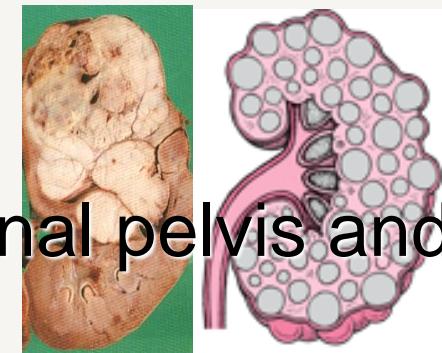


# Cystic diseases of kidney & Tumours of kidney and bladder

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At the end of this lecture the students should be able to

- Classify the types of cysts in kidney
- Describe the pathogenesis and pathology of autosomal dominant and autosomal recessive polycystic kidney disease
- Describe the epidemiology, pathogenesis, pathology and prognosis of following tumours
  - Wilm tumour
  - Renal cell carcinoma
  - Transitional cell carcinoma of renal pelvis and bladder



# Cystic diseases of kidney

- A heterogeneous group of diseases
  - Congenital
  - Developmental
  - Acquired

# Cystic diseases of kidney-Importance and relevance

- Some are clinically silent. Important in the differential diagnosis of a renal lesion
  - Clinicians
  - Radiologists
  - Pathologists
- Some are important cause of chronic renal disease
- Accurate diagnosis and appropriate counseling is necessary in genetically transmitted diseases

# Cysts in kidney-classification

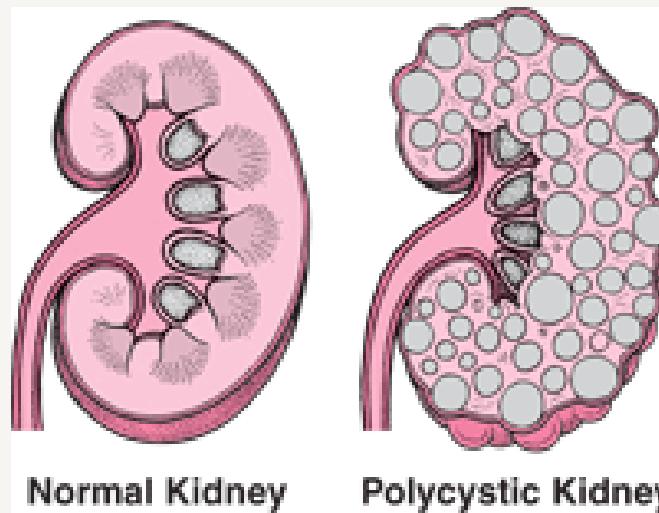
- Autosomal dominant polycystic kidney disease
- Autosomal recessive polycystic kidney disease
- Medullary cystic kidney
- Simple renal cysts
- Renal dialysis associated cysts

# Autosomal dominant polycystic kidney disease [ADPKD]

- An important cause of end stage renal disease in adults
- Inherited as an autosomal dominant trait with high degree of penetrance
- Mutation in PKD1, PKD2 and PKD3 genes
- Always bilateral
- Increased frequency of cysts in kidneys, liver, pancreas and spleen
- Berry aneurysms in cerebral arteries- subarachnoid haemorrhage

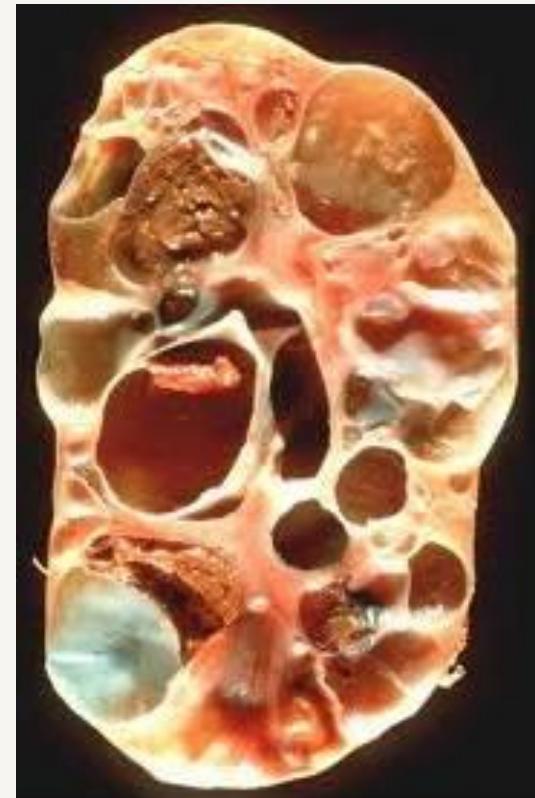
# ADPKD

- Can present at any age from childhood to late adult life
- Maintain the renal function till late disease
- Cysts in both cortex and medulla
- Cysts compress the adjacent parenchyma, causing ischemia and subsequent hypertension



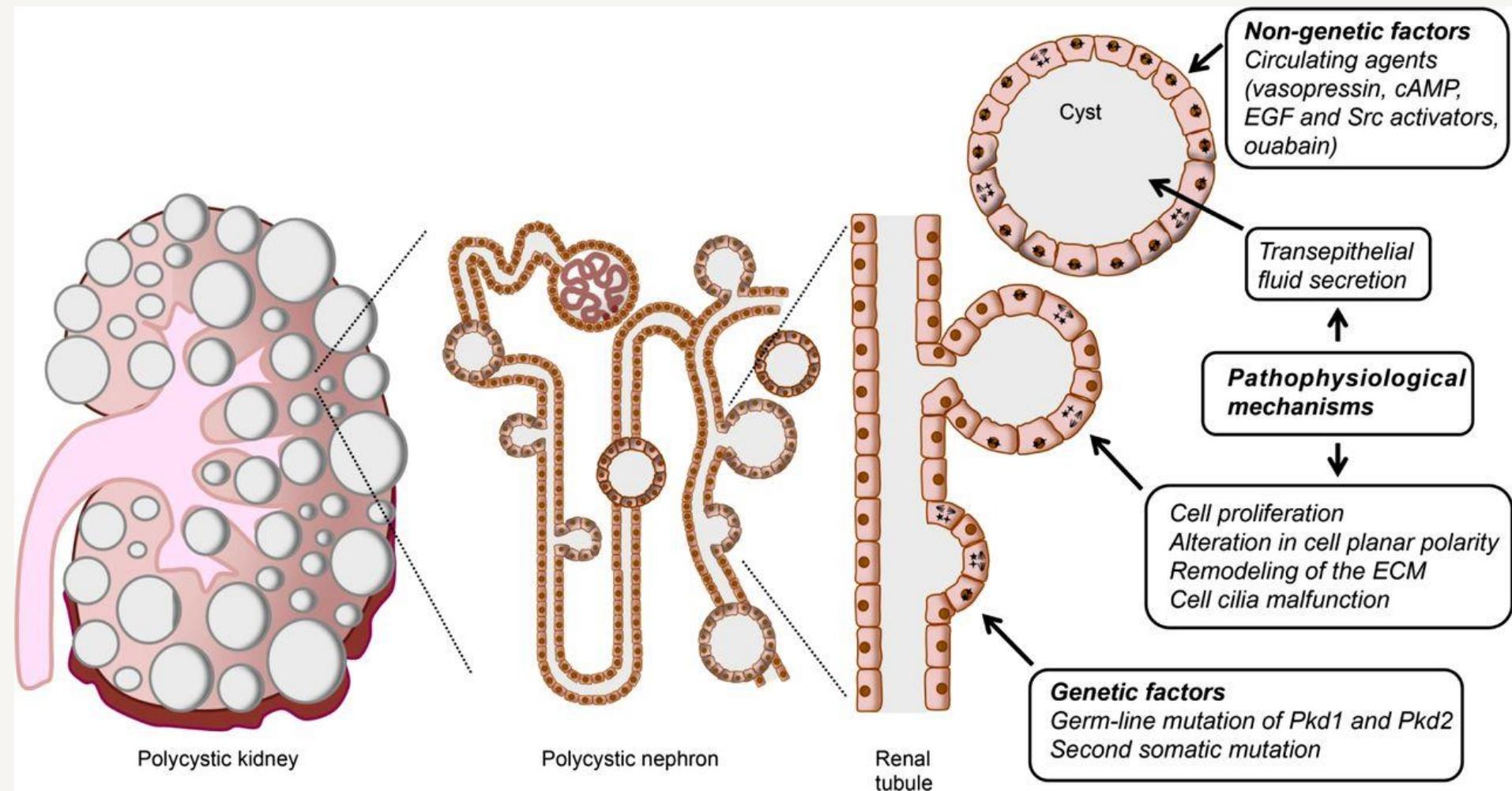
# APCKD-macroscopy

- Both kidneys are grossly enlarged [>1kg]
- Distorted by many cysts -few mm-10cm
- Thin bands of renal parenchyma in between the cysts
- The cysts are filled with
  - Clear fluid
  - blood



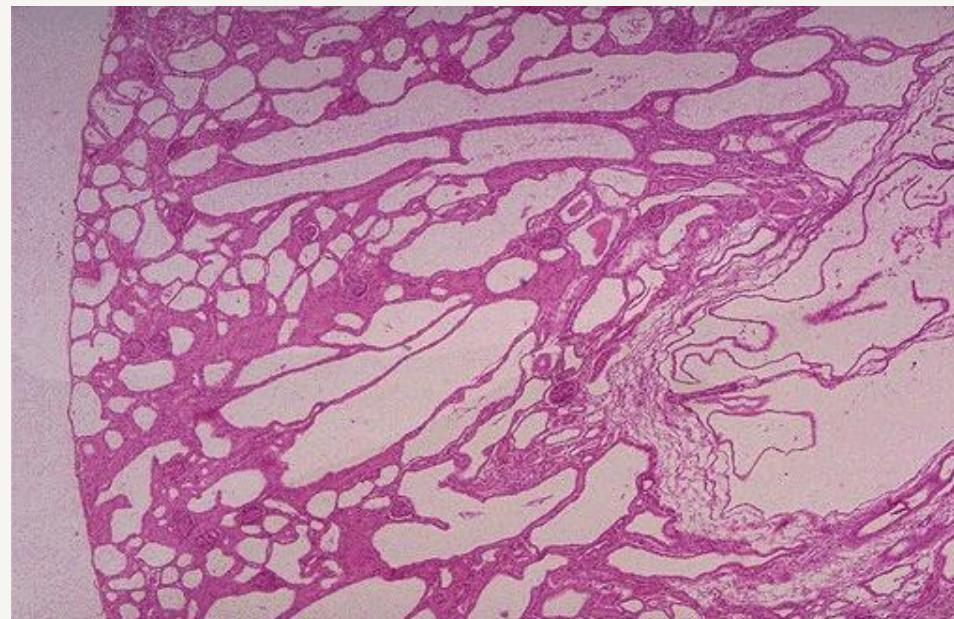
# APCKD-pathogenesis

- The cysts appear to arise from any point along the nephron



# APCKD-microscopy

- Cysts lined by columnar to cuboidal epithelium
- Normal renal parenchyma is found in between the cysts

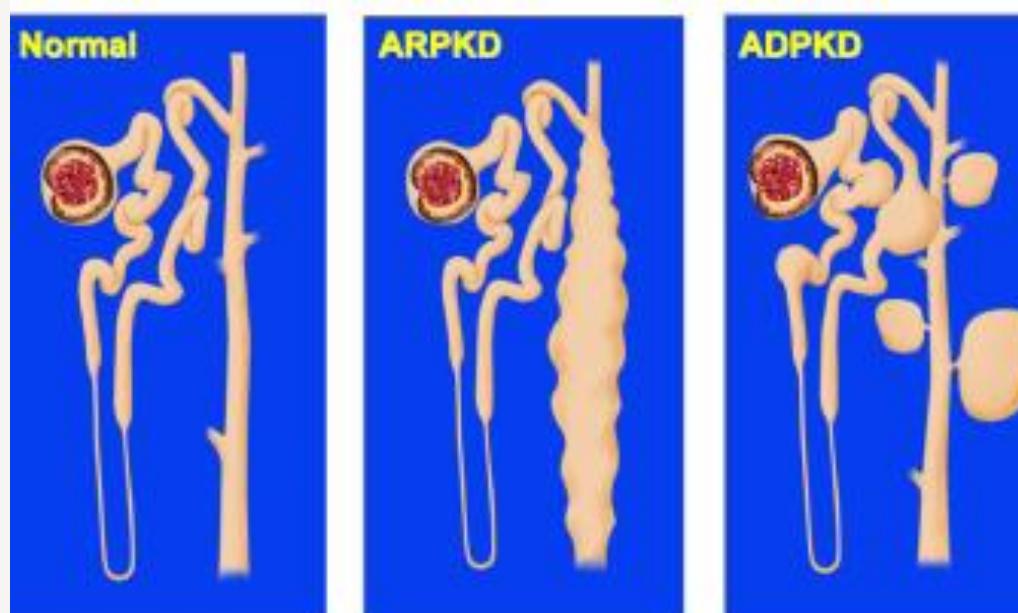


# Autosomal recessive polycystic kidney [ARPKD]

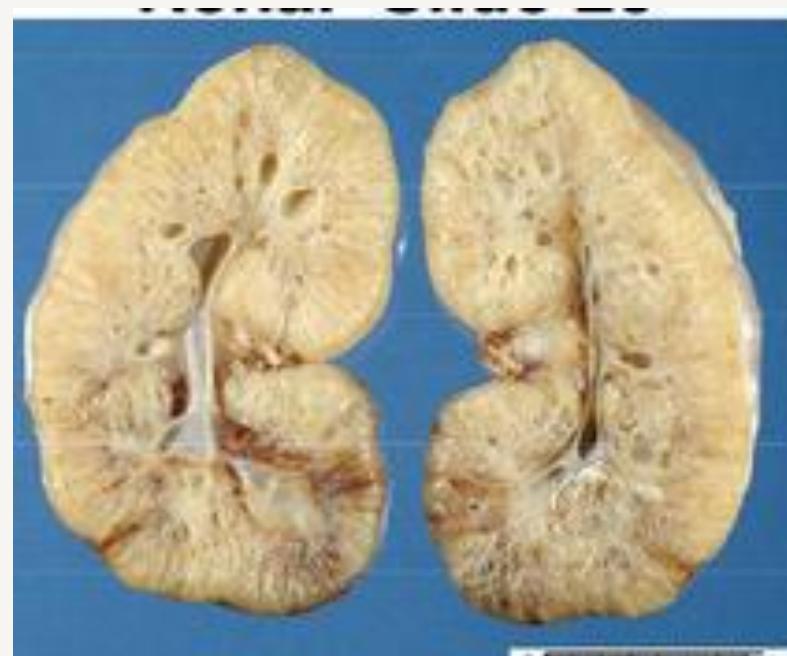
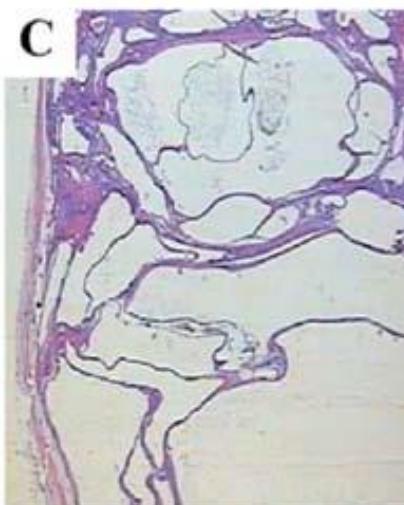
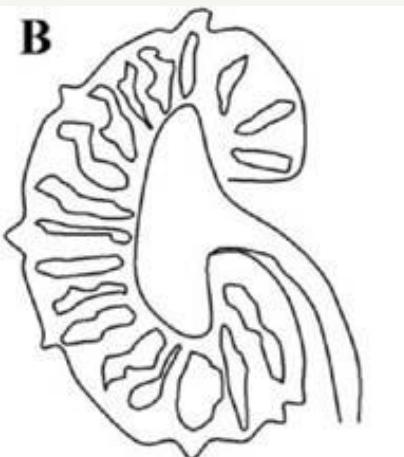
- Autosomal recessive inheritance
- Perinatal presentation
  - Severe form
  - 90% nephrons are involved
  - Still birth or death soon after birth
- Neonatal, infantile presentation
  - Less severe and long survival
- Associated with congenital hepatic fibrosis and bile duct hamartomas

# ARPKD morphology

- The external surface of the kidney is smooth
- The cut surface shows radially oriented fusiform cysts in the cortex & medulla
- The cysts are lined by cuboidal epithelium
- Cysts are derived of collecting duct epithelium



# ARPKD morphology



Compare clinical presentation,  
pathogenesis, pathology, outcome  
and complications of ADPKD and  
ARPKD

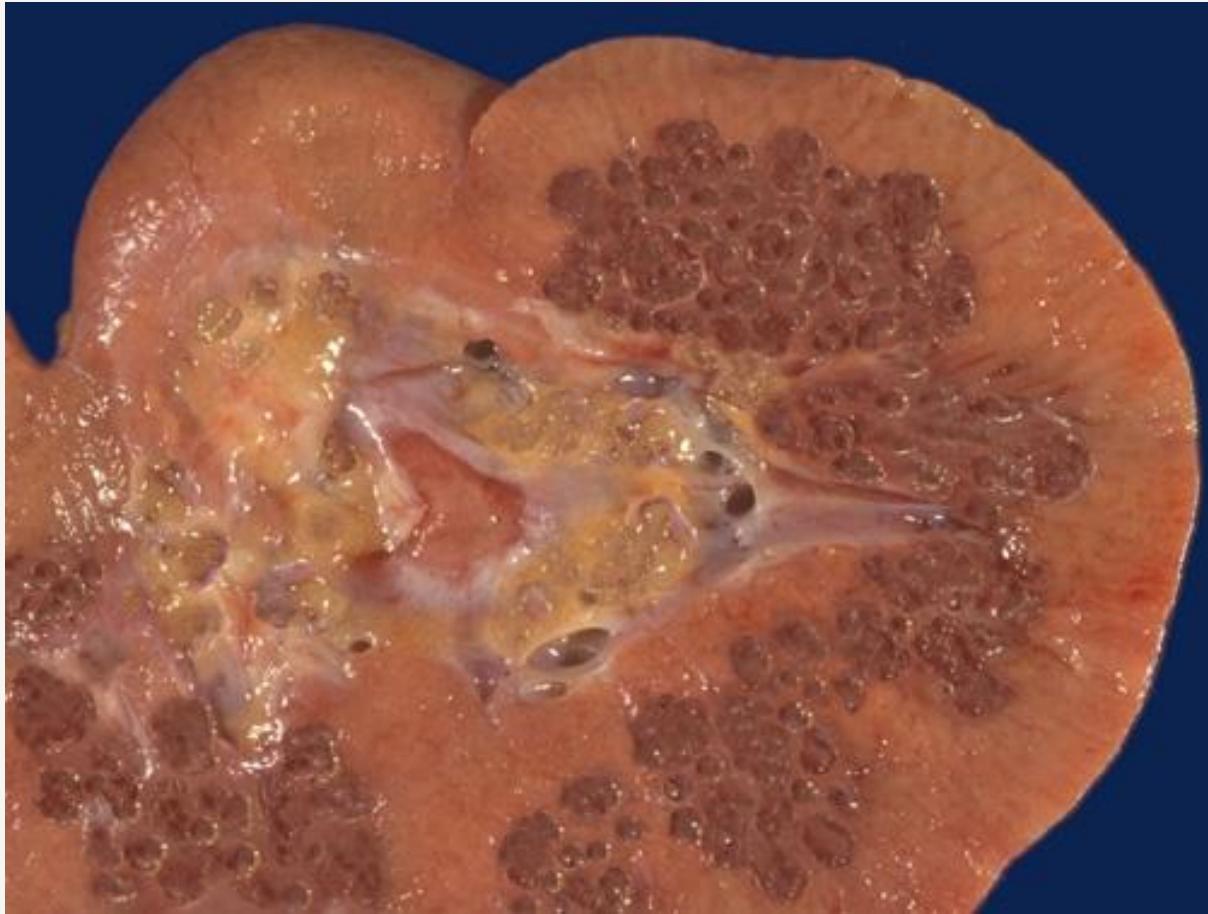
# Medullary cystic disease

- Medullary sponge kidney
- Medullary cystic disease complex-nephronophthisis / uraemic medullary cystic disease

# Medullary sponge kidney

- Small cysts (<5mm) in renal papillae
- Most patients 30-60 years
- The kidney may be enlarged ,normal or small
- The cysts are lined by cuboidal or transitional epithelium
- Pyelonephritis is a complication due to calculi formation

# Medullary sponge kidney



Cysts involving the inner medulla and papillary regions. The cortex appears normal.

# Medullary cystic disease complex- Nephronophthisis

- Accounts for 20-25% of CKD in children
- Familial- AD and AR inheritance
- Kidneys are contracted and granular
  - cysts at cortico medullary junction
  - Interstitial fibrosis

# Simple renal cysts

- Acquired lesions
- Common in older people over 50 years
- Usually solitary but may be multiple
- Cyst located in the cortex
- The cyst is lined by flattened epithelium
- Majority are clinically silent
  - Found at autopsy
  - Detected at ultrasonography
- Do not affect renal function
- Sometimes bleeding into cyst causes pain.

# Simple renal cysts



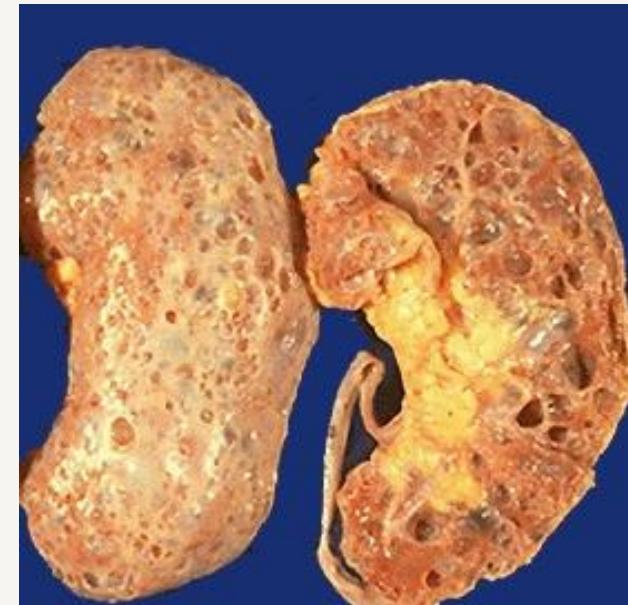
solitary



multiple

# Dialysis associated cysts

- Seen in CKD who have received dialysis therapy
- Multiple small cysts throughout the cortex and medulla
- Cysts arise due to obstruction of the tubules
  - by fibrosis
  - By oxalate crystals
- The size of the kidneys is usually not markedly increased



# Tumours of kidney

- Benign
- Malignant
- Malignant tumours are mostly primary
- Metastatic deposits also occurs , but are less frequent than would be expected in view of the high renal blood supply

# Childhood renal tumours

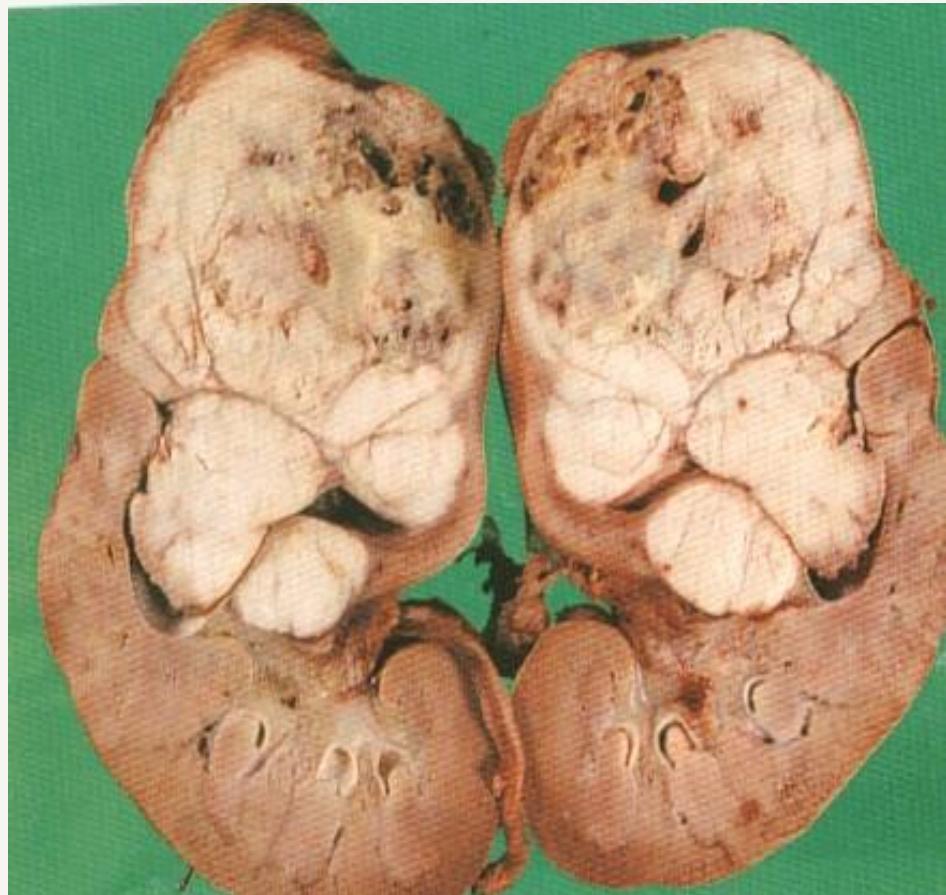
- Wilm tumour /Nephroblastoma
- Mesoblastic nephroma
- Multicystic nephroma
- Clear cell sarcoma
- Rhabdoid tumour

# Wilm tumour

- Commonest malignant renal tumour of childhood
- 50% of the cases occur before the age of 3
- 90 % of cases occur before the age of 10
- Bilateral in 10 %
- The commonest clinical manifestation is an abdominal mass
- Haematuria is uncommon
- Due to inactivation of WT1 onco suppressor gene

# Wilm tumour macroscopy

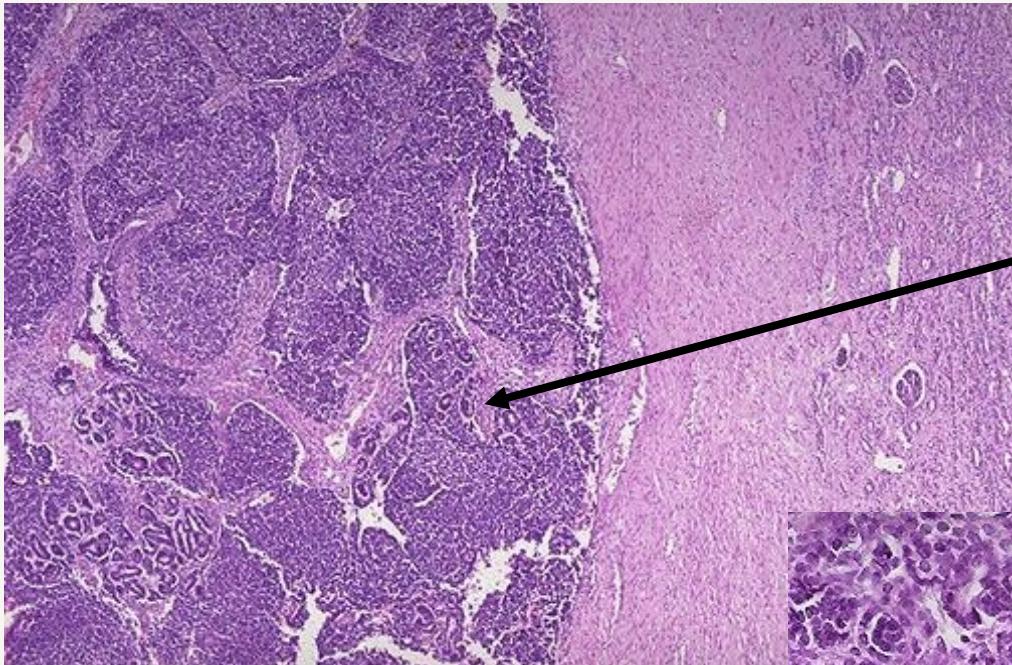
- Large well circumscribed tumours
- Firm in consistency
- Cut surface is solid and may show areas of necrosis and haemorrhage.
- Frequently extends beyond the renal capsule



# Wilm tumour microscopy

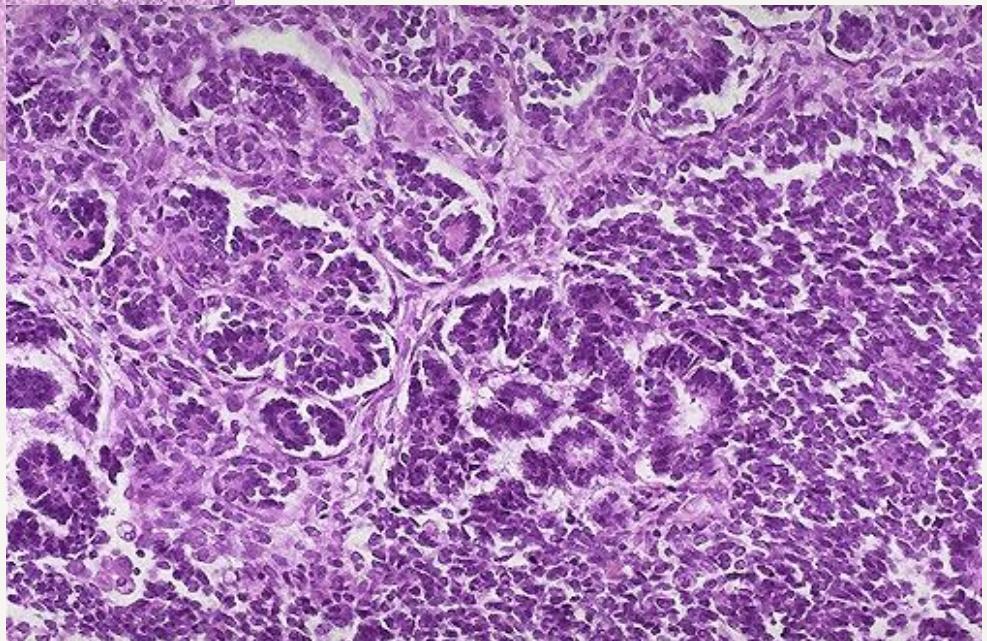
- Wilms tumor resembles the developing fetal nephrogenic zone of the kidney
- A blastemal tumour originating from mesonephric mesoderm.
- Major components are
  - **Undifferentiated blastema** –small oval/round cells
  - **Mesenchymal tissue** –smooth muscle ,skeletal muscle, cartilage
  - **epithelial tissue** –form tubular & glomeruloid structures
  - “a bizarre mixture”

# Wilm tumour microscopy



Undifferentiated  
blastema

Primitive glomerular and  
tubular structures.



# Wilm tumour -prognosis

- Aggressive and rapidly growing tumour.
- Pulmonary metastasis are seen in a high proportion of patients at the time of diagnosis.
- Aggressive therapy including surgery, chemotherapy and radiotherapy improve the prognosis.
- Unilateral tumours have a cure rate of 80%

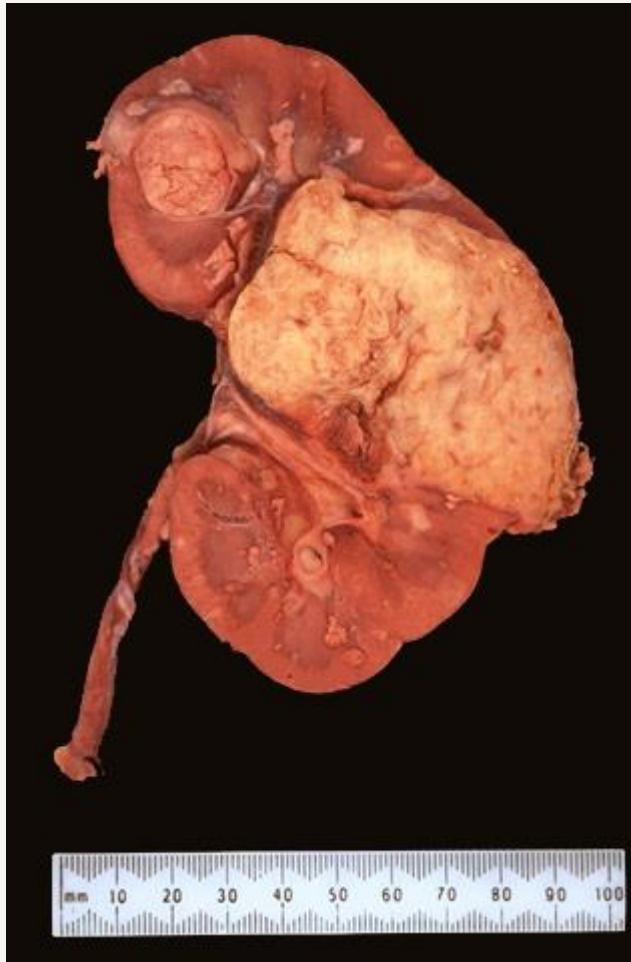
# Renal tumours of adults

- Benign
- Malignant

# Benign tumors of kidney

- Renal papillary adenoma}
- Renal fibroma } Asymptomatic
  
- Oncocytoma
- Angiomyolipoma

# Renal angiomyolipoma



- A hamartomatous lesion
- Associated with tuberous sclerosis
- Can mimic a malignancy

# Malignant tumors of kidney

- Renal cell carcinoma
- Transitional cell carcinoma of the renal pelvis

# Renal cell carcinoma

- Other terms- hypernephroma / adenocarcinoma of kidney
- Is the **commonest** renal cancer in adults.
- 1-3 % of all visceral tumours.
- Occurs most frequently over the age of 50.
- Male preponderance.
- Predisposing factors
  - Tobacco smoking
  - Genetic predisposition –Von Hippel Lindau disease

# Renal cell carcinoma –clinical presentation

- Haematuria
- Loin pain
- Loin mass
- Paraneoplastic syndrome
  - Hypercalcaemia
  - Hypertension
  - Polycythaemia
  - Cushing's syndrome
- Weight loss , anaemia, fever
- Amyloidosis
- Metastatic disease- lung & bone

Explain the pathophysiological basis of these symptoms/signs

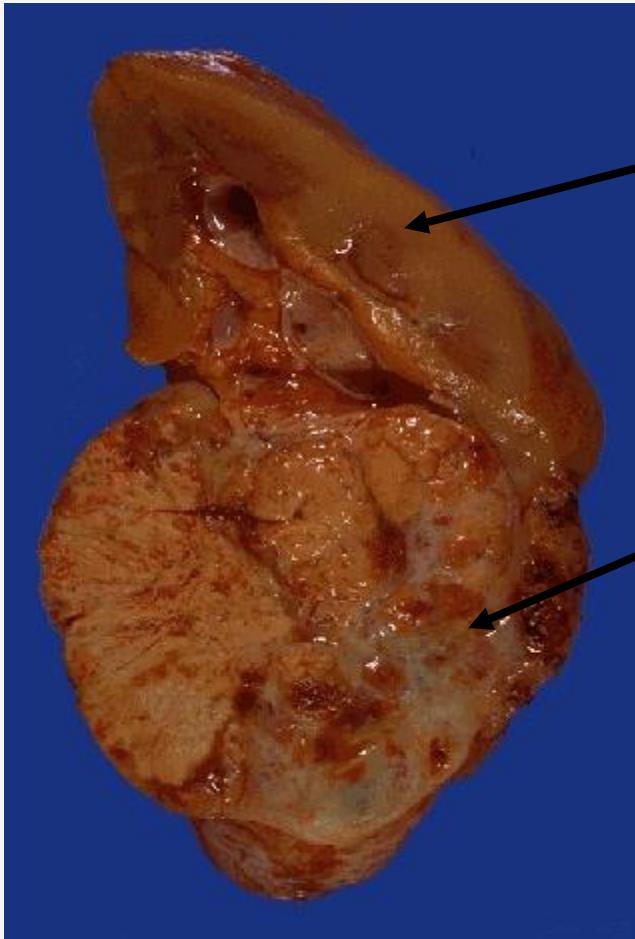
# Renal cell carcinoma -macroscopy

- May arise in any part of the kidney but the commonest site is upper pole.
- Unilateral solitary lesions
- Rarely(<1%) bilateral
- Large bosselated tumours destructing the kidney
- The margin of the tumour is well demarcated but can breach renal capsule and enter the perinephric fat.

# Renal cell carcinoma -macroscopy

- In the cut section the tumour has a variegated appearance.
  - Solid yellowish - grey tumour
  - Haemorrhagic areas
  - Necrotic areas
  - Cystic areas
- Widely infiltrating tumour
  - Extension into renal vein , IVC and sometimes into right atrium
  - Fungating growth into renal pelvis

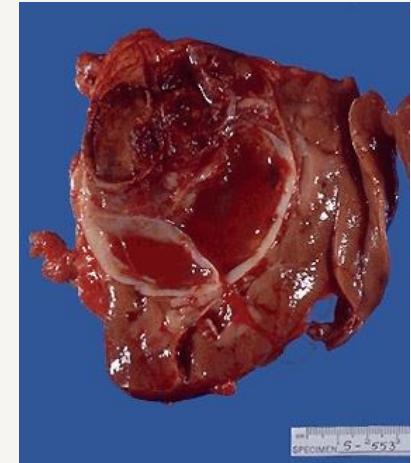
# Renal cell carcinoma -macroscopy



Renal tissue

- Variegated appearance
- Widely infiltrating.

Tumour with cystic appearance

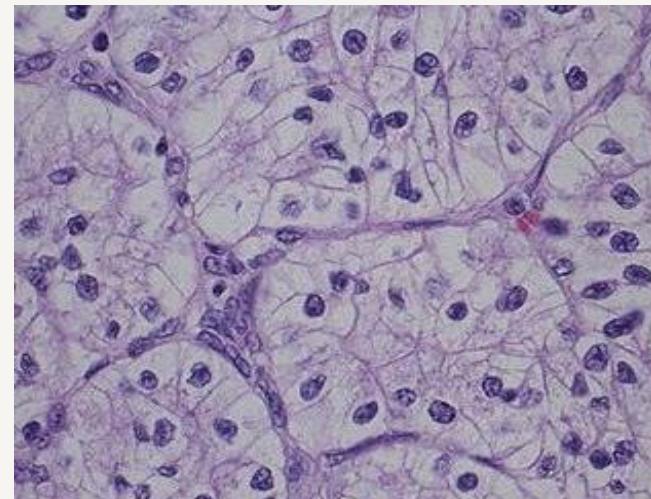
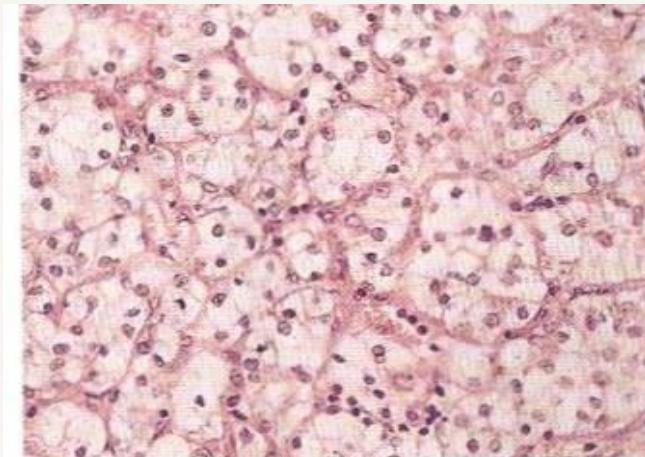


# Renal cell carcinoma -microscopy

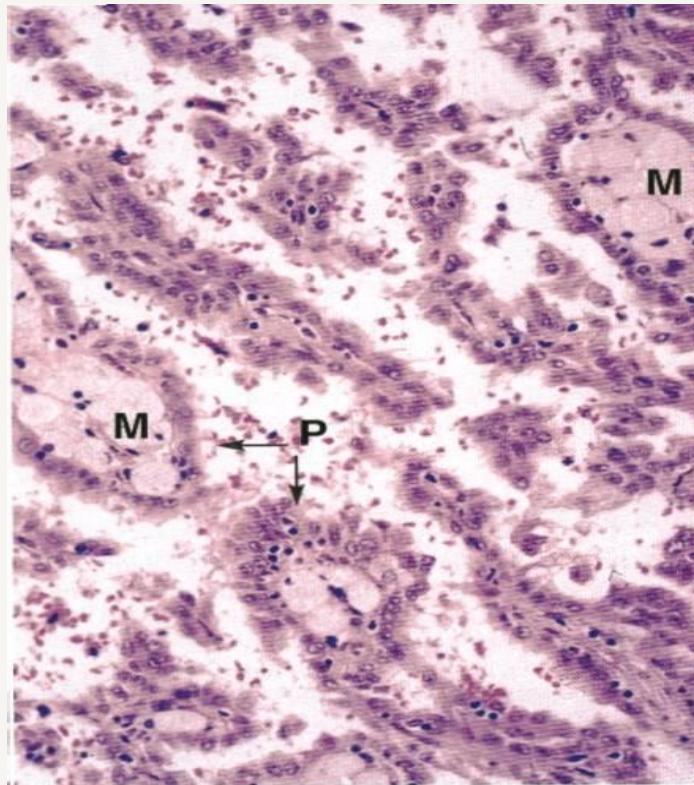
- It is an adenocarcinoma arising from the renal tubules.
- 4 main histological patterns
  - Clear cell carcinoma –commonest type
  - Papillary renal cell carcinoma
  - Chromophobe carcinoma
  - Collecting duct carcinoma –rare variant

# Clear cell carcinoma

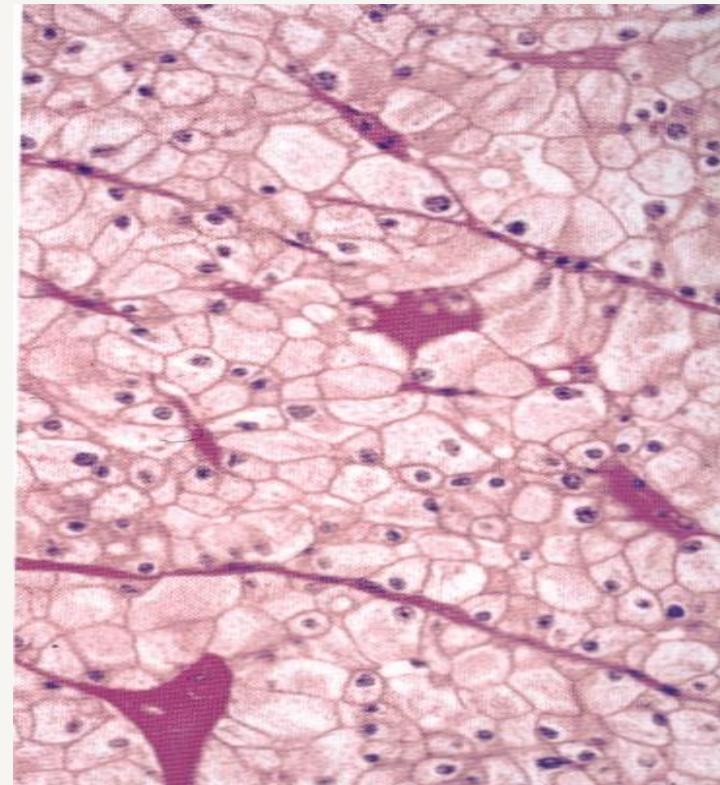
- Solid growth with nests of tumour cells separated by fibro vascular septae
- Tumour cells are large and polygonal
- Centrally placed round nuclei
- Clear cytoplasm due to accumulation of glycogen and fat.



# Renal cell carcinoma other microscopic types



Papillary renal cell carcinoma



Chromophobe renal cell carcinoma

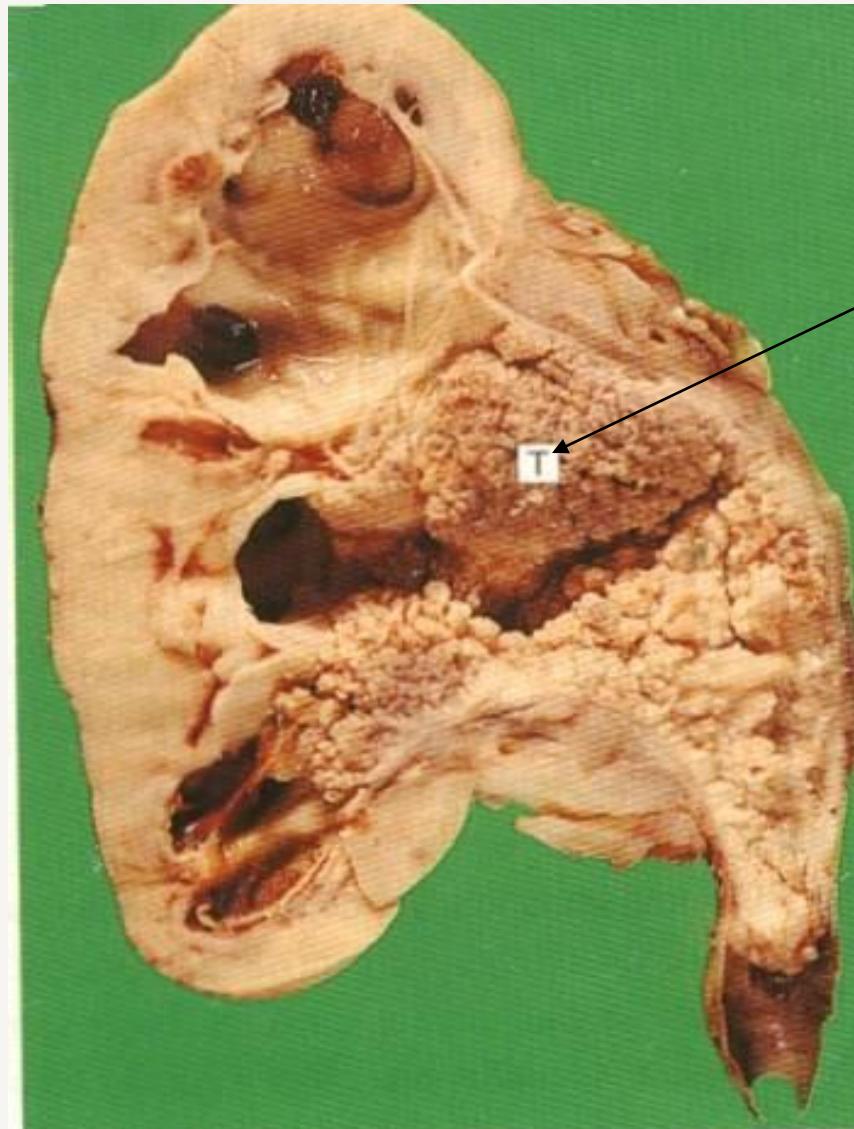
# Renal cell carcinoma -prognosis

- Prognosis depends on clinical staging and microscopic grading-
  - Fuhrman nuclear grading 1-4
- 5 year survival rate is about 70 % in the absence of distant metastases.
- Invade perinephric fat
- Metastatize to
  - regional lymphnodes
  - Lung
  - bone
- Renal vein invasion dramatically reduces the

# Urothelial carcinoma / transitional cell carcinoma

- Forms 5-10% of primary renal tumours
- Originates from the urothelium of renal pelvis.
- Grows as a fungating tumour in renal pelvis obstructing the upper urinary tract.
- Therefore becomes clinically apparent early.
  - Haematuria
  - Hydronephrosis
  - Loin pain
- Microscopically resembles urothelial carcinoma of lower urinary tract.

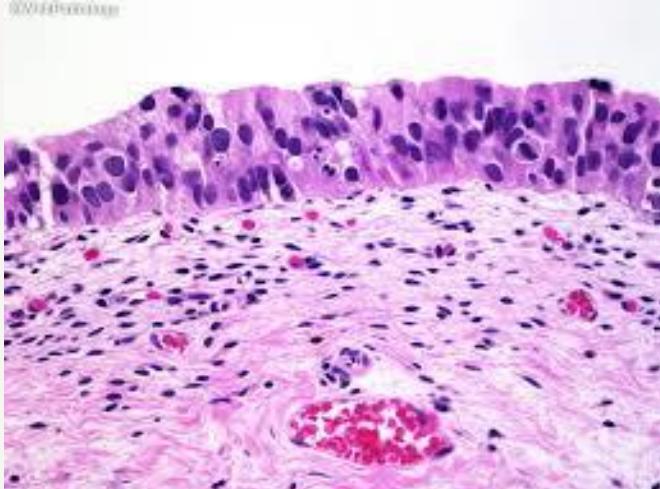
# Transitional cell carcinoma of renal pelvis



Tumour in the  
pelvis

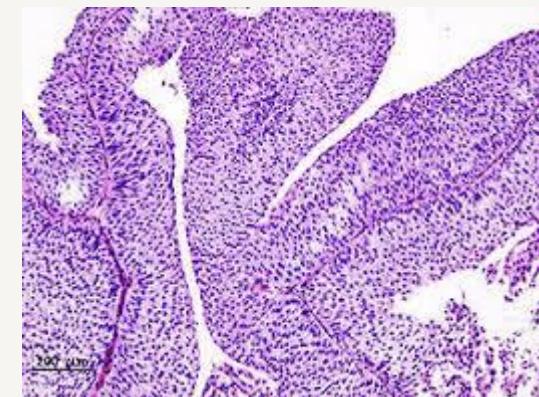
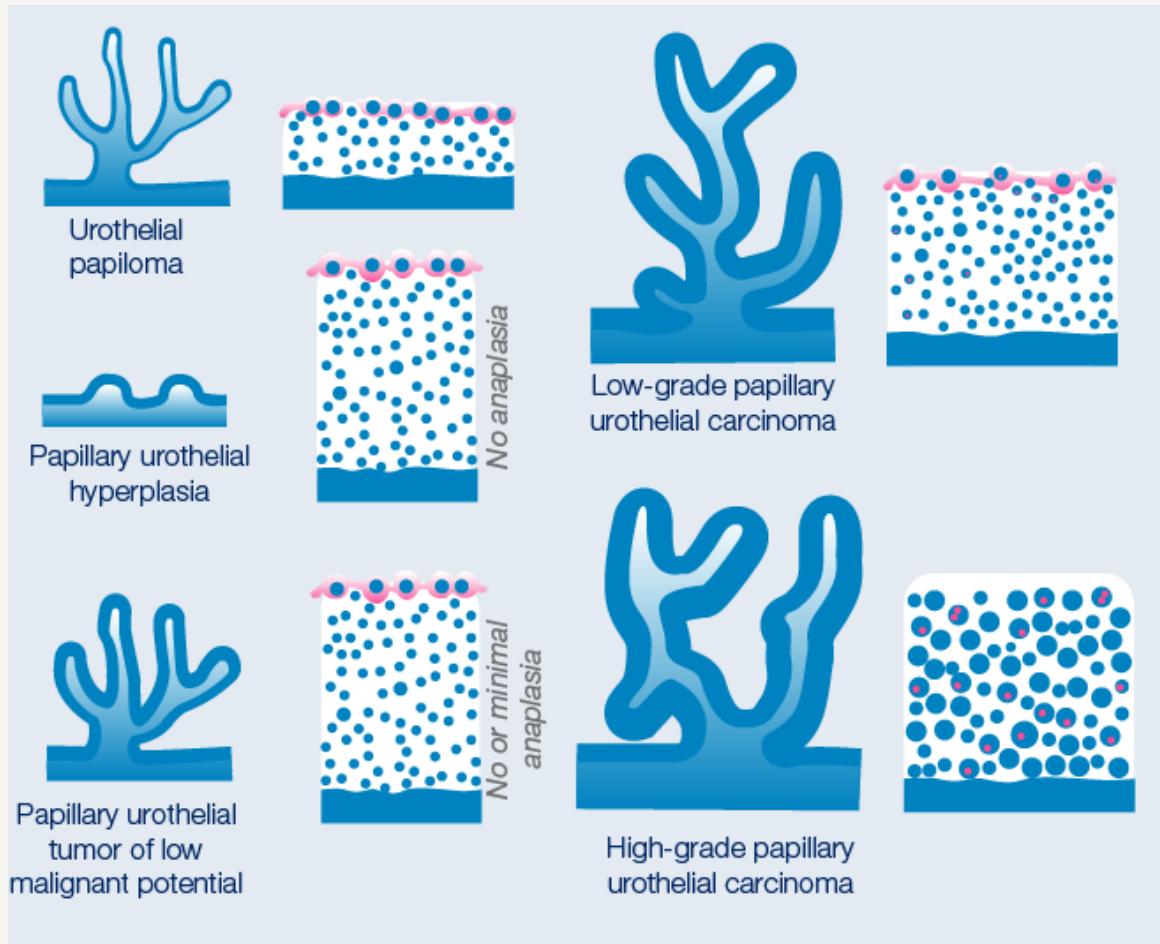
# Tumours of Bladder

- Majority of the bladder tumours arise from the transitional cell epithelium
  - Flat lesions with atypia
  - Papillary neoplasms
  - Invasive urothelial carcinomas
- Flat lesions with atypia
  - Urothelial dysplasias and carcinoma in situ



# Tumours of Bladder

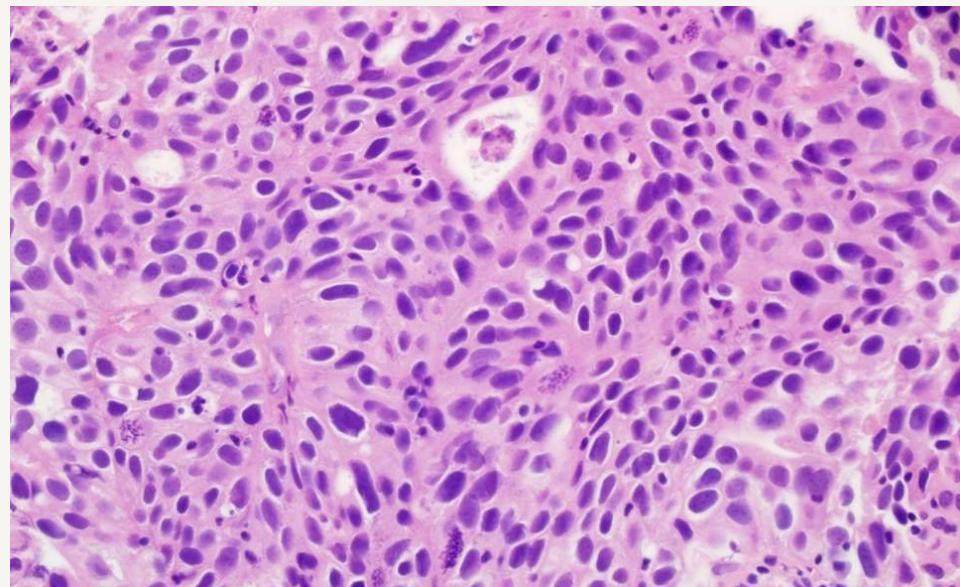
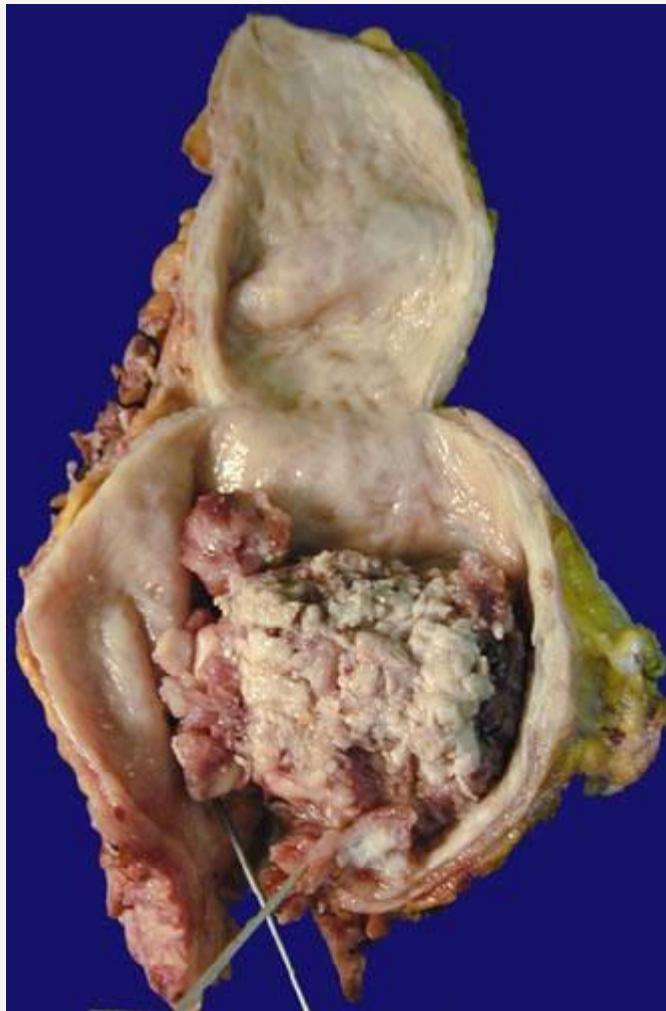
- Papillary urothelial neoplasms
  - Several categories with low to high grade cellular atypia



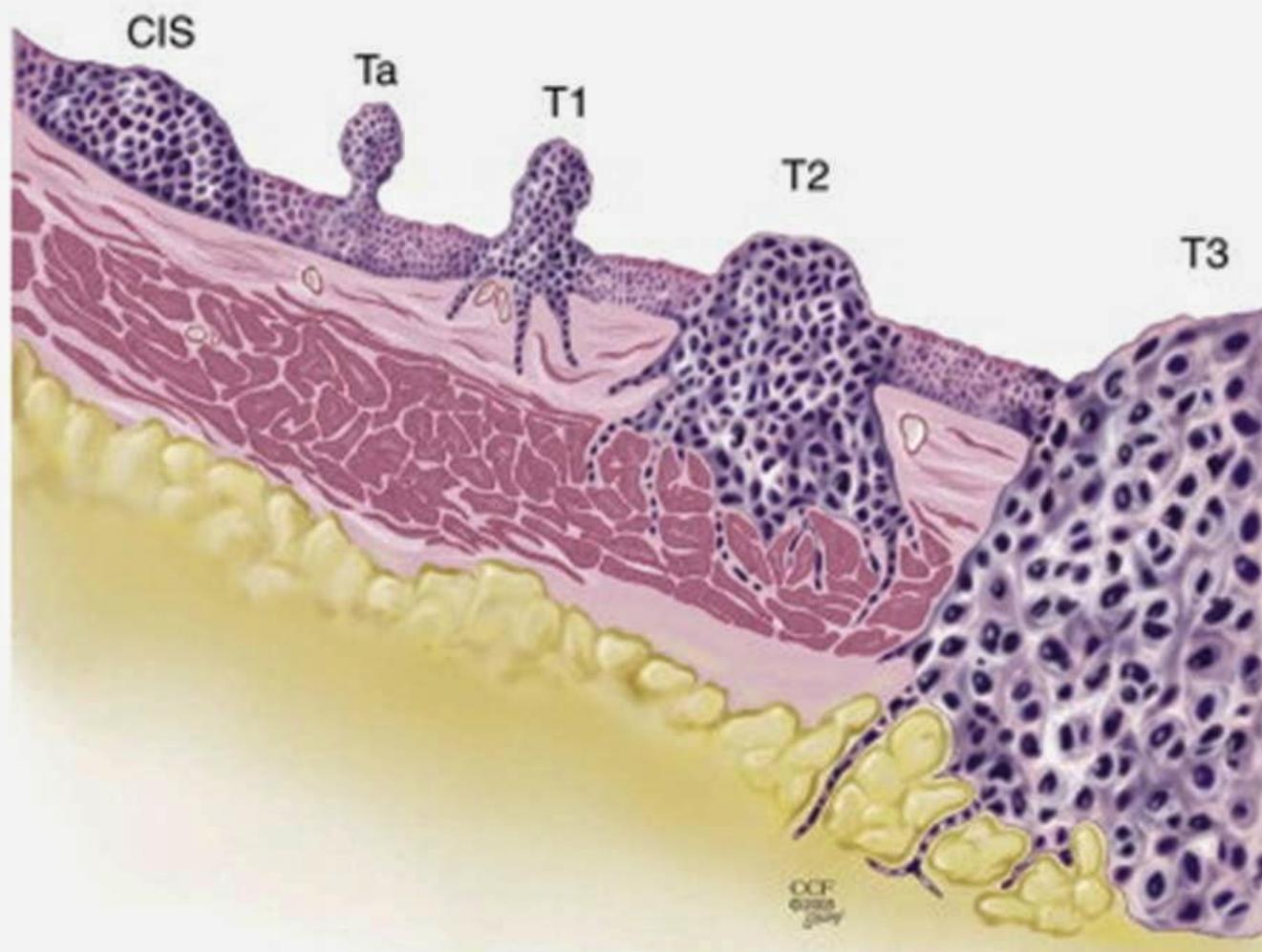
# Invasive urothelial carcinomas

- Elderly age group 50-80yrs
- Papillary ,polypoidal, nodular , ulcerative patterns
- Histological types
  - Majority –transitional cell carcinomas
  - Other-squamous cell carcinoma, adenocarcinoma

# Carcinoma of bladder



# Depth of invasion of bladder cancer



# Carcinoma of bladder- aetiological factors

- Cigarette smoking
- Industrial exposure- arylamines
- Parasitic- schistosoma haematobium
- Long term exposure to
  - Analgesics
  - Cyclophosphamides
- Prior irradiation

# Carcinoma of bladder

- What are the different clinical manifestations of bladder carcinoma?
- How do you investigate a patient with bladder carcinoma?