Congenital Spine Anomalies
Hedieh Khalatbari, MD

1. Sagittal T2-W and T1-W images of a 4-month old with a spinal dysraphism are shown. This anomaly may be classified as a:
   A. open spinal dysraphism
   B. closed spinal dysraphism with subcutaneous mass
   C. closed spinal dysraphism without a subcutaneous mass, simple type
   D. closed spinal dysraphism without a subcutaneous mass, complex type

   **Correct Answer: B**

2. Sagittal T2-W and T1-W images of a 4-month old with a spinal dysraphism are shown. The arrows point to a
   A. Meningocele
   B. Cystocele
   C. Hydromyelia
   D. syringocele

   **Correct Answer: D**

3. Sagittal T2-W and T1-W images of a 4-month old with a spinal dysraphism are shown. The diagnosis is
   A. Lipomyelomeningocele
   B. terminal lipomyelocystocele
   C. myelomeningocele
   D. lipomyelocele

   **Correct Answer: B**

**Rationale for Questions 1-3**
- Syringocele (red arrows)
- Meningocele (green arrows)
- Hydromyelia (blue arrows)
- Subcutaneous lipoma (yellow arrowhead)

Diagnosis: terminal lipomyelocystocele
4. Which of the following statements is correct regarding spinal leptomeningeal metastases of intracranial tumors?
   A. CSF cytology and spine MRI results are always concordant
   B. CSF cytology results have been reported to be a better predictor of survival
   C. MRI spine results have been reported to be a better predictor of survival
   D. CSF cytology is no longer necessary given advancements in spine MRI

Correct Answer: C

Reference

5. Based on the findings on this post-contrast spine MRI, what should one of the next steps be for this post-operative medulloblastoma patient?
   A. MR CSF flow study
   B. Radiation therapy for spinal metastases
   C. Repeat spine MRI in several weeks
   D. Laminectomy

Correct Answer: C

Reference

6. Which of the following statements regarding pediatric spine tumors is correct?
   A. Ependymoma is the most common histology
   B. Spinal pain is an uncommon presenting symptom
   C. Intramedullary tumors account for about 1/3 of cases
   D. Extradural tumors are least common

Correct Answer: C

Reference
Imaging of Pediatric Spinal Injury
Wendy A. Cohen, MD

7. Which of the following is true of cervical spine injuries in children?
   A. The anatomic distribution across age groups is similar
   B. The incidence across age groups is similar
   C. The mortality rate is greater in younger children
   D. Incidence of spinal cord injury associated with spine fracture is greater in adolescents than in young children

   Correct Answer: C

Rationale
A. Incorrect - The anatomic level of the injury varies across age groups. Upper cervical spine injuries (C0-C4) are more frequent than lower spine injuries in children <9 yr. This is not true in children > 9 yr.
   
<table>
<thead>
<tr>
<th>Age Group</th>
<th>Incidence</th>
</tr>
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<tbody>
<tr>
<td>0-3</td>
<td>70%</td>
</tr>
<tr>
<td>4-9</td>
<td>74%</td>
</tr>
<tr>
<td>10-14</td>
<td>52%</td>
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<tr>
<td>15-18</td>
<td>40%</td>
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</table>

B. Incorrect - The incidence of spinal injury is 2-fold greater in preadolescents and 5-fold greater in adolescents than in younger patients
C. Correct
D. Incorrect

References

8. Which of the following statements is true in children with SCIWORA (spinal cord injury without radiologic abnormality)?
   A. The incidence of SCIWORA compared to fracture/dislocations in the same in all pediatric age groups.
   B. Patients with a diagnosis of SCIWORA with abnormal neurologic exam can have a normal MRI
   C. Prognosis for recovery of neurologic function is poor.
   D. Only occurs in the cervical spine. Does not occur in the thoracic spine

   Correct Answer: B
Rationale
A. Incorrect. Incidence of SCIWORA is higher in children < 9 yr.
B. Correct. Reports vary as to frequency of normal spinal cord by MRI with an abnormal neurologic examination (35%, 78%).
C. Incorrect Prognosis of recovery depends on the severity of injury at initial presentation.
D. Incorrect Approximately 13% of SCIWORA cases occur in the thoracolumbar spine.

References

9. Many factors are thought to predispose young children (<8 yr.) to upper cervical spine injury when compared to older children. The following statement is not a characteristic of cervical spines in younger children
   A. Elasticity of the ligaments compared to the inelasticity of the spinal cord
   B. Fulcrum of rotation at C2-3 in younger children and at C4-5 in older children
   C. Anteriorly wedged vertebrae
   D. Vertically oriented facet joints

   **Correct Answer: D**

Rationale
A. Correct
B. Correct.
C. Correct.
D. Incorrect Facet joints are horizontally oriented in younger children.

Reference

Pediatric Brain Tumors: Update
*Tina Young Poussaint, MD, FACR*

10. The diffuse intrinsic brainstem glioma (now known as diffuse midline glioma) accounts for what percentage of brainstem gliomas?
   A. 10%
   B. 25%
   C. 35%
   D. 80%

   **Correct Answer: D**

Rationale
The majority of brainstem gliomas in childhood are the diffuse intrinsic type characterized by expansion of the pons and T2 prolongation.
Reference

11. Which tumor type is associated with basal cell nevus syndrome?
   A. Astrocytoma
   B. Subependymal giant cell tumor
   C. Hemangioblastoma
   D. Medulloblastoma

   Correct Answer: D

Rationale
The correct answer is D. Basal cell nevus syndrome is a syndrome caused by mutations in the PTCH1 gene on chromosome 9. In this syndrome there are the development of basal cell carcinomas, keratocysts in the maxilla and mandible and medulloblastoma of the desmoplastic type due to aberrations in the sonic hedgehog pathway.

Answer A is incorrect: Astrocytomas have an increased association with neurofibromatosis type 1, the most common of the phakomatoses. Tumors seen in these patients include neurofibromas and astrocytomas.

Answer B is incorrect: Subependymal giant cell astrocytoma is associated with tuberous sclerosis. These patients have subependymal nodules, white matter abnormalities, and cortical tubers as well.

Answer C is incorrect. Hemangioblastomas of the cerebellum, retina, and spine are associated with Von Hippel Lindau syndrome. Patients may also develop pancreatic cysts, islet cell tumors, pheochromocytomas, renal cysts, endolymphatic sac tumors, renal cell carcinoma, and epididymal cysts and cystadenomas.

References

12. Which genetic profile is associated with diffuse midline glioma?
   A. SMARCB1 tumor suppressor gene
   B. Sonic hedgehog pathway
   C. Wnt activated pathway
   D. K 27 M mutation in histone coding genes

   Correct Answer: D
Rationale
The correct answer is D. Diffuse midline gliomas have K27M mutations in the histone encoding genes of H3F3A or HIST1H3BC

A is Incorrect. This is the genetic aberration for atypical teratoid rhabdoid tumors

B is Incorrect. This pathway is associated with the molecular subtypes of medulloblastoma

C is Incorrect. This pathway is associated with the molecular subtypes of medulloblastoma

Reference

Pediatric Head and Neck Tumors
Hisham M. Dahmoush, MBBCh

13. What is the most common location of head and neck rhabdomyosarcomas?
   A. Carotid Space
   B. Buccal Space
   C. Masticator Space
   D. Paraspinal

   Correct Answer: C

Reference

14. What is the commonly affected gene in lipoblastoma?
   A. PLAG1
   B. GLUT1
   C. FOX01 to PAX7 fusion
   D. RB1

   Correct Answer: A

Reference

15. Regarding PET/MR assessment of head and neck tumors?
   A. Brown fat FDG uptake is most common in the spring
   B. Brown fat FDG uptake does not affect sensitivity of PET in identifying disease recurrence in the neck
   C. Echo-planar imaging (EPI) is the technique of choice for performing diffusion imaging in the neck
   D. Early increase in ADC values in malignant tumors during therapy on diffusion imaging suggests a favorable outcome

   Correct Answer: D
Reference

Metabolic Disease
Thierry A. G. M. Huisman, MD

16. Patterns of brain injury secondary to a metabolic disease as noted on MR imaging
   A. Allow to narrow down the differential diagnosis
   B. Increase the list of differential diagnosis
   C. Vary with the gender of the patient
   D. Vary with the applied magnetic field strength

Correct Answer: A

Reference

17. Inborn errors of metabolism
   A. Most frequently present during early pregnancy or at birth
   B. May present with a sudden neonatal death at 2-3 days of age
   C. Usually recover spontaneously
   D. Are usually linked to a static clinical picture over time

Correct Answer: B

Reference

18. The "Giant panda" on axial T2-weighted imaging of the mesencephalon suggests
   A. Tay syndrome
   B. Alexander disease
   C. Zellweger syndrome
   D. Wilson disease

Correct Answer: D

Reference