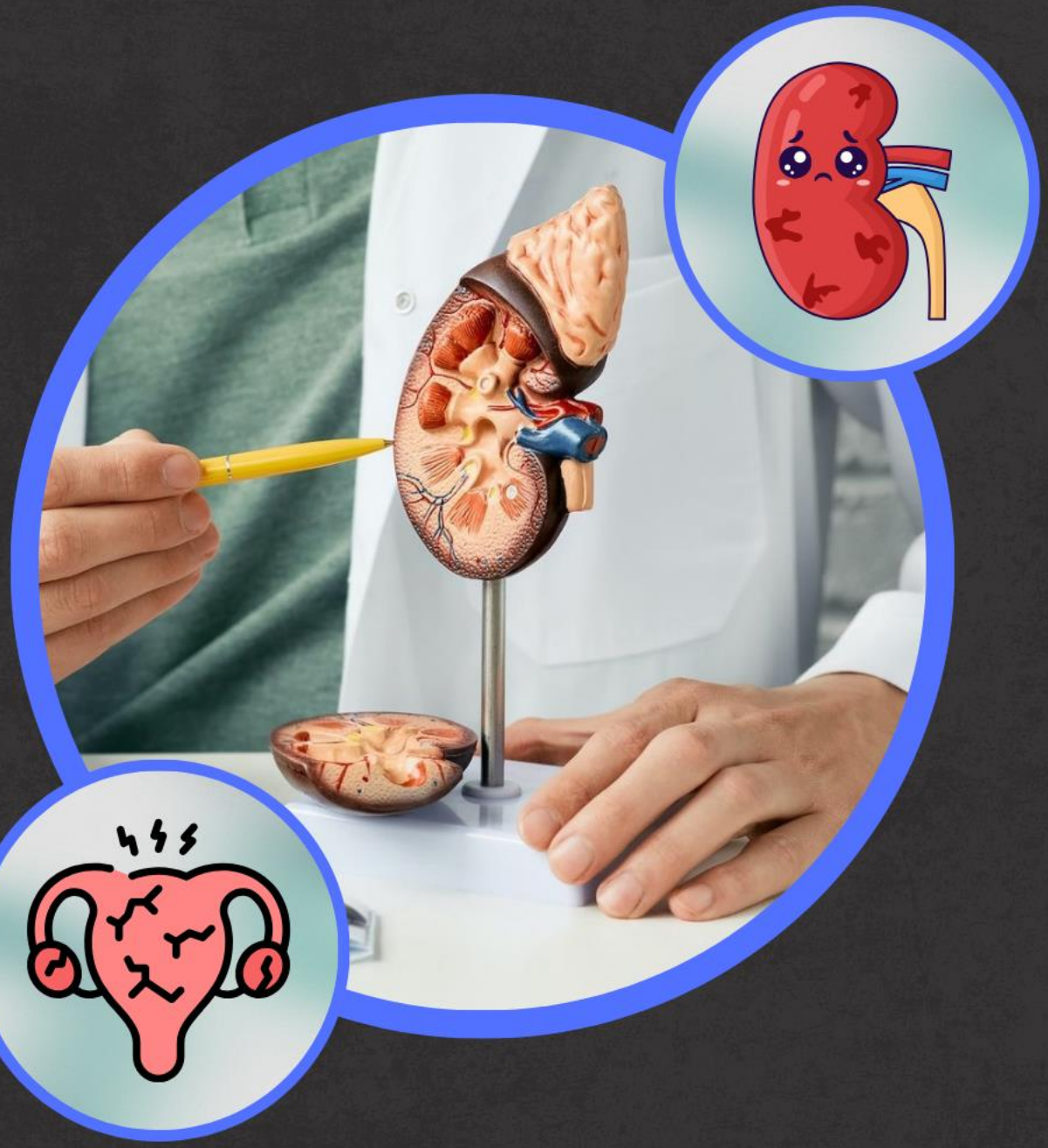


PATHO

MODIFIED NO. 5

الكُتّاب: إسماعيل العارضة وعمر صمادي

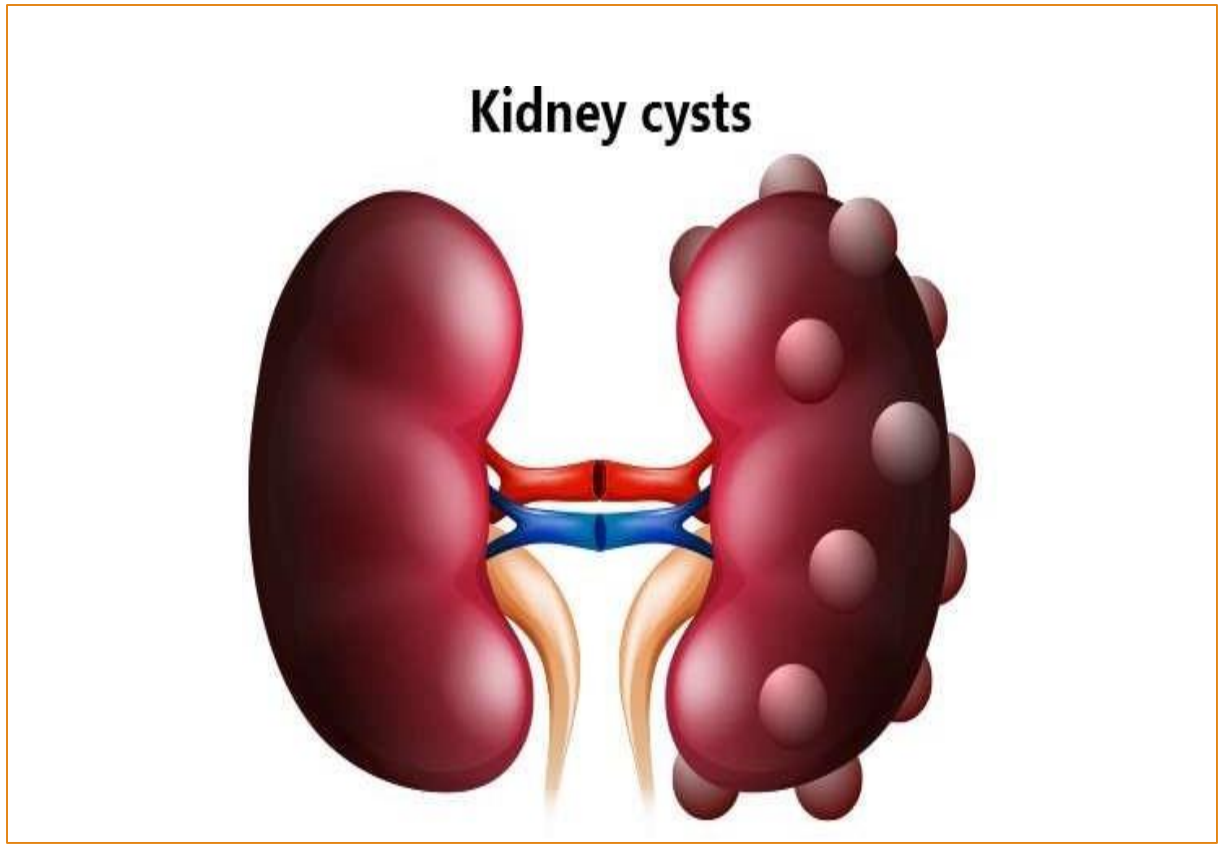
المدققين: محمد مجيد

الدكتور/ة: د.نسرين أبو شاهين

THE UNIVERSITY OF JORDAN

CYSTIC DISEASES of THE KIDNEY

Dr. Nisreen Abu Shahin



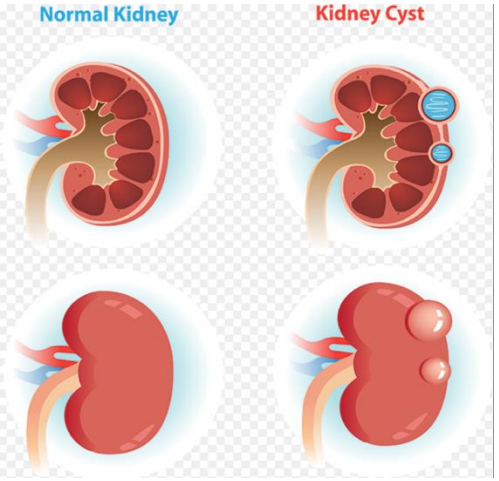
Color code

- Slides
- Doctor
- Additional info
- Important

- A cyst is a space that is filled with fluid.
- There are some different types of cysts that can involve the kidney, they range from completely innocent legions that have no clinical significance to others that inherited and that are leading to renal failure and can threaten the life of the patient.

Types of cysts

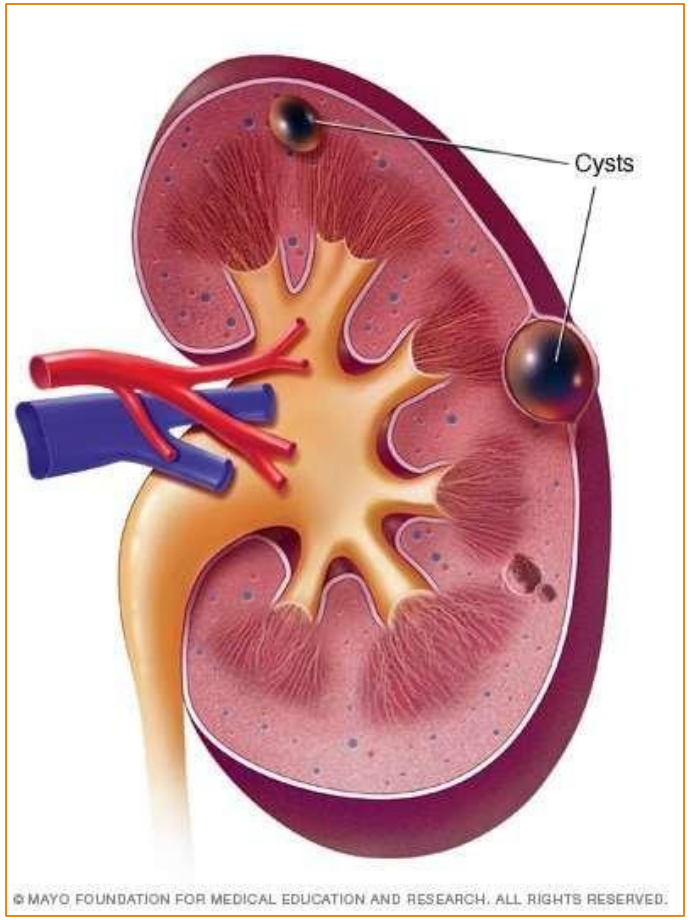
- Extra

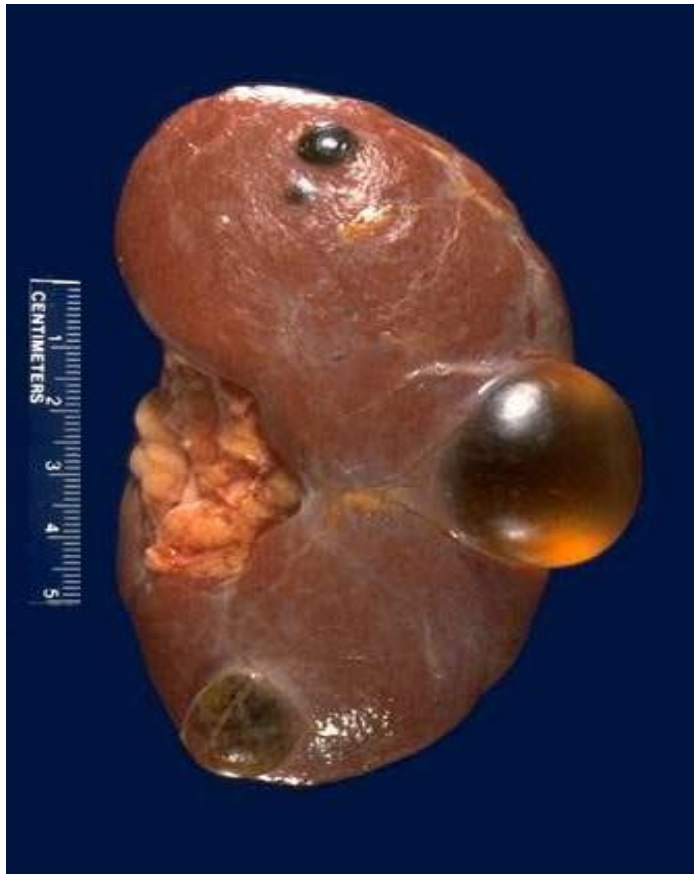


1) Simple Cysts
2) Dialysis-associated acquired cysts
3) Autosomal Dominant (Adult) Polycystic Kidney Disease
4) Autosomal Recessive (Childhood) Polycystic Kidney Disease
5) Medullary Cystic Disease

1- Simple Renal Cysts

- **Multiple or single**
- **1-5 cm in diameter**
- **filled with clear fluid.**
- **confined to the cortex.**
- **no clinical significance.** (Hence they were given the name simple)
- **Usually discovered incidentally or because of hemorrhage and pain**
- **Importance: to differentiate from kidney tumors**



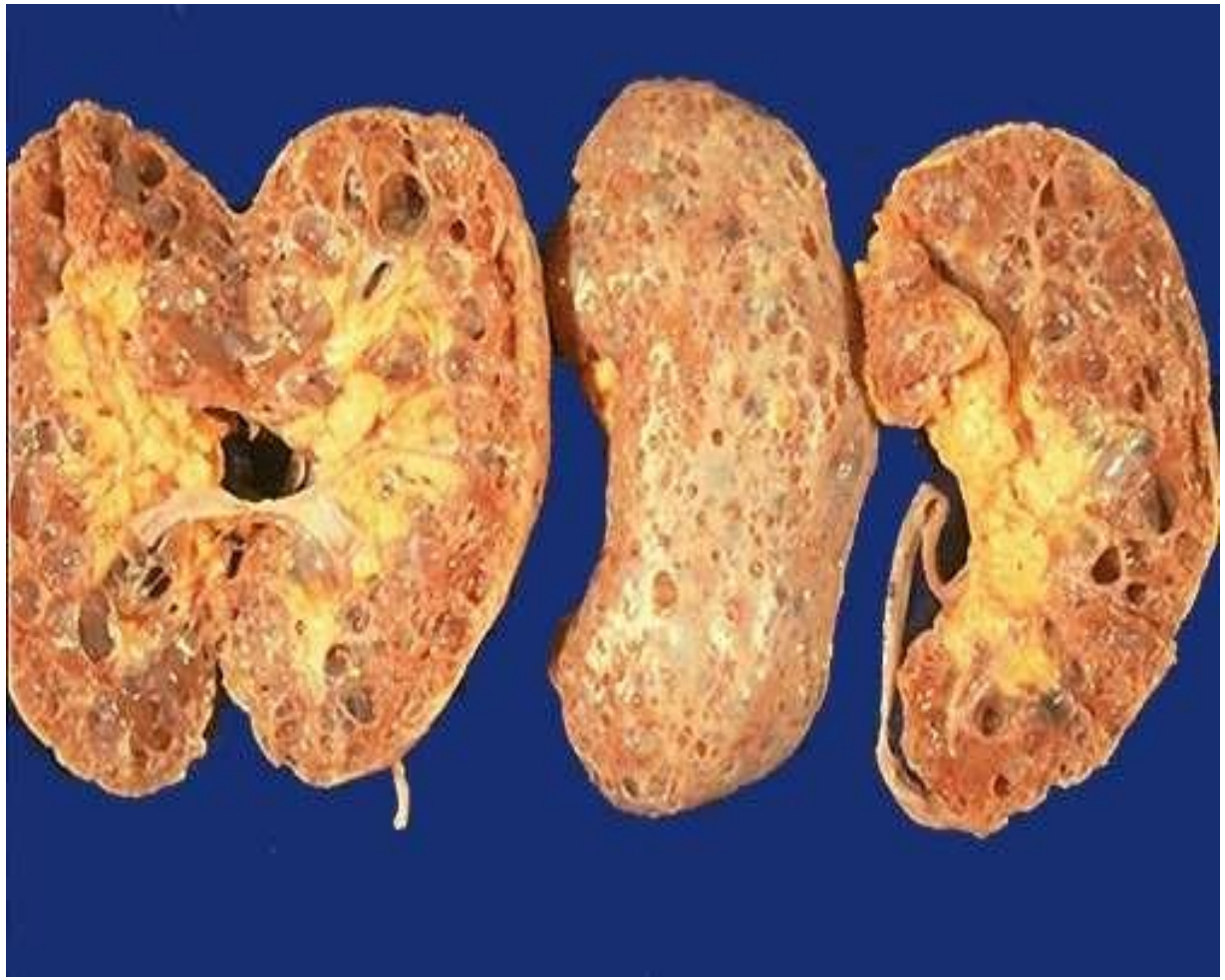


- These structures have usually a favorable prognosis.

2- Cysts Associated With Chronic Dialysis

- In this picture, we are look at end stage kidney.

- **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)

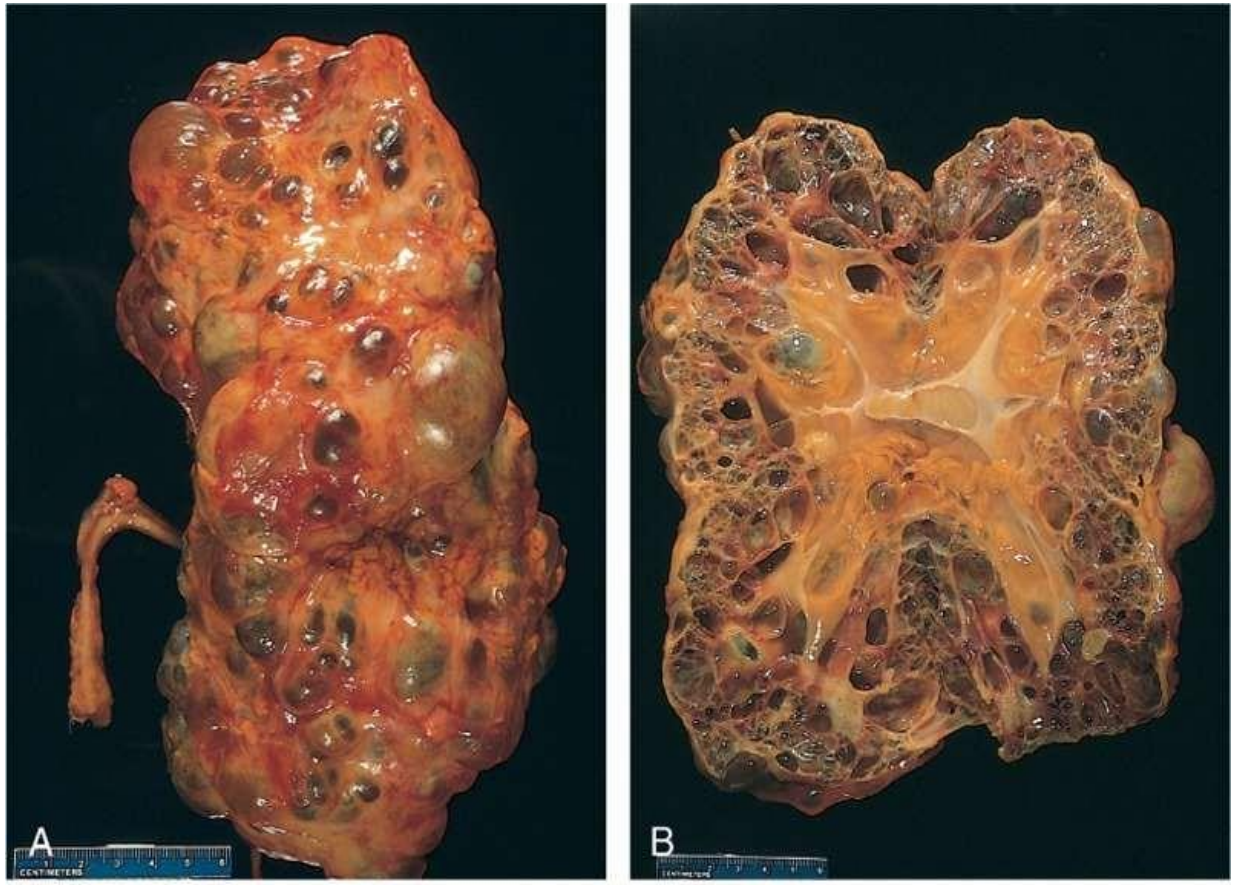


- How can these cysts develop? This is a result of chronic inflammation and irritation that develops within the atrophic renal parenchyma or degenerated renal parenchyma.
- This irritation and chronic inflammation might lead to a cell division there and with cell division the cell might acquire additional mutations and later on they might go on transform into a malignant cells.

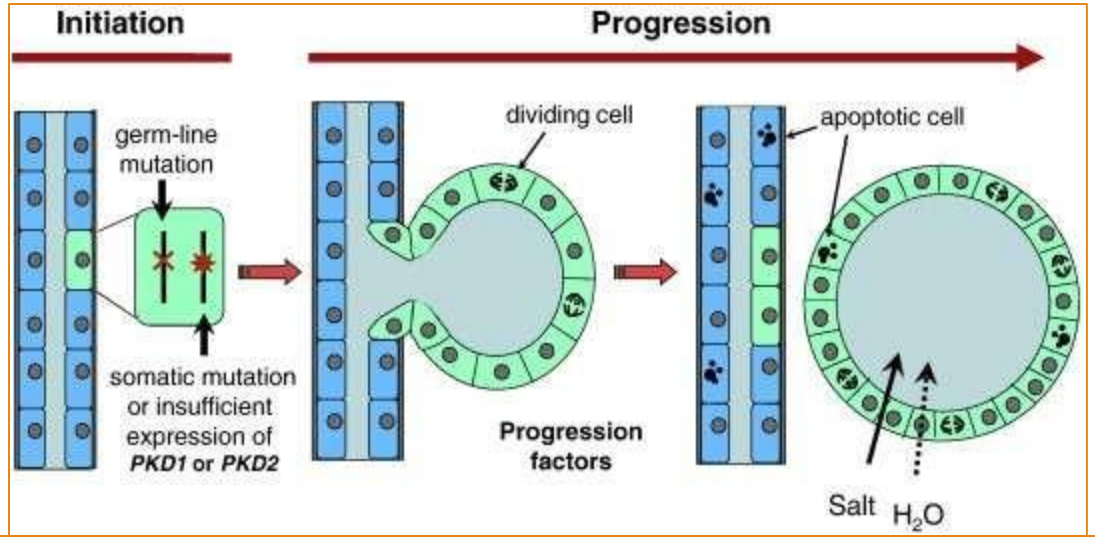
- The name adult polycystic kidney disease is coming from the fact that the symptoms and the manifestations of this condition doesn't appear to be evident or obvious until the patient reaches adulthood.
- In the picture, we are looking to a kidney from a patient with adult type of polycystic kidney disease and you can see the numerous number of cysts that are almost replacing every place or tissue within the parenchyma of this kidney.
- The size of the kidney is also larger than normal and as you can see also the cysts can be found both in the renal cortex as well as in the renal medulla.
- The size as well as the weight of this kidney is huge, it might reach up to 1000 or 1500 grams.

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)** (the most important one)
- ❑**(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

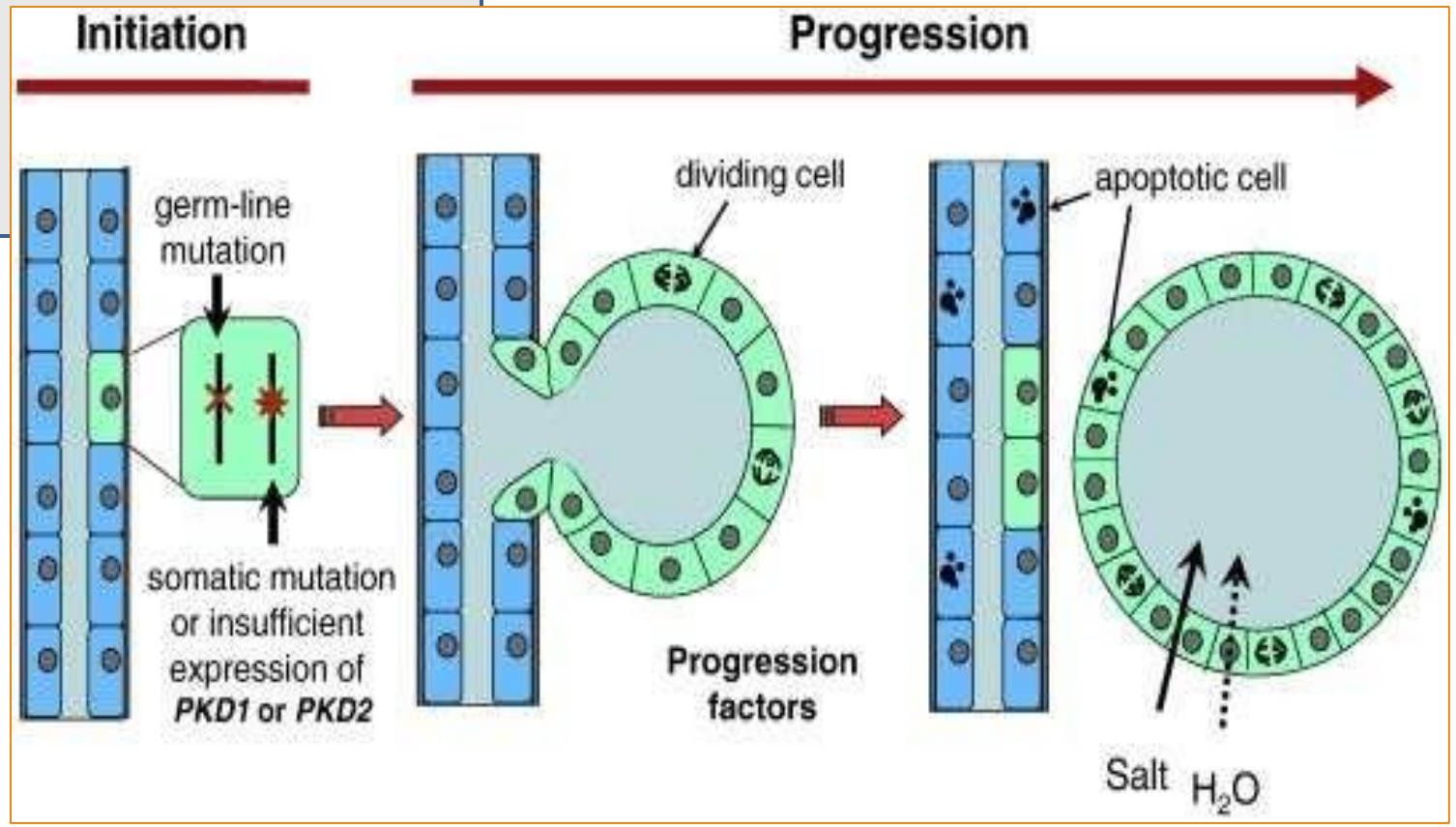
1 *hypertension* (75%)

2 *urinary tract infection, the most frequent complication, infection happens inside the cyst*

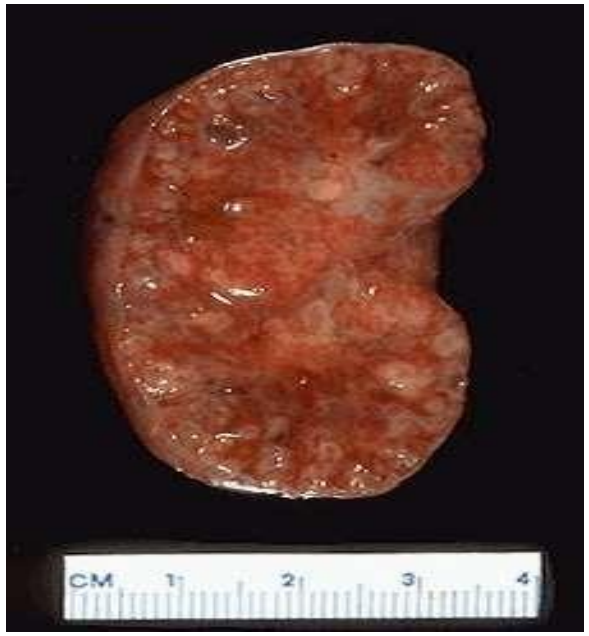
3 vascular aneurysms of circle of Willis (10% -30%)→→ (rupture -> subarachnoid hemorrhage)

4 chronic renal failure at age 50 (≈25% ; % increases with age) after 10 years of initiation of symptoms

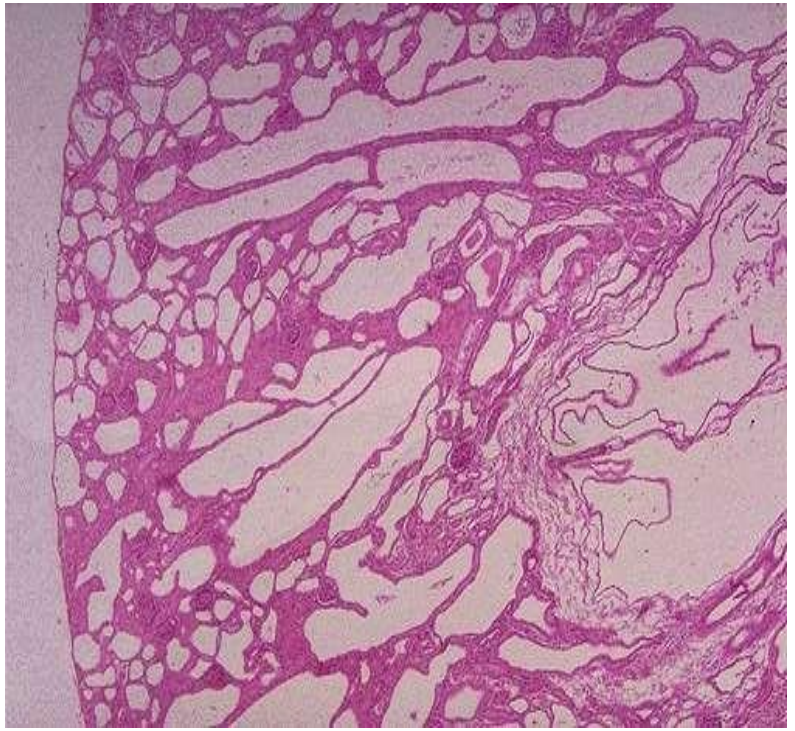
- The mutation in either PKD1 or PKD2 causes abnormal and progressive cell division in renal tubules cells, forming cysts.
- After the cyst grows, it will leave the original tubule and make an isolated cystic spaces.



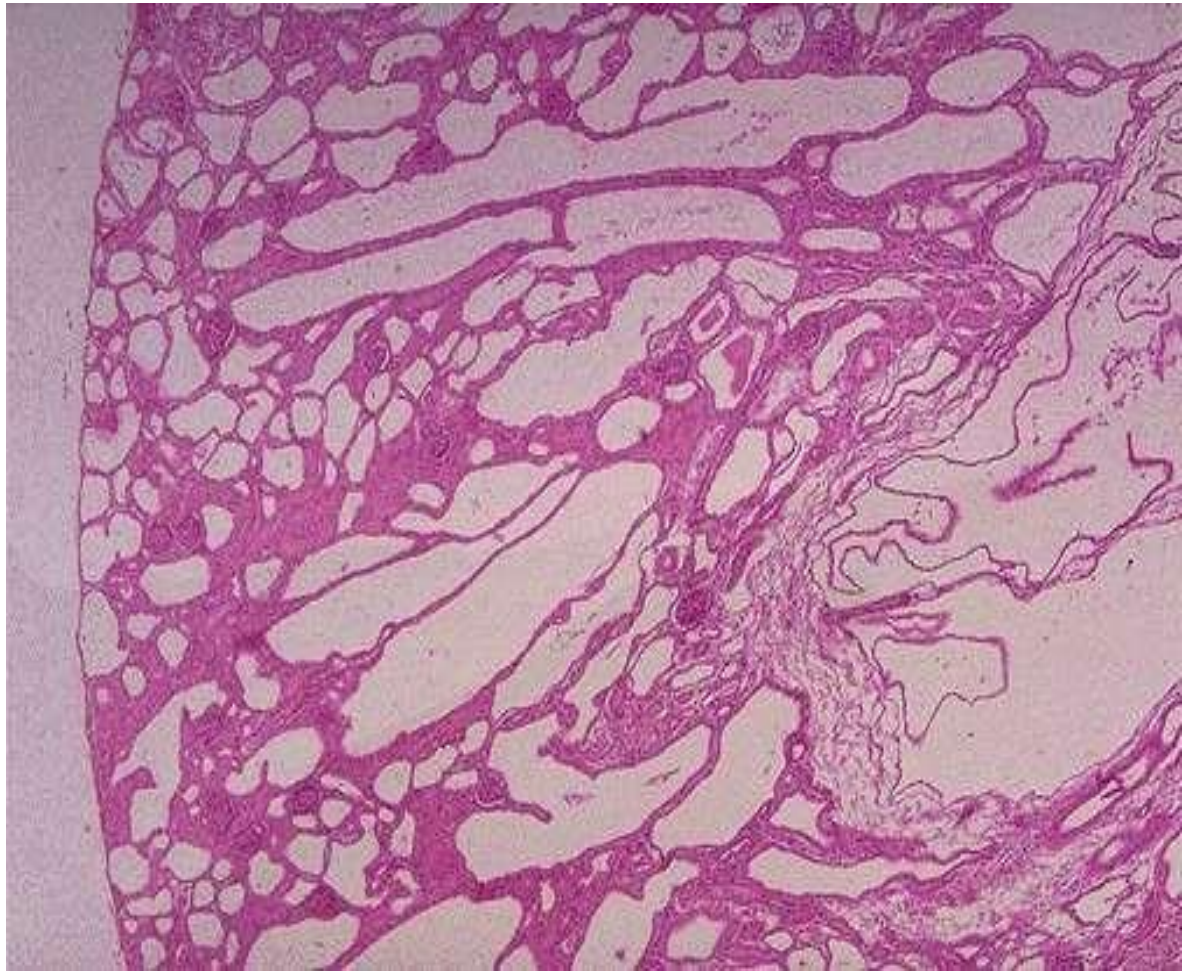
4-Autosomal Recessive (Childhood) Polycystic Kidney Disease



- **autosomal recessive, as the name indicates, begins at childhood**
- **1:20,000 live births.**
- **Types: perinatal, neonatal, infantile, and juvenile. According to the onset of symptoms.**
- **Presents early in life**
- **Associated with liver cysts and fibrosis**
- **Mutations in *PKHD1* gene coding for *fibrocystin*.**
- **Fibrocystin may be involved in the function of cilia in tubular epithelial cells .**



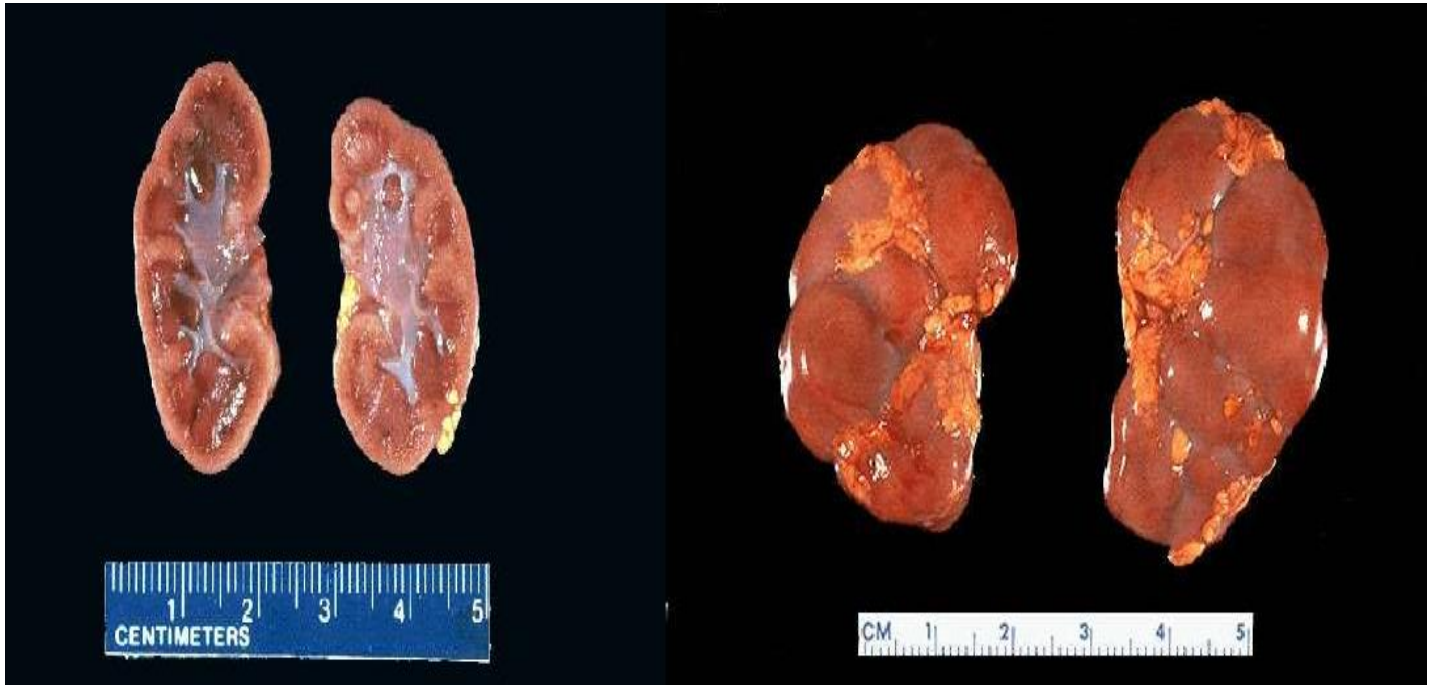
This picture shows ARPKD with cysts in both the cortex and medulla



ARPKD under the microscope, the parenchyma is replaced by cystic spaces (white spaces)

Normal vs childhood polycystic kidneys

NORMAL TERM INFANT KIDNEYS



CHILDHOOD POLYCYSTIC KIDNEYS

- Kidneys in ARPKD are bigger.
- They have small sized cysts in both the medulla and cortex (ADPKD show bigger sized cysts)

Adult vs childhood polycystic kidney disease

- As we said before, adult type has big cysts filling the whole kidney.
- Infantile type has small sized cysts.
- Both diseases could lead to renal failure.

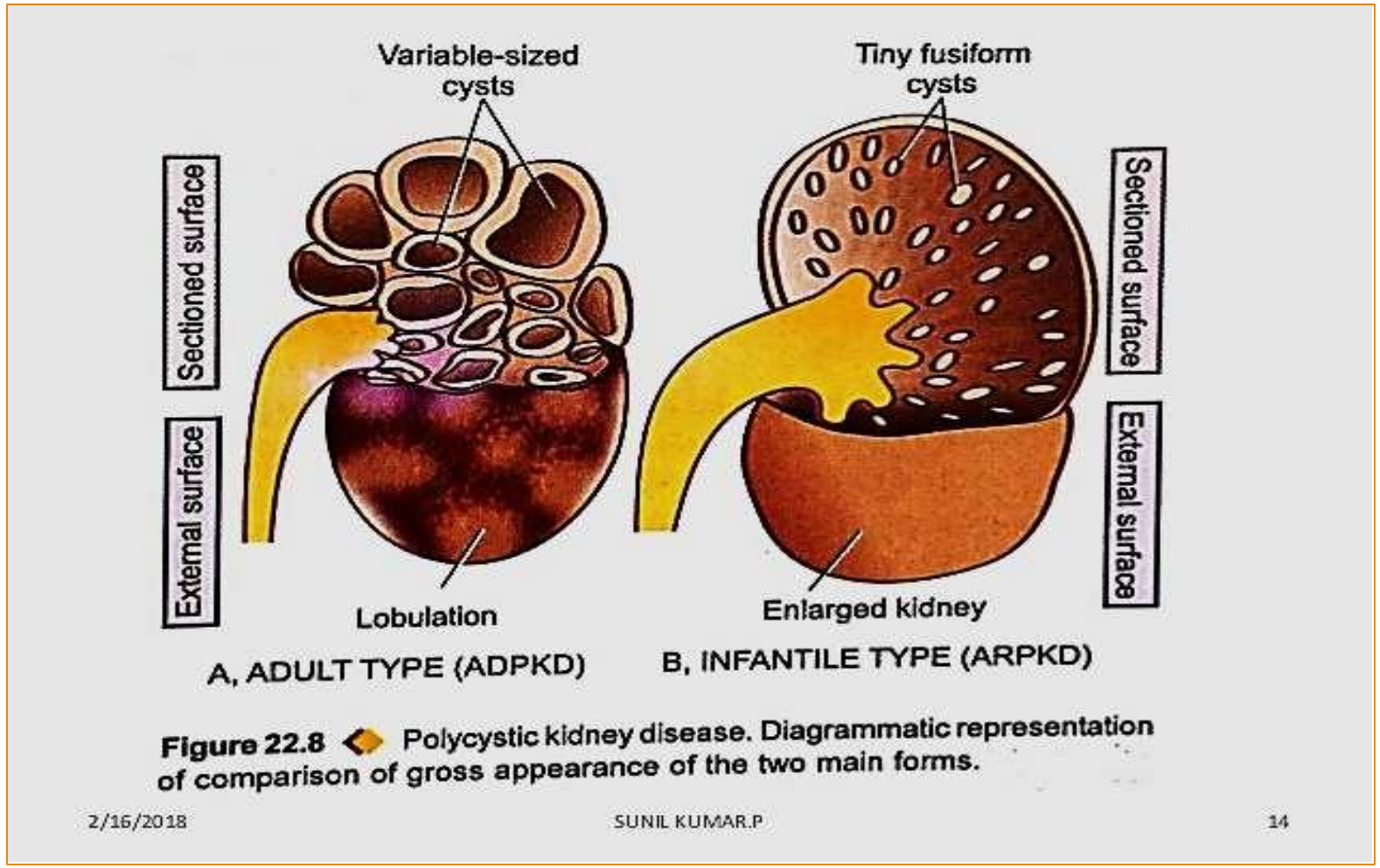


Figure 22.8 Polycystic kidney disease. Diagrammatic representation of comparison of gross appearance of the two main forms.

5- Medullary Cystic Disease

- **2 major types:**

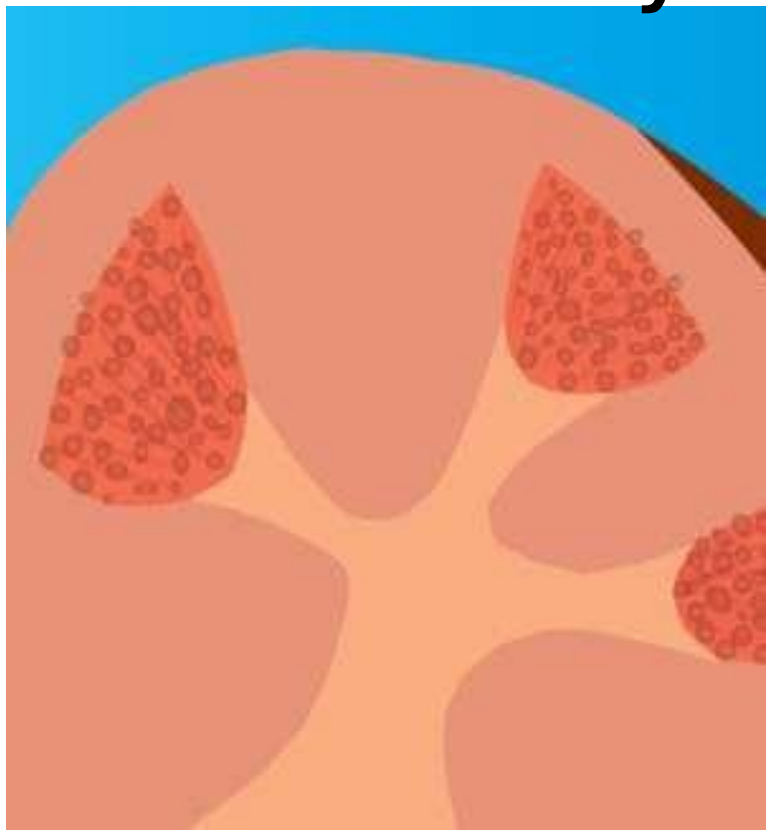
1 medullary sponge kidney

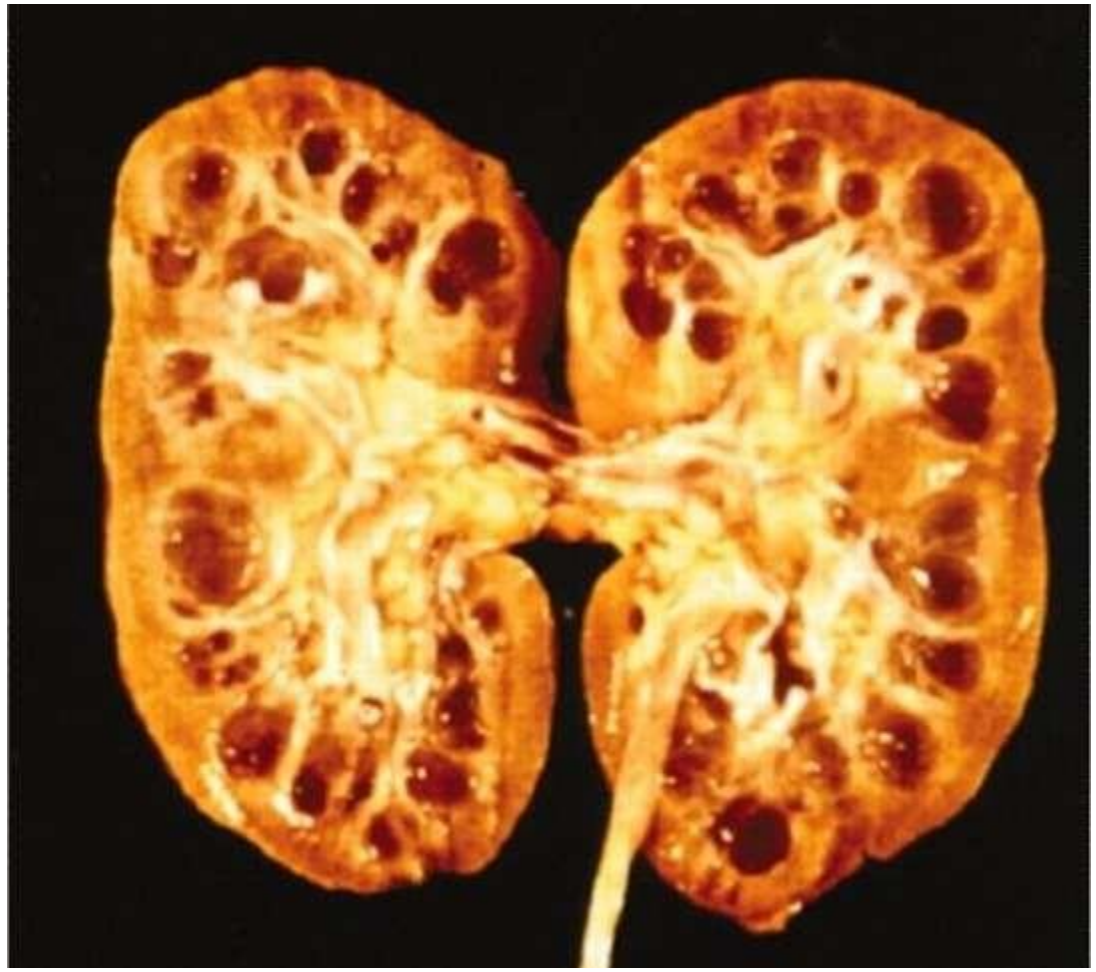
- **common and innocent condition**

2-*nephronophthisis-medullary cystic disease complex, or medullary-uremic*

- **almost always associated with renal dysfunction**
- **Less common**
- **usually begins in childhood**
- **Cysts are at cortico-medullary junction**

Medullary Sponge Kidney



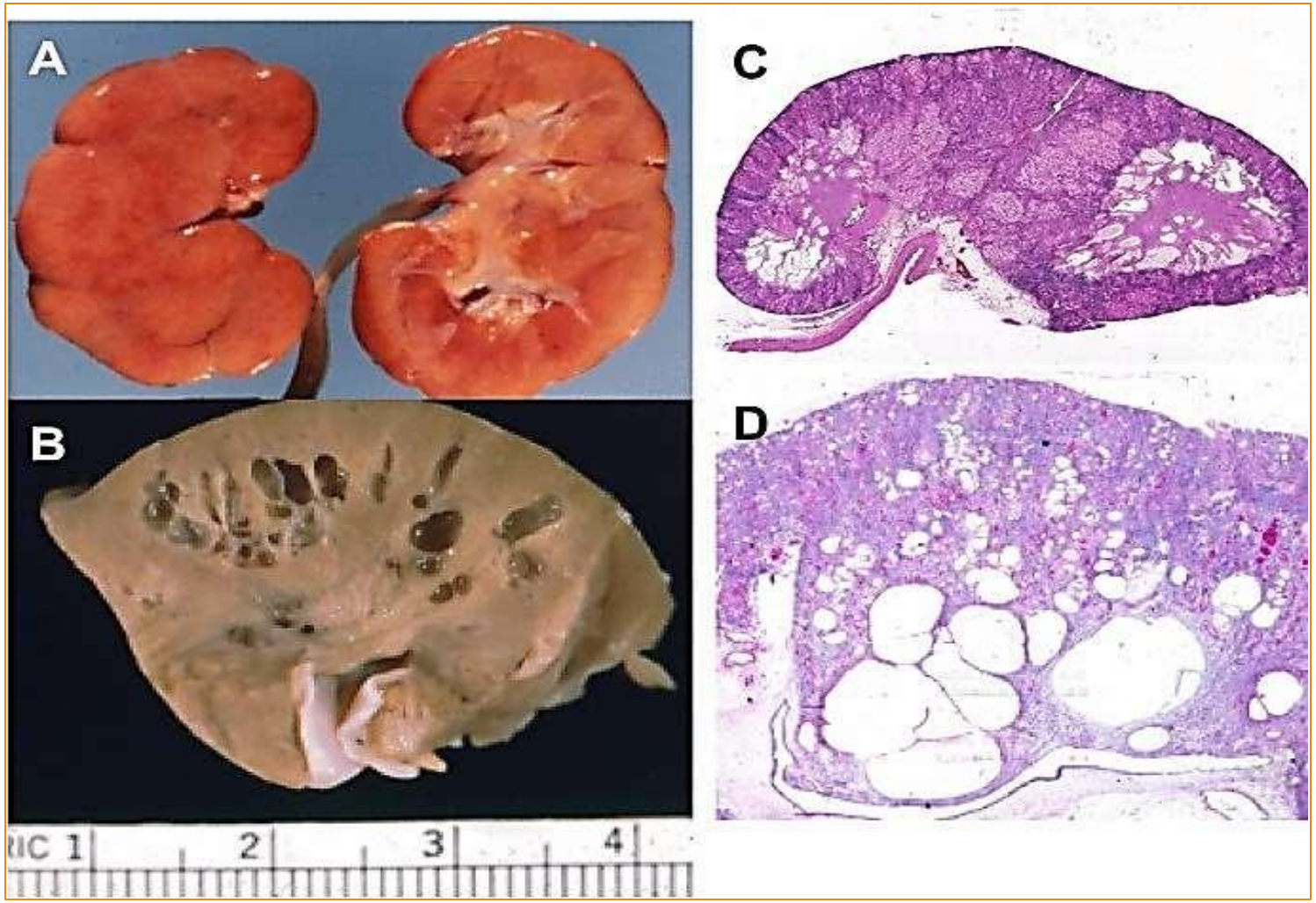


5- Medullary Cystic Disease

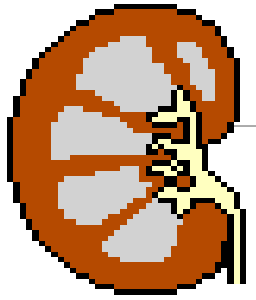
- 2- Nephronophthisis-medullary cystic disease complex (medullary-uremic type)
- ○A worse disease with progressive renal impairment
- ○polyuria and polydipsia (↓tubular function)
- ○ renal failure over 5-10 years
- ○ A positive family history and unexplained chronic renal failure in young patients should lead to suspicion of medullary cystic disease.

- The characteristic presentation is a child with polydypsia and polyuria associated with renal impairment and family history of renal failure.

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

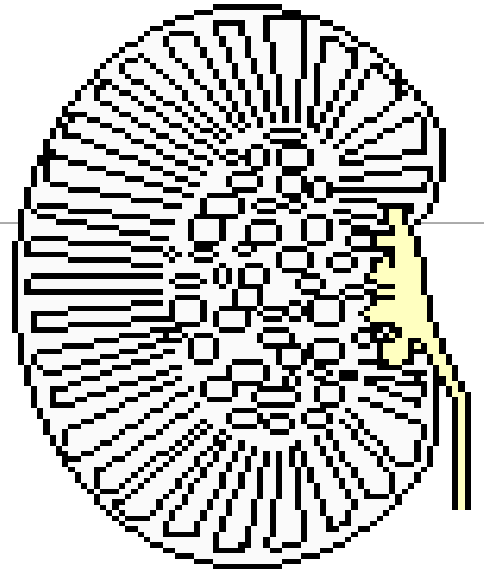


Kidney Cysts

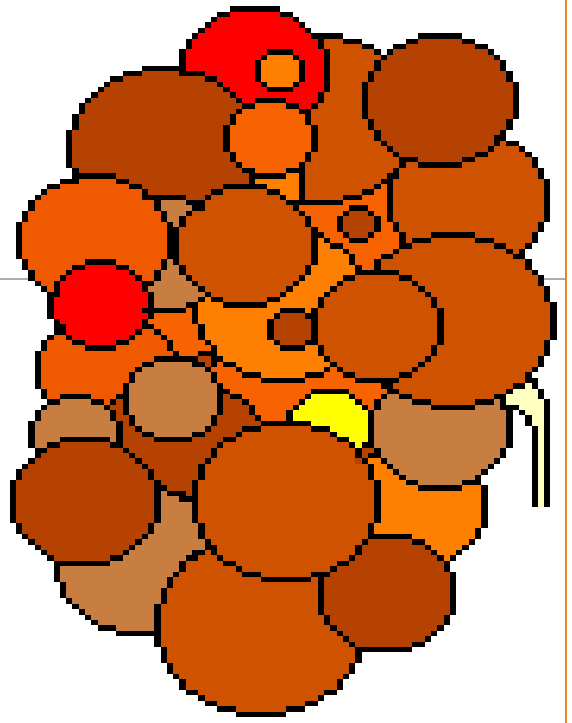


No cysts

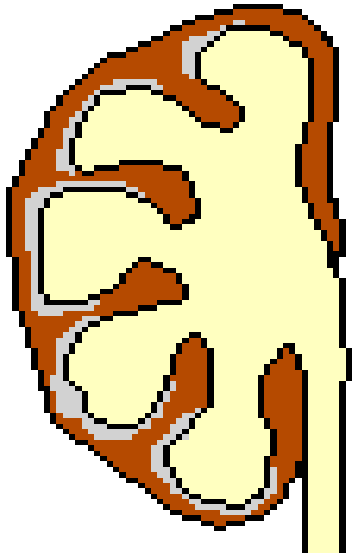
Simple cysts



Recessive polycystic



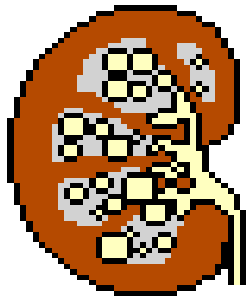
Dominant polycystic



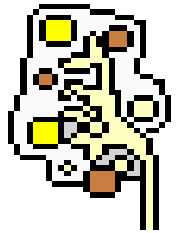
Hydronephrosis is not cysts

"Dysplasia"

Medullary sponge



Medullary uremic



Dialysis cystic

Additional sources

1. page 941-944 on robbins:

https://drive.google.com/file/d/1r2rNfH2NY4bwRJlNoFD7MGKzsfsA_8ch/view?usp=drivesdk

1. https://youtu.be/xfri-2LptWg?si=pKATsr_54Lr71-zZ
2. https://youtu.be/_F9600s_yBU?si=yglPanxIKYVOghsg

اذا ضاقت بك الحال، وارتشى الناس من حولك، تخيل النبي صلى الله عليه وسلم يقول لك:
اصبر حتى تلقاني عند الحوض، واذا نزل بك مرض أوجعك، وأنهك قواك، تخيل النبي صلى الله عليه وسلم يقول لم: اصبر حتى تلقاني عند الحوض، واذا خُلع الحجاب حولكِ إظهارًا لأنوثة او طمعًا بعريس، تمسكي بدينك، وتخيلي النبي صلى الله عليه وسلم يقول لكِ: اصبري حتى تلقيني عند الحوض.
اذا ظلمكم الأقربون، وهجركم المحبون، وقيل فيكم ما ليس فيكم، تخيلوا النبي صلى الله عليه وسلم يعزيكم: اصبروا حتى تلقوني عند الحوض.
اذا تزينت المعاصي، وراودتكم الدنيا عن دينكم، تذكروا أنها فانية، وانها دار عبور لا دار مُقامة، وليكن عزاؤكم في رحلة العمر كلها، اصبروا حتى تلقوني عند الحوض.

VERSIONS	SLIDE #	BEFORE CORRECTION	AFTER CORRECTION
V1→ V2			
V2→V3			

امسح الرمز و شاركنا بأفكارك لتحسين أدائنا !!