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WILMS TUMOR (nephroblastoma)

GENERALITIES

Most common paediatric renal tumor (80-90%)
6% of paediatric malignant tumors
Mean age at diagnosis 3.5 years old
5 malignant tumour in childhood
Origin in renal cortex

EMBRYONIC ORIGIN, NEPHROGENIC RESTS: increased risk of Wilms tumor in contralateral kidney
INTRALOBAR HIGUER RISK THAN PERILOBAR

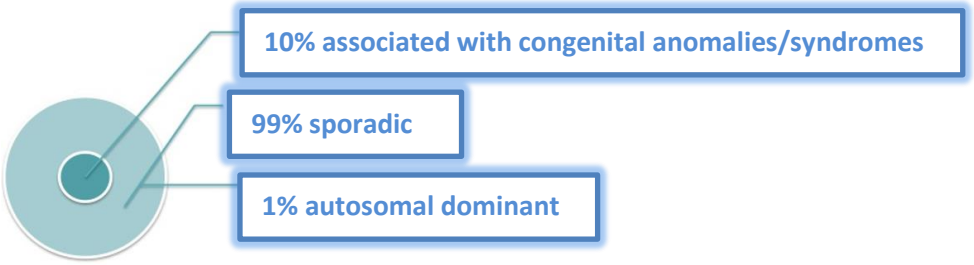
GENS: Wilms Tumor 1 (WT1) gene: chromosome 11p13
Wilms Tumor 2 (WT2) gene: chromosome 11p15

HISTOLOGY: Triphasic pattern (BLASTEMA, EPITHELIUM, STROMA).
80% heterogeneous (necrosis, haemorrhage). Solid or cystic
VARIANTS: Classic, anaplastic, diffusely anaplastic MOST IMPORTANT PROGNOSIS FACTOR

WILMS TUMOR DOES NOT CROSS MIDLINE (NEUBLASTOMA DOES CROSS)

DIAGNOSIS: ABDOMINAL MASS + PAIN + HEMATURIA. WELL-APPEARING *(NEUROBLASTOMA ILL-APEARING)*
TUMOR STAGING (based on surgery). Important prognosis factor.

STAGE	DESCRIPTION	%
1	Confined Getota´s fascia and completely resected	45%
2	Not confined within Getota´s fascia but completely resected	20%
3	Incomplete resection. Massive tumor spillage, preop biopsy, positive LN	20%
4	Distant Metastasis	20%
5	Bilateral	5%



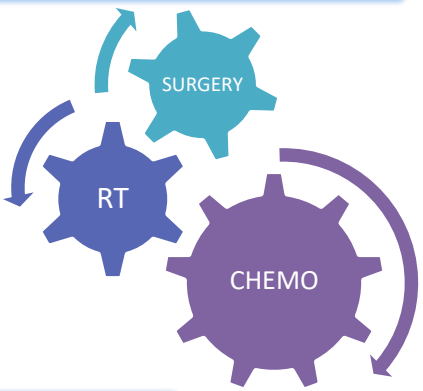
SYNDROME	CARACTERISTICS	LOCUS	WILMS
Beckwith-Wiedeman	Macroglossia, macrosomia, hemihyperplasia, visceromegaly, increased risk of tumors particularly Wilms' tumor, pancreatoblastoma and hepatoblastoma	11p15	>30%
Denys-Drash	Gonadal dysgenesis, nephropathy, and Wilms tumor	11p13	> 50%
WARG	Wilms tumour, Aniridia , Genitourinary anomalies, and mental Retardation	11p13	<5%

TREATMENT: multidisciplinary approach

- **Age, preop. extent, operative stage, histology**

WILMS TUMOR ARE HIGHLY CHEMO AND RADIOSENSITIVE

RADICAL NEPRHECTOMY + LYMPH NODE SAMPLIG + ADYUVANT CHEMOTHERAPY (consider RT if stage ≥ 3)



Neoadjuvant chemotherapy (Vincristine , Dactinomycin, and /or Doxorubicin) for bilateral or unresectable tumors or tumors in solitary kidney.
SIOP: In this consortium's trials, preoperative chemotherapy is administered prior to definitive resection for patients with renal tumours. This allows for fewer intraoperative tumour ruptures and a lower postoperative stage

MOST COMMON COMPLICACIONS: Small bowel obstruction, haemorrhage, infection or hernia

PROGNOSIS: 90% in localized disease and 70% metastatic disease