

SPR 2012 Annual Meeting – Neuroradiology Session
Susan Palasis, MD and Tina Young Poussaint, MD, Moderators

Pediatric Tumors: Brain

Tina Young Poussaint, MD

Which tumor type is associated with basal cell nevus syndrome?

- A. Astrocytoma
- B. Schwannoma
- C. Subependymal giant cell tumor
- D. Hemangioblastoma
- E. Medulloblastoma

Answer: E

Rationale:

The correct answer is E. 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: Schwannoma is associated with neurofibromatosis type 2 . Tumors found in NF-2 include multiple inherited schwannomas, meningiomas, and ependymomas.

Answer C 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 D 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:

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Pediatric Tumors: Spine

Andrea Rossi, MD

Which is the most common intramedullary tumor in the pediatric age group?

- A. Ependymoma
- B. Ganglioglioma
- C. Fibrillary Astrocytoma
- D. Pilocytic Astrocytoma
- E. Glioblastoma

Answer: D

Rationale:

Pilocytic astrocytomas account for 75% of all intramedullary tumors in the pediatric age group and typically affect children between 1 and 5 years of age.

Options A, B, C, and E are not correct. Intramedullary ependymomas are exceedingly rare in the pediatric age group outside the setting of neurofibromatosis type 2. Gangliogliomas are the second most common intramedullary tumor in the pediatric age group (15% of cases) and mostly affect children between 1 and 5 years of age, as do pilocytic astrocytomas. Fibrillary astrocytomas account for 7% and tend to occur in older children (around 10 years of age). Glioblastomas have only exceptionally been reported to occur in the spinal cord in children.

Reference:

Rossi A, Gandolfo C, Morana G, Tortori-Donati P. Tumors of the spine in children. *Neuroimaging Clin N Am* 2007; 17:17-35.

Pediatric Tumors: Head and Neck

Bernadette L. Koch, MD

Which of the following statement is true about infantile hemangiomas ?

- A. They do not respond to treatment with Propranolol.
- B. They are considered vascular malformations.
- C. They are usually visible at birth.
- D. They are Glut-1 negative.
- E. They are considered benign neoplasms.

Answer: E

Rationale:

Infantile hemangiomas are benign neoplasms, which usually undergo a proliferating phase followed by a spontaneous involution phase over a period of years. They typically are not present at birth but appear within the first few weeks of life.

Option A is not correct. Most hemangiomas are treated conservatively with expectant waiting. When treatment is necessary, options include intralesional steroids, systemic steroids and most recently there has been research supporting successful treatment with Propranolol.

Options B is not correct. Congenital vascular malformations include lymphatic malformations, venous malformations, arteriovenous malformations and arteriovenous fistulae.

Option C is not correct. Infantile hemangiomas are not typically present at birth. They appear within the first few weeks of life. Congenital hemangiomas are present at birth and are classified as rapidly involuting congenital hemangiomas (RICH) or noninvoluting congenital hemangiomas (NICH), based on their clinical behavior.

Option D is not correct. Glut-1 is a specific immunohistochemical marker that is expressed in all phases of hemangioma –proliferative, involuting and involuted phases. This marker is not present in vascular malformations.

References:

1. Mulliken JB and Glowacki J. (1982) Hemangiomas and vascular malformations in infants and children: A classification based on endothelial characteristics. *Plastic Reconstructive Surgery*; 69:412.
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Injury to the Immature Brain: Fetal Brain Injury

Orit A. Glenn, MD

Which of the following is not a cause of fetal brain injury?

- A. Congenital Infection
- B. Coagulopathy
- C. Mutations in collagen IV A1
- D. Cotwin demise
- E. In Vitro Fertilization

Answer: E

References:

1. Cheeran MC, Lokensgard JR, Schleiss MR. Neuropathogenesis of congenital cytomegalovirus infection: disease mechanisms and prospects for intervention. Clin Microbiol Rev. 2009 Jan;22(1):99-126.
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4. Jelin AC, Norton ME, Bartha AI, Fick AL, Glenn OA. Intracranial magnetic resonance imaging findings in the surviving fetus after spontaneous monochorionic cotwin demise. Am J Obstet Gynecol. 2008 Oct;199(4):398.e1-5.
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Fetal and Neonatal MR Sequences for Evaluation of Injury to the Immature Brain

Duan Xu, PhD

Which of the following pulse sequences is most sensitive for detection of subacute blood products?

- A. T1 SPGR
- B. Spin echo
- C. Gradient echo
- D. Diffusion

Answer: C

Rationale:

Gradient echo sequences with long TE are best for detection of local field inhomogeneities, especially in cases where ferritin and hemosiderin from subacute or chronic hemorrhage distorts the local magnetic field.

Option B: Spin echo uses RF pulses to rephase spins for imaging and mitigate T2* signal loss, and as a result does not prominently show local field distortions due to blood products.

Option A: T1 SPGR, although it is a gradient echo sequence that is sensitive to T2* effects, uses short values of TE for T1 contrast and is thus not as sensitive as option C.

Option D: Diffusion is acquired using spin-echo echo planar imaging, which is weighted by T2* effects but is dominated by T2 contrast, and has lower spatial resolution than option C.

References:

1. Tong KA, Ashwal S, Obenaus A, Nickerson JP, Kido D, Haacke EM. Susceptibility-weighted MR imaging: a review of clinical applications in children. *AJNR Am J Neuroradiol.* 2008 Jan;29(1):9-17. Epub 2007 Oct 9.
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Injury to the Immature Brain: Premature and Term Infants

A. James Barkovich, MD

Which of the following is NOT a significant factor in outcome due to brain injury in prematurely born infants?

- A. Damage to neurons in the subplate, just below the developing cortex
- B. Injury to periventricular and deep white matter
- C. Cerebral cortex infarctions
- D. Cerebellar hemorrhages
- E. Periventricular hemorrhagic infarctions

Answer: C

Rationale:

A. Damage to neurons in the subplate is almost certainly a cause of the cognitive and behavioral disorders found in about 50% of prematurely born neonates. The subplate is a region containing sparsely distributed neurons just below the cerebral cortex in the late second and early to mid third trimester. Thalamocortical axons enter the subplate after 24 postconceptional weeks (26 gestational weeks) and synapse with these neurons in the somatosensory, auditory, visual, and frontal cortex, forming actual circuits in the developing brain. Starting between 31 and 34 gestational weeks, these thalamocortical axons begin to move into the lower cortical plate and synapse in cortical layer IV, accompanied by a rapid growth of basal dendrites into layer III and pyramidal cells in layer V. Injury to the subplate at any of these times can impair cortex development and its connections with the remainder of the brain (Volpe, 2009, Kostovic and Judas, 2010).

B. Injury to the periventricular and deep white matter of the premature infant has been demonstrated by gross pathology, histology, and imaging. Although cavitory lesions have become very uncommon in modern nurseries, noncavitory lesions are still commonly seen. These are thought to be due to damage to oligodendrocyte precursors, likely as a result of ischemia and the production of lactate, which damages oligodendrocyte precursors, resulting in delayed myelination and axonal damage (Back *et al.*, 2002, Back, 2006, Segovia *et al.*, 2008, Volpe, 2009).

C. Infarctions of the cerebral cortex are not common in preterm infants and are not believed to be an important part of the developmental difficulties encountered by these infants. This is the correct answer to this question.

D. Cerebellar hemorrhages have only recently been shown to be a fairly common result of premature birth, being found in about 10% of published series. Large hemorrhages are associated with significant developmental delay. However, even small hemorrhages seem to be associated with abnormal neurologic examination at age 4.8 years. Intraventricular hemorrhage seems to be associated with cerebellar hypoplasia at term equivalent age (Tam *et al.*, 2011, Tam *et al.*, 2011).

E. Periventricular hemorrhagic infarction is a condition in which periventricular veins are compressed by germinal matrix hemorrhages. The venous compression impairs venous drainage, resulting in venous ischemia and infarction, which is almost always accompanied by hemorrhage. The combination of infarction and hemorrhage in the immature brain causes severe damage with liquefaction necrosis of involved areas. The impact on outcome depends on size and location of the infarct, and on uni- versus bilaterality. Large bilateral

PVHIs have devastating consequences, whereas small unilateral ones have much less severe neurodevelopmental consequences. Nearly all affected neonates, however, have neurodevelopmental effects (Bassan *et al.*, 2007, Maitre *et al.*, 2009).

References:

1. Back S, Han B, Luo N, Chricton C, Xanthoudakis S, Tam J, et al. Selective vulnerability of late oligodendrocyte progenitors to hypoxia-ischemia. *J Neurosci.* 2002; 22: 455-63.
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ABR Item Codes (required)	
Modality	C05.A_____
Primary Content	C05.E_____
Anatomy	_____C05.B__
Pathology	_____C05.L__