CLINICAL GUIDELINES

Pediatric Head Imaging Policy

Version 20.0.2018 Effective May 17, 2018



eviCore healthcare Clinical Decision Support Tool Diagnostic Strategies: This tool addresses common symptoms and symptom complexes. Imaging requests for individuals with atypical symptoms or clinical presentations that are not specifically addressed will require physician review. Consultation with the referring physician, specialist and/or individual's Primary Care Physician (PCP) may provide additional insight.

CPT® (Current Procedural Terminology) is a registered trademark of the American Medical Association (AMA). CPT® five digit codes, nomenclature and other data are copyright 2016 American Medical Association. All Rights Reserved. No fee schedules, basic units, relative values or related listings are included in the CPT® book. AMA does not directly or indirectly practice medicine or dispense medical services. AMA assumes no liability for the data contained herein or not contained herein.

Pediatric Head Imaging

Procedure Codes Associated with Pediatric Head Imaging	3
PEDHD-1: General Guidelines	5
PEDHD-2: Specialized Imaging Techniques	10
PEDHD-3: Pediatric Headache	13
PEDHD-4: Pediatric Head and Face Trauma	15
PEDHD-5: Sinusitis and Allergic Rhinitis	18
PEDHD-6: Epilepsy and Other Seizure Disorders	22
PEDHD-7: Macrocephaly, Microcephaly, and Hydrocephalus	26
PEDHD-8: Craniosynostosis	30
PEDHD-9: Chiari and Skull Base Malformations	32
PEDHD-10: Intracranial Aneurysms and AVM	36
PEDHD-11: Syncope	41
PEDHD-12: Pediatric Stroke	42
PEDHD-13: Benign Brain Lesions	45
PEDHD-14: Pediatric Demyelinating Diseases	47
PEDHD-15: Pituitary Dysfunction	50
PEDHD-16: Pediatric Ear Disorders	54
PEDHD-17: Autism Spectrum Disorders	58
PEDHD-18: Behavioral and Psychiatric Disorders	59
PEDHD-19: Intellectual Disability, Cerebral Palsy, and	
Developmental Motor Delay	60
PEDHD-20: Ataxia	62
PEDHD-21: Epistaxis	63
PEDHD-22: Pseudotumor Cerebri	65
PEDHD-23: Cranial Neuropathies	66
PEDHD-24: Pediatric Sleep Disorders	67
PEDHD-25: Temporomandibular Joint (TMJ) Imaging in Children	68
PEDHD-26: Tourette's Syndrome	69
PEDHD-27: Tuberous Sclerosis	70
PEDHD-28: Von Hippel Lindau Syndrome (VHL)	71
PEDHD-29: CNS Infection	72
PEDHD-30: Scalp and Skull Lesions	74
PEDHD-31: Eye Disorders	75

<u>Imaging Guidelines</u> V20.0.2018

Procedure Codes Associated with Pe- Head Imaging	
MRI	CPT [®]
Brain MRI without contrast	70551
Brain MRI with contrast (rarely used)	70552
Brain MRI without and with contrast	70553
Orbit, Face, Neck MRI without contrast	70540
Orbit, Face, Neck MRI with contrast (rarely used)	70542
Orbit, Face, Neck MRI without and with contrast	70543
Temporomandibular Joint (TMJ) MRI	70336
Functional MRI Brain not requiring physician or psychologist	70554
Functional MRI Brain requiring physician or psychologist	70555
MR Spectroscopy	76390
Unlisted MRI procedure (for radiation planning or surgical software)	76498
MRA	CPT [®]
Head MRA without contrast	70544
Head MRA with contrast	70545
Head MRA without and with contrast	70546
Neck MRA without contrast	70547
Neck MRA with contrast	70548
Neck MRA without and with contrast	70549
CT	CPT [®]
Head CT without contrast	70450
Head CT with contrast	70460
Head CT without and with contrast	70470
Orbits CT without contrast (includes temporal bone and mastoid)	70480
Orbits CT with contrast (includes temporal bone and mastoid)	70481
Orbits CT without and with contrast (includes temporal bone and mastoid)	70482
Maxillofacial CT without contrast (includes sinuses, jaw, and mandible)	70486
Maxillofacial CT with contrast (includes sinuses, jaw, and mandible)	70487
Maxillofacial CT without and with contrast (includes sinuses, jaw, and mandible)	70488
Neck CT without contrast (includes jaw, and mandible)	70490
Neck CT with contrast (includes jaw, and mandible)	70491
Neck CT without and with contrast (includes jaw, and mandible)	70492
CT Guidance for Stereotactic Localization (used for sinus surgery planning)	77011
CT Guidance for Placement of Radiation Therapy Fields	77014
Unlisted CT procedure (for radiation planning or surgical software)	76497
CTA	CPT [®]
Head CTA	70496
Neck CTA	70498

<u>Imaging Guidelines</u> V20.0.2018

Nuclear Medicine	CPT®
PET Brain Metabolic Evaluation	78608
PET Brain Perfusion Evaluation	78609
PET with concurrently acquired CT; limited area (this code rarely used in	78814
pediatrics)	70014
PET with concurrently acquired CT; whole body	78816
Brain Scintigraphy Static Limited	78600
Brain Scintigraphy Limited with Vascular Flow	78601
Brain Scintigraphy Complete Static	78605
Brain Scintigraphy Complete with Vascular Flow	78606
Brain Imaging SPECT	78607
Brain Imaging Vascular Flow	78610
Cisternogram	78630
Cerebrospinal Ventriculography	78635
Shunt Evaluation	78645
CSF Flow SPECT	78647
CSF Leakage Detection	78650
Radiopharmaceutical Dacryocystography	78660
Ultrasound	CPT [®]
Echoencephalography (Head or Cranial Ultrasound)	76506
Ophthalmic ultrasound, diagnostic; B-scan & quantitative A-scan performed same encounter	76510
	76511
Ophthalmic ultrasound, diagnostic; quantitative A-scan only Ophthalmic ultrasound, diagnostic; B-scan	76512
Ophthalmic ultrasound, diagnostic; anterior segment ultrasound, immersion	
(water bath) B-scan	76513
Ophthalmic ultrasound, diagnostic; corneal pachymetry, unilateral or bilateral	76514
Ophthalmic biometry by ultrasound, A-scan	76516
Ophthalmic biometry by ultrasound, A-scan, with lens power calculation	76519
Ophthalmic ultrasonic foreign body localization	76529
Soft tissues of head and neck Ultrasound (thyroid, parathyroid, parotid, etc.)	76536
Transcranial Doppler study of the intracranial arteries; complete study	93886
Transcranial Doppler study of the intracranial arteries; limited study	93888
Transcranial Doppler study of the intracranial arteries; vasoreactive study	93890
Transcranial Doppler study of the intracranial arteries; emboli detection without	93892
intravenous microbubble injection	
Transcranial Doppler study of the intracranial arteries;; emboli detection with intravenous microbubble injection	93893
Duplex scan of extracranial arteries; complete bilateral study	93880
Duplex scan of extracranial arteries; unilateral or limited study	93882
Non-invasive physiologic studies of extracranial arteries, complete bilateral study	93875

PEDHD-1: General Guidelines	
PEDHD-1.1: Pediatric Head Imaging Age Considerations	6
PEDHD-1.2: Pediatric Head Imaging Appropriate Clinical	
Evaluation	6
PEDHD-1.3: Pediatric Head Imaging Modality General	
Considerations	7

PEDHD-1.1: Pediatric Head Imaging Age Considerations

Many conditions affecting the head in the pediatric population are different diagnoses than those occurring in the adult population. For those diseases which occur in both pediatric and adult populations, minor differences may exist in management due to patient age, comorbidities, and differences in disease natural history between children and adults.

Patients who are < 18 years old should be imaged according to the pediatric head imaging guidelines and patients who are ≥ 18 years old should be imaged according to the adult head imaging guidelines, except where directed otherwise by a specific guideline section.

PEDHD-1.2: Pediatric Head Imaging Appropriate Clinical Evaluation

- A recent (within 60 days) face to face evaluation including a detailed history, physical examination with a thorough neurologic examination, and appropriate laboratory studies should be performed prior to considering advanced imaging (CT, MR, Nuclear Medicine), unless the patient is undergoing guideline-supported scheduled imaging evaluation.
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic patients for disorders involving the head is not supported. Advanced imaging of the head is only indicated in patients who have documented active clinical signs or symptoms of disease involving the head.
 - Advanced imaging of the head is not indicated for evaluation of recurrent isolated vomiting in patients without associated headache or focal neurologic findings unless a gastrointestinal workup (labs, imaging, and endoscopy) does not reveal a cause.
- Unless otherwise stated in a specific guideline section, repeat imaging studies of the head are not necessary unless there is evidence for progression of disease, new onset of disease, and/or documentation of how repeat imaging will affect patient management or treatment decisions.

Requests for Studies with Overlapping Fields

- ➤ There are many CPT® codes for imaging the head that have significantly overlapping fields. In the majority of cases where multiple head CPT® codes are requested, only one CPT® code should be approved unless there is clear documentation of a need for the additional codes to cover all necessary body areas.
- See <u>HD-1.1: General Guidelines Anatomic Issues</u> the correct coding of these studies.

PEDHD-1.3: Pediatric Head Imaging Modality General Considerations

MRI

- MRI is the preferred modality for imaging the pediatric head unless otherwise stated in a specific guideline section.
- Due to the length of time for image acquisition and the need for the patient to lie still, anesthesia is required for almost all infants except neonates and young children (age < 7 years), as well as older children with delays in development or maturity. In this patient population, MRI imaging sessions should be planned with a goal of minimizing anesthesia exposure adhering to the following considerations:
 - MRI should always be performed without and with contrast unless there is a specific contraindication to gadolinium use since the patient already has intravenous access for anesthesia.
 - Recent evidence based literature demonstrates the potential for gadolinium deposition in various organs including the brain, after the use of MRI contrast.
 - The U.S. Food and Drug Administration (FDA) has noted that there is currently no evidence to suggest that gadolinium retention in the brain is harmful and restricting gadolinium-based contrast agents (GBCAs) use is not warranted at this time. It has been recommended that GBCA use should be limited to circumstances in which additional information provided by the contrast agent is necessary and the necessity of repetitive MRIs with GBCAs should be assessed.
 - If requesting clinicians indicate that a non-contrast study is being requested with specific concern for gadolinium retention, the exam can be approved.
 - If multiple body areas are supported by eviCore guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session.

> CT

- CT is generally inferior to MRI for imaging the pediatric head, but has specific indications in which it is the preferred modality listed in specific sections of these guidelines.
 - CT should not be used to replace MRI in an attempt to avoid sedation unless listed as a recommended study in a specific guideline section.

Ultrasound

- Cranial ultrasound (CPT® 76506) is a non-invasive means of evaluating for intracranial abnormalities in infants with an open anterior fontanelle.
- Transcranial Doppler ultrasonography has some utility in select populations of older children with known or suspected intracranial vascular disease.

Nuclear Medicine

- Nuclear medicine studies other than metabolic PET imaging on the pediatric brain or head are rarely performed in an elective outpatient setting, but the following studies can be approved when requested for the following indications:
 - Brain Scintigraphy with or without vascular flow (any one of CPT[®] codes: CPT[®] 78600, CPT[®] 78601, CPT[®] 78605, or CPT[®] 78606)
 - Establish brain death (rarely done in outpatient setting).
 - Brain Imaging SPECT with Ioflupane I-23 (CPT® 78607)
 - Immunocompromised patients with mass lesion detected on CT or MRI for differentiation between lymphoma and infection.
 - Brain Imaging Vascular Flow (CPT® 78610)
 - Cerebral ischemia.
 - Establish brain death.
 - CSF Leakage Detection (CPT® 78650)
 - Evaluation of CSF rhinorrhea or otorrhea, or refractory post-lumbar puncture headache.
 - Radiopharmaceutical Dacryocystography (CPT® 78660)
 - Suspected obstruction of nasolacrimal duct due to excessive tearing.

The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.

- 1. Siegel MJ. Brain. In: Pediatric sonography. 5th ed. Philadelpia, Wolters Kluwer. 2018 (in press).
- 2. Nadgir R, and Yousem DM. Neuroradiology. The requisites. 4th Ed. Philadelphia, Mosby. 2017.
- 3. Latchaw RE, Kucharczyk J, and Moseley ME. Imaging of the nervous system diagnostic and therapeutic applications. Philadelphia, Elsevier. 2005. pp 2000. Accessed October 19, 2017. http://pubs.rsna.org/doi/full/10.1148/radiol.2412062583.
- 4. Louis ED, Mayer SA, and Rowland LP (Ed.). *Merritt's Neurology*. 13th Ed. Philadelphia, Lippincott. 2013.
- 5. Menkes JH, Sarnat HB, and Maria BL. Child Neurology. 7th Ed. Philadelphia, Lippincott. 2006.
- 6. Barkovich AJ. Diagnostic imaging: pediatric neuroradiology. *Amirsys*. 2nd Edition. Salt Lake City, UT. 2014.
- Prabhu SP,and Young-Poussaint Ty. Pediatric central nervous system emergencies. Neuroimag Clin N Am. 2010 Nov; 20 (4): 663-683. Accessed October 19, 2017. http://www.neuroimaging.theclinics.com/article/S1052-5149(10)00080-8/pdf.
- 8. Patra KP, Lancaster JD, Hogg J, et al. Pediatric MRI of the Brain: a primer. *Pediatr Rev.* 2014 Mar; 35 (3):106-113. Accessed October 19, 2017. http://pedsinreview.aappublications.org/content/35/3/106?download=true.
- 9. Riccabona M. Neonatal neurosonography. *Eur J Radiol.* 2014 Sep; 83 (9): 1495-1506. Accessed October 19, 2017. http://www.ejradiology.com/article/S0720-048X(14)00235-6/fulltext.
- Ing C, DiMaggio C, Whitehouse A, et al. Long-term differences in language and cognitive function after childhood exposure to anesthesia. *Pediatrics*. 2012 Sep; 130 (3): e476-e485. Accessed October 19, 2017. http://pediatrics.aappublications.org/content/pediatrics/130/3/e476.full.pdf.

11. Monteleone M, Khandji A, Cappell J, et al. Anesthesia in children: perspectives from nonsurgical pediatric specialists. *J Neurosurg Anesthesiol.* 2014; 26 (4): 396-398. Accessed October 19, 2017. https://www.medscape.com/medline/abstract/25191959.

- 12. DiMaggio C, Sun LS, and Li G. Early childhood exposure to anesthesia and risk of developmental and behavioral disorders in a sibling birth cohort. *Anesth Analg.* 2011; 113: 1143-1151. Accessed October 19, 2017. http://www.oalib.com/references/8560162.
- New York State Department of Health. Guidelines for determining brain death. December 2011, Available at: https://www.health.ny.gov/professionals/hospital administrator/letters/2011/brain death guidelines.ht m.
- 14. Donohoe KJ, Frey KA, Gerbaudo VH, et al. Society of nuclear medicine procedure guideline for brain death scintigraphy. Version 1.0. Approved February 25, 2003, available at: http://www.onelegacy.org/site/docs/SocietyNuclearMedicine_BrainDeathIdentification_022503.pdf.
- 15. Vlaar AMM, van Kroonenburgh MJPG, Kessels AGH, et al. Meta-analysis of the literature on diagnostic accuracy of SPECT in parkinsonian syndromes. *BMC Neurol.* 2007 Feb; 7: 27. Accessed October 19, 2017. https://www.researchgate.net/publication/6075009_Meta-analysis of the literature on diagnostic accuracy of SPECT in Parkinsonian syndromes.
- 16. Thrall JH, and Zeissman HA. Nuclear Medicine. The requisites. Mosby. 2001; 312-313.
- 17. Relkin N, Marmarou A, Klinge P, et al. Diagnosing idiopathic normal-pressure hydrocephalus. *Neurosurgery*. 2005; 57 (3 Suppl): S2-4-S2-16. Accessed October 19, 2017. https://www.ncbi.nlm.nih.gov/pubmed/16160425.
- MacDonald A, and Burrell S. Infrequently performed studies in nuclear medicine: Part 2. J Nucl Med Technol. 2009 Mar; 37: 1-13. Accessed October 19, 2017. http://tech.snmjournals.org/content/37/1/1.full.
- 19. Fraum TJ, Ludwig DR, Bashir MR, et al. Gadolinium-based contrast agents: a comprehensive risk assessment. *J Magn Reson Imaging*. 2017; 46:338–353. Accessed October 19, 2017. https://www.medscape.com/medline/abstract/28083913.
- 20. FDA Drug Safety Communication: FDA identifies no harmful effects to date with brain retention of gadolinium-based contrast agents for MRIs; review to continue. FDA Drug Safety Communication. May 22, 2017. https://www.fda.gov/Drugs/DrugSafety/ucm559007.htm.

<u>Imaging Guidelines</u> V20.0.2018

PEDHD-2: Specialized Imaging Techniques	
PEDHD-2.1: Magnetic Resonance Spectroscopy (MRS, CPT®	
76390)	11
PEDHD-2.2: Functional Magnetic Resonance Imaging (fMRI, CPT®	
70554 and CPT® 70555)	11
PEDHD-2.3: PET Brain Imaging (CPT® 78608 and CPT® 78609)	12

PEDHD-2.1: Magnetic Resonance Spectroscopy (MRS, CPT® 76390)

Magnetic Resonance Spectroscopy involves the analysis of the levels of certain chemicals in pre-selected voxels (small regions) on an MRI scan done at the same time.

NOTE: * Certain payers consider MRS investigational, and their coverage policies may take precedence over eviCore healthcare guidelines.

Uses in pediatric neuro-oncology: See <u>PEDONC-4: Pediatric CNS Tumors</u> for imaging indications.

Uses in Metabolic Disorders:

- These cases should be forwarded for medical director review.
- MRS is associated with disease-specific characteristics findings and is indicated for diagnosis and disease monitoring in the following metabolic disorders:
 - Canavan disease.
 - Creatine deficiency.
 - Nonketotic hyperglycinemia.
 - Maple Syrup Urine disease.
- MRS has nonspecific abnormal patterns that can aid in the diagnosis of the following metabolic disorders, but is not routinely indicated for disease monitoring:
 - Metachromatic leukodystrophy.
 - Pelizaeus-Merzbacher disease.
 - Hypomyelination and Congenital Cataract.
 - Globoid Cell Leukodystrophy (Krabbe disease).
 - X-linked adrenoleukodystrophy.
 - Mitochondrial disorders.
 - Alexander disease.
 - Megalencephalic leukoencephalopathy with subcortical cysts.
 - Vanishing White Matter disease.
 - MRS can be approved for disease monitoring of these diagnoses when recent MRI findings are inconclusive and a change in therapy is being considered.
- MRS is considered investigational for all other pediatric indications at this time.

PEDHD-2.2: Functional Magnetic Resonance Imaging (fMRI, CPT® 70554 and CPT® 70555)

- These cases should be forwarded for medical director review.
- fMRI is indicated to define eloquent areas of the brain as part of preoperative planning for epilepsy surgery or removal of a mass lesion.
 - The documentation should be clear that brain surgery is planned.
 - Can be approved concurrently with MRI Brain (CPT® 70551 or CPT® 70553) and/or PET Brain Metabolic (CPT® 78608).
- fMRI is considered investigational for all other pediatric indications at this time.

PEDHD-2.3: PET Brain Imaging (CPT® 78608 and CPT® 78609)

- These cases should be forwarded for medical director review.
- Uses in pediatric neuro-oncology: See <u>PEDONC-4: Pediatric CNS Tumors</u> for imaging indications.
- PET Brain is indicated to define active areas of the brain as part of preoperative planning for epilepsy surgery.
 - The documentation should be clear that brain surgery is planned.
 - Can be approved concurrently with MRI Brain (CPT® 70551 or CPT® 70553) and/or fMRI (CPT® 70554 or CPT® 70555).
- fMRI is considered investigational for all other pediatric indications at this time.

- Rossi A, and Biancheri R. Magnetic resonance spectroscopy in metabolic disorders. *Neuroimaging Clin N Am. 2013 Aug; 23 (3): 425–48.* Accessed October 19, 2017. http://www.neuroimaging.theclinics.com/article/S1052-5149(12)00217-1/pdf.
- 2. Hertz-Pannier L, Noulhaine M, Rodrigo S, et al. Pretherapeutic functional magnetic resonance imaging in children. *Neuroimag Clin N Am.* 2014 Nov; 24 (4): 639-653. Accessed October 19, 2017. http://www.neuroimaging.theclinics.com/article/S1052-5149(14)00071-9/pdf.
- 4. Schneider JF. MR Spectroscopy in children: protocols and pitfalls in non-tumorous brain pathology. *Pediatr Radiol.* 2016 Jun; 46 (7): 963-982. Accessed October 19, 2017. https://link.springer.com/article/10.1007/s00247-014-3270-z.
- 5. Dory CE, Coley BD, Karmazyn B, et al. Seizures—child. *ACR Appropriateness Criteria*®. 2012:1-10 Accessed October 19, 2017. https://acsearch.acr.org/docs/69441/Narrative/.
- Ramey WL, Martirosyan NL, Lieu CM, et al. Current management and surgical outcomes of medically intractable epilepsy. *Clin Neurol Neurosurg.* 2013 Dec; 115 (12): 2411-2418. Accessed October 19, 2017. http://www.sciencedirect.com/science/article/pii/S0303846713003892.

PEDHD-3: Pediatric Headache

Headache is a very common complaint in school aged children and adolescents. Many of these children have a family history of one of the primary headache disorders, such as migraine or tension headache.

- A recent (within 60 days) evaluation including a detailed headache history, physical examination with a thorough neurologic examination, and appropriate laboratory studies should be performed prior to considering advanced imaging.
- Advanced imaging is not indicated for pediatric patients with headache in the absence of red flag symptoms. Sensitivity and specificity of MRI are greater than that of CT for intracranial lesions.
- ➤ MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) is indicated for children with headaches and at least one of the following red flags:
 - Age ≤ 5 years.
 - Headaches awakening from sleep or always present in the morning.
 - Focal findings on neurologic examination including diplopia.
 - Clumsiness (common description of gait or coordination problems in young children).
 - Headaches associated with morning nausea/vomiting.
 - New onset of seizure activity with focal features.
 - Papilledema on physical exam.
 - Headache precipitated by coughing, sneezing, or Valsalva.
 - Exclusively occipital headache.
 - Progressive worsening in headache frequency and severity without period of temporary improvement.
 - Systemic symptoms such as persistent fever, weight loss, rash, or joint pain.
 - Immunocompromised patient.
 - Patient with hypercoagulable state or bleeding disorder.
 - Known history of cancer of any type.
 - Known autoimmune or rheumatologic disease.
 - Known genetic disorder with predisposition to intracranial mass lesions.
 - History of stable chronic headaches with recent significant change in frequency or severity.
 - Patients requiring sedation should generally not have non-contrast MRI studies.
 See PEDHD-1.3: Pediatric Head Imaging Modality General Considerations.
- CT Head poorly visualizes the posterior fossa in children and is generally insufficient to evaluate pediatric headaches with red flag symptoms. CT should not be approved in lieu of MRI solely to avoid sedation.
- CT Head without contrast is indicated for pediatric headache with one or more of the following:
 - Recent head trauma.
 - Suspected skull or other bony involvement.
 - Ventriculoperitoneal shunt with suspected shunt malfunction. See <u>PEDHD-7:</u> <u>Macrocephaly, Microcephaly, and Hydrocephalus</u> for additional imaging.
 - Sudden onset (thunderclap) headache with suspected intracranial hemorrhage.

- MRI is contraindicated due to implantable device or rapid clinical deterioration.
- MRA Brain or CTA Head are not generally medically necessary in the evaluation of headache in children unless a vascular lesion has been seen or suspected on a prior brain MRI Brain or CT Head.
 - Concurrent approval of both MRI and MRA is generally not indicated.
- MRV Head (CPT® 70544) is indicated in pediatric patients with papilledema and headache. See <u>PEDHD-22: Pseudotumor Cerebri</u> for additional imaging guidelines.

- 1. Hayes LL, Coley BD, Karmazyn B, et al. Headache—child. *ACR Appropriateness Criteria*®. 2012: 1-10. Accessed October 19, 2017. https://acsearch.acr.org/docs/69439/Narrative/.
- 2. Ryan ME, Palasis S, Saigal G, et al. Head trauma—child. *ACR Appropriateness Criteria*®. 2014: 1-13 Accessed October 19, 2017. https://acsearch.acr.org/docs/3083021/Narrative/.
- 3. Hershey A, Kabbouche MA, and O'Brien H. Headaches. *Nelson Textbook of Pediatrics, Chapter 595.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 2863-2874.
- 4. Blume HK. Pediatric headache: a review. *Pediatr Rev.* 2012 Dec; 33: 562-574. Accessed October 19, 2017. http://pedsinreview.aappublications.org/content/33/12/562.
- 5. De Bries A, Young PC, Wall E, et al. CT scan utilization patterns in pediatric patients with recurrent headache. *Pediatrics*. 2013 July; 132 (1); e1-e8. Accessed October 19, 2017. https://pdfs.semanticscholar.org/ab91/0b02850dd99a9c6607c45ad26dd6fcc2cc65.pdf.
- Lewis DW, Ashwal S, Dahl G, et al. Practice parameter: Evaluation of children and adolescents with recurrent headache. *Neurology*. 2002 Aug 27; 59 (4):490–498. https://www.ncbi.nlm.nih.gov/pubmed/12196640?dopt=Abstract.
- Trofimova A, Vey BL, Mullins ME, et al. Imaging of children with nontraumatic headaches. AJR Am J Roentgenol. 2017 Sep; 12: 1-10. Accessed October 19, 2017. http://www.ajronline.org/doi/abs/10.2214/AJR.17.18561.

PEDHD-4: Pediatric Head and Face	Trauma
PEDHD-4.1: Head Trauma	16
PEDHD-4.2: Facial Trauma	16

PEDHD-4.1: Head Trauma

In patients with recent head trauma, a history focused on the incident and careful examination of the head, neck, and neurological function should be performed prior to considering advanced imaging.

- CT Head without contrast (CPT® 70450) is the primary advanced imaging study in patients with acute head trauma.
 - CT Maxillofacial without contrast (CPT® 70486), Orbits/Temporal bone without contrast (CPT® 70480), or CT Cervical spine without contrast (CPT® 72125) is indicated if there has been associated injury to those structures.
- ➤ Brain MRI without contrast (CPT® 70551) is indicated for the following:
 - Children with an abnormal neurological exam that is not explained by the CT findings.
 - Children suspected of being the victims of physical abuse. See <u>PEDMS-7:</u> <u>Suspected Physical Child Abuse</u> for imaging considerations.
- ➤ Following a head injury, a repeat head CT Head without contrast (CPT® 70450) or MRI Brain without contrast (CPT® 70551) is indicated if the child develops fixed or fluctuating diminished mental acuity or alertness, or new abnormalities on neurological examination.
- Follow-up of known or treated subdural or epidural hematoma may require frequent imaging during the initial 8 weeks following injury, and these requests should generally be approved.
 - These cases should be forwarded for medical director review.
- Currently there is no well-validated pediatric version of the Canadian or New Orleans Head CT Rule to aid in deciding which children seen after recent head trauma would benefit from head CT.
- Advanced imaging is not indicated for children with minor head trauma and none of the following red flags:
 - Loss of consciousness.
 - Altered mental status.
 - Known or suspected skull fracture.
 - Glasgow Coma Score ≤ 13.

PEDHD-4.2: Facial Trauma

> CT without contrast is the preferred imaging study in facial trauma.

Coding of Facial Imaging

Both orbital/facial bone CT (CPT® 70480) and maxillofacial CT (CPT® 70486) cover the structures of the orbits, sinuses, and face. Unless there is a grounded suspicion of simultaneous involvement of more posterior lesions, especially of the region involving the middle or inner ear, one of these studies only should be sufficient.

Maxillofacial CT (CPT® 70486) is the usual study (except in obvious orbital or temporal bone trauma), but either study is appropriate.

- 1. Ryan ME, Palasis S, Saigal G, et al. Head trauma—child. *ACR Appropriateness Criteria*®. 2014:1-13. Accessed October 19, 2017. https://acsearch.acr.org/docs/3083021/Narrative/.
- 2. Osmond MH, Klassen TP, Wells GA, et al. CATCH: a clinical decision rule for the use of computed tomography in children with minor head injury. *CMAJ*. 2010 Mar 9; 182 (4): 341-348. Accessed October 19, 2017. http://www.cmaj.ca/content/182/4/341.full.
- Atabaki SM, Stiell IG, Bazarian JJ, et al. A clinical decision rule for cranial computed tomography in minor pediatric head trauma. Arch Pediatr Adolesc Med. 2008 May; 162 (5): 439-445. Accessed October 19, 2017.
 - http://journals.lww.com/pqs/Fulltext/2017/05000/Quality_Improvement_in_Pediatric_Head_Trauma_w ith.2.aspx.
- 4. Maguire JL, Boutis K, Uleryk EM, et al. Should a head-injured child receive a head CT scan? A systematic review of clinical prediction rules. *Pediatrics*. 2009; 124: e145-e154. Accessed October 19, 2017. https://pedclerk.bsd.uchicago.edu/sites/pedclerk.uchicago.edu/files/uploads/should.pdf.
- 5. Nigrovic LE, Stack AM, Mannix RC, et al. Quality improvement effort to reduce Cranial CTs for children with minor blunt head trauma. *Pediatrics*. 2015; 136 (1): e227-e233. Accessed October 19, 2017. http://pediatrics.aappublications.org/content/136/1/e227.full.

<u>Imaging Guidelines</u> V20.0.2018

PEDHD-5: Sinusitis and Allergic Rhinitis	
PEDHD-5.1: General Considerations	19
PEDHD-5.2: Imaging Indications in Sinusitis	19
PEDHD-5.3: Stereotactic CT Localization (CPT® 77011)	20
PEDHD-5.4: Requests for Both Head and Sinus Imaging	20
PEDHD-5.5: Allergic Rhinitis	20
PEDHD-5.6: Other Indications for Sinus Imaging	20

PEDHD-5.1: General Considerations

Acute sinusitis is a clinical diagnosis, and imaging is not indicated to establish a diagnosis. Acute bacterial sinusitis can be presumptively diagnosed in a child with acute upper respiratory infection (URI) symptoms and any of the following:

- Persistent symptoms lasting > 10 days without improvement.
- Worsening symptoms after initial period of improvement.
- Severe symptoms including purulent nasal discharge and fever > 102.2°F for at least 3 consecutive days.
- Presumed bacterial infections should be treated empirically with appropriate antibiotics.
- Imaging of any kind cannot distinguish bacterial from viral sinusitis.

PEDHD-5.2: Imaging Indications in Sinusitis

- Mild mucosal thickening in the paranasal sinuses or mastoids is an extremely common incidental finding noted on head imaging studies done for other indications. If there are no other abnormalities of facial structures noted, this finding is not an indication for advanced imaging of the sinuses or temporal bone.
- CT of the sinuses without contrast (CPT® 70486) is indicated if any of the following is present:
 - No improvement after 10 days of appropriate antibiotic treatment.
 - Generally this will be amoxicillin/clavulanate, amoxicillin, cefdinir, cefuroxime, cefpodoxime, or ceftriaxone.
 - Recurrence of a treated infection within 8 weeks of effective treatment.
 - Chronic sinusitis (persistent residual URI symptoms for > 90 days).
 - Known or suspected fungal sinusitis.
 - Preoperative evaluation to assess surgical candidacy.
- CT of the sinuses with contrast (CPT® 70487) can be performed if any of the following is present:
 - Orbital or facial cellulitis.
 - Proptosis.
 - Abnormal visual examination.
 - Ophthalmoplegia.
 - Cystic fibrosis.
 - Immunocompromised patient.
 - Fungal or vascular lesions visualized in nasal cavity.
- CT Head with contrast (CPT® 70460) or MRI Brain without and with contrast (CPT® 70553) is indicated if any of the following are present:
 - Focal neurologic findings.
 - Altered mental status.
 - Seizures.
- Repeat sinus imaging is generally not indicated for patients who have responded satisfactorily to treatment, but can be approved with clear documentation of the need for updated CT results to direct acute patient care decisions.
 - These cases should be forwarded for medical director review.

PEDHD-5.3: Stereotactic CT Localization (CPT® 77011)

Stereotactic CT localization is frequently obtained prior to sinus surgery. The dataset is then loaded into the navigational workstation in the operating room for use during the surgical procedure. The information provides exact positioning of surgical instruments with regard to the patient's 3D CT images. In most cases, the preoperative CT is a technical-only service that does not require interpretation by a radiologist.

- ➤ The imaging facility should report CPT® 77011 when performing a scan not requiring interpretation by a radiologist.
- ➤ If a diagnostic scan is performed and interpreted by a radiologist, the appropriate diagnostic CT code (e.g. CPT® 70486) should be used.
- ➤ It is not appropriate to report both CPT® 70486 and CPT® 77011 for the same CT stereotactic localization imaging session.
- ➤ 3D Rendering (codes CPT® 76376 or CPT® 76377) should not be reported in conjunction with CPT® 77011 (or CPT® 70486 if used). The procedure inherently generates a 3D dataset.
- Such operative studies are indicated when ordered by the operating surgeon for this purpose.

PEDHD-5.4: Requests for both Head and Sinus Imaging

- Head CT does not visualize all of the sinuses.
- ➤ Head MRI provides excellent visualization of the sinuses sufficient to recognize sinusitis, and addition of sinus CT for this purpose is unnecessary.
- ➤ In patients being evaluated for potential sinus surgery, separate sinus CT is often appropriate even after a head MRI in order to visualize obstructions to spontaneous mucous flow. See <u>PEDHD-5.3</u>: <u>Stereotactic CT Localization (CPT® 77011)</u>.
- Separate head imaging is not generally indicated in patients with a normal neurological examination who have headaches associated with sinus symptoms.
- Sinus CT or MRI is not indicated for the evaluation of headaches or neurological complaints without a more specific indication pointing to a sinus etiology.

PEDHD-5.5: Allergic Rhinitis

Advanced imaging is not indicated for diagnosis or management of patients with uncomplicated allergic rhinitis.

PEDHD-5.6: Other Indications for Sinus Imaging

See: **PEDHD-4.2: Facial Trauma** for imaging guidelines in trauma.

- Congenital anomalies of facial structures CT without contrast (CPT® 70486).
- 3-D CT reconstructed images (CPT® 76377) in conjunction with routine CT should be an integral part of the examination in evaluating craniofacial abnormalities.
- Tumors or other disorders of facial structures CT without and with contrast (CPT® 70488) or MRI Orbits/Face/Neck without and with contrast (CPT® 70543).

Obstructive sleep apnea—see <u>PEDHD-24: Sleep Disorders of Childhood</u> for imaging guidelines.

- Wald ER, Applegate KE, Bordley C, et al. Clinical practice guideline for the diagnosis and management of acute bacterial sinusitis in children aged 1 to 18 years. *Pediatrics*. 2013 Aug 5; 132 (1): e262-e280. Accessed October 19, 2017. https://www.medscape.com/medline/abstract/23796742.
- 2. Karmazyn B, Coley BD, Dempsey-Robertson ME, et al. Sinusitis—child. *ACR Appropriateness Criteria*®. 2012: 1-8. Accessed October 19, 2017. https://acsearch.acr.org/docs/69442/Narrative/.
- 3. Pappas DE, and Hendley JO. Sinusitis. *Nelson Textbook of Pediatrics, Chapter 380.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 2014-2017.
- 4. Magit A. Pediatric rhinosinusitis. *Otolaryngol Clin N Am.* 2014 Oct; 47 (5): 733-746. Accessed October 19, 2017. http://www.oto.theclinics.com/article/S0030-6665(14)00059-0/pdf.
- 5. Siedman MD, Gurgel RK, Lin SY, et al. Clinical practice guideline: allergic rhinitis executive summary. *Otolaryngol Head Neck Surg.* 2015 Feb 2; 152:197-206. Accessed October 19, 2017. http://journals.sagepub.com/doi/10.1177/0194599814562166.

PEDHD-6: Epilepsy and Other Seizure Disord	ers
PEDHD-6.1: Initial Imaging of Non-Febrile Seizures	23
PEDHD-6.2: Repeat imaging indications	23
PEDHD-6.3: Special Imaging Studies in Evaluation for Epilepsy	
Surgery	24
PEDHD-6.4: Febrile Seizures	24

A recent (within 60 days) face to face evaluation including a detailed history, physical examination with a thorough neurologic examination, and appropriate laboratory studies should be performed prior to considering advanced imaging, unless the patient is undergoing guideline-supported scheduled follow-up imaging evaluation. This clinical evaluation should also include family history and (whenever possible) the accounts of eyewitnesses to the event(s).

PEDHD-6.1: Initial Imaging of Non-Febrile Seizures

- ➤ MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) is indicated for the following:
 - First-time seizure in child ≥ 12 months of age that has no known cause and is not associated with fever.
 - Partial seizures.
 - Focal neurologic deficits.
 - Inconclusive findings on recent cranial ultrasound or CT Head.
 - If patient meets criteria for MRI imaging for initial imaging of non-febrile seizure, MRI is approvable even with a recent negative CT.
 - Patients requiring sedation should generally not have non-contrast MRI studies.
 See <u>PEDHD-1.3</u>: <u>Pediatric Head Imaging Modality General Considerations</u>.
- ➤ CT Head without contrast (CPT® 70450) is indicated for the following:
 - First-time seizure in child associated with recent head trauma.
 - Patient cannot safely undergo MRI (avoidance of sedation is not an indication).
- Cranial ultrasound (CPT® 76506) is indicated for the following:
 - First-time seizure in child < 12 months of age that has no known cause and is not associated with fever if the infant has an open fontanelle.
- The following imaging tests do not generally add valuable information initially and are not indicated for the initial evaluation of seizures in children:
 - CTA Head or Neck.
 - MRA Head or Neck.
 - MRI Cervical, Thoracic, or Lumbar Spine.

PEDHD-6.2: Repeat imaging indications

- Repeat MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) is indicated for the following:
 - Need to perform MRI using Epilepsy Protocol (typically 3T magnet with thin section angled slices through hippocampus and temporal lobes, either without or without and with contrast).
 - New or worsening focal neurologic deficits.
 - Increase in severity or frequency of seizures despite documented therapeutic antiepileptic drug levels.
 - Change in seizure type.
 - Preoperative evaluation for epilepsy surgery.
 - Patients requiring sedation should generally not have non-contrast MRI studies.
 See PEDHD-1.3: Pediatric Head Imaging Modality General Considerations.

PEDHD-6.3: Special Imaging Studies in Evaluation for Epilepsy Surgery

For patients with a previous brain MRI and documentation of intractable epilepsy for which surgical treatment or another interventional modality is under active consideration, any of the following are indicated for preoperative planning:

- These cases should be forwarded for medical director review
- ➤ PET Brain Metabolic (CPT® 78608).
- ➤ Functional MRI Brain (CPT® 70554 or CPT® 70555).
- MR Spectroscopy (CPT® 76390).
 - NOTE: Certain payers consider MR Spectroscopy investigational/experimental, and those coverage policies take precedence over eviCore Imaging Guidelines.

PEDHD-6.4: Febrile Seizures

A typical febrile seizure is a generalized seizure occurring in the presence of fever (T > 100.4°F) and no central nervous system infection in a child between the age of 6 months and 5 years.

- Neuroimaging should not be performed in the routine evaluation of children with simple febrile seizures.
- ➤ MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) is indicated for febrile seizures in the presence of one or more of the following:
 - Seizure lasting > 15 minutes.
 - Partial seizures.
 - Focal neurologic deficits.
 - Multiple seizures within 24 hours.
 - Macrocephaly
 - Signs and symptoms of increased intracranial pressure.

- 1. Dory CE, Coley BD, Karmazyn B et al. Seizures—child. *ACR Appropriateness Criteria*®. 2012: 1-10. Accessed October 19, 2017. https://acsearch.acr.org/docs/69441/Narrative/.
- 2. Mikati MA, and Hani AJ. Seizures in Childhood. *Nelson Textbook of Pediatrics, Chapter 593.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 2823-2857.
- 3. Sidhu R, Velayudam K, and Barnes G. Pediatric seizures. *Pediatr Rev.* 2013 Aug; 34 (8):333-341. https://www.ncbi.nlm.nih.gov/pubmed/23908360?dopt=Abstract.
- 4. Prabhu SP, and Young-Poissant T. Pediatric central nervous system emergencies. *Neuroimag Clin N Am.* 2010 Nov; 20 (4):663-683. Accessed October 19, 2017. http://www.neuroimaging.theclinics.com/article/S1052-5149(10)00080-8/pdf.
- 5. Ramey WL, Martirosyan NL, Lieu CM, et al. Current management and surgical outcomes of medically intractable epilepsy. *Clin Neurol Neurosurg.* 2013 Dec; 115 (12):2411-2418. Accessed October 19, 2017. https://www.ncbi.nlm.nih.gov/pubmed/24169149.
- St. Louis EK, and Cascino GD. Diagnosis of epilepsy and related episodic disorders. Continuum. 2016 Feb 1; 22: 15-37. Accessed October 19, 2017. https://mayoclinic.pure.elsevier.com/en/publications/diagnosis-of-epilepsy-and-related-episodic-disorders.
- 7. Hertz-Pannier L, Noulhaine M, Rodrigo S, et al. Pretherapeutic functional magnetic resonance imaging in children. *Neuroimag Clin N Am.* 2014 Nov; 24 (4): 639-653. Accessed October 19, 2017. Cv3r https://www.ncbi.nlm.nih.gov/pubmed/25441505.
- 8. Duffner PK, Berman PH, Baumann RJ, et al. Clinical practice guideline—febrile seizures: guideline for the neurodiagnostic evaluation of the child with a simple febrile seizure. *Pediatrics*. 2011 Feb; 127 (2): 389-394. Accessed October 19, 2017. http://pediatrics.aappublications.org/content/127/2/389.

PEDHD-7: Macrocephaly, Microcephaly, an	d
Hydrocephalus	
PEDHD-7.1: Macrocephaly	27
PEDHD-7.2: Microcephaly	27
PEDHD-7.3: Hydrocephalus	27

PEDHD-7.1: Macrocephaly

Macrocephaly is defined as head circumference that is greater than the 95th percentile for age and sex, established by use of measurements and CDC growth charts. An online calculator to determine head circumference percentile is available at: http://www.infantchart.com/cdc0to3headforage.php.

Birth to age 12 months:

- Ultrasound of the head (CPT® 76506) is indicated initially in patients with an open fontanelle.
- ➤ If hydrocephalus or hemorrhage is present on ultrasound, CT Head without contrast (CPT® 70450) is indicated.
- For any abnormality seen on ultrasound, MRI Brain without and with contrast (CPT® 70553) is indicated.

Age 13 months and older:

- ➤ MRI Brain without and with contrast (CPT® 70553) is indicated.
- CT is generally not indicated in this age group since uncomplicated hydrocephalus is less likely after early infancy.

PEDHD-7.2: Microcephaly

- Microcephaly is defined as head circumference that is less than the 5th percentile for age and sex, established by use of measurements and CDC growth charts. An online calculator to determine head circumference percentile is available at: http://www.infantchart.com/cdc0to3headforage.php.
- ➤ MRI Brain without and with contrast (CPT® 70553 is indicated for all patients.
 - CT is generally not recommended as that modality lacks the sensitivity to detect the relevant anatomical abnormalities.

PEDHD-7.3: Hydrocephalus

- This is the most common identifiable cause of macrocephaly. Almost all hydrocephalus is obstructive, except hydrocephalus due to choroid plexus papillomas. See <u>PEDONC-4.13: Choroid Plexus Tumors</u> for imaging guidelines for those lesions.
- Hydrocephalus is traditionally divided into non-communicating (the obstruction lies within the course of the brain's ventricular system) and communicating (the obstruction is distal to the ventricular system).
- Patients requiring sedation should generally not have non-contrast MRI studies. See PEDHD-1.3: Pediatric Head Imaging Modality General Considerations.

Initial Imaging Indications

Age 0-6 months:

- Screening head ultrasound examination (CPT® 76506)
- ➤ If ultrasound shows hydrocephalus, MRI Brain without and with contrast (CPT® 70553) is indicated.
- Serial US (CPT® 76506) can be used to monitor ventricular size to determine need and timing of placement of a ventricular catheter.

Greater than 6 months old:

➤ MRI Brain without and with contrast (CPT® 70553) is indicated.

Spine imaging:

MRI Spine without and with contrast (CPT® 72156, CPT® 72157, and CPT® 72158) may be indicated in individuals with Chiari malformation (multiple spine segments), Dandy-Walker malformation (cervical spine only), or malignant infiltration of the meninges.

Repeat Imaging Indications

- Rapid MRI Brain without contrast (CPT® 70551) or CT Head without contrast (CPT® 70450) is indicated for any new signs or symptoms suggesting shunt malfunction, including (but not limited to) sepsis, decreased level of consciousness, protracted vomiting, visual or neurologic deterioration, decline of mentation after initial improvement, or new or changing pattern of seizures. Rapid MRI Brain without contrast (CPT® 70551) or CT Head without contrast (CPT® 70450) is indicated following shunt placement and then every 12 months for patients with stable clinical findings.
 - Rapid MRI provides more anatomical detail and does not involve radiation exposure, but many providers use head CT as rapid MRI is not universally available.
 - For routine follow up imaging with CT a low dose protocol should be used.
- Shunting into the peritoneum (VP shunts) can give rise to abdominal complications, but these are generally symptomatic, so surveillance imaging of the abdomen is not indicated.
 - Abdominal ultrasound (CPT® 76700) can be approved for suspicion of CSF pseudocyst formation or distal shunt outlet obstruction.
- Familial screening is not indicated for hydrocephalus except in siblings of individuals with aqueductal stenosis, for whom a one-time CT Head without contrast (CPT® 70450) or Rapid MRI Brain without contrast (CPT® 70551) is indicated.

Additional Rarely Used Studies

- Cisternogram (CPT® 78630) is rarely done in children but can be approved for the following:
 - Known hydrocephalus with worsening symptoms.
 - Suspected obstructive hydrocephalus.

- Suspected normal pressure hydrocephalus with gait disturbance and either dementia or urinary incontinence.
- Cerebrospinal Ventriculography (CPT® 78635) is rarely done in children but can be approved for the following:
 - Evaluation of internal shunt, porencephalic cyst, or posterior fossa cyst.
- Nuclear Medicine Shunt Evaluation (CPT® 78645) and CSF Flow SPECT (CPT® 78647) are rarely done in children but can be approved for the following:
 - Suspected malfunction of ventriculoperitoneal, ventriculopleural, or ventriculovenous shunts.

- 1. Ashwal S, Michelson D, Plawner L, et al. Practice parameter: evaluation of the child with microcephaly (an evidence-based review). *Neurology*. 2009 Sep; 73: 887-897. Accessed October 19, 2017. https://www.aan.com/PressRoom/home/GetDigitalAsset/8479.
- 2. Kinsman SL, and Johnston MV. Hydrocephalus. *Nelson Textbook of Pediatrics, Chapter 591.11.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 2814-2817.
- 3. Boyle TP, Paldino MJ, Kimia AA, et al. Comparison of Rapid Cranial MRI to CT for Ventricular Shunt Malfunction. *Pediatrics*. 2014 July; 134 (1): e47-e54. Accessed October 19, 2017. http://pediatrics.aappublications.org/content/pediatrics/134/1/e47.full.pdf.

PEDHD-8: Craniosynostosis

PEDHD-8.1: Imaging

31

PEDHD-8.1: Imaging

Craniosynostosis is the premature closure of one or more cranial sutures, usually during infancy. Abnormal head shape is the common clinical feature.

- CT head without contrast (CPT® 70450) is indicated in the diagnosis of craniosynostosis, with reported sensitivity near 100%. CT also detects associated intracranial pathology.
- ➤ 3D rendering (CPT® 76377) is indicated with the initial diagnostic CT to evaluate the extent of synostosis and determine surgical candidacy or for preoperative planning.
- CT Maxillofacial (CPT® 70486) and CT Orbits (CPT® 70480) without contrast are generally not necessary to evaluate patients with craniosynostosis but are indicated if the craniosynostosis is part of a larger congenital defect which also involves the bones of the face or orbit.
- ➤ Head Ultrasonography (CPT® 76506) is an alternative method of assessing sutural patency in neonates and infants when radiographs are indeterminate.

- 1. Hall KM, Besachio DA, Moore MD, et al. Effectiveness of screening for craniosynostosis with ultrasound: a retrospective review. *Pediatr Radiol.* 2017; 47: 606-612.
- 2. Rozovsky K, Udjus K, Wilson N, et al. Cranial ultrasound as a first-line imaging examination for craniosynostosis. Pediatrics 2016. 137: e20152230
- 3. Kinsman SL, and Johnston MV. Craniosynostosis. *Nelson Textbook of Pediatrics, Chapter 591.12.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 2817-2819.
- Fearon JA. Evidence-based medicine: craniosynostosis. *Plast Reconstr Surg.* 2014 May; 133 (5): 1261-1275. Accessed October 19, 2017. http://journals.lww.com/plasreconsurg/Abstract/2014/05000/Evidence_Based_Medicine_Craniosynostosis.36.aspx.

PEDHD-9: Chiari and Skull Base Malformati	ons
PEDHD-9.1: Chiari I Malformations	33
PEDHD-9.2: Chiari II Malformations	33
PEDHD-9.3: Chiari III and IV Malformations	34
PEDHD-9.4: Basilar Impression	34
PEDHD-9.5: Platybasia	34

PEDHD-9.1: Chiari I Malformations

This is the most common, involving caudal displacement or herniation of the cerebellar tonsils. Chiari I is often associated with syringomyelia, and rarely with hydrocephalus. Most cases are asymptomatic and discovered incidentally on a head scan performed for another indication. When symptoms are present, they are usually nonspecific but can include lower cranial nerve palsies or sleep apnea.

- ➤ For initial evaluation, MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) and MRI of the entire spine without contrast (CPT® 72141, CPT® 72146, CPT® 72148) or without and with contrast (CPT® 72156, CPT® 72157, CPT® 72158) is indicated.
 - Patients requiring sedation should generally not have non-contrast MRI studies.
 See PEDHD-1.3: Pediatric Head Imaging Modality General Considerations.
- Repeat MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) is indicated for patients with a known Chiari I malformation when any of the following are present:
 - There are new or worsening signs or symptoms documented on a physical examination within 60 days of the imaging request.
 - A surgical procedure is actively being considered.
- Repeat MRI Spine imaging is not indicated for patients with normal initial spine imaging unless there are new or worsening signs or symptoms that suggest spinal cord pathology documented on a physical examination within 60 days of the imaging request.
 - These cases should be forwarded for medical director review.
- Repeat brain and spine imaging in individuals with Chiari I malformations and known syringomyelia or hydromyelia is highly individualized and is indicated at the discretion of the specialist coordinating the patient's care for this condition.
 - These cases should be forwarded for medical director review.
- Familial screening is not indicated for Chiari I Malformations.

PEDHD-9.2: Chiari II Malformations

These malformations are less common and more severe than Chiari I malformations. These patients usually present at birth. Myelomeningocele is always present, and syringomyelia and hydrocephalus are extremely common.

- Ultrasound is the initial examination in infants to determine ventricular size and associated anomalies and to provide a baseline for follow up evaluation.
- For initial advance imaging evaluation, MRI Brain without and with contrast (CPT[®] 70553) and MRI of the entire spine without and with contrast (CPT[®] 72156, CPT[®] 72157, CPT[®] 72158) is indicated.
- Repeat brain and spine imaging in individuals with Chiari II malformations is highly individualized and is indicated at the discretion of the specialist coordinating the patient's care for this condition.
 - These cases should be forwarded for medical director review.
- Familial screening is not indicated for Chiari II Malformations.

PEDHD-9.3: Chiari III and IV Malformations

Chiari III malformation includes cerebellar herniation into a high cervical myelomeningocele. Chiari IV malformation refers to complete cerebellar agenesis. Both Chiari III and IV malformations are noted at birth, and are rarely compatible with life.

- Repeat brain and spine imaging in individuals with Chiari III and IV malformations is highly individualized and is indicated at the discretion of the specialist coordinating the patient's care for this condition.
 - These cases should be forwarded for medical director review.
- Familial screening is not indicated for Chiari III or IV Malformations.

PEDHD-9.4: Basilar Impression

Basilar impression involves malformation of the occipital bone in relation to C1/2 (cervical vertebrae 1 and 2). The top of the spinal cord is inside the posterior fossa and the foramen magnum is undersized. Over time, this can lead to brain stem and upper spinal cord compression. Basilar impression can also be associated with the Chiari malformation, producing very complex anatomical abnormalities.

- MRI Brain (CPT® 70551) and cervical spine (CPT® 72141) without contrast are indicated.
- ➤ If surgery is being considered, CT Head (CPT® 70450) and cervical spine (CPT® 72125) without contrast are also indicated.
- Basilar impression appears to be genetic, and one-time screening of first-degree relatives with MRI Brain without contrast (CPT® 70551) can be approved.

PEDHD-9.5: Platybasia

Platybasia is a flattening malformation of the skull base, in which the clivus has a horizontal orientation.

Patients are usually asymptomatic, but either MRI Brain without contrast (CPT[®] 70551) or CT Head without contrast (CPT[®] 70450) is indicated to establish a positive diagnosis.

- 1. Siegel MJ. Brain. In: pediatric sonography. 5th ed. Philadelphia. Wolters Kluwer. 2018 (in press).
- 2. Kinsman SL, and Johnston MV. Congenital anomalies of the central nervous system. *Nelson Textbook of Pediatrics, Chapter 591.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 2802-2819.
- 3. Strahle J, Muraszko KM, Kapurch J, et al. Chiari malformation Type I and syrinx in children undergoing magnetic resonance imaging. *J Neurosurg Pediatr*. 2011 Aug; 8 (2): 205-213. Accessed October 19, 2017. http://theins.org/doi/10.3171/2011.5.PEDS1121.
- 4. Strahle J, Muraszko KM, Kapurch J, et al. Natural history of Chiari malformation Type I following decision for conservative treatment. *J Neurosurg Pediatr.* 2011 Aug; 8 (2): 214-221. Accessed October 19, 2017. http://theins.org/doi/10.3171/2011.5.PEDS1122.
- 5. Strahle J, Muraszko KM, Garton HJL, et al. Syrinx location and size according to etiology: identification of Chiari-associated syrinx. *J Neurosurg Pediatr.* 2015 July; 16 (1): 21-9 Epub 2015 Apr 3. Accessed October 19, 2017. https://www.ncbi.nlm.nih.gov/pubmed/25837888.
- 6. Strahle J, Smith BW, Martinez M, et al. The association between Chiari malformation Type I, spinal syrinx, and scoliosis. *J Neurosurg Pediatr.* 2015 Jun; 15 (6): 607-611. Accessed October 19, 2017. http://thejns.org/doi/full/10.3171/2014.11.PEDS14135.
- 7. Smoker WRK and Khanna G. Imaging the craniocervical junction. *Childs Nerv Syst.* 2008 Oct; 24 (10): 1123-1145. Accessed October 19, 2017. https://link.springer.com/article/10.1007/s00381-008-0601-0?no-access=true.

PEDHD-10: Intracranial Aneurysms and AVM
PEDHD-10.1: Pediatric Intracranial Aneurysms 37

PEDHD-10.2: Pediatric Intracranial Arteriovenous Malformations (AVM)

Imaging Guidelines

V20.0.2018

38

PEDHD-10.1: Pediatric Intracranial Aneurysms

Unlike adults, the majority of pediatric aneurysms are caused by genetic or developmental defects rather than environmental or lifestyle factors.

Pediatric aneurysms most commonly present with subarachnoid hemorrhage, headache, increased intracranial pressure, seizure activity, or focal neurologic findings.

- A recent (within 60 days) evaluation including a detailed history, physical examination with a thorough neurologic examination, and appropriate laboratory studies should be performed prior to considering advanced imaging, unless the patient is undergoing guideline-supported scheduled follow-up imaging evaluation.
- ➤ For patients presenting with suspected subarachnoid hemorrhage, CT Head without contrast (CPT® 70450) or MRI Brain without contrast (CPT® 70551) is indicated as an initial study.
 - If subarachnoid hemorrhage is present on CT or MRI, or lumbar puncture findings suggest hemorrhage, additional imaging with CTA Head (CPT® 70496) or MRA Head without contrast (CPT® 70544) is indicated.
- ➤ For patients presenting with headache, increased intracranial pressure, seizures, or focal neurologic findings, MRI without and with contrast (CPT® 70553) is indicated as an initial study.
 - If findings suspicious for intracranial aneurysm are present on MRI, additional imaging with CTA Head (CPT® 70496) or MRA Head without contrast (CPT® 70544) is indicated.
- ➤ For patients with known unruptured aneurysm presenting with headache, increased intracranial pressure, seizures, or focal neurologic findings, MRI without contrast (CPT® 70551) or without and with contrast (CPT® 70553) and MRA Head without contrast (CPT® 70544) are indicated.
- ➤ For patients with any of the following conditions and headache, increased intracranial pressure, seizures, or focal neurologic findings, MRI without contrast (CPT® 70551) or without and with contrast (CPT® 70553) and MRA Head without contrast (CPT® 70544) are indicated:
 - Polycystic kidney disease.
 - Fibromuscular dysplasia.
 - Ehlers-Danlos Syndrome.
 - Klippel-Trenaunay-Weber Syndrome.
 - Tuberous Sclerosis.
 - Moyamoya Syndrome.
 - Hereditary Hemorrhagic Telangiectasia (Osler-Weber-Rendu Syndrome).
 - Pseudoxanthoma elasticum.
 - Neurofibromatosis type 1.
 - Patients requiring sedation should generally not have non-contrast MRI studies.
 See PEDHD-1.3: Pediatric Head Imaging Modality General Considerations.
- The timing of follow-up imaging for intracranial aneurysms in children is similar to that in adults. See <u>HD-12.1: Intracranial Aneurysms</u> for follow-up imaging guidelines.

Screening MRI Brain or MRA Head for aneurysms is not supported in asymptomatic patients under age 20 since only 0.6 % of ruptured aneurysms occur in the pediatric age range.

Screening MRI Brain or MRA Head for aneurysms is not supported in patients with coarctation of the aorta repaired before age 3 since there is not an increased risk for intracranial aneurysm in this patient population.

<u>PEDHD-10.2: Pediatric Intracranial Arteriovenous Malformations</u> (AVM)

A recent (within 60 days) evaluation including a detailed history, physical examination with a thorough neurologic examination, and appropriate laboratory studies should be performed prior to considering advanced imaging, unless the patient is undergoing guideline-supported scheduled follow-up imaging evaluation.

Most intracranial AVMs are congenital, vary widely in their location and type, and are discovered at birth due to associated clinical findings or incidentally later in life. Certain hereditary conditions are associated with an increased risk for AVM development.

Vascular malformations include arteriovenous, venous, cavernous, and capillary malformations. The vein of Galen malformation is the most common arteriovenous malformation, presenting in neonates with signs of high output congestive heart failure or later in infancy of childhood with signs of hydrocephalus. Low flow venous, cavernous, and capillary malformations may be asymptomatic and discovered incidentally or they may present in childhood with seizures or neurologic findings secondary to intracranial hemorrhage.

Head ultrasound (CPT® 76506) is the study of choice for evaluation of a suspected vein of Galen malformation in the neonate. Once confirmed, MRI or conventional angiography are required to precisely identify the feeding arteries and draining vein, especially if embolization is planned.

MRA or CTA are indicated for diagnosis of low flow malformations.

- ➤ MRI Brain without and with contrast (CPT® 70553) is the initial study of choice for evaluation of suspected AVM after the neonate period.
 - Patients requiring sedation should generally not have non-contrast MRI studies.
 See PEDHD-1.3: Pediatric Head Imaging Modality General Considerations.
 - MRA, CTA, or CT are generally not indicated prior to completion of initial MRI.
- For patients with known AVM, MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553), and MRA Brain (CPT® 70544) or CTA Head (CPT® 70496) are indicated in the following circumstances:
 - New or worsening headaches, seizures, or focal neurologic symptoms.
 - Preoperative planning (including embolization).
- Head imaging for AVM screening is indicated for the following conditions:
 - Hereditary Hemorrhagic Telangiectasia (Osler-Weber-Rendu Syndrome).
 - MRI Brain without and with contrast (CPT® 70553) is indicated as an initial screening study for infants born to a parent with known HHT.

- MRI Brain without and with contrast (CPT® 70553) at the time of diagnosis, and a single repeat study after the age of 20.
- Ongoing surveillance imaging is not indicated for patients without new or worsening symptoms.
- Repeat MRI alone or with MRA or CTA (as above) is indicated for clinical signs or symptoms concerning for progression in a patient with a known AVM.
- Capillary Malformation-Arteriovenous Malformation (CM-AVM)
 - Caused by RASA1 mutations.
 - MRI Brain without and with contrast (CPT® 70553) at the time of diagnosis.
 - Ongoing surveillance imaging is not indicated for patients without new or worsening symptoms.
 - Repeat MRI alone or with MRA or CTA (as above) is indicated for clinical signs or symptoms concerning for progression in a patient with a known AVM.
 - See <u>PEDPVD-2: Vascular Anomalies</u>.
- Sturge-Weber Syndrome:
 - MRI Brain without and with contrast (CPT® 70553) and MRI Face/Neck (CPT® 70543) at the time of diagnosis.
 - Ongoing surveillance imaging is not indicated for patients without new or worsening symptoms.
 - Repeat MRI alone or with MRA or CTA (as above) is indicated for clinical signs or symptoms concerning for progression in a patient with a known AVM.
- Cerebral Cavernous Malformations:
 - Also known as cavernomas, cavernous angiomas, or cryptic vascular malformations.
 - MRI Brain without and with contrast (CPT® 70553) and MRI Cervical (CPT® 72156) and Thoracic (CPT® 72157) Spine without and with contrast at the time of diagnosis.
 - Ongoing surveillance imaging is not indicated for patients without new or worsening symptoms.
 - Repeat MRI alone or with MRA or CTA (as above) is indicated for clinical signs or symptoms concerning for progression in a patient with a known AVM.

- 1. Meyers PM, Halbach VD, and Barkovich AJ. Anomalies of cerebral vasculature: diagnostic and endovascular considerations. In: Barkovich AJ, Raybaud C eds. *Pediatric Neuroimaging*. 5th ed. Philadelphia PA. Wolters Kluwer. 2012; 1051-1108.
- 2. Barkovich AJ, and Raybaud CA. Congenital malformations of the brain and skull. In: Barkovich AJ, Raybaud C, eds. *Pediatric Neuroimaging*. 5th ed. Philadelphia PA. Wolters Kluwer. 2012; 367-568.
- 3. Gemmete JJ, Toma AK, Davagnanam I, et al. Pediatric cerebral aneurysms. *Neuroimag Clin N Am.* 2013 Nov; 23 (4): 771-779. Accessed October 19, 2017. http://www.neuroimaging.theclinics.com/article/S1052-5149(13)00033-6/abstract.
- 4. Beez T, Steiger H-J, and Hnggi D. Evolution of management of intracranial aneurysms in children: a systematic review of the modern literature. *J Child Neurol.* 2016; 31 (6): 773-783. Accessed October 20, 2017. https://www.medscape.com/medline/abstract/26516106.
- 5. Alvarez H, and Castillo M. Genetic markers and their influence on cerebrovascular malformations. *Neuroimag Clin N Am.* 2015 Feb; 25 (1): 69-82. Accessed October 20, 2017. http://www.neuroimaging.theclinics.com/article/S1052-5149(14)00102-6/pdf.
- 6. Nelson PK, et al. Neuroradiology in the imaging and therapeutic management of brain arteriovenous malformations (Ch 38). In Latchaw RE, Kucharczyk J, Moseley ME. *Imaging of the Nervous System*. Philadelphia, Elsevier. 2005.
- 7. Donti A, Spinardi L, Brighenti M, et al. Frequency of intracranial aneurysms determined by magnetic resonance angiography in children (mean age 16) having operative or endovascular treatment of coarctation of the aorta (mean age 13). *Am J Cardiol*. 2015 epub 2015 Aug; 116 (4):630-633. Accessed October 20, 2017. http://www.sciencedirect.com/science/article/pii/S0002914915013545.
- 8. McDonald J, and Pyeritz RE. Hereditary hemorrhagic telangiectasia. *GeneReviews™*, [Internet] eds. Pagon RA, Adam MP, Bird TD et al. version February 2, 2017. Accessed October 20, 2017. https://www.ncbi.nlm.nih.gov/books/NBK1351/.
- 9. Bayrak-Toydemir P and Stevenson D. *RASA1*-Related Disorders. *GeneReviews™* [Internet] eds. Pagon RA, Adam MP, Bird TD et al. version October 6, 2016. Accessed October 20, 2017. https://www.ncbi.nlm.nih.gov/books/NBK52764/.
- 10. Comi A and Pevsner J. Sturge-Weber syndrome. *OrphanetJ Rare Dis.* updated March 2014. https://rarediseases.org/rare-diseases/sturge-weber-syndrome/.
- 11. Morrison L, and Akers A. Cerebral cavernous malformation, familial. *GeneReviews™* [Internet] eds. Pagon RA, Adam MP, Bird TD et al. version August 4, 2016. Accessed October 20, 2017. https://www.ncbi.nlm.nih.gov/books/NBK1293/.
- 12. Linscott LL, Leach JL, Jones BV, et al. Developmental venous anomalies of the brain in children—imaging spectrum and update. *Pediatr Radiol.* 2016 Mar; 46 (3): 394-406. Accessed October 20, 2017. https://link.springer.com/article/10.1007%2Fs00247-015-3525-3.

PEDHD-11: Syncope

Syncope in children is almost always neurocardiogenic (vasovagal) in nature. Intracranial mass lesions do not cause isolated syncope. Syncope and seizure activity can often be challenging to distinguish for unwitnessed syncope.

Advanced imaging of the brain is not indicated for patients with isolated syncope without focal neurologic findings. See <u>PEDCD-5: Syncope</u> and <u>PEDHD-6: Epilepsy</u> and <u>Other Seizure Disorders</u> for additional imaging considerations.

- 1. Friedman KG, and Alexander ME. Chest pain and syncope in children: a practical approach to the diagnosis of cardiac disease. *J Pediatr*. 2013 Sep; 163 (3):896-901. Accessed October 20, 2017. http://www.jpeds.com/article/S0022-3476(13)00534-9/fulltext.
- 2. Cannon B, and Wackel P. Syncope. *Pediatr Rev.* 2016 Apr; 37 (4):159-168. Accessed October 20, 2017. http://pedsinreview.aappublications.org/content/37/4/159.

PEDHD-12: Pediatric Stroke	
PEDHD-12.1: General Considerations	43
PEDHD-12.2: Pediatric Stroke Initial Imaging	43
PEDHD-12.3: Pediatric Stroke Subsequent Imaging	43
PEDHD-12.4: Moyamoya Disease	43
PEDHD-12.5: Sickle Cell Disease	44
PEDHD-12.6: CNS Vasculitis and Stroke	44

PEDHD-12.1: General Considerations

Imaging indications are the same for neonates as for older children.

PEDHD-12.2: Pediatric Stroke Initial Imaging

- As pediatric strokes may be hemorrhagic, CT Head without contrast (CPT® 70450) is generally the initial study indicated.
 - MRI Brain without contrast (CPT® 70551) can be performed in lieu of initial CT if emergently available for evaluation of acute stroke symptoms.
- After the initial study, any of the following studies are indicated for initial evaluation of pediatric stroke:
 - MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553).
 - MRA Head without contrast (CPT® 70544) and Neck with contrast (CPT® 70548).
 - CTA Head (CPT® 70496) and Neck (CPT® 70498).
 - These cases should be forwarded for medical director review.

PEDHD-12.3: Pediatric Stroke Subsequent Imaging

- ➤ MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) is indicated for any new or worsening neurological findings or seizure activity.
- Most pediatric patients do not benefit from surveillance imaging after stroke, but specific surveillance imaging indications for specified conditions are listed in the disease-specific section.
 - MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553).
 - These cases should be forwarded for medical director review.

PEDHD-12.4: Moyamoya Disease

Initial imaging

▶ MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) MRA Head (CPT® 70544) and Neck (CPT® 70548) are indicated for all patients. Head and Neck CTA (CPT® 70496 and CPT® 70498) can be approved if MRI is contraindicated or not readily available.

Repeat imaging

- ➤ Head MRA (CPT® 70544) every 12 months. Head CTA (CPT® 70496) can be approved if MRI is contraindicated or not readily available.
- ➤ MRI Brain without contrast (CPT® 70551) every 12 months.

PEDHD-12.5: Sickle Cell Disease

Patients with sickle cell disease are at significantly increased risk for stroke and silent infarction, beginning at a very young age. Recent advances allow physicians to identify patients at high risk for stroke and begin a primary stroke prevention program.

- The following imaging is indicated for all sickle cell patients with a severe phenotype (Hgb SS or Hgb S β °):
 - Transcranial Doppler Ultrasound (CPT® 93886 or CPT® 93888) annually for all patients age 2 to 16.
 - A short interval repeat study is indicated for patients with conditional (170-199 cm/sec) flow results.
 - Transcranial Doppler is not indicated for patients with other phenotypes (Hgb SC, Hgb Sβ⁺).
 - Screening of asymptomatic sickle cell patients with MRI or MRA is no longer recommended.⁶

PEDHD-12.6: CNS Vasculitis and Stroke

- MRI Brain without and with contrast is the recommended initial study for all patients with vasculitis and suspected CNS involvement, whether primary or secondary.
 - A normal MRI Brain almost always completely excludes intracranial vasculitis
 - MRA Head (contrast as requested) is indicated for inconclusive MRI findings suggesting medium or large vessel vasculitis.
 - Patients with aggressive disease being treated with systemic therapy can have imaging approved for treatment response every 3 months during active treatment.
 - Annual surveillance imaging can be approved to detect progressive vascular damage that may require intervention

- 1. Adams RJ. TCD in sickle cell disease: an important and useful test. Pediatr Radiol. 2005; 35:229-234.
- 2. Kirton A, and deVeber G. Pediatric Stroke. *Nelson Textbook of Pediatrics, Chapter 601.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 2925-2933.
- 3. Rowland LP (Ed.). Merritt's Neurology.12th Ed. Philadelphia, Lippincott. 2010: 296-299
- Gemmete JJ, Davagnanam I, Toma AK, et al. Arterial ischemic stroke in children. *Neuroimag Clin N Am.* 2013 Nov; 23 (4): 781-798. Accessed October 20, 2017. http://www.neuroimaging.theclinics.com/article/S1052-5149(13)00033-6/pdf.
- 5. Stam J. Thrombosis of the cerebral veins and sinuses. *N Engl J Med.* 2005 Apr; 352: 1791-1798. Accessed October 20, 2017. http://www.nejm.org/doi/full/10.1056/NEJMra042354.
- 6. Scott RM and Smith ER. Moyamoya disease and moyamoya syndrome. *N Engl J Med.* 2009; 360:1226-1237. Accessed October 20, 2017. http://www.nejm.org/doi/full/10.1056/NEJMra0804622.
- 7. Buchanan GR, and Yawn BP (Co-Chairs). Evidence-based management of sickle cell disease expert panel report. 2014. National Heart, Lung, and Blood Institute. Accessed October 20, 2017. http://www.nhlbi.nih.gov/health-pro/quidelines/sickle-cell-disease-guidelines.
- 8. Moharir M, Shroff M, and Benseler SM. Childhood central nervous system vasculitis. *Neuroimag Clin N Am.* 2013 May; 23 (2): 293-308. Accessed October 20, 2017. http://www.neuroimaging.theclinics.com/article/S1052-5149(12)00212-2/pdf.
- Soliman M, Laxer R, Manson D, et al. Imaging of systemic vasculitis in childhood. *Pediatr Radiol.* 2015 Aug; 45 (8):1110-1125. Accessed October 20, 2017. https://link.springer.com/article/10.1007/s00247-015-3339-3?no-access=true.

PEDHD-13: Benign Brain Lesions	
PEDHD-13.1: Arachnoid Cysts	46
PEDHD-13.2: Pineal Cysts	46
PEDHD-13.3: Acoustic Neuromas	46

PEDHD-13.1: Arachnoid Cysts

Arachnoid cysts arise in the middle or posterior fossa, and the majority of lesions are discovered incidentally and do not require surgical intervention.

- MRI Brain without and with contrast (CPT® 70553) is indicated for initial evaluation of arachnoid cysts if not already completed.
- Repeat MRI Brain is not indicated for most patients with arachnoid cysts, but can be approved for the following:
 - Annual MRI Brain without and with contrast (CPT® 70553) until age 4 if diagnosed at a younger age.
 - New or worsening headache or focal neurologic deficits suggesting progression of cyst.
 - Preoperative planning.

PEDHD-13.2: Pineal Cysts

Pineal cysts are generally discovered incidentally and do not require surgical intervention.

- MRI Brain without and with contrast (CPT® 70553) is indicated for initial evaluation of pineal cysts if not already completed.
- Repeat MRI Brain is not indicated for most patients with pineal cysts, but can be approved for the following:
 - New or worsening headache or focal neurologic deficits suggesting progression of cyst.
 - Preoperative planning.

PEDHD-13.3: Acoustic Neuromas

> See PEDPND-2.2: Neurofibromatosis 2 for imaging guidelines in pediatric patients

- 1. Hervey-Jumper SL, Cohen-Gadol AA, and Maher CO. Neurosurgical management of congenital malformations of the brain. *Neuroimag Clin N Am.* 2011 Aug; 21 (3): 705-717. Accessed October 20, 2017. http://www.neuroimaging.theclinics.com/article/S1052-5149(11)00067-0/pdf.
- 2. Chtinis T, Guttman CR, Zaitsev A, et al. Quantitative MRI analysis in children with multiple sclerosis: a multicenter feasibility pilot study. *BMC Neurol*. 2013 Dec; 13: 173. Accessed October 20, 2017. https://link.springer.com/article/10.1186/1471-2377-13-173.
- 3. Al-Holou WN, Maher CO, Muraszko KM, et al. The natural history of pineal cysts in children and young adults. *J Neurosurg Pediatr.* 2010 Feb; 5 (2):162-166. Accessed October 20, 2017. http://thejns.org/doi/10.3171/2009.9.PEDS09297.
- 4. Raybaud C, and Barkovich AJ. Intracranial, orbital and neck masses in children. In: Barkovich AJ, Raybaud C, ed. *Pediatric Neuroimaging*. 5th ed. Philadelphia PA. Wolters Kluwer. 2012; 240-366.

PEDHD-14: Pediatric Demyelinating Diseas	ses
PEDHD-14.1: General Considerations	48
PEDHD-14.2: Multiple Sclerosis (MS)	48
PEDHD-14.3: Acute Disseminated Encephalomyelitis (ADEM)	48

PEDHD-14.1: General Considerations

MRI Brain without and with contrast (CPT® 70553) is the preferred imaging study for evaluation of pediatric demyelinating disease.

- MRI of the spinal cord (CPT® 72156 and CPT® 72157) without and with contrast is also indicated for evaluation of pediatric demyelinating disease.
- MRI of the lumbar spine (CPT® 72158) is not indicated unless the patient has a tethered cord or other anatomic abnormality causing caudal displacement of the filum terminalis.
- > CT imaging is generally not indicated in the evaluation of demyelinating disease.
- ➤ PET Brain (CPT® 78608 and CPT® 78609) and MR spectroscopy (CPT® 76390) are considered investigational for evaluation of pediatric demyelinating diseases.

PEDHD-14.2: Multiple Sclerosis (MS)

Multiple sclerosis is less common in children. About 4 % of MS cases are diagnosed before age 18, and only ~0.7 % of all MS cases begin before age 10.

Ataxia, optic neuritis, diplopia, and transverse myelitis are common presentations. MS can present as an acute encephalitis-like illness, especially in childhood.

Among children with suspected demyelinating diseases, the principal differential diagnosis is often between MS and acute disseminated encephalomyelitis.

- MRI (CPT® 70553) Brain and spinal cord (CPT® 72156 and CPT® 72157) without and with contrast is indicated for initial diagnosis in patients with clinical signs and/or symptoms suggestive of MS.
 - MRI (CPT® 70551) Brain and spinal cord (CPT® 72141 and CPT® 72146) without contrast can be approved if there is a contraindication to gadolinium administration.
- MRI (CPT® 70553) Brain and spinal cord (CPT® 72156 and CPT® 72157) without and with contrast is indicated every 6 months for disease monitoring.
 - MRI (CPT® 70551) Brain and spinal cord (CPT® 72141 and CPT® 72146) without contrast can be approved if there is a contraindication to gadolinium.

PEDHD-14.3: Acute Disseminated Encephalomyelitis (ADEM)

- ADEM has an acute onset, and is more common among younger children than MS, but the signs and symptoms overlap significantly, and distinguishing between MS and ADEM can be challenging based on clinical examination alone.
- MRI (CPT® 70553) Brain and spinal cord (CPT® 72156 and CPT® 72157) without and with contrast is indicated for initial diagnosis in patients with clinical signs and/or symptoms suggestive of ADEM.
 - MRI (CPT® 70551) Brain and spinal cord (CPT® 72141 and CPT® 72146) without contrast can be approved if there is a contraindication to gadolinium.
- MRI (CPT® 70553) Brain and spinal cord (CPT® 72156 and CPT® 72157) without and with contrast is indicated every 3 months for 1 year following diagnosis.
 - MRI (CPT® 70551) Brain and spinal cord (CPT® 72141 and CPT® 72146) without contrast can be approved if there is a contraindication to gadolinium.

 Most patients will have complete clinical recovery by 12 months, while stable MRI abnormalities (gliosis) may persist. These findings do not require additional imaging unless the patient develops new neurologic symptoms.

- Ness J. Demyelinating disorders of the central nervous system. Nelson Textbook of Pediatrics, Chapter 600. eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 2920-2925.
- 2. Verhey LH, Shroff M, and Banwell B. Pediatric multiple sclerosis pathological, clinical, and magnetic resonance imaging features. *Neuroimag Clin N Am.* 2013 May; 23 (2):227-243. Accessed October 20, 2017. http://www.neuroimaging.theclinics.com/article/\$1052-5149(12)00208-0/pdf.
- 3. Chtinis T, Guttman CR, Zaitsev A, et al. Quantitative MRI analysis in children with multiple sclerosis: a multicenter feasibility pilot study. *BMC Neurol.* 2013 Dec; 13: 173. Accessed October 20, 2017. https://link.springer.com/article/10.1186/1471-2377-13-173.
- 4. Van Haren K, and Waubant E. Therapeutic advances in pediatric multiple sclerosis. *J Pediatr.* 2013 Sep; 163 (3): 631-637. Accessed October 20, 2017. http://www.jpeds.com/article/S0022-3476(13)00445-9/fulltext.
- 5. Ketelslegers IA, Neuteboom RF, Boon M, et al. A comparison of MRI criteria for diagnosing pediatric ADEM and MS. *Neurology*. 2010 Mar; 74 (18): 1412; 1415. Accessed October 20, 2017. http://www.neurology.org/content/74/18/1412.
- Callen DJA, Shroff MM, Branson HM, et al. MRI in the diagnosis of pediatric multiple sclerosis. Neurology. 2009 Mar; 72 (11): 961-967. Accessed October 20, 2017. http://www.neurology.org/content/72/11/961.abstract.
- 7. Callen DJA, Shroff MM, Branson HM, et al. Role of MRI in the differentiation of ADEM from MS in children. *Neurology*. 2009 Mar; 72 (11): 968-973. Accessed October 20, 2017. http://www.neurology.org/content/72/11/968.
- 8. Marin SE, and Callen DJA. The magnetic resonance imaging appearance of monophasic acute disseminated encephalomyelitis: an update post application of the 2007 consensus criteria. *Neuroimag Clin N Am.* 2013 May; 23 (2): 245-266. Accessed October 20, 2017. http://www.neuroimaging.theclinics.com/article/S1052-5149(12)00209-2/fulltext.

<u>Imaging Guidelines</u> V20.0.2018

PEDHD-15: Pituitary Dysfunction	
PEDHD-15.1: General Considerations	51
PEDHD-15.2: Panhypopituitarism	51
PEDHD-15.3: Isolated Growth Hormone Deficiency	51
PEDHD-15.4: Diabetes Insipidus (DI) and Other Disorders of Anti-	
Diuretic Hormone	51
PEDHD-15.5: Precocious Puberty	52
PEDHD-15.6: Benign Pituitary Tumors	52
PEDHD-15.7: Pituitary Malignancies	53

PEDHD-15.1: General Considerations

The initial step in the evaluation of all potential pituitary masses is a detailed history, recent physical examination, and thorough neurological exam, including evaluation of the visual fields.

- Endocrine laboratory studies should be performed prior to considering advanced imaging.
- When pituitary imaging is indicated, MRI Brain without and with contrast (CPT® 70553) is the correct study.
 - One study (either brain MRI [CPT® 70553] or MRI Orbit, Face, Neck [CPT® 70543]) is adequate to image the pituitary. The ordering physician should specify that the study is specifically to evaluate the pituitary gland. The reporting of two CPT® codes, to image the pituitary, is not indicated.

PEDHD-15.2: Panhypopituitarism

Endocrine testing should be performed initially.

- MRI Brain without and with contrast (CPT® 70553) with special attention to the pituitary is indicated for newly diagnosed Panhypopituitarism.
- Patients with a normal pituitary on initial MRI do not need routine follow up imaging.
- Patients with mass lesions should have follow up imaging according to the guidelines for the specific diagnosis.

PEDHD-15.3: Isolated Growth Hormone Deficiency

Endocrine testing should be performed initially. For isolated growth hormone deficiency, two measurements of growth hormone with stimulation are performed.

- MRI Brain without and with contrast (CPT® 70553) with special attention to the pituitary is indicated for newly diagnosed isolated growth hormone deficiency.
- > Patients with a normal pituitary on initial MRI do not need routine follow up imaging.
- Patients with mass lesions should have follow up imaging according to the guidelines for the specific diagnosis.

PEDHD-15.4: Diabetes Insipidus (DI) and Other Disorders of Anti-Diuretic Hormone

The principal evaluation of ADH deficiency is by urine and blood electrolyte and osmolality testing - serum osmolality greater than 300 with urine osmolality less than 300. Deficiencies in ADH can either be central or nephrogenic.

Central Diabetes Insipidus (DI)

- ➤ MRI Brain without and with contrast (CPT® 70553) is indicated for newly diagnosed central DI.
- ➤ Head CT without contrast (CPT® 70450) with attention to the skull base may be approved with history of recent significant head trauma.
- Patients with a normal pituitary on initial MRI can have repeat MRI without and with contrast (CPT® 70553) every 12 months as germinomas may cause central DI while still too small to detect on imaging.
 - Serial measurement of β -hCG is also indicated for these patients, and MRI should be repeated if a significant rise in β -hCG is detected on screening.
- Patients with mass lesions should have follow up imaging according to the guidelines for the specific diagnosis.

Nephrogenic DI

Once this diagnosis is firmly established, further advanced imaging is usually not indicated.

Syndrome of Inappropriate Antidiuretic Hormone Secretion (SIADH)

Laboratory studies should be obtained prior to considering advanced imaging—urine osmolality should be high and serum osmolality low.

- ➤ MRI Brain without and with contrast (CPT® 70553) is indicated for initial evaluation of unexplained central SIADH.
- > Patients with a normal pituitary on initial MRI do not need routine follow up imaging.
- Patients with mass lesions should have follow up imaging according to the guidelines for the specific diagnosis.

PEDHD-15.5: Precocious Puberty

Defined as the appearance of secondary sexual characteristics before age 8 in girls and before age 9 in boys.

When precocious puberty is documented on physical examination, endocrine lab studies are not necessary prior to advanced imaging.

- ➤ Brain MRI Brain without and with contrast (CPT® 70553) is indicated for initial evaluation of any child with documented precocious puberty, following ultrasound of the abdomen (CPT® 76700) in both genders and ultrasound of the pelvis (CPT® 76856) in girls.
- Patients with a normal pituitary on initial MRI do not need routine follow up imaging.
- Patients with mass lesions should have follow up imaging according to the guidelines for the specific diagnosis.

PEDHD-15.6: Benign Pituitary Tumors

Benign pituitary tumor indications in pediatric patients are identical to those for adult patients. See <u>HD-19: Pituitary</u> for imaging guidelines.

PEDHD-15.7: Pituitary Malignancies

See <u>PEDONC-4.10: Craniopharyngioma and Pituitary Tumors or PEDONC-18:</u> <u>Histiocytic Disorders</u> for imaging guidelines

- 1. Seidenwurm DJ, Wippold FJ, Cornelius RS, et al. Neuroendocrine Imaging. *ACR Appropriateness Criteria*®. *J Am Coll Radiol.* 2012; 9 (5): 315-324. Accessed October 20, 2017. http://www.ncbi.nlm.nih.gov/pubmed/22554628.
- 2. Parks JS, and Felner EI. Disorders of the hypothalamus and pituitary gland. *Nelson Textbook of Pediatrics, Chapter 556.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 2635-2662.

PEDHD-16: Pediatric Ear Disorders	
PEDHD-16.1: Hearing Loss	55
PEDHD-16.2: Ear Pain	55
PEDHD-16.3: Cholesteatoma	56
PEDHD-16.4: Vertigo	56
PEDHD-16.5: Tinnitus	57

PEDHD-16.1: Hearing Loss

A recent (within 60 days) evaluation including a detailed history, physical examination (including otoscopic examination), and age-appropriate audiology testing should be performed on any child with known or suspected hearing loss prior to considering advanced imaging. The selection of imaging testing will depend on the age of the child and type of hearing loss.

- Temporal bone CT without contrast (CPT® 70480) is indicated for the following:
 - Conductive hearing loss of any cause.
 - Preoperative planning for resection of mass lesion or cochlear implant placement.
 - Sensorineural hearing loss in patients who cannot safely undergo MRI.
 - Mixed conductive and sensorineural hearing loss.
 - Congenital hearing loss.
 - Total deafness.
- ➤ MRI Brain without and with contrast (CPT® 70553) with attention to internal auditory canals (included in CPT® 70553 and does not require a separate CPT code) is indicated for the following:
 - Conductive hearing loss secondary to known or suspected mass lesion.
 - Preoperative planning for resection of mass lesion or cochlear implant placement.
 - Sensorineural hearing loss of any cause.
 - Mixed conductive and sensorineural hearing loss.
 - Congenital hearing loss.
 - Total deafness.
 - Patients requiring sedation should generally not have non-contrast MRI studies. See PEDHD-1.3: Pediatric Head Imaging Modality General Considerations.

PEDHD-16.2: Ear Pain

A recent (within 60 days) evaluation including a detailed history, physical examination (including otoscopic examination), should be performed on any child with ear pain prior to considering advanced imaging. Common causes of ear pain include external and middle ear infections, dental problems, sinus infection, neck problems, tonsillitis, and pharyngitis.

- Advanced imaging is not indicated in the overwhelming majority of pediatric patients with ear pain.
- > CT scan temporal bone without contrast (CPT® 70480) or without and with contrast (CPT® 70482), OR, MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553), OR MRI Orbits/Face/Neck without and with contrast (CPT® 70543) is indicated for the following:
 - Persistent ear pain without obvious cause.
 - Clinical suspicion for complicated or invasive infection such as mastoiditis.
 - Clinical suspicion of mass lesion causing ear pain.
 - Significant trauma with concern for hematoma formation.
 - Preoperative planning.

Patients requiring sedation should generally not have non-contrast MRI studies.
 See PEDHD-1.3: Pediatric Head Imaging Modality General Considerations.

PEDHD-16.3: Cholesteatoma

Cholesteatomas are expansive cysts of the middle ear filled with cellular debris. They can be congenital or arise from recurrent middle ear infections or trauma to the tympanic membrane. Hearing loss is usually conductive, although if the lesion is large enough combined conductive and sensorineural hearing loss may be present. Otoscopic exam findings and symptoms may include painless drainage from the ear or chronic/recurrent ear infections.

- ➤ CT scan temporal bone without contrast (CPT® 70480) or without and with contrast (CPT® 70482), OR MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553), OR MRI Orbits/Face/Neck without and with contrast (CPT® 70543) is indicated for preoperative evaluation in cholesteatoma patients.
- ➤ CT scan temporal bone without contrast (CPT® 70480) or without and with contrast (CPT® 70482), OR MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553), OR MRI Orbits/Face/Neck without and with contrast (CPT® 70543) is indicated one time post-operatively to exclude residual or regrown cholesteatoma to avoid the need for a second-look surgery.
 - Patients requiring sedation should generally not have non-contrast MRI studies.
 See PEDHD-1.3: Pediatric Head Imaging Modality General Considerations.

PEDHD-16.4: Vertigo

Isolated vertigo is an uncommon complaint during childhood. Middle ear/Eustachian tube problems are the most common cause of isolated vertigo in children. A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination (including otoscopic examination), should be performed on any child with vertigo prior to considering advanced imaging.

- If physical examination is otherwise normal and the vertigo responds to treatment, advanced imaging is not indicated.
- MRI Brain without and with contrast with attention to internal auditory canals (CPT[®] 70553) is indicated for the following:
 - Vertigo with associated headache or ataxia.
 - Vertigo associated with tinnitus.
 - Vertigo that does not respond to vestibular treatment.
 - Patients requiring sedation should generally not have non-contrast MRI studies.
 See <u>PEDHD-1.3: Pediatric Head Imaging Modality General Considerations</u>.

PEDHD-16.5: Tinnitus

Tinnitus without hearing loss is a less common complaint during childhood. Children with hearing loss and tinnitus should be imaged according to <u>PEDHD-16.1: Hearing Loss</u>. A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination (including otoscopic examination), and age-appropriate audiology testing should be performed on any child with known or suspected tinnitus prior to considering advanced imaging.

- Advanced imaging is not indicated in the overwhelming majority of pediatric patients with isolated tinnitus and normal hearing.
- CT scan temporal bone without contrast (CPT® 70480) or without and with contrast (CPT® 70482), OR MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553), OR MRI Orbits/Face/Neck without and with contrast (CPT® 70543) is indicated for the following:
 - Clinical suspicion of mass lesion causing tinnitus.
 - Persistent tinnitus after recent significant trauma.
 - Patients requiring sedation should generally not have non-contrast MRI studies.
 See PEDHD-1.3: Pediatric Head Imaging Modality General Considerations.

- 1. Haddad J, and Keesecker S. The ear. *Nelson Textbook of Pediatrics, Chapter 636.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 3069-3103.
- 2. Angtuaco EJ, Wippold FJ, Cornelius RS, et al. Hearing loss and/or vertigo. *ACR Appropriateness Criteria*®. 2013: 1-14. Accessed October 20, 2017. https://acsearch.acr.org/docs/69488/Narrative/.
- 3. Minovi A, and Dazert S. Diseases of the middle ear in childhood. *GMS Curr Top Otorhinolaryngol Head Neck Surg.* 2014 Dec; 13:1-29. Accessed October 20, 2017. http://www.egms.de/static/pdf/journals/cto/2014-13/cto000114.pdf.
- Savastano M, Marioni G, and de Filippis C. Tinnitus in children without hearing impairment. Int J Pediatr Otorhinolaryngol. 2009 Dec; 73S: S13-S15. Accessed October 20, 2017. https://www.sciencedirect.com/science/article/pii/S0165587609700035.

PEDHD-17: Autism Spectrum Disorders

The group of diagnoses, including Asperger syndrome, are classified as pervasive development disorders (PDD). These diagnoses are established on clinical criteria, and no imaging study can confirm the diagnosis.

Comprehensive evaluation for autism might include history, physical exam, audiology evaluation, speech, language, and communication assessment, cognitive and behavioral assessments, and academic assessment.

- MRI Brain without and with contrast (CPT® 70553) is indicated for new or worsening focal neurologic findings documented on a physical examination within 60 days of the imaging request.
 - Patients requiring sedation should generally not have non-contrast MRI studies.
 See PEDHD-1.3: Pediatric Head Imaging Modality General Considerations.
- PET imaging is considered investigational in the evaluation of patients with autism spectrum disorders.

- 1. Raviola G, Trieu ML, Walter HJ, et al. Autism spectrum disorder. *Nelson Textbook of Pediatrics, Chapter 30.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 176-183.
- 2. Baker E, and Jeste SS. Diagnosis and management of autism spectrum disorder in the era of genomics. *Pediatr Clin N Am.* 2015 June; 62 (3):607-618. Accessed October 20, 2017. http://www.pediatric.theclinics.com/article/S0031-3955(15)00023-1/pdf.
- 3. Zürcher NR, Bhanot A, McDougle CJ, et al. A systematic review of molecular imaging (PET and SPECT) in autism spectrum disorder: current state and future research opportunities. *Neuroscience and Biobehavioral Reviews* 2015; 52: 56-73.

PEDHD-18: Behavioral and Psychiatric Disorders

- Behavioral and psychiatric disorders of childhood or adolescence generally require no advanced imaging for diagnosis or management.
 - MRI Brain without and with contrast (CPT® 70553) is indicated for new or worsening focal neurologic findings documented on a physical examination within 60 days of the imaging request.
 - Patients requiring sedation should generally not have non-contrast MRI studies. See <u>PEDHD-1.3: Pediatric Head Imaging Modality General</u> Considerations.

References

1. Behavioral and Psychiatric Disorders. *Nelson Textbook of Pediatrics, Chapters 20-31.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 124-191.

FEDHU-19. Intellectual Disability, Celebral	
Palsy, and Developmental Motor Delay	
PEDHD-19.1: Intellectual Disability	61
PEDHD-19.2: Cerebral Palsy	61
PEDHD-19.3: Developmental Motor Delay	61

PEDHD-19.1: Intellectual Disability

Intellectual disability was formerly known as mental retardation, and may be primary or secondary to a variety of heterogeneous disorders.

- Brain MRI without and with contrast (CPT® 70553) is indicated for new or worsening focal neurologic findings documented on a physical examination within 60 days of the imaging request.
- Patients requiring sedation should generally not have non-contrast MRI studies. See PEDHD-1.3: Pediatric Head Imaging Modality General Considerations.

PEDHD-19.2: Cerebral Palsy

Many patients with intellectual disability also have cerebral palsy, but not all patients with cerebral palsy have intellectual disability.

Cerebral palsy is a static motor encephalopathy caused by a variety of entities spanning developmental, metabolic, genetic, infectious, ischemic, and other acquired etiologies.

- ➤ Brain MRI without and with contrast (CPT® 70553) is indicated for:
 - Initial evaluation of newly diagnosed cerebral palsy.
 - New or worsening focal neurologic findings documented on a physical examination within 60 days of the imaging request, including the presence of developmental delay.
 - Patients requiring sedation should generally not have non-contrast MRI studies.
 See PEDHD-1.3: Pediatric <u>Head Imaging Modality General Considerations</u>.

PEDHD-19.3: Developmental Motor Delay

There are many causes for developmental motor delay. Patients with motor delay can have decreased, normal, or increased muscular tone. Patients with low or normal tone do not require imaging unless they have focal neurologic findings.

- ➤ Brain MRI without and with contrast (CPT® 70553) is indicated for:
 - Initial evaluation of newly diagnosed developmental motor delay with increased muscle tone.
 - New or worsening focal neurologic findings documented on a physical examination within 60 days of the imaging request.
 - Patients requiring sedation should generally not have non-contrast MRI studies.
 See PEDHD-1.3: Pediatric Head Imaging Modality General Considerations.

- 1. Shapiro BK, and Batshaw ML. Intellectual Disability. *Nelson Textbook of Pediatrics, Chapter 36.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 216-222.
- 2. Johnston MV. Encephalopathies. *Nelson Textbook of Pediatrics, Chapter 598.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 2896-2910
- 3. Noritz GH, and Murphy NA. Motor delays: early identification and evaluation. *Pediatrics*. 2013 May; 131 (6). Accessed October 20, 2017. http://pediatrics.aappublications.org/content/131/6/e2016.

PEDHD-20: Ataxia

Ataxia refers to an abnormally ill-coordinated or unsteady gait for age. "Limb ataxia" refers to impaired coordination (for age) of limbs, especially arms. Developmental failure to acquire the ability to walk is a form of developmental delay, not ataxia.

(See: <u>PEDHD-19: Intellectual Disability, Cerebral Palsy, and Developmental Motor Delay</u>)

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination with a thorough neurologic examination, and appropriate laboratory studies should be performed prior to considering advanced imaging, unless the patient is undergoing guideline-supported scheduled follow-up imaging evaluation.
- ➤ Brain MRI without and with contrast (CPT® 70553) can be performed to evaluate ataxia, hereditary ataxia, and slowly progressive ataxia.
 - Cervical spine MRI without contrast (CPT® 72141) or without and with contrast (CPT® 72156) is indicated if brain MRI is non-diagnostic.
 - Patients requiring sedation should generally not have non-contrast MRI studies. See <u>PEDHD-1.3: Pediatric Head Imaging Modality General</u> Considerations.
- ➤ CT Head without and with contrast (CPT® 70470) or with contrast (CPT® 70460) is indicated for patients who have a contraindication to MRI.
 - CT should not be used in place of MRI solely to avoid sedation in young children because MRI is superior for imaging the posterior fossa.
- CT Head without contrast (CPT® 70450) or without and with contrast (CPT® 70470) or MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) is indicated for patients with acute ataxia following significant head trauma.

- 1. Broderick DF, Wippold FJ, Cornelius RS, et al. Ataxia. *ACR Appropriateness Criteria*®. 2012: 1-16. Accessed October 20, 2017. https://acsearch.acr.org/docs/69477/Narrative/.
- 2. Prabhu SP, and Young-Poussaint Ty. Pediatric central nervous system emergencies. *Neuroimag Clin N Am.* 2010 Nov; 20 (4):663-683. Accessed October 20, 2017. http://www.neuroimaging.theclinics.com/article/S1052-5149(10)00080-8/pdf.

64

PEDHD-21.1: Imaging

Initial evaluation of epistaxis (nosebleed), including recurrent epistaxis that is refractory to medical management is by direct or endoscopic visualization of the relevant portions of the upper airway.

- If a mass lesion is detected on direct visualization, any one of the following imaging studies is indicated:
 - CT Maxillofacial without contrast (CPT® 70486) or without and with contrast (CPT® 70488).
 - MRI Orbits/Face/Neck without and with contrast (CPT® 70543).

Reference

1. Haddad J, and Keesecker S. Acquired disorders of the nose. *Nelson Textbook of Pediatrics, Chapter* 377. eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 2008-2010

PEDHD-22: Pseudotumor Cerebri

Pseudotumor cerebri indications in pediatric patients are identical to those for adult patients. See <u>HD-17: Papilledema/Pseudotumor Cerebri</u> for imaging guidelines.

PEDHD-23: Cranial Neuropathies

- ➤ MRI Brain without and with contrast (CPT® 70553) is indicated for all patients with new or worsening specific cranial nerve abnormalities.
- ➤ MRI Neck without and with contrast (CPT® 70543) is also indicated for patients with abnormalities in cranial nerves IX, X, XI, or XII.

- 1. Wippold FJ, Cornelius RS, Aiken AH, et al. Cranial neuropathy. *ACR Appropriateness Criteria*®. 2017:1-22.
- 2. Rubin M. Overview of neuro-ophthalmologic and cranial nerve disorders. Merck Manual. 2014. Accessed October 20, 2017. https://www.merckmanuals.com/professional/neurologic-disorders/neuro-ophthalmologic-and-cranial-nerve-disorders/overview-of-neuro-ophthalmologic-and-cranial-nerve-disorders.

PEDHD-24: Pediatric Sleep Disorders

- See <u>Pediatric Sleep Guidelines</u> for sleep study indications.
- Advanced imaging is not indicated for the following:
 - Parasomnias.
 - Bed wetting (if child is otherwise neurologically normal).
 - Insomnia.
 - Narcolepsy.
 - Restless Leg Syndrome (polysomnography is useful).
- For Obstructive Sleep Apnea, endoscopic examination of the upper airway and lateral upper airway x-rays should be performed initially.
 - CT Maxillofacial without contrast (CPT® 70486) may be indicated for evaluation of obstructive anatomy if operative intervention is being considered.
- For Obstructive Sleep Apnea, endoscopic examination of the upper airway and lateral upper airway x-rays should be performed initially.
 - CT Maxillofacial without contrast (CPT® 70486) may be indicated.
- ➤ For Central Sleep Apnea, MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) is indicated if the clinical picture and/or polysomnography study suggests central sleep apnea.

References

1. Owens JA. Sleep medicine. *Nelson Textbook of Pediatrics, Chapter 19.* eds Kliegman RM, Stanton BF, St. Geme JW III, et al. 20th edition. 2016, pp 111-123.

PEDHD-25: Temporomandibular Joint (TMJ) Imaging in Children

There is a paucity of clinical symptoms and poor sensitivity of conventional x-rays in diagnosing TMJ arthritis in pediatric patients with arthritis.

TMJ MRI (CPT® 70336) is indicated annually for detecting silent TMJ arthritis in children with juvenile idiopathic arthritis (JIA).

- Zwir LM, Terreri MT, Sousa SA, et al. Are temporomandibular joint signs and symptoms associated with magnetic resonance imaging finings in juvenile idiopathic arthritis patients? A longitudinal study. *Clin Rheumatol*. 2015 Dec; 34 (12) 057-2063. Accessed October 20, 2017. https://link.springer.com/article/10.1007/s10067-015-2925-y.
- 2. Arabshahi B, and Cron RQ. Temporomandibular joint arthritis in juvenile idiopathic arthritis: the forgotten joint. *Curr Opin Rheumatol.* 2006 Sep; 18 (5): 490-495. Accessed October 20, 2017. https://www.ncbi.nlm.nih.gov/pubmed/16896288?dopt=Abstract.

PEDHD-26: Tourette's Syndrome

The diagnosis of Tourette's syndrome is made clinically and advanced neuroimaging is not indicated for either diagnosis or management.

References

 Serajee FJ, and Mahbubl AHM. Advances in tourette syndrome diagnosis and treatment. *Pediatr Clin N Am.* 2015 June; 62 (3): 687-701. Accessed October 20, 2017. http://www.pediatric.theclinics.com/article/S0031-3955(15)00027-9/pdf.

PEDHD-27: Tuberous Sclerosis

> See PEDONC-2.9: Tuberous Sclerosis Complex (TSC) for imaging guidelines.

PEDHD-28: Von Hippel Lindau Syndrome (VHL)

> See PEDONC-2.10: Von Hippel-Lindau Syndrome (VHL) for imaging guidelines.

PEDHD-29: CNS Infection

- CNS infection imaging indications in pediatric patients are similar to those for adult patients. See <u>HD-14: CNS Infection</u> for imaging guidelines.
- Pediatric-specific imaging considerations include suspected congenital brain infection and neonatal meningitis. The common causes of prenatal infections of the central nervous system are cytomegalovirus, *Toxoplasma gondii*, herpes simplex type 2 virus and most recently zika virus. The findings suggesting prenatal brain infection include microcephaly, microphthalmia, chorioretinitis, cataracts, hypotonia, and seizures. The following are performed for congenital brain infections:
 - The following imaging is considered for newborn infants with suspected prenatal brain infection regardless of inciting organism. (For additional information see CDC's Areas with risk of Zika site: https://www.cdc.gov/zika/geo/active-countries.html.)
 - Head ultrasound (CPT[©] 76506) can be approved as an initial imaging study.
 - If the ultrasound is abnormal, MRI Brain without and with contrast (CPT® 70553) is indicated.
 - Patients requiring sedation should generally not have only non-contrast MRI studies. See <u>PEDHD-1.3: Pediatric Head Imaging Modality</u> General Consideration.
 - Newborn infants with microcephaly should be evaluated as discussed in <u>PEDHD-7: Macrocephaly, Microcephaly, and Hydrocephalus</u>.
- Neonatal meningitis is most often caused by bacterial pathogens and usually occurs as a complication of sepsis in the first week of life. In older infants and children, meningeal inoculation occurs secondary to hematogenous spread or penetrating trauma.
- The following imaging is considered for newborns or older infants with and open fontanelle and suspected meningitis.
 - Head ultrasound (CPT[©] 76506) can be approved as an initial imaging study.
 - ◆ If the ultrasound is abnormal, MRI Brain without and with contrast (CPT® 70553) is indicated.

- 1. Hedlund G, Balfe JE, and Barkovich AJ. Infections of the developing and mature nervous system. In: Barkovich AJ, Raybaud C. eds *Pediatric Neuroimaging*. 5th ed. Philadelphia PA. Wolters Kluwer. 2012; 954-1050.
- 2. De Vries LS, and Volpe JJ. Viral, protozoan, and related intracranial infections. In: Volpe JJ, ed. *Volpe's Neurology of the Newborn*. 6th ed. Philadelphia: Elsevier. 2018; 973-1049.
- 3. Levine D, Jani JC, Castro-Aragon I, et al. How does imaging of congenital Zika compare with imaging of other TORCH infections? *Radiology*. 2017; 285: 744-761.
- 4. De Oliveria Melo AS, Aquiar RS, Amorim MM, et al. Congenital Zika virus infection: beyond neonatal microcephaly. *JAMA Neurol*.2016 Dec 1; 73: 1407-1416.
- 5. Vepraskas SA. Zika Virus an emerging arbovirus associated with fetal abnormalities. CDC's response to Zika. Accessed October 20, 2017. https://www.cdc.gov/zika/pdfs/pediatric-evaluation-follow-up-tool.pdf.
- 6. Rabe I, Meaney-Delman D, and Moore CA. "Zika Virus What Clinicians Need to Know." clinician outreach and communication activity call. Centers for Disease Control and Prevention. 26 Jan. 2016. Available at:
 - http://coursewareobjects.elsevier.com/objects/elr/ExpertConsult/Kliegman/nelson20e/updates/CDC_presentation_01262016.pdf

PEDHD-30: Scalp and Skull Lesions

- Scalp and skull lesion imaging indications in pediatric patients are identical to those for adult patients with the exception of neonates. See <u>HD-20: Scalp and Skull Lesions</u> for imaging guidelines.
 - In neonates and young infants, scalp masses include:
 - congenital lesions (cephalocele-discussed above, dermoid cysts, epidermoid cyst),
 - vascular lesions (hemangioma, sinus pericranii), and
 - extracranial hemorrhage related to birth trauma (caput succedaneum, cephalohematoma, subgaleal hematoma).
 - After the first year of life, malignant tumors, such as Langerhans cell histiocytosis metastases from neuroblastoma and rhabdomyosarcoma are an additional cause of a scalp mass.
- The following imaging is considered for newborns with palpable scalp and skull lesions.
 - Head ultrasound (CPT® 76506) can be approved as an initial imaging study.
 - If the ultrasound is abnormal and associated anomalies are suspected, CT or MRI Brain without and with contrast (CPT® 70553) is indicated.

References

1. Siegel MJ. Brain. In: *Pediatric sonography*. 5th ed. Philadelphia. Wolters Kluwer. 2018 (in press).

PEDHD-31: Eye Disorders

> Eye disorder imaging indications in pediatric patients are identical to those for adult patients. See **HD-32**: **Eye Disorders** for imaging guidelines.