL NARROWING



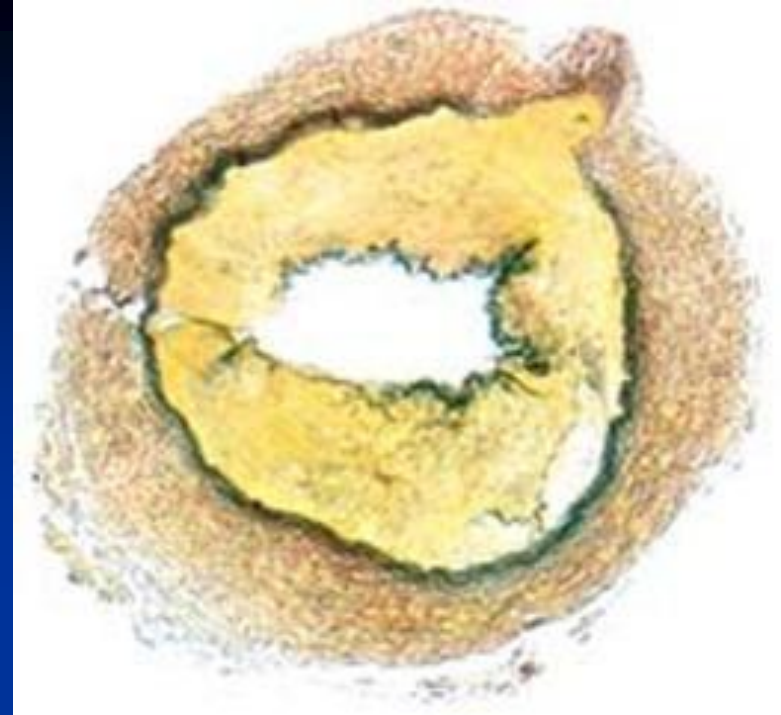
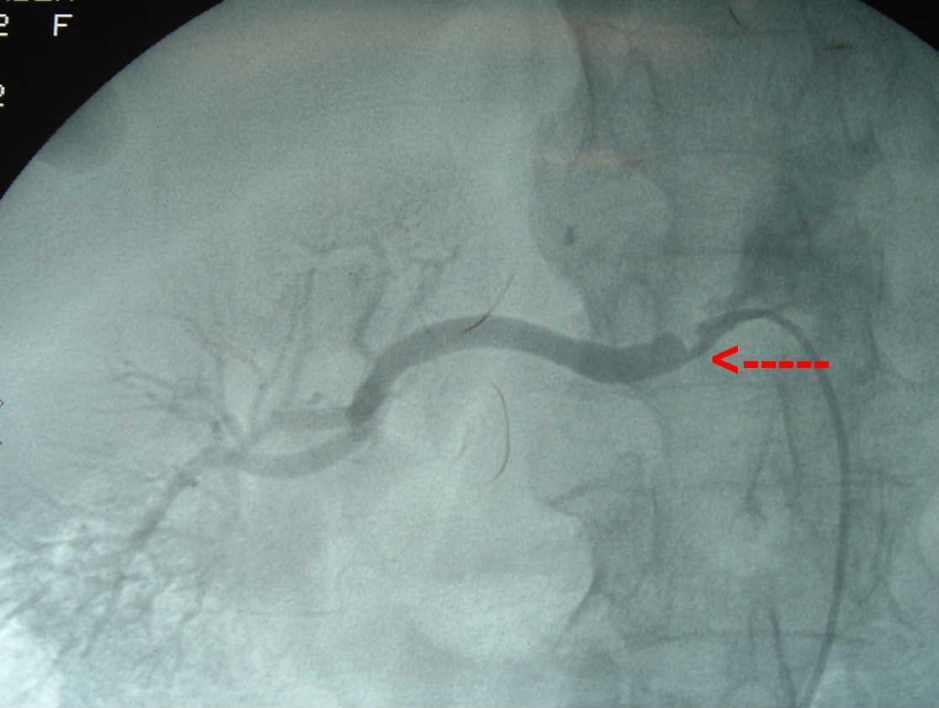
What is “onion-skinning”?

What is an onion?

What is “fibrinoid” necrosis?

Renal Artery Stenosis

- Rare cause of HTN
- SMALL Kidney
- 1) Plaque type is usual cause, yes regular old atherosclerosis
- 2) Fibromuscular “dysplasia” type:
 - INTIMAL HYPERPLASIA
 - MEDIAL HYPERPLASIA
 - ADVENTITIAL HYPERPLASIA
 - In younger women



PLAQUE, i.e.,
ATHEROSCLEROSIS

**FIBROMUSCULAR
DYSPLASIA**

MICROANGIOPATHIES

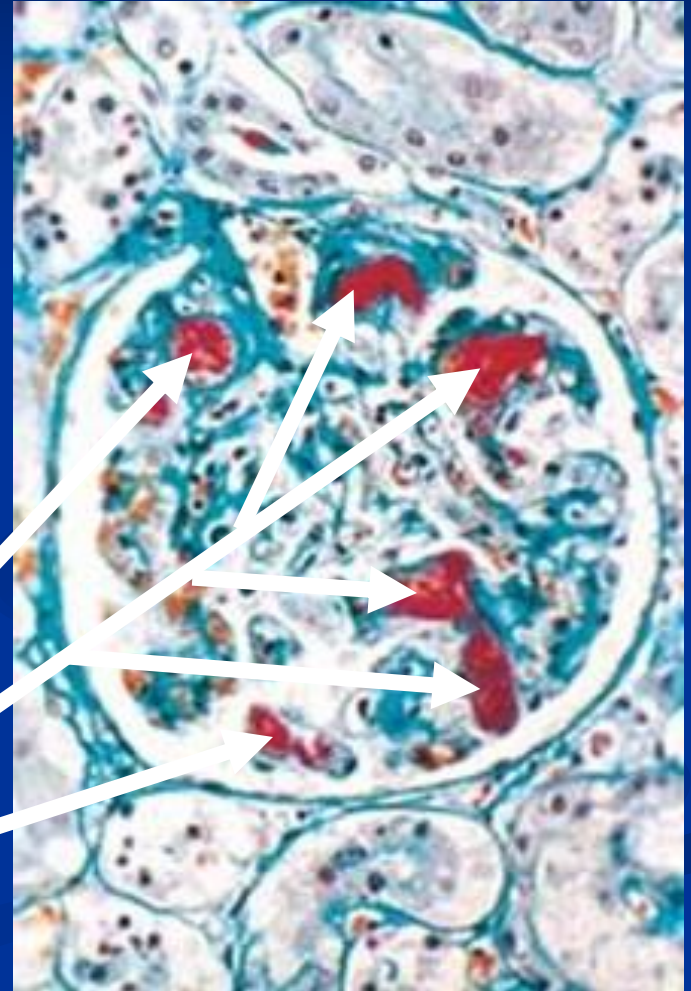
(thrombotic)

- Hemolytic-Uremic Syndrome
 - Familial
 - Childhood
 - Adult
- TTP (Thrombotic Thrombocytopenic Purpura), IDIOPATHIC

MICROANGIOPATHIES

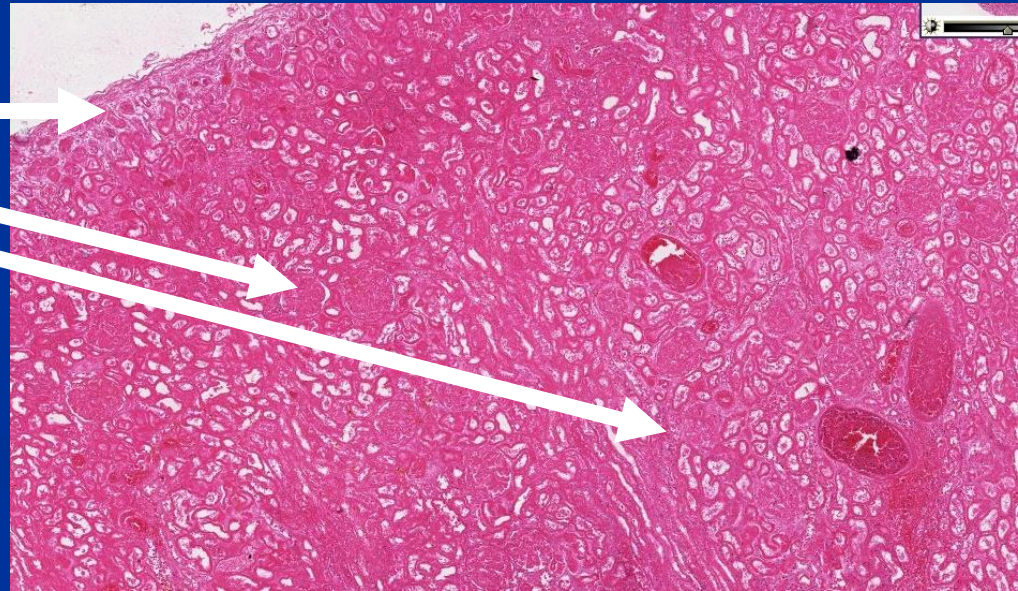
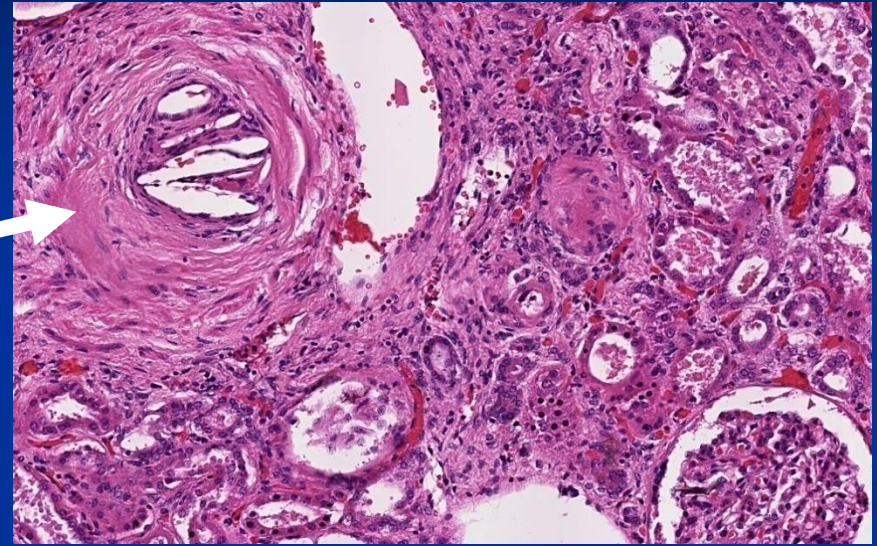
COMMON PROCESSES

- Hemolysis
- Thromboses in renal capillaries
- Thrombocytopenia (a “consumption” coagulopathy)
- FIBRIN PLUGS



OTHER VASCULAR

- Atherosclerosis
- Atheroemboli
- Sickle Cell
- Diffuse Cortical Necrosis

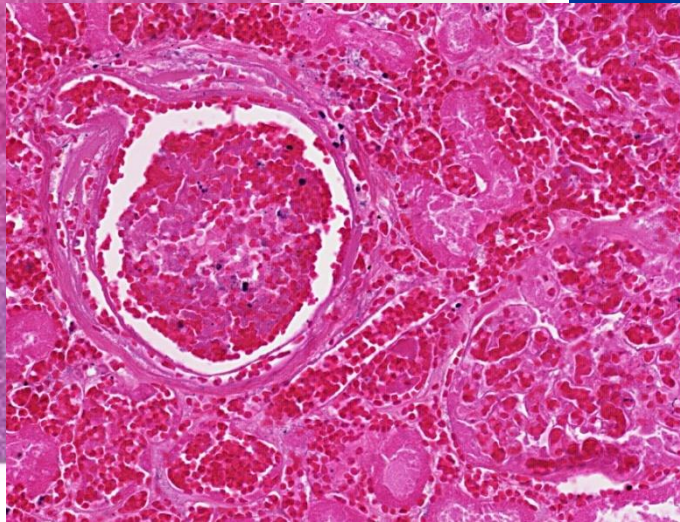
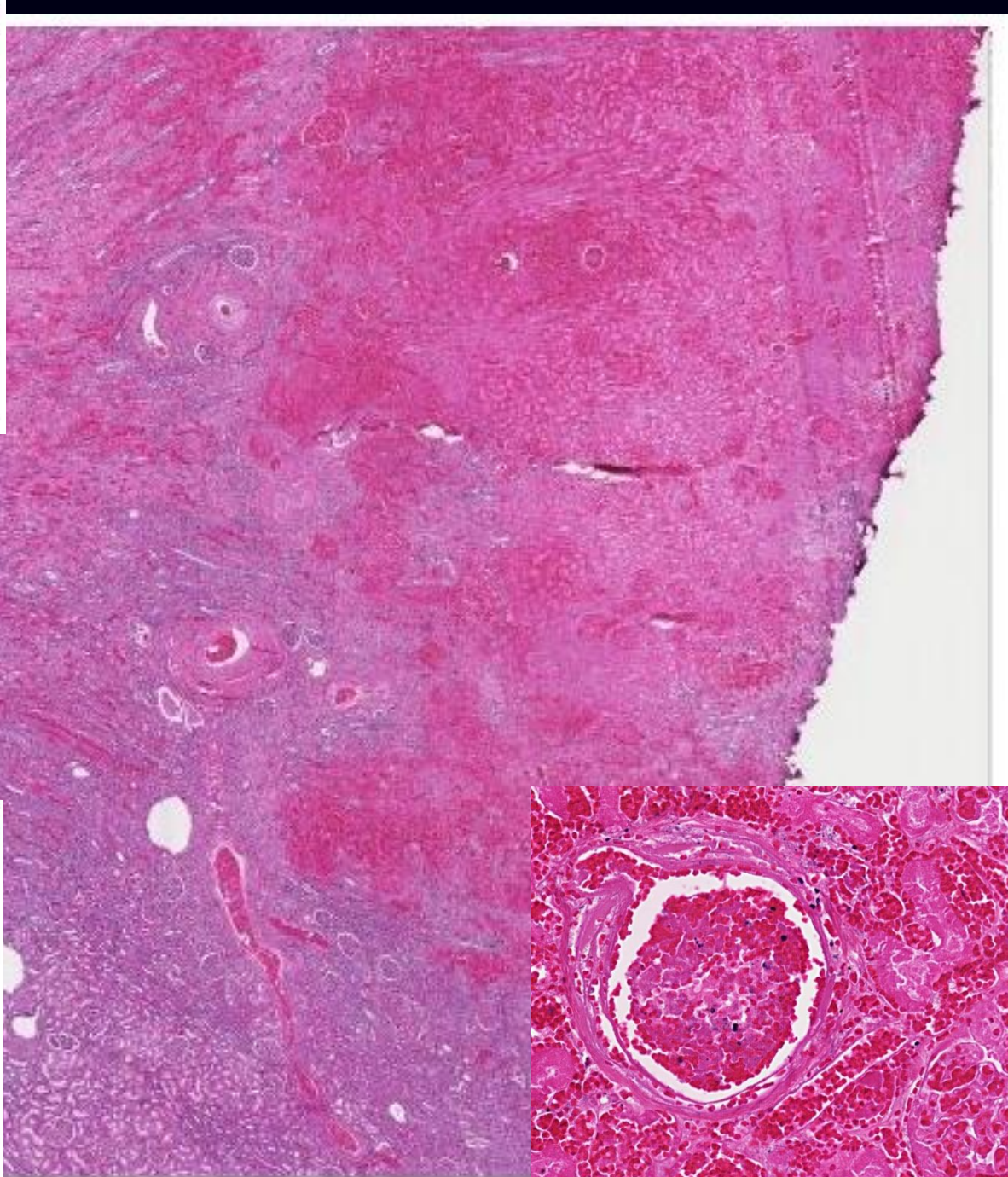
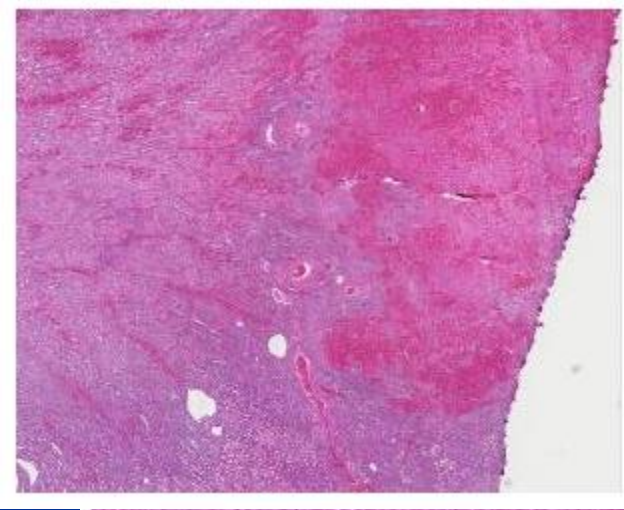


RENAL INFARCTS

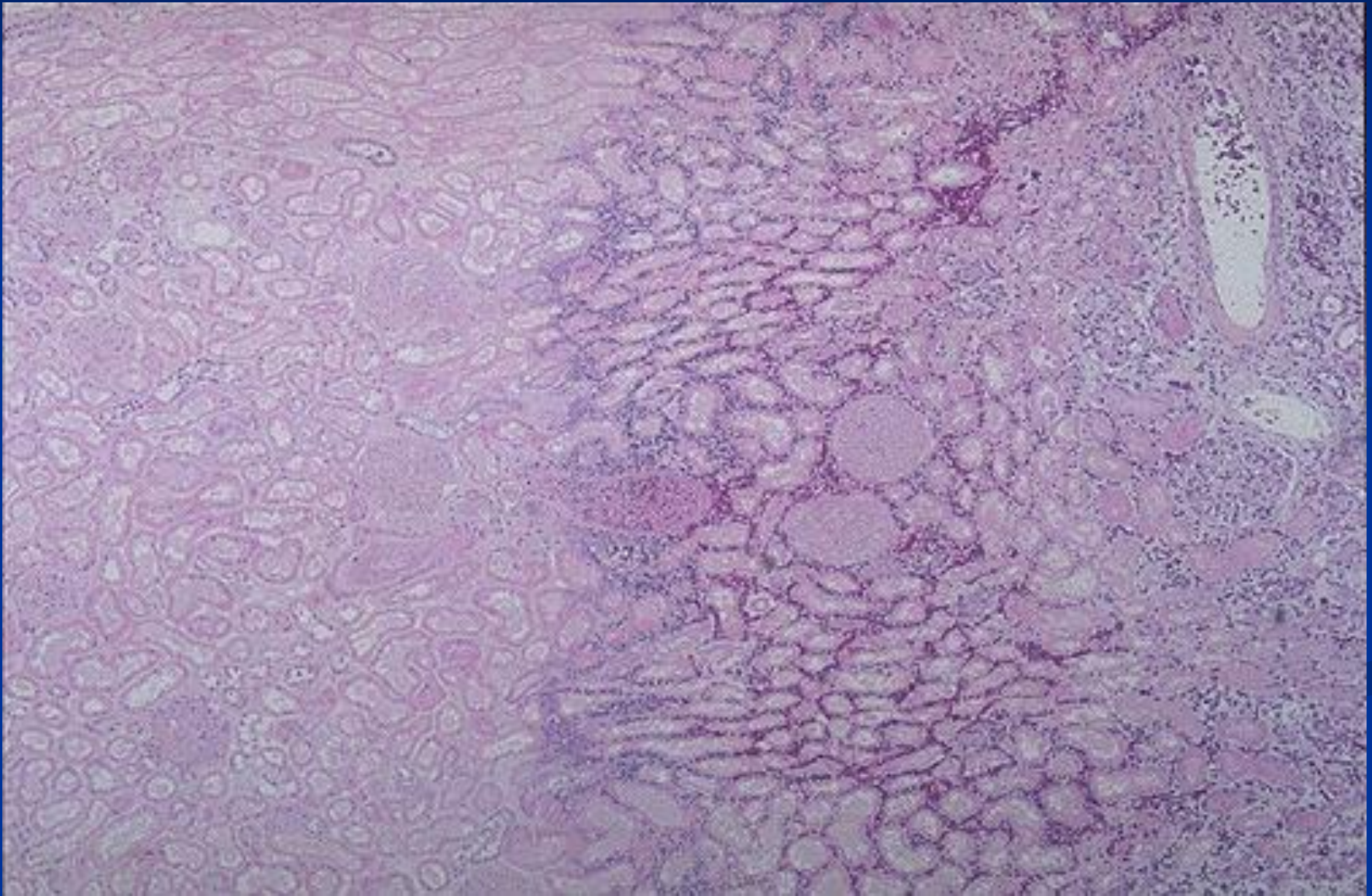
- WEDGE SHAPED
- WELL DELINEATED
- “WHITE” (anemic) INFARCT
OR “YELLOW”
- HEAL WITH A SCAR

- This is an acute renal infarction.
- Note the wedge shape of this zone of coagulative necrosis resulting from loss of blood supply with resultant tissue ischemia that produces the pale infarct.
- The small amount of blood supply from the capsule supplies the immediate subcortical zone.
- The remaining cortex is congested, as is the medulla.

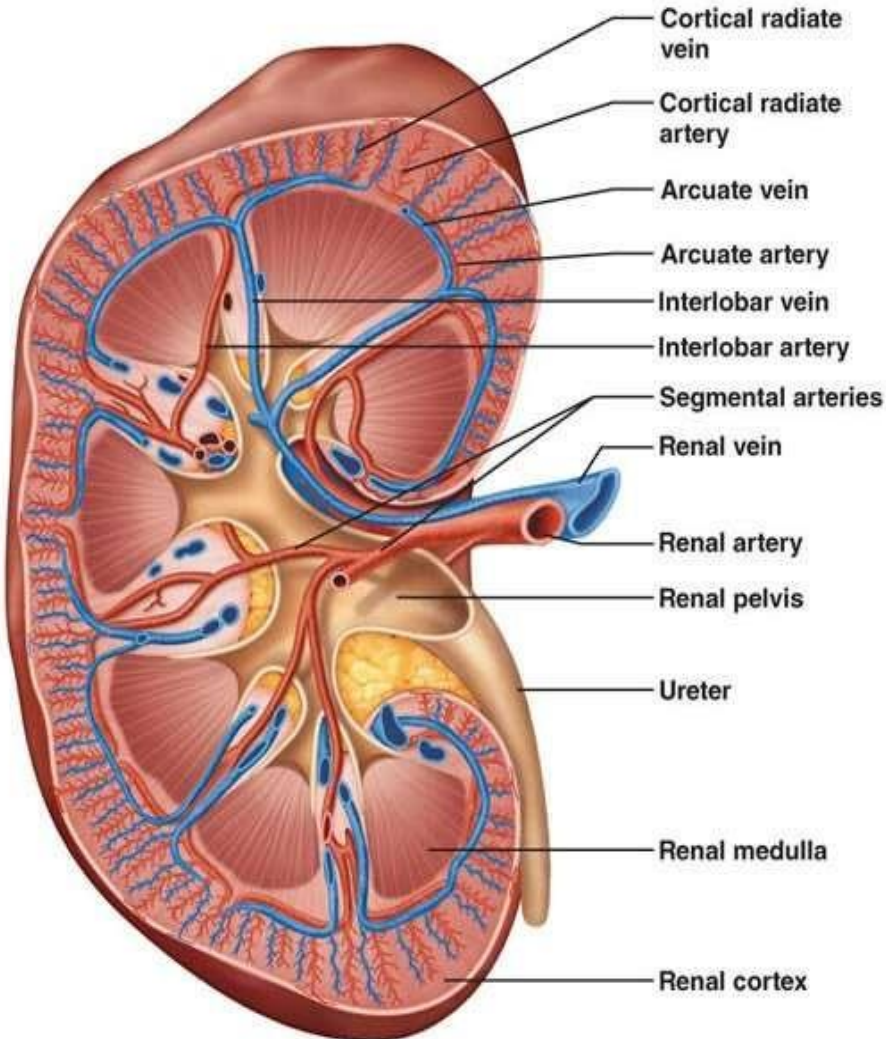




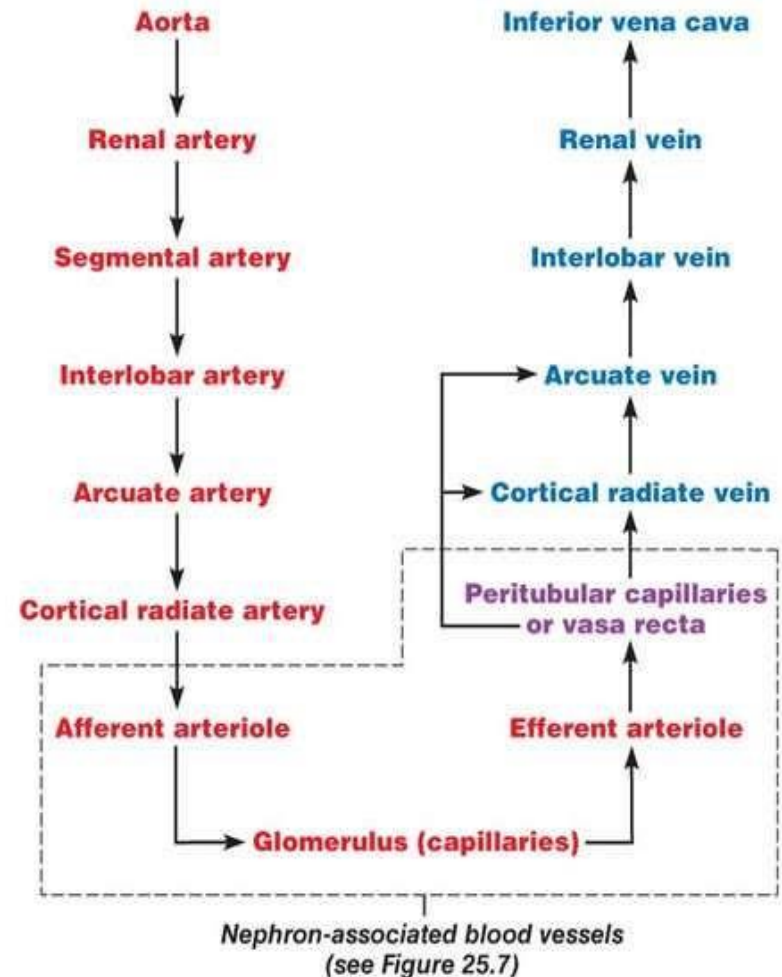
- This is the microscopic appearance of an acute renal infarct.
- At the far right is normal kidney, then to the left of that hyperemic kidney that is dying, then to the left of that pale pink infarcted kidney in which both tubules and glomeruli are dead.



BLOOD SUPPLY OF KIDNEY

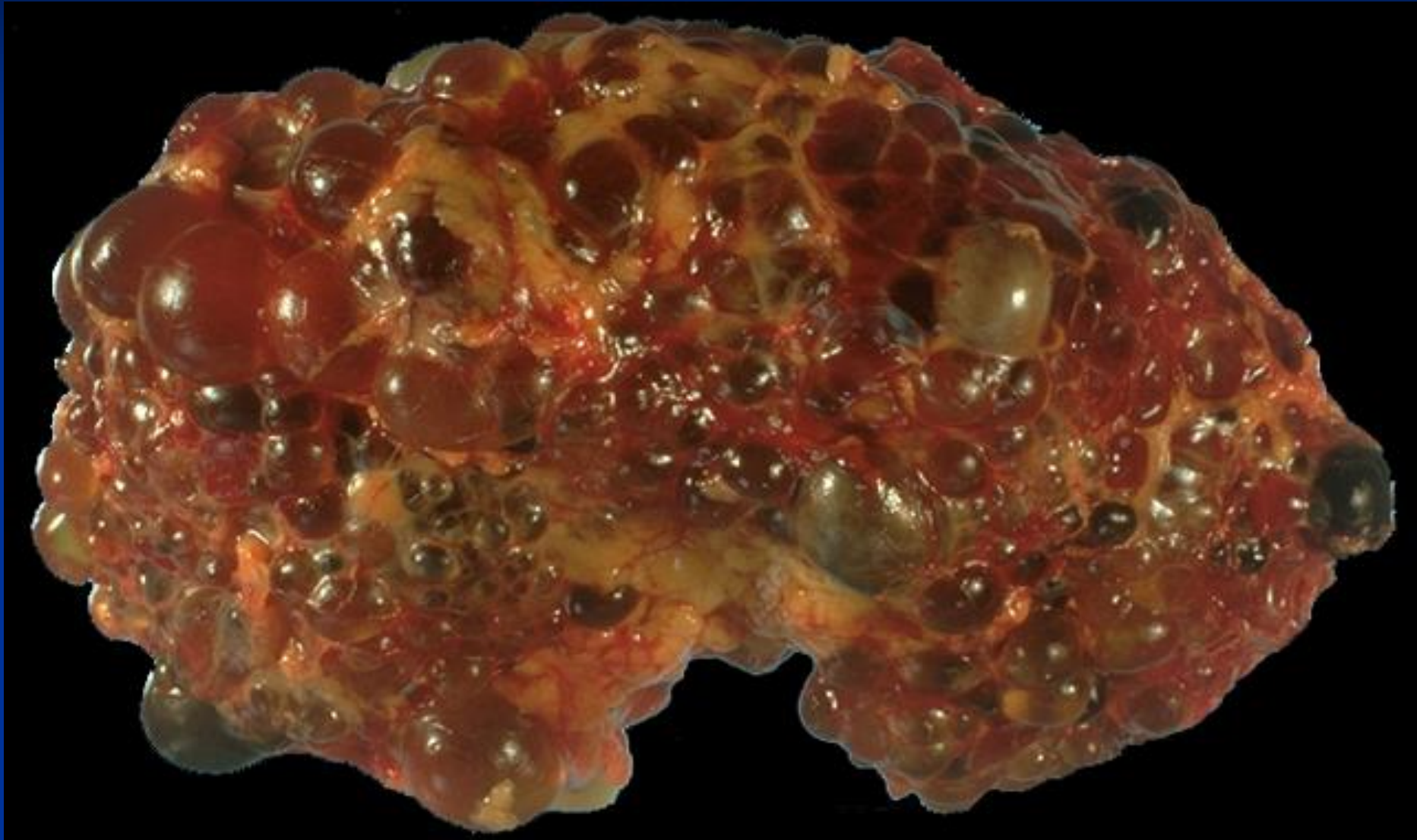


(a) Frontal section illustrating major blood vessels



(b) Path of blood flow through renal blood vessels

POLYCYSTIC KIDNEY DISEASE



DEFINITION

- ADPKD is a multisystem disorder characterized by multiple, **bilateral renal cysts** associated with **cysts in other organs**, such as liver, pancreas, and arachnoid membranes.
- It is a genetic disorder mediated primarily by mutations in **two different genes** and is expressed in an **autosomal dominant** pattern, with variable expression.



Markedly enlarged polycystic kidneys from a patient with ADPKD in comparison to a normal kidney in the middle.

Genes in PKD

Gene

Protein

ADPKD1

Polycystin 1

ADPKD2

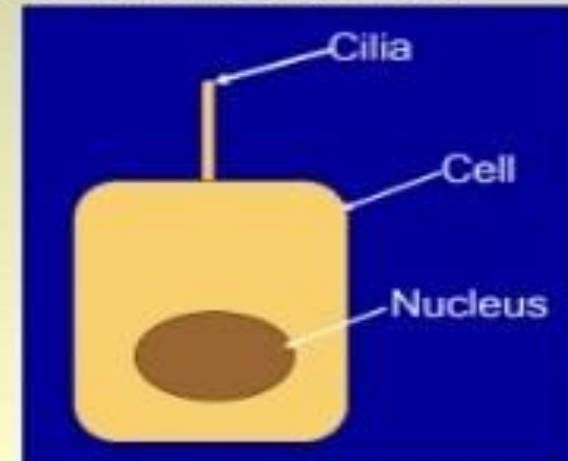
Polycystin 2

ARPKD

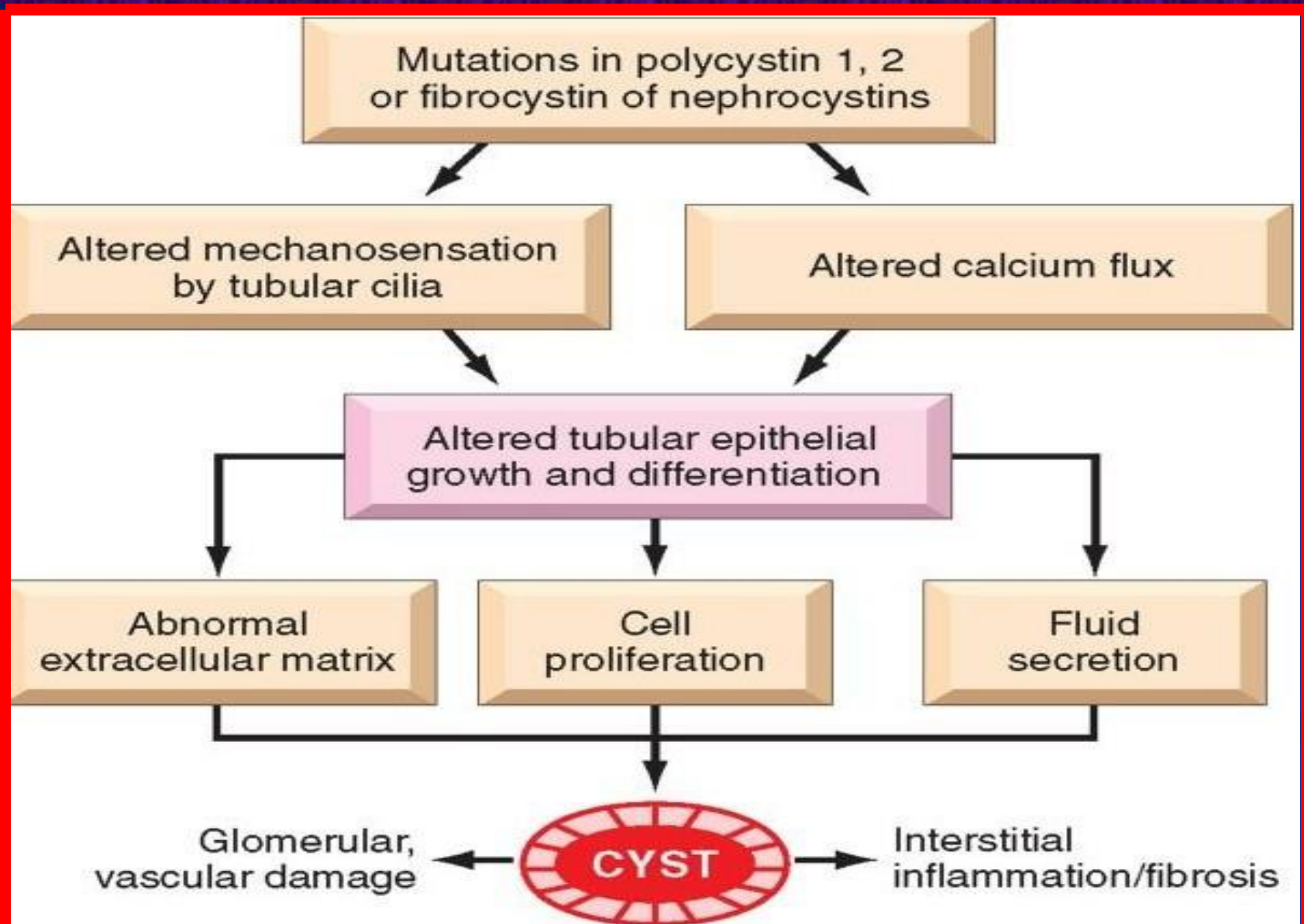
Fibrocystin/polyductin

Etiology and Pathogenesis

- The polycystic kidney disease (PKD) proteins now known as **polycystin 1 (PC1)** and **polycystin 2 (PC2)** play a critical role in the normal function of the **primary cilium** that is essential to maintaining the differentiated phenotype of tubular epithelium.
- **Disordered function of polycystins is the basis for cyst formation** in PKD by permitting a less differentiated tubular epithelial phenotype.



Possible mechanisms of cyst formation in polycystic kidney disease



Gross Morphology

The kidneys are usually bilaterally

ENLARGED and

may achieve enormous sizes; weights

as high as **4 kg** for each kidney
have been reported.

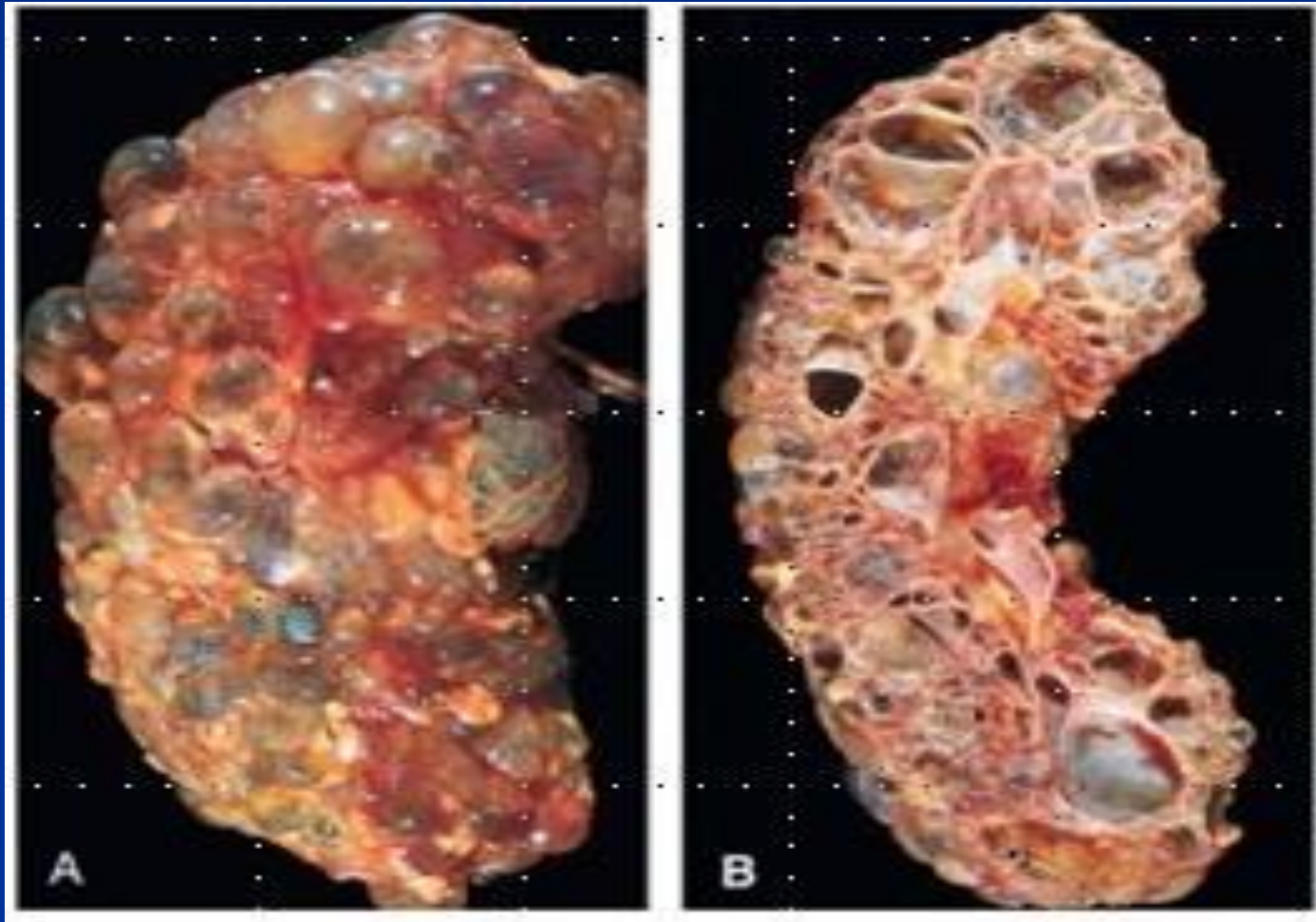
Morphology

- The external surface appears to be composed solely of a mass of cysts, up to 3 to 4 cm in diameter, with no intervening parenchyma. **However, microscopic examination reveals functioning nephrons dispersed between the cysts.**

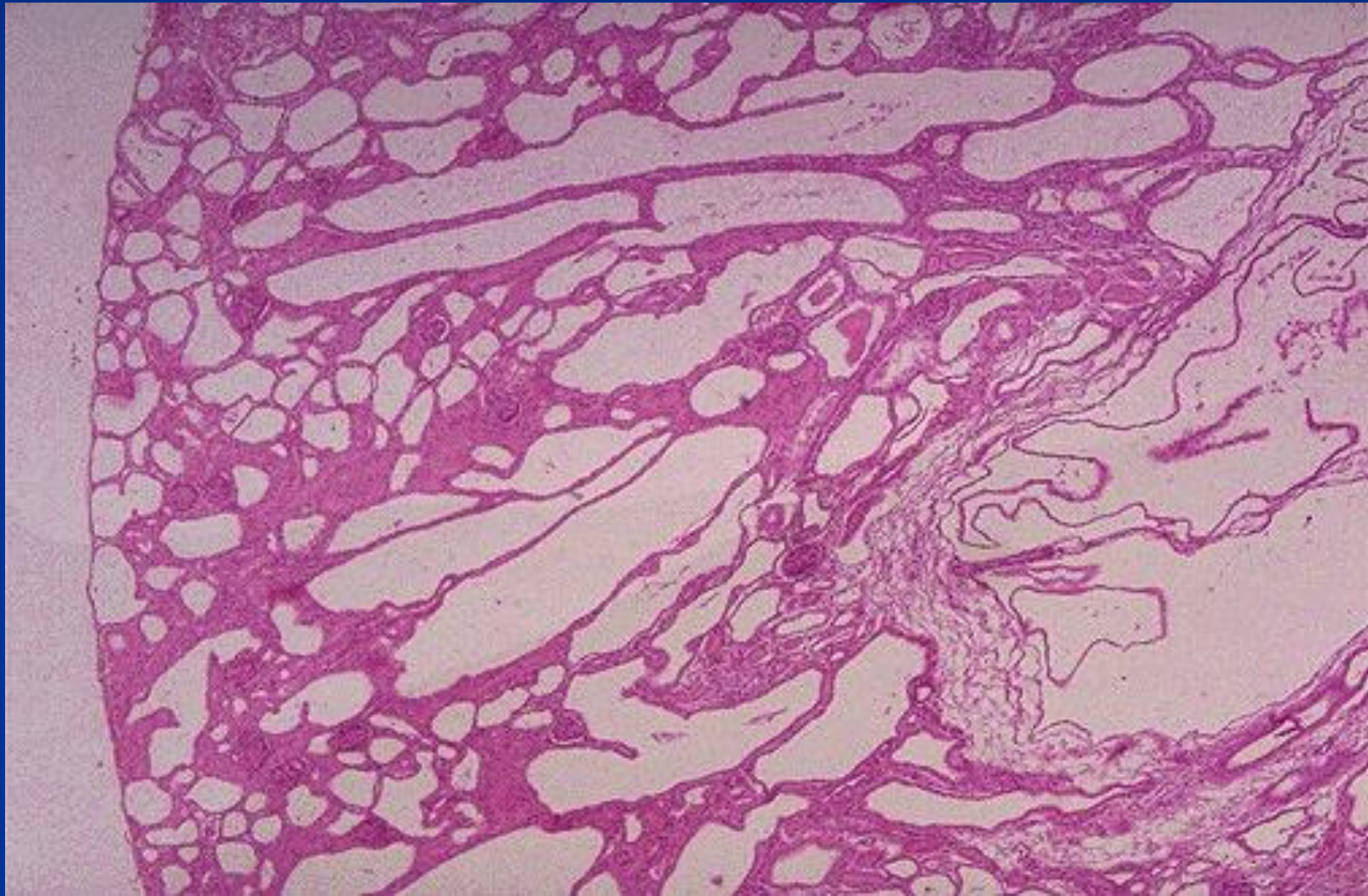
Morphology

- The cysts may be filled with
- a clear, serous fluid
- or,
- more usually, with turbid, red to brown,
- sometimes hemorrhagic fluid.

A and B, Autosomal-dominant adult polycystic kidney disease viewed from the external surface and bisected.
The kidney is markedly enlarged and contains numerous dilated cysts.



- Here is the microscopic appearance of recessive polycystic kidney disease (RPKD).
- Note that the cysts fill most of the parenchyma, and it is hard to find glomeruli.
- Many of the cysts are elongated and radially arranged from the center of the kidney on the right, much like spokes on a wagon wheel.



Clinical Features

Many of these patients remain asymptomatic until renal insufficiency announces the presence of the disease.

Clinical Features

- Hemorrhage or progressive dilation of cysts may produce **PAIN**.
- **Excretion of blood clots causes RENAL COLIC.** The enlarged kidneys, usually apparent on **abdominal palpation**, may induce a dragging sensation.

Clinical Features

The disease occasionally begins with the insidious onset of hematuria, followed by other features of progressive chronic kidney disease, such as proteinuria (rarely more than 2 gm/day), polyuria, and hypertension.

Clinical features

- Patients with *PKD2* mutations tend to have an older age at onset and later development of renal failure.

Clinical Features

- **This form of chronic renal failure is remarkable in that patients may survive for many years with azotemia slowly progressing to uremia.**

Renal Complications

- | | |
|----------------------|------------------|
| 1. Hypertension | 60-100% |
| 2. Gross hematuria | 50% |
| 3. Infection | common |
| 4. Nephrolithiasis | 20-25% |
| 5. Renal failure 50% | by age 60 (PKD1) |

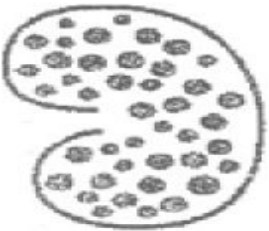
Liver/GI Complications

1. Liver cysts (94% > 35)
 - asymptomatic up to 80%
 - symptomatic uncommon (W:M 10:1)
2. Pancreatic cysts ~10%
3. Intestinal diverticuli ~80% pts with ESRD
4. Hernias ~10%

PROGNOSIS

- Ultimately,
- about 40% of adult patients die of coronary or hypertensive heart disease,
- 25% of infection,
- 15% of a ruptured berry aneurysm or hypertensive intracerebral hemorrhage, and the rest of other causes.

TABLE 20-11 -- Summary of Renal Cystic Diseases

	Inheritance	Pathologic Features	Clinical Features or Complications	Typical Outcome	Diagrammatic Representation
Adult polycystic kidney disease	Autosomal dominant	Large multicystic kidneys, liver cysts, berry aneurysms	Hematuria, flank pain, urinary tract infection, renal stones, hypertension	Chronic renal failure beginning at age 40–60 years	
Childhood polycystic kidney disease	Autosomal recessive	Enlarged, cystic kidneys at birth	Hepatic fibrosis	Variable, death in infancy or childhood	
Medullary sponge kidney	None	Medullary cysts on excretory urography	Hematuria, urinary tract infection, recurrent renal stones	Benign	
Familial juvenile nephronophthisis	Autosomal recessive	Corticomedullary cysts, shrunken kidneys	Salt wasting, polyuria, growth retardation, anemia	Progressive renal failure beginning in childhood	
Adult-onset medullary cystic disease	Autosomal dominant	Corticomedullary cysts, shrunken kidneys	Salt wasting, polyuria	Chronic renal failure beginning in adulthood	
Simple cysts	None	Single or multiple cysts in normal-sized kidneys	Microscopic hematuria	Benign	
Acquired renal cystic disease	None	Cystic degeneration in end-stage kidney disease	Hemorrhage, erythrocytosis, neoplasia	Dependence on dialysis	

Thank You