

AmeriHealth Caritas Louisiana

National Imaging Associates, Inc.*	
Clinical guidelines Original Date: September 1997	
BRAIN (HEAD) MRI	
BRAIN (HEAD) MRI with IAC (Internal Auditory Canal)	
CPT Codes:	Last Revised Date: April 2021
70551, 70552, 70553 – Brain MRI	
70540, 70542, 70543 - IAC	
Guideline Number: NIA_CG_001	Implementation Date: January 2022

INDICATIONS FOR BRAIN MRI

Brain MR/MRA are not approvable simultaneously unless they meet the criteria described below in the Indications for Brain MR/Brain MRA combination studies section.

For evaluation of headache

(ACR, 2019c; Holle, 2013; Quinones-Hinojosa, 2003; Schafer, 2007; Wilbrink, 2009)

- Chronic headache with a change in character/pattern (e.g., more frequent, increased severity, or duration)-
- Cluster headaches or other trigeminal-autonomic cephalgias, i.e., paroxysmal hemicrania, hemicrania continua, short-lasting unilateral neuralgiform headache attacks (SUNCT/SUNA) imaging is indicated once to eliminate secondary causes (IHS, 2018)
- New acute headache, sudden onset:
 - With a personal or family history (brother, sister, parent, or child) of brain aneurysm or AVM (arteriovenous malformation) OR
 - ← < 48 hours of "worst headache in my life" or "thunderclap" headache.
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- Note: The duration of a thunderclap type headache lasts more than 5 minutes. Sudden onset new headache reaching maximum intensity within 2-3 minutes.
- o Prior history of stroke or intracranial bleed
- Known coagulopathy or on anticoagulation
- New onset of headache with any of the following (ACR, 2019c; Micieli, 2020; Mitsikostas, 2016):

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^{1—}Brain (head) MRI

- Acute, new, or fluctuating neurologic deficits, such as sensory deficits, limb weakness, speech difficulties, visual loss*, lack of coordination, or mental status changes or with signs of increased intracranial pressure (papilledema)
 - * Not explained by underlying ocular diagnosis, glaucoma, or macular degeneration

* Not explained by underlying ocular diagnosis, glaucoma, or macular degeneration

Note: Neuroimaging warranted for atypical/complex migraine aura, but not for a typical migraine aura (see background)

- History of cancer or significantly immunocompromised
- o Fever
- Subacute head trauma
- Pregnancy or puerperium (Hamilton, 2020; Shobeiri, 2019)
- o Age > 50
- Severe unilateral headache with radiation to or from the neck, associated with suspicion of carotid or vertebral artery dissection.
- Related to activity or event (sexual activity, exertion, position) (new or progressively worsening)
- Persistent or progressively worsening during a course of physician_directed treatment (ACR, 2019c; Kuruvilla, 2015; Martin, 2011)

Note: Neuroimaging warranted for atypical/complex migraine aura, but not for a typical migraine aura (see background)

- Special considerations in the pediatric population with persistent headache (Trofimova, 2018):
 - Occipital location
 - Age < 6 years
 - Symptoms indicative of <u>increased</u> intracranial pressure, such as recurring headaches after waking with or without associated nausea/vomiting
 - Documented absence of family history of headache
 - Severe headache in a child with an underlying disease that predisposes to intracranial pathology (e.g., immune deficiency, sickle cell disease, neurofibromatosis, history of neoplasm, coagulopathy, hypertension, congenital heart disease)

For evaluation of neurologic symptoms or deficits

(ACR, 2019**2012a**)

- Acute, new, or fluctuating neurologic symptoms or deficits such as, sensory deficits, limb weakness, speech difficulties, visual loss*, lack of coordination, or mental status changes.
 - * Not explained by underlying ocular diagnosis, glaucoma, or macular degeneration

For evaluation of known or suspected stroke or vascular disease

(ACR <u>2012a</u>, 2017<u>a/2012a</u>, 2019; Jauch, 2013)

- Known or suspected stroke with any acute, new, or fluctuating symptoms or deficits such as sensory deficits, limb weakness, speech difficulties, visual loss*, lack of coordination, or mental status changes
 - *Not explained by underlying ocular diagnosis, glaucoma, or macular degeneration

- Suspected stroke with a personal or <u>first-degree</u> family history (brother, sister, parent, or child) of aneurysm or known coagulopathy or on anticoagulation.
- Symptoms of transient ischemic attack (TIA) (episodic neurologic symptoms such as sensory deficits, limb weakness, speech difficulties, visual loss, lack of coordination, or mental status changes)
- (episodic neurologic symptoms)
- Evaluation of suspected acute subarachnoid hemorrhage (SAH)-
- Follow_-up for known hemorrhage, hematoma, or vascular abnormalities.

 Note: MRI is the study of choice for detecting cavernous malformations (CCM). Follow-up imaging of known CCM should be done only to guide treatment decisions or to investigate new symptoms. First-degree relatives of patients with more than one family member with a CCM should have a screening MRI as well as genetic counseling (Akers, 2017; Velez, 2018; Zyck, 2021; Akers, 2017)
- Suspected central venous thrombosis see <u>background</u> (ACR, 2017<u>a</u>, Bushnell, 2014)
- Evaluation of neurological signs or symptoms in sickle cell disease (Mackin, 2014; Thust, 2014)

For evaluation of known or suspected trauma

(ACR, 2019f, Jagoda, 2008; Polinder, 2018)

- Known or suspected trauma or injury to the head with documentation of one or more of the following acute, new, or fluctuating:
 - Focal neurologic findings
 - Motor changes
 - Mental status changes
 - Amnesia
 - Vomiting
 - Seizures
 - Headache
 - Signs of increased intracranial pressure
- Known coagulopathy or on anticoagulation
- Known or suspected skull fracture by physical exam and <u>-or prior imaging positive x-ray.</u>
- Post concussive syndrome if persistent or disabling symptoms and imaging has not been performed
- Subacute or chronic traumatic brain injury with new cognitive and/or neurologic deficit

For evaluation of suspected brain tumor, mass, or metastasis

(Kerjnick, 2008; NCCN, 202019)

- Suspected brain tumor with any acute, new, or fluctuating neurologic symptoms or deficits such as sensory deficits, limb weakness, speech difficulties, visual loss*, lack of coordination, or mental status changes-
 - * Not explained by underlying ocular diagnosis, glaucoma, or macular degeneration
- Suspected brain metastasis or intracranial involvement in patients with a history of cancer based on neurological symptoms or examination findings (may include new or changing lymph nodes)

 Langerhans cell histiocytosis with visual, neurological, or endocrine abnormality; polyuria or polydipsia; suspected craniofacial bone lesions, aural discharge, or suspected hearing impairment/mastoid involvement (Haupt, 2013; NCCN, 2020)

Suspected Pituitary Tumors

(ACR, 2018; GHRS, 2000; Kannan, 2013; Majumdar, 2013); Shalender, 2018)

- With the following:
 - Neurologic findings (e.g., visual field deficit suggesting compression of the optic chiasm, diplopia, gaze palsy)
 - Suspected hypofunctioning pituitary gland based on hormonal testing, e.g., hypo
 pituitarism, growth hormone deficiency, hypogonadotropic hypogonadism [i.e., low
 gonadotropins (FSH/LH) and sex hormones*]
 - * Severe secondary hypogonadism with total testosterone persistently < 150 and low or normal LH/FSH OR
 - * Testosterone levels below normal range with low or normal LH/FSH; AND
 - Neurological signs- and and-symptoms; OR
 - Other pituitary hormonal abnormalities; OR
 - Consideration of reversible functional causes of gonadotropin suppression (e.g., obesity, opioid use, or comorbid illness)
 - Suspected hyperfunctioning pituitary gland based on hormonal testing, i.e., central hyperthyroidism (high TSH), Cushing disease (high ACTH), acromegaly/gigantism (high GH/IGF-1) or elevated prolactin (≥ 250 ng/mL or persistently elevated in the absence of another cause, e.g., stress, pregnancy, hypothyroidism, medication)
 - Central Diabetes Insipidus (low ADH)
 - Precocious puberty in a child (male < 9; female < 8), with hormonal studies suggesting a central cause and evidence of an accelerated bone age on xx-ray (Faizah, 2012)
 - o Pituitary apoplexy with sudden onset of neurological and hormonal symptoms

For evaluation of known brain tumor, mass, or metastasis

- Follow--up of known malignant brain tumor
- Suspected recurrence with prior history of CNS cancer based on neurological symptoms or examination findings
- Patient with history of CNS cancer (either primary or secondary) and a recent course of chemotherapy, radiation therapy (to the brain), or surgical treatment within the last two (2) years (NCCN, 2020)
- Follow_-up of known non-malignant brain tumor/lesion if symptomatic, new/changing signs or symptoms or complicating factors
- Follow--up of known meningioma (NHS, 2018)
 - o If <2cm or heavily calcified at 2 years and 5 years
 - > 2cm annually for 3 years and then scans at 5 years and 10 years-
 - Multiple meningiomas, annually
 - After treatment (surgery or radiotherapy), post-operative if concern for residual tumor, every 6-12 months, then annually for 3-5 years based on WHO Grade (see <u>background</u>)

- Follow-up of known of-pituitary adenoma
 - New neuroendocrine signs or symptoms
 - Functioning adenoma to assess response to treatment and 1-year follow-up after drug holiday (Stoller, 20152)
 - Asymptomatic Macroadenoma (≥ 10mm) follow_up every 6-18 months, post-surgical follow_up 1-2 years after surgery (Dekkers, 2008)
 - Asymptomatic, non-functioning Microadenoma < 10mm repeat in one year; if stable, repeat every 2-3 years (Lake, 2013)
- Tumor evaluation and monitoring in neurocutaneous syndromes—see background
- Follow<u>-up</u> of known pineal cyst (≥ 5mm) if there are atypical features or symptoms (e.g., headaches, gaze paresis, ataxia, papilledema, nausea/vomiting) (Cauley, 2009; Jussila 2017)
- Follow-up of known arachnoid cyst (Al-Holou, 2010, £2013; Mustansir, 2018)
 - < 4 years old, serial imaging is warranted</p>
 - > 4 years old, repeat imaging only if <u>newly</u> symptomatic, i.e., headaches, increased intracranial pressure, hydrocephalus, local mass effect, seizures, visual/endocrine dysfunction.
- Tumor evaluation and monitoring in neurocutaneous syndromes see background
- Langerhans cell histiocytosis (Haupt, 2013, NCCN, 2020)
 - To assess treatment response and surveillance of known brain lesions

For screening for known Non-CNS Cancer - see bBackground (NCCN, 2020)

- Default screening for
 - Kidney cancer
 - Lung cancer
 - Merkel cell carcinoma
 - Mucosal melanoma of the head and neck, especially of the oral cavity
 - Poorly differential neuroendocrine cancer (Large or Small cell/Unknown primary of neuroendocrine origin)
- Screening with preconditions
 - AML......Suspicion of leukemic meningitis
 - Cutaneous melanoma...... Stage IIIC or higher
 - o Testicular cancer-Seminoma...... High risk
 - o Gestational Trophoblastic Neoplasia..... Pulmonary metastasis
 - o Bladder cancer...... High risk, i.e., small cell
- All other cancer if CNS symptoms present

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For screening of Hereditary Cancer Syndromes

- Li Fraumeni syndrome- Annually (Cureus Kumar, 2018)
- Von Hippel Lindau Every 2 years, starting at age of 8 years (Rednam, 2017).
- Tuberous Sclerosis Every 1-3 years, until the age of 25 years (Krueeuger, 2013)

- MEN1 Every 3-5 years, starting at the age of 5 years (Brandi, 2001)
- NF-2- Brain IAC: Annually starting at the, from age of 10 years (Evans, 2017)
- Sturge Weber Syndrome: Once, after age 1 to rule out intracranial involvement after; in patients <1 year, only if symptomatic (Comi, 2011).

Indications for combination studies for the initial pre-therapy staging of cancer, OR active monitoring for recurrence as clinically indicated, OR evaluation of suspected metastases (NCCN, 202019)

 ≤ 5 concurrent studies to include CT or MRI of any of the following areas as appropriate depending on the cancer: Neck, Abdomen, Pelvis, Chest, Brain, Cervical Spine, Thoracic Spine or Lumbar Spine,

For evaluation of known or suspected seizure disorder

(ACR, 2019d; Cendes, 2016; Gaillard, 2009; Ho, 2013; Krumholz, 2007; Ramli, 2015)

- New onset of an unprovoked seizure in adults
- Newly identified change in seizure activity/pattern
- Known seizure disorder without previous imaging
- Medically refractory epilepsy
- Imaging indications for new onset seizures in the pediatric population (Hirtz, 2000; Kimia, 2012; Sadeq, 2016; ShaikhSahikj, 2019)
 - o Abnormal neurological exam, especially a postictal focal deficit
 - Significant developmental delay
 - Focal onset
 - EEG shows focal or suspected structural abnormalities
 - <1 year of age</p>

Note: Imaging is not indicated in simple febrile seizures

For evaluation of suspected multiple sclerosis (MS)

(CMSC, 2018; Thompson, 2017; Traboulsee, 2016)

- For evaluation of patient with neurologic symptoms or deficits suspicious for MS with
 - A clinically isolated syndrome (optic neuritis, transverse myelitis, or brain stem syndrome);
 OR
 - Recurrent episodes of variable neurological signs or symptoms not attributable to another cause
- To demonstrate dissemination in time for diagnosis (6-12 months for high risk, 12-24 months for low risk)

For evaluation of known multiple sclerosis (MS)

(CMSC, 2018)

- To establish a new baseline (no recent imaging, postpartum, or 6-12 months after switching disease modifying therapy)
- Prior to starting or switching disease-modifying therapy
- Every 1-2 years while on disease-modifying therapy to assess for subclinical disease activity, less frequently when stable for 2-3 years
- New signs or symptoms suggested of an exacerbation or unexpected clinical worsening

- Progressive Multifocal Leukoencephalopathy (PML) surveillance for patients on natalizumab (Tsyabri) (McGuigan, 2016)
 - 12 months after the start of treatment in all patients
 - Further surveillance MRI scanning timing is based on anti-JCV antibody status
 - If anti-JCV antibody negative, annually
 - If anti-JCV antibody positive and antibody index < 1.5. every 6 months</p>
 - If anti-JCV antibody positive and antibody index > 1.5, every 3-4 months

For evaluation of known or suspected infectious or inflammatory disease (e.g., meningitis or abscess) (Lummel, 2016; Oliveira, 2014)

- Suspected intracranial abscess or brain infection with acute altered mental status or with positive lab findings (such as elevated WBC's) OR follow_up assessment during or after treatment completed.
- Meningitis with positive signs and symptoms (such as fever, headache, mental status changes, stiff neck) OR with positive lab findings (such as elevated white blood cells or abnormal lumbar puncture fluid exam.)
- Suspected <u>e</u>Encephalitis with headache and altered mental status or follow-up as clinically warranted
- Endocarditis with suspected septic emboli-
- Suspected temporal arteritis in a patient ≥ 50 with temporal headache, abrupt visual changes, jaw claudication, temporal artery tenderness, constitutional symptoms or elevated ESR (D'Souza, 2016; Diamantopoulos, 2014; <u>D'Souza, 2016</u>; Klink, 2014; Salehi, 2016; <u>Yip 2020</u>); <u>AND</u>
 - Negative initial work-up (color Doppler ultrasonography or biopsy); OR
 - Atypical features, or failure to response to treatment with or concern for large vessel involvement intracranial involvement

Note: Protocol should include high-resolution contrast-enhanced imaging the temporal artery

- Central Nervous System (CNS) involvement in patients with known or suspected vasculitis or autoimmune disease with abnormal inflammatory markers or autoimmune antibodies
- Suspected primary CNS vasculitis based on neurological signs and symptoms with completed infectious/inflammatory lab work-up (Godasi, 2019; Zuccoli, 2011)-
- Central Nervous System (CNS) involvement in patients with known or suspected vasculitis or autoimmune disease with positive lab findings.
- Immunocompromised patient (e.g., transplant recipients, HIV with CD4<200, primary immunodeficiency syndromes, hematologic malignancies) with focal neurologic_-symptoms, headaches, behavioral, cognitive or personality changes-

For evaluation of clinical assessment documenting cognitive impairment of unclear cause (Harvey 2012; HQO, 2014; Narayanan, 2016)

Mental status score of either MMSE or MoCA of less than 26 or other similar mental status instruments*-/formal neuropsychological testing showing at least mild cognitive impairment AND a completed basic metabolic workup (such as thyroid function testing, liver function testing, complete blood count, electrolytes, and B12)-

*Other examples include: Ottawa 3DY (O3DY), Brief Alzheimer's Screen (BAS), Blessed Dementia Scale (BDS), caregiver-completed AD8 (cAD8), Brief Cognitive Rating Scale (BCRS), Clinical Dementia Rating (CDR) (Carpenter, 2011; McDougall, 1990)

FDA labeling for the drug Aduhelm (for Alzheimer's disease) requires baseline imaging and monitoring with Brain MRI. Criteria for coverage includes the following:

- o Baseline study within 1 year of initiating treatment
- o Prior to the 7th and 12th infusions
- o Monitoring if radiographic severe Amyloid Related Imaging Abnormalities (ARIA) is observed

For evaluation of movement disorders

(ACR, 2019a 2019e; Albanese, 2011; Mascalchi, 2012; McFarland, 2014; Pyatigorskaya, 2014; Sharifi, 2014)

- For evaluation of suspected Parkinson's with atypical feature or unresponsive to levodopa
- For evaluation of new non-Parkinson symptoms in known Parkinson's disease complicating the evaluation of the current condition.
- For the evaluation of other movement disorder to exclude a structural lesion (i.e., suspected Huntington disease, chorea, atypical parkinsonian syndromes, hemiballismus, atypical dystonia).

 Note: MRI not indicated in essential tremor or isolated focal dystonia (e.g., blepharospasm, cervical dystonia, laryngeal dystonia, oromandibular dystonia, writer's dystonia) (Alabanese, 2011; Comella, 2019; Sharfi, 2014)

For evaluation of cranial nerve and visual abnormalities

- Anosmia (loss of smell) or dysosmia documented by objective testing that is persistent and of unknown origin (Decker, 2013; Policeni, 2017; Rouby, 2011, Decker, 2013)
- Optic neuritis
- Abnormal eye findings on physical or neurologic examination (papilledema, pathologic nystagmus, optic atrophy, ocular nerve palsies, new onset anisocoria, visual field deficit, etc.) (Chang, 2019)
 Note: Not explained by underlying ocular diagnosis, glaucoma, or macular degeneration
- Binocular diplopia with concern for intracranial pathology (Iliescu, 2017)
- <u>Childhood strabismus with development delay or abnormal fundoscopic exam to rule out intracranial abnormalities (Kadom, 2008; Yoon, 2019; Kadom, 2008)</u>
- Horner's syndrome with symptoms localizing the lesion to the central nervous system (Lee, 2007).
- Trigeminal nNeuralgia or other trigeminal autonomic cephalgias if <40 years of age or atypical features (ie bilateral, hearing loss, dizziness/vertigo, visual changes, sensory loss, numbness, pain >2min, pain outside trigeminal nerve distribution, progression), notably in those with atypical presentation (Wilbrink, 2009; Bendtsen, 2019; Cruccu, 2016; Wilbrink, 2009)Cruccu, 2016)
- Bell's Palsy- if atypical signs, slow resolution beyond three weeks, no improvement at four months, or facial twitching/spasms prior to onset (Quesnel, 2010)
- Hemifacial spasm (Hermier, 2019)
- Other objective cranial nerve palsy (CN IX-XII) (ACR, 2017b; Mumtaz, 2014; Policeni, 2017)

- Bulbar symptoms, i.e., difficulty in chewing, weakness of the facial muscles, dysarthria, palatal weakness, dysphagia, and dysphonia and/or signs, i.e., atrophy and fasciculations of the tongue and absent gag reflex (Yedavelli, 2018)
- Pseudobulbar symptoms, i.e., dysphagia, dysarthria, facial weakness, sudden, stereotyped emotional outbursts that are not reflective of mood and/or signs, i.e., spastic tongue and exaggerated gag/jaw jerk (King, 2013)

For evaluation of known or suspected congenital abnormality (such as hydrocephalus, craniosynostosis, neural tube defects)

(Ashwal, 2009; Vinocur, 2010)

 Known or suspected congenital abnormality with any acute, new, or fluctuating neurologic, motor, or mental status changes.

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- Evaluation of macrocephaly in an infant/child <-with 18 with previously abnormal US, abnormal neurodevelopmental examination (Tan, 2018), signs of increased ICP or closed anterior fontanelle-
- Evaluation of microcephaly in an infant/child < 18 microcephaly.
- Evaluation of craniosynostosis and other skull deformities. CT is preferred imaging to assess bony structures; MRI imaging is preferred to assess intracranial soft tissue
- Evaluation of the corticomedullary junction in Achondroplasia (Dougherty, 2018; Kubota, 20201))
- Prior treatment OR treatment planned for congenital abnormality-

Note: For

evaluation of known or suspected hydrocephalus please see section on CSF abnormalities.

Cerebral Spinal Fluid (CSF) Abnormalities

- <u>Suspected or known hydrocephalus*</u> Evaluation of suspected hydrocephalus with any acute, new, or fluctuating neurologic, motor, or mental status changes
 <u>Suspected or kKnown hydrocephalus*</u>
- Initial imaging of For initial evaluation of a suspected or known. Arnold Chiari malformation **
- Follow--up imaging of a known type II or type III Arnold Chiari malformation ** (II/III). For Arnold Chiari type I, I-follow-up imaging only if new or changing signs/symptoms (Whitson, 2015)
- Initial evaluation for a known syrinx or syringomyelia
- Known or suspected normal pressure hydrocephalus (NPH) (Damasceno, 2015)
 - With symptoms of gait difficulty, cognitive disturbance, and urinary incontinence
- Follow--up shunt evaluation (Kamenova, 2018; Pople, 2002, Reddy, 2014; Wetzel, 2018)
 - Post operativity if indicated based on underlying disease <u>orand</u> pre-operative radiographic findings and/or
 - o 6-12 months after placement and/or
 - With neurologic symptoms that suggest shunt malfunction
- Evaluation of known or suspected cerebrospinal fluid (CSF) leakage (Severson, 2019)

- <u>Cisternography for intermittent and complex CSF rhinorrhea/otorrhea. CSF fluid should always</u>
 <u>be confirmed with laboratory testing (Beta-2 transferrin assay)</u> <u>-(Selcuk, 2010;</u>
 <u>KułakowskaMantur, 2011; Selcuk, 2010)</u>
- Suspected spontaneous intra-cranial hypotension with distinct postural headache (o→ther symptoms include; nausea, vomiting, dizziness, tinnitus, diplopia neck pain or imbalance) (Gordon 20049; NORD, 2017).
- CSF flow study for evaluation and management of CSF flow disorders (Bradley, 2016; Mohammad, 2019)
 - <u>*</u>*Often congenital, but can present later in life; or less commonly acquired secondary to tumor, stroke, trauma, infection, etc. (NORD, 2014)

Pre-operative/procedural evaluation for brain/skull surgery

Pre-operative evaluation for a planned surgery or procedure Pre-operative evaluation for a planned surgery or procedure if the imaging provides diagnostic information that is not available on prior studies (provider should be referred to the health plan for nondiagnostic surgical planning studies)

Post-operative/procedural evaluation

A follow-up study may be needed to help evaluate a patient's progress after treatment, procedure, intervention, or surgery. Documentation requires a medical reason that clearly indicates why additional imaging is needed for the type and area(s) requested.

Other Indications for a Brain MRI

- Vertigo associated with any of the following (<u>Kattah, 2009</u>; Welgampola, 2019; Yamada, 2019
 Kattah, 2009)
 - Signs or symptoms suggestive of a CNS lesion (ataxia, visual loss, double vision, weakness, or a change in sensation)
 - Progressive unilateral hearing loss
 - o Risk factors for cerebrovascular disease with concern for stroke
 - After full neurologic examination and vestibular testing with concern for central vertigo
 (i.e., skew deviation, vertical nystagmus, head thrust test, videonystagmography (VNG)/
 electronystagmography (ENG))

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- <u>Diagnosis of central sleep apnea on polysomnogram</u> <u>Diagnosis of central sleep apnea on polysomnogram</u>
 - Children > 1 year (Felix, 2016)
 - Adults in the absence of heart failure, chronic opioid use, high altitude, or treatment emergent central sleep apnea AND concern for a central neurological cause (Chiari malformation, tumor, infectious/inflammatory disease) OR with an abnormal neurological exam (Malhotra, 2010)
- Syncope with clinical concern for seizure or associated neurological signs or symptoms (Al-Nsoor, 2010; Strickberger, 2006)

- Cyclical vomiting syndrome or abdominal migraine with any localizing neurological symptoms (Angus-Leppan, 2018; Li-BUK, 2018; Thangam Venkatesan, 2019)
- Soft tissue mass of the head with nondiagnostic initial evaluation (ultrasound and/or radiograph) (ACR, 2017c; Kim, 2019; Zhang, 2018)
- Psychological changes with neurological deficits on exam or after completion of a full neurological assessment that suggests a possible neurologic cause (ACR, 2019b)
- Global developmental delay or developmental delay with abnormal neurological examination in a child < 18 years (Ali, 2015; Momen, 2011)
- Cerebral palsy if etiology has not been established <u>in</u> the neonatal period, there is change in the
 expected clinical or developmental profile or concern for progressive neurological disorder
 (Ashwal, 2004; NICE, 2020)
- Unexplained event (BRUE) formerly apparent life-threatening event (ALTE) in infants < 1 year with concern for neurological cause based on history and exam (Tieder, 2016)
 Note: Imaging is not indicated in low--risk patients

Indications for a Brain MRI with Internal Auditory Canal (IAC)

- Unilateral non-pulsatile tinnitus-
- Pulsatile tinnitus-
- Suspected acoustic neuroma (Schwannoma) or cerebellar pontine angle tumor with any of the following signs and symptoms: unilateral hearing loss by audiometry, headache, disturbed balance or gait, unilateral tinnitus, facial weakness, or altered sense of taste-
- Suspected cholesteatoma-
- Suspected glomus tumor-
- Asymmetric sensorineural hearing loss on audiogram-
- CSF otorrhea (MRI for intermittent leak, CT for active leaks) (Hiremath, 2019) CSF fluid should always be confirmed with laboratory testing (Beta-2 transferrin assay).
- Clinical <u>s</u>suspicion of acute mastoiditis as a complication of acute otitis media with intracranial complications (i.e., meningeal signs, cranial nerve deficits, focal neurological findings, altered mental status) (Patel, 2014; Platzek, 2014)
- Bell's Palsy for evaluation of the extracranial nerve course -if atypical signs, slow resolution beyond three weeks, no improvement at four months, or facial twitching/spasms prior to onset (Quesnel, 2010)

Indications for Combination Studies

(ACR, 2017<u>a</u>, 2019<u>a</u>)

• For approved indications as noted above and being performed in a child under 8 years of age who will need anesthesia for the procedure and there is a suspicion of concurrent intracranial pathology (Lawson, 2000).

Brain MRI/Neck MRA

- Recent ischemic stroke or transient ischemic attack
- Suspected carotid or vertebral artery dissection with focal or lateralizing neurological deficits

• Brain MRI/Brain MRA

- o Recent ischemic stroke or transient ischemic attack
- Thunderclap headache with continued concern for underlying vascular abnormality after initial negative work-up (Whitehead, 2019, Yeh, 2010, Yuan, 2005):
 - Negative Brain CT; AND
 - Negative Lumbar Puncture
- Acute, sudden onset of headache with personal history of a vascular abnormality or firstdegree family history of aneurysm
- Headache associated with exercise or sexual activity (IHS, 2018)

Suspected venous thrombosis (dural sinus thrombosis) – Brain MRV see <u>background</u>

Brain MRI/Brain MRA/Neck MRA

- Recent stroke or transient ischemic attack (TIA)
- Suspected carotid or vertebral artery dissection with focal or lateralizing neurological deficits
- Approved indications as noted above and being performed in a child under 8 years of age
 who will need anesthesia for the procedure and there is a suspicion of concurrent vascular
 and intracranial pathology (Lawson, 2000).

Brain MRI/Cervical MRI

- O For evaluation of Arnold Chiari Malformation.
- Suspected MS with new or changing symptoms consistent with cervical spinal cord disease.
- For follow-up of known multiple sclerosis (MS)
- Follow up to the initiation or change in medication for patient with known Multiple Sclerosis

Brain MRI/ Cervical MRI/Thoracic MRI (any combination)

- For evaluation of neuromyelitis optica spectrum disorders (recurrent or bilateral optic neuritis; recurrent transverse myelitis) (Wingerchuk, 2015)
- For known MS, prior to the initiation or change of disease modification treatments and assess disease burden (to establish a new baseline)
- Follow-up scans for known MS if patients have known spine disease (Kaunzner, 2017)
 - 6-12 months after starting/changing treatment
 - Every 1-2 years while on disease-modifying therapy to assess for subclinical disease activity, less frequently when stable for 2-3 years

• Brain MRI/ Cervical MRI/Thoracic MRI/Lumbar (any combination)

- Follow-up imaging of a known type II or type III Arnold Chiari malformation[†]. For Arnold Chiari type I, follow-up imaging only if new or changing signs/symptoms Follow-up imaging of a known Arnold Chiari malformation (II/III). For Chiari, I follow-up imaging only if new or changing signs/symptoms (Radic, 2018; Whitson, 2015; Radic, 2018)
- Suspected Leptomeningeal carcinomatosis (see background) -(Shah, 2011)
- Tumor evaluation and monitoring in neurocutaneous syndromes See bBackground

 CSF leak highly suspected and supported by patient history and/or physical exam findings (known or suspected spontaneous (idiopathic) intracranial hypotension (SIH), post lumbar puncture headache, post spinal surgery headache, orthostatic headache, rhinorrhea or otorrhea, or cerebrospinal-venous fistula)

• Brain MRI/Orbit MRI

- Optic neuropathy or unilateral optic disk swelling Unilateral papilledema optic disk swelling/optic neuropathy of unclear etiology to distinguish between a compressive lesion of the optic nerve, optic neuritis, ischemic optic neuropathy (arteritic or non-arteritic), central retinal vein occlusion or optic nerve infitrative disorders (Behbehani, 2007)
- Bilateral optic disk swelling (papilledema) with visual loss (Margolin, 2019)
- Optic Neuritis
 - -ilf atypical presentation, severe visual impairment, or poor recovery following initial onset or treatment onset (CMSC, 2018)
 - If needed to confirm optic neuritis and rule out compressive lesions
- Known or suspected neuromyelitis optica spectrum disorder with severe, recurrent, or bilateral optic neuritis (Wingerchuk, 2015)

• Brain MRI/FACE/SINUS/NECK MRI

Anosmia Anosmia or dysosmia on objective testing that is persistent and of unknown origin (Decker, 2013; Policeni, 2017; Zaghouani, 2013), Decker 2013)
 Granulomatosis with polyangiitis (Wegener's granulomatosis) disease (Pakalniskis, 2015)

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- Trigeminal Neuralgia or other trigeminal autonomic cephalgias, notably in those with atypical presentation Trigeminal neuralgia that meets the above criteria (Hughes, 2016; Policeni, 2017)
- Bells/hemifacial spasm that meets above criteria
- Objective cranial nerve palsy (CN IX-XII) (for evaluation of the extracranial nerve course)
 (Mumtaz, 2014; Policeni, 2017)
- Granulomatosis with polyangiitis (Wegener's granulomatosis) disease (Pakalniskis, 2015)

BACKGROUND

Brain (head) MRI is the procedure of choice for most brain disorders. It provides clear images of the brainstem and posterior brain, which are difficult to view on a CT scan. It is also useful for the diagnosis of demyelinating disorders (disorders-such as multiple sclerosis (MS) that cause destruction of the myelin sheath of the nerve). The evaluation of blood flow and the flow of cerebrospinal fluid (CSF) is possible with this non-invasive procedure.

MRI for Headache – Generally, magnetic resonance imaging is the preferred imaging technique for evaluating the brain parenchyma, and CT is preferable for evaluating subarachnoid hemorrhage. CT is faster and more readily available than MRI and is often used in urgent clinical situations. Neurologic

imaging is warranted in patients with headache disorders along with abnormal neurologic examination results or predisposing factors for brain pathology. Contrast—enhanced MRI is performed for evaluation of inflammatory, infectious, neoplastic, and demyelinating conditions.

Headache timeframes and other characteristics – Generally, acute headaches are present from hours to days, subacute from days to weeks and chronic headaches for more than 3 months. Acute severe headaches are more likely to be pathological (e.g., SAH, cerebral venous thrombosis) than non-acute (e.g., migraine, tension-type). Headaches can also be categorized as new onset or chronic/recurrent. Non-acute new onset headaches do not require imaginged unless there is a red flag as delineated above. Incidental findings lead to additional medical procedures and expense that do not improve patient well-being. Primary headache syndromes, such as migraine and tension headaches, are often episodic with persistent or progressive headache not responding to treatment requiring further investigation (e.g., new daily persistent headache). Imaging is indicated in chronic headaches if there is a change in the headache frequency (number of headaches episodes/month), duration of each episode, severity of the headaches or new characteristics, such as changing aura or associated symptoms (ACR, 2012b2019c; HISIHS, 2018; Jang, 2019; Spierings, 2003; Tyagi, 2012)

Migraine with aura

(Hadjikhani, 2019; IHS, 2018; Micieli, 2020) -- }

The headache phase of a migraine is preceded and/or accompanied by transient neurological symptoms referred to as aura in at least a third of migraine attacks. The most common aura consists of positive and/or negative visual phenomena, present in up to 99% of the patients. Somatosensory is the secondary most common type of aura (mostly paraesthesias in an upper limb and/or hemiface). Language/speech (mainly paraphasia and anomic aphasia) can also be affected. These neurological symptoms typically evolve over a period of minutes and may last up to 20 minutes or more. The gradual evolution of symptoms is thought to reflect spreading of a neurological event across the visual and somatosensory cortices. Characteristically, the aura usually precedes and terminates prior to headache, usually within 60 minutes. In others, it may persist or begin during the headache phase. ICHD-3 definition of the aura of migraine with typical aura consists of visual and/or sensory and/or speech/language symptoms, but no motor, brainstem or retinal symptoms and is characterized by gradual development, duration of each symptom no longer than one hour, a mix of positive and negative features and complete reversibility. Atypical or complex aura includes motor, brainstem, monocular visual disturbances, or ocular cranial nerve involvement (hemiplegic migraine, basilar migraine/brainstem aura, retinal migraine, ophthalmoplegic migraine) and secondary causes need to be excluded. Additional features of an aura that raise concern for an underlying vascular etiology include: late age of onset, short duration, evolution of the focal symptoms, negative rather than positive visual phenomenon, <u>and</u> history of vascular risk factors.

Table 1: Gait and brain imaging[‡]

<u>Gait</u>	<u>Characteristic</u>	Work up/Imaging
<u>Hemiparetic</u>	Spastic unilateral, circumduction	Brain and/or, Cervical spine imaging
		based on associated symptoms

<u>Diplegic</u>	Spastic bilateral, circumduction	Brain, Cervical and Thoracic Spine imaging
Myelopathic	Wide based, stiff, unsteady	Cervical and/or Thoracic spine MRI based on associated symptoms
<u>Ataxic</u>	Broad based, clumsy, staggering, lack of coordination, usually also with limb ataxia	Brain imaging
<u>Apraxic</u>	Magnetic, shuffling, difficulty initiating	Brain imaging
<u>Parkinsonian</u>	Stooped, small steps, rigid, turning en bloc, decreased arm swing	Brain Imaging
Choreiform	Irregular, jerky, involuntary movements	Medication review, consider brain imaging as per movement disorder Brain MR guidelines
Sensory ataxic	Cautious, stomping, worsening without visual input (ie + Romberg)	EMG, blood work, consider spinal (cervical or thoracic cord imaging) imaging based on EMG
Neurogenic	Steppage, dragging of toes	EMG, if there is foot drop, Lumbar spine MRI Pelvis MR appropriate evidence of plexopathy
<u>Vestibular</u>	Insecure, veer to one side, worse when eyes closed, vertigo	Consider Brain/IAC MRI as per GL

(*References: Chhetri, 2014; Clinch, 2021; Gait, 2021; Haynes, 2018; Marshall, 2012; Pirker, 2017)

Gait and brain imaging:

— Gait	<u>Characteristic</u>	
- Hemiparetic	— Spastic unilateral,	Brain and/or, Cervical spine imaging based on
	<u>circumduction</u>	associated symptoms
- Diplegic	Spastic bilateral,	Brain, Cervical and Thoracic Spine imaging
	<u>circumduction</u>	
	Wide based, stiff, unsteady	Cervical and/or Thoracic spine MRI based on
		associated symptoms
- Ataxic	Broad based, clumsy,	Brain imaging
	staggering, lack of	
	coordination, usually also with	
	<u>limb ataxia</u>	
<u>Apraxic</u>	Magnetic, shuffling, difficulty	Brain imaging
	<u>initiating</u>	
—— <u>Parkinsonian</u>	Stooped, small steps, rigid,	—Brain Imaging
	turning en bloc, decreased	
	arm swing	

<u>Choreiform</u>	Irregular, jerky, involuntary	Medication review, consider brain imaging as per
	movements	movement disorder Brain MR guidelines
Sensory ataxic	Cautious, stomping, worsening	EMG, blood work, consider spinal (cervical or
	without visual input (ie +	thoracic cord imaging) imaging based on EMG
	Romberg)	
Neurogenic	Steppage, dragging of toes	EMG, if there is foot drop Lumbar spine MR
		Pelvis MR appropriate if evidence of plexopathy
	Insecure, veer to one side,	Consider Brain/IAC MRI as per above GL
	worse when eyes closed,	
	vertigo	

Non-neurological causes of gait dysfunction include: pain (antalgic), side effects of drugs (analgesic, antihistamines, benzos, psych meds, antihypertensives), visual loss, hearing impairment, orthopedic disorders, rheumatologic disorders, psychogenic, and cardiorespiratory problems (orthostasis) (Foster 2021; Haynes, 2018; Marshall, 2012; Pirker 2017, Foster 2021, Haynes, 2018).

MRI and recent stroke or transient ischemic attack - k

A stroke or central nervous system infarction is defined as "brain, spinal cord, or retinal cell death attributable to ischemia, based on neuropathological, neuroimaging, and/or clinical evidence of permanent injury. ... Ischemic stroke specifically refers to central nervous system infarction accompanied by overt symptoms, whereas silent infarction causes no known symptoms" (Sacco, 2013). If imaging or pathology is not available, a clinical stroke is diagnosed by symptoms persisting for more than 24 hours. Ischemic stroke can be further classified by the type and location of ischemia and the presumed etiology of the brain injury. These include large-artery atherosclerotic occlusion (extracranial or intracranial), cardiac embolism, small-vessel disease and less commonly dissection, hypercoagulable states, sickle cell disease and undetermined causes (Kernan, 2014). TIAs in contrast, "are a brief episode of neurological dysfunction caused by focal brain or retinal ischemia, with clinical symptoms typically lasting less than one hour, and without evidence of acute infarction on imaging" (Easton, 2009). On average, the annual risk of future ischemic stroke after a TIA or initial ischemic stroke is 3–4%, with an incidence as high as 11% over the next 7 days and 24–29% over the following 5 years. This has significantly decreased in the last half century due to advances in secondary prevention (Hong, 2011).

Therefore, when revascularization therapy is not indicated or available in patients with an ischemic stroke or TIA, the focus of the work-up is on secondary prevention. This includes noninvasive vascular imaging to identify the underlying etiology, assess immediate complications and risk of future stroke. The majority of stroke evaluations take place in the inpatient setting. Admitting TIA patients is reasonable if they present within 72 hours and have an ABCD(2) score ≥3, indicating high risk of early recurrence, or the evaluation cannot be rapidly completed on an outpatient basis ((Easton, 2009). Minimally, both stroke and TIA should have an evaluation for high-risk modifiable factors, such as carotid stenosis atrial fibrillation as the cause of ischemic symptoms (Kernan, 2014). Diagnostic recommendations include÷ neuroimaging evaluation as soon as possible, preferably with magnetic resonance imaging, including DWI; noninvasive imaging of the extracranial vessels should be performed, and noninvasive imaging of intracranial vessels is reasonable (Wintermark, 2013).

Patients with a history of stroke and recent work—up with new signs or symptoms indicating progression or complications of the initial CVA should have repeat brain imaging as an initial study. Patients with remote or silent strokes discovered on imaging should be evaluated for high-risk modifiable risk factors based on the location and type of the presumed etiology of the brain injury.

MRI and Central Venous Thrombosis – a MV MR Venogram is indicated for the definite evaluation of a central venous thrombosis/dural sinus thrombosis. The most frequent presentations are isolated headache, intracranial hypertension syndrome, seizures, focal neurological deficits, and encephalopathy. Risk factors are hypercoagulable states inducing genetic prothrombotic conditions, antiphospholipid syndrome and other acquired prothrombotic diseases (such as cancer), oral contraceptives, pregnancy, puerperium (6-weeks postpartum), infections, and trauma. COVID-19 infection is associated with hypercoagulability, and a thromboinflammatory response, and an increased incidence of venous thromboembolic events (VTE) (Connors, 2020; Tu, 2020). Since venous thrombosis can cause SAH, infarctions, and hemorrhage, parenchymal imaging with MRI/CT is also appropriate (Bushnell, 2014; Courinho, 2015; Ferro, 2016).

MRI and benign tumors (e.g., schwannomas, choroid plexus papilloma, pineocytoma, gangliocytoma) A single follow_-up study is appropriate after the initial diagnosis to ensure stability. Follow_-up of known benign tumor <u>is indicated</u> if symptomatic, new/changing signs or symptoms, or complicating factors (Gupta, 2017). In neurocutaneous and hereditary cancer syndromes, follow-up surveillance may also be indicated (see below).

Galactorrhea and MRI - Imaging is not indicated in isolated galactorrhea without elevated prolactin (normoprolactinemic) (Atluri, 2018; Huang, 2012); Sridevi, 2018).

MRI and Meningioma (NHS, 2018) – For incidental meningiomas, most patients who progressed did so within 5 years of diagnosis (Islim, 2019). Small (<2cm) meningiomas rarely grow sufficiently to produce symptoms within 5 years. Heavily calcified meningiomas rarely grow. Patients with multiple meningiomas should have annual scans indefinitely, despite treatment, because of the possibility of further meningiomas developing.

For surveillance post--treatment:

- Solitary convexity WHO Grade 1 meningiomas MRI scan at 2½ years post-operatively.
- Solitary skull base or falcine origin WHO Grade 1 meningiomas- MRI scans at 1 year, 2 years, 3½ years and 5 years post-operatively. If a recurrence is detected, continue annual scans.
- WHO Grade 2 meningiomas- MRI scan at 6 months, 1 <u>year</u> then annually to 5 years. If a recurrence is detected, continue annual scans.
- WHO Grade 3 meningiomas 6-monthly MRI scans for 3 years, then annual scans to 5 years. If a recurrence is detected, continue annual scans.
- Patients who have had radiosurgery, including those being treated for a recurrence, should have scans at 6 months, then annually for 3 years, a scan at 5 years and a final scan at 10 years.

Table 2: MRI and staging screening in Non-CNS Cancers (NCCN, 2020)

(NONBRAIN/CNS) CANCER	PRECONDITION
Cutaneous melanoma	Stage IIIC or higher, default staging screening ≥ stage IIIC, surveillance with periodic brain MRI up to 3 years even if asymptomatic without prior brain mets; and if prior brain mets, surveillance every 3-6 months up to 3 years
Testicular cancer-Seminoma	If high risk, such as beta HCG >5000IU/L, or multiple lung or visceral mets, choriocarcinoma, neurological symptoms, or AFP>10,000ng/ml
Merkel cell carcinoma	Default staging screening, but especially for high risk (>stage IIIb, immunosuppression)
Lung cancer	Default staging screening brain MRI also for surveillance in small cell every 3 months for 2 years if they have had no prophylactic cranial radiation

MRI and Neurocutaneous Syndromes

- In NF-1, clinical evaluation appears to be more useful to detect complications than is screening imaging in asymptomatic patients. Imaging is indicated in evaluation of suspected tumors based on clinical evaluation and for follow-up of known intracranial tumors (Borofsky, 2013).
- Conversely in NF-2, routine MR imaging screening is always indicated, given the high prevalence of CNS tumors, especially vestibular schwannomas. In patients with NF-2, routine screening brain/IAC imaging is indicated annually starting from age 10 if asymptomatic or earlier with clinical signs/symptoms. Most individuals with NF2 eventually develop a spinal tumor, mostly commonly schwannomas, but meningioma and ependymomas are also seen. Spinal imaging at baseline and every 2 to 3 years is also advised with more frequent imaging, if warranted, based on sites of tumor involvement (Evans, 2017).
- In patients with Tuberous Sclerosis, Brain MRI should be obtained every 1-3 years up until age 25 for surveillance for CNS abnormalities (Krueger, 2013).
- In Von Hippel Lindau Syndrome, imaging of the brain and spinal cord for hemangioblastomas is recommended every 2 years (Rednam, 2017).
- In Sturge Weber Syndrome, Brain MRI can rule out intracranial involvement after only after age 1 and is recommended in patients <1 year only if symptomatic (Comi, 2011).

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MRI and Positron Emission Tomography (PET) for Chronic Seizures – When MRI is performed in the evaluation of patients for epilepsy surgery, almost a third of those with electrographic evidence of temporal lobe epilepsy have normal MRI scans. Interictal positron emission tomography (PET) may be used to differentiate patients with MRI-negative temporal lobe epilepsy.

Multiple Sclerosis (Rovira, 20154; Saguil, 2014; Thompson, 20187) —

The diagnosis of MS requires demonstration of lesions in the CNS disseminated in time and space and the absence of fever, infection, or other more likely etiologies. There is an expanding amount of available disease-modifying treatments that which are effective in slowing down disease

progression, especially in the early stages. Though they are expensive and These treatments can have serious side effects and can be costly; therefore, the accurate and expeditious diagnosis of MS is critical.

The diagnosis of MS can be made on clinical presentation alone with 2 clinical attacks and objective clinical evidence of more than 2 lesions. Attacks may be patient-reported or objectively observed and must last for a minimum of 24 hours and be 30 days apart. However, corroborating magnetic resonance imaging (MRI) is the diagnostic standard and is used, as well, to rule out other disorders. Additionally, MRI findings can replace certain clinical criteria in a substantial number of patients. In the revised McC-Donald Criteria, MRI findings can be used to establish dissemination in both time and space.

Table 3: Variable Symptoms and Signs of MS

<u>Symptoms</u>	<u>Signs</u>
Depressed mood	<u>Ataxia</u>
Memory loss/cognitive changes	<u>Dysmetria</u>
Dizziness or vertigo	Decreased sensation (pain, vibration, position)
<u>Fatigue</u>	Decreased strength

Hearing loss and tinnitus

Hyperreflexia, spasticity

Heat sensitivity (Uhthoff Phenomenon)

Nystagmus

Incoordination and gait disturbances

Lhermitte's sign

Sensory disturbances (dysesthesias, numbness, paresthesias)

<u>Visual defects (internuclear ophthalmoplegia, optic disc pallor, red color desaturation, reduced visual defects (internuclear ophthalmoplegia, optic disc pallor, red color desaturation, reduced visual defects (internuclear ophthalmoplegia, optic disc pallor, red color desaturation, reduced visual defects (internuclear ophthalmoplegia, optic disc pallor, red color desaturation, reduced visual defects (internuclear ophthalmoplegia, optic disc pallor, red color desaturation, reduced visual defects (internuclear ophthalmoplegia, optic disc pallor, red color desaturation, reduced visual defects (internuclear ophthalmoplegia) disc pallor, red color desaturation, reduced visual disc pallor (internuclear ophthalmoplegia) disc pallor (internuclear ophtha</u>

acuity)

nambriess, parestresias,

Pain

Urinary symptoms

<u>Visual disturbances (diplopia, oscillopsia)</u>

Weakness

In the presence of a clear, clinically isolated syndrome such as optic neuritis, transverse myelitis, or brain stem syndrome, brain MRI is the next diagnostic step. MS can also have variable and often subjective symptoms that come and go (see <u>Table 3below</u>). If there are recurrent episodes of variable neurological signs or symptoms not attributable to another cause with clinical concern for MS, imaging is warranted as well.

<u>Symptoms</u>	<u>Signs</u>
<u>Depressed mood</u>	<u>Ataxia</u>
Memory loss/cognitive changes	Dysmetria
<u>Dizziness or vertigo</u>	Decreased sensation (pain, vibration, position)

<u>Fatigue</u>	Decreased strength
Hearing loss and tinnitus	Hyperreflexia, spasticity
Heat sensitivity (Uhthoff Phenomenon)	<u>Nystagmus</u>
Incoordination and gait disturbances	<u>Lhermitte's sign</u>
<u>Pain</u>	Visual defects (internuclear ophthalmoplegia, optic disc pallor, red color desaturation, reduced visual acuity)
Sensory disturbances (dysesthesias, numbness, paresthesias)	
<u>Urinary symptoms</u>	
<u>Visual disturbances (diplopia,</u> <u>oscillopsia)</u>	
<u>Weakness</u>	

Symptoms
Depressed mood
Memory loss/cognitive changes
Dysmetria

Dizziness or vertigo Decreased sensation (pain, vibration, position)

Fatigue Decreased strength

Hearing loss and tinnitus Hyperreflexia, spasticity

Heat sensitivity (Uhthoff Phenomenon) Nystagmus

Incoordination and gait disturbances Lhermitte's sign

Pain

Visual defects (internuclear ophthalmoplegia,
optic disc pallor, red color desaturation, reduced

visual acuity)

Sensory disturbances (dysesthesias, numbness, paresthesias)

Urinary symptoms

Visual disturbances (diplopia, oscillopsia)

Weakness

MRI and Neuromyelitis optica spectrum disorders (NMOSD) (Wingerchuk, 2015) - NMOSD are inflammatory disorders of the central nervous system characterized by severe, immune-mediated demyelination and axonal damage predominantly affecting the optic nerves and spinal cord, but also the brain and brainstem. NMOSD can be distinguished from multiple sclerosis and other inflammatory disorders by the presence of the aquaporin-4 (AQP4) antibody. Features of NMOSD include attacks of bilateral or sequential optic neuritis acute transverse myelitis and the area postrema syndrome (with intractable hiccups or nausea and vomiting). The evaluation of suspected NMOSD entails brain and spinal cord neuroimaging. In contrast to MS (in which spinal cord involvement tends to be incomplete and asymmetric), NMOSD have a longer extent of spinal cord demyelination generally involving three or more vertebral segments.

Temporal Arteritis - Giant cell arteritis (GCA) is an inflammatory disorder that should be considered in patients over the age of 50 with the following signs or symptoms: new headaches, acute onset of visual disturbances (especially transient monocular visual loss), jaw claudication, constitutional symptoms, tenderness over the temporal artery, and elevated ESR and/or CRP. A diagnosis of polymyalgia rheumatica (PMR) is highly associated. Large vessel GCA denotes involvement of the aorta and its first-order branches, especially the subclavian arteries, and is common. Extra- and intracranial cerebral vasculitis can also be seen, but is more rare, and strokes are related to vasculitis of extracranial cerebral arteries causing vertebral or internal carotid arteries stenosis. Gold standard for diagnosis of GCA is temporal artery biopsy. Color Doppler ultrasound (CDUS) can be used as a surrogate for temporal artery biopsy in some cases. High-resolution magnetic resonance imaging (MRI) can visualize the temporal arteries when used with contrast. The presence of clinical manifestations unusual in GCA should prompt consideration of alternative diagnoses. Examples of such include: adenopathy, pulmonary infiltrates, digital cyanosis, ulceration, or gangrene, mononeuritis multiplex, stroke in the distribution of the middle cerebral artery, glomerulitis, or and/or rapidly rising creatinine (D'Souza, 2016; Diamantopoulos, 2014; D'Souza, 2016; Klink, 2014; Larivière, 2014; Salehi, 2016; Yip 2020; Larivièr, 2014).

MMSE - The Mini Mental State Examination (MMSE) is a tool that can be used to systematically and thoroughly assess mental status. It is an 11-question measure that tests five areas of cognitive function: orientation, registration, attention and calculation, recall, and language. The MMSE has been the most commonly used measure of cognitive function in dementia research, but researchers have recognized that it is relatively insensitive and variable in mildly impaired individuals. The maximum score is 30. A score of 23 or lower is indicative of cognitive impairment. The MMSE takes only 5-10 minutes to administer and is, therefore, practical to use repeatedly and routinely.

MoCA - The Montreal Cognitive Assessment (MoCA) was designed as a rapid screening instrument for mild cognitive dysfunction. It assesses different cognitive domains: attention and concentration, executive functions, memory, language, visuoconstructional skills, conceptual thinking, calculations, and orientation. MoCA differs from the MMSE mainly by including tests of executive function and abstraction, and by putting less weight on orientation to time and place. Ten of the MMSE's 30 points are scored solely on the time-place orientation test, whereas the MoCA assigns it a maximum of six points. The MoCA also puts more weight on recall and attention-calculation performance, while deemphasizing language skill. Time to administer the MoCA is approximately 10 minutes. The total possible score is 30 points; a score of 26 or above is considered normal.

MRI and Movement disorders - Atypical parkinsonian syndromes include progressive supranuclear palsy (PSP), multiple system atrophy (MSA), corticobasal degeneration (CBD), and dementia with Lewy bodies.

Anosmia - Nonstructural causes of anosmia include post_-viral symptoms, medications (AmitiptylineAmitriptyline, Enalapril, Nifedipine, Propranolol, Penicillamine, Sumatriptan, Cisplatin, Trifluoeuperazine, Propylthiouracil). These should be considered prior to advanced imaging to look for a structural cause_

-Anosmia and dysgeusia have been reported as common early symptoms in patients with COVID-19, occurring in greater than 80 percent of patients. For isolated anosmia, imaging is typically not needed once the diagnosis of COVID has been made given the high association. As such, COVID testing should be done prior to imaging (Geyer, 2008; Lechien, 2020; Saniasiaya, 20219).

Evaluation of olfactory function is essential to determine the degree of chemosensory loss and confirm the patient's complaint. It also allows monitoring of olfactory function over time, helps to detect malingerers, and to establish compensation for disability. There are two general types of olfactory testing: psychophysical and electrophysiologic testing. Psychophysical tests are used for clinical evaluation of olfactory loss; whereas, electrophysiologic tests, such as electro-olfactogram (EOG) or odor event—related potentials (OERPs) are used for research purposes only.

Olfactory threshold tests rely on measuring detection thresholds of a specific odorant, such as phenyl ethyl alcohol (PEA) or butyl alcohol. Odor identification tests are quantitative tests in which patients are asked to identify the odorants at the suprathreshold level. Eexamples include: -The Connecticut odor identification, The University of Pennsylvania Identification Test (UPSIT) and the Cross-Cultural Smell Identification Test (CC-SIT). In Europe, a commonly used test is a threshold- and odorant-identification forced-choice test that uses odorant-impregnated felt-tipped pens (Sniffin' Sticks). A simple olfactory screening test using a 70% isopropyl alcohol pad as a stimulant has also been well described in the literature (Wrobel, 2004).

MRI for Macrocephaly - Consider ultrasound in infants with macrocephaly and a normal neurological examination, no evidence of increased ICP and an open anterior fontanelle. If head US is normal, the infant should be monitored closely (Smith, 1998). The anterior fontanelle generally closes between 10 and 24 months of age, with 3% closing between 5-9 months and 11% after 24 months (Pindrik, 2014).

MRI and Normal Pressure Hydrocephalus (NPH) - Although diagnosis can be made based on CT findings alone, MRI is more accurate for disclosing associated pathologies (such as cerebrovascular disease), excluding other potential etiologies and for detecting NPH typical signs of prognostic value. A CT scan can exclude NPH and is appropriate for screening purposes and in patients who cannot undergo MRI (Damasceno, 2015).

MRI and Vertigo – The most common causes of vertigo seen are benign paroxysmal positional vertigo (BPPV), vestibular neuronitis (VN) and Ménière's disease. These peripheral causes of vertigo are benign, and treatment involves reassurance and management of symptoms. Central causes of vertigo, such as cerebrovascular accidents (CVAs), tumors and multiple sclerosis (MS), need to be considered if the patient presents with associated neurological symptoms, such as weakness, diplopia, sensory changes, ataxia, or confusion. Magnetic resonance imaging is appropriate in the evaluation of patients with vertigo who have neurologic signs and symptoms, progressive unilateral hearing loss or risk factors for cerebrovascular disease. MRI is more appropriate than CT for diagnosing vertigo due to its superiority in visualizing the posterior portion of the brain, where most central nervous system disease

that causes vertigo is found. A full neurologic and otologic evaluation including provocative maneuvers, vestibular function testing and audiogram can help evaluate vertigo of unclear etiology and differentiate between central and peripheral vertigo.

MRI and developmental delay – Significant developmental delay is defined as significant delay (more than two standard deviations below the mean) in one or more developmental domains: gross/fine motor, speech/language, cognition, social/personal, and activities of daily living. Isolated delay in social/language development is characteristic of autism spectrum disorders or hearing loss. Isolated delay in motor development is characteristic of cerebral palsy (a static encephalopathy) or myopathy. Global developmental delay (GDD) is: a subset of developmental delay defined as significant delay (by at least 2 SD's) in two or more developmental categories. Note that the term "GDD" is usually reserved for children <5 years old, whereas in older children >5 years, disability is quantifiable with IQ testing.

Low risk brief resolved unexplained event (BRUE) formerly apparent life-threatening event (ALTE) requires all the following:

- Age > 60 days
- Gestational age ≥ 32 weeks or older and corrected gestational age ≥ 45 weeks
- First brief event
- Event lasting < 1 minute
- No CPR required by the trained medical provider
- No concerning historical features or physical examination findings.

Combination MRI/MRA of the Brain – This is one of the most misused combination studies and other than what is indicated above these examinations should be ordered in sequence, not together. Vascular abnormalities can be visualized on the brain MRI.

Patients presenting with a new migraine with aura (especially an atypical or complex aura) can mimic a transient ischemic attack or an acute stroke. If there is a new neurologic deficit, imaging should be guided by concern for cerebrovascular disease, not that the patient has a headache (Whitehead Nahas, 2019).

Leptomeningeal Carcinomatosis (Andersen, 2019; Clarke, 2010; Maillie, 2021; Wang, 2018) —
Leptomeningeal metastasis is an uncommon and typically late complication of cancer with poor
prognosis and limited treatment options. Diagnosis is often challenging with nonspecific presenting
symptoms ranging from headache and confusion to focal neurologic deficits such as cranial nerve
palsies. Standard diagnostic evaluation involves a neurologic examination, MRI of the brain and
spine with gadolinium, and cytologic evaluation of the cerebral spinal fluid (CSF).
Hematologic malignancies (leukemia and lymphoma), primary brain tumors as well as solid
malignancies can spread to the leptomeninges. The most common solid tumors giving rise to LM are
breast cancer (12 - 35 %), small and non-small cell lung cancer (10-26 %), melanoma (5 -25 %),
gastrointestinal malignancies (4-14 %), and cancers of unknown primary (1-7 %).

POLICY HISTORY

Date POLICY HISTORY	Summary	
April-July 2021	Reordered Indications	
· <u> </u>	Updated references	
	Updated background section	
	Added	
	• Brain MR/MRA are not approvable simultaneously unless they meet the	
	criteria described below in the Indications for Brain MR/Brain MRA	
	combination studies section.	
	• Cluster headaches or other trigeminal-autonomic cephalgias i.e.	
	paroxysmal hemicrania, hemicrania continua, short-lasting unilateral	
	neuralgiform headache attacks (SUNCT/SUNA) imaging is indicated once	
	to eliminate secondary causes (IHS, 2018)	
	Note: MRI is the study of choice for detecting cavernous malformations	
	(CCM). Follow-up imaging of known CCM should be done only to guide	
	treatment decisions or to investigate new symptoms. First-degree	
	relatives of patients with more than one family member with a CCM	
	should also have a screening MRI as well as genetic counseling	
	• Langerhans cell histiocytosis with visual, neurological, or endocrine	
	abnormality; polyuria or polydipsia; suspected craniofacial bone lesions,	
	aural discharge, or suspected hearing impairment/mastoid involvement	
	• Langerhans cell histiocytosis -To assess treatment response and	
	surveillance of known brain lesions	
	 Progressive Multifocal Leukoencephalopathy (PML) surveillance for 	
	patients on natalizumab (Tsyabri)	
	 12 months after the start of treatment in all patients 	
	 Further surveillance MRI scanning timing is based on anti-JCV 	
	antibody status	
	If anti-JCV antibody negative, annually	
	If anti-JCV antibody positive and antibody index < 1.5. every 6	
	<u>months</u>	
	If anti-JCV antibody positive and antibody index > 1.5, every 3-	
	4 months	
	Temporal Arteritis: Note: Protocol should include high-resolution	
	contrast-enhanced imaging the temporal artery	
	• similar mental status instruments */formal neuropsychological *Other	
	examples include Ottawa 3DY (O3DY), Brief Alzheimer's Screen (BAS),	
	Blessed Dementia Scale (BDS), caregiver-completed AD8 (cAD8), Brief	
	Cognitive Rating Scale (BCRS), Clinical Dementia Rating (CDR) (Carptenter,	
	2011; McDougall, 1990)	

- FDA labeling for the drug Aduhelm (for Alzheimer's disease) requires
 baseline imaging and monitoring with Brain MRI. Criteria for coverage includes the following:
 - Baseline study within 1 year of initiating treatment
 - Prior to the 7th and 12th infusions
 - Monitoring if radiographic severe Amyloid Related Imaging Abnormalities (ARIA) is observed
- Optic atrophy as an abnormal eye finding
- Childhood strabismus with development delay or abnormal fundoscopic exam to rule out intracranial abnormalities
- Bulbar symptoms ie. difficulty in chewing, weakness of the facial muscles, dysarthria, palatal weakness, dysphagia, and dysphonia and/or signs i.e. atrophy and fasciculations of the tongue and absent gag reflex
- Pseudobulbar symptoms i.e. dysphagia, dysarthria, facial weakness,
 sudden, stereotyped emotional outbursts that are not reflective of mood
 and/or signs i.e. spastic tongue and exaggerated gag/jaw jerk
- Evaluation of the corticomedullary junction in Achondroplasia
- Evaluation of suspected hydrocephalus with any acute, new, or fluctuating neurologic, motor, or mental status changes (separated this from known hydrocephalus)
- Cisternography for intermittent and complex CSF rhinorrhea/otorrhea.
 CSF fluid should always be confirmed with laboratory testing (Beta-2 transferrin assay).
- Suspected carotid or vertebral artery dissection with focal or lateralizing neurological deficits to Brain MRI/Brain MRA/Neck MRA combo
- Headache associated with exercise or sexual activity (Brain MRI/Brain MRA combo)
- Pre-operative evaluation for a planned surgery or procedure
 Brain MRI/ Cervical MRI/Thoracic MRI (any combination)
 - For evaluation of neuromyelitis optica spectrum disorders (recurrent or bilateral optic neuritis; recurrent transverse myelitis)
 - For known MS, prior to the initiation or change of disease
 modification treatments and assess disease burden (to establish a new baseline)
 - Follow -up scans for known MS if patients have known spine disease:
 - 6-12 months after starting/changing treatment
 - Every 1-2 years while on disease-modifying therapy to assess
 for subclinical disease activity, less frequently when stable for
 2-3 years

Brain MRI/ Cervical MRI/Thoracic MRI/Lumbar (any combination)

- Follow up imaging of a known Arnold Chiari malformation (II/III). For
 Chiari, I follow-up imaging only if new or changing signs/symptoms
 - Suspected Leptomeningeal carcinomatosis (LC)
 - Tumor evaluation and monitoring in neurocutaneous syndromes See
 Background
 - CSF leak highly suspected and supported by patient history and/or physical exam findings (known or suspected spontaneous (idiopathic) intracranial hypotension (SIH), post lumbar puncture headache, post spinal surgery headache, orthostatic headache, rhinorrhea or otorrhea, or cerebrospinal-venous fistula)

Brain MRI/Orbit MRI Optic Neuritis- If needed to confirm optic neuritis and rule out compressive lesions

Clarified

- Symptoms indicative of increased intracranial pressure, such as recurring headaches after waking with or without associated nausea/vomiting
- Suspected stroke with a personal or first-degree family history (brother, sister, parent, or child) of aneurysm or known coagulopathy or on anticoagulation
- Symptoms of transient ischemic attack (TIA) (episodic neurologic symptoms such as sensory deficits, limb weakness, speech difficulties, visual loss, lack of coordination, or mental status changes)
- Known or suspected skull fracture by physical exam and/or prior imaging
- Neurologic findings (e.g. visual field deficit suggesting compression of the optic chiasm, diplopia, gaze palsy) – Pituitary
- Follow-up known of pituitary adenoma New neuroendocrine signs or symptoms
- Follow of known arachnoid cyst (Al-Holou, 2010, 2013; Mustansir, 2018)
 - > 4 years old, repeat imaging only if newly symptomatic i.e.
 headaches, increased intracranial pressure, hydrocephalus, local mass effect, seizures, visual/endocrine dysfunction.
- Temporal Arteritis Atypical features, failure to response to treatment or concern for intracranial involvement
- Central Nervous System (CNS) involvement in patients with known or suspected vasculitis or autoimmune disease with abnormal inflammatory markers or autoimmune antibodies
- Suspected primary CNS vasculitis based on neurological signs and symptoms with completed infectious/inflammatory lab work-up
- Anosmia or dysosmia on objective testing that is persistent and of unknown origin (also in combo section)

- Trigeminal Neuralgia or other trigeminal autonomic cephalgias, notably in those with atypical presentation (also in combo section)
- Clarified age < 18 for imaging of microcephaly and macrocephaly
- For initial evaluation of a suspected Arnold Chiari malformation
- For follow up imaging of a known Arnold Chiari malformation (II/III). For Chiari I follow-up imaging only if new or changing signs/symptoms
- After full neurologic examination and vestibular testing with concern for central vertigo (i.e. skew deviation, vertical nystagmus, head thrust test, videonystagmography (VNG)/electronystagmography (ENG))
- Clarified age < 18 for imaging of developmental delay
- Brain with IAC CSF fluid should always be confirmed with laboratory testing (Beta-2 transferrin assay).
- Optic neuropathy or unilateral optic disk swelling of unclear etiology (Brain MRI/Orbit MRI)

Deleted

- Approved indications as noted above and being performed in a child under 8 years of age who will need anesthesia for the procedure and there is a suspicion of concurrent vascular and intracranial pathology (redundant)
- Brain MRI/Cervical MRI combo section (included in Brain MRI/ Cervical MRI/Thoracic MRI/Lumbar combos)

See word document

May 2020

Clarified:

- New onset headache with (neurologic deficit) or with signs of increased intracranial pressure (papilledema)
- Special additional considerations in the pediatric population with persistent headache
 - Documented absence of family history of headache
- For evaluation of known or suspected stroke or vascular disease:
- Suspected brain tumor
- Suspected brain metastasis or intracranial involvement in patients with a history of cancer based on neurological symptoms or examination findings
- Follow up of known malignant brain tumor

Clarified:

Patient with history of CNS cancer (either primary or secondary)
 and a recent course of chemotherapy, radiation therapy (to the
 brain), or surgical treatment within the last two (2) years

- Follow up of known non-malignant brain tumor/lesion if symptomatic, new/changing signs or symptoms or complicating factors
- New onset of an unprovoked seizure in adults
- Suspected intracranial abscess or brain infection
- Suspected Encephalitis with headache and altered mental status or follow-up as clinically warranted
- Mental status score of either MMSE or MoCA of less than 26 or other similar mental status instruments/neuropsychological testing

Clarified:

- Anosmia (loss of smell) documented by objective testing that is persistent and of unknown origin
- Chiari malformation/syrinx Often congenital, but can present later in life; or less commonly acquired secondary to tumor, stroke, trauma, infection etc.
- Vertigo associated with any of the following
 - Risk factors for cerebrovascular disease with concern for stroke
 - After full neurologic examination and vestibular testing with concern for central vertigo
- Combo Brain MRI/Orbit MRI
 - Reworded: Unilateral optic disk swelling/optic neuropathy
 of unclear etiology to distinguish between a compressive
 lesion of the optic nerve, optic neuritis, ischemic optic
 neuropathy (arteritic or non-arteritic), central retinal vein
 occlusion or optic nerve infiltrative disorders
 - Bilateral optic disk swelling (papilledema) with vision loss

Added:

- Visual loss (as a neurological deficit) Not explained by underlying ocular diagnosis, glaucoma or macular degeneration
- Under New acute headache, sudden onset:
 - With a personal or family history of brain aneurysm or AVM (arteriovenous malformation)
 - Known coagulopathy or on anticoagulation
- Under New onset of headache and any of the following
 - <u> Fever</u>
 - Subacute head trauma
 - Pregnancy or puerperium
 - o Age > 50

 Neurological deficits - Note: Neuroimaging warranted for atypical/complex migraine aura, but not for a typical migraine aura (see background)

Added:

- Special additional considerations in the pediatric population with persistent headache
 - Symptoms indicative of intracranial pressure, such as recurring headaches after waking with or without associated nausea/vomiting
 - Severe headache in a child with an underlying disease that predisposes to intracranial pathology (e.g.; immune deficiency, sickle cell disease neurofibromatosis, history of neoplasm, coagulopathy, hypertension, congenital heart disease)
- Suspected stroke with a personal or family history (brother, sister, parent or child) of aneurysm or known coagulopathy/anticoagulation

Added:

- Suspected Pituitary Tumors:
 - With the following:
 - Neurologic findings (e.g. visual field deficit suggesting compression of the optic chiasm)
 - Suspected hypofunctioning pituitary gland based on hormonal testing e.g., hypo pituitarism, growth hormone deficiency, hypogonadotropic hypogonadism
 [i.e. low gonadotropins (FSH/LH) and sex hormones*]
 - * severe secondary hypogonadism with total testosterone persistently < 150 and low or normal LH/FSH OR
 - * testosterone levels below normal range with low or normal LH/FSH AND
 - neurological sign and symptoms OR
 - other pituitary hormonal abnormalitiesOR
 - consideration of reversible functional causes of gonadotropin suppression (e.g. obesity, opioid use, or comorbid illness)

Added:

 Suspected hyperfunctioning pituitary gland based on hormonal testing i.e., central hyperthyroidism (high TSH), Cushing disease

- (high ACTH), acromegaly/gigantism (high GH/IGF-1) or elevated prolactin (>250 ng/mL or persistently elevated in the absence of another cause eg. stress, pregnancy, hypothyroidism, medication)
- Note: Galactorrhea without elevated prolactin, imaging is not indicated
- Central Diabetes Insipidus (low ADH)
- Precocious puberty in a child (male < 9; female < 8), with hormonal studies suggesting a central cause and evidence of an accelerated bone age on X-ray
- Pituitary apoplexy with sudden onset of neurological and hormonal symptoms
- Suspected recurrence with prior history of CNS cancer based on neurological symptoms or examination

Added:

- Follow up of known meningioma
 - If <2cm or heavily calcified at 2 years and 5 years
 - > 2cm annually for 3 years and then scans at 5 years and 10 years.
 - Multiple meningiomas, annually
 - After treatment (surgery or radiotherapy), post-operative if concern for residual tumor, every 6-12 months then annually for 3-5 years based on WHO Grade (see background)
- Follow-up known of pituitary adenoma
 - New signs or symptoms
 - Functioning adenoma to assess response to treatment and
 1-year follow-up after drug holiday

Added:

- Follow of known pineal cyst (> 5mm) if there are atypical features
 or symptoms (e.g., headaches, gaze paresis, ataxia, papilledema, nausea/vomiting)
- Follow of known arachnoid cyst
 - < 4 years old, serial imaging is warranted</p>
 - > 4 years old, repeat imaging is approvable if newly symptomatic i.e. headaches, increased intracranial pressure, hydrocephalus, local mass effect, seizures, visual/endocrine dysfunction
- For screening for known Non-CNS Cancer
 - Default screening for
 - Kidney cancer

- Lung cancer
- Merkel cell carcinoma

Added:

- Mucosal melanoma of the head and neck, especially of the oral cavity
- Poorly differential neuroendocrine cancer (Large or Small cell/Unknown primary of neuroendocrine origin)
- Screening with preconditions
 - AML.....Suspicion of leukemic meningitis
 - Cutaneous melanoma....Stage IIIC or higher
 - Testicular Cancer-Seminoma..... High risk
 - Gestational Trophoblastic Neoplasia...Pulmonary metastasis
 - o Bladder cancer......High risk, i.e. small cell
- All other cancer if CNS symptoms presentAdded:
- For screening of Hereditary Cancer Syndromes
 - Li Fraumeni syndrome- Annually
 - Von Hippel Lindau Every 2 years, starting at age of 8 years
 - Tuberous Sclerosis Every 1-3 years, until the age of 25
 years
 - MEN1 Every 3-5 years, starting at the age of 5 years
 - O NF-2- Brain IAC: Annually starting, from age of 10 years
 - Sturge Weber Syndrome: Once, after age 1 to rule out intracranial involvement after; in patients <1 year, only if symptomatic
- Known seizure disorder without previous imaging

Added:

- Imaging indications for new onset seizures in the pediatric population
 - Abnormal neurological exam, especially a postictal focal deficit
 - Significant developmental delay
 - Focal onset
 - EEG shows focal or suspected structural abnormalities

Note: Imaging is not indicated in simple febrile seizures

- Suspected temporal arteritis in a patient > 50 with temporal
 headache, abrupt visual changes, jaw claudication, temporal artery
 tenderness, constitutional symptoms or elevated ESR AND
 - Negative initial work-up (color Doppler ultrasonography or biopsy) OR

 Atypical features or failure to response to treatment with concern for large vessel involvement

Added:

- MRI indicted for atypical dystonia. Note: MRI not indicated in essential tremor or isolated focal dystonia (e.g., blepharospasm, cervical dystonia, laryngeal dystonia, oromandibular dystonia, writer's dystonia)
- Binocular diplopia with concern for intracranial pathology
- Hemifacial spasm
- Other objective cranial nerve palsy (CN IX-XII)
- Follow up shunt evaluation (Pople, 2002, Reddy, 2014, Kamenova, 2018)
 - Post operatively if indicated based on underlying disease
 and pre-operative radiographic findings and/or
 - 6-12 months after placement and/or
 - With neurologic symptoms that suggest shunt malfunction

Added:

- Suspected spontaneous intra-cranial hypotension with distinct postural headache other symptoms include: nausea, vomiting, dizziness, tinnitus, diplopia neck pain or imbalance
- CSF flow study for evaluation and management of CSF flow disorders
- Diagnosis of central sleep apnea on polysomnogram
 - Ochildren > 1 year
 - Adults in the absence of heart failure, chronic opioid use, high altitude, or treatment emergent central sleep apnea
 AND concern for a central neurological cause (Chiari malformation, tumor, infectious/inflammatory disease) OR with an abnormal neurological exam
- Syncope with clinical concern for seizure or associated neurological signs or symptoms
- Cyclical vomiting syndrome or abdominal migraine with any localizing neurological symptoms
- Soft tissue mass of the head with nondiagnostic initial evaluation (ultrasound and/or radiograph)

Added:

 Cerebral palsy if etiology has not been established the neonatal period, there is change in the expected clinical or developmental profile or concern for progressive neurological disorder Unexplained event (BRUE) formerly apparent life-threatening event
 (ALTE) in infants < 1 year with concern for neurological cause based
 on history and exam

Note: Imaging is not indicated in low risk patients

- Under Indications for a Brain MRI with Internal Auditory Canal (IAC):
 - CSF otorrhea (MRI for intermittent leak, CT for active leaks)
 - Clinical Suspicion of acute mastoiditis as a complication of acute otitis media with intracranial complications (i.e. meningeal signs, cranial nerve deficits, focal neurological findings, altered mental status)
 - Bell's Palsy for evaluation of the extracranial nerve course if atypical signs, slow resolution beyond three weeks, no
 improvement at four months, or facial twitching/spasms
 prior to onset

Added:

- Combo Brain MRI/MRA
 - Thunderclap headache with continued concern for underlying vascular abnormality after initial negative workup
 - Negative Brain CT;
 - AND Negative Lumbar Puncture
 - Acute, sudden onset of headache with personal history of a vascular abnormality or first-degree family history of aneurysm
- Combo Brain MRI/Orbit MRI
 - Optic Neuritis if atypical presentation, severe visual impairment or poor recovery following initial onset or treatment onset
- Combo Brain MRI/Face/Sinus/Neck MRI
 - Bells/hemifacial spasm that meets above criteria
 - Objective cranial nerve palsy (CN IX-XII) (for evaluation of the extracranial nerve course
 - Granulomatosis with polyangiitis (Wegener's granulomatosis) disease

Deleted:

- Under New onset of headache and any of the following
 - Temporal headache in person > 55, with sedimentation rate
 (ESR) > 55 with tenderness over the temporal artery.

- Known or suspected pituitary tumor with corroborating physical exam (i.e., galactorrhea or acromegaly) neurologic findings and/or lab abnormalities.
- Known brain tumor and new onset of headache.
- Follow up shunt evaluation within six (6) months of placement or one (1) year follow up and/or with neurologic symptoms
- From combo Brain MRI/MRA Clinical suspicion of subarachnoid hemorrhage (SAH) ie thunderclap headache

August 2019

- For evaluation of patient with neurologic symptoms or deficits suspicious for MS: Added: "clinically isolated syndrome OR recurrent episodes of variable neurological signs or symptoms not attributable to another cause; And Removed time frame of 'within the last 4 weeks'
- Removed: Stable condition with no prior imaging within the past ten (10) months or within the past six (6) months if patient has relapsing disease
- Removed: Exacerbation of symptoms or change in symptom characteristics such as frequency or type and demonstrated compliance with medical therapy.
- For evaluation of MS. added:
 - To establish a new baseline (no recent imaging, postpartum, or 6-12 months after switching disease modifying therapy)
 - Prior to starting or switching disease-modifying therapy
 - Every 1-2 years while on disease-modifying therapy to assess for subclinical disease activity, less frequently when stable for 2-3 years
 - New signs or symptoms suggested of an exacerbation or unexpected clinical worsening
 - PML surveillance for patients on natalizumab
- For evaluation of known or suspected seizure disorder, added:
 - Newly identified change in seizure activity/pattern
- Renamed Parkinson's section to: Movement disorders and added:
 - For the evaluation of other movement disorder to exclude a structural lesion (i.e., suspected Huntington disease, chorea, atypical parkinsonian syndromes, hemiballismus, secondary dystonia).
 - * MRI not indicated in essential tremor or primary dystonia
 - For suspected Parkinson's, added 'with atypical feature or unresponsive to levodopa

- For evaluation of neurologic symptoms or deficits, added: visual loss
- For trauma, added:
 - On anticoagulation
 - Post concussive syndrome if persistent or disabling symptoms and imaging has not been performed
 - Subacute or chronic traumatic brain injury with new cognitive and/or neurologic deficit
- For evaluation of headache, added or removed:
 - Prior history of stroke or intracranial bleed with sudden onset of severe headache (moved)
 - New headache and signs of increased intracranial pressure
 - Related to activity or event (sexual activity, exertion, position) (new or progressively worsening)
 - New headache and persistent or progressively worsening during a course of physician directed treatment
 - Special considerations in the pediatric population with persistent headache:
 - Occipital location
 - Age < 6 years</p>
 - No family history of headache
- For evaluation of brain tumor:
 - Specified 'malignant' for f/u of known tumor
 - Added: Follow up of known benign tumor if symptomatic, new/changing signs or symptoms or complicating factors;
 Follow up of known meningioma; and tumor evaluation and monitoring in neurocutaneous syndromes
 - Removed: Known lung cancer or rule out metastasis and/or preoperative evaluation, Metastatic melanoma (not all melanomas)
- For evaluation of suspected stroke:
 - Moved 'patient with history of a known stroke with new and sudden onset of severe headache'
 - Separated: Family history of aneurysm
- For evaluation inflammatory disease or infections:
 - Changed meningitis with positive signs and symptoms from 'And' positive lab findings to 'OR' positive labs
 - For suspected encephalitis removed 'severe' headache
- For evaluation of congenital abnormality:
 - Modified the age restriction of > 6 months age for eval of macrocephaly to include 'in an infant/child with previously

- <u>abnormal US, abnormal neurodevelopmental exam, signs of increased ICP or closed anterior fontanelle'</u>
- For known or suspected normal pressure hydrocephalus (NPH):
 - Added With symptoms of gait difficulty, cognitive disturbance and urinary incontinence
- Other Indications:
 - Added detail to Vertigo including:
 - Signs or symptoms suggestive of a CNS lesion (ataxia, visual loss, double vision, weakness or a change in sensation)
 - Progressive unilateral hearing loss
 - Risk factors for cerebrovascular disease
 - After full neurologic examination and ENT work-up with concern for central vertigo
 - Modified developmental delay to include: Global developmental delay or developmental delay with abnormal neurological examination
 - Added:
 - Horner's syndrome with symptoms localizing the lesion to the central nervous system
 - Trigeminal Neuralgia if <40 years of age or atypical features (ie bilateral, hearing loss, dizziness/vertigo, visual changes, sensory loss, numbness, pain >2min, pain outside trigeminal nerve distribution, progression)
 - Bell's Palsy- if atypical signs, slow resolution beyond three weeks, no improvement at four months, or facial twitching/spasms prior to onset.
 - Psychological changes with neurological deficits on exam or after completion of a full neurological assessment that suggests a possible neurologic cause
 - New onset anisocoria
 - Removed Objective cranial nerve palsy; and Cholesteatoma
 (duplicated)
- For Brain MRI/Neck MRA: deleted 'confirmed carotid occlusion >
 60%, surgery or angioplasty candidate' and added 'Suspected
 carotid or vertebral artery dissection with focal or lateralizing
 neurological deficits'
- Added Brain MRI/Brain MRA section, including: Clinical suspicion of subarachnoid hemorrhage (SAH) ie thunderclap headache; and Suspected venous thrombosis (dural sinus thrombosis)

- Added Brain MRI/Brain MRA/Neck MRA section, including: Recent stroke or transient ischemic attack (TIA); and Approved indications as noted above and being performed in a child under 8 years of age who will need anesthesia for the procedure and there is a suspicion of concurrent vascular and intracranial pathology
- For Brain MRI/Cervical MRI, added: Suspected MS with new or changing symptoms consistent with cervical spinal cord disease; and Follow up to the initiation or change in medication for patient with known Multiple Sclerosis
- For Brain MRI/Orbit MRI, added: Bilateral papilledema with visual loss; and Known or suspected neuromyelitis optica spectrum disorder with severe, recurrent or bilateral optic neuritis; AND changed age restriction from 3 years to 8 years for children requiring anesthesia for the procedure with suspicion of concurrent orbital and intracranial pathology or tumor
- Added section for Brain MRI/Face/Sinus/Neck MRI, including:
 Anosmia on objective testing; and Trigeminal neuralgia or cranial nerve palsy that meets the above criteria
- Updated background information and references

August 2019

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 "clinically isolated syndrome OR recurrent episodes of variable neurological signs or symptoms not attributable to another cause; And Removed time frame of 'within the last 4 weeks'
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 - Specified 'malignant' for f/u of known tumor
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changed age restriction from 3 years to 8 years for children requiring anesthesia for the procedure with suspicion of concurrent orbital and intracranial pathology or tumor

- Added section for Brain MRI/Face/Sinus/Neck MRI, including: Anosmia on objective testing; and
 Trigeminal neuralgia or cranial nerve palsy that meets the above criteria
- Updated background information and references

May 2020

Clarified:

- New onset headache with (neurologic deficit) or with signs of increased intracranial pressure (papilledema)
- Special additional considerations in the pediatric population with persistent headache
 - Documented absence of family history of headache
- For evaluation of known or suspected stroke or vascular disease:
- Suspected brain tumor
- Suspected brain metastasis or intracranial involvement in patients with a history of cancer based on neurological symptoms or examination findings
- Follow up of known malignant brain tumor

Clarified:

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 - Neurological deficits Note: Neuroimaging warranted for atypical/complex migraine aura, but not for a typical migraine aura (see background)

Added:

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 - Symptoms indicative of intracranial pressure, such as recurring headaches after waking with or without associated nausea/vomiting
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- Suspected Pituitary Tumors:
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consideration of reversible functional causes of gonadotropin suppression (e.g. obesity, opioid use, or comorbid illness)

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- Suspected hyperfunctioning pituitary gland based on hormonal testing i.e., central hyperthyroidism (high TSH), Cushing disease (high ACTH), acromegaly/gigantism (high GH/IGF 1) or elevated prolactin (>250 ng/mL or persistently elevated in the absence of another cause eg. stress, pregnancy, hypothyroidism, medication)
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 - o Kidney cancer
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Added:

Mucosal melanoma of the head and neck, especially of the oral cavity

- Poorly differential neuroendocrine cancer (Large or Small cell/Unknown primary of neuroendocrine origin)
- Screening with preconditions
- o AML Suspicion of leukemic meningitis
- o Cutaneous melanoma Stage IIIC or higher
- o Testicular Cancer Seminoma High risk
- o Gestational Trophoblastic Neoplasia Pulmonary metastasis
- o Bladder cancer High risk, i.e. small cell
- All other cancer if CNS symptoms present

Added:

- For screening of Hereditary Cancer Syndromes
 - Li Fraumeni syndrome- Annually
 - Von Hippel Lindau Every 2 years, starting at age of 8 years
 - Tuberous Sclerosis Every 1-3 years, until the age of 25 years
 - MEN1 Every 3-5 years, starting at the age of 5 years
 - NF-2 Brain IAC: Annually starting, from age of 10 years
 - Sturge Weber Syndrome: Once, after age 1 to rule out intracranial involvement after; in patients <1 year, only if symptomatic
- Known seizure disorder without previous imaging

Added:

- Imaging indications for new onset seizures in the pediatric population
 - Abnormal neurological exam, especially a postictal focal deficit
 - Significant developmental delay
 - Focal onset
 - EEG shows focal or suspected structural abnormalities

Note: Imaging is not indicated in simple febrile seizures

- Suspected temporal arteritis in a patient > 50 with temporal headache, abrupt visual changes, jaw claudication, temporal artery tenderness, constitutional symptoms or elevated ESR AND
 - Negative initial work up (color Doppler ultrasonography or biopsy) OR
 - Atypical features or failure to response to treatment with concern for large vessel involvement

Added:

- MRI indicted for atypical dystonia. Note: MRI not indicated in essential tremor or isolated focal dystonia (e.g., blepharospasm, cervical dystonia, laryngeal dystonia, oromandibular dystonia, writer's dystonia)
- Binocular diplopia with concern for intracranial pathology
- Hemifacial spasm
- Other objective cranial nerve palsy (CN IX-XII)
- Follow up shunt evaluation (Pople, 2002, Reddy, 2014, Kamenova, 2018)
 - Post operatively if indicated based on underlying disease and pre-operative radiographic findings and/or

- With neurologic symptoms that suggest shunt malfunction

Added:

- Suspected spontaneous intra-cranial hypotension with distinct postural headache other symptoms include: nausea, vomiting, dizziness, tinnitus, diplopia neck pain or imbalance
- CSF flow study for evaluation and management of CSF flow disorders
- Diagnosis of central sleep apnea on polysomnogram

 - Adults in the absence of heart failure, chronic opioid use, high altitude, or treatment emergent central sleep apnea AND concern for a central neurological cause (Chiari malformation, tumor, infectious/inflammatory disease) OR with an abnormal neurological exam
- Syncope with clinical concern for seizure or associated neurological signs or symptoms
- Cyclical vomiting syndrome or abdominal migraine with any localizing neurological symptoms
- Soft tissue mass of the head with nondiagnostic initial evaluation (ultrasound and/or radiograph)

Added:

- Cerebral palsy if etiology has not been established the neonatal period, there is change in the
 expected clinical or developmental profile or concern for progressive neurological disorder
- Unexplained event (BRUE) formerly apparent life threatening event (ALTE) in infants < 1 year with concern for neurological cause based on history and exam
 Note: Imaging is not indicated in low risk patients
- Under Indications for a Brain MRI with Internal Auditory Canal (IAC):
 - CSF otorrhea (MRI for intermittent leak, CT for active leaks)
 - Clinical Suspicion of acute mastoiditis as a complication of acute otitis media with intracranial complications (i.e. meningeal signs, cranial nerve deficits, focal neurological findings, altered mental status)
 - Bell's Palsy for evaluation of the extracranial nerve course—if atypical signs, slow resolution beyond three weeks, no improvement at four months, or facial twitching/spasms prior to onset

Added:

- Combo Brain MRI/MRA
 - Thunderclap headache with continued concern for underlying vascular abnormality after initial negative work up
 - Negative Brain CT:
 - AND Negative Lumbar Puncture
 - Acute, sudden onset of headache with personal history of a vascular abnormality or firstdegree family history of aneurysm
- Combo Brain MRI/Orbit MRI
 - Optic Neuritis if atypical presentation, severe visual impairment or poor recovery following initial onset or treatment onset

- Combo Brain MRI/Face/Sinus/Neck MRI
 - O Bells/hemifacial spasm that meets above criteria
 - Objective cranial nerve palsy (CN IX XII) (for evaluation of the extracranial nerve course
 - o Granulomatosis with polyangiitis (Wegener's granulomatosis) disease

Deleted:

- Under New onset of headache and any of the following
 - Temporal headache in person > 55, with sedimentation rate (ESR) > 55 with tenderness over the temporal artery.
- Known or suspected pituitary tumor with corroborating physical exam (i.e., galactorrhea or acromegaly) neurologic findings and/or lab abnormalities.
- Known brain tumor and new onset of headache.
- Follow up shunt evaluation within six (6) months of placement or one (1) year follow up and/or with neurologic symptoms
- From combo Brain MRI/MRA Clinical suspicion of subarachnoid hemorrhage (SAH) ie thunderclap headache

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Reviewed / Approved by NIA Clinical Guideline Committee

GENERAL INFORMATION

It is an expectation that all patients receive care/services from a licensed clinician. All appropriate supporting documentation, including recent pertinent office visit notes, laboratory data, and results of any special testing must be provided. If applicable: All prior relevant imaging results and the reason that alternative imaging cannot be performed must be included in the documentation submitted.

Reviewed / Approved by M. Auf Khalid M.D., Medical Director, Radiology

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