

# Cigna Medical Coverage Policies – Radiology Pediatric Head Imaging Guidelines

Effective February 01, 2024



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## Instructions for use

The following coverage policy applies to health benefit plans administered by Cigna. Coverage policies are intended to provide guidance in interpreting certain standard Cigna benefit plans and are used by medical directors and other health care professionals in making medical necessity and other coverage determinations. Please note the terms of a customer's particular benefit plan document may differ significantly from the standard benefit plans upon which these coverage policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a coverage policy.

In the event of a conflict, a customer's benefit plan document always supersedes the information in the coverage policy. In the absence of federal or state coverage mandates, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of:

1. The terms of the applicable benefit plan document in effect on the date of service
2. Any applicable laws and regulations
3. Any relevant collateral source materials including coverage policies
4. The specific facts of the particular situation

Coverage policies relate exclusively to the administration of health benefit plans. Coverage policies are not recommendations for treatment and should never be used as treatment guidelines.

This evidence-based medical coverage policy has been developed by eviCore, Inc. Some information in this coverage policy may not apply to all benefit plans administered by Cigna.

These guidelines include procedures eviCore does not review for Cigna. Please refer to the [Cigna CPT code list](#) for the current list of high-tech imaging procedures that eviCore reviews for Cigna.

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# General Guidelines (PEDHD-1)

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# Procedure Codes Associated with Head Imaging

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| Procedure Codes Associated with Head Imaging                         |       |
|--|-------|
| MRI  | CPT®  |
| MRI Brain without contrast   | 70551 |
| MRI Brain with contrast (rarely used)                                | 70552 |
| MRI Brain without and with contrast                                  | 70553 |
| MRI Orbit, Face, Neck without contrast                               | 70540 |
| MRI Orbit, Face, Neck with contrast (rarely used)                    | 70542 |
| MRI Orbit, Face, Neck without and with contrast                      | 70543 |
| MRI Temporomandibular Joint (TMJ)                                    | 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®  |
| MRA Head without contrast  | 70544 |
| MRA Head with contrast   | 70545 |
| MRA Head without and with contrast                                   | 70546 |
| MRA Neck without contrast  | 70547 |
| MRA Neck with contrast   | 70548 |
| MRA Neck without and with contrast                                   | 70549 |
| CT   | CPT®  |
| CT Head without contrast   | 70450 |
| CT Head with contrast  | 70460 |
| CT Head without and with contrast                                    | 70470 |

| Procedure Codes Associated with Head Imaging                                     |                        |
|--|------------------------|
| CT Orbits without contrast (includes temporal bone and mastoid)                  | 70480                  |
| CT Orbits with contrast (includes temporal bone and mastoid)                     | 70481                  |
| CT Orbits without and with contrast (includes temporal bone and mastoid)         | 70482                  |
| CT Maxillofacial without contrast (includes sinuses, jaw, and mandible)          | 70486                  |
| CT Maxillofacial with contrast (includes sinuses, jaw, and mandible)             | 70487                  |
| CT Maxillofacial without and with contrast (includes sinuses, jaw, and mandible) | 70488                  |
| CT Neck without contrast (includes jaw, and mandible)                            | 70490                  |
| CT Neck with contrast (includes jaw, and mandible)                               | 70491                  |
| CT Neck 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                  |
| <b>CTA</b>   | <b>CPT<sup>®</sup></b> |
| CTA Head   | 70496                  |
| CTA Neck   | 70498                  |

| <b>Ultrasound</b>   | <b>CPT<sup>®</sup></b> |
|---|------------------------|
| Echoencephalography (Head or Cranial Ultrasound)  | 76506                  |
| Ophthalmic ultrasound, diagnostic; B-scan & quantitative A-scan performed same encounter      | 76510                  |
| Ophthalmic ultrasound, diagnostic; quantitative A-scan only                                   | 76511                  |
| 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 |
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| Transcranial Doppler study of the intracranial arteries; emboli detection without intravenous microbubble injection | 93892 |
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# General Guidelines (PEDHD-1.0)

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- A pertinent clinical evaluation including a detailed history, physical examination with a thorough neurologic examination since the onset or change in signs and/or symptoms<sup>29</sup>, appropriate laboratory studies and basic imaging such as plain radiography or ultrasound should be performed prior to considering advanced imaging (CT, MR, Nuclear Medicine), unless the individual is undergoing guideline-supported scheduled imaging evaluation. A meaningful technological contact (telehealth visit, telephone call, electronic mail or messaging) since the onset or change in signs and/or symptoms<sup>29</sup>, can serve as a pertinent clinical evaluation.
  - A detailed neurological exam is required prior to advanced imaging except in the following scenarios:
    - Individual is undergoing a guideline-supported scheduled follow-up imaging evaluation
    - Tinnitus, TMJ, Sinus or mastoid disease, ear pain, hearing loss, eye disease, papilledema<sup>30</sup>, dental requests and epistaxis. (A relevant physical exam is still required.)
    - The request is from a neurologist, neurosurgeon, endocrinologist, otolaryngologist, or ophthalmologist who has evaluated the individual since onset of symptoms.
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic individuals for disorders involving the head is not supported. Advanced imaging of the head is only indicated in individuals who have documented active clinical signs or symptoms of disease involving the head.
- 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 individual management or treatment decisions.



# Pediatric Head Imaging Age Considerations (PEDHD-1.1)

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- 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 individual age, comorbidities, and differences in disease natural history between children and adults.
- Individuals who are 18 years old or younger<sup>31</sup> and any conditions not specifically discussed in the General Head Imaging Guidelines should be imaged according to the Pediatric Head Imaging Guidelines. Any conditions not specifically discussed in the Pediatric Head Imaging Guidelines should be imaged according to the General Head Imaging Guidelines. Individuals who are >18 years old should be imaged according to the General Head Imaging Guidelines, except where directed otherwise by a specific guideline section.

# Pediatric Head Imaging Appropriate Clinical Evaluation (PEDHD-1.2)

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## 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 is appropriate unless there is clear documentation of a need for the additional codes to cover all necessary body areas.
- See General Guidelines - Anatomic Issues (HD-1.1) in the Head Imaging Guidelines for the correct coding of these studies.

# Pediatric Head Imaging Modality General Considerations (PEDHD-1.3)

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- 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 required for MRI acquisition and the need to minimize individual movement, anesthesia is usually required for almost all infants (except neonates) and young children (age <7 years) as well as older children with delays in development or maturity. This anesthesia may be administered via oral or intravenous routes. In this individual population, MRI sessions should be planned with a goal of minimizing anesthesia exposure by adhering to the following considerations:
    - MRI procedures can be performed without and/or with contrast use as supported by these condition based guidelines. If intravenous access will already be present for anesthesia administration and there is no contraindication for using contrast, imaging without and with contrast may be appropriate if requested. By doing so, the requesting provider may avoid repetitive anesthesia administration to perform an MRI with contrast if the initial study without contrast is inconclusive.
  - 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 multiple body areas are supported by eviCore guidelines for the clinical condition being evaluated, MRI/MRA 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.
  - CT Head without contrast (CPT® 70450) may be indicated for:
    - Mass effect

- Blood/blood products
- Urgent/emergent settings due to availability and speed of CT
- Trauma
- Recent hemorrhage, whether traumatic or spontaneous
- Bony structures of the head evaluations including dystrophic calcifications
- Hydrocephalus evaluation and follow-up
  - Some centers use limited non-contrast “fast or rapid MRI” (CPT® 70551) to minimize radiation exposure in children - these requests are appropriate.
- Prior to lumbar puncture in individuals with cranial complaints
- Scenarios in which MRI is contraindicated (i.e. pacemakers, ICDs, cochlear implants, aneurysm clips, orbital metallic fragments, etc.)
- CT and MR Angiography (CTA and MRA) Head and Neck
  - MRA Head may be performed without contrast (CPT® 70544), with contrast (CPT® 70545), or without and with contrast (CPT® 70546).
  - CTA Head is performed without and with contrast (CPT® 70496).
  - MRA Neck may be done either without contrast (CPT® 70547), with contrast (CPT® 70548), or without and with contrast (CPT® 70549), depending on facility preference and protocols and type of scanner.
  - CTA Neck is done with and without contrast (CPT® 70496)
- Indications for CTA and MRA Head and Neck vessels include, but are not limited to the following:
  - MRA is the preferred study in children unless contraindicated:
    - Pulsatile tinnitus
    - Hemifacial spasm if consideration for surgical decompression
    - Evaluation of stroke or TIA (See Pediatric Stroke Initial Imaging (PEDHD-12.2), Pediatric Stroke Subsequent Imaging (PEDHD-12.3), Moyamoya Disease (PEDHD-12.4), Sickle Cell Disease (PEDHD-12.5) and CNS Vasculitis and Stroke (PEDHD-12.6) including collateral assessment)
    - Follow up of known cerebral artery stenosis
    - Trigeminal neuralgia failed medical therapy
    - Cerebral sinus thrombosis suspected with increased intracranial pressure (refractory headaches, papilledema, diagnosis of pseudotumor cerebri)
    - Aneurysm suspected with acute “thunderclap” headache syndrome and appropriate screening or evaluation of known subarachnoid hemorrhage and pseudoaneurysms (may be appropriate to limit CTA to include only the head to avoid unnecessary radiation to the individual) (See Pediatric Intracranial Aneurysms (PEDHD-10.1))
    - Noninflammatory vasculopathy, including radiation vasculopathy (See Long Term Pediatric Cancer Survivors (PEDONC-19) in the Pediatric Oncology Imaging Guidelines)
    - Traumatic vascular injuries

- Vascular malformations, vascular anatomic variants and fistulas (See Pediatric Intracranial Arteriovenous Malformations (AVM) (PEDHD-10.2))
- Arterial, including carotid dissections
- Tumors of vascular origin or involving vascular structures
- Surgical and radiation therapy localization, planning and neuronavigation
- Evaluation for vascular intervention and follow-up including postsurgical/posttreatment vascular complications
- Intra-cranial pre-operative planning if there is concern of possible vascular involvement or risk for vascular complication from procedure
- Vasculitis and collagen vascular disease (See CNS Vasculitis and Stroke (PEDHD-12.6))
- Sickle cell disease (See Sickle Cell Disease (PEDHD-12.5))
- Moyamoya disease (See Moyamoya Disease (PEDHD-12.4))
- MRA Head without contrast (CPT® 70544), MRA head with contrast (CPT® 70545) **OR** MRA head without and with contrast (CPT® 70546) **OR** CTA Head (CPT® 70496) for follow up of aneurysm clipping or coiling procedures (See Intracranial Aneurysms (HD-12.1) in the Head Imaging Guidelines)
- CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart (there is no specific code for CT/MR venography):
  - If arterial and venous CT or MR studies are both performed in the same session, only one CPT® code should be used to report both procedures
- MRA without and with contrast with venous sinus thrombosis to differentiate total from subtotal occlusion
- NOTE: Evaluation of posterior circulation disease requires both neck and head MRA/CTA to visualize the entire vertebral-basilar system.
- 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.
- 3D Rendering
  - CPT® 76377 (3D rendering requiring image post-processing on an independent workstation) can be considered in the following clinical scenarios:
    - Bony conditions:
      - Evaluation of congenital skull abnormalities in newborns, infants, and toddler (usually for preoperative planning)
      - Complex joint fractures or pelvis fractures
      - Spine fractures (usually for preoperative planning)
      - Complex facial fractures
    - Preoperative planning for other complex surgical cases
    - Cerebral angiography

- 3D Rendering (CPT® 76377) may be used for surgical planning and surgical follow up after craniotomy when ordered by surgical specialist.
- 3D Rendering indications in pediatric head imaging are identical to those in the general imaging guidelines. See 3D Rendering (Preface-4.1) in the Preface Imaging Guidelines
- 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.

### **Background and Supporting Information**

- “The U.S. Food and Drug Administration (FDA) is warning that repeated or lengthy use of general anesthetic and sedation drugs during surgeries or procedures in children younger than 3 years or in pregnant women during their third trimester may affect the development of children’s brains. ...Published studies in pregnant animals and young animals have shown the use of general anesthetic and sedation drugs for more than 3 hours caused widespread loss of nerve cells in the brain. ... All the studies in children had limitations, and it is unclear whether any negative effects seen in children’s learning or behavior were due to the drugs or to other factors, such as the underlying medical condition that led to the need for the surgery or procedure.”<sup>28</sup>

# General Guidelines-Other Imaging Situations (PEDHD-1.4)

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- MRI Brain without contrast (CPT® 70551) or MRI Brain with and without contrast (CPT® 70553) can be performed for nausea and vomiting, persistent, unexplained and a negative GI evaluation
- Screening for metallic fragments before MRI should be done initially with Plain x-ray.
  - The use of CT Orbital to rule out orbital metallic fragments prior to MRI is rarely necessary
  - Plain x-rays are generally sufficient; x-ray detects fragments of 0.12 mm or more, and CT detects those of 0.07 mm or more
  - Plain x-ray is generally sufficient to screen for aneurysm clips
- CPT® 76377 (3D rendering requiring image post-processing on an independent workstation) can be considered when performed in conjunction with conventional angiography (i.e.: conventional 4 vessel cerebral angiography).
- MRI Brain with and without contrast (CPT® 70553) is appropriate in consideration of neurosarcoidosisCT or MRI Perfusion (See CT or MRI Perfusion (HD-24.5) in the Head Imaging Guidelines)
  - Performed as part of a CT Head or MRI Brain examination in the evaluation of individuals with very new strokes or brain tumors.
  - Category III 0042T - “cerebral perfusion analysis using CT”. The study is generally limited to evaluation of acute stroke (<24 hours), to help identify individuals with stroke-like symptoms most likely to benefit from thrombolysis or thrombectomy, to assist in planning and evaluating the effectiveness of therapy for cervical or intracranial arterial occlusive disease and/or chronic cerebral ischemia, identifying cerebral hyperperfusion syndrome following revascularization and following aneurysmal subarachnoid hemorrhage. Other indications are usually regarded as investigational and experimental. Individual health plan policies should be confirmed. (See Moyamoya Disease (PEDHD-12.4))
  - There is no specific CPT® code for MRI Perfusion. Perfusion weighted images are not coded separately from a MRI Brain examination. If MRI Brain is planned, no additional CPT® codes are necessary or appropriate to perform MRI perfusion.
- MRI Perfusion may be obtained with MRI Brain (CPT® 70551 **OR** CPT® 70552 **OR** CPT® 70553) no additional CPT® codes are necessary or appropriate to perform MRI perfusion.<sup>31</sup>

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# Specialized Imaging Techniques (PEDHD-2)

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# Magnetic Resonance Spectroscopy (MRS, CPT<sup>®</sup> 76390) (PEDHD-2.1)

HDP.SI.0002.1.C

v1.0.2024

- 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.
- Uses in pediatric neuro-oncology: See Pediatric CNS Tumors (PEDONC-4) in the Pediatric Oncology Imaging Guidelines.
- MRS is indicated in individuals with neonatal hypoxic ischemic encephalopathy to help estimate the age of the injury.
- Uses in Metabolic Disorders:
  - See Neurometabolic and Neurogenetic Disorders (PEDHD-19.4)
- MRS is considered investigational for all other pediatric indications at this time.

# Functional Magnetic Resonance Imaging (fMRI, CPT<sup>®</sup> 70554 and CPT<sup>®</sup> 70555) (PEDHD-2.2)

HDP.SI.0002.2.A

v1.0.2024

- MRI 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 performed concurrently with MRI Brain (CPT<sup>®</sup> 70551 or CPT<sup>®</sup> 70553) and/or PET Brain Metabolic (CPT<sup>®</sup> 78608 or CPT<sup>®</sup> 78609).
- fMRI is considered investigational for all other pediatric indications at this time.

# PET Brain Imaging (CPT® 78608) (PEDHD-2.3)

HDP.SI.0002.3.A

v1.0.2024

- Uses in pediatric neuro-oncology: See Pediatric CNS Tumors (PEDONC-4) in the Pediatric Oncology Imaging Guidelines.
- Metabolic (FDG) 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 appropriate concurrently with MRI Brain (CPT® 70551 or CPT® 70553) and/or fMRI (CPT® 70554 or CPT® 70555).
- Metabolic (FDG) PET Brain/MRI is generally not supported for neurologic conditions due to lack of standardization in imaging technique and interpretation. However, it can be appropriate in certain pediatric individuals when ALL of the following criteria are met:
  - The individual meets guideline criteria for Metabolic (FDG) PET/CT Brain AND
  - Metabolic (FDG) PET/CT Brain is not available at the treating institution AND
  - The provider requests Metabolic (FDG) PET Brain/MRI in lieu of Metabolic (FDG) PET/CT Brain
- Metabolic (FDG) PET Brain/MRI, when the above criteria are met, are reported using the code combination of Metabolic (FDG) PET Brain (CPT® 78608) and MRI Brain (CPT® 70551 or CPT® 70553). All other methods of reporting Metabolic (FDG) PET Brain/MRI are inappropriate
  - When clinically appropriate, diagnostic MRI codes can be appropriate at the same time as the Metabolic (FDG) PET Brain/MRI code combination.
- Metabolic (FDG) PET Brain is considered investigational for all other pediatric indications at this time.

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# Pediatric Headache (PEDHD-3)

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# Pediatric Headache (PEDHD-3.1)

HDP.PH.0003.1.A

v1.0.2024

- 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 pertinent clinical evaluation including a detailed history, physical examination with a thorough neurologic examination, since the onset or change in signs and/or symptoms, and appropriate laboratory studies should be performed prior to considering advanced imaging.
- Advanced imaging is not indicated for pediatric individuals 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** MRI Brain 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, and/or symptoms on neurologic examination including diplopia.
  - Clumsiness (common description of gait or coordination problems in young children).
  - Headaches associated with morning nausea/vomiting.
  - Seizures
  - Papilledema on physical exam.
  - Headache precipitated by coughing, sneezing, physical exertion<sup>12</sup> or Valsalva.
  - Thunderclap 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 individual.
  - Individual 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.
  - Focal neurological complaints including dizziness, visual change, acute hypertension or altered mental status.



- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is appropriate in the presence of neurological signs and/or symptoms, including headache, after COVID-19 infection
- If concern for CNS infection – See **CNS Infection (PEDHD-29)**
- CT Head poorly visualizes the posterior fossa in children and is generally insufficient to evaluate pediatric headaches with red flag symptoms. CT is not supported in lieu of MRI solely to avoid sedation.
- CT Head without contrast (CPT® 70450) is indicated for pediatric headache with one or more of the following:
  - Sudden severe headache including thunderclap headache
  - Acute setting of suspected intracranial infection prior to lumbar puncture (CT Head with contrast CPT® 70460 if intracranial spread of disease is suspected to detect suppurative fluid collections) (See General Guidelines-Other Imaging Situations (PEDHD-1.4))
  - To exclude new hemorrhage, significant mass effect, or hydrocephalus in cases including rapid clinical deterioration.
  - Recent head trauma.
  - Suspected skull or other bony involvement.
  - If MRI is contraindicated
  - Ventriculoperitoneal shunt with suspected shunt malfunction. See **Macrocephaly, Microcephaly, and Hydrocephalus (PEDHD-7)** for additional imaging.
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is appropriate if a recent CT is inconclusive.
- MRI Brain without and with contrast (CPT® 70553) is appropriate if an abnormality is identified on a noncontrasted MRI performed greater than 2 weeks, otherwise a MRI Brain with contrast (CPT® 70552) is appropriate
- MRA Head 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 MRI Brain or CT Head.
  - Concurrent MRI and MRA is generally not indicated.
- MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546) is indicated in pediatric individuals with papilledema and headache. See Papilledema/Pseudotumor Cerebri (HD-17.1) in the Head Imaging Guidelines

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# Pediatric Head and Face Trauma (PEDHD-4)

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# Head Trauma (PEDHD-4.1)

HDP.PS.0004.1.A

v1.0.2024

- In individuals with recent head trauma, a history focused on the incident and careful examination of the head, neck, and neurological function should be performed since the onset or change in signs and/or symptoms prior to considering advanced imaging.
- Advanced imaging is indicated for children with head trauma with ANY of the following red flags:
  - Loss of consciousness
  - Altered mental status or abnormal behavior
  - Known or suspected skull fracture
  - Glasgow Coma Score <15
  - Age younger than 2 years
  - Vomiting
  - Severe mechanism of injury
  - Severe or worsening headache
  - Amnesia
  - Nonfrontal scalp hematoma
- CT Head without contrast (CPT® 70450) is the primary advanced imaging study in individuals with acute head trauma.
  - CT Maxillofacial without contrast (CPT® 70486), CT 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.
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is indicated for the following:
  - Children with an abnormal neurological exam that is not explained by the CT findings.
  - Subacute (8 days to one month after initial traumatic event) or chronic blunt head trauma with new or worsening neurological signs or cognitive symptoms
  - Children suspected of being the victims of physical abuse. See Suspected Physical Child Abuse (PEDMS-7) in the Pediatric Musculoskeletal Imaging Guidelines.
- Following a head injury, a repeat 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 parenchymal subdural or epidural hematoma may require frequent imaging during the initial 8 weeks following injury, and these requests should generally appropriate.

## Facial Trauma (PEDHD-4.2)

HDP.PS.0004.2.A

v1.0.2024

- CT Maxillofacial without contrast (CPT® 70486) is the preferred imaging study in facial trauma.

### Coding of Facial Imaging

- Both CT Orbital/Facial/Temporal Bone without contrast (CPT® 70480) and CT Maxillofacial (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.
- CT Maxillofacial without contrast (CPT® 70486) is the usual study (except in obvious orbital or temporal bone trauma), but either study is appropriate.

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v1.0.2024

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# Sinusitis and Allergic Rhinitis (PEDHD-5)

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# Sinus And Facial Imaging General Considerations (PEDHD-5.1)

HDP.AR.0005.1.A

v1.0.2024

- 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.



# Imaging Indications in Sinusitis (PEDHD-5.2)

HDP.AR.0005.2.A

v1.0.2024

- 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 Maxillofacial 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, including nasal obstruction, facial pressure/pain or cough<sup>11</sup> for >90 days)
  - Known or suspected fungal sinusitis
    - MRI Orbit/Face/Neck without and with contrast (CPT® 70543) is appropriate if requested instead of CT Maxillofacial
  - Preoperative evaluation to assess surgical candidacy
- CT Maxillofacial with contrast (CPT® 70487) can be performed if ANY of the following is present:
  - Orbital or facial cellulitis
  - Proptosis.
  - Abnormal visual examination
  - Ophthalmoplegia
  - Immunocompromised individual
  - Fungal or vascular lesions visualized in nasal cavity
- CT Head with contrast (CPT® 70460) **OR** MRI Brain without and with contrast (CPT® 70553) **OR** MRI Orbit/Face/Neck with and without contrast (CPT® 70543) is indicated if ANY of the following are present:
  - Focal neurologic findings
  - Altered mental status
  - Seizures
  - Concern for orbital complications
  - Concern for invasive fungal sinusitis
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR** CTA Head (CPT® 70496) is appropriate with these findings as well if there is clinical concern for

associated vascular complications including but not limited to mycotic aneurysm or venous sinus thrombosis.

- Repeat sinus imaging is generally not indicated for individuals who have responded satisfactorily to treatment, but is appropriate with clear documentation of the need for updated CT results to direct acute patient care decisions.

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

HDP.AR.0005.3.A

v1.0.2024

- 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 individual's 3D CT images. In most cases, the preoperative CT is a technical-only service that does not require interpretation by a radiologist.
- For treatment planning for sinus surgery CPT® 77011: A stereotactic CT localization scan 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 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 (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.

# Requests for both Head and Sinus Imaging (PEDHD-5.4)

HDP.AR.0005.4.A

v1.0.2024

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

# Allergic Rhinitis (PEDHD-5.5)

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HDP.AR.0005.5.A

v1.0.2024

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

## Other Indications for Sinus Imaging (PEDHD-5.6)

HDP.AR.0005.6.A

v1.0.2024

- See **Facial Trauma (PEDHD-4.2)** for imaging guidelines in trauma.
- CT Maxillofacial without contrast (CPT® 70486) - Congenital anomalies of facial structures.
- Cleft lip and palate can be associated with brain malformations and abnormal brain development.
  - MRI Brain (CPT® 70551) is appropriate in cases of cleft lip and/or palate.
  - MRI Orbits/Face/Neck without contrast (CPT® 70540) or MRI Orbits/Face/Neck with and without contrast (CPT® 70543)<sup>19</sup> is appropriate if requested by surgeon or any provider in consultation with the surgeon. See **Facial Anomalies (PEDHD-8.2)**
- 3D CT reconstructed images (CPT® 76377) **OR** CPT® 76376) in conjunction with routine CT should be an integral part of the examination in evaluating craniofacial abnormalities.
- CT Maxillofacial without and with contrast (CPT® 70488) **OR** MRI Orbits/Face/Neck without and with contrast (CPT® 70543) - Tumors or other disorders of facial structures.
- Obstructive sleep apnea See **Pediatric Sleep Disorders (PEDHD-24.1)** for imaging guidelines.
- See Sinus and Facial Imaging (HD-29) for conditions not addressed in Sinus and Facial Imaging (PEDHD-5)

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**v1.0.2024**

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# Epilepsy and Other Seizure Disorders (PEDHD-6)

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# Epilepsy and Other Seizure Disorders (PEDHD-6.0)

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HDP.EP.0006.0.A

v1.0.2024

- A pertinent evaluation including a detailed history, physical examination with a thorough neurologic examination, since the onset or change in signs and/or symptoms, and appropriate laboratory studies should be performed prior to considering the use of an advanced imaging (CT, MRI, Nuclear Medicine) procedure. An exception can be made if the individual is undergoing guideline-supported, scheduled follow-up imaging evaluation or request is from or in consultation with a neurologist or neurosurgeon who has seen the individual since onset of symptoms. This clinical evaluation should also include family history and (whenever possible) the accounts of eyewitnesses to the event(s).

# Initial Imaging of Non-Febrile Seizures (PEDHD-6.1)

HDP.EP.0006.1.A

v1.0.2024

- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is indicated for the following:
  - First-time seizure in child that has no known cause and is not associated with fever
  - Partial seizures
  - New onset primary generalized epilepsy (e.g., absence epilepsy or juvenile myoclonic epilepsy)<sup>1</sup> in those who are neurologically abnormal (e.g. developmental delay)
  - Focal neurologic deficits
  - Inconclusive findings on recent cranial ultrasound or CT Head
    - If individual meets criteria for MRI imaging for initial imaging of non-febrile seizure, MRI is appropriate even with a recent negative CT.
  - MRI Brain with and without contrast (CPT® 70553) is appropriate if there are history or examination findings concerning for a mass lesion or demyelinating disease.
- CT Head without contrast (CPT® 70450) is indicated for the following:
  - First-time seizure in child associated with recent head trauma, barrier to obtaining a neuroimaging study in a timely manner and should not preclude MRI imaging when requested.(Late post traumatic seizures may be better evaluated by MRI Brain without contrast (CPT® 70551) See **Head Trauma (PEDHD-4.1)**)
  - Individual cannot safely undergo MRI (avoidance of sedation is not an indication) or in urgent situations.
  - Identification of blood and calcifications
- Cranial ultrasound (CPT® 76506) for the following:
  - First-time seizure in child <30 days of age that has no known cause and is not associated with fever if the infant has an open fontanelle.
  - Cranial ultrasound is not required before MRI Brain without (CPT® 70551) for hypoxic ischemic encephalopathy (HIE) and congenital malformations.
- 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

# Repeat imaging indications (PEDHD-6.2)

HDP.EP.0006.2.A

v1.0.2024

- Repeat MRI Brain without contrast (CPT® 70551) **OR** MRI Brain 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
  - Refractory or drug resistant seizures (See Background and Supporting Information below)
  - Change in seizure type
  - Repeat imaging for persistent seizures as per specialist request or any provider in consultation with a specialist
  - MRI Brain with contrast (CPT® 70552) **OR** MRI Brain without and with contrast (CPT® 70553) to clarify an abnormality on noncontrast MRI or if considering infection or inflammation

## Background and Supporting Information

- Drug Resistant synonyms may include “Refractory”, “Intractable” or “Pharmacoresistant”
- Drug Resistant requires only 2 trials of antiepileptic medications

Repeat imaging indications (PEDHD-6.2)

# Special Imaging Studies in Evaluation for Epilepsy Surgery (PEDHD-6.3)

HDP.EP.0006.3.A

v1.0.2024

- Individuals with a previous MRI Brain and documentation of intractable epilepsy for which surgical treatment or another interventional modality is under active consideration, below are examples of, but not all inclusive, include:
  - Focal Resection
  - Temporal Lobe Resection
  - Extratemporal Resection
  - Lesionectomy
  - Multiple Subpial Transections
  - Laser Interstitial Thermal Therapy
  - Anatomical or Functional Hemispherectomy and Hemispherotomy
  - Corpus Callosotomy
  - Stereotactic Radiosurgery
  - Neurostimulation Device Implantations including,
    - Vagus Nerve Stimulation (VNS)
    - Responsive Neurostimulation
    - Deep Brain Stimulation
- The following requests are appropriate for presurgical evaluation:
  - MRI Brain without contrast (CPT® 70551) **OR** MRI Brain with and without contrast 3T/7T (CPT® 70553)
  - Ictal SPECT (CPT® 78803)
  - Functional MRI (f-MRI) (CPT® 70555 or CPT® 70554) See Functional MRI (fMRI) (HD-24.2) in the Head Imaging Guidelines
  - Metabolic (FDG) PET Brain (CPT® 78608)
    - Metabolic (FDG) PET Brain/MRI is generally not supported for neurologic conditions due to lack of standardization in imaging technique and interpretation. However, it may be appropriate in certain pediatric individuals when ALL of the following criteria are met:
      - The individual meets guideline criteria for Metabolic (FDG) PET/CT Brain **AND**
      - Metabolic (FDG) PET/CT Brain is not available at the treating institution **AND**
      - The provider requests Metabolic (FDG) PET Brain/MRI in lieu of Metabolic (FDG) PET/CT Brain
    - Metabolic (FDG) PET Brain/MRI, when the above criteria are met, are reported using the code combination of:

- Metabolic (FDG) PET Brain (CPT® 78608) **AND**
- MRI Brain without contrast (CPT® 70551 **OR** MRI Brain without and with contrast CPT® 70553).
- All other methods of reporting Metabolic (FDG) PET Brain/MRI are inappropriate
- When clinically appropriate, diagnostic MRI codes may be appropriate at the same time as the Metabolic (FDG) PET Brain/MRI code combination.
- MR Spectroscopy (CPT® 76390).
  - NOTE: Certain payors consider MR Spectroscopy investigational/experimental, and those coverage policies take precedence over eviCore Imaging Guidelines
- See Primary Central Nervous System Tumors-General Considerations (ONC-2.1) in the Oncology Imaging Guidelines for additional imaging requests for surgery and/or Neurosurgical Imaging (HD-28) in the Head Imaging Guidelines
- When noninvasive EEG monitoring is insufficient, intracranial monitoring with stereo-EEG or grids/strips and electrodes may be required with appropriate additional imaging for neuronavigation with one of each of the following after consulting the health plan direction for unlisted codes:
  - MRI Brain with and without (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551)
  - CT Head, contrast as requested CPT
  - If previous head imaging is considered inadequate or additional sequences/protocols are required **OR** is greater than 6 months old, diagnostic head imaging may be appropriate.
- Due to variances with techniques currently available for neuronavigation, the following are appropriate:
  - CTA Head (CPT® 70496) **OR** MRA Head (CPT® 70544, CPT® 70545 or CPT® 70546)
  - Post-operative imaging including after intracranial (EEG) monitoring is appropriate per neurosurgeon's request.

## Febrile Seizures (PEDHD-6.4)

HDP.EP.0006.4.A

v1.0.2024

- A typical febrile seizure is a generalized seizure occurring in the presence of fever ( $T > 100.4^{\circ}\text{F}/38^{\circ}\text{C}$ ) 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<sup>®</sup> 70551) **OR** MRI Brain without and with contrast (CPT<sup>®</sup> 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 (Head circumference that is greater than the 95<sup>th</sup> percentile for age and sex, established by use of measurements and CDC growth charts. See **Macrocephaly (PEDHD-7.1)**)
  - Signs and symptoms of increased intracranial pressure
  - Developmental delay
  - If CT Head without contrast (CPT<sup>®</sup> 70450) was performed for an initial evaluation for new onset seizure, MRI (as described above) is indicated for additional evaluation
- CT of head without contrast (CPT<sup>®</sup> 70450) is indicated for:
  - Evaluation of structural findings in seizure etiologies that contain dystrophic calcifications, such as with oligodendrogliomas and tuberous sclerosis.
  - Acute setting of seizure evaluation.

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# Macrocephaly, Microcephaly, and Hydrocephalus (PEDHD-7)

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# Macrocephaly (PEDHD-7.1)

HDP.MC.0007.1.A

v1.0.2024

- Macrocephaly is defined as head circumference that is greater than the 95<sup>th</sup> percentile for age and sex, or head circumference increasing in percentiles over two visits 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 6 months:
  - Ultrasound Head (CPT® 76506) is indicated initially in individuals with an open fontanelle.
  - CT Head without contrast (CPT® 70450) is indicated if hydrocephalus or hemorrhage is present on ultrasound.
  - MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) is indicated for any abnormality seen on ultrasound.
- Age 7 months and older, or with closed fontanelle:
  - MRI Brain without contrast (CPT® 70551) or 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.

## Microcephaly (PEDHD-7.2)

HDP.MC.0007.2.A

v1.0.2024

- Microcephaly is defined as head circumference that is less than the 5<sup>th</sup> percentile for age and sex, or head circumference decreasing in percentiles over two visits 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 individuals.
  - CT is generally not recommended as that modality lacks the sensitivity to detect the relevant anatomical abnormalities

# Hydrocephalus (PEDHD-7.3)

HDP.MC.0007.3.A

v1.0.2024

- This is the most common identifiable cause of macrocephaly. Almost all hydrocephalus is obstructive, except hydrocephalus due to choroid plexus papillomas. See Choroid Plexus Tumors (PEDONC-4.13) in the Pediatric Oncology 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).
- Ventriculomegaly refers to enlarged ventricular spaces. It is often initially found on fetal ultrasound. It can be from an obstructive cause or can be relative secondary to small brain volume. It can remain stable and may be monitored with serial ultrasound (CPT® 76506) to assess stability or MRI Brain with and without contrast (CPT® 70553) if over age 6 months. If ventriculomegaly progresses to hydrocephalus, follow imaging timelines listed below for hydrocephalus.
- Benign external hydrocephalus (aka benign extra-axial fluid collection among other names) is defined as a rapid increase in head circumference in an infant with enlarged frontal subarachnoid spaces. It is a common cause of macrocephalus and is commonly secondary to a familial large head size. See **Macrocephaly (PEDHD-7.1)** for initial imaging guidelines. It typically requires no intervention. Once diagnosed and confirmed with MRI Brain with and without contrast (CPT® 70553) no additional imaging is required unless new neurological symptoms appear, worsen or persist beyond age 4 years. If developmental motor delay See **Developmental Motor Delay (PEDHD 19.3)**.
- For CSF flow imaging See CSF Flow Imaging (HD-24.4) in the Head Imaging Guidelines

## Initial Imaging Indications

- Age 0-6 months:
  - Screening head ultrasound examination (CPT® 76506)
  - MRI Brain without and with contrast (CPT® 70553) is indicated if ultrasound shows hydrocephalus.
  - Serial US (CPT® 76506) can be used to monitor ventricular size to determine need and timing of placement of a ventricular catheter, or performance of an endoscopic third ventriculostomy (ETV).
- Greater than 6 months old:
  - MRI Brain without and with contrast (CPT® 70553) is indicated.

| Spine Imaging Indication   | Imaging Study   |
|--|---|
| Including, but not limited to: <ul style="list-style-type: none"> <li>• Chiari Malformation</li> <li>• Malignant infiltration of meninges</li> </ul> | MRI Cervical Spine without and with contrast (CPT® 72156)<br><b>OR</b><br>MRI Cervical Spine without contrast (CPT® 72141)<br><br><b>AND/OR</b><br>MRI Thoracic Spine without contrast (CPT® 72146) <b>OR</b><br>MRI Thoracic Spine without and with contrast (CPT® 72157)<br><br><b>AND/OR</b><br>MRI Lumbar Spine without and with contrast (CPT® 72158)<br><b>OR</b><br>MRI Lumbar Spine without contrast (CPT® 72148) |
| Dandy-Walker malformation  | MRI Cervical Spine without and with contrast (CPT® 72156)<br><b>OR</b><br>MRI Cervical Spine without contrast (CPT® 72141)  |

### **Repeat Imaging Indications including CSF flow shunting and Ventriculostomy**

- 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 or ETV 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 after shunt setting adjustments or as ordered by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon.
- Rapid MRI Brain without contrast (CPT® 70551) **OR** CT Head without contrast (CPT® 70450) is indicated in the postoperative period following shunt placement or ETV, with further follow-up imaging 6-12 months after the procedure and then every 12 months for individuals with stable clinical findings.
  - Rapid MRI provides more anatomical detail and does not involve radiation exposure, but many providers use CT Head 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) is appropriate 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 appropriate 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 appropriate for the following:
  - Evaluation of internal shunt, porencephalic cyst, or posterior fossa cyst.
- Nuclear Medicine Shunt Evaluation (CPT® 78645) and CSF Flow SPECT (CPT® 78803) are rarely done in children but can be appropriate for the following:
  - Suspected malfunction of ventriculoperitoneal, ventriculopleural, or ventriculovenous shunts.

### **Background and Supporting Information**

- Head ultrasound can be performed while the fontanelles are still open and has excellent spatial and anatomic resolution, particularly within the first 2 months of life. After 6 months, smaller acoustic windows due to closing sutures limit the sensitivity of the examination.
- Rapid MRI Brain without contrast (CPT® 70551) provides more anatomical detail and does not involve radiation exposure, but many providers use CT Head as rapid MRI is not universally available

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**v1.0.2024**

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# Craniosynostosis (PEDHD-8)

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# Craniosynostosis Imaging (PEDHD-8.1)

HDP.CS.0008.1.C

v1.0.2024

- Craniosynostosis is the premature closure of one or more cranial sutures, usually during infancy. Craniosynostosis may be caused by a genetic condition, such as Apert, Pfeiffer or Crouzon syndrome to name a few.<sup>18</sup> Abnormal head shape **is the common clinical feature**.
- Skull x-rays and/or ultrasound should be obtained prior to considering advanced imaging. In cases of very strong consideration of craniosynostosis with surgical planning in progress, x-rays and/or ultrasound are not required.
- CT Head without contrast (CPT® 70450) is indicated in the diagnosis of craniosynostosis.
- 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 without contrast (CPT® 70486) **AND** CT Orbits/Temporal Bone without contrast (CPT® 70480) are generally not necessary to evaluate individuals with craniosynostosis but are indicated if the craniosynostosis is part of a larger congenital defect which also involves the bones of the face or orbits.
- CT Maxillofacial without contrast (CPT® 70486) **AND/OR** CT Orbits/Temporal Bone without contrast (CPT® 70480) is/are supported in certain types of craniosynostosis and is/are supported when ordered by surgical specialties or in consultation with surgical specialties during surgical evaluation and planning.
- Ultrasound Head (CPT® 76506) is supported as an alternative method of assessing sutural patency in neonates and infants when radiographs are indeterminate. If inconclusive or for pre-operative planning, CT with 3D rendering is supported as discussed previously in this section.
- CT Head without contrast (CPT® 70450) is supported postoperatively at the discretion of or in consultation with the surgical specialist coordinating the individual's care.
- Positional plagiocephaly typically does NOT require advanced imaging.<sup>13,15,16</sup>



## Facial Anomalies (PEDHD-8.2)

HDP.CS.0008.2.A

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- Congenital anomalies of facial structures
  - CT Maxillofacial without contrast (CPT<sup>®</sup> 70486) **OR** MRI Orbits/Face/Neck without contrast (CPT<sup>®</sup> 70540) **OR** MRI Orbits/Face/Neck with and without CPT<sup>®</sup> 70543)<sup>19</sup>.
- Facial Anomalies (such as, cleft lip and palate) can be associated with brain malformations and abnormal brain development.
  - MRI Brain without contrast (CPT<sup>®</sup> 70551) is supported in individuals with cleft lip and/or palate.
  - MRI Orbits/Face/Neck without contrast (CPT<sup>®</sup> 70540) **OR** MRI Orbits/Face/Neck with and without contrast (CPT<sup>®</sup> 70543)<sup>19</sup> is supported if requested by surgeon or any provider in consultation with the surgeon.

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# Chiari and Skull Base Malformations (PEDHD-9)

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# Chiari I Malformations (PEDHD-9.1)

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HDP.CH.0009.1.A

v1.0.2024

- Ultrasound can be utilized for initial examination in infants to determine ventricular size and associated anomalies and to provide a baseline for follow up evaluation but is not required prior to MRI.

| Indication   | Imaging Study  |
|--|--|
| <ul style="list-style-type: none"> <li>Initial evaluation for suspected or known Chiari malformations</li> </ul> | <p>Any of the following sets of imaging:</p> <p>MRI Brain without contrast (CPT® 70551)<br/><b>OR</b></p> <p>MRI Brain without contrast (CPT® 70551)<br/><b>OR</b></p> <p>MRI Brain without and with contrast (CPT® 70553)</p> <p><b>AND/OR</b></p> <p>MRI Cervical Spine without contrast (CPT® 72141) <b>OR</b></p> <p>MRI Cervical Spine without and with contrast (CPT® 72156)</p> <p><b>AND/OR</b></p> <p>MRI Thoracic Spine without contrast (CPT® 72146) <b>OR</b></p> <p>MRI Thoracic Spine without and with contrast (CPT® 72157)</p> <p><b>AND/OR</b></p> <p>MRI Lumbar Spine without contrast (CPT® 72148) <b>OR</b></p> <p>MRI Lumbar Spine without and with contrast (CPT® 72158)</p> |

| Indication   | Imaging Study  |
|--|--|
| <ul style="list-style-type: none"> <li>Repeat imaging for any of the following:               <ul style="list-style-type: none"> <li>At the discretion of or in consultation with the neurologist and/or neurosurgeon coordinating the individual's care</li> <li>New or worsening signs or symptoms</li> <li>Surgical procedure is actively being considered</li> </ul> </li> </ul> | Any of the following sets of imaging:<br><br>MRI Cervical Spine without contrast (CPT® 72141) <b>OR</b><br><br>MRI Cervical Spine without and with contrast (CPT® 72156)<br><br><b>AND/OR</b><br><br>MRI Thoracic Spine without contrast (CPT® 72146) <b>OR</b><br><br>MRI Thoracic Spine without and with contrast (CPT® 72157)<br><br><b>AND/OR</b><br><br>MRI Lumbar Spine without contrast (CPT® 72148) <b>OR</b><br><br>MRI Lumbar Spine without and with contrast (CPT® 72158) |

- Familial screening is not indicated for Chiari I Malformations
- For CSF flow imaging, see **CSF Flow Imaging (HD-24.4)** in the Head Imaging Guidelines

### Background and Supporting Information

- Chiari I malformations involve caudal displacement or herniation of the cerebellar tonsils. Chiari I may be 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 headache, lower cranial nerve palsies, or sleep apnea.
- Chiari II malformations are more severe than Chiari I malformations. These individuals usually present at birth. Myelomeningocele is always present, and syringomyelia and hydrocephalus are extremely common.
- Chiari III malformations include cerebellar herniation into a high cervical myelomeningocele. Chiari IV malformations refer 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 I malformations and known syringomyelia or hydromyelia is highly individualized.

## Basilar Impression (PEDHD-9.4)

HDP.CH.0009.4.A

v1.0.2024

- 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 without contrast (CPT® 70551) **AND** Cervical Spine without contrast (CPT® 72141) are indicated.
- CT Head without contrast (CPT® 70450) **AND** Cervical Spine without contrast (CPT® 72125) are also indicated if surgery is being considered.
- Basilar impression appears to be genetic, and one-time screening of first-degree relatives with MRI Brain without contrast (CPT® 70551) can be appropriate.

# Platybasia (PEDHD-9.5)

HDP.CH.0009.5.A

v1.0.2024

- Platybasia is a flattening malformation of the skull base, in which the clivus has a horizontal orientation.
- MRI Brain without contrast (CPT® 70551) **OR** CT Head without contrast (CPT® 70450) is indicated to establish a diagnosis when clinically suspected, individuals are usually asymptomatic



## References (HDP-9)

**v1.0.2024**

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# Intracranial Aneurysms and AVM (PEDHD-10)

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# Pediatric Intracranial Aneurysms (PEDHD-10.1)

HDP.IA.0010.1.A

v1.0.2024

- A pertinent 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 individual is undergoing guideline-supported scheduled follow-up imaging evaluation or request is from a neurologist or neurosurgeon who has seen the individual since onset of symptoms
- Initial study for suspected subarachnoid hemorrhage:
  - CT Head without contrast (CPT® 70450) **AND OR**
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553)
- If subarachnoid hemorrhage is present on CT or MRI, or lumbar puncture findings suggest hemorrhage, then the following is/are supported
  - MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR**
  - CTA Head (CPT® 70496) **AND/OR**
  - CT Brain Perfusion (CPT® 0042T) or MRI Brain Perfusion (CPT® 70553)
  - See also: **Background and Supporting Information**
- If findings suspicious for intracranial aneurysm on MRI, then the following is supported
  - MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR**
  - CTA Head (CPT® 70496)
- Initial study for individuals presenting with headache, increased intracranial pressure, seizures, or focal neurologic findings:
  - CT Head without contrast (CPT® 70450) **AND/OR**
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553)
  - If findings suspicious for intracranial aneurysm on prior imaging, then the following is supported:
    - MRA head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR**
    - CTA Head (CPT® 70496)
- Imaging for individuals with known aneurysm presenting with headache, increased intracranial pressure, seizures, or focal neurologic findings:
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553) **AND/OR**
  - MRA Head (CPT® 70544, CPT® 70545 **OR** CPT® 70546) **OR**
  - CTA Head (CPT® 70496)

- CTA Head (CPT® 70496) for individuals with treated aneurysms is preferred.
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) **AND/OR** MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) is/ are indicated for individuals with any of the following conditions (including but not limited to the conditions below) and headache, increased intracranial pressure, seizures, or focal neurologic findings:
  - 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
  - Kawasaki disease
  - Coarctation of the aorta
  - Patent ductus arteriosus
  - Hyper-IgE syndrome
  - 4 forms of Loeys-Dietz syndrome
  - Klinefelter syndrome
  - Alpha-glucosidase deficiency
  - Noonan syndrome
  - Pheochromocytoma
  - Alpha-1-antitrypsin deficiency
  - Presence of an azygos anterior cerebral artery
  - Marfan Syndrome
  - Bicuspid aortic valve
  - Microcephalic osteodysplastic primordial dwarfism Type II
  - Multisystemic Smooth Muscle Syndrome (MSMS)/Smooth Muscle Dysfunction Syndrome (SMDS)/ACTA2 Mutations<sup>(Regalado,Cuoco)</sup> (See **PEDHD 12.8**)
- MRI Perfusion may be obtained with MRI Brain (CPT® 70551 **OR** CPT® 70552 **OR** CPT® 70553)
  - No additional CPT® codes are necessary or appropriate to perform MRI perfusion.<sup>22</sup>
- The timing of follow-up imaging for intracranial aneurysms in children is similar to that in adults. See Intracranial Aneurysms (HD-12.1) in the Head Imaging Guidelines.
- Screening MRI Brain or MRA Head for aneurysms is generally not supported in asymptomatic individuals under age 20 since only 0.6 % of ruptured aneurysms

occur in the pediatric age range. However, for Multisystemic Smooth Muscle Syndrome (MSMS)/Smooth Muscle Dysfunction Syndrome (SMDS)/ACTA2 Mutations see **PEDHD 12.8**.)

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

### **Background and Supporting Information**

- Prior CT Head (CPT® 70450) does not exclude indication for MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551)
- 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.

# Pediatric Intracranial Arteriovenous Malformations (AVM) (PEDHD-10.2)

HDP.IA.0010.2.A

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- A pertinent 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 individual is undergoing guideline-supported scheduled follow-up imaging evaluation or request is from or in consultation with a neurologist or neurosurgeon who has seen the individual since onset of symptoms.
- Vascular malformations include arteriovenous, venous, cavernous, and capillary malformations and vein of Galen Malformations.

| Disorders   | Indications (any of the following)   | Supported Imaging   |
|---|--|---|
| <ul style="list-style-type: none"> <li>• Any aneurysmal and/or AVM Disorder listed in this guideline</li> </ul> | <ul style="list-style-type: none"> <li>• When MRI is contraindicated</li> <li>• Any emergency setting</li> </ul>   | <ul style="list-style-type: none"> <li>• CT Head without contrast (CPT® 70450) AND OR</li> <li>• CTA Head (CPT® 70496) AND OR</li> <li>• CTA Neck (CPT® 70498)</li> </ul>   |
| <ul style="list-style-type: none"> <li>• Suspected Vein of Galen malformation in the neonate</li> </ul>         | <ul style="list-style-type: none"> <li>• Confirmation by Ultrasound Head (CPT® 76506)</li> </ul>   | <ul style="list-style-type: none"> <li>• MRI Brain without contrast (CPT® 70551) OR</li> <li>• MRI Brain without and with contrast (CPT® 70553) OR</li> <li>• Catheter Angiography to precisely identify the feeding arteries and draining vein, especially if embolization is planned</li> </ul> |
| <ul style="list-style-type: none"> <li>• Low Flow Malformations</li> </ul>                                      | <ul style="list-style-type: none"> <li>• When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon</li> </ul> | <ul style="list-style-type: none"> <li>• MRA Head (CPT® 70544, CPT® 70545, OR CPT® 70546) OR</li> <li>• CTA Head (CPT® 70496)</li> </ul>  |

| Disorders  | Indications (any of the following)   | Supported Imaging   |
|--|--|---|
| <ul style="list-style-type: none"> <li>Suspected AVM after the neonate period</li> </ul>   | <ul style="list-style-type: none"> <li>When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon</li> </ul>   | <ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553)OR</li> <li>MRI Brain without contrast (CPT® 70551)</li> </ul>   |
| <ul style="list-style-type: none"> <li>Known AVM</li> </ul>  | <ul style="list-style-type: none"> <li>When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon</li> </ul>   | <ul style="list-style-type: none"> <li>MRI Brain without contrast (CPT® 70551)OR</li> <li>MRI Brain without and with contrast (CPT® 70553)AND/OR</li> <li>MRA Head (CPT® 70544, CPT® 70545, <b>OR</b> CPT® 70546)OR</li> <li>CTA Head (CPT® 70496)</li> </ul> |
| <ul style="list-style-type: none"> <li>Hereditary Hemorrhagic Telangiectasia</li> <li>(HHT; Osler-Weber-Rendu Syndrome)</li> </ul> | <ul style="list-style-type: none"> <li>Suspected based on Family History with at least one affected first-degree relative. (biological parent or sibling)</li> <li>At diagnosis, especially if confirmed by Genetic Testing</li> <li>Screening for confirmed HHT</li> <li>Clinical signs or symptoms concerning for disease progression</li> <li>When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon or geneticist</li> </ul> | <ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553)OR</li> <li>MRI Brain without contrast (CPT® 70551)AND/OR</li> <li>MRA Head (CPT® 70544, CPT® 70545 <b>OR</b> CPT® 70546)OR</li> <li>CTA Head (CPT® 70496)</li> </ul>  |

| Disorders  | Indications (any of the following)  | Supported Imaging   |
|--|---|---|
| <ul style="list-style-type: none"> <li>Capillary Malformation-Arteriovenous Malformation (CM-AVM)</li> </ul> | <ul style="list-style-type: none"> <li>Suspected based on Family History with at least one affected first-degree relative (biological parent or sibling)</li> <li>At diagnosis, especially if confirmed by Genetic Testing</li> <li>Screening for confirmed CM-AVM</li> <li>Clinical signs or symptoms concerning for disease progression When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon or geneticist</li> </ul> | <ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) <b>OR</b></li> <li>MRI Brain without contrast (CPT® 70551) AND/OR</li> <li>MRA Head (CPT® 70544, CPT® 70545 <b>OR</b> CPT® 70546) <b>OR</b> CTA Head (CPT® 70496) AND/OR</li> <li>MRI Cervical Spine without and with contrast (CPT® 72156) <b>OR</b> MRI Cervical Spine without contrast (CPT® 72141) AND/OR</li> <li>MRI Thoracic Spine without and with contrast (CPT® 72157) <b>OR</b> MRI Thoracic Spine without contrast (CPT® 72146)<sup>( Bayrak-Toydemir)</sup></li> </ul> |



| Disorders  | Indications (any of the following)   | Supported Imaging  |
|--|--|--|
| <ul style="list-style-type: none"> <li>Cerebral Cavernous Malformations (CCM)</li> </ul> | <ul style="list-style-type: none"> <li>Suspected based on Family History with at least one affected first-degree relative (biological parent or sibling).</li> <li>At diagnosis, especially if confirmed by Genetic Testing</li> <li>Screening for confirmed CCM</li> <li>Clinical signs or symptoms concerning for disease progression</li> <li>When requested by a neurologist or neurosurgeon or geneticist or any provider in consultation with a neurologist or neurosurgeon or geneticist</li> </ul> | <ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) <b>OR</b></li> <li>MRI Brain without contrast (CPT® 70555)AND/OR</li> <li>MRA Head (CPT® 70544, CPT® 70545 <b>OR</b> CPT® 70546) <b>OR</b></li> <li>CTA Head (CPT® 70496)AND/OR</li> <li>MRI Cervical Spine without and with contrast (CPT® 72156) <b>OR</b></li> <li>MRI Cervical Spine without contrast (CPT® 72141)AND/OR</li> <li>MRI Thoracic Spine without and with contrast (CPT® 72157) <b>OR</b></li> <li>MRI Thoracic Spine without contrast (CPT® 72146)</li> </ul> |

| Disorders   | Indications (any of the following)  | Supported Imaging   |
|---|---|---|
| <ul style="list-style-type: none"> <li>Microcephalic Osteodysplastic Primordial Dwarfism, Type II (MOPDII)<sup>(Bober)</sup></li> </ul> | <ul style="list-style-type: none"> <li>Suspected based on Family History with at least one affected first-degree relative (biological parent or sibling)</li> <li>At diagnosis, especially if confirmed by Genetic Testing</li> <li>Screening for confirmed MOPDII, repeated annually</li> <li>Clinical signs or symptoms concerning for disease progression</li> <li>When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon or geneticist</li> </ul> | <ul style="list-style-type: none"> <li>MRI Brain without contrast (CPT® 70551) <b>OR</b></li> <li>MRI Brain without and with contrast (CPT® 70553)AND/OR</li> <li>MRA Head (CPT® 70544, CPT® 70545, <b>OR</b> CPT® 70546) <b>OR</b></li> <li>CTA Head (CPT® 70496)AND/OR</li> <li>MRA Neck without contrast (CPT® 70547, CPT® 70548 <b>OR</b> CPT® 70549) <b>OR</b></li> <li>CTA Neck (CPT® 70498)</li> </ul> |
| <ul style="list-style-type: none"> <li>Sturge-Weber Syndrome</li> </ul>   | <ul style="list-style-type: none"> <li>At diagnosis</li> <li>Clinical signs or symptoms concerning for disease progression</li> <li>When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon</li> </ul>   | <ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) <b>OR</b></li> <li>MRI Brain without contrast (CPT® 70551)AND/OR</li> <li>MRI Orbits/Face/Neck without and with contrast (CPT® 70543) <b>OR</b></li> <li>MRI Orbits/Face/Neck without contrast (CPT® 70540)</li> </ul>  |

**Additional indications**

- CT Head without contrast (CPT® 70450) may be indicated for:
  - Mass effect
  - Urgent/emergent settings due to availability and speed of CT

- Trauma
- Recent hemorrhage, whether traumatic or spontaneous
- Prior to lumbar puncture in individuals with cranial complaints
- Scenarios in which MRI is contraindicated (i.e. pacemakers, ICDs, cochlear implants, aneurysm clips, orbital metallic fragments, etc.)

### **Background and Supporting Information**

- Prior CT Head (CPT® 70450) does not exclude indication for MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551)
- 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.
- Hereditary AVMs usually have an autosomal dominant pattern of inheritance.<sup>5,6,13, 20</sup>

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**v2.0.2023**

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# Syncope (PEDHD-11)

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# Syncope (PEDHD-11.1)

HDP.SY.0011.1.A

v1.0.2024

- 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 individuals with isolated syncope without focal neurologic findings. See Syncope (PEDCD-5) in the Pediatric Cardiac Imaging Guidelines and **Epilepsy and Other Seizure Disorders (PEDHD-6)** for additional imaging considerations.
- There is no role for advanced neuroimaging for Postural Tachycardia Syndrome (POTS).

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# Pediatric Stroke (PEDHD-12)

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# Pediatric Stroke General Considerations (PEDHD-12.1)

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HDP.PS.0012.1.A

v1.0.2024

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

# Pediatric Stroke Initial Imaging (PEDHD-12.2)

HDP.PS.0012.2.A

v2.0.2023

- Children may not present with typical stroke findings. MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) is appropriate in the presence of neurological signs and/or symptoms with concern for stroke.<sup>4</sup>
- ANY of the following studies are indicated for evaluation.
  - CT Head without contrast (CPT® 70450) **OR** MRI Brain without contrast (CPT® 70551)
  - MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR** CTA Head (CPT® 70496)
  - For suspected carotid dissection CTA Neck (CPT® 70498) **OR** MRA Neck (CPT® 70547, CPT® 70548 **OR** CPT® 70549)
  - Note: Both MRA **OR** CTA Head **AND** Neck are needed to visualize the posterior vertebrobasilar circulation for evaluation of the vertebrobasilar stroke/TIA (vertigo associated with diplopia, dysarthria, bifacial numbness or ataxia) or concern for arterial dissection (risks may include premature stroke [under age 50], head or neck trauma, fibromuscular dysplasia, Ehlers-Danlos syndrome, and chiropractic neck manipulation)
- In individuals with COVID-19, See **COVID-19 and Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDHD-12.7)**

# Pediatric Stroke Subsequent Imaging (PEDHD-12.3)

HDP.PS.0012.3.A

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- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is indicated for any new or worsening neurological findings or seizure activity.
- Repeat imaging for follow up and resolution of stroke or hemorrhage as determined by a neurology specialist or any provider in consultation with a neurology specialist.
  - MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553)
  - MRA Head/MRV Head (CPT® 70544, CPT® 70545 **OR** CPT® 70546) **OR** CTA Head/CTV Head (CPT® 70496) for follow-up of known cerebral artery stenosis or thrombosis<sup>4,7</sup>
  - Other surveillance imaging indications after stroke are listed in the disease-specific sections.

## Background and Supporting Information

- CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart (there is no specific code for CT/MR venography):
  - If arterial and venous CT or MR studies are both performed in the same session, only **one** CPT® code is used to report both procedures
  - If an arterial CTA **OR** MRA study has been performed and subsequently a repeat study is needed to evaluate the venous anatomy, then this study is supported
  - If a venous CTV **OR** MRV study has been performed and subsequently a repeat study is needed to evaluate the arterial anatomy, then this study is supported
  - MRA without and with contrast with venous sinus thrombosis to differentiate total from subtotal occlusion

# Moyamoya Syndrome/Disease (PEDHD-12.4)

HDP.PS.0012.4.A

v1.0.2024

## Initial imaging

- Below are indicated for initial evaluation of Moyamoya Syndrome Disease:
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553) **AND/OR**
  - MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **AND/OR**
  - MRA Neck (CPT® 70547, CPT® 70548, **OR** CPT® 70549)
    - If MRA is contraindicated or not readily available, then CTA Head (CPT® 70496) **AND/OR** CTA Neck (CPT® 70498) are supported.
- MRI Perfusion may be obtained with MRI Brain (CPT® 70551 **OR** CPT® 70552 **OR** CPT® 70553), No additional CPT® codes are necessary or appropriate to perform MRI perfusion<sup>33</sup>

## Repeat imaging for Moyamoya Syndrome/Disease

- MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) every 12 months.
  - CTA Head (CPT® 70496) can be appropriate if MRI/MRA is contraindicated or not readily available
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) every 12 months
- Radiopharmaceutical Localization Imaging SPECT (CPT® 78803) with vasodilating agent acetazolamide (Diamox) challenge can be appropriate when surgery or other vascular intervention is being considered.
- CT Perfusion (CPT® 0042T) **OR** MRI Perfusion (CPT® 70551 **OR** CPT® 70552 **OR** CPT® 70553)<sup>33</sup> (see **Background and Supporting Information**) is indicated for the following:
  - Prior to change in treatment
  - Post-surgical
  - When requested by any provider coordinating care with a neurologist or neurosurgeon<sup>(Choudri2014,ACR2017)</sup>
- CT or MRI Perfusion see General Guidelines-Other Imaging Situations (PEDHD-1.4)
- MRI Perfusion may be obtained with MRI Brain (CPT® 70551 **OR** CPT® 70552 **OR** CPT® 70553), No additional CPT® codes are necessary or appropriate to perform MRI perfusion.<sup>33</sup>

Screening imaging for Moyamoya Disease<sup>(HAN2014, KIM2016)</sup>

- Screening is **NOT** indicated for Moyamoya SYNDROME.

- See Background and Supporting Information for Moyamoya DISEASE versus Moyamoya SYNDROME
- Screening with any of the following for Moyamoya Disease is indicated for first degree relatives (biological parent, full sibling, or biological child) of individuals with Moyamoya Disease:
  - MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR**
  - Transcranial Doppler (TCD) Ultrasound (CPT® 93886 or CPT® 93888)
    - See Transcranial Doppler (CPT®93886) (HD-24.8) in the Head Imaging Guideline
  - If MRA is contraindicated or not readily available, then CTA Head (CPT® 70496) is supported
    - See **General Guidelines-Other Imaging Situations (PEDHD-1.4)**
- MRI Perfusion may be obtained with MRI Brain (CPT® 70551 **OR** CPT® 70552 **OR** CPT® 70553), No additional CPT® codes are necessary or appropriate to perform MRI perfusion.<sup>33</sup>

### **Background and supporting information**

- Moyamoya disease (MMD) is a rare cerebrovascular disease characterized by progressive spontaneous bilateral occlusion of the intracranial internal carotid arteries (ICA) and their major branches (middle cerebral artery [MCA], and anterior cerebral artery [ACA]) with compensatory capillary collaterals as an expression of pathologically increased angiogenic activity resembling a "puff of smoke" (Japanese: Moyamoya) on cerebral angiography.<sup>26</sup> Moyamoya Disease is most prevalent in individuals with East Asian ancestry. Up to 15% of individuals with Moyamoya Disease may have a family history of Moyamoya Disease.<sup>23, 24</sup>
- Moyamoya Disease is distinguished from Moyamoya Syndrome (MMS). MMD is a primary disease process. MMS is a secondary process that occurs in response to another underlying pathological process that causes stenosis of intracranial blood vessels.<sup>25</sup> There are two peaks of incidence with different clinical presentations, at around 10 years and 30-40 years. The peak appears to occur later in women than men. In children, ischemic symptoms, especially transient ischemic attacks, are predominant. Intellectual decline, seizures, and involuntary movements are also more common in this age group. In contrast, adult patients present with intracranial hemorrhage more often than pediatric patients.<sup>24</sup>

# Sickle Cell Disease (PEDHD-12.5)

HDP.PS.0012.5.A

v1.0.2024

- The following imaging is indicated for all sickle cell individuals with a severe phenotype (Hgb SS or Hgb S $\beta^0$ )
  - Transcranial Doppler (TCD) Ultrasound (CPT<sup>®</sup> 93886 or CPT<sup>®</sup> 93888) annually for all individuals age 2 to 16:
    - A short interval repeat study is indicated for individuals with conditional (170-199 cm/sec) flow results, or with individuals undergoing transfusion therapy.
  - TCD for children aged 17 years old may be appropriate on a case-by-case basis.
    - See **Transcranial Doppler (CPT<sup>®</sup> 93886) (HD-24.8)** in the Head Imaging Guidelines for other indications for this modality and **Stroke/TIA (HD-21.1)** in the Head Imaging Guidelines
  - After 17 years old, for individuals with a history of abnormal TCDs, TCDs may be repeated every 3 months.
  - TCD is not indicated for individuals with other phenotypes (Hgb SC, Hgb S $\beta^+$ ).
  - See indications below for advanced imaging with MR or CT.
- MRI Brain without contrast (CPT<sup>®</sup> 70551) **OR** MRI Brain without and with contrast (CPT<sup>®</sup> 70553) is indicated for any of the following:
  - 2 non-imaging Transcranial Doppler (TCD) measurements of  $\geq 200$  cm/sec or a single measurement of  $>220$  cm/sec or 2 assessment TCD measurements  $\geq 185$  cm/sec or a single measurement  $>205$  cm/sec.
  - Persistently abnormal TCD velocities
    - For more regarding TCD, see **Background and Supporting Information**
  - Screening to detect silent cerebral infarcts
  - New symptoms or cognitive impairment occurs or a change in academic performance
  - After an infarct-like lesion is identified, repeat every 12-24 months to assess for cerebral infarct progression
  - Prior to any change in therapy<sup>20-25</sup>

## **Background and Supporting Information**

- TCD is used to screen for overt and silent infarctions and monitor response to transfusion therapy
- Individuals 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 individuals at high risk for stroke and begin a primary stroke prevention program.

- Identification of silent cerebral infarction is important because treatment with prophylactic red cell transfusions to maintain hemoglobin S levels at <30% of total hemoglobin may reduce recurrent stroke and extent of neurologic damage.



# CNS Vasculitis and Stroke (PEDHD-12.6)

HDP.PS.0012.6.A

v1.0.2024

- MRI Brain without and with contrast (CPT® 70553) is the recommended initial study for all individuals with vasculitis and suspected CNS involvement, whether primary or secondary.
  - A normal MRI Brain almost always completely excludes intracranial vasculitis
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) is indicated for inconclusive MRI findings suggesting medium or large vessel vasculitis.
  - Individuals with aggressive disease being treated with systemic therapy can have imaging for treatment response every 3 months during active treatment.
  - Annual surveillance imaging is appropriate to detect progressive vascular damage that may require intervention.

# COVID-19 and Multisystem Inflammatory Syndrome in Children (MIS-C) (PEDHD-12.7)

**HDP.PS.0012.7.A**  
**v1.0.2024**

- Symptoms of MIS-C may include some or all of the following:
  - Headache, irritability
  - Mucocutaneous changes similar to Kawasaki disease (i.e. strawberry tongue, red cracked lips, rash of hands and/or feet)
  - Polymorphous or vasculitic rash
  - Non-exudative conjunctivitis
  - Tachycardia, hypotension
  - Cough, shortness of breath
  - Abdominal pain, vomiting, diarrhea
  - Lymphadenopathy, joint pain, sore throat
- MRA is the preferred study in children however, CTA Head (CPT® 70496) and/or Neck (CPT® 70498) is appropriate if MRA is contraindicated

| Indication   | Imaging Study   |
|--|---|
| <ul style="list-style-type: none"> <li>• Imaging for neurological signs and/or symptoms, including headache, after known or presumed COVID-19 infection</li> </ul> | <ul style="list-style-type: none"> <li>• Any or all of the following sets of imaging:                             <ul style="list-style-type: none"> <li>• MRI Brain without contrast (CPT® 70551) <b>OR</b></li> <li>• MRI Brain without and with contrast (CPT® 70553)</li> <li>• MRA Head (CPT® 70544, CPT®70545 <b>OR</b> CPT® 70546) <b>OR</b></li> <li>• CTA Head (CPT® 70496) <b>AND/OR</b></li> <li>• MRA Neck (CPT® 70547 <b>OR</b> CPT® 70548 <b>OR</b> CPT® 70549) <b>OR</b></li> <li>• CTA Neck (CPT® 70498)</li> </ul> </li> </ul> |

- If concern for CNS infection – See **CNS Infection (PEDHD-29.1)**
- See Multisystem inflammatory syndrome in children (MIS-C) (PEDCD-12) in the Pediatric Cardiac Imaging Guidelines

**Background and Supporting Information**

- COVID-19 infections in children are generally mild in comparison to that of adults, however a post viral syndrome in children has become increasingly noted.

Multisystem Inflammatory Syndrome in Children (MIS-C) can cause an inflammatory vasculopathy, prothrombotic state and/or post viral myocarditis in children who have had a COVID-19 infection caused by SARS-CoV-2. The child may have had minor symptoms or been asymptomatic at the time of COVID-19 infection but the virus can trigger endothelial injury and activation of the IL-1 pathway similar to that in Kawasaki disease and acute rheumatic fever.

# Multisystemic Smooth Muscle Syndrome (MSMS)/Smooth Muscle Dysfunction Syndrome (SMDS)/ACTA2 Mutations (PEDHD-12.8)

HDP.PS.0012.8.A

v1.0.2024

## Head/Neck Evaluations for MSMS/SMDS

### Initial Evaluations

- Confirmed ACTA2 Mutation
  - MRI Brain without and with contrast (CPT® 70553) with **OR** without Perfusion **AND/OR**
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **AND/OR**
  - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549)

### As requested by neurologist and/or neurosurgeon and/or geneticist and/or provider coordinating care with a neurologist and/or neurosurgeon and/or geneticist

- MRI Brain without and with contrast (CPT® 70553) with **OR** without Perfusion **AND/OR**
  - MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **AND/OR**
  - MRA Neck (CPT® 70547, CPT® 70548, **OR** CPT® 70549)
- MRI Perfusion may be obtained with MRI Brain (CPT® 70551 **OR** CPT® 70552 **OR** CPT® 70553), No additional CPT® codes are necessary or appropriate to perform MRI perfusion.<sup>33</sup>
  - Because radiation is a known risk factor for moyamoya disease. MRI/MRA Head is recommended instead of Computed Tomography (CT/CTA).<sup>29</sup>
  - Conventional catheter angiogram should be reserved for patients with focal neurologic symptoms or evidence on MRA or transcranial Doppler (TCD) of critical or progressive narrowing of the cerebral arteries.<sup>31</sup>
  - See Screening for Suspected Peripheral Artery Disease/Aneurysmal Disease (PVD-2).

**Background and Supporting Information**

- Smooth muscle dysfunction syndrome (SMDS)/Multisystemic Smooth Muscle Syndrome (MSMS) presents with a recognizable pattern of complications, including congenital mydriasis, patent ductus arteriosus (PDA), pulmonary arterial hypertension, aortic and other arterial aneurysms, moyamoya-like cerebrovascular disease, intestinal hypoperistalsis and malrotation, and hypotonic bladder.<sup>29</sup>
- SMDS/MSMS is caused by heterozygous mutations of the ACTA2 altering arginine 179, most commonly p.Arg179His. With a single exception, all cases are due to de novo mutations.<sup>29</sup>

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# Benign Brain Lesions (PEDHD-13)

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# Arachnoid Cysts (PEDHD-13.1)

HDP.BL.0013.1.A

v1.0.2024

- 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 contrast (CPT® 70551) **OR** 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 individuals with arachnoid cysts, but is appropriate for the following:
  - Annual MRI Brain without contrast (CPT® 70551) **OR** 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
  - When requested by a specialist or any provider in consultation with a specialist

## Pineal/Colloid Cysts (PEDHD-13.2)

HDP.BL.0013.2.A

v1.0.2024

- Pineal cysts are generally discovered incidentally and do not require surgical intervention.
- MRI Brain without contrast (CPT® 70551) **OR** 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 individuals with pineal cysts, but MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is appropriate for the following:
  - New or worsening headache or focal neurologic deficits suggesting progression of cyst
  - Preoperative planning
- Repeat MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is appropriate for colloid cysts for the following:
  - In the presence of symptoms including syncope
  - Evaluation of CSF flow (CPT® 70551)
  - When requested by a specialist or any provider in consultation with a specialist

# Acoustic Neuromas (PEDHD-13.3)

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HDP.BL.0013.3.A

v1.0.2024

- See Neurofibromatosis 2 (PEDPND-2.2) in the Pediatric Peripheral Nerve Disorders Imaging Guidelines

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**v1.0.2024**

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# Pediatric Demyelinating Diseases (PEDHD-14)

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# Pediatric Demyelinating Disease

## General Considerations (PEDHD-14.1)

HDP.DD.0014.1.A

v1.0.2024

- MRI Brain without and with contrast (CPT® 70553) is the preferred imaging study for evaluation of pediatric demyelinating disease.
  - MRI Cervical Spine without and with contrast (CPT® 72156 **AND** MRI Thoracic Spine without and with contrast CPT® 72157) is also indicated for evaluation of pediatric demyelinating disease.
  - MRI Lumbar Spine without and with contrast (CPT® 72158) is not indicated unless the individual 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.
- Metabolic (FDG) PET Brain (CPT® 78608) and MR Spectroscopy (CPT® 76390) are considered investigational for evaluation of pediatric demyelinating diseases.
- See **Neurometabolic and Neurogenetic Disorders (PEDHD-19.4)**

# Multiple Sclerosis (MS) (PEDHD-14.2)

HDP.DD.0014.2.A

v1.0.2024

- 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 Brain without and with contrast (CPT® 70553) **AND** Cervical Spine (CPT® 72156 **AND** Thoracic Spine without and with contrast (CPT® 72157) is indicated for initial diagnosis in individuals with clinical signs and/or symptoms suggestive of MS.
  - MRI Brain (CPT® 70551) and Spinal Cord (CPT® 72141 and CPT® 72146) without contrast is appropriate if there is a contraindication to gadolinium administration.
- MRI Brain without contrast (CPT® 70551), which is preferred, OR MRI Brain without and with contrast (CPT® 70553) is indicated every 6 months for disease monitoring whether or not receiving treatment.<sup>19</sup>
  - MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** Cervical Spine without contrast (CPT® 72141) **AND/OR** CPT® 72146) is (are) indicated every 12 months or for new signs or symptoms.<sup>19</sup>
  - MRI Orbits/Face/Neck without and with contrast (CPT® 70543) may be considered if optic neuritis is suspected, in addition to the above scenario
  - Symptoms suggestive of Progressive Multifocal Leukoencephalopathy (PML) during Tysabri therapy (or other medications with similar risk).
    - Screening for individuals on natalizumab (Tysabri) or other drugs with risk of PML (Progressive Multifocal Leukoencephalopathy)
      - MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) every 6 months while on treatment
      - MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) every 3-6 months for high risk individuals positive for serum JC virus antibody and >18 months natalizumab exposure
- If a non-contrast study shows incidental evidence of possible demyelinating disease, repeat with MRI Brain with contrast (CPT® 70552) is appropriate within 2 weeks of previous non-contrast study as the presence of enhancing lesions may be helpful in confirming the diagnosis. If non-contrast study was performed more than 2 weeks prior to the performance of the repeat imaging, an MRI Brain with and without contrast (CPT® 70553) is appropriate.

## Background and Supporting Information

- Medications with similar risks of PML as Tysabri include: Dimethyl fumarate (Tecfidera), Fingolimod (Gilenya), Ocrelizumab (Ocrevus), Mavenclad (cladribine),

Vumerity (diroximel fumarate), Soliris (eculizaumab), Zeposia (ozanimod), Lemtrada (alemtuzumab), Bafiertam (monomethyl fumarate), Rituxan (rituximab).

- 3D imaging in the evaluation of Multiple Sclerosis is not supported as a separate code as most scanners are capable of 3D acquisitions or other imaging sequences may be done.



# Acute Disseminated Encephalomyelitis (ADEM) and Other Pediatric Demyelinating Disorders (PEDHD-14.3)

HDP.DD.0014.3.C

v1.0.2024

- MRI Brain (CPT® 70553) and Spinal Cord (CPT® 72156 and CPT® 72157) without and with contrast is indicated for initial diagnosis in individuals with clinical signs and/or symptoms suggestive of ADEM.
  - MRI Brain (CPT® 70551) and Spinal Cord (CPT® 72141 and CPT® 72146) without contrast is indicated if there is a contraindication to gadolinium.
- MRI Brain without and with contrast (CPT® 70553) **AND** MRI Cervical Spine (CPT® 72156 **AND** MRI Thoracic Spine without contrast CPT® 72157) is/are indicated every 3 months for 1 year following diagnosis or if ordered out of sequence or beyond one year by a neurologist or any provider in consultation with a neurologist.
  - MRI Brain without contrast (CPT® 70551) **AND** MRI Cervical Spine (CPT® 72141) **AND** MRI Thoracic Spine without contrast (CPT® 72146) is/are indicated if there is a contraindication to gadolinium.
  - Most individuals will have complete clinical recovery by 12 months, while stable MRI abnormalities (gliosis) may persist. These findings do not require additional imaging unless the individual develops new neurologic symptoms. Prolonged symptoms or return of symptoms may represent a different demyelinating disorder.
- There are other pediatric demyelinating disorders that are less common but have clinical overlap with multiple sclerosis and ADEM such as (but not limited to):
  - Neuromyelitis optica (NMO) spectrum disorders (See Neuromyelitis Optica and NMO Spectrum Disorders (HD 16.2))
  - Anti-MOG syndromes (anti-myelin oligodendrocyte glycoprotein) (See Anti-MOG Syndromes (HD 16.3))
  - Demyelination secondary to infectious or inflammatory disorders (e.g. transverse myelitis) (See General Guidelines – Other Imaging Situations (HD 1.7))
- These conditions may require a different treatment regimen than multiple sclerosis and may require repeat imaging to monitor treatment response as the diagnosis becomes more clear. Repeat imaging with MRI Brain and/or MRI Cervical Spine and MRI Thoracic Spine as requested by neurology or infectious disease may be approved.
- See Neurometabolic and Neurogenetic Disorders (PEDHD-19.4)

**Background and Supporting Information**

- 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.

# Transverse Myelitis (PEDHD-14.4)

HDP.DD.0014.4.A

v1.0.2024

- Transverse myelitis is an inflammatory disorder of the spine and can be:
  - Idiopathic
  - Associated with autoimmune central nervous system inflammatory diseases
  - First event of multiple sclerosis (MS)
  - Neuromyelitis optica (NMO)
  - MOG (Myelin Oligodendrocyte Glycoprotein) antibody disorder
  - Associated with connective tissue autoimmune disease
  - Systemic Lupus Erythematosus (SLE)
  - Systemic Sclerosis
  - Rheumatoid Arthritis (RA)
  - Sjogren's Syndrome (SS)
  - Neuro-Sarcoidosis (NS)
  - Post-infectious or post-vaccine neurological syndrome
- See Multiple Sclerosis (MS) and Related Conditions (HD-16)

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# Pituitary Dysfunction (PEDHD-15)

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# Pituitary Dysfunction General Considerations (PEDHD-15.1)

HDP.PD.0015.1.A

v1.0.2024

- 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 initial advanced imaging.
- MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) when pituitary imaging is indicated.
  - Pituitary Gland: one study (either MRI Brain without and with contrast [CPT® 70553] **OR** MRI Orbits/Face/Neck [CPT® 70543]) is adequate to report the imaging of the pituitary. The reporting of two CPT® codes, to image the pituitary, is not indicated.
- If a previous MRI Brain was reported to show a possible pituitary tumor with supporting evidence of pituitary disease or is inconclusive, a repeat MRI with dedicated pituitary protocol may be performed. If a prior MRI Brain was without contrast a follow up scan either MRI Brain with contrast (CPT® 70552) **OR** MRI Brain with and without contrast (CPT® 70553) is appropriate
- For association between pituitary dysfunction and optic nerve issues see Eye Disorders and Visual Loss (HD- 32.1) in the Head Imaging Guidelines
- For repeat imaging, See Pituitary (HD-19.1) in the Head Imaging Guidelines

# Panhypopituitarism (PEDHD-15.2)

HDP.PD.0015.2.A

v1.0.2024

- Endocrine testing should be performed initially.
- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) with special attention to the pituitary is indicated for newly diagnosed Panhypopituitarism.
- Individuals with a normal pituitary on initial MRI do not need routine follow up imaging.
- Individuals with mass lesions should have follow up imaging according to the guidelines for the specific diagnosis

# Isolated Growth Hormone Deficiency (PEDHD-15.3)

HDP.PD.0015.3.A

v1.0.2024

- Clinical features include: height below the normal range (<3<sup>rd</sup> percentile), subnormal growth velocity or the child's height is below the range expected based on parental height.
- Risk factors include: a history of brain tumor, cranial irradiation or other congenital/organic hypothalamic-pituitary abnormality as well as an incidental finding of a hypothalamic-pituitary abnormality on MRI.
- Endocrine testing should be performed initially.
- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) with special attention to the pituitary is indicated for any of the following:
  - Both IGF1 and IGFBP3 are below the laboratory reference range for age/sex or Tanner stage.
  - 2 measurements of growth hormone stimulation with different stimulation agents (glucagon, clonidine, arginine, insulin, levodopa) performed on the same day or separate days produce maximal GH levels <10ng/mL See Background and Supporting Information
- Individuals with a normal pituitary on initial MRI do not need routine follow up imaging.
- Individuals with mass lesions should have follow up imaging according to the guidelines for the specific diagnosis.

## Background and Supporting Information

- Growth hormone stimulation testing is limited by poor specificity and requires failure on 2 tests to diagnose growth hormone deficiency.
- Controversy exists as to the cutoff level which differentiates a normal response from a deficient response on provocative testing. Some experts support GH <7 ng/mL however many pediatric endocrinologists consider a peak GH level <10 ng/mL to be indicative of growth hormone deficiency and may identify children with partial GHD.



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

HDP.PD.0015.4.A

v1.0.2024

- Laboratory testing should be performed initially. Diabetes insipidus is characterized by serum osmolality >300mOsm/kg and urine osmolality <300 mOsm/kg.
- Central diabetes insipidus (CDI) is caused by diminished synthesis or secretion of vasopressin in the hypothalamus and nephrogenic diabetes insipidus (NDI) is caused by renal resistance to vasopressin.

## Central Diabetes Insipidus (DI)

- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) is indicated for newly diagnosed central DI
- Individuals with a normal pituitary on initial MRI can have repeat MRI Brain without and with contrast (CPT® 70553) every 3-6 months for the first 2 years as germinomas may cause central DI while still too small to detect on imaging.
  - Serial measurement of  $\beta$ -hCG is also indicated for these individuals, and MRI should be repeated if a significant rise in  $\beta$ -hCG is detected on screening.
- Individuals 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) **OR** MRI Brain without contrast (CPT® 70551) is indicated for initial evaluation of unexplained central (hypothalamic pituitary) SIADH.
- Individuals with a normal pituitary on initial MRI do not need routine follow up imaging.
- Individuals with mass lesions should have follow up imaging according to the guidelines for the specific diagnosis.

## **Background and Supporting Information**

- See Small Cell Lung Cancer-Suspected/Diagnosis (ONC-7.1) and Paraneoplastic Syndromes (ONC-30.3) in the Oncology Imaging Guidelines.
- Pulmonary diseases including infection (tuberculosis, viral/bacterial pneumonia), acute respiratory infections, mechanical ventilation and others can cause SIADH

although the mechanism is unclear. Individuals with lung disease should have chest imaging according to the guidelines for the specific diagnosis.

# Precocious Puberty (PEDHD-15.5)

HDP.PD.0015.5.C

v1.0.2024

- Endocrine laboratory studies (baseline LH, FSH and either estradiol or testosterone) are used to determine if the etiology of precocious puberty is central (gonadotropin dependent) or peripheral (gonadotropin independent). Estradiol and testosterone levels will often be elevated to a pubertal range.

## **Central Precocious Puberty (CPP)**

- An LH >0.3 U/L on a random blood sample is the most reliable screening test for central precocious puberty. If LH <0.3 U/L and CPP is suspected, a stimulation test with a GnRH analog is the gold standard.
- Neuroimaging should always follow hormonal studies that suggest a central origin of precocious puberty.
- MRI Brain without and with contrast (CPT® 70553, preferred study) **OR** MRI Brain without contrast (CPT® 70551) is indicated for evaluation of any child with documented central precocious puberty.
- MRI is appropriate irrespective of age and gender in individuals with precocious puberty and concurrent CNS symptoms of severe headache, visual changes or seizures.
- Individuals with a normal pituitary on initial MRI do not need routine follow up imaging.
- Individuals with mass lesions should have follow up imaging according to the guidelines for the specific diagnosis. (Benign Pituitary Tumors (PEDHD-15.6) and Pituitary Malignancies (PEDHD-15.7))

## **Peripheral Precocious Puberty**

- The differential diagnosis of peripheral precocious puberty (LH suppressed or in the pre-pubertal range with elevated estradiol, testosterone and/or adrenal androgens) is broad and may include ovarian, testicular, adrenal and other sources of excessive hormonal production
- Ultrasound Abdomen (CPT® 76700) in both genders and Ultrasound Pelvis (CPT® 76856) in females and Scrotal ultrasound (CPT® 76870) in males depending on the suspected source of hormonal excess for initial imaging.
- See CNS Germinomas and Non-Germinomatous Germ Cell Tumors (PEDONC-4.7) in the Pediatric Oncology Imaging Guidelines for evaluation of HCG secreting CNS tumors
- See Hepatoblastoma (PEDONC-11.2) in the Pediatric Oncology Imaging Guidelines for evaluation of HCG secreting hepatic tumors
- See Pediatric Germ Cell Tumors (PEDONC-10) in the Pediatric Oncology Imaging Guidelines and Testicular, Ovarian and Extragonadal Germ Cell Tumors (ONC-20) in the Oncology Imaging Guidelines for evaluation of Leydig Cell tumors.
- See Adrenal Cortical Lesions (AB-16.1) in the Abdomen Imaging Guidelines for evaluation of adrenal virilizing tumors

**Background and Supporting Information**

- Defined as the appearance of secondary sexual characteristics before age 8 in females and before age 9 in males. The diagnosis is made clinically using Tanner staging and often will include a bone age assessment (hand/wrist radiographs) and/or abdominal and/or pelvic ultrasound (See Peripheral Precocious Puberty).

# Benign Pituitary Tumors (PEDHD-15.6)

HDP.PD.0015.6.A

v1.0.2024

- Benign pituitary tumor indications in pediatric individuals are identical to those for adult individuals. See Pituitary (HD-19) in the Head Imaging Guidelines.

# Pituitary Malignancies (PEDHD-15.7)

HDP.PD.0015.7.A

v1.0.2024

- See Craniopharyngioma and Other Hypothalamic/Pituitary Region Tumors (PEDONC-4.10) or Histiocytic Disorders (PEDONC-18) in the Pediatric Oncology Imaging Guidelines

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**v1.0.2024**

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12. IGFMS - Clinical: Insulin-Like Growth Factor-1, LC-MS, Serum. [www.mayocliniclabs.com](http://www.mayocliniclabs.com). <https://www.mayocliniclabs.com/test-catalog/Clinical+and+Interpretive/62750>

# Pediatric Ear Disorders (PEDHD-16)

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# Hearing Loss (PEDHD-16.1)

HDP.ED.0016.1.A

v1.0.2024

- A pertinent 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.
- CT Orbits/Temporal Bone 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 individuals 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.
  - Hearing loss associated with tinnitus see **Tinnitus (PEDHD-16.5)**
- Both modalities (CT and MRI) are supported simultaneously for evaluation and surgical planning if ordered by or in consultation with an ENT or Neurosurgical specialist
- Limited MRI Brain with attention to internal auditory canals (CPT® 70540, CPT® 70542, or CPT® 70543) is supported when requested by the provider in place of a complete MRI Brain. Note: Limited MRI codes should not be used in addition to MRI Brain codes; IAC views are performed as additional sequences as part of the brain study. (See General Guidelines – Anatomic Issues (HD-1.1) in the Head Imaging Guidelines)

## Ear Pain (PEDHD-16.2)

HDP.ED.0016.2.A

v1.0.2024

- A pertinent 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, and referred pain from the oral pharynx, tonsillitis, and pharyngitis.
- Advanced imaging is not indicated in the overwhelming majority of pediatric individuals with ear pain.

| Indications  | Imaging Study  |
|--|--|
| <ul style="list-style-type: none"> <li>• Any of the following               <ul style="list-style-type: none"> <li>• Persistent ear pain without obvious cause</li> <li>• Clinical suspicion for complicated or invasive infection such as mastoiditis</li> <li>• Clinical suspicion of mass lesion causing ear pain</li> <li>• Significant trauma with concern for hematoma formation</li> <li>• Preoperative planning</li> </ul> </li> </ul> | <ul style="list-style-type: none"> <li>• ONE of the following:               <ul style="list-style-type: none"> <li>• CT Orbits/Temporal Bone without contrast (CPT® 70480) <b>OR</b></li> <li>• CT Orbits/Temporal Bone without and with contrast (CPT® 70482) <b>OR</b></li> <li>• MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553) <b>OR</b></li> <li>• MRI Orbits/Face/Neck without and with contrast (CPT® 70543)</li> </ul> </li> </ul> |

# Cholesteatoma (PEDHD-16.3)

HDP.ED.0016.3.A

v1.0.2024

- One of the following study is indicated for preoperative evaluation in children with cholesteatoma:
  - CT Orbits/Temporal Bone without contrast (CPT® 70480) **OR**
  - CT Orbits/Temporal Bone 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)
- ONE of the following study is indicated one time post-operatively to exclude residual or regrown cholesteatoma to avoid the need for a second-look surgery:
  - CT Orbits/Temporal Bone without contrast (CPT® 70480) **OR**
  - CT Orbits/Temporal 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)

## Background and Supporting Information

- 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 a white mass in the middle ear cleft, painless drainage from the ear or chronic/recurrent ear infections. Advanced imaging for the diagnosis and management of suspected cholestatoma, in particular, should be reserved for the otolaryngologist or provider in consultation with the otolaryngologist.

# Vertigo (PEDHD-16.4)

HDP.ED.0016.4.C

v1.0.2024

- A pertinent 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.

## Background and Supporting Information

- Isolated vertigo is an uncommon complaint during childhood. Middle ear/Eustachian tube problems are the most common cause of isolated vertigo in children.

# Tinnitus (PEDHD-16.5)

HDP.ED.0016.5.A

v1.0.2024

- Tinnitus without hearing loss is a less common complaint during childhood.
- Children with hearing loss and tinnitus should be imaged according to **Hearing Loss (PEDHD-16.1)**. A pertinent 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 individuals with isolated tinnitus and normal hearing.

| Indications   | Imaging Study  |
|---|--|
| <ul style="list-style-type: none"> <li>• ANY of the following                             <ul style="list-style-type: none"> <li>• Clinical suspicion of mass lesion causing tinnitus</li> <li>• Persistent tinnitus after recent significant trauma</li> </ul> </li> </ul> | <ul style="list-style-type: none"> <li>• ONE of the following                             <ul style="list-style-type: none"> <li>• CT Orbits/Temporal Bone without contrast (CPT® 70480) <b>OR</b></li> <li>• CT Orbits/Temporal Bone without and with contrast (CPT® 70482), <b>OR</b></li> <li>• MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553) <b>OR</b></li> <li>• MRI Brain without contrast with attention to internal auditory canals (CPT® 70553) <b>OR</b></li> <li>• MRI Orbits/Face/Neck without and with contrast (CPT® 70543)</li> </ul> </li> </ul> |
| <ul style="list-style-type: none"> <li>• ANY of the following:                             <ul style="list-style-type: none"> <li>• Pulsatile tinnitus</li> <li>• Suspicion for vascular lesions</li> </ul> </li> </ul>   | <ul style="list-style-type: none"> <li>• MRA Head (CPT® 70544, CPT® 70545 <b>OR</b> CPT® 70546) <b>OR</b></li> <li>• CTA Head (CPT® 70496)</li> </ul> <p><b>AND/OR</b></p> <ul style="list-style-type: none"> <li>• MRA Neck (CPT® 70547, CPT® 70548 <b>OR</b> CPT® 70549) <b>OR</b></li> <li>• CTA Neck (CPT® 70498)</li> </ul>   |

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# Autism Spectrum Disorders (PEDHD-17)

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# Autism Spectrum Disorders (PEDHD-17.1)

HDP.AS.0017.1.A

v1.0.2024

- 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) **OR** MRI Brain without contrast (CPT® 70551) is indicated for new or worsening focal neurologic findings documented on a pertinent physical. Consider advanced imaging if there is loss of developmental milestones and/or regression in two or more areas of development.
- PET imaging is considered investigational in the evaluation of individuals with autism spectrum disorders.



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**v1.0.2024**

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# Behavioral and Psychiatric Disorders (PEDHD-18)

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# Behavioral and Psychiatric Disorders (PEDHD-18.1)

HDP.BD.0018.1.A

v1.0.2024

- Behavioral and psychiatric disorders of childhood or adolescence, to include Attention Deficit Hyperactivity Disorder (ADHD), generally require no advanced imaging for diagnosis or management.<sup>2</sup>
  - MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) **OR** CT Head without contrast (CPT® 70450) is indicated for:
    - Acute onset mental status change
    - New or worsening focal neurologic findings
    - Presentation of acute psychiatric symptoms with comorbid serious medical illness
    - Non-auditory hallucinations (e.g., visual, tactile, olfactory) with no known etiology
    - Nonresponse to adequate medication trials
    - Symptoms of an organic brain disorder (e.g., focal deficits, severe headache, or seizures)
    - Prior to Electro-Convulsive Therapy (ECT) to screen for intracranial disease  
See **HD 4.2: Mental Health Related Disorders**
- For concerns of PANS (Pediatric acute-onset neuropsychiatric syndrome) and PANDAS (Pediatric autoimmune neuropsychiatric disorder associated with streptococcal infection) see **Movement Disorders including Tourette's Syndrome (PEDHD-26)**

## Reference (HDP-18)

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**v1.0.2024**

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# Developmental Disorders (PEDHD-19)

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# Intellectual Disability (PEDHD-19.1)

HDP.ID.0019.1.A

v1.0.2024

- Intellectual disability may be primary or secondary to a variety of heterogeneous disorders. See Background and Supporting Information.
- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) is indicated for new or worsening focal neurologic findings and/or new or worsening cognitive decline.<sup>9</sup>

## Background and Supporting Information

- Intellectual disability is a condition characterized by significant limitations in both intellectual functioning and adaptive behavior that originates before the age of 22.<sup>13</sup> One way to measure intellectual functioning is an IQ test. Generally, an IQ test score of around 70 or as high as 75 indicates a significant limitation in intellectual functioning.<sup>13</sup>

# Cerebral Palsy (PEDHD-19.2)

HDP.ID.0019.2.A

v1.0.2024

- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) is indicated for:
  - Initial evaluation of newly diagnosed cerebral palsy.
  - New or worsening focal neurologic findings documented on a physical examination, including the presence of developmental delay.
  - Re-evaluation after 24 months of age due to rapid myelination during the first 2 years of life.
- For spinal imaging requests See Myelopathy (SP-7.1) in the Spine Imaging Guidelines

## Background and Supporting Information

- Many individuals with intellectual disability also have cerebral palsy, but not all individuals 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

# Developmental Motor Delay (PEDHD-19.3)

HDP.ID.0019.3.A

v1.0.2024

- There are many causes for developmental motor delay. Individuals with motor delay can have decreased, normal, or increased muscular tone. Individuals with normal tone do not require imaging unless they have focal neurologic findings.
- MRI Brain without contrast (CPT® 70553) **OR** MRI Brain without and with contrast (CPT® 70553) is indicated for:
  - Initial evaluation of newly diagnosed developmental motor delay with abnormal muscle tone.
  - Toe walking, when associated with upper motor neuron signs including hyperreflexia, abnormal tone (spasticity/hypotonia), or positive Babinski sign.
  - New or worsening focal neurologic findings.
  - Re-evaluation after 24 months of age due to rapid myelination during the first 2 years of life.
- For spinal imaging requests See Myelopathy (SP-7.1) in the Spine Imaging Guidelines and Tethered Cord (PEDSP-5) in the Pediatric Spine Imaging Guidelines.



# Neurometabolic and Neurogenetic Disorders (PEDHD-19.4)

HDP.ID.0019.4.A

v1.0.2024

| Imaging Supported   | Indications  | Suspected or known Neurometabolic and/or neurogenetic disorders, but not limited to  |
|---|--|--|
| <p>MRI Brain without and with contrast (CPT® 70553)</p> <p><b>OR</b></p> <p>MRI Brain without contrast (CPT® 70551)</p> <p><b>AND/OR</b></p> <p>Magnetic Resonance Spectroscopy (MRS, CPT® 76390)</p> | <p>Requested by a neurologist or geneticist, or any provider in consultation with a neurologist or geneticist, for:</p> <ul style="list-style-type: none"> <li>• Initial evaluation</li> </ul> <p><b>AND/OR</b></p> <ul style="list-style-type: none"> <li>• Disease monitoring</li> </ul> <p><b>AND/OR</b></p> <ul style="list-style-type: none"> <li>• New or worsening symptoms</li> </ul> <p><b>AND/OR</b></p> <ul style="list-style-type: none"> <li>• Change in therapy is being considered</li> </ul> | <ul style="list-style-type: none"> <li>• X-linked adrenoleukodystrophy (X-ALD, CALD)</li> <li>• Canavan disease</li> <li>• Creatine deficiency</li> <li>• Nonketotic hyperglycinemia</li> <li>• Maple Syrup Urine disease</li> <li>• Metachromatic Leukodystrophy (MCL)</li> <li>• Pelizaeus-Merzbacher disease (PMD)</li> <li>• Hypomyelination and Congenital Cataract</li> <li>• Globoid Cell Leukodystrophy (Krabbe disease)</li> <li>• Mitochondrial disorders (such as, but not limited to Leigh’s syndrome, Kearns-Sayre syndrome, Mitochondrial Encephalopathy with Lactic Acidosis and Stroke-Like Episodes (MELAS))</li> <li>• Alexander disease (ALX, AXD, dysmyelinogenic leukodystrophy)</li> <li>• Megalencephalic leukoencephalopathy with subcortical cysts</li> <li>• Vanishing White Matter (VWM) disease (Leukoencephalopathy with VWM, Childhood Ataxia with CNS Hypomyelination (CACH) syndrome)</li> </ul> |

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**v1.0.2024**

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# Ataxia (PEDHD-20)

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# Ataxia (PEDHD-20.1)

HDP.AT.0020.1.A

v1.0.2024

- 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 also **Developmental Disorders (PEDHD-19)**
- A pertinent evaluation including a detailed history and physical examination with a thorough neurologic examination, should be performed prior to considering advanced imaging, unless the individual is undergoing guideline-supported scheduled follow-up imaging evaluation or request is from or in consultation with a neurologist or neurosurgeon who has seen the individual since onset of symptoms.
- If spinal etiology of ataxia is suspected the following may be indicated
  - MRI Cervical Spine (CPT® 72141 or CPT® 72156) and/or
  - MRI Thoracic Spine (CPT® 72146 or CPT® 72157) and/or
  - MRI Lumbar Spine (CPT® 72148 or CPT® 72158)

| Indications   | Imaging Studies   |
|---|---|
| <ul style="list-style-type: none"> <li>• ANY of the following:                             <ul style="list-style-type: none"> <li>• Ataxia</li> <li>• Hereditary Ataxia</li> <li>• Slowly progressive ataxia</li> </ul> </li> </ul> | <ul style="list-style-type: none"> <li>• ONE of the following:                             <ul style="list-style-type: none"> <li>• MRI Brain without and with contrast (CPT® 70553) <b>OR</b></li> <li>• MRI Brain without contrast (CPT® 70551)</li> </ul> </li> </ul>  |
| <ul style="list-style-type: none"> <li>• Suspected Spinal Etiology</li> </ul>   | <ul style="list-style-type: none"> <li>• MRI Cervical Spine without contrast (CPT® 72141) <b>OR</b></li> <li>• MRI Cervical Spine without and with contrast (CPT® 72156)AND/OR</li> <li>• MRI Thoracic Spine without contrast (CPT® 72146) <b>OR</b></li> <li>• MRI Thoracic Spine without and with contrast (CPT® 72157)AND/OR</li> <li>• MRI Lumbar Spine without contrast (CPT® 72148) <b>OR</b></li> <li>• MRI Lumbar Spine without and with contrast (CPT® 72158)</li> </ul> |

| Indications   | Imaging Studies  |
|---|--|
| <ul style="list-style-type: none"> <li>Acute ataxia following significant head trauma</li> </ul>  | <ul style="list-style-type: none"> <li>ONE of the following:               <ul style="list-style-type: none"> <li>CT Head without contrast (CPT® 70450) <b>OR</b></li> <li>CT Head without and with contrast (CPT® 70470) <b>OR</b></li> <li>MRI Brain without contrast (CPT® 70551) <b>OR</b></li> <li>MRI Brain without and with contrast (CPT® 70553)</li> </ul> </li> </ul>  |
| <ul style="list-style-type: none"> <li>Contraindication to MRI</li> </ul>   | <ul style="list-style-type: none"> <li>ONE of the following:               <ul style="list-style-type: none"> <li>CT Head without and with contrast (CPT® 70470) <b>OR</b></li> <li>CT Head with contrast (CPT® 70460)</li> </ul> </li> </ul>  |
| <ul style="list-style-type: none"> <li>BOTH of the following:               <ul style="list-style-type: none"> <li>Contraindication to MRI <b>AND</b></li> <li>Suspected Spinal Etiology</li> </ul> </li> </ul> | <ul style="list-style-type: none"> <li>CT Cervical Spine without contrast (CPT® 72125) <b>OR</b></li> <li>CT Cervical Spine without and with contrast (CPT® 72127) <b>AND/OR</b></li> <li>CT Thoracic Spine without contrast (CPT® 72128) <b>OR</b></li> <li>CT Thoracic Spine without and with contrast (CPT® 72130) <b>AND/OR</b></li> <li>CT Lumbar Spine without contrast (CPT® 72131) <b>OR AND/OR</b></li> <li>CT Cervical Spine without and with contrast (CPT® 72133)</li> </ul> |

- 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.
- If there is a contraindication to contrast and a spinal etiology is suspected the following may be indicated:
  - CT Cervical Spine (CPT® 72125 or CPT® 72127) **AND/OR**
  - CT Thoracic Spine (CPT® 72128 or CPT® 72130) **AND/OR**
  - CT Lumbar Spine (CPT® 72131 or CPT® 72133)
- Repeat imaging may be appropriate no more frequently than every 12 months when requested by a neurologist and/or neurosurgeon or any provider in consultation with a neurologist and/or neurosurgeon unless there are new signs or symptoms.

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**v1.0.2024**

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# Epistaxis (PEDHD-21)

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# Epistaxis Imaging (PEDHD-21.1)

HDP.ET.0021.1.A

v1.0.2024

- 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** CT Maxillofacial without and with contrast (CPT® 70488)
  - MRI Orbits/Face/Neck without and with contrast (CPT® 70543)



## References (HDP-21)

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# Pseudotumor Cerebri (PEDHD-22)

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# Pseudotumor Cerebri (PEDHD-22.1)

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HDP.PC.0022.1.A

v1.0.2024

- Pseudotumor cerebri indications in pediatric individuals are identical to those for adult individuals. See Papilledema/Pseudotumor Cerebri (HD-17.1) in the Head Imaging Guidelines.

# Cranial Neuropathies (PEDHD-23)

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# Cranial Neuropathies (PEDHD-23.1)

HDP.CN.0023.1.A

v1.0.2024

- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) is indicated for all individuals with new or worsening specific cranial nerve abnormalities.
- MRI ORbits/Face/Neck without and with contrast (CPT® 70543) **OR** MRI Orbits/Face/Neck without contrast (CPT® 70540) is also indicated for individuals with abnormalities in cranial nerves IX, X, XI, or XII.

## References (HDP-23)

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**v1.0.2024**

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# Pediatric Sleep Disorders (PEDHD-24)

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# Pediatric Sleep Disorders (PEDHD-24.1)

HDP.SD.0024.1.A

v1.0.2024

- See Pediatric Sleep Guidelines (SL-3) in the Sleep Apnea and Treatment Clinical Guidelines
- For over 18 years of age OR regarding Oral Appliance OR Hypersomnolence
  - See **Sleep-Related Imaging Guidelines (HD-37)**
- For Obstructive Sleep Apnea:
  - There is NO indication for imaging prior to tonsillectomy (with or without adenoidectomy) for obstructive sleep apnea
  - Initially, endoscopic examination of the upper airway should be performed.
    - CT Maxillofacial without contrast (CPT® 70486) is supported for evaluation of obstructive anatomy if operative intervention, other than a tonsillectomy (with or without adenoidectomy), is being considered.
- For suspected Central Sleep Apnea, the following is supported:
  - MRI Brain without contrast (CPT® 70551) OR
  - MRI Brain without and with contrast (CPT® 70553)
- Advanced imaging is NOT indicated for the following:
  - Confusional arousals
  - Sleep terrors
  - Nightmare disorder
  - Sleep walking (Somnambulism)
  - Bed wetting (Enuresis)
  - Insomnia
  - Narcolepsy (without or with cataplexy)
  - Restless Leg Syndrome/Periodic Limb Movement Disorder
- For suspected sleep-related seizures, see **Epilepsy and Other Seizure Disorders (PEDHD-6)**



## References (HDP-24)

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**v1.0.2024**

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# Temporomandibular Joint (TMJ)/Dental/Maxillofacial Imaging in Children (PEDHD-25)

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# Temporomandibular Joint Imaging (PEDHD-25.1)

HDP.TJ.0025.1.A

v1.0.2024

- Temporomandibular Joint (TMJ) Imaging in Children indications in pediatric individuals are very similar to those for adult individuals. See Temporomandibular Joint Disease (TMJ) (HD-30.1) in the Head Imaging Guidelines.
- Pediatric-specific imaging considerations include the following:
  - There is a paucity of clinical symptoms and poor sensitivity of conventional x-rays in diagnosing TMJ arthritis in pediatric individuals with arthritis
- MRI TMJ (CPT® 70336) is indicated annually for detecting silent TMJ arthritis in children with juvenile idiopathic arthritis (JIA) as requested by a rheumatologist and/or oral/maxillofacial surgeon (OMS) and/or any provider in consultation with a rheumatologist or OMS
- Repeat imaging with MRI TMJ (CPT® 70336) in patients with JIA is indicated for any of the following:
  - Change in signs or symptoms suggesting progression of disease
  - To monitor the effects of treatment<sup>8</sup>
- Bone Scintigraphy/Bone Scan 3 Phase Study (CPT® 78315) in individuals over 12 years of age<sup>7</sup> is appropriate in anticipation or consideration of surgery<sup>7</sup>
- Unilateral condylar hyperplasia is manifested by slow growth in areas of the mandible causing facial asymmetry. It is usually a self-limiting condition seen predominantly in 12–30 year olds.

# Dental/Periodontal/Maxillofacial Imaging (PEDHD-25.2)

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HDP.TJ.0025.2.A

v1.0.2024

- See Dental/Periodontal/Maxillofacial Imaging (HD 30.2)

## References (HDP-25)

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# Movement Disorders including Tourette's Syndrome (PEDHD-26)

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# Tourette's Syndrome (PEDHD-26.1)

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HDP.MD.0026.1.A

v1.0.2024

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

## Movement Disorders (PEDHD-26.2)

HDP.MD.0026.2.A

v1.0.2024

- Movement disorders are hyperkinetic and hypokinetic movements that are involuntary. The majority are diagnosed based on a clinical diagnosis and do not require imaging.
- Typically Benign Movement disorders include:
  - Stereotypies, repetitive rhythmic movements
  - Tics that are vocal or motor with typical onset and course
  - Tourette Syndrome
  - Essential Tremor or tremors of anxiety or weakness
  - Restless Leg Syndrome
- MRI Brain without contrast (CPT® 70551), **OR** MRI Brain without and with contrast (CPT® 70553) is considered in the following clinical scenarios:
  - Atypical clinical features for example, movements that persist in sleep, onset outside of typical age at onset (4-6 years for tics), rapid progression, incomplete or uncertain medication responsiveness, or clinical diagnostic uncertainty, limbic encephalitis
  - Dystonia, intermittent involuntary muscle contractions
  - Chorea, continual irregular movements
  - Ballism, involuntary high amplitude movements
  - Athetosis, slow writhing continuous movements
  - Myoclonus, involuntary muscle jerks (not sleep myoclonus)
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is supported for concerns of:
  - PANS (Pediatric acute-onset neuropsychiatric syndrome) and/or
  - PANDAS (Pediatric autoimmune neuropsychiatric disorder associated with streptococcal infection)
    - Only after a complete medical workup including labs, acute infection, and other comorbid psychiatric disorders (examples, such as Obsessive Compulsive Disorder (OCD), Attention Deficit Hyperactivity Disorder (ADHD) and Autism Spectrum Disorder (ASD) have been investigated.
    - Routine brain imaging is not routinely recommended for OCD, ADHD or ASD.
- See Movement Disorders (HD-15.1) in the Head Imaging Guidelines for the following
  - Suspected Huntington Disease
  - Evaluation for surgical treatment of Essential Tremor or Parkinson's disease, including Deep Brain Stimulator (DBS) placement



- Post-op imaging is supported when ordered by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon for either procedure.

### **Background and Supporting Information**

- There is little evidence to support the use of MRA/CTA and PET in the evaluation of movement disorders.
- Tourette syndrome (TS) is a neurological disorder characterized by repetitive, stereotyped, involuntary movements and vocalizations called tics. The first symptoms of TS are almost always noticed in childhood. Some of the more common tics include eye blinking and other vision irregularities, facial grimacing, shoulder shrugging, and head or shoulder jerking. Perhaps the most dramatic and disabling tics are those that result in self-harm such as punching oneself in the face, or vocal tics including coprolalia (uttering swear words) or echolalia (repeating the words or phrases of others). Many with TS experience additional neurobehavioral problems including inattention, hyperactivity and impulsivity, and obsessive-compulsive symptoms such as intrusive thoughts/worries and repetitive behaviors

Reference: <https://www.ninds.nih.gov/Disorders/All-Disorders/Tourette-Syndrome-Information-Page>

## References (HDP-26)

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# Tuberous Sclerosis (PEDHD-27)

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# Tuberous Sclerosis (PEDHD-27.1)

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HDP.TS.0027.1.A

v1.0.2024

- See Tuberous Sclerosis Complex (TSC) (PEDONC-2.9) in the Pediatric Oncology Imaging Guidelines

# Von Hippel-Lindau Syndrome (VHL) (PEDHD-28)

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# Von Hippel-Lindau Syndrome (VHL) (PEDHD-28.1)

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HDP.VL.0028.1.A

v1.0.2024

- See Von Hippel-Lindau Syndrome (VHL) (PEDONC-2.10) in the Pediatric Oncology Imaging Guidelines

# CNS Infection (PEDHD-29)

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# CNS Infection (PEDHD-29.1)

HDP.CI.0029.1.A

v1.0.2024

- CNS infection imaging indications in pediatric individuals are similar to those for adult individuals. See CNS and Head Infection/Neuro-COVID-19 (HD-14) in the Head Imaging Guidelines.
- CT Head (as per General Guidelines – CT Head (HD-1.4) in the Head Imaging Guidelines) may be considered in Pediatric CNS Infection.
- The following studies are supported for suspected intracranial infection if any signs of CNS infection are present:
  - MRI Brain without and with contrast (CPT® 70553) (preferred) **OR** MRI Brain without contrast (CPT® 70551)
  - CT Head (CPT® 70450, CPT® 70460, **OR** CPT® 70470) in cases where MRI is contraindicated or urgently required prior to lumbar puncture to evaluate for meningitis.
- Repeat imaging is supported if requested by infectious disease specialist, neurologist, ophthalmologist, neuro-ophthalmologist or neurosurgeon or any provider coordinating care with an infectious disease specialist, neurologist, ophthalmologist, neuro-ophthalmologist or neurosurgeon.
- 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://wwwnc.cdc.gov/travel/page/zika-information>)
- Ultrasound Head (CPT® 76506) is supported as an initial imaging study
  - MRI Brain without and with contrast (CPT® 70553) is indicated if the ultrasound is abnormal.
  - Newborn infants with microcephaly should be evaluated as discussed in **Macrocephaly, Microcephaly, and Hydrocephalus (PEDHD-7)**
- The following imaging is considered for newborns or older infants with an open fontanelle and suspected meningitis:
  - Ultrasound Head (CPT® 76506) as an initial imaging study, but is not required
  - MRI Brain without and with contrast (CPT® 70553)<sup>2</sup> is indicated if the ultrasound is abnormal
- A wide spectrum of neurological diseases have been observed in children with COVID-19 infection in children including, but not limited to, Multisystem Inflammatory Syndrome.<sup>10,11</sup>



- MRI Brain without and with contrast (CPT® 70553) **AND/OR**
- MRA Head (CPT® 70544, CPT®70545 **OR** CPT® 70546) **OR** CTA Head (CPT® 70496) **AND/OR**
- MRA Neck (CPT® 70547 **OR** CPT® 70548 **OR** CPT® 70549) **OR** CTA Neck (CPT® 70498) **AND/OR**
  - If Acute Necrotizing Myelitis is suspected, the following are indicated:
    - MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR**
    - MRI Thoracic Spine without and with contrast (CPT® 72157) **AND/OR**
    - MRI Lumbar Spine without and with contrast (CPT® 72158)
- Metabolic (FDG) Brain PET (CPT® 78608) is appropriate to evaluate individuals suspected of having encephalitis, including autoimmune encephalitis, if diagnosis remains unclear after evaluation with MRI Brain, CSF analysis, and lab testing including serology.<sup>12</sup>

### ***Background and Supporting Information***

- Neonatal meningitis most often is 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.

## References (HDP-29)

**v1.0.2024**

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# Scalp and Skull Lesions (PEDHD-30)

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# Scalp and Skull Lesions (PEDHD-30.1)

HDP.SL.0030.1.A

v1.0.2024

- Scalp and skull lesion imaging indications in pediatric individuals are identical to those for adult individuals with the exception of neonates. See Scalp and Skull Lesions (HD-20.1) in the Head Imaging Guidelines.
  - In neonates and young infants, scalp masses include:
    - Congenital lesions (cephalocele-discussed above, dermoid cysts, epidermoid cyst)
    - Vascular lesions (hemangioma, sinus pericranii)
    - 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.
    - Ultrasound Head (CPT® 76506) is supported as an initial imaging study.
    - MRI Brain without and with contrast (CPT® 70553) (preferred) or CT Head without and with contrast (CPT® 70470) is indicated if the ultrasound is abnormal and associated anomalies are suspected
  - The following imaging is indicated for children and adults with Pott Puffy Tumor:
    - MRI Brain without and with contrast (CPT® 70553) OR CT Head without and with contrast (CPT® 70470)<sup>8</sup>
    - Repeat imaging is supported if requested by neurologist, neurosurgeon, otolaryngologist (ENT) and/or oral maxillofacial surgery (OMS) or any provider coordinating care with a neurologist, neurosurgeon, otolaryngologist (ENT) and/or oral maxillofacial surgery (OMS).

## **Background and Supporting Information**

- Pott Puffy Tumor is an abscess involving the frontal bone with adjacent osteomyelitis as the result of a frontal sinus infection that spreads contiguously through the wall of the sinus or through hematogenous spread via the veins that drain sinus mucosa.<sup>8</sup>

## References (HDP-30)

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# Eye Disorders (PEDHD-31)

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## Eye Disorders (PEDHD-31.1)

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- Eye disorder imaging indications in pediatric individuals are close to identical to those for adult individuals. See Eye Disorders and Visual Loss (HD-32.1) in the Head Imaging Guidelines

| Indication  | Imaging Studies   |
|---|---|
| <ul style="list-style-type: none"> <li>• Imaging is supported in the evaluation for congenital disorders or disorders that begin early in life, such as, but not limited to               <ul style="list-style-type: none"> <li>• Optic Nerve Hypoplasia</li> <li>• Septo-Optic Dysplasia</li> <li>• Infantile Nystagmus Syndrome<sup>4</sup></li> </ul> </li> </ul> | <ul style="list-style-type: none"> <li>• MRI Orbits/Face/Neck without contrast (CPT<sup>®</sup> 70540) <b>OR</b></li> <li>• MRI Orbits/Face/Neck without and with contrast (CPT<sup>®</sup> 70543) <b>OR</b></li> <li>• CT Orbits/Temporal Bone with contrast (CPT<sup>®</sup> 70481) <b>OR</b></li> <li>• CT Orbits/Temporal Bone without contrast (CPT<sup>®</sup> 70480)AND/OR</li> <li>• MRI Brain without contrast (CPT<sup>®</sup> 70551) <b>OR</b></li> <li>• MRI Brain with and without contrast (CPT<sup>®</sup> 70553)<sup>2</sup></li> </ul> |

- Repeat imaging is supported if requested by a neurologist, ophthalmologist, neuro-ophthalmologist or neurosurgeon or any provider coordinating care with a neurologist, ophthalmologist, neuro-ophthalmologist or neurosurgeon.
- For traumatic retinal hemorrhages as seen in suspected shaken baby syndrome (See **(PEDHD-4.1)**)

## References (HDP-31)

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