

Evolent Clinical Guideline 004-22011 for Brain Magnetic Resonance Angiography (MRA-MRV)

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STATEMENT

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.~~
- *Where a specific clinical indication is not directly addressed in this guideline, medical necessity determination will be made based on widely accepted standard of care criteria. These criteria are supported by evidence-based or peer-reviewed sources such as medical literature, societal guidelines and state/national recommendations.*
- *The guideline criteria in the following sections were developed utilizing evidence-based and peer-reviewed resources from medical publications and societal organization guidelines as well as from widely accepted standard of care, best practice recommendations.*

Purpose

Indications for performing magnetic resonance angiography (MRA) ~~or Magnetic Resonance Venography (MRV) of~~ in the head/brain region.

Special Note

Brain MRI/MRA are not approvable simultaneously unless they meet the criteria described below in the indications for Brain MRI/Brain MRA combination studies section. If there is a combination request* for an overlapping body part, either requested at the same time or sequentially (within the past 3 months) the results of the prior study should be show one or more of the following:

- Inconclusive or show a need for additional or follow up imaging evaluation-**OR**
- The office notes should clearly document an indication why overlapping imaging is needed and how it will change management for the patient.

(*Unless approvable in the combination section See Combination Studies as noted in the guidelines)

section for indicated combinations below)

NOTE: Authorization for MR Angiography covers both arterial and venous imaging. The term angiography refers to both arteriography and venography.

INDICATIONS FOR BRAIN MR ANGIOGRAPHY/~~MR VENOGRAPHY~~

Evaluation of Suspected Intracranial Vascular Disease ^(1,2)

Aneurysm Screening

- Screening for intracranial aneurysm if two or more first-degree family members (parent, brother, sister, or child) with history of intracranial aneurysm ^(1,3)
 - **Note:** Repeat study is recommended every 5-7 years ⁽³⁾
- For one first degree relative with aneurysm, asymptomatic screening is not indicated - and would require a neurological sign or symptom supporting clinical concern for aneurysm ⁽⁴⁾
- Screening for aneurysm in high-risk populations ^{(1),5,6,7,8,9,10}
 - KNOWN genetic syndromes (see Genetic Syndromes and Rare Diseases)Imaging in Known Genetic Conditions)
 - Bicuspid aortic valve
 - Known aortic diseases (aneurysm, coarctation, dissection)

Suspected Vascular Abnormalities

- Suspected high flow vascular malformation (arteriovenous malformation (AVM) or dural arteriovenous fistula) in patient with previous or indeterminate imaging study ^{4(1,2)}
 - **Note:** MRI is the study of choice for detecting low-flow vascular malformations such as cavernomas, developmental venous anomalies and capillary telangiectasia ⁽²⁾
- Thunderclap headache with continued concern for underlying vascular abnormality (i.e., aneurysm or reversible cerebral vasoconstriction syndrome) after initial negative brain imaging ^{11,12,13,14(5-7)}
 - **Note:** Negative brain CT < 6 hours after headache onset excludes subarachnoid hemorrhage in neurologically intact patients. ¹³⁽⁷⁾ MRI lacks sensitivity in excluding subarachnoid hemorrhage less than 24 hours after headache onset. ^{11,15(5,8)}
- Headache associated with exercise, exertion, Valsalva or sexual activity ^{44(5,9)}
- Isolated third nerve palsy (oculomotor) with pupil involvement to evaluate for aneurysm ^{16,17(10,11)}
 - Horner's syndrome, non-central (miosis, ptosis, and anhidrosis) ⁴⁸
- Non-Central Horner's Syndrome (Secondary/preganglionic or tertiary/post-ganglionic) to evaluate for a vascular source (Such as dissection, aneurysm, arteritis)
 - **NOTE:** CTA/MRA of the chest and neck may also be indicated
- Pulsatile tinnitus to identify a suspected arterial vascular etiology ^{19,20(12-14)}

Note: MRI is the study of choice for detecting cavernomas, developmental venous anomalies and capillary telangiectasia (see background section) ⁽²⁾

Cerebrovascular Disease

Ischemic

- Recent ischemic stroke or transient ischemic attack (See background section)^{(24,22)(15,16)}
- Note:** For remote strokes with no prior vascular imaging, imaging can be considered based on location/type of stroke and documented potential to change management
- Known or suspected vertebrobasilar insufficiency (VBI) in patients with symptoms such as dizziness, vertigo, headaches, diplopia, blindness, vomiting, ataxia, weakness in both sides of the body, or abnormal speech.^{(23,24,25,26)(17-19)}
- Suspected carotid or vertebral artery dissection: secondary to trauma or spontaneous due to weakness of vessel wall^{(27,28)(16,20,21)}

Hemorrhagic

- Known subarachnoid hemorrhage (SAH) CTA is favored over MRA^(1,2)
- Known cerebral intraparenchymal hemorrhage with concern for underlying vascular abnormality^{(2,16)22}

Venous* MRV ^{(4)(16,22)}**

- Suspected central venous thrombosis (dural sinus and ANY ONE of the following:
 - Patient has a hypercoagulable state such as pregnancy, post-partum, prothrombotic conditions (acquired or genetic), malignancy, oral contraceptive use, recent infection, recent trauma or covid-19
 - Documentation of concern for central venous thrombosis) is specified
 - Papilledema or signs/symptoms of increased intracranial pressure
- Distinguishing benign intracranial hypertension (pseudotumor cerebri) from dural sinus thrombosis

Sickle cells disease (ischemic and/or hemorrhagic)⁽²⁹⁾

- Neurological signs or symptoms in sickle cell disease
- Stroke risk in sickle cell patients (2 - 16 years of age) with a transcranial doppler velocity ≥ 200

Vasculitis and Other Intracranial Vascular Disease

- Suspected Known vasculitis or autoimmune disease with concern for secondary CNS vasculitis based on neurological signs or symptoms in the setting of an underlying systemic disease with abnormal inflammatory markers or autoimmune antibodies⁽⁴⁾
- Suspected primary CNS vasculitis based on neurological signs and symptoms with completed infectious/inflammatory lab work-up^{(1,23)30,31}
- Large vessel vasculitis:
 - (Giant cell or Takayasu arteritis) with suspected intracranial involvement^{(32,33,34,35,36)(24)}
 - Takayasu's Arteritis⁽²⁵⁾
 - At initial diagnosis

- Every 6 months for the first 2 years while on therapy
- Annually after the first 2 years
- Suspected Moyamoya disease ^{(2,26)37}
- Suspected reversible cerebral vasoconstriction syndrome ^(14,38)_(22,27)
- For patients with fibromuscular dysplasia (FMD) ^(28,29):
 - One-time vascular study from brain to pelvis
- Spontaneous coronary arteries dissection (SCAD) ⁽³⁰⁾:
 - One-time vascular study from brain to pelvis

Note: Vessel wall MRI (ordered as Brain MRI) can also be performed in the evaluation of vasculitides ⁽³⁹⁾

Evaluation of Known Intracranial Vascular Disease ^(1,2)

- Known intracranial aneurysm, treated aneurysm, or known vascular malformation (i.e., AVM or dural arteriovenous fistula)
- Known vertebrobasilar insufficiency with new or worsening signs or symptoms (VBI) ^(23,24,26)_(17,19,31)
- Follow-up of known carotid or vertebral artery dissection within with any ONE of the following ^(16,32,33):
 - At 3-6 months post dissection (for evaluation of recanalization and/or to guide anticoagulation treatment ^(40,41))
 - When documentation is provided that the results will be used to guide anticoagulation treatment
 - When there is recurrent pain, headache or new neurologic deficits that suggest progression
- Known vasculitis, reversible cerebral vasoconstriction syndrome or Moyamoya disease ^{(2,26,27,34)31,42,43,44,45}

Pre-operative/procedural **PREOPERATIVE OR POSTOPERATIVE ASSESSMENT**

When not otherwise specified in the guideline:

Preoperative Evaluation ⁽¹⁵⁾:

- Pre-operative evaluation for a planned surgery or procedure
- Refractory trigeminal neuralgia or hemifacial spasm when done for surgical evaluation ^(46,47)₍₃₅₎
- Post-operative/procedural Imaging of the area requested is needed to develop a surgical

plan

Postoperative Evaluation^(48,49):

- 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.
- Known or suspected complications
- A clinical reason is provided how imaging may change management

NOTE: This section applies only within the first few months following surgery

FURTHER EVALUATION OF INDETERMINATE FINDINGS

Unless follow up is otherwise specified within the guideline:

- For initial evaluation of an inconclusive finding on a prior imaging report (i.e., x-ray, ultrasound or CT) that requires further clarification
- One follow-up exam of a prior indeterminate MR/CT finding to ensure no suspicious interval change has occurred. (No further surveillance unless specified as highly suspicious or change was found on last follow-up exam.)

IMAGING IN KNOWN GENETIC SYNDROMES AND RARE DISEASES CONDITIONS

- Fibromuscular dysplasia (FMD):^(50,51)
 - One-time vascular study from brain to pelvis
- Vascular Ehlers-Danlos syndrome:^(52,53)
- ADPKD (Autosomal Dominant Polycystic Kidney Disease)⁽³⁶⁾:
 - Every 10 years (including at diagnosis and then every)
- Fabry Disease⁽³⁷⁾:
 - Every 2 years (including at diagnosis) starting at age 18 months OR
 - More frequently if symptomatic
- Loeys-Dietz⁽³⁸⁾:
 - Every two years (including at diagnosis) OR

- More frequently if abnormalities are found
- Loeys-Dietz:⁽⁵⁴⁾
- Vascular Ehlers-Danlos syndrome (vEDS):⁽³⁹⁾
 - Every 18 months (including at diagnosis) OR
 - As clinically indicated to follow known vascular abnormalities

Combination Studies for Known Genetic Conditions

NOTE: When medical necessity is met for an individual study **AND** conscious sedation is required (such as for young pediatric patients or patients with significant developmental delay), the entire combination is indicated)

Brain MRI and MRA

- Fabry Disease:⁽³⁷⁾
 - Every 2 years (including at diagnosis) starting at age 18 OR
 - More frequently if symptomatic
- Sickle Cell Disease:^(40,41)
 - When needed to screen for silent stroke
 - Abnormal Transcranial Doppler Velocity > 200 cm/s
 - New neurologic or cognitive concerns (including TIA, no formal testing required)
 - When cessation or changing frequency of transfusions is under consideration

Brain/Neck/Chest/Abdomen/Pelvis MRA

- then Loeys-Dietz:⁽³⁸⁾
 - Every two years (including at diagnosis) OR
 - More frequently if abnormalities are found
- Spontaneous coronary arteries dissection (SCAD):⁽⁵⁵⁾
- One-time Vascular Ehlers-Danlos syndrome (vEDS):⁽³⁹⁾
 - Every 18 months (including at diagnosis) OR
 - As clinically indicated to follow known vascular study from brain to pelvis abnormalities

FABRY DISEASE ANNUAL NEUROLOGIC ASSESSMENT WITH BRAIN MRI/MRA EVERY TWO TO THREE YEARS BEGINNING AT AGE 18 YEARS⁽⁵⁶⁾

FOR OTHER SYNDROMES AND RARE DISEASES NOT OTHERWISE ADDRESSED IN THE GUIDELINE, COVERAGE IS BASED ON A CASE-BY-CASE BASIS USING SOCIETAL GUIDANCE OTHER COMBINATION STUDIES WITH BRAIN MRA AND

NOTE: When medical necessity is met for an individual study **AND** conscious sedation is required (such as for young pediatric patients or patients with significant developmental delay), the entire combination is indicated)

Brain/Neck MRA

- Recent ischemic stroke or transient ischemic attack^(21,22)attack (TIA)^(15,16)
 - **Note:** For remote strokes with no prior vascular imaging, imaging can be considered based on location/type of stroke and documented potential to change management
- Known or suspected vertebrobasilar insufficiency (VBI) in patients with symptoms such as dizziness, vertigo, headaches, diplopia, blindness, vomiting, ataxia, weakness in both sides of the body, or abnormal speech ^(23,24,25,26)_(17-19,31)
- Suspected carotid ⁽⁵⁷⁾₍₄₂₎ or vertebral ⁽⁵⁸⁾₍₄₃₎ artery dissection (secondary to trauma ⁽⁵⁹⁾₍₄₄₎ or spontaneous) due to weakness of vessel wall ^(22,27,28)_(16,20,45)
- FollowFollow-up of known carotid or vertebral artery dissection withinwith any ONE of the following^(16,32,33)
 - At 3-6 months post dissection (for evaluation of recanalization **and**/or to guide anticoagulation treatment ^(22,60,61))
 - Horner's syndrome, non-central (miosis, ptosis, and anhidrosis)⁽⁴⁸⁾
 - Large vessel vasculitis (When documentation is provided that the results will be used to guide anticoagulation treatment)
 - When there is recurrent pain, headache or new neurologic deficits that suggest progression
- Giant cell or Takayasuarteritis with suspected intracranial and extracranial involvement ⁽²⁴⁾
- Asymptomatic patients with an abnormal ultrasound of the neck or carotid duplex imaging (e.g., internal carotid stenosis $\geq 70\%$, technically limited study, aberrant direction of flow in the carotid or vertebral arteries) and patient is surgery or angioplasty candidate ^(62,63,64)_(16,46)
- Symptomatic patients with an abnormal ultrasound of the neck or carotid duplex imaging (e.g., internal carotid stenosis $\geq 50\%$, technically limited study, aberrant direction of flow in the carotid or vertebral arteries) and patient is surgery or angioplasty candidate ^(62,65)_(16,46)

- Pulsatile tinnitus to identify a suspected arterial vascular etiology ^{(19,20)(12,13)}

Brain MRI and Brain MRA

- Recent ischemic stroke or transient ischemic attack (TIA) ^{(21,22)(15,16)}
- Thunderclap headache with continued concern for underlying vascular abnormality (i.e., aneurysm or reversible cerebral vasoconstriction syndrome) after initial negative brain imaging ^{(11,12,13,14)(5-7,22)}
 - **Note:** Negative brain CT < 6 hours after headache onset excludes subarachnoid hemorrhage in neurologically intact patients. ⁽¹³⁾ ⁽⁷⁾ MRI lacks sensitivity in excluding subarachnoid hemorrhage less than 24 hours after headache onset. ^{(11,15)(5,8)}
- Acute, sudden onset of headache with personal history of a vascular abnormality or first-degree family history of aneurysm ^{(1,22)(14)}
- Headache associated with exercise, exertion, Valsalva or sexual activity ⁽¹¹⁾⁽⁵⁾
- SuspectedSuspected central venous thrombosis (dural sinus and ANY ONE of the following) ⁽¹⁶⁾
 - Patient has a hypercoagulable state such as pregnancy, post-partum, prothrombotic conditions (acquired or genetic), malignancy, oral contraceptive use, recent infection, recent trauma or Covid-19
 - Documentation of concern for central venous thrombosis ⁽²²⁾ MRI/MRV** is specified
 - Neurological Papilledema or signs/symptoms or symptoms in sickle cell patients ⁽²⁹⁾
 - High stroke risk in sickle cell patients (2-16 years of age) with a transcranial doppler velocity > 200 ⁽²⁹⁾ increased intracranial pressure
- **See Imaging in Known Genetic Conditions** for additional indications
- Known Moyamoya disease ^{(2,26)37} or reversible cerebral vasoconstriction with any new or changing neurological signs or symptoms ^{(14,38)(22,27)}
- Suspected secondary CNS vasculitis based on neurological signs or symptoms in the setting of an underlying systemic disease with abnormal inflammatory markers or autoimmune antibodies ⁽¹⁾
- Suspected primary CNS vasculitis based on neurological signs and symptoms with completed infectious/inflammatory lab work-up ^{(1,23,47)30,31}
- Giant cell arteritis with suspected intracranial involvement ⁽²⁴⁾
 - Fabry disease annual neurologic assessment with Brain MRI/MRA every two to three years beginning at age 18 years ⁽⁵⁶⁾

Brain MRI/ and Brain-MRA/Neck MRA ^(2,16)

- Recent ischemic stroke or transient ischemic attack (TIA) ^{(21,22)(15,16)}
- History of stroke and ONE of the following:

- No prior workup
- New neurologic signs or symptoms
- Suspected or known carotid or vertebral artery dissection with focal or lateralizing neurological deficits
- Pulsatile tinnitus with concern for a suspected arterial vascular and/or intracranial etiology ^(19,20) ₍₁₂₋₁₄₎
 - **NOTE:** For the indication of pulsatile tinnitus the Brain MRI of the combination should include the Internal Auditory Canal (IAC)
- Giant cell arteritis with suspected intracranial and extracranial involvement ⁽²⁴⁾
- ~~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~~ ⁽⁶⁶⁾

Note: CTA and MRA are generally comparable noninvasive imaging alternatives each with their own advantages and disadvantages. Brain MRI can be combined with Brain CTA/Neck CTA.

Brain/Neck/Chest MRA

- Non central Horner's syndrome (secondary/preganglionic or tertiary/post-ganglionic) for evaluation of underlying vascular source (such as dissection, aneurysm, arteritis) ^(48,49)

Brain/Neck/Chest/Abdomen/Pelvis MRA

Brain/Neck/Chest/Abdomen/Pelvis MRA

- For patients with fibromuscular dysplasia (FMD), a one-time vascular study from brain to pelvis ^(50,51) is indicated ^(28,29)
- ~~Vascular Ehlers-Danlos syndrome: At diagnosis and then every 18 months; more frequently if abnormalities are found~~ ^(52,53)
- ~~Loeys-Dietz: at diagnosis and then every two years, more frequently if abnormalities are found~~ ⁽⁵⁴⁾
- For assessment in patients with spontaneous coronary artery dissection (SCAD), (SCAD is a common initial diagnostic event for underlying fibromuscular dysplasia (FMD)) ⁽⁵⁰⁾
 - **NOTE:** Body vascular imaging for SCAD can be done performed at the time of coronary angiography ⁽⁶⁷⁾
- Takayasu's Arteritis ⁽²⁵⁾:
 - At initial diagnosis
 - Every 6 months for the first 2 years while on therapy
 - Annually after the first 2 years

CODING AND STANDARDS

Coding

CPT Codes

70544, 70545, 70546

Applicable Lines of Business

<input checked="" type="checkbox"/>	CHIP (Children's Health Insurance Program)
<input checked="" type="checkbox"/>	Commercial
<input checked="" type="checkbox"/>	Exchange/Marketplace
<input checked="" type="checkbox"/>	Medicaid
<input checked="" type="checkbox"/>	Medicare Advantage

BACKGROUND

General Overview

Magnetic resonance angiography (MRA) or magnetic resonance venography (MRV) can be used as a first-line investigation of intracranial vascular disease. It is an alternative to invasive intra-catheter angiography that was once the mainstay for the investigation of intracranial vascular disease. MRA/MRV may use a contrast agent, gadolinium, which is non-iodine-based, for better visualization. It can be used in patients who have history of contrast allergy and who are at high risk of kidney failure. A single authorization covers both MRA and MRV.

MRA and Non-Aneurysmal Vascular Malformations

Non-aneurysmal vascular malformations can be divided in low flow vascular malformations and high flow vascular malformations. Low flow vascular malformations include dural venous anomalies (DVA), cavernomas, and capillary telangiectasias. High flow vascular malformations include AVM and dural arteriovenous fistulas (dAVF). For low flow malformations, MRI is the study of choice. There is limited medical literature to support vascular imaging (CTA or MRA). CTA plays a limited role in the assessment of cavernoma but may be used to demonstrate a DVA. MRA is not usually helpful in the assessment of cavernoma, capillary telangiectasia, and DVA. Vascular imaging is indicated in high flow vascular malformations.^(1,2,68)

There is no evidence to support screening of first-degree relatives for AVMs.⁽⁶⁹⁾ The risk of having an AVM may be higher than in the general population, but absolute risk is low.

Pulsatile tinnitus

Pulsatile tinnitus has many etiologies, and the choice of study should be based on accompanying signs and symptoms. For general screening MRI brain with IAC/MRA brain and neck is approvable. If HHIIH is suspected (typically with headache and vision changes in a younger woman with a high BMI), MRI/MRV brain is indicated. If there is concern for vascular etiology, CTA or MRA brain/neck is indicated. If there is associated hearing loss and neurological signs/symptoms, MRI brain with IAC is indicated. If the temporal bone is suspected to be involved and/or retrotympatic lesion seen on otoscopy, CT temporal bone/IAC is indicated. If there is concurrent concern for boney and a vascular issue, CTA of the head and neck can be used to evaluate both.

MRA and Recent Stroke or Transient Ischemic Attack

- 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. 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.⁽⁷⁰⁾ 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.⁽⁷¹⁾
- 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.

MRA vs CTA for CVA

Preferred vascular imaging of the head and neck includes non-contrast head MRA and contrast-enhanced neck MRA. MRA may not be able to be performed in patients with claustrophobia, morbid obesity, or implanted device, but it can be useful in patients with renal failure or contrast allergies. In patients with high radiation exposure, MRA as an alternative should be considered. For acute stroke, CTA is preferred after CT (to rule of hemorrhage) and to look for thrombus/possible intervention that is time sensitive.^(2,22)

MRA and Intracerebral Hemorrhage⁽⁷²⁾

MRA is useful as a screening tool for an underlying vascular abnormality in the evaluation of spontaneous intracerebral hemorrhage (ICH). Etiologies of spontaneous ICH include tumor, vascular malformation, aneurysm, hypertensive arteriopathy, cerebral amyloid angiopathy, venous thrombosis, vasculitis, RCVS, drug induced vasospasm, venous sinus thrombosis, Moyamoya disease, anticoagulant use and hemorrhagic transformation of an ischemic infarct. History can help point to a specific etiology. Possible risk factors for the presence of underlying vascular abnormalities include age younger than 65, female, lobar or intraventricular location, and the absence of hypertension or impaired coagulation.

MRV and Central Venous Thrombosis **

MR Venogram is indicated for the evaluation of a central venous thrombosis/dural sinus thrombosis. The most frequent presentations are isolated headache, intracranial hypertension syndrome (headache, nausea/vomiting, transient visual obscurations, pulsatile tinnitus, CN VI palsy, papilledema)⁽⁷³⁾, 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, a thromboinflammatory response, and an increased incidence of venous thromboembolic events (VTE)^(74,75). Since venous thrombosis can cause SAH, infarctions, and hemorrhage, parenchymal imaging with MRI/CT is also appropriate^(76,77,78).

MRA and Dissection

Craniocervical dissections can be spontaneous or traumatic. Patients with blunt head or neck trauma who meet Denver Screening criteria should be assessed for cerebrovascular injury (although about 20% will not meet criteria). The criteria include focal or lateralizing neurological deficits (not explained by head CT); infarct on head CT; face, basilar skull, or cervical spine fractures; cervical hematomas that are not expanding; Glasgow coma score less than 8 without CT findings; massive epistaxis; cervical bruit or thrill.^(27,79,80)

Craniocervical dissections can be spontaneous or traumatic.

Spontaneous dissection presents with headache, neck pain with neurological signs or symptoms. There is often minor trauma or precipitating factor (i.e., exercise, neck manipulation). Dissection is thought to occur due to weakness of the vessel wall, and there may be an

underlying connective tissue disorder... Dissection of the extracranial vessels can extend intracranially and/or lead to thrombus which can migrate into the intracranial circulation, causing ischemia. Therefore, MRA of the head and neck is warranted. ^{(28,81)(20,51)}

Moyamoya Disease

Family members of Moyamoya Disease (MMD) patients may also have MMD, but not have any obvious symptoms. Routine screening should be implemented for all family members of MMD patients. TCD may be the preferred choice for screening because it is inexpensive and safe and as a high diagnostic agreement with MRA.

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. ⁽¹⁴⁾

Contraindications and Preferred Studies

- Contraindications and reasons why a CT/CTA cannot be performed may include: impaired renal function, significant allergy to IV contrast, pregnancy (depending on trimester).
- Contraindications and reasons why an MRI/MRA cannot be performed may include: impaired renal function, claustrophobia, non-MRI compatible devices (such as non-compatible defibrillator or pacemaker), metallic fragments in a high-risk location, patient exceeds weight limit/dimensions of MRI machine.

Acronyms / Abbreviations

ADPKD: Autosomal Dominant Polycystic Kidney Disease

AVM: Arteriovenous Malformation

CNS: Central Nervous System

CTA: Computed Tomography Angiography

CTV: Computed Tomography Venography

CVA: Cerebrovascular Accident

dAVF: Dural Arteriovenous Fistulas

DVA: Dural Venous Anomalies

IAC: Internal Auditory Canal

ICH: Intracerebral Hemorrhage

MDCTA: Multidetector CT Angiography

MMD: Moyamoya Disease
MRA: Magnetic Resonance Angiography
MRI: Magnetic Resonance Imaging
MRV: Magnetic Resonance Venography
SCAD: Spontaneous Coronary Arteries Dissection
SAH: Subarachnoid Hemorrhage
VBI: Vertebrobasilar Insufficiency
TIA: Transient Ischemic Attack

SUMMARY OF EVIDENCE

ACR Appropriateness Criteria® Cerebrovascular Diseases-Aneurysm, Vascular Malformation, and Subarachnoid Hemorrhage ⁽¹⁾

Study Design: The study design involves a detailed literature review and expert panel recommendations to establish imaging guidelines for various cerebrovascular conditions. The criteria are based on the latest evidence and expert consensus to ensure appropriate imaging procedures are selected for different clinical scenarios.

Target Population: The target population includes patients with cerebrovascular diseases such as aneurysms, vascular malformations, and SAH. Specific variants address different clinical presentations, including known acute SAH, suspected cerebral vasospasm, untreated cerebral aneurysms, previously treated cerebral aneurysms, high-risk cerebral aneurysm screening, known high-flow vascular malformations, and suspected CNS vasculitis.

Key Factors:

Imaging Recommendations: The document outlines the appropriateness of various imaging modalities, including arteriography, CTA, MRA, MRI, and ultrasound, for different clinical scenarios. Each variant provides specific recommendations based on the clinical presentation and the relative radiation level associated with each imaging procedure.

Clinical Presentations: The criteria cover a wide range of clinical presentations, from acute SAH to surveillance monitoring of untreated and treated aneurysms, as well as screening for high-risk populations and evaluation of suspected CNS vasculitis.

Expert Panel: The recommendations are developed by an expert panel on neurological imaging, including specialists from various institutions and organizations. The panel's collaboration ensures a comprehensive and well-rounded approach to imaging guidelines.

Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders, 3rd edition ⁽⁵⁾

Study Design: The ICHD-3 is a systematic classification of headache disorders based on extensive research and clinical studies. The classification is hierarchical, allowing for detailed

diagnosis from the first-digit level to the fifth. The criteria for each headache type are based on clinical features, diagnostic criteria, and evidence from field-testing studies.

Target Population: The target population includes individuals experiencing various types of headaches, ranging from primary headaches like migraines and tension-type headaches to secondary headaches attributed to other disorders. The classification is intended for use by healthcare professionals, including neurologists, general practitioners, and researchers, to diagnose and manage headache disorders.

Key Factors:

Primary Headaches: The document classifies primary headaches into categories such as migraines, tension-type headaches, and trigeminal autonomic cephalgias. Each category includes specific diagnostic criteria, clinical features, and comments on pathophysiology and treatment.

Secondary Headaches: These are headaches attributed to other disorders, such as trauma, vascular disorders, infections, and psychiatric disorders. The classification provides criteria for diagnosing secondary headaches based on the temporal relationship between the headache and the underlying disorder.

Diagnostic Criteria: The criteria for each headache type include the number of attacks, duration, pain characteristics, associated symptoms, and exclusion of other diagnoses. For example, migraine without aura requires at least five attacks lasting 4-72 hours with specific pain characteristics and associated symptoms like nausea and photophobia.

Field Testing: The classification includes results from field-testing studies that validate the diagnostic criteria. These studies involve large populations and use advanced diagnostic methods like neuroimaging and genetic testing.

Clinical and Research Applications: The ICHD-3 is designed for both clinical practice and research. It helps clinicians diagnose and manage headache disorders and provides a standardized framework for researchers to study headache epidemiology, pathophysiology, and treatment.

ACR Appropriateness Criteria® Cerebrovascular Diseases-Stroke and Stroke-Related Conditions⁽¹⁶⁾

Study Design: The document is a guideline developed by the American College of Radiology (ACR) Appropriateness Criteria Expert Panel on Neurological Imaging. It is based on a systematic analysis of medical literature from peer-reviewed journals and follows established methodology principles such as the Grading of Recommendations Assessment, Development, and Evaluation (GRADE) and the RAND/UCLA Appropriateness Method.

Target Population: The guidelines are intended for use by radiologists, radiation oncologists, and referring physicians in making decisions regarding radiologic imaging and treatment for patients with cerebrovascular diseases, including stroke and stroke-related conditions.

Key Factors:

Conditions Covered: The guidelines encompass a wide range of cerebrovascular diseases, including carotid stenosis, carotid dissection, intracranial large vessel

occlusion, and cerebral venous sinus thrombosis. They also address complications such as intraparenchymal hemorrhage and completed ischemic strokes.

Imaging Recommendations: The document provides evidence-based guidelines for appropriate imaging examinations for diagnosis and treatment of specified medical conditions. It includes recommendations for various imaging modalities such as CT, MRI, MRA, and ultrasound.

Clinical Scenarios: The guidelines cover different clinical scenarios, including transient ischemic attack (TIA), acute ischemic stroke, recent ischemic infarct, and known intraparenchymal hemorrhage, among others.

Methodology: The guideline development and revision process involves a multidisciplinary expert panel and supports the systematic analysis of medical literature. In instances where peer-reviewed literature is lacking or equivocal, expert opinions are used to formulate recommendations.

ANALYSIS OF EVIDENCE

Shared Conclusions ^(1,5,16):

- Diagnostic Imaging:** All three articles emphasize the importance of diagnostic imaging in identifying and managing cerebrovascular conditions. They discuss various imaging modalities such as CT, MRI, MRA, and CTA, highlighting their roles in diagnosing conditions like stroke, aneurysms, and vascular malformations.
- Clinical Guidelines:** The articles provide clinical guidelines for the management of cerebrovascular diseases. They stress the need for evidence-based approaches and the use of standardized criteria to ensure accurate diagnosis and effective treatment.
- Risk Factors:** Each article discusses the risk factors associated with cerebrovascular diseases, including hypertension, smoking, and genetic predispositions. They highlight the importance of identifying these risk factors to prevent and manage conditions effectively.

POLICY HISTORY SUMMARY

Date	Summary
<u>July 2025</u>	<ul style="list-style-type: none"> Fixed a sentence-spacing error in the Background Edited the policy history for June 2025 to better reflect the changes that were presented at committee. No clinical changes
<u>June 2025</u>	<ul style="list-style-type: none"> Guideline name changed from Brain MRA_MRV to Brain Magnetic Resonance Angiography (MRA) Guideline number changed from 004-2 to 2011

Date	Summary
	<ul style="list-style-type: none"> ● <u>Added new bullet-point to the General Statement section</u> ● <u>Checked the Medicare Advantage box in the Applicable Lines of Business table</u> ● <u>Added a Summary of Evidence and Analysis of Evidence</u> ● <u>Updated references</u> ● <u>Updated background section</u> ● <u>Updated combination section</u> <p><u>Updated G and RD section</u></p> <ul style="list-style-type: none"> ● <u>Updated and rearranged the genetic section</u> ● <u>Removed headache with Valsalva</u> <p><u>Removed Horner's and added to combo section</u></p> <ul style="list-style-type: none"> ● <u>Clarified carotid dissection follow-up</u> <u>Clarified low and high flow vascular malformation</u> ● <u>Clarified central Horner's</u> ● <u>Clarified CVT</u> ● <u>Clarified secondary CNS vasculitis</u> ● <u>Clarified follow-up of known carotid or vertebral artery dissection</u> ● <u>Added intervals for imaging of Takayasu arteritis</u> ● <u>Added history of stroke and no prior workup or new neurologic signs or symptoms</u>
June 2024	<ul style="list-style-type: none"> ● Updated references ● Updated background section ● Updated combination section ● Clarified <ul style="list-style-type: none"> ○ Frequency of screening in genetic syndromes ● Added <ul style="list-style-type: none"> ○ Screening for aneurysm in high-risk populations ○ Bicuspid aortic valve ○ Known aortic diseases (aneurysm, coarctation, dissection) ○ Suspected carotid or vertebral artery dissection; secondary to trauma or spontaneous due to weakness of vessel wall

Date	Summary
	<p>(already in combo)</p> <ul style="list-style-type: none"> ○ Follow-up of known carotid or vertebral artery dissection within 3-6 months for evaluation of recanalization and/or to guide anticoagulation treatment (already in combo) ○ Horner's syndrome, non-central (miosis, ptosis, and anhidrosis) - also in combo section ○ Vessel wall MRI (ordered as Brain MRI) can also be performed in the evaluation of vasculitides ○ Genetic syndromes and rare disease section. ○ Refractory trigeminal neuralgia or hemifacial spasm when done for surgical evaluation ○ Known Moyamoya disease or reversible cerebral vasoconstriction with any new or changing neurological signs or symptoms (Brain MRA/MRI combo) ○ Suspected secondary CNS vasculitis based on neurological signs or symptoms in the setting of an underlying systemic disease with abnormal inflammatory markers or autoimmune antibodies (Brain MRA /MRI combo) ○ Suspected primary CNS vasculitis based on neurological signs and symptoms with completed infectious/inflammatory lab work-up (Brain MRA /MRI combo) ○ Large vessels vasculitis with suspected intracranial and extracranial involvement (Brain MRA /Neck/ Brain MRI combo) ○ Giant cell arteritis with suspected intracranial involvement (combos) ● Deleted <ul style="list-style-type: none"> ○ MRI Brain with IAC/MRA Head/MRA Neck section
May 2023	<ul style="list-style-type: none"> ○ Updated and reformatted references ○ Updated background section ○ Added: <ul style="list-style-type: none"> ○ Section on further evaluation of indeterminate or questionable findings on prior imaging ○ Follow-up of known carotid or vertebral artery dissection within 3-6 months for evaluation of recanalization and/or to guide anticoagulation treatment (Combo Brain/Neck MRA) ○ Note: For remote strokes with no prior vascular imaging, imaging can be considered based on location/type of

Date	Summary
	<p>stroke and documented potential to change management (also in combo section)</p> <ul style="list-style-type: none"> o Note on CTA VS MRA o Clarified: <ul style="list-style-type: none"> o Screening for aneurysm in polycystic kidney disease (in adults) o Screening for intracranial aneurysm if two or more first-degree family members (parent brother, sister, or child) with history of intracranial aneurysm o For one first degree relative with aneurysm, asymptomatic screening is not indicated – would require a neurological sign or symptom supporting clinical concern for aneurysm. o Thunderclap headache with continued concern for underlying vascular abnormality (i.e. aneurysm or reversible cerebral vasoconstriction syndrome) after initial negative brain imaging o Note: MRI lacks sensitivity in excluding subarachnoid hemorrhage less than 24 hours after headache onset (also in Combo Brain MRI/MRA section) o Headache associated with exercise, exertion, Valsalva or sexual activity (Also in Combo Brain MRI/MRA) o Known subarachnoid hemorrhage (SAH) – CTA is favored over MRA o Deleted: <ul style="list-style-type: none"> o Vascular abnormality visualized on previous brain imaging that is equivocal or needs further evaluation

LEGAL AND COMPLIANCE

Guideline Approval

Committee

Reviewed / Approved by Evolent Specialty Services Clinical Guideline Review Committee

Disclaimer

Evolent Clinical Guidelines do not constitute medical advice. Treating health care professionals are solely responsible for diagnosis, treatment, and medical advice. Evolent uses Clinical Guidelines in accordance with its contractual obligations to provide utilization management. Coverage for services varies for individual members according to the terms of their health care coverage or government program. Individual members' health care coverage may not utilize some Evolent Clinical Guidelines. Evolent clinical guidelines contain guidance



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Evolent Clinical Guidelines are comprehensive and inclusive of various procedural applications for each service type. Our guidelines may be used to supplement Medicare criteria when such criteria is not fully established. When Medicare criteria is determined to not be fully established, we only reference the relevant portion of the corresponding Evolent Clinical Guideline that is applicable to the specific service or item requested in order to determine medical necessity.

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