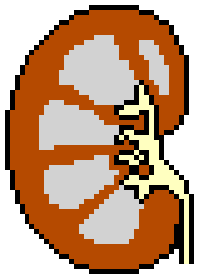
Cystic Diseases of Kidneys

Cystic renal diseases;

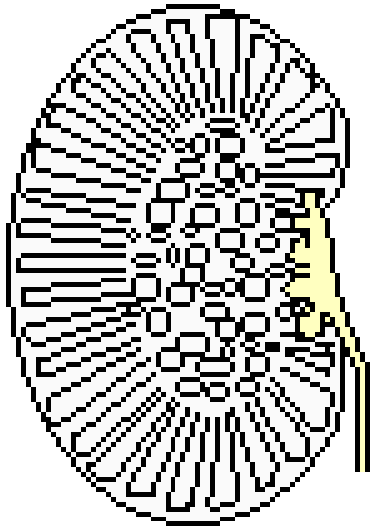
1. Cystic renal dysplasia (Potter II)
2. Polycystic kidney disease
    - 2.1. Adult (autosomal dominant) (Potter III)
    - 2.2. Infantile (autosomal recessive) (Potter I)
3. Medullary cystic disease
    - 3.1. Medullary sponge kidney
    - 3.2. Nephronophtisis
4. Dialysis associated cystic disease
5. Simple renal cysts
6. Parenchymal renal cysts
7. Cystic Change with Obstruction (Potter IV) **(Perihilar renal cysts)**

Kidney Cysts

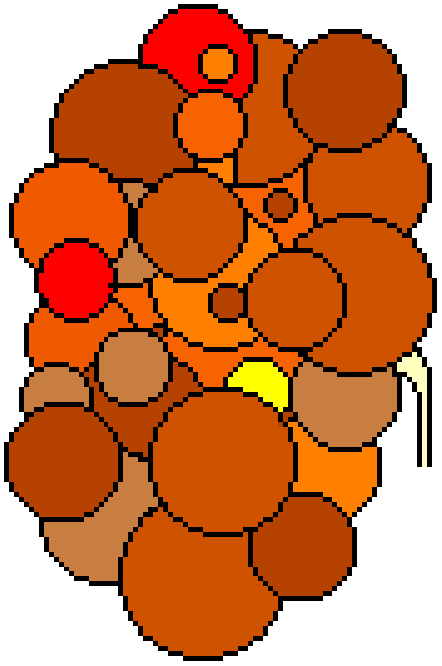


No cysts

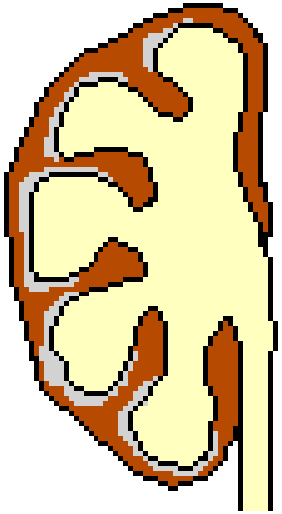
Simple cysts



Recessive polycystic

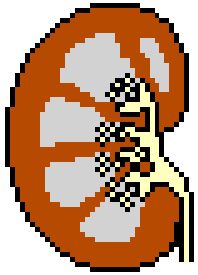


Dominant polycystic

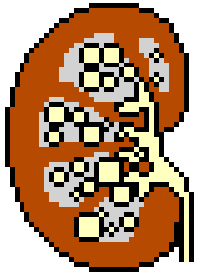


Hydronephrosis is not cysts

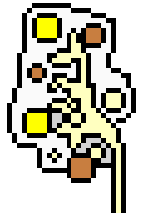
"Dysplasia"



Medullary sponge



Medullary uremic



Dialysis cystic

1. Cystic renal dysplasia (Potter II)

- **The most common form of inherited cystic renal disease**

  abnormal differentiation of the metanephric parenchyma during embryologic development of the kidney

- Unilateral (affected person survives in many cases)

  absence of one functional kidney from birth → the other kidney undergoes compensatory hyperplasia → may reach a size similar to the combined weight of two kidneys (400 to 500 g)

- **There are two main subgroups:**

  Type IIa: affected kidney is large

  Type IIb: affected kidney is quite small (hypodysplasia)

- **Combinations:**

  In unilateral cases:

  - one kidney or part of one kidney
  - Type II a or Type IIb

  In bilateral cases:

  - both can be large
  - both can be small
  - one can be larger and the other small

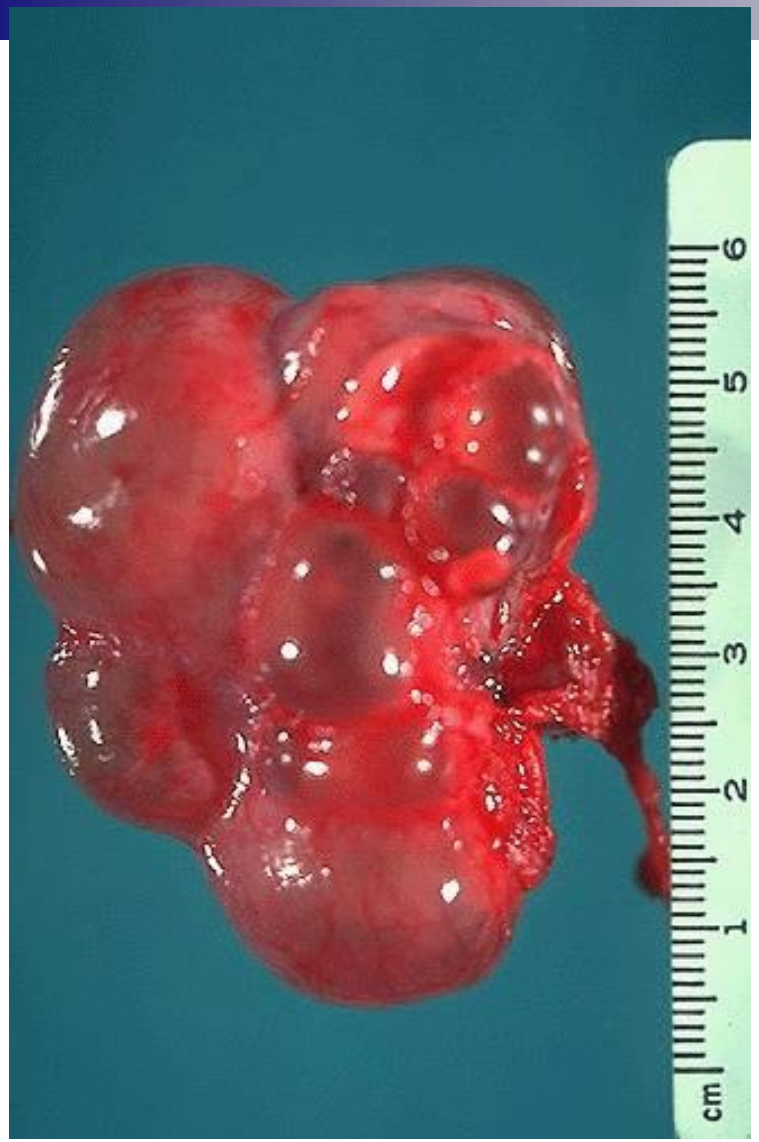
Gross:

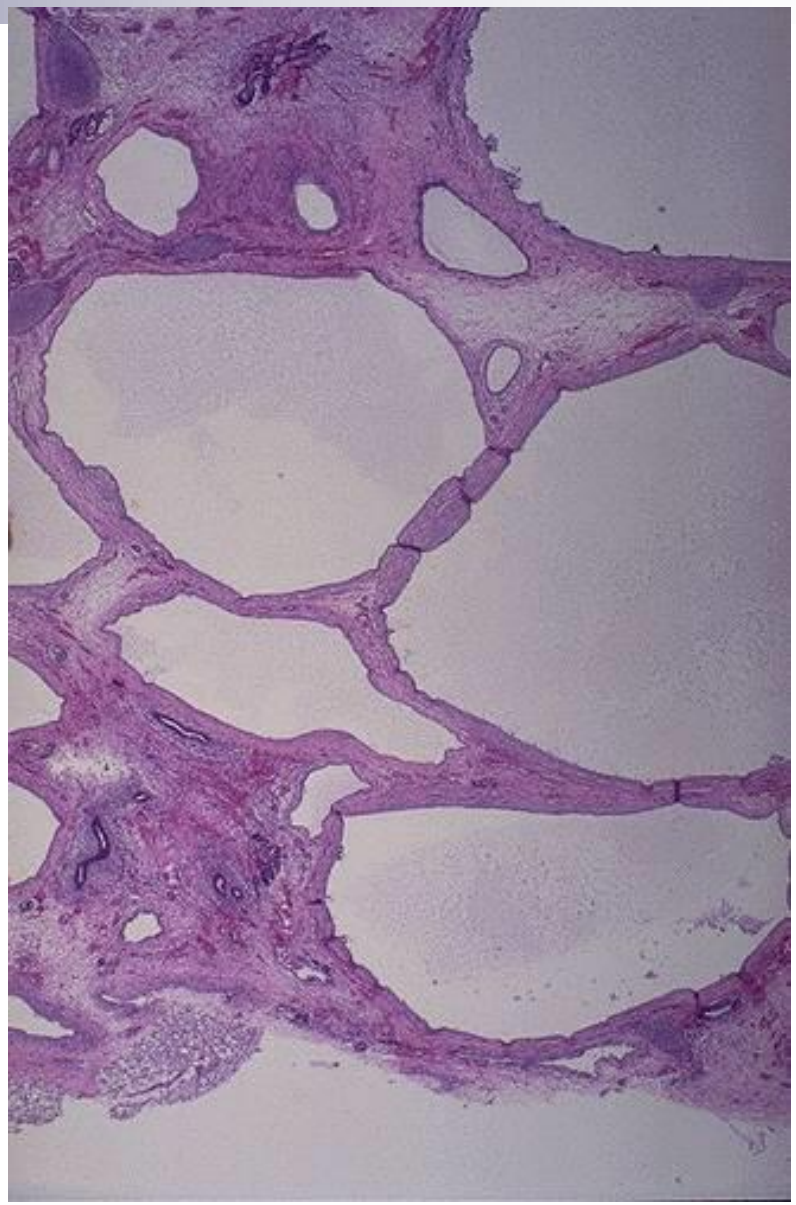
- The cysts
  - are variably sized (from 1 mm to 1 cm)
  - filled with clear fluid

Microscopy:

- Few recognizable glomeruli and tubuli (nephron)
- The hallmark:

  presence of "primitive ducts" lined by cuboidal to columnar epithelium and surrounded by a collagenous stroma
- Increased stroma may contain small islands of cartilage
- The liver will not show congenital hepatic fibrosis
  - Differential diagnosis: Infantile (autosomal recessive) (Potter I)





Cystic renal dysplasia

2. Polycystic kidney disease

2.1. Adult (autosomal dominant): Potter III

2.2. Childhood (autosomal recessive): Potter I

(1:30,000)

2.1. Autosomal dominant polycystic kidney disease (ADPKD); Adult polycystic kidney disease

- Common (1:1,000)
- Autosomal dominant pattern
- High recurrence risk in affected families → 50%
- Rarely manifests itself before middle age
  - diagnosis by ultrasound
  - renal hypertension
  - progressive renal failure as the cysts become larger

    middle aged → older adults
- Half of these patients are on dialysis or transplanted

Pathology

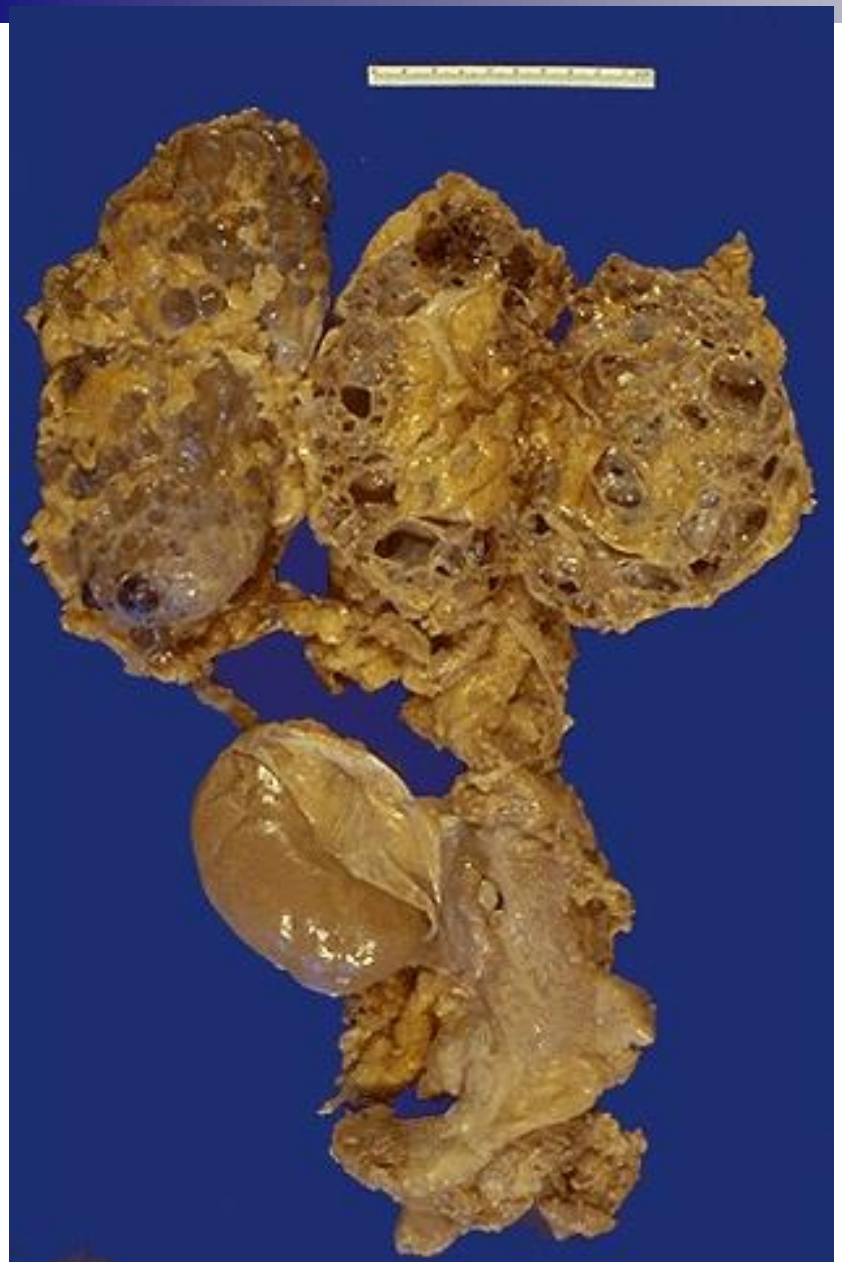
- Very large kidneys (3 or 4 kg or more)
- Hundreds of fluid-filled cysts (up to 4 cm in diameter)
- Hemorrhage into some cysts
- The surrounding normal kidney tissue undergoes pressure atrophy
- Surprisingly, they may still be working

ADPKD **Associated Conditions**

- Liver cysts (30%)
- Splenic cysts (10%)
- Pancreatic cysts (5%)
- Cerebral aneurysms (20%)
- Diverticulosis coli

Outstandig Clinical Findings:

- high blood pressure
- renal failure
- intracranial hemorrhage (ruptured berry aneurysms)
- cyst infections (nosocomial)
- harmless hepatic, splenic and pancreatic cysts (US)



2.2. Autosomal recessive polycystic kidney disease (ARPKD); Infantile (Childhood) polycystic kidney disease

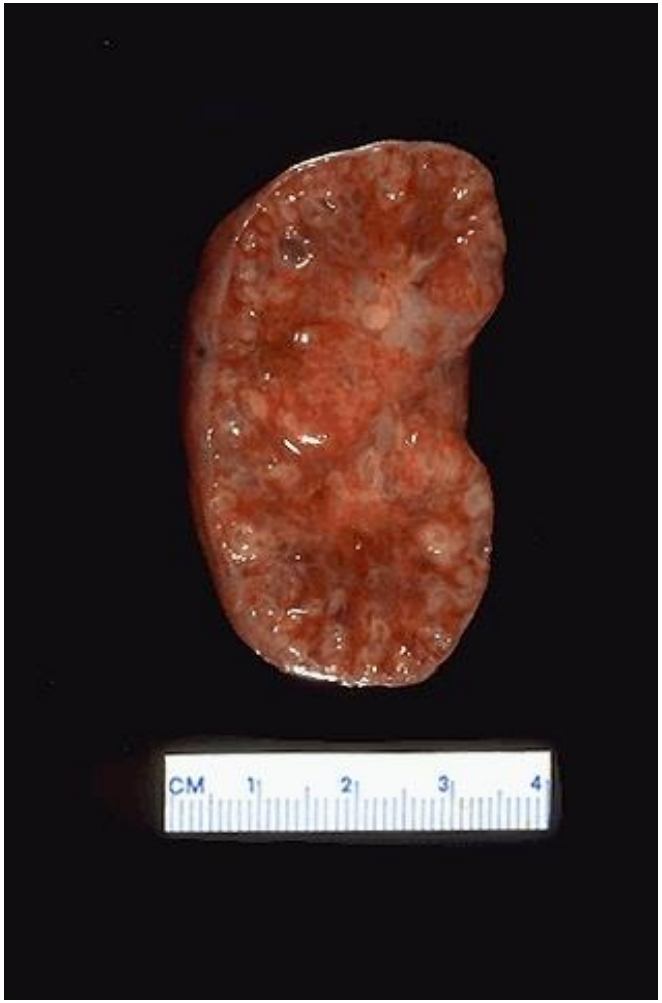
- Autosomal recessive pattern
- Bilateral
- In utero → poor renal function and failure to form significant amounts of fetal urine → oligohydramnios + pulmonary hypoplasia
  - so that newborns do not have sufficient lung capacity to survive

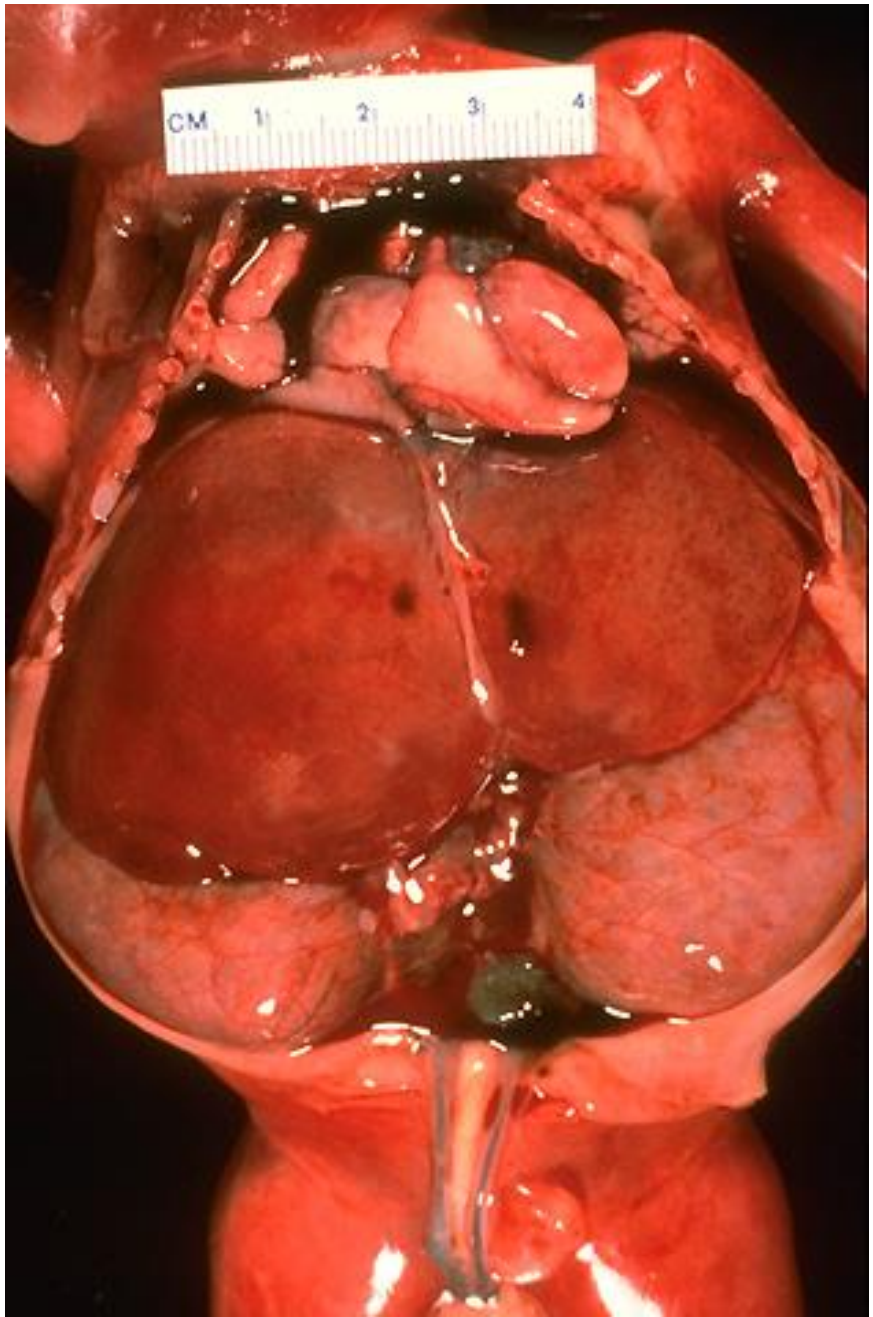
- Huge, white, smooth-surfaced kidneys at birth
- Cysts 1-2 mm in diameter (from the collecting ducts)
- They are arranged in a radial, "sun-ray" pattern perpendicular to the capsule (because the collecting ducts are dilated)
- Fatal in infancy or early childhood
- Enormous kidneys → restrict the ability of the lungs and gut to function
  - Differential diagnosis: Cystic renal dysplasia (Potter II)

congenital portal fibrosis of the liver –present in

- The liver will show congenital hepatic fibrosis
  - Differential diagnosis: Cystic renal dysplasia (Potter II)

- Infantile type/autosomal recessive polycystic kidney disease (ARPKD).





3. Medullary cystic disease

3.1. Medullary sponge kidney

3.2. Nephronophtisis

3.1. Medullary sponge kidney

- Idiopathic
- Very common (1 in 200 people)
- Normal renal functions
- Dilated distal portions of collecting ducts superficially resemble cysts:
  - Urinary stones within the "cysts“
  - Superimposed infection (pyelonephritis)
- Chronic back pain

3.2. Nephronophthisis

Uremic medullary cystic disease

- Most common cause of endstage renal disease in children and young adults
- Pathology:
  - Cysts in the medulla
  - Cortical tubular atrophy
  - Interstitial fibrosis
- The initial manifestations are inability to retain sodium and water

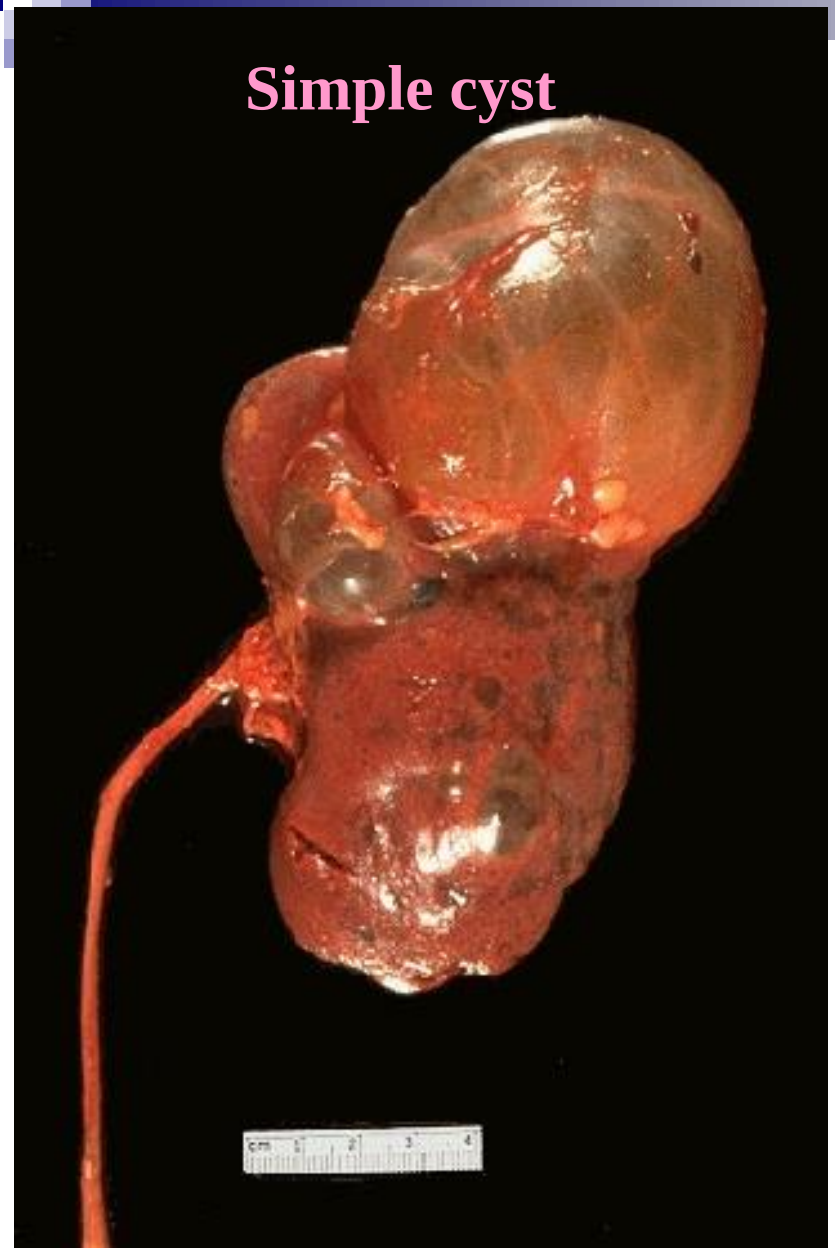
4. Dialysis associated cystic disease ("trans-stygian kidney")

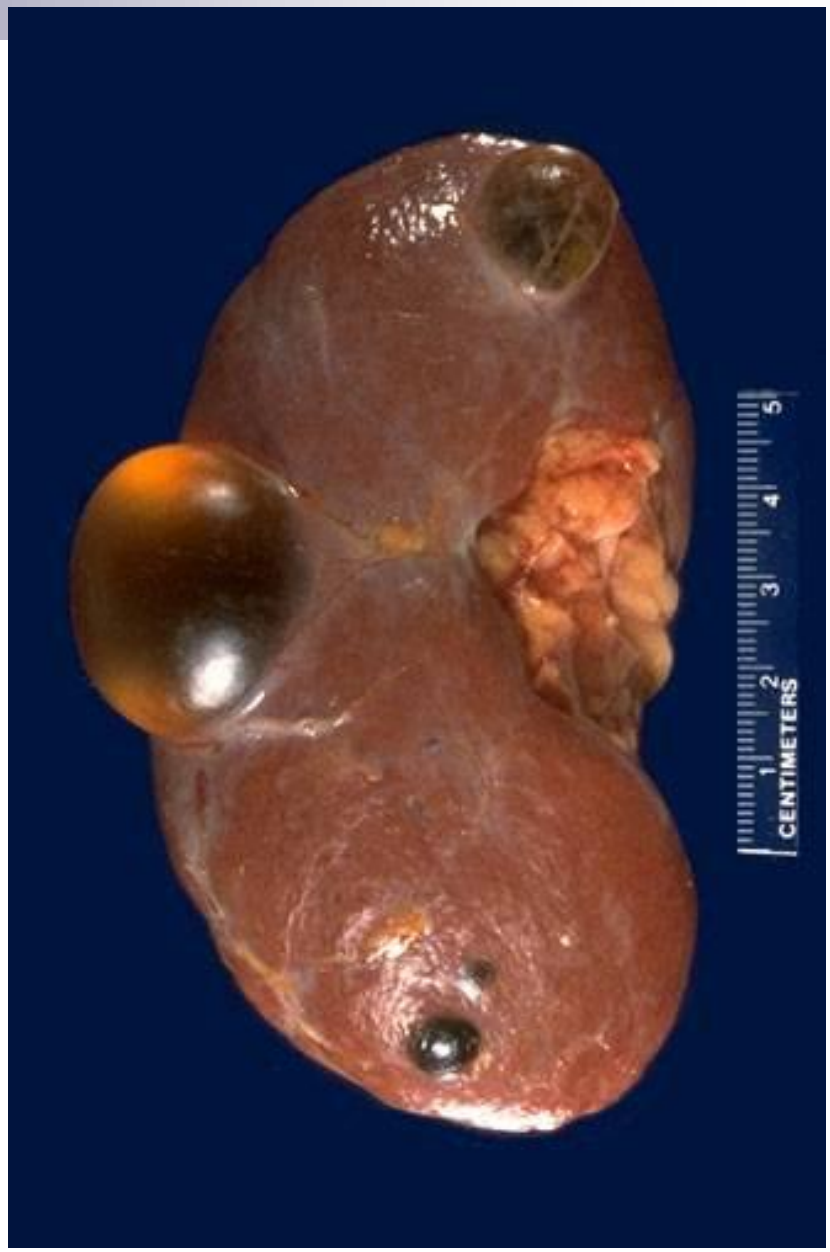
- Styx
- A few remaining tubules stretched wide open ("cysts")
  - stones
  - painful bleeding
  - aggressive carcinomas
- Pathology
  - fibrosis and a few chronic inflammatory cells
  - oxalate crystals in the tubules
  - fibromuscular masses in the blood vessels
  - cortical adenomas and renal cell carcinomas

5. Simple renal cysts

- A few cysts in a kidney
- Old person
- Commonest incidental finding at autopsy
- Often develop after small kidney infarcts ("arterial nephrosclerosis")

Simple cyst





6. Parenchymal renal cysts

- 1. Associated with infection:

  TB

  Echinococcus
- 2. Associated with tumor

  Cystic degeneration of a carcinoma
- 3. Traumatic intrarenal hematoma

7. Cystic Change with Obstruction

- Potter IV
- Fetus and newborn with urinary tract obstruction → renal cystic changes
  - hydroureter
  - hydronephrosis
  - bladder dilation

- Unilateral /Bilateral
  - depends upon the point of obstruction

    For example, posterior urethral valves in a male fetus, or urethral atresia in a male or female fetus, will cause bladder outlet obstruction so that both kidneys are involved

Pathology

The cysts may be no more than 1 mm in size

The cysts tend to be in a cortical location

"cortical microcysts"

Disease	Heredity	Age	Uni- or Bilateral	Gross Features	Microscopic Features	Associated Malformation or Disease
Dysgenesis						
Total renal dysgenesis	None	Usually infants	Either	Nonreniform "cluster of grapes"	Macrocysts, primitive mesenchyme; cartilage	Cardiovascular, gastrointestinal, central nervous system, and urinary tract abnormalities
Segmental dysgenesis	None	Any	Usually unilateral	Irregular cysts with scarring		
Polycystic renal disease						
Adult polycystic disease	Autosomal dominant	Adults; rarely children	Bilateral	Large, bosselated, reniform; cysts in cortex and medulla	Glomerular cysts and nondescript cysts anywhere along nephron	Gross cysts of liver, pancreas, lung; cerebral aneurysms
Infantile polycystic disease	Autosomal recessive	Infants, children	Bilateral	Large smooth kidney with radial fusiform cysts in cortex and medulla	Flat, cuboidal epithelium	Congenital hepatic fibrosis
Medullary cystic disease						
Medullary sponge kidney	None	Any age; usually adults	Either	Cysts at tip of papillae	Papillary cysts lined by flattened epithelium; medullary calcification	Renal stones
Uremic Medullary cystic disease	Variable	Older children and adolescents	Bilateral	Small coarsely scarred kidneys with 1- to 20-mm cysts at corticomedullary junction	Flat epithelial lining of cysts; glomerular sclerosis with interstitial fibrosis and atrophy; patchy inflammatory infiltrate	None
Glomerulocystic disease	None	Newborns, infants, and children	Bilateral	Enlarged, reniform with 1- to 8-mm cysts throughout cortex	Glomerular cysts; collapsed tuft in glomerular cysts	None
Simple cyst	None	Adults; rarely children	Usually unilateral	Single cyst or few cysts usually in the cortex	Nondescript lining	None
Dialysis cystic disease	None	Adults on long-term dialysis	Bilateral	Multiple cysts throughout cortex	Flattened tubular epithelial lining	Increased incidence of renal adenocarcinoma