CIGNA MEDICAL COVERAGE POLICIES - RADIOLOGY Head Imaging Guidelines

Effective Date: February 1, 2025





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 <u>Cigna CPT</u> <u>code list</u> for the current list of high-tech imaging procedures that EviCore reviews for Cigna.

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Head Imaging Guidelines	V1
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General Guidelines (HD-1)

Guideline

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Head Imaging Guidelines

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Abbreviations for Head Imaging Guidelines

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Abbrevia	tions for Head Imaging Guidelines	
ACTH	adrenocorticotropic hormone	
AD	Alzheimer's Disease	1
ADH	antidiuretic hormone	
AION	arteritic ischemic optic neuritis	1
AVM	arteriovenous malformation	
СВСТ	Cone-beam computerized tomography	
CMV	Cytomegalovirus	
CSF	cerebrospinal fluid	
СТ	computed tomography	
СТА	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]

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Abbrevia	tions for Head Imaging Guidelines	
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	
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)	1
SAH	subarachnoid hemorrhage	
SIADH	Syndrome of Inappropriate Antidiuretic Hormone Secretion	1

Head Imaging Guidelines

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Abbrevia	Abbreviations for Head Imaging Guidelines		
SLE	systemic lupus erythematosus		
TIA	transient ischemic attack		
TMJ	temporomandibular joint disease		
TSH	thyroid-stimulating hormone		
VBI	vertebrobasilar insufficiency		
VP	ventriculoperitoneal		
XRT	radiation therapy		

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General Guidelines (HD-1.0)

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

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General Guidelines – Anatomic Issues (HD-1.1)

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- If two studies using the same modality both cover the anatomic region of clinical interest, only one is generally needed, with the exception of the following scenarios:
 - CT Maxillofacial (CPT[®] 70486, CPT[®] 70487, or CPT[®] 70488) or CT 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.

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General Guidelines – Modality (HD-1.2)

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- MRI is preferable to CT for most indications. For exceptions, See <u>General</u> <u>Guidelines – CT Head (HD-1.4)</u>
- 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) 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.

Evidence Discussion (HD-1.2)

- MRI of the brain is the appropriate initial imaging study for diagnosis, characterization and surveillance of a variety of neurologic conditions, including, but not limited to: neoplastic conditions, evaluation of the brain parenchyma, meninges, ischemia and infarction, neurodegenerative disorders, hydrocephalus, demyelinating conditions, post-traumatic brain injury, inflammatory and autoimmune disorders and infectious disorders.
- MRI brain has some benefit over CT for determining age of intracranial hemorrhage, early stroke (via Diffusion imaging sequences), and detection of micro hemorrhage.
- MRI is also indicated for further characterization of abnormalities detected on other imaging tests such as CT or sonography.
- Limitations to MRI include artifacts due to motion and susceptibility effects, contrast complications, contraindication due to ferromagnetic devices or implants. Additionally, severe claustrophobia may require sedation in order to complete the study.

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General Guidelines – MRI Brain (HD-1.3)

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- MRI Brain with contrast (CPT[®] 70552) should not be ordered except to follow-up on a very recent abnormal or equivocal non-contrast MRI Brain.²⁹
- 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.

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General Guidelines – CT Head (HD-1.4)

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- Scenarios in which MRI is contraindicated (i.e. pacemakers, ICDs, cochlear implants, aneurysm clips, orbital metallic fragments, etc.)
- In urgent cases, CT Head, contrast as requested is supported [CT Head without and with contrast (CPT[®] 70470), CT Head with contrast (CPT[®] 70460) or CT Head without contrast (CPT[®] 70450)]
- CT Head without contrast (CPT[®] 70450) is supported for:
 - Mass effect
 - Blood/blood products
 - Urgent/emergent settings due to availability and speed of CT
 - Trauma
 - Recent hemorrhage, whether traumatic or spontaneous
 - Bony structures of the head evaluations including dystrophic calcifications
 - Hydrocephalus evaluation and follow-up (some centers use limited non-contrast "fast or rapid MRI" (CPT[®] 70551) to minimize radiation exposure in children).
 - Prior to lumbar puncture in individuals
 - 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 <u>Papilledema/Pseudotumor Cerebri</u> (HD-17.1) and <u>Eye Disorders and Visual Loss (HD-32.1)</u>

Evidence Discussion (HD-1.4)

- CT head is the preferred modality for evaluation of bony structures, acute intracranial hemorrhage, trauma, and detection of abnormalities associated with calcifications.
- This modality is also useful for follow up of intracranial hemorrhage, hydrocephalus shunts, and post-operative follow up.
- CT head provides more rapid detection of intracranial abnormalities in urgent or emergent situations.
- CT has less motion artifact than MRI due to its faster acquisition and better spatial resolution than MRI.
- Limitations of CT include lower early detection rates for occult fracture than MRI, ionizing radiation exposure, and lower contrast resolution than MRI.

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

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- MRA Head may be performed without contrast (CPT[®] 70544), with contrast (CPT[®] 70545), or without and with contrast (CPT[®] 70546)
- MRA Neck may be done 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 <u>Stroke/TIA (HD-21.1)</u>) including collateral assessment
 - Trigeminal neuralgia having failed medical therapy (see <u>Trigeminal Neuralgia and</u> 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 post-surgical/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

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- Eagle Syndrome Dynamic/positional CTA to assess for vascular compression (also known as bow-hunter's syndrome)¹² (see <u>Eagle Syndrome (Neck-10.3)</u>)
- 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 <u>Intracranial</u> <u>Aneurysms (HD-12.1)</u>)
- 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

Evidence Discussion (HD-1.5)

 Indications for cervicocerebral computed tomography angiography (CTA) of the head and neck vessels include the diagnosis, characterization and/or surveillance of a variety of vascular conditions, including, but not limited to, arterial aneurysms, dissections, ischemic stroke and transient ischemic attacks, vasculitis, traumatic vascular injuries, pulsatile tinnitus, tumors of vascular origin, and prior to surgical intervention. CTA may refer to arterial vessels (CTA) or evaluation of venous structures (CTV). Head Imaging Guidelines

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- CTA may be used as the initial imaging modality or as a follow up study for characterizing known disease or assessing changes over time.
- Depending on the indication, CTA may be limited to the head to avoid unnecessary radiation to the patient. Examples include surveillance of intracranial aneurysms (that are not located in the posterior circulation).
- Risks of CTA include exposure to ionizing radiation, thus, magnetic resonance angiography (MRA) is available as an alternative to reduce radiation exposure. In addition, MRA is an alternative for patients with iodinated contrast allergies or other contraindications to iodinated contrast.
- Magnetic resonance angiography (MRA) indications also cover a variety of vascular conditions of the head and neck, for diagnosis, characterization and surveillance, and may be used to evaluate either arterial (MRA) or venous structures (MRV).
- MRA, as an alternative modality, is noninvasive, and does not require iodinated contrast. Limitations include artifacts due to motion, slow or turbulent flow, and susceptibility effects, and claustrophobia. MRA may be performed without contrast or with gadolinium contrast. Gadolinium contrast administration is limited to those without renal impairment or known gadolinium contrast allergy. Additionally, MRA may not be a feasible option for those with contraindications to MRI such as incompatible pacemakers, cochlear implants, neurostimulators or other devices. In these scenarios, CTA may be the appropriate alternative.

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General Guidelines – PET Coding Notes (HD-1.6)

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

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General Guidelines – Other Imaging Situations (HD-1.7)

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Persistent Nausea and Vomiting

Screening for Metallic Fragments Before MRI

Gender Affirming Care Head and Neck Surgical Planning

3D Rendering

Eagle Syndrome

CSF Leak with or without Headache

Evidence Discussion (HD-1.7)

Persistent Nausea and Vomiting

 Nausea and vomiting, persistent, unexplained and a negative GI evaluation: MRI Brain without contrast (CPT[®] 70551) or MRI Brain without and with contrast (CPT[®] 70553) is supported

Screening for Metallic Fragments Before MRI

- 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

Gender Affirming Care Head and Neck Surgical Planning

- For gender affirming care procedure planning:
 - ANY or ALL of the following pre-operative CT requests are supported if the individual has a health plan benefit covering the gender affirming surgeries:^{31,32}
 - CT Maxillofacial without contrast (CPT[®] 70486)

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- CT Orbits/Temporal bone without contrast (CPT[®] 70480)
- CT Neck with contrast (CPT[®] 70491)
- CT Head without (CPT[®] 70450)
- 3D rendering (CPT[®] 76376 or CPT[®] 76377)
- Pre-operative imaging is not supported if the gender affirming surgeries are not health plan covered benefits.
- Requesting providers are encouraged to confirm eligibility with the member's health plan prior to service.

3D Rendering

- CPT[®] 76377 (3D rendering requiring image post-processing on an independent workstation) or CPT[®] 76376 (3D rendering not 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 pre-operative planning)
 - Complex joint fractures or pelvis fractures
 - Spine fractures (usually for pre-operative planning)
 - Complex facial fractures
 - Pre-operative planning for other complex surgical cases
 - Cerebral angiography: 3D rendering when performed in conjunction with conventional angiography (i.e.: conventional 4 vessel 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 or CPT[®] 76376) 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

Eagle Syndrome

- See Eagle Syndrome (Neck-10.3) in the Neck Imaging Guidelines.
- See <u>General Guidelines CT and MR Angiography (CTA and MRA) (HD-1.5)</u> for vascular imaging related to Eagle Syndrome.¹⁵

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CSF Leak with or without Headache

 CSF Leak with or without headache, see <u>Low Pressure Headache and CSF Leak</u> (HD-11.15)

Evidence Discussion (HD-1.7)

Neurologic evaluation of Nausea and vomiting

• In the evaluation of persistent, unexplained nausea and vomiting, an MRI brain is supported after a negative GI evaluation. Nausea and vomiting were reported as the initial symptom of a brain tumor in 5% of brain tumor cases. During the time course until diagnosis, nausea and vomiting is present in 25% of brain tumor cases.

Screening for metallic fragments

The American College of Radiology White Paper on MR safety advises that all
patients who have a history of orbit trauma by a potential ferromagnetic foreign body
for which they sought medical attention are to have their orbits cleared by either
a plain x-ray orbit films (2 views) or by a radiologist's review and assessment of a
prior CT or MR images obtained since the suspected traumatic event. Screening
for the presence of a metallic aneurysm clips with plain films of the skull is also is
recommended. Although CT is more sensitive than plain films, the radiation dose is
greatly increased.

3D Rendering

• 3-D/rotational angiography, as part of cerebral angiography, is also useful for radiation dose reduction during diagnostic and interventional neuroradiology procedures.

Gender affirming head and neck surgeries

 As the field has evolved, more centers are using frontal sinus setback as 90% of patients require frontal bone osteotomy and setback, based on their frontal bone anatomy. For individuals requiring bony manipulation, a fine-cut, non-contrast craniofacial CT scan from the vertex to the hyoid bone is used to map the bony framework. Virtual surgical planning improves efficiency, safety, and accuracy for frontal sinus setback and mandibular angle reduction surgeries. CT neck is indicated for laryngoplasty surgeries.

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Taste and Smell Disorders (HD-2)

Guideline

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HD.TS.0002.1.A v1.0.2025

- MRI Brain without and with contrast (CPT[®] 70553) or MRI Brain without contrast (CPT[®] 70551) AND/OR MRI Orbit/Face/Neck without (CPT[®] 70540) or MRI Orbit/Face/Neck 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)¹
- 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: <u>Neuro-COVID-19 and Sars-CoV-2 Vaccines (HD-14.2)</u> and <u>Stroke/TIA (HD-21.1)</u>)

Evidence Discussion (HD-2.1)

- Initial imaging of the olfactory nerve and pathway for unexplained unilateral or bilateral anosmia or for dysgeusia should utilize MRI brain and/or MRI orbits, face and neck. These imaging studies are supported by clinical evidence for the identification and characterization of a potential cranial nerve lesion.
- CT of the sinuses and face may be superior to identify fractures, inflammatory sinus disease, and other bony lesions in some cases.

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Ataxia (HD-3)

Guideline

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Head Imaging Guidelines

Ataxia (HD-3.1)

HD.AX.0003.1.A v1.0.2025

- Common manifestations include: poor coordination, an abnormal (including widebased) 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 <u>Multiple Sclerosis (MS) (HD-16.1)</u>
 - 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: <u>Head Trauma (HD-13.1)</u>)
- If brain tumor is suspected, see <u>Primary Central Nervous System Tumors</u> (<u>ONC-2.1</u>) in the Oncology Imaging Guidelines.
- For suspected Normal Pressure Hydrocephalus, see <u>Normal Pressure</u> <u>Hydrocephalus (NPH) (HD-8.4)</u>

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.

Evidence Discussion (HD-3.1)

- MRI brain is the preferred initial imaging modality for evaluation of ataxia when a central nervous system cause is suspected. MRI of the spinal cord, to include the cervical and thoracic spine, may also be added if clinically indicated.
- CT head is not recommended for the initial evaluation of non-traumatic ataxia due to inferior soft tissue resolution when compared to MRI Brain. In addition, MRI brain provides better visualization of the cerebellum and posterior fossa and is more sensitive for the detection of posterior fossa infarcts.

©2024 EviCore by EVERNORTH 400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 • In general, MRI is preferred over CT, unless there is a history of acute trauma or a contraindication to MRI.

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

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Autism Spectrum Disorders (HD-4.0)

HD.BD.0004.0.A

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- 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) OR MRI Brain without contrast (CPT[®] 70551) is indicated for ANY of the following:
 - New or worsening cognitive decline or focal neurologic findings documented on a pertinent physical²
 - PET imaging is considered not medically necessary in the evaluation of individuals with autism spectrum disorders.

Evidence Discussion (HD-4.0)

- While the diagnosis of Autism Spectrum Disorder is based on behavioral signs and symptoms, MRI brain with and without contrast is indicated for new or worsening focal neurological findings and/or loss of developmental milestones and/or regression. In these clinical situations, advanced imaging may be used to adjust a patient's treatment plan, without which their development may continue to regress.
- PET is considered not currently medically necessary in the evaluation of individuals with autism spectrum disorder PET imaging in this scenario would unnecessarily expose patients to radiation and provide no clinical utility related to autism spectrum disorder.

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Mental Health Related Disorders (HD-4.1)

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- Mental health diagnoses, to include Attention Deficit Hyperactivity Disorder (ADHD), do not routinely require advanced imaging.¹¹
- 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 refractory Obsessive Compulsive Disorder (OCD).¹⁰
 - 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)

Evidence Discussion (HD-4.1)

- There is no role for advanced imaging in Mental health workup (including ADHD). Unnecessary imaging has detrimental effects in that it provides no positive impact on outcomes/management and does expose patients to unnecessary radiation, contrast, and financial strain.
- It would be appropriate to utilize Advanced imaging (CT or MRI) in the following conditions.
 - Acute mental status change, disturbance in consciousness or arousal state

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- Psychiatric disorders with the following clinical presentations:
 - Acute psychosis
 - Late onset over age 40
 - Presentation of acute psychiatric symptoms with comorbid serious medical illness
 - Non-auditory hallucinations of unknown etiology
 - Nonresponse to adequate medication trials
 - Symptoms of an organic brain disorder (e.g., focal deficits, severe headache, or seizures)
- Advanced imaging may be medically necessary for electroconvulsive therapy clearance and prior to deep brain stimulation for medically refractory Obsessive Compulsive Disorder.

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Mental Status Change (HD-4.2)

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After a detailed history, which includes onset, duration, and timeframe (i.e., constant vs intermittent) **AND** bedside neurologic exam that includes a mental status evaluation providing a description of the level of alertness, other characteristics and/or cognitive testing, the following are supported:

Indication	Supported Imaging
Acute or worsening mental status change, initial or repeat imaging	 CT Head without contrast (CPT[®] 70450) OR If setting is urgent, CT Head contrast as requested (CPT[®] 70450 OR CPT[®] 70460 OR CPT[®] 70470) OR If MRI is contraindicated, CT Head contrast as requested (CPT[®] 70450 OR CPT[®] 70460 OR CPT[®] 70470) CT Head permitted even with prior MRI Brain imaging
	OR
	 MRI Brain without contrast (CPT[®] 70551) OR MRI Brain without and with contrast (CPT[®] 70553) <i>MRI Brain permitted even with prior head CT imaging</i>

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Indication	Supported Imaging
 *Presence of any Red Flag, including: Sudden language, focal motor, or sensory deficit – <u>Stroke/TIA (HD-21.1)</u> Headache – <u>Headaches with Red</u> <u>Flags (HD-11.2)</u> Hypertensive urgency – <u>Stroke/TIA</u> (HD-21.1) and <u>Sudden Onset of</u> <u>Headache (HD-11.3)</u> 	*See relevant guideline
 Fever/tachycardia, possible meningitis, or other CNS infection – <u>CNS and Head</u> <u>Infection (HD-14.1)</u> 	
 COVID-19 – <u>Neuro-COVID-19 and</u> <u>Sars-CoV-2 Vaccines (HD-14.2)</u> 	
Coagulopathy or anticoagulant use- Abnormal Blood Clotting (HD-11.9)	
 Pregnancy or post-partum – <u>Pregnancy</u> (HD-11.10) 	
 Known malignancy – <u>Low Grade</u> <u>Gliomas (ONC-2.2)</u>, <u>High Grade</u> <u>Gliomas (ONC-2.3)</u> and <u>Brain</u> <u>Metastases (ONC-31.3)</u> 	
 Trauma- <u>Head Trauma (HD-13.1)</u> 	
 Non-auditory hallucinations – <u>Mental</u> <u>Health Related Disorders (HD-4.1)</u> 	
 Suspected increased intracranial pressure – <u>Papilledema/Pseudotumor</u> <u>Cerebri (HD-17.1)</u> and <u>Hydrocephalus</u> <u>Shunts (HD-11.14)</u> 	
 Seizure/suspected seizure – <u>Epilepsy/</u> <u>Seizures (HD-9.1)</u> 	

Background and Supporting Information

This section refers to acute/subacute mental status change, which generally involves signs and symptoms which begin over minutes to days, and includes changes in behavior and alertness, agitation, and/or confusion – as opposed to chronic, progressive cognitive decline, as in dementia.

Head Imaging Guidelines

©2024 EviCore by EVERNORTH 400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 Page 35 of 326 www.EviCore.com 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.

Delirium and psychosis are defined as follows:

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

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

Of note, even a seemingly mild, reversible brain insult superimposed upon a chronic pathophysiologic process may cause a sudden mental status change, and head imaging may or may not be necessary, depending on the provider's pre-test suspicion of a significant new diagnosis.

Vagal Nerve Stimulators (VNS), which are FDA approved for treatment of depression, are included among potential treatments, which also include medication trials.

Evidence Discussion (HD-4.2)

- Advanced brain imaging is supported for acute onset of mental status change, or worsening symptoms in the setting of a known intracranial process with MRI brain with or without a previous CT head.
- Advanced imaging supported for mental status change with precipitating factors including suspected seizure, COVID related symptoms, head trauma, stroke, mass or known malignancy, suspected increased intracranial pressure, intracranial infection, hypertensive emergency, presence of coagulopathy, pregnancy and postpartum period, associated headache.
- According to the ACR, advantages of MRI for altered mental state include: 1) higher sensitivity for detection of ischemia, encephalitis, subtle cases of SAH; and 2) enhancement of pathology compared with CT. The disadvantages of MRI in this clinical scenario are the same as with MRI in general, including patient inconvenience

(longer examination time), imaging quality is susceptible to patients' movements, and implanted devices that are not MRI safe.

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Chiari and Skull-Base Malformations (HD-5)

Guideline

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Chiari Malformations (HD-5.1)

HD.CM.0005.1.A

v1.0.2025

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

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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.

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

HD.CM.0005.2.A v1.0.2025

• See Chiari Malformations (HD-5.1)

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Chiari III and IV Malformations (HD-5.3)

HD.CM.0005.3.A v1.0.2025

• See Chiari Malformations (HD-5.1)

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Basilar Impression/Basilar Invagination (HD-5.4)

HD.CM.0005.4.A v1.0.2025

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).¹³
- 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

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Platybasia (HD-5.5)

HD.CM.0005.5.A v1.0.2025

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 ($CPT^{\mathbb{R}}$ 70544, $CPT^{\mathbb{R}}$ 70545, or $CPT^{\mathbb{R}}$ 70546) **OR**
 - CTA Head (CPT[®] 70496) AND/OR
 - MRA Neck (CPT[®] 70547, CPT[®] 70548, or CPT[®] 70549) **OR**
 - CTA Neck (CPT[®] 70498)¹³

Background and Supporting Information

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

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Evidence Discussion (HD-5)

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- A CT scan of the head is less sensitive than an MRI of the brain for evaluation of intracranial structures, including major structural abnormalities of the posterior fossa.
- Neuroimaging in the initial evaluation of Chiari malformation should include the spinal cord due to the common occurrence or increased frequency of associated conditions such as cervical syrinx and tethered cord.
- For initial evaluation, treatment planning, and follow up, MRI is the preferred modality for malformations of the brain and cervicocranial junction. MRI is ideal for evaluating soft tissues, neural structures, and ligaments.
- As congenital brainstem and cerebellar anomalies are associated with spinal anomalies, MRI of the complete spine is helpful for diagnosis, follow up and treatment planning.
- A phase-contrast CSF flow study at the craniocervical junction is a supportive study for evaluation of Chiari malformation.
- Evaluation of cervicojunction anomalies, including basilar invagination and platybasia, may require more than one modality for diagnosis and surgical planning. CT characterizes osseous anatomy and may be helpful for surgical planning. MRI is preferred for evaluation of the soft tissues, neural structures and ligaments for these conditions. As craniocervical junction anomalies may lead to compression of adjacent vascular structures, CT- or MR-Angiography of the head and neck are useful for surgical planning.

Head Imaging Guidelines

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

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Facial Palsy (HD-6.1)

HD.FP.0006.1.A

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- MRI Brain without and with contrast (CPT[®] 70553) (with attention to posterior fossa and IACs) or MRI Brain without contrast (CPT[®] 70551) AND/OR MRI Orbit/Face/ Neck without contrast (CPT[®] 70540) or MRI Orbit/Face/Neck without and with 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 <u>Autoimmune/</u> <u>Paraneoplastic Encephalitis & NeuroInflammatory Disorders (HD-14.3)</u>)
- 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, presurgical 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 stroke² (see <u>General Guidelines- CT and MR Angiography (CTA and MRA)</u> (HD-1.5) and <u>Stroke/TIA (HD-21.1)</u>).

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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.

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Hemifacial Spasm (HD-6.2)

HD.FP.0006.2.A v1.0.2025

- 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
- For tardive dyskinesia, see Movement Disorders (HD-15.1)

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Evidence Discussion (HD-6)

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- Facial nerve palsy/Bell's Palsy, commonly referred to as Bell's Palsy, does not routinely require imaging as recommended by the American Academy of Neurology and the American Academy of Otolaryngology Head and Neck Surgery Foundation. Complete recovery typically occurs within 3 to 6 months.
- When imaging is indicated, MRI is the preferred modality for evaluating the facial nerve from its origin in the brainstem, through its intracranial and extracranial segments. This would include imaging of the brain, face or both areas concurrently. MRI is useful to exclude structural causes of facial nerve paralysis in the setting of red flags.
- Imaging is reserved for cases with "red flags," which include atypical, recurrent or persistent cases. Limiting imaging to those with "red flags" avoids unnecessary radiation exposure, identification of incidental findings, contrast reactions, and unnecessary costs. The risk of limiting imaging includes missing identifiable and treatable causes of facial paralysis. To mitigate this risk, clinical follow up is recommended at 3 months.
- MRI has sensitivity ranging from 73% to 100% in detecting peripheral spread of tumor.
- As the facial nerve courses through the temporal bone, CT temporal bone is useful to identify temporal bone fractures, bony anatomy, bone erosion and for surgical planning.
- Vascular imaging is helpful if stroke is clinically suspected.
- For evaluation of hemifacial spasm, MRA allows characterization of vascular loops compressing the facial nerve, with sensitivity >95% and correlates well with surgical findings.

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

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See <u>Recurrent Laryngeal Nerve Palsy in Neck-7.1</u>

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Dementia (HD-8)

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Dementia (HD-8.1)

HD.DM.0008.1.A

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- For acute mental status change, see <u>Mental Status Change (HD-4.2)</u> and <u>Stroke/</u> <u>TIA (HD-21.1)</u>
- 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 with 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 Eightitem Informant Interview to Differentiate Aging and Dementia (AD8) Dementia Score > 2.²⁴
- 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 <u>Normal Pressure</u> <u>Hydrocephalus (HD-8.4)</u>
- Quantitative Magnetic Resonance Image (MRI) Analysis of the Brain
 - Volumetric or quantitative analysis of the brain or temporal lobes and hippocampus may be ordered as Quantitative MRI Analysis of the Brain (CPT[®] 0865T or CPT[®] 0866T) or 3D rendering (CPT[®] 76376 and CPT[®] 76377).

Head Imaging Guidelines

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Background and Supporting Information

Mild Cognitive Impairment (MCI), also referred to as mild neurocognitive disorder, is marked by focal or multifocal cognitive impairment with minimal impairment of instrumental activities of daily living that do not cross the threshold for dementia.¹⁶

Dementia, or major neurocognitive disorder, as defined by the Diagnostic and Statistical Manual of Mental Disorders (DSM-5), refers to significant cognitive decline, with impairment in cognitive performance in domains including complex attention, executive function, learning and memory, language, perceptual-motor skills, or social cognition.¹⁶

Evidence Discussion (HD-8.1)

- The primary role of neuroimaging in the work up of patients diagnosed with dementia is to exclude other serious differential diagnosis such as tumors, subdural hematomas, and normal pressure hydrocephalus. The American Academy of Neurology (AAN) recommends the use of noncontrast CT or Brain MRI for aiding in the diagnosis of dementia. Cross sectional imaging may also identify characteristic brain atrophy patterns found in common neurodegenerative diseases and vascular insults. CT imaging may also be used when MRI scans are contraindicated.
- Volumetric MRI brain for the diagnosis of dementia is not currently recommended for routine clinical use by the AAN. There remains a significant evidence gap in the literature regarding clinical validation of volumetric MRI in the diagnosis of dementia. Their use remains limited to research studies.

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Dementia - PET (HD-8.2)

HD.DM.0008.2.A

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- Prior to consideration of Brain 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 > 2.²⁴
- 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.

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
 - 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.

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- 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 <u>Imaging Related to</u> <u>Alzheimer's Treatment with Amyloid Reduction Medications (HD-8.5)</u>).
 - Otherwise, these studies are **NOT** considered medically necessary for any of the following scenarios:
 - Screening for dementia
 - Diagnosis of dementia
 - 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.^{21,22,23}

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.
- Recent research has examined the utility of PET/MRI for evaluation of patients with Dementia. Due to the prolonged acquisition 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 patients 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.

Evidence Discussion (HD-8.2)

- Diagnosis is based on clinical features, neuropsychological testing, and brain imaging (preferably MRI) to rule out other structural disease. FDG-PET accurately discriminates Alzheimer's disease patients from normal subjects with a sensitivity of 96% and specificity of 100%.
- Metabolic (FDG) PET Brain is helpful by demonstrating patterns of abnormality more consistent with FTD than Alzheimer's disease. FDG-PET Brain has a sensitivity of 86% and specificity of 97.6% in evaluating individuals with FTD.2 The use of FDG-PET increases diagnostic accuracy and confidence for both AD and FTD.2 It is particularly helpful in cases of diagnostic uncertainty.
- Recent research has examined the utility of PET/MRI for evaluation of patients with dementia. Due to the prolonged acquisition 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. Utilization of PET/MRI provides greater confidence in reading images by permitting greater structural correlation. A recent study compared FDG-PET/CT and FDG-PET/MRI in a memory disorders clinic. The main findings were that FDG PET/MRI revealed more vascular pathology in 35% of patients, induced a change of the interpretation of FDG PET in 17% of patients, and was considered to influence patient management in 22% of patients.

Head Imaging Guidelines

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- 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 > $2^{.24}$

- 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

Evidence Discussion (HD-8.3)

- For suspected Lewy Body Dementia (LBD), a CT or MRI head is appropriate as the initial imaging study.
- To increase diagnostic accuracy of LBD, SPECT modalities are helpful for differentiating LBD from Alzheimer's dementia.
- Functional imaging of the dopamine transporter (DAT) (lodine-123 loflupane) using SPECT shows a deficiency in the nigrostriatal pathway in LBD. This is considered a second line imaging test after cross-sectional imaging has excluded other pathology, such as vascular lesions along the nigrostriatal pathway, which can lead to abnormal DAT images with false positive results.
- An abnormal DAT-SPECT scan has a sensitivity of 77.7% and a specificity of 90.4% for probable LBD.

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Normal Pressure Hydrocephalus (NPH) (HD-8.4)

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- 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^{18,19,20}
- 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 > 2.²⁴
- 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 pre-surgical 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 <u>Hydrocephalus Shunts (HD-11.14)</u> or <u>Low Pressure Headache and CSF Leak</u> (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.

Head Imaging Guidelines

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Evidence Discussion (HD-8.4)

- Initial neuroimaging for the evaluation of suspected Normal Pressure Hydrocephalus (NPH) includes CT head or MRI brain in patients with clinical symptoms and no explanation for hydrocephalus.
- Only a single modality study is indicated. The initial best modality is MRI Brain because of its higher intrinsic soft tissue resolution and because it can often be used as a pre-surgical exam.
- By using the appropriate single best test we avoid duplicate imaging and unnecessary radiation to the lens of the eye and other Head and neck structures.
- Cine MRI showing hyperdynamic aqueductal CSF flow can also help in identifying shunt-responsive NPH patients. The benefit of this exam is that it offers us functional information about CSF flow and can help improve patient outcomes.

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

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

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Donanemab (Kisunla®)

Indication	Supported Imaging
 Consideration of Donanemab (Kisunla[®]) therapy and ALL of the following are met:^{17,18} Patient age ≥59 years of age and ≤ 86 years of age MCI or Mild dementia due to AD Mini-Mental State Examination (MMSE) score ≥20 and ≤28 Progressive change in memory function for at least 6 months No history of prior intracerebral hemorrhage greater than 1 cm, severe white matter disease OR vasogenic edema Not currently taking another amyloid reducing drug The medication is prescribed by a neurologist 	 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 78814)
On Donanemab therapy prior to the 2 nd , 3 rd , 4 th and 7 th infusions ¹⁷	 MRI Brain without contrast (CPT[®] 70551) OR MRI Brain without and with contrast (CPT[®] 70553)
Follow up while on Donanemab 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

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Indication	Supported Imaging
Neurologic signs and/or symptoms occurring while on treatment with Donanemab ¹⁷	 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.
Follow up imaging during treatment at 6, 12 and 18 months ^{17,18}	Amyloid PET Brain (CPT [®] 78811 or 78814)

Lecenamab (Leqembi®)

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Indication	Supported Imaging	
Consideration of Lecanemab (Leqembi) therapy and ALL of the following are met: ^{27,30}	Baseline MRI Brain (<i>within 3 months of medication initiation</i>)	
 Patient is ≥ 50 years of age and ≤ 90 years of age 	 MRI Brain without contrast (CPT[®] 70551) OR 	
 MCI or Mild dementia due to AD Qualifying test scores include Mini- 	 MRI Brain without and with contrast (CPT[®] 70553) 	
Mental Status Exam (MMSE) with score ≥ 22, Clinical Dementia Rating global	AND/OR	
score 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	 Amyloid PET Brain (CPT[®] 78811 or CPT[®] 78814) 	
 Patient has no history of brain hemorrhage, bleeding disorder or recent history (within 12 months) of stroke or transient ischemic attacks or any history of seizures 		
 Not currently taking another amyloid reducing drug 		
 The medication is prescribed by a neurologist 		

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Indication	Supported Imaging
On Lecanemab therapy prior to 5 th , 7 th and 14 th 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)

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.²⁵

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.²⁵

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

Page 69 of 326 www.EviCore.com the imaging time frame determined by the treating physician. Resumption of dosing is guided by clinical judgment.²⁵

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.²⁵

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.²⁵ The risk of ARIA is increased in apolipoprotein E #4 (ApoE #4) homozygotes.²⁵

Evidence Discussion (HD-8.5)

- Structural brain imaging in the work up of patients diagnosed with dementia is
 primarily to exclude other significant intracranial abnormalities. A brain MRI will assist
 with the diagnosis of dementia by excluding structural pathology such as tumors or
 subdural hematomas.
- Amyloid related imaging abnormalities (ARIA) have been associated with treatment by amyloid reduction medications. ARIA usually occurs early in treatment and may be asymptomatic although serious and life-threatening events may occur. Screening brain MRI 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.
- Amyloid PET brain is a form of molecular imaging, which uses a tracer that binds to amyloid plaques in the brain. At the present time, the use of Amyloid PET brain is limited to confirming the presence of amyloid in the brain, in those with mild cognitive impairment due to Alzheimer's disease or mild dementia due to Alzheimer's disease, prior to treatment with amyloid reducing medications.

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Epilepsy/Seizures (HD-9)

Guideline

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Epilepsy/Seizures (HD-9.1)

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- 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
 - New neurologic deficit or no return to previous neurologic baseline¹
 - 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 or neurosurgeon, or any provider in consultation with a neurologist or neurosurgeon.
 - 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 <u>Head</u> <u>Trauma (HD-13.1)</u>
- 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[®] 76376 and CPT[®] 76377).¹²

Evidence Discussion (HD-9.1)

 The use of advanced imaging is indicated for the initial evaluation of adults with seizure. Unenhanced CT is more readily available so is usually the initial imaging examination performed for adults presenting with first seizure. In the acute setting this primary exam is utilized to exclude conditions requiring urgent or emergent intervention, such as a bleed. CT is also appropriate if MRI is contraindicated and to evaluate seizure foci that contain dystrophic calcifications, such as oligodendrogliomas and tuberous sclerosis, yet the overall success of CT in detecting focal lesions in epilepsy is low, at approximately 30%. In studies where patients were

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evaluated with both MRI and CT, CT failed to detect potentially epileptogenic lesions identified on MRI 16-42% of the time1.Therefore, MRI of the brain is the study of choice to evaluate new-onset seizures (when available), refractory or drug resistant seizures, prior to discontinuation of anti-epileptic therapy, and known seizure with change in semiology.

• If CT is initially performed, it can be followed by an MRI. If an MRI not using the "Epilepsy Protocol" is initially performed, it can be followed by an MRI with the Epilepsy Protocol for greater sensitivity of detection of epileptogenic lesions. The failure rate for detection of lesions improving from 39% to 91% with epilepsy-trained radiologist reading MRI images obtained using a specialized, epilepsy protocol.

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

HD.EP.0009.2.A

v1.0.2025

- The following requests are supported for consideration of potential surgery:
 - MRI Brain without contrast (CPT[®] 70551) OR MRI Brain with and without contrast (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)
 - 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
 - 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 <u>Functional MRI (fMRI) (HD-24.2)</u>)
 - 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) or CPT[®]76376 (3D rendering not 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 <u>Neurosurgical Imaging (HD-28.1)</u> and <u>Neuronavigation</u> (HD-28.2)
 - Post-operative imaging including after intracranial (EEG) monitoring per neurosurgeon or neurologist or any provider in consultation with neurosurgeon or neurologist.

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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, the guidelines only require review 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)

Evidence Discussion (HD-9.2)

- MRI head for the initial imaging of patients with known seizure disorder requiring surgical planning to identify the seizure focus including tumor, hippocampal sclerosis and vascular lesions. Follow-up MRI after a previous standard protocol study if performed with special "Epilepsy Protocol" can provide additional information.
- FDG-PET/CT brain may be complementary as a functional tool to structural imaging using MRI to localize the focus of refractory seizure activity, with reported sensitivities of PET in the assessment of temporal lobe epilepsy ranging from 87% to 90% and extra-temporal lobe epilepsy ranging from 38% to 55%.
- PET/MRI, performed as MRI brain without contrast, or with and without contrast, coregistered with FDG-PET brain, increased the sensitivity of brain MRI in 60% of non-

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lesional patients and is therefore supported for pre-surgical evaluation of refractory seizures.

 Ictal SPECT, Functional MRI (fMRI) and MRI brain co-registered with Magnetoencephalography (MEG) are also useful to further identify the seizure focus as well as eloquent areas of the cortex that are essential for language, motor function and memory in surgical candidates when done as a replacement for the higher risk Wada test or direct electrical stimulation mapping.

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

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Trigeminal Neuralgia/Trigeminal Neuropathy (HD-10.1)

HD.TM.0010.1.A

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- 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 neuropathy⁵
 - Trigeminal neuralgia
 - Trigeminal neuralgia which involves the ophthalmic nerve, (periorbital or forehead pain), once post-herpetic neuralgia (a complication of shingles) 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 foramina⁵
- Contrast-enhanced navigation protocol CT (CPT[®] 76497) for gamma knife stereotactic radiosurgery for trigeminal neuralgia⁵, (see also, <u>Neuronavigation</u> (<u>HD-28.2</u>) and <u>Post Operative Imaging (HD-28.3</u>)) for post-treatment imaging studies
- MRI Cervical spine without contrast (CPT[®] 72141) OR MRI Cervical spine without and with contrast (CPT[®] 72156) for suspected lesion of the spinal trigeminal tract and nucleus.⁵
- 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

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The spinal trigeminal tract and nucleus extend from the midpons caudally into the upper cervical cord at the C2-4 levels. For suspected lesions of the spinal trigeminal tract and nucleus, imaging the brain stem and the cervical spinal cord is supported.⁵

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Glossopharyngeal Neuralgia/ Glossopharyngeal Neuropathy (HD-10.2)

HD.TM.0010.2.A

v1.0.2025

- 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 neuropathy⁵
- 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 ligament⁵ (see also <u>Eagle Syndrome (Neck-10.3)</u>)
- 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 neuralgia⁵

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

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Evidence Discussion (HD-10)

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- The American Academy of Neurology recommends routine use of MRI in the evaluation of patients with trigeminal neuralgia. Neuroimaging identifies structural causes in up to 15% of patients. The most commonly identified abnormalities include cerebellopontine angle tumors and multiple sclerosis plaques.
- MRI brain and/or MRI orbits, face and neck are necessary for direct visualization of the entire course of the trigeminal nerve.
- MRA head, when combined with MRI brain for evaluation of vascular compression of the trigeminal nerve, has sensitivity of 97-100% and specificity of 100%. CTA is less commonly performed concurrently with MRI of the trigeminal nerve.
- CT maxillofacial may be complementary to MRI in characterizing skull base erosions, calcifications, and skull base foramina.
- In the evaluation of glossopharyngeal neuralgia, MRI of the brain and/or MRI orbits, face and neck, allows direct visualization of the entire course of the glossopharyngeal nerve. Imaging should include the pharynx and larynx to exclude a neck mass. To further evaluate bony anatomy, calcifications, and the stylohyoid ligament, CT neck is also appropriate. MRA head and neck is helpful to exclude neurovascular compression in patients with glossopharyngeal neuralgia

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Headache (HD-11)

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Headache General Guidelines (HD-11.0)

HD.HA.0011.0.C v1.0.2025

- 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 <u>Headaches with Red Flags (HD-11.2)</u> and <u>Advanced Imaging</u> <u>Indications Related to Migraines (HD-11.17)</u>)
 - 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

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Headache and Suspected Vascular Dissection (HD-11.1)

HD.HA.0011.1.A

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- CTA Neck (CPT[®] 70498) and MRA Neck (CPT[®] 70547, CPT[®] 70548, or CPT[®] 70549) are indicated 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 vertebro-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 <u>Stroke/TIA (HD-21.1)</u>, <u>Sudden Onset of Headache (HD-11.3)</u>, <u>New Headache Onset Older than Age 50 (HD-11.7)</u>, <u>Abnormal Blood Clotting (HD-11.9)</u>, <u>Pregnancy (HD-11.10)</u>, <u>Physical Exertion (HD-11.11)</u>, and <u>Systemic Infections (HD-11.13)</u>

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Headaches with Red Flags (HD-11.2)

HD.HA.0011.2.A

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- 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 <u>Primary Central Nervous System Tumors – General Considerations</u> (ONC-2.1) in the Oncology Imaging Guidelines and <u>Stroke/TIA (HD-21.1)</u>)
 - 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 <u>Cancer or Immunosuppression</u> (HD-11.8))
 - Sudden onset (see <u>Sudden Onset of Headache (HD-11.3)</u>)
 - New onset age >50 (see <u>New Headache Onset Older than Age 50 (HD-11.7)</u> and <u>Migraine Exceptions (HD-11.17)</u>)
 - History of head trauma (see <u>Headaches Associated with Head Trauma</u> (HD-11.12), and <u>Head and Facial Trauma (HD-13)</u>)
 - Headache precipitated by cough or valsalva, physical exertion, or sexual activity (see <u>Physical Exertion (HD-11.11)</u>)
 - Currently pregnant (including pregnancy and the immediate postpartum period) (see <u>Pregnancy (HD-11.10)</u>)
 - Hypercoagulable state or bleeding disorder (see <u>Abnormal Blood Clotting</u> (HD-11.9))
 - New persistent headache (see <u>Migraine Exceptions (HD-11.17)</u>)
 - Headache awakens individual from sleep (see <u>Sudden Onset of Headache</u> <u>(HD-11.3)</u>)
- 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

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- 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)

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V1.0.2025

- 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 (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 artery 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 high⁸
- 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 vascular imaging indications for headaches require additional information.
 - See <u>Stroke/TIA (HD-21.1)</u>, <u>New Headache Onset Older than Age 50 (HD-11.7)</u>, <u>Abnormal Blood Clotting (HD-11.9)</u>, <u>Pregnancy (HD-11.10)</u>, <u>Physical Exertion</u> (HD-11.11), <u>Intracranial Aneurysms (HD-12.1)</u> and <u>Systemic Infections</u> (HD-11.13)

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Trigeminal Autonomic Cephalgias (HD-11.4)

HD.HA.0011.4.A v1.0.2025

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

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 paroxsysmal and continua.

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

HD.HA.0011.5.A

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- Skull base, orbital, periorbital or oromaxillary¹ 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 (CPT[®] 70553 and/or CPT[®] 70543) OR
 - MRI Brain and/or Orbits/Face/Neck without contrast (CPT[®] 70551 and/or CPT[®] 70540)
 - 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)

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

HD.HA.0011.6.A v1.0.2025

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

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- For new onset headache in individuals older than 50 years of age:
 - MRI Brain without and with contrast (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 <u>Cerebral Vasculitis (HD-22)</u>

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Cancer or Immunosuppression (HD-11.8)

HD.HA.0011.8.A

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

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Abnormal Blood Clotting (HD-11.9)

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- 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.).

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Pregnancy (HD-11.10)

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- 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 preeclampsia, reversible cerebral vasoconstriction syndrome, and cerebral venous thrombosis, as well as idiopathic intracranial hypertension^{1,6}
- For post LP/epidural anesthesia, see <u>Low Pressure Headache and CSF Leak</u> (HD-11.15)

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Physical Exertion (HD-11.11)

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- 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 (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

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Headaches Associated With Head Trauma (HD-11.12)

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- 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 injury¹⁴ that does not meet criteria under <u>Head and Facial Trauma (HD-13)</u>, other subsections may apply including, but not exclusive to: <u>Headaches with Red</u> <u>Flags (HD-11.2)</u> and <u>Sudden Onset of Headache (HD-11.3)</u>

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Systemic Infections (HD-11.13)

HD.HA.0011.13.A

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- Headaches in the setting of acute, subacute, or chronic systemic infections:
 - MRI Brain without and with contrast (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 <u>General Guidelines CT</u> <u>Head (HD-1.4)</u> 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 <u>Neuro-COVID-19 and Sars-CoV-2 Vaccines (HD-14.2)</u> for headache related to neuro-COVID-19 or SARS-CoV-2 vaccines

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Hydrocephalus Shunts (HD-11.14)

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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.

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- Nuclear Medicine Shunt Evaluation (CPT[®] 78645) and CSF Flow SPECT (CPT[®] 78803) for the following:
 - Suspected malfunction of ventriculoperitoneal, ventriculopleural, or ventriculovenous shunts.
- 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 noncommunicating hydrocephalus can be difficult with MRI Brain or CT Head alone, and modalities which visualize CSF flow may be useful such as cisternography or CT cisternography.

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Low Pressure Headache and CSF Leak (HD-11.15)

HD.HA.0011.15.A

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

Supported Imaging	
MRI brain without and with contrast (CPT [®] 70553)	
 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) 	
CT Cervical Spine with contrast (CPT [®] 72126)	
AND/OR	
CT Thoracic Spine with contrast (CPT [®] 72129)	S
AND/OR	De
CT Lumbar Spine with contrast (CPT [®] 72132)	el:
Radionucleotide cisternogram (CPT [®] 78630)	ead Imaging Guidelines
<u>م</u>	
CT Head with contrast (CPT [®] 70460)	
CT Maxillofacial with contrast (CPT [®] 70487)	
OR	
CT Temporal Bone with contrast (CPT [®] 70481)	0
	 MRI brain without and with contrast (CPT[®] 70553) 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) CT Cervical Spine with contrast (CPT[®] 72126) AND/OR CT Thoracic Spine with contrast (CPT[®] 72129) AND/OR CT Lumbar Spine with contrast (CPT[®] 72132) Radionucleotide cisternogram (CPT[®] 78630) CT Head with contrast (CPT[®] 70460) OR CT Maxillofacial with contrast (CPT[®] 70487)

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Indication	Supported Imaging
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 <u>Hydrocephalus Shunts (HD-11.14)</u>)

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

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

HD.HA.0011.16.A

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- Brain imaging should follow applicable sections in <u>Headache (HD-11)</u>
- 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 <u>Red Flag Indications (SP-1.2)</u>, and clinical re-evaluation after treatment period.
 - See <u>Neck (Cervical Spine) Pain Without/With Neurological Features</u> (Including Stenosis) (SP-3.1) and <u>Neck (Cervical Spine) Trauma (SP-3.2)</u> 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)
 - **<u>Red Flag Indications (SP-1.2)</u>** 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 <u>Myelopathy (SP-7.1)</u> 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).

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- 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)

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Advanced Imaging Indications Related To Migraines (HD-11.17)

HD.HA.0011.17.A

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- 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 <u>Headaches with Red Flags (HD-11.2)</u>).
 - See below for advanced imaging indications related to migraines.
- New migraine with age ≥50 (see <u>New Headache Onset Older than Age 50</u> (HD-11.7))
- Change in frequency or severity of migraine (see <u>Headaches with Red Flags</u> (HD-11.2))
- MRI Brain without (CPT[®] 70551) or MRI Brain with and without (CPT[®] 70553) or CT Head without (CPT[®] 70450) for the following:
 - Unusual, prolonged or persistent aura (greater than 60 minutes) (See <u>Background</u> and <u>Supporting Information</u>)
 - 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 <u>Head Trauma (HD-13.1)</u> and <u>Headaches Associated with Head Trauma</u> (HD-11.12)

Head Imaging Guidelines

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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 (double vision) and retinal (visual complaints)

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Evidence Discussion (HD-11)

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- The majority of headaches are due to benign causes and are self-limited. The yield of positive findings on a CT head for evaluation of headache in the emergency department setting ranged from 7% to 13%.
- The American College of Radiology in the Choosing Wisely campaign recommends against imaging for primary headache syndromes in the absence of red flags and with a normal neurologic exam. The American Headache Society and Choosing Wisely Task Force stated that the overuse or misuse of imaging studies for headache was the most commonly mentioned problem. Overuse of CT head was identified as the main concern. The authors note that a single CT scan of the head exposes patients to an average of 2 mSV of radiation, the equivalent of 8 months of background radiation.
- Incidental findings are common and can result in anxiety for the patient, additional referrals and specialist consults, and more imaging studies. Incidental findings on MRI occur in 2% of the general population.
- The American Headache Society and the American Academy of Neurology recommend neuroimaging in patients with headaches with atypical features, red flags and/or abnormal neurologic exam findings.
- The presence of neurologic or systemic signs, new headaches over age 50, or headaches in the setting of malignancy or immunosuppression, always require further evaluation with advanced imaging, and are considered "red flags," due to the higher likelihood of intracranial pathology. CT head in the presence of red flags is helpful to exclude intracranial hemorrhage. However, MRI brain has higher contrast resolution than CT head and is preferred for evaluation of structural pathologies, particularly in non-urgent settings.
- Subarachnoid hemorrhage due to ruptured cerebral aneurysm accounts for 4-12% of acute severe headaches. CT head is indicated as initial imaging for thunderclap headache. CT head had a negative predictive value between 99.9-100% in detecting aneurysmal subarachnoid hemorrhage within 6 hours of headache onset. The sensitivity is over 90% when CT head is performed within the first 24 hours. CT Angiography (CTA) head obtained concurrently or in follow up may identify cerebral aneurysm, dissection, and reversible cerebral vasoconstriction syndrome.
- In selected cases, CT head is supported for evaluation and follow up of headache caused by subdural or epidural hemorrhage, skull fracture, sinus infection or subarachnoid hemorrhage.
- New headaches in the setting of pregnancy and the postpartum period require special consideration.
- Over a third of pregnant women presenting to the hospital with headache have a secondary cause. Of patients with headache in the immediate post-partum period, 41% had an abnormal MRI brain.

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- Imaging in this scenario includes MRI brain, MR Venogram (MRV) head, and/or MR Angiography (MRA) head. Gadolinium contrast is relatively contraindicated during pregnancy and should be avoided.
- Trigeminal autonomic cephalgias, including cluster headaches, are required to have MRI brain to exclude pathology in the pituitary region. MRI should include the brain and the pituitary region.
- Headaches concerning for raised intracranial pressure or intracranial hypotension, required additional evaluation with neuroimaging. To exclude hydrocephalus, a mass, or cerebral venous sinus thrombosis, MRI brain, orbits and venogram are indicated in the setting of papilledema and/or intracranial hypertension. In urgent cases, a CT head can rapidly diagnose causes such as mass, edema or hydrocephalus.
- MRI is also useful to evaluate for structural causes of headache due to intracranial hypotension and csf leaks. Depending on the suspected source of the leak, imaging the brain and spinal cord may be required. Spinal imaging may include MRI of the spinal cord, or CT myelogram.
- CTA head or MRA head in the evaluation of headache are indicated for suspicion of carotid or vertebral arterial dissections, AVMs and cerebral aneurysm, as secondary causes of headache.
- CT Venogram (CTV) head or MRV head in the evaluation of headache are supported for suspicion of cerebral venous sinus thrombosis or stenosis in select cases, included suspected headache associated with pregnancy and the post-partum period, headache with papilledema, intracranial hypertension, and the trigeminal autonomic cephalgias.

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Aneurysm and AVM (HD-12)

Guideline

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Intracranial Aneurysms (HD-12.1)

HD.AN.0012.1.C v1.0.2025

 Creening 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 the individual. Autosomal dominant polycystic kidney disease Alpha-1-antitrypsin deficiency Azygos anterior cerebral artery (presence of) Coarctation of the aorta or bicuspid aortic valve Fibromuscular dysplasia (one screening study after confirmed diagnosis) Ehlers-Danlos Syndrome Type 4 (Vascular) Glucocorticoid-remediable aldosteronism (GRA)²⁵ Hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu Syndrome) See Screening for Vascular related genetic 		
 MRA Head (CPT® 70544, CPT® 70545, or CPT® 70546) MRA Head (CPT® 70546) 	Disorders and Indications (Any of the following)	Supported Imaging
See Screening for Vascular related genetic	 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 the individual. Autosomal dominant polycystic kidney disease Alpha-1-antitrypsin deficiency Alpha-glucosidase deficiency Azygos anterior cerebral artery (presence of) Coarctation of the aorta or bicuspid aortic valve Fibromuscular dysplasia (one screening study after confirmed diagnosis) Ehlers-Danlos Syndrome Type 4 (Vascular) Glucocorticoid-remediable aldosteronism (GRA)²⁵ Hereditary hemorrhagic telangiectasia (Osler- 	• MRA Head (CPT® 70544,
connective tissue Disorders (PVD-2.2)	· · ·	

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Disorders and Indications (Any of the following)	Supported Imaging
 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) - <i>CONTINUED</i>: Kawasaki disease Klinefelter syndrome Klippel-Trenaunay-Weber Syndrome Loeys-Dietz Syndrome Marfan Syndrome Microcephalic osteodysplastic primordial dwarfism Neurofibromatosis Type 1 Noonan Syndrome Pheochromocytoma Pseudoxanthoma elasticum Tuberculosis sclerosis 	 CTA Head CPT[®] 70496) OR MRA Head (CPT[®] 70544, CPT[®] 70545, or CPT[®] 70546)
See <u>Screening for Vascular related genetic</u> connective tissue Disorders (PVD-2.2)	
 New or worsening clinical symptoms or signs of cerebral aneurysm, including: Thunderclap headache (see <u>Sudden Onset of Headache (HD-11.3)</u>) 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 	 CTA Head (CPT[®] 70496) OR MRA Head (CPT[®] 70544, CPT[®] 70545, CPT[®] 70546) AND/OR MRI Brain without contrast (CPT[®] 70551) or MRI Brain without and with contrast (CPT[®] 70553)
Questionable or equivocal findings on an initial MRA Head	CTA Head (CPT® 70496)

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Disorders and Indications (Any of the following)	Supported Imaging
For suspected or confirmed cerebral aneurysm, ruptured or unruptured, for initial evaluation, treatment, intervention or follow up OR If initial catheter angiography is negative, repeat imaging is indicated. ²²	3D Rendering (CPT [®] 76377 or CPT [®] 76376) with cervicocerebra angiography/arteriography and/ or cerebral angiography ²² (See <u>General Guidelines-Other</u> <u>Imaging Situations (HD-1.7)</u>)
Follow up of known cerebral aneurysm: The optimal interval and duration for radiologic follow-up has not been determined. Radiographic follow-up for unruptured or treated intracranial aneurysms upon request by the neurosurgeon or team managing the intracranial aneurysm. ²²	 CTA Head (CPT[®] 70496) OR MRA Head (CPT[®] 70544, CPT[®] 70545, CPT[®] 70546) AND/OR MRI Brain without contrast (CPT[®] 70551) or MRI Brain without and with contrast (CPT[®] 70553)
 Additional physical characteristics of a known aneurysm: 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 	MRI Brain without contrast (CPT [®] 70551) or MRI Brain without and with contrast (CPT [®] 70553)
Follow up of cerebral aneurysm located in the vertebro-basilar circulation OR	MRA Neck (CPT [®] 70547, CPT [®] 70548, or CPT [®] 70549) OR CTA Neck (CPT [®] 70498)
If intracranial etiology of SAH has not been found	

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Disorders and Indications (Any of the following)	Supported Imaging
Subacute complications (i.e. vasospasm, delayed cerebral ischemia, and hydrocephalus), beginning days to weeks, arising from a subarachnoid hemorrhage and/or aneurysm treatment, upon request from the neurosurgeon and/or team managing the episode	CT Head OR MRI Brain contrast as requested
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	MRI Spine (Cervical without and with contrast CPT [®] 72156, AND / OR Thoracic without and with contrast CPT [®] 72157, AND/OR Lumbar without and with contrast CPT [®] 72158)
Catheter angiogram negative in SAH patient with remaining suspicion for cerebral aneurysm and these studies have not yet been performed	 CTA Head (CPT[®] 70496) AND/ OR MRA Head (CPT[®] 70544, CPT[®] 70545, or CPT[®] 70546)

- 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 vascular imaging indications for headaches require additional information. See <u>Stroke/TIA (HD-21.1), Sudden Onset of Headache (HD-11.3), New Headache</u> <u>Onset Older than Age 50 (HD-11.7), Abnormal Blood Clotting (HD-11.9),</u> <u>Pregnancy (HD-11.10), Physical Exertion (HD-11.11)</u>, and <u>Systemic Infections</u> (HD-11.13)

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

HD.AN.0012.2.A

v1.0.2025

Disorders and Indications (Any of the following)	Supported Imaging	
Any aneurysmal and/or AVM disorders listed in this guideline	 CT head without contrast (CPT[®] 70450) 	
 When MRI contraindicated²⁹ 	AND/OR	
Any urgent setting	CTA head (CPT [®] 70496)	
	AND/OR	
	• CTA Neck (CPT [®] 70498)	
Known AVM	MRI Brain without contrast (CPT [®] 70551)	
 When requested by a neurologist, neurosurgeon or any provider in 	OR	
consultation with a neurologist or neurosurgeon	 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	 Imaging as listed above in "known AVM" 	
 When requested by a neurologist, neurosurgeon or any provider in 	AND/OR	
consultation with a neurologist or neurosurgeon	 MRA Neck (CPT[®] 70547, CPT[®] 70548, OR CPT[®] 70549) 	
	OR	
	• CTA Neck (CPT [®] 70498)	
		1
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Disorders and Indications (Any of the following)	Supported Imaging
 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)
 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)

Head Imaging Guidelines

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Disorders and Indications (Any of the following)	Supported Imaging
Capillary Malformation-Arteriovenous Malformation (CM-AVM)	 MRI Brain without and with contrast (CPT[®] 70553)
 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 	 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)³¹

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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)

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Disorders and Indications (Any of the following)	Supported Imaging
Microcephalic Osteodysplastic Primordial Dwarfism, Type II (MOPD II) ¹⁹	 MRI Brain without contrast (CPT[®] 70551)
 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 	 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, OR CPT[®] 70549) OR CTA Neck (CPT[®] 70498)
 Sturge-Weber Syndrome At diagnosis Clinical signs or symptoms concerning for 	 MRI Brain without contrast (CPT[®] 70551) OR
 disease progression When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon 	 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}

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- For concerns related to stroke, see Stroke/TIA HD-21.1
- 3D imaging (CPT[®] 76376 or CPT[®] 76377) with MRI Brain without and with contrast (CPT[®] 70553) OR MRI Brain without contrast (CPT[®] 70551) is supported
- 3D Rendering (CPT[®] 76377 or CPT[®] 76376) 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 <u>General Guidelines Other Imaging Situations (HD-1.7)</u> 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 <u>Functional</u> <u>MRI (fMRI) (HD-24.2)</u>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}

Evidence Discussion (HD-12)

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- After the initial identification of a subarachnoid hemorrhage, the search for a ruptured cerebral aneurysm begins with imaging of the cerebral vessels with CT Angiopraphy (CTA), MR Angiography (MRA) or diagnostic cervico-cerebral catheter angiography.
- CTA head has over 90% sensitivity and specificity for the diagnosis of cerebral aneurysm in the setting of subarachnoid hemorrhage.
- MRA head has a sensitivity of 95% and a specificity of 89% for diagnosis of cerebral aneurysm.
- Diagnostic cervico-cerebral catheter angiography has the highest spatial and temporal resolution of any vascular imaging study, however, is invasive and requires use of contrast. It has a sensitivity and specificity over 98% for identification of aneurysm and can also diagnose vascular abnormalities in up to 13% with subarachnoid hemorrhage and a negative CTA. In addition, this modality can identify an aneurysm in 25% of previously negative studies and repeat studies are supported for this reason.
- In select cases of subarachnoid hemorrhage when an intracranial aneurysm is not identified, imaging the neck vessels and spinal vessels is appropriate.
- Aneurysm growth ranges from 4% to 14% on follow up imaging. For surveillance of incidentally identified cerebral aneurysms or ruptured and/or treated aneurysms, the less invasive modalities, CTA and/or MRA are supported, over the more invasive diagnostic cervico-cerebral angiography.
- Screening for cerebral aneurysms in high risk patient populations is also recommended with the less invasive modalities, CTA head or MRA head. This includes patients with autosomal dominant polycystic kidney disease (ADPKD), who had a prevalence of cerebral aneurysm ranging from 10-11.5%. The American Heart Association recommends screening those with at least 2 family members with cerebral aneurysm or subarachnoid hemorrhage. Screening is also recommended for conditions with known increased risk of cerebral aneurysm.
- Although vascular imaging is the primary focus of neuroimaging in the diagnosis and follow up of cerebral aneurysms, parenchymal imaging with MRI brain may be helpful in select clinical scenarios including giant aneurysms, posterior fossa aneurysms, in the setting of cranial neuropathies or focal neurologic findings and suspected stroke.
- Cervicocerebral angiography is the gold standard for imaging arteriovenous malformations (AVM) and arteriovenous fistulas (AVF). CT Angiography head (CTA) has a 90% sensitivity for the overall detection of AVMs and 100% for AVMs > 3 cm in size. In the evaluation of pulsatile tinnitus, CTA has a sensitivity of 86% with a specificity of 100% in identifying high flow AVFs. MR Angiography (MRA) is an alternative modality in these scenarios.

Head Imaging Guidelines

©2024 EviCore by EVERNORTH 400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 Page 126 of 326 www.EviCore.com • MRI brain for diagnosis and follow up of AVM has an overall sensitivity of 89% and 100% for lesions > 3 cm in size.

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Head and Facial Trauma (HD-13)

Guideline

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Head Trauma (HD-13.1)

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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 <u>Headache Associated with Head Trauma (HD-11.12)</u>)

For subacute head trauma (7 days to 3 months post-trauma)⁷ and chronic head trauma (greater than 3 months post-trauma) symptoms⁷

- 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, or CPT[®] 70546)
 - CT and MR Venography (CTV and MRV) are reported with the same codes as the CTA/MRA counterpart. If arterial and venous CT or MR studies are both performed in the same session, only one CPT[®] code should be used to report both procedures (see <u>General Guidelines - CT and MR Angiography (CTA and MRA)</u> (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 <u>Neck (Cervical Spine) Pain Without/With Neurological Features (Including</u> <u>Stenosis) and Trauma (SP-3.2)</u> 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}

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Head Imaging Guidelines

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.

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Facial Trauma (HD-13.2)

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- CT Maxillofacial without contrast (CPT[®] 70486) and/or CT Head without contrast (CPT[®] 70450) indicated for any concern regarding significant injury to facial structures including but not limited to:
 - Concern for orbital, maxillary, or mandibular fractures¹⁶
 - 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)
- MRI Maxillofacial without contrast (CPT[®] 70540) OR MRI Maxillofacial without and with contrast (CPT[®] 70543) for evaluation of cranial nerve deficits not explained or incompletely characterized on CT.¹⁶
- If concern for CSF leak and CT Maxillofacial or Temporal bone is inconclusive⁷, (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.

Evidence Discussion (HD-13)

v1.0.2025

- For evaluation of acute mild head trauma, less than 10% will have positive findings on CT head, and of this group, less than 1% will require neurosurgical intervention.
- The American College of Emergency Physicians and the Choosing Wisely Initiative recommend avoiding CT scans of the head in emergency department patients with minor head injury who are at low risk based on validated decision rules. This recommendation was based on the concern for patient exposure to ionizing radiation and the increased life time risk of cancer with such exposure.
- Selective CT scanning is recommended by validated clinical practice guidelines, including the New Orleans Criteria and the Canadian CT Head Rule. Both guidelines are 100% sensitive for mild head trauma requiring neurosurgical intervention. The New Orleans Criteria has a sensitivity > 97% for any traumatic finding on CT, with a specificity less than 6%. The Canadian CT Head Rule has a sensitivity between 83.4% 87.2% with a specificity between 37.2% 39.7%. The Canadian CT Head Rule has a 100% sensitivity and 29% specificity in cases of intracranial hemorrhage.
- When imaging is indicating by a validated clinical decision rule, CT head is the preferred imaging modality for evaluation of acute head trauma.
- If the initial CT head confirms subdural hematoma, follow up CT head is supported to monitor progression.
- For follow up in patients with persistent neurologic deficits without etiology identified on initial CT head, MRI brain is more sensitive, and can visualize cortical contusions, subdural hematomas, and white matter lesions in diffuse axonal injury. Up to 27% of patients with mild traumatic brain injury (TBI) with normal initial CT head show abnormalities on MRI brain.
- When vascular injury is suspected in the setting of head trauma, CT Angiography (CTA) head and neck is a non-invasive, rapid, and useful modality to evaluate for arterial injury. MR Angiography is an alternative option. For suspected intracranial venous injury, CT Venography (CTV) is indicated, with MR Venography (MRV) an alternative option.
- There is insufficient evidence to support the use of single-photon emission computed tomography (SPECT), FDG-PET/CT brain, CT/MRI-Perfusion, MR spectroscopy (MRS), functional MRI (fMRI), or diffusion tensor imaging (DTI) in the evaluation of head trauma.
- Patients with head trauma are also at risk for orbital, facial and temporal bone injuries. CT of the orbit can diagnose fractures, displaced fracture fragments, foreign bodies, traumatic hematoma, and extraocular muscle injury. CT head is also recommended in the evaluation of suspected orbital fractures due to concomitant intracranial injury incidence of 9%.

Head Imaging Guidelines

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- CT maxillofacial is useful in diagnosing maxillofacial injuries including non-displaced fractures. CT provides multiplanar and 3-D image reconstructions, allowing for better characterization of complex fractures, which is useful for surgical planning.
- Over one-third of patients with frontal sinus fractures are likely to have a concomitant intracranial injury, thus concurrent CT head imaging is recommended in patient with suspected frontal sinus fractures. In addition, between 8% to 10% of patients with frontal sinus fractures have subdural or epidural hematomas requiring surgical treatment.
- High resolution CT (HRCT) facial and temporal bone are sensitive modalities for subtle or non-displaced skull base defects, with sensitivity of 92% for identifying cerebrospinal fluid leak.

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

Guideline

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CNS and Head Infection (HD-14.1)

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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) OR MRI Brain without contrast (CPT[®] 70551) OR
 - CT Head (CPT[®] 70450, CPT[®] 70460, or CPT[®] 70470) in cases where MRI is contraindicated, in urgent scenarios, or prior to lumbar puncture, see <u>General</u> <u>Guidelines-CT Head (HD-1.4)</u>
 - If vascular involvement is suspected, in addition to MRI Brain, the following are supported²¹:
 - 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)

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 <u>General Guidelines CT Head (HD-1.4)</u> regarding additional indications for CT Head.
 - See <u>Skull Base Osteomyelitis (SBO) (HD-20.1)</u>, <u>Sinus and Facial Imaging</u> (HD-29.1), <u>Dental/Periodontal/Maxillofacial Imaging (HD-30.2)</u>, <u>Mental Status</u> <u>Change (HD-4.2)</u>, and <u>Eye Disorders and Visual Loss (HD-32.1)</u>

Evidence Discussion (HD-14.1)

- A head CT is recommended for quickly assessing intracranial infections in urgent cases. However, MRI is more effective for examining conditions that affect the cranial nerves, brain tissue, and meninges. Although MRI is superior in detecting minor changes in the brain associated with infections, CT scans can promptly identify pathophysiological changes that may influence the patient's prognosis.
- Acute bacterial meningitis often presents with a normal CT scan. However, due to the high mortality rate of up to 50% when left untreated, a CT scan is necessary to rule out other causes of encephalopathy or neurologic deficits. In certain clinical situations, CT is also required to exclude increased intracranial pressure before performing a lumbar puncture. Venous thromboses increase the risk of hemorrhage and are linked to high mortality. Consequently, both CT and MRI scans, including T1-weighted sequences, are recommended. These scans help detect high signal changes in several venous sinuses and can be complemented with CT- or MRvenography.
- Similar considerations apply when diagnosing non-bacterial central nervous system (CNS) infections. CT scans are advantageous due to quick access to care, faster diagnosis, and earlier treatment initiation, which can reduce morbidity. However, MRI is superior in detecting patterns of vasogenic versus cytotoxic edema, contrast enhancement, and the distribution of involvement, whether multifocal or unifocal/ unihemispheric. These distinctions are crucial in differentiating between differential diagnoses, such as systemic infections with hematogenous spread versus head/neck infections with a direct spread pattern.
- Many patients present with neurologic signs and symptoms that are indicative of either a cortical or subcortical syndrome. Often, these cannot be fully characterized by clinical presentation alone. As a result, radiologic evaluation becomes essential in diagnosing the etiology of the underlying process. The potential causes are varied and include meningoencephalitis, acute cerebrovascular disease, hemorrhagic necrotizing encephalopathy, immune-mediated (Bickerstaff) encephalitis, and demyelinating diseases such as acute disseminated encephalomyelitis (ADEM) and multiple sclerosis (MS). Therefore, prompt and accurate diagnosis is critical to select the most appropriate imaging method (MRI vs. CT) for each clinical scenario.

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

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- 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 <u>Stroke/TIA (HD-21.1)</u> for vascular imaging
 - See <u>Transverse Myelitis (HD-16.4)</u> 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).
- 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}

Head Imaging Guidelines

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-inducedimmunethrombotic-thrombocytopenia). (see Full EUA Prescribing Information).

Evidence Discussion (HD-14.2)

- A head CT is recommended for quickly assessing intracranial infections in urgent cases. However, MRI is more effective for examining conditions that affect the cranial nerves, brain tissue, and meninges. Although MRI is superior in detecting minor changes in the brain associated with infections, CT scans can promptly identify pathophysiological changes that may influence the patient's prognosis.
- Acute bacterial meningitis often presents with a normal CT scan. However, due to the high mortality rate of up to 50% when left untreated, a CT scan is necessary to rule out other causes of encephalopathy or neurologic deficits. In certain clinical situations, CT is also required to exclude increased intracranial pressure before performing a lumbar puncture. Venous thromboses increase the risk of hemorrhage and are linked to high mortality. Consequently, both CT and MRI scans, including T1-weighted sequences, are recommended. These scans help detect high signal changes in several venous sinuses and can be complemented with CT- or MRvenography.
- Similar considerations apply when diagnosing non-bacterial central nervous system (CNS) infections. CT scans are advantageous due to quick access to care, faster diagnosis, and earlier treatment initiation, which can reduce morbidity. However, MRI is superior in detecting patterns of vasogenic versus cytotoxic edema, contrast enhancement, and the distribution of involvement, whether multifocal or unifocal/ unihemispheric. These distinctions are crucial in differentiating between differential diagnoses, such as systemic infections with hematogenous spread versus head/neck infections with a direct spread pattern.
- Many patients present with neurologic signs and symptoms that are indicative of either a cortical or subcortical syndrome. Often, these cannot be fully characterized by clinical presentation alone. As a result, radiologic evaluation becomes essential

in diagnosing the etiology of the underlying process. The potential causes are varied and include meningoencephalitis, acute cerebrovascular disease, hemorrhagic necrotizing encephalopathy, immune-mediated (Bickerstaff) encephalitis, and demyelinating diseases such as acute disseminated encephalomyelitis (ADEM) and multiple sclerosis (MS). Therefore, prompt and accurate diagnosis is critical to select the most appropriate imaging method (MRI vs. CT) for each clinical scenario.

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

HD.HI.0014.3.A v1.0.2025

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 or in consultation with a neurologist, neurosurgeon, psychiatrist, oncologist, rheumatologist, or infectious disease specialist.²⁶

Initial Imaging²⁶:

- MRI Brain without and with contrast (CPT[®] 70553) 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 unavailable or contraindicated or for bony pathology concerns
- 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 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 to²⁶:

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

Head Imaging Guidelines

©2024 EviCore by EVERNORTH 400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 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 Information²⁶

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

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- 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 hypometabolism 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)

Evidence Discussion (HD-14.3)

 The American College of Radiology (ACR) Appropriateness Criteria® provides guidance on the appropriateness of CT versus MRI Brain in certain clinical scenarios relating to altered mental status. For acute, undifferentiated presentations and with focal symptoms, CT head is an appropriate modality, but with known intracranial process, suspected medical illness or toxic-metabolic etiology, and/or psychosis, MRI Brain, is appropriate and in some cases may be preferable.

Head Imaging Guidelines

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- In the appropriate clinical scenarios, recognition of structural and functional imaging patterns of brain involvement using CT head, MRI brain, and brain PET, in autoimmune encephalitis (including paraneoplastic and non-rheumatologic inflammatory disorders) can facilitate rapid access to appropriate treatment, as well as avoid invasive diagnostic procedures such as brain biopsy.
- FDG-PET/CT brain performed at a median 4 weeks of symptom onset was more often abnormal than initial MRI, EEG, or laboratory cerebrospinal fluid testing, in patients with suspected autoimmune encephalitis (AE), with focal hypometabolism, the most common PET/CT finding.
- Spine MRI shows abnormalities in up to 45% of cases of paraneoplastic myeloneuropathy.
- The three-dimensional (3D) constructive interference in steady state (CISS) is a gradient-echo MRI or Fast Imaging Employing Steady-state Acquisition Cycled Phases (FIESTA-C) on GE MRI systems are widely employed for over a decade and have been shown to have utility in demonstration of contrast between cerebrospinal fluid and brain parenchymal structure. Therefore, these have particular utility in the examination of cranial nerves, the ventricular system, cavernous sinus, and other structures which are commonly involved in neuro-inflammatory conditions.
- Neuropsychiatric lupus and other neuro-inflammatory conditions have been described to mimic vascular disease, such as vasculitis and small vessel cerebrovascular disease, and in these cases vessel imaging with CT Angiography (CTA) and MR Angiography (MRA) Brain can contribute to meaningful diagnosis.
- In the evaluation of a first episode of psychosis when an autoimmune cause is suspected, up to 4% of cases have abnormalities on MRI brain.
- Sarcoidosis can manifest with neurologic complications in every part of the neural axis, with diagnostic challenges represented by multiple pathophysiologic pathways and frequently lack of specific histopathologic diagnosis. In clinically suspected neurosarcodoisis on the basis of synthesized clinical history and physical examination findings, the demonstration of neuro-inflammation using cerebrospinal fluid testing and contrast-enhanced MRI is useful. In difficult or complicated cases, FDG-PET and Gallium-67 imaging have been useful for identification of targets for biopsy.

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Movement Disorders (HD-15)

Guideline

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Movement Disorders (HD-15.1)

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- The majority of movement disorders are diagnosed based on a clinical diagnosis and do not require imaging. These include:
 - Typical Parkinson's Disease
 - Essential Tremor or tremors of anxiety or weakness
 - **Restless Leg Syndrome**
 - Tics or spasms which can be duplicated at will 0
 - Tourette syndrome²²
 - Tardive dyskinesia^{19,20,21}
- 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
 - Anti-psychotic drug-induced Parkinsonism or Atypical Tardive dyskinesia^{19,20,21} 0
 - 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

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Page 153 of 326 www.EviCore.com 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 <u>Post-Operative Imaging (HD-28.3)</u> indications
- MRI Brain with and without (CPT[®] 70553) for initial imaging for suspected motor neuron disease, see <u>Motor Neuron Disease/Amyotrophic Lateral Sclerosis (ALS)</u> (PND-8.1) in the Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines
- Dementia associated with movement disorder, see <u>Lewy Body Dementia (LBD) –</u> <u>SPECT Brain Scan (HD-8.3)</u>

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 <u>Lewy</u> <u>Body Dementia (LBD) - SPECT Brain Scan (HD-8.3)</u>)

Evidence Discussion (HD-15)

- The majority of movements disorders are diagnosed based on history and clinical examination findings and do not require imaging. For cases of diagnostic uncertainty, incomplete response to medication, for atypical Parkinsonism or drug-induced parkinsonism, and for suspected Huntington disease, MRI brain is the preferred imaging modality.
- Structural imaging with MRI Brain is usually normal in patients with Parkinson's disease but is useful to diagnose causes of secondary parkinsonism, such as stroke, iron deposition, normal pressure hydrocephalus, and neoplasm.
- CT head is not preferred due to its limited soft-tissue characterization when compared to MRI.
- Functional imaging studies assessing dopaminergic function in Parkinson's disease include single-photon emission computed tomography (SPECT) and positron emission tomography (PET) imaging.1,2,3 These studies are used as an adjunct diagnostic test.

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- Both Dopamine Transporter single-photon emission computed tomography (DAT-SPECT) and [18F]-fluorodopa (F-DOPA) PET brain are useful to differentiate suspected Parkinsonian syndromes from non-neurodegenerative disorders such as Essential Tremor, drug-induced tremors, vascular parkinsonism, and/or psychogenic tremors.
- DAT-SPECT has a sensitivity of 91% with a specificity of 100% for Essential Tremor, a sensitivity of 86.2% with a specificity of 93.8% for drug-induced parkinsonism, and a sensitivity of 86.2% with a specificity of 82.9% for vascular parkinsonism.
- F-DOPA PET brain has a sensitivity of 73% with a specificity of 91% for evaluation of parkinsonian syndrome vs non-neurodegenerative parkinsonian syndrome.
- Neither DAT-SPECT scans nor F-DOPA PET brain scans are useful for the differentiation between subtypes of Parkinsonian syndromes, to monitor progression of disease nor to predict the risk of development of disease.
- There is insufficient evidence for the routine use of FDG-PET brain in the diagnosis of Parkinsonian syndromes.

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

Guideline

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Multiple Sclerosis (MS) (HD-16.1)

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Establishing a New Diagnosis of Multiple Sclerosis

Repeat Imaging for Unclear Diagnosis

New Neurologic Symptoms in an Individual with Multiple Sclerosis

Baseline Imaging with Disease Modifying Therapy (DMT)

Current Treatment with High Risk Disease Modifying Therapy (DMT)

Annual Imaging on Low Risk DMT or No Treatment

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

Prolonged Treatment with Tysabri (natalizumab)

Progressive Multifocal Leukoencephalopathy (PML) Evaluation

Background and Supporting Information

- 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.
- Computed Tomography (CT) scans of the head and/or spine are NOT indicated for the evaluation of multiple sclerosis due to inferior soft tissue resolution when compared to MRI.⁴⁷
- 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[®] 76376 and CPT[®] 76377).¹
- Quantitative Magnetic Resonance Image (MRI) Analysis of the Brain
 - Volumetric or quantitative analysis of the brain or temporal lobes and hippocampus may be ordered as Quantitative MRI Analysis of the Brain (CPT[®] 0865T or CPT[®] 0866T) or 3D rendering (CPT[®] 76376 and CPT[®] 76377).
 - 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.

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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 OR 	 MRI Brain without and with contrast (CPT[®] 70553) 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)
 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 hours⁴³ Baseline exclusion of appropriate alternative conditions that can mimic MS OR Radiologically Isolated Syndrome (RIS)* based on ALL of the following: Individual with brain MRI obtained for unrelated reason with findings conspicuous for demyelinating disease⁴¹ 	 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)

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Indication	Supported Imaging
 Baseline exclusion of appropriate alternative conditions that can mimic MS 	
*For more information about CIS and RIS, see Background and Supporting Information	

Repeat Imaging for Unclear Diagnosis

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

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New Neurologic Symptoms in an Individual with Multiple Sclerosis

Indication	Supported Imaging
New neurologic signs or symptoms in an individual with Multiple Sclerosis and/or concern for a possible diagnosis of	 MRI Brain without contrast (CPT[®] 70551) OR MRI Brain without and with contrast (CPT[®] 70553)
demyelinating disease	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 <u>Eye Disorders and Visual Loss</u> (<u>HD-32.1)</u>
	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)

Head Imaging Guidelines

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Baseline Imaging with Disease Modifying Therapy (DMT)

Indication	Supported Imaging
 Before starting OR changing disease modifying therapy (DMT)¹ 	 MRI Brain without contrast (CPT[®] 70551) OR MRI Brain without and with contrast (CPT[®] 70553)
AND/OR	
 3-6 months after starting or changing DMT to establish a new MRI treatment baseline 	
AND/OR	
 If there are new abnormal MRI Brain findings without clinical symptoms, an additional follow up MRI Brain is supported after 6 months¹ 	

Current Treatment with High Risk Disease Modifying Therapy (DMT)

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

Head Imaging Guidelines

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Annual Imaging on Low Risk DMT or No Treatment

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

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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	 MRI Brain without contrast (CPT[®] 70551) OR MRI Brain without and with contrast (CPT[®] 70553)
Patient with history of Radiologically Isolated Syndrome* (RIS) ¹	If there are new or worsening symptoms concerning for spinal cord involvement, the following imaging is ALSO indicated: ¹
*For more information about CIS or RIS, 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)

Head Imaging Guidelines

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Prolonged 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)

Progressive Multifocal Leukoencephalopathy (PML) Evaluation

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

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 event suggestive of central nervous system (CNS) demyelination with

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evidence of separation of space and time on MRI as well as reasonably excluding other possible conditions that could account for the clinical and imaging findings.^{1,45}

- Multiple sclerosis commonly begins with a relapsing-remitting course with partial or complete neurologic recovery following acute events.
 - An acute demyelinating event lasts at least 24 hours or longer
 - Common types of MS relapses include:
 - Unilateral optic neuritis
 - Brainstem or cerebellar syndrome (i.e. trigeminal neuralgia, diplopia or intranuclear ophthalmoplegia (INO), and/or ataxia)
 - Partial transverse myelitis
 - Individuals with multiple sclerosis are most often diagnosed during their twenties or thirties.
 - Females are more frequently diagnosed with multiple sclerosis compared to males.
- The first event concerning for demyelinating disease without meeting criteria for separation of time is known as a clinically isolated syndrome (CIS).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}
- Progressive Multifocal Leukoencephalopathy (PML) is a progressive multi-focal disease of the central nervous system that can occur in individuals treated with immunosuppressive or immunomodulatory medications.⁴⁶
 - In individuals treated with natalizumab, there is an increased risk of developing PML in individuals who:
 - Received prior immunosuppressive medication, and/or
 - Have a high JC virus antibody index, and/or
 - Received natalizumab for ≥ 18 months¹
- 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}
- 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 associated with a high risk of PML and/or other CNS opportunistic infections (i.e. herpes encephalitis, cryptococcal meningitis) include (but are not limited to): Tysabri[®] (natalizumab), 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)

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Neuromyelitis Optica Spectrum Disorders (HD-16.2)

HD.MS.0016.2.A

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

Indication	Supported Imaging
Clinical concern for optic neuritis	 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	 MRI Brain without and with contrast (CPT[®] 70553) OR MRI Brain without contrast (CPT[®] 70551)

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Indication	Supported Imaging
Clinical concern for transverse myelitis	 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)

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Indication	Supported Imaging
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[®] 72141)

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

Indication	Supported Imaging
Clinical concern for optic neuritis	 MRI Orbit without and with contrast (CPT[®] 70543) OR MRI Orbit without contrast (CPT[®] 70540)
New neurologic signs or symptoms concerning for brain involvement	 MRI Brain without and with contrast (CPT[®] 70553) OR MRI Brain without contrast (CPT[®] 70551)

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Indication	Supported Imaging		
Clinical concern for transverse myelitis	 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 	 MRI Brain without and with contrast (CPT[®] 70553) OR MRI Brain without contrast (CPT[®] 70551) AND/OR 		
 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 or ophthalmologist or ophthalmologist 	 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) 		

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- For Neuromyelitis Optica Spectrum Disorder with concern for occult neoplasm, see **Paraneoplastic Syndromes (ONC-30.3)** in the Oncology Imaging Guidelines.⁵³
- Computed Tomography (CT) scans of the head and/or spine are not recommended for the evaluation of NMOSD due to inferior soft tissue resolution when compared to MRI.⁴⁷
- Quantitative Magnetic Resonance Image (MRI) Analysis of the Brain
 - Volumetric or quantitative analysis of the brain or temporal lobes and hippocampus may be ordered as Quantitative Analysis of the Brain (CPT[®] 0865T or CPT[®] 0866T) or 3D rendering (CPT[®] 76376 and CPT[®] 76377).
 - 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 episodes in patients with NMOSD.
 - Even after a single event, severe permanent disability can occur, especially if 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
- Core clinical characteristics of NMOSD include⁷
 - Optic neuritis
 - Frequently bilateral optic nerve involvement with severe vision loss
 - Longitudinally extensive transverse myelitis
 - Extends \geq 3 complete vertebral segments 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 lesions⁷
 - Rarely paraneoplastic syndromes occur with NMO spectrum disorder
 - Medications used for the treatment of NMO spectrum disorders include (but are not limited to) azathioprine, Enspryng[®] (satralizumab), mycophenolate, Soliris[®] (eculizumab), rituximab³⁷, Uplizna[®] (inebilizumab) and Ultomiris[®] (ravulizumab)⁵⁴
 - Possible adverse reactions associated with treatment include risk of PML and meningococcal infections.

©2024 EviCore by EVERNORTH 400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 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

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

HD.MS.0016.3.A v1.0.2025

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

Indication	Supported Imaging
Clinical concern for optic neuritis	 MRI Orbit without and with contrast (CPT[®] 70543) OR MRI Orbit without contrast (CPT[®] 70540)
Neurologic signs or symptoms concerning for brain involvement	 MRI Brain without and with contrast (CPT[®] 70553) OR MRI Brain without contrast (CPT[®] 70551)
Clinical concern for transverse myelitis	 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)

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

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

Indication	Supported Imaging
Clinical concern for optic neuritis	 MRI Orbit without and with contrast (CPT[®] 70543) OR MRI Orbit without contrast (CPT[®] 70540)
Neurologic signs or symptoms concerning for brain involvement	 MRI Brain without and with contrast (CPT[®] 70553) OR MRI Brain without contrast (CPT[®] 70551)

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Indication	Supported Imaging
Clinical concern for transverse myelitis	 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:	 MRI Brain without and with contrast (CPT[®] 70553) OR
 Re-establish baseline after starting treatment (typically 3-6 	• MRI Brain without contrast (CPT [®] 70551)
months after last MRI)	AND/OR
 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 or ophthalmologist³⁴ 	 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/OR MRI Thoracic Spine without and with contrast (CPT[®] 72157)
	OR
	 MRI Cervical Spine without contrast (CPT[®] 72141) AND/OR MRI Thoracic Spine without contrast (CPT[®] 72146)

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- For MOG (myelin oligodendrocyte glycoprotein) Antibody-Associated Disease with concern for occult neoplasm, see <u>Paraneoplastic Syndromes (ONC-30.3)</u> in the Oncology Imaging Guidelines.⁵²
- Computed Tomography (CT) scans of the head and/or spine are not recommended for the evaluation of MOG (myelin oligodendrocyte glycoprotein) Antibody-Associated Disease due to inferior soft tissue resolution when compared to MRI.⁴⁷

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
 - Relapses are more common in the first six months after the first episode.
 - An acute relapse is considered when an individual with MOGAD develops new neurologic signs or symptoms at least 30 days following the previous event.
- Diagnosis is based on the clinical presentation, MRI findings, and the presence of auto-antibodies.
- Clinical features of individuals with MOGAD include³⁴
 - 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 improves quickly with return to normal or near normal visual acuity following treatment with intravenous corticosteroids.³⁴
 - Transverse myelitis
 - Cauda equina 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 MRI³⁴
 - Most T2 lesions resolve or reduce in size substantially on follow up MRI
 - Brainstem encephalitis
 - Encephalitis with seizures⁴⁵
 - Acute disseminated encephalomyelitis (ADEM)
 - Occurs mainly in children but can occur in adults.
 - Tumefactive brain lesions
 - Cranial neuropathies

©2024 EviCore by EVERNORTH 400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 Unlike multiple sclerosis, it is rare for individuals with MOGAD to develop asymptomatic lesions within the brain, optic nerves and/or spinal cord.³⁴

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

Clinical Concern	for T	Fransverse	Myelitis
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Indication	Supported Imaging
Clinical concern for transverse myelitis	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)

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New Neurologic Signs or Symptoms

Indication	Supported Imaging
New neurologic signs or symptoms	 MRI Cervical Spine without and with contrast (CPT[®] 72156) OR
Symptoms	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)

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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)

- For transverse myelitis with concern for occult neoplasm, see <u>Paraneoplastic</u> <u>Syndromes (ONC-30.3</u>) in the Oncology Imaging Guidelines.²²
- 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 process⁴²

 See <u>Multiple Sclerosis (MS) (HD-16.1)</u>, <u>Neuromyelitis Optica Spectrum</u> 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

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- 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 <u>Neuromyelitis Optica Spectrum Disorders (HD-16.2)</u>)
 - Initial event of MOG antibody-associated disease (MOGAD) (see <u>MOG Antibody-Associated Diseases (MOGAD) (HD-16.3)</u>)
 - May be associated with connective tissue disease
 - Systemic lupus erythematous (SLE)
 - Rheumatoid Arthritis (RA)
 - Sjögren's syndrome
 - Systemic sclerosis
 - Manifestation of neurosarcoidosis (see <u>Autoimmune/Paraneoplastic</u> <u>Encephalitis & Neuroinflammatory Disorders (HD-14.3)</u>)
 - Post-infectious and/or post-vaccination related
 - COVID-19 and COVID-19 post-vaccination myelitis cases have been reported (see <u>Neuro-COVID-19 and Sars-COV-2 Vaccines (HD-14.2)</u>)
 - May have a prodromal syndrome with fever, respiratory and/or gastrointestinal symptoms⁴⁰
 - May be associated with headache, neck stiffness or recurrence of fever⁴⁰

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Evidence Discussion (HD-16)

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- Magnetic resonance imaging (MRI) is the recommended imaging modality for the diagnosis and monitoring of multiple sclerosis (MS) and other inflammatory diseases of the central nervous system (CNS). Its high sensitivity for the evaluation of inflammatory and neurodegenerative processes in the brain and spinal cord has made it the gold standard for the evaluation of patients with MS.
- Computed Tomography Scan (CT) is not recommended for the evaluation of MS due to inferior soft tissue resolution when compared to MRI.
- MRI plays an important role in the following clinical scenarios:
 - establishing the diagnosis of multiple sclerosis (MS) by establishing evidence for dissemination in space and time.
 - diagnostic workup. Approximately 50-90% of patients with MS have spinal cord lesions.
 - detecting optic nerve abnormalities in patients with symptoms concerning for optic neuritis.
 - assessment of treatment response and in monitoring for potential treatment related safety concerns. Management and surveillance intervals are primarily consensus based and have been addressed in several evidence and practice based guidelines.
 - evaluation of other central nervous system (CNS) inflammatory diseases, including autoimmune disseminated encephalomyelitis (ADEM), neuromyelitis optica (NMO), myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) and transverse myelitis (TM).
 - detecting conus medullaris involvement in patients with myelin oligodendrocyte glycoprotein-associated disease (MOGAD) and transverse myelitis.

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Papilledema/Pseudotumor Cerebri (HD-17)

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Papilledema/Pseudotumor Cerebri (HD-17.1)

HD.PP.0017.1.A v1.0.2025

- See Eye Disorders and Visual Loss (HD-32.1)
- Papilledema and Pseudotumor Cerebri (Idiopathic Intracranial Hypertension, Benign 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 <u>General Guidelines CT Head (HD-1.4)</u> regarding required use of CT Head prior to lumbar puncture and/or spinal tap.
 - See <u>Eye Disorders and Visual Loss (HD-32.1)</u> 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 <u>Hydrocephalus Shunts (HD-11.14)</u>)
 - 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)

Evidence Discussion (HD-17)

- In the evaluation of suspected or known intracranial hypertension and/or exam findings of papilledema, neuroimaging is helpful for diagnosis, excluding other structural causes, and for the identification of venous outflow obstruction.
- MRI Brain allows detection of findings supportive of intracranial hypertension and detection of structural abnormalities such as mass, edema, or hydrocephalus. MR

Venogram allows identification of venous sinus stenosis and thrombosis for treatment planning in these scenarios.

- Alternatively, CT Head allows exclusion of secondary causes such as hydrocephalus, mass or edema, particularly in urgent scenarios.CT Venogram allows direct vessel visualization to exclude venous outflow obstruction in these cases.
- In addition, orbital symptoms may be evaluated with either CT Orbits or MRI Orbits, with CT providing superior bony anatomy evaluation and calcification detection and MRI providing superior soft tissue resolution and evaluation of the optic nerve

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Paresthesias and/ or Weakness (HD-18)

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Sensory/Weakness Complaints (HD-18.1)

HD.PS.0018.1.A v1.0.2025

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:

RI Brain without contrast (CPT [®] 551) OR RI Brain without and with contrast PT [®] 70553) OR
Al Cervical Spine without contrast PT [®] 72141) OR Al Cervical Spine without and with htrast (CPT [®] 72156) OR Al Thoracic Spine without contrast PT [®] 72146) OR Al Thoracic Spine without and with htrast (CPT [®] 72157)
-

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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 torso⁴ 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
	MRI Lumbar Spine without contrast

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- 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 <u>Head Trauma (HD-13.1)</u> 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 <u>Muscle Diseases (PN-8.5)</u> and <u>Gaucher Disease</u> (Storage Disorders) (PN-8.6)
 - Motor Neuron Disease or Amyotrophic Lateral Sclerosis (ALS), see <u>Motor Neuron</u> <u>Disease/Amyotrophic Lateral Sclerosis (ALS) (PN-8.1)</u>
 - Neuromuscular Junction Disorders, see <u>Neuromuscular Junction Disorders</u> (PN-8.4)
 - Multifocal Motor Neuropathy (MMN) and Chronic Inflammatory Demyelinating Polyneuropathy (CIDP), see <u>Polyneuropathy (PN-3.1)</u>
 - Polyneuropathy, see Polyneuropathy (PN-3.1)
 - Neuropathy with concern for malignancy, see <u>Paraneoplastic Syndromes</u> (<u>ONC-30.3</u>) in the Oncology Imaging Guidelines
 - Proximal asymmetric and concern for plexopathy, see <u>Brachial Plexus (PN-4.1)</u> and/or <u>Lumbar and Lumbosacral Plexus (PN-5.1)</u>
 - Sensory and/or motor symptoms localized to a single nerve, see <u>Focal</u> <u>Neuropathy (PN-2.1)</u>
 - Thoracic Outlet Syndrome, see <u>Thoracic Outlet Syndrome (CH-31.1)</u> in the Chest Imaging Guidelines
 - Radiculopathy, see appropriate level in the Spine Imaging Guidelines
 - Cauda Equina Syndrome, see <u>Red Flag Indications (SP-1.2)</u> 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

©2024 EviCore by EVERNORTH 400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 Page 195 of 326 www.EviCore.com 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 and Neuromuscular Disorders 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 <u>Polyneuropathy (PN-3.1)</u> in the Peripheral Nerve and Neuromuscular Disorders (PNND) 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

Evidence Discussion (HD-18)

- The imaging modality of choice for the evaluation of signs or symptoms localizing to the spinal cord is with MRI. MRI allows visualization of the soft tissues and structures that comprise the neural axis. Imaging of the cervical and thoracic segments are sufficient to view the entire spinal cord.
- MRI of the lumbar spine is reserved for the evaluation of conus and the cauda equina.
- For evaluation of isolated distal symmetric polyneuropathy, MRI of the brain and/ or spine rarely change management in these patients despite being frequently performed. MRI has little role in these scenarios as it evaluates the central nervous system.

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Pituitary (HD-19)

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Head Imaging Guidelines

Pituitary (HD-19.1)

HD.PT.0019.1.A

- v1.0.2025
- 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 <u>General Guidelines Anatomic Issues (HD-1.1)</u> 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 <u>Secondary Amenorrhea (PV-3.1)</u> 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.

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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

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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.

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Indication	Supported 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: 	
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 discontinuation²² 	

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Indication	Supported 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)

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Indication	Supported 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 Glucagor 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 TSI 	
Hypopituitarism (deficiency of one or more	MRI Brain without and with contrast (CPT [®] 70553) o MRI Brain without contrast (CPT [®] 70551)	
pituitary hormones)		

Head Imaging Guidelines

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Indication	Initial Imaging	Repeat Imaging for Non-Operative Care
Diabetes Insipidus (DI) - ADH deficiency	 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 hyperostosis. 	MRI to evaluate for

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

©2024 EviCore by EVERNORTH 400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 Page 205 of 326 www.EviCore.com 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

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

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- For imaging in the immediate post-operative period or for acute surgical complications
 See <u>Primary Central Nervous System Tumors (ONC-2.1)</u> 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.

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Empty Sella Turcica (HD-19.3)

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

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

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 See <u>Craniopharyngioma and Other Hypothalamic/Pituitary Region Tumors</u> (PEDONC-4.10)

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Evidence Discussion (HD-19)

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- MRI imaging of the Sella region using high-resolution pituitary protocols is the preferred diagnostic imaging modality for evaluation of the pituitary and sellar regions and is considered the gold standard for imaging the pituitary gland when there is suspicion of hypothalamic pituitary disease.
- Both the anatomy and pathology of the pituitary gland and surrounding areas including optic chiasm, infundibulum and vascular structures, as well as an empty sella, are reliably depicted on MRI.
- MRI is the most sensitive imaging study for evaluating pituitary disease.
- CT of the Sella can be used to detect bone destructive lesions of the skull base, such as craniopharyngiomas, menigiomas, or larger pituitary macroadenomas, but CT is insensitive when compared to MRI for pituitary pathology.
- MRI utilizes a magnetic field and radio waves with computer processing to produce detailed images whereas CT uses ionizing radiation. Radiation dosages vary based on many factors and can be harmful to tissues. Thus, from radiation safety perspective MRI should be utilized when appropriate and supported by existing literature.

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Scalp and Skull (HD-20)

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Scalp and Skull Lesions (HD-20.1)

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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 the initial imaging of scalp lesions⁶
- X-ray is the initial imaging of skull (bony) lesions⁶
- 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.
 - When bony lesions are detected on physical examination with any of the following:⁶
 - Signs or symptoms of Langerhan's cell histiocytosis
 - Signs or symptoms of multiple myeloma
 - History of a cancer condition with a suspicion of metastasis
 - History of Paget's disease
 - History of radiation therapy to the head region
- MRI Brain without contrast (CPT[®] 70551) or MRI Brain 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 surgeon (OMS) or any provider coordinating care with a neurologist, neurosurgeon, otolaryngologist (ENT) and/or oromaxillofacial surgeon (OMS)

Background and Supporting Information

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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

Evidence Discussion (HD-20.1)

- The ACR Practice Parameter or the Performance of Computed Tomography (CT) of the head and of the extracranial head and neck gives a broad description of some of the pathologies that would be beneficially imaged using this modality, and points out that the reason for imaging must be for a valid medical reason and should be done with the aim for using only the minimum necessary radiation. This in some cases requires the use of additional modalities.
- The majority of skull lesions is benign, but advanced imaging characteristics may aid in defining the lesion as having a relatively high pretest probability of malignancy (prior to histological confirmation). However, clinical contextual information is necessary to help decide which patients would benefit from advanced imaging including the patient's age and features of the patient's presenting history.
- Ultrasound offers many radiographic advantages for the characterization of scalp masses, which are not visualized by CT or MRI, the primary goal being to differentiate benign vs. malignant scalp masses. Cancers of the scalp represent 2% of all skin cancers. In both squamous and basal cell carcinomas, the US shows hypoechoic solid tumors with increased vascularity, and basal cell pathology can also consist of hyperechoic spots internally.
- In folliculotropic mycosis fungoides (FMF), the most common manifestation of cutaneous T-cell lymphoma, the sonographic features include skin thickening, and hypoechoic upper dermis and hair follicles, with large surrounding hyperechoic deposits.
- Skull (bony) lesions are most often discovered incidentally either clinically or as a result of CT or MRI of the brain performed for another indication, and these skull masses can be either malignant or benign. The patient history is essential to understand along with the imaging characteristics in order to obtain accurate diagnosis. Radiographic features, both CT- and MRI- specific can be used to differentiate between benign and malignant lesions, identifying whether lesions have well-defined borders, sclerotic margins and a narrow transition zone.
- The presence of bony destruction is a useful observation, periosteal reaction, soft tissue component, and intracranial or extracranial extension can be identified as malignant features in addition, and patterns such as lytic vs. sclerotic, dingle vs multiple, homogeneous vs varied composition also can give helpful information for diagnosis, and various patterns are recognizable that may support the tissue type of origin such as fibrogenic, chondrogenic, osteogenic, vascular, etc.
- Plain radiograph can identify some of these features as a first diagnostic study, but advanced imaging may be necessary, in conjunction with x-ray, and at times CT and MRI are useful as complementary studies.
- Pott puffy tumor is a rare complication of sinusitis or trauma, and early diagnosis is important since it is treatable with broad spectrum antibiotics, therefore advanced imaging is indicated with a clinical suspicion.

Skull Base Osteomyelitis (SBO) (HD-20.2)

HD.SK.0020.2.A

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- 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 (CPT[®] 78800 or 78801, and 78803, 78831, 78830 or 78832)²
 - Bone Scan (CPT[®] 78830 or 78832)²
 - Skull base osteomyelitis: + Gallium and + Bone scan
 - Necrotizing otitis externa: + Gallium and Bone scan
 - Indium WBC (CPT[®] 78800 or 78801, and 78803, 78831, 78830 or 78832) 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.²
- Treatment response: Gallium-67 Scan every 4-6 weeks till scan is negative²
- Surveillance Scanning: Gallium-67 Scan at 4 weeks and 3 months post-treatment²

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

Evidence Discussion (HD-20.2)

- CT, although involves radiation, is more readily available and provides superior information regarding bony erosion and/or demineralization in the patient with suspected osteomyelitis.
- MRI can assist with early detection of bone changes in as early as 3-5 days from onset of osteomyelitis. In cases of diabetic osteomyelitis MRI provides a sensitivity of

90% and a specificity of 79%. MRI provides superior soft tissue detail and intracranial involvement secondary to its superior resolution when compared to CT for evaluation of skull based osteomyelitis.

Both nuclear imaging by means of Technetium 99m (^{99m}Tc) and Gallium 67 (⁶⁷Ga) scan can assist in localizing infection. The Gallium scan is often used to determine the resolution of the infection and thus the end of antibiotic therapy. The Technetium 99m scan can be useful for detecting the infection however often times remains positive prolonged period of time, and thus should not be used to determine resolution of infection.

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Stroke/TIA (HD-21.1)

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Indications	Supported Imaging
 Acute ischemic stroke (within the first 24 hours) Transient ischemic attacks (TIA) Suspected Hemorrhagic stroke Suspected 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 	 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
Venous vascular imaging for evaluation of venous infarcts	 CTA Neck (CPT[®] 70498) MR or CT Venography (MRA Head [CPT[®] 70544, CPT[®] 70545, or CPT[®] 70546] OR CTA Head (CPT[®] 70496)
Cerebral Angiography for stroke evaluation	

Head Imaging Guidelines

Indications	Supported Imaging
 Stroke in Pregnancy and other hypercoagulable states⁴³ 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 	imaging options and vascular imaging
Repeat imaging for follow up and resolution of stroke or hemorrhage	As requested by a neurologist, neurosurgeon, hematologist or physiatrist (PM&R) or any provider in consultation with a neurologist, neurosurgeon, hematologist, or physiatrist
Reversible Cerebral Vasoconstriction Syndrome	See <u>Sudden Onset of Headache</u> (HD-11.3)
Neurologic signs and/or symptoms, including headaches, associated with COVID-19 infection and/or COVID-19 vaccination	 MRI Brain without contrast (CPT[®] 70551) OR MRI Brain without and with contrast (CPT[®] 70553)
(Strokes may be arterial or venous)	See also <u>General Guidelines-CT head</u> (HD-1.4), Abnormal Blood Clotting (HD-11.9) and <u>Neuro-Covid-19 (HD-14.2)</u>
Adults with HbSS (Sickle cell disease) or HbSb Thalassemia	One time MRI Brain without contrast (CPT [®] 70551) or MRI Brain 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)

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Indications	Supported Imaging
Documented Stroke or TIA	Transcranial Doppler Studies
Moyamoya Disease, when surgery or other vascular intervention is being considered	See <u>Moyamoya Syndrome/Disease</u> (HD-21.5)
Evaluation of paradoxical venous thromboembolism in cryptogenic stroke with PFO	See <u>Acute Limb Swelling (PVD-12.2)</u> and <u>Cryptogenic Stroke (HD-21.3)</u>
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 <u>Dementia PET (HD-8.2)</u>
Multisystem Smooth Muscle Syndrome/ Smooth Muscle Dysfunction Syndrome	See <u>Multisystem Smooth Muscle</u> 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 hypercoagulable 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

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Cryptogenic Stroke (HD-21.3)

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- 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 <u>Acute Limb Swelling (PVD-12)</u> 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 <u>Paraneoplastic Syndromes (ONC-30.3)</u> in the Oncology Imaging Guidelines.
 - Cardiac CT (CPT[®] 75574 or CPT[®] 75572) instead of TEE if TTE is inconclusive

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Transient Global Amnesia (HD-21.4)

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- 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 amnesia³⁹
 - 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 <u>Stroke/TIA</u> (HD-21.1)
- For suspected seizure, see Epilepsy/Seizures (HD-9.1)

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Moyamoya Syndrome/Disease (HD-21.5)

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Initial imaging for Moyamoya Syndrome/Disease

- .Below are indicated for initial evaluation of Moyamoya Syndrome/Disease³⁶:
 - 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/Disease³⁶

- MRA Head (CPT[®] 70544, CPT[®] 70545, 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, CPT[®] 78830 or CPT[®] 78832)¹² with vasodilating agent acetazolamide (Diamox) challenge is supported when surgery or other vascular intervention is considered. Follow up or repeat testing per neurologist, neurosurgeon, hematologist or in consultation with a neurologist, neurosurgeon or hematologist.
- 3D Rendering (CPT[®] 76377 or CPT[®] 76376) with cerebral angiography to define the presence, location, and anatomy of intracranial and cervical vascular malformations.²²
 - See <u>General Guidelines Other Imaging Situations (HD-1.7)</u> and <u>3D Rendering</u> (<u>Preface-4.1</u>) in the Preface Imaging Guidelines³⁷
- 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

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• 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

Head Imaging Guidelines

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- MRI Brain without contrast (CPT[®] 70551) OR MRI Brain without and with contrast (CPT[®] 70553) indications:
 - Screening to detect silent cerebral infarcts⁸
 - New symptoms or cognitive impairment occurs or a change in academic performance⁸
 - 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 Brain⁸
 - 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 S β^+).⁸

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

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Indications	Supported Imaging
Initial evaluation for confirmed ACTA2 mutation ⁴⁹	 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	 MRI Brain without and with contrast (CPT[®] 70553) with OR without MRI perfusion
coordinating care with a neurologist and/or neurosurgeon and/or geneticist ⁴⁹	AND/OR
	 MRA Head (CPT[®] 70544, CPT[®] 70545, or CPT[®] 70546)
	AND/OR
	 MRA Neck (CPT[®] 70547, CPT[®] 70548, or CPT[®] 70549)

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- No additional CPT[®] codes are necessary or appropriate to perform MRI perfusion. $_{51}^{51}$
- 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
- Conventional catheter angiogram 3D rendering (CPT[®] 76376 or 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 <u>Screening for Suspected Peripheral Artery Disease/Aneurysmal Disease</u> (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

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Evidence Discussion (HD-21)

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- Guidelines from the American Heart Association (AHA) and American Stroke Association support the role of neuroimaging in stroke triage and patient selection for endovascular therapy in the management of acute stroke. Current AHA guidelines also recommend non-invasive imaging of the carotid arteries for patients with TIA or stroke who may be candidates for carotid endarterectomy or stenting. This includes CT Angiography (CTA) of the head and neck or MR Angiography (MRA) of the head and neck.
- For clinically suspected stroke, initial imaging includes CT head to exclude intracranial hemorrhage, exclude other structural causes and assess for early ischemic changes. CT Angiography (CTA) head is indicated during the initial evaluation to assess for large vessel occlusion and has high sensitivity of 93% and specificity of 100%. CTA neck is also a rapid modality for imaging the extracranial vasculature to identify carotid stenosis, occlusion and vertebral-basilar disease.
- CT Perfusion (CTP) can identify patients with large vessel occlusion who may be candidates for endovascular therapy in the acute stroke setting.
- A CT stroke protocol that includes unenhanced CT, CTA head, and CTP has effective radiation doses between 10-15 mSvm, with newer generation scanners and optimized sequences with lower radiation doses closer to 2 mSv.5
- MRI brain with Diffusion Weighted Imaging (DWI) is the most sensitive test to detect an acute ischemic infarct, with a sensitivity of 94% and a specificity of 97%.
- MR Angiography (MRA) head and neck is an alternative to CTA for identification of vascular lesions in the setting of a stroke evaluation. Diagnostic cervicocerebral catheter angiography has the highest spatial and temporal resolution of any vascular imaging study, however, is invasive and requires use of contrast.
- Compared with CT, a full stroke protocol with MRI is longer to acquire and susceptible to motion artifacts, in addition to contraindication with metallic devices, and certain implants. MRI, however, does have the advantage of increased sensitivity for acute ischemia, including in transient ischemic attack, and does not require radiation.
- In the delayed stroke evaluation, CT head may identify complications such as hemorrhagic conversion, mass effect and herniation. MRI brain in this scenario can confirm the extent of an ischemic stroke, evaluate for underlying pathology and identify any complications.
- For clinically suspected transient ischemic attack (TIA), CT head is useful to exclude hemorrhage and other intracranial abnormalities. CT perfusion can identify abnormalities in the setting of TIA in up to one-third of cases. CT Angiography (CTA) head and neck is a rapid modality for evaluating intracranial and extracranial vascular lesions. MRA head and neck is an alternative modality, preferred in those with renal impairment and iodine contrast allergy. MRI brain is the most sensitive modality for

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acute ischemic infarct. MRI brain with Diffusion Weighted Imaging (DWI) sequences can identify ischemic changes in approximately 40% of patients with TIA.

- For clinically suspected venous sinus thrombosis, imaging is indicated to identify the clot and assess for complications, such as venous infarction or hemorrhagic transformation. In addition to imaging previously reviewed for the stroke protocol, CT Venogram (CTV) or MR Venogram (MRV) are appropriate to localize the clot within the venous system.
- For stroke or hemorrhage related to Cerebral amyloid angiopathy (CAA), Amyloid PET brain has a sensitivity that ranges from 82% to 91%, however, it's specificity is poor, ranging from 44% to 55%, therefore, this modality is not recommended.

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Cerebral Vasculitis (HD-22)

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Cerebral Vasculitis (HD-22.1)

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- When CNS vasculitis is suspected MRI Brain without and with contrast (CPT[®] 70553) is supported
 - MRA Head ($CPT^{\mathbb{B}}$ 70544, $CPT^{\mathbb{B}}$ 70545, or $CPT^{\mathbb{B}}$ 70546) **AND/OR**
 - MRA Neck (CPT[®] 70547, CPT[®] 70548, or CPT[®] 70549); **OR**
 - CTA Head (CPT[®] 70496) AND/OR
 - CTA Neck (CPT[®] 70498) are supported concurrently with brain imaging
- Primary CNS vasculitis includes Giant Cell Arteritis also known as Temporal Arteritis (see <u>New Headache Onset Older than Age 50 (HD-11.7)</u>)
- If initial vascular imaging is suspicious for vasculitis, 3D rendering (CPT[®] 76377 or CPT[®] 76376) with cervicocerebral angiography/arteriography (see <u>General</u> <u>Guidelines- Other Imaging Situations (HD-1.7)</u>).
- Transcranial Doppler Studies for individuals with documented vasculitis or concern for vasospasm
- FDG-PET/CT Brain (CPT[®] 78608) is not supported due to lack of peer reviewed literature or expert consensus supporting the study for vasculitis.⁷
- 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.

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Dominant Vessel Involved	Primary	Secondary
Large arteries	Giant cell arteritisTakayasu's arteritis	Aortitis with rheumatoid disease; Infection (e.g. syphilis)
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)

Evidence Discussion (HD-22)

- Noninvasive neuroimaging modalities play a role in the diagnostic evaluation of central nervous vasculitis by providing supportive imaging findings and guiding biopsy. The preferred modality for the evaluation of central nervous system vasculitis is MRI, which provides superior soft-tissue resolution. MRI brain is abnormal in > 95% of patients with CNS vasculitis. MRI brain shows infarcts in up to 50% of cases and white matter hyperintensities in 42% of cases.
- MRA head was found to be abnormal in 81% of patients with angiographic findings of vasculitis and normal in 100% of patients with a normal angiogram.
- CT Angiography is an alternative non-invasive modality that also provides visualization of blood vessels.
- FDG-PET/CT brain is not supported due to the high physiologic FDG uptake in the brain and limited resolution of the camera system.6 Atherosclerosis may also interfere with the FDG-PET interpretation.

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Dizziness, Vertigo and Syncope (HD-23)

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Dizziness/Vertigo (HD-23.1)

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ndications	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: <u>Headaches with Red Flags</u> (HD-11.2) <u>Multiple Sclerosis and Related Conditions (HD-16)</u> Brain Metastases (ONC-31.3)

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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 <u>Head Trauma</u> (HD-13.1)
	 AND/OR CT Orbit/Temporal bone without contrast (CPT[®] 70480)

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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

Head Imaging Guidelines

©2024 EviCore by EVERNORTH 400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 Page 242 of 326 www.EviCore.com 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

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- Individual has cervical spinal disease (i.e., cervical stenosis, severe kyphoscoliosis, limited cervical range of motion, Down's syndrome, severe rheumatoid arthritis, cervical radiculopathies, Paget's disease, ankylosing spondylitis, low back dysfunction, spinal cord injuries, spinal fractures)
- Individual unable to lay flat (i.e., severe heart disease)
- Individual has severe atherosclerotic disease or recent dissection involving the anterior or posterior cerebral circulation
- Unable to be seated in exam chair (i.e., morbidly obese), or maneuver cannot be safely performed given morbid obesity
- Ehlers Danlos/Marfans/Connective tissue disorder due to risk of cranio spinal instability/dissection
- Triggered episodic vestibular syndrome (t-EVS) usually last seconds to minutes with the most common triggers (vs. exacerbating factors) are head motion or change in body position. In the Emergency Department, benign paroxysmal positional vertigo (BPPV) is the second most common cause of t-EVS after orthostatic hypotension.
 Far lateral rotation of the neck leads to mechanical occlusion of one or both vertebral arteries causing temporary symptoms of vertigo and nystagmus when this position is maintained and may occur with the individual upright.
- Diagnoses or conditions associated with OH or nOH include: Parkinson Disease (PD), Multiple System Atrophy (MSA), Pure Autonomic Failure (PAF) or Dementia with Lewy Bodies (DLB), unexplained fall or syncope, peripheral neuropathies secondary to diabetes, amyloidosis and HIV), individuals ≥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 <u>Polyneuropathy (PN-3.1)</u> in the Peripheral Nerve and Neuromuscular Disorders Imaging Guidelines, <u>Multiple Myeloma and Plasmacytomas</u> (<u>ONC-25</u>) in the Oncology Imaging Guidelines, and <u>Paraneoplastic Syndromes</u> (<u>ONC-30.3</u>) 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

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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.

Evidence Discussion (HD-23.1)

- MRI brain is the preferred initial imaging modality for evaluation of persistent vertigo, vertigo associated with an abnormal neurologic exam, and vertigo due to a suspected central cause.
- CT head is not recommended for the initial evaluation of dizziness due to inferior soft tissue resolution when compared to MRI Brain. In addition, MRI brain provides better visualization of the cerebellum and posterior fossa and is more sensitive for the detection of posterior fossa infarcts. For suspected superior semicircular canal dehiscence, CT temporal bone is the appropriate initial imaging study.
- In the evaluation of dizziness or vertigo in the emergency department, the positivity rate of CT head was 2%, for MRI brain 4%, with the diagnostic yield increasing to 12% for MRI brain if neurologic findings were present.
- For dizziness due to suspected vertebral-basilar insufficiency, MRA sensitivity reaches 97% when performed with contrast-enhancement.
- For suspected vertebral artery dissection, CTA had the highest sensitivity 100%, followed by MRA 77%, and Doppler ultrasound at 71%.
- Vascular imaging should include the entire vertebral artery from the origin at the aortic arch to the basilar artery.

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Syncope (HD-23.2)

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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	a Guidelines
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)	laad Imaain
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Evidence Discussion (HD-23.2)

- The Choosing Wisely Campaign Best Practices, supported by the American College of Emergency Physicians, American College of Physicians, and the American Academy of Neurology, recommend against neuroimaging in the evaluation of simple syncope and a normal neurologic evaluation.
- The initial evaluation for patients with syncope includes a detailed history, physical exam and electrocardiography. Neuroimaging has a low diagnostic yield of 5% to 6.4% of an acute abnormality on CT head. Clinical factors associated with abnormal scans include head trauma or a focal neurologic deficit on exam.
- In select cases when neuroimaging is indicated, structural brain imaging with either CT head or MRI brain may be useful, along with vascular imaging, depending on the suspected underlying pathology.
- Inappropriate imaging studies may identify incidental findings, incorrectly assumed to be the cause of syncope, leading to further delay in the identification of the true cause and risk additional unnecessary procedures.
- Situational syncope does not require advanced imaging.

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Other Imaging Studies (HD-24)

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Head Imaging Guidelines

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HD.OI.0024.1.A v1.0.2025

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.

TMS is typically utilized for behavioral health purposes.

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Functional MRI (fMRI) (HD-24.2)

HD.OI.0024.2.A v1.0.2025

- 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 <u>Unlisted Procedures/Therapy Treatment Planning (Preface-4.3)</u> in the Preface Imaging Guidelines if MRI Unlisted is requested for surgical planning

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Magnetic Resonance Spectroscopy (MRS) (HD-24.3)

HD.OI.0024.3.A

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- MRS (CPT[®] 76390) involves analysis of the levels of certain chemicals in a preselected 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 (pre-treatment and posttreatment)
 - Grading of primary glial neoplasm, particularly high-grade versus low-grade glioma
 - Evidence or suspicion of brain infection, especially cerebral abscess (pre-treatment and post-treatment) 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 <u>Primary Central Nervous System Tumors General Considerations</u> (ONC-2.1), Low Grade Gliomas (ONC-2.2), and <u>High Grade Gliomas (ONC-2.3)</u> in the Oncology Imaging Guidelines

Head Imaging Guidelines

CSF Flow Imaging (HD-24.4)

HD.OI.0024.4.A v1.0.2025

- 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.

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CT or MRI Perfusion (HD-24.5)

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- 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.
 - $^\circ$ CTA Head and/or Neck is indicated in conjunction with the CT Perfusion study (CPT $^{\textcircled{R}}$ 0042T)
- CPT[®] 0042T "cerebral perfusion analysis using CT".
 - To evaluation of acute stroke (<24 hours) to help identify individuals with strokelike symptoms and to help identify those most likely to benefit from thrombolysis or thrombectomy
 - Follow up for acute cerebral ischemia or infarction and/or reperfusion in the subacute or chronic phase of recovery
 - To assist in planning and evaluating the effectiveness of therapy for cervical or intracranial arterial occlusive disease (as an isolated test or in combination with a cerebrovascular reserve challenge) and/or chronic cerebral ischemia
 - · Identifying cerebral hyperperfusion syndrome following revascularization
 - Evaluation of the vascular status of solid tumors where MRI is degraded due to susceptibility artifact from air-containing spaces, surgical clips, or dental work
 - Follow up of tumor response to therapy
- 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

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- 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.

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Magnetic Resonance Neurography (MRN) (HD-24.6)

HD.OI.0024.6.A

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• See <u>Magnetic Resonance Neurography (MRN) (PN-7.1)</u> in the Peripheral Nerve and Neuromuscular Disorders (PNND) Imaging Guidelines.

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

HD.OI.0024.7.A v1.0.2025

- 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 <u>Temporomandibular Joint Disease (TMJ) (HD-30.1)</u> and <u>Dental/Periodontal/</u> <u>Maxillofacial Imaging (HD-30.2)</u>

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Evidence Discussion (HD-24)

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- Functional magnetic resonance imaging (fMRI) is useful for localizing eloquent cortex in relation to a focal brain lesion, for pre-surgical planning and therapeutic follow up. Overall functional MRI imaging sensitivity is 83% with a specificity of 82%, for mapping language and motor functions. Functional MRI is a useful tool for predicting post-operative outcomes in patients with a single brain tumor. Overall, fMRI studies are used in preoperative decision making in 89% of tumor patients and in 91% of epilepsy surgery patients. In 63% of epilepsy patients undergoing surgical evaluation, fMRI imaging results helped to avoid further studies, including the Wada test.
- For cases when conventional imaging by magnetic resonance imaging or computed tomography provides limited information regarding specific clinical questions, magnetic resonance spectroscopy (MRS) provides further characterization of brain tumors, radiation treatment changes, cerebral abscess, seizure disorders, and inherited metabolic disorders. MRS has a 90% sensitivity and 86% specificity in distinguishing tumoral tissue from non-tumoral tissue.
- Pulse-gated MRI imaging or MRI CINE is performed as part of an MRI brain study and allows qualitative and quantitative analysis of oscillatory cerebrospinal fluid (CSF) movement in normal and abnormal conditions. This imaging technique is useful for evaluation of hydrocephalus, Chiari syndromes, Normal Pressure Hydrocephalus, intracranial hypertension, and spontaneous intracranial hypotension.
- MRI perfusion is useful for the diagnosis and characterization of mass lesions, surgical planning and therapeutic follow up. MR perfusion allows localization of tumor for higher yield on stereotactic biopsy and noninvasive differentiation between radiation necrosis from recurrent tumor when conventional MR findings are equivocal.
- CT perfusion has multiple uses including in stroke diagnosis and treatment planning, characterization of neoplastic disease and response to treatment, and is alternative modality for those with contraindication to MRI-based perfusion imaging.
- The American Heart Association/American Stroke Association guidelines for acute stroke management recommend CT Perfusion for selecting candidates for mechanical thrombectomy within 24 hours after last known well.

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Epistaxis (HD-25)

Guideline

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Epistaxis (HD-25.1)

HD.EX.0025.1.A

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- 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/Neck without and with contrast (CPT[®] 70543)
- Patients who have failed initial management with cauterization and packing and have persistent or recurrent epistaxis despite these primary interventions, should be referred to a clinician who can evaluate the patient for their candidacy for surgical ligation or endovascular embolization.³
- Prior to embolization with surgical or endovascular technique, CT Maxillofacial without contrast (CPT[®] 70486) OR CT Maxillofacial with contrast (CPT[®] 70487) is supported when requested by the clinician 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

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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%).

Evidence Discussion (HD-25)

- The American Academy of Otolaryngology Head & Neck Surgery (AAO-HNS) recommends, 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 to 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 AAO-HNS Guideline without the exam findings (anterior rhinoscopy and/or nasal endoscopy) or the procedural needs of the patient, directing the need for such studies. If anterior rhinoscopy does not reveal the source of bleeding, it is recommended that the clinician perform nasal endoscopy, first.
- Further characterization of any mass lesions suspected on initial nasal endoscopy may be evaluated with CT Maxillofacial, either with OR without contrast (CPT® 70487 or CPT® 70486), AND/OR MRI Orbit, Face, and/or Neck without and with contrast (CPT® 70543)
- Because of the risks involved in embolization procedures (blindness, stroke and others), CT or MRI imaging is supported prior to any planned intervention.
- 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).

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Mastoid Disease or Ear Pain (HD-26)

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Mastoid Disease or Ear Pain (HD-26.1)

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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)
- 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 preoperative 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).

Evidence Discussion (HD-26)

 Contrast enhanced CT is commonly used for evaluation of head and neck infections due to its accessibility and short examination time. MRI provides better sensitivity of soft tissue infections in the setting of cholesteatoma, when there is concern for abscess formation or intracranial complications.

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Hearing Loss and Tinnitus (HD-27)

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Hearing Loss (HD-27.1)

HD.HL.0027.1.A

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- 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 <u>General Guidelines (HD-1.0)</u>
- 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 <u>Mastoid Disease or Ear Pain (HD-26.1)</u>)
 - 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.

Head Imaging Guidelines

Evidence Discussion (HD-27.1)

- A complete history and otologic exam should be performed prior to advanced imaging for the workup of hearing loss. Formal audiometric testing is also necessary to determine whether the hearing loss is conductive, sensorineural or mixed.
- MRI brain is generally preferred for sensorineural and mixed hearing loss, particularly for unilateral hearing loss, congenital loss, or for surgical planning.
- CT orbits/temporal bone is preferred for cases of conductive hearing loss, trauma, or suspected bony or middle ear disorders.
- Both may be supported for surgical planning (cochlear implants, petrous apex disorders)

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Head Imaging Guidelines

Tinnitus (HD-27.2)

HD.HL.0027.2.A v1.0.2025

- 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 <u>General</u> <u>Guidelines (HD-1.0)</u>)

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 symptoms⁶
- 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

©2024 EviCore by EVERNORTH 400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 Page 273 of 326 www.EviCore.com 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.

Evidence Discussion (HD-27.2)

- A targeted history and clinical examination should be performed as the initial evaluation of a patient with tinnitus and determination as to whether the tinnitus is bothersome or not should be made before any imaging is considered.
- Both MRI and CT have utility in diagnosing the etiology of tinnitus, particularly for concerns of mass lesions, or for tinnitus in conjunction with hearing loss or trauma.
- MRA or CTA of the head and neck are also useful in the workup of pulsatile tinnitus or for suspicion of a vascular lesion

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Neurosurgical Imaging (HD-28)

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- 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

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Neuronavigation (HD-28.2)

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- Neurosurgical navigation is "image-based" meaning that the necessary preoperative CT and MRI images are used for navigation in the operating room (image acquisition). Accurate registration (a process to match the 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 <u>Unlisted Procedures/Therapy Treatment Planning (Preface-4.3)</u> in the Preface Imaging Guidelines and <u>Unlisted Procedure Codes (ONC-1.5)</u> 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 or CPT[®] 76376) (see <u>General Guidelines Other Imaging</u> <u>Situations (HD-1.7)</u>)
 - 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

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- 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 <u>Special Imaging Studies in Evaluation for Epilepsy Surgery</u> (<u>PEDHD-6.3</u>) in the Pediatric Head Imaging Guidelines
 - See <u>Modality General Considerations (PEDONC-1.3)</u> and <u>Pediatric CNS</u> <u>Tumors (PEDONC-4)</u> in the Pediatric and Special Populations Oncology Guidelines
 - Epilepsy
 - See Presurgical Work-Up for Drug-Resistant Epilepsy (HD-9.2)
 - Movement Disorders
 - See <u>Movement Disorders (HD-15.1)</u>
 - Pituitary or Sella Surgery
 - See <u>Pituitary (HD-19.1)</u>
 - Acoustic Neuroma and Other Cerebellopontine Angle Tumors
 - See Acoustic Neuroma and Other Cerebellopontine Angle Tumors (HD-33.1)
 - Central Nervous System Tumors
 - See <u>Primary Central Nervous System Tumors (ONC-2)</u> and <u>Brain</u> <u>Metastases (ONC-31.3)</u> in the Oncology Imaging Guidelines

Evidence Discussion (HD-28)

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- Imaging modalities for neurosurgical planning include MRI brain and CT head, along with vascular imaging.
- MRI brain is the preferred modality for the evaluation of intracranial neoplasms and other conditions affecting the brain parenchyma, meninges or cranium, due to its superior soft tissue resolution when compared to CT head.
- CT head is the preferred modality for evaluation of bony structures in the preoperative setting.
- For localization of relevant vascular anatomy prior to surgery, MRI angiography (MRA) head and neck or CT angiography (CTA) head and neck, may be appropriate.
- Pre-surgical navigational imaging, whether by CT or MRI, allows a spatially accurate anatomical patient model for use in the treatment-planning process.
- The requirements for surgical planning images differ from the requirements for diagnostic images, especially regarding the spatial accuracy of the images in the stereotactic coordinates used for localization and targeting.
- Navigation based on an immediate preoperative scan optimizes the accuracy of data used for initial surgical planning. Navigation systems reduce length of surgery, lower incidence of wound infection and shorten length of post-operative hospital stay.
- For post-operative imaging, CT head is also useful for follow up of intracranial hemorrhage, edema, hydrocephalus, shunts, and general post-operative follow up. CT head has the benefit of providing rapid evaluation if a post-operative complication is suspected. Post-operative MRI brain provides superior soft tissue resolution in less urgent scenarios.

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Sinus and Facial Imaging (HD-29)

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Sinus and Facial Imaging (HD-29.1)

HD.SI.0029.1.C

- v1.0.2025
- 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
 - 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 <u>Taste and Smell Disorders (HD-2.1)</u> 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 imaging studies not performed for the purpose of evaluating sinus pathology, such as MRI Brain for headache, when requested by ENT for clinical correlation.
- Studies requested for the sole 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 <u>Cough (CH-3.1)</u> in the Chest Imaging Guidelines.

- CT Maxillofacial with contrast (CPT[®] 70487)* is indicated in the immunocompromised individual with symptoms of sinusitis³, and suspicion for ANY of the following:
 - Orbital or facial cellulitis
 - Proptosis
 - Abnormal visual examination
 - Ophthalmoplegia
 - Fungal or vascular lesions visualized in nasal cavity
- *Contrast level as requested when ordered by the surgeon or in consultation with the surgeon (i.e. ENT or ophthalmologist)³
- CT Maxillofacial without contrast (CPT[®] 70486) OR CT Maxillofacial with contrast (CPT[®] 70487) OR MRI Orbits/Face/Neck without and with contrast (CPT[®] 70543). However, CT Maxillofacial without contrast (CPT[®] 70486) may also be requested with MRI Orbits/Face/Neck without and with contrast (CPT[®] 70543) for surgical planning or osseous involvement).^{1,3,4}
 - Sinonasal obstruction, polyp, or suspected mass
 - Suspected orbital complication
 - Suspected invasive fungal sinusitis
 - Cystic fibrosis
 - Osteomyelitis and odontogenic infections, see <u>Skull Base Osteomyelitis (SBO)</u> (HD-20.2) and <u>Dental/Periodontal/Maxillofacial Imaging (HD-30.2)</u> for additional imaging modalities
- 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 with contrast (CPT[®] 70481) performed alone or added to CT Maxillofacial for:
 - Suspected orbital complications⁴
- For Skull Base Osteomyelitis (SBO), see <u>Skull Base Osteomyelitis (SBO) (HD-20.2)</u>
- 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:
 - The following imaging is indicated: CT Maxillofacial without contrast (CPT[®] 70486) OR limited CT Sinus without contrast (CPT[®] 76380)^{1,3,4}
 - There is a new abnormality on exam such as obstructing mass
 - The following imaging is indicated: CT Maxillofacial without contrast (CPT[®] 70486) **OR** CT Maxillofacial with contrast (CPT[®] 70487)^{1,3,4}

- If sinus surgery is planned (including but not limited to Balloon Sinus Ostial Dilation or Functional Endoscopic Sinus Surgery) AND the most recent diagnostic CT Maxillofacial without contrast (CPT[®] 70486) is greater than 6 months old OR there is a change in clinical status as described above (i.e interval completion of provider-prescribed medical management after the last CT was performed), a repeat diagnostic CT Maxillofacial without contrast (CPT[®] 70486) is supported for surgical planning.^{1,3,4}
 - Repeat CT Maxillofacial solely for the use of navigation during the sinus surgery (i.e. the most recent diagnostic CT Maxillofacial performed within the prior six months was only inadequate due to lacking anatomic landmarks or insufficient thinness of cuts) should be requested with CPT[®] 77011, not the diagnostic CPT[®] 70486.
 - 3D Rendering (CPT[®] 76376 or CPT[®] 76377) should not be reported in conjunction with CPT[®] 77011 (or CPT[®] 70486 if used). The procedure inherently generates a 3D dataset.
- Complications of ABRS (acute bacterial rhinosinusitis) are 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
 - Orbital signs (cellulitis, impaired extraocular motility, decrease in vision or proptosis)
- Complications of ABRS are best assessed using iodine contrast-enhanced CT Maxillofacial with contrast (CPT[®] 70487) OR gadolinium based MR imaging (MRI Orbits/Face/Neck without and with contrast (CPT[®] 70543) to identify extra-sinus extension or involvement^{1,3,4}
 - CT Maxillofacial without contrast (CPT[®] 70486) may also be requested with MRI Orbits/Face/Neck without and with contrast (CPT[®] 70543) for surgical planning or osseous involvement.^{1,3,4}
 - Suspected complications are the only indication for MR imaging of the paranasal sinuses in the setting of ABRS.

For additional medical necessity criteria for CT maxillofacial, see Cone Beam Imaging, see Cone Beam Computed Tomography (CBCT) (HD-24.7) and Dental/Peridontal/ Maxillofacial Imaging (HD-30.2))

Evaluation of potential candidates for Eustachian Tube balloon dilation procedure is with a one-time CT of the temporal bone without contrast (CPT[®] 70480). See medical necessity discussion in <u>Mastoid Diseae or Ear Pain (HD-26.1)</u>. CT Sinus/Maxillofacial irrespective of contrast level is not supported if the sole indication for medical necessity

is to evaluate a potential candidate for Eustachian Tube balloon dilation procedure, without meeting other HD-29.1 medical necessity criteria.^{7,8}

Background and Supporting Information

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

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Evidence Discussion (HD-29.1)

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- The American Academy of Otolaryngology Head & Neck Surgery (AAO-HNS) recommends that clinicians should not obtain radiographic imaging for patients with suspected uncomplicated Acute Rhinosinusitis (ARS), with imaging reserved for cases with clinically suspected complication. ARS refers to inflammation of the nasal cavity and paranasal sinuses lasting <4 weeks' duration.
- Contrasted CT maxillofacial is first line imaging for rhinosinusitis with suspected complications (orbital or intracranial). There is up to 91% accuracy with CT to detect orbital complications vs clinical exam alone. CT also is preferred for surgical planning. However, CT is often more useful for surgical planning and easier to perform. Noncontrasted CT sinus is not preferred, but may be useful for surgical navigation. There is no relevant literature to support pre- and post-contrast CT imaging. MRI head or orbits/face/neck can be complementary with CT. MRI is more accurate than CT in the evaluation of soft tissues regarding intra-orbital and intracranial complications.
- Chronic rhinosinusitis, acute recurrent bacterial sinusitis, non-invasive fungal sinusitis, and/or sinonasal polyposis are best evaluated initially with non-contrast CT maxillofacial. CT is critical for surgical planning. Contrast is not necessary unless complications are suspected. MRI is not useful as the first-line study because of the lack of bony detail. In select cases, evaluation with MRI without and with IV contrast may be helpful to differentiate fluid secretions from inflamed mucosa and exclude an underlying obstructing mass.
- Urgent CT maxillofacial, either without or with IV contrast is first line imaging for any suspected invasive fungal sinusitis, as delay in diagnosis and surgical debridement could increase the already high risk of mortality. In cases of invasive fungal sinusitis, MRI without and with IV contrast of the head and/or orbits/face/neck is adjunctive to look for invasion into surrounding soft tissues as well as vascular complications.
- CT and MRI are considered complimentary imaging modalities in the evaluation of a sinonasal mass—localizing and characterizing the lesions to determine their extent for treatment planning. If an MRI is planned, the CT may be performed without IV contrast since the main purpose of the CT is to evaluate osseous involvement.

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

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Temporomandibular Joint Disease (TMJ) (HD-30.1)

HD.TJ.0030.1.A

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- 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. The exception to the conservative management requirement includes recent trauma, dislocation, severe malocclusion, dental infection or abscess.^{6,8}
- 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 <u>Temporomandibular Joint (TMJ) Imaging in Children (PEDHD-25)</u> 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 treatment¹¹
 - 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 surgery¹³

Evidence Discussion (HD-30.1)

 MRI is preferred for evaluation of the temporomandibular joint (TMJ) due to its superior contrast resolution and it ability to acquire dynamic imaging for demonstration of the functionality of the joint.

- MRI is the imaging modality of choice for the diagnosis of internal derangement with an accuracy of 95% in assessing the disc position and form and 93% accuracy in assessing the osseous changes.
- MRI is reserved for patients with persistent symptoms in whom conservative measures have been ineffective, or in those with suspected internal joint derangement. Imaging the TMJ prematurely may lead to harms including unnecessary surgery.
- CT is the alternative modality for evaluating bony anatomy of the TMJ, fractures, degenerative changes, erosions, infections, congenital anomalies, acute and chronic inflammatory conditions, pre-operative evaluation and follow up after surgery.
- For pre-operative planning of unilateral condylar hyperplasia, bone scintigraphy is useful to predict ongoing condylar growth.
- The diagnosis of chronic rheumatoid arthritis of the TMJ is established with contrastenhanced MRI. It is the preferred imaging study for diagnosis, disease progression, treatment monitoring and annual surveillance of TMJ arthritis in juvenile idiopathic arthritis (JIA).

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Dental/Periodontal/Maxillofacial Imaging (HD-30.2)

HD.TJ.0030.2.A

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- 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 <u>Cone Beam Computed Tomography (CBCT)</u> (<u>HD-24.7</u>))
- 3-D rendering (CPT[®] 76376 or CPT[®] 76377) should **NOT** be reported separately
- Cone beam CT (CBCT) may also be called i-CAT scanner or mini-CAT scanner

Evidence Discussion (HD-30.2)

- CT is the radiologic modality for evaluating the bony anatomy of the head, acute and chronic inflammatory conditions, paranasal sinuses, pre-operative evaluation and follow up after surgery.
- Recommendations by the American Association of Endodontists and the American Academy of Oral and Maxillofacial Radiology support the use of cone beam CT (CBCT) as a supplemental imaging technique when conventional radiography fails to answer the clinical question and for surgical planning.

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Cranial Neuropathies (HD-31)

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Cranial Neuropathies (HD-31.1)

HD.CN.0031.1.A v1.0.2025

Cranial Neuropathies Imaging Indications

- MRI Brain without and with contrast (CPT[®] 70553) OR MRI Brain without contrast (CPT[®] 70551) is indicated for all individuals with new or worsening specific cranial nerve abnormalities.²
- MRI Orbit/Face/Neck without and with contrast (CPT[®] 70543) OR MRI Orbit/ Face/Neck 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.^{1,2}
- CT Neck with contrast (CPT[®] 70491) is supported for evaluation of abnormalities involving cranial nerves IX, X, XI, or XII.²
- Imaging of the Brain and Orbit, Face and/or Neck may be performed concurrently when requested.²
- For specific cranial neuropathies², see the corresponding guideline section listed below:
 - CN I: Olfactory nerve (see Taste and Smell Disorders (HD-2.1))
 - CN II, III, IV, VI: Optic, Oculomotor, Trochlear and Abducens (see <u>Eye Disorders</u> and Visual Loss (HD-32.1))
 - CN V: Trigeminal nerve (see <u>Trigeminal Neuralgia and other Centrally Mediated</u> <u>Facial Pain Syndromes (HD-10.1)</u>)
 - CN VII: Facial nerve (see Facial Palsy (HD-6.1))
 - CN VIII: Vestibulocochlear nerve (see <u>Dizziness/Vertigo (HD-23.1)</u>, <u>Hearing Loss</u> (HD-27.1), <u>Tinnitus (HD-27.2)</u>, <u>Acoustic Neuroma and Other Cerebellopontine</u> <u>Angle Tumors (HD-33.1)</u>. For isolated nystagmus (see <u>Eye Disorders and Visual</u> <u>Loss (HD-32.1)</u>)
 - CN IX: Glossopharyngeal nerve (see <u>Glossopharyngeal Neuralgia/</u> <u>Glossopharyngeal Neuropathy HD-10.2)</u>)
 - CN X: Vagal nerve, imaging as detailed above (see also <u>Recurrent Laryngeal</u> <u>Palsy/Vocal Cord Palsy (Neck-7.1)</u>)
 - 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 <u>General Guidelines - CT and MR</u> <u>Angiography (CTA and MRA) (HD-1.5)</u>, <u>Headache and Suspected Vascular</u> <u>Dissection (HD-11.1)</u> and <u>Stroke/TIA (HD-21.1)</u>

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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.

The spinal trigeminal tract and nucleus extend from the midpons caudally into the upper cervical cord at the C2-4 levels. For suspected lesions of the spinal trigeminal tract and nucleus, imaging the brain stem and cervical spinal cord is supported.² See <u>Trigeminal</u> <u>Neuralgia and other Centrally Mediated Facial Pain Syndromes (HD-10.1)</u>.

Number	Cranial Nerve Name	Nerve dysfunction on exam	Guideline Section in HD
ł	Olfactory (smell)	Anosmia, hyposmia, parosmia, phatosmia	2
H	Optic (vision)	Optic neuritis, disc edema, papilledema, afferent pupillary defect APD)	16, 17, 32
111	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
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

Head Imaging Guidelines

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Number	Cranial Nerve Name	Nerve dysfunction on exam	Guideline Section in HD
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
XII	Hypoglossal (tongue movement)	Tongue deviation, atrophy, fasciculation	1.1

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Head Imaging Guidelines

Number	Cranial Nerve Name	Nerve dysfunction on exam	Guideline Section in HD
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

Evidence Discussion (HD-31.1)

- Imaging of each body section along the entire course of the relevant cranial nerve may be indicated if detailed clinical evaluation is unable to localize the site of the lesion. 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, indicating need to image multiple body sections. MRI brain, orbits, face, neck, or any combination may be necessary depending on the clinical need.
- MRI is the standard modality for imaging the cranial nerves.
- CT Neck is useful to exclude neck masses when evaluating either isolated or multiple lower cranial neuropathies. CT may be complementary to MRI in characterizing skull base erosions, calcifications, and skull base foramina.

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Eye Disorders and Visual Loss (HD-32)

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Eye Disorders and Visual Loss (HD-32.1)

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- 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, 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 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 <u>Cranial Neuropathies</u> (HD-31.1) and <u>Papilledema/Pseudotumor Cerebri (HD-17.1)</u>)
 - 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 <u>Cranial Neuropathies (HD-31.1)</u>)
 - Nystagmus²¹
- 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 <u>Multiple Sclerosis</u> (MS) (HD-16.1) and <u>Neuromyelitis Optica Spectrum Disorders (HD-16.2)</u>)
- Visual field defects are associated with retrochiasmal pathology (see <u>Stroke/TIA</u> (HD-21.1) or <u>Primary Central Nervous System Tumors (ONC-2)</u> in the Oncology Imaging Guidelines or <u>Brain Metastasis (ONC- 31.3)</u> 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

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- 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 <u>Intracranial</u> <u>Aneurysms (HD-12.1)</u>)
- Amaurosis Fugax (see <u>Stroke/TIA (HD-21.1)</u>)
 - Individuals describe a transient darkening or loss of vision, typically monocular
- Central Retinal Artery Occlusion, Branch Retinal Artery Occlusion, and Ophthalmic Artery Occlusion (see <u>Stroke/TIA (HD-21.1)</u>)
 - 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:

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- Retinoblastoma (see **Retinoblastoma (PEDONC-12)** in the Pediatric and Special
- Uveal (choroidal) melanoma (see **Ocular Melanoma (ONC-5.9)** in the Oncology Imaging Guidelines)
- Vasculitis including Temporal Arteritis (Giant Cell Arteritis) (see <u>Cerebral Vasculitis</u>) (HD-22.1))

Background and Supporting Information

Imaging Non-Indications

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- Imaging is not necessary if visual loss or ocular symptom/sign is due to known intrinsic eve disease, such as refractive errors, amblyopia, ptervolum. 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.

These guidelines apply to services or supplies managed by EviCore for Cigna as outlined by the Cigna CPT list.

- 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) or relapses, worsening proptosis, diplopia, lid retraction, or optic neuropathy
- For surgical planning for orbital decompression, strabismus surgery or lid surgery
- Autoimmune Retinopathy ٠
 - 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
 - Populations Oncology Imaging Guidelines)

Biopsy results are not required before initial staging

List of Abbreviations and Meanings:

Abbreviation	Meaning
AC	Anterior chamber
APD	Afferent pupillary defect (see RAPD)
BCVA	Best-corrected visual acuity
C3F8	Gas bubble injected into vitreous cavity during retina surgery
сс	With correction (current new or old glasses or contact lenses)
СР	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
Ε'	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

Head Imaging Guidelines

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Head Imaging Guidelines

Abbreviation	Meaning	
НТ	Hypertropia	
HVF	Humphrey visual field test (automated perimetry)	
l	Iris	
Ishihara	Commonly used color plates	
IOP	Intraocular pressure	
К	Cornea	
LF	Levator function	
LFH	Lid fissure height	
LLL	Lids, lashes, lacrimal gland	
М	Macula	
ME	Macular edema	
MH	Macular hole	
MP	Membrane peel	
MRD1	Margin-reflex distance from upper lid margin to pupillary light reflex	
MRx	Manifest refraction	
NI	No improvement	
NLDO	Nasolacrimal duct obstruction	
NSC or NS	Nuclear sclerotic cataract	-
OD	Right eye	
OS	Left eye	-
ortho	Eyes are aligned on the same target	
ОСТ	Optical Coherence Tomography	
P	Periphery	

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Abbreviation	Meaning
PD	Prism diopter
ph or PH	Pinhole (crude assessment of best-corrected visual acuity)
PPV or PPVx	Pars plana vitrectomy
PVD	Posterior vitreous detachment
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
Та	Applanation tonometry (intraocular pressure measurement)
Тр	Tonopen tonometry (intraocular pressure measurement)
V	Vessels
Va	Visual acuity
VF	Visual field testing (formal automated perimetry versus confrontation visual field testing)
х	Exophoria at distance
Χ'	Exophoria at near
ХТ	Exotropia
X(T)	Intermittent exotropia at distance
XT'	Exotropia at near

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Head Imaging Guidelines

Abbreviation	Meaning
X(T)'	Intermittent exotropia at near

Evidence Discussion (HD-32.1)

- When evaluating suspected or known issues involving the eye, orbit, and/or brain, consideration must be given to:
 - whether or not imaging is required,
 - which body area should be imaged, i.e. brain, orbits, or both, and
 - which modality, MRI or CT, would best provide the information needed while exposing an individual to the least risk.
- The body area imaged should be reasonably expected to be potentially involved in the suspected condition. The angles of and distance between each view differ between brain imaging and orbital imaging. There are circumstances in which imaging of both the brain and orbits may be useful, as in conditions that can affect both locations or for which evaluation by the different techniques provides useful information.
- · Soft tissue detail such as neural tissue is well-visualized by MRI.
- · Calcification, bone, and hemorrhages are well-visualized by CT.
- MRI carries no risk of radiation exposure but is sensitive to motion, takes longer, and may require sedation or anesthesia for a longer duration than would be required for CT. Certain populations may have psychological or physical difficulty undergoing MRI scans, including children, those with obesity, movement disorders, anxiety or claustrophobia.
- CT carries risk of radiation exposure but is less sensitive to motion and has a shorter duration than MRI. Imaging more than one body area increases the exposure dose. Certain populations may carry higher risk of detrimental effects from exposure, including children.
- Radiation exposure of the ocular lens contributes to cataract formation. Radiation doses vary between CT scans due to differences in scanning technique, number of images taken per CT, body area scanned, CT machines used, and facility protocols. The cancer risk of radiation exposure in diagnostic CT is considered extremely low, and the benefit of an appropriately indicated CT examination far outweighs the potential risk. Cataract formation is among the earliest radiation associated pathologies in the eye. The Beaver Dam Eye Study, a population-based study of common age-related eye diseases, found that nuclear sclerosis and posterior subcapsular opacity were significantly associated with CAT scans.

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Pupillary Abnormalities Including Horner's Syndrome (HD-32.2)

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- 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 <u>Eye Disorders and Visual</u> 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
- MRI Brachial plexus for Horner syndrome with traction or trauma to the brachial plexus²
 - Any **ONE** of the following:
 - MRI Upper Extremity other than joint without contrast (CPT[®] 73218)
 - MRI Upper Extremity other than joint without and with contrast (CPT[®] 73220)
 - MRI Chest without contrast (CPT[®] 71550)
 - MRI Chest without and with contrast (CPT[®] 71552)
 - MRI Neck without contrast (CPT[®] 70540)
 - MRI Neck without and with contrast (CPT[®] 70543)
- CT Chest with contrast (CPT[®] 71260) or MRI Chest without and with contrast (CPT[®]71552) for suspected chest mass^{4,24}
- CT Neck with contrast (CPT[®] 70491) or MRI Face/Neck/Orbits without and with contrast (CPT[®] 70543) for suspected neck mass⁴
- CTA Neck without and with contrast (CPT[®] 70498) or MRA Neck (CPT[®] 70547, CPT[®] 70548, or CPT[®] 70549) for suspected carotid injury or dissection

©2024 EviCore by EVERNORTH 400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 Page 309 of 326 www.EviCore.com 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

Evidence Discussion (HD-32.2)

- In the evaluation of Horner Syndrome, if a detailed clinical evaluation is unable to localize the site of the lesion, imaging of the entire course of the relevant oculosympathetic pathway is required, as symptoms may result from pathology affecting the nerve fibers at any point along the course of the pathway, requiring multiple imaging modalities. MRI brain, MRI Orbits/Face/Neck, MRI cervical spine and/or MRI Brachial plexus studies may be necessary, depending on the clinical presentation.
- CT Neck is useful to exclude neck masses. CT may be complementary to MRI in characterizing skull base erosions, calcifications, and skull base foramina.
- For suspected lung masses associated with Horner syndrome, such as for evaluation of Pancoast tumors, chest imaging is recommended. A mass may be diagnosed on a CT chest or an MRI chest. CT scans provide 60% sensitivity, 65% specificity, and 63% accuracy in defining the local extent of tumor, in contrast to MRI with a sensitivity of 88%, a specificity of 100%, and an accuracy of 94%. MRI of the chest is a more accurate preoperative examination in identifying the local extent of a Pancoast tumor.
- For suspected carotid injury or dissection, vascular imaging with either CT Angiography (CTA) neck or MR Angiography (MRA) neck is indicated, depending on the individual's risk and benefit profile.

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

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

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- · Acoustic neuroma and vestibular schwannoma may be used interchangeably
- Initial diagnosis is usually made during evaluation for asymmetric hearing loss and/ or vertigo (see <u>Dizziness, Vertigo and Syncope (HD-23)</u> and <u>Hearing Loss and</u> <u>Tinnitus (HD-27)</u>) 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 neurologist, neurosurgeon, or otolaryngologist, or any provider in consultation with a neurologist, neurosurgeon, or otolaryngologist.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.
- 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 <u>General Guidelines Anatomic Issues (HD-1.1)</u>)
- See <u>Primary Central Nervous System Tumors- General Considerations</u> (<u>ONC-2.1</u>) in the Oncology Imaging Guidelines for additional imaging requests for surgery

Evidence Discussion (HD-33)

 MRI brain is the preferred initial imaging modality for evaluation of persistent vertigo, vertigo associated with an abnormal neurologic exam, vertigo due to a suspected central cause, pulsatile or asymmetric tinnitus, and/or hearing loss.

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- CT head is not recommended for the initial evaluation of suspected acoustic neuroma due to inferior soft tissue resolution when compared to MRI Brain. In addition, MRI brain provides better visualization of the cerebellum, posterior fossa and cranial nerves.
- MRI brain, in this clinical scenario, is performed using specialized internal auditory canal (IAC) protocols, which include thin-section sequences to evaluate for vascular loops and small vestibular schwannomas.
- MRI brain can diagnose lesions in the cerebellopontine angle including schwannoma, meningioma, and other posterior fossa tumors.
- 3D-Fast imaging employing steady state acquisition (3D-FIESTA) demonstrates significantly higher spatial resolution with superior imaging contrast between cranial nerves and CSF with a shorter acquisition time than conventional MRI scan.
- Follow up imaging is recommended 6 months after diagnosis to evaluate for rapid growth, then annually for 5 years. After 5 years, tumor growth that has remained stable is unlikely but may still occur, therefore, lifelong surveillance is advised with longer imaging intervals.
- Follow up imaging after surgical resection and/or stereotactic radiosurgery to assess residual tumor and treatment response is performed at 6-12 months with additional follow up annually for 5 years and every 2 years thereafter.

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Pineal/Colloid Cysts (HD-34)

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Pineal/Colloid Cysts (HD-34.1)

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Pineal Cysts

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

- MRI Brain without contrast (CPT[®] 70551) or MRI Brain without and with contrast (CPT[®] 70553) is indicated for initial evaluation of pineal cysts if not already completed.
- Repeat MRI Brain is not indicated for most individuals with pineal cysts, but MRI Brain without contrast (CPT[®] 70551) or MRI Brain without and with contrast (CPT[®] 70553) for the following:
 - New or worsening headache or focal neurologic deficits suggesting progression of cyst
 - Pre-operative planning

Colloid Cysts

- MRI Brain without contrast (CPT[®] 70551) or MRI Brain without and with contrast (CPT[®] 70553) is indicated for the initial evaluation of colloid cysts if not already completed.
- 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 neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon

Evidence Discussion (HD-34)

- MRI brain is the preferred modality for the evaluation of intracranial cysts, due to its superior soft tissue resolution when compared to CT head.
- Follow up imaging of pineal cysts is supported for new or worsening headaches, focal neurologic deficits, and/or for surgical planning, otherwise, routine follow up is not supported.
- In contrast to pineal cysts, colloid cysts may lead to sudden obstruction of cerebrospinal fluid flow at the foramen of Monro, resulting in neurologic symptoms, including syncope. Other than this scenario, follow up imaging indications are similar to pineal cysts.

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Arachnoid Cysts (HD-35)

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Arachnoid Cysts (HD-35.1)

HD.AR.0035.1.A v1.0.2025

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

- MRI Brain without contrast (CPT[®] 70551) or MRI Brain without and with contrast (CPT[®] 70553) is indicated for initial evaluation of arachnoid cysts if not already completed.
- Repeat MRI Brain is not indicated for most individuals with arachnoid cysts, except in the following scenarios:
 - New or worsening headache or focal neurologic deficits suggesting progression of cyst
 - Pre-operative planning
 - When requested by a neurologist or neurosurgeon or any provider in consultation with a neurologist or neurosurgeon

Evidence Discussion (HD-35)

- MRI brain is the preferred modality for evaluation and follow up of intracranial arachnoid cysts, due to its superior soft tissue resolution when compared to CT head.
- Most intracranial arachnoid cysts remain asymptomatic and follow up imaging is not routinely supported. Surgical intervention is reserved for those with symptoms.

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Sleep-Related Imaging (HD-37)

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General Guidelines Sleep-Related Imaging (HD-37.1)

HD.SL.0037.1.A v1.0.2025

- 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 (pre-treatment 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 <u>Epilepsy and Other Seizure Disorders</u> (HD-9))

Evidence Discussion (HD-37.1)

 Patient management is rarely impacted by structural brain imaging in the evaluation of unexplained hypersomnolence. Instead, a thorough evaluation can result in an accurate diagnosis while safeguarding patients from unnecessary exposure to radiation and over-reliance on incidental imaging findings as potential contributors to the symptom(s). Head Imaging Guidelines

©2024 EviCore by EVERNORTH 400 Buckwalter Place Boulevard, Bluffton, SC 29910 (800) 918-8924 Page 324 of 326 www.EviCore.com The appropriate step in care for patients with disordered sleep is to evaluate their breathing with polysomnography. Advanced imaging can lead to gaps in care and ineffective treatment of disordered sleep patterns. Instead, emphasis should be placed on holistic evaluation, including sleep history and sleep testing. Radiography (X-Rays), 3D Advanced Imaging, or dynamic nasopharyngoscopy are not supported by evidence to being superior over polysomnography at this time.

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Head Imaging Guidelines

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