

# Cigna Medical Coverage Policies – Radiology Head Imaging Guidelines

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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 (HD-1)

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# Abbreviations for Head Imaging Guidelines

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Abbreviations for Head Imaging Guidelines	
<b>ACTH</b>	adrenocorticotrophic hormone
<b>AD</b>	Alzheimer's Disease
<b>ADH</b>	antidiuretic hormone
<b>AION</b>	arteritic ischemic optic neuritis
<b>AVM</b>	arteriovenous malformation
<b>CBCT</b>	Cone-beam computerized tomography
<b>CMV</b>	Cytomegalovirus
<b>CSF</b>	cerebrospinal fluid
<b>CT</b>	computed tomography
<b>CTA</b>	computed tomography angiography
<b>DNA</b>	deoxyribonucleic acid
<b>DWI</b>	diffusion weighted imaging (for MRI)
<b>EEG</b>	electroencephalogram
<b>ENT</b>	Ear, Nose, Throat
<b>ESR</b>	erythrocyte sedimentation rate
<b>FDG</b>	fluorodeoxyglucose
<b>FSH</b>	follicle-stimulating hormone
<b>FTD</b>	Frontotemporal Dementia
<b>GCA</b>	giant cell arteritis
<b>GCS</b>	Glasgow Coma Scale
<b>HIV</b>	human immunodeficiency virus
<b>LH</b>	luteinizing hormone
<b>MMSE</b>	mini mental status examination
<b>MRA</b>	magnetic resonance angiography
<b>MRI</b>	magnetic resonance imaging
<b>MRN</b>	magnetic resonance neurography
<b>MS</b>	multiple sclerosis

Abbreviations for Head Imaging Guidelines	
<b>MSI</b>	magnetic source imaging
<b>NAION</b>	non-arteritic ischemic optic neuritis
<b>NPH</b>	normal pressure hydrocephalus
<b>PET</b>	positron emission tomography
<b>PML</b>	progressive multifocal leukoencephalopathy
<b>PNET</b>	primitive neuro ectodermal tumor
<b>PWI</b>	perfusion weighted imaging (for MRI)
<b>SAH</b>	subarachnoid hemorrhage
<b>SIADH</b>	Syndrome of Inappropriate Antidiuretic Hormone Secretion
<b>SLE</b>	systemic lupus erythematosus
<b>TIA</b>	transient ischemic attack
<b>TMJ</b>	temporomandibular joint disease
<b>TSH</b>	thyroid-stimulating hormone
<b>VBI</b>	vertebrobasilar insufficiency
<b>VP</b>	ventriculoperitoneal
<b>XRT</b>	radiation therapy



# General Guidelines (HD-1.0)

HD.GG.0001.0.A

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- A pertinent clinical evaluation including a detailed history, physical examination including a neurological examination since the onset or change in symptoms, and appropriate laboratory studies should be performed prior to considering the use of an advanced imaging (CT, MR, Nuclear Medicine) procedure.
  - A pertinent clinical evaluation furnished via telehealth since the onset or change in symptoms, is treated the same as an in-person clinical evaluation.
  - An exception to a pertinent clinical evaluation can be made if the individual is undergoing a guideline-supported, scheduled follow-up imaging evaluation.
    - Scheduled follow-up of known problems such as, multiple sclerosis, tumors, or hydrocephalus, scheduled surveillance with no new symptoms, screening asymptomatic individual due to family history or otherwise meet criteria for repeat imaging, as well as appropriate laboratory studies and non-advanced imaging modalities
  - A detailed neurological exam is required prior to advanced imaging except in the following scenarios:
    - Tinnitus, TMJ, sinus or mastoid disease, ear pain, hearing loss, eye disease, pituitary disease, and epistaxis. (A pertinent clinical evaluation since onset of symptoms is still required)
    - The request is from a neurologist, neurosurgeon, endocrinologist, otolaryngologist, or ophthalmologist who has seen the individual since onset of symptoms
  - Other meaningful contact (telephone call, electronic mail or messaging) since the onset or change in symptoms, with an established individual can substitute for a face-to-face clinical evaluation

# General Guidelines – Anatomic Issues

## (HD-1.1)

HD.GG.0001.1.A

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- If two studies using the same modality both cover the anatomic region of clinical interest, only one is generally needed, with the exception of the following scenarios:
  - CT Maxillofacial (CPT® 70486, CPT® 70487, or CPT® 70488) or CT Orbital/Temporal bone (CPT® 70480, CPT® 70481, or CPT® 70482): both cover the structures of the orbits, sinuses, and face. Two separate imaging studies are only supported if there is suspicion of simultaneous involvement of more posterior lesions, especially of the region involving the middle or inner ear.
  - Pituitary Gland: one study (either MRI Brain [CPT® 70553] or MRI Orbit, Face, Neck [CPT® 70543]) is adequate to report the imaging of the pituitary. If a previous routine MRI Brain was reported to show a possible pituitary tumor, a repeat MRI with dedicated pituitary protocol is supported.
  - Internal Auditory Canal: (IAC) MRI can be reported as a limited study with one code from the set (CPT® 70540, CPT® 70542, or CPT® 70543), but should not be used in conjunction with MRI Brain codes (CPT® 70551, CPT® 70552, or CPT® 70553) if IAC views are performed as part of the brain.
  - Mandible (jaw): CT Maxillofacial (CPT® 70486, CPT® 70487, or CPT® 70488) or CT Neck (CPT® 70490, CPT® 70491, or CPT® 70492) can be used to report imaging of the mandible. CT Neck will also image the submandibular space.
    - If MRI is indicated, MRI Orbit, Face, Neck (CPT® 70540, CPT® 70542, or CPT® 70543) can be used to report imaging of the mandible and submandibular space.
    - MRI Temporomandibular Joint(s) (TMJ) is reported as CPT® 70336. This code is inherently bilateral and should not be reported twice on the same date of service.
  - MRI Brain without and with contrast (CPT® 70553) is indicated for all individuals with new or worsening specific cranial nerve abnormalities. For Bell's palsy, See **Facial Palsy (HD-6.1)**.
  - MRI Neck without and with contrast (CPT® 70543) is also indicated for individuals with abnormalities in cranial nerves IX, X, XI, or XII<sup>29</sup>

# General Guidelines – Modality (HD-1.2)

HD.GG.0001.2.A

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- MRI is preferable to CT for most indications. For exceptions, See **General Guidelines – CT Head (HD-1.4)**
- MRI for these indications following an initial CT:
  - MRI Brain without and with contrast (CPT® 70553) to follow-up abnormalities seen on CT Head without contrast (CPT® 70450) when a mass, lesion, or infection is found.
  - MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) (preferred) to follow-up abnormalities seen on CT Head without contrast (CPT® 70450) when there is suspected Multiple Sclerosis or other demyelinating disease.
  - MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) to follow up on stroke or TIA when initial CT Head was done on emergent basis.
  - MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) for evaluation of new onset seizures.

# General Guidelines – MRI Brain (HD-1.3)

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- MRI Brain with contrast (CPT® 70552) should not be ordered except to follow-up on a very recent non-contrast MRI Brain (within two weeks).
- The AMA CPT manual does not describe nor assign any minimum or maximum number of sequences for any CT or MRI study. Both MRI and CT imaging protocols are often influenced by the individual clinical situation of the individual and additional sequences are not uncommon. There are numerous MRI sequences that are performed to evaluate specific clinical questions, and this technology is constantly undergoing development. Additional sequences, however, are still performed and coded under the routine MRI Brain CPT® 70551, CPT® 70552, or CPT® 70553.

# General Guidelines – CT Head (HD-1.4)

HD.GG.0001.4.A

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- Scenarios in which MRI is contraindicated (i.e. pacemakers, ICDs, cochlear implants, aneurysm clips, orbital metallic fragments, etc.)
- In urgent cases, CT Head, contrast as requested is supported [CT Head without and with contrast (CPT® 70470), CT Head with contrast (CPT® 70460) or CT Head without contrast (CPT® 70450)]
- CT Head without contrast (CPT® 70450) is supported 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).
  - Prior to lumbar puncture in individuals with cranial complaints (without contrast) (CPT® 70450)
  - Evaluation of optic disc edema and/or papilledema, a non-contrast CT Head is useful to assess for space-occupying processes such as intracranial hemorrhage, mass effect, and hydrocephalus, See **Papilledema/Pseudotumor Cerebri (HD-17.1)** and **Eye Disorders and Visual Loss (HD-32.1)**

# General Guidelines – CT and MR Angiography (CTA and MRA) (HD-1.5)

HD.GG.0001.5.A

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- MRA Head may be performed without contrast (CPT® 70544), with contrast (CPT® 70545), or without and with contrast (CPT® 70546).
- 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 Head is performed without and with contrast (CPT® 70496)
- CTA Neck is performed with and without contrast (CPT® 70498)
- Indications for CTA or MRA Head and Neck vessels include, but are not limited to the following:<sup>12,24</sup>
  - Pulsatile tinnitus
  - Hemifacial spasm if consideration for surgical decompression
  - Evaluation of stroke or TIA (See **Stroke/TIA (HD-21.1)** ) including collateral assessment
  - Trigeminal neuralgia failed medical therapy
  - Cerebral venous 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 (appropriate to limit CTA to include only the head to avoid unnecessary radiation to the individual)
  - Noninflammatory vasculopathy, including radiation vasculopathy
  - Traumatic vascular injuries
  - Vascular malformations, vascular anatomic variants and fistulas
  - Arterial 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
  - Eagle Syndrome - Dynamic/positional CTA to assess for vascular compression (also known as bow-hunter's syndrome)<sup>12</sup>
  - NOTE: Evaluation of posterior circulation disease requires both neck and head MRA/CTA to visualize the entire vertebral-basilar system.

- MRA Head without, with, or without and with contrast or CTA Head for follow up of aneurysm clipping or coiling procedures (See **Intracranial Aneurysms (HD-12.1)** )
- MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496, CPT® 70498) **AND/OR** MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498) is indicated if arterial dissection is suspected, or known and re-evaluation is needed (as directed by neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon)<sup>12,24</sup>
  - There are high risk scenarios including but not exclusive to: Fibromuscular dysplasia (FMD), Marfan Disease, motor vehicle accident (MVA) with whiplash, or chiropractic manipulation
- Other vascular imaging indications for headaches require additional information. See **Stroke/TIA (HD-21.1)** , **Sudden Onset of Headache (HD-11.3)** , **New Headache Onset Older than Age 50 (HD-11.7)** , **Abnormal Blood Clotting (HD-11.9)** , **Pregnancy (HD-11.10)** , **Physical Exertion (HD-11.11)** , and **Systemic Infections (HD-11.13)**
- 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 is supported

# General Guidelines – PET Coding Notes (HD-1.6)

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- Metabolic Brain PET should be reported as Metabolic Brain PET (CPT® 78608)
- Amyloid Brain PET should be reported as limited PET (CPT® 78811) or limited PET/CT (CPT® 78814)



# General Guidelines – Other Imaging Situations (HD-1.7)

HD.GG.0001.7.C

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- Nausea and vomiting, persistent, unexplained and a negative GI evaluation: MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) is supported.
- 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 without and with contrast (CPT® 70553) and/or MRI Cervical Spine without and with contrast (CPT® 72156) and/or MRI Thoracic Spine without and with contrast (CPT® 72157) is appropriate for consideration of neurosarcoidosis.<sup>4, 32,33,34,35</sup> For non-neurologic imaging related to sarcoidosis, See **Sarcoid (CH-15.1)**
- 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 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 or as ordered by a specialist (neurologist or neurosurgeon) or any provider in consultation with a specialist
- 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. If symptomatic, abdomen imaging (MRI or CT) may be indicated as ordered by a specialist or any provider in consultation with a specialist
- CT scans represent the gold standard for diagnosis of an elongated styloid process<sup>31</sup>. CT Maxillofacial and/or CT Neck with contrast or without contrast (CPT® 70487 or CPT® 70486 and/or CPT® 70491 or CPT® 70490) are supported for evaluation of Eagle Syndrome<sup>30,31</sup>. See **General Guidelines – CT and MR Angiography (CTA and MRA) (HD-1.5)** for vascular imaging related to Eagle Syndrome<sup>15</sup>
- For facial feminization/masculinization procedural planning:
  - Preoperative CT requests for CT Maxillofacial without contrast (CPT® 70486) with or without 3D rendering (CPT® 76377), and/or CT Neck with contrast (CPT®

70491) are supported if the individual has a health plan benefit covering the facial feminization/masculinization and laryngoplasty surgeries and the surgery has been approved.

- Additionally CT Head without (CPT® 70450) for the following:
  - History of prior cranial surgery
  - History of head trauma
  - Presence of neurological signs and symptoms
- Preoperative imaging is not supported if the facial feminization/masculinization and laryngoplasty surgeries are not health plan covered benefits
- 3D Rendering
  - CPT® 76377 (3D rendering requiring image post-processing on an independent workstation) is supported 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 See **Intracranial Aneurysms (HD-12.1)**, **Arteriovenous Malformations (AVMs) and Related Lesions (HD-12.2)**, **Stroke/TIA (HD-21.1)**, and **Cerebral Vasculitis (HD-22.1)**<sup>26</sup>
  - 3D Rendering (CPT® 76377) for surgical planning and surgical follow up after craniotomy when ordered by surgical specialist or any provider in consultation with a 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

## Background and Supporting Information

- Eagle syndrome is due to a calcified stylohyoid ligament or an elongated styloid process. It may cause neck, face or jaw pain and may cause compression of the vessels that carry blood to the brain, neck and face (carotid artery).

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**v1.0.2023**

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# Taste and Smell Disorders (HD-2)

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## Taste and Smell Disorders (HD-2.1)

HD.TS.0002.1.A

v1.0.2023

- MRI Brain without and with contrast (CPT® 70553) or without contrast (CPT® 70551) and/or MRI Orbit, Face, and Neck without (CPT® 70540) or without and with contrast (CPT® 70543) is indicated with unexplained unilateral or bilateral anosmia (inability to perceive odor) or dysgeusia (complete or partial loss of taste)
- CT Maxillofacial (CPT® 70486, CPT® 70487 or CPT® 70488) is indicated initially if sinus or facial bone disorders are suspected
- For individuals who test positive for SARS-CoV-2, MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) is appropriate for neurologic symptoms or signs, other than change in taste or smell, for consideration of other pathology. See **Neuro-COVID-19 and Sars-CoV-2 Vaccines (HD-14.2)** and **Stroke/TIA (HD-21.1)**

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

In those individuals with consideration of COVID-19 due to signs/symptoms, testing to identify for SARS-CoV-2 is encouraged.

## References (HD-2)

**v1.0.2023**

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# Ataxia (HD-3)

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# Ataxia (HD-3.1)

HD.AX.0003.1.A

v1.0.2023

- Common manifestations include: poor coordination, an abnormal (including wide-based) gait, abnormal finger to nose testing, abnormal rapid alternating movements, abnormal eye movements, and/or difficulty with navigation of stairs and around corners.<sup>3</sup>
- MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) is appropriate in all individuals with ataxia:
  - MRI Cervical, Thoracic and/or Lumbar Spine without contrast or with and without contrast (CPT® 72141 or CPT® 72156, CPT® 72146 or CPT® 72157, CPT® 72148 or CPT® 72158) if spinal disease is suspected
  - If these symptoms are acute and stroke is suspected, See **Stroke/TIA (HD-21.1)**
  - If MS is suspected, See **Multiple Sclerosis (MS) (HD-16.1)**
  - CT Head without contrast (CPT® 70450) and/or CT Temporal Bone without contrast (CPT® 70480) added if these symptoms are acute following head trauma. See **Head Trauma (HD-13.1)**
- If brain tumor is suspected, See **Primary Central Nervous System Tumors (ONC-2.1)** in the Oncology Imaging Guidelines.
- MRI Brain without contrast (CPT® 70551), or CT Head without contrast (CPT® 70450) if there is a contraindication to MRI, for those with gait abnormalities, cognitive impairment and/or urinary symptoms (e.g. urgency, frequency and/or incontinence) for the evaluation of Normal Pressure Hydrocephalus. See Normal Pressure Hydrocephalus (NPH) (HD-8.4)

## **Background and Supporting Information**

- In general, MRI is preferred over CT, unless there is a history of acute trauma or contraindication to MRI. For all other causes, MRI provides better visualization of the cerebellum and posterior fossa.

## References (HD-3)

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**v1.0.2023**

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# Behavioral Disorders (HD-4)

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# Behavioral Disorders – General Information (HD-4.0)

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HD.BD.0004.0.A

v1.0.2023

- Autism
  - The group of diagnoses, including Asperger syndrome, are classified as pervasive development disorders (PDD). These diagnoses are established on clinical criteria, and no imaging study can confirm the diagnosis.
  - Comprehensive evaluation for autism might include history, physical exam, audiology evaluation, speech, language, and communication assessment, cognitive and behavioral assessments, and academic assessment.
    - MRI Brain without and with contrast (CPT® 70553) is indicated:
      - New or worsening focal neurologic findings documented on a pertinent physical
      - Loss of developmental milestones and/or regression
    - PET imaging is considered investigational in the evaluation of individuals with autism spectrum disorders.

# Behavioral Disorders and Mental Status Change (HD-4.1)

HD.BD.0004.1.C

v1.0.2023

- Psychiatric diagnoses do not routinely require advanced imaging
- MRI Brain without contrast (CPT® 70551), or MRI Brain without and with contrast (CPT® 70553), or CT Head without contrast (CPT® 70450)
  - Acute mental status change, disturbance in consciousness or arousal state
  - Psychotic disorders (including schizophrenia), bipolar disorder and related disorders in the following clinical presentations:
    - Acute first episode onset
    - Late onset over age 40
    - 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 ECT treatment, utilize to screen for intracranial disease: either MRI Brain without contrast (CPT® 70551) or CT Head without contrast (CPT® 70450)
- Deep Brain Stimulation Therapy for psychiatric disorders is considered investigation and experimental, so imaging is not indicated.

## References (HD-4)

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# Chiari and Skull-Base Malformation (HD-5)

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# Chiari I Malformations (HD-5.1)

HD.CM.0005.1.A

v1.0.2023

This involves 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.

- For initial evaluation, MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) and MRI of the entire spine without contrast (CPT® 72141, CPT® 72146, CPT® 72148) or without and with contrast (CPT® 72156, CPT® 72157, CPT® 72158) is indicated.
- For CSF flow imaging See **CSF Flow Imaging (HD-24.4)**
- Repeat imaging at the discretion of or in consultation with the specialist coordinating the individual's care for this condition.
- Repeat MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) is indicated for individuals with a known Chiari I malformation when any of the following are present:
  - There are new or worsening signs or symptoms
  - A surgical procedure is actively being considered.
- Repeat MRI Spine imaging is not indicated for individuals with normal initial spine imaging unless there are new or worsening signs or symptoms from baseline that suggest spinal cord pathology.
- Repeat brain and spine imaging in individuals with Chiari I malformations and known syringomyelia or hydromyelia is highly individualized
- Familial screening is not indicated for Chiari I Malformations.

# Chiari II Malformations (Arnold Chiari Malformation) (HD-5.2)

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HD.CM.0005.2.A

v1.0.2023

These 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.

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

## Chiari III and IV Malformations (HD-5.3)

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HD.CM.0005.3.A

v1.0.2023

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

- Repeat brain and spine imaging in individuals with Chiari III and IV malformations is highly individualized and is indicated at the discretion of or in consultation with the specialist coordinating the individual's care for this condition.
- Familial screening is not indicated for Chiari III or IV Malformations.

## Basilar Impression (HD-5.4)

HD.CM.0005.4.A

v1.0.2023

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

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

# Platybasia (HD-5.5)

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**HD.CM.0005.5.A****v1.0.2023**

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

- Individuals are usually asymptomatic, but either MRI Brain without contrast (CPT® 70551) or CT Head without contrast (CPT® 70450) is indicated to establish a diagnosis when clinically suspected.

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**v1.0.2023**

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# Facial Palsy (Bell's Palsy)/Hemifacial Spasm (HD-6)

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## Facial Palsy (HD-6.1)

HD.FP.0006.1.A

v1.0.2023

Typical features of Bell's palsy include variable initial ipsilateral temporal and auricular pain before facial weakness, onset over 72 hours, ipsilateral complete facial weakness, and an otherwise normal neurological and systemic examination. There is usually slow improvement over several months. Unless "red flags" are present, imaging is not necessary.

- MRI Brain without and with contrast (CPT® 70553) (with attention to posterior fossa and IACs) or without contrast (CPT® 70551) and/or MRI Orbit, Face and Neck without contrast (CPT® 70540) or with and without contrast (CPT® 70543) are supported with the following "red flags" of unexplained facial paresis/paralysis in clinical scenarios with:
  - Trauma to the temporal bone
  - History of tumor, systemic cancer, HIV or Lyme disease
  - No improvement in 8 weeks
  - No full recovery in 3 months
  - Gradual onset over weeks to months
  - Vertigo or hearing loss
  - Bilateral involvement
  - Other atypical or inconsistent features including:
    - Second episode of paralysis on the same side
    - Paralysis of isolated branches of the facial nerve
    - Paralysis associated with other cranial nerves
- MRI Brain without and with contrast (CPT® 70553) for known sarcoidosis with suspected neurosarcoid or CNS involvement is supported



## Hemifacial Spasm (HD-6.2)

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HD.FP.0006.2.A

v1.0.2023

- MRI Brain without and with contrast (CPT® 70553)
- Add CTA Head (CPT® 70496) or MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) for consideration of vascular decompression surgical procedure to clarify the vascular anatomy in individuals who have failed conservative medical management

## References (HD-6)

**v1.0.2023**

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# Recurrent Laryngeal Palsy/Vocal Cord Palsy (HD-7)

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# Recurrent Laryngeal Palsy/Vocal Cord Palsy (HD-7.1)

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HD.RL.0007.1.A

v1.0.2023

- The following are supported with unilateral vocal cord/fold palsy identified by laryngoscopy:
  - MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) **and** MRI Orbit, Face and Neck with and without contrast (CPT® 70543) or CT Neck with contrast (CPT® 70491)
  - CT Chest with contrast (CPT® 71260) added with **left vocal cord palsy**

## References (HD-7)

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**v1.0.2023**

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# Dementia (HD-8)

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# Dementia (HD-8.1)

HD.DM.0008.1.C

v1.0.2023

- For acute mental status change See **Behavioral Disorders and Mental Status Change (HD-4.1)** and Stroke/TIA (HD-21.1)
- For Medicare members with Mild Cognitive Impairment (MCI) or Mild Alzheimer's Dementia being considered for enrollment in an approved clinical trial under Coverage with Evidence Development (CED) investigating amyloid-reduction therapy. See Medicare National Coverage Determinations (NCD) Manual, Section 220.6.20
- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) or CT Head without contrast (CPT® 70450) is supported after an initial clinical diagnosis of dementia has been established.
  - The following components are required:
    - A detailed neurological exam is not required when dementia is diagnosed with abnormal bedside mental status testing by score results
    - Established diagnosis of dementia: date of onset of symptoms with documentation of 6 months of cognitive decline based on a detailed history of memory loss with impairment of day-to-day activities confirmed by family members or others with knowledge of the individual's status
    - OR
    - Results of bedside testing and/or neuropsychological testing can be performed when history and bedside mental status examination cannot provide a confident diagnosis.
      - Examples of abnormal bedside mental status testing such as Mini-Mental Status Exam (MMSE) with score <26, Montreal Cognitive Assessment Survey (MoCA) with score <26, Memory Impairment Screen (MIS) with score <5, or the St. Louis University Mental Status (SLUMS) with score <21.
    - Presumptive causes or etiology/ies of dementia
      - Cannot occur exclusively during bouts of delirium
      - Cannot be explained by another mental disorder
- MRI Brain without contrast (CPT® 70551), or CT Head without contrast (CPT® 70450) if there is a contraindication to MRI and/or contrast, for those with gait abnormalities, cognitive impairment and/or urinary symptoms (e.g. urgency, frequency and incontinence) for the evaluation of Normal Pressure Hydrocephalus. (See **Normal Pressure Hydrocephalus (HD-8.4)**).
- 3D Brain imaging in dementia:
  - 3D analysis of the temporal lobes and hippocampus (also known as volumetric analysis or Neuro Quant) (CPT® 76377) lacks sufficient specificity and sensitivity to be clinically useful in the evaluation or follow up of individuals with dementia. Its use is limited to research studies and it is otherwise considered to be

investigational and experimental in routine clinical practice. Advanced imaging studies, or other procedures, are considered investigational and experimental if there is a paucity of supporting evidence; if the evidence has not matured to exhibit improved health parameters or; the advanced imaging study/procedure lacks a collective opinion of support.



## Dementia - PET (HD-8.2)

HD.DM.0008.2.C

v3.0.2022

- Prior to consideration of PET imaging for a diagnosis of dementia, all of the following components are required:
  - Established diagnosis of dementia: date of onset of symptoms with documentation of 6 months of cognitive decline based on a detailed history of memory loss with impairment of day-to-day activities confirmed by family members or others with knowledge of the individual's status  
OR
  - Results of bedside testing and/or neuropsychological testing can be performed when history and bedside mental status examination cannot provide a confident diagnosis.
    - Examples of abnormal bedside mental status testing such as Mini-Mental State Exam (MMSE) with score <26, Montreal Cognitive Assessment Survey (MoCA) with score <26, Memory Impairment Screen (MIS) with score <5, or the St. Louis University Mental Status (SLUMS) with score <21.
  - Results of any structural imaging (MRI or CT Head) performed.
  - Relevant laboratory tests (For example, but not limited to, B-12, thyroid function tests).
  - Presumptive causes or etiology/ies of dementia
    - Cannot occur exclusively during bouts of delirium
    - Cannot be explained by another mental disorder

CPT® 78608 is used to report FDG PET metabolic brain studies for dementia, seizure disorders, and dedicated PET tumor imaging studies of the brain.

CPT® 78609 is used to report PET Brain perfusion studies that are not performed with FDG. These scans are nationally noncovered by Medicare.

CPT® 78811 (limited PET) or CPT® 78814 (limited PET/CT) are used to report Amyloid Brain PET (these codes are for static images to measure amyloid, as opposed to the FDG PET which is a metabolic study).

- FDG PET for Dementia and Neurodegenerative Diseases
  - For Medicare members See the Medicare National Coverage Determinations (NCD) Manual, Section 220.6.13 for the coverage policy
  - FDG Brain PET (CPT® 78608) is useful in distinguishing between Alzheimer's disease (AD) and Frontotemporal dementia (FTD). It is otherwise considered investigational and experimental for the purpose of diagnosis and management of mild cognitive impairment (MCI) and other forms of dementia including, but not limited to, Lewy Body disease, Parkinson's disease, Normal Pressure

Hydrocephalus and Chronic Traumatic Encephalopathy. Advanced imaging studies, or other procedures, are considered investigational and experimental if there is a paucity of supporting evidence; if the evidence has not matured to exhibit improved health parameters or; the advanced imaging study/procedure lacks a collective opinion of support. Appropriate documentation should support concern for one of the variants of Frontotemporal dementia (Behavioral Variant or Primary Progressive Aphasia type FTD) based on a detailed history and exam findings (which includes neuropsychological testing) and meet the following criteria:

- Meets diagnostic criteria for AD and FTLD (frontotemporal lobar dementia); and
  - Has a documented cognitive decline of at least 6 months; and
  - Evaluation has ruled out specific alternative neurodegenerative disease or causative factors; and
  - Cause of clinical symptoms is uncertain; and
  - The results are expected to help clarify the diagnosis between FTLD and AD and help guide future treatment.
- Amyloid Brain PET
    - For Medicare members See Medicare National Coverage Determinations (NCD) Manual, Section 220.6.20 for coverage policy
    - Amyloid Brain PET (CPT® 78811 or CPT® 78814) imaging is considered experimental and investigational in the diagnosis of Alzheimer's disease and in differentiating between Alzheimer's disease and other neurodegenerative/neurologic disorders. Advanced imaging studies, or other procedures, are considered investigational and experimental if there is a paucity of supporting evidence; if the evidence has not matured to exhibit improved health parameters or; the advanced imaging study/procedure lacks a collective opinion of support.
    - For Cerebral Amyloid Angiopathy See **Stroke/TIA (HD-21.1)**

#### Background and Supporting Information

- The frontotemporal dementias (FTDs) are a group of neurodegenerative disorders that differ from Alzheimer's disease. The basic pathology involves accumulation of tau proteins in the brain rather than amyloid. Onset tends to be younger (less than 65) and progression usually more rapid than in senile dementia-Alzheimer type (SDAT). There is no treatment, and the medications used to help memory in Alzheimer's disease are not effective.
- There are several subtypes of FTD; most common are the behavioral variant with early loss of executive functions, impaired judgment disinhibition and impulsivity, and the semantic variant with primary and progressive loss of language ability. Other less common subtypes include progressive supranuclear palsy, corticobasal syndrome, and FTD associated with motor neuron disease.

- Diagnosis is based on clinical features, neuropsychological testing, and brain imaging (preferably MRI) to rule out other structural disease. Metabolic (FDG) PET Brain is helpful by demonstrating patterns of abnormality more consistent with FTD than Alzheimer's disease.

# Lewy Body Dementia (LBD) - SPECT Brain Scan (HD-8.3)

HD.DM.0008.3.A

v1.0.2023

- Dementia with Lewy bodies is often hard to diagnose because its early symptoms may resemble those of Alzheimer's or a psychiatric illness. Over time people with LBD often develop similar symptoms due to the presence of Lewy bodies in the brain.
  - Clinicians and researchers may use the "1-year rule" to help make a diagnosis. If cognitive, psychiatric, emotional, and/or personality symptoms appear at the same time as or at least a year before movement problems/parkinsonism, the diagnosis is dementia with Lewy bodies. If cognitive problems develop more than a year after the onset of movement problems, Parkinson's disease, the diagnosis is Parkinson's disease dementia (PDD).
- Core Clinical Symptoms
  - Dementia
  - Movement problems/parkinsonism
  - Cognitive fluctuations
  - Visual hallucinations
  - REM sleep behavior disorder
- Supportive Clinical Symptoms
  - Extreme sensitivity to antipsychotic medications
  - Falls, fainting
  - Severe problems with involuntary functions (maintaining blood pressure, incontinence, constipation, loss of smell)
  - Changes in personality and mood (depression, apathy, anxiety)
- Prior to consideration of SPECT Brain Scan for a diagnosis of LBD, all of the following components are required:
  - Established diagnosis of dementia: date of onset of symptoms with documentation of 6 months of cognitive decline based on a detailed history of memory loss with impairment of day-to-day activities confirmed by family members or others with knowledge of the individual's status OR
  - Results of bedside testing and/or neuropsychological testing can be performed when history and bedside mental status examination cannot provide a confident diagnosis.
    - Examples of abnormal bedside mental status testing such as Mini-Mental State Exam (MMSE) with score <26, Montreal Cognitive Assessment Survey (MoCA) with score <26, Memory Impairment Screen (MIS) with score <5, or the St. Louis University Mental Status (SLUMS) with score <21.

- Results of any structural imaging (MRI or CT Head) performed
- Relevant laboratory tests (Such as B-12, thyroid function tests)
- SPECT Brain Scan (CPT® 78803 or CPT® 78830) is supported after all of the above criteria are met
- PET Brain is not indicated for LBD

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

#### Test Results Supporting Diagnosis

- Abnormal 123iodine-MIBG myocardial scintigraphy showing reduced communication of cardiac nerves
- Sleep study confirming REM sleep behavior disorder without loss of muscle tone

# Normal Pressure Hydrocephalus (NPH) (HD-8.4)

HD.DM.0008.4.C

v1.0.2023

- CT Head without contrast (CPT® 70450) or MRI Brain without contrast (CPT® 70551) is indicated if the individual has at least two symptoms involving gait abnormality (See Background and Supporting Information), urinary incontinence, or dementia AND
  - The clinical symptoms cannot be completely explained by other neurological or non-neurological disease, AND
  - There is no apparent preceding disorder that would cause hydrocephalus<sup>24,25,26</sup>
- The components of Dementia are delineated in Dementia (HD 8.1), but include:
  - Results of testing and/or neuropsychological testing can be performed when history and mental status examination cannot provide a confident diagnosis.
  - Examples of abnormal mental status testing such as Mini-Mental State Exam (MMSE) with score <26, Montreal Cognitive Assessment Survey (MoCA) with score <26, Memory Impairment Screen (MIS) with score <5, or the St. Louis University Mental Status (SLUMS) with score <21.
  - Relevant laboratory tests (For example, but not limited to, B-12, thyroid function tests, etc.)
  - Presumptive causes or etiology/ies of dementia
    - Cannot occur exclusively during bouts of delirium
    - Cannot be explained by another mental disorder
- MRI Brain (CPT® 70551, CPT® 70552, or CPT® 70553) is not generally indicated for the diagnosis of NPH if a CT has been performed. However, MRI Brain is indicated if needed for presurgical planning.
  - After neuro imaging the next step is CSF sampling, drainage, and dynamics
- Follow-up imaging for individuals diagnosed with NPH with a shunt should follow **Hydrocephalus Shunts (HD-11.14)**, or **Low Pressure Headache and CSF Leak (HD-11.15)**

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

Normal Pressure Hydrocephalus (NPH) seen typically in the elderly. It comprises a triad of symptoms: cognitive dysfunction, incontinence of urine, and gait disturbance (typically a “magnetic”, small-step, or broad based gait). The reported neuroradiologic marker for this is ventriculomegaly (enlarged ventricles) in the brain. Unfortunately, these symptoms and this neuroradiologic finding is common in the elderly, making the diagnosis of NPH in any given individual problematic. It is radiographically common and clinically rare.

## References (HD-8)

**v1.0.2023**

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# Epilepsy/Seizures (HD-9)

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# Epilepsy/Seizures (HD-9.1)

HD.EP.0009.1.A

v1.0.2023

- MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) for:
  - Evaluation of new onset seizures
  - Refractory or drug resistant seizures
  - Change in the type of seizure
  - If CT Head without contrast (CPT® 70450) was performed for an initial evaluation for new onset seizure, MRI (as described above) is indicated for additional evaluation
  - Follow-up studies after a previous routine normal study if performed with special “Epilepsy Protocol” (typically 3T magnet, thin sections with angled slices through hippocampus and temporal lobes)
- CT Head without contrast (CPT® 70450) for:
  - Evaluation of structural findings in seizure etiologies that contain dystrophic calcifications, such as with oligodendrogliomas and tuberous sclerosis.
  - Acute setting of seizure evaluation
- CT Head (contrast as requested) when:
  - MRI is contraindicated

# Presurgical Work-Up for Drug-Resistant Epilepsy (HD-9.2)

HD.EP.0009.2.C

v1.0.2023

- The following requests are supported for consideration of potential surgery:
  - MRI Brain with and without 3T/7T (CPT® 70551 or CPT® 70553)
  - FDG PET (CPT® 78608)
    - Medicare covers FDG PET for pre-surgical evaluation for the purpose of localization of a focus of refractory seizure activity. The complete coverage policy is found in the Medicare National Coverage Determinations (NCD) Manual, Section 220.6.9
  - Ictal SPECT (CPT® 78803)
  - Functional MRI (fMRI) (CPT® 70555 or CPT® 70554) See **Functional MRI (fMRI) (HD-24.2)**
- When noninvasive EEG monitoring is insufficient, intracranial monitoring with stereo-EEG or grids/strips and electrodes is indicated with additional imaging for neuronavigation. See **Neurosurgical Imaging (HD-28.1)** and **Neuronavigation (HD-28.2)**
  - Post-operative imaging including after intracranial (EEG) monitoring per neurosurgeon or any provider in consultation with neurosurgeon.
- See **Primary Central Nervous System Tumors-General Considerations (ONC-2.1)** in the Oncology Imaging Guidelines and/or **Neurosurgical Imaging (HD-28.1)** for additional imaging requests for surgery

## *Background and Supporting Information*

Below are examples of surgical treatment or an interventional modality that may be under active consideration for individuals with intractable epilepsy (not all inclusive):

- 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 (Neuromodulation) including

- Vagus Nerve Stimulation (VNS)
- Responsive Neurostimulation (RNS) system also known as NeuroPace
- Deep Brain Stimulation

## References (HD-9)

**v1.0.2023**

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# Facial Pain/Trigeminal Neuralgia (HD-10)

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# Facial Pain/Trigeminal Neuralgia (HD-10.1)

HD.TM.0010.1.C

v1.0.2023

- MRI Brain without and with contrast (CPT® 70553) (with special attention to the skull base), and/or facial imaging, MRI Orbit without and with contrast (CPT® 70543) for:
  - Suspected tic douloureux or one of its cranial nerve variants such as glossopharyngeal neuralgia (CN IX)
  - Concern about an underlying diagnosis of multiple sclerosis
  - Trigeminal neuralgia which involves the ophthalmic nerve, (periorbital or forehead pain), once post-herpetic neuralgia (a complication of shingles), facial pain consistent with trigeminal branch nerve involvement (infra-orbital or mental nerve) has been excluded by history
- MRA Head (CPT® 70544, CPT® 70545 or CPT® 70546) or CTA Head (CPT® 70496) for:
  - Failed medical treatment
  - Surgical planning

## Background and Supporting Information

The differential diagnosis of facial pain is extensive, complex, and difficult, and there is considerable case-to-case variation in optimal imaging pathway.

## References (HD-10)

**v1.0.2023**

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# Headache (HD-11)

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# Headache General Guidelines (HD-11.0)

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HD.HA.0011.0.C

v1.0.2023

- Advanced imaging of the head is NOT indicated for any of the following:
  - Primary headache disorder in the absence of focal neurological deficits or “red flags” (See *Headaches with Red Flags* (HD-11.2) and *Migraine Exceptions* (HD-11.17))
  - Newly diagnosed migraine or tension-type headache with a normal neurologic exam or for chronic stable headache including migraine with no neurologic deficit.

## Background and Supporting Information

- The yield of detecting abnormal, treatable lesions by CT or MRI in individuals with headache but normal neurological exam has been found to be low.

# Headache and Suspected Vascular Dissection (HD-11.1)

HD.HA.0011.1A

v1.0.2023

- CTA Neck (CPT® 70498) and MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) are appropriate in the evaluation for headache with suspected carotid or vertebral artery dissection and in certain high risk scenarios including, but not exclusive to: Fibromuscular dysplasia (FMD), Marfan Disease, acute MVA with whiplash, and acute headache and/or neck pain due to chiropractic manipulation.
  - CTA Head (CPT® 70496) or MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) is appropriate if there is concern for extension of a carotid dissection to the skull base or above
  - Evaluation of posterior circulation disease requires both neck and head MRA/CTA to visualize the entire vertebral-basilar system
- MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498) if arterial dissection is suspected, or known and re-evaluation is needed (as directed by neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon)
- MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496, or CPT® 70498) if arterial dissection is suspected, or known and re-evaluation is needed (as directed by neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon)
- Other vascular imaging indications for headaches require additional information. See **Stroke/TIA (HD-21.1)**, **Sudden Onset of Headache (HD-11.3)**, **New Headache Onset Older than Age 50 (HD-11.7)**, **Abnormal Blood Clotting (HD-11.9)**, **Pregnancy (HD-11.10)**, **Physical Exertion (HD-11.11)**, and **Systemic Infections (HD-11.13)**

## Headaches with Red Flags (HD-11.2)

HD.HA.0011.2A

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- Red Flags - If any of the below unusual symptoms or history are present advanced imaging studies are supported (see relevant section):
  - Cancer history or immunosuppression See **Cancer or Immunosuppression (HD-11.8)**
  - Sudden onset See **Sudden Onset of Headache (HD-11.3)**
  - Headache accompanied by seizures, vomiting, focal neurological complaints including dizziness, visual change, acute hypertension or altered mental status (See **Primary Central Nervous System Tumors – General Considerations (ONC-2.1)** in the Oncology Imaging Guidelines and **Stroke/TIA (HD-21.1)**)
  - New onset age >50 See **New Headache Onset Older than Age 50 (HD-11.7)** and **Migraine Exceptions (HD-11.17)**
  - History of head trauma (See **Headaches Associated with Head Trauma (HD-11.12)**, and **Head and Facial Trauma (HD-13)**)
  - Headache precipitated by cough or valsalva, physical exertion, or sexual activity See **Physical Exertion (HD-11.11)**
  - Currently pregnant (including pregnancy and the immediate postpartum period) See **Pregnancy (HD-11.10)**
  - Hypercoagulable state or bleeding disorder See **Abnormal Blood Clotting (HD-11.9)**
  - New persistent headache See **Migraine Exceptions (HD-11.17)**
  - Headache awakens individual from sleep
- MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) or CT Head without contrast (CPT® 70450):
  - Unusual symptoms or history as detailed in the “Red Flag” sections above
  - Abnormal examination findings (altered mental status, papilledema, focal signs or symptoms including unilateral weakness or sensory loss, loss of coordination, seizures, gait disturbance, cranial nerve abnormality, vision loss, nystagmus, dysarthria, dysphagia, fever, meningismus)
- Chronic headache with significant change in character, severity or frequency of headache (For example: progressively worsening headache over a period of days or weeks, transformation of established migraine to chronic daily headaches):
  - MRI Brain without and with contrast (CPT® 70553); or
  - MRI Brain without contrast (CPT® 70551); or
  - CT Head without contrast (CPT® 70450)
  - MRA/MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA/CTV Head (CPT® 70496) can be added to evaluate the recent onset of a progressive,

severe, daily headache, with or without papilledema and concern for cerebral venous sinus thrombosis.

- CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. 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
- For papilledema: See **Papilledema/Pseudotumor Cerebri (HD-17.1)**

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

Aura symptoms may accompany or precede a headache within 60 minutes and may include, but are not exclusive to the following symptoms:

- Visual (flashing lights, loss of vision)
- Sensory (paresthesia)
- Speech and/or language (difficulty speaking)
- Motor (any weakness)
- Brainstem (dizziness, double vision) and retinal (visual complaints)

# Sudden Onset of Headache (HD-11.3)

HD.HA.0011.3.A

v1.0.2023

- For sudden onset of headache including:
  - Worst, most severe headache ever experienced or thunderclap-type (example: awakening from sleep)
  - Sudden onset unilateral headache, suspected carotid or vertebral dissection or ipsilateral Horner's syndrome
  - Consideration of reversible cerebral vasoconstriction syndrome (RCVS) (typically bilateral headache)
- If any of these onset of headache features are present, the following are supported:
  - CT Head without contrast (preferred study) (CPT® 70450) **or**
  - MRI Brain without contrast (CPT® 70551) **or**
  - MRI Brain without and with contrast (CPT® 70553) **and/or**
  - CTA Head (CPT® 70496) **or** MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546)
  - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498) if arterial dissection is suspected
    - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. 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
- Repeat MRA/CTA Head and Neck imaging in 2-4 weeks if suspicion of Reversible Cerebral Vasoconstriction Syndrome (RCVS) is high
- MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498) if arterial dissection is suspected, or known and re-evaluation is needed (as directed by neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon)
- MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496, or CPT® 70498) if arterial dissection is suspected, or known and re-evaluation is needed (as directed by neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon)
- High risk scenarios including, but not exclusive to: Fibromuscular Dysplasia (FMD), Marfan Disease, MVA with whiplash, and chiropractic manipulation
- Other vascular imaging indications for headaches require additional information. See Stroke/TIA (HD-21.1), **New Headache Onset Older than Age 50 (HD-11.7)**, **Abnormal Blood Clotting (HD-11.9)**, **Pregnancy (HD-11.10)**, **Physical Exertion (HD-11.11)**, and **Systemic Infections (HD-11.13)**
- See **Intracranial Aneurysms (HD-12.1)** and **Stroke/TIA (HD-21.1)**



# Trigeminal Autonomic Cephalgias (HD-11.4)

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HD.HA.0011.4.A

v1.0.2023

- Trigeminal autonomic cephalgias includes cluster headache, short-lasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndromes; short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA) and hemicrania paroxysmal and continua.
  - May also include pituitary screening
- Cluster Headache (may also include pituitary)
- For trigeminal autonomic cephalgias and cluster headache, the following studies are indicated:
  - MRI Brain without and with contrast (preferred study) (CPT® 70553); or
  - MRI Brain without contrast (CPT® 70551)
- See **Facial Pain/Trigeminal Neuralgia (HD-10.1)**

# Skull Base, Orbit, Periorbital or Oromaxillary (HD-11.5)

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HD.HA.0011.5.A

v1.0.2023

- Skull base, orbital, periorbital or oromaxillary<sup>1</sup> imaging is appropriate for concern of skull base tumors in individuals with head and neck cancers, other skull base abnormalities seen on previous imaging, any invasive sinus infections as well as sinus tumors or orbital tumors with intracranial extension.
- In these clinical scenarios, the following studies are indicated:
  - MRI Brain and/or Orbits without and with contrast (preferred study) (CPT® 70553 and/or CPT® 70543); **or**
  - MRI Brain and/or Orbits without contrast (CPT® 70551 and/or CPT® 70540); **or**
  - CT Head and/or Orbits without and with contrast (CPT® 70470 and/or CPT® 70482); **or**
  - CT Head and/or Orbits with contrast (CPT® 70460 and/or CPT® 70481)

# Suspected Intracranial Extension of Sinusitis or Mastoiditis (HD-11.6)

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HD.HA.0011.6.A

v1.0.2023

- For suspected intracranial extension of sinusitis or mastoiditis:
  - MRI Brain without and with contrast (CPT® 70553) See **Mastoid Disease or Ear Pain (HD-26.1)** and **Skull Base, Orbit, Periorbital or Oromaxillary (HD-11.5)**

# New Headache Onset Older than Age 50 (HD-11.7)

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HD.HA.0011.7.A

v1.0.2023

- For new onset headache in individuals older than 50 years of age:
  - MRI Brain without and with contrast (preferred study) (CPT® 70553); **or**
  - MRI Brain without contrast (CPT® 70551); **or**
  - CT Head without contrast (CPT® 70450)
  - If Giant Cell Arteritis, also known as Temporal Arteritis, is suspected, MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546)

# Cancer or Immunosuppression (HD-11.8)

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HD.HA.0011.8A

v1.0.2023

- For new headache in individuals with cancer or who are immunocompromised:
  - MRI Brain without and with contrast (preferred study) (CPT® 70553); **or**
  - MRI Brain without contrast (CPT® 70551)

# Abnormal Blood Clotting (HD-11.9)

HD.HA.0011.9A

v1.0.2023

- MRI Brain without and with contrast (CPT® 70553); **or** MRI Brain without (CPT® 70551); **or** CT Head without contrast (CPT® 70450):
  - New onset headaches in individual with hypercoagulable states or bleeding disorder including pregnancy and the immediate postpartum period
    - MRA/MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA/CTV Head (CPT® 70496) if there is concern for venous sinus thrombosis
      - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. 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
  - Individuals with potential for bleeding diathesis
    - Taking anticoagulants or two or more antiaggregants or having a medical condition that predisposes to bleeding (for example, but not limited to: thrombocytopenia, liver failure, Idiopathic Thrombocytopenic Purpura (ITP), etc.).

## Pregnancy (HD-11.10)

HD.HA.0011.10.A

v1.0.2023

- For new onset headache during pregnancy or immediate post-partum period (within 3 months after delivery):
  - MRI Brain without contrast (Gadolinium relatively contraindicated in pregnancy) (CPT® 70551), Postpartum: MRI Brain without and with contrast (CPT® 70553) if not breastfeeding, if unsure, MRI Brain without contrast (CPT® 70551)
- Important causes of secondary headache include vascular disorders, such as pre-eclampsia, reversible cerebral vasoconstriction syndrome, and cerebral venous thrombosis, as well as idiopathic intracranial hypertension<sup>1,4,5,6,7</sup>
  - MRA/MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA/CTV Head (CPT® 70496)
    - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. 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. (Gadolinium relatively contraindicated in pregnancy)
    - Vascular imaging can be performed concurrently with brain imaging
- For post LP/epidural anesthesia - See Low Pressure Headache and CSF Leak (HD-11.15)

## Physical Exertion (HD-11.11)

HD.HA.0011.11.A

v1.0.2023

- For onset of headache with Valsalva maneuver, cough, physical exertion, change in position, **or** sexual activity, but not merely a worsening of a pre-existing headache with these activities, the following procedures are supported:
  - MRI Brain without and with contrast (preferred study) (CPT® 70553); **or**
  - MRI Brain without contrast (CPT® 70551); **or**
  - CT Head without contrast (CPT® 70450); **AND/OR**
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **or**
  - CTA Head without and with contrast (CPT® 70496)
  - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498) if arterial dissection or aneurysm is suspected



# Headaches Associated with Head Trauma (HD-11.12)

HD.HA.0011.12.A

v1.0.2023

- Acute head trauma with headache, See **Head Trauma (HD-13.1)**,
- Acute headache attributed to traumatic injury to the head that developed within 7 days of injury<sup>14</sup> that does not meet criteria under **Head and Facial Trauma (HD-13)**, other subsections may apply including, but not exclusive to: **Headaches with Red Flags (HD-11.2)** and **Sudden Onset of Headache (HD-11.3)**
- New or progressively worsening headache with subacute head trauma, defined as within 7 days to three months post-trauma, with or without unexplained cognitive or neurologic deficits:<sup>14</sup>
  - CT Head without contrast (CPT® 70450); **or**
  - MRI Brain without contrast (CPT® 70551)
- Persistent headaches attributed to traumatic injury to the head persisting for longer than 3 months following the injury, with or without unexplained cognitive or neurologic deficits:<sup>14</sup>
  - MRI Brain without contrast (CPT® 70551); **or**
  - MRI Brain without and with contrast (CPT® 70553)

# Systemic Infections (HD-11.13)

HD.HA.0011.13.A

v1.0.2023

- Headaches in the setting of acute, subacute, or chronic systemic infections:
  - MRI Brain without and with contrast (preferred study) (CPT® 70553); or MRI Brain without contrast (CPT® 70551)
  - MRA/MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546)
    - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. 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
  - CT Head, contrast as requested, when MRI Brain is contraindicated. See **General Guidelines – CT Head (HD-1.4)** for additional CT Head indications.
  - CT Head without (CPT® 70450) prior to performance of Lumbar Puncture (aka spinal tap)
- See **CNS and Head Infection (HD-14.1)**

# Hydrocephalus Shunts (HD-11.14)

HD.HA.0011.14.C

v1.0.2023

- Shunted Hydrocephalus may present with headaches: thus imaging is indicated
- 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).
- For CSF flow imaging See **CSF Flow Imaging (HD-24.4)**
- For Hydrocephalus Shunts, See **General Guidelines – Other Imaging Situations (HD-1.7)** and **General Guidelines – CT Head (HD-1.4)**

## Initial Imaging Indications

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

## Repeat Imaging Indications including CSF flow shunting and Ventriculostomy

- 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 (Endoscopic third ventriculostomy) malfunction, including (but not limited to) sepsis after shunt setting adjustments, decreased level of consciousness, protracted vomiting, visual or neurologic deterioration, decline of mentation after initial improvement or new or changing pattern of seizures or as ordered by neurologist or neurosurgeon or any provider in consultation with neurologist or neurosurgeon
- 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.
- 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) for suspicion of CSF pseudocyst formation or distal shunt outlet obstruction.
- See General Guidelines – Other Imaging Situations (HD-1.7)

## Additional Rarely Used Studies

- Cisternogram (CPT® 78630) for the following:
  - Known hydrocephalus with worsening symptoms.
  - Suspected obstructive hydrocephalus.
  - Suspected normal pressure hydrocephalus with gait disturbance and either dementia or urinary incontinence.
  - CSF Leak (See Low Pressure Headache and CSF Leak (HD-11.15))
- Cerebrospinal Ventriculography (CPT® 78635) 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) for the following:
  - Suspected malfunction of ventriculoperitoneal, ventriculopleural, or ventriculovenous shunts.

### **Background and Supporting Information**

- Ventriculomegaly is the condition where ventricles are enlarged, and this may be due to 1) hydrocephalus, a condition of increased intracranial pressure (ICP) (imaging shows ventricles are disproportionately enlarged compared to sulci), or 2) brain atrophy, most commonly related to age or trauma, which is not associated with increased ICP (imaging shows ventricles and sulci are proportionately enlarged).
- Hydrocephalus is divided into obstructive/non-communicating vs. communicating types, and these usually have different etiologies and radiographic features.
- Obstructive or non-communicating hydrocephalus classically involves an intraventricular obstruction in which CSF flow over the convexities and between the ventricles is reduced, and the proximal ventricle(s) is/are dilated. This is a medical emergency.
- Communicating hydrocephalus involves extraventricular obstruction, poor absorption or overproduction of CSF. There is normal intracranial CSF flow and absence of disproportionate ventricular dilation, yet there is still a mildly increased CSF pressure. Normal pressure hydrocephalus is an example of this type.
- Distinguishing between ventriculomegaly due to brain atrophy and non-communicating hydrocephalus can be difficult with MRI Brain or CT Head alone, and modalities which visualize CSF flow may be useful such as cisternography or CT cisternography.

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# Low Pressure Headache and CSF Leak (HD-11.15)

HD.HA.0011.15.C

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- CSF leaks may occur in:
  - CSF shunt overdrainage
  - Traumatic CSF leaks
    - Thecal holes and rents from lumbar punctures and epidural catheterizations
    - Spinal and cranial surgeries including skull base and some sinus surgeries
    - Proximal brachial plexus and nerve root avulsion injuries
  - Spontaneous leaks may occur in, but not exclusive to:
    - Pre-existing weakness of the dural sac including:
      - Disorders of connective tissue matrix including Marfan syndrome, Marfanoid features
      - Joint hypermobility
    - Trivial trauma in the setting of preexisting dural weakness
    - Spondylotic spurs, herniated discs
- Evaluation of suspected CSF leak (rhinorrhea/otorrhea) or refractory post-lumbar puncture/low pressure headache:
  - MRI Brain without and with contrast (CPT® 70553) and
  - MRI Cervical, Thoracic and Lumbar Spine, which according to facility protocols can be completed without contrast (CPT® 72141, CPT® 72146, and CPT® 72148), with and without contrast (CPT® 72156, CPT® 72157, and CPT® 72158) or with contrast only (CPT® 72142, CPT® 72147, and CPT® 72149) or CT myelography (CT Cervical, Thoracic, and Lumbar Spine with contrast [CPT® 72126, CPT® 72129, CPT® 72132])
- CT Head without contrast (CPT® 70450), CT Maxillofacial without contrast (CPT® 70486) or CT Temporal Bone without contrast (CPT® 70480) if concern for CSF rhinorrhea
- For CSF leak detection, CSF Leakage Detection (CPT® 78650) and/or Cisternogram (CPT 78630) for the following: [See Facial Trauma (HD-13.2)]
  - Evaluation of CSF rhinorrhea or otorrhea
  - Refractory headache post-lumbar puncture
- Additional Cisternogram (CPT® 78630) indications:
  - Known hydrocephalus with worsening symptoms (for example headache)
  - Suspected obstructive hydrocephalus
- Individuals with a Shunt, See Hydrocephalus Shunts (HD-11.14)

# Cervicogenic Headaches including Occipital Neuritis/Neuralgia (HD-11.16)

HD.HA.0011.16A

v1.0.2023

- Cervicogenic Headache
  - Headache caused by a disorder of the cervical spine, usually accompanied by neck pain or other signs and symptoms of cervical disease. Typical findings include reduced cervical range of motion, side-locked pain, and symptoms exacerbated by provocative maneuvers such as head movement or digital pressure.
- Occipital Neuralgia/Neuritis - Occipital neuralgia is classified unilateral or bilateral paroxysmal, shooting or stabbing pain in the posterior part of the scalp, in the distribution(s) of the greater, lesser and/or third occipital nerves, sometimes accompanied by diminished sensation or dysaesthesia in the affected area and commonly associated with tenderness over the involved nerve(s).
  - Pain has at least two of the following three characteristics:
    - Recurring in paroxysmal attacks lasting from a few seconds to minutes
    - Severe in intensity
    - Shooting, stabbing or sharp in quality
  - Pain is associated with both of the following:
    - Dysaesthesia and/or allodynia apparent during innocuous stimulation of the scalp and/or hair
    - Either or both of the following:
      - Tenderness over the affected nerve branches
      - Trigger points at the emergence of the greater occipital nerve or in the distribution of C2
  - Pain is eased temporarily by local anaesthetic block of the affected nerve(s)
- MRI Cervical Spine without contrast (CPT® 72141) or CT Cervical Spine without contrast (CPT® 72125)
  - Imaging should follow **Neck (Cervical Spine) Pain Without/With Neurological Features (Including Stenosis) (SP-3.1)** and **Neck (Cervical Spine) Trauma (SP-3.2)** in the Spine Imaging Guidelines including 6 weeks of physician directed care within the last 3 months with re-evaluation.
    - Exemptions to the 6 weeks of conservative care include:
      - In cases of Cervical Spine injury, results of plain x-rays of the cervical spine and a 6 week trial of provider-directed treatment and clinical reevaluation are not required for individuals with a high risk mechanism of cervical spine injury within the last 3 months (See **Neck (Cervical Spine)**)

**Trauma (SP-3.2)** in the Spine Imaging Guidelines in the Spine Imaging Guidelines)

- **Red Flag Indications (SP-1.2)** in the Spine Imaging Guidelines
  - For ANY of the following:
    - Bony abnormalities: Atlanto-axial dislocations/instability (including but not limited to: Down's syndrome, Ehlers-Danlos and Marfan syndromes and rheumatoid arthritis), platybasia, osteomas, callous formation of the posterior C1/2 arches
    - Posterior fossa lesions, Chiari malformations, demyelinating disease
    - Myelopathy/myelitis (See **Myelopathy (SP-7.1)** in the Spine Imaging Guidelines)
- Brain imaging should follow applicable sections in **Headache (HD-11)**

# Advanced Imaging Indications Related to Migraines (HD-11.17)

HD.HA.0011.17A

v1.0.2023

- Advanced imaging of the head is NOT indicated for newly diagnosed migraine with a normal neurological exam or chronic stable migraine with no neurological deficit and/or no red flags (See **Headaches with Red Flags (HD-11.2)**). See below for advanced imaging indications related to migraines.
- MRI Brain without (CPT® 70551) preferred or MRI Brain with and without (CPT® 70553) or CT Head without (CPT® 70450) for the following:
  - New migraine with age ≥50 (See **New Headache Onset Older than Age 50 (HD-11.7)**)
  - Change in frequency or severity of migraine (See **Headaches with Red Flags (HD-11.2)**)
  - Unusual, prolonged or persistent aura (greater than 60 minutes) (See **Background and Supporting Information**)
  - Worst migraine
  - Hemiplegic migraine
    - Migraine with any motor weakness.
  - Migrainous accompaniments
    - Passing neurological symptoms that can affect vision, speech, movement, and behavior-“mimic stroke”
  - Migraine aura without headache
    - Migraine with an aura in which the aura is neither accompanied nor followed by a headache within 60 minutes.
  - Side-locked migraine (unilateral)
    - Unilateral hemicranial pain – includes primary and secondary causes.
      - New daily persistent headache (new daily headache present greater than three months)
      - Trigeminal autonomic cephalgias includes cluster headache short-lasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndromes; short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA) and hemicrania paroxysmal and continua are covered in **Trigeminal Autonomic Cephalgias (HD-11.4)**
  - Post-traumatic migraine
    - See **Head Trauma (HD-13.1)** and **Headaches Associated with Head Trauma (HD-11.12)**



***Background and Supporting Information***

- Aura symptoms may accompany or precede a headache within 60 minutes and may include, but are not exclusive to, the following symptoms:
  - Visual (flashing lights, loss of vision)
  - Sensory (paresthesia)
  - Speech and/or language (difficulty speaking)
  - Motor (any weakness)
  - Brainstem (dizziness, double vision) and retinal (visual complaints)

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# Aneurysm and AVM (HD-12)

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# Intracranial Aneurysms (HD-12.1)

HD.AN.0012.1.C

v1.0.2023

- CTA Head (CPT® 70496) or MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) in ANY of the following clinical scenarios:
  - Symptoms or signs of cerebral aneurysm, including:
    - "Thunderclap headache" See Sudden Onset of Headache (HD-11.3)
    - Third nerve palsy with pupillary involvement (pupil-sparing third nerve palsies are not caused by external compression)
    - Suspicion of aneurysm bleed [CT Head or MRI Brain or CSF exam showing evidence of subarachnoid hemorrhage (SAH) or intracerebral hemorrhage]
    - Abnormal CT Head or MRI Brain suggesting possible aneurysm
  - Screening for High Risk Populations as defined by the following criteria (screening usually begins at age 20 unless unusual circumstances as aneurysms are uncommon in children and adolescents):
    - Positive Family History: Two or more first degree relatives (parent, sibling, or child) with history of cerebral aneurysm or SAH: screening every 5 years beginning at age 20
      - One first degree relative (parent, sibling, or child) with history of cerebral aneurysm or SAH can have one screening study but risks and benefits should be discussed with individual
    - Autosomal dominant polycystic kidney disease
    - Coarctation of the aorta or bicuspid aortic valve
    - Neurofibromatosis Type 1
    - Type 4 (Vascular) Ehlers-Danlos Syndrome
    - Marfan Syndrome
    - Loeys-Dietz Syndrome
    - Microcephalic osteodysplastic primordial dwarfism
    - Presence of an azygos anterior cerebral artery
    - Diagnosis of fibromuscular dysplasia (one screening study after confirmed diagnosis)
    - Pseudoxanthoma elasticum
    - Hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu Syndrome)
    - Alpha-1-antitrypsin deficiency
    - Pheochromocytoma
    - Klinefelter syndrome
    - Tuberous sclerosis
    - Noonan syndrome
    - Alpha-glucosidase deficiency

- Klippel-Trenaunay-Weber Syndrome
- Kawasaki disease
- CTA Head (CPT® 70496) to confirm questionable or equivocal findings on an initial MRA Head.
- For suspected or confirmed cerebral aneurysm, ruptured or unruptured, for initial evaluation, treatment, intervention or follow up, 3D rendering (CPT® 76377) with cervicocerebral angiography/arteriography and/or cerebral angiography.<sup>22</sup> (See **General Guidelines - Other Imaging Situations (HD-1.7)**)
- Follow up of known cerebral aneurysm:
  - The optimal interval and duration for radiologic follow-up has not been determined. Radiographic follow-up with MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA (CPT® 70496) for unruptured or treated intracranial aneurysm upon request by the neurosurgeon or team managing the unruptured intracranial aneurysm<sup>22</sup>
- MRI Brain without contrast (CPT® 70551) or with and without (CPT® 70553) in the following scenarios:
  - If there are new signs, symptoms or clinical findings
  - To evaluate and treat a giant aneurysm (>2.5 cm)
  - Posterior fossa aneurysms
  - Thrombosed or partially thrombosed aneurysms
  - To evaluate the relationship of the aneurysm to the dura
  - To evaluate for the presence of calcification
  - Other surveillance criteria as per the neurosurgeon or team managing the aneurysm repair
- Head imaging (CT Head or MRI Brain contrast as requested) to assess for subacute complications, (i.e. vasospasm, delayed cerebral ischemia and hydrocephalus), beginning days to weeks arising from a subarachnoid hemorrhage and aneurysm treatment upon request from the neurosurgeon and team managing the episode.
- MRI Spinal (Cervical, Thoracic, Lumbar (without and with contrast) [CPT® 72156, CPT® 72157, CPT® 72158]) is appropriate to evaluate individuals with SAH and negative studies for brain aneurysm in whom spinal abnormalities (i.e. AVM) may be suspected as the cause of hemorrhage.
- MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498) are not supported for screening and for follow-up on surgically treated cerebral aneurysms, except if they are located in the vertebral-basilar system.
- Initial catheter arteriography can be negative in 10-20% of cases of subarachnoid hemorrhage (SAH). CTA Head (CPT® 70496) and/or MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) if these had not been initially performed. If initial catheter angiography is negative, repeat imaging is appropriate.<sup>22</sup>

- If an intracranial etiology for SAH has not been found, CTA (CPT® 70498) or MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) to evaluate for less common causes of SAH.
- High risk scenarios for vascular dissection include, but are not limited to: Fibromuscular dysplasia (FMD), Marfan Disease, MVA with whiplash, and chiropractic manipulation.
  - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498) if arterial dissection is suspected, or known and re-evaluation is needed (as directed by neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon)
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496, CPT® 70498) if arterial dissection is suspected, or known and re-evaluation is needed (as directed by neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon)
- .
- Other indications for headaches require additional information. See **Stroke/TIA (HD-21.1)**, **Sudden Onset of Headache (HD-11.3)**, **New Headache Onset Older than Age 50 (HD-11.7)**, **Abnormal Blood Clotting (HD-11.9)**, **Pregnancy (HD-11.10)**, **Physical Exertion (HD-11.11)**, and **Systemic Infections (HD-11.13)**

# Arteriovenous Malformations (AVMs) and Related Lesions (HD-12.2)

HD.AN.0012.2.C

v1.0.2023

- MRI Brain without and with contrast (CPT® 70553) or without contrast (CPT® 70551) in the following clinical scenarios:
  - AVM is suspected based on a history of SAH
  - Screening for:
    - Hereditary hemorrhagic telangiectasia syndrome (Osler Weber Rendu)
    - Familial cavernous malformation: Screening should include MRI Brain without or without and with contrast (with gradient echo images)
  - 3D imaging (CPT® 76377) with MRI Brain is supported
- CTA Head (CPT® 70496) or MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) if screening MRI Brain is positive.
- If a cerebral cavernous malformation is diagnosed in the brainstem or presented with a focal neurological deficit (ex. seizure) or intracranial hemorrhage, repeated vessel and head imaging (MRI Brain without and with contrast (CPT® 70553) or without contrast (CPT® 70551), AND/OR MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496)) is supported.
- MRI Brain without and with contrast (CPT® 70553) or without contrast (CPT® 70551), AND/OR MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496) for repeat advanced imaging when requested by a specialist or any provider in consultation with a specialist.
- MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70496) is supported for screening or for follow-up on surgically treated AVMs, if they are located in the vertebral-basilar system.<sup>22</sup> See General Guidelines – CT and MR Angiography (CTA and MRA) (HD-1.5)
- 3D Rendering (CPT® 76377) with cerebral angiography to define the presence, location, and anatomy of intracranial and cervical vascular malformations.<sup>22</sup> See **General Guidelines - Other Imaging Situations (HD-1.7)** and Background and Supporting Information

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

- Trauma is the most common reason for subarachnoid hemorrhage. Ruptured berry aneurysm is the most common reason for non-traumatic subarachnoid hemorrhage in adults
- Small aneurysms are present in about 1% to 2% of adults, but very few ever reach a size for which bleeding is a risk (>5 mm). Small (<3 to 4 mm) unruptured aneurysms in those with no personal history of SAH have a 0.1% to 0.5% a year rate of bleeding. The risk of cerebral aneurysm with family history ranges from 2% with one first degree relative to 30% to 35% for identical twin or two parents.



The risks and benefits of screening these populations need to be considered before advanced imaging.

- AVMs most often come to clinical notice either by bleeding or by acting as a seizure focus. They are usually congenital, recognized later in life and have an initial risk of bleeding of 2% per year.
- Cerebral angiography is a form of angiography which provides images of blood vessels in and around the brain and/or neck. This is a catheter based procedure, using x-ray imaging guidance and iodine-based contrast to visualize blood vessels.

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# Head and Facial Trauma (HD-13)

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# Head Trauma (HD-13.1)

HD.TR.0013.1.A

v1.0.2023

Individuals with head trauma are at risk for facial and cervical trauma.

Subacute head trauma is defined as within 7 days to 3 months post-trauma.<sup>8</sup>

SPECT, PET, CT/MRI perfusion, DTI (diffusion tensor imaging), functional MRI, and MR spectroscopy are not considered routine clinical practice at this time.<sup>3,4,8</sup>

See **Neck (Cervical Spine) Pain Without/With Neurological Features (Including Stenosis) and Trauma (SP-3)** in the Spine Imaging Guidelines

See **General Guidelines – CT and MR Angiography (CTA and MRA) (HD-1.5)** for traumatic vascular injuries

- CT Head without contrast (CPT® 70450) is the primary imaging in individuals with acute head trauma and ANY of the following modified Canadian CT Head Rule/New Orleans Criteria for those with loss of consciousness, amnesia or disorientation accompanying blunt head trauma within 24 hours.<sup>1,8,10</sup>
- CT Head is indicated when one of the following is present:<sup>8</sup>
  - Taking one anticoagulant or two antiaggregants, (e.g., aspirin and Plavix)
  - Known platelet or clotting disorder
  - Glasgow coma scale (GCS) score of less than 15 at 2 hours following injury
  - >30 minutes of amnesia before impact
  - Regardless of documented or stated head impact, any "dangerous mechanism of injury" either direct or indirect, including, but not exclusive to:
    - Fall greater than 5 steps down stairs
    - Fall from height greater than 3 feet
    - Any pedestrian motor vehicle accident
    - High impact motor vehicle accident
  - Suspected open skull fracture
  - Signs of basilar skull fracture (Battle's sign, Raccoon eyes, CSF rhinorrhea, cranial nerve palsy, hemotympanum, acute hearing loss)
  - Vomiting
  - Individual >60 years old
  - Alcohol or drug intoxication
  - Visible trauma above clavicles
  - Deficits in short term memory, altered level of alertness, abnormal behavior or focal neurological deficit

- Seizure
- Headache [See **Headache Associated with Head Trauma (HD-11.12)**]
- MRI Brain without contrast (CPT® 70551) or CT Head without (CPT® 70450) are appropriate for the initial imaging of individuals with subacute or chronic head trauma and unexplained cognitive or neurologic deficits.<sup>8</sup>
- MRI Brain without and with contrast (CPT® 70553) if post-traumatic infection is suspected
- Follow-up imaging, MRI or CT, for known subdural hematomas, intracerebral hemorrhage, or contusions can be done at the discretion of ordering specialist or any provider in consultation with a specialist. Short term follow-up imaging of acute TBI without neurologic deterioration, noncontrast CT is the most appropriate imaging study, but only in individuals with risk factors (such as subfrontal/temporal intraparenchymal contusions, anticoagulation, >65 years or intracranial hemorrhage). MRI as a complementary study when neurological findings or symptoms are not sufficiently explained by CT or in subacute and chronic TBI for new, persistent, or slowly progressive symptoms.<sup>8</sup>
- Follow up imaging for known subdural hematomas, intracerebral hemorrhages or contusions can be completed with one of the following:
  - MRI Brain without and with contrast (CPT® 70553) or
  - MRI Brain without contrast (CPT® 70551) or
  - CT Head without and with contrast (CPT® 70470) or
  - CT Head without contrast (CPT® 70450)
- For suspected intracranial venous or arterial injury, CTA/CTV Head (CPT® 70496) and MRA/MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546)
  - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. 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. See **General Guidelines - CT and MR Angiography (CTA and MRA) (HD-1.5)**

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

Recent studies have shown that Diffusion tensor MRI tractography may be more sensitive in demonstrating abnormalities such as axonal injury in closed head injury than conventional MRI, but these techniques are best described presently as research tools and their use in clinical practice is not determined.<sup>3,4,8</sup>

Decisions regarding return to normal activities, including sports, are made based on the clinical status of the individual and repeat imaging is unnecessary.

In cases of post-traumatic infection, contrast-enhanced MRI or CT may be helpful

## Facial Trauma (HD-13.2)

HD.TR.0013.2.C

v1.0.2023

- CT Maxillofacial without contrast (CPT® 70486) indicated for any concern regarding significant injury to facial structures including but not limited to:
  - Concern for orbital, maxillary, or mandibular fractures
  - Trauma with associated symptoms of anosmia, hearing, vision or speech changes, vertigo, facial numbness
  - Physical exam findings of CSF rhinorrhea (suspected post traumatic CSF leak), malocclusion, severe focal facial tenderness, focal loss of facial sensation
- CT Orbits/Temporal Bone without contrast (CPT® 70480):
  - Concern for orbital injury or orbital wall fracture
  - Symptoms of diplopia, blurred vision, vision loss
  - Physical exam findings of enophthalmos, entrapment of extraocular muscle(s)
  - Suspicion for temporal bone fracture
  - Suspected post-traumatic (CSF leak)

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**Note** Initial x-rays are not required before advanced imaging for the above indications

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- CT Head cisternography with contrast if CT Maxillofacial or Temporal bone is inconclusive<sup>8</sup> (See **Low Pressure Headache and CSF Leak (HD-11.15)** )

### Background and Supporting Information

Imaging is not necessary in the evaluation of simple nasal fractures if tenderness and swelling is limited to the nasal bridge, the individual can breathe through each naris, and there is no septal hematoma.

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**v1.0.2023**

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# CNS and Head Infection/ Neuro-COVID-19 (HD- 14)

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# CNS and Head Infection (HD-14.1)

HD.HI.0014.1.A

v1.0.2023

- Signs of intracranial infection include: 1) headaches, seizures, meningeal signs (neck stiffness), or new focal neurological deficits in a setting of fever or elevated white blood cell count (WBC); 2) known infection elsewhere; 3) or immunosuppression. ONE of the following studies for suspected intracranial infection if any of these signs of 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
    - See **General Guidelines – CT Head (HD-1.4)** regarding additional indications for CT Head.
- See **Skull Base Osteomyelitis (SBO) (HD-20.1)** , **Sinus and Facial Imaging (HD-29.1)** , **Dental/Periodontal/Maxillofacial Imaging (HD-30.2)** , and **Eye Disorders and Visual Loss (HD-32.1)**
- FDG Brain PET (CPT® 78608) 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, if appropriate.

# Neuro-COVID-19 and Sars-CoV-2 Vaccines (HD-14.2)

HD.HI.0014.2.A

v1.0.2023

- The findings observed in the central nervous system in the acute-phase of COVID-19 may extend into a prolonged symptomatic phase of Neuro-COVID in long haulers with chronic COVID syndrome. Symptoms may include, but are not inclusive to: “brain fog”, dizziness, inability to concentrate, psychiatric symptoms, and confusion.<sup>8,9</sup>
- Acute-phase neurologic manifestations of COVID-19 include: headache, dizziness, taste and smell dysfunction, impaired consciousness (described as confusion or agitation), cerebrovascular events (ischemic stroke, cerebral venous sinus thrombosis, cerebral hemorrhage), seizures, meningoencephalitis, and immune-mediated neurologic diseases (Guillan-Barre syndrome, Miller-Fisher syndrome, polyneuritis cranialis, transverse myelitis).<sup>10,11,15,16,20</sup>
- MRI Brain without contrast (CPT® 70551) or MRI Brain with and without contrast (CPT® 70553), or CT Head without contrast (CPT® 70450) for the evaluation of acute or chronic Neuro-COVID-19 syndrome. See **Stroke/TIA (HD-21.1)** for vascular imaging. CT Head without and with contrast (CPT® 70470) if there is a contraindication to MRI. Cervical and/or Thoracic spinal cord imaging (MRI Cervical and/or Thoracic Spine without and with contrast (CPT® 72156 and CPT® 72157) if suspected transverse myelitis.
- Repeat imaging considered on a case-by-case basis for a change in neurological symptoms or signs on the neurological exam and/or change in the treatment.
- Neurologic adverse reactions in those receiving SARS-CoV-2 vaccines, including mRNA vaccines (Pfizer, Moderna), have been reported, and include, although not limited to: headache, Guillan-Barre syndrome, transverse myelitis, facial nerve palsy, small fiber neuropathy, autoimmune encephalitis, reversible cerebral vasoconstriction syndrome, multiple sclerosis, neuromyelitis optica, intracerebral bleeding, cerebral venous sinus thrombosis, hypophysitis, epilepsy, encephalopathy, and acute disseminated encephalomyelitis.<sup>13,14,17,18,19,21</sup>
- MRI Brain without contrast (CPT® 70551) or MRI Brain with and without contrast (CPT® 70553), or CT Head without contrast (CPT® 70450) and/or MRI Cervical and/or Thoracic Spine without and with contrast (CPT® 72156 and CPT® 72157), are supported for evaluation of suspected neurologic adverse reactions after SARS-CoV-2 vaccination. CT Head without and with contrast (CPT® 70470) if there is a contraindication to MRI. See **Stroke/TIA (HD-21.1)** for vascular imaging.
- According to the Centers for Disease Control (CDC), there is a plausible causal relationship between the Johnson & Johnson/Janssen COVID-19 vaccine and a rare and serious adverse event, blood clots with low platelets (thrombosis with thrombocytopenia syndrome TTS).<sup>12</sup> It occurs at a rate of 7 per 1 million vaccinated females between 18-49 years old. For females 50 years and older and males of all

ages, this adverse event is even more rare. Imaging listed above is appropriate if this condition is suspected. See **Abnormal Blood Clotting (HD-11.9)**

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# Movement Disorders (HD-15)

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# Movement Disorders (HD-15.1)

HD.MD.0015.1.C

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- The majority of movement disorders are diagnosed based on a clinical diagnosis and do not require imaging. These include:
  - Typical Parkinson's Disease
  - Essential Tremor or tremors of anxiety or weakness
  - Restless Leg Syndrome
  - Tics or spasms which can be duplicated at will
- MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) in the following clinical scenarios:
  - Atypical Parkinsonism (Parkinson's Plus Syndromes – See Background and Supporting Information) because of unusual clinical features (for example, persistent unilateral signs and symptoms, young onset under age of 50, rapid progression), incomplete or uncertain medication responsiveness, or clinical diagnostic uncertainty.
  - Suspected Huntington Disease
- Evaluation for surgical treatment of Essential Tremor, Parkinson's disease, and/or Spasmodic Torticollis/Dystonia See **Torticollis and Dystonia (Neck-10.2)** in the Neck Imaging Guidelines
  - Deep Brain Stimulation (DBS) therapy
    - MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) and unlisted CT procedure code (CPT® 76497)
  - MR guided Focused Ultrasound:
    - CT Head without contrast (CPT® 70450) to evaluate bone density and MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553)
  - Repeat imaging studies, MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) and CT Head without contrast (CPT® 70450), when ordered by a specialist or any provider in consultation with a specialist if greater than 6 months old **and/or** for new symptoms/signs
  - Post op imaging when ordered by a specialist or any provider in consultation with a specialist for either procedure
- MRI Brain with and without (CPT® 70553) for initial imaging for suspected motor neuron disease. See **Motor Neuron Disease/Amyotrophic Lateral Sclerosis (ALS) (PND-8.1)** in the Peripheral Nerve Disorders Imaging Guidelines
- Dementia associated with movement disorder, See **Lewy Body Dementia (LBD) – SPECT Brain Scan (HD-8.3)**

## Background and Supporting Information

- There is little evidence to support the use of MRA/CTA and PET in the evaluation of movement disorders.
- Parkinson's Plus Syndromes are a group of disorders characterized by atypical parkinsonism. They are NOT Parkinson's disease. They represent different neurodegenerative diseases with features of PD, and may be confused with PD. These syndromes include, but are not limited to:
  - Multiple system atrophy: orthostatic hypotension (dysautonomia), dysphonia, dysarthria
  - Progressive Supranuclear Palsy: balance difficulties, vertical gaze paresis
  - Corticobasal Syndrome: dysphasia, apraxia, myoclonus, alien-limb phenomenon
- These are distinct entities. Care must be taken to determine if there are unusual features present that will suggest atypical parkinsonian syndrome.
- Dementia with Lewy bodies (DLB): dementia prior to movement disorder. See **Lewy Body Dementia (LBD) - SPECT Brain Scan (HD-8.3)**.

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# Multiple Sclerosis (MS) and Related Conditions (HD-16)

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# Multiple Sclerosis (MS) (HD-16.1)

HD.MS.0016.1.C

v1.0.2023

- MRI Brain without and with contrast (CPT® 70553) is indicated in these clinical scenarios requires:
  - Clinical suspicion based on recurrent episodes of variable neurological signs and symptoms or clinically isolated syndromes
  - MRI Brain without and with contrast (CPT® 70553) is the preferred study for initial imaging to establish the diagnosis of MS. However, MRI Brain without contrast (CPT® 70551) is indicated if there is a contraindication to gadolinium.
  - 3T MRI is preferable to 1.5 T MRI if available
  - Baseline exclusion of appropriate alternative conditions that can mimic MS
- Repeat MRI Brain without contrast (CPT® 70551) (preferred study) or MRI Brain without and with contrast (CPT® 70553) is supported for an individual with an established diagnosis of MS in the following scenarios:
  - New episode of neurological deficit or re-evaluation of the diagnosis
  - Every 3-6 months until stable on disease modifying therapy (DMT)
  - Re-establish baseline when instituting or changing disease modifying therapy (typically 6 months after the start of a new therapy)
  - **EVERY 6 MONTHS** for individuals treated with disease modifying therapy associated with either risk of progressive multifocal leukoencephalopathy (PML) and/or other CNS opportunistic infections
  - MRI every 3 months for high risk individuals that are JC virus antibody positive and treated 18 or more months with natalizumab (Tysabri®)
  - **Annual** MRI surveillance for stable individuals that are not on disease modifying therapy or are treated with beta interferon or glatiramer acetate medications (see Background and Supporting Information for list of medications)
  - Symptoms suggestive of Progressive Multifocal Leukoencephalopathy (PML) during treatment with natalizumab (Tysabri®) therapy or medications with similar risk
- MRI Cervical Spine without or MRI Cervical Spine without and with contrast (CPT® 72141 or CPT® 72156) and/or MRI Thoracic Spine without or MRI Thoracic Spine without and with contrast (CPT® 72146 or CPT® 72157) is indicated in these clinical scenarios:
  - Clinical suspicion of demyelinating disease and/or establishing baseline imaging at diagnosis
  - Annual surveillance or new signs/symptoms concerning for spinal cord involvement (worsening weakness, numbness/tingling, spasticity, Lhermitte's sign, sensory level, or change in bladder and/or bowel functioning)

- MRI Orbit without contrast (CPT® 70540) or without and with contrast (CPT® 70543) if optic neuritis is suspected with supporting documentation, in addition to the above scenario
- MRI Brain with contrast (CPT® 70552) is supported within 2 weeks of previous non-contrast study, if a non-contrast study shows incidental evidence of possible demyelinating disease, as the presence of enhancing lesions may be helpful in confirming the diagnosis
  - MRI Brain with and without contrast (CPT® 70553) is appropriate, if non-contrast study was performed more than 2 weeks prior to repeat imaging.
- MRI Lumbar Spine is not needed since Cervical and Thoracic studies will usually visualize the entire spinal cord. If the clinical concern is for lumbosacral radiculopathy, See **Lower Extremity Pain with Neurological Features (Radiculopathy, Radiculitis, or Plexopathy and Neuropathy) with or without Low Back (Lumbar Spine) Pain (SP-6.1)** in the Spine Imaging Guidelines.
- Family members need not be screened, unless they exhibit suspicious signs or symptoms suggestive of MS.
- 3D FLAIR sequences are useful in improving lesion detection for the diagnosis and monitoring of multiple sclerosis. 3D FLAIR sequences do not require an additional CPT® for 3D rendering (CPT® 76377).<sup>1</sup>
- Volumetric and quantitative MRI measures that may include 3D analysis or rendering (CPT® 76377) require further validation before it can be determined to be clinically useful. Its use is limited to research studies and it is otherwise considered to be investigational and experimental in routine clinical practice.<sup>1</sup> Advanced imaging studies, or other procedures, are considered investigational and experimental if there is a paucity of supporting evidence; if the evidence has not matured to exhibit improved health parameters or; the advanced imaging study/procedure lacks a collective opinion of support.

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

- Multiple Sclerosis is common and variable with more females affected and at a younger age than males. MS tends to be relapsing-remitting (improves between episodes), relapsing-progressive (worsens with attacks) and chronic progressive (gradual and steady neurological decline).
- Ataxia, diplopia, optic neuritis and partial transverse myelitis are common symptoms that occur with multiple sclerosis.
- Sagittal MRI Spinal Cord with phased array detector coil (CPT® 72156 or CPT® 72157) is an alternative spinal imaging.
- Interferon beta medications include (but not limited to): Avonex®, Betaseron®, Extavia®, Plegridy®, Rebif®
- Glatiramer acetate medications include (but not limited to): Copaxone®, Glatopa®
- Medications with high risks of PML as Tysabri® (natalizumab) and/or other CNS opportunistic infections (i.e. herpes encephalitis) include (but not limited to): Tecfidera® (dimethyl fumarate), Gilenya® (fingolimod), Ocrevus® (ocrelizumab), Kesimpta® (ofatumumab), Mavenclad® (cladribine), Vumerity® (diroximel fumarate),

Zeposia® (ozanimod), Lemtrada® (alemtuzumab), Bafiertam® (monomethyl fumarate), Rituxan® (rituximab)



# Neuromyelitis Optica and NMO Spectrum Disorders (HD-16.2)

HD.MS.0016.2.C

v1.0.2023

- Neuromyelitis optica (NMO, Devic's disease) is a chronic inflammatory autoimmune disease that involves the optic nerve, spinal cord and brain. Diagnosis is based on the clinical presentation, MRI findings, and presence of auto-antibodies.
- MRI Brain without and with contrast (CPT® 70553), and/or MRI Cervical and Thoracic Spine without and with contrast (CPT® 72156, CPT® 72157) are recommended for the **initial** imaging studies. However, if there is an allergy or significant concerns to gadolinium, (GFR is compromised), then unenhanced studies are supported
- MRI Orbit without and with contrast (CPT® 70543) preferred, or MRI Orbit without contrast (CPT® 70540) if optic neuritis is suspected with supporting documentation, in addition to the above scenario.
- Repeat imaging for MRI Brain without contrast (CPT® 70551) or with and without contrast (CPT® 70553) and/or MRI Cervical Spine without contrast (CPT® 72141) or without and with contrast (CPT® 72156) and/or MRI Thoracic Spine without contrast (CPT® 72146) or without and with contrast (CPT® 72157) for an established diagnosis of Neuromyelitis Optica spectrum disorders, in the following scenarios:
  - New symptoms or signs of neurological deficit or re-evaluation of the diagnosis
  - Annual Surveillance
- MRI Orbit without contrast (CPT® 70540) or without and with contrast (CPT® 70543) indicated for new vision or worsening vision complaints concerning for optic neuritis or for follow-up of known optic neuritis
  - When requested by a neurology specialist or any provider in consultation with a neurology specialist in the treatment of this condition
  - Non-contrast studies when requested by a neurology specialist or any provider in consultation with a neurology specialist
- Repeat imaging MRI Brain without contrast or with and without contrast (CPT® 70551 or CPT® 70553) for follow up when requested by a neurology specialist or any provider in consultation with a neurology specialist
- 3D analysis of the temporal lobes and hippocampus (also known as volumetric analysis or Neuro Quant) (CPT® 76377) lacks sufficient specificity and sensitivity to be clinically useful in the evaluation or follow up of individual with multiple sclerosis. Its use is limited to research studies and it is otherwise considered to be investigational and experimental in routine clinical practice. Certain imaging studies are considered investigational by various payors, and their coverage policies take precedence over eviCore's guidelines. Advanced imaging studies, or other procedures, are considered investigational and experimental if there is a paucity of supporting evidence; if the evidence has not matured to exhibit improved health

parameters or; the advanced imaging study/procedure lacks a collective opinion of support.

#### Background and Supporting Information

- Neuromyelitis Spectrum disorder can be associated with optic neuritis (frequently bilateral involvement with severe vision loss), long segment transverse myelitis, brainstem syndromes, and area postrema syndrome: otherwise unexplained episode of hiccups or nausea and vomiting. Rarely, paraneoplastic syndromes occur with NMO spectrum disorder
- Medications used for the treatment of NMO spectrum disorders include (but are not limited to) azathioprine, Encoring® (satralizumab), mycophenolate, Soliris® (eculizumab), and Uplizna® (inebilizumab). Possible adverse reactions associated with treatment include risk of PML and meningococcal infections.

## Anti-MOG Syndromes (HD-16.3)

HD.MS.0016.3.A

v1.0.2023

- MOG (myelin oligodendrocyte glycoprotein)-IgG disorders are CNS inflammatory diseases, distinct from multiple sclerosis and NMO-spectrum disorders. MOG-IgG disorders can be associated with optic neuritis, transverse myelitis, brainstem encephalitis, encephalitis with seizures and acute disseminated encephalomyelitis (ADEM, occurs mainly in children but can occur in adults). Rarely individuals can present with intractable nausea or hiccups. There may be involvement of the conus therefore, lumbar spine imaging is appropriate. Diagnosis is based on the clinical presentation, MRI findings and presence of autoantibodies.
- MRI Brain without and with contrast (CPT® 70553) and/or MRI Cervical and Thoracic Spine without and with contrast (CPT® 72156 and CPT® 72157) are recommended for the **initial** imaging studies. However, if there is an allergy or significant concerns to gadolinium, (GFR is compromised), then unenhanced studies are supported.
- Due to involvement of the conus that can occur with this syndrome, MRI Lumbar Spine without and with (CPT® 72158) or MRI Lumbar Spine without contrast (CPT® 72148) is supported.
- MRI Orbit without and with contrast (CPT® 70543) preferred, or MRI Orbit without contrast (CPT® 70540) if optic neuritis is suspected with supporting documentation, in addition to the above scenario
- Repeat imaging MRI Brain without contrast (CPT® 70551) or with and without contrast (CPT® 70553) and/or Spine imaging, MRI Cervical Spine without contrast (CPT® 72141) preferred, or without and with contrast (CPT® 72156) and MRI Thoracic Spine without contrast (CPT® 72146) preferred, or without and with contrast (CPT® 72157) for an established diagnosis MOG IgG disorder for the following scenarios:
  - New symptoms or signs in an individual with known anti-MOG syndrome (these may include loss or blurred vision, loss of color vision, weakness of a limb or limbs, including paraparesis or complete paralysis, loss of sensation, loss of bladder or bowel control, profound bladder retention, and seizures).
  - Evaluation for recurrent disease should occur with any new or progressive neurologic signs or symptoms
  - Annual yearly surveillance
  - MRI Orbit without and with contrast (CPT® 70543) preferred, or MRI Orbit without contrast (CPT® 70540) for new or worsening visual complaints concerning for optic neuritis

# Transverse Myelitis (HD-16.4)

HD.MS.0016.4.A

v1.0.2023

- Clinical symptoms may include one or more of the following: bilateral limb weakness and/or weakness involving the upper and lower extremity on the same side, numbness and/or paresthesias/dyesthesias, urinary incontinence/retention, worsening constipation and/or bowel urgency/incontinence and/or erectile dysfunction.
- Examination findings may include loss of manual dexterity, weakness of extensor muscles in an upper extremity and/or weakness of flexor muscles in the lower extremity and/or sensory or motor symptoms involving the limbs on the same side of the body, spasticity, sensory level, Lhermitte's sign, hyperreflexia and/or upgoing toes (positive Babinski), Hoffman's sign, clonus, and/or ataxia.
- There may be involvement of the conus, therefore, initial imaging of the lumbar spine is appropriate
- MRI Brain without and with contrast (CPT® 70553) and/or MRI Cervical, Thoracic, and Lumbar Spine without and with contrast (CPT® 72156, CPT® 72157, and CPT® 72158) are recommended for the initial imaging studies. If there is a contraindication to gadolinium-based contrast, then unenhanced studies (MRI Brain without contrast (CPT® 70551) and/or MRI Cervical, Thoracic, and Lumbar Spine without contrast (CPT® 72141, CPT® 72146, and CPT® 72148)<sup>32,33</sup>
- Repeat imaging for MRI Brain without contrast, preferred, or with and without contrast (CPT® 70551 or CPT® 70553) and/or spine imaging, MRI Cervical Spine without contrast, preferred, or without and with contrast (CPT® 72141 and CPT® 72156) and MRI Thoracic Spine without contrast, preferred, or without and with contrast (CPT® 72146 and CPT® 72157) are supported in the following scenarios:
  - Evaluation for recurrent disease should occur with any new neurologic signs or symptoms<sup>30</sup>
  - Annual surveillance<sup>30</sup>

## Background and Supporting Information

- Transverse myelitis is an inflammatory disorder of the spinal cord.

Potential etiologies include, but not limited to:

- Autoimmune central nervous system inflammatory disease
  - First event of multiple sclerosis
  - Neuromyelitis optica (NMO)
  - MOG (Myelin Oligodendrocyte Glycoprotein) antibody disorder
- Associated with connective tissue autoimmune diseases.

- Systemic lupus erythematosus
- Systemic sclerosis
- Rheumatoid arthritis
- Sjögren's syndrome
- Neuro-sarcoidosis
- Post-infectious or post-vaccine neurological syndrome<sup>32</sup>

## References (HD-16)

v1.0.2023

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# Papilledema/ Pseudotumor Cerebri (HD-17)

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# Papilledema/Pseudotumor Cerebri (HD-17.1)

HD.PP.0017.1.A

v1.0.2023

- See **Eye Disorders and Visual Loss (HD-32.1)**
- Papilledema and Pseudotumor Cerebri:
  - MRI Brain without and with contrast (CPT® 70553) (preferred) and/or CT Head without contrast (CPT® 70450), when MRI contraindicated or for urgent evaluation, when there is suspected elevated intracranial pressure and papilledema such as with pseudotumor cerebri (idiopathic intracranial hypertension) to exclude cerebral mass lesions, obstructive hydrocephalus, etc. See **General Guidelines – CT Head (HD-1.4)** regarding required use of CT Head prior to lumbar puncture and/or spinal tap.
  - MRI Orbit without and with contrast (CPT® 70543) or CT Orbit without and with contrast (CPT® 70482) if there is concern for orbital pseudotumor or a primary bilateral orbital disorder. See **Eye Disorders and Visual Loss (HD-32.1)** regarding concern for orbital pseudotumor or primary orbital disorder.
  - Repeat imaging to evaluate either:
    - Shunt dysfunction in those individuals who have had ventriculoperitoneal (VP) or lumboperitoneal (LP) shunts (See Hydrocephalus Shunts (HD-11.14))
    - Clinical deterioration (with worsening or new neurological signs and symptoms)
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496) for suspected venous sinus thrombosis or venous stenosis.<sup>2</sup>
    - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. 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
    - See **Stroke/TIA (HD-21.1)**

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# Paresthesias and/or Weakness (HD-18)

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# Sensory/Weakness Complaints (HD-18.1)

HD.PS.0018.1.A

v1.0.2023

- Advanced imaging for general complaints **specific** only for sensory and/or weakness that are unaccompanied by other signs or symptoms and have the following: (1) a thorough history and neurological exam, (including the symptomatic area), (2) documentation localizes to the central nervous system and (3) the use of imaging can verify a specific diagnosis.
- Imaging decisions are based on the exam and documentation provided:
  - Findings supportive of advanced imaging of the brain and/or spinal cord:
    - One or more of the following abnormal exam findings: hyperreflexia, Babinski/Hoffman sign, increased tone in an affected limb, weakness of extensor muscles in an upper extremity and/or weakness of flexor muscles in the lower extremity and/or sensory or motor symptoms limited to two limbs on the **same side** of the body.
      - MRI Brain with and without contrast (CPT® 70553) or without contrast (CPT® 70551) and/or MRI Cervical Spine with and without contrast (CPT® 72156) or without contrast (CPT® 72141) and/or MRI Thoracic Spine with and without contrast (CPT® 72157) or without contrast (CPT® 72146)
  - Findings supportive of advanced imaging of the spinal cord:
    - Decreased pinprick sensation on one side, weakness and diminished proprioception on the other side
    - Sensory level on the trunk with sensory loss in both legs
      - MRI Brain with and without contrast (CPT® 70553) or without contrast (CPT® 70551) and/or MRI Cervical Spine with and without contrast (CPT® 72156) or without contrast (CPT® 72141) and/or MRI Thoracic Spine with and without contrast (CPT® 72157) or without contrast (CPT® 72146)
      - MRI Lumbar Spine is not needed since Cervical and Thoracic studies will usually visualize the entire spinal cord.
      - MRI Lumbar Spine without (CPT® 72148) or with and without (CPT® 72158) is supported if there is specific concern for involvement of the conus medullaris (the terminal end of the spinal cord). Symptoms suggestive of conus medullaris syndrome include, but are not limited to, saddle anesthesia, urinary retention, bowel incontinence, and lower limb paresthesia and/or weakness.
  - Findings NOT consistent with a spinal cord localization and do not warrant spinal cord imaging include the following:
    - Sensory loss that involves the hands and feet and not the trunk
    - Diminished reflexes in an affected limb

- Limb pain
- Weakness and diminished pain sensation in the same limb may be due to either a peripheral or brain/brainstem lesion
- For generalized pure motor syndromes, (such as generalized weakness), refer to the Peripheral Nerve Disorders (PND) Imaging Guidelines:
  - Myopathy or Inflammatory Muscle Diseases
  - Clinical exam, lab testing, and EMG/NCV are typically required prior to imaging for myopathy or myositis. See **Muscle Diseases (PN-6.2)** and **Gaucher Disease (Storage Disorders) (PN-6.3)**
  - Motor Neuron Disease or Amyotrophic Lateral Sclerosis (ALS) See **Motor Neuron Disease/Amyotrophic Lateral Sclerosis (ALS) (PN-8.1)**
  - Neuromuscular (Junction) Disease See **Neuromuscular Junction Disorders (PN-6.1)**
  - Multifocal Motor Neuropathy (MMN) and Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)
    - EMG/NCV required initially. See **Polyneuropathy (PN-3.1)**
- For pure sensory symptoms:
  - Proximal and distal symmetric pattern is supportive of spinal cord involvement.
    - MRI Cervical Spine with and without contrast (CPT® 72156) or without contrast (CPT® 72141) and/or MRI Thoracic Spine with and without contrast (CPT® 72157) or without contrast (CPT® 72146)
    - MRI Lumbar Spine is not needed since Cervical and Thoracic studies will usually visualize the entire spinal cord. See **Myelopathy (SP-7.1)** in the Spine Imaging Guidelines
  - Polyneuropathy
    - EMG/NCV is required initially. See **Polyneuropathy (PN-3.1)** in the Peripheral Nerve Disorders (PND) Imaging Guidelines
  - Proximal and distal asymmetric (20% may occur as part of a paraneoplastic syndrome) See **Polyneuropathy (PN-3.1)** in the Peripheral Nerve Disorders (PND) Imaging Guidelines and **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines
- For mixed sensory and motor symptoms refer to the Peripheral Nerve Disorders (PND) Imaging Subsections:
  - Proximal asymmetric symptoms may be due to a polyradiculopathy or radiculoplexopathy, See **Brachial Plexus (PN-4.1)**, **Lumbar and Lumbosacral Plexus (PN-5.1)**, and appropriate Spine Imaging Guidelines
  - For proximal and distal symmetric symptoms See **Polyneuropathy (PN-3.1)**
- Focal symptoms:
  - Radiculopathy for the appropriate level in the Spine Imaging Guidelines

- Plexopathy to Brachial Plexus or Lumbar and Lumbosacral Plexus See **Brachial Plexus (PN-4.1)** or **Lumbar and Lumbosacral Plexus (PN-5.1)** in the Peripheral Nerve Disorders (PND) Imaging Guidelines
- Thoracic Outlet Syndrome See **Thoracic Outlet Syndrome (CH-31.1)** in the Chest Imaging Guidelines
- Mononeuropathy or Focal Neuropathy See **Focal Neuropathy (PN-2.1)** in the Peripheral Nerve Disorders (PND) Imaging Guidelines.

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

- Paresthesia refers to an abnormal sensation that is associated with nervous system dysfunction and may be described as a tingling, pricking, pins and needles, or a burning sensation. The priority is to determine whether the etiology is due to pathology of the peripheral or central nervous system.
- A thorough clinical history, including symptom location and time course, can be helpful to differentiate between the two. For example, paresthesia affecting one side of the face and/or body (i.e. hemisensory deficit) points strongly towards central nervous system dysfunction. Therefore, MRI Brain (CPT® 70551 or CPT® 70553), MRI Cervical Spine (CPT® 72141 or CPT® 72156) and/or MR Thoracic Spine (CPT® 72146 or CPT® 72157) could be warranted, especially based on the location of symptoms. Typically, lumbar spine imaging is not indicated unless there is sphincter involvement, saddle anesthesia, and/or cauda equina syndrome is suspected. In contrast, an insidious course of distal, symmetric limb paresthesia is more commonly associated with peripheral nerve abnormalities. In such case, NCS/ EMG testing results should be completed prior to advanced imaging. (See **Peripheral Nerve Imaging Guidelines** ).
- A detailed neurological exam is most essential in determining whether advanced imaging is indicated. The presence of upper motor neuron signs (e.g. increased tone, hyperreflexia, presence of Babinski or Hoffman signs) necessitates central nervous system imaging. Conversely, lower motor neuron signs (e.g. decreased tone, hypo- or areflexia, muscle atrophy) can indicate that nerve conduction and needle EMG testing should be completed in order to evaluate for neuropathy or other peripheral nervous system diseases. It is important to note that both peripheral and central nervous system disease can co-exist. As a result, if both upper and lower motor neuron signs are observed simultaneously, advanced imaging is appropriate regardless of NCS/EMG testing results. See **Polyneuropathy (PN-3.1)** in the Peripheral Nerve Disorders (PND) Imaging Guidelines.

## References (HD-18)

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# Pituitary (HD-19)

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## Pituitary (HD-19.1)

HD.PT.0019.1.A

v1.0.2023

- Endocrine laboratory studies should be performed prior to considering advanced imaging, except in the cases of stable, non-functioning microadenomas or macroadenomas and cysts.
- MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) with a specific pituitary protocol that includes fine cuts through the sella is the primarily performed pituitary imaging:
  - MRI Orbit, Face, Neck without and with contrast (CPT® 70543) or CT Head without and with contrast (CPT® 70470) are alternatives
  - CT Head without contrast (CPT® 70450) or without and with contrast (CPT® 70470) and/or CT Maxillofacial without contrast (CPT® 70486) in addition to MRI to visualize perisellar bony structures in the preoperative evaluation of certain sellar tumors and for preoperative planning for transphenoidal approaches
    - See **General Guidelines – Anatomic Issues (HD-1.1)** as CT Temporal bone (CPT® 70480) is supported instead of CT Maxillofacial per surgeon's preference and contrast level
  - CTA Head (CPT® 70496) or MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) for surgical planning
  - MRI Brain without and with contrast (CPT® 70553) covers both brain and dedicated pituitary if performed at the same time; no additional CPT® codes are needed
- Repeat imaging for incidentally found lesions on other studies:
  - MRI Brain without and with contrast (CPT® 70553) or MRI Orbit/Face/Neck without and with contrast (CPT® 70543) follow-up dedicated pituitary study obtained if a pituitary abnormality is reported incidentally on a MRI Brain or CT Head performed for other reasons (MRI Brain without and with contrast [CPT® 70553] covers both brain and dedicated pituitary if performed at the same time; no additional CPT® codes are needed); further evaluation and subsequent imaging dependent on specific imaging and biochemical laboratory evaluation findings.
- Repeat Imaging in the setting of worsening clinical status or new neurologic symptoms
- For Amenorrhea: See **Secondary Amenorrhea (PV-3.1)** in the Pelvic Imaging Guidelines

### Pituitary Imaging

Indication	Initial Imaging	Repeat Imaging
<b>Microadenoma:</b> Nonfunctioning, unexplained pituitary asymmetries, or incidentally found small tumors (<10 mm)	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)</li> </ul>	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) at 12 months and then (if stable in size), every 1-2 years for 3 years, and less frequently thereafter based on clinical status</li> </ul>
<b>Macroadenoma (≥10 mm):</b> Nonfunctioning and/or not surgically removed including those with a post-operative remnant	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)</li> </ul>	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) every 6 months for the first year and then (if stable in size), every year for 3 years, and less frequently thereafter based on clinical status (longer if craniopharyngioma)</li> </ul>
<b>Acromegaly*</b> (Elevated IGF-1 confirmed by lack of suppression of growth hormone on glucose suppression testing)	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)</li> </ul>	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)</li> <li>At least 12 weeks after surgery to evaluate for residual tumor</li> <li>If treated with Pegvisomant, 6 to 12 months after treatment initiated, then annually if stable</li> <li>Long-term follow-up imaging based on clinical and biochemical status at the request of a specialist or any provider in consultation with a specialist</li> </ul>

Indication	Initial Imaging	Repeat Imaging
<b>Cushing's Disease**</b> (Pituitary ACTH excess leading to hypercortisolism)	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)</li> </ul>	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)</li> <li>At least 12 weeks after surgery as new baseline</li> <li>Annually after bilateral adrenalectomy for Cushing's disease or ectopic ACTH production</li> <li>Long-term follow-up imaging based on clinical and biochemical status at the request of a specialist or any provider in consultation with a specialist</li> </ul>
<b>Rathke's cleft cyst/Simple cyst</b>	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)</li> </ul>	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) in one year; if stable and without mass effect or invasion into surrounding structures, no further imaging is required.</li> </ul>

Indication	Imaging
<b>Prolactinomas***</b>	<ul style="list-style-type: none"> <li>• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) with: <ul style="list-style-type: none"> <li>• Diagnosis: <ul style="list-style-type: none"> <li>• Unexplained prolactin level above the normal range</li> </ul> </li> <li>• On Dopamine Agonist (DA) therapy with good response: <ul style="list-style-type: none"> <li>• Macroadenomas 3 months after start of DA therapy</li> <li>• Microadenomas 1 year after start of DA therapy</li> <li>• To decide on stoppage of therapy after ~2 years if in “remission” (normal PRL and no visible tumor on MRI)</li> </ul> </li> <li>• On Dopamine Agonist therapy with suboptimal response: <ul style="list-style-type: none"> <li>• PRL levels rise</li> <li>• New symptoms develop (galactorrhea, vision changes, headaches, pituitary deficiency)</li> <li>• If on high dose maximal DA and no plans for surgery/radiation therapy use guideline for microadenoma or macroadenoma</li> </ul> </li> <li>• After Dopamine Agonist therapy: <ul style="list-style-type: none"> <li>• Rise in PRL level</li> <li>• For DA stoppage at menopause, use guideline for microadenoma or macroadenoma</li> </ul> </li> <li>• Galactorrhea/nipple discharge with normal prolactin and thyroid function levels: See <b><u>Nipple Discharge/Galactorrhea (BR-6.1)</u></b> in the Breast Imaging Guidelines</li> </ul> </li> </ul>
<b>Medication-induced Prolactinemia****</b>	<ul style="list-style-type: none"> <li>• To differentiate between medication-induced hyperprolactinemia and hyperprolactinemia due to a pituitary or hypothalamic mass if the medication cannot be discontinued or hyperprolactinemia persists after medication discontinuation<sup>22</sup></li> </ul>

Indication	Imaging
<b>TSH, FSH, or LH producing adenomas (inappropriate pituitary hypersecretion of TSH, FSH or LH)*****</b>	<ul style="list-style-type: none"> <li>• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) when hormone levels are inappropriately elevated and there is a concern for a pituitary lesion.</li> <li>• Refer to appropriate post-operative, or Microadenoma/Macroadenoma guidelines based on the size of the lesion and initial management.</li> <li>• Long-term follow-up imaging based on clinical and biochemical status at the request of a specialist or any provider in consultation with a specialist</li> </ul>
<b>Male Hypogonadism*****</b>	<ul style="list-style-type: none"> <li>• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) if ONE of the following: <ul style="list-style-type: none"> <li>• Severe secondary hypogonadism (as indicated by morning serum testosterone level &lt;150 ng/dl and low or normal LH and FSH levels) ( <b><u>Background and Supporting Information</u></b> )</li> <li>• Below normal testosterone level (serum total testosterone, free testosterone and/or bioavailable morning testosterone) AND low or normal LH and FSH levels, in an individual with either: <ul style="list-style-type: none"> <li>• Panhypopituitarism</li> <li>• Hyperprolactinemia</li> <li>• Signs of tumor mass effect (headache, visual impairment, or visual field deficit)</li> <li>• Elevated sex hormone binding globulin (SHBG)</li> </ul> </li> </ul> </li> </ul>
<b>Hypopituitarism</b>	<ul style="list-style-type: none"> <li>• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)</li> </ul>

Indication	Initial Imaging	Repeat Imaging for Non-Operative Care
<b>Diabetes Insipidus (DI)</b>	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) if: <ul style="list-style-type: none"> <li>Laboratory testing consistent with DI (serum osmolality should be high and urine osmolality should be low) and etiology uncertain</li> </ul> </li> </ul>	NA
<b>Syndrome of Inappropriate ADH (SIADH)</b>	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) ) or MRI Brain without contrast (CPT® 70551) if: <ul style="list-style-type: none"> <li>Etiology remains uncertain or is thought to be in the nervous system;</li> <li>Urine osmolality should be high and serum osmolality low</li> </ul> </li> </ul>	NA
<b>Other Pituitary Region Tumors</b>	<ul style="list-style-type: none"> <li>Evaluation may require CT in addition to MRI to evaluate for hyperostosis.</li> </ul>	

### **Background and Supporting Information**

- **\*Acromegaly:** A serum level of growth hormone greater than 1ng/mL when measured two hours following an oral glucose load confirms acromegaly.
- **\*\*Cushing's Disease:** It is important to differentiate Cushing's syndrome (hypercortisolism from any source) from Cushing's disease which is ACTH hypersecretion from the pituitary gland. Hypercortisolism is quantified by 24hour urine cortisol collection, low dose dexamethasone suppression test and/or late night salivary cortisol measurement. ACTH is elevated in Cushing's disease and ectopic sources of ACTH production, but suppressed in other causes of hypercortisolism. A high dose dexamethasone suppression test can help determine if the elevated ACTH is from a pituitary or ectopic source. Petrosal sinus sampling may be required for tumor localization preoperatively in the setting of a normal pituitary MRI or a small adenoma. These tumors may be managed with surgery, medical therapy, radiation and/or bilateral adrenalectomy.
- **\*\*\*Prolactinoma:** To establish the diagnosis of hyperprolactinemia, a single measurement of serum prolactin is recommended; a level above the upper limit of normal confirms the diagnosis as long as the serum sample was obtained without excessive venipuncture stress. Pregnancy and primary hypothyroidism should be excluded as physiologic causes of prolactin elevation and medications that may be

contributing to prolactin elevation should be considered. Dopamine agonist therapy is typically stopped during pregnancy, monitoring of prolactin levels ceases. Routine imaging surveillance during pregnancy is not recommended due to risk to fetus. Repeat imaging with MRI without gadolinium can be performed however for new or worsening symptoms, such as headaches or visual symptoms.

- **\*\*\*\* Medication-induced prolactin elevation:** Medication induced hyperprolactinemia is seen most commonly with antipsychotics/neuroleptics and antidepressants, but may also be seen with some anti-emetics and antihypertensive agents. In individuals on prolactin elevating drugs, a prolactin level should be repeated after withdrawal of medications for 72 h, however, this approach may not be safe if this treatment is offered for psychiatric indications. If stopping the drug is not feasible, pituitary MRI is advised to rule out a sellar/parasellar tumor.<sup>22</sup>
- **\*\*\*\*\*TSH, FSH, or LH producing adenomas:** These are the least common of all hormonally active pituitary tumors. Individuals with TSH secreting adenomas have inappropriate TSH elevation in the setting of hyperthyroidism (elevated thyroid hormone levels). Almost all gonadotroph adenomas are clinically non-functioning. The infrequent presentation of a functioning gonadotroph adenoma should be differentiated clinically from appropriate FSH and LH elevation seen in low estrogen states (including menopause) as well as primary hypogonadism (testicular failure). Functioning TSH, FSH or LH pituitary adenomas may be managed with surgical, radiation and/or medical therapies.
- **\*\*\*\*\*Male Hypogonadism:** Alterations in sex hormone-binding globulin (SHBG) can impact testosterone levels. Free or bioavailable testosterone concentrations should be measured when total testosterone concentrations are close to the lower limit of the normal range and when altered SHBG levels are suspected (e.g. moderate obesity, nephrotic syndrome, hypo- and hyperthyroidism, use of glucocorticoids, progestins, estrogens, and androgenic steroids, anticonvulsants, acromegaly, diabetes mellitus, aging, HIV disease, liver cirrhosis, hepatitis). LH and FSH should be obtained to evaluate for secondary (central) hypogonadism, once low testosterone level is confirmed. Morning testosterone level is drawn anytime before 10 am for a typical sleep-wake cycle.
- "Central hypothyroidism is an anatomic or functional disorder of the pituitary gland or the hypothalamus, resulting in altered TSH secretion. Diagnosis is usually made biochemically with low circulating free T4 (FT4) concentrations associated with low/normal serum TSH levels."<sup>24</sup>



# Post-Operative and Repeat Imaging Indications (HD-19.2)

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- For imaging in the immediate post-operative period or for acute surgical complications, See **Primary Central Nervous System Tumors (ONC-2.1)** in the Oncology Imaging Guidelines.
- A routine post-operative MRI is generally done at 3 months and/or at the discretion of, or in consultation with, a specialist.
- Frequency of follow-up imaging depends on the post-operative size and/or functional status of the pituitary adenoma. Refer to the grid sections for Microadenoma/Macroadenoma as well as those for disorders of pituitary hormone excess.
- Individuals with hyper-functioning tumors such as acromegaly, Cushing's disease, and excess TSH secretion may be treated with a combination of surgery, medical therapy and radiation. Long-term monitoring of clinical status and repeat imaging at the discretion of, or in consultation with, a specialist is appropriate.

## Empty Sella Turcica (HD-19.3)

HD.DPT.0019.3.A

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- Enlarged/Empty Sella Turcica: An enlarged sella turcica without evident tumor is an incidental finding on MRI Brain or CT Head from a defect in the dural diaphragm of the sella (especially if there is elevated intracranial pressure from another cause), pituitary surgery, or as a result of a pituitary tumor which has expanded the sella and then infarcted (pituitary apoplexy).
- MRI Brain with and without contrast (pituitary protocol) (CPT® 70553) with thin sections of pituitary or MRI Brain without contrast (CPT® 70551) is supported. CT Head with and without contrast (CPT® 70470) – If MRI is contraindicated.
  - Primary Empty Sella:
    - Incidentally found on other studies, asymptomatic and no related abnormalities: follow up at 2 years. No further imaging unless clinical symptoms develop (neuro-/ophthalmological symptoms, intracranial hypertension, or endocrine/hormonal abnormalities).
    - Following medical or surgical treatment of related endocrine, neurological, or ophthalmological problems: follow-up imaging every 6 months in the year after treatment and/or at the request of a specialist or any provider in consultation with a specialist. See **Papilledema/Pseudotumor Cerebri (HD-17.1)** for additional imaging recommendations
  - Secondary Empty Sella
    - Imaging according to the cause or if clinical disease progression (such as adenomas, infiltrative or malignant disorders, hormonal abnormalities, neuro-/ophthalmological symptoms)

# Craniopharyngioma and Other Hypothalamic/Pituitary Region Tumors (HD-19.4)

HD.DPT.0019.4.A

v1.0.2023

- PET Brain Metabolic Imaging and MR Spectroscopy do not have a defined role in the evaluation of craniopharyngioma

Indication	Imaging Study
Initial staging for all individuals	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553)</li> <li>Concurrent CT Head without contrast (CPT® 70450) in addition to MRI if craniopharyngioma is suspected</li> </ul>
<ul style="list-style-type: none"> <li>Additional initial staging for individuals with: <ul style="list-style-type: none"> <li>Multicentric tumors</li> <li>Clinical signs or symptoms suggesting spinal cord involvement</li> </ul> </li> </ul>	<ul style="list-style-type: none"> <li>MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)</li> </ul>
Operative planning or image guidance	<ul style="list-style-type: none"> <li>MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546)</li> </ul> <p>OR</p> <ul style="list-style-type: none"> <li>CTA Head (CPT® 70496)</li> </ul>
Baseline imaging following resection	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553)</li> </ul>
Completion of radiotherapy	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553)</li> </ul>
Treatment response to chemotherapy	<ul style="list-style-type: none"> <li>MRI Brain without and with contrast (CPT® 70553) every 2 cycles during active treatment and at the end of planned chemotherapy</li> </ul>
Additional treatment response imaging during induction chemotherapy for individuals with measurable spinal cord disease on MRI	<ul style="list-style-type: none"> <li>MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158) every 2 cycle</li> </ul>

Indication	Imaging Study
Surveillance	<ul style="list-style-type: none"> <li>• MRI Brain without and with contrast (CPT® 70553) every 3 months for 1 year, then every 4 months for 1 year, then every 6 months for 1 year, then annually until 10 years after completion of therapy as late progressions can occur</li> <li>• For additional imaging guidelines for individuals in long term follow up after CNS tumor treatment that included radiation therapy, See <b><u>Second Malignant Neoplasms (SMN) (PEDONC-19.3)</u></b></li> </ul>
Suspected spinal cord recurrence	<ul style="list-style-type: none"> <li>• MRI Spine without and with contrast (Cervical-CPT® 72156, Thoracic-CPT® 72157, Lumbar-CPT® 72158)</li> </ul>

## Background and Supporting Information

### General Considerations:

- Imaging guidelines and treatment approaches for pediatric pituitary tumors other than craniopharyngioma are consistent with those used for adults with pituitary tumors
  - For these tumors follow guidelines in **Pituitary (HD-19.1)**
- Craniopharyngiomas are less common, accounting for 6 to 8% of pediatric CNS tumors.
- Most commonly affects children in the preadolescent ages
- Several key imaging findings can be used to differentiate the tumors in this region including the presence of calcifications, cysts, and T1/T2 enhancement patterns in craniopharyngiomas
  - These are best evaluated using a COMBINATION of both MRI and CT modalities. Preoperative prediction is much more successful when BOTH modalities are obtained prior to biopsy.
- Other less common tumors in the optic chiasm, sellar and suprasellar region may include Germ Cell Tumors (Germinomatous Germ Cell Tumors (GCT) See **CNS Germinomas and Non-Germinomatous Germ Cell Tumors (NGGCT) (PEDONC-4.7)** and **Langerhans Cell Histiocytosis (LCH) (PEDONC-18.2)** in the **Pediatric Oncology Imaging Guidelines**

### Treatment Considerations:

- Surgical resection is curative for many individuals

- Those with a complete resection should then be imaged according to surveillance guidelines after post-resection imaging is completed
- Individuals with incomplete resection and receiving adjuvant radiation therapy can have a single MRI Brain (CPT® 70553) at completion of radiotherapy and should then be imaged according to surveillance guidelines

## References (HD-19)

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# Scalp and Skull (HD-20)

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# Scalp and Skull Lesions (HD-20.1)

HD.SK.0020.1.A

v1.0.2023

The majority of these are benign soft tissue or bony lesions easily defined by physical examination or with skull x-rays or ultrasound.

- Ultrasound is initial imaging of scalp or skull lesions
- CT Head without or without and with contrast (CPT® 70450 or CPT® 70470) is appropriate for the following scenarios:
  - Any lesion on physician examination and skull x-ray or ultrasound which is not clearly benign.
  - In cases where surgical planning is in progress, x-rays and/or ultrasound are not required.
  - Langerhans' cell histiocytosis, myeloma, and metastatic cancer, when symptoms suggest bony lesions.
- MRI Brain without contrast (CPT® 70551) or with and without contrast (CPT® 70553) if there is concern for intracranial extension.
- See **Dental/Periodontal/Maxillofacial Imaging (HD-30.2)** for mandibular masses

# Skull Base Osteomyelitis (SBO) (HD-20.2)

HD.SK.0020.2.A

v1.0.2023

- Note: SBO may occur from the temporal bones or paranasal sinuses and imaging should be of the region of origin
- Neuroimaging is indicated in the diagnosis and treatment of skull base osteomyelitis and necrotizing external otitis. The following advanced imaging studies for the diagnosis of skull base osteomyelitis and necrotizing external otitis:
  - MRI Brain without and with contrast (CPT® 70553)
    - Will be positive earliest in disease
  - CT Head without contrast (CPT® 70450), CT Temporal bone without contrast (CPT® 70480), CT Temporal bone with contrast (CPT® 70481), CT Maxillofacial without contrast (CPT® 70486), CT Maxillofacial with contrast (CPT® 70487) or CT Neck with (CPT® 70491)
    - Will best define bony destruction, but is positive later in disease
  - Gallium-67 Scan
  - Bone Scan
    - Skull base osteomyelitis: + Gallium and + Bone scan
    - Necrotizing otitis externa: + Gallium and - Bone scan
  - Indium WBC may be substituted for or used in addition to Gallium scanning to evaluate response to therapy and especially in cases that have undergone surgical debridement.
- Treatment response: Gallium-67 Scan every 4-6 weeks till scan is negative
- Surveillance Scanning: Gallium-67 Scan at 4 weeks and 3 months post treatment

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**v1.0.2023**

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# Stroke/TIA (HD-21)

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# Stroke/TIA (HD-21.1)

HD.HL.0021.1.C

v1.0.2023

- CT Head without contrast (CPT® 70450), CTA Head without and with contrast (CPT® 70496) and CTA Neck (CPT® 70498) and CT perfusion (CPT® 0042T):
  - Acute stroke (within the first 24 hours)
  - Transient ischemic attacks (TIA)
  - Concern for intracerebral or subdural hemorrhage
- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) to evaluate concern for new stroke or TIA. MRI is preferred for evaluation concern for new stroke or TIA. MRI is preferred for evaluation of stroke/TIA, with or without a previous CT Head.
- A repeat CT Head without contrast (CPT® 70450) or with and without contrast (CPT® 70470) any time if there has been a change in clinical status, such as clinical deterioration, or concern for hemorrhagic conversion, or new onset seizure, etc.
- CT Head without contrast (CPT® 70450) or with and without contrast (CPT® 70470) is supported after 24 hours if there is a contraindication to MRI.
- TIA includes Amaurosis fugax or ocular microembolism (optic nerve/retinal arterial or Hollenhorst plaques seen on exam)
- MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496) AND MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498) added to CT Head or MRI Brain for evaluation of stroke or TIA. A previously performed Duplex Ultrasound Carotid Arteries (CPT® 93880), should not preclude these studies. Duplex Ultrasound Carotid Arteries (CPT® 93880) is not sufficient to image the vertebral arteries.
  - 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) See **Intracranial Aneurysms (HD-12.1)**
- MR or CT Venography (MRA Head [CPT® 70544, CPT® 70545, or CPT® 70546] or CTA Head [CPT® 70496]) to evaluate venous infarcts after diagnosis on MRI Brain or CT Head.
  - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. 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.
- 3D Rendering (CPT® 76377) performed with cerebral angiography is supported as part of the stroke evaluation. See **General Guidelines- Other Imaging Situations (HD-1.7)**.

- For consideration of Reversible Cerebral Vasoconstriction Syndrome (See **Sudden Onset of Headache (HD-11.3)** )
- One time MRI Brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) screening to detect silent cerebral infarcts in adults with HbSS or HbSb thalassemia.
- Transcranial Doppler Studies for individuals with documented stroke or TIA.
- Repeat imaging for follow up and resolution of stroke or hemorrhage as determined by a neurology or neurosurgery specialist or any provider in consultation with a neurology or neurosurgery specialist.
- Radiopharmaceutical Localization Imaging SPECT (CPT® 78803 or CPT® 78830) with vasodilating agent acetazolamide (Diamox) challenge when surgery or other vascular intervention is being considered for Moyamoya disease
- Evaluation of paradoxical venous thromboembolism in cryptogenic stroke with PFO, See **Acute Limb Swelling (PVD-12.2)** in the PVD Imaging Guidelines
- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) in the presence of neurological signs and/or symptoms, including headache, after COVID-19 infection and/or COVID-19 vaccination. See **General Guidelines – CT Head (HD-1.4)**, **Abnormal Blood Clotting (HD-11.9)**, and **Neuro-COVID-19 (HD-14.2)**
- Amyloid-PET Brain (CPT® 78811 or CPT® 78814) has been proposed to evaluate individuals with suspected Cerebral Amyloid angiopathy (CAA), a malady that causes weakening of blood vessels in the brain and results in small hemorrhages. Different types of amyloid are found in other conditions such as dementia with Lewy bodies, Parkinson's disease and Huntington's disease. Amyloid in CAA has low uptake of PET tracers when compared to these other conditions. Due to this low specificity, use of amyloid-PET is not considered appropriate in the evaluation of CAA. Medicare does not cover amyloid PET for this condition and there are no current CED studies available.<sup>31,32</sup> See **Dementia - PET (HD-8.2)**

## Cryptogenic Stroke (HD-21.3)

HD.ST.0021.3.A

v1.0.2023

- 25% of individuals with ischemic stroke have no probable cause and is considered cryptogenic after a standard workup including an echocardiogram, inpatient cardiac telemetry or 24-Holter monitoring, CT or MRI Brain and vessel imaging of the brain or neck arteries and hematologic tests.
- A stroke may also be considered cryptogenic after a standard evaluation fails to yield an etiology in a person <50 years of age without risk factors with more extensive testing.
- Most cryptogenic sources are embolic in etiology from venous or arterial sources with investigations from disturbances in coagulation and sources of embolism including patent foramen ovale (PFO) and paroxysmal atrial fibrillation.
- Specialized evaluation with the following documentation:
  - MRI/CT Brain with results of stroke
  - Results of MRA/CTA Head and Neck
  - TTE or TEE
  - 24-Hr Holter monitor or Inpatient cardiac telemetry and 12-Lead ECG
- Hematologic testing to include: CBC, Platelet count, INR, PT, PTT, D-Dimer and Arterial and Venous Hypercoagulability tests
  - MRA or CTA Pelvis for the evaluation of paradoxical venous thromboembolism with PFO, See **Acute Limb Swelling (PVD-12)** in the Peripheral Vascular Disease (PVD) Imaging Guidelines.
  - Workup for occult cancer, CT Chest Abdomen and/or Pelvis with contrast after the previously indicated tests with results are provided. See **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines.
  - Cardiac CT (CPT® 75574 or CPT® 75572) instead of TEE if TTE is inconclusive

## Transient Global Amnesia (HD-21.4)

HD.ST.0021.4.A

v1.0.2023

- Transient Global Amnesia (TGA) is a clinical diagnosis with the differential diagnosis including, but not exclusive to: ischemic events, migraine headaches, and transient epileptic amnesia.
- Characteristics of TGA may include the following:
  - Witnessed episode
  - There must be anterograde amnesia during the attack
  - Cognitive impairment is limited to amnesia
  - No clouding of consciousness or loss of personal identity
  - No focal neurological signs/symptoms
  - No epileptic features
  - Attack must resolve within 24 hours
  - No recent head injury or active epilepsy
- Head and vessel imaging for ischemic etiology work-up should follow **Stroke/TIA (HD-21.1)**



## References (HD-21)

**v1.0.2023**

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# Cerebral Vasculitis (HD-22)

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# Cerebral Vasculitis (HD-22.1)

HD.CV.0022.1.C

v1.0.2023

- The diagnosis of primary central nervous system vasculitis is challenging because of its nonspecific and varied symptoms. Central nervous system vasculitis typically presents with headache, followed by encephalopathy and behavioral changes. Focal neurologic deficits, including but not limited to, visual loss, unilateral weakness, language impairment, sensory loss, incoordination, occurs in 20% to 30% of individuals. Seizures and intracranial hemorrhage may also occur. With a strong clinical suspicion, brain imaging is important for supporting the diagnostic process and directing biopsy.<sup>6</sup>
- Primary central nervous system vasculitis includes Giant Cell Arteritis also known as Temporal Arteritis. See **New Headache Onset Older than Age 50 (HD-11.7)**
- MRI Brain without and with contrast (CPT® 70553) is supported when CNS vasculitis is suspected
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) and MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549); OR CTA Head (CPT® 70496) and CTA Neck (CPT® 70498) in addition to MRI Brain
- If initial vascular imaging is suspicious for vasculitis, 3D rendering (CPT® 76377) with cervicocerebral angiography/arteriography (See **General Guidelines- Other Imaging Situations (HD-1.7)**).
- Transcranial Doppler Studies for individuals with documented vasculitis or concern for vasospasm
- FDG-PET is not supported due to lack of peer reviewed literature or expert consensus supporting the study for vasculitis. Certain imaging studies are considered investigational by various payors, and their coverage policies take precedence over eviCore's guidelines. Advanced imaging studies, or other procedures, are considered investigational and experimental if there is a paucity of supporting evidence; if the evidence has not matured to exhibit improved health parameters or; the advanced imaging study/procedure lacks a collective opinion of support
- For extra-cranial giant cell arteritis evaluation, See Giant Cell Arteritis (PVD-6.9.2)

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

Classification of vasculitides based on vessel size adapted from Younger. MRA and CTA are useful for the evaluation of the large proximal arteries; evaluation of a possible small vessel vasculitis may be beyond the resolution of routine MRA and CTA Head. However, other abnormalities, such as atherosclerotic disease, arterial dissection, Moyamoya disease, or reversible cerebral vasoconstriction may be demonstrated. Conventional angiogram is superior to MRA and CTA in demonstrating abnormalities in smaller vessels and is considered the "gold standard" in the evaluation of primary small vessel CNS vasculitis

Dominant Vessel Involved	Primary	Secondary
Large arteries	<ul style="list-style-type: none"> <li>Giant cell arteritis</li> <li>Takayasu's arteritis</li> </ul>	Aortitis with rheumatoid disease; Infection (e.g. syphilis)
Medium arteries	<ul style="list-style-type: none"> <li>Classical polyarteritis nodosa</li> <li>Kawasaki disease</li> </ul>	Infection (e.g. hepatitis B)
Small vessels and medium arteries	<ul style="list-style-type: none"> <li>Wegener's granulomatosis</li> <li>Churg–Strauss syndrome</li> <li>Microscopic polyangiitis</li> </ul>	Vasculitis with rheumatoid disease, systemic lupus erythematosus (lupus cerebritis), Sjögren's syndrome, drugs, infection (e.g. HIV)
Small vessels	<ul style="list-style-type: none"> <li>Henoch-Schönlein purpura</li> <li>Essential cryoglobulinemia</li> <li>Cutaneous leukocytoclastic vasculitis</li> </ul>	Drugs (e.g. sulphonamides, etc.) Infection (e.g. hepatitis C)

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# Dizziness, Vertigo and Syncope (HD-23)

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# Dizziness/Vertigo (HD-23.1)

HD.DZ.0023.1.A

v1.0.2023

- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) when history and exam suggest non-benign dizziness such as:
  - Episodes lasting hour(s) or are continuous
  - Inconclusive positional testing or equivocal or unusual nystagmus findings
  - Visual disturbances including loss and diplopia
  - Headache
  - Hearing loss
  - Unilateral tinnitus
  - Abnormal cranial nerve findings
  - Ataxia
  - Positive Romberg sign
  - Absent head thrust sign
  - Focal neurologic deficits
  - Dysarthria
  - Drop attacks
  - Weakness, including unilateral or hemibody weakness
  - Failure to respond to vestibular therapy or is unable to participate due to clinical condition
  - Consideration of vestibular migraine as it is a diagnosis of exclusion.
  - See **Stroke/TIA (HD-21.1)** , **Headaches with Red Flags (HD-11.2)** and **Multiple Sclerosis (MS) and Related Conditions (HD-16)**
- If ENG/VNG (Electronystagmography/Video-nystagmography) test results are abnormal and support a central cause for vertigo, MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) is indicated
- CT Head without contrast (CPT® 70450) or without and with contrast (CPT® 70470) if concern for acute stroke (See **Stroke/TIA (HD-21.1)** ) and/or if MRI is contraindicated.
- For imaging indicated in the setting of head trauma, See **Head Trauma (HD-13.1)**
- Dizziness with asymmetric hearing loss (See **Hearing Loss and Tinnitus (HD-27)** ) and concern for vestibular schwannoma or possible Meniere's disease. (Note: MRI Brain should be performed with thin sections of IACs). Limited MRI Brain with attention to internal auditory canals (CPT® 70540, CPT® 70542, or CPT® 70543) 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)** ).

- CTA Head (CPT® 70496) and CTA Neck (CPT® 70498) or MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) and MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) added if concern for vertebrobasilar disease including dissection (acute onset vertigo and associated symptoms or signs of weakness, ataxia, drop attacks, visual loss, diplopia, dysarthria). See **General Guidelines – CT and MR Angiography (CTA and MRA) (HD-1.5)**, **Headache and Suspected Vascular Dissection (HD-11.1)** and **Intracranial Aneurysms (HD-12.1)** )
- CT Temporal bone without contrast (CPT® 70480) added if history of head trauma or concern for semicircular canal dehiscence, temporal bone fractures in individuals with post-traumatic vertigo and diagnosing erosion in the bony labyrinth from inflammatory or iatrogenic causes. (See **Background and Supporting Information** below).

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

- Dizziness, a common complaint, with benign and dangerous causes, may be continuous, triggered, or spontaneous.
- For the continuously dizzy individual with nystagmus at the time of evaluation, a head impulse test and a test of skew should be performed to determine if dizziness is due to a peripheral cause or a posterior circulation stroke. Abnormalities on exam may be indications for imaging as detailed below.
- For triggered dizziness, positional testing such as the Dix-Hallpike maneuver, and/or orthostatic blood pressure measurements, should be performed. If symptoms are reproduced on examination, triggered dizziness is confirmed. Imaging as indicated in the relevant sections below.
- Spontaneous dizziness may be due to vestibular migraine, TIA, or Meniere's disease, among other causes. A detailed neurologic examination should be performed, and imaging as detailed below.
- The Dix-Hallpike maneuver should be performed or the individual should be referred to a clinician who could perform the procedure if Benign Paroxysmal Positional Vertigo (BPPV)
- The Head Impulse Test (HIT) is also known as the Head thrust test. It is designed to evaluate the vestibular-ocular reflex in an individual with concern for a peripheral vestibulopathy due to ACUTE spontaneous vertigo. The individual is instructed to look at the examiner during the entire test. The individual's head is then quickly turned or rotated to one side and then the other. If normal, the individual's eyes should remain locked on the examiner. If abnormal, the eyes will move in the direction of the head rotation and then quickly correct. This saccade indicates peripheral vestibular hypofunction on the side of the direction that the head is turned. The HIT test is abnormal in individuals with vestibular neuronitis, and normal in individuals with a posterior circulation stroke.
- Posterior Canal BPPV (85-95% of BPPV cases) is defined as:
  - Individual reports repeated episodes of vertigo with changes in head position relative to gravity.
  - Each of the following criteria is fulfilled on physical exam:

- Vertigo associated with torsional (rotatory), upbeat (toward the forehead) nystagmus is provoked by the Dix-Hallpike test.
- There is a latency period between the completion of the Dix-Hallpike maneuver and the onset of vertigo and nystagmus.
- The provoked vertigo and nystagmus increase and then resolve within 60 seconds from the onset of the nystagmus.
- Lateral or Horizontal Canal BPPV (5-15% of BPPV cases) will have horizontal or no nystagmus to which a supine roll test assess for this condition.
- Exclusions for Dix-Hallpike maneuver
  - Individual previously diagnosed with BPPV and who on date of encounter in calendar year does not have positional dizziness or vertigo consistent with active BPPV
  - Individual has declined Dix-Hallpike maneuver
  - Individual has cervical spinal disease (i.e., cervical stenosis, severe kyphoscoliosis, limited cervical range of motion, Down's syndrome, severe rheumatoid arthritis, cervical radiculopathies, Paget's disease, ankylosing spondylitis, low back dysfunction, spinal cord injuries, spinal fractures)
  - Individual unable to lay flat (i.e., severe heart disease)
  - Individual has severe atherosclerotic disease or recent dissection involving the anterior or posterior cerebral circulation
  - Unable to be seated in exam chair (i.e., morbidly obese), or maneuver cannot be safely performed given morbid obesity
  - Ehlers Danlos/Marfans/Connective tissue disorder due to risk of cranio spinal instability/dissection
- Triggered episodic vestibular syndrome (t-EVS) usually last seconds to minutes with the most common triggers (vs. exacerbating factors) are head motion or change in body position. In the Emergency Department, benign paroxysmal positional vertigo (BPPV) is the second most common cause of t-EVS after orthostatic hypotension. Far lateral rotation of the neck leads to mechanical occlusion of one or both vertebral arteries causing temporary symptoms of vertigo and nystagmus when this position is maintained and may occur with the individual upright.
- Diagnoses or conditions associated with OH or nOH include: Parkinson Disease (PD), Multiple System Atrophy (MSA), Pure Autonomic Failure (PAF) or Dementia with Lewy Bodies (DLB), unexplained fall or syncope, peripheral neuropathies secondary to diabetes, amyloidosis and HIV), individuals  $\geq 70$  years of age and frail and on multiple medications and individuals with postural (orthostatic) dizziness or nonspecific symptoms that occur when standing. Symptoms may include: lightheadedness or dizziness, the sensation of blacking out, cognitive dysfunction, mental dulling, generalized weakness, neck pain or discomfort in the suboccipital and paracervical region (coat hanger) or platypnea (dyspnea while standing).
- Secondary or advanced laboratory testing is considered for use in select individuals for paraneoplastic syndromes (paraneoplastic panel) and serum and urine protein

electrophoresis for monoclonal gammopathy for peripheral neuropathy. See **Polyneuropathy (PN-3.1)** in the Peripheral Nerve Disorders Imaging Guidelines, **Multiple Myeloma and Plasmacytomas (ONC-25)** in the Oncology Imaging Guidelines, and **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines.

- Semicircular canal dehiscence (SCD) is a rare syndrome caused by dehiscence in the bony covering of the affected superior, posterior or lateral semicircular canal. When present, it can result in vestibular symptoms of vertigo associated with auditory symptoms including oscillopsia evoked by noise and conductive hearing loss. The vestibular symptoms in SCD can be debilitating. Individuals may note that loud noises cause them to see things moving or that they experience a similar sensation when they cough, sneeze, or strain to lift something heavy. The signs of vestibular abnormalities in SCD relate directly to the effect of the dehiscence which has created a third mobile window of the inner ear. Some individuals have a conductive hearing loss for low-frequency sounds that can resemble the pattern in otosclerosis.
- Occlusive carotid artery disease does not cause fainting but rather causes focal neurologic deficits such as unilateral weakness. Thus, carotid imaging will not identify the cause of the fainting and increases cost. Fainting is a frequent complaint, affecting 40% of people during their lifetime.

## Syncope (HD-23.2)

HD.DZ.0023.2.A

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- Advanced imaging (CT Head (CPT® 70450) or MRI Brain (CPT® 70551 or CPT® 70553) and vessel imaging (Carotid dopplers (CPT® 93880) and CTA Head (CPT® 70496) or CTA Neck (CPT® 70498) or MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) and/or MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) is not indicated for simple syncope without focal signs of a neurological deficit, external evidence of head trauma or symptoms of stroke.<sup>6,15</sup> A cardiac evaluation should be performed in the absence of focal signs and symptoms including a detailed history and examination (e.g. orthostatics), an EKG and/or additional evaluations including, but not exclusive to cardiac echocardiogram, tilt table testing, holter monitor, external loop recorder, etc.
- Exceptions of cases that require additional evaluation include frequent recurrent syncope with risk of injury or identified injury related to syncope, such as head trauma<sup>6,15</sup> See **Head Trauma (HD-13.1)**
- Situational syncope is not an indication for advanced imaging. This includes, precipitating factors prior to syncope including, but not limited to, coughing, defecation, eating, laughing and urination.
- **Myoclonic jerks** are frequently seen in vasovagal syncope and often misinterpreted as a sign of epilepsy. Loss of tone is usually seen in syncope whereas prolonged amnesia/confusion and tongue biting are symptoms and signs associated with a seizure. See **Epilepsy/Seizure (HD-9.1)**
- See **Stroke/TIA (HD-21.1)** and **Head Trauma (HD-13.1)**

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# Other Imaging Studies (HD-24)

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# Transcranial Magnetic Stimulation (TMS) (HD-24.1)

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HD.OI.0024.1.C

v1.0.2023

In TMS, an electromagnetic coil placed on the surface of the skull overlying the motor cortex depolarizes the motor axons, creating a motor evoked potential (MEP), which is recorded via superficial skin electrodes as it passes through the upper and lower motor pathways to an innervated muscle.

## Functional MRI (fMRI) (HD-24.2)

HD.OI.0024.2.A

v1.0.2023

- fMRI is useful in pre-operative scenarios to define the “eloquent” areas of brain
  - The ordering physician must be a neurologist, neurosurgeon or radiation oncologist or any provider in consultation with one of these specialists.
- Primary indications for fMRI include, but are not limited to, the following:
  - Assessment of intracranial neoplasm and other targeted lesions
  - Presurgical planning and operative risk assessment
  - Assessment of eloquent cortex (e.g., language, sensory, motor, visual centers) in relation to a tumor or another focal lesion
  - Surgical planning (biopsy or resection)
  - Therapeutic follow-up, as a one-time, post-operative, follow up study
  - Evaluation of preserved eloquent cortex
  - Assessment of eloquent cortex for epilepsy surgery
  - Assessment of radiation treatment planning and post-treatment evaluation of eloquent cortex
- fMRI is appropriate with PET Brain in epilepsy surgery planning
- Procedure codes for functional MRI:
  - CPT® 70554 MRI Brain, functional MRI, including test selection and administration of repetitive body part movement and/or visual stimulation, not requiring physician or psychologist administration
  - CPT® 70555 MRI Brain, functional MRI; requiring physician or psychologist administration of entire neurofunctional testing
  - MRA Head without contrast (CPT® 70544) may be erroneously ordered in place of fMRI, as the CPT codes are similar.
- MRI Brain (CPT® 70551 or CPT® 70553) and/or fMRI (CPT® 70554 or CPT® 70555) are appropriate concurrently. (See **Unlisted Procedures/Therapy Treatment Planning (Preface-4.3)** in the Preface Imaging Guidelines if MRI Unlisted is requested for surgical planning)

# Magnetic Resonance Spectroscopy (MRS) (HD-24.3)

HD.OI.0024.3.C

v1.0.2023

- Advanced imaging studies, or other procedures, are considered investigational and experimental if there is a paucity of supporting evidence; if the evidence has not matured to exhibit improved health parameters or; the advanced imaging study/procedure lacks a collective opinion of support
- MRS (CPT® 76390) involves analysis of the levels of certain chemicals in a pre-selected voxels (small regions) on an MRI scan done at the same time.
- When conventional imaging by magnetic resonance imaging (MRI) or computed tomography (CT) provides limited information regarding specific clinical questions, indications for MRS in adults and children include, but are not limited to, the following and is evaluated on a case-by-case basis:
  - Distinguish recurrent brain tumor from radiation necrosis as an alternative to PET (CPT® 78608)
  - Diagnosis of certain rare inborn errors of metabolism affecting the CNS (primarily pediatric individuals)
  - Evidence or suspicion of primary or secondary neoplasm (pretreatment and posttreatment)
  - Grading of primary glial neoplasm, particularly high-grade versus low-grade glioma
  - Evidence or suspicion of brain infection, especially cerebral abscess (pretreatment and posttreatment) and HIV-related infections
  - Seizures, especially temporal lobe epilepsy

## *Background and Supporting Information*

- Evaluation of certain primary brain tumors where diagnostic accuracy has been established in peer-reviewed literature. See Primary Central Nervous System Tumors – General Considerations (ONC-2.1), Low Grade Gliomas (ONC-2.2), and High Grade Gliomas (ONC-2.3) in the Oncology Imaging Guidelines

## CSF Flow Imaging (HD-24.4)

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HD.OI.0024.4.A

v1.0.2023

- Pulse-gated MRI imaging or MRI CINE is generally performed as a part of a MRI Brain study. It is not coded separately for preoperative evaluation of hydrocephalus, Chiari syndromes, Normal Pressure Hydrocephalus, Idiopathic Intracranial Hypertension (also known as pseudotumor cerebri), and spontaneous intracranial hypotension.
- There is no specific or unique procedure code for this study; it is done as a special sequence of a routine MRI Brain without contrast (CPT® 70551).
- If not previously performed as part of recent study, a second study for the purpose of evaluating CSF flow may be performed.

## CT or MRI Perfusion (HD-24.5)

HD.OI.0024.5.C

v3.0.2022

- Performed as part of a CT Head or MRI Brain examination in the evaluation of individuals with very new strokes or brain tumors.
- A CT perfusion study, if performed in conjunction with a CT angiogram of the intracranial and/or cervical vessels, can be performed before, after, or concurrent with the CT angiogram. A CTA Head and/or Neck is appropriate in conjunction with the CT Perfusion study (CPT® 0042T).
- CPT® 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 and to help identify those most likely to benefit from thrombolysis or thrombectomy.
- There is no specific CPT® code for MRI Perfusion. Perfusion weighted images are obtained with contrast and are not coded separately from a contrasted MRI Brain examination. If MRI Brain without and with contrast is appropriate, no additional CPT® codes are necessary or appropriate to perform MRI perfusion.
- Primary indications for perfusion magnetic resonance imaging (MRI) include the following:
  - Diagnosis and Characterization of Mass Lesions
    - Differential diagnosis (tumor versus tumor mimic)
    - Diagnosis of primary neoplasms (may include grading)
    - Surgical planning (biopsy or resection)
    - Targeting locations for biopsy
    - Guiding resection extent
  - Therapeutic follow-up
    - Radiation necrosis versus recurrent or residual tumor
    - Chemonecrosis versus recurrent or residual tumor
    - Pseudoprogession and pseudoresponse
    - Monitor potential transformation of non-resectable low grade neoplasms to higher grade
  - Assessment of Neurovascular Disease
    - Acute stroke (assessment of ischemic penumbra)
    - Assessment of the hemodynamic significance of cervical or intracranial vascular stenosis
    - Assessment of cervical or intracranial revascularization efficacy
    - Assessment of vasospasm
- Secondary indications include, but are not exclusive to:

- Follow-up of acute cerebral ischemia or infarction and/or reperfusion in the subacute or chronic phase of recovery
- To assist in planning and evaluating the effectiveness of therapy for cervical or intracranial arterial occlusive disease (as an isolated test or in combination with a cerebrovascular reserve challenge) and/or chronic cerebral ischemia
- Identifying cerebral hyperperfusion syndrome following revascularization
- Evaluation of the vascular status of solid tumors where MRI is degraded due to susceptibility artifact from air-containing spaces, surgical clips, or dental work
- Follow-up of tumor response to therapy
- Other indications are usually regarded as investigational and experimental. Advanced imaging studies, or other procedures, are considered investigational and experimental if there is a paucity of supporting evidence; if the evidence has not matured to exhibit improved health parameters or; the advanced imaging study/procedure lacks a collective opinion of support.

#### *Background and Supporting Information*

- Pre-contrasted images for MRI perfusion are needed with contrasted images in case there is blood, calcification or to determine if there are artifacts if the area in question is close to bone. MRI Brain with and without should be the requested study for MRI Perfusion.

# Magnetic Resonance Neurography (MRN) (HD-24.6)

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HD.OI.0024.6.C

v1.0.2023

- MRN is currently considered investigational. Advanced imaging studies, or other procedures, are considered investigational and experimental if there is a paucity of supporting evidence; if the evidence has not matured to exhibit improved health parameters or; the advanced imaging study/procedure lacks a collective opinion of support
- See **Magnetic Resonance Neurography (MRN) (PN-7.1)** in the Peripheral Nerve Disorders (PND) Imaging Guidelines.

# Cone Beam Computed Tomography (CBCT) (HD-24.7)

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HD.OI.0024.7.A

v1.0.2023

- CPT® Codes: CPT® 70486, CPT® 70487, CPT® 70488, CPT® 70480, CPT® 70482 (No separate 3-D rendering codes should be reported)
- An alternative to traditional CT imaging is in-office cone beam testing and possible decreased radiation dosage. The indications for office-based CT imaging are the same as for traditional scanners, and they should not be used for diagnosing or managing uncomplicated acute bacterial rhinosinusitis (ABRS).
- See **Temporomandibular Joint Disease (TMJ) (HD-30.1)** and **Dental/Periodontal/Maxillofacial Imaging (HD-30.2 )**



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# Epistaxis (HD-25)

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## Epistaxis (HD-25.1)

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HD.EX.0025.1.A

v1.0.2023

- CT Maxillofacial without or with contrast (CPT® 70486 or CPT® 70487) and/or MRI Orbit, Face, and/or Neck without and with contrast (CPT® 70543) are appropriate based on endoscopic findings of mass lesion during ENT examination.

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**v1.0.2023**

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# Mastoid Disease or Ear Pain (HD-26)

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# Mastoid Disease or Ear Pain (HD-26.1)

HD.MA.026.1.A

v1.0.2023

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

- Advanced imaging is not indicated in the overwhelming majority of individuals with ear pain.
- CT Temporal Bone without contrast (CPT® 70480) or without and with contrast (CPT® 70482), OR, MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553), OR MRI Orbits/Face/Neck without and with contrast (CPT® 70543) is indicated for the following:
  - Persistent ear pain without obvious cause.
  - Clinical suspicion for complicated or invasive infection such as mastoiditis.
  - Clinical suspicion of mass lesion causing ear pain.
  - Significant trauma with concern for hematoma formation.
  - Preoperative planning
- Cholesteatomas are expansive cysts of the middle ear filled with cellular debris. They can be congenital or arise from recurrent middle ear infections or trauma to the tympanic membrane. Hearing loss is usually conductive, although if the lesion is large enough combined conductive and sensorineural hearing loss may be present. Otoscopic exam findings and symptoms may include painless drainage from the ear or chronic/recurrent ear infections.
  - CT Temporal Bone without contrast (CPT® 70480) or without and with contrast (CPT® 70482), OR MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553), OR MRI Orbits/Face/Neck without and with contrast (CPT® 70543) is indicated for preoperative evaluation in cholesteatoma individuals.
  - CT Temporal Bone without contrast (CPT® 70480) or without and with contrast (CPT® 70482), OR MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553), OR MRI Orbits/Face/Neck without and with contrast (CPT® 70543) is indicated one time post-operatively to exclude residual or regrown cholesteatoma to avoid the need for a second-look surgery.

## References (HD-26)

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**v1.0.2023**

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# Hearing Loss and Tinnitus (HD-27)

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# Hearing Loss (HD-27.1)

HD.HL.0027.1.A

v1.0.2023

- An initial evaluation including hearing tests, by bedside testing or by formal audiology, is necessary to determine whether an individual's hearing loss is conductive (external or middle ear structures) or sensorineural (inner ear structures, such as cochlea or auditory nerve) hearing loss. See **General Guidelines (HD-1.0)**
- CT Temporal Bone without (CPT® 70480) or MRI Brain without and with contrast (with IAC views) (CPT® 70553) or without contrast (CPT® 70551):
  - Conductive hearing loss should have a CT Temporal Bone initially in the absence of an evident mass in the middle ear
  - Mixed conductive (MC)/Sensorineural (SN) hearing loss or any sensorineural hearing loss (MRI generally preferred for SN - See **Background and Supporting Information** )
  - Unilateral fluctuating or asymmetric hearing loss
  - Cholesteatoma [See **Mastoid Disease or Ear Pain (HD-26.1)**]
  - Congenital hearing loss
  - Surgical planning, including cochlear implants (both CT Temporal Bone and MRI Brain for surgical planning if requested by surgeon or any provider in consultation with the surgeon)
  - Hearing loss with vertigo [See **Dizziness/Vertigo (HD-23.1)**]
- CT Temporal Bone with contrast (CPT® 70481):
  - Glomus tumors or other vascular tumors of the middle ear, and/or surgical planning
  - Acquired sensorineural hearing loss if MRI unavailable or contraindicated
- Limited MRI Brain with attention to internal auditory canals (CPT® 70540, CPT® 70542, or CPT® 70543) 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)** )

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

- Sensorineural (SN) hearing loss – MRI is generally preferable to CT. CT Temporal bone is appropriate in post-traumatic SN hearing loss, to evaluate for bony remodeling of the IAC due to vestibular schwannoma and labyrinthine ossification resulting from prior infection and for consideration of otospongiosis, a common cause of MC and SN hearing loss.

# Tinnitus (HD-27.2)

HD.HL.0027.2.A

v1.0.2023

- A hearing evaluation is not required prior to imaging for tinnitus.
- The history in individuals with tinnitus should include a description of the tinnitus (episodic or constant, pulsatile or non-pulsatile, rhythmicity, pitch, quality of the sound), as well as inciting or alleviating factors. Continuous and pulsatile tinnitus are more concerning for an underlying and significant disorder. Audiometric assessment can be used as initial diagnostic testing particularly in individuals with tinnitus that is unilateral, persistent (>6 months) or associated with hearing difficulties. See **General Guidelines (HD-1.0)**.
- MRI Brain and internal auditory canal (IAC) without and with contrast (CPT® 70553), or MRI Brain and internal auditory canal (IAC) without contrast (CPT® 70551), or CT temporal bone without contrast (CPT® 70480) for:
  - Pulsatile tinnitus
  - Asymmetric or unilateral non-pulsatile tinnitus (i.e. tinnitus that localizes to one ear)
  - Tinnitus associated with focal neurologic abnormalities, including asymmetric hearing loss
  - Suspicion for vascular lesions
- Imaging not supported for bilateral non-pulsatile tinnitus without other neurologic signs or symptoms<sup>6</sup>
- MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA/CTV Head (CPT® 70496) **AND/OR** MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498) **AND/OR** CT Temporal Bone without contrast (CPT® 70480) or CT Temporal Bone with contrast (CPT® 70481):
  - Pulsatile tinnitus or suspicion for vascular lesions
- Limited MRI Brain with attention to internal auditory canals (CPT® 70540, CPT® 70542, or CPT® 70543) 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)**.)
- CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. 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.

## Background and Supporting Information

- Nonpulsatile tinnitus may be described as ringing, buzzing, or clicking sensations which is constant and nonsynchronous.

- Pulsatile tinnitus is a repetitive sound coinciding with the individual's heartbeat. The symptom may be subjective or objective

## References (HD-27)

v1.0.2023

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# Neurosurgical Imaging (HD-28)

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## Neurosurgical Imaging (HD-28.1)

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**v1.0.2023**

- Typically advanced imaging for monitoring disease for mass lesions occurs after biopsy (histologic) confirmation. This ensures appropriate determination related to phase of oncology imaging and alignment to appropriate diagnosis-specified guideline section.
  - However, repeat imaging by neurosurgeons or others of the management team for areas of the central nervous system (CNS) where permanent neurologic damage would be excessive with even a limited biopsy attempt is supported. Examples would include, but are not exclusive to: medically fragile individual, and tumors of the brainstem, eloquent areas of the brain, deep gray matter areas of the brain (ex. thalamus), and cavernous sinus.
- Repeat diagnostic head imaging:
  - Previous diagnostic head imaging is determined to be inadequate or additional imaging sequences/protocols are required by the neurosurgeon or the treatment team
  - Prior imaging is greater than 6 months old

# Neuronavigation (HD-28.2)

HD.NI.0028.2.C

v1.0.2023

- Neuronavigation
  - Neurosurgical navigation is “image-based” meaning that the necessary preoperative CT and MRI images are used for navigation in the operating room (image acquisition). Accurate registration (a process to match the preoperative images to the individual position) of preoperative images is necessary to guide surgery regardless of the navigation system that is used. Registration can be point-based or surface matched routines to allow the surgeon to view the overlapping data sets and the current situation to allow navigation.
  - The process of registration for neuronavigation via the acquisition of preoperative CT and MRI images does not require a radiologist interpretation.
    - It is not appropriate to request diagnostic imaging codes for the purpose of registration for neuronavigation.
    - Can be referenced by proprietary brand systems such as Brainlab or Stealth imaging procedures
    - See **Unlisted Procedures/Therapy Treatment Planning (Preface-4.3)** in the Preface Imaging Guidelines and **Unlisted Procedure Codes (ONC-1.5)** in Oncology in the Oncology Imaging Guidelines
- Advanced imaging for neuronavigation (image acquisition for registration for surgery) with one of each of the following as unlisted codes apply:
  - Unlisted MRI procedure code (CPT® 76498)
  - Unlisted CT procedure code (CPT® 76497)
  - Due to variances with techniques currently available for neuronavigation, the following are appropriate:
    - CTA Head without and with contrast (CPT® 70496) or MRA Head (CPT® 70544, CPT® 70545 or CPT® 70546) (to avoid arterial and venous structures)
    - 3D (CPT® 76377) See General Guidelines – Other Imaging Situations (HD-1.7)
  - Diagnostic imaging codes are only appropriate if radiological supervision and interpretation of imaging is necessary with supporting documentation
    - MRI Brain without contrast (CPT® 70551), or MRI Brain with contrast (CPT® 70552), or MRI Brain without and with contrast (CPT® 70553) (contrast as requested) and/or CT Head without contrast (CPT® 70450), or CT Head with contrast (CPT® 70460), or CT Head without and with contrast (CPT® 70470) (contrast as requested)



## Post Operative Imaging (HD-28.3)

HD.NI.0028.3.A

v1.0.2023

- Post-operative imaging including MRI Brain without contrast (CPT® 70551), or MRI Brain with contrast (CPT® 70552), or MRI Brain without and with contrast (CPT® 70553) (contrast as request) or CT Head without contrast (CPT® 70450), or CT Head with contrast (CPT® 70460), or CT Head without and with contrast (CPT® 70470) (contrast as request) per neurosurgeon's or in concert with management team's request that includes, but not exclusive to:
  - Within 24-72 hours following brain surgery including to document the need for repeat surgery or if adjuvant intervention is necessary, concern or rule out for complication(s), evaluation if incomplete resection vs. consideration for plan for gross resection
  - Signs or symptoms indicating concern of clinical deterioration
  - Development of new neurological signs or symptoms
  - Follow-up on blood products, edema, and/or concern of cerebrospinal fluid leak
  - Follow up imaging per condition based guideline
- See additional condition-based guidelines:
  - Pediatric Neurosurgeries
    - See **Special Imaging Studies in Evaluation for Epilepsy Surgery (PEDHD-6.3)** in the Pediatric Head Imaging Guidelines
    - See **Modality General Considerations (PEDONC-1.3)** and **Pediatric CNS Tumors (PEDONC-4)** in the Pediatric Oncology Guidelines
  - Epilepsy.
    - See Presurgical Work-Up for Drug-Resistant Epilepsy (HD-9.2)
  - Movement Disorders
    - See Movement Disorders (HD-15.1)
  - Pituitary or Sella Surgery.
    - See **Pituitary (HD-19.1)**
  - Acoustic Neuroma and Other Cerebellopontine Angle Tumors
    - See **Acoustic Neuroma and Other Cerebellopontine Angle Tumors (HD-33.1)**
  - Central Nervous System Tumors
    - See **Primary Central Nervous System Tumors (ONC-2)** and **Brain Metastases (ONC-31.3)** in the Oncology Imaging Guidelines

## References (HD-28)

**v1.0.2023**

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# Sinus and Facial Imaging (HD-29)

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# Sinus and Facial Imaging (HD-29.1)

HD.SI.0029.1.C

v1.0.2023

- CT Maxillofacial without contrast (CPT® 70486) or limited CT Sinus without contrast (CPT® 76380) is supported for ANY of the following:<sup>3</sup>
  - Acute sinusitis with no improvement in symptoms after a minimum of 4 weeks of treatment
  - Concern for potential or suspected complicated sinusitis, which is sinusitis with orbital or intracranial extension (See **Background and Supporting Information** below)
  - Recurrent sinusitis (4 or more episodes of acute sinusitis within the past 12 months without symptoms or signs between episodes)
  - Chronic sinusitis (≥12 weeks sinusitis) with at least two of the following signs and symptoms:
    - Mucopurulent drainage
    - Nasal obstruction or congestion
    - Facial pain, pressure, and/or fullness (may involve the anterior face, periorbital region, or manifest with headache that is localized or diffuse)
    - Decreased sense of smell
    - (**Note:** A trial of antibiotic therapy is not required prior to imaging if individual meets criteria for chronic sinusitis)
- Surgical candidate (See **Unlisted Procedures/Therapy Treatment Planning (Preface-4.3)** in the Preface Imaging Guidelines if unlisted code is requested for surgical planning).
- Studies requested for the purpose of navigation for sinus surgery should be coded CPT® 77011 (CT guidance for stereotactic localization). It is not appropriate to report both CPT® 70486 and CPT® 77011 for the same CT stereotactic localization imaging session.
- For unexplained cough See **Cough (CH-3.1)** in the Chest Imaging Guidelines.
- CT Maxillofacial with contrast (CPT® 70487) if ANY of the following is present:
  - Orbital or facial cellulitis
  - Proptosis
  - Abnormal visual examination
  - Ophthalmoplegia
  - Cystic fibrosis
  - Immunocompromised individual
  - Fungal or vascular lesions visualized in nasal cavity
- CT Maxillofacial without contrast (CPT® 70486) or CT Maxillofacial with contrast (CPT® 70487) or MRI Maxillofacial without and with contrast (CPT® 70543):

- Sinonasal obstruction, polyp, or suspected mass
- Suspected orbital complication
- Suspected invasive fungal sinusitis
- Osteomyelitis and odontogenic infections (MRI is the preferred modality) See **Skull Base Osteomyelitis (SBO) (HD-20.2)** and Dental/Periodontal/Maxillofacial Imaging (HD-30.2)
- MRI Brain with and without contrast (CPT® 70553) for suspected intracranial complication
- CT Orbit without contrast (CPT® 70480) or CT Orbit without and with contrast (CPT® 70482) performed alone or added to CT Maxillofacial for:
  - Suspected orbital complications
- For Skull Base Osteomyelitis (SBO) See **Skull Base Osteomyelitis (SBO) (HD-20.2)**
- Repeat imaging for ANY of the following scenarios:
  - An ENT specialist or any provider in consultation with an ENT specialist requests the imaging **and** ONE or more of the following:
    - There has been a follow-up visit since the previous imaging and there is no improvement after an additional 3 weeks of conservative treatment after initial imaging was completed
    - There is a new abnormality on exam such as obstructing mass
    - Planned sinus surgery (including but not limited to Balloon Sinus Ostial Dilation or Functional Endoscopic Sinus Surgery)
- Complication of ABRS (acute bacterial rhinosinusitis) is suspected based on:
  - Severe headache
  - Facial Swelling
  - Cranial nerve palsies
  - Photophobia
  - Orbital signs (cellulitis, impaired extraocular motility, decrease in vision or proptosis)
  - Fever
- CT findings that correlate with ABRS include opacification, air-fluid level, and moderate to severe mucosal thickening. Complications of ABRS are best assessed using iodine contrast-enhanced CT or gadolinium based MR imaging to identify extra-sinus extension or involvement. Suspected complications are the only indication for MR imaging of the paranasal sinuses in the setting of ABRS

For Cone Beam Imaging, See **Cone Beam Computed Tomography (CBCT) (HD-24.7)**

### *Background and Supporting Information*

- Rhinosinusitis is defined as inflammation of the nasal cavity and adjacent paranasal sinuses. Acute sinusitis refers to symptom duration <4 weeks, subacute 4 to 12 weeks, and chronic >12 weeks. Complicated sinusitis refers to symptoms suggesting spread of disease into adjacent structures, including orbital or intracranial complications.
- There is no evidence to support advanced imaging of acute (<4 weeks) and subacute (4 to 12 weeks) uncomplicated rhinosinusitis.
- There is no evidence to support routine follow-up advanced imaging after treatment with clinical improvement of sinusitis.

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**v1.0.2023**

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# Temporomandibular Joint Disease (TMJ) and Dental/Periodontal/Maxill ofacial Imaging (HD-30)

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# Temporomandibular Joint Disease (TMJ) (HD-30.1)

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HD.TJ.0030.1.A

v1.0.2023

- MRI TMJ (CPT® 70336) is the diagnostic study of choice and should be reserved for those who fail a minimum of 6 weeks of non-surgical treatment and who are actively being considered for TMJ surgery
- CT Maxillofacial without contrast (CPT® 70486) or without and with contrast (CPT® 70488) when there is suspicion of bony involvement based on prior x-ray or MRI
- Ultrasound (CPT® 76536) can be used to look for the presence of a joint effusion and to evaluate cartilage and disk displacement with open and closed mouth imaging and to guide injections
- TMJ imaging in children with Juvenile Rheumatoid Arthritis, See **Temporomandibular Joint (TMJ) Imaging in Children (PEDHD-25)** in the Pediatric Head Imaging Guidelines
- Jaw Asymmetry - 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. CPT® 78315 Bone Scan 3 Phase Study is appropriate in anticipation or consideration of surgery<sup>13</sup>

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

HD.TJ.0030.2.C

v1.0.2023

- Cone beam CT for surgical planning when plain x-rays alone are insufficient. Potential indications include but are not limited to:
  - Impacted teeth
  - Supernumerary teeth
  - Dentoalveolar trauma
  - Root resorption
  - Foreign body
  - Odontogenic cysts, tumors, or other jaw pathology
  - Cleft pathology
  - Orthognathic surgery for dentofacial anomalies
  - Osteomyelitis and odontogenic infections (X-ray not required)
  - Bisphosphonate-related osteonecrosis of the jaw (X-ray not required)
  - Salivary gland stones
  - Maxillofacial bone graft planning
  - Dental implants related to tooth loss from injury, trauma, or jaw pathology such as cysts, tumors, or cancer
- Cone Beam CT: Report with CPT® Codes: CPT® 70486, CPT® 70487, CPT® 70488, CPT® 70480, CPT® 70482
- 3-D rendering (CPT® 76377) should NOT be reported separately
- Cone beam CT (CBCT) may also be called i-CAT scanner or mini-CAT scanner

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# Eye Disorders and Visual Loss (HD-32)

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# Eye Disorders and Visual Loss (HD-32.1)

HD.VL.0032.1.A

v1.0.2023

- For specific conditions - See **Background and Supporting Information** that include table of abbreviations
- Examination of ocular complaints and visual loss may include evaluation of pupillary responses, extraocular motility, visual acuity, visual field testing, intraocular pressures, external examination, slit lamp examination, and/or fundoscopic exam of retinae.
- MRI Orbits without contrast (CPT® 70540) or MRI Orbits without and with contrast (CPT® 70543) or CT Orbits with contrast (CPT® 70481) or CT Orbits without contrast (CPT® 70480) and/or MRI Brain without contrast (CPT® 70551) or MRI Brain with and without contrast (CPT® 70553):<sup>1</sup>
  - Unexplained vision loss
  - Optic atrophy
  - Optic neuropathy
  - Papilledema/optic disc swelling - See **Papilledema/Pseudotumor Cerebri (HD-17.1)**
  - Afferent Pupillary Defect (APD)
  - Chiasmal symptoms/signs (including bitemporal field deficit)
  - Ophthalmoplegia, Diplopia, and/or Cranial nerve palsy
- For optic disc edema/papilledema, CT Head without contrast (CPT® 70450) is helpful to assess for space-occupying processes such as intracranial hemorrhage, mass effect and hydrocephalus.<sup>16</sup>
- For suspected optic neuritis, MRI is preferred modality - See **Multiple Sclerosis (MS) (HD-16.1)** and **Neuromyelitis Optica and NMO Spectrum Disorders (HD-16.2)**
- Homonymous defects are associated with retrochiasmal pathology - See **Stroke/TIA (HD-21.1)** ; or **Primary Central Nervous System Tumors (ONC-2)** in the Oncology Imaging Guidelines or **Brain Metastasis (ONC- 31.3)** in the Oncology Imaging Guidelines
- MRI Orbits without contrast (CPT® 70540) or MRI Orbits without and with contrast (CPT® 70543) or CT Orbits with contrast (CPT® 70481):
  - Exophthalmos (including thyroid eye disease), enophthalmos or nontraumatic orbital asymmetry
  - Suspected orbital cellulitis or atypical pre-septal cellulitis, uveitis or scleritis
  - Orbital mass or metastasis
  - Orbital inflammatory syndrome (orbital pseudotumor) and dacryocystitis or dacryoadenitis

- CT Orbit without contrast (CPT® 70480) and/or CT Head without contrast (CPT® 70450)
  - Orbital trauma with visual defect
  - Exophthalmos (including thyroid eye disease)
- When requested by the surgeon or in consultation with surgeon, contrast level as requested. This includes requests from Ophthalmologists and Oculoplastic surgeons. Contrast level preference may vary per institutional protocol.
- Binocular Diplopia from Cranial Nerve Palsies or Intracranial Disease<sup>1</sup>
  - MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) and/or MRI Orbits without and with contrast (CPT® 70543) or MRI Orbits without contrast (CPT® 70540) or CT Orbits without contrast (CPT® 70480):
    - Fourth Nerve Palsy
    - Sixth Nerve Palsy
    - Internuclear Ophthalmoplegia or Skew deviation
    - Third nerve palsy with pupillary involvement or suspicion of aneurysm
      - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496) is indicated. (See **Intracranial Aneurysms (HD-12.1)** )
- Amaurosis Fugax - See **Stroke/TIA (HD-21.1)**
  - Individuals describe a monocular transient darkening or loss of vision
- Central Retinal Artery Occlusion, Branch Retinal Artery Occlusion, and Ophthalmic Artery Occlusion - See **Stroke/TIA (HD-21.1)**
  - Individuals describe a sudden monocular loss of vision or visual field. Etiology is usually embolic and is considered a stroke to the retina
- There is currently no data to support advanced imaging while on Tepezza® (teprotumumab) unless there are neurologic symptoms or ophthalmologic symptoms.<sup>19,20</sup> Additional imaging indications include:
  - Reassess compressive optic neuropathy (including APD, decreased vision, and/or visual field defects)
  - Non-responders with relapses and/or worsening proptosis, diplopia, lid retraction, or optic neuropathy
  - Surgical planning for orbital decompression, strabismus surgery or lid surgery

### **Background and Supporting Information**

- Imaging Non-Indications
  - Imaging is not necessary if visual loss or ocular symptom/sign is due to known intrinsic eye disease, such as refractive errors, amblyopia, pterygium, subconjunctival hemorrhage, conjunctivitis, cataracts, macular degeneration, central serous retinopathy, retinal vein occlusion, retinal detachment, etc. Monocular diplopia is not an indication for imaging. Physiologic anisocoria

- (difference in pupil diameter between the two eyes of 2 mm or less) and surgically distorted pupils are not indications for imaging.
- Imaging is not typically necessary in cases of ptosis without concern for Horner's or 3rd nerve palsy
- Advanced imaging of the brain and orbit are not routinely paired. Appropriateness for each region is needed to image both regions, based on suspicion of these disorders.
- Orbital imaging alone may be sufficient unless other signs or symptoms suggest brain involvement.
- Thyroid function and iodine contrast: thyroid dysfunction can occur in susceptible individuals after iodine exposure.
- Autoimmune Retinopathy
  - Suspicion for CAR (Cancer associated retinopathy) or MAR (melanoma associated retinopathy) syndromes - See **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines
- Oncologic conditions
  - Retinoblastoma - See **Retinoblastoma (PEDONC-12)** in the Pediatric Oncology Imaging Guidelines
  - Uveal (choroidal) melanoma - See **Ocular Melanoma (ONC-5.9)** in the Oncology Imaging Guidelines
  - Biopsy results are not required before initial staging
- Temporal Arteritis (Giant Cell Arteritis) - See **Cerebral Vasculitis (HD-22.1)**
- List of Abbreviations and Meanings:

Abbreviation	Meaning
AC	Anterior chamber
APD	Afferent pupillary defect
BCVA	Best-corrected visual acuity
C3F8	Gas bubble injected into vitreous cavity during retina surgery
cc	With correction (current new or old glasses or contact lenses)
CP	Color plates
C/S	Conjunctiva/sclera
CSME	Clinically significant macular edema
CVF	Confrontation visual field (testing of gross field of view)
D	Disc, optic nerve head
DBH	Dot blot hemorrhages
DCR	Dacrocystorhinostomy
DFE	Dilated fundus exam



Abbreviation	Meaning
E	Esophoria at distance
E'	Esophoria at near
EOM	Extraocular movements
ERM	Epiretinal membrane
ET	Esotropia at distance
E(T)	Intermittent esotropia at distance
ET'	Esotropia at near
E(T)'	Intermittent esotropia at near
GVF	Goldmann visual field test
HT	Hypertropia
HVF	Humphrey visual field test (automated perimetry)
I	Iris
Ishihara	Commonly used color plates
IOP	Intraocular pressure
K	Cornea
LF	Levator function
LFH	Lid fissure height
LLL	Lids, lashes, lacrimal gland
M	Macula
ME	Macular edema
MH	Macular hole
MP	Membrane peel
MRD1	Margin-reflex distance from upper lid margin to pupillary light reflex
MRx	Manifest refraction
NI	No improvement
NSC or NS	Nuclear sclerotic cataract
OD	Right eye
OS	Left eye
ortho	Eyes are aligned on the same target
OCT	Ocular Coherence Tomography
P	Periphery

Abbreviation	Meaning
PD	Prism diopter
ph or PH	Pinhole (crude assessment of best-corrected visual acuity)
PPV or PPVx	Pars plana vitrectomy
PVD	Posterior vitreous detachment
RD	Retinal detachment
RT	Retinal tear
SB	Scleral buckle
sc	Without correction
SF6	Gas bubble injected into vitreous cavity during retina surgery
SLE	Slit lamp examination
SO	Silicone oil
SRF	Subretinal fluid
Ta	Applanation tonometry (intraocular pressure measurement)
Tp	Tonopen tonometry (intraocular pressure measurement)
V	Vessels
Va	Visual acuity
VF	Visual field testing (formal automated perimetry versus confrontation visual field testing)
X	Exophoria at distance
X'	Exophoria at near
XT	Exotropia
X(T)	Intermittent exotropia at distance
XT'	Exotropia at near
X(T)'	Intermittent exotropia at near

# Pupillary Abnormalities including Horner's Syndrome (HD-32.2)

HD.VL.0032.2.A

v1.0.2023

- Anisocoria and Other Pupillary Disorders
  - Physiologic anisocoria (difference in pupil diameter between the two eyes of typically 2 mm or less) and surgically distorted pupils are not indications for advanced imaging.
  - Dilated pupil from suspected Third nerve palsy - See **Eye Disorders and Visual Loss (HD-32.1)**
  - Horner's Syndrome (See below)
- Horner's Syndrome (anisocoria, ptosis, and ipsilateral anhidrosis) is caused by disruption of sympathetic innervation to the eye and face. Definitive diagnosis may be established by pharmacologic testing of the pupillary response with eye drops. Evaluation and imaging depends on determining whether the cause is a central lesion (brainstem or cervical spinal cord), preganglionic lesion (spinal cord or sympathetic chain in the chest), or postganglionic lesion (neck or carotid artery).
- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) for suspected intracranial or brainstem lesions
- MRI Cervical Spine without contrast (CPT® 72141) or MRI Cervical Spine without and with contrast (CPT® 72156) for suspected spinal cord abnormality
- CT Chest with contrast (CPT® 71260) for suspected chest mass
- CT Neck with contrast (CPT® 70491) for suspected neck mass
- CTA Neck without and with contrast (CPT® 70498) or MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) for suspected carotid injury or dissection
- MRI Orbits without contrast (CPT® 70540), MRI Orbits without and with contrast (CPT® 70543) or CT Orbit with contrast (CPT® 70481) for suspected orbital lesion or mass

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v1.0.2023

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# Acoustic Neuroma and Other Cerebellopontine Angle Tumors (HD-33)

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# Acoustic Neuroma and Other Cerebellopontine Angle Tumors (HD-33.1)

HD.AC.0033.1.A

v1.0.2023

- Acoustic neuroma and vestibular schwannoma may be used interchangeably
- Initial diagnosis is usually made during evaluation for asymmetric hearing loss and/or vertigo. See **Dizziness, Vertigo and Syncope (HD-23)** and **Hearing Loss and Tinnitus (HD-27)** for evaluation of those problems
- MRI Brain without and with contrast (CPT® 70553) which should be done with attention to the internal auditory canals for initial diagnosis.
- MRI Brain without contrast (CPT® 70551) if performed with FIESTA protocol
- MRI Orbits, Neck, or Face without and with contrast (CPT® 70543) with audiologic or clinical features of retrocochlear hearing loss and a negative MRI Brain and in the rare individual in whom a detailed search is indicated for both a lesion of the cerebellopontine angle **and** lesions of the cerebral hemispheres
- Repeat MRI Brain (contrast as requested) 6 months after diagnosis, then annually for 5 years and thereafter per specialist or any provider in consultation with a specialist.<sup>7</sup>
- MRI Brain without and with contrast with attention to the internal auditory canals (CPT® 70553) is performed after surgical resection and following stereotactic radiation therapy at 6 to 12 months to document the completeness of tumor removal and to serve as a baseline for further follow-up. Additional follow up is done annually for 5 years and every 2 years thereafter.
- See **Primary Central Nervous System Tumors- General Considerations (ONC-2.1)** in the Oncology Imaging Guidelines for additional imaging requests for surgery

## References (HD-33)

**v1.0.2023**

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# Pineal/Colloid Cysts (HD-34)

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# Pineal/Colloid Cysts (HD-34.1)

HD.PT.0034.1.A

v1.0.2023

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

- MRI Brain without contrast (CPT® 70551) or 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 without and with contrast (CPT® 70553) 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 without and with contrast (CPT® 70553) 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

## References (HD-34)

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**v1.0.2023**

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# Arachnoid Cysts (HD-35)

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# Arachnoid Cysts (HD-35.1)

HD.AR.0035.1.A

v1.0.2023

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 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, except in the following scenarios:
  - 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

## References (HD-35)

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# Sleep-Related Imaging (HD-37)

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# General Guidelines Sleep-Related Imaging (HD-37.1)

HD.SL.0037.1.A

v1.0.2023

- Oral Appliance: There is a lack of published case-controlled clinical studies in Sleep literature validating the use of advanced imaging with respect to oral appliance therapy (pretreatment assessment). Previous literature has demonstrated support for cephalometric studies (x-ray)<sup>1</sup> in predicting treatment success. Nasoendoscopy (sedated and non-sedated with provocative maneuvers such as Mueller maneuver) has been helpful as well in this regard.<sup>2</sup> Routine use of advanced imaging is not supported at this time
- Hypersomnolence: MRI Brain with and without contrast (CPT® 70553) when there are focal neurologic signs or suspicion for an inflammatory neurologic process as the etiology. Recognition and treatment of a comorbid sleep disorders is paramount, and a complete neurologic history and examination should precede any request for advanced imaging
- Central Sleep Apnea: MRI Brain with and without contrast (CPT® 70553) for unexplained central sleep apnea syndrome when a primary CNS etiology is suspected; i.e., unassociated with CHF, COPD or other potential etiology. Specific etiologies should be stated for imaging requests, including but not limited to, suspected Chiari malformation, stroke, CNS demyelinating disease, posterior fossa lesion, anoxia or infection



## References (HD-37)

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**v1.0.2023**

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