Imaging of Melanoma
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1. Which of the following statements is true regarding pediatric malignant melanoma?

A. The main predictor of outcome is the stage of disease at the time of diagnosis
B. Melanoma is a disease seen only in Caucasians
C. Spitzoid melanomas who lack the TERT promoter mutation have a poorer prognosis
D. The majority of congenital melanocytic nevi undergo malignant degeneration to melanoma

Correct Answer: A. The main predictor of outcome is the stage of disease at the time of diagnosis

Rationale: Disease stage at the time of diagnosis is the main predictor of outcome. The lower the stage, the better the outcome. Option B is not correct. Melanoma is more common in Caucasians but can develop in any race. It is 5-times more common in Caucasians than Hispanics and 20-times more common in Caucasians than African Americans. Option C is not correct. Spitzoid melanomas who lack the TERT promoter mutation have a much better prognosis than those with the TERT promoter mutation. The latter mutation is associated with aggressive behavior and a higher tendency for metastasis and death from disease. Option D is not correct. Only about 5-10% of congenital melanocytic nevi become melanoma.

References:


Pleuropulmonary Blastoma and DICER1 Syndrome
R. Paul Guillerman, MD

2. Which of the following raises the likelihood that a lung cyst in a child represents a cystic congenital lung malformation rather than a cystic pleuropulmonary blastoma (PPB)?

A. Cyst multifocality
B. Spontaneous pneumothorax
C. Presence on mid-2nd-trimester prenatal ultrasound
D. Family history of lung cysts

**Correct Answer:** C. Presence on mid-2nd-trimester prenatal ultrasound

**Rationale:** Since cystic PPBs and cystic congenital lung malformations are very similar in appearance on imaging but the decision regarding surgical management of these lesions may differ, particularly in an asymptomatic child, it is important to recognize features that favor one versus the other. Congenital cystic lung malformations are often manifest on mid-2nd-trimester prenatal ultrasound exams, while cystic PPBs are typically first identified postnatally or on third trimester prenatal ultrasound exams. Cyst multifocality, spontaneous pneumothorax, or a family history of lung cysts or other DICER1 syndrome-related conditions increases the likelihood that a lung cyst in a child represents a cystic PPB rather than a cystic congenital lung malformation.

**References:**


**Desmoplastic Small Round Cell Tumor: What the Radiologist Needs to Know**

**Anita P. Price, MD, FACR**

3. Desmoplastic small round cell tumor is associated with which of the following gene fusions:

   A. EWS - ATF1
   B. EWS - WT1
   C. TFE3 - ASPL
   D. SSX1 – SYT

**Correct Answer:** B. EWS - WT1

**Rationale:** Fusion of the Ewing sarcoma gene (EWS) with Wilms Tumor1 suppressor gene (WT1), resulting from a characteristic translocation (11;22) (p13;q12) translocation. Options A, C and D are incorrect. Clear cell sarcoma is associated with EWS-ATF1 gene fusion. Alveolar soft part sarcoma is associated with TFE3- ASPL gene fusion. Synovial sarcoma is associated with SSX1 – SYT.

**References:**

4. Axial and coronal CT images of a 7 yo male with no previous medical history. Which of the following is the least likely diagnosis?

A. Burkitt Lymphoma
B. DSRCT
C. Nephroblastomosis
D. Angiomyolipoma

Correct Answer: D. Angiomyolipoma

Rationale: Angiomyolipoma is an uncommon benign renal neoplasm in children containing mature adipose tissue, smooth muscle, and abnormal blood vessels. Varying amounts of fat are present. 80% patients with Tuberous Sclerosis have angiomyolipoma. 90% of angiomyolipomas which are not associated with Tuberous Sclerosis present as solitary unilateral lesions. The lesions in this child are bilateral and do not contain fat. The child has no significant prior medical history to suggest tuberous sclerosis. A. Burkitt lymphoma may involve the kidney as multifocal hypodense renal lesions. B. DSRCT can involve the kidney as hypodense lesions. A delayed right nephrogram raises the question of a pelvic mass causing distal ureteral obstruction. Axial image through the pelvis of same patient demonstrates large heterogeneous retrovesical pelvic mass with punctate calcification. Patient has pathologically confirmed DSRCT. C. Nephroblastomatosis can present as hypodense peripheral cortical lesions.

References:


**Ovarian Tumors of Childhood**
*Shailee V. Lala, MD*

5. The most common ovarian tumor in children is:

   - A. Sclerosing stromal tumor
   - B. Mature cystic teratoma
   - C. Dysgerminoma
   - D. Serous cystadenoma

**Correct Answer:** B. Mature cystic teratoma

**References:**


6. What percentage of ovarian dysgerminomas are bilateral?

   - A. 0-5%
   - B. 10-15%
   - C. 45-50%
   - D. 70-75%

**Correct Answer:** B. 10-15%

**References:**