Cigna Medical Coverage Policies – Radiology
Head Imaging Guidelines

Effective February 12, 2024

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.

Table of Contents

Guideline Page

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

General Guidelines (HD-1)

Abbreviations for Head Imaging Guidelines

v2.0.2024

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

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

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

General Guidelines (HD-1.0)

HD.GG.0001.0.A
v2.0.2024

- 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, neuro-opthalmologist, endocrinologist, gynecologist, otolaryngologist, or ophthalmologist who has seen the individual since onset of symptoms, or any provider in consultation with one of the above specialists.
- 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
- CT head contrast as requested (CPT® 70450 OR CPT® 70460 OR CPT® 70470) is supported when MRI is contraindicated.

General Guidelines – Anatomic Issues (HD-1.1)

HD.GG.0001.1.A
v2.0.2024

- 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 Orbit/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.
- Cranial Neuropathies
  - MRI Brain without and with contrast (CPT ® 70553) or without contrast (CPT® 70551) is indicated for all individuals with new or worsening specific cranial nerve abnormalities. [29]
  - MRI Orbit/Face/Neck without and with contrast (CPT® 70543) or without contrast (CPT® 70540) is also indicated for individuals with abnormalities in cranial nerves I, II, III, IV, V, VI, VII, IX, X, XI, or XII[13, 29]
  - CT Neck with contrast (CPT® 70491) is supported for evaluation of abnormalities involving cranial nerves IX, X, XII, or XII[29]
  - Imaging of the Brain and Orbit, Face and/or Neck may be performed concurrently when requested.[29]
  - For specific cranial neuropathies[29], see the corresponding guideline section listed below:

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

- CN I: Olfactory nerve (see **Taste and Smell Disorders (HD-2.1)**)
- CN II, III, IV, VI: Optic, Oculomotor, Trochlear and Abducens (see **Eye Disorders and Visual Loss (HD-32.1)**)
- CN V: Trigeminal nerve (see **Trigeminal Neuralgia and other Centrally Mediated Facial Pain Syndromes (HD-10.1)**)
- CN VII: Facial nerve (see **Facial Palsy (HD-6.1)**)
- CN VIII: Vestibulocochlear nerve (see **Dizziness/Vertigo (HD-23.1)**, **Hearing Loss (HD 27.1)**, **Tinnitus (HD 27.2)**, **Acoustic Neuroma and Other Cerebellopontine Angle Tumors (HD 33.1)**. For isolated nystagmus (see **Eye Disorders and Visual Loss (HD-32.1)**)
- CN IX: Glossopharyngeal nerve (see **Glossopharyngeal Neuralgia/ Glossopharyngeal Neuropathy (HD-10.2)**)
- CN X: Vagal nerve, imaging as detailed above (see also **Recurrent Laryngeal Palsy/Vocal Cord Palsy (Neck-7.1)**)
- CN XI: Spinal accessory nerve, imaging as indicated above
- CN XII: Hypoglossal nerve, imaging as indicated above

- For cranial neuropathies, whether isolated or multiple, due to clinically suspected stroke and/or vascular dissection (see **General Guidelines - CT and MR Angiography (CTA and MRA) (HD-1.5)**, **Headache and Suspected Vascular Dissection (HD-11.1)** and **Stroke/TIA (HD-21.1)**)

Background and Supporting Information

If a detailed clinical evaluation is unable to localize the site of the lesion, imaging of the entire course of the relevant cranial nerve is required, as cranial neuropathy can result from pathology affecting the nerve fibers at any point along the course of the nerve, from the cranial nerve origin in the brainstem to the end organ supplied by the nerve, requiring multiple imaging modalities.

Number	Cranial Nerve Name	Nerve dysfunction on exam	Guideline Section in HD
I	Olfactory (smell)	Anosmia, hyposmia, parosmia, phatosmia	2
II	Optic (vision)	Optic neuritis, disc edema, papilledema, afferent pupillary defect APD)	16, 17, 32
III	Oculomotor (eye and pupil movement)	Eye "down and out", +/- dilated pupil, ptosis, diplopia	32
IV	Trochlear (depresses the eye)	Inability to depress the eye, diplopia	32

Number	Cranial Nerve Name	Nerve dysfunction on exam	Guideline Section in HD
V	Trigeminal (sensation, mastication, taste)	Pain, numbness, corneal reflex loss, jaw deviation, trigeminal neuralgia, loss of taste	10
VI	Abducens (lateral movement of the eye)	Eye turns medially, inability to abduct, lateral rectus palsy, diplopia	32
VII	Facial (movement facial muscles, taste at 2/3, salivation/lacrimation)	Inability to close eyelid, smile, nasolabial fold flattening, hyperacusis, impaired taste, salivation, lacrimation	6
VIII	Auditory, Vestibular, Vestibulochochlear (hearing and balance)	Hearing loss, tinnitus, vertigo, nystagmus, abnormal gait/balance, sway on Romberg	23, 27, 33
IX	Glossopharyngeal (swallow, sensation, pharynx, posterior 1/3 tongue, parotid salivary gland)	Depressed gag reflex and palate, dysphagia, uvula deviation, throat pain	10.2
X	Vagus (swallow, speech, parasympathetic to heart, lungs, GI tract)	Vocal cord paralysis, recurrent laryngeal nerve palsy, spasmodic dysphonia	7.1, 1.1
XI	Spinal Accessory (motor function neck/shoulder)	Sternocleidomastoid (SCM) weakness when turning head opposite, shoulder elevation, winging scapula	1.1

Number	Cranial Nerve Name	Nerve dysfunction on exam	Guideline Section in HD
XII	Hypoglossal (tongue movement)	Tongue deviation, atrophy, fasciculation	1.1
INO	Internuclear Opthalmoplegia (lesion of medial longitudinal fasciculus, CN III, CN VI)	Impaired adduction of ipsilateral eye with nystagmus of abducting eye	16, 21, 22
Horner Syndrome	Disruption of sympathetic innervation to eye and face	Ptosis, miosis (constricted pupil), facial anhidrosis (absence of sweating)	32.2, 11.3

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

General Guidelines – Modality (HD-1.2)

HD.GG.0001.2.A
v2.0.2024

- 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.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

General Guidelines – MRI Brain (HD-1.3)

HD.GG.0001.3.A
v2.0.2024

- MRI Brain with contrast (CPT® 70552) should not be ordered except to follow-up on a very recent non-contrast MRI Brain.
- After an MRI Brain without contrast (CPT® 70551), a follow up MRI brain with contrast (CPT® 70552) may be performed at the discretion of a neurologist, a neurosurgeon, or a neuro-ophthalmologist, or any provider in consultation with a neurologist, neurosurgeon, or neuro-ophthalmologist, and/or at the recommendation of the radiologist. [32]
- Gadolinium is relatively contraindicated in pregnancy, MRI Brain without contrast (CPT® 70551) is supported.[33]
- 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
v2.0.2024

- 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
v2.0.2024

- 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 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:[12,24]
  - 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 having failed medical therapy (see **Trigeminal Neuralgia and other Centrally Mediated Facial Pain Syndromes (HD-10.1)**)
  - 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)
  - Non-inflammatory 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)[12] (see **Eagle Syndrome (Neck-10.3)**)
  - NOTE: Evaluation of posterior circulation disease requires both neck and head MRA/CTA to visualize the entire vertebral-basilar system.

- MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496) is indicated 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) **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)[12,24]
    - 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)

HD.GG.0001.6.A
v2.0.2024

- 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)

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

General Guidelines – Other Imaging Situations (HD-1.7)

HD.GG.0001.7.C
v2.0.2024

- 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).
- Eagle Syndrome: See **Eagle Syndrome (Neck-10.3)**. See **General Guidelines – CT and MR Angiography (CTA and MRA) (HD-1.5)** for vascular imaging related to Eagle Syndrome[15]
- 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)** [26]

- 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

References (HD-1)

v2.0.2024

1. Grossman RI, Yousem DM. Neuroradiology. Philadelphia, PA: Mosby Elsevier; 2010.
2. Latchaw RE, Kucharczyk J, Moseley ME. Imaging of the nervous system: diagnostic and therapeutic applications. Philadelphia: Elsevier Mosby; 2005
3. Elan Lewis, Stephan Mayer, Lewis Rowland 13th edition. Merritt's neurology. Philadelphia, PA: Lippincott Williams & Wilkins; 2015
4. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck
5. Hornby PJ. Central neurocircuitry associated with emesis. The American Journal of Medicine. 2001;111(8):106-112. doi:10.1016/s0002-9343(01)00849-x
6. Shosha E, Dubey D, Palace J, et al. Area postrema syndrome. Neurology. 2018;91(17). doi:10.1212/wnl.0000000000006392
7. Singh P, Yoon SS, Kuo B. Nausea: a review of pathophysiology and therapeutics. Therapeutic Advances in Gastroenterology. 2015;9(1):98-112. doi:10.1177/1756283x15618131
8. Gutkowski P, Rot S, Fritsch M, Meier U, Gölz L, Lemcke J. Secondary deterioration in patients with normal pressure hydrocephalus after ventriculoperitoneal shunt placement: a proposed algorithm of treatment. Fluids and Barriers of the CNS. 2020;17(1). doi:10.1186/s12987-020-00180-w
9. Capitán L, Santamaría JG, Simon D, et al. Facial Gender Confirmation Surgery. Plastic and Reconstructive Surgery. 2020;145(4). doi:10.1097/prs.0000000000006686
10. Hatcher-Martin JM, et al. Telemedicine in Neurology. Telemedicine Work Group of the American Academy of Neurology update. Neurology® 2020;94:30-38. doi:10.1212/WNL.0000000000008708
11. ACR ASNR SPR Practice Parameter for the Performance and Interpretation of Cervicocerebral Computed Tomography Angiography (CTA) Revised 2020
12. ACR ASNR SPR Practice Parameter for the Performance and Interpretation of Magnetic Resonance Imaging (MRI) of the Brain. Revised 2019
13. Expert Panel on Neurologic Imaging:, Kennedy TA, Corey AS, et al. ACR Appropriateness Criteria® Orbits Vision and Visual Loss. J Am Coll Radiol. 2018;15(5S):S116-S131. doi:10.1016/j.jacr.2018.03.023
14. Ederies A, Demchuk A, Chia T, Gladstone DJ, Dowlatshahi D, Bendavit G, Wong K, Symons SP, Aviv RI. Postcontrast CT extravasation is associated with hematoma expansion in CTA spot negative patients. Stroke. 2009 May;40(5):1672-6. doi: 10.1161/STROKEAHA.108.541201
15. Chuang WC, Short JH, McKinney AM, Anker L, Knoll B, McKinney ZJ. Reversible left hemispheric ischemia secondary to carotid compression in Eagle syndrome: surgical and CT angiographic correlation. AJNR Am J Neuroradiol 2007;28:143-5
16. Chou DW, Tejani N, Kleinberger A, Shih C. Initial Facial Feminization Surgery Experience in a Multicenter Integrated Health Care System. Otolaryngology–Head and Neck Surgery. 2020;163(4):737-742. doi:10.1177/0194599820924635
17. Raffaini M, Perello R, Tremolada C, Agostini T. Evolution of Full Facial Feminization Surgery. Journal of Craniofacial Surgery. 2019;30(5):1419-1424. doi:10.1097/scs.0000000000005221
18. Eggerstedt M, Hong YS, Wakefield CJ, Westrick J, Smith RM, Revenaugh PC. Setbacks in Forehead Feminization Cranioplasty: A Systematic Review of Complications and Patient-Reported Outcomes. Aesthetic Plastic Surgery. 2020;44(3):743-749. doi:10.1007/s00266-020-01664-8
19. Spiegel JH. Facial Feminization for the Transgender Patient. Journal of Craniofacial Surgery. 2019;30(5):1399-1402. doi:10.1097/scs.0000000000005645
20. Callen AL, Badiee RK, Phelps A, Potigailo V, Wang E, Lee S, Talbott J, Glastonbury C, Pomerantz JH, Narvid J. Facial Feminization Surgery: Key CT Findings for Preoperative Planning and Postoperative Evaluation. AJR 2020 Dec 30 [published online].Accepted manuscript. doi:10.2214/AJR.20.25228
21. James SE, Herman JL, Rankin S, Keisling M, Mottet L, Anafi M. (2016). The Report of the 2015 U.S. Transgender Survey. Washington, DC: National Center for Transgender Equality
22. Spiegel JH. Gender affirming and aesthetic cranioplasty: what's new?Curr Opin Otolaryngol Head Neck Surg 2020, 28:201-205. doi:10.1097/MOO.0000000000000640
23. Pasternak JJ and Abcejo AS. Anesthesia and the brain after concussion. Curr Opin Anesthesiol 2020, 33:639–645. doi:10.1097/ACO.0000000000000906
24. PRACTICE PARAMETER 1 Cervicocerebral MRA. https://www.acr.org/-/media/ACR/Files/Practice-Parameters/cervicocerebralmra.pdf?la=en
25. ACR-ASNR-SIR-SNIS Practice Prameter for the Performance of Diagnostic Cervicocerebral Catheter Angiography in Adults. Revised 2021. (Resolution 4) https://www.acr.org/-/media/ACR/Files/Practice-Parameters/cervicocerebralcathangio.pdf?la=en

26. Expert Panel on Neurologic Imaging, Whitehead MT, Cardenas AM, et al. ACR Appropriateness Criteria® Headache. *J Am Coll Radiol*. 2019;16(11S):S364-S377. doi:10.1016/j.jacr.2019.05.030
27. Expert Panel on Neurological Imaging: Luttrull MD, Boulter DJ, et al. ACR Appropriateness Criteria® Acute Mental Status Change, Delirium, and New Onset Psychosis. J Am Coll Radiol. 2019;16(5S):S26-S37. doi:10.1016/j.jacr.2019.02.024
28. Expert Panel on Neurological Imaging: Ledbetter LN, Burns J, et al. ACR Appropriateness Criteria® Cerebrovascular Diseases-Aneurysm, Vascular Malformation, and Subarachnoid Hemorrhage. J Am Coll Radiol. 2021;18(11S):S283-S304. doi:10.1016/j.jacr.2021.08.012
29. Expert Panel on Neurological Imaging, Rath TJ, Policeni B, et al. ACR Appropriateness Criteria® Cranial Neuropathy: 2022 Update. *J Am Coll Radiol*. 2022;19(11S):S266-S303. doi:10.1016/j.jacr.2022.09.021
30. Badhey, A et al. Eagle syndrome: A comprehensive review. Clin Neurol Neurosurg. 2017 159:34-38. doi: 10.1016/j.clineuro.2017.04.021
31. Jamal B, Jalisi S, Grillone G. Surgical management of long-standing eagle's syndrome. Annals of Maxillofacial Surgery. 2017;7(2):232. doi:10.4103/ams.ams_53_17
32. Tillema JM. Imaging of Central Nervous System Demyelinating Disorders. *Continuum (Minneap Minn)*. 2023;29(1):292-323. doi:10.1212/CON.0000000000001246
33. American College of Radiology. ACR Manual on Contrast Media. Available at: https://www.acr.org/-/media/ACR/Files/Clinical-Resources/Contrast_Media.pdf. Accessed June 22, 2023.

Taste and Smell Disorders (HD-2)

Taste and Smell Disorders (HD-2.1)

HD.TS.0002.1.A
v2.0.2024

- MRI Brain without and with contrast (CPT® 70553) or without contrast (CPT® 70551) AND/OR MRI Orbits/Face/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 (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)

HD.TS.0002.2.A

v2.0.2024

1. Expert Panel on Neurological Imaging, Rath TJ, Policeni B, et al. ACR Appropriateness Criteria® Cranial Neuropathy: 2022 Update. J Am Coll Radiol. 2022;19(11S):S266-S303. doi:10.1016/j.jacr.2022.09.021
2. Devere R. Disorders of Taste and Smell. CONTINUUM: Lifelong Learning in Neurology. 2017;23(2):421-446. doi:10.1212/con.0000000000000463
3. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck
4. Politi LS, Salsano E, Grimaldi M. Magnetic Resonance Imaging Alteration of the Brain in a Patient With Coronavirus Disease 2019 (COVID-19) and Anosmia. *JAMA Neurology*. 2020. doi:10.1001/jamaneurol.2020.2125
5. Symptoms of Coronavirus. Centers for Disease Control and Prevention. https://www.cdc.gov/coronavirus/2019-ncov/symptoms-testing/symptoms.html. Published May 13, 2020
6. Soler ZM, Patel ZM, Turner JH, Holbrook EH. A primer on viral-associated olfactory loss in the era of COVID-19. International Forum of Allergy & *Rhinology*. 2020;10(7):814-820. doi:10.1002/alr.22578

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Ataxia (HD-3)

Ataxia (HD-3.1)

HD.AX.0003.1.A
v2.0.2024

- 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.[3]
- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) is indicated in all individuals with ataxia:
  - MRI Cervical without contrast or without and with contrast (CPT® 72141 or CPT® 72156) **AND/OR** MRI Thoracic without contrast or without and with contrast (CPT® 72146 or CPT® 72157) **AND/OR** MRI Lumbar Spine without contrast or without and with contrast (CPT® 72148 or CPT® 72158) may be added 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 Orbit/Temporal Bone without contrast (CPT® 70480) may be added if these symptoms are acute following head trauma, (see also: **Head Trauma (HD-13.1)**)
- If brain tumor is suspected, see **Primary Central Nervous System Tumors (ONC-2.1)** in the Oncology Imaging Guidelines.
- For suspected 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)

HD.AX.A
v2.0.2024

1. Expert Panel on Neurologic Imaging: Juliano AF, Policeni B, et al. ACR Appropriateness Criteria® Ataxia. *J Am Coll Radiol.* 2019;16(5S):S44-S56. doi:10.1016/j.jacr.2019.02.021
2. Graff-Radford NR, Jones DT. Normal Pressure Hydrocephalus. *CONTINUUM: Lifelong Learning in Neurology*. 2019;25(1):165-186. doi:10.1212/con.0000000000000689
3. Ashizawa T, Xia G. Ataxia. *CONTINUUM: Lifelong Learning in Neurology*. 2016;22(4):1208-1226. doi:10.1212/con.0000000000000362

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Mental Health Disorders and Mental Status Change (HD-4)

Autism Spectrum Disorders (HD-4.0)

HD.BD.0004.0.A
v2.0.2024

- This group of diagnoses, including Asperger syndrome, is classified as pervasive development disorders (PDD). These diagnoses are established on clinical criteria, and no imaging study can confirm the diagnosis.
- Comprehensive evaluation for autism might include history, physical exam, audiology evaluation, speech, language, and communication assessment, cognitive and behavioral assessments, and academic assessment.
  - MRI Brain without and with contrast (CPT® 70553) is indicated for **ANY** of the following:
    - New or worsening focal neurologic findings documented on a pertinent physical
    - Loss of developmental milestones and/or regression
  - PET imaging is considered not medically necessary in the evaluation of individuals with autism spectrum disorders.

Mental Health Related Disorders (HD-4.1)

HD.BD.0004.1.A
v2.0.2024

- Mental health diagnoses, to include Attention Deficit Hyperactivity Disorder (ADHD), do not routinely require advanced imaging.[12]
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) **OR** CT Head without contrast (CPT® 70450) may be indicated for the exceptions listed below:
  - 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 psychosis
    - 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 electroconvulsive therapy (ECT) treatment, the following may be utilized to screen for intracranial disease: MRI Brain without contrast (CPT® 70551) **OR** CT Head without contrast (CPT® 70450)
- Deep Brain Stimulation Therapy for psychiatric disorders is considered not medically necessary, except for medically refactory Obsessive Compulsive Disorder (OCD).[11]
  - Imaging supported prior to Deep Brain Stimulation (DBS) therapy for medically refractory Obsessive Compulsive Disorder (OCD):
    - MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) **AND/OR** unlisted CT procedure code (CPT® 76497)

Mental Status Change (HD-4.2)

HD.BD.0004.2.A
v2.0.2024

- An initial assessment should be performed prior to imaging requests. The initial assessment should include a history describing the onset, duration, and timeframe (constant versus intermittent or episodic nature) of symptoms.
- Bedside neurologic exam should include a mental status evaluation that provides a description of the level of alertness, other characteristics and/or cognitive testing.
- CT Head without contrast (CPT® 70450) **OR** MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553**)** is supported for **ANY** of the following[2]:
  - Acute or worsening (this includes repeat imaging) mental status change
  - Presence of any Red Flag, including:
    - Language, focal motor or sensory deficit (see **Stroke/TIA (HD-21.1)**)
    - Headache associated with acute mental status or other cognitive change. (see **Headaches with Red Flags (HD-11.2)**)
    - Presence of fever and/ or tachycardia (see **CNS and Head Infection (HD-14.1)**)
    - History of COVID-19 (see **Neuro-COVID-19 and Sars-CoV-2 Vaccines (HD 14.2)**)
    - History of hypertensive urgency associated with the mental status change (see **Stroke/TIA (HD-21.1)** and **Sudden Onset of Headache (HD 11.3)**)
    - Presence of coagulopathy or anticoagulant use (see **Abnormal Blood Clotting (HD 11.9)**)
    - Altered mental status in pregnancy and postpartum period (see **Pregnancy (HD 11.10)**)
    - History of significant antecedent trauma (see **Head Trauma (HD 13.1)**)
    - History of known underlying malignancy (see **Brain Metastases (ONC 31.3))**
    - Fluctuating alertness or consciousness
    - Glasgow Coma Scale (GCS) score of less than 15 (see **Head Trauma (HD 13.1)**) in the setting of antecedent trauma
  - Acute onset of mental status change or worsening symptoms (this includes repeat imaging) in the setting of **known intracranial process** (mass, recent hemorrhage, recent infarct, central nervous system infection, etc.)
  - MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) is supported with or without a previous CT Head.
  - CT Head without and with contrast (CPT 70470) is supported if clinical concern exists for initial diagnosis or progression of intracranial infection (such as abscesses or empyema), tumor, hemorrhage/stroke and/or inflammatory conditions.(See **CNS and Head Infection (HD 14.1)**, **Neuro-COVID-19 and Sars-CoV-2 Vaccines (HD 14.2)**, **Stroke/TIA (HD-21.1)** and/or **Brain Metastases (ONC 31.3)**).

- CT Head contrast as requested (CPT® 70450 **OR** CPT® 70460 **OR** CPT® 70470) is supported when:
  - MRI is contraindicated[13]
  - Clinical urgent setting when head imaging is otherwise supported.
- Condition specific imaging is listed in the associated guideline. These may include but are not limited to:
  - Seizure or suspected seizure. A description of events may include transient alteration of awareness, any neurologic deficit with rapid onset and offset of symptoms, episodic occurrence of symptoms, and/or abnormal rhythmic body movements. (See **Epilepsy/Seizures (HD 9.1)**)
  - Post-COVID syndrome/Long haul COVID/COVID-related neurocognitive syndrome, including associated brain fog, (See **Neuro-COVID-19 and Sars-CoV-2 Vaccines (HD 14.2)**)
  - History of significant antecedent head trauma or possible head trauma is present (See **Head Trauma (HD 13.1)**)
  - Concern for ischemic stroke, intracranial hemorrhage, or focal motor or sensory deficits (See **Stroke/TIA (HD-21.1)**)
  - Concern for mass (See **Low Grade Gliomas (ONC 2.2)**, **High Grade Gliomas (ONC 2.3)** and/or **Brain Metastases (ONC 31.3)**)
  - Suspected increased intracranial pressure (See **Papilledema/Pseudotumor Cerebri (HD 17.1)** and/or **Hydrocephalus Shunts (HD 11.14)**)
  - Hallucinations and/or delusions are present (See **Mental Health Related Disorders (HD-4.1)**)

Background and Supporting Information

This section refers to acute and subacute mental status change, generally implicating signs and symptoms occurring over minutes to days.

Acute mental status change or encephalopathy is characterized by changes in behavior or alertness, agitation, confusion, as opposed to chronic, progressive cognitive decline, such as dementia related disorders.

The terms delirium and psychosis are narrowly defined as follows:

- Delirium refers to acute onset of deficits in attention, awareness, and cognition that fluctuate in severity over time, often associated with psychomotor disturbance, altered sleep cycle, and emotional variability. These disturbances may be hyperactive (restlessness, agitation) or hypoactive (psychomotor retardation, lethargy). There may be accompanying fever, and autonomic symptoms (tachycardia, sweating) depending on underlying etiology.
- Psychosis refers to a disorder of impaired reality testing characterized by the presence of hallucinations and/ or delusions, or both without (without insight into their pathologic nature). This may be associated with disorganized behavior, thought blocking, illogicality, tangentiality, perseveration, neologisms.

The purpose of the initial assessment is to define the category of the etiology. These may include: toxic/ metabolic (hypoglycemic, drug exposures), structural (trauma, stroke, hypoxic-ischemic, hydrocephalus, tumor), paroxysmal (seizure, psychiatric), inflammatory (infectious, autoimmune).

Of note even a mild or trivial, acute insult superimposed upon a chronic pathophysiologic process may cause acute mental status change, and head imaging may or may not be necessary, depending on the provider's pretest suspicion of a new significant diagnosis.

Non response to adequate medication trials may include, but is not limited to, implantation of Vagal Nerve Stimulator (VNS), which is FDA approved for treatment of depression.

References (HD-4)

v2.0.2024

1. Uzelac A. Imaging of Altered Mental Status. *Radiologic Clinics of North America*. 2020;58(1):187-197. doi:10.1016/j.rcl.2019.08.002
2. Expert Panel on Neurological Imaging: Luttrull MD, Boulter DJ, et al. ACR Appropriateness Criteria® Acute Mental Status Change, Delirium, and New Onset Psychosis. J Am Coll Radiol. 2019;16(5S):S26-S37. doi:10.1016/j.jacr.2019.02.024
3. Andrea S, Papirny M, Raedler T. Brain Imaging in Adolescents and Young Adults With First-Episode Psychosis. *The Journal of Clinical Psychiatry*. 2019;80(6). doi:10.4088/jcp.18m12665
4. Bridgemohan CF.Chapter 54: Autism spectrum disorder. In: Kliegman RM, St. Geme JW III, Blum NJ, Shah SS, Tasker RC, Wilson KM. eds. *Nelson Textbook of Pediatrics* 21st ed. 2020: 294-302
5. Baker E, and Jeste SS. Diagnosis and Management of Autism Spectrum Disorder in the Era of Genomics. *Pediatric Clinics of North America*. 2015;62(3):607-618. doi:10.1016/j.pcl.2015.03.003
6. Zürcher NR, Bhanot A, McDougle CJ, Hooker JM. A systematic review of molecular imaging (PET and SPECT) in autism spectrum disorder: Current state and future research opportunities. *Neuroscience & Biobehavioral Reviews*. 2015;52:56-73. doi:10.1016/j.neubiorev.2015.02.002.
7. Julayanont P, Suryadevara U. Psychosis. *Continuum (Minneap Minn)*. 2021;27(6):1682-1711. doi:10.1212/CON.0000000000001013
8. Keepers GA. American Psychiatric Association.The American Psychiatric Association practice guideline for the treatment of patients with schizophrenia / Guideline Writing Group, Systematic Review Group, Committee on Practice Guidelines.3rd edition. American Psychiatric Association; 2021
9. Rapinesi C, Kotzalidis GD, Ferracuti S, Sani G, Girardi P, Del Casale A. Brain Stimulation in Obsessive-Compulsive Disorder (OCD): A Systematic Review. Curr Neuropharmacol. 2019;17(8):787-807. doi: 10.2174/1570159X17666190409142555. PMID: 30963971; PMCID: PMC7059162.
10. Ali SA, Mathur N, Malhotra AK, Braga RJ. Electroconvulsive Therapy and Schizophrenia: A Systematic Review. Mol Neuropsychiatry. 2019 Apr;5(2):75-83. doi: 10.1159/000497376. Epub 2019 Apr 2. PMID: 31192220; PMCID: PMC6528094.
11. Staudt MD, Pouratian N, Miller JP, et al. Congress of Neurological Surgeons Systematic Review and Evidence-Based Guidelines for Deep Brain Stimulations for Obsessive-Compulsive Disorder: Update of the 2014 Guidelines. Neurosurgery. 2021;88(4):710-712. doi:10.1093/neuros/nyaa596
12. Pereira-Sanchez V, Castellanos FX. Neuroimaging in attention-deficit/hyperactivity disorder. Curr Opin Psychiatry. 2021;34(2):105-111. doi:10.1097/YCO.0000000000000669
13. Expert Panel on MR Safety, Kanal E, Barkovich AJ, et al. ACR guidance document on MR safe practices: 2013. *J Magn Reson Imaging*. 2013;37(3):501-530. doi:10.1002/jmri.24011

Chiari and Skull-Base Malformations (HD-5)

Chiari Malformations (HD-5.1)

HD.CM.0005.1.A
v2.0.2024

Indication	Supported Imaging
Initial Evaluation for suspected or known Chiari malformations:	• MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRI Cervical Spine without contrast (CPT® 72141) or MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) or MRI Thoracic Spine without and with contrast (CPT® 72157) **AND/OR** • MRI Lumbar Spine without contrast (CPT® 72148) or MRI Lumbar Spine without and with contrast (CPT® 72158)

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Indication	Supported Imaging
Repeat imaging for one of the following: • New or worsening signs or symptoms • Surgical procedure is actively being considered • At the discretion of or in consultation with a neurologist and/or neurosurgeon coordinating the individual's care	• MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRI Cervical Spine without contrast (CPT® 72141) or MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) or MRI Thoracic Spine without and with contrast (CPT® 72157) **AND/OR** • MRI Lumbar Spine without contrast (CPT® 72148) or MRI Lumbar Spine without and with contrast (CPT® 72158)

- Familial screening is NOT indicated for Chiari Malformations.
- For CSF flow imaging (see **CSF Flow Imaging (HD-24.4)**)

Background and Supporting Information

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

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

Chiari III malformations include cerebellar herniation into a high cervical myelomeningocele. Chiari IV 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 I malformations and known syringomyelia or hydromyelia is highly individualized.

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

HD.CM.0005.2.A
v2.0.2024

- See **Chiari Malformations (HD-5.1)**

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Chiari III and IV Malformations (HD-5.3)

HD.CM.0005.3.A
v2.0.2024

- See **Chiari Malformations (HD-5.1)**

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Basilar Impression/Basilar Invagination (HD-5.4)

HD.CM.0005.4.A
v2.0.2024

Imaging indications for suspected or known Basilar Impression or Basilar Invagination:

- MRI Brain (CPT® 70551) **AND/OR** MRI Cervical Spine (CPT® 72141) without contrast
- If surgery is being considered, CT Head (CPT® 70450) **AND/OR** CT Cervical Spine (CPT® 72125) without contrast are also indicated **AND/OR** MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR** CTA Head (CPT® 70496) **AND/OR** MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) **OR** CTA Neck (CPT® 70498).[14]
- One-time screening of first-degree relatives with MRI Brain without contrast (CPT® 70551) is supported.

Background and Supporting Information

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.

Basilar invagination is an abnormality at the craniovertebral junction, either congenital or degenerative, resulting in the odontoid prolapsing into the already limited space of the foramen magnum. It is commonly associated with conditions such as Chiari malformation, syringomyelia, and Klippel-Feil syndrome.[12]

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Platybasia (HD-5.5)

HD.CM.0005.5.A
v2.0.2024

Imaging indications for suspected or known Platybasia:

- MRI Brain without contrast (CPT®70551) or CT Head without contrast (CPT®70450)
- If surgery is being considered,
  - CT Head (CPT®70450) **AND/OR**
  - CT Cervical Spine without contrast (CPT®72125) **AND/OR**
  - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR**
  - CTA Head (CPT® 70496) **AND/OR**
  - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) **OR**
  - CTA Neck (CPT® 70498)[14]

Background and Supporting Information

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

References (HD-5)

v2.0.2024

1. Strahle J, Muraszko KM, Kapurch J, et al. Chiari malformation Type I and syrinx in children undergoing magnetic resonance imaging. *J Neurosurg Pediatr*. 2011 Aug; 8 (2): 205-213
2. Strahle J, Muraszko KM, Kapurch J, et al. Natural history of Chiari malformation Type I following decision for conservative treatment. *J Neurosurg Pediatr.* 2011 Aug; 8 (2): 214-221
3. Strahle J, Muraszko KM, Garton HJL, et al. Syrinx location and size according to etiology: identification of Chiari-associated syrinx. *J Neurosurg Pediatr*. 2015 July; 16 (1): 21-9 Epub 2015 Apr 3
4. Strahle J, Smith BW, Martinez M, et al. The association between Chiari malformation Type I, spinal syrinx, and scoliosis. *J Neurosurg Pediatr*. 2015 Jun; 15 (6): 607-611
5. Victorio MC, Khoury CK. Headache and Chiari I Malformation in Children and Adolescents. *Seminars in Pediatric Neurology*. 2016;23(1):35-39
6. Radic JAE, Cochrane DD. Choosing Wisely Canada: Pediatric Neurosurgery Recommendations. *Paediatrics & Child Health*. 2018;23(6):383-387. doi:10.1093/pch/pxy012
7. Smoker WRK and Khanna G. Imaging the craniocervical junction. Childs Nerv Syst. 2008 Oct; 24 (10): 1123-1145
8. Kinsman SL and Johnston MV. Congenital anomalies of the central nervous system. Nelson Textbook of Pediatrics, Chapter 609. eds Kliegman RM, St. Geme JW III, Blum NJ, Shah SS, Tasker RC, Wilson KM. 21st edition. 2020, pp 3063-3082
9. Dantas FLR, Dantas F, Caires AC, Botelho RV. Natural History and Conservative Treatment Options in Chiari Malformation Type I in Adults: A Literature Update. Cureus. 2020;12(12):e12050. Published 2020 Dec 13. doi:10.7759/cureus.12050
10. Holly LT, Batzdorf U. Chiari malformation and syringomyelia. J Neurosurg Spine. 2019;31(5):619-628. doi:10.3171/2019.7.SPINE181139
11. Rosenblum JS, Pomeraniec IJ, Heiss JD. Chiari Malformation (Update on Diagnosis and Treatment). Neurol Clin. 2022;40(2):297-307. doi:10.1016/j.ncl.2021.11.007
12. Donnally III CJ, Munakomi S, Varacallo M. Basilar Invagination. [Updated 2022 Nov 19]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK448153/
13. Brito JNPO, Santos BAD, Nascimento IF, Martins LA, Tavares CB. Basilar invagination associated with chiari malformation type I: A literature review. Clinics (Sao Paulo). 2019;74:e653. doi:10.6061/clinics/2019/e653
14. Pinter NK, McVige J, Mechtler L. Basilar Invagination, Basilar Impression, and Platybasia: Clinical and Imaging Aspects. Curr Pain Headache Rep. 2016;20(8):49. doi:10.1007/s11916-016-0580-x

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

Facial Palsy (HD-6.1)

HD.FP.0006.1.A
v2.0.2024

- 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/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[2]:
  - 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, (see also: **Autoimmune/ Paraneoplastic Encephalitis & NeuroInflammatory Disorders (HD-14.3)**)
- CT Orbit/Temporal Bone without contrast (CPT® 70480), in the presence of red flags, to assess osseous integrity of the temporal bone, to characterize fractures, pre-surgical anatomy, inflammatory middle ear disease, bone tumor, facial canal foraminal expansion and/or bone erosion.[2]
- CT Orbit/Temporal Bone with contrast (CPT® 70481), in the presence of red flags, for suspected tumors and/or infection.[2]
- CT Maxillofacial without contrast (CPT® 70486) to assess bony facial nerve canal **OR** with contrast (CPT® 70487) when infection or tumor are suspected, if requested per institutional protocol.[2]
- MRA Head without contrast (CPT® 70544), with contrast (CPT® 70545), or without and with contrast (CPT® 70546) **AND/OR** MRA Neck without contrast (CPT® 70547), with contrast (CPT® 70548), or without and with contrast (CPT® 70549) **OR** CTA Head (CPT® 70496) **AND/OR** CTA Neck (CPT® 70498) for clinically suspected stroke.[2] (see also: **General Guidelines- CT and MR Angiography (CTA and MRA) (HD-1.5)** and **Stroke/TIA (HD-21.1)**

Background and Supporting Information

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.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Hemifacial Spasm (HD-6.2)

HD.FP.0006.2.A
v2.0.2024

- For hemifacial spasm, facial synkinesis, or blepharospasm:
  - 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

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

References (HD-6)

v2.0.2024

1. Baugh RF, Basura GJ, Ishii LE, et al. Clinical practice guideline. Bell's Palsy Executive Summary. *Otolaryngology–Head and Neck Surgery*. 2013;149(5):656-663. doi:10.1177/0194599813506835
2. Expert Panel on Neurological Imaging, Rath TJ, Policeni B, et al. ACR Appropriateness Criteria® Cranial Neuropathy: 2022 Update. J Am Coll Radiol. 2022;19(11S):S266-S303. doi:10.1016/j.jacr.2022.09.021
3. Yaltho TC, Jankovic J. The many faces of hemifacial spasm: Differential diagnosis of unilateral facial spasms. Movement Disorders. 2011;26(9):1582-1592. doi:10.1002/mds.23692
4. Reich SG. Bell's Palsy. CONTINUUM: Lifelong Learning in Neurology. 2017;23(2):447-466. doi:10.1212/con.0000000000000447
5. Stern BJ, Royal W, Gelfand JM, et al. Definition and Consensus Diagnostic Criteria for Neurosarcoidosis. JAMA Neurology. 2018;75(12):1546. doi:10.1001/jamaneurol.2018.2295

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

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

HD.RL.0007.1.A
v2.0.2024

- See **Recurrent Laryngeal Nerve Palsy in Neck-7.1**

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Dementia (HD-8)

Dementia (HD-8.1)

HD.DM.0008.1.C
v2.0.2024

- For acute mental status change, see **Mental Status Change (HD-4.2)** and **Stroke/ TIA (HD-21.1)**
- For members being considered for amyloid reducing medications for the treatment of Mild Cognitive Impairment (MCI) due to Alzheimer's disease or mild dementia due to Alzheimer's disease see **Imaging related to Alzheimer's Treatment and Amyloid Reduction Medications (HD-8.5)**.
- 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, the St. Louis University Mental Status (SLUMS) with score <21, or the Eight-item Informant Interview to Differentiate Aging and Dementia (AD8) Dementia Score > 2[31].
    - Presumptive causes or etiology/ies of dementia
      - Cannot occur exclusively during bouts of delirium
      - Cannot be explained by another mental disorder
- For the evaluation of Normal Pressure Hydrocephalus, see **Normal Pressure Hydrocephalus (HD-8.4)**).
- Quantitative Magnetic Resonance Image (MRI) Analysis of the Brain
  - Volumetric analysis of the temporal lobes and hippocampus or Neuro Quant may be ordered as 3D rendering (CPT® 76377) or quantitative analysis of the brain (CPT® 0865T or CPT® 0866T). These studies lack sufficient specificity and sensitivity to be clinically useful in the evaluation or follow up of individuals with dementia. Their use is limited to research studies and are otherwise considered to be not medically necessary in routine clinical practice.

Dementia - PET (HD-8.2)

HD.DM.0008.2.A
v2.0.2024

- 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, the St. Louis University Mental Status (SLUMS) with score <21 or the Eight-item Informant Interview to Differentiate Aging and Dementia (AD8) Dementia Score > 2[31].
  - Results of any structural imaging (MRI or CT Head) performed.
  - 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 not medically necessary 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.

  - 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
  - Amyloid Brain PET (CPT® 78811 or CPT® 78814) imaging is only indicated for treatment with amyloid-reducing medications (see **Imaging Related to Alzheimer's Treatment and Amyloid Reduction Medications (HD-8.5)**).
  - Otherwise, these studies are considered not medically necessary in the diagnosis and/or treatment of Alzheimer's disease and in differentiating between Alzheimer's disease and other neurodegenerative/neurologic disorders.
- For Cerebral Amyloid Angiopathy, see **Stroke/TIA (HD-21.1)**
- FDG-PET(CPT® 78608)/MRI Brain without contrast (CPT® 70551) OR MRI Brain without and with contrast (CPT® 70553) imaging may be considered on a case by case basis for those imaging centers that will utilize FDG-PET/MRI during an initial evaluation (instead of MRI alone) and who also have a standardization of imaging protocol.[27, 28, 29, 30]

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.
- Health plans may have specific criteria that differ in their coverage policies

- Recent research has examined the utility of PET/MRI for evaluation of patients with Dementia. Due to the prolonged accusation time, motion during a PET may lead to artifacts such as blurring of the images. Use of co-registration of PET with MRI can lead to better PET assessment especially with quantitative measurements[27,30] . Utilization of PET/MRI provides greater confidence in imaging reading by permitting greater structural correlation. A recent study compared FDG-PET/CT and FDG-PET/MRI in a memory disorders clinic. This study identified more patient with cerebrovascular disease (stroke) and better cortical atrophy characterization [28]. The authors found that PET/MRI provided significant improvement in diagnosis and management of patients in which dementia is a consideration. Finally a Canadian study suggested that FDG-PET/MRI is "financial justifiable"[29].

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

HD.DM.0008.3.A
v2.0.2024

- 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, the St. Louis University Mental Status (SLUMS) with score <21, or the Eight-item

Informant Interview to Differentiate Aging and Dementia (AD8) Dementia Score > 2[31].

- Results of any structural imaging (MRI or CT Head) performed

- 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
v2.0.2024

- 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[24,25,26]
- 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, the St. Louis University Mental Status (SLUMS) with score <21, or the Eight-item Informant Interview to Differentiate Aging and Dementia (AD8) Dementia Score > 2[31].
  - 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.

Imaging Related to Alzheimer's Treatment and Amyloid Reduction Medications (HD-8.5)

HD.DM.0008.5.A
v2.0.2024

Health plans may have specific criteria that differ in their coverage policies.

A pertinent clinical evaluation including a detailed history, mental status testing results, and appropriate laboratory studies should be performed prior to considering treatment with amyloid reduction medications.

Medical records should be provided that support a clinical diagnosis of Mild Cognitive Impairment (MCI) due to Alzheimer's Dementia (AD) or early Alzheimer's Dementia (AD). Other conditions such as Dementia with Lewy Bodies (DLB), Frontotemporal Dementia (FTD), vascular dementia, pseudodementia due to mood disorder, vitamin B12 deficiency, untreated thyroid disease, traumatic brain injury, and/or encephalopathy, have been excluded.

Results of bedside testig and/or neuropsychological testing can be performed when history and mental status examination cannot provide a confident diagnosis.

Prior to treatment initiation, when ALL criteria are met:

- Patient is ≥ 50 years of age and ≤ 85 years of age **AND**
- A clinical diagnosis of Mild Cognitive Impairment (MCI) due to Alzheimer's Dementia (AD) or early Alzheimer's Dementia (AD) **AND**
- Qualifying test scores include Mini-Mental Status Exam (MMSE) with score ≥ 22, Clinical Dementia Rating global socre of 0.5 or 1.0, Clinical Dementia Rating-Sum of Boxes (CDR-SB) ≥ 0.5 and/or a Memory Box score of 0.5 or greater **AND**
- Patient has no history of brain hemorrhage, bleeding disorder or recent history (within 12 months) of stroke or transient ischemic attachks or any history of seizures **AND**
- Patient is not taking anticoagulant or antiplatelet agents (except aspirin for prevention of cardiovascular or thromboembolic events) **AND**
- Not currently taking another amyloid reducing drug **AND**
- The medication is prescribed by a neurologist

Lecenamab (Leqembi)

Indication	Supported Imaging
Consideration of Lecanemab (Leqembi) therapy	Baseline MRI Brain (*within 3 months of medication initiation*) • MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **AND/OR** • Amyloid PET Brain (CPT® 78811 or CPT® 78814)
On Lecanemab therapy prior to 5th, 7th and 14th infusions	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)
Follow up while on Lecanemab therapy with radiographically observed Amyloid-Related Imaging Abnormality (ARIA) See **Background and Supporting Information**	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) per the treating neurologist
Neurologic signs and/or symptoms occurring while on treatment with Lecanemab	• CT Head without contrast (CPT® 70450) **OR** • MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) A follow-up MRI Brain is appropriate after a CT Head if requested
Post-treatment imaging at 18 months	• Amyloid PET Brain (CPT® 78811 or CPT® 78814)

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Aducanumab (Aduhelm)

Indication	Supported Imaging
For consideration of Aducanumab (Aduhelm) therapy	Baseline MRI Brain (*within 3 months of medication initiation*) • MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **AND/OR** • Amyloid PET Brain (CPT® 78811 or CPT® 78814)
On Aducanumab therapy prior to the 5th, 7th, 9th, and 12th infusions	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)
Follow up while on therapy with radiographically observed Amyloid-Related Imaging Abnormality (ARIA) See **Background and Supporting Information**	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) per the treating neurologist
Neurologic signs and/or symptoms occurring while on treatment with Aducanumab	• CT Head without contrast (CPT® 70450) **OR** • MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) A follow-up MRI Brain is appropriate after a CT Head if requested
Post-treatment imaging at 18 months	• Amyloid PET Brain (CPT® 78811 OR CPT® 78814)

Background and Supporting Information

Amyloid reduction medications are indicated for the treatment of Mild Cognitive Impairment (MCI) due to Alzheimer's disease and mild, early stage Alzheimer's disease[32,33].

These medications are monoclonal antibodies that selectively bind to aggregated forms of beta amyloid. The accumulation of amyloid plaques in the brain is a defining pathophysiologic feature of Alzheimer's disease. In clinical trials, these medications reduce amyloid beta plaque compared with placebo[32,33].

Amyloid related imaging abnormalities (ARIA) have been caused by these medications. ARIA usually occurs early in treatment and may be asymptomatic although serious and life-threatening events may occur. Screening MRI brain prior to treatment initiation and periodic monitoring during treatment is recommended. For moderate to severe ARIA, treatment may be suspended. Once ARIA is identified on a brain MRI, follow up MRIs are indicated to assess for radiographic resolution and/or symptom resolution with the imaging time frame determined by the treating physician. Resumption of dosing is guided by clinical judgment[32,33].

ARIA may be further characterized as ARIA with edema (ARIA-E) or ARIA with hemosiderin (ARIA-H). ARIA-E presents on MRI as brain edema or sulcal effusions. ARIA-H includes microhemorrhage and superficial siderosis. ARIA-E and ARIA-H may occur simultaneously[32,33].

Although ARIA is usually asymptomatic, symptoms associated with ARIA include headache, confusion, visual changes, dizziness, nausea, aphasia, weakness, gait difficulty and seizures, including status epilepticus. Focal neurologic deficits may also occur[32,33]. The risk of ARIA is increased in apolipoprotein E Ɛ4 (ApoE Ɛ4) homozygotes[33].

References (HD-8)

v2.0.2024

1. McKhann GM, Knopman DS, Chertkow H, et al. The diagnosis of dementia due to Alzheimer's disease: Recommendations from the National Institute on Aging-Alzheimer's Association workgroups on diagnostic guidelines for Alzheimer's disease. Alzheimer's & Dementia. 2011;7(3):263-269. doi:10.1016/j.jalz.2011.03.005
2. Lewis SL. Dementia Untangled. CONTINUUM: Lifelong Learning in Neurology. 2019;25(1):12-13. doi:10.1212/01.con.0000553293.87198.a4
3. Decision Memo for Positron Emission Tomography (FDG) for Alzheimer's Disease/Dementia (CAG-00088N). CMS.gov. Centers for Medicare & Medicaid Services. https://www.cms.gov/medicare-coverage-database/details/nca-decision-memo.aspx?NCAId=64&fromdb=true
4. Wippold FJ 2nd, Brown DC, Broderick DF, et al. ACR Appropriateness Criteria Dementia and Movement Disorders. J Am Coll Radiol. 2015;12(1):19-28. doi:10.1016/j.jacr.2014.09.025
5. Knopman DS, DeKosky ST, Cummings JL, et al. Practice parameter: Diagnosis of dementia (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology. Neurology. 2001;56(9):1143-1153. doi:10.1212/wnl.56.9.1143
6. Johnson KA, Minoshima S, Bohnen NI, et al. Appropriate use criteria for amyloid PET: A report of the Amyloid Imaging Task Force, the Society of Nuclear Medicine and Molecular Imaging, and the Alzheimer's Association. Alzheimer's & Dementia. 2013;9(1):E1-E16. doi:10.1016/j.jalz.2013.01.002
7. Decision Memo for Positron Emission Tomography (FDG) and Other Neuroimaging Devices for Suspected Dementia (CAG-00088R). CMS.gov. Centers for Medicare & Medicaid Services. https://www.cms.gov/medicare-coverage-database/details/nca-decision-memo.aspx?NCAId=104&NcaName=Positron+Emission+Tomography+(FDG)+and+Other+Neuroimaging+Devices+for+Suspected+Dementia+(1st+Recon)&bc=AiAAAAAAEAAA&
8. NCD for FDG PET for Dementia and Neurodegenerative Diseases (220.6.13), Effective date 4/3/2009, Implementation date 10/30/2009. http://www.cms.gov/medicare-coverage-database/details/ncd-details.aspx?NCDId=288&ncdver=3&bc=BAABAAAAAAAA&
9. Rabinovici GD, Gatsonis C, Apgar C, et al. Association of Amyloid Positron Emission Tomography With Subsequent Change in Clinical Management Among Medicare Beneficiaries With Mild Cognitive Impairment or Dementia. *Jama*. 2019;321(13):1286-1294. doi:10.1001/jama.2019.2000
10. Subramaniam RM, Frey KA, Hunt CH, et al. ACR–ACNM Practice Parameter for the Performance of Dopamine Transporter (DaT) Single Photon Emission Computed Tomography (SPECT) Imaging for Movement Disorders. Clinical Nuclear Medicine. 2017;42(11):847-852. doi:10.1097/rlu.0000000000001815
11. Graff-Radford NR, Jones DT. Normal Pressure Hydrocephalus. CONTINUUM: Lifelong Learning in Neurology. 2019;25(1):165-186. doi:10.1212/con.0000000000000689
12. Tartaglia MC, Rosen HJ, Miller BL. Neuroimaging in Dementia. Neurotherapeutics. 2011;8(1):82-92. doi:10.1007/s13311-010-0012-2
13. ACR ACNM ASNR SNMMI Practice Parameter for Brain PET-CT Imaging in Dementia. 2020
14. Consensus Recommendations for the Postmortem Diagnosis of Alzheimer's Disease. Neurobiology of Aging. 1997;18(4):S1-S2. doi:10.1016/s0197-4580(97)00057-2
15. Approaches and Tools for Primary Care Providers Developed by the GSA Workgroup on Cognitive Impairment Detection and Earlier Diagnosis. https://www.geron.org/images/gsa/kaer/gsa-kaer-toolkit.pdf
16. Lombardi G, Crescioli G, Cavedo E, et al. Structural magnetic resonance imaging for the early diagnosis of dementia due to Alzheimer's disease in people with mild cognitive impairment. Cochrane Database of Systematic Reviews. Published online March 2, 2020. doi:10.1002/14651858.cd009628.pub2
17. Yousaf T, Dervenoulas G, Valkimadi P-E, Politis M. Neuroimaging in Lewy body dementia. Journal of Neurology. 2019;266(1):1-26. doi:10.1007/s00415-018-8892-x
18. Goto H, Ishii K, Uemura T, et al. Differential Diagnosis of Dementia with Lewy Bodies and Alzheimer Disease Using Combined MR Imaging and Brain Perfusion Single-Photon Emission Tomography. American Journal of Neuroradiology. 2010;31(4):720-725. doi:10.3174/ajnr.a1926
19. McCleery J, Morgan S, Bradley KM, Noel-Storr AH, Ansorge O, Hyde C. Dopamine transporter imaging for the diagnosis of dementia with Lewy bodies. Cochrane Database of Systematic Reviews. Published online January 30, 2015. doi:10.1002/14651858.cd010633.pub2
20. Armstrong MJ. Lewy Body Dementias. CONTINUUM: Lifelong Learning in Neurology. 2019;25(1):128-146. doi:10.1212/con.0000000000000685
21. American Psychiatric Association. Diagnostic and Statistical Manual of Mental Disorders. 5th ed. American Psychiatric Association; 2013
22. Diagnosis of Dementia. www.aan.com. https://www.aan.com/Guidelines/home/GuidelineDetail/42

23. Zukotynski K, Kuo PH, Mikulis D, et al. PET/CT of Dementia. American Journal of Roentgenology. 2018;211(2):246-259. doi:10.2214/ajr.18.19822
24. Nakajima M, Yamada S, Miyajima M, et al. Guidelines for Management of Idiopathic Normal Pressure Hydrocephalus (Third Edition): Endorsed by the Japanese Society of Normal Pressure Hydrocephalus. Neurologia medico-chirurgica. 2021;61(2):63-97. doi:10.2176/nmc.st.2020-0292
25. Capone PM, Bertelson JA, Ajtai B. Neuroimaging of Normal Pressure Hydrocephalus and Hydrocephalus. Neurologic Clinics. 2020;38(1):171-183. doi:10.1016/j.ncl.2019.09.003
26. Park HY, Park CR, Suh CH, Kim MJ, Shim WH, Kim SJ. Prognostic Utility of Disproportionately Enlarged Subarachnoid Space Hydrocephalus in Idiopathic Normal Pressure Hydrocephalus Treated with Ventriculoperitoneal Shunt Surgery: A Systematic Review and Meta-analysis. American Journal of Neuroradiology. 2021;42(8):1429-1436. doi:10.3174/ajnr.a7168
27. Chen KT, Salcedo S, Chonde DB, et al. MR-assisted PET motion correction in simultaneous PET/MRI studies of dementia subjects. J Magn Reson Imaging. 2018;48(5):1288-1296. doi:10.1002/jmri.26000
28. Kaltoft NS, Marner L, Larsen VA, Hasselbalch SG, Law I, Henriksen OM. Hybrid FDG PET/MRI vs. FDG PET and CT in patients with suspected dementia - A comparison of diagnostic yield and propagated influence on clinical diagnosis and patient management. PLoS One. 2019;14(5):e0216409. Published 2019 May 2. doi:10.1371/journal.pone.0216409
29. Prato FS, Pavlosky WF, Foster SC, Thiessen JD, Beaujot RP. Screening for Dementia Caused by Modifiable Lifestyle Choices Using Hybrid PET/MRI. J Alzheimers Dis Rep. 2019;3(1):31-45. Published 2019 Feb 4. doi:10.3233/ADR-180098
30. Patel KP, Wymer DT, Bhatia VK, Duara R, Rajadhyaksha CD. Multimodality Imaging of Dementia: Clinical Importance and Role of Integrated Anatomic and Molecular Imaging. Radiographics. 2020;40(1):200-222. doi:10.1148/rg.2020190070
31. Svensson A, Granvik E, Sjögren Forss K. Performance of the Eight-item Informant Interview to Differentiate Aging and Dementia within a context similar to the Swedish primary healthcare sector: a systematic review of diagnostic test accuracy studies. Scand J Prim Health Care. 2020;38(4):454-463. doi:10.1080/02813432.2020.1844370
32. Aduhelm® intravenous infusion [prescribing information]. Cambridge, MA: Biogen; August 2023.
33. Leqembi® intravenous infusion [prescribing information]. Nutley, NJ: Eisai; July 2023.
34. Cummings J, Apostolova L, Rabinovici GD, et al. Lecanemab: Appropriate Use Recommendations.*J Prev Alzheimers Dis*. 2023;10(3):362-377. doi:10.14283/jpad.2023.30
35. Swanson CJ, Zhang Y, Dhadda S, et al. A randomized, double-blind, phase 2b proof-of-concept clinical trial in early Alzheimer's disease with lecanemab, an anti-Aβ protofibril antibody [published correction appears in Alzheimers Res Ther. 2022 May 21;14(1):70].*Alzheimers Res Ther*. 2021;13(1):80. Published 2021 Apr 17. doi:10.1186/s13195-021-00813-8

Epilepsy/Seizures (HD-9)

Epilepsy/Seizures (HD-9.1)

HD.EP.0009.1.C
v2.0.2024

- 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 was performed for an initial evaluation for new onset seizure, MRI (as described above) is indicated for additional evaluation
  - Follow-up MRI Brain with “Epilepsy Protocol” is supported.
- Repeat imaging at discretion of the neurologist
- MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) **OR** CT Head without contrast (CPT® 70450)[1]
- 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) (CPT®70450, CPT®70460 **OR** CPT®70470) when:
  - MRI is contraindicated
  - Request is urgent
- For Seizure and/or Altered Mental Status associated with Head Trauma, see **Head Trauma (HD-13.1)**
- 3D T1 and/or FLAIR sequences are useful in improving lesion detection for the diagnosis and monitoring of epilepsy. 3D T1 and FLAIR sequences do not require an additional CPT® for 3D rendering (CPT® 76377).[12]

Perioperative Evaluations for Drug-Resistant Epilepsy (HD-9.2)

HD.EP.0009.2.C
v2.0.2024

- The following requests are supported for consideration of potential surgery:
  - MRI Brain without contrast (CPT® 70551) OR MRI Brain with and without (CPT® 70553)
    - Follow-up MRI Brain after a previous routine study if performed with special "Epilepsy Protocol" (typically 3T or 7T magnet, thin sections with angled slices through hippocampus and temporal lobes)
  - 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
    - PET/MRI is MRI Brain without contrast (CPT® 70551) **OR** MRI Brain with and without (CPT® 70553) co-registered **WITH** FDG-PET Brain (CPT® 78608) and is supported for pre-surgical evaluation of refractory seizure when requested by neurosurgeon or neurologist or any provider in consultation with a neurosurgeon or neurologist[25,27]
  - Ictal SPECT (CPT® 78803)
  - Functional MRI (fMRI) (CPT® 70555 or CPT® 70554)
    - If MRA Head (CPT® 70544) is indicated but Functional MRI (CPT® 70554 or CPT® 70555) was erroneously ordered, then CPT® 70544 may be substituted when appropriate. See **Functional MRI (fMRI) (HD-24.2)**
  - MRI Brain without contrast (CPT® 70551) **OR** MRI Brain with and without (CPT® 70553)
    - Indicated if co-registered with Magnetoencephalography (MEG)[1]
  - 3D rendering CPT® 76377 (3D rendering requiring image post-processing on an independent workstation) is not necessary for epilepsy surgery alone, since 3D rendering can be obtained as part of the MRI Brain epilepsy protocol, unless complicated surgical repair considerations involving craniotomy are required.[12]
- When non-invasive EEG monitoring is insufficient, intracranial monitoring with stereo-EEG or grids/strips and depth 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 neurologist or any provider in consultation with neurosurgeon or neurologist.

- 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

- Magnetoencephalography (MEG) plays an important role in clarifying the significance of abnormalities seen on both structural and functional imaging, for the purpose of epileptogenic zone localization for surgical planning. When used in conjunction with other techniques, MEG plays a major role in the non-invasive epilepsy surgery evaluation. Currently, eviCore reviews only for the MRI co-registered with MEG.
- MEG followed by co-registration with Brain MRI is referred to as Magnetic Source Imaging (MSI).[20]

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 (LITT)
- 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 (DBS)

References (HD-9)

v2.0.2024

1. Expert Panel on Neurological Imaging, Lee RK, Burns J, et al. ACR Appropriateness Criteria® Seizures and Epilepsy. J Am Coll Radiol. 2020;17(5S):S293-S304. doi:10.1016/j.jacr.2020.01.037
2. Krumholz A, Wiebe S, Gronseth GS, et al. Evidence-based guideline: Management of an unprovoked first seizure in adults: Report of the Guideline Development Subcommittee of the American Academy of Neurology and the American Epilepsy Society. Neurology. 2015;84(16):1705-1713. doi:10.1212/wnl.0000000000001487
3. Hirtz D, Berg A, Bettis D, et al. Practice parameter: treatment of the child with a first unprovoked seizure: Report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. *Neurology*. 2003;60(2):166-175. doi:10.1212/01.wnl.0000033622.27961.b6
4. Lapalme-Remis S, Nguyen DK. Neuroimaging of Epilepsy. *Continuum (Minneap Minn)*. 2022;28(2):306-338. doi:10.1212/CON.0000000000001080
5. Tranvinh E, Lanzman B, Provenzale J, Wintermark M. Imaging Evaluation of the Adult Presenting With New-Onset Seizure. *AJR Am J Roentgenol*. 2019;212(1):15-25. doi:10.2214/ajr.18.20202
6. Ho K, Lawn N, Bynevelt M, Lee J, Dunne J. Neuroimaging of first-ever *Neurol Clin Pract*. 2013;3(5):398-403. doi:10.1212/CPJ.0b013e3182a78f25
7. Knowlton RC, Elgavish RA, Bartolucci A, et al. Functional imaging: II. Prediction of epilepsy surgery outcome. *Ann Neurol.* 2008;64(1):35-41. doi:10.1002/ana.21419
8. Weil S, Noachtar S, Arnold S, Yousry TA, Winkler PA, Tatsch K. Ictal ECD-SPECT differentiates between temporal and extratemporal epilepsy: confirmation by excellent postoperative seizure control. *Nucl Med Commun*. 2001;22(2):233-237. doi:10.1097/00006231-200102000-00016.
9. Qiu J, Cui Y, Qi B, Sun L, Zhu Z. The application of preoperative computed tomography angiogram for hemispherectomy. *Clin Pract*. 2017;7(4). doi:10.4081/cp.2017.992.
10. Guedj E, Varrone A, Boellaard R, et al. EANM procedure guidelines for brain PET imaging using [^{18}F]FDG, version 3 [published correction appears in Eur J Nucl Med Mol Imaging. 2022 Mar 7;:]. *Eur J Nucl Med Mol Imaging*. 2022;49(2):632-651. doi:10.1007/s00259-021-05603-w
11. Correction to: EANM procedure guidelines for brain PET imaging using [^{18}F]FDG, version 3.Guedj E, Varrone A, Boellaard R, Albert NL, Barthel H, van Berckel B, Brendel M, Cecchin D, Ekmekcioglu O, Garibotto V, Lammertsma AA, Law I, Peñuelas I, Semah F, Traub-Weidinger T, van de Giessen E, Van Weehaeghe D, Morbelli S.Eur J Nucl Med Mol Imaging. 2022 May;49(6):2100-2101. doi: 10.1007/s00259-022-05755-3.
12. Bernasconi A, Cendes F, Theodore WH, et al. Recommendations for the use of structural magnetic resonance imaging in the care of patients with epilepsy: A consensus report from the International League Against Epilepsy Neuroimaging Task Force.*Epilepsia*. 2019;60(6):1054-1068. doi:10.1111/epi.15612
13. Passaro EA. Neuroimaging in Adults and Children With Epilepsy.*Continuum (Minneap Minn)*. 2023;29(1):104-155. doi:10.1212/CON.0000000000001242
14. Szaflarski JP, Gloss D, Binder JR, et al. Practice guideline summary: Use of fMRI in the presurgical evaluation of patients with epilepsy: Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology.*Neurology*. 2017;88(4):395-402. doi:10.1212/WNL.0000000000003532
15. Ponisio MR, Zempel JM, Day BK, et al. The Role of SPECT and PET in Epilepsy.*AJR Am J Roentgenol*. 2021;216(3):759-768. doi:10.2214/AJR.20.23336
16. Rampp S, Stefan H, Wu X, et al. Magnetoencephalography for epileptic focus localization in a series of 1000 cases.*Brain*. 2019;142(10):3059-3071. doi:10.1093/brain/awz23
17. Culler GW 4th, Jobst BC. Surgical Treatments for Epilepsy.*Continuum (Minneap Minn)*. 2022;28(2):536-558. doi:10.1212/CON.0000000000001106
18. Delev D, Quesada CM, Grote A, et al. A multimodal concept for invasive diagnostics and surgery based on neuronavigated voxel-based morphometric MRI postprocessing data in previously nonlesional epilepsy. *J Neurosurg*. 2018;128(4):1178-1186. doi:10.3171/2016.12.jns161676.
19. Englot DJ, Nagarajan SS, Imber BS, et al. Epileptogenic zone localization using magnetoencephalography predicts seizure freedom in epilepsy surgery.*Epilepsia*. 2015;56(6):949-958. doi:10.1111/epi.13002
20. Laohathai C, Ebersole JS, Mosher JC, et al. Practical Fundamentals of Clinical MEG Interpretation in Epilepsy.*Front Neurol*. 2021;12:722986. Published 2021 Oct 14. doi:10.3389/fneur.2021.722986
21. Carrette E, Stefan H. Evidence for the Role of Magnetic Source Imaging in the Presurgical Evaluation of Refractory Epilepsy Patients.*Front Neurol*. 2019;10:933. Published 2019 Sep 10. doi:10.3389/fneur.201
22. Spencer D. MRI (minimum recommended imaging) in epilepsy.*Epilepsy Curr*. 2014;14(5):261-263. doi:10.5698/1535-7597-14.5.261

23. Wellmer J, Quesada CM, Rothe L, Elger CE, Bien CG, Urbach H. Proposal for a magnetic resonance imaging protocol for the detection of epileptogenic lesions at early outpatient stages.*Epilepsia*. 2013;54(11):1977-1987. doi:10.1111/epi.12375
24. Oldan JD, Shin HW, Khandani AH, Zamora C, Benefield T, Jewells V. Subsequent experience in hybrid PET-MRI for evaluation of refractory focal onset epilepsy.*Seizure*. 2018;61:128-134. doi:10.1016/j.seizure.2018.07.022
25. Salamon N, Kung J, Shaw SJ, et al. FDG-PET/MRI coregistration improves detection of cortical dysplasia in patients with epilepsy. Neurology. 2008;71(20):1594-1601. doi:10.1212/01.wnl.0000334752.41807.2f
26. Johnson R, Rizk G, Kaur H, Ibekwe H, Atta M, Gayed I. Refractory seizures: Prediction of outcome of surgical intervention based on results from PET-CT, PET-MRI and electroencephaology.*Neuroradiol J*. 2020;33(1):57-65. doi:10.1177/1971400919881464
27. Tóth M, Barsi P, Tóth Z, et al. The role of hybrid FDG-PET/MRI on decision-making in presurgical evaluation of drug-resistant epilepsy.*BMC Neurol*. 2021;21(1):363. Published 2021 Sep 18. doi:10.1186/s12883-021-02352-z

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Trigeminal Neuralgia and other Centrally Mediated Facial Pain Syndromes (HD-10)

Trigeminal Neuralgia/Trigeminal Neuropathy (HD-10.1)

HD.TM.0010.1.C
v2.0.2024

- MRI Brain without and with contrast (CPT® 70553) (with special attention to the skull base) or MRI Brain without contrast (CPT® 70551) **AND/OR** facial imaging, MRI Orbit/ Face/Neck without contrast (CPT® 70540) or MRI Orbit/Face/Neck with and without contrast (CPT® 70543)[5] for:
  - Symptoms of trigeminal neuropathy[5]
  - Suspected trigeminal neuralgia or one of its cranial nerve variants such as glossopharyngeal neuralgia (CN IX), (see **Glossopharyngeal Neuralgia/Glossopharyngeal Neuropathy (HD-10.2)**)
  - 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
- CT Maxillofacial without contrast (CPT® 70486) or CT Maxillofacial with contrast (CPT®70487) for evaluating the skull base and neural foramina[5]
- Contrast-enhanced navigation protocol CT (CPT® 76497) for gamma knife stereotactic radiosurgery for trigeminal neuralgia[5], (see also, **Neuronavigation (HD-28.2)** and **Post Operative Imaging (HD-28.3)**) for post-treatment imaging studies
- MRA Head (CPT® 70544, CPT® 70545 or CPT® 70546) or CTA Head (CPT® 70496) for:
  - Trigeminal neuralgia (vascular imaging may be obtained concurrently with structural brain imaging)[5]
  - 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.

Symptoms of trigeminal neuropathy include facial pain, facial numbness, and/or weakness of the muscles of mastication.

Trigeminal neuralgia, also known as tic douloureux (the involuntary wincing associated with the occurrence of pain), refers to sudden, severe, shooting "electrical" pains along one or more sensory divisions of the trigeminal nerve, provoked by movements such as chewing, or by external stimuli, such as wind blowing or touching the face.

Glossopharyngeal Neuralgia/Glossopharyngeal Neuropathy (HD-10.2)

HD.TM.0010.2.A
v2.0.2024

- MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) **AND/OR** MRI Orbit/Face/Neck without and with contrast (CPT® 70543) or MRI Orbit/Face/Neck without contrast (CPT® 70540) for suspected glossopharyngeal neuralgia or neuropathy[5]
- CT Neck with contrast (CPT® 70491) to delineate skull base erosion, deep space neck masses, calcifications, the skull base bony anatomy and/or the stylohyoid ligament[5] (see also **Eagle Syndrome (Neck-10.3)**)
- MRA Head with contrast (CPT® 70545), or MRA Head without and with contrast (CPT® 70546) **AND/OR** MRA Neck with contrast (CPT® 70548), or MRA Neck without and with contrast (CPT® 70549), to assess for neurovascular compression for the evaluation of glossopharyngeal neuralgia[5]

Background and Supporting Information

- Glossopharyngeal neuralgia presents as severe pain in the throat and neck, classically triggered by swallowing[5].
- Glossopharyngeal neuropathy may present with pain, dysphagia, loss of gag reflex, impaired taste, and impaired sensation along posterior one-third of the tongue and/or inability to elevate the palate[5].

References (HD-10)

v2.0.2024

1. Goh BT, Poon CY, Peck RHL. The importance of routine magnetic resonance imaging in trigeminal neuralgia diagnosis. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology*. 2001;92(4):424-429. doi:10.1067/moe.2001.115130
2. Yaltho TC, Jankovic J. The many faces of hemifacial spasm: Differential diagnosis of unilateral facial spasms. *Movement Disorders*. 2011;26(9):1582-1592. doi:10.1002/mds.23692
3. Cruccu G. Trigeminal Neuralgia. *CONTINUUM: Lifelong Learning in Neurology*. 2017;23(2):396-420. doi:10.1212/con.0000000000000451
4. AAN Practice Parameter: The Diagnostic Evaluation and Treatment of Trigeminal Neuralgia. October 2008. Reaffirmed 7/21/2018
5. Expert Panel on Neurological Imaging, Rath TJ, Policeni B, et al. ACR Appropriateness Criteria® Cranial Neuropathy: 2022 Update. J Am Coll Radiol. 2022;19(11S):S266-S303. doi:10.1016/j.jacr.2022.09.021

Headache (HD-11)

Headache General Guidelines (HD-11.0)

HD.HA.0011.0.C
v2.0.2024

- 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 **Advanced Imaging Indications Related to Migraines (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.[16]

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.[16]

Headache and Suspected Vascular Dissection (HD-11.1)

HD.HA.0011.1A
v2.0.2024

- 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 indicated 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
v2.0.2024

- MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) or CT Head without contrast (CPT® 70450) supported for any of the following:
  - Headache accompanied by seizures, vomiting, focal neurological complaints including dizziness, visual change, altered mental status, or acute hypertension (see **Primary Central Nervous System Tumors – General Considerations (ONC-2.1)** in the Oncology Imaging Guidelines and **Stroke/TIA (HD-21.1)**)
  - Abnormal examination findings (including, but not limited to, altered mental status, papilledema, focal signs or symptoms including unilateral weakness or sensory loss, hyperreflexia, clonus, increased tone, Hoffman or Babinski sign, loss of coordination, seizures, gait disturbance, cranial nerve abnormality, vision loss, nystagmus, dysarthria, dysphagia, fever, meningismus)
- Headaches with any of the following 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)**)
  - 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 (see **Sudden Onset of Headache (HD-11.3)**)
- 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:[28]

- 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
v2.0.2024

- 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)
  - High risk scenarios including Fibromuscular Dysplasia (FMD), Marfan Disease, MVA with whiplash, and chiropractic manipulation
- 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 carotid or vertebral 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[8]
- 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)**, **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)**, **Intracranial Aneurysms (HD-12.1)**, and **Systemic Infections (HD-11.13)**

Trigeminal Autonomic Cephalgias (HD-11.4)

HD.HA.0011.4.A
v2.0.2024

- For trigeminal autonomic cephalgias and cluster headache[27]:
  - MRI Brain without and with contrast (preferred study) (CPT® 70553) **OR**
  - MRI Brain without contrast (CPT® 70551)
  - May also include pituitary screening (see **Pituitary (HD-19)**)
- For facial pain (see **Trigeminal Neuralgia and other Centrally Mediated Facial Pain Syndromes (HD-10))**

Background and Supporting Information

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.

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

HD.HA.0011.5.A
v2.0.2024

- Skull base, orbital, periorbital or oromaxillary[1] imaging is indicated 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/Face/Neck without and with contrast (preferred study) (CPT® 70553 and/or CPT® 70543) **OR**
  - MRI Brain and/or Orbits/Face/Neck without contrast (CPT® 70551 and/or CPT® 70540) **OR**
  - CT Head and/or Orbits/Temporal bone without and with contrast (CPT® 70470 and/or CPT® 70482) **OR**
  - CT Head and/or Orbits/Temporal bone with contrast (CPT® 70460 and/or CPT® 70481)

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

HD.HA.0011.6.A
v2.0.2024

- 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)**

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

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

HD.HA.0011.7.A
v2.0.2024

- 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), see **Cerebral Vasculitis (HD-22)**

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Cancer or Immunosuppression (HD-11.8)

HD.HA.0011.8A
v2.0.2024

- 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
v2.0.2024

- 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
    - MRA/MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA/CTV Head (CPT® 70496) may be added for venogram when requested.
      - 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
v2.0.2024

- 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)
  - MRA/MRV Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA/CTV Head (CPT® 70496) when venogram is requested
    - 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
- 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[1,6]
- For post LP/epidural anesthesia, see **Low Pressure Headache and CSF Leak (HD-11.15)**

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Physical Exertion (HD-11.11)

HD.HA.0011.11.A
v2.0.2024

- 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:[26]
  - 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 carotid or vertebral artery dissection or aneurysm is suspected

Headaches Associated With Head Trauma (HD-11.12)

HD.HA.0011.12.A
v2.0.2024

- 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:[14]
  - 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:[14]
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553)
- 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[14] 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)**

Systemic Infections (HD-11.13)

HD.HA.0011.13.A
v2.0.2024

- 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 without contrast (CPT® 70450) or CT Head without and with contrast (CPT® 70470) 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)**
- See **Neuro-COVID-19 and Sars-CoV-2 Vaccines (HD-14.2)** for headache related to neuro-COVID-19 or SARS-CoV-2 vaccines

Hydrocephalus Shunts (HD-11.14)

HD.HA.0011.14.C
v2.0.2024

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) for any of the following:
  - New signs or symptoms suggesting shunt malfunction or endoscopic third ventriculostomy (ETV) malfunction
    - Symptoms may include but are 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.
  - Requests ordered by a neurologist, neurosurgeon, or any provider in consultation with a neurologist or neurosurgeon.
- MRI Brain without contrast (CPT® 70551) or CT Head without contrast (CPT® 70450) is indicated in the post-operative 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.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

- For CSF flow imaging, see **CSF Flow Imaging (HD-24.4)**
- See also **General Guidelines - CT Head (HD-1.4)**

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.

..

Low Pressure Headache and CSF Leak (HD-11.15)

HD.HA.0011.15A
v2.0.2024

- Evaluation of suspected CSF leak (rhinorrhea/otorrhea) or refractory post-lumbar puncture or low pressure headache:[15]

Indication	Supported Imaging
Intracranial imaging	• MRI Brain without and with contrast (CPT® 70553)
Spinal imaging (MRI)	• 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) **AND/OR** • MRI Lumbar Spine without contrast (CPT® 72148) or without and with contrast (CPT® 72158)
Spinal imaging, post-myelogram	• CT Cervical Spine with contrast (CPT® 72126) **AND/OR** • CT Thoracic Spine with contrast (CPT® 72179) **AND/OR** • CT Lumbar Spine with contrast (CPT® 72132)
Cisternogram, radionucleotide (111 In-DTPA)	• Radionucleotide cisternogram (CPT® 78630)

Indication	Supported Imaging
Cisternogram, post-myelogram (iodinated contrast)	• CT Head with contrast (CPT® 70451) **OR** • CT Maxillofacial with contrast (CPT® 70487) **OR** • CT Temporal Bone with contrast (CPT® 70481)
Symptoms of CSF rhinorrhea or otorrhea	• CT Head without contrast (CPT® 70450) **AND/OR** • CT Maxillofacial without contrast (CPT® 70486) **OR** • CT Temporal Bone without contrast (CPT® 70480)

- 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)**)

Background and Supporting Information

- Common radiological findings of CSF leaks include: abnormalities of the cribiform plate or ethmoid sinus, dural dehiscence at the anterior skull base, pneumatization of the sphenoid sinus, and fluid within the middle ear.
- 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

Cervicogenic Headaches Including Occipital Neuritis/Neuralgia (HD-11.16)

HD.HA.0011.16A
v2.0.2024

- Brain imaging should follow applicable sections in **Headache (HD-11)**
- MRI Cervical Spine without contrast (CPT® 72141) or CT Cervical Spine without contrast (CPT® 72125)
  - Failure of recent (within 3 months) 6-week trial of provider-directed treatment (unless presence of a red flag) as defined in **Red Flag Indications (SP-1.2)**, and clinical re-evaluation after treatment period.
    - See **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
      - Exemptions to the 6 weeks of conservative care include:
        - 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
      - **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)

Background and Supporting Information

- 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)

Advanced Imaging Indications Related To Migraines (HD-11.17)

HD.HA.0011.17A
v2.0.2024

- 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:[28]
  - 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)

References (HD-11)

v2.0.2024

1. Expert Panel on Neurologic Imaging, Pallavi S, Utukuri MD, et al. ACR Appropriateness Criteria® Headache. Available at https://acsearch.acr.org/docs/69482/Narrative/ American College of Radiology. 2022.
2. Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders, 3rd edition. Cephalalgia. 2018;38(1):1-211
3. Thurtell MJ. Idiopathic Intracranial Hypertension. CONTINUUM: Lifelong Learning in Neurology. 2019;25(5):1289-1309. doi:10.1212/con.0000000000000770
4. Burch R. Headache in Pregnancy and the Puerperium. Neurologic Clinics. 2019;37(1):31-51. doi:10.1016/j.ncl.2018.09.004
5. Jamieson DG, Mcvige JW. Imaging of Neurologic Disorders in Pregnancy. Neurologic Clinics. 2020;38(1):37-64. doi:10.1016/j.ncl.2019.09.001
6. Rayhill M. Headache in Pregnancy and Lactation. *Continuum (Minneap Minn)*. 2022; 28(1): 72-92. doi: 10.1212/CON.0000000000001070
7. Kamel H, Navi BB, Sriram N, Hovsepian DA, Devereux RB, Elkind MS. Risk of a Thrombotic Event after the 6-Week Postpartum Period. New England Journal of Medicine. 2014;370(14):1307-1315. doi:10.1056/nejmoa1311485
8. Perillo T, Paolella C, Perrotta G, Serino A, Caranci F, Manto A. Reversible cerebral vasoconstriction syndrome: review of neuroimaging findings. *Radiol Med*. 2022; 127(9): 981-990. doi: 10.1007/s11547-022-01532-2
9. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck
10. Dobrocky T, Nicholson P, Häni L, et al. Spontaneous intracranial hypotension: searching for the CSF leak. *Lancet Neurol*. 2022; 21(4): 369-380. doi: 10.1016/S1474-4422(21)00423-3.
11. Expert Panel on Neurologic Imaging:, Salmela MB, Mortazavi S, et al. ACR Appropriateness Criteria® Cerebrovascular Disease. J Am Coll Radiol. 2017;14(5S):S34-S61. doi:10.1016/j.jacr.2017.01.051
12. Pruitt AA. Central Nervous System Infections Complicating Immunosuppression and Transplantation. CONTINUUM: Lifelong Learning in Neurology. 2018;24(5):1370-1396. doi:10.1212/con.0000000000000653
13. Evans, RW, Burch RC, Frishberg BM, et al. Neuroimaging for Migraine: The American Headache Society Systematic Review and Evidence-Based Guideline. Headache: The Journal of Head and Face Pain. 2020;60(2):318-336. doi:10.1111/head.13720
14. Expert Panel on Neurological Imaging, Shih RY, Burns J, et al. ACR Appropriateness Criteria® Head Trauma: 2021 Update. J Am Coll Radiol. 2021;18(5S):S13-S36. doi:10.1016/j.jacr.2021.01.006
15. ACR ASNR SPR Practice Parameter for the Performance of Myelography and Cisternography. Revised 2019
16. Jordan JE, Flanders AE. Headache and Neuroimaging: Why We Continue to Do It. *AJNR Am J Neuroradiol*. 2020; 41(7): 1149-1155. doi: 10.3174/ajnr.A6591
17. Sweet JA, Mitchell LS, Narouze S, et al. Occipital Nerve Stimulation for the Treatment of Patients With Medically Refractory Occipital Neuralgia. Neurosurgery. 2015;77(3):332-341. doi:10.1227/neu.0000000000000872
18. https://ichd-3.org/13-painful-cranial-neuropathies-and-other-facial-pains/13-4-occipital-neuralgia/
19. https://ichd-3.org/11-headache-or-facial-pain-attributed-to-disorder-of-the-cranium-neck-eyes-ears-nose-sinuses-teeth-mouth-or-other-facial-or-cervical-structure/11-2-headache-attributed-to-disorder-of-the-neck/11-2-1-cervicogenic-headache/
20. Doddamani RS, Meena RK, Sawarkar D, Aggarwal D, Chandra PS. Management Options in Occipital Neuralgia: A. Journal of Peripheral Nerve Surgery Vol. 2020;4(1)
21. O'Neill F, Nurmikko T, Sommer C. Other facial neuralgias. Cephalalgia. 2017;37(7):658-669. doi:10.1177/0333102417689995
22. Barmherzig R, Kingston W. Occipital Neuralgia and Cervicogenic Headache: Diagnosis and Management. Current Neurology and Neuroscience Reports. 2019;19(5). doi:10.1007/s11910-019-0937-8
23. Labastida-Ramírez A, Benemei S, Albanese M, et al. Persistent post-traumatic headache: a migrainous loop or not? The clinical evidence. The Journal of Headache and Pain. 2020;21(1). doi:10.1186/s10194-020-01122-5
24. Henderson FC, Austin C, Benzel E, et al. Neurological and spinal manifestations of the Ehlers-Danlos syndromes. American journal of medical genetics Part C, Seminars in medical genetics. 2017;175(1):195-211. doi:10.1002/ajmg.c.31549

25. Chou DE. Secondary Headache Syndromes. CONTINUUM: Lifelong Learning in Neurology. 2018;24(4):1179-1191. doi:10.1212/con.0000000000000640
26. Smith JH. Other Primary Headache Disorder. *Continuum (Minneap Minn)*. 2021; 27(3): 652-664. doi: 10.1212/CON.0000000000000960
27. Nahas SJ. Cluster Headache and Other Trigeminal Autonomic Cephalalgias. *Continuum (Minneap Minn)*. 2021; 27(3): 633-651. doi: 10.1212/CON.0000000000000965
28. Recober A. Pathophysiology of Migraine. *Continuum (Minneap Minn)*. 2021; 27(3): 586-596. doi: 10.1212/COM.000000000000983

Aneurysm and AVM (HD-12)

Intracranial Aneurysms (HD-12.1)

HD.AN.0012.1.C
v2.0.2024

- 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
    - Glucocorticoid-remediable aldosteronism (GRA)[25]
  - 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.[22] (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 intracranial aneurysm.[22]
- 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 indicated 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 indicated.[22]
- 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
v2.0.2024

Disorders and Indications (Any of the following)	Supported Imaging
Any aneurysmal and/or AVM disorders listed in this guideline • When MRI contraindicated[29] • Any emergency setting	• CT Head without contrast (CPT® 70450) **AND/OR** • CTA Head (CPT® 70496) **AND/OR** • CTA Neck (CPT® 70498)
Known AVM • When requested by a neurologist, neurosurgeon or any provider in consultation with a neurologist or neurosurgeon	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, CPT® 70546) **OR** • CTA Head (CPT® 70496)
Known AVM in the vertebral-basilar system[22] • When requested by a neurologist, neurosurgeon or any provider in consultation with a neurologist or neurosurgeon	• Imaging as listed above in "known AVM" **AND/OR** • MRA Neck (CPT® 70547, CPT® 70548, OR CPT® 70549) **OR** • CTA Neck (CPT® 70498)
Subarachnoid Hemorrhage (SAH) • AVM is suspected based on a history of SAH	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551)

Disorders and Indications (Any of the following)	Supported Imaging
Hereditary Hemorrhagic Telangiectasia (HHT; Osler-Weber-Rendu Syndrome) • Suspected based on family history with at least one affected first-degree relative (biological parent or sibling) • At diagnosis, especially if confirmed by genetic testing • Screening for confirmed HHT • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist, neurosurgeon, geneticist, or any provider in consultation with a neurologist, neurosurgeon or geneticist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, CPT® 70546) **OR** • CTA Head (CPT® 70496)
Capillary Malformation-Arteriovenous Malformation (CM-AVM) • Suspected based on family history with at least one affected first-degree relative (biological parent or sibling) • At diagnosis, especially if confirmed by genetic testing • Screening for confirmed CM-AVM • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist, neurosurgeon, geneticist, or any provider in consultation with a neurologist, neurosurgeon or geneticist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, CPT® 70546) **OR** • CTA Head (CPT® 70496) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146)

Disorders and Indications (Any of the following)	Supported Imaging
Cerebral Cavernous Malformations (CCM) • At diagnosis, especially if confirmed by genetic testing • Screening for confirmed CCM • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist, neurosurgeon, geneticist, or any provider in consultation with a neurologist, neurosurgeon or geneticist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, CPT® 70546) **OR** • CTA Head (CPT® 70496) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146)
Microcephalic Osteodysplastic Primordial Dwarfism, Type II (MOPD II)[19] • At diagnosis, especially if confirmed by genetic testing • Screening for confirmed MOPD II, repeated annually • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist, neurosurgeon, geneticist, or any provider in consultation with a neurologist, neurosurgeon or geneticist	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, CPT® 70546) **OR** • CTA Head (CPT® 70496) **AND/OR** • MRA Neck (CPT® 70547, CPT® 70548, CPT® 70549) **OR** • CT Neck (CPT® 70498)

Disorders and Indications (Any of the following)	Supported Imaging
Sturge-Weber Syndrome • At diagnosis • Clinical signs or symptoms concerning for disease progression • When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRI Orbits/Face/Neck without and with contrast (CPT® 70543) **OR** • MRI Orbits/Face/Neck without contrast (CPT® 70540)

- MRI Brain without and with contrast (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 (CPT® 70496) supported for symptoms including headache, seizure, and/or focal neurologic deficits[11,20,26]
- For concerns related to stroke, see **Stroke/TIA (HD-21.1)**
- 3D imaging (CPT® 76377) with MRI Brain without and with contrast (CPT® 70553) OR MRI Brain without contrast (CPT® 70551) is supported
- 3D Rendering (CPT® 76377) with cerebral angiography to define the presence, location, and anatomy of intracranial and cervical vascular malformations at diagnosis and for follow up, including post-treatment[11,26]. See **General Guidelines - Other Imaging Situations (HD-1.7)** and **Background and Supporting Information**
- See **General Guidelines – CT and MR Angiography (CTA and MRA) (HD-1.5)**
- Functional MRI (CPT® 70554 OR CPT® 70555) for surgical planning, see **Functional MRI (fMRI) (HD-24.2)** [11]

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.
- Most intracranial AVMs are congenital, vary widely in their location and type, and are discovered at birth due to associated clinical findings or incidentally later in life. Certain hereditary conditions are associated with an increased risk for AVM development.
- Vascular malformations include arteriovenous, venous, cavernous, and capillary malformations.
- Hereditary AVMs usually have an autosomal dominant pattern of inheritance.[10,19,31,33]

References (HD-12)

HD.AN.0012.3.A
v2.0.2024

1. Rozenfeld MN, Ansari SA, Shaibani A, Russell EJ, Mohan P, Hurley MC. Should Patients with Autosomal Dominant Polycystic Kidney Disease Be Screened for Cerebral Aneurysms? *American Journal of Neuroradiology*. 2013;35(1):3-9. doi:10.3174/ajnr.a3437
2. Vlak MHM, Rinkel GJE, Greebe P, Greving JP, Algra A. Lifetime risks for aneurysmal subarachnoid haemorrhage: multivariable risk stratification. *Journal of Neurology, Neurosurgery & Psychiatry*. 2013;84(6):619-623. doi:10.1136/jnnp-2012-303783
3. Nguyen TN. Management of Unruptured Intracranial Aneurysms and Brain Arteriovenous Malformations.*Continuum (Minneap Minn)*. 2023;29(2):584-604. doi:10.1212/CON.0000000000001247
4. Thompson BG, Brown RD, Amin-Hanjani S, et al. Guidelines for the Management of Patients With Unruptured Intracranial Aneurysms. *Stroke*. 2015;46(8):2368-2400. doi:10.1161/str.0000000000000070
5. Chu LC, Johnson PT, Dietz HC, Fishman EK. CT Angiographic Evaluation of Genetic Vascular Disease: Role in Detection, Staging, and Management of Complex Vascular Pathologic Conditions. *American Journal of Roentgenology*. 2014;202(5):1120-1129. doi:10.2214/ajr.13.11485
6. Hishikawa T, Date I, Tokunaga K, et al. Risk of rupture of unruptured cerebral aneurysms in elderly patients. *Neurology*. 2015;85(21):1879-1885. doi:10.1212/wnl.0000000000002149
7. Backes, D, Rinkel GJE, Greving JP, et al. ELAPSS score for prediction of risk of growth of unruptured intracranial aneurysms. *Neurology*. 2017;88(17):1600-1606. doi:10.1212/wnl.0000000000003865
8. Ding D, Etminan N. A model for predicting the growth of unruptured intracranial aneurysms. *Neurology*. 2017;88(17):1594-1595. doi:10.1212/wl.0000000000003874
9. Kadian-Dodov D, Gornik HL, Gu X, et al. Dissection and Aneurysm in Patients With Fibromuscular Dysplasia. *Journal of the American College of Cardiology*. 2016;68(2):176-185. doi:10.1016/j.jacc.2016.04.044
10. McDonald J, Stevenson DA. Hereditary Hemorrhagic Telangiectasia. In: Adam MP, Mirzaa GM, Pagon RA, et al., eds.*GeneReviews®*. Seattle (WA): University of Washington, Seattle; June 26, 2000. [Updated 2021 Nov 24]
11. Derdeyn CP, Zipfel GJ, Albuquerque FC, et al. Management of Brain Arteriovenous Malformations: A Scientific Statement for Healthcare Professionals From the American Heart Association/American Stroke Association. *Stroke*. 2017;48(8). doi:10.1161/str.0000000000000134.
12. Expert Panel on Neurologic Imaging: Salmela MB, Mortazavi S, et al. ACR Appropriateness Criteria® Cerebrovascular Disease. J Am Coll Radiol. 2017;14(5S):S34-S61. doi:10.1016/j.jacr.2017.01.051
13. Rosser T. Neurocutaneous Disorders. CONTINUUM: Lifelong Learning in Neurology. 2018;24(1):96-129. doi:10.1212/con.0000000000000562
14. Horne MA, Flemming KD, Su I-C, et al. Clinical course of untreated cerebral cavernous malformations: a meta-analysis of individual patient data. *The Lancet Neurology*. 2016;15(2):166-173. doi:10.1016/s1474-4422(15)00303-8
15. Vella M, Alexander M, Mabray M, et al. Comparison of MRI, MRA, and DSA for Detection of Cerebral Arteriovenous Malformations in Hereditary Hemorrhagic Telangiectasia. *American Journal of Neuroradiology*. 2020;41(6):969-975. doi:10.3174/ajnr.a6549
16. Lawton MT and Vates GE. Subarachnoid Hemorrhage. *N Engl J Med* 2017;377:257-66. doi: 10.1056/NEJMcp1605827
17. Connolly ES, Rabinstein AA, Carhuapoma JR, et al. Guidelines for the Management of Aneurysmal Subarachnoid Hemorrhage. *Stroke*. 2012;43(6):1711-1737. doi:10.1161/str.0b013e3182587839
18. Meschia JF, Bushnell C, Boden-Albala B, et al. Guidelines for the Primary Prevention of Stroke. *Stroke*. 2014;45(12):3754-3832. doi:10.1161/str.0000000000000046
19. Michael B.Bober and Andrew P. Jackson, Microcephalic Osteodysplastic Primordial Dwarfism Type II: A clinical review Current Osteoporosis Report (2017) 15:61-69 doi: 10.1007/s11914-017-0348-1
20. Chen, C-J et al. Brain arteriovenous malformations: A review of natural history, pathobiology, and interventions *Neurology* 2020 95(20):917-927. doi: 10.1212/WNL.0000000000010968
21. ACR-ASNR-SIR-SNIS Practice Prameter for the Performan of Diagnostic Cervicocerebral Catheter Angiography in Adults. Revised 2016. (Resolution 13) https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CervicoCerebralCathAngio.pdf
22. Expert Panel on Neurological Imaging, Ledbetter LN, Burns J, et al. ACR Appropriateness Criteria® Cerebrovascular Diseases-Aneurysm, Vascular Malformation, and Subarachnoid Hemorrhage. J Am Coll Radiol. 2021;18(11S):S283-S304. doi:10.1016/j.jacr.2021.08.012
23. Nesvick CL, Oushy S, Ravindran K, et al. Repeat Catheter Angiography in Patients with Aneurysmal-Pattern Angiographically Negative Subarachnoid Hemorrhage. Neurocritical Care. Published online June 28, 2021. doi:10.1007/s12028-021-01247-8

24. Rosenberg TL, Suen JY, Richter GT. Arteriovenous Malformations of the Head and Neck. Otolaryngologic Clinics of North America. 2018;51(1):185-195. doi:10.1016/j.otc.2017.09.005
25. Litchfield WR, Anderson BF, Weiss RJ, Lifton RP, Dluhy RG. Intracranial aneurysm and hemorrhagic stroke in glucocorticoid-remediable aldosteronism. *Hypertension*. 1998;31(1 Pt 2):445-450. doi:10.1161/01.hyp.31.1.445
26. Johnson MD, Staarmann B, Zuccarello M. A Rational Approach to the Management of Cerebral Arteriovenous Malformations. *World Neurosurg*. 2022;159:338-347. doi:10.1016/j.wneu.2021.08.045
27. Hoh BL, Ko NU, Amin-Hanjani S, et al. 2023 Guideline for the Management of Patients With Aneurysmal Subarachnoid Hemorrhage: A Guideline From the American Heart Association/American Stroke Association [published online ahead of print, 2023 May 22]. Stroke. 2023;10.1161/STR.0000000000000436. doi:10.1161/STR.0000000000000436
28. Faughnan ME, Mager JJ, Hetts SW, et al. Second International Guidelines for the Diagnosis and Management of Hereditary Hemorrhagic Telangiectasia. *Ann Intern Med*. 2020;173(12):989-1001. doi:10.7326/M20-1443
29. Expert Panel on MR Safety, Kanal E, Barkovich AJ, et al. ACR guidance document on MR safe practices: 2013. *J Magn Reson Imaging*. 2013;37(3):501-530. doi:10.1002/jmri.24011
30. Sabeti S, Ball KL, Bhattacharya SK, et al. Consensus Statement for the Management and Treatment of Sturge-Weber Syndrome: Neurology, Neuroimaging, and Ophthalmology Recommendations. *Pediatr Neurol*. 2021;121:59-66. doi:10.1016/j.pediatrneurol.2021.04.013
31. Bayrak-Toydemir P, Stevenson DA. Capillary Malformation-Arteriovenous Malformation Syndrome. In: Adam MP, Mirzaa GM, Pagon RA, et al., eds. *GeneReviews*®. Seattle (WA): University of Washington, Seattle; February 22, 2011. Updated: September 12, 2019.
32. Hammill AM, Wusik K, Kasthuri RS. Hereditary hemorrhagic telangiectasia (HHT): a practical guide to management. *Hematology Am Soc Hematol Educ Program*. 2021;2021(1):469-477. doi:10.1182/hematology.2021000281
33. Morrison L, Akers A. Cerebral Cavernous Malformation, Familial. In: Adam MP, Mirzaa GM, Pagon RA, et al., eds. *GeneReviews*®. Seattle (WA): University of Washington, Seattle; February 24, 2003.

Head and Facial Trauma (HD-13)

Head Trauma (HD-13.1)

HD.TR.0013.1.A
v2.0.2024

For acute head trauma (0 to 7 days post-trauma)[7]

- CT Head without contrast (CPT® 70450) is preferred in individuals with **ANY** of the following modified Canadian CT Head Rule/New Orleans Criteria.[1,7,9]
  - Regardless of documented or stated head impact, ANY "dangerous mechanism of injury", either direct or indirect, including, but not exclusive to:
    - Fall from height greater than 3 feet
    - Fall greater than 5 steps down stairs
    - Any pedestrian motor vehicle accident
    - High impact motor vehicle accident
  - Individual >60 years old
  - Loss of consciousness, amnesia, or disorientation accompanying blunt head trauma within 24 hours
  - 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
  - Suspected open skull fracture
  - Signs of basilar skull fracture (Battle's sign, Raccoon eyes, CSF rhinorrhea, cranial nerve palsy, hemotympanum, acute hearing loss)
  - Vomiting
  - 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)**]

For subacute head trauma (7 days to 3 months post-trauma)[7] and chronic head trauma (greater than 3 months post-trauma) symptoms[7]

- MRI Brain without contrast (CPT® 70551) or CT Head without contrast (CPT® 70450) is indicated for the initial imaging of individuals with subacute or chronic head trauma and unexplained cognitive or neurologic deficits.[7]
- MRI Brain without and with contrast (CPT® 70553) if post-traumatic infection is suspected

Repeat and follow-up imaging

- Follow-up imaging for known subdural hematomas, intracerebral hemorrhage, or contusions can be done at the discretion of the ordering provider 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 short term follow-up imaging of acute traumatic brain injury (TBI) without neurologic deterioration, CT Head without contrast (CPT® 70450) is the most appropriate imaging study in individuals with ANY of the following risk factors
  - subfrontal/temporal intraparenchymal contusions
  - anticoagulation
  - age >65 years
  - intracranial hemorrhage
- MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) can be approved 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.[7]

For suspected intracranial venous or arterial injury

- CTA/CTV Head (CPT® 70496) **OR** MRA/MRV Head (CPT® 70544, CPT® 70545, CPT® 70546)

or

  - 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))**

SPECT, PET, CT/MRI perfusion, DTI (diffusion tensor imaging), functional MRI, and MR spectroscopy are not considered routine clinical practice at this time.[3,7]

See **Neck (Cervical Spine) Pain Without/With Neurological Features (Including Stenosis) and Trauma (SP-3.2)** in the Spine Imaging Guidelines

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

Background and Supporting Information

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

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.[3,8]

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.A
v2.0.2024

- 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) and/or CT Head without contrast (CPT® 70450)[11]:
  - 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
  - Physical exam findings of CSF otorrhea (suspected post-traumatic CSF leak)
- If concern for CSF leak and CT Maxillofacial or Temporal bone is inconclusive[7] (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.

References (HD-13)

HD.TR.0013.3.A

v2.0.2024

1. Stiell IG, Wells GA, Vandemheen K, et al. The Canadian CT Head Rule for patients with minor head injury. *Lancet*. 2001;357(9266):1391-1396. doi:10.1016/s0140-6736(00)04561-x
2. Giza CC, Kutcher JS, Ashwal S, et al. Summary of evidence-based guideline update: Evaluation and management of concussion in sports: Report of the Guideline Development Subcommittee of the American Academy of Neurology. Neurology. 2013;80(24):2250-2257. doi:10.1212/wnl.0b013e31828d57dd
3. Silverberg ND, Iaccarino MA, Panenka WJ, et al. Management of Concussion and Mild Traumatic Brain Injury: A Synthesis of Practice Guidelines. *Arch Phys Med Rehabil*. 2020;101(2): 382-393. doi: 10.1016/j.apmr.2019.10.179.
4. Hoffmann JF. An Algorithm for the Initial Management of Nasal Trauma. *Facial Plast Surg*. 2015;31(3): 183-193. doi: 10.1055/s-0035-1555618.
5. Sun JK, Lemay DR. Imaging of facial trauma. *Neuroimaging Clin N Am*. 2002;12(2):295-309. doi:10.1016/s1052-5149(02)00002-3
6. Harmon KG, Clugston JR, Dec K, et al. American Medical Society for Sports Medicine position statement on concussion in sport. *B J Sports Med*. 2019;53(4):213-225. doi:10.1136/bjsports-2018-100338
7. Expert Panel on Neurological Imaging, Shih RY, Burns J, et al. ACR Appropriateness Criteria® Head Trauma: 2021 Update. J Am Coll Radiol. 2021;18(5S):S13-S36. doi:10.1016/j.jacr.2021.01.006
8. Wintermark M, Sanelli PC, Anzai Y, et al. Imaging Evidence and Recommendations for Traumatic Brain Injury: Conventional Neuroimaging Techniques. *J A Coll Radiol*. 2015;12(2):e1-e14. doi:10.1016/j.jacr.2014.10.014
9. Papa L, Stiell IG, Clement CM, et al. Performance of the Canadian CT Head Rule and the New Orleans Criteria for predicting any traumatic intracranial injury on computed tomography in a United States Level I trauma center. *Acad Emerg Med*. 2012;19(1): 2-10. doi: 10.1111/j.1553-2712.2011.01247.x.
10. Reljic T, Mahony H, Djulbegovic B, et al. Value of Repeat Head Computed Tomography after Traumatic Brain Injury: Systematic Review and Meta-Analysis. *J Neurotrauma*. 2014;31(1):78-98. doi:10.1089/neu.2013.2873
11. Expert Panel on Neurologic Imaging:, Kennedy TA, Corey AS, et al. ACR Appropriateness Criteria® Orbits Vision and Visual Loss. J Am Coll Radiol. 2018;15(5S):S116-S131. doi:10.1016/j.jacr.2018.03.023
12. Mower WR, Hoffman JR, Herbert M, et al. Developing a Decision Instrument to Guide Computed Tomographic Imaging of Blunt Head Injury Patients. *J Trauma*. 2005;59(4):954-959. doi:10.1097/01.ta.0000187813.79047.42
13. Haydel MJ, Preston CA, Mills TJ, Luber S, Blaudeau E, Deblieux PM. Indications for Computed Tomography in Patients with Minor Head Injury. *N Engl J Med*. 2000;343(2):100-105. doi:10.1056/nejm200007133430204
14. https://www.cdc.gov/traumaticbraininjury/pdf/tbi_clinicians_factsheet-a.pdf
15. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck

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

CNS and Head Infection (HD-14.1)

HD.HI.0014.1.A
v2.0.2024

INITIAL IMAGING

- Signs of intracranial infection include, but are not limited to
  - headaches, seizures, meningeal signs (neck stiffness)
  - new focal neurological deficits in a setting of fever or elevated white blood cell count (WBC)
  - known infection elsewhere 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) **OR**
  - CT Head (CPT® 70450, CPT® 70460, or CPT® 70470) in cases where MRI is contraindicated
  - If vascular involvement is suspected, in addition to MRI Brain, the following are supported[21]:
    - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR**
    - CTA Head (CPT® 70496) **AND/OR**
    - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) **OR**
    - CTA Neck (CPT® 70498)
    - (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)
  - Concern for vasculitis, see **Cerebral Vasculitis (HD-22.1)**

REPEAT IMAGING

- As requested by an infectious disease specialist, neurologist, neurosurgeon, radiologist or any provider coordinating care with an infectious disease specialist, neurologist, neurosurgeon or radiologist
- Repeat imaging would refer to any of the CPT codes listed above as initial imaging.
  - 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)**, **Mental Status Change (HD-4.2)**, and **Eye Disorders and Visual Loss (HD-32.1)**

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

HD.HI.0014.2.A
v2.0.2024

- The following studies are supported for evaluation of:
  - Acute or chronic Neuro-COVID-19 syndrome
    - MRI Brain without contrast (CPT® 70551) **OR**
    - MRI Brain without and with contrast (CPT® 70553) **OR**
    - CT head without contrast (CPT® 70450) **OR**
    - CT head without and with contrast (CPT® 70470) is supported if there is a contraindication to MRI **AND/OR**
    - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) OR CTA Head (CPT® 70496) **AND/OR**
    - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498)
    - 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 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
  - Suspected neurologic adverse reactions after SARS- CoV-2 vaccination:
    - MRI Brain without contrast (CPT® 70551) **OR**
    - MRI Brain without and with contrast (CPT® 70553) **OR**
    - CT head without contrast (CPT® 70450) **OR**
    - CT head without and with contrast (CPT® 70470) is supported if there is a contraindication to MRI **AND/OR**
    - MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) OR CTA Head (CPT® 70496) **AND/OR**
    - MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) or CTA Neck (CPT® 70498)
    - 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 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

  - If suspected transverse myelitis and/or COVID infection, then ANY the following are supported:
    - MRI Cervical without and with contrast (CPT® 72156)
    - MRI Thoracic without and with contrast (CPT® 72157)
    - MRI Lumbar Spine without and with contrast (CPT® 72158)[35,36]
    - See **Stroke/TIA (HD-21.1)** for vascular imaging
    - See **Transverse Myelitis (HD-16.4)** regarding spine imaging to evaluate for post-vaccination neurological syndrome
- 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.

Background and Supporting Information

- 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.[8,9]
- 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).[10,11,15,16,20]
- 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.[13,14,17,18,19,21]
- Cases of Thrombosis with Thrombocytopenia Syndrome (TTS) following administration of the Johnson & Johnson/Janssen COVID-19 Vaccine have been reported in males and females, in a wide age range of individuals 18 years and

older, with the highest reporting rate (approximately 8 cases per 1,000,000 doses administered) in females ages 30-49 years; overall, approximately 15% of TTS cases have been fatal. Currently available evidence supports a causal relationship between TTS and the Johnson & Johnson/Janssen COVID-19 Vaccine. The clinical course of these events shares features with autoimmune heparin-induced thrombocytopenia. In individuals with suspected TTS following administration of the Johnson & Johnson/Janssen COVID-19 Vaccine, the use of heparin may be harmful and alternative treatments may be needed. Consultation with hematology specialists is strongly recommended. The American Society of Hematology has published considerations relevant to the diagnosis and treatment of TTS following administration of the Janssen COVID-19 Vaccine (https://www.hematology.org/covid-19/vaccine-induced-immunethrombotic-thrombocytopenia). (see Full EUA Prescribing Information).

- Janssen COVID-19 Vaccine EUA Fact Sheet for Healthcare Providers 03132023 (fda.gov)

Autoimmune/Paraneoplastic Encephalitis & Neuroinflammatory Disorders (HD-14.3)

HD.HI.0014.3.A
v2.0.2024

Indications:

When acute/ subacute or rapid progression (< 3 months) of altered mental status, focal findings including cranial nerve, motor or sensory symptoms or memory loss or psychiatric symptoms, seizure, and/ or focal CNS findings are present.[26]

OR

There is a stated concern for neuro-inflammatory encephalitis from a neurologist, neurosurgeon or psychiatrist[26]:

Initial Imaging [26]:

- MRI Brain without and with contrast (CPT® 70553; preferred study) **OR** MRI Brain without contrast (CPT® 70551) **OR**
- CT Head without contrast (CPT® 70450) **OR** CT Head without and with contrast (CPT® 70470) when MRI is contraindicated or for bony pathology concerns
- CTA Head (CPT® 70496) **AND/OR** CTA Neck (CPT® 70498) for evaluating large vessel obstructions, aneurysms and vascular malformations, dissection, vasospasm, and vasculopathies such as CNS vasculitis (see **Cerebral Vasculitis (HD 22.1)**, **Intracranial Aneurysms (HD 12.1)**, **Arteriovenous Malformations (AVMs) and Related Lesions (HD 12.2)**, **Stroke/TIA (HD 21.1)**)

Repeat Imaging:

MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551) when specialized sequences are needed such as, but not limited to[26]:

- High T2 contrast sequences (CISS, FIESTA) sequences to identify blood (SWI) or
- To identify acute cytotoxic edema (DWI) or
- When requested by a neurologist, oncologist, rheumatologist or infectious disease specialist

Metabolic (FDG) Brain PET (CPT® 78608) is indicated to evaluate individuals suspected of having encephalitis, including autoimmune encephalitis, if diagnosis remains unclear after evaluation with MRI Brain, CSF analysis, and/or lab testing including serology.[26]

Neurosarcoidosis [31,32,33,34]:

- Supported for known or suspected neurosarcoidosis.
  - MRI Brain without and with contrast (CPT® 70553)

AND/OR

- If spinal cord involvement suspected, then
  - MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR**
  - MRI Thoracic Spine without and with contrast (CPT® 72157) **AND/OR**
  - MRI Lumbar Spine without and with contrast (CPT® 72158)

  AND/OR
- If peri-orbital involvement suspected, then
  - MRI Orbits/Face/Neck without and with contrast (CPT® 70543)
- Repeat imaging supported if requested by neurologist, rheumatologist, ophthalmologist, oncologist or radiologist or provider in consultation with a neurologist, rheumatologist, ophthalmologist, oncologist or radiologist.

- For non-neurologic imaging related to sarcoidosis (see **Sarcoid (CH-15.1)**)

Background and Supporting Information [26]

Supportive studies in the evaluation of Autoimmune/Paraneoplastic Encephalopathy include:

- CSF pleocytosis (>5 WBC/μL) or
- EEG changes or
- Supporting labs (including positive CSF antibody positivity and/or serologies)

Potential etiologies:

- Paraneoplastic
  - NMDA Receptor encephalitis
  - LGI1 antibody encephalitis
- Autoimmune
  - Neurosarcoidosis can involve any of the following:
    - Brain, Cranial Nerves, Spinal Cord and/or Peripheral Nerves
    - Acute Disseminated Encephalomyelitis (ADEM), Anti-MOG Syndrome, Multiple Sclerosis (MS), Neuromyelitis Optica (NMO)
    - IgG4 related disease
    - CNS histiocytosis
- Neuro-rheumatologic
  - ANCA related disease
  - Behcet's disease
  - Sjogren Syndrome +/- Rheumatoid Arthritis (RA)

FDG-PET imaging of the brain for paraneoplastic and autoimmune encephalitis may be more sensitive than Brain MRI (87% vs. 56%) but is nonspecific. Areas of hypometablism are seen in neurodegenerative disorders such as dementias. However,

topographic patterns of hypometabolism may help characterize the disorder as autoimmune/ paraneoplastic encephalitis, in a way that may help clarify diagnosis and alter management strategies. For example, anterior to posterior gradient of hypometabolism is seen in NMDA Receptor encephalitis. Hemispheric hypometabolism out of proportion to atrophy characterizes Rasmussen encephalitis.[26]

Non-head Imaging

- MRI is helpful in determining the length of spine lesion (short versus longitudinally extensive transverse myelitis), width (partial versus transverse), and location (eccentric, central, hemicord, anterior versus posterior, conus, tracts, or meningeal).
  - See **Myelopathy (SP 7.1)** and **Anti-MOG Syndromes (HD 16.3)**
- The Trident Sign on axial MRI, which has been described in relation to neurosarcoidosis, demonstrates leptomeningeal or dorsal subpial enhancement that may or may not involve the central canal.
  - See **Myelopathy (SP 7.1)**
- Involvement of the conus medullaris is a clue to Anti-MOG (Myelin Oligodendrocyte Glycoprotein-associated disorder) as the cause of longitudinally extensive transverse myelitis.
  - See **Transverse Myelitis (HD 16.4)**
- CT of the chest, abdomen, and pelvis with contrast is a generally accepted first method of screening for occult malignancy or systemic inflammation (e.g., sarcoidosis).
  - See **Paraneoplastic Syndromes (ONC 30.3)** and **Sarcoid (CH 15.1)**

References (HD-14)

v2.0.2024

1. Abdalkader M, Xie J, Cervantes-Arslanian A, Takahashi C, Mian AZ. Imaging of Intracranial Infections. *Seminars in Neurology*. 2019;39(03):322-333. doi:10.1055/s-0039-1693161
2. Probasco JC, Solnes L, Nalluri A, et al. Abnormal brain metabolism on FDG-PET/CT is a common early finding in autoimmune encephalitis. *Neurology - Neuroimmunology Neuroinflammation*. 2017;4(4). doi:10.1212/nxi.0000000000000352
3. Rubin R. As Their Numbers Grow, COVID-19 "Long Haulers" Stump Experts. *JAMA*. 2020;324(14):1381–1383. doi:10.1001/jama.2020.17709
4. E. M. Liotta et al. Frequent neurologic manifestations and encephalopathy-associated morbidity in Covid-19 patients. *Annals of Clinical and Translational Neurology* 2020; 7(11): 2221–2230 doi: 10.1002/acn3.51210.
5. Chen X, Laurent S, Onur OA, Kleineberg NN, Fink GR, Schweitzer F, Warnke C. A systematic review of neurological symptoms and complications of COVID-19. *Journal of Neurology*. 2021 Feb;268(2):392-402. doi: 10.1007/s00415-020-10067-3
6. Finsterer J Neurological side effects of SARS-CoV-2 vaccinations Acta Neurol Scand. 2022 145(1): 5–9. doi: 10.1111/ane.13550
7. Kaulen LD, Doubrovinskaia S, Mooshage C, Jordan B, Purrucker J, Haubner C, Seliger C, Lorenz HM, Nagel S, Wildemann B, Bendszus M, Wick W, Schönenberger S. Neurological autoimmune diseases following vaccinations against SARS-CoV-2: a case series. Eur J Neurol. 2022 Feb;29(2):555-563. doi: 10.1111/ene.15147
8. Maury A, Lyoubi A, Peiffer-Smadja N, de Broucker T, Meppiel E. Neurological manifestations associated with SARS-CoV-2 and other coronaviruses: A narrative review for clinicians. Rev Neurol (Paris). 2021 Jan-Feb;177(1-2):51-64. doi: 10.1016/j.neurol.2020.10.001
9. Moreno-Escobar MC, Kataria S, Khan E, Subedi R, Tandon M, Peshwe K, Kramer J, Niaze F, Sriwastava S. Acute transverse myelitis with Dysautonomia following SARS-CoV-2 infection: A case report and review of literature. J Neuroimmunol. 2021 Apr 15;353:577523. doi: 10.1016/j.jneuroim.2021.577523
10. Patone M, Handunnetthi L, Saatci D, Pan J, Katikireddi SV, Razvi S, Hunt D, Mei XW, Dixon S, Zaccardi F, Khunti K, Watkinson P, Coupland CAC, Doidge J, Harrison DA, Ravanan R, Sheikh A, Robertson C, Hippisley-Cox J. Neurological complications after first dose of COVID-19 vaccines and SARS-CoV-2 infection. Nat Med. 2021 Dec;27(12):2144-2153. doi: 10.1038/s41591-021-01556-7
11. Rosenblum HG, Hadler SC, Moulia D, Shimabukuro TT, Su JR, Tepper NK, Ess KC, Woo EJ, Mba-Jonas A, Alimchandani M, Nair N, Klein NP, Hanson KE, Markowitz LE, Wharton M, McNally VV, Romero JR, Talbot HK, Lee GM, Daley MF, Mbaeyi SA, Oliver SE. Use of COVID-19 Vaccines After Reports of Adverse Events Among Adult Recipients of Janssen (Johnson & Johnson) and mRNA COVID-19 Vaccines (Pfizer-BioNTech and Moderna): Update from the Advisory Committee on Immunization Practices - United States, July 2021. MMWR Morb Mortal Wkly Rep. 2021 Aug 13;70(32):1094-1099. doi: 10.15585/mmwr.mm7032e4
12. Rosenblum HG, Gee J, Liu R, Marquez PL, Zhang B, Strid P, Abara WE, McNeil MM, Myers TR, Hause AM, Su JR, Markowitz LE, Shimabukuro TT, Shay DK. Safety of mRNA vaccines administered during the initial 6 months of the US COVID-19 vaccination programme: an observational study of reports to the Vaccine Adverse Event Reporting System and v-safe. Lancet Infect Dis. 2022 Mar 7:S1473-3099(22)00054-8. doi: 10.1016/S1473-3099(22)00054-8
13. Vasconcelos TMF, Oliveira DN, Ferreira GM, Torres FC, Castro JDV, Braga-Neto P, Sobreira-Neto MA. Covid-19 post-infectious acute transverse myelitis responsive to corticosteroid therapy: report of two clinical cases. J Neurovirol. 2021 Oct;27(5):791-796. doi: 10.1007/s13365-021-01010-x 12
14. Frontera JA, Tamborska AA, Doheim MF, Garcia-Azorin D, Gezegen H, Guekht A, Yusof Khan AHK, Santacatterina M, Sejvar J, Thakur KT, Westenberg E, Winkler AS, Beghi E; contributors from the Global COVID-19 Neuro Research Coalition. Neurological Events Reported after COVID-19 Vaccines: An Analysis of VAERS. Ann Neurol. 2022 Mar 2. doi: 10.1002/ana.26339
15. ACR–ASNR–SPR PRACTICE PARAMETER FOR THE PERFORMANCE OF COMPUTED TOMOGRAPHY (CT) OF THE HEAD https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head.pdf
16. Expert Panel on Neurological Imaging, Rath TJ, Policeni B, et al. ACR Appropriateness Criteria® Cranial Neuropathy: 2022 Update.*J Am Coll Radiol*. 2022;19(11S):S266-S303. doi:10.1016/j.jacr.2022.09.021
17. Anand P. Neurologic Infections in Patients on Immunomodulatory and Immunosuppressive Therapies.*Continuum (Minneap Minn)*. 2021;27(4):1066-1104. doi:10.1212/CON.0000000000000985
18. Chow F. Neurosyphilis.*Continuum (Minneap Minn)*. 2021;27(4):1018-1039. doi:10.1212/CON.0000000000000982

19. Roos KL. Neurologic Complications of Lyme Disease.*Continuum (Minneap Minn)*. 2021;27(4):1040-1050. doi:10.1212/CON.0000000000001015
20. Singh SK, Hasbun R. Neuroradiology of infectious diseases.*Curr Opin Infect Dis*. 2021;34(3):228-237. doi:10.1097/QCO.0000000000000725
21. Weidauer S, Wagner M, Enkirch SJ, Hattingen E. CNS Infections in Immunoincompetent Patients : Neuroradiological and Clinical Features.*Clin Neuroradiol*. 2020;30(1):9-25. doi:10.1007/s00062-019-00837-6
22. Sakai M, Higashi M, Fujiwara T, et al. MRI imaging features of HIV-related central nervous system diseases: diagnosis by pattern recognition in daily practice.*Jpn J Radiol*. 2021;39(11):1023-1038. doi:10.1007/s11604-021-01150-4
23. Corrêa DG, de Souza SR, Freddi TAL, Fonseca APA, Dos Santos RQ, Hygino da Cruz LC Jr. Imaging features of neurosyphilis.*J Neuroradiol*. 2023;50(2):241-252. doi:10.1016/j.neurad.2023.01.003
24. Beghi E, Helbok R, Ozturk S, et al. Short- and long-term outcome and predictors in an international cohort of patients with neuro-COVID-19.*Eur J Neurol*. 2022;29(6):1663-1684. doi:10.1111/ene.15293
25. Premraj L, Kannapadi NV, Briggs J, Seal SM, Battaglini D, Fanning J, Suen J, Robba C, Fraser J, Cho SM. Mid and long-term neurological and neuropsychiatric manifestations of post-COVID-19 syndrome: A meta-analysis. J Neurol Sci. 2022 Mar 15;434:120162. doi: 10.1016/j.jns.2022.120162. Epub 2022 Jan 29. PMID: 35121209; PMCID: PMC8798975.
26. Wahed LA, Cho TA. Imaging of Central Nervous System Autoimmune, Paraneoplastic, and Neuro-rheumatologic Disorders. Continuum (Minneap Minn). 2023 Feb 1;29(1):255-291. doi: 10.1212/CON.0000000000001244. PMID: 367958880.
27. Bordonne, M., Chawki, M.B., Doyen, M. *et al.* Brain 18F-FDG PET for the diagnosis of autoimmune encephalitis: a systematic review and a meta-analysis. *Eur J Nucl Med Mol Imaging* 48, 3847–3858 (2021). https://doi.org/10.1007/s00259-021-05299-y
28. Graus F, Titulaer MJ, Balu R, et al. A clinical approach to diagnosis of autoimmune encephalitis. Lancet Neurol. 2016;15(4):391-404. doi:10.1016/S1474-4422(15)00401-9
29. Budhram A, Leung A, Nicolle MW, Burneo JG. Diagnosing autoimmune limbic encephalitis.*CMAJ*. 2019;191(19):E529-E534. doi:10.1503/cmaj.181548
30. Solnes LB, Jones KM, Rowe SP, et al. Diagnostic Value of^{18}F-FDG PET/CT Versus MRI in the Setting of Antibody-Specific Autoimmune Encephalitis.*J Nucl Med*. 2017;58(8):1307-1313. doi:10.2967/jnumed.116.184333
31. Stern BJ, Royal W, Gelfand JM, et al. Definition and Consensus Diagnostic Criteria for Neurosarcoidosis: From the Neurosarcoidosis Consortium Consensus Group. JAMA Neurology. 2018;75(12):1546. doi:10.1001/jamaneurol.2018.2295.
32. Bradshaw MJ, Pawate S, Koth LL, Cho TA, Gelfand JM. Neurosarcoidosis: Pathophysiology, Diagnosis, and Treatment. Neurol Neuroimmunol Neuroinflamm. 2021 Oct 4;8(6):e1084. doi: 10.1212/NXI.0000000000001084
33. Fritz D, van de Beek D, Brouwer MC. Clinical features, treatment and outcome in neurosarcoidosis: systematic review and meta-analysis. BMC Neurol. 2016 Nov 15;16(1):220. doi: 10.1186/s12883-016-0741-x
34. Pawate S. Sarcoidosis and the Nervous System. *Continuum (Minneap Minn)*. 2020 Jun;26(3):695-715. doi: 10.1212/CON.0000000000000855
35. Khan E, Shrestha AK, Colantonio MA, Liberio RN, Sriwastava S. Acute transverse myelitis following SARS-CoV-2 vaccination: a case report and review of the literature. *J Neurol*. 2022.269(3):1121-1132. doi:10.1007/s00415-021-10785-2
36. Ismail II, Salama S. Association of CNS demyelination and COVID-19 infection: an updated systematic review. *J Neurol*. 2022:269(2):541-576. doi:10.1007/s00415-021-10752-x

Movement Disorders (HD-15)

Movement Disorders (HD-15.1)

HD.MD.0015.1.C
v2.0.2024

- 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 MRI Brain without and with contrast (CPT® 70553) in the following clinical scenarios:
  - Clinical diagnostic uncertainty
  - Incomplete or uncertain response to medication
  - Atypical Parkinsonism suspected because of unusual clinical features. These may include, but are not limited to:
    - Persistent unilateral signs or symptoms
    - Onset under age 50
    - Rapid progression
    - See **Background and Supporting Information** for further information on atypical parkinsonism and Parkinson's Plus Syndromes
  - 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/OR** unlisted CT procedure code (CPT® 76497)
  - MR guided Focused Ultrasound:
    - CT Head without contrast (CPT® 70450) to evaluate bone density **AND/OR** MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553)
  - Repeat imaging studies for pre-surgical evaluation, MRI Brain without contrast (CPT® 70551) or MRI Brain without and with contrast (CPT® 70553) **AND/OR** CT Head without contrast (CPT® 70450), when ordered by a Neurosurgeon or Neurologist or any provider in consultation with a Neurosurgeon or Neurologist if greater than 6 months old **and/or** for new symptoms/signs
  - Post op imaging when ordered by a Neurosurgeon or Neurologist or any provider in consultation with a Neurosurgeon or Neurologist for either procedures, see also **Post-Operative Imaging (HD-28.3)** indications

- 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

- 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)**)

References (HD-15)

v2.0.2024

1. Expert Panel on Neurological Imaging, Harvey HB, Watson LC, et al. ACR Appropriateness Criteria® Movement Disorders and Neurodegenerative Diseases. J Am Coll Radiol. 2020;17(5S):S175-S187. doi:10.1016/j.jacr.2020.01.042
2. Thaler A, Alcalay RN. Diagnosis and Medical Management of Parkinson Disease.*Continuum (Minneap Minn)*. 2022;28(5):1281-1300. doi:10.1212/CON.0000000000001152
3. Maiti B, Perlmutter JS. Imaging in Movement Disorders.*Continuum (Minneap Minn)*. 2023;29(1):194-218. doi:10.1212/CON.0000000000001210
4. Subramaniam RM, Frey KA, Hunt CH, et al. ACR-ACNM Practice Parameter for the Performance of Dopamine Transporter (DaT) Single Photon Emission Computed Tomography (SPECT) Imaging for Movement Disorders. Clinical Nuclear Medicine. 2017;42(11):847-852. doi:10.1097/rlu.0000000000001815
5. Bega D, Gonzalez-Latapi P, Zadikoff C, Spies W, Simuni T. Is There a Role for DAT-SPECT Imaging in a Specialty Movement Disorders Practice? Neurodegenerative Diseases. 2015;15(2):81-86. doi:10.1159/000370116
6. Mohammed N, Patra D, Nanda A. A meta-analysis of outcomes and complications of magnetic resonance–guided focused ultrasound in the treatment of essential tremor. Neurosurgical Focus. 2018;44(2). doi:10.3171/2017.11.focus17628
7. Schreglmann SR, Krauss JK, Chang JW, Bhatia KP, Kägi G. Functional lesional neurosurgery for tremor: a systematic review and meta-analysis. Journal of Neurology, Neurosurgery & Psychiatry. 2018;89(7):717-726. doi:10.1136/jnnp-2017-316302
8. Halpern CH, Santini V, Lipsman N, et al. Three-year follow-up of prospective trial of focused ultrasound thalamotomy for essential tremor. Neurology. 2019;93(24). doi:10.1212/wnl.0000000000008561
9. Pouratian N, Baltuch G, Elias WJ, Gross R. American Society for Stereotactic and Functional Neurosurgery Position Statement on Magnetic Resonance-Guided Focused Ultrasound for the Management of Essential Tremor. Neurosurgery. 2019. doi:10.1093/neuros/nyz510
10. Shah BR, et.al. Advanced MRI techniques for transcranial high intensity focused ultrasound targeting. Brain 2020:1-9. doi:10.1093/brain/awaa107
11. Elias JW. A randomized Trial of Focused Ultrasound Thalamotomy for Essential Tremor. N Engl J Med 2016;375:730-9. doi: 10.1056/NEJMoa1600159
12. Rughani A, Schwalb JM, Sidiropoulos C, et al. Congress of Neurological Surgeons Systematic Review and Evidence-Based Guideline on Subthalamic Nucleus and Globus Pallidus Internus Deep Brain Stimulation for the Treatment of Patients With Parkinson's Disease: Executive Summary. Neurosurgery. 2018;82(6):753-756. doi:10.1093/neuros/nyy037.
13. Y. Xiao, J. C. Lau, D. Hemachandra, G. Gilmore, A. Khan and T. M. Peters, "Image guidance in deep brain stimulation surgery to treat Parkinson's disease: a comprehensive review," in IEEE Transactions on Biomedical Engineering, doi: 10.1109/TBME.2020.3006765.
14. Sakamoto F, Shiraishi S, Ogasawara K, et al. A diagnostic strategy for Lewy body disease using DAT-SPECT, MIBG and Combined index. Annals of Nuclear Medicine. 2020;34(6):415-423. doi:10.1007/s12149-020-01464-9.
15. Humanitarian Device Exemption. U.S. Food and Drug Administration (FDA). Page Last Updated: 07/12/2021. https://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfhde/hde.cfm?id=H020007
16. Fluorodopa F18 Injection Package Insert. Highlights of prescribing information. U.S. Food and Drug Administration Website. https://www.accessdata.fda.gov/drugsatfda_docs/label/2019/200655s000lbl.pdf. Revised 10/2019.
17. Dhawan V, Niethammer MH, Lesser ML, et al. Prospective F-18 FDOPA PET Imaging Study in Human PD. *Nucl Med Mol Imaging.* 2022;56(3):147-157.doi:10.1007/s13139-022-00748-4
18. Morbelli S, Esposito G, Arbizu J, et al. EANM practice guideline/SNMMI procedure standard for dopaminergic imaging in Parkinsonian syndromes 1.0.*Eur J Nucl Med Mol Imaging*. 2020;47(8):1885-1912. doi:10.1007/s00259-020-04817-8

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

Multiple Sclerosis (MS) (HD-16.1)

HD.MS.0016.1.C
v2.0.2024

Establishing a New Diagnosis of Multiple Sclerosis

Indication	Supported Imaging
Establishing a new diagnosis of Multiple Sclerosis is based on the following: • Clinical suspicion based on recurrent episodes of variable neurological signs and/or symptoms **AND** • Baseline exclusion of appropriate alternative conditions that can mimic MS	• MRI Brain without and with contrast (CPT® 70553) (preferred study) **OR** • MRI Brain without contrast (CPT® 70551) if there is a contraindication to gadolinium If optic neuritis** is suspected the following imaging is **ALSO** indicated: • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) ****For additional information related to optic neuritis see Eye Disorders and Visual Loss (HD-32.1)** **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146)

Unclear Diagnosis

Indication	Supported Imaging
Diagnosis of Multiple Sclerosis remains unclear or equivocal after initial MRI • May repeat imaging 3- 6 months after initial MRI Brain	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Clinically Isolated Syndrome (CIS)

Indication	Supported Imaging
Clinically Isolated Syndrome (CIS)* based on **ALL** of the following**:** • First episode of neurologic symptoms and neurologic deficits concerning for possible demyelinating disease. • Symptoms last ≥ 24 hours[43] • Initial episode of neurologic symptoms and neurologic deficits • Baseline exclusion of appropriate alternative conditions that can mimic MS *For more information about CIS, see **Background and Supporting Information**	• MRI Brain without and with contrast (CPT® 70553) (preferred study) **OR** • MRI Brain without contrast (CPT® 70551) if there is a contraindication to gadolinium If optic neuritis is suspected **, the following imaging is **ALSO** indicated: • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) ****For additional information related to optic neuritis, see Eye Disorders and Visual Loss (HD-32.1)** **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146)

Radiologically Isolated Syndrome (RIS)

Indication	Supported Imaging
Radiologically Isolated Syndrome (RIS)* based on **ALL** of the following: • Individual with brain MRI obtained for unrelated reason with findings conspicuous for demyelinating disease[41] • No history of recurrent neurologic symptoms suggestive of CIS or RRMS • Baseline exclusion of appropriate alternative conditions that can mimic MS *For more information about RIS, see **Background and Supporting Information**	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146)

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

New Episode of Neurological Deficit in an Individual with Multiple Sclerosis and/or Concern for Possible Diagnosis of Demyelinating Disease

Indication	Supported Imaging
New episode of neurological deficit in an individual with Multiple Sclerosis and/or concern for a possible diagnosis of demyelinating disease	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) If optic neuritis is suspected**, the following imaging is **ALSO** indicated: • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) ****For additional information related to optic neuritis, see Eye Disorders and Visual Loss (HD-32.1)** If there are new or worsening symptoms concerning for spinal cord involvement, the following imaging is **ALSO** indicated: • MRI Cervical Spine without contrast (CPT® 72141) **OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) **OR** • MRI Thoracic Spine without and with contrast (CPT® 72157)

Baseline Imaging with Disease Modifying Therapy (DMT)

Indication	Supported Imaging
• Before starting or changing disease modifying therapy (DMT)[1] **AND/OR** • 3-6 months after starting or changing DMT to establish a new MRI treatment baseline **AND/OR** • If new abnormal MRI Brain findings without clinical symptoms, an additional follow up MRI Brain is supported after 6 months[1]	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and without contrast (CPT® 70553)

Current Treatment with Disease Modifying Therapy (DMT)

Indication	Supported Imaging *Every 3-6 Months*	Supported Imaging *Annually*
Individuals treated with DMT* associated with either the risk progressive multifocal leukoencephalopathy (PML) **AND/OR** other CNS opportunistic infections * For list of medications, see **Background and Supporting Information**	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)	• MRI Cervical Spine without contrast (CPT® 72141) **OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) **OR** • MRI Thoracic Spine without and with contrast (CPT® 72157)

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Annual Supported Imaging

Indication	Supported Imaging *Annually*
Individuals with diagnosed Multiple Sclerosis with **EITHER** of the following: • Not treated with disease modifying therapy (DMT)* **OR** • Treated with beta interferon or glatiramer acetate medications * For list of DMT medications, see **Background and Supporting Information**	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **AND/OR** • MRI Cervical Spine without contrast (CPT® 72141) **OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) **OR** • MRI Thoracic Spine without and with contrast (CPT® 72157)

Treatment with Tysabri® (natalizumab)

Indication	Supported Imaging *Every 3-6 Months*	Supported Imaging *Annually*
Individuals treated with Tysabri® (natalizumab) with the following medical history: • ≥ 18 months of treatment • During Tysabri® (natalizumab) treatment and up to 9-12 months after transitioning off Tysabri® (natalizumab)[1] **AND** • JC virus antibody positive	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)	• MRI Cervical Spine without contrast (CPT® 72141) **OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) **OR** • MRI Thoracic Spine without and with contrast (CPT® 72157)

PML Symptoms during Treatment with Tysabri® (natalizumab) or other Medication with Similar Risk

Indication	Supported Imaging
Symptoms suggestive of Progressive Multifocal Leukoencephalopathy (PML)* during treatment with Tysabri® (natalizumab) or other medication with similar risk * For more information about PML, see **Background and Supporting Information**	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and without contrast (CPT® 70553)

History of Clinically Isolated Syndrome (CIS) or Radiologically Isolated Syndrome (RIS)

Indication	Supported Imaging *Annually*
Patient with history of Clinically Isolated Syndrome* (CIS)[1] **OR** Patient with history of Radiologically Isolated Syndrome* (RIS)[1] *For more information about CIS or RIS, see **Background and Supporting Information**	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)

- 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).[1]
- Quantitative Magnetic Resonance Image (MRI) Analysis of the Brain
  - Volumetric analysis of the temporal lobes and hippocampus or Neuro Quant may be ordered as 3D rendering (CPT® 76377) or quantitative analysis of the brain (CPT® 0865T or CPT® 0866T). These studies lack sufficient specificity and

sensitivity to be clinically useful in the evaluation or follow up of individuals with Multiple Sclerosis. Their use is limited to research studies and are otherwise considered to be not medically necessary in routine clinical practice.

Background and Supporting Information

- Multiple sclerosis is an autoimmune disease that is associated with inflammation, demyelination, and neurodegenerative changes within the optic nerves, brain and spinal cord (i.e. central nervous system (CNS)).
- A diagnosis of multiple sclerosis can be established after an individual has at least one clinical attack suggestive of central nervous system (CNS) demyelination with evidence of separation of space and time as well as reasonably excluding other possible conditions that could account for the clinical and imaging findings.[1,45]
    - MRI lesions in multiple sclerosis are round or ovoid T2-hyperintense lesions that are well circumscribed and 3mm^2 or greater in size.
    - MRI findings that can establish dissemination of space include:
        - Involvement in two or more of the following locations:
            - ≥ 1 brainstem lesion
            - ≥ 1 juxtacortical (abutting the cortex) or cortical lesion
            - ≥ 1 periventricular lesion (abutting the ventricle)
            - ≥ 1 spinal cord lesion
    - MRI findings that can establish dissemination of time:
        - New T2-hyperintense lesion irrespective of timing
        - Presence of simultaneous enhancing and non-enhancing lesions on MRI
    - Multiple sclerosis commonly begins with a relapsing-remitting course with partial or complete neurologic recovery following attacks.
        - An acute attack lasts at least 24 hours or longer
        - Common types of MS attacks include:
            - Unilateral optic neuritis
            - Brainstem or cerebellar syndrome (i.e. trigeminal neuralgia, diplopia or intranuclear ophthalmoplegia (INO), and/or ataxia)
            - Partial transverse myelitis
        - Females are more frequently diagnosed with relapsing-remitting multiple sclerosis (RRMS) compared to males.
            - Individuals are most often diagnosed during their twenties or thirties.
            - Individuals with relapsing multiple sclerosis may later transition into a more progressive phase of the disease that is characterized by insidious cognitive and/or physical decline.
        - Primary progressive multiple sclerosis (PPMS) is characterized by progressive neurologic decline in the absence of acute attacks.
            - The incidence of primary progressive multiple sclerosis is equal among males and females.

- It is often diagnosed at 45-50 years of age.

- The first event concerning for demyelinating disease without meeting criteria for separation of time is known as a clinically isolated syndrome (CIS).[43]
  - A diagnosis of multiple sclerosis can occur when an individual with CIS has a second attack and/or develops a new lesion on MRI.
- Natural history studies and clinical trials of disease modifying therapies (DMT) have shown that individuals with CIS with characteristic MRI brain lesions carry a high risk for meeting diagnostic criteria for multiple sclerosis.[43]
- Clinical trials of MS disease modifying therapy in CIS show that fewer individuals treated with a disease modifying therapy develop a second attack and have reduced MRI activity.[43]
- Individuals who undergo a brain MRI for other indications (i.e. headaches, trauma, seizure) which incidentally reveals abnormalities that are characteristic for demyelination in the absence of clinical symptoms is known as radiologically isolated syndrome (RIS).[41,43,45]
  - A diagnosis of RIS is established by the following: [45]
    - Absence of a clinical attack suggestive of demyelination.
    - MRI abnormalities not related to the effects of substances (recreational drugs, toxic exposure) or other medical condition.
    - The central nervous system (CNS) MRI abnormalities cannot be accounted for by another disease process.
      - MRI white matter abnormalities associated with a vascular pattern of disease.
  - Factors associated with a higher risk for transitioning to multiple sclerosis include [41]
    - Younger age at diagnosis
    - Male patients
    - Individuals with spinal cord and/or brain stem lesions
    - Presence of oligoclonal bands in the cerebrospinal fluid (CSF)
  - A recent randomized, placebo-controlled trial for individuals with radiologically isolated syndrome (RIS) using dimethyl fumarate showed potential benefit in delaying clinical events and MRI activity.
  - Recent studies on radiologically isolated syndrome suggests that this is likely a pre-clinical phase for individuals with multiple sclerosis.
- Progressive Multifocal Leukoencephalopathy (PML) is a progressive multi-focal disease of the central nervous system that can occur in individuals with treated with immunosuppressive or immunomodulatory medications.[46]
  - There is a relatively high incidence of PML in individuals treated with natalizumab although other disease modifying therapies have been associated with PML.[1,46]
    - Increased risk of developing PML has been associated with individuals treated with natalizumab who received prior immunosuppressive

medication, have a high JC virus antibody index, and/or have received natalizumab for ≥ 18 months.[1]
- More frequent MRI monitoring has been associated with lower PML lesion volume at diagnosis and a better outcome than annual monitoring.[1]
- Frequent MRI surveillance is recommended after discontinuing natalizumab due to the potential of carry-over PML that can occur.[1]

▪ A diagnosis of PML is established by neurological symptoms, characteristic MRI abnormalities and positive PCR for the JC virus in cerebrospinal fluid (CSF).
▪ Common symptoms of PML may include but are not limited to hemiparesis, ataxia, gait disorder, visual deficits (i.e. homonymous hemianopia), and/or seizures.[46]

○ Sagittal MRI Spinal Cord with phased array detector coil (CPT® 72156 or CPT® 72157) is an alternative spinal imaging.
○ Interferon beta medications include (but are not limited to): Avonex®, Betaseron®, Extavia®, Plegridy®, Rebif®
○ Glatiramer acetate medications include (but are not limited to): Copaxone, Glatopa®
○ Medications with high risks of PML as Tysabri® (natalizumab) and/or other CNS opportunistic infections (i.e. herpes encephalitis, cryptococcal meningitis) include (but are not limited to): Tecfidera® (dimethyl fumarate), Gilenya® (fingolimod), Tascenso® ODT (fingolimod), Aubagio® (teriflunomide), Ocrevus® (ocrelizumab), Kesimpta® (ofatumumab), Mavenclad® (cladribine), Mayzent® (siponimod), Ponvory® (ponesimod), Vumerity® (diroximel fumarate), Zeposia® (ozanimod), Lemtrada® (alemtuzumab), Bafiertam® (monomethyl fumarate), Briumvi® (ublituximab), Rituxan® (rituximab)

Neuromyelitis Optica and NMO Spectrum Disorders (HD-16.2)

HD.MS.0016.2.C
v2.0.2024

Initial evaluation of Neuromyelitis Optica Spectrum Disorders (NMOSD) with any of the following:

Indication	Supported Imaging
Clinical concern for optic neuritis when requested by a neurologist, neuro-ophthalmologist, ophthalmologist or any provider in consultation with a neurologist, neuro-ophthalmologist, or ophthalmologist	MRI Orbit without and with contrast (CPT® 70543) **OR** MRI Orbit without contrast (CPT® 70540)
Recurrent hiccups or intractable nausea and/or vomiting (clinical concern for area postrema syndrome)	MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551)
Other neurologic signs or symptoms concerning for brain involvement ordered by a neurologist or any provider in consultation with a neurologist	MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551)
Clinical concern for transverse myelitis when ordered by a neurologist or any provider in consultation with a neurologist	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** Due to potential for conus involvement, • MRI Lumbar spine without and with contrast (CPT® 72158) **OR** • MRI Lumbar spine without contrast (CPT® 72148)

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Indication	Supported Imaging
Positive NMO antibody test when ordered by a neurologist or any provider in consultation with a neurologist[37]	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146)

Patient with established diagnosis of (NMOSD) with any of the following:

Indication	Supported Imaging
Clinical concern for optic neuritis when requested by a neurologist, neuro-opthamologist, ophthalmologist or any provider in consultation with a neurologist, neuro-opthamologist, or ophthalmologist	MRI Orbit without and with contrast (CPT® 70543) **OR** MRI Orbit without contrast (CPT® 70540)
New neurologic signs or symptoms concerning for brain involvement when requested by a neurologist or any provider in consultation with a neurologist	MRI Brain without and with contrast (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551)

Indication	Supported Imaging
Clinical concern for transverse myelitis when ordered by a neurologist or any provider in consultation with a neurologist	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** Due to potential for conus involvement, • MRI Lumbar spine without and with contrast (CPT® 72158) **OR** • MRI Lumbar spine without contrast (CPT® 72148)
Repeat imaging may be supported for **ANY** of the following: • Re-establish baseline after starting treatment (typically 3-6 months after last MRI) • Changing disease modifying therapy (DMT) • As requested when ordered by a neurologist, neuro-ophthalmologist, ophthalmologist or any provider in consultation with a neurologist, neuro-ophthalmologist or ophthalmologist[37]	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146)

- Quantitative Magnetic Resonance Image (MRI) Analysis of the Brain
  - Volumetric analysis of the temporal lobes and hippocampus or Neuro Quant may be ordered as 3D rendering (CPT® 76377) or quantitative analysis of the brain (CPT® 0865T or CPT® 0866T). These studies lack sufficient specificity and

sensitivity to be clinically useful in the evaluation or follow up of individuals with NMOSD. Their use is limited to research studies and are otherwise considered to be not medically necessary in routine clinical practice.

Background and Supporting Information

- Neuromyelitis optica spectrum disorder (NMOSD, Devic's disease) is a chronic inflammatory autoimmune disease that involves the optic nerves, spinal cord and brain.
- Accrual of disability occurs during acute attacks in patients with NMOSD.
  - Even after a single attack, severe permanent disability can occur, especially if the attack is not treated immediately and appropriately.
  - Unlike multiple sclerosis, it is rare for individuals with NMOSD to develop asymptomatic lesions within the brain, optic nerves and/or spinal cord.[34]
- Diagnosis is based on the clinical presentation, MRI findings, and the presence of auto-antibodies.
- Core clinical characteristics of NMOSD include[7]
  - Optic neuritis
    - Frequently bilateral optic nerve involvement with severe vision loss
    - Long unilateral and/or bilateral lesion on MRI (more than half of the distance from the orbit to the chiasm and those involving the posterior aspects of the optic chiasm)
  - Longitudinally extensive transverse myelitis
    - ≥ 3 complete, contiguous vertebral segments of the spinal cord are involved
    - More than 70% of the lesion resides within the central gray matter of the spinal cord
  - Area postrema syndrome
    - Otherwise unexplained episode of recurrent hiccups or intractable nausea and vomiting
  - Brainstem or cerebral syndrome with NMOSD typical brain lesions[7]
    - Lesions involve periependymal surfaces of the 3rd and 4th ventricles in the brainstem and cerebellum
    - Hypothalamic or thalamic lesions
    - Large, confluent unilateral or bilateral subcortical or deep white matter lesions[7]
    - Long (≥ 1/2 the length of the corpus callosum) with diffuse, heterogeneous or edematous corpus callosum lesions
    - Long corticospinal tract lesions, involving unilateral or bilateral, contiguously involving internal capsule and cerebral peduncle
    - Extensive periependymal lesions, often with gadolinium enhancement
  - 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), rituximab[37], and Uplizna® (inebilizumab)
  - Possible adverse reactions associated with treatment include risk of PML and meningococcal infections
- Several medications that are effective in multiple sclerosis, including interferon β, fingolimod, alemtuzumab, and natalizumab are associated with severe outcomes, including catastrophic exacerbations in patients with NMOSD.[35]

MOG Antibody-Associated Disease (MOGAD) (HD-16.3)

HD.MS.0016.3.A
v2.0.2024

Initial evaluation of MOG (myelin oligodendrocyte glycoprotein) antibody-associated diseases (MOGAD) with any of the following:

Indication	Supported Imaging
Clinical concern for optic neuritis when requested by a neurologist, ophthalmologist or any provider in consultation with a neurologist or ophthalmologist	• MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540)
Neurologic signs or symptoms concerning for brain involvement when ordered by a neurologist or any provider in consultation with a neurologist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551)
Clinical concern for transverse myelitis when ordered by a neurologist or any provider in consultation with a neurologist	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** Due to potential for conus involvement: • MRI Lumbar spine without and with contrast (CPT® 72158) **OR** • MRI Lumbar spine without contrast (CPT® 72148)

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Indication	Supported Imaging
Positive MOG antibody test when ordered by a neurologist or any provider in consultation with a neurologist[34]	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) and MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Cervical Spine without contrast (CPT® 72141) and MRI Thoracic Spine without contrast (CPT® 72146)

Patients with established diagnosis of (MOGAD) with any of the following:

Indication	Supported Imaging
Clinical concern for optic neuritis when requested by a neurologist, ophthalmologist or any provider in consultation with a neurologist or ophthalmologist	• MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540)
Neurologic signs or symptoms concerning for brain involvement when ordered by a neurologist or any provider in consultation with a neurologist	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551)

Indication	Supported Imaging
Clinical concern for transverse myelitis when ordered by a neurologist or any provider in consultation with a neurologist	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** Due to potential for conus involvement: • MRI Lumbar spine without and with contrast (CPT® 72158) **OR** • MRI Lumbar spine without contrast (CPT® 72148)
Repeat imaging may be supported for **ANY** of the following: • Re-establish baseline after starting treatment (typically 3-6 months after last MRI) • Changing disease modifying therapy (DMT) • As requested when ordered by a neurologist, neuro-ophthalmologist, ophthalmologist or any provider in consultation with a neurologist, neuro-ophthalmologist or ophthalmologist[34]	• MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) **AND/OR** • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) **AND/OR** • MRI Cervical Spine without and with contrast (CPT® 72156) and MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Cervical Spine without contrast (CPT® 72141) and MRI Thoracic Spine without contrast (CPT® 72146)

- Acute relapse is considered when an individual with MOGAD develops new neurologic signs or symptoms at least 30 days following the onset of a previous attack.[34]

Background and Supporting Information

- MOG (myelin oligodendrocyte glycoprotein)-IgG disorders are CNS inflammatory diseases, distinct from multiple sclerosis and NMO-spectrum disorders.
- Unlike multiple sclerosis and neuromyelitis optica spectrum disorder (NMOSD), individuals with MOG antibody-associated disease (MOGAD) can have a monophasic or relapsing course.[34]
- Diagnosis is based on the clinical presentation, MRI findings, and the presence of auto-antibodies
- Clinical features of individuals with MOGAD include[34]
    - Optic neuritis
        - Bilateral optic neuritis is common at onset, and seems to be more frequent in individuals with MOGAD than with those with multiple sclerosis or neuromyelitis optica spectrum disorder (NMOSD).[34]
        - Vision returns quickly with return to normal or near normal visual acuity following treatment with intravenous corticosteroids.[34]
    - Transverse myelitis
        - May be short segment
        - Longitudinally extensive transverse myelitis (≥3 vertebral segments of the spinal cord)
        - Cauda equine and peripheral nerve root involvement can occur (lumbar spine imaging is indicated)[45]
        - Can occur as an isolated episode of transverse myelitis, as a component of ADEM or in conjunction with optic neuritis.[34]
        - T2 spinal cord lesions often are centrally located and can be restricted to the grey matter producing the “H sign” on MRI
            - 20%-25% of spinal cord lesions in individuals with MOGAD do not involved the grey matter.[34]
        - Most T2 lesions resolve or reduce in size substantially on follow up MRI
    - Brainstem encephalitis
    - Encephalitis with seizures
    - May be associated with cortical edema and leptomeningeal enhancement[45]
    - Acute disseminated encephalomyelitis (ADEM)
    - Occurs mainly in children but can occur in adults.
    - Tumefactive brain lesions
    - Cranial neuropathies

Relapses are more common in the first six months after the first attack.

- Unlike multiple sclerosis and neuromyelitis optica spectrum disorder (NMOSD), individuals with MOG antibody-associated disease (MOGAD) can have a monophasic or relapsing course.[34]
- Unlike multiple sclerosis, it is rare for individuals with MOGAD to develop asymptomatic lesions within the brain, optic nerves and/or spinal cord.[34]

- An acute relapse is considered when an individual with MOGAD develops new neurologic signs or symptoms at least 30 days following the onset of a previous attack.
  - Relapses are more common in the first six months after the first attack.[34]
  - New symptoms or signs in an individual with known MOGAD may include
    - Blurred vision, vision loss and/or loss of color vision
    - Motor weakness of a limb or limbs, including paraparesis or complete paralysis
    - Motor weakness may include:
      - Loss and/or worsening of manual dexterity
      - New or worsening foot drag
    - Change in sensation in a limb or limbs that may be associated with paresthesias and/or dysesthesias
    - Urinary urgency, incontinence and/or urinary retention
    - Worsening constipation and/or bowel urgency/incontinence
    - Sexual dysfunction
    - Lhermitte's phenomenon
    - New or worsening spasticity
    - New or worsening gait difficulties (i.e. spastic and/or ataxic gait) and/or sexual dysfunction
    - Seizures

Transverse Myelitis (HD-16.4)

HD.MS.0016.4.A
v2.0.2024

An initial assessment, to include a pertinent history and neurologic exam, should be performed prior to imaging requests.

Clinical Concern for Transverse Myelitis

Indication	Supported Imaging
Clinical concern for transverse myelitis when ordered by a neurologist or radiologist or any provider in consultation with a neurologist or radiologist	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** Due to potential for conus involvement, • MRI Lumbar spine without and with contrast (CPT® 72158) **OR** • MRI Lumbar spine without contrast (CPT® 72148) **AND/OR** • MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) If optic neuritis is suspected*, the following imaging is **ALSO** indicated: • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) ***For additional information related to optic neuritis see Eye Disorders and Visual Loss (HD-32.1)**

New Neurologic Signs or Symptoms

Indication	Supported Imaging
New neurologic signs or symptoms	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** • MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551) If optic neuritis is suspected*, the following imaging is **ALSO** indicated: • MRI Orbit without and with contrast (CPT® 70543) **OR** • MRI Orbit without contrast (CPT® 70540) ***For additional information related to optic neuritis, see Eye Disorders and Visual Loss (HD-32.1)**

History of Transverse Myelitis

Indication	Supported Imaging *Annually for 5 years* [44]
Individual with a history of transverse myelitis • Ordered by a neurologist or any provider in consultation with a neurologist	• MRI Cervical Spine without and with contrast (CPT® 72156) **OR** • MRI Cervical Spine without contrast (CPT® 72141) **AND/OR** • MRI Thoracic Spine without and with contrast (CPT® 72157) **OR** • MRI Thoracic Spine without contrast (CPT® 72146) **AND/OR** • MRI Brain without and with contrast (CPT® 70553) **OR** • MRI Brain without contrast (CPT® 70551)

- Individuals with transverse myelitis present with various symptoms of sensory, motor and/or autonomic dysfunction.
  - Bilateral signs and/or symptoms (although not necessarily symmetrical)[42]
  - Examination findings may include but are not limited to any of the following:
    - Bilateral limb weakness
    - Loss of manual dexterity
    - New or worsening foot drop
    - Sensory abnormalities
    - Sensory level
    - Hyperreflexia (including upgoing toes, positive Babinski, Hoffman's sign, clonus)
    - Gait abnormality (spastic or ataxic gait)
    - See also: **Background and Supporting Information**
  - If inflammation is identified within the spinal cord suggestive of transverse myelitis, a brain MRI is recommended to evaluate for a multifocal inflammatory process[42]
- See **Multiple Sclerosis (MS) (HD-16.1)**, **Neuromyelitis Optica and NMO Spectrum Disorders (HD-16.2)**, **MOG Antibody-Associated Diseases (MOGAD) (HD-16.3)**

Background and Supporting Information

- Symptoms may include but are not limited to the following:
  - Motor weakness of a limb or limbs, including paraparesis and/or complete paralysis

  - Change in sensation in a limb or limbs that may be associated with paresthesias and/or dysthesias.
  - Urinary urgency, incontinence and/or urinary retention
  - Worsening constipation and/or bowel urgency/incontinence
  - Sexual dysfunction
  - Lhermitte's sign
  - New or worsening spasticity
- Acute transverse myelitis is defined as an acute inflammatory syndrome leading to motor and/or sensory impairment, with or without sphincter dysfunction, secondary to a variety of autoimmune or inflammatory diseases.[42]
- Diagnosed by spinal MRI and/or cerebrospinal fluid.
- Individuals typically progress to maximal neurological deficits within 4 weeks.
- Longitudinally extensive transverse myelitis (≥ 3 vertebral segments) is more commonly associated with neuromyelitis optica spectrum disorders (NMOSD) and/or MOG antibody-associated diseases (MOGAD)[34,44]
- Transverse myelitis:
  - May be idiopathic
  - Initial event of multiple sclerosis (see **Multiple Sclerosis (MS) (HD-16.1)**
  - Initial event of neuromyelitis optica spectrum disorder (NMOSD) (see **Neuromyelitis Optica and NMO Spectrum Disorders (HD-16.2)**)
  - Initial event of MOG antibody-associated disease (MOGAD) (see **MOG Antibody-Associated Diseases (MOGAD) (HD-16.3)**)
  - May be associated with connective tissue disease
    - Systemic lupus erythematous (SLE)
    - Rheumatoid Arthritis (RA)
    - Sjögren's syndrome
    - Systemic sclerosis
  - Manifestation of neurosarcoidosis (see **Autoimmune/Paraneoplastic Encephalitis & Neuroinflammatory Disorders (HD-14.3)**)
  - Post-infectious and/or post-vaccination related
    - COVID-19 and COVID-19 post-vaccination myelitis cases have been reported (see **Neuro-COVID-19 and Sars-COV-2 Vaccines (HD-14.2)**)
    - May have a prodromal syndrome with fever, respiratory and/or gastrointestinal symptoms[40]
    - Neurologic symptoms may be associated with headache, neck stiffness or recurrence of fever[40]

References (HD-16)

v2.0.2024

1. Wattjes MP, Ciccarelli O, Reich DS, et al. 2021 MAGNIMS-CMSC-NAIMS consensus recommendations on the use of MRI in patients with multiple sclerosis. *The Lancet Neurology*. 2021;20(8):653-670. doi:10.1016/S1474-4422(21)00095-8
2. Thompson AJ, Banwell BL, Barkhof F, et al. Diagnosis of multiple sclerosis: 2017 revisions of the McDonald criteria. *The Lancet Neurology*. 2018;17(2):162-173. doi:10.1016/s1474-4422(17)30470-2
3. Kaunzner UW, Gauthier SA. MRI in the assessment and monitoring of multiple sclerosis: an update on best practice. *Therapeutic Advances in Neurological Disorders*. 2017;10(6):247-261. doi:10.1177/1756285617708911
4. FDA Drug Safety Communication: New risk factor for Progressive Multifocal Leukoencephalopathy (PML) associated with Tysabri (natalizumab). Originally issued February 13, 2018. U S Food and Drug Administration Home Page. https://www.fda.gov/drugs/drug-safety-and-availability/fda-drug-safety-communication-new-risk-factor-progressive-multifocal-leukoencephalopathy-pml
5. Rae-Grant A, Day GS, Marrie RA, et al. Comprehensive systematic review summary: Disease-modifying therapies for adults with multiple sclerosis. *Neurology*. 2018;90(17):789-800. doi:10.1212/wnl.0000000000005345
6. Shosha E, Dubey D, Palace J, et al. Area postrema syndrome. *Neurology*. 2018;91(17). doi:10.1212/wnl.0000000000006392
7. Wingerchuk DM, Banwell B, Bennett JL, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. *Neurology*. 2015;85(2):177-189. doi:10.1212/wnl.0000000000001729
8. Kaunzner UW, Gauthier SA. MRI in the assessment and monitoring of multiple sclerosis: an update on best practice. *Therapeutic Advances in Neurological Disorders*. 2017;10(6):247-261. doi:10.1177/1756285617708911
9. Expert Panel on Neurologic Imaging:, Kennedy TA, Corey AS, et al. ACR Appropriateness Criteria® Orbits Vision and Visual Loss. *J Am Coll Radiol*. 2018;15(5S):S116-S131. doi:10.1016/j.jacr.2018.03.023
10. Hornby PJ. Central neurocircuitry associated with emesis. *The American Journal of Medicine*. 2001;111(8):106-112. doi:10.1016/s0002-9343(01)00849-x
11. Ciccarelli O, Cohen JA, Reingold SC, et al. Spinal cord involvement in multiple sclerosis and neuromyelitis optica spectrum disorders. *The Lancet Neurology*. 2019;18(2):185-197. doi:10.1016/s1474-4422(18)30460-5
12. Ciron J, Audoin B, Bourre B, et al. Recommendations for the use of Rituximab in neuromyelitis optica spectrum disorders. *Revue Neurologique*. 2018;174(4):255-264. doi:10.1016/j.neurol.2017.11.005
13. Rudie JD, Mattay RR, Schindler M, et al. An Initiative to Reduce Unnecessary Gadolinium-Based Contrast in Multiple Sclerosis Patients. *Journal of the American College of Radiology*. 2019;16(9):1158-1164. doi:10.1016/j.jacr.2019.04.005
14. Major EO. Progressive Multifocal Leukoencephalopathy Lesions and JC Virus. *JAMA Neurology*. 2018;75(7):789. doi:10.1001/jamaneurol.2018.0004
15. Vukusic S, Rollot F, Casey R, et al. Progressive Multifocal Leukoencephalopathy Incidence and Risk Stratification Among Natalizumab Users in France. *JAMA Neurology*. 2020;77(1):94. doi:10.1001/jamaneurol.2019.2670
16. Rae-Grant A, et al. Practice guideline: Disease-modifying therapies for adults with multiple sclerosis. . Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology. https://download.lww.com/wolterskluwer_vitalstream_com/PermaLink/WNL/A/WNL_2018_04_19_RAEGRANT_NEUROLOGY2017835181R1_SDC3.pdf
17. Wattjes MP, Barkhof F. Diagnosis of natalizumab-associated progressive multifocal leukoencephalopathy using MRI. *Current Opinion in Neurology*. 2014;27(3):260-270. doi:10.1097/wco.0000000000000099
18. Bloomgren G, Richman S, Hotermans C, et al. Risk of Natalizumab-Associated Progressive Multifocal Leukoencephalopathy. *New England Journal of Medicine*. 2012;366(20):1870-1880. doi:10.1056/nejmoa1107829
19. Hegen H, Reindl M. Recent developments in MOG-IgG associated neurological disorders. *Ther Adv Neurol Disord*. 2020 Jul 31;13:1756286420945135. doi: 10.1177/1756286420945135
20. De Stefano N, Battaglini M, Pareto D, et al. MAGNIMS recommendations for harmonization of MRI data in MS multicenter studies. Neuroimage Clin. 2022;34:102972. doi:10.1016/j.nicl.2022.102972
21. Reich DS, Lucchinetti CF, Calabresi PA. Multiple Sclerosis. N Engl J Med. 2018;378(2):169-180. doi:10.1056/NEJMra1401483
22. Lopez Chiriboga S, Flanagan EP. Myelitis and Other Autoimmune Myelopathies. *CONTINUUM: Lifelong Learning in Neurology*. 2021;27(1):62-92. doi:10.1212/con.0000000000000900

23. Genovese AV, Hagemeier J, Bergsland N, et al. Atrophied Brain T2 Lesion Volume at MRI Is Associated with Disability Progression and Conversion to Secondary Progressive Multiple Sclerosis. *Radiology*. 2019;293(2):424-433. doi:10.1148/radiol.2019190306
24. Jakimovski D, Zivadinov R, Bergsland N, Ramasamy DP, Hagemeier J, Genovese AV, Hojnacki D, Weinstock-Guttman B, Dwyer MG. Clinical feasibility of longitudinal lateral ventricular volume measurements on T2-FLAIR across MRI scanner changes. *Neuroimage Clin*. 2021;29:102554. doi: 10.1016/j.nicl.2020.102554
25. Saslow L, Li DKB, Halper J, et al. An International Standardized Magnetic Resonance Imaging Protocol for Diagnosis and Follow-up of Patients with Multiple Sclerosis. *International Journal of MS Care*. 2020;22(5):226-232. doi:10.7224/1537-2073.2020-094
26. Berger B, Hottenrott T, Rauer S, Stich O. Screening for onconeural antibodies in neuromyelitis optica spectrum disorders. *BMC Neurology*. 2017;17(1). doi:10.1186/s12883-016-0779-9
27. Carnero Contentti E, Correale J. Neuromyelitis optica spectrum disorders: from pathophysiology to therapeutic strategies. *Journal of Neuroinflammation*. 2021;18(1). doi:10.1186/s12974-021-02249-1
28. Juryńczyk M, Weinshenker B, Akman-Demir G, et al. Status of diagnostic approaches to AQP4-IgG seronegative NMO and NMO/MS overlap syndromes. *Journal of Neurology*. 2015;263(1):140-149. doi:10.1007/s00415-015-7952-8
29. Winkelmann A, Loebermann M, Reisinger EC, Hartung H-P, Zettl UK. Disease-modifying therapies and infectious risks in multiple sclerosis. *Nature Reviews Neurology*. 2016;12(4):217-233. doi:10.1038/nrneurol.2016.21
30. Gastaldi M, Marchioni E, Banfi P, et al. Predictors of outcome in a large retrospective cohort of patients with transverse myelitis. Mult Scler. 2018;24(13):1743-1752. doi:10.1177/1352458517731911
31. Lavi ES, Pal A, Bleicher D, Kang K, Sidani C. MR Imaging of the Spine: Urgent and Emergent Indications. Semin Ultrasound CT MR. 2018;39(6):551-569. doi:10.1053/j.sult.2018.10.006
32. Sarbu N, Lolli V, Smirniotopoulos JG. Magnetic resonance imaging in myelopathy: a pictorial review. Clin Imaging. 2019;57:56-68. doi:10.1016/j.clinimag.2019.05.002 29
33. Stern BJ, Royal W 3rd, Gelfand JM, et al. Definition and Consensus Diagnostic Criteria for Neurosarcoidosis: From the Neurosarcoidosis Consortium Consensus Group. JAMA Neurol. 2018;75(12):1546-1553. doi:10.1001/jamaneurol.2018.2295
34. Banwell B, Bennett JL, Marignier R, et al. Diagnosis of myelin oligodendrocyte glycoprotein antibody-associated disease: International MOGAD Panel proposed criteria.*Lancet Neurol*. 2023;22(3):268-282. doi:10.1016/S1474-4422(22)00431-8
35. Holmoy T, Høglund RA, Illes Z, Myhr KM, Torkildsen Ø. Recent progress in maintenance treatment of neuromyelitis optica spectrum disorder.*J Neurol*. 2021;268(12):4522-4536. doi:10.1007/s00415-020-10235-5
36. Ismail II, Salama S. Association of CNS demyelination and COVID-19 infection: an updated systematic review. *J Neurol*. 2022:269(2):541-576. doi:10.1007/s00415-021-10752-x
37. Jarius S, Aktas O, Ayzenberg I, et al. Update on the diagnosis and treatment of neuromyelits optica spectrum disorders (NMOSD)- revised recommendations of the Neuromyelitis Optica Study Group (NEMOS). Part I: Diagnosis and differential diagnosis. (published online ahead of print, 2023 Apr 6).*J Neurol*. 2023;10.1007/s00415-023-11634-0. doi:10.1007/s00415-023-11634-0
38. Khan E, Shrestha AK, Colantonio MA, Liberio RN, Sriwastava S. Acute transverse myelitis following SARS-CoV-2 vaccination: a case report and review of the literature. *J Neurol*. 2022.269(3):1121-1132. doi:10.1007/s00415-021-10785-2
39. Marrodan M, Hernandez MA, Kohler AA, Correale J. Differential diagnosis in acute inflammatory myelitis. *Multi Scler Relat Disord* 2020:46:102481. doi doi:10.1016/j.msard.2020.102481
40. Murphy OC, Messacar K, Benson L, et al. Acute flaccid myelitis: cause, diagnosis, and management. Lancet. 2021:397(10271):334-346. doi:10.1016/S0140-6736(20)32723-9
41. Okuda DT, Kantarci O, Lebrun-Frénay C, et al. Dimethyl Fumarate Delays Multiple Sclerosis in Radiologically Isolated Syndrome.*Ann Neurol*. 2023;93(3):604-614. doi:10.1002/ana.26555
42. Transverse Myelitis Consortium Working Group. Proposed diagnostic criteria and nosology of acute transverse myelitis.*Neurology*. 2002;59(4):499-505. doi:10.1212/wnl.59.4.499
43. Lublin FD, Reingold SC, Cohen JA, et al. Defining the clinical course of multiple sclerosis: the 2013 revisions.*Neurology*. 2014;83(3):278-286. doi:10.1212/WNL.0000000000000560
44. Bulut E, Shoemaker T, Karakaya J, et al. MRI Predictors of Recurrence and Outcome after Acute Transverse Myelitis of Unidentified Etiology.*AJNR Am J Neuroradiol*. 2019;40(8):1427-1432. doi:10.3174/ajnr.A6121
45. Tillema JM. Imaging of Central Nervous System Demyelinating Disorders.*Continuum (Minneap Minn)*. 2023;29(1):292-323. doi:10.1212/CON.0000000000001246
46. Kartau M, Sipilä JO, Auvinen E, Palomäki M, Verkkoniemi-Ahola A. Progressive Multifocal Leukoencephalopathy: Current Insights. Degener Neurol Neuromuscul Dis. 2019;9:109-121. Published 2019 Dec 2. doi:10.2147/DNND.S203405

Papilledema/ Pseudotumor Cerebri (HD-17)

Papilledema/Pseudotumor Cerebri (HD-17.1)

HD.PP.0017.1.A
v2.0.2024

- See **Eye Disorders and Visual Loss (HD-32.1)**
- Papilledema and Pseudotumor Cerebri (Idiopathic Intracranial Hypertension):
  - MRI Orbits/Face/Neck without contrast (CPT® 70540) **OR** MRI Orbits/Face/Neck without and with contrast (CPT® 70543) **OR** CT Orbits/Temporal bone with contrast (CPT® 70481) **OR** CT Orbit/Temporal bone without contrast (CPT® 70480) **AND/OR** MRI Brain without contrast (CPT® 70551) **OR** MRI Brain with and without contrast (CPT® 70553):
    - Suspected elevated intracranial pressure **AND/OR** papilledema
    - CT Head without contrast (CPT® 70450) can be approved when MRI is contraindicated or for urgent evaluation
    - See **General Guidelines – CT Head (HD-1.4)** regarding required use of CT Head prior to lumbar puncture and/or spinal tap.
    - 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) may be added for venogram when requested.[2]
    - 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)**

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

References (HD-17)

HD.PP.0017.2.A

v2.0.2024

1. Friedman DI. Papilledema and Idiopathic Intracranial Hypertension. CONTINUUM: Lifelong Learning in Neurology. 2014;20:857-876. doi:10.1212/01.con.0000453314.75261.66
2. Expert Panel on Neurologic Imaging, Whitehead MT, Cardenas AM, et al. ACR Appropriateness Criteria® Headache. J Am Coll Radiol. 2019;16(11S):S364-S377. doi:10.1016/j.jacr.2019.05.030
3. Thurtell MJ. Idiopathic Intracranial Hypertension. CONTINUUM: Lifelong Learning in Neurology. 2019;25(5):1289-1309. doi:10.1212/con.0000000000000770
4. Wall M. Update on Idiopathic Intracranial Hypertension. *Neurologic Clinics*. 2017;35(1):45-57. doi:10.1016/j.ncl.2016.08.004
5. Costello F, Scott JN. Imaging in Neuro-ophthalmology. CONTINUUM: Lifelong Learning in Neurology. 2019;25(5):1438-1490. doi:10.1212/con.0000000000000783
6. Aylward SC, Reem RE. Pediatric Intracranial Hypertension. *Pediatr Neurol*. 2017 Jan;66:32-43. doi: 10.1016/j.pediatrneurol.2016.08.010

Paresthesias and/or Weakness (HD-18)

Sensory/Weakness Complaints (HD-18.1)

HD.PS.0018.1.A
v2.0.2024

Advanced imaging for complaints of sensory loss and/or paresthesias (see **Background and Supporting Information**) and/or weakness that are unaccompanied by other symptoms and not preceded by trauma must have the following: a thorough clinical history and a detailed neurological exam (including the symptomatic area).

Imaging for sensory and weakness complaints may be indicated with the following findings:

Findings Specific to the Brain and/or Spinal Cord	Supported Imaging

ANY of the following:
• Hyperreflexia
• Babinski/Hoffman sign*
• Increased tone in affected limb
• Bladder and/or bowel dysfunction[4]
• Motor symptoms in ANY of the following patterns:
  • Two limbs on same side of body
  • Face and limb involvement
• Sensory symptoms in ANY of the following patterns:
  • Two limbs on same side of body
  • Face and limb involvement
*See **Background and Supporting Information**
	• MRI Brain without contrast (CPT® 70551) **OR**
	• MRI Brain without and with contrast (CPT® 70553)
	AND/OR
	• MRI Cervical Spine without contrast (CPT® 72141) **OR**
	• MRI Cervical Spine without and with contrast (CPT® 72156)
	AND/OR
	• MRI Thoracic Spine without contrast (CPT® 72146) **OR**
	• MRI Thoracic Spine without and with contrast (CPT® 72157)

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Findings Specific to the Spinal Cord	Supported Imaging
ANY of the following: • Decreased pinprick sensation on one side of the body with weakness and decreased proprioception on the other side • Sensory level (also called spinal cord level) on the trunk with sensory loss in both legs • Tight band around the trunk or torso[4] • Pure sensory symptoms with proximal and distal involvement and a symmetric pattern • Decreased or absent reflexes AND noted concern for spinal cord shock or acute spinal cord injury*[4] *See **Background and Supporting Information**	• MRI Cervical Spine without contrast (CPT® 72141) **OR** • MRI Cervical Spine without and with contrast (CPT® 72156) **AND/OR** • MRI Thoracic Spine without contrast (CPT® 72146) **OR** • MRI Thoracic Spine without and with contrast (CPT® 72157)

Findings Specific to the Terminal End of the Spinal Cord	Supported Imaging

Concern for conus medullaris syndrome.*
• Symptoms may include, but are not limited to:
  • Saddle anesthesia
  • Urinary retention
  • Bowel incontinence
  • Lower limb paresthesias
  • Lower limb weakness
 	
*See **Background and Supporting Information**
	• MRI Lumbar Spine without contrast (CPT® 72148) **OR**
	• MRI Lumbar Spine without and with contrast (CPT® 72158)

- MRI Lumbar Spine is not typically indicated to visualize the spinal cord except in the clinical scenarios noted above. MRI Cervical Spine and MRI Thoracic Spine will image the entire spinal cord.
- Findings NOT consistent with central nervous system localization and NOT supporting brain or spinal cord imaging include:

  - Sensory loss that involves the hands and feet and not the trunk
  - Limb pain
- For symptoms after trauma, refer to **Head Trauma (HD-13.1)** and/or the appropriate level in the Spine Imaging Guidelines
- For generalized weakness, polyneuropathy, and/or other patterns of sensory and/or motor symptoms not referenced above, refer to the following guidelines:
  - 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 Disorders, see **Neuromuscular Junction Disorders (PN-6.1)**
  - Multifocal Motor Neuropathy (MMN) and Chronic Inflammatory Demyelinating Polyneuropathy (CIDP), see **Polyneuropathy (PN-3.1)**
  - Polyneuropathy, see **Polyneuropathy (PN-3.1)**
  - Neuropathy with concern for malignancy, see **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines
  - Proximal asymmetric and concern for plexopathy, see **Brachial Plexus (PN-4.1)** and/or **Lumbar and Lumbosacral Plexus (PN-5.1)**
  - Sensory and/or motor symptoms localized to a single nerve, see **Focal Neuropathy (PN-2.1)**
  - Thoracic Outlet Syndrome, see **Thoracic Outlet Syndrome (CH-31.1)** in the Chest Imaging Guidelines
  - Radiculopathy, see appropriate level in the Spine Imaging Guidelines
  - Cauda Equina Syndrome, see **Red Flag Indications (SP-1.2)** in the Spine 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 nervous system (PNS) or central nervous system (CNS).
- A thorough clinical history, including symptom location and time course, can be helpful to differentiate PNS pathologies from CNS. For example, paresthesia affecting one side of the face and/or body (i.e. hemisensory deficit) points strongly towards central nervous system dysfunction. Therefore, brain and/or spinal cord imaging may be supported based on the location of symptoms. Typically, lumbar spine imaging is not supported 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 cases, NCS/EMG testing results should be

completed prior to advanced imaging. (See **Peripheral Nerve Imaging Guidelines)**.

- Upper motor neuron signs (e.g. increased tone, hyperreflexia, presence of Babinski or Hoffman signs) may support a need for central nervous system imaging.
- Lower motor neuron signs (e.g. decreased tone, hypo- or areflexia, muscle atrophy) may support evaluation for peripheral nervous system diseases. Nerve conduction and needle EMG testing should be completed prior to advanced imaging.
- 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 may be supported regardless of NCS/EMG testing results (see **Polyneuropathy (PN-3.1)** in the Peripheral Nerve Disorders (PND) Imaging Guidelines).
- Babinski sign - presence of an upgoing big toe with stimulation of the lateral plantar region of the foot.[14]
- Hoffman sign - involuntary flexion of the fingers, particularly the thumb and index fingers, triggered by flicking the distal segment of the middle finger.[14]
- Spinal cord shock/acute spinal cord injury - occurs after hyperacute or acute injury to the cord and presents with flaccid areflexia below the level of injury. May be associated with hypotension and/or bradycardia if loss of sympathetic tone occurs. Signs may last from days to weeks before upper motor neuron findings develop.[4]
- Conus Medullaris Syndrome - compressive damage to the spinal cord from T12-L2. Symptoms suggestive of conus medullaris syndrome include saddle anesthesia, urinary retention, bowel incontinence, and/or lower extremity motor or sensory changes.[13]

References (HD-18)

HD.PS.0018.2.A
v2.0.2024

1. Paresthesia Information Page. National Institute of Neurological Disorders and Stroke. https://www.ninds.nih.gov/Disorders/All-Disorders/Paresthesia-Information-Page
2. Levin MC, By, Professional.Manuals.TopicPage.LastRevisionDate| Content last modified Jan 2019. Numbness - Neurologic Disorders. Merck Manuals Professional Edition. https://www.merckmanuals.com/professional/neurologic-disorders/symptoms-of-neurologic-disorders/numbness
3. London ZN. A Structured Approach to the Diagnosis of Peripheral Nervous System Disorders. CONTINUUM: Lifelong Learning in Neurology. 2020;26(5):1130-1160. doi:10.1212/con.0000000000000922
4. Hardy TA. Spinal Cord Anatomy and Localization. CONTINUUM: Lifelong Learning in Neurology. 2021;27(1):12-29. doi:10.1212/con.0000000000000899
5. Larson ST and Wilbur J. Muscle Weakness in Adults: Evaluation and Differential Diagnosis. Am Fam Physician. 2020;101(2):95-108
6. Filippakis A, Jara J, Ventura N, Scala S, Scopa C, Ruthazer R, Karakis I, Srinivasan J, Russell JA, Ho DT. A prospective study of benign fasciculation syndrome and anxiety. Muscle & nerve. 2018 Dec;58(6):852-4
7. Callaghan BC, Price RS, Feldman EL. Distal Symmetric Polyneuropathy. JAMA. 2015;314(20):2172. doi:10.1001/jama.2015.13611
8. Hughes R. Investigation of peripheral neuropathy. BMJ. 2010;341(nov05 1):c6100-c6100. doi:10.1136/bmj.c6100.6
9. Campbell WW. DeJong's The Neurologic Examination, 7th ed, Lippincott Williams & Wilkins, Philadelphia 2013
10. Wattjes MP, Ciccarelli O, Reich DS, et al. 2021 MAGNIMS-CMSC-NAIMS consensus recommendations on the use of MRI in patients with multiple sclerosis. The Lancet Neurology. 2021;20(8):653-670. doi:10.1016/S1474-4422(21)00095-8
11. Roth CJ, Angevine PD, Aulino JM, et. al. Expert Panel on Neurologic Imaging. ACR Appropriateness Criteria®: Myelopathy. American College of Radiology (ACR); Date of Origin: 1996. Last Review: 2020. https://acsearch.acr.org/docs/69484/Narrative/
12. Bykowski J, Aulino JM, Berger KL, et al. (2016). ACR Appropriateness Criteria® Plexopathy. American College of Radiology (ACR).
13. Rider LS, Marra EM. Cauda Equina and Conus Medullaris Syndromes. 2022 Aug 8. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan-.
14. Shahrokhi M, Asuncion RMD. Neurologic Exam. 2023 Jan 16. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan-.

Pituitary (HD-19)

Pituitary (HD-19.1)

HD.PT.0019.1.A
v2.0.2024

- Endocrine laboratory studies should be performed prior to considering advanced imaging, except in the cases of stable, non-functioning microadenomas or macroadenomas, cysts and/or for incidentally found lesions.
- 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 pre-operative evaluation of certain sellar tumors and for pre-operative 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
- See **Secondary Amenorrhea (PV-3.1)** in the Pelvic Imaging Guidelines for initial lab and imaging work up to exclude other causes. See Female Hypogonadism or Prolactinoma or other relevant sections in the grid if suspicion for pituitary tumor/disease.

Pituitary Imaging

Indication	Initial Imaging	Repeat Imaging
Microadenoma: Nonfunctioning, unexplained pituitary asymmetries, or incidentally found small tumors (<10 mm)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)	• 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
Macroadenoma (≥10 mm): Nonfunctioning and/ or not surgically removed including those with a post-operative remnant	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)	• 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)

Acromegaly* (Elevated IGF-1 confirmed by lack of suppression of growth hormone on glucose suppression testing)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)
		  • At least 12 weeks after surgery to evaluate for residual tumor
		  • If treated with Pegvisomant, 6 to 12 months after treatment initiated, then annually if stable
		  • 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

Indication	Initial Imaging	Repeat Imaging

Cushing's Disease** (Pituitary ACTH excess leading to hypercortisolism)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)
		  • At least 12 weeks after surgery as new baseline
		  • Annually after bilateral adrenalectomy for Cushing's disease or ectopic ACTH production
		  • 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

Rathke's cleft cyst/ Simple cyst	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)	• 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.

Indication	Imaging

Prolactinomas* **	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) with:
	  • Diagnosis:
	  • Unexplained prolactin level above the normal range
	  • On Dopamine Agonist (DA) therapy with good response:
	  • Macroadenomas 3 months after start of DA therapy
	  • Microadenomas 1 year after start of DA therapy
	  • To decide on stoppage of therapy after ~2 years if in “remission” (normal PRL and no visible tumor on MRI)
	  • On Dopamine Agonist therapy with suboptimal response:
	  • PRL levels rise
	  • New symptoms develop (galactorrhea, vision changes, headaches, pituitary deficiency)
	  • If on high dose maximal DA and no plans for surgery/radiation therapy use guideline for microadenoma or macroadenoma
	  • After Dopamine Agonist therapy:
	  • Rise in PRL level
	  • For DA stoppage at menopause, use guideline for microadenoma or macroadenoma
	  • Galactorrhea/nipple discharge with normal prolactin and thyroid function levels: See **Nipple Discharge/Galactorrhea (BR-6.1)** in the Breast Imaging Guidelines

Medication-induced Prolactinemia**	• 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[22]

Indication	Imaging
TSH, FSH, or LH producing adenomas (inappropriate pituitary hypersecretion of TSH, FSH or LH)***	• 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. • Refer to appropriate post-operative, or Microadenoma/Macroadenoma guidelines based on the size of the lesion and initial management. • 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

Male Hypogonadism****	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) if **ONE** of the following:
	  • Severe secondary hypogonadism (as indicated by morning serum testosterone level <150 ng/dl and low or normal LH and FSH levels) (See **Background and Supporting Information**)
	  • 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:
	  • Panhypopituitarism
	  • Hyperprolactinemia
	  • Signs of tumor mass effect (headache, visual impairment, or visual field deficit)
	  • Elevated sex hormone binding globulin (SHBG)

Female Hypogonadism (Secondary Amenorrhea may be a feature)[25]	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) for normal or low FSH with low estradiol (LH may be normal or low also)

Indication	Imaging
Growth Hormone Deficiency (Adult onset) [25]	MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) for the following: • Low Growth Hormone (GH) **OR** • Low IGF-1 **AND** • One abnormal provocative test (likely will be Glucagon Stimulation test as GNRH is unavailable and Insulin Tolerance test poses risks) • If 3 or more pituitary hormones are deficient (including GH), then provocative test is not needed.
Secondary (Central) Adrenal Insufficiency [25]	MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) for the following: • ACTH is low or normal at 10 or lower **AND** • Low baseline cortisol level < 3 µg/dL **OR** • abnormal ACTH stimulation test with suboptimal cortisol stimulation where cortisol does not reach above 18 µg/dL
Central Hypothyroidism [25]	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) for the following: • Low free T4 with normal, low or mildly elevated TSH
Hypopituitarism **(deficiency of one or more pituitary hormones)**	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551)

Indication	Initial Imaging	Repeat Imaging for Non-Operative Care
Diabetes Insipidus (DI)	• MRI Brain without and with contrast (CPT® 70553) or MRI Brain without contrast (CPT® 70551) if: • Laboratory testing consistent with DI (serum osmolality should be high and urine osmolality should be low) and etiology uncertain	NA

Syndrome of Inappropriate ADH (SIADH)	• MRI Brain without and with contrast (CPT® 70553)) or MRI Brain without contrast (CPT® 70551) if:
	  • Etiology remains uncertain or is thought to be in the nervous system;
	  • Urine osmolality should be high and serum osmolality low
		NA

Other Pituitary Region Tumors	• Evaluation may require CT in addition to MRI to evaluate for hyperostosis.	

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 or inappropriately normal in Cushing's disease and ectopic sources of ACTH production, but suppressed in other causes of hypercortisolism[26]. 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 pre-operatively 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.[22]
- *****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.[24]

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

HD.PT.0019.2.A
v2.0.2024

- 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 an Endocrinologist, Neurologist, Neurosurgeon, ENT, Ophthalmologist, Neuro-Ophthalmologist or Radiation Oncologist.
- 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 an Endocrinologist, Neurologist, Neurosurgeon, ENT, Ophthalmologist, Neuro-Ophthalmologist or Radiation Oncologist.

Empty Sella Turcica (HD-19.3)

HD.DPT.0019.3.A
v2.0.2024

- 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
v2.0.2024

- See **Craniopharyngioma and Other Hypothalamic/Pituitary Region Tumors (PEDONC-4.10)**

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

References (HD-19)

v2.0.2024

1. American Association of Clinical Endocrinologists Medical Guidelines for Clinical Practice for the Evaluation and Treatment of Hypogonadism in Adult Male Patients—2002 Update. *Endocrine Practice*. 2002;8(6):439-456. doi:10.4158/ep.8.6.439
2. Katznelson L, Laws ER, Melmed S, et al. Acromegaly: An Endocrine Society Clinical Practice Guideline. *The Journal of Clinical Endocrinology & Metabolism*. 2014;99(11):3933-3951. doi:10.1210/jc.2014-2700
3. Melmed S, Casanueva FF, Hoffman AR, et al. Diagnosis and Treatment of Hyperprolactinemia: An Endocrine Society Clinical Practice Guideline. *The Journal of Clinical Endocrinology & Metabolism*. 2011;96(2):273-288. doi:10.1210/jc.2010-1692
4. Hoang JK Hoffman AR, González RG, et al. Management of Incidental Pituitary Findings on CT, MRI, and 18 F-Fluorodeoxyglucose PET: A White Paper of the ACR Incidental Findings Committee. *Journal of the American College of Radiology*. 2018;15(7):966-972. doi:10.1016/j.jacr.2018.03.037
5. Marinis LD, Bonadonna S, Bianchi A, Maira G, Giustina A. Primary Empty Sella. *The Journal of Clinical Endocrinology & Metabolism*. 2005;90(9):5471-5477. doi:10.1210/jc.2005-0288
6. Chiloiro S, Giampietro A, Bianchi A, et al. DIAGNOSIS OF ENDOCRINE DISEASE: Primary empty sella: a comprehensive review. *European Journal of Endocrinology*. 2017;177(6). doi:10.1530/eje-17-0505
7. Freda PU, Beckers AM, Katznelson L, et al. Pituitary Incidentaloma: An Endocrine Society Clinical Practice Guideline. *The Journal of Clinical Endocrinology & Metabolism*. 2011;96(4):894-904. doi:10.1210/jc.2010-1048
8. Expert Panel on Neurologic Imaging:, Burns J, Policeni B, et al. ACR Appropriateness Criteria® Neuroendocrine Imaging. J Am Coll Radiol. 2019;16(5S):S161-S173. doi:10.1016/j.jacr.2019.02.017
9. Thompson CJ et al.eds. Melmed S et al. Chapter 10: Posterior Pituitary. In: *Williams Textbook of Endocrinology*, 14th ed., 2019: 303-330
10. Cooke DW et al.eds. Melmed S et al. Chapter 25: Normal and Aberrant Growth in Children. In: *Williams Textbook of Endocrinology*, 14th ed. 2019: 937-1022
11. Styne DM. eds. Melmed S et al. Chapter 26: Physiology and Disorders of Puberty. In: *Williams Textbook of Endocrinology*, 14th ed. 2019: 1023-1164
12. Bhasin S, Brito JP, Cunningham GR, et al. Testosterone Therapy in Men With Hypogonadism: An Endocrine Society* Clinical Practice Guideline. *The Journal of Clinical Endocrinology & Metabolism*. 2018;103(5):1715-1744. doi:10.1210/jc.2018-00229
13. Chen CC, Carter BS, Wang R, et al. Congress of Neurological Surgeons Systematic Review and Evidence-Based Guideline on Preoperative Imaging Assessment of Patients With Suspected Nonfunctioning Pituitary Adenomas. *Neurosurgery*. 2016;79(4). Pp E524-526. doi:10.1227/neu.0000000000001391
14. Nieman LK, Biller BMK, Findling JW, et al. Treatment of Cushing's Syndrome: An Endocrine Society Clinical Practice Guideline. *The Journal of Clinical Endocrinology & Metabolism.* 2015;100(8):2807-2831. doi:10.1210/jc.2015-1818
15. Woodmansee WW, Carmichael J, Kelly D, Katznelson L. American Association Of Clinical Endocrinologists And American College Of Endocrinology Disease State Clinical Review: Postoperative Management Following Pituitary Surgery. *Endocrine Practice*. 2015;21(7):832-838. doi:10.4158/ep14541.dscr
16. Ziu M, Dunn IF, Hess C, et al. Congress of Neurological Surgeons Systematic Review and Evidence-Based Guideline on Posttreatment Follow-up Evaluation of Patients With Nonfunctioning Pituitary Adenomas. *Neurosurgery*. 2016;79(4):E541-E543. doi:10.1227/neu.0000000000001392
17. Jane JA, Jr. Surgical Treatment of Pituitary Adenomas. (Updated 10/4/2019). In: Feingold KR, Anawalt B, Boyce A, et al. eds. Endotext [Internet]. South Dartmouth (MA): MD Text com, Inc; 2000
18. Cardinale F, Pero G, Quilici L, et al. Cerebral Angiography for Multimodal Surgical Planning in Epilepsy Surgery: Description of a New Three-Dimensional Technique and Literature Review. *World Neurosurgery*. 2015;84(2):358-367. doi:10.1016/j.wneu.2015.03.028
19. Prevedello D, Otto B, Carrau R, de Lara D, Ditzel Filho LeoFS. Application of Image Guidance in Pituitary Surgery. *Surgical Neurology International*. 2012;3(3):73. doi:10.4103/2152-7806.95418
20. Guo Z, Liu C, Hou H, et al. Preoperative Computed Tomography (CT) Evaluation of Anatomical Abnormalities in Endonasal Transsphenoidal Approach in Pituitary Adenoma. *Medical Science Monitor*. 2018;24:1268-1275. doi:10.12659/msm.904402
21. Aghi MK, Chen CC, Fleseriu M, et al. Congress of Neurological Surgeons Systematic Review and Evidence-Based Guidelines on the Management of Patients With Nonfunctioning Pituitary Adenomas. *Neurosurgery*. 2016;79(4):521-523. doi:10.1227/neu.0000000000001386
22. Samperi I, Lithgow K, Karavitaki N. Hyperprolactinaemia. *Journal of Clinical Medicine*. 2019;8(12):2203. doi:10.3390/jcm8122203

23. Esposito D, Olsson DS, Ragnarsson O, Buchfelder M, Skoglund T, Johannsson G. Non-functioning pituitary adenomas: indications for pituitary surgery and post-surgical management. *Pituitary*. 2019;22(4):422-434. doi:10.1007/s11102-019-00960
24. Persani L. Central Hypothyroidism: Pathogenic, Diagnostic, and Therapeutic Challenges. *The Journal of Clinical Endocrinology & Metabolism*. 2012;97(9):3068-3078. doi:10.1210/jc.2012-1616
25. Fleseriu M, Hashim IA, Karavitaki N, et al. Hormonal Replacement in Hypopituitarism in Adults: An Endocrine Society Clinical Practice Guideline.*J Clin Endocrinol Metab*. 2016;101(11):3888-3921. doi:10.1210/jc.2016-2118
26. Sharma ST; AACE Adrenal Scientific Committee. AN INDIVIDUALIZED APPROACH TO THE EVALUATION OF CUSHING SYNDROME.*Endocr Pract*. 2017;23(6):726-737. doi:10.4158/EP161721.RA

Scalp and Skull (HD-20)

Scalp and Skull Lesions (HD-20.1)

HD.SK.0020.1.A
v2.0.2024

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 indicated 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
- The following imaging is indicated for children and adults with Pott Puffy Tumor:
  - MRI Brain without and with contrast (CPT®70553) or CT Head without and with contrast (CPT® 70470)[4]
  - Repeat imaging is supported if requested by a neurologist, neurosurgeon, otolaryngologist (ENT) and/or oromaxillofacial surgery (OMS) or any provider coordinating care with a neurologist, neurosurgeon, otolaryngologist (ENT) and/or oromaxillofacial surgery (OMS)

Background and Supporting Information

Pott Puffy Tumor is an abscess involving the frontal bone with adjacent osteomyelitis as the result of a frontal sinus infection that spreads contiguously through the wall of the sinus or through hematogenous spread via the veins that drain sinus mucosa.[4]

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Skull Base Osteomyelitis (SBO) (HD-20.2)

HD.SK.0020.2.A
v2.0.2024

- 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

Background and Supporting Information

Skull based osteomyelitis is a rare complication of otitis externa. It occurs most commonly among the immunocompromised, older members (greater than 65 years of age) and members with diabetes.[5]

References (HD-20)

v2.0.2024

1. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck
2. Khan M, Quadri SQ, Kazmi A, et al. A comprehensive review of skull base osteomyelitis: Diagnostic and therapeutic challenges among various presentations. *Asian Journal of Neurosurgery*. 2018;13(4):959. doi:10.4103/ajns.ajns_90_17
3. Expert Panel on Neurologic Imaging:, Kirsch CFE, Bykowski J, et al. ACR Appropriateness Criteria® Sinonasal Disease. *J Am Coll Radiol*. 2017;14(11S):S550-S559. doi:10.1016/j.jacr.2017.08.041
4. Barnett RR, Piazza MG, Elton SW. Pediatric Neurosurgery in Primary Care: Masses of the Scalp and Skull in Children.*Pediatr Clin North Am*. 2021;68(4):743-757. doi:10.1016/j.pcl.2021.04.003
5. Treviño González JL, Reyes Suárez LL, Hernández de León JE. Malignant otitis externa: An updated review. Am J Otolaryngol. 2021 Mar-Apr;42(2):102894. doi: 10.1016/j.amjoto.2020.102894. Epub 2021 Jan 5. PMID: 33429178

Stroke/TIA (HD-21)

Stroke/TIA (HD-21.1)

HD.HL.0021.1.C
v2.0.2024

Indication	Supported Imaging
• Acute ischemic stroke (within the first 24 hours) • Transient ischemic attacks (TIA) • Hemorrhagic stroke • Subdural hemorrhage	**Any ANY** or **ALL** may be approved: • CT head without contrast (CPT® 70450) • CTA head (CPT® 70496) • CTA Neck (CPT® 70498) • CT Perfusion (CPT® 0042T)
Concern for new stroke or TIA (MRI is preferred for evaluation of stroke/TIA, with or without a previous CT head)	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553)
Contraindication to MRI	• CT head without contrast (CPT® 70450) **OR** • CT head without and with contrast (CPT® 70470)
Arterial Vascular Imaging supported for TIA/Stroke evaluation including dissection: • Supported concurrently with brain imaging • Both MRA or CTA Head and Neck are needed to visualize the posterior vertebrobasilar circulation for evaluation of vertebrobasilar stroke/TIA	• MRA head (CPT® 70544, CPT® 70545, or CPT® 70546) **OR** • CTA head (CPT® 70496) **AND/OR** • MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549) OR • CTA Neck (CPT® 70498)
Venous vascular imaging for evaluation of venous infarcts	• MR or CT Venography (MRA Head [CPT® 70544, CPT® 70545, or CPT® 70546] **OR** • CTA Head (CPT® 70496)
Cerebral Angiography for stroke evaluation	3D Rendering (CPT® 76377)

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Indication	Supported Imaging
Stroke in Pregnancy and other hypercoagulable states[43] • See arterial and venous vascular imaging studies above for vascular imaging request • See **Background and Supporting Information**	• MRI Brain without contrast (CPT® 70551) OR • CT head without contrast (CPT® 70450)
Amaurosis Fugax or Ocular Microembolism • May include optic nerve/retinal arterial or Hollenhorst plaques on exam	See above for TIA or New Stroke brain imaging options and vascular imaging
Repeat imaging for follow up and resolution of stroke or hemorrhage	As requested by a neurologist, neurosurgeon, or physiatrist (PM&R) or any provider in consultation with a neurologist, neurosurgeon or physiatrist
Reversible Cerebral Vasoconstriction Syndrome	See **Sudden Onset of Headache (HD-11.3)**
Neurologic signs and/or symptoms, including headaches, associated with COVID-19 infection and/or COVID-19 vaccination (Strokes may be arterial or venous)	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) See also **General Guidelines-CT head (HD-1.4), Abnormal Blood Clotting (HD-11.9)** and **Neuro-Covid-19 (HD-14.2)**
Adults with HbSS (Sickle cell disease) or HbSb Thalassemia	One time MRI brain without contrast (CPT® 70551) or without and with contrast (CPT® 70553) for screening to deter silent cerebral infarcts Follow up or repeat testing per Neurologist or Hematologist or in consultation with a Neurologist or Hematologist See also **Sickle Cell Disease (HD-21.6)**
Documented Stroke or TIA	Transcranial Doppler Studies

Indication	Supported Imaging
Moyamoya Disease, when surgery or other vascular intervention is being considered	• Radiopharmaceutical Localization Imaging SPECT (CPT® 78803 or CPT® 78830) with vasodilating agent acetazolamide (Diamox) Follow up or repeat testing per Neurologist or Neurosurgeon or in consultation with a Neurologist or Neurosurgeon See **Moyamoya Syndrome/Disease (HD-21.5)**
Evaluation of paradoxical venous thromboembolism in cryptogenic stroke with PFO	See **Acute Limb Swelling (PVD-12.2)** and **Cryptogenic Stroke (HD-21.3)**
Cerebral Amyloid Angiopathy (CAA)[31,32,38]	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **OR** • CT Head without contrast (CPT® 70450) Amyloid-PET Brain (CPT® 78811 or CPT® 78814) is considered not medically necessary investigational and experimental for stroke evaluation. See **Dementia PET (HD-8.2)**
Multisystem Smooth Muscle Syndrome/Smooth Muscle Dysfunction Syndrome	See **Multisystem Smooth Muscle Syndrome/Smooth Muscle Dysfunction Syndrome (HD-21.7)**

Background and Supporting Information

- Pregnancy is an independent risk factor for stroke. Additional risk factors are not required for assessment of a stroke/TIA with acute focal neurological deficits.
- Additional arterial and venous hypercoaguable states that impose a stroke risk include:
  - Antiphospholipid syndrome
  - Hyperhomocysteinemia
  - Factor V Leiden mutation
  - Prothrombin gene mutation
  - Protein S deficiency
  - Protein C deficiency
  - Anti-thrombin deficiency

Cryptogenic Stroke (HD-21.3)

HD.ST.0021.3.A
v2.0.2024

- 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
v2.0.2024

- 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:
  - Inability to retain new information, lasting for several hours with preservation of alertness and all other cognitive functions with repetitive queries and amnesia[39]
  - 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)**
- For suspected seizure, see **Epilepsy/Seizures (HD-9.1)**

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Moyamoya Syndrome/Disease (HD-21.5)

HD.ST.0021.5.C
v2.0.2024

Initial imaging for Moyamoya Syndrome/Disease

- .Below are indicated for initial evaluation of Moyamoya Syndrome/Disease[36]:
  - MRI Brain without contrast (CPT® 70551) **OR**
  - MRI Brain without and with contrast (CPT® 70553) **AND/OR**
  - MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **AND/OR**
  - MRA Neck (CPT® 70547, CPT® 70548 **OR** CPT® 70549)
    - If MRA is contraindicated or not readily available, then CTA Head (CPT® 70496) **AND/OR** CTA Neck (CPT® 70498) is/are supported

Repeat imaging for Moyamoya Syndrome/Disease [36]

- MRA Head (CPT® 70544, CPT® 70545, or OR CPT® 70546) every 12 months **AND/OR**
- MRA Neck (CPT® 70547, CPT® 70548 **OR** CPT® 70549)
  - If MRA is contraindicated or not readily available, then CTA Head (CPT® 70496) **AND/OR** CTA Neck (CPT® 70498) is/are supported
- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) every 12 months[33,36]
- Radiopharmaceutical Localization Imaging SPECT (CPT® 78803) with vasodilating agent acetazolamide (Diamox) challenge is supported when surgery or other vascular intervention is considered.
- 3D Rendering (CPT® 76377) with cerebral angiography to define the presence, location, and anatomy of intracranial and cervical vascular malformations.[22]
  - See **General Guidelines - Other Imaging Situations (HD-1.7)** and **3D Rendering (Preface-4.1)** in the Preface Imaging Guidelines[37]
- CT Perfusion (CPT® 0042T) **OR** MRI Perfusion (CPT® 70551 OR CPT® 70552 OR CPT® 70553)[51] indicated:
  - When requested by neurologist and/or neurosurgeon
  - Prior to change in treatment
  - Post-surgical[33,36]

Screening imaging for Moyamoya Disease [34,35]

- Screening not indicated for Moyamoya Syndrome
  - See **Background and Supporting Information**
- Screening for Moyamoya Disease is indicated for:

  - First degree relatives (biological parent, full sibling, or biological child) of individuals with Moyamoya Disease when requested by, or any provider in consultation with a neurologist, geneticist or neurosurgeon
- Below are indicated for screening evaluation of Moyamoya Disease:
  - MRA Head (CPT® 70544, CPT® 70545, **OR** CPT® 70546) **OR** Transcranial Doppler (TCD) Ultrasound (CPT® 93886 or CPT® 93888)
  - If MRA is contraindicated or not readily available, then CTA Head (CPT® 70496) is supported

CT Perfusion (CPT ® 0042T)

- Is supported if requested by a neurologist, neurosurgeon or any provider coordinating care with a neurologist or neurosurgeon.[36]

MRI Perfusion

- MRI Perfusion may be obtained with MRI Brain (CPT® 70551 OR CPT® 70552 OR CPT® 70553)
  - No additional CPT® codes are necessary or appropriate to perform MRI perfusion.[33]

Background and Supporting Information

Moyamoya disease (MMD) is a rare cerebrovascular disease characterized by progressive spontaneous bilateral occlusion of the intracranial internal carotid arteries (ICA) and their major branches (middle cerebral artery, MCA, and anterior cerebral artery, ACA) with compensatory capillary collaterals as an expression of pathologically increased angiogenic activity resembling a "puff of smoke" (Japanese: Moyamoya) on cerebral angiography.[41] Moyamoya Disease is most prevalent individuals with East Asian ancestry. Up to 15% of individuals with Moyamoya Disease may have a family history of Moyamoya Disease. [34,35]

Moyamoya Disease is distinguished from Moyamoya Syndrome (MMS). MMD is a primary disease process. MMS is a secondary process that occurs in response to another underlying pathological process that causes stenosis of intracranial blood vessels.[40] There are two peaks of incidence with different clinical presentations, at around 10 years and 30-40 years. The peak appears to occur later in women than men. In children, ischemic symptoms, especially transient ischemic attacks, are predominant. Intellectual decline, seizures, and involuntary movements are also more common in this age group. In contrast, adult patients present with intracranial hemorrhage more often than pediatric patients.[35]

Sickle Cell Disease (HD-21.6)

HD.ST.0021.6.C
v2.0.2024

- MRI Brain without contrast (CPT® 70551) **OR** MRI Brain without and with contrast (CPT® 70553) indications:
  - Screening to detect silent cerebral infarcts
  - New symptoms or cognitive impairment occurs or a change in academic performance
  - Prior to any change in therapy[42, 44, 45, 46,52]
- MRA Head (CPT® 70544, CPT® 70545 OR CPT® 70546) **OR** CTA Head (CPT® 70496) indications:
  - Any new, indeterminate or equivocal findings on MRI Brain
  - Prior to any change in therapy[42, 44, 45, 46,52]

Background and Supporting Information

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

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

- TCD for children aged 17 years old may be appropriate on a case-by-case basis.
- See also **Stroke/TIA (HD-21.1)** in the Head Imaging Guidelines.
- After 17 years old, for individuals with a history of abnormal TCDs, TCDs may be repeated every 3 months.[47]
- TCD is not indicated for individuals with other phenotypes (Hgb SC, Hgb Sβ$^+$).[8]

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Multisystemic Smooth Muscle Syndrome (MSMS)/Smooth Muscle Dysfunction Syndrome (SMDS)/ACTA2 Mutations[49] (HD-21.7)

HD.ST.0021.7.C
v2.0.2024

Indications	Supported Imaging
Initial evaluation for confirmed ACTA2 mutation	• MRI Brain without and with contrast (CPT® 70553) with OR without MRI perfusion **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **AND/OR** • MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549)
Repeat imaging if requested by neurologist and/or neurosurgeon and/or geneticist and/or provider coordinating care with a neurologist and/or neurosurgeon and/or geneticist	• MRI Brain without and with contrast (CPT® 70553) with OR without MRI perfusion **AND/OR** • MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) **AND/OR** • MRA Neck (CPT® 70547, CPT® 70548, or CPT® 70549)

- MRI Perfusion may be obtained with MRI Brain (CPT® 70551 OR CPT® 70552 OR CPT® 70553)
  - No additional CPT® codes are necessary or appropriate to perform MRI perfusion. [51]
- Because radiation is a known risk factor for development of moyamoya, MRI/MRA Head is recommended instead of Computed Tomography (CT)/CTA.[49]
  - See **Background and Supporting Information**

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

- Conventional catheter angiogram 3D rendering (CPT® 76377) should be reserved for patients with focal neurologic symptoms or evidence on MRA or transcranial Doppler (TCD) of critical or progressive narrowing of the cerebral arteries.[49]
    - See **Screening for Suspected Peripheral Artery Disease/Aneurysmal Disease (PVD-2)**

Background and Supporting Information

Smooth muscle dysfunction syndrome (SMDS)/Multisystemic Smooth Muscle Syndrome (MSMS) presents with a recognizable pattern of complications, including congenital mydriasis, patent ductus arteriosus (PDA), pulmonary arterial hypertension, aortic and other arterial aneurysms, moyamoya-like cerebrovascular disease, intestinal hypoperistalsis and malrotation, and hypotonic bladder.[49]

SMDS/MSMS is caused by heterozygous mutations of the ACTA2 altering arginine 179, most commonly p.Arg179His. With a single exception, all cases are due to de novo mutations.[49]

References (HD-21)

v2.0.2024

1. Expert Panel on Neurologic Imaging:, Salmela MB, Mortazavi S, et al. ACR Appropriateness Criteria® Cerebrovascular Disease. J Am Coll Radiol. 2017;14(5S):S34-S61. doi:10.1016/j.jacr.2017.01.051
2. Kovacs MJ. Letter by Kovacs Regarding Article, "Diagnosis and Management of Cerebral Venous Thrombosis: A Statement for Healthcare Professionals From the American Heart Association/American Stroke Association." Stroke. 2011;42(7). doi:10.1161/strokeaha.111.619437
3. Stam J. Thrombosis of the Cerebral Veins and Sinuses. New England Journal of Medicine. 2005;352(17):1791-1798. doi:10.1056/nejmra042354
4. Schievink WI. Spontaneous dissection of the carotid and vertebral arteries. The New England Journal of Medicine. 2001;344(12):898-906. doi:10.1056/NEJM200103223441206
5. Arnold M, Bousser M-G. Carotid and vertebral artery dissection. Practical Neurology. 2005;5(2):100-109. doi:10.1111/j.1474-7766.2005.00292.x
6. Powers WJ, Rabinstein AA, Ackerson T, et al. Guidelines for the Early Management of Patients With Acute Ischemic Stroke: 2019 Update to the 2018 Guidelines for the Early Management of Acute Ischemic Stroke: A Guideline for Healthcare Professionals From the American Heart Association/American Stroke Association. Stroke. 2019;50(12). doi:10.1161/str.0000000000000211
7. Burton TM, Bushnell CD. Reversible Cerebral Vasoconstriction Syndrome. Stroke. 2019;50(8):2253-2258. doi:10.1161/strokeaha.119.024416
8. Debaun MR, Jordan LC, King AA, et al. American Society of Hematology 2020 guidelines for sickle cell disease: prevention, diagnosis, and treatment of cerebrovascular disease in children and adults. Blood Advances. 2020;4(8):1554-1588. doi:10.1182/bloodadvances.2019001142
9. Osgood M, Budman E, Carandang R, Goddeau JRP, Henninger N. Prevalence of Pelvic Vein Pathology in Patients with Cryptogenic Stroke and Patent Foramen Ovale Undergoing MRV Pelvis. Cerebrovascular Diseases. 2015;39(3-4):216-223. doi:10.1159/000376613
10. Messé SR, Gronseth GS, Kent DM, et al. Practice advisory update summary: Patent foramen ovale and secondary stroke prevention. Neurology. 2020;94(20):876-885. doi:10.1212/wnl.0000000000009443
11. Demeestere J, Wouters A, Christensen S, Lemmens R, Lansberg MG. Review of Perfusion Imaging in Acute Ischemic Stroke. Stroke. 2020;51(3):1017-1024. doi:10.1161/strokeaha.119.028337
12. Belani P, Schefflein J, Kihira S, et al. COVID-19 Is an Independent Risk Factor for Acute Ischemic Stroke. American Journal of Neuroradiology. 2020. doi:10.3174/ajnr.a6650
13. Merkler AE, Parikh NS, Mir S, et al. Risk of Ischemic Stroke in Patients With Coronavirus Disease 2019 (COVID-19) vs Patients With Influenza. JAMA Neurology. 2020. doi:10.1001/jamaneurol.2020.2730
14. Guzik A, Bushnell C. Stroke Epidemiology and Risk Factor Management. CONTINUUM: Lifelong Learning in Neurology. 2017;23(1):15-39. doi:10.1212/con.0000000000000416
15. Tsivgoulis G, Alexandrov AV. Ultrasound in Neurology. CONTINUUM: Lifelong Learning in Neurology. 2016;22(5):1655-1677. doi:10.1212/con.0000000000000374
16. Meschia JF, Bushnell C, Boden-Albala B, et al. Guidelines for the Primary Prevention of Stroke. Stroke. 2014;45(12):3754-3832. doi:10.1161/str.0000000000000046
17. Lawton MT and Vates GE. Subarachnoid Hemorrhage. N Engl J Med 2017;377:257-66. doi: 10.1056/NEJMcp1605827
18. ACR AIUM SPR SRU Practice Parameter for the Performance of an Ultrasound Examination of the Extracranial Cerebrovascular System. 2016
19. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck
20. Kamel, H, et al. Tailoring the Approach to Embolic Stroke of Undetermined Source. A Review. JAMA Neurol;76(7):855-861. doi:10.1001/jamaneurol.2019.0591
21. Navi BB and and Iadecola C. Ischemic Stroke in Cancer Patients: A Review of an Underappreciated Pathology. Ann Neurol. 2018 May ; 83(5): 873–883. doi:10.1002/ana.25227
22. Saver, JL. Cryptogenic Stroke. NEngl J Med 2016;374:2065-74. doi:10.1056/NEJMcp1503946
23. Scwarzbach CJ, et al. Stroke and Cancer. The Importance of Cancer-Associated Hypercoagulation as a Possible Stroke Etiology Stroke. 2012;43:3029-3034. doi: 10.1161/STROKEAHA.112.658625
24. Mangla A, Navi BB, Layton K, Kamel H. Transient global amnesia and the risk of ischemic stroke. Stroke. 2014;45(2):389-393. doi:10.1161/STROKEAHA.113.003916
25. Spiegel DR, Smith J, Wade RR, et al. Transient global amnesia: current perspectives. Neuropsychiatric Disease and Treatment. 2017;Volume 13:2691-2703. doi:10.2147/ndt.s130710
26. Chandra A, Stone CR, Du X, Li WA, Huber M, Bremer R, Geng X, Ding Y. The cerebral circulation and cerebrovascular disease III: Stroke. Brain circulation. 2017 Apr;3(2):66

27. Hakimi R, Sivakumar S. Imaging of Carotid Dissection. Current Pain and Headache Reports. 2019;23(1). doi:10.1007/s11916-019-0741-9
28. Ghoneim A, Straiton J, Pollard C, Macdonald K, Jampana R. Imaging of cerebral venous thrombosis. Clinical Radiology. 2020;75(4):254-264. doi:10.1016/j.crad.2019.12.009
29. Dmytriw AA, Song JSA, Yu E, Poon CS. Cerebral venous thrombosis: state of the art diagnosis and management. Neuroradiology. 2018;60(7):669-685. doi:10.1007/s00234-018-2032-2
30. ACR-ASNR-SIR-SNIS Practice Prameter for the Performan of Diagnostic Cervicocerebral Catheter Angiography in Adults. Revised 2016. (Resolution 13) https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CervicoCerebralCathAngio.pdf
31. McCarter SJ, Lesnick TG Lowe V et al. Cerebral Amyloid Angiopathy Pathology and Its Association With Amyloid-β PET Signal Neurology 2021;97 (18) :e1799-e1808. doi:10.1212/WNL.0000000000012770
32. Baron JC, Farid K, Dolan E, et al. Diagnostic utility of amyloid PET in cerebral amyloid angiopathy-related symptomatic intracerebral hemorrhage. J Cereb Blood Flow Metab. 2014;34(5):753-758. doi:10.1038/jcbfm.2014.43
33. ACR-ASNR- SPR Practice Parameters for the performance of Computed Tomography (CT) perfusion in neuroradiologic imaging. Revised 2017. Resolution 18. https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Perfusion.pdf
34. Han C, Feng H, Han YQ, et al. Prospective screening of family members with moyamoya disease patients.*PLoS One*. 2014;9(2):e88765. Published 2014 Feb 19. doi:10.1371/journal.pone.0088765
35. Kim JS. Moyamoya Disease: Epidemiology, Clinical Features, and Diagnosis.*J Stroke*. 2016;18(1):2-11. doi:10.5853/jos.2015.01627
36. Choudri A, Zaza A, Auschwitz T, Mossa-Basha M. Noninvasive vascular imaging of moyamoya: Diagnosis, followup, and surgical planning. Journal of Pediatric Neuroradiology 3 (2014) 13–20. doi:10.3233/PNR-14082
37. Expert Panel on Neurological Imaging, Ledbetter LN, Burns J, et al. ACR Appropriateness Criteria® Cerebrovascular Diseases-Aneurysm, Vascular Malformation, and Subarachnoid Hemorrhage. J Am Coll Radiol. 2021;18(11S):S283-S304. doi:10.1016/j.jacr.2021.08.012
38. Szidonya L, Nickerson JP. Cerebral Amyloid Angiopathy.*Radiol Clin North Am*. 2023;61(3):551-562. doi:10.1016/j.rcl.2023.01.009
39. Ropper AH. Transient Global Amnesia.*N Engl J Med*. 2023;388(7):635-640. doi:10.1056/NEJMra2213867
40. Berry JA, Cortez V, Toor H, Saini H, Siddiqi J. Moyamoya: An Update and Review.*Cureus*. 2020;12(10):e10994. Published 2020 Oct 16. doi:10.7759/cureus.10994
41. Mertens R, Graupera M, Gerhardt H, et al. The Genetic Basis of Moyamoya Disease.*Transl Stroke Res*. 2022;13(1):25-45. doi:10.1007/s12975-021-00940-2
42. Krishnamurti L. Hematopoietic Cell Transplantation for Sickle Cell Disease. Front Pediatr. 2021;8:551170. Published 2021 Jan 5. doi:10.3389/fped.2020.551170
43. Roeder HJ, Lopez JR, Miller EC. Ischemic stroke and cerebral venous sinus thrombosis in pregnancy.*Handb Clin Neurol*. 2020;172:3-31. doi:10.1016/B978-0-444-64240-0.00001-5
44. Kanter J, Liem RI, Bernaudin F, et al. American Society of Hematology 2021 guidelines for sickle cell disease: stem cell transplantation. Blood Adv. 2021;5(18):3668-3689. doi:10.1182/bloodadvances.2021004394C
45. Jordan LC, Kassim AA, Wilkerson KL, Lee CA, Waddle SL, Donahue MJ. Using novel magnetic resonance imaging methods to predict stroke risk in individuals with sickle cell anemia. Hematol Oncol Stem Cell Ther. 2020;13(2):76-84. doi:10.1016/j.hemonc.2019.12.009
46. Hirtz D, Kirkham FJ. Sickle Cell Disease and Stroke. Pediatr Neurol. 2019;95:34-41. doi:10.1016/j.pediatrneurol.2019.02.018
47. Bernaudin F, Verlhac S, Arnaud C, et al. Long-term treatment follow-up of children with sickle cell disease monitored with abnormal transcranial Doppler velocities. Blood. 2016;127(14):1814-1822. doi:10.1182/blood-2015-10-675231.
48. Kirkham FJ, Lagunju IA. Epidemiology of Stroke in Sickle Cell Disease.*J Clin Med*. 2021;10(18):4232. Published 2021 Sep 18. doi:10.3390/jcm10184232
49. Regalado ES, Mellor-Crummey L, De Backer J, et al. Clinical history and management recommendations of the smooth muscle dysfunction syndrome due to ACTA2 arginine 179 alterations. *Genet Med.* 2018;20(10):1206-1215. doi:10.1038/gim.2017.245
50. Cuoco JA, Busch CM, Klein BJ, et al. ACTA2 Cerebral Arteriopathy: Not Just a Puff of Smoke. *Cerebrovasc Dis.* 2018;46(3-4):161-171. doi:10.1159/000493863.
51. ACR–ASNR–SPR PRACTICE PARAMETER FOR THE PERFORMANCE OF INTRACRANIAL MAGNETIC RESONANCE PERFUSION IMAGING. Revised 2022 (Resolution 24). *PRACTICE PARAMETER MR_Perfusion*.https://www.acr.org/-/media/ACR/Files/Practice-Parameters/mr-perfusion.pdf? la=en.
52. Nickel RS, Kamani NR. Ethical Challenges in Hematopoietic Cell Transplantation for Sickle Cell Disease. Biol Blood Marrow Transplant. 2018;24(2):219-227. doi:10.1016/j.bbmt.2017.08.034

Cerebral Vasculitis (HD-22)

Cerebral Vasculitis (HD-22.1)

HD.CV.0022.1.C
v2.0.2024

- When CNS vasculitis is suspected MRI Brain without and with contrast (CPT® 70553) is supported
  - 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) are supported concurrently with brain imaging
- Primary CNS vasculitis includes Giant Cell Arteritis also known as Temporal Arteritis (see **New Headache Onset Older than Age 50 (HD-11.7)**)
- 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.
- For extra-cranial giant cell arteritis evaluation (see **Giant Cell Arteritis (PVD-6.9.2))**

Background and Supporting Information

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.[6]

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	• Giant cell arteritis • Takayasu's arteritis	Aortitis with rheumatoid disease; Infection (e.g. syphilis)

Dominant Vessel Involved	Primary	Secondary
Medium arteries	• Classical polyarteritis nodosa • Kawasaki disease	Infection (e.g. hepatitis B)
Small vessels and medium arteries	• Wegener's granulomatosis • Churg–Strauss syndrome • Microscopic polyangiitis	Vasculitis with rheumatoid disease, systemic lupus erythematosus (lupus cerebritis), Sjögren's syndrome, drugs, infection (e.g. HIV)
Small vessels	• Henoch-Schönlein purpura • Essential cryoglobulinemia • Cutaneous leukocytoclastic vasculitis	Drugs (e.g. sulphonamides, etc.) Infection (e.g. hepatitis C)

References (HD-22)

HD.CV.0022.2.A
v2.0.2024

1. Younger DS. Epidemiology of Neurovasculitis. *Neurologic Clinics*. 2016;34(4):887-917. doi:10.1016/j.ncl.2016.06.006
2. Soun JE, Song JW, Romero JM, Schaefer PW. Central Nervous System Vasculopathies. *Radiologic Clinics of North America*. 2019;57(6):1117-1131. doi:10.1016/j.rcl.2019.07.005
3. Salmela MB, Mortazavi S, Jagadeesan BD, et al. ACR Appropriateness Criteria ® Cerebrovascular Disease. *Journal of the American College of Radiology*. 2017;14(5). doi:10.1016/j.jacr.2017.01.051
4. Okazaki T, Shinagawa S, Mikage H. Vasculitis syndrome-diagnosis and therapy. *Journal of General and Family Medicine*. 2017;18(2):72-78. doi:10.1002/jgf2.4
5. Ikeda T, Furukawa F, Kawakami T, et al. Outline of guidelines for the management of vasculitis and vascular disorders in Japan, 2016 revised edition. *The Journal of Dermatology*. 2017;45(2):122-127. doi:10.1111/1346-8138.14086
6. Expert Panel on Neurological Imaging, Ledbetter LN, Burns J, et al. ACR Appropriateness Criteria® Cerebrovascular Diseases-Aneurysm, Vascular Malformation, and Subarachnoid Hemorrhage. J Am Coll Radiol. 2021;18(11S):S283-S304. doi:10.1016/j.jacr.2021.08.012

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Dizziness, Vertigo and Syncope (HD-23)

Dizziness/Vertigo (HD-23.1)

HD.DZ.0023.1.A
v2.0.2024

Indications	Supported Imaging

Red Flags:
• History of malignancy
• Associated symptoms:
  • Headache
  • Hearing loss
  • Unilateral tinnitus
  • Visual disturbances
  • Drop attacks
  • Vestibular migraine
  • Weakness
• Duration of episode:
  • Episodes lasting hour(s) or
  • Continuous
• Exam findings:
  • Inconclusive positional testing or equivocal or unusual nystagmus findings (Negative Dix-Hallpike)
  • Visual disturbances including loss and diplopia
  • Hearing loss
  • Abnormal cranial nerve findings
  • Ataxia
  • Positive Romberg sign
  • Absent head thrust sign
  • Focal neurologic deficits
  • Dysarthria
  • Weakness, including unilateral or hemibody weakness
• Failed treatment:
  • Failure to respond to vestibular therapy or unable to participate due to clinical condition
• Abnormal test results:
  • ENG/VNG results support central cause
	• MRI Brain without contrast (CPT 70551 **OR**
	• MRI Brain without and with contrast (CPT 70553) **OR**
	• CT head without contrast (CPT 70450)
	
	If MRI contraindicated:
	• CT head without contrast (CPT 70450) **OR**
	• CT head without and with contrast (CPT 70470)
	
	See also:
	• **Headaches with Red Flags (HD-11.2)**
	• **Multiple Sclerosis and Related Conditions (HD-16)**
	• **Brain Metastases (ONC-31.3)**

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Indications	Supported Imaging
Stroke/TIA	See **Stroke/TIA (HD-21.1)**
Acoustic Neuroma/Vestibular Schwannoma	• MRI Brain without and with contrast (with IAC views) (CPT® 70553) **OR** without contrast (CPT® 70551) • 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 See also • **Acoustic Neuroma (HD-33.1)** • **Peripheral Nerve Sheath Tumors (PN-9.1)**
Head trauma / Temporal Bone Fracture / Post-traumatic vertigo	• CT Head without contrast (CPT® 70450) • See **Head Trauma (HD-13.1)** **AND/OR** • CT Orbit/Temporal bone without contrast (CPT® 70480)

Indications	Supported Imaging
Vertebrobasilar disease/ Vertebrobasilar Insufficiency/ Dissection	• CTA Head (CPT® 70496 **AND/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) See also: • **General Guidelines - CT and MR Angiography (CTA and MRA) (HD-1.5)** • **Headache and Suspected Vascular Dissection (HD-11.1)** • **Intracranial Aneurysms (HD-12.1)**
Semicircular canal dehiscence	• CT Orbit/Temporal bone without contrast (CPT® 70480)
Meniere's Disease	• MRI Brain without and with contrast (with IAC views) (CPT® 70553) **OR** without contrast (CPT® 70551) • 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

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) is suspected.
- 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), upbeating (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 circulationUnable 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/dissectio
- 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 ≥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 playpnea (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 boney 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

v2.0.2024

Indications	Supported Imaging
Syncope with focal signs of a neurologic deficit **OR** Syncope without focal signs of a neurological deficit AND negative or inconclusive Electrocardiogram (EKG)	• MRI Brain without contrast (CPT® 70551) **OR** • MRI Brain without and with contrast (CPT® 70553) **OR** • CT head without contrast (CPT® 70450) **AND/OR** • CTA head (CPT® 70496) **OR** • MRA head (CPT® 70544, CPT® 70545, or CPT® 70546) **AND/OR** • CTA neck (CPT® 70498) **OR** • MRA neck (CPT® 70547, CPT® 70548, or CPT® 70549)
Recurrent syncope with risk of head injury or head trauma related to syncope[6,15]	See **Head Trauma (HD-13.1)**
Situational syncope, including precipitating factors to syncope such as coughing, defecation, eating, laughing, or urination Myoclonic jerks without symptoms or signs associated with seizure, including but not limited to prolonged amnesia/confusion, tongue biting.	Advanced imaging is not indicated
Loss of consciousness with other symptoms or signs of seizure, including but not limited to, prolonged amnesia/confusion, tongue biting, and/or urinary incontinence.	See **Epilepsy/Seizure (HD-9.1)**

References (HD-23)

v2.0.2024

1. Runser LA, Gauer RL and Houser A. Syncope: Evaluation and Differential Diagnosis. *Am Fam Physician*. 2017;95(5):303-312
2. Expert Panel on Neurologic Imaging:, Sharma A, Kirsch CFE, et al. ACR Appropriateness Criteria® Hearing Loss and/or Vertigo. *J Am Coll Radiol*. 2018;15(11S):S321-S331. doi:10.1016/j.jacr.2018.09.020
3. Cheshire WP. Syncope. *CONTINUUM: Lifelong Learning in Neurology*. 2017;23(2):335-358. doi:10.1212/con.0000000000000444
4. Bhattacharyya N, Gubbels SP, Schwartz SR, et al. Clinical Practice Guideline: Benign Paroxysmal Positional Vertigo (Update). *Otolaryngology–Head and Neck Surgery*. 2017;156(3_suppl). doi:10.1177/0194599816689667
5. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck
6. Shen W-K, Sheldon RS, Benditt DG, et al. 2017 ACC/AHA/HRS Guideline for the Evaluation and Management of Patients With Syncope: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society. Circulation. 2017;136(5). doi:10.1161/cir.0000000000000499
7. Basura GJ, Adams ME, Monfared A, et al. Clinical Practice Guideline: Ménière's Disease. *Otolaryngology–Head and Neck Surgery*. 2020;162(2_suppl). doi:10.1177/0194599820909438
8. Gibbons CH, Schmidt P, Biaggioni I, et al. The recommendations of a consensus panel for the screening, diagnosis, and treatment of neurogenic orthostatic hypotension and associated supine hypertension. *Journal of Neurology*. 2017;264(8):1567-1582. doi:10.1007/s00415-016-8375-x
9. Choosing Wisely. An initiative of the ABIM Foundation. *American Academy of Neurology*. Released February 21, 2013; Last reviewed 2019
10. Choosing Wisely. An initiative of the ABIM Foundation. *American College of Emergency Physicians*. October 27, 2014
11. Scott JW, Schwartz AL, Gates JD, Gerhard-Herman M, Havens JM. Choosing Wisely for Syncope: Low-Value Carotid Ultrasound Use. *Journal of the American Heart Association*. 2014;3(4). doi:10.1161/jaha.114.001063
12. Dix-Hallpike maneuver performed for paitents with BPPV. www.aan.com. https://www.aan.com/policy-and-guidelines/quality/quality-measures2/quality-measures/other/dix-hallpike-maneuver-performed-for-paitents-with-BPPV
13. Baloh RW. Vestibular Migraine I: Mechanisms, Diagnosis, and Clinical Features. *Seminars in Neurology*. 2020;40(01):076-082. doi:10.1055/s-0039-3402735
14. Tehrani ASS, Kattah JC, Kerber KA, et al. Diagnosing Stroke in Acute Dizziness and Vertigo. *Stroke*. 2018;49(3):788-795. doi:10.1161/strokeaha.117.016979
15. Expert Panels on Cardiac Imaging and Neurological Imaging, Kligerman SJ, Bykowski J, et al. ACR Appropriateness Criteria® Syncope. *J Am Coll Radiol*. 2021;18(5S):S229-S238. doi:10.1016/j.jacr.2021.02.021
16. Shmuely S, et al. Differentiating Motor Phenomena in Tilt-Induced Syncope and Convulsive Seizures. *Neurology*. 2018;90:e1339-e1346. doi:10.1212/WNL.0000000000005301
17. Henderson FC, Austin C, Benzel E, et al. Neurological and spinal manifestations of the Ehlers-Danlos syndromes. American journal of medical genetics Part C, Seminars in medical genetics. 2017;175(1):195-211. doi:10.1002/ajmg.c.31549
18. Edlow JA, Gurley KL, Newman-Toker DE. A New Diagnostic Approach to the Adult Patient with Acute Dizziness. *The Journal of Emergency Medicine*. 2018;54(4):469-483. doi:10.1016/j.jemermed.2017.12.024
19. Edlow JA. The timing-and-triggers approach to the patient with acute dizziness. *Emerg Med Pract*. 2019 Dec;21(12):1-24. Epub 2019 Dec 1. PMID: 31765116
20. Krishnan K, Bassilious K, Eriksen E, et al. Posterior circulation stroke diagnosis using HINTS in patients presenting with acute vestibular syndrome: A systematic review. *European Stroke Journal*. Published online April 10, 2019:239698731984370. doi:10.1177/2396987319843701
21. Fife TD. Approach to the History and Evaluation of Vertigo and Dizziness. *CONTINUUM: Lifelong Learning in Neurology*. 2021;27(2):306-329. doi:10.1212/con.0000000000000938
22. Hain TC, Cherchi M. Vestibular Testing. *CONTINUUM: Lifelong Learning in Neurology*. 2021;27(2):330-347. doi:10.1212/con.0000000000000978
23. Steenerson KK. Acute Vestibular Syndrome. *CONTINUUM: Lifelong Learning in Neurology*. 2021;27(2):402-419. doi:10.1212/con.0000000000000958

Other Imaging Studies (HD-24)

Transcranial Magnetic Stimulation (TMS) (HD-24.1)

HD.OI.0024.1.C
v2.0.2024

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.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Functional MRI (fMRI) (HD-24.2)

HD.OI.0024.2.A
v2.0.2024

- 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 indicated 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
  - If MRA Head (CPT® 70544) is indicated but Functional MRI (CPT® 70554 or CPT® 70555) was erroneously ordered, then CPT® 70544 may be substituted when appropriate
- 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.A
v2.0.2024

- 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)

HD.OI.0024.4.A
v2.0.2024

- Pulse-gated MRI imaging or MRI CINE is generally performed as a part of a MRI Brain study. It is not coded separately for pre-operative 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.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

CT or MRI Perfusion (HD-24.5)

HD.OI.0024.5.A
v2.0.2024

- Performed as part of a CT Head or MRI Brain examination in the evaluation of individuals with very new strokes or brain tumors.
- 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.
  - CTA Head and/or Neck is indicated in conjunction with the CT Perfusion study (CPT® 0042T).
- CPT® 0042T - “cerebral perfusion analysis using CT”.
  - 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
  - Follow up fo acute cerebral ischemic 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 hypoperfusion 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
- MRI Perfusion may be obtained with MRI Brain (CPT® 70551 OR CPT® 70552 OR CPT® 70553).
  - No additional CPT® codes are necessary or appropriate to perform MRI perfusion.[9]
- Indications for perfusion magnetic resonance imaging (MRI) MRI Perfusion (CPT® 70551 OR CPT® 70552 OR CPT® 70553)[9] 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
    - Pseudoprogression 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
- Other indications are usually regarded as not medically necessary.

Magnetic Resonance Neurography (MRN) (HD-24.6)

HD.OI.0024.6.A
v2.0.2024

- See **Magnetic Resonance Neurography (MRN) (PN-7.1)** in the Peripheral Nerve Disorders (PND) Imaging Guidelines.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

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

HD.OI.0024.7.A
v2.0.2024

- 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)**

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

References (HD-24)

v2.0.2024

1. Tsivgoulis G, Alexandrov AV. Ultrasound in Neurology. *CONTINUUM: Lifelong Learning in Neurology*. 2016;22(5):1655-1677. doi:10.1212/con.0000000000000374
2. *PRACTICE PARAMETER CT_Perfusion*. https://www.acr.org/-/media/ACR/Files/Practice-Parameters/ct-perfusion.pdf
3. Connolly ES, Rabinstein AA, Carhuapoma JR, et al. Guidelines for the Management of Aneurysmal Subarachnoid Hemorrhage. *Stroke*. 2012;43(6):1711-1737. doi:10.1161/str.0b013e3182587839
4. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck
5. Demeestere J, Wouters A, Christensen S, Lemmens R, Lansberg MG. Review of Perfusion Imaging in Acute Ischemic Stroke. *Stroke*. 2020;51(3):1017-1024. doi:10.1161/strokeaha.119.028337
6. *PRACTICE PARAMETER Transcranial Doppler Ultrasound*. https://www.acr.org/-/media/ACR/Files/Practice-Parameters/us-transcranial.pdf?la=en
7. *PRACTICE PARAMETER FMRI*. https://www.acr.org/-/media/ACR/Files/Practice-Parameters/fmr-brain.pdf?la=en
8. *PRACTICE PARAMETER 1 MR Spectroscopy*. https://www.acr.org/-/media/ACR/Files/Practice-Parameters/mr-spectroscopy.pdf?la=en
9. ACR–ASNR–SPR PRACTICE PARAMETER FOR THE PERFORMANCE OF INTRACRANIAL MAGNETIC RESONANCE PERFUSION IMAGING. Revised 2022 (Resolution 24). *PRACTICE PARAMETER MR_Perfusion*.https://www.acr.org/-/media/ACR/Files/Practice-Parameters/mr-perfusion.pdf? la=en
10. Bradley WG. Magnetic Resonance Imaging of Normal Pressure Hydrocephalus. *Seminars in Ultrasound, CT and MRI*. 2016;37(2):120-128. doi:10.1053/j.sult.2016.01.005
11. Farb R, Rovira À. Chapter 2: Hydrocephalus and CSF Disorders. In: Hodler J, Kubik-Huch RA, von Schulthess GK. eds. *Hydrocephalus and CSF Disorders--Diseases of the Brain, Head and Neck, Spine 2020–2023: Diagnostic Imaging*. 2020 Feb 15
12. Antipova D, Eadie L, Macaden AS, Wilson P. Diagnostic value of transcranial ultrasonography for selecting subjects with large vessel occlusion: a systematic review. *The Ultrasound Journal*. 2019;11(1). doi:10.1186/s13089-019-0143-6
13. Batra A, Clark JR, LaHaye K, et al. Transcranial Doppler Ultrasound Evidence of Active Cerebral Embolization in COVID-19. *Journal of Stroke and Cerebrovascular Diseases*. 2021;30(3):105542. doi:10.1016/j.jstrokecerebrovasdis.2020.105542
14. Purkayastha S, Sorond F. Transcranial Doppler Ultrasound: Technique and Application. *Seminars in neurology*. 2012;32(4):411-420. doi:10.1055/s-0032-1331812
15. Feng Y, Su X, Zheng C, Lu Z. The Noninvasive Diagnostic Value of MRN for CIDP: A Research from Qualitative to Quantitative. *Spine*. 2020;45(21):1506-1512. doi:10.1097/brs.0000000000003599
16. AIUM Practice Guideline for the Performance of a Transcranial Doppler Ultrasound Examination for Adults and Children. *Journal of Ultrasound in Medicine*. 2012;31(9):1489-1500. doi:10.7863/jum.2012.31.9.1489
17. Expert Panel on Pediatric Imaging, Robertson RL, Palasis S, et al. ACR Appropriateness Criteria® Cerebrovascular Disease-Child. J Am Coll Radiol. 2020;17(5S):S36-S54. doi:10.1016/j.jacr.2020.01.036
18. McGirr A, Vila-Rodriguez F, Cole J, et al. Efficacy of Active vs Sham Intermittent Theta Burst Transcranial Magnetic Stimulation for Patients With Bipolar Depression. *JAMA Network Open*. 2021;4(3):e210963. doi:10.1001/jamanetworkopen.2021.0963
19. Lacomis D, Gooch C. Upper motor neuron assessment and early diagnosis in ALS. *Neurology*. 2019;92(6):255-256. doi:10.1212/wnl.0000000000006867
20. Rosenfeld RM, Piccirillo JF, Chandrasekhar SS, et al. Clinical Practice Guideline (Update): Adult Sinusitis. *Otolaryngology–Head and Neck Surgery*. 2015;152(2_suppl):S1-S39. doi:10.1177/0194599815572097

Epistaxis (HD-25)

Epistaxis (HD-25.1)

HD.EX.0025.1.A
v2.0.2024

- After initial nasal endoscopy by ENT, if there are findings suspicious for a mass lesion:
  - 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)
- Patients who have failed initial management with cauterization and packing and have persistent or recurrent epistaxis despite primary interventions, should be referred to a clinical who can evaluate for candidacy for surgical ligation or endovascular embolization.[3]
- Prior to embolization with surgical or endovascular technique, CT Maxillofacial (without contrast CPT® 70486 or without contrast CPT® 70487), is supported when requested by the clinical performing embolization or referring for embolization. If endovascular embolization is planned, CTA Head (CPT® 70496) **AND/OR** CTA Neck (CPT® 70498) may be requested ahead of the interventional radiologic procedure.[5]

Background and Supporting Information

The American Academy of Otolaryngology Head and Neck Surgery recommend, in its most recent 2020 Clinical Practice Guidelines on Epistaxis, that the clinician should perform, or should refer to a clinician who can perform, nasal endoscopy to identify the site of bleeding and guide further management in patients with recurrent nasal bleeding, despite prior treatment with packing or cautery, or with recurrent unilateral nasal bleeding. No recommendations for advanced imaging are outlined in this Guideline without the exam findings (anterior rhinoscopy and/or nasal endoscopy) or the procedural needs of the patient indicating the need for such studies. If anterior rhinoscopy does not reveal the source of bleeding, it is recommended that the clinician perform nasal endoscopy, or refer to a clinician who can perform nasal endoscopy, first.[3]

Embolization procedures have shown an average nosebleed control rate of 87%, with minor transient complications in 20% (transient nasal ischemia, temporal-facial pain or numbness, headache, swelling, jaw claudication, trismus, and access site complications not requiring additional therapy) and major complications in up to 2.1% to 3.8% (skin/nasal necrosis, permanent facial nerve paralysis, monocular blindness, and stroke).

Detailed angiography, including internal and external carotid angiography, and precise embolization techniques are required. Despite use of meticulous techniques and knowledge of external carotid-internal carotid anastomoses, blindness and stroke are the most feared complications of endovascular embolization. These complications are rare but are more frequent than in patients undergoing surgical arterial ligation. In one

study, similar transient ischemic attacks are demonstrated across all groups but there is increased risk of stroke in the groups who underwent endovascular embolization alone (0.9%) or combined with surgical ligation (1.6%) as compared with surgical ligation alone (0.1%).[3,4,5]

References (HD-25)

v2.0.2024

1. Expert Panel on Neurologic Imaging:, Kirsch CFE, Bykowski J, et al. ACR Appropriateness Criteria® Sinonasal Disease. *J Am Coll Radiol*. 2017;14(11S):S550-S559. doi:10.1016/j.jacr.2017.08.041
2. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck
3. Tunkel DE, Anne S, Payne SC, et al. Clinical Practice Guideline: Nosebleed (Epistaxis). *Otolaryngology–Head and Neck Surgery*. 2020;162(1_suppl). doi:10.1177/0194599819890327
4. Strach K, Schröck A, Wilhelm K, et al. Endovascular treatment of epistaxis: indications, management, and outcome.*Cardiovasc Intervent Radiol*. 2011;34(6):1190-1198. doi:10.1007/s00270-011-0155-5
5. Brinjikji W, Kallmes DF, Cloft HJ. Trends in epistaxis embolization in the United States: a study of the Nationwide Inpatient Sample 2003-2010.*J Vasc Interv Radiol*. 2013;24(7):969-973. doi:10.1016/j.jvir.2013.02.035

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Mastoid Disease or Ear Pain (HD-26)

Mastoid Disease or Ear Pain (HD-26.1)

HD.MA.026.1.A
v2.0.2024

A pertinent clinical evaluation including a detailed history, physical examination (including otoscopic examination), must 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.

Indications (Any one of the following)	Supported Imaging
• Persistent ear pain without obvious cause • Clinical suspicion for complicated or invasive infection such as mastoiditis • Clinical suspicion for complications from otitis media • Clinical suspicion of mass lesion causing ear pain • Significant trauma with concern for hematoma formation • Pre-operative planning	• CT Orbits/Temporal Bone without contrast (CPT® 70480) **OR** • CT Orbits/Temporal Bone without and with contrast (CPT® 70482) **OR** • MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553) **OR** • MRI Orbits/Face/Neck without and with contrast (CPT® 70543)

- Advanced imaging is not indicated in the overwhelming majority of individuals with ear pain.
- Advanced imaging for the diagnosis and management of suspected cholesteatoma, in particular, should be reserved for the otolaryngologist or in consultation with the otolaryngologist
- Imaging indicated for pre-operative evaluation for cholesteatoma surgery:
  - CT Orbits/Temporal Bone without contrast (CPT® 70480) **OR**
  - CT Orbits/Temporal Bone without and with contrast (CPT® 70482) **AND/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)
- Indicated one time post-operatively to exclude residual or regrown cholesteatoma to avoid the need for a second-look surgery:
  - CT Orbits/Temporal Bone without contrast (CPT® 70480) **OR**
  - CT Orbits/Temporal Bone without and with contrast (CPT® 70482) **AND/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)

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

- Eustachian Tube Dilation:(endoscopic balloon dilatation of the Eustachian Tube, to treat persistent Eustachian tube dysfunction)[3, 4]
  - CT Orbit/Temporal Bone without contrast (CPT® 70480) can be approved for pre-operative evaluation of possible aberrant carotid
- Concern for Petrous Apex Lesions when requested by the Otolaryngologist or in consultation with the Otolaryngologist, the following are supported[6]:
  - CT Orbit/Temporal bone without contrast (CPT® 70480) **OR**
  - CT Orbit/Temporal bone without and with contrast (CPT® 70482) **AND/OR**
  - MRI Brain without and with contrast (CPT® 70553) **OR**
  - MRI Orbits/Face/Neck without or with contrast (CPT® 70543)
- For concern related to non-resolving otalgia with chronic otorrhea:
  - See **Skull Base Osteomyelitis (SBO) (HD-20.2)**

Background and Supporting Information

- Common causes of ear pain include external and middle ear infections, dental problems, sinus infection, neck problems, and referred pain from the oral pharynx.
- Clinical suspicion for complications from otitis media such as coalescent mastoiditis, resulting in: subperiosteal abscess formation/Bezold's abscess, acute facial nerve paralysis, and intracranial abscess formation.
- Cholesteatomas are expansive cysts of the middle ear filled with cellular debris. They can be congenital or arise from recurrent middle ear infections or trauma to the tympanic membrane. Hearing loss is usually conductive, although if the lesion is large enough combined conductive and sensorineural hearing loss may be present. Otoscopic exam findings and symptoms may include a white mass in the middle ear cleft, painless drainage from the ear or chronic/recurrent ear infections.
- Petrous apex lesions/infections may include: cholesteatoma, cephelocele, mucocele, and cholesterol granuloma and can present with symptoms of pain, hearing loss, headache, vertigo, and Cranial nerve insults(including CN V VI, VII, IX, X, XI).

References (HD-26)

v2.0.2024

1. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck
2. Rosenfeld RM, Shin JJ, Schwartz SR, et al. Clinical Practice Guideline: Otitis Media with Effusion (Update). Otolaryngology–Head and Neck Surgery. 2016;154(1_suppl):S1-S41. doi:10.1177/0194599815623467
3. Micucci S, Keschner DB, Liang J. Eustachian Tube Balloon Dilation: Emerging Practice Patterns for a Novel Procedure. Ann Otol Rhinol Laryngol. 2018 Nov;127(11):848-855. doi: 10.1177/0003489418798858.
4. Tucci DL, McCoul ED, Rosenfeld RM, Tunkel DE, Batra PS, Chandrasekhar SS, Cordes SR, Eshraghi AA, Kaylie D, Lal D, Lee J, Setzen M, Sindwani R, Syms CA 3rd, Bishop C, Poe DS, Corrigan M, Lambie E. Clinical Consensus Statement: Balloon Dilation of the Eustachian Tube. Otolaryngol Head Neck Surg. 2019 Jul;161(1):6-17. doi: 10.1177/0194599819848423.
5. Treviño González JL, Reyes Suárez LL, Hernández de León JE. Malignant otitis externa: An updated review. Am J Otolaryngol. 2021 Mar-Apr;42(2):102894. doi: 10.1016/j.amjoto.2020.102894. Epub 2021 Jan 5. PMID: 33429178
6. 6. Potter GM, Siripurapu R. Imaging of Petrous Apex Lesions. Neuroimaging Clin N Am. 2021;31(4):523-540. doi:10.1016/j.nic.2021.06.005

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

Hearing Loss and Tinnitus (HD-27)

Hearing Loss (HD-27.1)

HD.HL.0027.1.A
v2.0.2024

- 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 Orbits/Temporal Bone without (CPT® 70480) **OR** MRI Brain without and with contrast (with IAC views) (CPT® 70553) **OR** MRI Brain without contrast (CPT® 70551):
  - 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 Orbits/Temporal Bone without contrast (CPT® 70480):
  - Conductive hearing loss should have a CT Temporal Bone initially in the absence of an evident mass in the middle ear
- CT Orbits/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 indicated 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
v2.0.2024

- 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)**)

Indications (Any one of the following)[1,5,6]	Supported Imaging
• Clinical suspicion of mass lesion causing 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 • Persistent tinnitus after recent significant trauma. • Pulsatile tinnitus with or without concern for vascular lesion	• CT Orbits/Temporal Bone without contrast (CPT® 70480) **OR** • CT Orbits/Temporal Bone without and with contrast (CPT® 70482) **OR** • MRI Brain without and with contrast with attention to internal auditory canals (CPT® 70553) **OR** • MRI Brain without contrast with attention to internal auditory canals (CPT® 70551) **OR** • MRI Orbits/Face/Neck without contrast (CPT® 70540), with contrast CPT® 70542, or without and with contrast (CPT® 70543)
• Pulsatile tinnitus • Suspicion for vascular lesions	• MRA Head (CPT® 70544, CPT® 70545 OR CPT® 70546) **OR** • CTA Head (CPT® 70496) **AND/OR** • MRA Neck (CPT® 70547, CPT® 70548 or CPT® 70549) **OR** • CTA Neck (CPT® 70498)

- Imaging not supported for bilateral non-pulsatile tinnitus without other neurologic signs or symptoms[6]
- 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

- Non-pulsatile tinnitus may be described as ringing, buzzing, or clicking sensations which is constant and non-synchronous.
- Pulsatile tinnitus is a repetitive sound coinciding with the individual's heartbeat. The symptom may be subjective or objective.

References (HD-27)

HD.HL.0027.3.A

v2.0.2024

1. Expert Panel on Neurologic Imaging:, Sharma A, Kirsch CFE, et al. ACR Appropriateness Criteria® Hearing Loss and/or Vertigo. J Am Coll Radiol. 2018;15(11S):S321-S331. doi:10.1016/j.jacr.2018.09.020
2. Isaacson J, Vora NM. Differential diagnosis and treatment of hearing loss. *American Family Physician*. 2003 Sep 15;68(6):1125-32
3. Chandrasekhar SS, Do BST, Schwartz SR, et al. Clinical Practice Guideline: Sudden Hearing Loss (Update). *Otolaryngology–Head and Neck Surgery*. 2019;161(1_suppl). doi:10.1177/0194599819859885
4. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck
5. Expert Panel on Neurologic Imaging: Jain, V, Policeni B, et al. ACR Appropriateness Criteria® Tinnitus. 2023 https://acsearch.acr.org/docs/3094199/Narrative/
6. Tunkel DE, Bauer CA, Sun GH, Rosenfeld RM, Chandrasekhar SS, Cunningham ER Jr, Archer SM, Blakley BW, Carter JM, Granieri EC, Henry JA, Hollingsworth D, Khan FA, Mitchell S, Monfared A, Newman CW, Omole FS, Phillips CD, Robinson SK, Taw MB, Tyler RS, Waguespack R, Whamond EJ. Clinical practice guideline: tinnitus executive summary. Otolaryngol Head Neck Surg. 2014 Oct;151(4):533-41. doi: 10.1177/0194599814547475

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Neurosurgical Imaging (HD-28)

Neurosurgical Imaging (HD-28.1)

HD.NI.0028.1.A
v2.0.2024

- 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
v2.0.2024

- Neurosurgical navigation is “image-based” meaning that the necessary pre-operative CT and MRI images are used for navigation in the operating room (image acquisition). Accurate registration (a process to match the pre-operative images to the individual position) of pre-operative 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 pre-operative CT and MRI images does not require a radiologist interpretation.
  - Diagnostic imaging codes are not indicated 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 indicated:
    - 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 indicated 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
v2.0.2024

- 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)

v2.0.2024

1. Orringer DA, Golby A, Jolesz F. Neuronavigation in the surgical management of brain tumors: current and future trends. Expert Review of Medical Devices. 2012;9(5):491-500. doi:10.1586/erd.12.42
2. Rughani A, Schwalb JM, Sidiropoulos C, et al. Congress of Neurological Surgeons Systematic Review and Evidence-Based Guideline on Subthalamic Nucleus and Globus Pallidus Internus Deep Brain Stimulation for the Treatment of Patients With Parkinson's Disease: Executive Summary. Neurosurgery. 2018;82(6):753-756. doi:10.1093/neuros/nyy037
3. Kotecha R, Sahgal A, Rubens M, et al. Stereotactic radiosurgery for non-functioning pituitary adenomas: meta-analysis and International Stereotactic Radiosurgery Society practice opinion. Neuro-Oncology. 2019;22(3):318-332. doi:10.1093/neuonc/noz225
4. Xiao Y, Lau JC, Hemachandra D, Gilmore G, Khan AR, Peters TM. Image Guidance in Deep Brain Stimulation Surgery to Treat Parkinson's Disease: A Comprehensive Review. IEEE Transactions on Biomedical Engineering. 2021;68(3):1024-1033. doi:10.1109/tbme.2020.3006765
5. Delev D, Quesada CM, Grote A, et al. A multimodal concept for invasive diagnostics and surgery based on neuronavigated voxel-based morphometric MRI postprocessing data in previously nonlesional epilepsy. Journal of Neurosurgery. 2018;128(4):1178-1186. doi:10.3171/2016.12.jns161676
6. Yang I, Udawatta M, Prashant GN, et al. Stereotactic Radiosurgery for Neurosurgical Patients: A Historical Review and Current Perspectives. World Neurosurgery. 2019;122:522-531. doi:10.1016/j.wneu.2018.10.193
7. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck
8. Fitzpatrick JM. The role of registration in accurate surgical guidance. Proceedings of the Institution of Mechanical Engineers, Part H: Journal of Engineering in Medicine. 2009;224(5):607-622. doi:10.1243/09544119jeim589
9. Maurer CR, Fitzpatrick JM, Wang MY, Galloway RL, Maciunas RJ, Allen GS. Registration of head volume images using implantable fiducial markers. IEEE Transactions on Medical Imaging. 1997;16(4):447-462. doi:10.1109/42.611354
10. Pfisterer WK, Papadopoulos S, Drumm DA, Smith K, Preul MC. Fiducial Versus Nonfiducial Neuronavigation Registration Assessment and Considerations of Accuracy. Operative Neurosurgery. 2008;62(suppl_1):ONS201-ONS208. doi:10.1227/01.neu.0000317394.14303.99
11. Gumprecht HK, Widenka DC, Lumenta CB. Brain Lab VectorVision Neuronavigation System: Technology and Clinical Experiences in 131 Cases. Neurosurgery. 1999;44(1):97-104. doi:10.1097/00006123-199901000-00056
12. Grunert P, Darabi K, Espinosa J, Filippi R. Computer-aided navigation in neurosurgery. Neurosurgical Review. 2003;26(2):73-99. doi:10.1007/s10143-003-0262-0
13. Mezger U, Jendrewski C, Bartels M. Navigation in surgery. Langenbeck's Archives of Surgery. 2013;398(4):501-514. doi:10.1007/s00423-013-1059-4
14. Omay SB, Barnett GH. Surgical navigation for meningioma surgery. Journal of Neuro-Oncology. 2010;99(3):357-364. doi:10.1007/s11060-010-0359-6
15. Maciunas R. Computer-assisted neurosurgery. Clin Neurosurg. 2006;(53):267-271
16. Kelly PJ, Kall BA, Goerss SJ. Results of Computed Tomography-based Computer-assisted Stereotactic Resection of Metastatic Intracranial Tumors. Neurosurgery. 1988;22(1):7-17. doi:10.1227/00006123-198801000-00002
17. Wang MY, Maurer CR, Fitzpatrick JM, Maciunas RJ. An automatic technique for finding and localizing externally attached markers in CT and MR volume images of the head. IEEE Transactions on Biomedical Engineering. 1996;43(6):627-637. doi:10.1109/10.495282
18. American College of Radiology. ACR Practice Parameter for the performance of brain sterotactic radiosurgery
19. American College of Radiology. ACR–ASNR–SPR practice parameter for the performance and interpretation of magnetic resonance imaging (MRI) of the brain
20. PRACTICE PARAMETER 1 Cervicocerebral MRA. https://www.acr.org/-/media/ACR/Files/Practice-Parameters/cervicocerebralmra.pdf?la=en
21. PRACTICE PARAMETER 1 Cervicocerebral CTA. https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CervicoCerebralCTA.pdf?la=en

Sinus and Facial Imaging (HD-29)

Sinus and Facial Imaging (HD-29.1)

HD.SI.0029.1.C
v2.0.2024

- CT Maxillofacial without contrast (CPT® 70486) or limited CT Sinus without contrast (CPT® 76380) is supported for ANY of the following:[3]
  - Acute sinusitis without resolution of symptoms after a minimum of 4 weeks of treatment (Treatment can include an appropriate course and duration of empiric oral antibiotic, topical intranasal steroid, and/or nasal saline rinses.)
  - Concern for potential or suspected complicated sinusitis, which is sinusitis with actual or threatened orbital or intracranial extension (See **Background and Supporting Information** below)
  - Recurrent sinusitis (4 or more episodes of acute bacterial rhinosinusitis within the past 12 months without symptoms or signs between episodes)
    - In practice, recurrent acute exacerbations of chronic rhinosinusitis are seen as well as recurrent acute rhinosinusitis with disease free intervals between the acute episodes. CT Maxillofacial without contrast (CPT® 70486) may still be indicated under chronic sinusitis definitions.[6]
  - 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 (see **Taste and Smell Disorders (HD-2.1)** if anosmia, hyposmia, or dysosmia is an isolated symptom)
    - (**Note**: A trial of antibiotic therapy is not required prior to imaging if individual meets criteria for chronic sinusitis)
  - Follow up on incidentally noted sinus pathology (i.e. mucosal thickening, partial opacification of a sinus, or other indeterminate finding in incompletely visualized sinuses) on other studies not performed for the purpose of evaluating sinus pathology, such as MRI Brain for headache, when requested by ENT for clinical correlation.
- 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 as the main symptom, and suspected Upper Airway Cough Syndrome (UACS) as the etiology, see **Cough (CH-3.1)** in the Chest Imaging Guidelines.

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com

- CT Maxillofacial with contrast (CPT® 70487) if ANY of the following is present:
  - Orbital or facial cellulitis
  - Proptosis
  - Abnormal visual examination
  - Ophthalmoplegia
  - Immunocompromised individual
  - Fungal or vascular lesions visualized in nasal cavity
- CT Maxillofacial without contrast (CPT® 70486) **OR** CT Maxillofacial with contrast (CPT® 70487) **OR** MRI Orbits/Face/Neck without and with contrast (CPT® 70543):
  - Sinonasal obstruction, polyp, or suspected mass
  - Suspected orbital complication
  - Suspected invasive fungal sinusitis
  - Cystic fibrosis
  - Osteomyelitis (MRI is the preferred modality) and odontogenic infections, 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 Orbits/Temporal bone without contrast (CPT® 70480) or CT Orbits/Temporal bone 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)
      - If the most recent CT maxillofacial scan is greater than 6 months old or there is a change in clinical status as described above, a repeat diagnostic CT Maxillofacial without contrast (CPT® 70486) is supported for surgical planning.
      - Repeat CT Maxillofacial solely for the use of navigation during the sinus surgery (i.e. the most recent diagnostic CT Maxillofacial was not adequate due to lacking anatomic landmarks or insufficient thinnes of

cuts) should be requested with CPT® 77011, not the diagnostic CPT® code 70486.
    - 3D Rendering (CPT® 76377) should not be reported in conjunction with CPT® 77011 (or CPT® 70486 if used). The procedure inherently generates a 3D dataset.
- Complication of ABRS (acute bacterial rhinosinusitis) is suspected based on:
    - Any constellation of symptoms worrisome for intracranial extension of infection or meningitis (i.e. severe headache, photophobia, fever, neck stiffness)
    - 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)** and **Dental/Peridontal/Maxillofacial Imaging (HD-30.2)**

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.

References (HD-29)

v2.0.2024

1. Rosenfeld RM, Piccirillo JF, Chandrasekhar SS, et al. Clinical Practice Guideline (Update): *Adult Sinusitis*. Otolaryngology–Head and Neck Surgery. 2015;152(2_suppl):S1-S39. doi:10.1177/0194599815572097
2. Desrosiers M, Evans GA, Keith PK, et al. Canadian clinical practice guidelines for acute and chronic rhinosinusitis. *Allergy, Asthma & Clinical Immunology*. 2011;7(1). doi:10.1186/1710-1492-7-2
3. Expert Panel on Neurologic Imaging:, Kirsch CFE, Bykowski J, et al. ACR Appropriateness Criteria® Sinonasal Disease. J Am Coll Radiol. 2017;14(11S):S550-S559. doi:10.1016/j.jacr.2017.08.041
4. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck
5. Abdalkader M, Xie J, Cervantes-Arslanian A, Takahashi C, Mian AZ. Imaging of Intracranial Infections. *Seminars in Neurology*. 2019;39(03):322-333. doi:10.1055/s-0039-1693161
6. Wu D, Bleier B, Wei Y. Definition and characteristics of acute exacerbation in adult patients with chronic rhinosinusitis: a systematic review.*J Otolaryngol Head Neck Surg*. 2020;49(1):62. Published 2020 Aug 18. doi:10.1186/s40463-020-00459-w

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Temporomandibular Joint Disease (TMJ) and Dental/Periodontal/Maxillofacial Imaging (HD-30)

Temporomandibular Joint Disease (TMJ) (HD-30.1)

HD.TJ.0030.1.A
v2.0.2024

- 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
- For TMJ imaging in patients with Juvenile Idiopathic Arthritis (see **Temporomandibular Joint (TMJ) Imaging in Children (PEDHD-25)** in the Pediatric Head Imaging Guidelines)
    - MRI TMJ (CPT® 70336) is indicated annually for detecting silent TMJ arthritis in children and young adults with juvenile idiopathic arthritis as requested by a rheumatologist and/or oral/maxillofacial surgeon (OMS) and/or any provider in consultation with a rheumatologist or OMS.
    - Repeat imaging with MRI TMJ (CPT® 70336) in patients with JIA is indicated for any of the following:
        - Change in signs or symptoms suggesting progression of disease
        - To monitor the effects of treatment[11]
    - Bone Scintigraphy/Bone Scan 3 Phase Study (CPT® 78315) in individuals over 12 years of age is indicated in anticipation or consideration of surgery.
- 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 indicated in anticipation or consideration of surgery[13]

Dental/Periodontal/Maxillofacial Imaging (HD-30.2)

HD.TJ.0030.2.C
v2.0.2024

- 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
  - Post-operative imaging, including dental implants[14,15]
- Cone Beam CT: Report with CPT® Codes: CPT® 70486, CPT® 70487, CPT® 70488, CPT® 70480, CPT® 70482 (see **Cone Beam Computed Tomography (CBCT) (HD-24.7)**)
- 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

References (HD-30)

v2.0.2024

1. De Vos W, Casselman J, Swennen GRJ. Cone-beam computerized tomography (CBCT) imaging of the oral and maxillofacial region: A systematic review of the literature. *International Journal of Oral and Maxillofacial Surgery*. 2009;38(6):609-625. doi:10.1016/j.ijom.2009.02.028
2. Scrivani SJ, Keith DA, Kaban LB. Temporomandibular Disorders. *New England Journal of Medicine*. 2008;359(25):2693-2705. doi:10.1056/nejmra0802472
3. Bag AK. Imaging of the temporomandibular joint: An update. *World Journal of Radiology*. 2014;6(8):567. doi:10.4329/wjr.v6.i8.567
4. Horner K, O'Malley L, Taylor K, Glenny A-M. Guidelines for clinical use of CBCT: a review. *Dentomaxillofacial Radiology*. 2015;44(1):20140225. doi:10.1259/dmfr.20140225
5. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck.pdf
6. Guidelines for Diagnosis and Management of Disorders Involving the Temporomandibular Joint and Related Musculoskeletal Structures. *Cranio* ®. 2003;21(1):68-76. doi:10.1080/08869634.2003.11746234
7. Mercuri LG. Management of temporomandibular joint disorders. Journal of Oral Biology and Craniofacial Research. 2012;2(3):141-142. doi:10.1016/j.jobcr.2012.10.010
8. Gauer R, Semidey M. Diagnosis and Treatment of Temporomandibular Disorders. *Am Fam Physician*. 2015 Mar 15;91(6):378-386
9. National Academies of Sciences. Temporomandibular Disorders: Priorities for Research and Care. Priorities for Research and Care | The National Academies Press. https://doi.org/10.17226/25652. Published March 12, 2020
10. Whyte A, Boeddinghaus R, Bartley A, Vijeyaendra R. Imaging of the temporomandibular joint. Clin Radiol. 2021 Jan;76(1):76.e21-76.e35. doi: 10.1016/j.crad.2020.06.020
11. Schmidt C, Ertel T, Arbogast M, et al. The Diagnosis and Treatment of Rheumatoid and Juvenile Idiopathic Arthritis of the Temporomandibular Joint. Dtsch Arztebl Int. 2022;119(4):47-54. doi:10.3238/arztebl.m2021.0388
12. Kim IH, Singer SR, Mupparapu M. Review of cone beam computed tomography guidelines in North America. Quintessence Int. 2019 Jan 25;50(2):136-145. doi: 10.3290/j.qi.a41332
13. Almeida FT, Pacheco-Pereira C, Flores-Mir C, Le LH, Jaremko JL, Major PW. Diagnostic ultrasound assessment of temporomandibular joints: a systematic review and meta-analysis. Dentomaxillofac Radiol. 2019 Feb;48(2):20180144. doi: 10.1259/dmfr.20180144 9
14. Weiss R 2nd, Read-Fuller A. Cone Beam Computed Tomography in Oral and Maxillofacial Surgery: An Evidence-Based Review.*Dent J (Basel)*. 2019;7(2):52. Published 2019 May 2. doi:10.3390/dj7020052
15. Jacobs R, Salmon B, Codari M, Hassan B, Bornstein MM. Cone beam computed tomography in implant dentistry: recommendations for clinical use.*BMC Oral Health*. 2018;18(1):88. Published 2018 May 15. doi:10.1186/s12903-018-0523-5
16. Liu P, Shi J. Growth trends analysis of unilateral condylar hyperplasia followed up with planar scintigraphy: Retrospective overview of 249 cases. *Medicine (Baltimore)*. 2021;100(51):e28226. doi:10.1097/MD.0000000000028226

Eye Disorders and Visual Loss (HD-32)

Eye Disorders and Visual Loss (HD-32.1)

HD.VL.0032.1.A
v2.0.2024

- 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. An exam performed by a Neuro-Ophthalmologist, Neurologist, or an Optometrist meets this requirement.
- MRI Orbits/Face/Neck without contrast (CPT® 70540) **OR** MRI Orbits/Face/Neck without and with contrast (CPT® 70543) **OR** CT Orbits/Temporal bone with contrast (CPT® 70481) **OR** CT Orbits/Temporal bone without contrast (CPT® 70480) **AND/OR** MRI Brain without contrast (CPT® 70551) **OR** MRI Brain with and without contrast (CPT® 70553):[1]
  - Unexplained vision loss
  - Optic atrophy (Cranial Nerve II)
  - Optic neuropathy (Cranial Nerve II)
  - Papilledema/optic disc swelling (Cranial Nerve II) (see **Cranial Neuropathies (HD-1.1)** and **Papilledema/Pseudotumor Cerebri (HD-17.1)**)
  - Afferent Pupillary Defect (APD) or Relative Afferent Pupillary Defect (RAPD)
  - Chiasmal symptoms/signs (including bitemporal field deficit)
  - Ophthalmoplegia, Diplopia, and/or Cranial nerve palsy (Specifically CN III, IV, and VI, see **Cranial Neuropathies HD-1.1)**)
  - Nystagmus[21]
- 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.[16]
- For suspected optic neuritis, MRI is the preferred modality (see **Multiple Sclerosis (MS) (HD-16.1)** and **Neuromyelitis Optica and NMO Spectrum Disorders (HD-16.2)**)
- Visual field 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/Face/Neck without contrast (CPT® 70540) or MRI Orbits/Face/Neck without and with contrast (CPT® 70543) or CT Orbits/Temporal bone with contrast (CPT® 70481):
  - Exophthalmos (including thyroid eye disease), enophthalmos or non-traumatic 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 Orbits/Temporal bone without contrast (CPT® 70480) and/or CT Head without contrast (CPT® 70450):
  - Orbital trauma with visual defect
  - Exophthalmos (including thyroid eye disease)
- CT Maxillofacial without and with contrast (CPT® 70488) or CT Maxillofacial without contrast (CPT® 70486) or CT Maxillofacial with contrast (CPT® 70487)[22,23]
  - For pre-operative planning for procedures including dacryocystorhinostomy (DCR) to correct nasolacrimal duct obstruction (NLDO)[22,23]
- 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.
- MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) or CTA Head (CPT® 70496) for suspicion of intracranial aneurysm, including Third nerve palsy with pupillary involvement (see **Intracranial Aneurysms (HD-12.1)**
- MRA Head (CPT®70544, CPT®70545, or CPT®70546) or CTA Head (CPT®70496) **AND/OR** MRA Neck (CPT®70547, CPT®70548, or CPT®70549) or CTA Neck (CPT® 70498) for evaluation of diplopia due to suspected stroke or TIA (see **Intracranial Aneurysms (HD-12.1)**)
- Amaurosis Fugax (see **Stroke/TIA (HD-21.1)**)
  - Individuals describe a transient darkening or loss of vision, typically monocular
- 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.[19,20] Any one of the following are supported when additional imaging is indicated:
  - MRI Orbits/Face/Neck without contrast (CPT® 70540)
  - MRI Orbits/Face/Neck without and with contrast (CPT® 70543)
  - CT Orbits/Temporal bone with contrast (CPT® 70481)
  - CT Orbits/Temporal bone without contrast (CPT® 70480)
  - CT head without contrast (CPT® 70450)
- Additional imaging indications include:
  - To reassess compressive optic neuropathy (Symptoms/Signs of compressive optic neuropathy include APD, decreased visual acuity, and/ or visual field defects)

  - For non-response to Tepezza (Teprotumumab), relapses, worsening proptosis, diplopia, lid retraction, or optic neuropathy
  - For surgical planning for orbital decompression, strabismus surgery or lid surgery
- 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
- Vasculitis including Temporal Arteritis (Giant Cell Arteritis) (see **Cerebral Vasculitis (HD-22.1)**)

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.
  - Suspicion for disorders involving both regions is needed to image both regions.
  - 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.

List of Abbreviations and Meanings:

Abbreviation	Meaning
AC	Anterior chamber
APD	Afferent pupillary defect (see RAPD)
BCVA	Best-corrected visual acuity

Abbreviation	Meaning
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	Dacryocystorhinostomy
DFE	Dilated fundus exam
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

Abbreviation	Meaning
MP	Membrane peel
MRD1	Margin-reflex distance from upper lid margin to pupillary light reflex
MRx	Manifest refraction
NI	No improvement
NLDO	Nasolacrimal duct obstruction
NSC or NS	Nuclear sclerotic cataract
OD	Right eye
OS	Left eye
ortho	Eyes are aligned on the same target
OCT	Optical Coherence Tomography
P	Periphery
PD	Prism diopter
ph or PH	Pinhole (crude assessment of best-corrected visual acuity)
PPV or PPVx	Pars plana vitrectomy
PVD	Posterior vitreous detachment
RAPD	Relative Afferent Pupillary Defect (see APD)
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

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

Abbreviation	Meaning
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
v2.0.2024

- 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/Face/Neck without contrast (CPT® 70540), MRI Orbits/Face/Neck without and with contrast (CPT® 70543) or CT Orbits/Temporal bone with contrast (CPT® 70481) for suspected orbital lesion or mass

References (HD-32)

HD.VL.0032.3.A

v2.0.2024

1. Expert Panel on Neurologic Imaging:, Kennedy TA, Corey AS, et al. ACR Appropriateness Criteria® Orbits Vision and Visual Loss. J Am Coll Radiol. 2018;15(5S):S116-S131. doi:10.1016/j.jacr.2018.03.023
2. Lee JH, Lee HK, Lee DH, Choi CG, Kim SJ, Suh DC. Neuroimaging Strategies for Three Types of Horner Syndrome with Emphasis on Anatomic Location. *American Journal of Roentgenology*. 2007;188(1):W74-W81. doi:10.2214/ajr.05.1588
3. Szatmáry G. Imaging in Patients With Visual Symptoms. CONTINUUM: Lifelong Learning in Neurology. 2016;22(5):1499-1528. doi:10.1212/con.0000000000000375
4. Kawasaki AK. Diagnostic Approach to Pupillary Abnormalities. CONTINUUM: Lifelong Learning in Neurology. 2014;20:1008-1022. doi:10.1212/01.con.0000453306.42981.94
5. Prasad S. Diagnostic Neuroimaging in Neuro-ophthalmic Disorders. CONTINUUM: Lifelong Learning in Neurology. 2014;20:1023-1062. doi:10.1212/01.con.0000453305.65851.1c
6. ACR-ASNR-SPR Practice Parameter for the Performance of Computed Tomography (CT) of the Extracranial Head and Neck Revised 2021. (Resolution 5). https://www.acr.org/-/media/ACR/Files/Practice-Parameters/CT-Head-Neck
7. Tamhankar MA, Volpe NJ. Management of acute cranial nerve 3, 4 and 6 palsies: role of neuroimaging. *Curr Opin Ophthalmol*. 2015;26(6):464–468. doi:10.1097/ICU.0000000000000200
8. Tamhankar MA, Biousse V, Ying GS, et al. Isolated third, fourth, and sixth cranial nerve palsies from presumed microvascular versus other causes: a prospective study. *Ophthalmology*. 2013;120(11):2264–2269. doi:10.1016/j.ophtha.2013.04.009
9. Pineles SL, Velez FG. Isolated Ocular Motor Nerve Palsies. *J Binocul Vis Ocul Motil*. 2018;68(3):70–77. doi:10.1080/2576117X.2018.1481266
10. Flaxel CJ, Adelman RA, Bailey ST, et al. Retinal and Ophthalmic Artery Occlusions Preferred Practice Pattern®. *Ophthalmology*. 2020;127(2):P259–P287. doi:10.1016/j.ophtha.2019.09.028
11. Dagi LR, Velez FG, Archer SM, et al. Adult Strabismus Preferred Practice Pattern®. *Ophthalmology*. 2020;127(1):P182–P298. doi:10.1016/j.ophtha.2019.09.023
12. Sadaka A, Schockman SL, Golnik KC. Evaluation of Horner Syndrome in the MRI Era. *Journal of Neuro-Ophthalmology.* 2017;37(3):268-272. doi:10.1097/wno.0000000000000503
13. Glisson CC. Approach to Diplopia. CONTINUUM: Lifelong Learning in Neurology. 2019;25(5):1362-1375. doi:10.1212/con.0000000000000786
14. Gross JR, Mcclelland CM, Lee MS. An approach to anisocoria. *Current Opinion in Ophthalmology*. 2016;27(6):486-492. doi:10.1097/icu.0000000000000316
15. Costello F, Scott JN. Imaging in Neuro-ophthalmology. CONTINUUM: Lifelong Learning in Neurology. 2019;25(5):1438-1490. doi:10.1212/con.0000000000000783
16. Expert Panel on Neurologic Imaging, Whitehead MT, Cardenas AM, et al. ACR Appropriateness Criteria® Headache. J Am Coll Radiol. 2019;16(11S):S364-S377. doi:10.1016/j.jacr.2019.05.030
17. Lee SY, Rhee CM, Leung AM, Braverman LE, Brent GA, Pearce EN. A review: Radiographic iodinated contrast media-induced thyroid dysfunction. J Clin Endocrinol Metab. 2015;100(2):376-383. doi:10.1210/jc.2014-3292
18. van der Molen AJ, Thomsen HS, Morcos SK; Contrast Media Safety Committee, European Society of Urogenital Radiology (ESUR). Effect of iodinated contrast media on thyroid function in adults. Eur Radiol. 2004 May;14(5):902-7. doi: 10.1007/s00330-004-2238-z
19. Teo HM, Smith TJ, Joseph SS Efficacy and Safety of Teprotumumab in Thyroid Eye Disease Ther Clin Risk Manag. 2021 17:1219-1230. doi: 10.2147/TCRM.S303057
20. Bednarczuk Z, Pearce, SH The knowns and unknowns of teprotumumab for thyroid eye disease Lancet Diabetes Endocrinol. 2021 9:323-325. doi: 10.1016/S2213-8587(21)00076-0
21. Lee AG, Brazis PW. Localizing forms of nystagmus: symptoms, diagnosis, and treatment.*Curr Neurol Neurosci Rep*. 2006;6(5):414-420. doi:10.1007/s11910-996-0022-y
22. Freitag SK, Roos JC. Preoperative imaging should be performed prior to surgery in all cases of acquired nasolacrimal obstruction-Yes.*Eye (Lond*). 2017;31(3):351-352. doi:10.1038/eye.2016.237
23. Choi SC, Lee S, Choi HS, Jang JW, Kim SJ, Lee JH. Preoperative Computed Tomography Findings for Patients with Nasolacrimal Duct Obstruction or Stenosis.*Korean J Ophthalmol*. 2016;30(4):243-250. doi:10.3341/kjo.2016.30.4.243

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

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

HD.AC.0033.1.A
v2.0.2024

- 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/Face/Neck 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.[7]
- 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)

v2.0.2024

1. Kesavadas C, Thomas B, Kapilamoorthy T, Hingwala D, Chatterjee S. Applications of 3D CISS sequence for problem solving in neuroimaging. *Indian Journal of Radiology and Imaging*. 2011;21(2):90. doi:10.4103/0971-3026.82283
2. Camelio S, Schmid UD, Horsfield MA, et al. Visualization of cranial nerves I-XII: value of 3D CISS and T2-weighted FSE sequences. *European Radiology*. 2000;10(7):1061-1067. doi:10.1007/s003300000452
3. Olson JJ, Kalkanis SN, Ryken TC. Congress of Neurological Surgeons Systematic Review and Evidence-Based Guidelines on the Treatment of Adults With Vestibular Schwannomas: Executive Summary. *Neurosurgery.* 2017;82(2):129-134. doi:10.1093/neuros/nyx586
4. Zou J, Hirvonen T. “Wait and scan” management of patients with vestibular schwannoma and the relevance of non-contrast MRI in the follow-up. *Journal of Otology.* 2017;12(4):174-184. doi:10.1016/j.joto.2017.08.002
5. Lin EP, Crane BT. The Management and Imaging of Vestibular Schwannomas. *American Journal of Neuroradiology*. 2017;38(11):2034-2043. doi:10.3174/ajnr.a5213
6. Goldbrunner R, Weller M, Regis J, et al. EANO guideline on the diagnosis and treatment of vestibular schwannoma. *Neuro-Oncology*. 2019;22(1):31-45. doi:10.1093/neuonc/noz153
7. Somers T, Kania R, Waterval J, Havenbergh TV. What is the Required Frequency of MRI Scanning in the Wait and Scan Management? J Int Adv Otol 2018; 14(1): 85-9. doi: 10.5152/iao.2018.5348

Pineal/Colloid Cysts (HD-34)

Pineal/Colloid Cysts (HD-34.1)

HD.PT.0034.1.A
v2.0.2024

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
  - Pre-operative 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

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

References (HD-34)

v2.0.2024

1. Ajtai B, Bertelson JA. Imaging of Intracranial Cysts. *CONTINUUM: Lifelong Learning in Neurology*. 2016;22(5):1553-1573. doi:10.1212/con.0000000000000372
2. Tanaka T, Arnold L, Gabriela Mazuru D, Golzy M, Carr SB, Litofsky NS. Pineal cysts: Does anyone need long-term follow up? *Journal of Clinical Neuroscience*. 2021;83:146-151. doi:10.1016/j.jocn.2020.10.051
3. Jussila M-P, Olsén P, Salokorpi N, Suo-Palosaari M. Follow-up of pineal cysts in children: is it necessary? *Neuroradiology*. 2017;59(12):1265-1273. doi:10.1007/s00234-017-1926-8

Arachnoid Cysts (HD-35)

Arachnoid Cysts (HD-35.1)

HD.AR.0035.1.A
v2.0.2024

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

- MRI Brain without contrast (CPT® 70551) or 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
  - Pre-operative planning
  - When requested by a specialist or any provider in consultation with a specialist

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924

References (HD-35)

v2.0.2024

1. Ajtai B, Bertelson JA. Imaging of Intracranial Cysts. *CONTINUUM: Lifelong Learning in Neurology*. 2016;22(5):1553-1573. doi:10.1212/con.0000000000000372
2. Hall S, Smedley A, Sparrow O, Mathad N, Waters R, Chakraborty A, Tsitouras V. Natural History of Intracranial Arachnoid Cysts. *World Neurosurg*. 2019 Jun;126:e1315-e1320. doi: 10.1016/j.wneu.2019.03.087

Sleep-Related Imaging (HD-37)

General Guidelines Sleep-Related Imaging (HD-37.1)

HD.SL.0037.1.A
v2.0.2024

- Hypersomnolence:
  - 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.
    - MRI Brain with and without contrast (CPT® 70553) **OR**
    - MRI Brain without contrast (CPT® 70551)
- Central Sleep Apnea:
  - 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.
    - MRI Brain with and without contrast (CPT® 70553) **OR**
    - MRI Brain without contrast (CPT® 70551)
- 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)[1] in predicting treatment success.
  - Nasoendoscopy (sedated and non-sedated with provocative maneuvers such as Mueller maneuver) has been helpful as well in this regard.[2]
  - Routine use of advanced imaging is not supported at this time.
- For suspected sleep-related seizures (see **Epilepsy and Other Seizure Disorders (HD-9)**)

References (HD-37)

HD.SL.0037.2.A

v2.0.2024

1. Guarda-Nardini L, Manfredini D, Mion M, Heir G, Marchese-Ragona R. Anatomically Based Outcome Predictors of Treatment for Obstructive Sleep Apnea with Intraoral Splint Devices: A Systematic Review of Cephalometric Studies. *Journal of Clinical Sleep Medicine*. 2015;11(11):1327-1334. doi:10.5664/jcsm.5198
2. Sutherland K, Vanderveken OM, Tsuda H, et al. Oral Appliance Treatment for Obstructive Sleep Apnea: An Update. *Journal of Clinical Sleep Medicine*. Published online February 15, 2014. doi:10.5664/jcsm.3460
3. Deak MC, Kirsch DB. Sleep-Disordered Breathing in Neurologic Conditions. *Clinics in Chest Medicine*. 2014;35(3):547-556. doi:10.1016/j.ccm.2014.06.009
4. Trotti LM, Bliwise DL. Brain MRI findings in patients with idiopathic hypersomnia.*Clin Neurol Neurosurg*. 2017;157:19-21. doi:10.1016/j.clineuro.2017.03.010
5. Kotuła J, Kuc AE, Lis J, Kawala B, Sarul M. New Sagittal and Vertical Cephalometric Analysis Methods: A Systematic Review.*Diagnostics (Basel)*. 2022;12(7):1723. Published 2022 Jul 15. doi:10.3390/diagnostics12071723
6. Ramar K, Dort LC, Katz SG, et al. Clinical Practice Guideline for the Treatment of Obstructive Sleep Apnea and Snoring with Oral Appliance Therapy: An Update for 2015.*J Clin Sleep Med*. 2015;11(7):773-827. Published 2015 Jul 15. doi:10.5664/jcsm.4858
7. Chen H, Eckert DJ, van der Stelt PF, Guo J, Ge S, Emami E, Almeida FR, Huynh NT (2020) Phenotypes of responders to mandibular advancement device therapy in obstructive sleep apnea patients: a systematic review and meta-analysis. Sleep Med Rev. 49:101229

400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 www.eviCore.com