1. In the International Neuroblastoma Risk Group Staging System (INRGSS), what is staging of localized tumors based on?

   A. Age at presentation and clinical symptomatology
   B. Physical examination and laboratory values
   C. Presence or absence of imaging defined risk factors
   D. Surgical resectability and lymph node assessment

**Correct Answer:** C. Presence or absence of imaging defined risk factors

**Rationale:** In the International Neuroblastoma Risk Group Staging System (INRGSS), staging of localized tumors is based on the presence or absence of imaging defined risk factors. Patient age at presentation is not used for staging of localized tumors, although it does play a role in differentiating stage M and stage MS disease. Clinical symptomatology, physical examination, and laboratory values are not used for disease staging. Surgical resectability and lymph node assessment are not used in the International Neuroblastoma Risk Group Staging System (INRGSS), but are instead used for localized tumor staging in the International Neuroblastoma Staging System (INSS).

**References:**


2. Based on the MIBG scan shown, what INRGSS stage of disease does this 2-year-old child have?

   A. Stage L1
   B. Stage L2
   C. Stage M
   D. Stage MS

**Correct Answer:** C. Stage M

**Rationale:** The MIBG scan in this 2-year-old child shows focal uptake in the primary left suprarenal mass. Abnormal MIBG uptake is also seen in distant metastatic sites within the skeleton (including the right lateral orbit, spine, pelvis, bilateral proximal humeri, and bilateral proximal femora). The answer is stage M disease, which is defined as distant metastatic disease (except as defined for stage MS). The answer cannot be Stage L1.
or L2 disease given the presence of distant metastases. Stage MS disease is not correct because of the patient’s age (greater than 18-months), as well as the skeletal metastases visible on the MIBG scan.

References:


**Image Defined Risk Factors**

*Andrew T. Trout, MD*

3. Which of the following is correct about Image Defined Risk Factors (IDRFs):

   A. Number of IDRFs determines disease stage
   B. Presence of IDRFs determines disease stage
   C. IDRFs determine whether tumors are resected
   D. MIBG scans can be used to define IDRFs

**Correct Answer:** B. Presence of IDRFs determines disease stage

**Rationale:** The presence (or absence) of IDRFs defines neuroblastoma stage. The number of IDRFs does not impact staging. While IDRFs relate to resectability, the presence of IDRFs does not define whether a tumor will be resected or not. IDRFs must be defined by MRI or CT. MIBG is in important part of disease staging but cannot be used to define IDRFs.

References:


4. Which of the following is an Image Defined Risk Factor?

   A. Lung nodule(s)
   B. Liver nodule(s)
   C. Tumor encasing the superior mesenteric artery origin
   D. Bone marrow disease

**Correct Answer:** C. Tumor encasing the superior mesenteric artery origin

**Rationale:** Lung nodules, liver nodules and bone marrow disease are all findings of distant metastatic disease (which have implications on staging) but are not IDRFs. Vascular encasement is an IDRF.

References:

5. With regard to the revised International Neuroblastoma Response Criteria (INRC) one of the following statements is true:

A. The criteria do not apply to phase I and II studies
B. Functional imaging is omitted from the criteria
C. The INRC only apply to patients enrolled in COG trials in North America
D. The revised INRC will incorporate the RECIST guidance and specific nuclear medicine modalities to define tumor response

**Correct Answer:** D. The revised INRC will incorporate the RECIST guidance and specific nuclear medicine modalities to define tumor response

**Rationale:** The revised INRC will be applied worldwide in all cooperative pediatric oncology trials, including phase I and II studies. Functional nuclear medicine imaging with metiodobenzylguanidine (MIBG) and FDG PET-CT scanning are included in the response criteria. The primary tumour need only be measured in one dimension (in any orthogonal plane) as per the RECIST 1.1 guidance.

**References:**


6. Regarding neuroblastic tumors in general, which of the following statement is true:

A. Neuroblastic tumors constitute a homogenous set of neoplasms
B. Neuroblastoma accounts for 12% of deaths associated with cancer in children younger than 12 years of age
C. Outcome is poor in infancy
D. Bone marrow disease is only assessed by bone marrow trephine and biopsy
Correct Answer: B. Neuroblastoma accounts for 12% of deaths associated with cancer in children younger than 12 years of age

Rationale: Neuroblastoma, a cancer of the sympathetic nervous system, accounts for 12% of deaths associated with cancer in children younger than 12 years of age. It is a heterogeneous disease with nearly 50% of patients having a high-risk phenotype. The prognosis is best in the first year of life with some tumors showing spontaneous regression. Bone marrow disease can also be assessed by MIBG or FDG PET-CT scanning and by MRI.

References:


Clinical and Imaging Features

Ethan A. Smith, MD

7. In the study by Flynt, et al. what was a statistically significant difference between adrenocortical carcinoma and adrenal adenoma?

A. Larger size
B. Right sided tumor
C. Patient gender
D. Hormonal activity

Correct Answer: A. Larger Size

Rationale: In this study, size was the only statistically significant difference in the primary tumor (excluding the presence of metastatic disease) between malignant lesions (ACC) and adenoma. Hormonal activity, tumor side and gender are not predictors of benign versus malignant lesions.

Reference:

8. Which of the following has the highest risk of developing adrenocortical carcinoma?

A. DICER1 mutation  
B. Tuberous Sclerosis complex  
C. Rothmund-Thomson syndrome  
D. Li-Fraumeni syndrome  

Correct Answer: D. Li-Fraumeni syndrome  

Rationale: Patients with Li-Fraumeni syndrome, a cancer predisposition syndrome, are at increased risk for adrenal cortical carcinoma. DICER1 mutation is a risk factor for pleuropulmonary blastoma, cystic nephroma and ovarian tumors, not adrenal malignancy. Tuberous sclerosis patients are at risk for several benign abdominal lesions (mostly renal) and a CNS abnormalities, but not for adrenal cortical carcinoma. Rothmund-Thomson syndrome is a syndrome primarily affected the skin, eyes and GI tract. Patients are at increased risk for osteosarcoma and skin cancer. There is no increased risk of adrenal malignancy associated this syndrome.

Reference:

Imaging and Clinical Features
Andrew T. Trout, MD

9. The most common location for pheochromocytoma in the pediatric/young adult population is:

A. The adrenal gland  
B. The organ of Zuckerkandl  
C. Carotid bifurcation  
D. Para-aortic thoracic  

Correct Answer: A. The adrenal gland  

Rationale: The adrenal gland is the most common location for pheochromocytoma in children (as in adults). Pheochromocytomas can occur in the other listed areas but are less common.

References:
10. Which imaging study is likely to be LEAST useful in the search for suspected pheochromocytoma?

A. I-123 MIBG  
B. F-18 FDG PET  
C. Whole body MRI  
D. Abdominal ultrasound

Correct Answer: D. Abdominal ultrasound

Rationale: MIBG, FDG-PET and whole body MRI have all been described as imaging modalities to search for occult Pheochromocytoma in patients with either symptoms or risk factors for pheochromocytoma. While pheochromocytoma are most commonly localized in the adrenal glands, ultrasound of the abdomen will miss small adrenal tumors, may miss extraadrenal tumors and does not screen for tumors outside of the abdomen.

References: