Pediatric Cancer Subtypes

- Leukemia: 33%
- CNS: 9%
- Neuroblastoma: 6%
- Hodgkin's lymphoma: 7%
- Non-Hodkin's lymphoma: 5%
- Kidney (Wilm's): 4%
- Liver (hepatoblastoma): 2%
- Bone (Osteosarcoma and Ewing's): 8%
- Soft tissue sarcoma: 2%
ALL
(Acute Lymphoblastic leukemia)

- 75% of all Leukemias
- Can present with generalized bone pain, fatigue
- Bruising, nose bleeds
- Unusual fevers, infection
- Lymphadenopathy, hepatosplenomegaly
Figure 21.6 Leukaemic blast cells on a bone marrow smear.
Signs and symptoms of acute leukaemia

General
- Malaise, anorexia

Bone marrow infiltration
- Anaemia → Pallor, lethargy
- Neutropenia → Infection
- Thrombocytopenia → Bruising, petechiae, nose bleeds → Bone pain

Reticulo-endothelial infiltration
- Hepatosplenomegaly
- Lymphadenopathy
- Superior mediastinal obstruction (uncommon)

Other organ infiltration*
- Central nervous system → Headaches, vomiting, nerve palsies
- Testes → Testicular enlargement

*Rare at diagnosis, more often at relapse
ALL Treatment

- **Induction**: 4-6 weeks,
- **Consolidation /delayed Intensification**: 6-12 months; rotating drugs.

- **Maintenance**: Daily oral 6-MP, weekly MTX, Monthly pulses of Vincristine and Steroid.
- Imatinib mesylate-Tyrosine Kinase Inhibitor- Ph Chromosome positive patients
- **CNS prophylaxis**: Intrathecal chemo
- **CNS Therapy**: RT + Intensive systemic chemo
- **Testicular disease**: RT
- **SANCTUARY- CNS, Testis**
<table>
<thead>
<tr>
<th>Prognostic factor</th>
<th>High-risk features</th>
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<tbody>
<tr>
<td>Age</td>
<td>&lt;1 year or &gt;10 years</td>
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<tr>
<td>Tumour load (measured by the white cell count, WBC)</td>
<td>&gt;50×10⁹/L</td>
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<tr>
<td>Cytogenetic/molecular genetic abnormalities in tumour cells</td>
<td>e.g. MLL rearrangement, t(4;11), hypodiploidy (&lt;44 chromosomes)</td>
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<td>Speed of response to initial chemotherapy</td>
<td>Persistence of leukaemic blasts in the bone marrow</td>
</tr>
<tr>
<td>Minimal residual disease assessment (MRD) (submicroscopic levels of leukaemia detected by PCR)</td>
<td>High</td>
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</table>
Brain Tumors- Important to know

- 20% of all malignancies in children
- Age 3-7 years
- Most often infratentorial
- Sx: Persistent vomiting, headache, gait imbalance, diplopia, ataxia, vision loss, school deterioration, growth deceleration
- Associations with Inherited Genetic disorders:
  - Neurofibromatosis, Tuberous sclerosis, Von-Hippel-Lindau disease, Li-Fraumeni (glioma), Turcot syndrome
Brain tumours almost always primary infratentorial.

The types of brain tumour are:

Astrocytoma (~40%)

Medulloblastoma (~20%)

Ependymoma (~8%)

Brainstem glioma
Craniopharyngioma (4%)
Lymphomas

Lymphomas are malignancies of the cells of the immune system.

Types

Hodgkin and non-Hodgkin lymphoma (NHL).
Lymphoma

Hodgkin

- Ages 15-34 yrs and 50+
- Persistent, painless, rubbery lymphadenopathy
- B symptoms
- Reed-Sternberg cell
- Treatment: chemotherapy and radiation based on response and risk group

Non-Hodgkin

- Incidence peak 7-11 yrs
- Rapidly growing with distant mets
- Types
  - Lymphoblastic lymphoma
  - Burkitt’s
    - Endemic (EBV) vs. sporadic
  - Diffuse large B cell
  - Anaplastic large T cell
- Primary treatment with chemotherapy
Hodgkin lymphoma

Clinical features

B symptoms
Hodgkin's disease

- Pathology
- Reed Sternberg cell is the Hallmark feature
Non-Hodgkin lymphoma

T-cell malignancies characterised by a mediastinal mass with varying degrees of bone marrow infiltration. The mediastinal mass may cause superior vena caval obstruction.

B-cell malignancies present more commonly as localised lymph node disease usually in the head and neck or abdomen. Abdominal disease presents with pain from intestinal obstruction, a palpable mass or even intussusception in cases with involvement of the ileum.
Abdominal Mass

Wilms’ Tumour

- Most common primary renal neoplasm of childhood
- 2-5 years of age
- Asymptomatic, unilateral abdominal mass
- Renal symptoms: HTN, hematuria, abdo pain, vomiting
- Treatment:
  - Surgical with nephrectomy
  - Chemotherapy +/- radiation
  - 90% long term survival

Neuroblastoma

- Neural crest cells arising from sympathetic tissues
- Originate from any site in sympathetic nervous system
  - Presentation based on location of mass
  - Common to have mets
- Treatment can vary
  - Observation to....
  - Chemo/Rads/Surgery/ Bone marrow transplant/ Targeted therapy
Neuroblastoma

Neuroblastoma and related tumours arise from neural crest tissue in the adrenal medulla and sympathetic nervous system. It is a biologically unusual tumour in that spontaneous regression sometimes occurs in very young infants. There is a spectrum of disease from the benign (ganglioneuroma) to the highly malignant (neuroblastoma). Neuroblastoma is most common before the age of 5 years.
**Figure 21.11** Transverse MRI image showing a large left-sided primary neuroblastoma arising from the adrenal region and distorting coeliac and mesenteric blood vessels.

**Figure 21.12** The MIBG scan ‘maps’ metastatic tumour marrow. This image shows the lower half of the abdomen, pelvis and legs. The dark areas are evidence of high isotope uptake and the pattern is consistent with widespread metastatic disease. (Normal uptake from excretion of isotope into urine in the bladder has been blocked in this exposure.)
**Box 21.1** Presentation of neuroblastoma

<table>
<thead>
<tr>
<th>Common</th>
<th>Less common</th>
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<tbody>
<tr>
<td>Pallor</td>
<td>Paraplegia</td>
</tr>
<tr>
<td>Weight loss</td>
<td>Cervical</td>
</tr>
<tr>
<td>Abdominal mass</td>
<td>lymphadenopathy</td>
</tr>
<tr>
<td>Hepatomegaly</td>
<td>Proptosis</td>
</tr>
<tr>
<td>Bone pain</td>
<td>Periorbital bruising</td>
</tr>
<tr>
<td>Limp</td>
<td>Skin nodules</td>
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Wilms tumour (nephroblastoma) originates from embryonal renal tissue and is the commonest renal tumour of childhood. Over 80% of patients present before 5 years of age, very rarely seen after 10 years of age.

**Figure 21.13** Large Wilms tumour arising within the left kidney, showing characteristic cystic and solid tissue densities.
## Box 21.2 Presentation of Wilms tumour

<table>
<thead>
<tr>
<th>Common</th>
<th>Uncommon</th>
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<tbody>
<tr>
<td>Abdominal mass</td>
<td>Abdominal pain</td>
</tr>
<tr>
<td></td>
<td>Anorexia</td>
</tr>
<tr>
<td></td>
<td>Anaemia (haemorrhage into mass)</td>
</tr>
<tr>
<td></td>
<td>Haematuria</td>
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<tr>
<td></td>
<td>Hypertension</td>
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About 5% have bilateral disease at diagnosis.
Retinoblastoma -

- **Presentation:** Leukocoria (cats eye reflex), dilated pupil, esotropia, strabismus

- **Unilateral 75%** (could be hereditary/non)
  - 60% unilateral and *non hereditary*
  - 15% unilateral and *hereditary (RB1 mutation)*

- **Bilateral 25%**
  - 25% are bilateral and hereditary, *have RB1 mutation*
  - Earlier age, 11mos, Can develop in each eye separately
  - Higher incidence of sarcoma, melanoma, brain tumors.