## Adult Cancers

<table>
<thead>
<tr>
<th>Women</th>
<th>Men</th>
</tr>
</thead>
<tbody>
<tr>
<td>Breast Cancer</td>
<td>182,800</td>
</tr>
<tr>
<td>Lung Cancer</td>
<td>74,600</td>
</tr>
<tr>
<td>Colon Cancer</td>
<td>50,400</td>
</tr>
<tr>
<td>Uterine Cancer</td>
<td>36,100</td>
</tr>
<tr>
<td>Ovarian Cancer</td>
<td>23,100</td>
</tr>
<tr>
<td>Melanoma</td>
<td>20,400</td>
</tr>
<tr>
<td>Bladder Cancer</td>
<td>14,900</td>
</tr>
<tr>
<td>Pancreatic Cancer</td>
<td>14,600</td>
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<tr>
<td>Prostate Cancer</td>
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<tr>
<td>Lung Cancer</td>
<td>89,500</td>
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<tr>
<td>Colon Cancer</td>
<td>43,400</td>
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<tr>
<td>Bladder Cancer</td>
<td>38,300</td>
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<tr>
<td>Melanoma</td>
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</tr>
<tr>
<td>Kidney Cancer</td>
<td>18,800</td>
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<tr>
<td>Pancreatic Cancer</td>
<td>13,700</td>
</tr>
<tr>
<td>Stomach Cancer</td>
<td>13,400</td>
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</table>
Declaration

I have nothing to declare
## Cancer in Children

<table>
<thead>
<tr>
<th>Type</th>
<th>% Total</th>
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<tbody>
<tr>
<td>Leukemia</td>
<td>30</td>
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<tr>
<td>Brain tumors (meduloblastoma)</td>
<td>25</td>
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<tr>
<td>Lymphoma</td>
<td>15</td>
</tr>
<tr>
<td>Neuroblastoma</td>
<td>8</td>
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<tr>
<td>Sarcoma</td>
<td>7</td>
</tr>
<tr>
<td>Wilms’ tumor</td>
<td>6</td>
</tr>
<tr>
<td>Osteosarcoma</td>
<td>5</td>
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<tr>
<td>Retinoblastoma</td>
<td>3</td>
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<tr>
<td>Liver tumors</td>
<td>1</td>
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</tbody>
</table>
Pediatric Tumors
Neuroblastoma
Epidemiology

Most common extracranial solid tumor in children
Prevalence is 1 per 7,000 live births
Approximately 550 new cases in the United States per year
Median age is 22 months
CT findings include small calcifications, vessel encasement, displacement of other organs and involvement of the neuroforamen
Current Management of Neuroblastoma

Presentation

Multiple manifestations depending on location and presence of paraneoplastic syndromes

50 - 75% present with an abdominal mass

25 % present with hypertension secondary to catecholamine secretion (patients often look sick)

Thoracic tumors may present with Horner’s syndrome (ptosis, miosis and enophthalmos)
Current Management of Neuroblastoma

Presentation

Anatomic sites of origin (autonomic nervous system)

- Adrenal 50%
- Mediastinal 22%
- Paraspinal 20%
- Pelvic 4%
- Neck 4%
Current Management of Neuroblastoma

Cellular classification

Neuroblastoma is one of the small blue round cell neoplasms

Three classic histologic patterns

- Neuroblastoma
- Ganglioneuroblastoma
- Ganglioneuroma

Can mature into benign tumor
Current Management of Neuroblastoma

Neuroblastoma
Neuroblastoma

*Staging System ~ The INSS*

**Stage 1:** localized tumor with complete gross excision, with or without microscopic residual disease; representative ipsilateral lymph nodes negative for tumor microscopically (nodes attached to and removed with the primary tumor may be positive).

**Stage 2A:** localized tumor with *incomplete gross excision*; representative ipsilateral nonadherent lymph nodes negative for tumor microscopically.

**Stage 2B:** localized tumor with or without complete gross excision, with ipsilateral nonadherent lymph nodes *positive for tumor*. Enlarged contralateral lymph nodes must be negative microscopically.
Neuroblastoma

*Staging System ~ The INSS*

**Stage 3:** Unresectable unilateral tumor infiltrating across the midline, with or without regional lymph node involvement; or localized unilateral tumor with contralateral regional lymph node involvement

**Stage 4:** any primary tumor with dissemination to distant lymph nodes, bone, bone marrow, liver, skin, and/or other organs (except as defined for stage 4S).

**Stage 4S:** localized primary tumor (as defined for stage 1, 2A, or 2B), with dissemination limited to skin, liver, and/or bone marrow (limited to infants less than 1 year of age).
Current Management of Neuroblastoma

Biologic Staging

Serum markers

- Ferritin
- Neuron specific enolase
- LDH
- Urine- HVA and VMA

Genetic markers

- Hyperdiploid karyotype
- N-MYC amplification (>10 copies)
- Chromosome 1 deletion

Histology- very important in prognosis (Shimada classification)
Stage 1. Survival rate for this is 90%, or more

Stage 2. (A) Cancer is still in the area where it started, but ALL tumors seen at this stage cannot be taken out by surgery. There is a 75% - 90% survival rate in this stage.

Stage 2. (B) Positive Nodes: Survival rate at this stage is 50% - 70% in babies, and 80% in children over a year old.

Stage 3. The cancer has started to spread now. It will cross the spine to the other side of the body. It will be impossible to get all the tumors by surgery. Survival is 50% - 70% for babies, and 80% for children.

Stage 4. The cancer is getting to other parts of the body now. Bone, the marrow of the bone, liver are just a few of the body parts effected now. Survival rates are now beginning to diminish somewhat. 50% - 80%.

Stage 4. (S) 10% - 40%. 
Neuroblastoma

After chemotherapy
Wilms’ Tumor

Residual right kidney
IVC tumor thrombus
Atrial tumor thrombus
Wilms’ Tumor

- most common malignant renal tumor of childhood
- 450-500 cases/year in USA
- diagnosed between the ages of 1 and 4 years
- responsible genes: WT1 (11p13), WT2 (11p15), 16q, 1p
- associated malformations (hemihypertrophy, aniridia) and Beckwith-Wiedemann syndrome
Wilms’ Tumor

- 1/3 of patients will present with microscopic hematuria
- Obstruction of the left renal vein can produce an acute varicocele due to gonadal vein obstruction
- There is no specific serum tumor marker for Wilms’
- CT and MRI are complimentary as may give more information of vessel involvement
Staging

Stage I
tumor limited to the kidney, intact capsule

Stage II
tumor extends beyond the kidney, complete excision

Stage III
not completely resectable
(infiltrated peritoneum, lymph node, IVC, atrium)

Stage IV
hematogenous metastases

Stage V
bilateral renal involvement
Histology

• Histology is as important as a factor as staging
• Favorable histology lesions: no anaplasia
• Unfavorable histology lesions: anaplasia
• Loss of heterozygosity 1p 16q - worse prognosis
Pre-operative Chemo: Controversial issues

- The value of preoperative chemotherapy?
- Whether pre-resection biopsy is indicated and if so, how is it best performed—may upstage a tumor from stage 1 to 3
- Additional risk to patients who could be cured with surgery alone
- 2 groups NWTS and SIOP
NWTSG recommendations

• primary nephrectomy
• adjuvant chemotherapy and/or irradiation
• preoperative chemotherapy only in case of
  – massive, nonresectable unilateral tumor
  – caval extension above the hepatic vein
  – bilateral tumors
Goal of preoperative therapy

- decrease surgical morbidity
- decrease the risk of intraoperative tumor rupture
- improve the outcome
- treat non-visible metastases very early
- the effect of the chemotherapy serves as a prognostic factor
SIOP studies

• preoperative chemotherapy:
  – reduces tumor rupture
  – induces favorable stage distribution
• recurrence-free survival is higher, though survival rates are the same
• postoperative irradiation can be reduced
• 4 weeks of treatment is optimal
• less surgical complications
<table>
<thead>
<tr>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
</thead>
<tbody>
<tr>
<td>provides easier operation</td>
<td>can be administered to a benign tumor</td>
</tr>
<tr>
<td>less perioperative complications</td>
<td>unreliable staging</td>
</tr>
<tr>
<td>prevents tumor rupture</td>
<td>toxic reactions</td>
</tr>
<tr>
<td>produces favorable staging</td>
<td></td>
</tr>
<tr>
<td>less irradiation</td>
<td></td>
</tr>
<tr>
<td>treats non visible metastases early</td>
<td></td>
</tr>
<tr>
<td>prognostic parameter</td>
<td></td>
</tr>
<tr>
<td>prolongs the recurrent free survival</td>
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</tbody>
</table>
Wilms’ Tumor - Survival

- Stage 1: 96%
- Stage 2: 92%
- Stage 3: 73%
- Stage 4: 73%
Other Renal Tumors

- Clear Cell Sarcoma - good survival
- Rhabdoid - rare and very poor survival
Hepatoblastoma
Epidemiology

- Most common primary malignant hepatic tumor in children.
- Rare after the age of three years; most occur between the age of one and two years (97% occur before age 10 years).
- Males are more affected than females 1.5:1
- Increased incidence in FAP, Beckwith-Wiedemann, hemihypertrophy
- They are found in otherwise normal liver tissue—unlike hepatocellular carcinoma which usually occurs in cirrhotic liver
Presentation

- Abdominal distention/fullness
- RUQ mass
- Constitutional symptoms of fever, weight loss
- Jaundice
- Precocious puberty
Diagnostic Evaluation

• Laboratory
  – CBC- thrombocytosis
  – LFT- elevation in alkaline phosphatase
  – Alpha Fetoprotein, B- hCG
  – Hepatitis Panel (B and C)
Non cirrhotic liver
Predilection for right lobe
Tan, lobulated
Pseudocapsule with areas of focal necrosis
Hepatoblastoma

Pathology
Pathology

• Pure Epithelial Cell Type - may have a better prognosis

• Mesenchymal Cell Type

• Undifferentiated
Hepatoblastoma

Pathology

Histologically the tumors may be pure *epithelial*, or, less commonly, *mixed* (with mesenchymal elements).
Staging

• **Stage I**
  – No metastases, tumor completely resected

• **Stage II**
  – No metastases, tumor grossly resected with microscopic residual disease (i.e., positive margins); or tumor rupture, or tumor spill at the time of surgery

• **Stage III**
  – No distant metastases, tumor unresectable or resected with gross residual tumor, or positive lymph nodes

• **Stage IV**
  – There are distant metastases regardless of the extent of liver involvement
Prognosis

• Stage 1 and 2 - excellent 5 yr survival (80-90%)
• With stage 3 disease - preoperative chemotherapy will render most resectable
• With use of chemotherapy plus surgery long term disease free survival has increased from 20-30% to 80%
Teratoma; pancreas
Teratoma

- From the latin teraton- meaning monster
- Arise from pluripotent cells and contain a wide variety of tissues foreign to the site from which they arise
- Often are benign, but may contain malignant elements
- Contain all three germ cell layers- neural tissue, skin and intestinal tissue
- Most frequent location: sacrococcygeal, ovary, chest, testicle, retroperitoneum
Teratoma

• Frequently are congenital tumors, but can present at any age
• Very large SCT can produce hydrops and subsequent fetal demise
• Histology: mature, immature and malignant
• Malignant- most common is yolk sac variety
• Can produce beta-HCG and produce virilization
• Produce alpha-fetoprotein- follow levels to look for recurrence
Teratoma

Sacrococcygeal Teratoma

- Type 1 – most common
- Type 4 can present later in life with history of constipation - important to feel posterior when doing rectal exam
- Must remove coccyx to prevent recurrence
Teratoma
Teratoma
Teratoma
Teratoma
Teratoma
Teratoma
Teratoma
Neuroblastoma

- Is the most common extracranial solid tumor of childhood—most often arising from the adrenal gland
- Often presents with advanced disease
- Has small calcifications, encases vessels and often invades the neuroforamen
- Secretes vasoactive substances (catecholamines) and patients often appear sick
- Prognosis: tumor biology (i.e. N-myc) is as important as anatomic stage
- Can differentiate into benign ganglioneuroma
Wilm’s Tumor

- Arises from the kidney, associated with other syndromes—hemihypertrophy and Beckwith-Wiedemann syndrome
- Is malignant, but has an excellent prognosis
- Anatomic stage is most important prognostic determinant
- Can be treated with pre-operative chemotherapy if originally unresectable
Hepatoblastoma

• Most common primary malignant hepatic tumor in children.
• Rare after the age of three years;
• Increased incidence in FAP, Beckwith-Wiedemann, hemihypertrophy
• Not associated with cirrhosis
• Long term survival only with complete resection
Teratoma

- Most often a benign tumor
- Contains mature ectopic tissues
- Most frequent congenital tumor
- Characterized by large calcifications and heterotopic appearance
- For SCT must resect the coccyx and follow AFP for signs of recurrence
Survivorship