Pediatric Oncology for Otolaryngologists

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Basic Facts About Childhood Cancer

- 12,400 children are diagnosed with cancer each year
- 1 in 300 boys will develop cancer
- 1 in 333 girls will develop cancer
- Leading cause of death by disease in children
- 2,500 children die of cancer each year
- 75% of children with cancer will survive
Major Pediatric Malignancies

- Leukemia
- Central nervous system tumor
- Lymphoma
- Sarcoma
- Neuroblastoma
- Wilms tumor
- Germ cell tumor
- Retinoblastoma
- Hepatoblastoma
Trends in age-specific incidence rates for all childhood cancers by age

ALL RACES, BOTH SEXES, SEER, 1975-1995

<table>
<thead>
<tr>
<th>AGE</th>
<th>Line Color</th>
</tr>
</thead>
<tbody>
<tr>
<td>under 5 yrs</td>
<td>green</td>
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<tr>
<td>5-9 yrs</td>
<td>orange</td>
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<tr>
<td>10-14 yrs</td>
<td>red</td>
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<tr>
<td>15-19 yrs</td>
<td>blue</td>
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</table>
Total childhood cancer age-specific incidence rates


AVERAGE ANNUAL RATE PER MILLION

Age (in years) at diagnosis
Age-adjusted* incidence rates for cancer by ICC** group and race/ethnicity

Under 20, both sexes, SEER, 1990-1995

*Adjusted to the 1970 US standard population
**International Classification of Childhood Cancer
Number of cases of all childhood cancers by ICCC and age group

ALL RACES, BOTH SEXES, SEER, 1975-1995

ICCC GROUP

- Leukemia
- Lymphoma
- Brain / CNS
- Sympathetic Nerv.
- Retinoblastoma
- Renal
- Hepatic
- Bone
- Soft tissue
- Germ cell
- Carcinomas
- Other

Number of cases (in thousands)

AGE

- under 1 yr.
- 1-4 yrs
- 5-9 yrs
- 10-14 yrs
- 15-19 yrs
Figure XIV.1: Trends in childhood cancer age-adjusted* rates, all races, both sexes, age <20
SEER incidence & US mortality, 1975-95

*Adjusted to the 1970 US standard population
Trends in age-specific cancer mortality rates by type

15-19 YRS, ALL RACES, BOTH SEXES, USA, 1975-1995

**AVERAGE ANNUAL RATE PER MILLION**

**CANCER TYPES**
- Green: Brain & CNS
- Orange: Leukemia
- Red: Bones & joints
- Blue: Soft tissue
- Yellow: Non-Hodgkins lymphomas

Year:
- 1975-1977
- 1981-1983
- 1987-1989
- 1993-1995
Clinical Trials for Childhood Cancers

- Began 50 years ago when cure rate was less than 10%
- Basis for modern treatment of childhood cancers
- Key to success in pediatric oncology
Head and Neck Tumors

- Hodgkin lymphoma
- Rhabdomyosarcoma
- Neuroblastoma
- Nasopharyngeal carcinoma
- Esthesioneuroblastoma
- Thyroid carcinoma
- Other
Hodgkin Lymphoma

- Most common lymphoma in children
- More common in children older than 15 years of age
- Associated with Epstein-Barr virus and immunodeficiency
- 60-80% of cases involve neck
- Local invasion of regional lymph node
- Metastatic spread to lung, liver, bone, and bone marrow
- B symptoms
  - Fever
  - Night sweats
  - Weight loss
Hodgkin Lymphoma

- Histological subtypes
  - Nodular sclerosis
  - Mixed cellularity
  - Lymphocyte depletion
  - Lymphocyte rich classical
  - Lymphocyte predominance

- Surgery does not have major role
Hodgkin Lymphoma

- Chemotherapy with radiotherapy is standard approach
  - Doxorubicin
  - Bleomycin
  - Vincristine
  - Etoposide
  - Prednisone
  - Cyclophosphamide
- Event-free survival is 80-90%
Lymph Node Biopsy Indications

- Lymph node >2.5 cm in size
- Lymph nodes that are enlarging
- Lymph nodes that have not diminished after 5-6 weeks or normalized after 10-12 weeks
- Lymph nodes associated with abnormal chest radiograph
Rhabdomyosarcoma

- Most common sarcoma in children
- Associated with Li-Fraumeni syndrome and neurofibromatosis
- 40% of cases involve head and neck
  - Parameningeal site (infratemporal and pterygopalatine fossae, middle ear, paranasal sinus, and nasopharynx)
  - Orbit
  - Other (scalp, cheek, parotid gland, oral cavity, oropharynx, and larynx)
- Local invasion of bone, meninges, and brain
- Metastatic spread to lung and bone
Rhabdomyosarcoma

- Histological subtypes
  - Embryonal
  - Alveolar
  - Botryoid
  - Spindle cell
- Surgery or radiotherapy for local control
Rhabdomyosarcoma

- Chemotherapy for systemic control
  - Vincristine
  - Actinomycin
  - Cyclophosphamide
  - Irinotecan
  - Doxorubicin
  - Ifosfamide
  - Etoposide

- Event-free survival is 25-86%
Neuroblastoma

- Most common extracranial tumor in infants
- 3% of cases involve neck
  - Horner syndrome
- Local invasion of cervical lymph node
- Metastatic spread to distant lymph node, liver, bone, bone marrow, and skin
- May cause paraneoplastic syndrome
  - Opsoclonus-myoclonus syndrome
  - Catecholamine secretion
  - Vasoactive intestinal peptide secretion
Neuroblastoma

• Surgery or radiotherapy for local control
• Chemotherapy for systemic control
  – Cyclophosphamide
  – Doxorubicin
  – Vincristine
  – Cisplatin
  – Carboplatin
  – Etoposide
• Event-free survival is 40-95%
Nasopharyngeal Carcinoma

- More common in children older than 10 years of age
- More frequent in North Africa and Southeast Asia
- Associated with Epstein-Barr virus
- Arises in nasopharynx
- Local invasion of nose, mouth, pharynx, skull base, and cervical lymph node
- Metastatic spread to lung, liver, and bone
- Advanced disease at diagnosis is common
Nasopharyngeal Carcinoma

- Histological subtypes
  - Squamous cell carcinoma (type 1)
  - Nonkeratinizing carcinoma (type 2)
  - Undifferentiated carcinoma (type 3)
- Surgery often has limited role
- Chemotherapy with radiotherapy is standard approach
  - Cisplatin
  - Fluorouracil
  - Methotrexate
  - Bleomycin
- Event-free survival is 77-84%
Esthesioneuroblastoma

- More common in adolescents
- More common in boys
- Arises in nasopharynx
- Local invasion of orbit, sinus, skull base, and cervical lymph node
- Metastatic spread to lung, bone, and bone marrow
- Advanced stage at diagnosis is common
Esthesioneuroblastoma

- Distinct from primitive neuroectodermal tumor
- Surgery with radiotherapy is standard approach
- Chemotherapy may improve outcomes
  - Vincristine
  - Doxorubicin
  - Cyclophosphamide
  - Ifosfamide
  - Etoposide
- Event-free survival is 60%
Thyroid Carcinoma

- More common in children older than 15 years of age
- More common in girls
- Part of multiple endocrine neoplasia syndrome type 2
- Arises in neck
- Local invasion of cervical lymph node
- Metastatic spread to lung and bone
- Open biopsy may be more diagnostic than fine needle aspiration
Thyroid Carcinoma

• Histological subtypes
  – Papillary
  – Follicular
  – Medullary
  – Anaplastic

• Surgery with radioactive iodine therapy and thyroxine replacement is standard approach

• Event-free survival is 61-67%
Other

- Oral tumor
  - Lymphoma
  - Sarcoma
  - Mucoepidermoid carcinoma
  - Squamous cell carcinoma

- Salivary gland tumor
  - Mucoepidermoid carcinoma
  - Acinic cell carcinoma
  - Adenocarcinoma
  - Undifferentiated carcinoma
  - Rhabdomyosarcoma
Other

• Laryngeal tumor
  – Rhabdomyosarcoma
  – Squamous cell carcinoma

• Midline carcinoma