Cystic diseases

Edited based on 019 Slides

Cystic diseases of the kidney	Inherited/ Acquired	Size and number	Clinical appearance	Location	Complications	Notes
Simple Cysts		- Single or multiple - 1-5 cm in diameter	- No clinical Significance - Hemorrhage - Pain - Filled with clear fluid - **Favorable** Prognosis	**- Confined to the cortex**		Discovered incidentally or because of hemorrhage and pain Imp to differentiate it from kidney tumors
Dialysis -associated Acquired Cysts	**Acquired** -In patients with renal failure who have **prolonged dialysis**	Numerous/ multiple		-Cortex -Medulla	**-Renal Carcinoma** (100 times greater than in the general population) - Pain - Hematuria (due to hemorrhage)	Can transform into a malignant tumor.
Autosomal dominant (Adult) Polycystic Kidney Disease	**Inherited** Autosomal dominant 1. PKD1 gene 85-90% (MORE COMMON) – Polycystin 1 2. PKD2 gene - 10-15% Polycystin2	**-Multiple** **-Bilateral** **-Varies in size** (mostly large)	- Onset of symptoms in Adults (4th decade) - Increase in kidney weight and size - Multiple bilateral cysts replacing renal parenchyma. - Forms lobules on the external surface of the kidney **Symptoms**: 1. Flank **pain**	- Cortex - Medulla	**-Hypertension (70%)** -Urinary infection -Vascular aneurysms of circle of Willis → **Subarachnoid hemorrhage** **Most feared complication:** - renal failure at age 50	**Represents 10% of chronic renal failure** -eventually destroy the renal parenchyma

			2. **Heavy dragging sensation** 3. Abdominal mass - Hemorrhage 4. **Obstruction** (stones/hemorrhage of the cyst) 5. Intermittent gross **hematuria**			
Autosomal Recessive (Childhood) Polycystic Kidney Disease	Inherited Autosomal recessive - PKHD1 gene- Fibrocystin mutation (function of cilia in tubular cells)	-Multiple **-Tiny fusiform cysts**	-Onset of symptoms in **childhood** Presents early in life **- Associated with liver cysts** - Enlarged kidney	-Thrown and seen within the parenchyma	**-Chronic renal failure**	Types: 1. Prenatal 2. Neonatal 3. Infantile 4. Juvenile
Medullary Cystic Disease (Nephronophthisis-medullary cystic disease complex (medullary-uremic type)	-Present with positive family history		-Always associated with renal dysfunction - Begins in **children** -Polyuria	-**Cortico-medullary junction**	**Renal failure over 5-10 yrs**	**(LESS COMMON-WORST TYPE)**

			-Polydipsia (decreased tubular function) **IMPORTANT** - Positive family history+ UNEXPLAINED chronic renal failure in young patients → **suspicion of medullary cystic disease**			
Medullary cystic disease (medullary spongy kidney)		Small , give kidney spongy like appearance	- rare , non threatening condition - most pateint are asymptotic	Ecatic (dilatory)With in dilating collecting tubules of renal medulla		-Most common type - diagnosed based on incidental finding after radiological investigation for other reasons