

AmeriHealth Caritas Louisiana

National Imaging Associates, Inc.*	
Clinical guidelines BRAIN (HEAD) MRA/MRV	Original Date: September 1997
CPT Codes: 70544, 70545, 70546	Last Revised Date: May 2020
Guideline Number: NIA(CG)_004-2	Implementation Date: <u>January 2021</u><u>TBD</u>

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. All prior relevant imaging results, and the reason that alternative imaging (gold standard, protocol, contrast, etc.) cannot be performed must be included in the documentation submitted.

INDICATIONS FOR BRAIN (HEAD) MR_Angiography/MR_Venography:

For evaluation of known intracranial vascular disease:

~~(ACR, 2017; Sanelli, 2014; Obusez, 2014; Jageer, 2000; Singhal, 2016; Ancelet, 2015)~~

- ~~Known intracranial aneurysm or arteriovenous malformation (AVM).~~
- ~~Known vertebrobasilar insufficiency (VBI).~~
- ~~Vascular abnormality visualized on previous brain imaging.~~
- ~~Known vasculitis, reversible cerebral vasoconstriction syndrome or Moyamoya disease (Ancelet, 2015; Tarasow, 2011)~~

For evaluation of suspected intracranial vascular disease ~~:(ACR-CVD, 2017, 2019)~~

~~(ACR, 2017; Chalouhi, 2011; Khan, 2007; Lin, 2006; Ayanzen, 2000; Hofmann, 2013; Abboud, 2003; Ryan, 2003; Zuccoli, 2011)~~

Aneurysm screening

- **Screening for suspected intracranial aneurysm in patient with a first-degree familial history (parent brother, sister, or child) of intracranial aneurysm.**
Note: Repeat study is recommended every 5 years (Chalouhi, 2011).
- **Screening for aneurysm in polycystic kidney disease (after age 30), Ehlers-Danlos syndrome, Loeys-Dietz syndrome, fibromuscular dysplasia, spontaneous coronary arteries dissection (SCAD).**

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neurofibromatosis, or known aortic coarctation (Macaya, 2019; Hayes, 2018; Hitchcock, 2016; Macaya, 2019)

Vascular abnormalities

- Suspected high flow vascular malformation (arteriovenous malformation (AVM) or dural arteriovenous fistula) in patient with previous or indeterminate imaging study.
- Thunderclap headache with continued concern after initial negative work-up for underlying vascular abnormality after initial negative work-up (Whitehead, 2019, Yeh, 2010, Yuan, 2005):
 - Negative Brain (CT; AND/
 - Negative Lumbar Puncture; OR
 - Negative Brain or MRI) and continued concern for underlying vascular abnormality (Whitehead, 2019, Yeh, 2010, Yuan, 2005).
- Isolated third nerve palsy (oculomotor) with pupil involvement to evaluate for aneurysm (Pula, 2016).
- Pulsatile tinnitus to identify a vascular etiology (Hofmann, 2013; Pegge, 2017; Hofmann, 2013).

Cerebrovascular Disease

Ischemic

- Recent ischemic new-onset stroke or transient ischemic attack (See Background section) (Sanelli, 2014; Wintermark, 2013).
- Known or suspected vertebrobasilar insufficiency (VBI) in patients with symptoms such as dizziness, vertigo, headaches, diplopia, blindness, vomiting, ataxia and weakness in both sides of the body, or abnormal speech (Lima-Neto, 2017; Pirau, 2019; Lima-Neto, 2017; Searls, 2012).

Screening for suspected intracranial aneurysm in patient with a first-degree familial history (parent brother, sister, or child) of intracranial aneurysm. Note: Repeat study is recommended every 5 years.

Screening for aneurysm in polycystic kidney disease, Ehlers-Danlos syndrome, fibromuscular dysplasia, neurofibromatosis, or known aortic coarctation. Hemorrhagic

- Clinical suspicion of subarachnoid hemorrhage (SAH) (i.e., thunderclap headache) (Yeh, 2010)
- Known Non-traumatic Known subarachnoid hemorrhage (SAH)
- Spontaneous Non-traumatic Known intra-cerebral intraparenchymal hemorrhage with concern for underlying vascular abnormality

Known or suspected vertebrobasilar insufficiency (VBI) in patients with symptoms such as dizziness, vertigo, headaches, diplopia, blindness, vomiting, ataxia and weakness in both sides of the body, or abnormal speech (Lima-Neto, 2017; Searls, 2012)

Suspected arteriovenous malformation (AVM) in patient with previous or indeterminate imaging study. Venous- MRV*

- Suspected central venous thrombosis (dural sinus thrombosis) - ordered as MRV see background

— Distinguishing benign intracranial hypertension (pseudotumor cerebri) from dural sinus thrombosis (Agarwal, 2010; Aldossary, 2018; Agarwal, 2010).

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Sickle cells disease (ischemic and/or hemorrhagic) (Abboud, 2003; Thust, 2014; Abboud, 2003)

- Neurological signs or symptoms in sickle cell patients
- High stroke risk in sickle cell patients (2 - 16 years of age) with a transcranial doppler velocity > 200.

Pulsatile tinnitus to identify a vascular etiology.

Other Vasculitis intracranial vascular disease with initial laboratory workup (such as ESR, CRP, plasma viscosity) (Berlit, 2014)

- Suspected secondary CNS vasculitis based on neurological sign or symptoms in the setting of an underlying systemic disease with abnormal inflammatory markers or autoimmune antibodies WITH
 - with abnormal lab results suggesting acute inflammation in inflammatory markers or autoimmune antibodies (Berlit, 2014).
- Suspected primary CNS vasculitis based on neurological signs and symptoms WITH with
 - with completed infectious/inflammatory lab work-up (Godasi, 2019; Zucoli, 2011).

Suspected , reversible cerebral vasoconstriction syndrome or (Singhal, 2016).

Other intracranial vascular disease

- Suspected Moyomoya disease -(Ancelet, 2015; Tarasow, 2011).
- Suspected reversible cerebral vasoconstriction syndrome (Singhal, 2016).

(Ancelet, 2015; Godasi, 2019; Singhal, 2016; Tarasow, 2011)

Stroke risk in sickle cell patients (2 - 16 years of age) with a transcranial doppler velocity > 200.

Neurological signs or symptoms in sickle cell disease

Refractory trigeminal neuralgia when done for surgical planning (Leal, 2010)

For evaluation of known intracranial vascular disease (ACR-CVD, 2017, ; 2019):

- Known intracranial aneurysm or high flow vascular malformation (AVM or dural arteriovenous fistula)
- -Vascular abnormality visualized on previous brain imaging that is equivocal or needs further evaluation
- Known vertebrobasilar insufficiency (VBI) (Lima-Neto, 2017; Searls, 2012).
- Known vasculitis, reversible cerebral vasoconstriction syndrome or Moyomoya disease (Ancelet, 2015; Godasi, 2019; Obusez, 2014; Singhal, 2016; Ancelet, 2015; Obusez, 2014; Tarasow, 2011).

Other indications for a Brain MRA:

- Refractory trigeminal neuralgia when done for surgical planning (Leal, 2010).

Pre-operative/procedural evaluation for treatment, procedure, intervention, or brain/skull surgery (Farsad, 2009).

Post-operative/procedural evaluation (Lee, 2015; Serafin, 2012; Lee, 2015; Wong, 2007; Lee, 2009)

- 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.

Indications for Brain MRA/Neck MRA combination studies (ACR-CVD, 2017; 2019):

- Recent ischemic new onset stroke or transient ischemic attack (TIA) (Sanelli, 2014).
- Known or suspected vertebrobasilar insufficiency (VBI) in patients with symptoms such as dizziness, vertigo, headaches, diplopia, blindness, vomiting, ataxia and weakness in both sides of the body, or abnormal speech (Lima-Neto, 2017; Pirau, 2019; Lima-Neto, 2017; Searls, 2012).
- Known or suspected carotid or cerebral artery occlusion in patients with a sudden onset of one-sided weakness or numbness, abnormal speech, vision defects, incoordination or severe dizziness.
- Known or suspected vertebrobasilar insufficiency (VBI) in patients with symptoms such as dizziness, vertigo, headaches, diplopia, blindness, vomiting, ataxia and weakness in both sides of the body, or abnormal speech (Lima-Neto, 2017; Searls, 2012)
- Head trauma in a patient with closed head injury with suspected carotid or vertebral artery dissection; due to trauma or spontaneous injuries due to weakness of vessel wall leading to dissection. (Franz, 2012; Shakir, 2016; Franz, 2012).
- Pulsatile tinnitus to identify vascular etiology (Pegge, 2017).
- Asymptomatic patients with an abnormal ultrasound of the neck or carotid duplex imaging (e.g., carotid stenosis $\geq 70\%$, technically limited study, aberrant direction of flow in the carotid or vertebral arteries) and patient is surgery or angioplasty candidate (Brott, 2011; Brott, 2011; DaCosta, 2019; Brott, 2011; Marquardt, 2010)
- Symptomatic patients with an abnormal ultrasound of the neck or carotid duplex imaging (e.g., carotid stenosis $\geq 50\%$, technically limited study, aberrant direction of flow in the carotid or vertebral arteries) and patient is surgery or angioplasty candidate (AAN, 2010; Brott, 2011; Rerkasem, 2011)
- Pulsatile tinnitus to identify vascular etiology (Hofmann, 2013; Pegge, 2017; Hofmann, 2013). (Pegge, 2017).
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Indications for Brain MRI/Brain MRA combination studies (ACR-CVD, 2017; 2019)

- Recent ischemic new onset stroke or transient ischemic attack
- Clinical suspicion of subarachnoid hemorrhage (SAH) ie thunderclap headache
- Thunderclap headache after initial negative work-up (CT/LP) and continued concern for underlying vascular abnormality (Whitehead, 2019; Yeh, 2010; Yuan, 2005).

- 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 first-degree family history of aneurysm
- Suspected venous thrombosis (dural sinus thrombosis) – MRV*

Indications for Brain MRI/Brain MRA/Neck MRA combination studies:

- Recent stroke or transient ischemic attack (TIA)
- Recent ischemic new onset stroke or transient ischemic attack (TIA) (ACR-CVD, 2017, 2019)
- 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). 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).

BACKGROUND:

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.

Three different techniques of MRA/MRV are: time of flight (both 2D and 3D TOF), phase contrast (PC), and contrast enhanced angiography. Time of flight MRA takes advantage of the phenomena of flow related enhancement and is the preferred MRA technique due to the speed at which the exam can be acquired.

Recent stroke or transient ischemic attack (See Background section)

MRA and Cerebral Aneurysms – Studies that compared MRA with catheter angiography in detecting aneurysms found that MRA could find 77% - 94% of the aneurysms previously diagnosed by catheter angiography that were larger than 5 mm. For aneurysms smaller than 5 mm, MRI detected only 10% - 60% of those detected with catheter angiography. On the other hand, aneurysms that were missed by catheter angiography in patients with acute subarachnoid hemorrhage were detected with MRA, due to the much larger number of projections available with MRA (Chen, 2018).-

MRA and Cerebral Arteriovenous Malformations (AVM) – Brain arteriovenous malformation (AVM) may cause intracranial hemorrhage and is usually treated by surgery. 3D TOF-MRA is commonly used during the planning of radio-surgery to delineate the AVM nidus, but it is not highly specific for the detection of a small residual AVM after radio-surgery.

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 imagining (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. (ACR-EVD, 2017; Lee, 2012).

MRA and recent stroke or transient ischemic attack-

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.

MRA and Intracerebral Hemorrhage – MRA is useful as a screening tool for an underlying vascular (Bekelis, 2012; Delgado, 2009) 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, Moyomoya 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.

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.~~

MRV - A pitfall of the TOF technique, particularly 3D TOF, is that in areas of slowly flowing blood, turbulence or blood which flows in the imaging plane there can be regions of absent or diminished signal. The signal loss can be confused with vascular occlusion or thrombi. To avoid this pitfall MRA performed after the intravenous administration of gadolinium based contrast agents is utilized at many facilities.

Intracranial magnetic resonance venography (MRV) is used primarily to evaluate the patency of the venous sinuses. The study can be performed with TOF, Phase contrast and IV contrast enhanced techniques. Delayed images to allow for enhancement of the venous system are required to obtain images when intravenous gadolinium enhanced studies are undertaken.

Saturation pulses are utilized in studies not undertaken with intravenous contrast to help eliminate flow related signal in a specified direction and thus display the desired arterial or venous structures on their own. In cranial applications, saturation pulses applied at the inferior margin of the imaging field eliminate signal from arterial flow in order to visualize the veins. Conversely, superior saturation pulses are used to eliminate venous flow related enhancement when evaluation of the arterial structures is desired. (Ayanzen, 2000).

MRV and Central Venous Thrombosis* – a 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, 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. Since venous thrombosis can cause SAH, infarctions and hemorrhage parenchymal imaging with MRI/CT is also appropriate ([Ferro, 2016](#); Bushnell, 2014; Courinho, 2015; [Ferro, 2016](#)).

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, 2019).

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 (Franz, 2012; Liang, 2013; Mundinger, 2013; Simon, 2019). Spontaneous dissection presents with headache, neck pain with neurological signs or symptoms. There is often minor trauma or precipitating factor (ie 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 (Nash, 2019; Shakir, 2016).

POLICY HISTORY:

Review Date: July 2019

Review Summary:

- Added:
 - Reversible cerebral vasoconstriction syndrome or Moyomoya disease
 - Clinical suspicion of subarachnoid hemorrhage (SAH) (i.e., thunderclap headache)
 - Spontaneous intracerebral hemorrhage with concern for underlying vascular abnormality
 - Suspected primary CNS vasculitis with infectious/inflammatory lab work-up, reversible cerebral vasoconstriction syndrome or Moyomoya disease
 - Refractory trigeminal neuralgia when done for surgical planning
- Further clarified:
 - Suspected vertebrobasilar insufficiency (VBI) symptoms
 - MRV for suspected central venous thrombosis

- For Brain MRA/Neck MRA combo:
 - Removed the past two-week restriction from 'recent stroke or TIA'
 - Clarified CVA symptoms to include - known or suspected carotid or cerebral artery occlusion with sudden onset of numbness or incoordination
 - Added spontaneous injuries due to weakness of vessel wall leading to dissection
 - Added asymptomatic patients with an abnormal ultrasound of the neck or carotid duplex imaging (e.g. carotid stenosis $\geq 70\%$, technically limited study, aberrant direction of flow in the carotid or vertebral arteries) and patient is surgery or angioplasty candidate
 - Added symptomatic patients with an abnormal ultrasound of the neck or carotid duplex imaging (e.g. carotid stenosis $\geq 50\%$, technically limited study, aberrant direction of flow in the carotid or vertebral arteries) and patient is surgery or angioplasty candidate
- Added section for Brain MRI/Brain MRA combination studies, including:
 - Recent stroke or transient ischemic attack
 - Clinical suspicion of subarachnoid hemorrhage (SAH) ie thunderclap headache
 - Suspected venous thrombosis (dural sinus thrombosis)
- Added section for Brain MRI/Brain MRA/Neck MRA combination studies, including:
 - Recent stroke or transient ischemic attack (TIA)
 - 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
- Updated background info and refs

Review Date: May 2020

Review Summary:

- Updated background information references
- Reordered and categorized indications and background information

Clarified:

- Screening for aneurysm: polycystic kidney disease (after age 30)
- Suspected or known dural arteriovenous fistula as an example of a vascular malformation
- Recent ischemic stroke or transient ischemic attack (also in all combo sections)
- Cerebral intraparenchymal hemorrhage
- Suspected secondary CNS vasculitis based on neurological sign or symptoms in the setting of an underlying systemic disease
- Suspected primary CNS vasculitis based on neurological signs and symptoms
- Vascular abnormality visualized on previous brain imaging that is equivocal or needs further evaluation
- Reworded- Suspected carotid or vertebral artery dissection; due to trauma or spontaneous due to weakness of vessel wall leading to dissection – in the combo Neck/Brain MRA section

Added:

- Screening for aneurysm: Loeys-Dietz syndrome
- Thunderclap headache with continued concern for underlying vascular abnormality after initial negative work-up
 - Negative Brain CT; AND
 - Negative Lumbar Puncture; OR
 - Negative Brain MRI
- Isolated third nerve palsy (oculomotor) with pupil involvement to evaluate for aneurysm
- Vasculitis with initial laboratory workup (such as ESR, CRP, plasma viscosity)
- Thunderclap headache with continued concern for underlying vascular abnormality after initial negative work-up – in combo Brain MRI/MRA section
 - Negative Brain CT; AND
 - Negative Lumbar Puncture; OR
- Acute, sudden onset of headache with personal history of a vascular abnormality or first-degree family history of aneurysm – in combo Brain MRI/MRA section

Deleted

- Screening for aneurysm: Ehlers-Danlos syndrome, neurofibromatosis
- Clinical suspicion of subarachnoid hemorrhage (SAH) (i.e., thunderclap headache)
- Known or suspected carotid or cerebral artery occlusion in patients with a sudden onset of one-sided weakness or numbness, abnormal speech, vision defects, incoordination or severe dizziness - in the combo Neck/Brain MRA section
- Clinical suspicion of subarachnoid hemorrhage (SAH) (i.e., thunderclap headache) - in the combo MRI/MRA section

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