Neuroblastoma: Image Defined Risk Factors

Hervé Brisse, MD, PhD

1. 3.5-year-old boy with right lumbar paravertebral dumbbell ganglioneuroblastoma. How many Image Defined Risk Factors (IDRF) can you identify on these pictures?
   A. No IDRF
   B. Two IDRF
   C. Three IDRF
   D. Five IDRF
   E. Six IDRF

Correct Answer: C

References

Rationale
IDRF are surgical risk factors detected on imaging that make total tumor excision risky or difficult at diagnosis.
✓ IDRF 1: spinal canal invasion > 1/3 in axial plane
✓ IDRF 2: infiltration of the diaphragm
✓ IDRF 3: multicompartamental extension (i.e., abdomen + mediastinum)
   • Contact with renal pedicle (not shown) = additional IDRF
   • Abdominal aorta and IVC displaced but not encased (not IDRF)
   • Right kidney displaced but not invaded (not IDRF)

Neuroblastoma: INRGSS

Susan Cohn, MD

1. The survival of children with neuroblastoma is associated with:
   A. Image defined risk factors
   B. Amplification of the c-myc oncogene
   C. Translocation of the ALK-EML4 genes
   D. BRCA1 mutations
   E. Family history of neuroblastoma

Correct Answer: A

References
1. I-131 MIBG therapy for neuroblastoma:
   A. May be considered in all high risk patients based on results of the bone scan and MRI
   B. Can be performed in any hospital that performs diagnostic MIBG scans
   C. Requires special shielding in the room and education for all staff who may come in contact with the patient
   D. Can only be given once

   Correct Answer: C

References

Rationale
Prior to considering I-131 MIBG therapy in a patient with neuroblastoma, the tumor must show MIBG-avidity. Therefore, a diagnostic I-123 or I-131 MIBG scan must be performed and if there is MIBG-avidity, therapy can be considered. The results of the bone scan and MRI cannot assess this.

Performing I-131 MIBG therapy requires a complex coordination of care by the entire team treating the patient. This includes cooperation and education of the radiology/nuclear medicine department, radiation safety, oncology, nursing, child life, house staff, the code team and even dietary. Safety of the staff and all hospital members is important.

Prior to starting a MIBG therapy program, radiation safety should be consulted to assess the room and the amount of shielding required to safely house the patient on a ward where other patients are staying. Ideally the patient should be in a corner room with outside walls. Shielding is based on both occupancy and exposure rate.

This shielding requirements should be determined by a qualified radiation physicist and can be assessed based on the following calculations:

Exposure Rate: \( DR = \Gamma \times A/d^2 \)

\( \Gamma = 7.64 \times 10^{-5} \text{ mSv MBq per m}^2 \text{ h} \)

\( \text{Pb} = \text{HVL}/\ln(2) \times \ln(\text{CL}/20) \)

10 cm concrete~ 0.6 cm Pb

I-131 MIBG therapy has been given as tandem therapy and in fact, has been shown to be effective in association with a tandem approach and rescue stem cell transplant, although not curative.
Retinoblastoma Imaging Update
Hervé Brisse, MD, PhD

1. 8-month-old girl with unilateral retinoblastoma.
MRI & ocular fundus.
Which of the following is CORRECT?
A. The tumor is mainly endophytic
B. Extra-scleral invasion is visible
C. Optic nerve is invaded
D. Anterior chamber is invaded
E. Tumor size/volume is a significant risk factor of post-laminar optic nerve invasion

Correct Answer: E

References

Rationale
There is a strong association between tumor size (or volume) and post-laminar optic nerve invasion.
A. Not Correct. Total retinal detachment is associated with exophytic tumors.
B. Not Correct: Extra-scleral invasion at diagnosis is rare (unless delayed diagnosis). The low signal intensity of the normal sclera is nicely visible in front of the tumor.
C. Not correct. The optic nerve is normal in size and signal and no abnormal enhancement is seen. The NPV of high-resolution MRI is high, allowing to rule out advanced optic nerve invasion before enucleation. The sensitivity of HR-MRI in normal-sized optic nerves is about 60%.
D. Not Correct. Isolated enhancement of the iris is a reactive process (neovascularization).

Pediatric Soft Tissue Sarcomas
Beth McCarville, MD

1. Regarding rhabdomyosarcoma, which of the following is considered a favorable site?
A. Extremity
B. Parameningeal
C. Middle ear
D. Prostate
E. Orbit

Correct Answer: E
References

SPECT-CT
Susan E. Sharp, MD

1. What is the left retroperitoneal uptake seen on SPECT/CT due to?
   A. residual retroperitoneal tumor
   B. skeletal metastatic disease
   C. physiologic renal activity
   D. physiologic adrenal activity
   E. physiologic bowel activity

   Correct Answer: D

Reference

Rationale
The SPECT/CT images show that the left retroperitoneal uptake is due to physiologic adrenal activity.

The CT scan shows a normal adrenal gland at the site of MIBG uptake. No residual tumor is seen in this region. The uptake is anterior to the kidney and therefore cannot be renal activity. The uptake is medial and posterior to bowel and is therefore not bowel activity. The uptake does not correspond to the spine or another bony structure and is therefore not a skeletal metastasis.

Hybrid Imaging in the Era of Personal Oncogenomics
Helen R. Nadel, MD, FRCPC

1. 18F-FDG uptake on PET/CT scan in patient with metastatic germ cell tumor is targeted to expressing;
   A. apoptosis
   B. angiogenic proliferation
   C. Glut-1 transporter
   D. SS receptor

   Correct Answer: C

References
(http://www.sciencedirect.com/science/article/pii/S0092867411001279)
Rationale
Glut-1 transporter is upregulated during cancer growth. Deoxy-glucose is a glucose analog that enters the cell using GLUT-1. Once within the cytoplasm, DG is phosphorylated to FG-6-phosphate but does not appear to be further metabolised in tumor cells expressing Glut-1.

A and B are in correct as expression of these processes on PET CT would be with other radiotracers. Apoptosis would be targeted with I-124 Annexin-V or Cu-64–annexin-V. Angiogenesis would be targeted with 18F-galacto-RGD peptide. D is incorrect as SSR-somatostatin receptor expression is for assessment of neuroendocrine tumors and uses Ga-68 DOTA radiotracers.

Thyroid Carcinoma - Imaging and Therapy
Marguerite T. Parisi, MD, MS

1. Which of the following is TRUE regarding differentiate thyroid cancer in children?
A. The incidence of thyroid nodules is higher in children than in adults.
B. The likelihood of a thyroid nodule harboring a malignancy is higher in children than adults.
C. Children with differentiated thyroid cancer present with less advanced disease than adults.
D. Prognosis for children with differentiated thyroid cancer is worse than that in adults.

Correct Answer: B

References:
2. La Franchi S. Inaugural Management Guidelines for Children with Thyroid Nodules and Differentiated Thyroid Cancer: Children Are Not Small Adults. Thyroid 2015; 25(7):713-715.

Rationale:
Correct Answer is B: While the incidence of thyroid nodules is lower in children than in adults, the percentage of thyroid nodules harboring cancer is much higher in children. In adults, approximately 5% of solitary thyroid nodules harbor malignancy compared to between 14%-61% (average 26%) in children.

A is incorrect.
The incidence of thyroid nodules is greater in adults than in children. Thyroid nodules occur in 1%-5% of children compared to 10% in young adults and >50% in adults aged 60 years and older.
C is incorrect. Unlike adults, children with differentiated thyroid cancer (DTC) typically present with advanced disease at diagnosis. Extensive regional lymph node involvement is present in 60%-80% of children with DTC compared to 30% of adults. There is also a higher incidence of distant metastases in children with DTC compared to adults.

D is incorrect. Despite the fact that children with DTC present with more advanced disease than their adult counterparts, overall prognosis is excellent and better than that in adults.

Pediatric Oncologic Interventions - American Approach

Kamlesh Kukreja, MD

1. Largest organization dedicated to pediatric and adolescent cancer research in the US?
   A. Plowing for Cancer
   B. Cancer spankers
   C. Tumorators
   D. Answer for cancer
   E. Children’s Oncology Group

Correct Answer: E

Reference
https://www.childrensoncologygroup.org/

Rationale
The Children’s Oncology Group (COG), a National Cancer Institute supported clinical trials group, is the world’s largest organization devoted exclusively to childhood and adolescent cancer research. The COG unites more than 9,000 experts in childhood cancer at more than 200 leading children’s hospitals, universities, and cancer centers across North America, Australia, New Zealand, and Europe in the fight against childhood cancer.

Pediatric Oncologic Interventions - European Approach

Fernando Gómez Muñoz

1. What are the potential advantages of transarterial chemoembolization using doxorubicin for the treatment of non-surgical candidates with liver hepatoblastoma?
   A. Higher intratumoural concentration
   B. Higher grade of tumour necrosis
   C. Reduced toxicity due to lower systemic exposure
   D. Higher risk of cardiac toxicity
   E. A, B and C are correct

Correct Answer: E

References

Rationales:
Answer is E: The three first answers are right
Option A is correct: Loadable beads allow direct intra-arterial infusion of doses of doxorubicin as high as 150 mg. Indeed, almost all the tumor vascularization depends on the artery
Option B is correct: Higher grade of tumour necrosis is achieved as both, anoxia and cytotoxic effect of doxorubicin favour tumoral lysis
Option C is correct: Doxorubicin is released from the beads along two weeks and remains in the liver mainly.
Option D is not correct: The risk of cardiac toxicity is diminished as systemic exposure to the drug is low.

**Limitations of RECIST in Pediatric Oncology**

*Kieran McHugh, MD*

1. With regard to version 1.1 of the Response Evaluation Criteria in Solid Tumors (RECIST) guidance, one of the following statements is true:
   A. Five lesions per organ should be measured
   B. A lymph node with a short axis diameter of ≥10 but ≤15mm in short axis is considered normal
   C. Non-target lesions must be measured on each occasion
   D. A lymph node with a diameter of ≥15mm in short axis is considered pathological
   E. Leptomeningeal disease is measurable disease

   **Correct Answer: C**

**Reference**

New response evaluation criteria in solid tumors: revised RECIST guideline (version 1.1)

**Rationale**

*Option 4 is correct.* A lymph node with a short axis diameter of ≥15mm is measurable disease.

*Option 1 is not correct.* Two lesions per organ, representative of all involved organs, should be measured (these are deemed ‘target lesions’).

*Option 2 is not correct.* A lymph node with a short axis diameter of ≥10 but ≤15mm is considered pathological, but is recorded as a non-target lesion.

*Option 3 is not correct.* Non-target lesions are noted on each occasion but they are not measured.

*Option 5 is not correct.* Leptomeningeal disease, like pleural effusions and ascites, is non-measurable disease.

**Pediatric Lymphoma-PET Imaging**

*Steve Yoon-Ho Cho, MD*

1. The Deauville criteria or 5-point scale for FDG PET Lymphoma Staging and Response uses which reference organs
   A. Mediastinal Blood Pool and Brain
   B. Brain and Liver
   C. Liver and Mediastinal Blood Pool
   D. Muscle and Liver

   **Correct Answer: C**

**References**

**Hepatoblastoma Update**  
*Alexander J. Towbin, MD*

1. The staging system used for risk stratification in hepatoblastoma in current collaborative trials is called:
   A. Evans  
   B. PRETEXT  
   C. POSTTEXT  
   D. SIOPEL  

**Correct Answer: B**

**Reference**  

**Rationale**  
The PRETEXT or PRETreatment EXTent of disease staging system was designed by SIOPEL for use in their trials. Currently all collaborative groups are using this scoring system.

**Advances in Pediatric Bone Tumor Assessment**  
*Philippe Petit, MD*

1. Concerning long bone osteosarcoma in children which one is correct?
   A. The MR exploration must be only focused on the tumoral bone lesion  
   B. There are more false negative results to define the intraosseous tumor extension with the SE T1 sequence than with the STIR sequence.  
   C. CT scan of the lesion must be added to the MR exploration.  
   D. On MR, the feathery edema like pattern around the well-limited bulky lesion must not be considered as potential tumor.  
   E. DWI performed at mid-course of chemotherapy is a solid technique to detect early the poor responders.

**Correct Answer: B**

**References**

PET-MR
Geetika Khanna, MD, MS

1. Which of the following is TRUE regarding PET-MR compared to PET-CT?
   A. Higher radiation dose
   B. Higher pulmonary resolution
   C. Better attenuation correction of cortical bone
   D. Better soft tissue contrast

Correct Answer: D

References:

Rationale
A - Incorrect. Current data suggests that with PET-MR radiation dose can be decreased by 30-70% in children compared to PET-CT. B - Incorrect. Detection of sub centimeter pulmonary nodules is limited by PET-MR compared to PET-CT. C - Incorrect. Since cortical bone is hypointense on most MR sequences, attenuation correction of cortical bone is challenging with standard Dixon based methodology used in MR attenuation correction. D - Correct. Better soft tissue contrast due to T2 prolongation, diffusion restriction and enhancement properties of most tumors allows for improved soft tissue contrast on PET-MR compared to PET-CT. This aids in detection of visceral lesions.

Late Effects
Eline Deurloo, MD, PhD

1. Figure 1 shows an ultrasound image of the liver of a 16 year old girl, with a history of nephroblastoma at the age of 4. An ultrasound of the abdomen was performed for increasing abdominal pain. Figure 2 shows some of the MRI images.

Which one of the following is the MOST likely etiology of the lesion marked by the callipers?
A. Cyst
B. Hemangioma
C. Nephroblastoma metastasis
D. FNH / regeneration nodule
E. Hepatocellular carcinoma

Correct Answer: D

References:

Rationale
Option A is not correct. The lesion is solid, not cystic. Option B is not correct. Although the lesion is partly hyperechoic on US, it does not show the typical features of hemangioma on contrast-enhanced MRI. Option C is not correct. The likelihood of nephroblastoma metastasis occurring 12 years after the initial diagnosis is very low.
Furthermore, the imaging characteristics (especially the MRI images) do not show suspicious characteristics. **Option D is correct.** There are multiple lesions in the liver, many years after a treated nephroblastoma. Some of the lesions show a typical spoke wheel appearance on MRI. **Option E is not correct.** There is no increased risk to develop hepatocellular carcinomas after therapy for nephroblastoma. Furthermore, hepatocellular carcinoma is usually a solitary lesion.

2. A coronal T1 weighted image of the pelvis is shown of a 14 year old patient with an oncologic disease (fig.3).

What is the most likely diagnosis of the bone marrow abnormalities that are shown?

A. Bone infarcts due to treatment for leukemia
B. Metastases
C. Fatty replacement due to radiotherapy on the pelvis
D. Osteochondromas after radiotherapy
E. Normal bone marrow signal intensity for a 14 year old.

**Correct Answer: A**

Reference

Rationale
The correct answer is A: there are multiple lesions in the ilium and sacrum that are sharply delineated and hyperintense on T1. This is the typical appearance of bone infarcts as a result of treatment for leukemia. **Answer B is not correct.** The lesions are sharply delineated and almost symmetrical, which is very unlikely for metastases. Also, the lesions are hyperintense on T1 weighted images, whereas metastases would be hypointense. **Answer C is not correct.** Fatty replacement due to radiotherapy on the pelvis occurs in the whole irradiated area, and not in focal areas. Also, the fatty replacement is not delineated by low signal intensity margins. **Answer D is not correct,** because the lesions are within the bone, hence they are not osteochondromas. Furthermore, they do not show features of osteochondromas. **Answer E is not correct** because there are well demarcated lesions in the bone, which is not normal.

**Imaging in Bone Marrow Transplant Patients**
*Ethan A. Smith, MD*

1. What is the most commonly affected organ or organ system in acute graft-versus-host disease?
A. Liver
B. Kidney
C. Skin
D. Lung
E. Pancreas

**Correct Answer: C**

Reference

Rationale
The skin is affected in approximately 81% of cases of GVHD and is usually the first organ involved. The characteristic finding is a pruritic, maculopapular rash. The liver is affected in approximately 50% of patients. The kidney, lung and pancreas are not typically involved in acute GVHD.