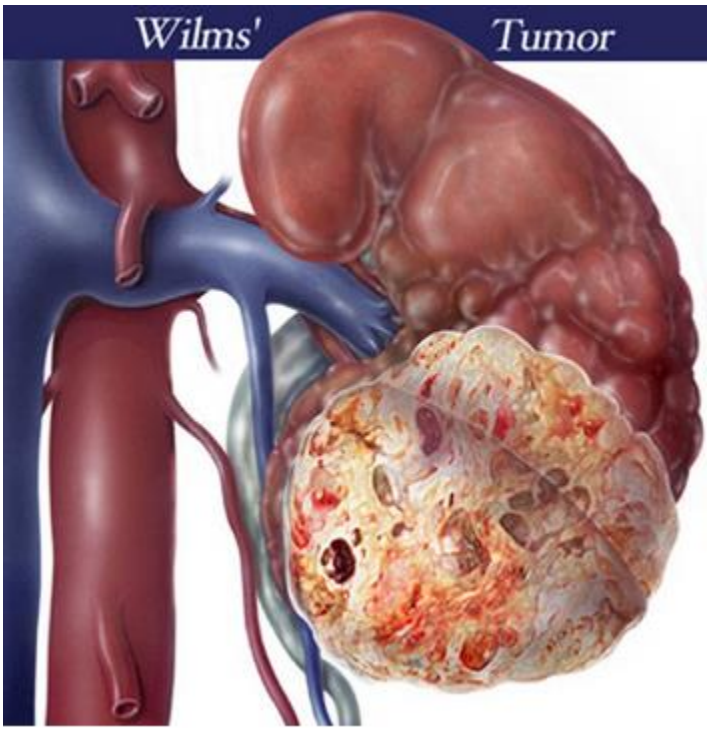
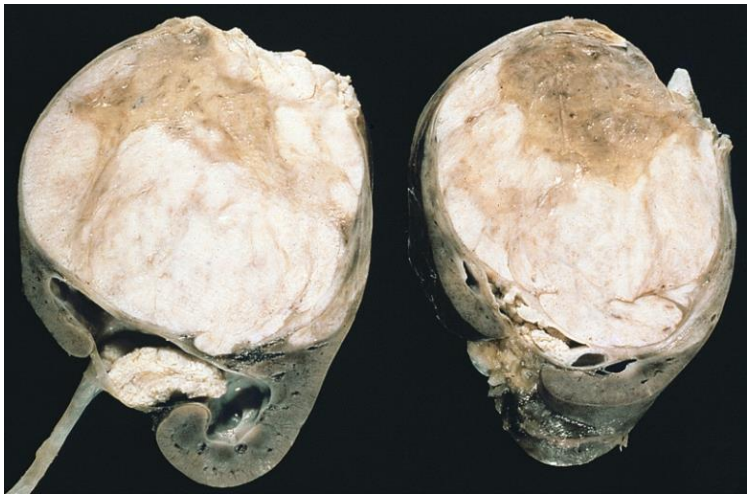
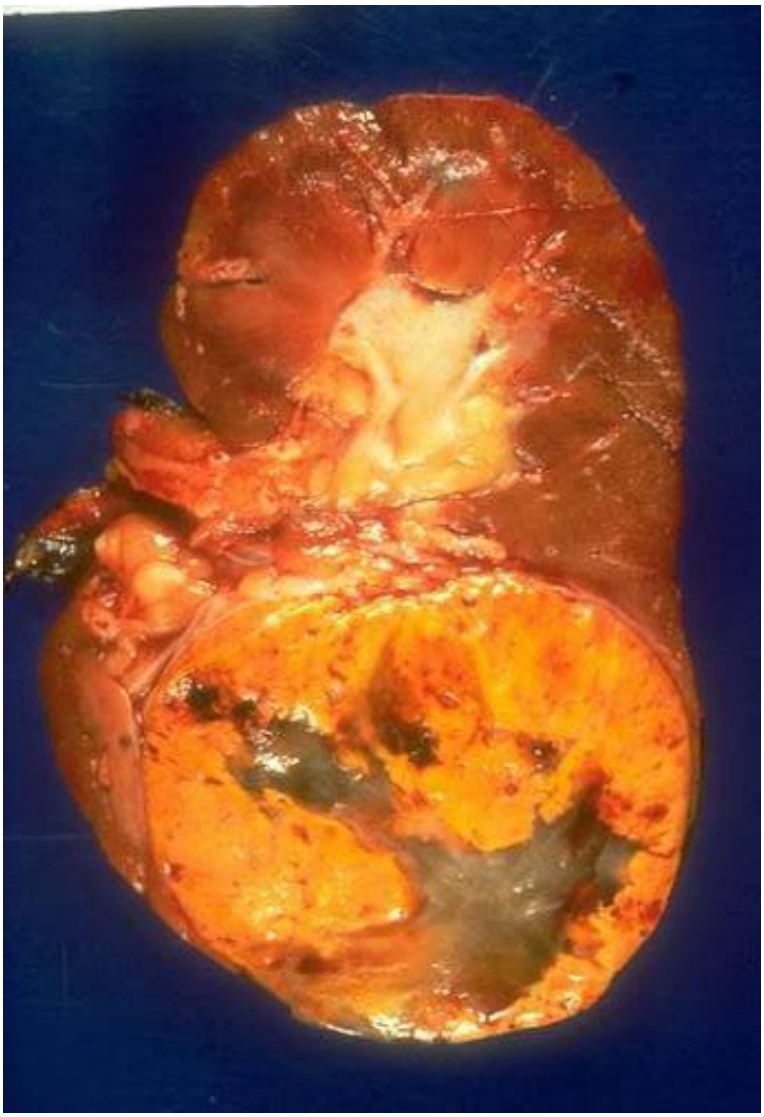
	I- Wilm's tumour (Embryoma or Nephroblastoma)	II- Renal cell carcinoma (Hypernephroma)
★ **Incidence :**	1- 10% of childhood malignancy 2- Usually below 4 years. 3- More in males. 4- May be bilateral in 10% .	1- 80% of renal tumours. 2- Usually above 40 years. 3- More in males. 4- Bilateral (2%)
★ **Predisposing Factors :**		1-Use of **tobacco products** . 2- Defect in chromosome 3 3-**Von Hipple-Lindau disease** (renal & pancreatic cyst , cerebellar haemangio-blastoma , retinal angioma & pheochromocytoma)
★ **Pathology :**		
1- Origin:	▪ Arise from embryonic nephrogenic multipotent mesodermal cells.	▪ From the epithelium of the proximal convoluted tubules.
2- Site:	▪ In one pole of the kidney, usually the upper pole.	
3-Gross picture	▪ Large smooth mass infiltrates & destroys the renal parenchyma → early loss of renal function ▪ The pelvicatyceal system resists and becomes stretched and compressed → haematuria is late.	▪ A mass variable in size. Compresses the renal parenchyma →late loss of renal functions ▪ Early infiltration of the pelvicalyceal system and early haematuria.





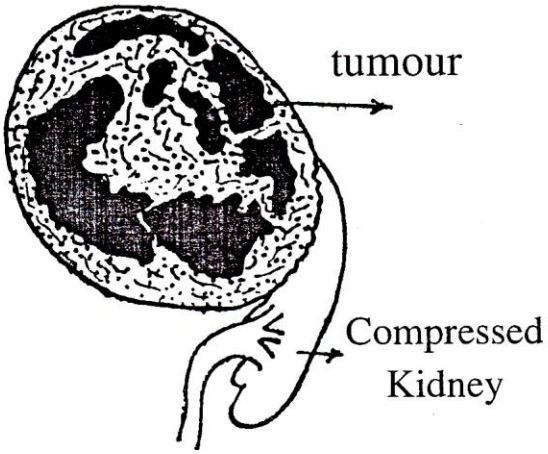
Renal cell carcinoma



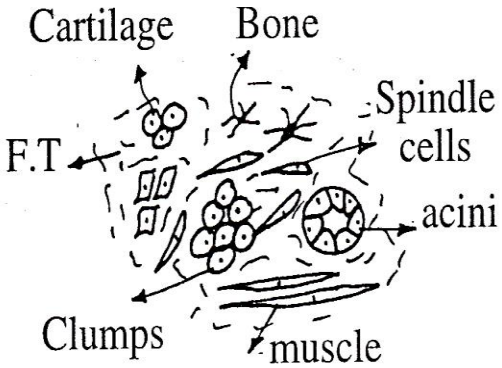
* Wilm's Tumour *

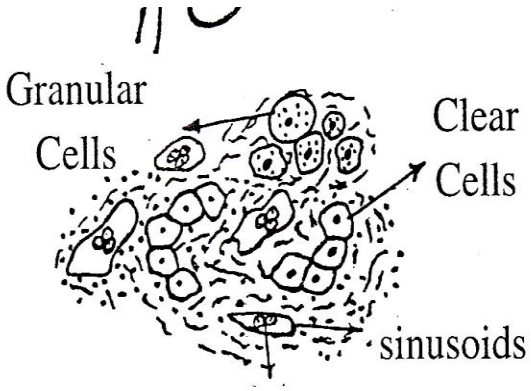
Cartilage Bone

* Renal Cell Carcinoma *

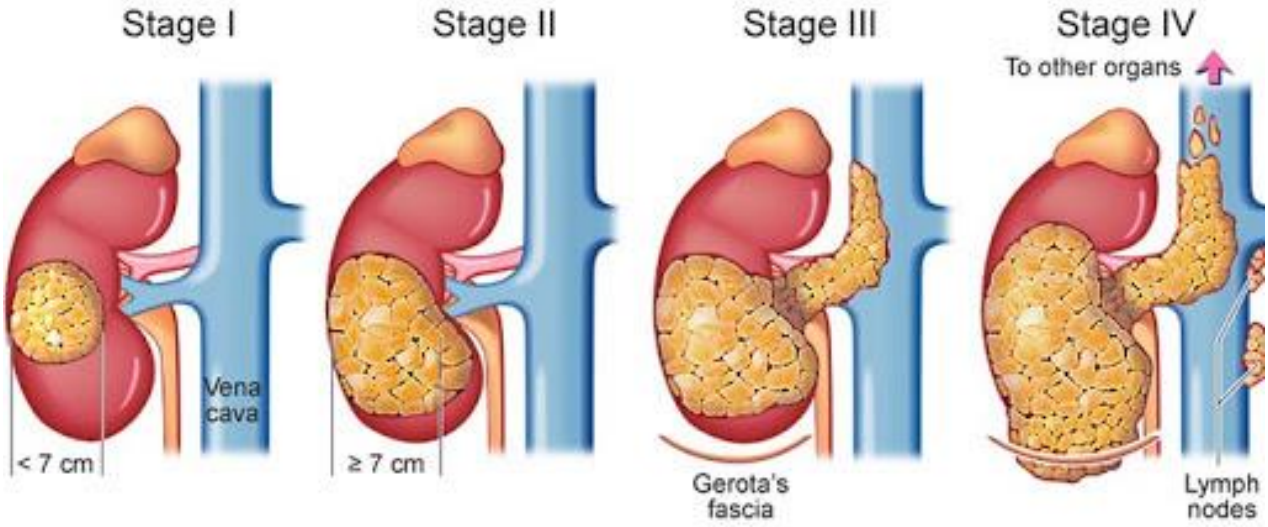


4- Cut section:	▪ Circumscribed pseudo-capsulated greyish tumour with areas of hge. , necrosis & degeneration.	▪ Circumscribed pseudo-capsulated (compresed normal tissues) tumour which is golden yellow (high lipid content), with areas of hge & necrosis. ▪ The tumour is divided into many lobules by fibrous septa.
5- Microscopically	▪ Mixed epithelial & C.T. tumour consists of clumps of cells, acini, fibroblasts, fibrous tissue, muscle fibres, cartilage , spindle and rounded cells. ▪ The cells have variable degree of differentiation .	▪ Adenocarcinoma in which the malignant cells are arranged in irregular or solid acini. ▪ The cells are large, polyhydral or cubical & vacuolated (due to high lipid & glycogen contents) ▪ Granular cells (increased mitochondria in cytoplasm) ▪ The blood vessels are malformed & infiltrated with malignant cells.

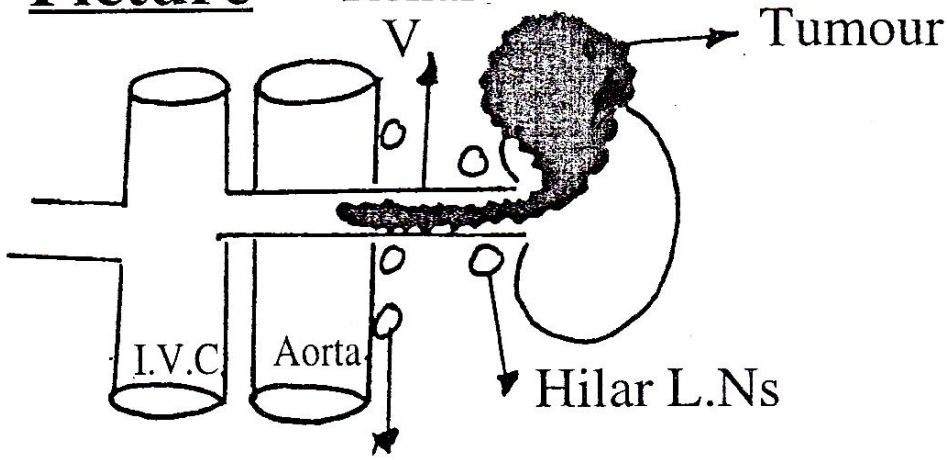




* Microscopic *



Picture



* Spread *

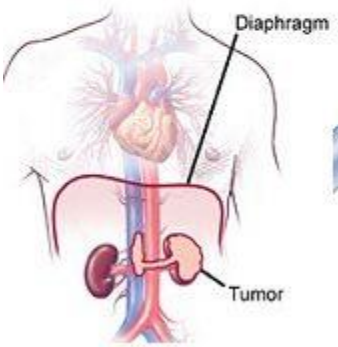
6- Staging:	I: Limited to kidney, completely excised II: Extending outside kidney, completely excised III: Extending outside kidney, incompletely excised IV: Distant metastases V: Bilateral renal tumors	▪ **T : 1ry tumor** ➢ T1:tumor less than 7cm , limited to the kidney. ➢ T2: tumor more than 7cm , limited to the kidney. ➢ T3: spread to major veins , adrenal gland or perinephric tissues . ➢ T4:Spread beyond Gerota's fascia . ▪ **N: Regional lymph nodes** ➢ N0: no regional LNs metastases ➢ N1: one regional LNs metastases ➢ N2: more than one regional LNs metastases ▪ **M : Distal metastases** ➢ M0:No distal metastases . ➢ M1:p distal metastases .
★**Complications:**		
I)Spread :	▪ **Mainly direct** spread.	▪ **Mainly blood** spread which may be: a. By **embolisation** to the lungs, bones & brain. b. By **permeation** to the lumen of the renal vein → I.V.C. → malignant thrombus.
1-Direct :	▪ To the surrounding renal tissue & organs.	
2-Lymphatic :	▪Less commonly lymphatic spread , to the hilar L.Ns →	

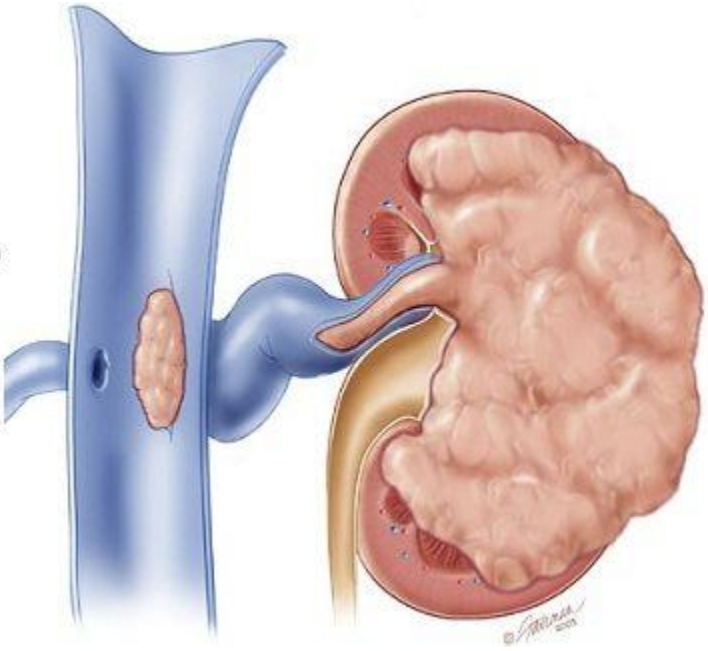
	para-aortic L.Ns → thoracic duct → Virchow's gland.	
3-Blood	▪ Mainly & early to the lungs and less commonly bones & brain.	
II)Destruction of renal tissues	▪Early loss of renal function .	▪Late .
III) Infection , anaemia, cachexia & death on late cases		
★ **Clinical picture :**		
I)Haematuria :	▪Late	▪ The **earliest presentation** is causeless, painless, spontaneous total and recurrent haernaturia .
**** Any patient with haematuria should be investigated throughly for the possiblity of neoplasms of the urinary tract.***		
II)Renal swelling :	▪ The **earliest and main** presentation (90%). ▪ Rapidly growing swelling in the loin, does not extend across the midline. ▪ Smooth & usually firm.	▪ Late presentation. ▪ Hard, irregular, ill-defined renal swelling.
	▪ Restricted mobility or fixed	

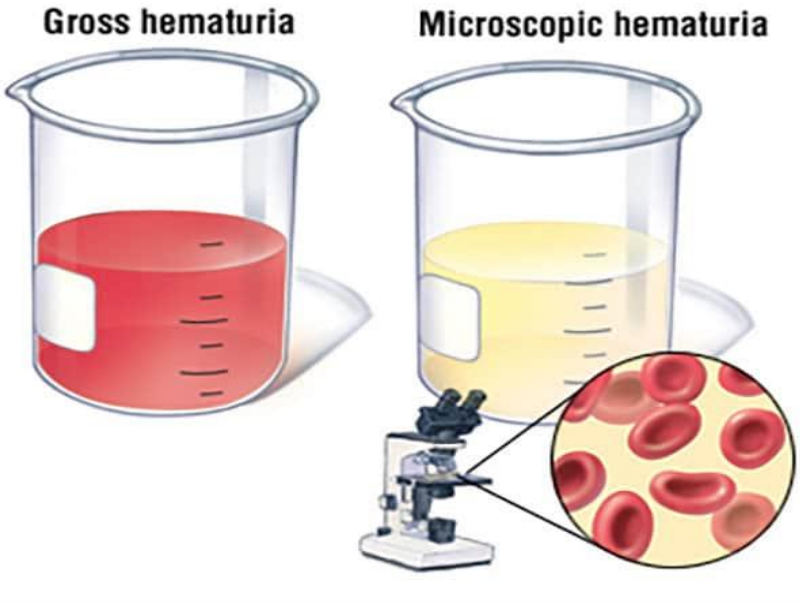
Spread of

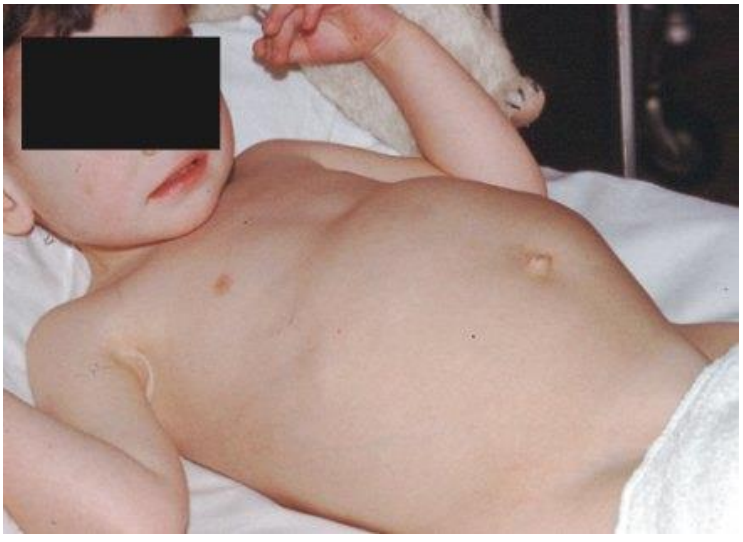
renal cell carcinoma

Into renal vein & IVC



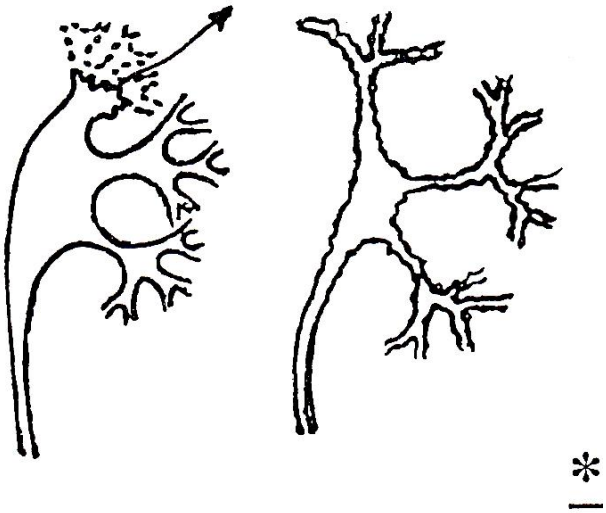




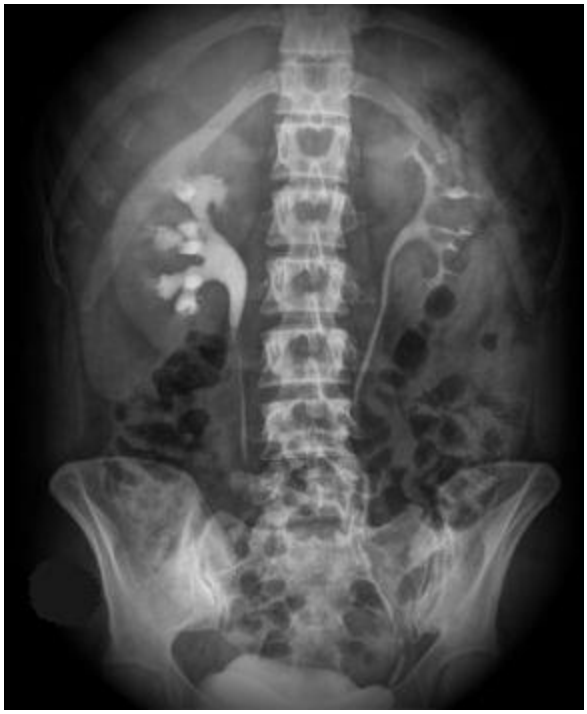


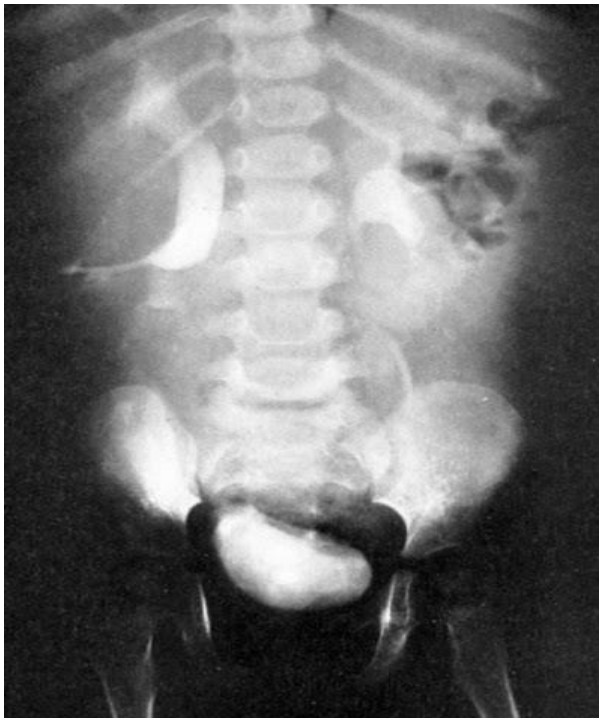
<table>
<tr><td>III)Pain :</td><td>▪ Late presentation due to:
a. Dull aching due to stretch of the renal capsule or infiltration of the surrounding structures.
b. Ureteric colic due to passage of clots,
c. Referred pain due to lumbar nerve infiltration,
d. Dragging pain in the loin.</td></tr>
<tr><td colspan="2">▪ The classical triad of haernaturia, pain & renal mass is rare usually indicate an advanced disease.</td></tr>
<tr><td>IV) Features of metastases</td><td colspan="2">▪ May be the first presentation with silent primary tumour. (Mention)</td></tr>
<tr><td rowspan="2">V) Atypical Manifestations</td><td colspan="2">▪ Hypertension due to encroachment of renal blood vessels → renal ischaemia → excess rennin production.
▪ Fever , malaise , anorexia , loss of weight and anaemia</td></tr>
<tr><td>▪ Associated anomalies may be present as neurofibromatosis, genito-urinary tract anomalies, visceromegally…etc</td><td>▪ 2 ry. varicocele on the left side only due to obstruction of left testicular vein. It is rapidly progressive & does not empty by elevation of scrotum.
▪ Polycythaemia due to secretion of erythropoietin by the tumor .
▪ Hypercalcaemia due to bone</td></tr>
</table>

		metastases or the tumor secret parathormone like substances .
★ **Investigations :**		
1-Urin analysis	▪ Haematuria is late .	▪ Haematuria is early
2-Renal functions	▪ Early loss of renal function in bilateral cases .	▪ Late loss of renal function
3- Plain X-ray:	▪ Abdomen : Large soft tissue shadow obliterates the psoas shadow. ▪ Chest : Cannon ball lung metastases are common.	
4- IVU :	▪ Gross deformity of pelvicalyceal system ▪ Early loss of renal function	▪ Show elongation, compression, displacement or amputation of a calyx.
5- U/S	▪ It is the initial investigation of choice to differentiate solid tumor from hydronephrosis .	
6- CT scan	▪ Show site, size and extend of the tumour involvement of renal capsule or surrounding organs , L.N.s, renal vein & I.V.C, liver metastasis differentiate cystic from solid swellings.	
7- **Investigations to detect metastases** (metastatic work up as breast cancer)		

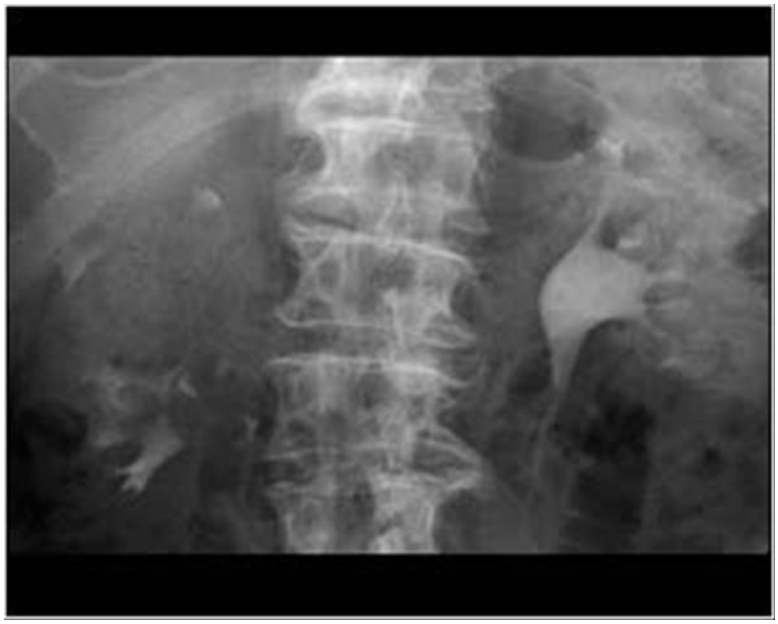


*** I.V.P. ***



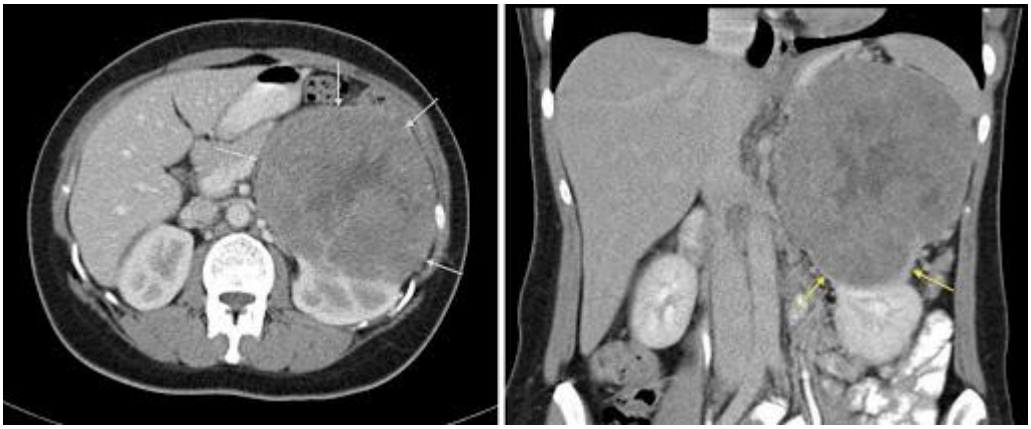


Wilms' Tumor



Renal Cell Carcinoma

CT show renal tumor



★ **D.D :**	▪ Other causes of renal or abdomenial swellings in children specially: 1-Neuroblastoma (hard , irregular & tends to cross the midline) 2- Polycystic kidney, solitary renal cyst & hydronephrosis.	▪ Other causes of haematuria

★ Treatment:

I- Operable cases:

- **Features: (as usual)**
- **Methods:**

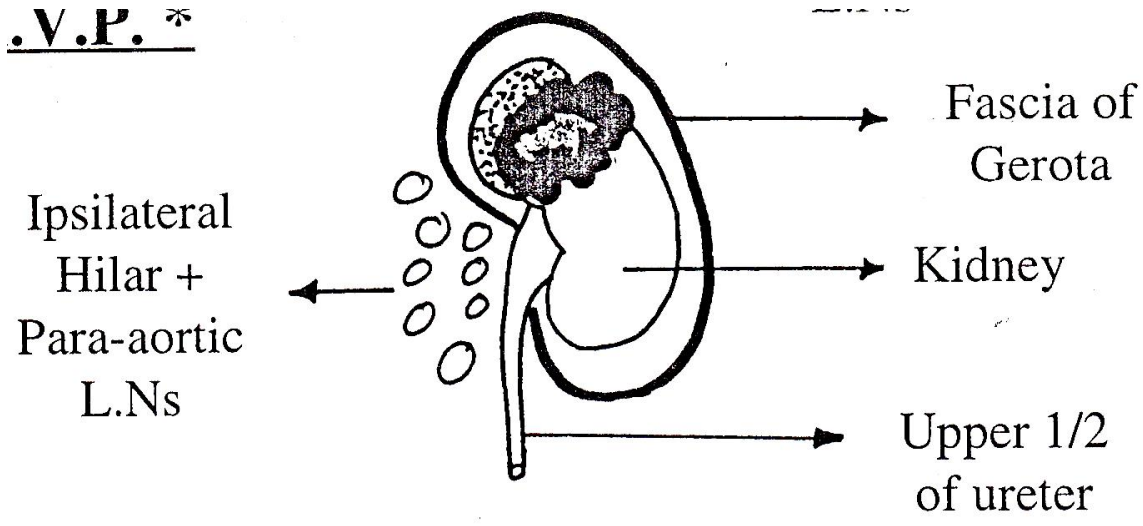
A. Neo-adjuvant (Preoperative) chemotherapy : only for large unresectable Wilms' tumor to shrink the tumor which then can be removed and put surgical clips to direct post-operative radiotherapy

B. Radical nephrectomy through anterior transperitoneal approach:

- It includs en block removal of the kidney within the surrounding fascia of Gerota (include suprarenal gland), upper 1/2 of ureter and ipsilateral hilar L. Ns & para-aortic L.Ns.

- ***Nephrectomy should be through anterior transperitoneal because:***
  - It allows full abdominal exploration
  - It allows preliminary ligation of the renal vessels thus minimizing hge. and dissemination.
  - If the renal vein contains a tumour thrombus it can be ligated flush with I.V.C. If this thrombus extends into l.V.C , resection of the affected segment is needed.
  - Huge tumour can be easily delivered.
  - Infiltrated viscera are dealt with .

C- Post-operative immunotherapy and target therapy for renal cell carcinoma and chemotherapy & radiotherapy for Wilmis' tumor.



Structures Removed In Radical Nephrectomy

II- Inoperable cases:

- **Features:** (as usual)
- **Methods:**

  1-Palliative partial or total nephrectomy

  2- Renal cell carcinoma is resistant to both chemotherapy & radiotherapy therefore **immunotherapy and target therapy** are the alternatives .