Anomalies and cystic diseases of the kidney

– Anomalies

① Agenesis: Complete absence of renal tissue Unilateral or bilateral
associated with Insulin resistance, DM

② Horseshoe Kidney: Most common Congenital anomaly / 90% fused at lower Pole

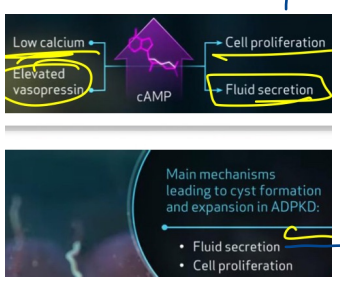
③ Hypoplasia: Reduced nephrons and Pyramids 6 or less (Normal Pyramids = more than 7
Associated with PAX2 mutation

Autosomal dominant Polycystic Kidney disease

– Mutations in Polycystin1 (PKD1) / Polycystin2 (PKD_2)
– associated with TSC2
– 1–2/1000 birth
– Male = female
– ADULT

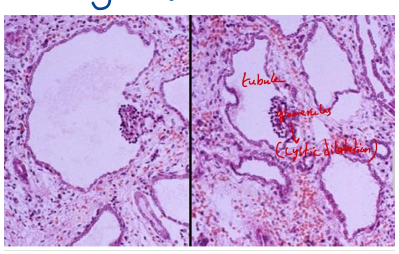
both Present in renal tubular epith cells
* 85% located in Primary cilia + cell membrane of renal tubular epith cells
15% Primary cilia and endoplasmic reticula

* defect in PKD_1 or PKD_2
→ ↓ Calcium + ↑ vasopressin → ↑ cAMP



→ cystic dilatation

* histologic description
– Cysts are lined by Cuboidal / low culomnar or flattened epith
* 3rd Common Cause of end stage renal disease
* Ass with Von Meyeburg Complexes in liver (bile duct dilatation)
+ Hepatic cysts, aneurysms
* Autosomal ressesive more Ass with hepatic cysts And cyst only in liver + Pancreas
AD → cysts in multiple organs
* most common Cause of death → Hypertention + Heart disease
* early stage GFR Normal

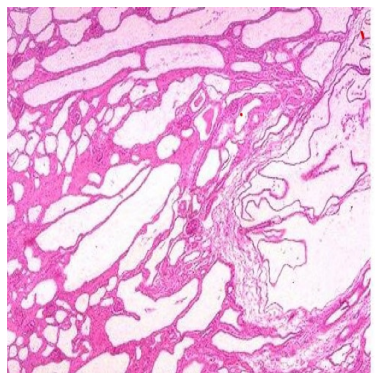


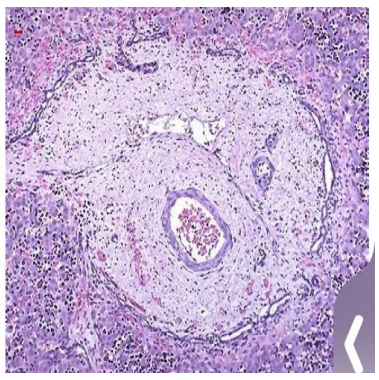
Autosomal recessive Polycystic Kidney disease

– Mutation ⇒ PKHD1 (fibrocystin / Polyductin)
– Children
– Portal fibrosis
– Markedly enlarged Kidney with smooth surface
– small cysts in Cortex and medulla
* Histologic description → Cysts lined by low culomnar, cuboidal or flattend cell

* كلهم نفس الهستولوجي
فاجتد أفرق بينهم غيرجع للكلينيكال manifestation

* اذا جابلي حصوة وحكالي
– fibrosis in liver = AR
– Adult / Aneurysm = AD



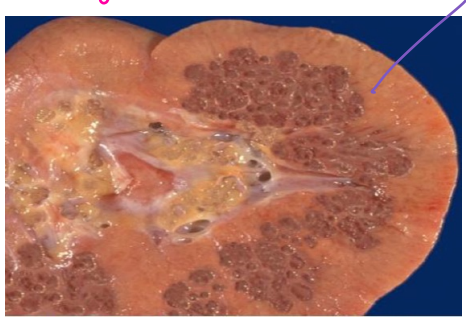


Acquired Cystic Kidney disease

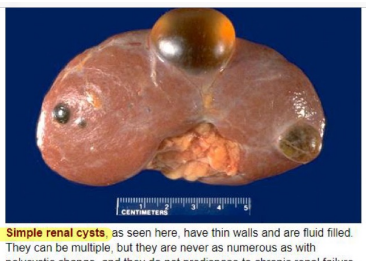
– 3 or more cysts Per Kidney in Patients with longstanding hemo or Peritoneal dyalysis
– Male > Female
– Adult > children
* Risk of Renal cell Carcinoma
– 40% of Kidney with cysts
(AD + AR = (all Kidney with cysts)
– Histologic discription ⟹ Low Culmnar / Cuboidal
– cyst may Contain oxalate crystals

Medullary Spongy Kidney

– Sporadic cystic disease
– bilateral cystic dilations of medullary collecting ducts (Normal Cortex)
– Ass with hemihypertrophy of body
– usually in Adult Marfan's Syndrome Caroli // Ehlers Danlos Syndrome
* Normal sized Kidney with Multiple small cysts in medullary Pyramids ⇒ Spongylike appearance
* Mostly bilateral



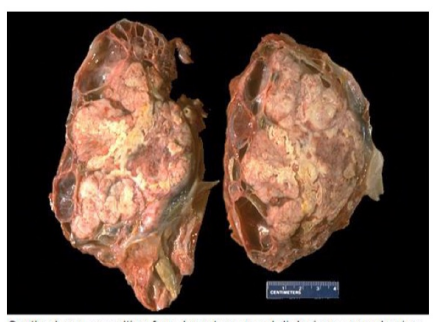
* Cuboidal epith
* Managment ⇒ Cranberry Juice


Simple renal cysts, as seen here, have thin walls and are fluid filled. They can be multiple, but they are never as numerous as with polycystic change, and they do not predispose to chronic renal failure or to neoplasia. Such simple cysts become more common as persons become older.

cyst less than normal
1–2 small size


This is a multicystic dysplastic kidney. This condition must be distinguished from ARPKD because it occurs only sporadically and not with a defined inheritance pattern, though it is more common than ARPKD. The cysts of multicystic renal dysplasia are larger and more variably sized than those of ARPKD. Often, multicystic renal dysplasia is unilateral. If bilateral, it is often asymmetric. If bilateral, oligohydramnios and its complications can ensue, just as with ARPKD.

Sporadic


Cystic change resulting from long-term renal dialysis may rarely give rise to renal cell carcinoma. A large irregular tan variegated **mass** is seen here on sectioning of a kidney that has large **cysts** arranged around the mass.

Cyst + mass
=
Renal cell Carcinoma
Ass with acquired cystic KD

* لما نت ♡