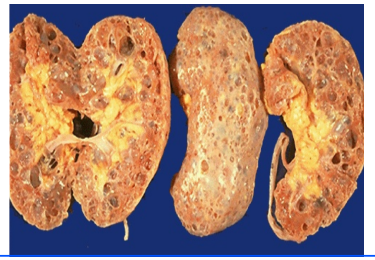
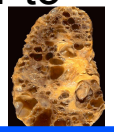
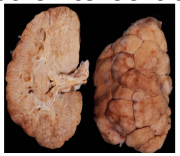
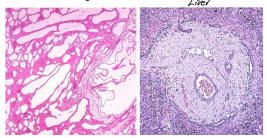
	Medullary sponge kidney	Autosomal dominant polycystic kidney disease	Autosomal recessive polycystic kidney disease	Acquired cystic kidney disease
	Sporadic cystic disease characterized by bilateral cystic dilations of medullary collecting ducts; normal cortex	↓intracellular calcium ↑CAMP Normal renal function is maintained until mid adulthood in most patients ↑Vasopressin (AVP or ADH)powerful modulator Cell proliferatio Fluid secretion	*limited urine output(oligohydramnios) *Potter sequence *joint deformities *pulmonary hypoplasia	Three or more cysts per kidney in patients on longstanding hemo- or peritoneal dialysis for end stage renal disease (unrelated to underlying renal pathology) .Also occurs in patients with long term uremia prior to dialysis
mutations	Associated with : *hemihypertrophy of body (25% of cases), *Marfan's, *Caroli's *Ehlers-Danlos syndrome	in genes coding for # polycystin 1 (PKD1, chromosome 16p, most common) Mutation accounts for-85% of ADPKD case #polycystin 2 (PDK2, chromosome 4q) 15%	Mutations in PKHDE1 gene (Polycystic Kidney and Hepatic Disease 1, produces fibrocystin / polyductin) at 6p12, expressed in kidney,pancreas and liver	
	Usually presents in adulthood Patients are usually asymptomatic with normal renal function Calcifications on X-ray, stones, hematuria and infection at age 30+ years Does not progress to end stage renal disease .Diagnosed with intravenous pyelography	*Third most common cause of end stage renal disease *hematuria, abdominal pain, hypertension, *urinary tract infection or Urolithiasis *Associated with Von Meyenburg Complexes in liver; hepatic cysts *berry aneurysms *mitral valve prolapse *cysts in pancreas, lung, spleen, pineal gland and seminal vesicles *aortic aneurysms; hepatic fibrosis and intestinal diverticula *die from infection *heart disease *stroke	Early mortality is most common, usually due to pulmonary complications *no cysts other than kidney and liver but liver is always affected *(every portal triad, every case) herring duct cysts (ductal plate malformation) congenital hepatic fibrosis *Patients later develop hypertension, renal insufficiency, portal hvpertension with splenomegalyor cholangitis	(Males > females during first ten years of dialysis Not restricted to adults; occurs in children and young adults on dialysis .Increased risk of renal cell carcinoma but death is rare 
Prognostic factors		Poor prognostic factors: sickle cell trait, male sex, early disease onset, .early hypertension onset and proteinuria		

Gross description	Normal sized kidneys with multiple, small cysts in medullary pyramids and papillae, giving medulla a sponge-like appearance Most often bilateral	Markedly enlarged kidneys (up to 8 kg) composed of sub-capsula cysts up to 4 cm Cysts contain clear to brown fluid . 	Markedly enlarged kidneys smooth surface Small cysts in cortex and medulla (collecting ducts) Dilated channels are perpendicular to cortical surface 	Moderately enlarged kidneys (usually < 800 g) with cortical and .medullary cysts containing clear fluid replacement of kidney with cysts 40% <
Microscopic (histologic) description	Medullary cysts lined by cuboidal epithelium or urothelium May have concretions adherent to cyst wall Often severe inflammation and scarring in interstitium, with tubular atrophy near papillary tips cysts involving the inner medullary and papillary regions . the cortex appears normal. It is often bilateral occurs sporadically recurrent hematuria urinary tract infection Renal failure is unlikely to occur, but may result from severe pyelonephritis.	*Cysts are lined by cuboidal or flattened epith . *papillary projections or polyps *Functional nephrons exist between cysts: 1)global sclerosis 2)tubular atrophy 3)interstitial fibrosis 4)chronic inflammation *Infants may show primarily cystic dilatation of Bowman's space *have renal adenomas 20%	*Radially arranged elongated cysts that form as dilations of all collecting tubules *fluid accumulation *Cysts lined by cuboidal or flattened cells from collecting tubules *Normal nephrons without cystic change / interstitial fibrosis are present in between the cysts *The liver shows portal fibrosis with complex bile ductular profiles 	*Cysts lined by flattened or cuboidal epithelium that may show focal *pseudo-papillae with nuclear enlargement and loss of polarity (Cysts may contain oxalate crystals scattered small cysts, none of which is over 2 cm in size. *Surrounding parenchyma shows global glomerulosclerosis, interstitial fibrosis and tubular atrophy *signe oF renal Fainler
				risk of renal cell carcinoma