SPR 2018 Pediatric Neuroradiology Session
Saturday Workshop
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SAM References

Eye(s) Have a Clue

Tim N. Booth, MD

1. **Which is the CORRECT statement concerning Morning Glory Disc Anomaly?**
    A. There is no elevation of the adjacent retinal surface.
    B. There can be associated enhancement of the optic nerve.
    C. Magnetic resonance angiography is not needed.
    D. There is an association with occipital encephaloceles.

Correct Answer: B
Rationale: Answer B is correct. There can be enhancement of the distal optic nerve, which should not be mistaken for tumor. Answer A is incorrect. Explanation: In MGDA, there can be elevation of the retina surrounding a funnel shaped defect within the optic disc. Answer C is incorrect. Explanation: Ipsilateral arterial stenosis/occlusion has been commonly reported associated with MGDA and MRA should be obtained. Answer D is incorrect. Explanation: Sphenoid encephaloceles are associated, not occipital. The sphenoid encephalocele may obstruct the nasal airway.
References:

- Elika S, Robson CD, Heidary G, Palidino MJ. Morning Glory Disc Anomaly: Characteristic MR Imaging Findings. AJNR Am J Neuroradiol 2013; 34: 2010-2014
- Masaro M, Thorarensen O, Liu GT, et al. Morning Glory Disc Anomaly and Moyamoya Vessels. Arch Opthalmol 1998; 116: 253-254
- Koenig SB, Naidich TP, Lissner G. The Morning Glory Syndrome Associated with Sphenoidal Encephalocele. Ophthalmology 1982; 89: 1368-1373

2. **Imaging abnormalities in CHARGE association include which of the following?**
    1. Olfactory bulb enlargement
    2. Enlarged vestibule
    3. Absent parotid glands
    4. Absent cochlea

Correct Answer: C
Rationale: Answer C is correct. Absent parotid glands have been reported recently with submandibular glands usually present in CHARGE association. Answer A is incorrect. Absent, not enlarged, olfactory bulbs have been reported in CHARGE and may indicate pituitary dysfunction. Answer B is incorrect. The vestibule is small with absent semicircular canals and can be associated with oval window atresia. Answer D is incorrect. The cochlea is present, but usually dysplastic.
References:

- Hoch MJ, Patel SH, Jethanamest D, et al. Head and Neck MRI Findings in CHARGE Syndrome. AJNR Am J Neuroradiol 2017; 38: 2357-2363
- Blustain J, Kirsch, Panigrphy A, Netchine I. Olfactory Anomalies in CHARGE Syndrome: Imaging Findings of a Potential Major Diagnostic Criterion. AJNR Am J Neuroradiol 2008; 29: 1266-1269

Imaging Approach of Congenital Ear Anomalies

Caroline D. Robson, MBChB

3. **Enlarged vestibular aqueducts (EVA) are frequently associated with:**
    A. Cochlear hypoplasia
    B. Thyroid goiter
    C. Dysmorphic cochlear scalar chambers
    D. Cochlear incomplete partition type III anomaly

Correct answer: C
Rationale: There is variable cochlear anomaly associated with EVA. Cochlear scalar chambers are usually asymmetric and sometimes there is deficiency of the interscalar septum between the cochlear apical and middle turns (IP-II anomaly). The resulting dysmorphism of the scalar chambers is measurable on MR. Goiter can occur in Pendred syndrome in association with EVA but usually only in areas where Iodine is deficient in the diet.
Reference:

- Reinshagen KL, Curtin HD, Quesnel AM and Juliano AF. Measurement for Detection of Incomplete Partition Type II Anomalies on MR Imaging. AJNR 2017 Oct;38(10):2003-2007. doi: 10.3174/ajnr.A5335. Epub 2017 Aug 3.

4. **Branchio-oto-renal (BOR) syndrome is usually characterized by:**
    A. An incomplete partition anomaly of the cochlea
    B. Hypoplastic and offset middle and apical cochlear turns
    C. Small Eustachian tubes
    D. Renal cysts and branchial cleft cysts in all patients

Correct answer: B
Rationale: The characteristic anomaly of the cochlea in BOR is hypoplastic and offset middle and apical cochlear turns. The Eustachian tubes are usually large. Not all patients are found to have the same or complete phenotypic manifestations of BOR.
Reference:

- Propst EJ, Blaser S, Gordon KA, Harrison RV, Papsin BC. Temporal bone findings on computed tomography imaging in branchio-oto-renal syndrome. Laryngoscope. 2005 Oct;115(10):1855-62

Current Concepts of Autoimmune Encephalitis

Manohar Shroff, MD

5. **Which of the following is the most common Autoimmune Encephalitis?**
    A. Limbic Encephalitis
    B. Anti-N-Methyl-D-aspartate receptor encephalitis
    C. Rasmussen Encephalitis
    D. Opsoclonus-Myoclonus

Correct Answer: B
Rationale: Anti-NMDA receptor encephalitis is the most frequent autoimmune encephalitis in children and adolescents. Approximately 40% of patients are younger than 18 years.
Reference:

- The frequency of autoimmune N-methyl-D-aspartate receptor encephalitis surpasses that of individual viral etiologies in young individuals enrolled in the California Encephalitis Project. Gable MS, Sheriff H, Dalmau J, Tilley DH, Glaser CA.Clin Infect Dis. 2012 Apr;54(7):899-904. doi: 10.1093/cid/cir1038

6. Which of the following is typically associated with presence of an ovarian teratoma?

1. Limbic Encephalitis
2. Ophelia Syndrome
3. Anti-N-Methyl-D-aspartate receptor encephalitis
4. Rapid onset obesity with hypothalamic dysfunction, hypoventilation and autonomic dysregulation (ROHHAD)

Correct Answer: C

Rationale: Anti-NMDA receptor encephalitis is usually associated with ovarian teratoma, but this is reported more often with women over 18 years of age. In children and teenage girls, the presence of an associated ovarian teratoma in Anti-NMDA receptor encephalitis though described, is uncommon. Ophelia syndrome is a form of limbic encephalitis which occurs in Hodgkin's lymphoma. Though limbic encephalitis is the most well described paraneoplastic encephalitis in older adults, in children it is uncommon. ROHHAD is associated with neural crest tumors (ganglioneuroma, neuroblastoma, or ganglioneuroblastoma).

Reference:

- Armangue T, Petit-Pedrol M, Dalmau J. Autoimmune encephalitis in children. J Child Neurol. 2012 Nov;27(11):1460-9

Emergent Neuroimaging for Suspected Acute Pediatric Stroke

Susan Palasis, MD

7. Emergent treatment of an acute ischemic stroke is contraindicated by which of the following?

1. SWI DWI mismatch.
2. Presence of gross hemorrhage.
3. The insular "dot" sign.
4. Presence of large to medium vessel occlusion.

Correct Answer: B

Rationale: Emergent treatment for acute ischemic stroke is contraindicated when gross hemorrhage is present on CT or MR. Answer A is incorrect. SWI provides a rough estimate of tissue perfusion with the prominent hypointense cortical veins in the ischemic territory due to increased concentration of deoxyhemoglobin. A greater mismatch between asymmetric cortical veins on SWI and a small DWI area (SWI–DWI mismatch) are more likely to have a favorableoutcome from reperfusion strategy or recanalization. Answer C is incorrect. The insular dot sign indicates the presence of thrombus in insular vessels and is not a contraindication to treatment. Answer D is incorrect. Patients with a large vessel occlusion (cervical or intracranial internal carotid artery, M1 or M2 segment of the MCA) are most likely to benefit from acute stroke intervention.

References:

- Hsu et al. The Neuroradiology Journal. 2017, Vol. 30(2) 109–119
- Heit JJ, Wintermark M. New developments in clinical ischemic stroke prevention and treatment and their imaging implications. J Cereb Blood Flow Metab. 2017 Jan 1 : 271678X17694046

8. **What is the most sensitive method for detection of acute pediatric ischemic stroke?**

1. Non-enhanced head CT
2. Non-enhanced head CT and CT angiography (CTA)
3. Diffusion weighted imaging (DWI)
4. Catheter angiography

Correct Answer: C

Rationale: DWI can reveal acute diffusion restrictions from ischemia within minutes of an acute ischemic event. Answer A is incorrect. CT may be negative up to 12 hours following symptoms of an acute ischemic stroke. Answer B is incorrect. MRI with DWI is the most sensitive imaging method of detecting acute ischemic stroke. CT with CTA is reserved for evaluation of suspected acute ischemic stroke only when emergent MRI is not available.

Answer D is incorrect. Emergent catheter angiography is reserved for cases where neurointervention is a therapeutic option.

References:

- Quick Brain Magnetic Resonance Imaging With Diffusion-Weighted Imaging as a First Imaging Modality in Pediatric Stroke. Christy A. et al. Pediatric Neurology (2018) Volume 78 , 55 – 60
- Imaging of Pediatric Vascular Emergencies. Tang, Y. et al. Emerg Radiol (2018). https://doi.org/10.1007/s10140-017-1576-5

The Child with Intractable Epilepsy

Erin Simon Schwartz, MD, FACR

9. Which of the following statements is true regarding focal cortical dysplasia (FCD)?

1. Type Ia FCD is frequently detected with clinical brain MRI
2. Type IIa FCD is histologically characterized by dysmorphic neurons and balloon cells
3. Type IIb FCD is histologically characterized by dysmorphic neurons
4. Type IIb is associated with transmantle dysplasia

Correct Answer: D

Rationale: Transmantle cortical dysplasias are histologically type IIb FCDs. Answer A is incorrect as type Ia FCD is commonly missed or not detectable with typical clinical MRI examinations. Answer B is incorrect as type IIa FCD is histologically characterized by dysmorphic neurons. Answer C is incorrect as type IIa FCD is histologically characterized by dysmorphic neurons and balloon cells.

Reference:

- Blumcke I, et al. The clinicopathologic spectrum of focal cortical dysplasias: a consensus classification proposed by an ad hoc Task Force of the ILAE Diagnostic Methods Commission. Epilepsia 2011; 52: 158–74.

10. Which of the following is true regarding magnetoencephalography (MEG) and pediatric epilepsy?

1. MEG data can help guide intracranial EEG placement for improved detection of seizure foci
2. MEG is invasive and frequently requires sedation for children
3. MEG can only show abnormal activity when the structural brain MRI is abnormal
4. Multimodal integration, with MEG, MRI, DTI and/or PET, does not improve surgical outcome

Correct Answer: A

Rationale: MEG data can definitely help guide intracranial EEG placement for improved detection of seizure foci, both subdural electrode and stereoscopically placed depth electrodes. Answer B is incorrect as MEG is entirely noninvasive and rarely requires anesthesia for successful completion. Answer C is incorrect as MEG results are independent of MRI findings, and resection of MEG "lesions" has been shown to have equivalent clinical outcome to MRI lesions. Answer D is incorrect as multimodal integration has been shown to improve clinical outcome following surgery for intractable epilepsy.

References:

- Schwartz ES, et al. Magnetoencephalography. Pediatr Radiol 2010;40(1):50-58.
- Wilenius J, et al. Epilepsy Research 2013;105:337-348.
- Tan Y-L, et al. Quantitative surface analysis of combined MRI and PET enhances detection of focal cortical dysplasias. NeuroImage 2018;166:10-18.

The New Molecular Classification of Pediatric Brain Tumors: Implications for the Radiologist

Zoltan Patay, MD, PhD

11. Which of the following statements is true for the 2016 WHO classification of CNS tumors?

1. CNS PNET (primitive neuroectodermal tumor) is not an entity but a pattern now
2. The term (entity) CNS PNET has been removed from the group of embryonal tumors
3. CNS PNET has been replaced by ETANTR (embryonal tumor with abundant neuropil and true rosettes), which now also comprises ependymoblastoma and medulloepithelioma
4. ATRT does not belong to the group of embryonal tumors any more

Correct Answer: B

Rationale: The 2017 WHO classification does not recognize PNET as a separate entity, hence the term primitive neuroectodermal tumor (or PNET) has been removed from the ”diagnostic lexicon”. Answer A is incorrect. Explanation: While the 2016 WHO classification continues to describe entities, variants and patterns, the term PNET has been deleted completely. Answer C is incorrect. Explanation: ETANTR has never been included in the WHO classification. Ependymoblastoma and most medulloepitheliomas are now consolidated into the new entity named ”Embryonal tumour with multilayered rosettes, C19MC-altered”, which also comprises tumors which were diagnosed as ETANTR in the past. Answer D is incorrect. Explanation: ATRT remains a recognized embryonal tumor entity in the latest WHO classification of CNS tumors.

References:

- David N. Louis et al. The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary Acta Neuropathol (2016) 131, 803–820
- Sturm D et al. New Brain Tumor Entities Emerge from Molecular Classification of CNS-PNETs Cell (2016) 164, 1060–1072
- Korshunov A et al. Embryonal tumor with abundant neuropil and true rosettes (ETANTR), ependymoblastoma, and medulloepithelioma share molecular similarity and comprise a single clinicopathological entity. Acta Neuropathol. (2014) 128, 279-89.

12. Which of the following statements is <u>not</u> true for medulloblastoma?

1. In the 2000 edition of the WHO Classification of Tumours medulloblastoma was defined as a malignant (WHO grade IV), invasive embryonal tumor of the cerebellum with predominantly neuronal differentiation.
2. In the 2016 update of the WHO classification of Tumours of the CNS, medulloblastomas are classified according to molecular characteristics in addition to histopathological features.
3. Embryonal tumors formerly referred to as “medulloblastoma” include 4 “transcriptionally” different subtypes with different molecular features and often different demographic and clinical features too.
4. Group 1 and 2 tumors are always midline intraventricular, whereas Group 3 and 4 may be found in any location in the posterior fossa.

Correct Answer: D

Rationale: Correct (actually the *“incorrect”*) answer is: D. Group 3 and 4 medulloblastoma are midline, intraventricular. Group 1 (WNT) and 2 (SHH) medulloblastoma can also be midline, intraventricular, but Group 1 are commonly located within the CP angle cistern and the foramen of Luschka, whereas Group 2 are typically hemispheric in the cerebellum. Answer A is correct. Explanation: This is the conventional histopathological definition of medulloblastoma. Answer B is correct. Explanation: The new WHO classification uses both ”old” histopathological and ”new” molecular features to describe entities. This concept is referred to as ”integrated phenotypic and genotypic parameters”. Answer C is correct. Explanation: Cerebellar tumors characterized by the medulloblastoma histopathological phenotype are distilled into four principal groups (Groups 1-4). A updated version of the classification recognizes further subgroups totaling 12 at present (WNT a, b; SHH a, b, g, d; Group 3a, b, g and Group 4a, b, g).

References:

- David N. Louis et al. The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary Acta Neuropathol (2016) 131, 803–820
- Taylor MD et al. Molecular subgroups of medulloblastoma: the current consensus. Acta Neuropathol (2012) 123, 465-472
- Perreault S et al. MRI Surrogates for Molecular Subgroups of Medulloblastoma. Am J Neuroradiol (2014), 35, 1263-1269

Treatment Related Complications in Pediatric Brain Tumors

Dennis W. Shaw, MD

13. Pseudoprogression typically seen within the first 3 months following radiation therapy is;

1. Thought to represent a local inflammatory response with increased permeability.
2. Associated with decreased survival.
3. Symptomatic in most patients.
4. Less commonly seen with methylation of the MGMT promotor.

Correct Answer: A

Rationale: Answer B is incorrect. Explanation: The presence of pseudoprogression is associated with increased survival. Answer C is incorrect. Explanation: more typically pseudoprogression is asymptomatic. Answer D is incorrect. Explanation: MGMT promotor methylation is highly correlated with pseudoprogression.

Reference:

- Pseudoprogression and Pseudoresponse: Imaging Challenges in the Assessment of Postreatment Glioma. Hygino da Cruz Jr et al; AJNR 2011.

14. Radiation necrosis following radiotherapy for brain tumors;

1. Is likely in a new lesion appearing greater than 3 years following therapy.
2. Never occurs with less than 60 Gy.
3. Has a higher incidence in someone with diabetes.
4. Is frequently intermixed with tumor.

Correct Answer: D

Rationale: Answer A is incorrect. Explanation: radiation necrosis is usually seen between 2 and 32 months following therapy, 85% occurring within 2 years. Answer B is incorrect. Explanation: though most cases are seen with greater than 60 Gy, radiation necrosis can be seen at lower doses. Answer C is incorrect. Explanation: diabetes is not associated with increased likelihood of radiation necrosis.

Reference:

- Radiation Necrosis in the Brain: Imaging Features and Differentiation from Tumor Recurrence. Shah et al; Radiographics 2012

Neuroimaging Evaluation of Hydrocephalus and Increased Intracranial Pressure

Asim F. Choudhri, MD

15. Which of the following causes of hydrocephalus is commonly an indication for hydrocephalus:

A. Venous sinus thrombosis
B. Tectal glioma (Correct answer)
C. Colloid cyst
D. Choroid plexus carcinoma of the lateral ventricle

Correct Answer: B

Rationale: Explanation: A tectal glioma can obstruct the aqueduct of Sylvius, impairing CSF flow from the third ventricle to the fourth ventricle. An endoscopic third ventriculostomy will bypass this obstruction. Answer A is incorrect. Explanation: Venous sinus thrombosis results in a elevation of intracranial pressures without

obstruction to flow. Answer C is incorrect. Explanation: A colloid cyst will typically obstruct the foramina of Monro resulting in impaired CSF flow from the lateral ventricles to the third ventricle. An ETV would not be expected to address this obstruction. Answer D is incorrect. Explanation: A choroid plexus carcinoma of the lateral ventricle causes hydrocephalus due to increased production of CSF, not impaired transit. Accordingly, an ETV would not be expected to address this abnormality.

Reference:

- Griessenauer CJ, Rizk E, Miller JH, Hendrix P, Tubbs RS, Dias MS, Riemenschneider K, Chern JJ. Pediatric tectal plate gliomas: clinical and radiological progression, MR imaging characteristics, and management of hydrocephalus. J Neurosurg Pediatr. 2014 Jan;13(1):13-20

16. Pseudotumor cerebri is associated with which of the following?

A. Papilledema (Correct answer)
B. Enlarged pituitary gland
C. Carotid terminus stenosis
D. Pachymeningeal enhancement

Correct Answer: A

Rationale: Explanation: Papilledema is a common feature of pseudotumor cerebri, related to elevated CSF pressures and resultant distention of the optic nerve sheath and optic nerve head edema. Answer B is incorrect. Explanation: Pseudotumor cerebri is often associated with a partial empty sella, but is not associated with an enlarged pituitary gland. Answer C is incorrect. Explanation: Pseudotumor cerebri is associated with transverse sinus stenosis. Carotid terminus stenosis is a feature of moyamoya vasculopathy. Answer D is incorrect. Explanation: Pachymeningeal enhancement is a feature associated with intracranial hypotension, and is not feature of pseudotumor cerebri.

Reference:

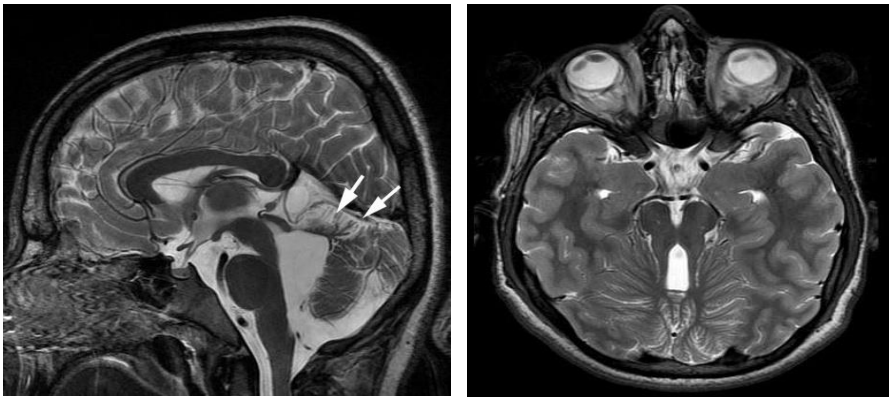
- Mandelstam S, Moon A. MRI of optic disc edema in childhood idiopathic intracranial hypertension. Pediatr Radiol. 2004 Apr;34(4):362

Update on Posterior Fossa Malformations

Thierry A. G. M. Huisman, MD

17. The most likely diagnosis in this young patient with episodic tachypnea, ocular motor apraxia, ataxia and intellectual disability is:

A. Dandy Walker Malformation
B. Joubert Boltshauser Syndrome
C. Muscle-Eye-Brain Disease
D. Meckel Gruber Syndrome



Correct Answer: B

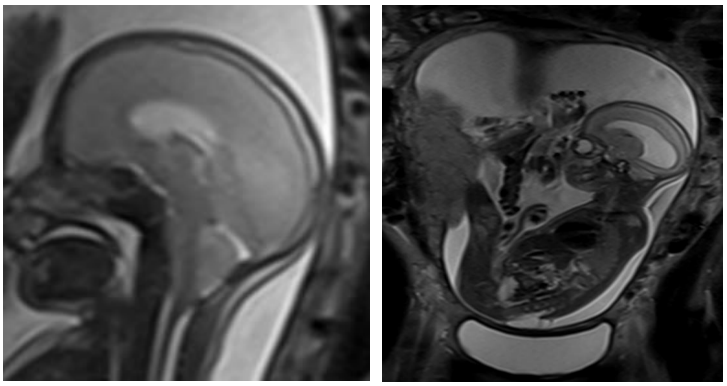
Rationale: Clinical presentation as well as the characteristic Molar Tooth appearance of the mesencephalon/brainstem on axial imaging is considered pathognomonic for the Joubert-Boltshauser syndrome. Answer Dandy walker Syndrome is incorrect. Explanation: No cystic enlargement of the IV ventricle. Answer Muscle Eye Brain Disease is incorrect. Explanation: Normal pons, no cerebellar cysts, no Z-shaped brainstem on sagittal imaging Answer Meckel Gruber syndrome is incorrect. Explanation: Syndrome primarily presents with renal cysts, hypoplastic lungs and arthrogryposis next to occipital encephalocele

Reference:

- Mandelstam S, Moon A. MRI of optic disc edema in childhood idiopathic intracranial hypertension. Pediatr Radiol. 2004 Apr;34(4):362

18. The most likely diagnosis in this fetus is:

A. Chiari 1 in skin covered myelomeningocele
B. Chiari 1 in non-skin covered myelomeningocele
C. Chiari 2 in skin covered myelomeningocele
D. Chiari 2 in non-skin covered myelomeningocele



Correct Answer: C

Rationale: A Chjari 2 malformation of the posterior fossa is considered to be associated with or secondary to a non-skin covered spinal dysraphia allowing for leakage of cerebrospinal fluid out of the spinal canal during development of the posterior fossa structures. This is summarized as the “unified theory”. All other answers are incorrect. A Chiari 1 malformation is characterized by an isolated low positioning of the cerebellar tonsils into the upper cervical spinal canal. A Chiari 2 malformation is more complex and in nearly 100% of cases associated with a non-skin covered spinal dysraphia.

Reference:

- Mandelstam S, Moon A. MRI of optic disc edema in childhood idiopathic intracranial hypertension. Pediatr Radiol. 2004 Apr;34(4):362