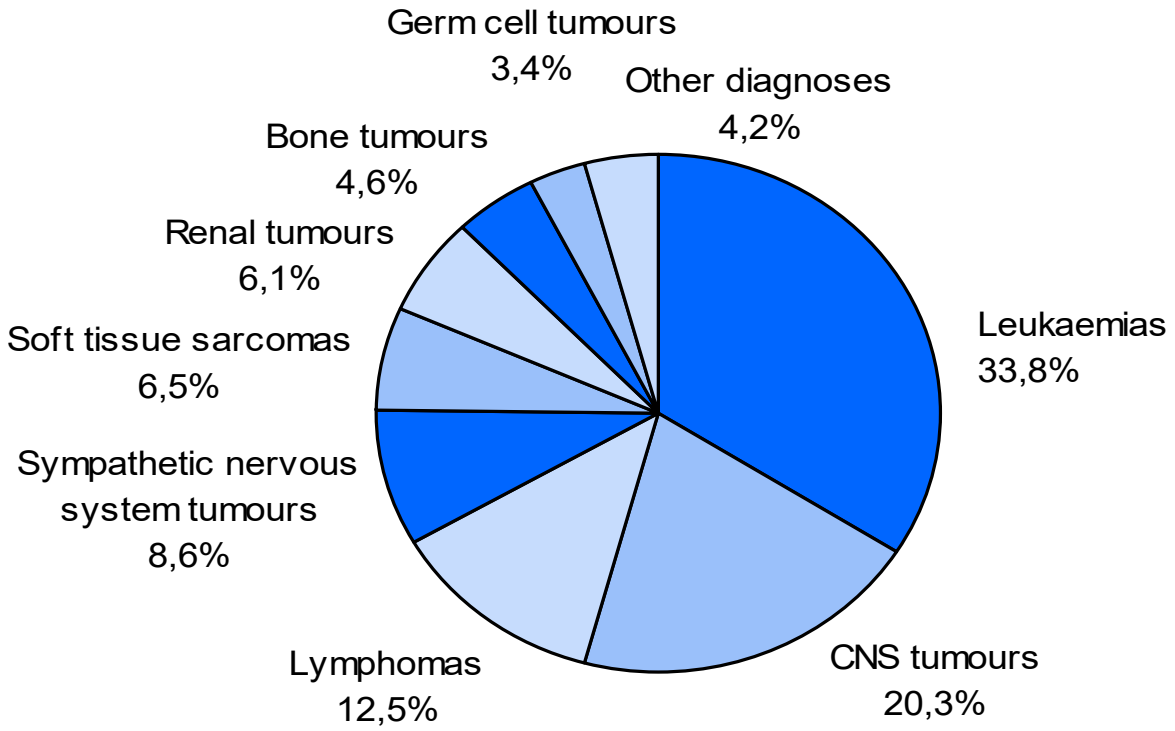
PEDIATRIC MALIGNANT SOLID TUMORS

By

Eman Mohamed Fahmy

Clasification of Pediatric Malignancies



Systemic Neoplasms (Leukaemias and Lymphomas) : Solid Tumors = 1:1.

Malignant Solid Tumors (50%)

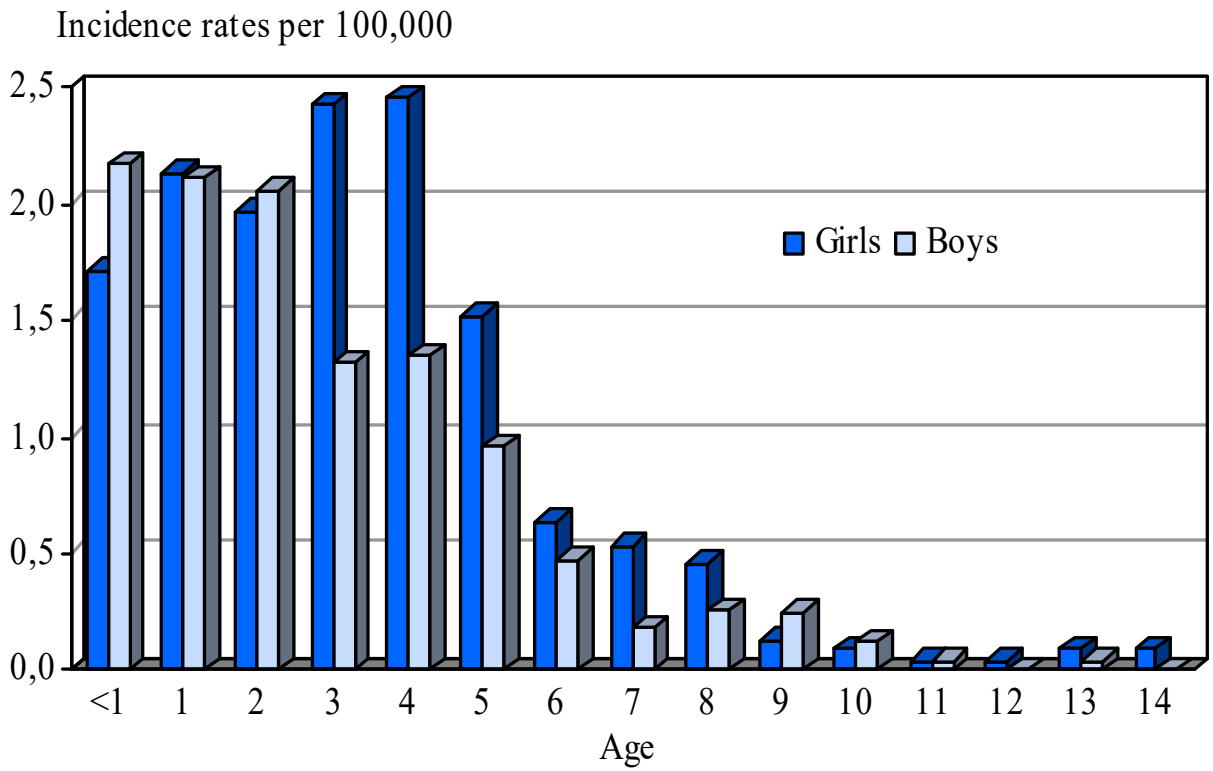
- Brain Tumors : 17% (Astrocytoma,Medulloblastoma....)
- Embrional Tumors : 17% (Neurollastoma (8%), Nephroblastoma (7%), Retinollastoma(1.5%), Hepatoblastoma (0.5%))

- Soft Tissue Sarcomas 8% (Rhabdomyosarcoma.)
- Bone Tumors - 4% (Osteosarcoma, , EwingSarcoma ...)
- Germ Cell Tumors - 2%
- Epithelial Tumors - Carcinomas and other very Rare Tumors - 2%

Wilms" Tumors (Nephroblastoma)

- Epidemiology - 7% of Neoplastic Diseases in Children
- Unfavonrable Histology (Focal or Diffuse Anaplasia) - 10%
- Favourable Histology (Multicystic and with Fibroadenomatous Structures) - 90%
- Gene Mutations - 11p 13 for WT1 and 11p 15 for WT2
- BW syndrome

Incidence


Incidence rates per 100,000
2,5
2,0
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Girls
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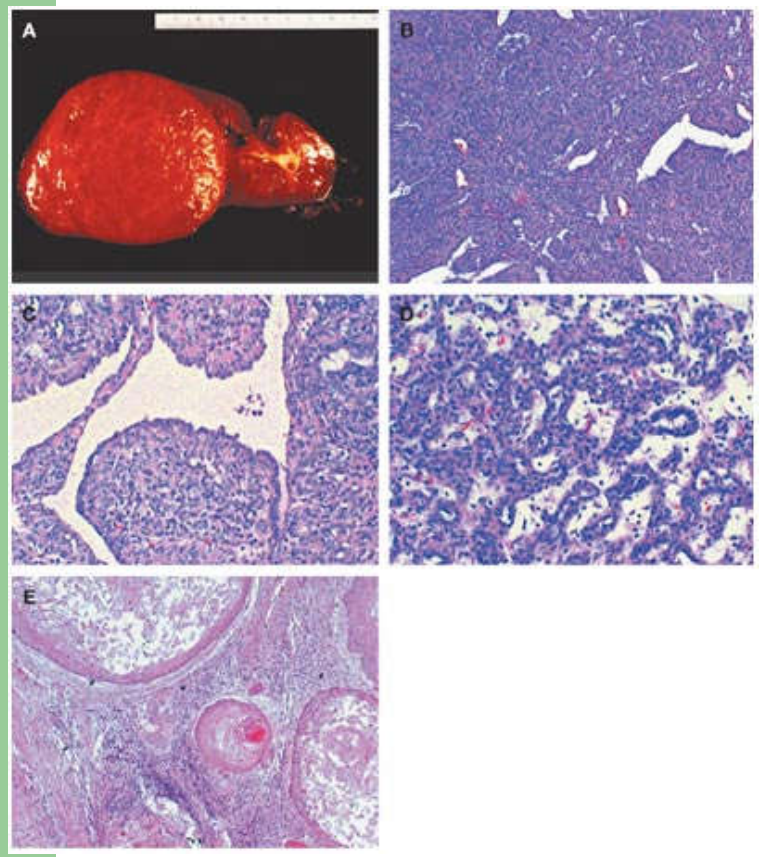
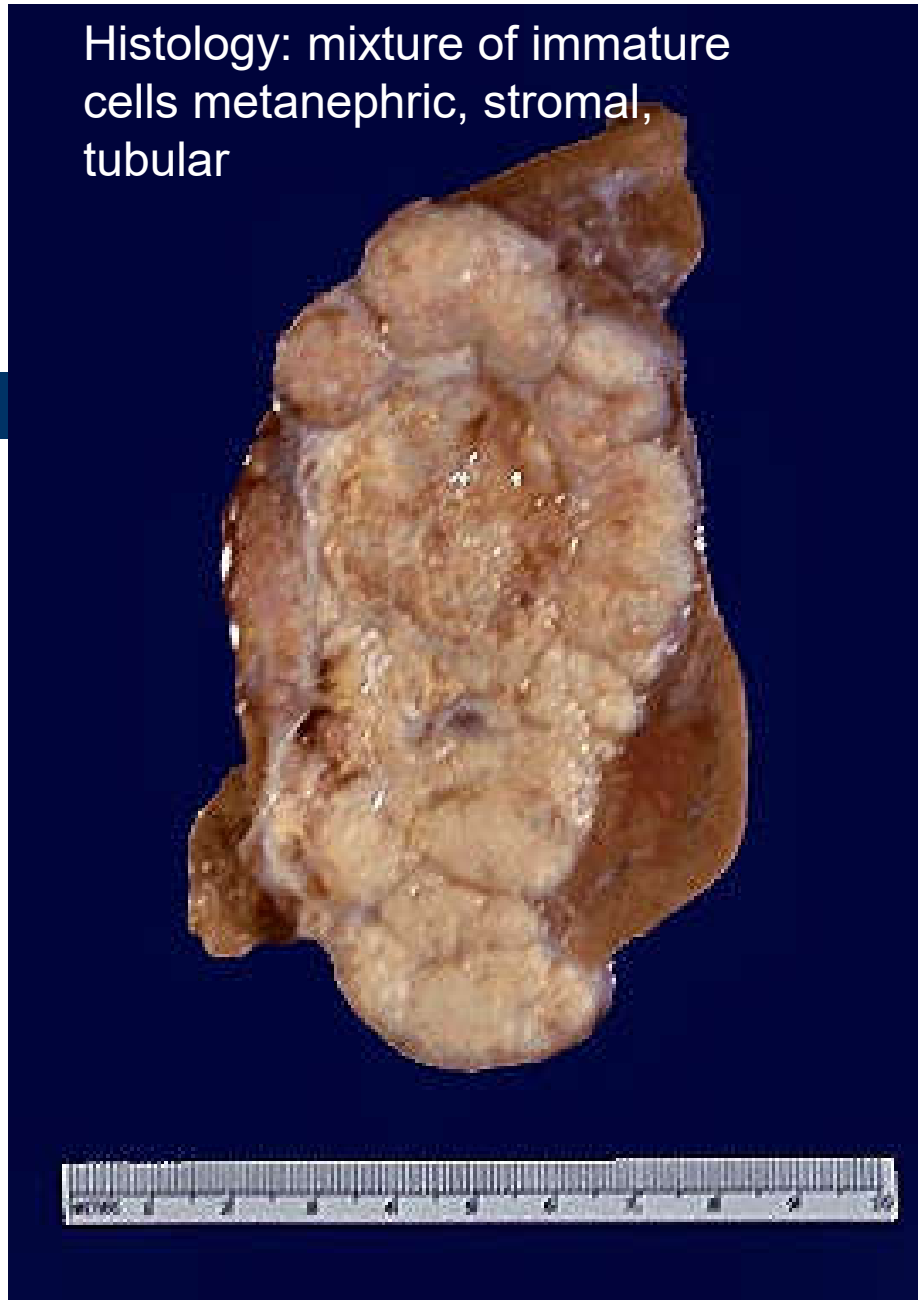
Age

Wilms Tumor:





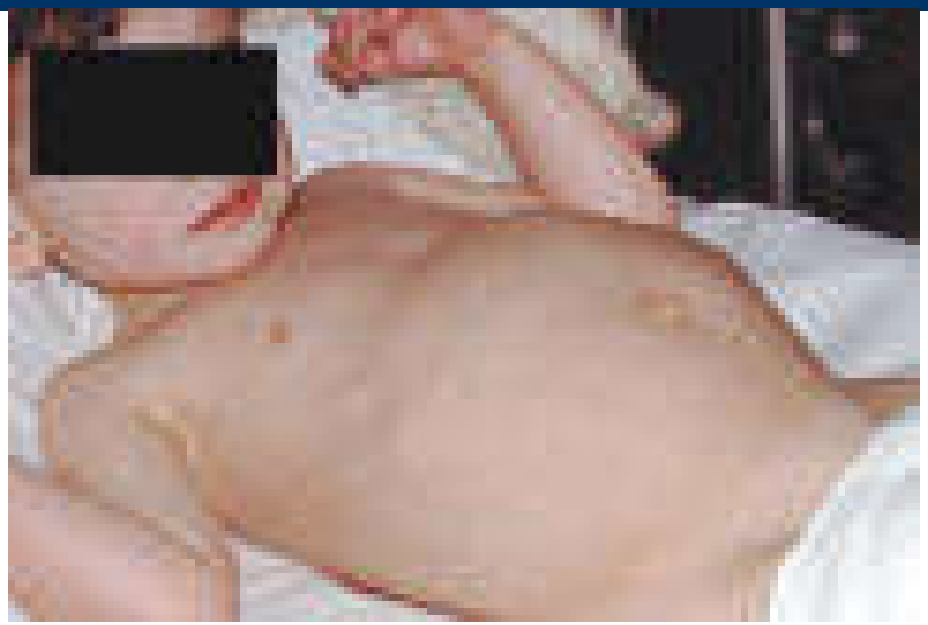
Cajaiba MM *et al.* (2006) Rhabdomyosarcoma, Wilms tumor, and deletion of the *patched* gene in Gorlin syndrome *Nat Clin Pract Oncol* **3**: 575–580 doi:10.1038/ncponc0608

Clinical Manifestation

- Good Clinical State
- Haematuria - 25%
- Abdominal pain - 35%
- High blood pressure - 35%
- An abdominal Tumor mass **- discovered accidentally**

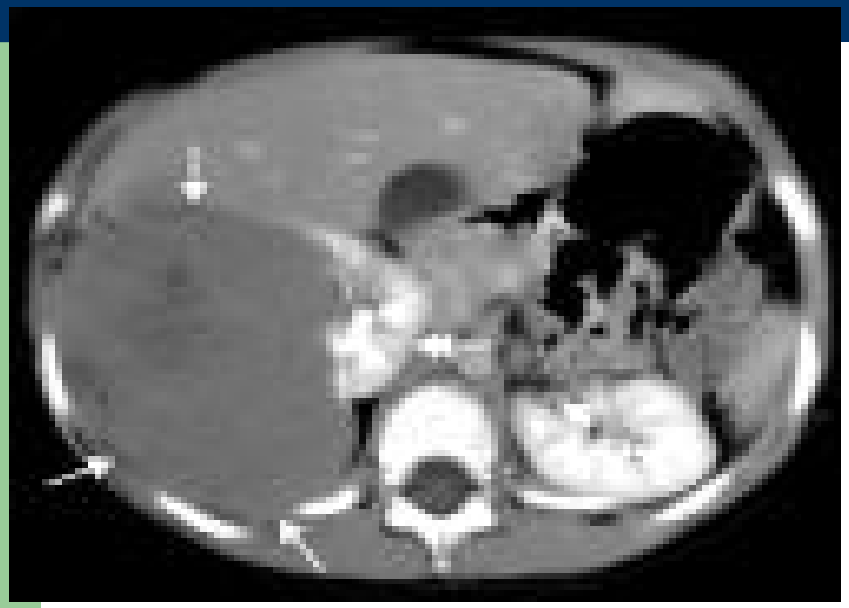
Clinical stages

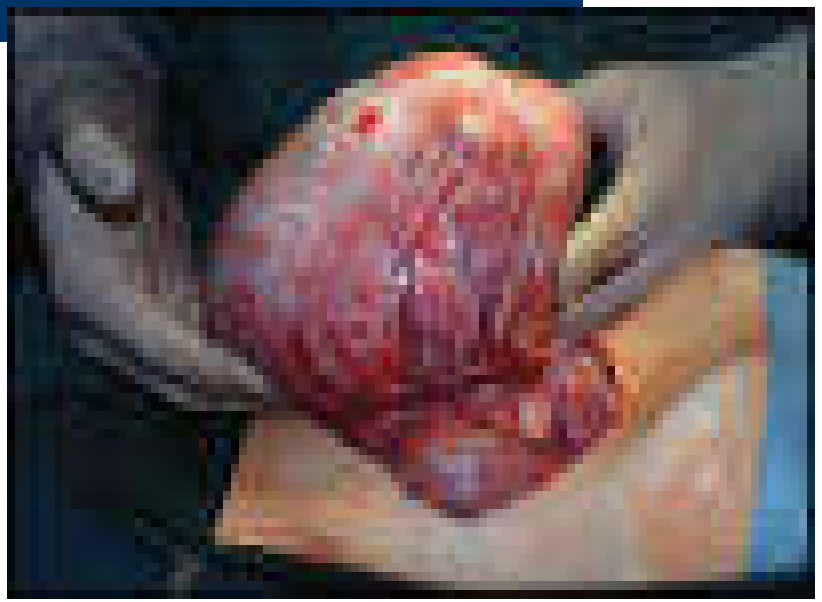
- I. Tumor limited to the Kidney, in size - 5 cm. Intact Renal Capsule, Excised Completly.
- II. Tumor extends outside the Kidney in size - 10 cm, Excised Completly.
- III. Tumor over 10 cm in size. Infiltrated other Organs in Ablomen, without Hematogenous Mts, Complete Excision Impossible.
- IV. Tumor with Hematogenous Mts (Lungs - 10% liver - 1% ets.)
- V. Bilateral Renal Tumors



Laboratory and RadiologicalExaminations

- Hb, Plt, WBS,
- LDH
- Urine analysis CT and Abdominal Ultrasound
- Chest X-ray
- **Tumor Histology**





Risk Factors

- Low Risk - Cases withl Favourable Histology, in I and II Stages, under 3 Year of Age

- High Risk - Cases with Unfavourable Histology, in III, IV and V Stages, over 3 Year of Age

Treatment

- Surgery - Nephrectomy, Lymphadenotomy, Excision when it is possibly of lung or liver Mts. In V stage -partial resection.
- Radiotherapy. No RT in I and II stage. In III and IV stage RT in Tumor Region with Dose 20-30 GY. In Mts regions - 15 GY
- Chemotherapy

Prognosis

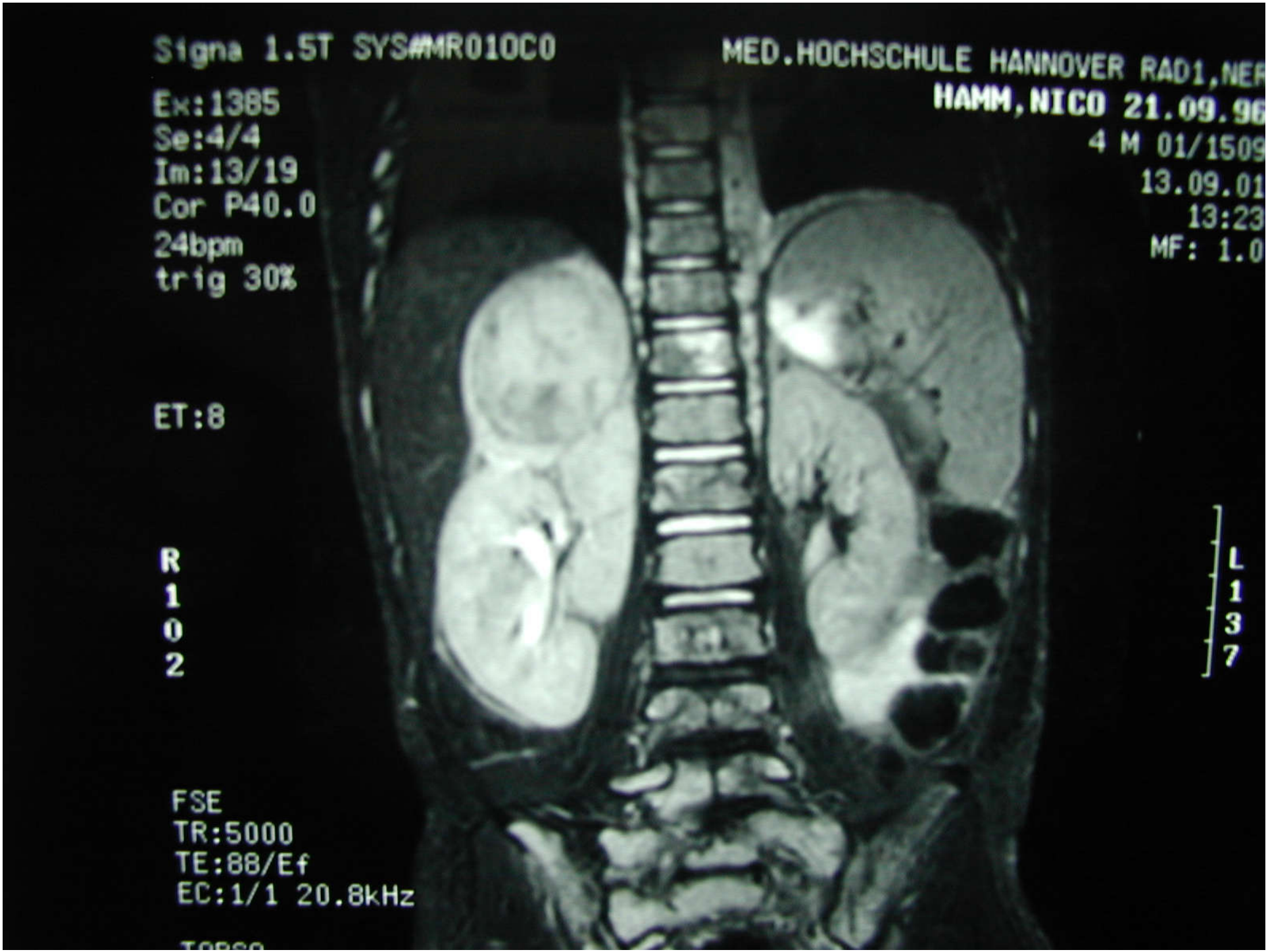
- Low Risk - 85% survival
- High Risk - 40% survival

Neuroblastoma

- Epidemiology - 8% of Neoplastic Diseases in children
- Unfavourable Histology (Neuroblastoma) - 90%
- Favourable Histology (Ganglioneuroblastoma) - 10%

Neuroblastoma Localization -

- 70% Abdomen (1/2 of Cases from neural crest tissue suprarenal gl.)
- 15% sympathatic chain in Mediastinum,
- 3% Neck,
- 8% Paravertebral Region,
- 4% Other Rare Regions (Olfactory Region, Multiple Primary Tumors C.N.S ets)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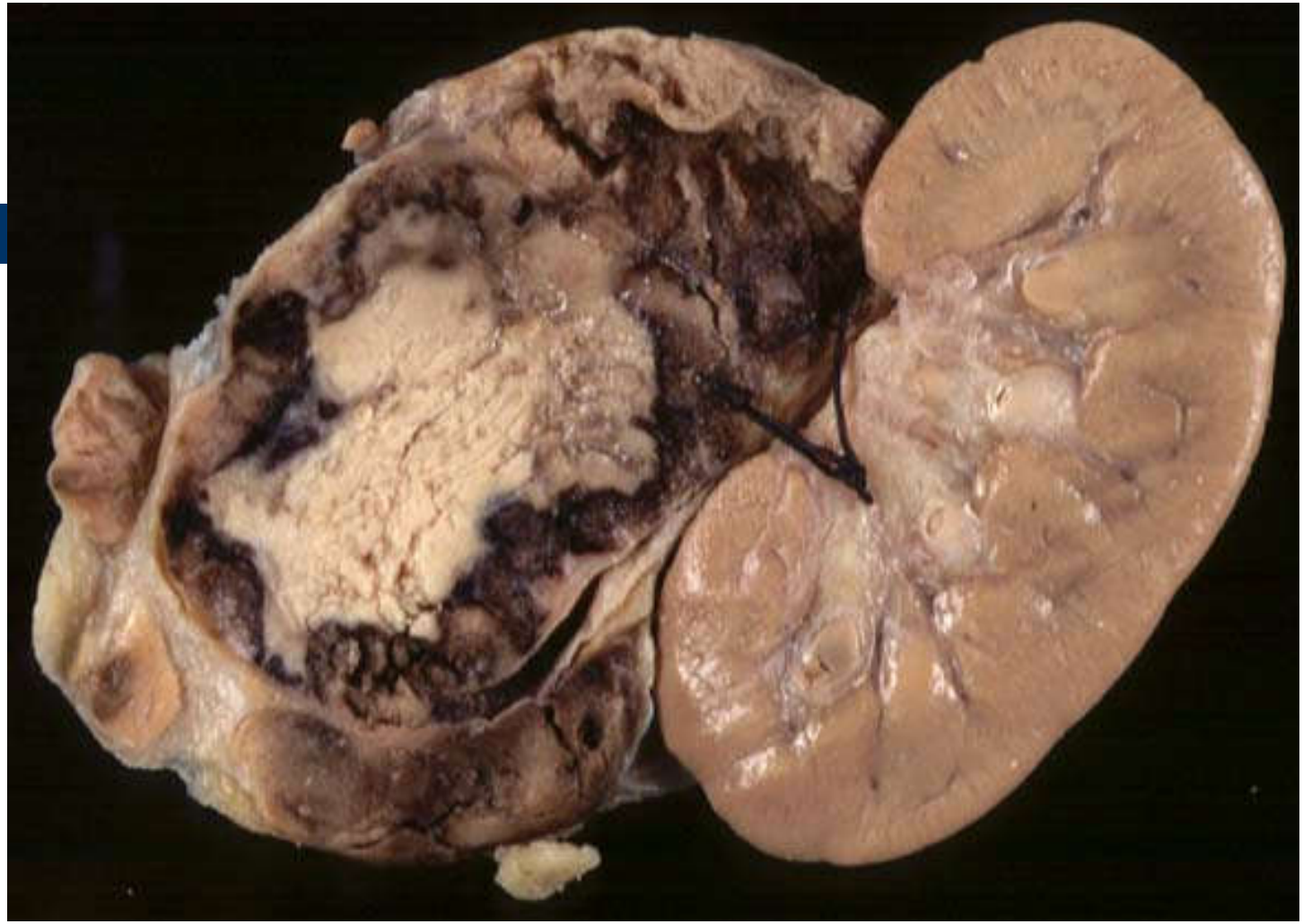
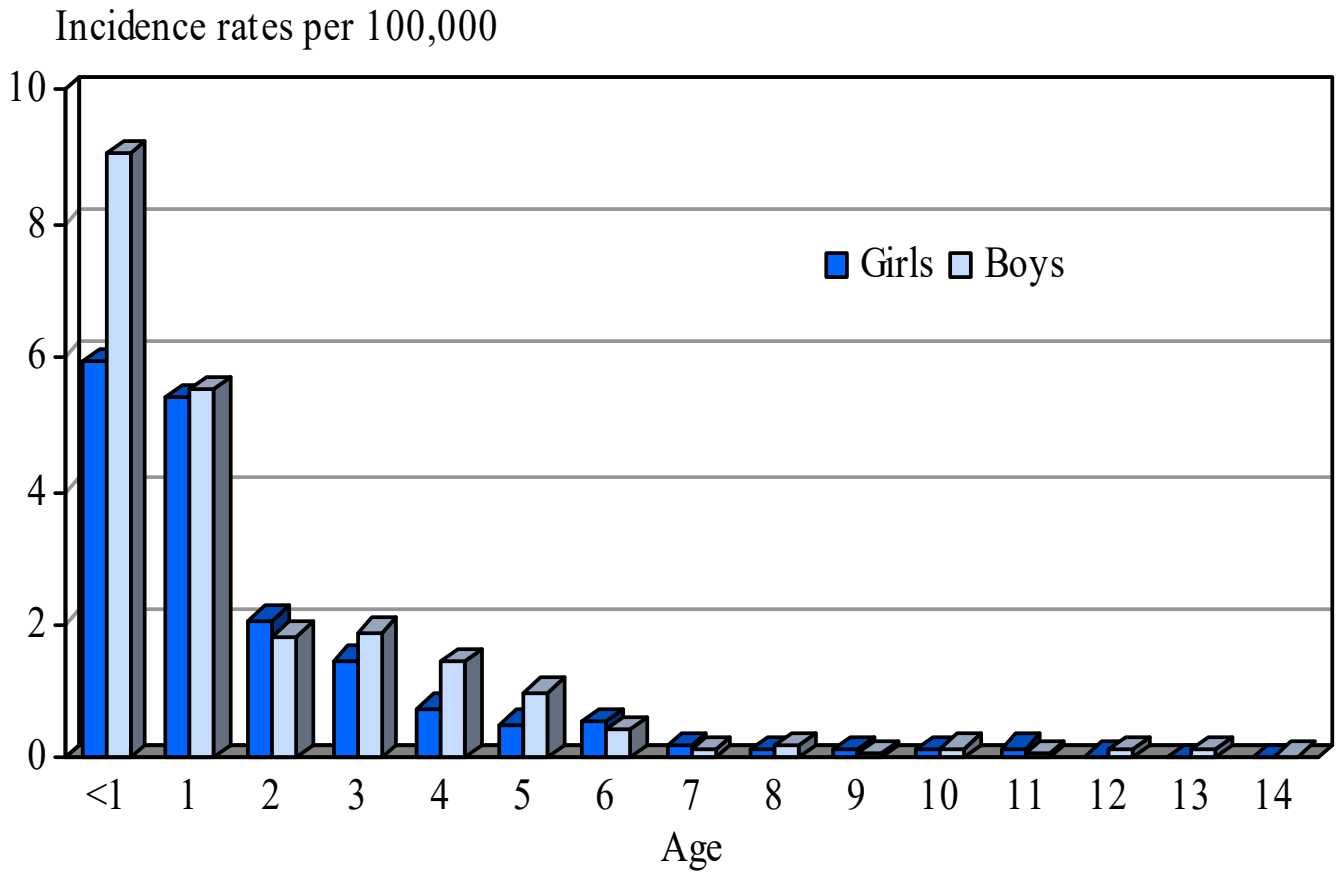


Incidence


Incidence rates per 100,000
10
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Girls
Boys
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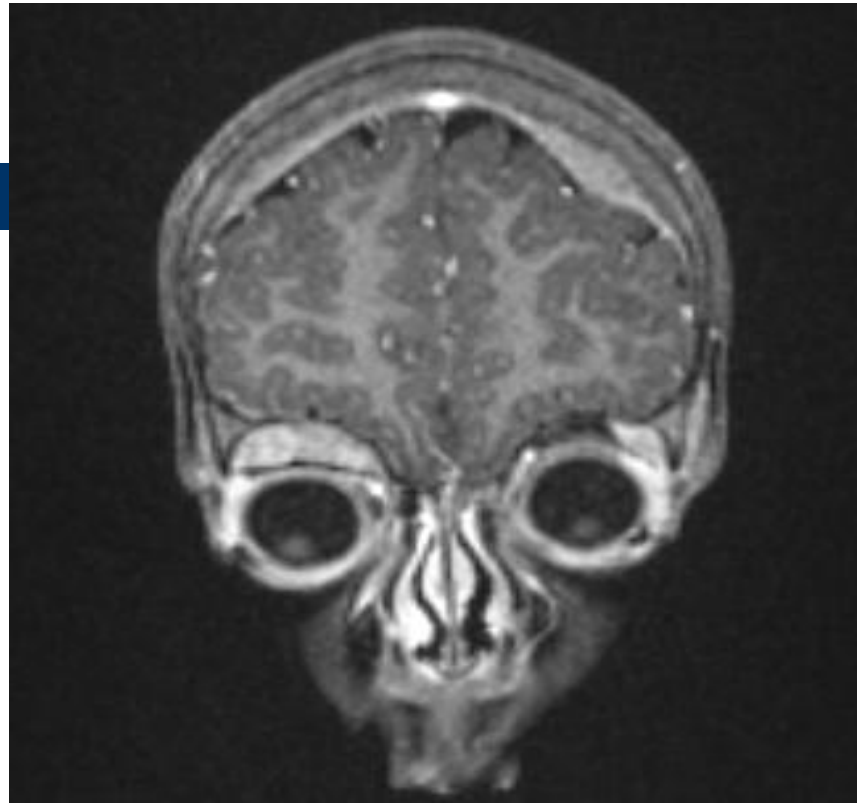
Age

Clinical Manifestation

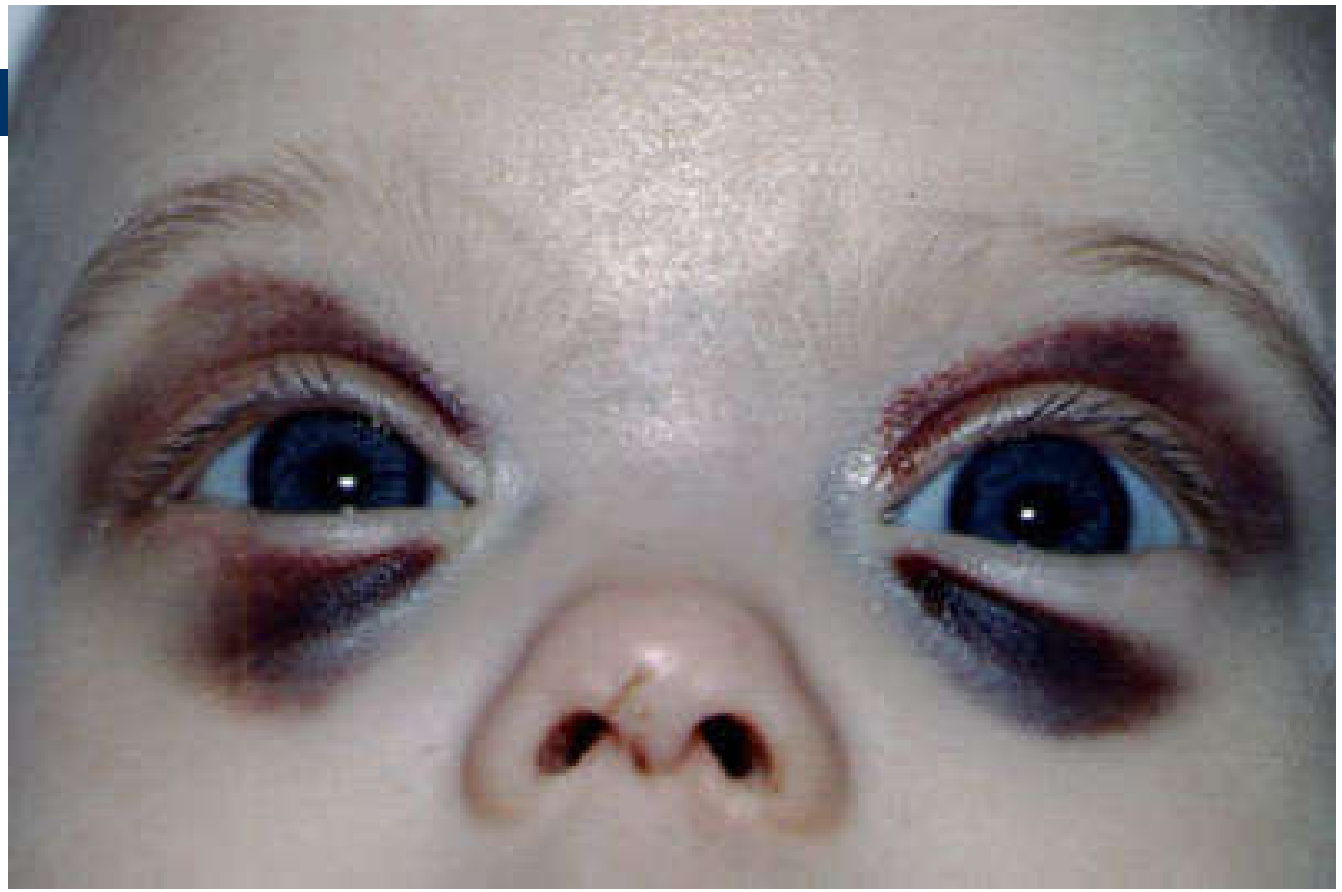
- Poor clinical State(weight loss,poor appetite,fever)
- Symptoms of Primary Localization(abdomenal pain and mass, hepatomegally)
- Symptoms of Metastatic Localization(bone, bone marrow, skin, internal organs)
- Paraneoplastic Symptoms(hypertension, sweating, secretory diarrae)

There are orbital and skull vault metastases,

soft-tissue masses.
The skull lesions are extradural masses which deform the underlying brain.
The right orbital lesion forms a superior extraconal mass,
depressing the right globe.



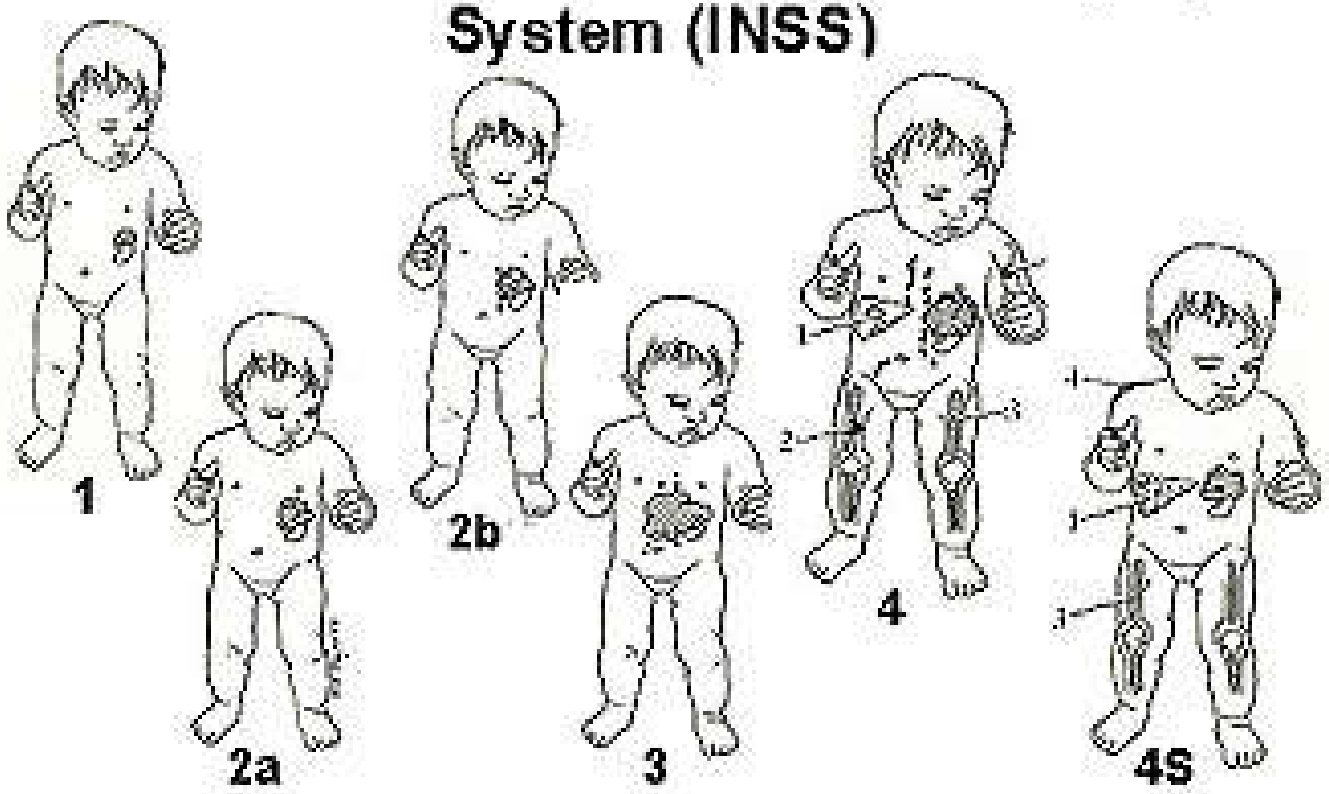
bilateral ecchymosis in a child with metastatic neuroblastoma.



Clinical Stages (Evans et al.)

- I Tumor Limited to the Organ or Structure of Origin. Excised Comletely.
- II Tumor with Regional Spread, not Crossing the Midline.
- III Tumor Crossing the Midline, Bilateral Lymph Nodes May by involved. Complete Excision imposible.
- IV Tumor with Distant Mts (Bone - 50% of cases, Lymph Nodes, Organs, Soft Tissues)
- IV-S Tumor in I and II Cl Stage, with Limited Dissemination to Liver, Skin and Bone Marrow (without Bones), Infants under 2 Years of Age, especially under 1 Year of Age.

International Neuroblastoma Staging System (INSS)

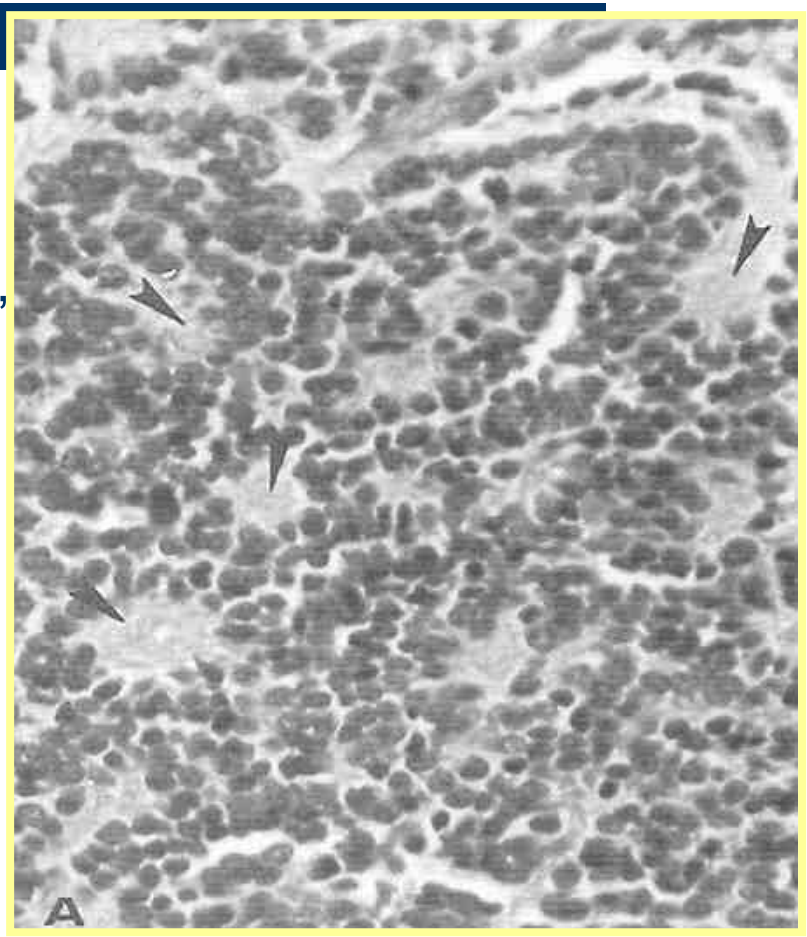


Laboratory and Radiological Examinations

- Hb, Plt, WBC, LDH,
- Bone Marrow Aspiration
- Urinanalysis
- CT and Abdominal Ultrasound
- Bone Isotope Scanning, Scanning with 131I-MIBG
- Chest X-ray
- Tumor Markers - N-myc, Ferritin, NSE, Cantecholamines' Metabolites(VMA,HVA)
- Tumor Histology

Neuroblastoma

- "Small blue round cell" tumor
- Immunohistochemical stains: neurofilament proteins, synaptophysin, NSE
- Electron microscopy: neurosecretory granules, microtubules and filaments
- Chromosome 1 deletions or *N-myc* oncogene amplification



From, Principles and Practice of Pediatric Oncology, Lippincott Williams & Wilkins, p 903.

Risk Factors

- Low Risk - Cases with Ganglioneuroblastoma, in I, II and IV-S Stages, under 1 Year of age, with Neck and Mediastinum Localisation

- High Risk - Cases with Neuroblastoma, in III and IV Stages (Bone Mts), over 2 Years of age, with High Levels of LDH, NSE and Fevritin, with Abdominal and Paravertebral Localisation, with Amplification of N-myc.

Treatment

- Surgery - Survival is better when Radical Excision is done.

- Radiotherapy - No RT in I, II and IV-S Stages
- **In III and IV Stages RT in Tumor and Mts Redions with Dose 15-35 GY.**

- Chemotherapy

Prognosis

- Low Risk - 80% survival
- High Risk - 35% survival