

**American College of Radiology
ACR Appropriateness Criteria®
Cerebrovascular Disease-Child**

Variant: 1 Child age older than 6 months. Emergent imaging for clinical presentation suggestive of acute nonsickle-cell related stroke. New focal fixed or worsening neurologic defect lasting less than 24 hours from last seen normal state. No contraindications to emergent intervention. Initial imaging.

Procedure	Appropriateness Category	Peds Relative Radiation Level
MRA head without IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
CT head without IV contrast	Usually Appropriate	⊕⊕⊕
MRI head perfusion with IV contrast	May Be Appropriate	O
MRI head perfusion without IV contrast	May Be Appropriate	O
MRI head without and with IV contrast	May Be Appropriate	O
CTA head with IV contrast	May Be Appropriate	⊕⊕⊕⊕
Arteriography cervicocerebral	Usually Not Appropriate	⊕⊕⊕⊕
MRA head with IV contrast	Usually Not Appropriate	O
MRI head with IV contrast	Usually Not Appropriate	O
CT head perfusion with IV contrast	Usually Not Appropriate	
CT head with IV contrast	Usually Not Appropriate	⊕⊕⊕
CT head without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕

Variant: 2 Child. Clinical presentation suggestive of acute stroke, not a candidate for emergent intervention. Initial imaging.

Procedure	Appropriateness Category	Peds Relative Radiation Level
MRA head without IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
CT head without IV contrast	Usually Appropriate	⊕⊕⊕
MRI head perfusion with IV contrast	May Be Appropriate	O
MRI head perfusion without IV contrast	May Be Appropriate	O
MRI head without and with IV contrast	May Be Appropriate	O
CTA head with IV contrast	May Be Appropriate	⊕⊕⊕⊕
US duplex Doppler transcranial	Usually Not Appropriate	O
Arteriography cervicocerebral	Usually Not Appropriate	⊕⊕⊕⊕
MRA head with IV contrast	Usually Not Appropriate	O
MRI head with IV contrast	Usually Not Appropriate	O
CT head perfusion with IV contrast	Usually Not Appropriate	
CT head with IV contrast	Usually Not Appropriate	⊕⊕⊕
CT head without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕

Variant: 3 Child. Clinical presentation suggestive of acute stroke, known or suspected arteriopathy, or moyamoya. Not a candidate for emergent treatment. Initial imaging.

Procedure	Appropriateness Category	Peds Relative Radiation Level
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MRA head without IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
MRI head perfusion with IV contrast	May Be Appropriate	O
MRI head perfusion without IV contrast	May Be Appropriate	O
MRI head without and with IV contrast	May Be Appropriate	O
CT head without IV contrast	May Be Appropriate	⊕⊕⊕⊕
CTA head with IV contrast	May Be Appropriate	⊕⊕⊕⊕⊕
US duplex Doppler transcranial	Usually Not Appropriate	O
Arteriography cervicocerebral	Usually Not Appropriate	⊕⊕⊕⊕⊕
MRA head with IV contrast	Usually Not Appropriate	O
MRI head with IV contrast	Usually Not Appropriate	O
CT head perfusion with IV contrast	Usually Not Appropriate	
CT head with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕
CT head without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕⊕
SPECT or SPECT/CT brain perfusion	Usually Not Appropriate	⊕⊕⊕⊕⊕

Variant: 4 Child. Known or suspected cervicocranial arterial dissection based on clinical or imaging findings. Next imaging study.

Procedure	Appropriateness Category	Peds Relative Radiation Level
MRA head and neck without IV contrast	Usually Appropriate	O
MRI head and neck without IV contrast	Usually Appropriate	O
CTA head and neck with IV contrast	Usually Appropriate	⊕⊕⊕⊕⊕
Arteriography cervicocerebral	May Be Appropriate	⊕⊕⊕⊕⊕
MRA head and neck with IV contrast	May Be Appropriate (Disagreement)	O
MRI head and neck with IV contrast	May Be Appropriate (Disagreement)	O
MRI head and neck without and with IV contrast	May Be Appropriate	O
CT head without IV contrast	May Be Appropriate	⊕⊕⊕⊕
US duplex Doppler transcranial and carotid artery	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕
CT head without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕⊕

Variant: 5 Child. Clinical presentation suggestive of acute stroke, known or suspected central nervous system vasculitis. Initial imaging.

Procedure	Appropriateness Category	Peds Relative Radiation Level
MRA head without IV contrast	Usually Appropriate	O
MRI head without and with IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
CTA head with IV contrast	Usually Appropriate	⊕⊕⊕⊕⊕
Arteriography cervicocerebral	May Be Appropriate	⊕⊕⊕⊕⊕
MRA head with IV contrast	May Be Appropriate	O
MRI head with IV contrast	May Be Appropriate (Disagreement)	O
CT head without IV contrast	May Be Appropriate	⊕⊕⊕⊕
US duplex Doppler transcranial	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕⊕

CT head without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕
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Variant: 6 Child. Nontraumatic intraparenchymal hemorrhage (hematoma) found on CT or MRI. Unknown etiology. Next imaging study.

Procedure	Appropriateness Category	Peds Relative Radiation Level
Arteriography cervicocerebral	Usually Appropriate	⊕⊕⊕⊕
MRA head without IV contrast	Usually Appropriate	O
MRI head without and with IV contrast	Usually Appropriate	O
CTA head with IV contrast	Usually Appropriate	⊕⊕⊕⊕
MRA head with IV contrast	May Be Appropriate	O
MRI head with IV contrast	May Be Appropriate (Disagreement)	O
MRI head without IV contrast	May Be Appropriate (Disagreement)	O
US duplex Doppler transcranial	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕
CT head without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕
CT head without IV contrast	Usually Not Appropriate	⊕⊕⊕

Variant: 7 Child. Nontraumatic subarachnoid hemorrhage (SAH) detected by noncontrast CT. Next imaging study.

Procedure	Appropriateness Category	Peds Relative Radiation Level
Arteriography cervicocerebral	Usually Appropriate	⊕⊕⊕⊕
MRA head without IV contrast	Usually Appropriate	O
CTA head with IV contrast	Usually Appropriate	⊕⊕⊕⊕
MRA head with IV contrast	May Be Appropriate	O
MRI head without and with IV contrast	May Be Appropriate	O
MRI head without IV contrast	May Be Appropriate	O
US duplex Doppler transcranial	Usually Not Appropriate	O
MRI head with IV contrast	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕
CT head without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕
CT head without IV contrast	Usually Not Appropriate	⊕⊕⊕

Variant: 8 Child. Clinical presentation suggestive of acute stroke, known or suspected high-flow vascular anomaly. Initial imaging.

Procedure	Appropriateness Category	Peds Relative Radiation Level
MRA head without IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
CT head without IV contrast	Usually Appropriate	⊕⊕⊕
CTA head with IV contrast	Usually Appropriate	⊕⊕⊕⊕
Arteriography cervicocerebral	May Be Appropriate (Disagreement)	⊕⊕⊕⊕
MRA head with IV contrast	May Be Appropriate	O
MRI head without and with IV contrast	May Be Appropriate	O
US duplex Doppler transcranial	Usually Not Appropriate	O
MRI head with IV contrast	Usually Not Appropriate	O

CT head with IV contrast	Usually Not Appropriate	⊕⊕⊕
CT head without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕

Variant: 9 Child. Clinical presentation suggestive of acute stroke, known or suspected low-flow vascular anomaly. Initial imaging.

Procedure	Appropriateness Category	Peds Relative Radiation Level
MRI head without IV contrast	Usually Appropriate	O
CT head without IV contrast	Usually Appropriate	⊕⊕⊕
MRA head without IV contrast	May Be Appropriate (Disagreement)	O
MRI head without and with IV contrast	May Be Appropriate	O
CTA head with IV contrast	May Be Appropriate (Disagreement)	⊕⊕⊕⊕
US duplex Doppler transcranial	Usually Not Appropriate	O
Arteriography cervicocerebral	Usually Not Appropriate	⊕⊕⊕⊕
MRA head with IV contrast	Usually Not Appropriate	O
MRI head with IV contrast	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	⊕⊕⊕
CT head without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕

Variant: 10 Child. Clinical presentation suggestive of acute stroke, known or suspected cortical vein or dural venous sinus thrombosis. Initial imaging.

Procedure	Appropriateness Category	Peds Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
MRV head with IV contrast	Usually Appropriate	O
MRV head without IV contrast	Usually Appropriate	O
CT head without IV contrast	Usually Appropriate	⊕⊕⊕
CTV head with IV contrast	Usually Appropriate	⊕⊕⊕⊕
US duplex Doppler transcranial	Usually Not Appropriate	O
Arteriography cervicocerebral	Usually Not Appropriate	⊕⊕⊕⊕
MRI head with IV contrast	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	⊕⊕⊕
CT head without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕

Variant: 11 Child. Clinical presentation suggestive of acute stroke, sickle cell disease. New focal fixed or worsening neurologic defect. Initial imaging.

Procedure	Appropriateness Category	Peds Relative Radiation Level
MRA head without IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
CT head without IV contrast	Usually Appropriate	⊕⊕⊕
US duplex Doppler transcranial	Usually Not Appropriate	O
Arteriography cervicocerebral	Usually Not Appropriate	⊕⊕⊕⊕
MRA head with IV contrast	Usually Not Appropriate	O
MRI head with IV contrast	Usually Not Appropriate	O
MRI head without and with IV contrast	Usually Not Appropriate	O

CT head with IV contrast	Usually Not Appropriate	⊕⊕⊕
CT head without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕
CTA head with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕

Panel Members

Richard L. Robertson, MD^a, Susan Palasis, MD^b, Michael J. Rivkin, MD^c, Sumit Pruthi, MD, MBBS^d, Twyla B. Bartel, DO, MBA^e, Nilesh K. Desai, MD^f, Nadja Kadom, MD^g, Abhaya V. Kulkarni, MD^h, H.F. Samuel Lam, MD, MPHⁱ, Mohit Maheshwari, MD^j, Sarah S. Milla, MD^k, David M. Mirsky, MD^l, John S. Myseros, MD^m, Sonia Partap, MD, MSⁿ, Rupa Radhakrishnan, MD^o, Bruno P. Soares, MD^p, Andrew T. Trout, MD^q, Unni K. Udayasankar, MD^r, Matthew T. Whitehead, MD^s, Boaz Karmazyn, MD^t

Summary of Literature Review

Introduction/Background

Stroke is an infrequent but an important and under-recognized cause of morbidity in children. Strokes may be due to either brain ischemia or intracranial hemorrhage. The incidence of pediatric acute stroke is estimated to be between 1.5 and 13/100,000 children per year [1-3]. There is a slight male predominance for both ischemic (58%) and hemorrhagic (65%) strokes [4]. Common symptoms of pediatric acute stroke include headache (56%), vomiting (36%), focal weakness (35%), numbness (24%), visual disturbance (23%), seizures (21%), and altered consciousness (21%) [5]. There are a number of stroke mimics in children with "stroke-like" symptoms, including migraine (38%), seizures with postictal paralysis (15%), and Bell's palsy (10%) [5]. Because of stroke mimics in children and the frequent lack of specificity in symptoms, the diagnosis may be delayed, especially in infants and young children [6]. Pediatric stroke mortality ranges from 5% to 20%, depending on the cause of the stroke, and is divided into perinatal (28 weeks gestation to 28 days of life) and childhood (29 days to 19 years of age) [3,4,7]. Imaging plays a critical role in the assessment of children with possible stroke and especially in children who present with acute onset of stroke symptoms.

Perinatal stroke (<6 months of age), although the most common acute stroke in children (20 to 62.5/100,000 live births), will not be discussed in this topic. As in many cases, the diagnosis is retrospective when the child presents later in life with new onset seizure, asymmetric motor function, or failure of developmental milestones [8]. The majority of perinatal stroke is likely to be caused by thromboembolism from the placenta through a patent foramen ovale or fetal heart defect [8].

Discussion of Procedures by Variant

Variant 1: Child age older than 6 months. Emergent imaging for clinical presentation suggestive of acute nonsickle-cell related stroke. New focal fixed or worsening neurologic defect lasting less than 24 hours from last seen normal state. No contraindications to emergent intervention. Initial imaging.

Given the morbidity and mortality associated with acute stroke in children, thrombolysis has a high potential for benefit that should be balanced with the risk for intracranial hemorrhage. Because

there are little data on benefits and risks of children treated with thrombolysis, there is significant controversy on the management of these children. As a result of lack of safety and efficacy data and because thrombolysis is not approved by the FDA for use in childhood stroke, <2% of children with acute ischemic stroke receive tissue-type plasminogen activator. Both intravenous (IV) and intra-arterial tissue-type plasminogen activator have been used in a few small case series [2,9,10]. In 2010, the National Institute of Neurological Disorders and Stroke initiated a prospective treatment trial in acute pediatric stroke, the Thrombolysis in Pediatric Stroke trial [2,11]. In this trial, 46% had confirmed arterial ischemic stroke and 44% had stroke mimic, 22% had medical contraindication to thrombolysis, and 25% presented outside of the therapeutic window [2]. The study was closed by the NIH for lack of patient accrual [2].

To date, there is insufficient information about acute pediatric stroke to confirm the appropriate window for initiation of thrombolytic therapy in children [12]. A window of 24 hours from stroke onset to initiation of treatment is used in this clinical scenario. This treatment is considered in children >2 years of age [13]. The risk of symptomatic hemorrhage into an ischemic infarction in adults treated with IV tissue-type plasminogen activator is 6.4% and is unknown in children [2]. Recently, interest in the use of intra-arterial treatment for stroke in children has developed because of the documented success of this type of therapy in adults. However, currently, no systematic study of such therapy in children has been performed, and only case reports exist [14]. Adult studies have shown good results with mechanical thrombectomy up to 6 hours or longer on a case-by-case basis from acute stroke symptom onset [15].

Variant 1: Child age older than 6 months. Emergent imaging for clinical presentation suggestive of acute nonsickle-cell related stroke. New focal fixed or worsening neurologic defect lasting less than 24 hours from last seen normal state. No contraindications to emergent intervention. Initial imaging.

A. CT Head

CT of the head is frequently the front-line imaging study for the assessment of a child with a suspected acute stroke. Although CT is less sensitive than MRI for the early depiction of acute ischemic infarction, the technique is rapid, usually does not require the child to be sedated, and can be useful in evaluating for hemorrhage or in excluding other treatable pathologies [16].

Variant 1: Child age older than 6 months. Emergent imaging for clinical presentation suggestive of acute nonsickle-cell related stroke. New focal fixed or worsening neurologic defect lasting less than 24 hours from last seen normal state. No contraindications to emergent intervention. Initial imaging.

B. CTA Head

CT angiography (CTA) can provide a useful assessment of intracranial vessels in arteriopathies and thromboembolic disease [17].

Variant 1: Child age older than 6 months. Emergent imaging for clinical presentation suggestive of acute nonsickle-cell related stroke. New focal fixed or worsening neurologic defect lasting less than 24 hours from last seen normal state. No contraindications to emergent intervention. Initial imaging.

C. MRI Head

The imaging procedure of choice for acute pediatric stroke detection is emergent MRI with diffusion-weighted imaging (DWI) [6,16-19]. MRI with DWI in the first 3 hours after symptom onset has been shown to be more sensitive for the demonstration of acute ischemic infarction (MRI, 77%,

versus CT, 16%) and is considered superior to CT for the detection of infarction for up to 12 hours after symptom onset [19]. The same clinical considerations around emergent stroke intervention in children noted for emergent CT evaluation hold true for emergent MRI evaluation. In addition, MRI is able to detect significant intracranial hemorrhage as readily as CT [18,20-23]. Susceptibility-weighted imaging (SWI) may improve the detection of blood products and the depiction of cerebral venous structures [20,24]. Children often present with stroke mimics like postictal paralysis of complicated migraine or migraine-like headaches, and SWI can sometimes demonstrate characteristic susceptibility changes that can help make the diagnosis [25]. "Blooming" of susceptibility artifact within a vessel suggests intravascular thrombus. In adults, the size of the parenchymal abnormality on DWI correlates strongly with final stroke volume [19].

Variant 1: Child age older than 6 months. Emergent imaging for clinical presentation suggestive of acute nonsickle-cell related stroke. New focal fixed or worsening neurologic defect lasting less than 24 hours from last seen normal state. No contraindications to emergent intervention. Initial imaging.

D. MRA Head

MR angiography (MRA) can provide information on the intracranial vasculature and is particularly helpful in noninvasive assessment of arteriopathies [17]. MRA is susceptible to flow-related artifacts that may simulate regions of stenosis, especially in the setting of turbulence that may occur with anemia at vessel branch points [26]. MRI and MRA of the cervical vessels in cases of unexplained stroke should be considered, with attention paid to the time constraints of the window of therapeutic intervention [27]. MRA can be performed with IV contrast to improve vessel delineation, although contrast is not typically required to produce diagnostic imaging in acute stroke.

Variant 1: Child age older than 6 months. Emergent imaging for clinical presentation suggestive of acute nonsickle-cell related stroke. New focal fixed or worsening neurologic defect lasting less than 24 hours from last seen normal state. No contraindications to emergent intervention. Initial imaging.

E. CT Head Perfusion

Perfusion CT in children is feasible but requires repetitive imaging of the brain [28].

Variant 1: Child age older than 6 months. Emergent imaging for clinical presentation suggestive of acute nonsickle-cell related stroke. New focal fixed or worsening neurologic defect lasting less than 24 hours from last seen normal state. No contraindications to emergent intervention. Initial imaging.

F. MR Head Perfusion

Perfusion-weighted MR can be performed with either dynamic susceptibility contrast administration or arterial spin-label techniques without IV contrast and can provide information on the adequacy of cerebral blood flow [17,19,29]. Perfusion MR is not necessary to proceed to emergent acute stroke thrombolysis.

Variant 1: Child age older than 6 months. Emergent imaging for clinical presentation suggestive of acute nonsickle-cell related stroke. New focal fixed or worsening neurologic defect lasting less than 24 hours from last seen normal state. No contraindications to emergent intervention. Initial imaging.

G. Arteriography Cervicocerebral

Cerebral angiography remains the most definitive means of imaging abnormalities of the cerebral vasculature in children. In experienced centers, there is a low incidence of neurologic (0%–1%) or

other serious complications [30,31]. Cerebral angiography is not necessary for emergent acute thrombolysis. It may be performed as part of thrombectomy in the setting of emergent acute stroke intervention if this is a viable therapeutic option for the child.

Variant 2: Child. Clinical presentation suggestive of acute stroke, not a candidate for emergent intervention. Initial imaging.

Suspected acute nonsickle cell-related stroke with neurologic symptoms lasting >24 hours from last seen normal state does not meet the criteria for Variant 1. If clinical or imaging findings of conditions in Variants 3 through 11 are present, the imaging should be tailored according to those specific variants. Ischemic stroke risk factors in children differ from those in neonates and adults and include arteriopathies (53%), cardiac disorders (31%), infection (24%), acute or chronic head and neck disorders (23%), acute or chronic systemic conditions (41%), and other causes (24%), [1,5]. Fifty-two percent of children have multiple risk factors [5]. Ischemic strokes affect the anterior circulation in 83% of children with isolated posterior circulation involvement in 15% [27]. Basilar circulation strokes account for 4.6% and are most often due to basilar artery occlusion or vertebral artery dissection [32,33].

Variant 2: Child. Clinical presentation suggestive of acute stroke, not a candidate for emergent intervention. Initial imaging.

A. CT Head

CT of the head is frequently the initial imaging study for the assessment of a child with a suspected acute stroke. Although CT is less sensitive than MRI for the early depiction of acute ischemic infarction, the technique is rapid, usually does not require the child to be sedated, and can be useful in evaluating for hemorrhage or in excluding other treatable pathologies [16].

Variant 2: Child. Clinical presentation suggestive of acute stroke, not a candidate for emergent intervention. Initial imaging.

B. CTA Head

CTA can provide a useful assessment of intracranial vessels in arteriopathies and thromboembolic disease [17].

Variant 2: Child. Clinical presentation suggestive of acute stroke, not a candidate for emergent intervention. Initial imaging.

C. MRI Head

The imaging procedure of choice for pediatric stroke detection is MRI with DWI [6,16-19]. MRI with DWI in the first 3 hours after symptom onset has been shown to be more sensitive for the demonstration of acute ischemic infarction (MRI, 77%, versus CT, 16%) and is considered superior to CT for the detection of infarction for up to 12 hours after symptom onset [19]. In addition, MRI is able to detect significant intracranial hemorrhage as readily as CT [18,20-23]. SWI may improve the detection of blood products and the depiction of cerebral venous structures [20,24].

"Blooming" of susceptibility artifact within a vessel suggests intravascular thrombus. In adults, the size of the parenchymal abnormality on DWI correlates strongly with final stroke volume [19]. Very high resolution MRI is now being used to directly image the vessel wall and may show thickening or enhancement of the wall in arteriopathies.

Variant 2: Child. Clinical presentation suggestive of acute stroke, not a candidate for emergent intervention. Initial imaging.

D. MRA Head

MRA can provide information on the intracranial vasculature and is particularly helpful in

noninvasive assessment of arteriopathies [17]. MRA is susceptible to flow-related artifacts that may simulate regions of stenosis, especially in the setting of turbulence that may occur with anemia at vessel branch points [26]. MRI and MRA should include the cervical vessels in cases of unexplained stroke because cerebral arterial abnormalities are found in 25% of patients [27].

Variant 2: Child. Clinical presentation suggestive of acute stroke, not a candidate for emergent intervention. Initial imaging.

E. CT Head Perfusion

Perfusion CT in children is feasible but requires repetitive imaging of the brain [28].

Variant 2: Child. Clinical presentation suggestive of acute stroke, not a candidate for emergent intervention. Initial imaging.

F. MR Head Perfusion

Perfusion-weighted MR can be performed with either dynamic susceptibility contrast administration or arterial spin-label techniques without IV contrast and can provide information on the adequacy of cerebral blood flow [17,19,29].

Variant 2: Child. Clinical presentation suggestive of acute stroke, not a candidate for emergent intervention. Initial imaging.

G. Arteriography Cervicocerebral

Cerebral angiography remains the most definitive means of imaging abnormalities of the cerebral vasculature in children. In experienced centers, there is a low incidence of neurologic complications (0%–1%) or other serious complications [30,31].

Variant 2: Child. Clinical presentation suggestive of acute stroke, not a candidate for emergent intervention. Initial imaging.

H. US Duplex Doppler Transcranial

Using an open fontanel as an acoustic window, ultrasound (US) can be used to diagnose infarction in the neonate but MRI, as in the older child, is more definitive. US shows 68% of infarctions in the first 3 days of life and 87% within the first 2 weeks [34]. Detailed evaluation of the brain parenchyma is generally not possible with US following the closure of the fontanel, but Doppler US can be used to interrogate flow in the intracranial vessels [35].

Variant 3: Child. Clinical presentation suggestive of acute stroke, known or suspected arteriopathy, or moyamoya. Not a candidate for emergent treatment. Initial imaging.

Arteriopathies, defined as intrinsic vessel stenosis, irregularity, pseudoaneurysm, banding, or dissection flap, account for 18% to 64% of pediatric ischemic stroke cases [36]. Common causes of arteriopathy include moyamoya (22%), arterial dissection (15% to 20%), vasculitis (12%), and sickle cell disease (SCD) arteriopathy (8%) [27,36]. Genetic mutation constitutes an increasingly represented etiology of cerebral arteriopathy as demonstrated by mutations in the ACTA2 or CERC 1 genes. Arteriopathies are a strong indicator of recurrent stroke risk (66%) [36-38]. Moyamoya refers to the angiographic appearance of a progressive stenosis or occlusion of the internal carotid artery apex and proximal branches of the circle of Willis with the development of stereotypical collaterals. Moyamoya occurs in approximately one in every 1 million children in the United States and accounts for 6% of all pediatric strokes [39]. The underlying cause is unclear, but it is likely that many different factors, both genetic and environmental, contribute to developing the arteriopathy. The moyamoya syndrome may be idiopathic (moyamoya disease) or occur in association with other conditions (moyamoya syndrome). Up to 40% of children with SCD may show moyamoya-like changes on imaging [39]. Moyamoya disease is, by definition, bilateral but may be asymmetric

in severity [39]. Unilateral involvement is considered moyamoya syndrome. As many as 12% of patients with sickle cell anemia will have a clinically detected stroke by the age of 20 [40,41]. Importantly, at a minimum, 85% of patients with sickle cell anemia who present with frank stroke will have evidence of cerebral arteriopathy on neuroimaging [41,42]. Most strokes in moyamoya in children are ischemic and most frequently occur in the vascular border zone territories, but cortical infarctions can occur as well.

Variant 3: Child. Clinical presentation suggestive of acute stroke, known or suspected arteriopathy, or moyamoya. Not a candidate for emergent treatment. Initial imaging.

A. CT Head

CT is less sensitive than MRI for demonstrating acute infarctions but can be useful in evaluating rare instances of intracranial hemorrhage in moyamoya [43].

Variant 3: Child. Clinical presentation suggestive of acute stroke, known or suspected arteriopathy, or moyamoya. Not a candidate for emergent treatment. Initial imaging.

B. CTA Head

CTA, including multiphase CTA, is an alternative to MRA and catheter angiography in the initial diagnosis and follow-up of children with moyamoya.

Variant 3: Child. Clinical presentation suggestive of acute stroke, known or suspected arteriopathy, or moyamoya. Not a candidate for emergent treatment. Initial imaging.

C. MRI Head

Proposed screening guidelines for moyamoya include diagnostic imaging using MRI/MRA in at-risk populations, particularly if there are symptoms of cerebral ischemia. Screening of first-degree relatives of patients with moyamoya is probably not warranted in the absence of symptoms or unless multiple family members are already known to be affected. T2-weighted fluid-attenuated inversion-recovery imaging may show high signal in the sulci (ivy sign), indicative of slow flow in affected vascular territories. A similar appearance may be evident on T1-weighted contrast-enhanced studies. SWI may be used to demonstrate microhemorrhage that has been reported in up to 52% of patients [44]. Arteriopathy can also be further evaluated with the technique of vessel-wall imaging [15,45].

Variant 3: Child. Clinical presentation suggestive of acute stroke, known or suspected arteriopathy, or moyamoya. Not a candidate for emergent treatment. Initial imaging.

D. MRA Head

MRA is an alternative to CTA in the initial and follow-up imaging evaluation of children with moyamoya and is the preferred noninvasive vascular imaging modality. MRA is typically performed together with brain MRI to evaluate both the cerebral vasculature and the brain parenchyma.

Variant 3: Child. Clinical presentation suggestive of acute stroke, known or suspected arteriopathy, or moyamoya. Not a candidate for emergent treatment. Initial imaging.

E. CT Head Perfusion

The use of perfusion CT, including xenon-enhanced CT, in children with moyamoya is feasible but requires repetitive imaging of the brain [28].

Variant 3: Child. Clinical presentation suggestive of acute stroke, known or suspected arteriopathy, or moyamoya. Not a candidate for emergent treatment. Initial imaging.

F. MR Head Perfusion

MR cerebral blood flow evaluation includes perfusion-weighted MR, either using arterial spin-

labeling techniques without IV contrast or dynamic susceptibility contrast techniques with IV contrast. Perfusion imaging is also used to assess functional improvement after treatment [39,46-49].

Variant 3: Child. Clinical presentation suggestive of acute stroke, known or suspected arteriopathy, or moyamoya. Not a candidate for emergent treatment. Initial imaging.

G. HMPAO SPECT or SPECT/CT Brain

Single-photon-emission computed tomography (SPECT) studies can be used to assess perfusion in patients with moyamoya and can be augmented with acetazolamide challenge to assess perfusion instability and vascular reserve. Perfusion imaging is also used to assess functional improvement after treatment [39].

Variant 3: Child. Clinical presentation suggestive of acute stroke, known or suspected arteriopathy, or moyamoya. Not a candidate for emergent treatment. Initial imaging.

H. Arteriography Cervicocerebral

Catheter angiography is required for definitive diagnosis of moyamoya and is typically obtained as part of a preoperative assessment prior to surgical revascularization or following revascularization to assess the development of surgically created collaterals.

Variant 3: Child. Clinical presentation suggestive of acute stroke, known or suspected arteriopathy, or moyamoya. Not a candidate for emergent treatment. Initial imaging.

I. US Duplex Doppler Transcranial

Head US does not typically play a role in the management of children with moyamoya.

Variant 4: Child. Known or suspected cervicocranial arterial dissection based on clinical or imaging findings. Next imaging study.

Cervicocranial artery dissection occurs in 2.5/100,000 children per year and is up to 4 times more common in children than in adults [50-52]. Purely intracranial dissections are also more common in children than adults [50]. Patients with cervical artery dissection can present with headache, neck pain, or ischemic infarction due to emboli, whereas patients with intracranial artery dissection can present with ischemic infarction, subarachnoid hemorrhage (SAH), or symptoms due to local mass effect [50,52]. A history of antecedent trauma may or may not be present.

Variant 4: Child. Known or suspected cervicocranial arterial dissection based on clinical or imaging findings. Next imaging study.

A. CT Head

CT can show areas of infarction or SAH although the sensitivity of CT is less than that of MRI with DWI for demonstrating acute infarction [43]. A crescent-shaped hyperattenuating area in a cranial or cervical artery is consistent with an intramural hematoma and is highly suggestive of dissection [52].

Variant 4: Child. Known or suspected cervicocranial arterial dissection based on clinical or imaging findings. Next imaging study.

B. CTA Head and Neck

CTA can demonstrate vessel narrowing, pseudoaneurysm, or an intimal flap in dissection [50,52].

Variant 4: Child. Known or suspected cervicocranial arterial dissection based on clinical or imaging findings. Next imaging study.

C. MRI Head and Neck

MRI is the preferred study for the demonstration of infarction due to cervicocranial arterial dissection [50,52,53]. DWI and perfusion-weighted MRI provide information on regions of infarction. SWI can demonstrate emboli, and increased oxygen extraction leads to prominence of the veins in infarcted regions. T1-weighted imaging with fat suppression or T2-weighted imaging may demonstrate an intramural hematoma in 76% to 91% of patients with dissection [50,52].

Variant 4: Child. Known or suspected cervicocranial arterial dissection based on clinical or imaging findings. Next imaging study.

D. MRA Head and Neck

MRA without or with IV contrast may be used to assess for cervicocranial artery dissection to demonstrate an intimal flap, vessel narrowing, or pseudoaneurysm formation [52,53].

Variant 4: Child. Known or suspected cervicocranial arterial dissection based on clinical or imaging findings. Next imaging study.

E. Arteriography Cervicocerebral

Catheter angiography remains the gold standard for the radiologic evaluation of dissection but is typically reserved for patients in whom dissection is suspected but not confirmed on noninvasive imaging [52,53]. Angiography may reveal vessel narrowing, pseudoaneurysm formation, or an intimal flap in regions of dissection and distal vessel occlusions due to emboli.

Variant 4: Child. Known or suspected cervicocranial arterial dissection based on clinical or imaging findings. Next imaging study.

F. US Duplex Doppler Transcranial and Carotid Artery

Doppler US may be used to evaluate the cervical internal carotid arteries for dissection. The technique is of limited utility for carotid dissections at or above the skull base and in the evaluation of the vertebral arteries because of a lack of a good acoustic window [52].

Variant 5: Child. Clinical presentation suggestive of acute stroke, known or suspected central nervous system vasculitis. Initial imaging.

Childhood vasculitis is an inflammatory process only affecting the intracranial vessels and can result in stroke. It can present as childhood primary angiitis of the central nervous system or as a secondary phenomenon in systemic rheumatologic infections and neoplastic conditions [54]. Childhood primary angiitis of the central nervous system is subcategorized into large-medium vessel vasculitis (angiography positive) or small vessel (vessel abnormality too small to be demonstrated on MRA, CTA, or conventional angiography) best diagnosed by brain biopsy [55]. The diagnosis of childhood primary angiitis of the central nervous system requires an acquired neurologic deficit, angiographic or histologic features of central nervous system vasculitis, and no evidence of a systemic condition associated with the central nervous system findings.

Variant 5: Child. Clinical presentation suggestive of acute stroke, known or suspected central nervous system vasculitis. Initial imaging.

A. CT Head

Evaluation of central nervous system vasculitis by CT is typically negative [56].

Variant 5: Child. Clinical presentation suggestive of acute stroke, known or suspected central nervous system vasculitis. Initial imaging.

B. CTA Head

CTA can provide a noninvasive assessment of intracranial vessels in arteriopathies as an alternative to catheter angiography in some patients but is typically negative in small-vessel vasculitis (eg,

childhood primary angiitis of the central nervous system) [17].

Variant 5: Child. Clinical presentation suggestive of acute stroke, known or suspected central nervous system vasculitis. Initial imaging.

C. MRI Head

MRI typically shows progressive multifocal parenchymal lesions on T2-weighted imaging. Gadolinium enhancement of lesions is inconsistent.

Variant 5: Child. Clinical presentation suggestive of acute stroke, known or suspected central nervous system vasculitis. Initial imaging.

D. MRA Head

MRA can provide a noninvasive assessment of intracranial vessels in arteriopathies as an alternative to catheter angiography in some patients but is typically negative in small vessel-vasculitis (eg, childhood primary angiitis of the central nervous system) [17].

Variant 5: Child. Clinical presentation suggestive of acute stroke, known or suspected central nervous system vasculitis. Initial imaging.

E. Arteriography Cervicocerebral

Catheter angiography is the most sensitive imaging method in the assessment of cerebral vasculitis but is typically negative in small-vessel vasculitis (eg, childhood primary angiitis of the central nervous system) [17].

Variant 5: Child. Clinical presentation suggestive of acute stroke, known or suspected central nervous system vasculitis. Initial imaging.

F. US Duplex Doppler Transcranial

Head US is not usually indicated in the evaluation of cerebral vasculitis.

Variant 6: Child. Nontraumatic intraparenchymal hemorrhage (hematoma) found on CT or MRI. Unknown etiology. Next imaging study.

Approximately half of all strokes beyond the perinatal period are hemorrhagic, whereas only 6.5% to 13% of strokes in adults are hemorrhagic [57,58]. Hemorrhagic strokes are associated with a variety of etiologies, including arteriovenous fistula or arteriovenous malformation (48%), brain tumors (15%), genetic or acquired coagulopathy (9%), thrombocytopenia (6%), cavernous malformation (5%), and coagulopathy and aneurysm (2%) [58]. Presenting symptoms of hemorrhagic stroke differ with respect to the age of the child but may include mental status changes, seizures, or focal neurologic deficit.

Variant 6: Child. Nontraumatic intraparenchymal hemorrhage (hematoma) found on CT or MRI. Unknown etiology. Next imaging study.

A. CT Head

Unenhanced head CT is useful to assess the location and size of hemorrhage, evidence of mass effect, and hydrocephalus [59]. IV contrast may be helpful in demonstrating an underlying cause for the hemorrhage, such as vascular malformation, aneurysm, or tumor.

Variant 6: Child. Nontraumatic intraparenchymal hemorrhage (hematoma) found on CT or MRI. Unknown etiology. Next imaging study.

B. CTA Head

CTA may be used to delineate the vascular anatomy and may be used to demonstrate a vascular malformation or aneurysm but lacks the temporal information available from catheter

angiography.

Variant 6: Child. Nontraumatic intraparenchymal hemorrhage (hematoma) found on CT or MRI. Unknown etiology. Next imaging study.

C. MRI Head

Unenhanced MRI can delineate the location and size of hemorrhage, evidence of mass effect, and hydrocephalus and may show evidence of an underlying structural lesion, such as vascular malformation, aneurysm, or tumor. SWI and arterial spin-labeling imaging may improve the delineation of the draining veins and arteriovenous shunting, respectively, compared with conventional MR sequences [24]. Contrast may be required to demonstrate small malformations, such as those that may occur in hereditary hemorrhagic telangiectasia, or other causes of parenchymal hematoma, such as tumor.

Variant 6: Child. Nontraumatic intraparenchymal hemorrhage (hematoma) found on CT or MRI. Unknown etiology. Next imaging study.

D. MRA Head

MRA may be used to delineate the vascular anatomy and may be used to demonstrate a vascular malformation or aneurysm but lacks the temporal information available from catheter angiography.

Variant 6: Child. Nontraumatic intraparenchymal hemorrhage (hematoma) found on CT or MRI. Unknown etiology. Next imaging study.

E. Arteriography Cervicocerebral

Catheter angiography is the gold standard for imaging of high-flow vascular malformations and aneurysms [60]. Although developmental venous anomalies (DVAs) can be demonstrated on catheter angiography, cavernoma and capillary telangiectasia are angiographically occult [61].

Variant 6: Child. Nontraumatic intraparenchymal hemorrhage (hematoma) found on CT or MRI. Unknown etiology. Next imaging study.

F. US Duplex Doppler Transcranial

Head US may be used to evaluate the size and location of hemorrhage, evidence of mass effect, and hydrocephalus in neonates and infants with open fontanelles but is limited in its ability to distinguish infarction from hemorrhage and underlying structural causes of intracranial hemorrhage.

Variant 7: Child. Nontraumatic subarachnoid hemorrhage (SAH) detected by noncontrast CT. Next imaging study.

Fifty-seven percent of SAH in children is due to aneurysm; however, only 2% to 15% of hemorrhagic strokes in children are due to aneurysm [58,62]. Aneurysms in children account for <1% of SAH due to aneurysms in all age groups [62]. The mortality in pediatric aneurysm has been reported to be 1.3%, with morbidity including infarction (8%) and seizures (4%) [63]. Only 0.6% of ruptured aneurysms occur in patients <19 years of age [51,60]. Aneurysms in children are idiopathic (45%), post-traumatic (20%), or due to a variety of conditions causing abnormal vessel-wall hemodynamic stress [62]. In contrast to adults, aneurysms in children are more likely to be giant (>25 mm) or fusiform [62]. Children with a positive family history of aneurysm account for <5% of pediatric aneurysms, and fewer than 2% of patients with a positive family history of aneurysm develop an aneurysm in the first two decades of life [62]. Fusiform aneurysms are more likely to increase in size over time than are saccular aneurysms [64].

Variant 7: Child. Nontraumatic subarachnoid hemorrhage (SAH) detected by noncontrast CT.

Next imaging study.

A. CT Head

The initial imaging study in suspected SAH is a noncontrast head CT. If the CT is negative for SAH, lumbar puncture may be performed. Unenhanced head CT is useful in ruptured aneurysms to assess the location and size of hemorrhage, evidence of mass effect, and hydrocephalus [59]. Small aneurysms are difficult to evaluate with unenhanced CT whereas a giant aneurysm, if present, appears as a hyperdense mass.

Variant 7: Child. Nontraumatic subarachnoid hemorrhage (SAH) detected by noncontrast CT.

Next imaging study.

B. CTA Head

CTA is a noninvasive alternative to catheter angiography to show an underlying vascular cause of SAH with reported sensitivity (96.5%) and specificity (88%) for aneurysms of all sizes with sensitivity (98.4%) and specificity (100%) for aneurysms >3 mm [65]. CTA is used to confirm the presence of aneurysm and may be used as an adjunct to catheter angiography in the pretreatment assessment of aneurysm. CTA provides an alternative to catheter angiography for assessing residual or recurrent aneurysm but may be limited by streak artifact from the treatment device. Using digital subtraction angiography as the gold standard, CTA has a reported sensitivity of 74% and specificity of 96% for the demonstration of residual aneurysm post-treatment [66].

Variant 7: Child. Nontraumatic subarachnoid hemorrhage (SAH) detected by noncontrast CT.

Next imaging study.

C. MRI Head

MRI is not typically an initial imaging study in patients with suspected SAH. However, SWI is sensitive to the presence of subarachnoid blood. MRI may be useful in classifying the aneurysms into saccular, dissecting, giant, and infectious [60]. MRI may be used to evaluate potential complications of treatment, including cerebral infarction or hemorrhage [63]. Only patients with MRI-safe aneurysm clips should be imaged with MR.

Variant 7: Child. Nontraumatic subarachnoid hemorrhage (SAH) detected by noncontrast CT.

Next imaging study.

D. MRA Head

MRA is a noninvasive alternative to catheter angiography to show an underlying vascular cause of SAH. MRA of the brain is the examination of choice for children with unruptured aneurysms being followed prior to treatment [51]. A meta-analysis of the literature shows the sensitivity and specificity of MRA for the detection of intracranial aneurysm to be 95% and 89%, respectively [67]. Given the limited number of children who will develop an aneurysm under the age of 20, routine screening with MRA of children with a positive family history of aneurysm is not supported in the literature [68].

Variant 7: Child. Nontraumatic subarachnoid hemorrhage (SAH) detected by noncontrast CT.

Next imaging study.

E. Arteriography Cervicocerebral

Catheter angiography remains the gold standard for the demonstration of high-flow vascular anomalies and aneurysms as a cause of SAH. Catheter angiography can be performed for pretreatment planning and for post-treatment assessment. Catheter angiography is associated with a low incidence of procedural complications in children [51,60]. Catheter angiography is the gold standard for the diagnosis and pretreatment evaluation of intracranial aneurysm and may also

be required for post-treatment follow-up in some patients [60].

Variant 7: Child. Nontraumatic subarachnoid hemorrhage (SAH) detected by noncontrast CT. Next imaging study.

F. US Duplex Doppler Transcranial

Head US is not sensitive for the presence of subarachnoid blood or vascular causes of intracranial hemorrhage. Head US provides limited information on intracranial aneurysm, but Doppler US may be used to confirm the vascular nature of a mass seen on head US performed in neonates and young infants with open fontanelles.

Variant 8: Child. Clinical presentation suggestive of acute stroke, known or suspected high-flow vascular anomaly. Initial imaging.

High-flow vascular anomalies (arteriovenous fistula and arteriovenous malformation), comprised of arteriovenous shunts, are the most common cause of spontaneous intracranial hemorrhage in children presenting with stroke symptoms [57-59]. Abnormal communications between the supplying artery and draining vein can consist of one or more macroscopic fistulae (arteriovenous fistula) or a vascular nidus (arteriovenous malformation). Intracranial arteriovenous shunts may occur in isolation or may be part of a syndrome or genetic condition [59]. In children, 48% of hemorrhagic strokes are due to arteriovenous shunts [58]. Children are more likely than adults to present with hemorrhage due to arteriovenous shunts (59% versus 41%, respectively), but only 18% of arteriovenous shunts become symptomatic before 15 years of age [59,69,70]. The annual hemorrhage rate is estimated to be between 2% to 4% [57,59]. In addition to intracranial hemorrhage, high-flow arteriovenous shunts may cause parenchymal ischemic brain injury either because of shunting of blood away from the brain parenchyma or because of venous hypertension caused by high-flow venopathy.

Variant 8: Child. Clinical presentation suggestive of acute stroke, known or suspected high-flow vascular anomaly. Initial imaging.

A. CT Head

Unenhanced head CT is useful in hemorrhagic high-flow vascular anomalies to assess the location and size of hemorrhage, evidence of mass effect, and hydrocephalus [59]. High-flow vascular malformations that have not bled are slightly hyperdense relative to brain on unenhanced head CT. The detection of a high-flow vascular anomaly that has not bled may be improved with the use of IV contrast.

Variant 8: Child. Clinical presentation suggestive of acute stroke, known or suspected high-flow vascular anomaly. Initial imaging.

B. CTA Head

CTA is often used to delineate the anatomy of a high-flow vascular anomaly but lacks the temporal information available from catheter angiography. CTA may be used to monitor the lesion prior to treatment or to assess for growth or recurrence after treatment.

Variant 8: Child. Clinical presentation suggestive of acute stroke, known or suspected high-flow vascular anomaly. Initial imaging.

C. MRI Head

MRI is used to determine the location and size of high-flow vascular anomalies and is often preferred to CT for use in children. SWI and arterial spin-labeling imaging may improve the delineation of the draining veins and arteriovenous shunting, respectively, compared with conventional MRI sequences [24]. Contrast may be required to demonstrate small malformations.

Variant 8: Child. Clinical presentation suggestive of acute stroke, known or suspected high-flow vascular anomaly. Initial imaging.**D. MRA Head**

MRA, like CTA, can be used to delineate the anatomy of a high-flow vascular anomaly but lacks the temporal information available from catheter angiography. MRA is often preferable to CTA in children for following vascular anomalies serially. The use of IV contrast may be required to assess the venous outflow of the anomaly. MRA may be used to monitor the anomaly prior to treatment or to assess for growth or recurrence after treatment.

Variant 8: Child. Clinical presentation suggestive of acute stroke, known or suspected high-flow vascular anomaly. Initial imaging.**E. Arteriography Cervicocerebral**

With the increased accuracy of the noninvasive imaging studies CTA and MRA, catheter angiography is not typically used as the initial diagnostic imaging for suspected high-flow vascular anomaly. Catheter angiography can be used for pretreatment evaluation and post-treatment assessment of any residual anomaly [59,71].

Variant 8: Child. Clinical presentation suggestive of acute stroke, known or suspected high-flow vascular anomaly. Initial imaging.**F. US Duplex Doppler Transcranial**

Head US with Doppler may be used to show the presence of large high-flow vascular malformations, such as vein of Galen malformation and dural arteriovenous fistula or malformation in neonates and young infants. US is useful in very young children because it can be performed without sedation or anesthesia, which may be required for MRI examinations.

Variant 9: Child. Clinical presentation suggestive of acute stroke, known or suspected low-flow vascular anomaly. Initial imaging.

Low-flow vascular anomalies include cavernous malformation, DVA, and capillary telangiectasia. Cavernomas are comprised of endothelial spaces containing venous blood and account for 17% of hemorrhagic stroke in children [18]. In a large series, 62% of children with cavernomas presented with hemorrhage, 35% with seizures with or without hemorrhage, and 26% had cavernomas discovered incidentally [72]. Patients with cavernoma (15%) have multiple lesions and 10% have a family history of cavernoma and may have mutations in CCM1, CCM2, and CCM3 genes [72-74]. Although 9% are associated with prior cranial irradiation [72], 86% are supratentorial and 14% infratentorial in location. Cavernomas have an annual hemorrhage rate of 3.3% to 4.5% [72,75]. DVA is an anomalous vein draining normal brain parenchyma, and 20% of DVA are associated with cavernoma [72]. Most hemorrhage in patients with DVA is believed to be due to bleeding from an associated cavernoma [61,72].

Variant 9: Child. Clinical presentation suggestive of acute stroke, known or suspected low-flow vascular anomaly. Initial imaging.**A. CT Head**

Noncontrast CT can show acute hemorrhage with cavernoma. Nonhemorrhagic cavernomas and DVA may be faintly hyperdense on noncontrast imaging [61]. IV contrast increases the conspicuity of DVA.

Variant 9: Child. Clinical presentation suggestive of acute stroke, known or suspected low-flow vascular anomaly. Initial imaging.**B. CTA Head**

CTA plays a limited role in the assessment of cavernoma but may be used to demonstrate a DVA.

Variant 9: Child. Clinical presentation suggestive of acute stroke, known or suspected low-flow vascular anomaly. Initial imaging.

C. MRI Head

MRI is the imaging study of choice for cavernoma. Various stages of hemorrhage may be evident. T2-weighted gradient-echo imaging or SWI may show additional cavernomas not seen on spin-echo imaging. The presence of acute and subacute blood products increases the likelihood of future hemorrhage [75]. DVAs are visible on both spin-echo and gradient-echo imaging and are occasionally associated with gliosis or cortical malformation. Contrast may increase the conspicuity of DVA.

Variant 9: Child. Clinical presentation suggestive of acute stroke, known or suspected low-flow vascular anomaly. Initial imaging.

D. MRA Head

MRA is not usually helpful in the assessment of cavernoma, capillary telangiectasia, and DVA.

Variant 9: Child. Clinical presentation suggestive of acute stroke, known or suspected low-flow vascular anomaly. Initial imaging.

E. Arteriography Cervicocerebral

Catheter angiography is rarely required for the evaluation of incidentally discovered DVA but may be used to exclude the possibility of an associated high-flow vascular malformation. An isolated DVA appears as an abnormal cluster of veins draining into a single collector and appears only on the venous phase of the arteriogram [61]. Cavernomas and capillary telangiectasia are usually angiographically occult.

Variant 9: Child. Clinical presentation suggestive of acute stroke, known or suspected low-flow vascular anomaly. Initial imaging.

F. US Duplex Doppler Transcranial

Head US is not usually helpful in the assessment of cavernoma or capillary telangiectasia. Head US may occasionally be used to demonstrate large DVA in neonates and young infants with open fontanelles.

Variant 10: Child. Clinical presentation suggestive of acute stroke, known or suspected cortical vein or dural venous sinus thrombosis. Initial imaging.

Cerebral sinovenous thrombosis has an incidence of 1/200,000 children per year. It is less common in children than is arterial ischemic or hemorrhagic stroke and is most often associated with infection, trauma, dehydration, cancer, oral contraceptives, and prothrombotic disorders [76,77]. The risk is highest in the first year of life, with neonates accounting for 61% of cerebral sinovenous thrombosis [76-78]. Prothrombotic disorders are present in more than half of children, and multiple risk factors are commonly present. An acute illness with sepsis and dehydration is present in up to one-third of patients. Trauma is more common in older children. Cerebral sinovenous thrombosis can cause elevation in venous pressure, increased intracranial pressure, and venous infarction. Hemorrhagic infarction (40%) and hydrocephalus (10%) may complicate cerebral sinovenous thrombosis [77]. Infarctions are more frequently hemorrhagic in neonates (72%) than in older children (48%) [79]. The lateral dural venous sinuses (73%) and superior sagittal sinus (35%) are most commonly affected [76-78].

Variant 10: Child. Clinical presentation suggestive of acute stroke, known or suspected cortical vein or dural venous sinus thrombosis. Initial imaging.

A. CT Head

Unenhanced CT shows increased density, often accompanied by distension, of the thrombosed sinus or vein [77,80]. CT is less sensitive than MRI for the demonstration of early ischemic infarction but readily shows areas of brain hemorrhage [77,79].

Variant 10: Child. Clinical presentation suggestive of acute stroke, known or suspected cortical vein or dural venous sinus thrombosis. Initial imaging.

B. CTV Head

CT venography (CTV) shows the "empty delta" sign because of a lack of enhancement within the affected venous structure.

Variant 10: Child. Clinical presentation suggestive of acute stroke, known or suspected cortical vein or dural venous sinus thrombosis. Initial imaging.

C. MRI Head

MRI without and with IV contrast is used to diagnose and follow cortical vein and dural venous sinus thrombosis and is preferred over CT/CTV for serial examinations. Short echo-time gradient-echo MRI with IV contrast shows a similar "empty delta" sign to contrast-enhanced CT or CTV within the thrombosed sinus or vein [77,80]. Gradient-echo T1-weighted postcontrast MRI has been shown to have slightly greater sensitivity (92.5%) and specificity (100%) compared with unenhanced MR venography (MRV), sensitivity (89.6%) and specificity (91.8%), for the demonstration of dural venous sinus and cortical venous thrombosis [81].

Variant 10: Child. Clinical presentation suggestive of acute stroke, known or suspected cortical vein or dural venous sinus thrombosis. Initial imaging.

D. MRV Head

MRV is used to diagnose and follow cortical vein and dural venous sinus thrombosis and is typically performed in conjunction with anatomic MRI without and with IV contrast for a comprehensive assessment of the intracranial venous drainage. MRV without IV contrast is commonly used to confirm absence of flow in a thrombosed dural venous sinus. MRV with IV contrast is less susceptible to the flow artifacts that may occur because of turbulent flow in the dural venous sinuses with noncontrast MRV [80].

Variant 10: Child. Clinical presentation suggestive of acute stroke, known or suspected cortical vein or dural venous sinus thrombosis. Initial imaging.

E. Arteriography Cervicocerebral

Catheter angiography is not routinely used for the diagnosis of cortical venous or dural venous sinus thrombosis but is used when endovascular treatment is required.

Variant 10: Child. Clinical presentation suggestive of acute stroke, known or suspected cortical vein or dural venous sinus thrombosis. Initial imaging.

F. US Duplex Doppler Transcranial

Head US with Doppler may be used to assess patency of the dural venous sinuses in the neonate. Following closure of the fontanel, CT and MRI are more commonly used.

Variant 11: Child. Clinical presentation suggestive of acute stroke, sickle cell disease. New focal fixed or worsening neurologic defect. Initial imaging.

SCD constitutes one of the main etiologies of pediatric stroke. Eleven percent of children with SCD not receiving primary stroke prevention therapy will have a stroke by 20 years of age [36,82,83]. Most infarctions are due to vasculopathy involving the supraclinoid portion of the internal carotid

artery or branches of the circle of Willis and occurring in a watershed distribution. Clinically silent infarctions are known to occur in 17% of children with SCD [84]. The National Heart Lung and Blood institute recommends that children with SCD between 2 and 16 years of age undergo transcranial Doppler US screening every 6 months. The treatment of acute stroke symptoms in children with SCD is transfusion.

Variant 11: Child. Clinical presentation suggestive of acute stroke, sickle cell disease. New focal fixed or worsening neurologic defect. Initial imaging.

A. CT Head

CT is relatively insensitive compared with MRI for the demonstration of infarction and is typically only used in the acute setting if MRI is unavailable or to document hemorrhage or evaluate for mass effect [26].

Variant 11: Child. Clinical presentation suggestive of acute stroke, sickle cell disease. New focal fixed or worsening neurologic defect. Initial imaging.

B. CTA Head

CTA is not used in routine neurovascular screening in children with SCD. CTA can be used in children with contraindication to MRA.

Variant 11: Child. Clinical presentation suggestive of acute stroke, sickle cell disease. New focal fixed or worsening neurologic defect. Initial imaging.

C. MRI Head

MRI can show T2 prolongation in watershed vascular territories or cortex in patients with SCD.

Variant 11: Child. Clinical presentation suggestive of acute stroke, sickle cell disease. New focal fixed or worsening neurologic defect. Initial imaging.

D. MRA Head

MRA is the study of choice to screen for vessel stenosis in children with SCD with elevated transcranial Doppler US velocities in the intracranial vessels [39,44]. Although MRA can accurately show vessel stenosis, it is susceptible to false-positive results when long echo times are used or when severe anemia is present, resulting in turbulent blood flow and localized artefactual signal loss resembling focal stenosis [26,84,85]. Although gadolinium-enhanced MRA can be used, it is generally not required and is often avoided because of concerns that it may potentiate hemolysis in patients with SCD [26]. MRI of the brain without IV contrast is typically performed during the same examination as the MRA to assess the brain parenchyma for ischemic injury.

Variant 11: Child. Clinical presentation suggestive of acute stroke, sickle cell disease. New focal fixed or worsening neurologic defect. Initial imaging.

E. Arteriography Cervicocerebral

Catheter angiography is generally not indicated except in uncommon circumstances, including preoperative assessment for revascularization for moyamoya syndrome and suspected aneurysm complicating SCD [26].

Variant 11: Child. Clinical presentation suggestive of acute stroke, sickle cell disease. New focal fixed or worsening neurologic defect. Initial imaging.

F. US Duplex Doppler Transcranial

The National Heart Lung and Blood institute recommends that children with SCD between 2 and 16 years of age to undergo transcranial Doppler US screening every 6 months [35]. Velocity measurements from the distal internal carotid artery or proximal middle cerebral artery are used to

assess stroke risk. Time averaged means of maximum velocity of <170 cm/sec are associated with a low risk of stroke, velocities of 170 to 199 cm/sec are associated with an intermediate risk of stroke, and velocities of >200 cm/sec are associated with a high risk of stroke (40%) in the next 3 years [35,86].

Summary of Highlights

- **Variant 1:** MRI head without IV contrast, CT head without IV contrast, or MRA head without IV contrast is usually appropriate for the initial emergent imaging of children >6 months of age with a clinical presentation suggestive of acute nonsickle cell-related stroke with new focal or worsening neurologic defect lasting <24 hours from the last seen normal state with no contraindications to emergent intervention. MRI of the head without IV contrast and CT head without IV contrast are appropriate initial imaging alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care). MRA head without IV contrast provides complementary information and should be performed in conjunction with MRI head without IV contrast in this clinical scenario.
- **Variant 2:** MRI head without IV contrast, CT head without IV contrast, or MRA head without IV contrast is usually appropriate for the initial imaging of a child with clinical presentation suggestive of acute stroke who are not candidates for emergent intervention. MRI of the head without IV contrast and CT head without IV contrast are appropriate initial imaging alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care). MRA head without IV contrast provides complementary information and should be performed in conjunction with MRI head without IV contrast in this clinical scenario.
- **Variant 3:** MRA head without IV contrast or MRI head without IV contrast is usually appropriate for the initial imaging of a child (not a candidate for emergent treatment) with clinical presentation suggestive of acute stroke, known or suspected arteriopathy, or moyamoya. MRA head without IV contrast provides complementary information and should be performed in conjunction with MRI head without IV contrast in this clinical scenario.
- **Variant 4:** MRI head and neck without IV contrast, CTA head and neck with IV contrast, or MRA head and neck without IV contrast is usually appropriate as the next imaging study for children with known or suspected cervicocranial arterial dissection based on clinical or imaging findings. These procedures are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care). The panel did not agree on recommending MRA head and neck with IV contrast or MRI head and neck with IV contrast for this clinical scenario. There is insufficient medical literature to conclude whether or not these patients would benefit from these procedures for this clinical scenario. Follow-up imaging in this patient population is controversial but may be appropriate.
- **Variant 5:** MRA head without IV contrast, MRI head without IV contrast, CTA head with IV contrast, or MRI head without and with IV contrast is usually appropriate for the initial imaging of children with clinical presentation suggestive of acute stroke, known or suspected central nervous system vasculitis. MRA head without IV contrast provides complementary information and should be performed in conjunction with MRI head without IV contrast or MRI head without and with contrast in this clinical scenario. These procedures are otherwise equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care). The panel did not agree on

recommending MRI head with IV contrast for this clinical scenario. There is insufficient medical literature to conclude whether or not these patients would benefit from this procedure for this clinical scenario. Imaging in this patient population is controversial but may be appropriate.

- **Variant 6:** MRA head without IV contrast, CTA head with IV contrast, arteriography cervicocerebral, or MRI head without and with IV contrast is usually appropriate as the next imaging study for children with nontraumatic intraparenchymal hemorrhage (hematoma) found on CT or MRI of unknown etiology. MRA head without IV contrast provides complementary information and should be performed in conjunction with MRI head without IV contrast or MRI head without and with contrast in this clinical scenario. These procedures are otherwise equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care). The panel did not agree on recommending MRI head with IV contrast or MRI head without IV contrast for this clinical scenario. There is insufficient medical literature to conclude whether or not these patients would benefit from these procedures for this clinical scenario. Follow-up imaging in this patient population is controversial but may be appropriate.
- **Variant 7:** MRA head without IV contrast, CTA head with IV contrast, or arteriography cervicocerebral is usually appropriate as the next imaging study for children with nontraumatic SAH detected by noncontrast CT. MRA head without IV contrast provides complementary information and should be performed in conjunction with MRI head without IV contrast in this clinical scenario. These procedures are otherwise equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care).
- **Variant 8:** CT head without IV contrast, CTA head with IV contrast, MRA head without IV contrast, or MRI head without IV contrast is usually appropriate for the initial imaging of children with clinical presentation suggestive of acute stroke, known or suspected high-flow vascular anomaly. MRA head without IV contrast provides complementary information and should be performed in conjunction with MRI head without IV contrast in this clinical scenario. These procedures are otherwise equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care). The panel did not agree on recommending arteriography cervicocerebral for this clinical scenario. There is insufficient medical literature to conclude whether or not these patients would benefit from arteriography cervicocerebral for this clinical scenario. Imaging in this patient population is controversial but may be appropriate.
- **Variant 9:** MRI head without IV contrast or CT head without IV contrast is usually appropriate for the initial imaging of children with clinical presentation suggestive of acute stroke, known or suspected low-flow vascular anomaly. Either procedure is a usually appropriate initial imaging alternative. The panel did not agree on recommending CTA head with IV contrast or MRA head without IV contrast for this clinical scenario. There is insufficient medical literature to conclude whether or not these patients would benefit from these procedures for this clinical scenario. Imaging in this patient population is controversial but may be appropriate.
- **Variant 10:** MRV head with IV contrast, CTV head with IV contrast, MRI head without and with IV contrast, MRV head without IV contrast, CT head without IV contrast, or MRI head without IV contrast is usually appropriate for the initial imaging of children with clinical presentation suggestive of acute stroke, known or suspected cortical vein or dural venous sinus thrombosis. MRV head with IV contrast and MRV without IV contrast provide

complementary information too, and should be performed in conjunction with MRI head without IV contrast or MRI head without and with IV contrast. These procedures are otherwise equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care).

- **Variant 11:** MRA head without IV contrast, MRI head without IV contrast, or CT head without IV contrast is usually appropriate for the initial imaging of children with clinical presentation suggestive of acute stroke and sickle cell disease with new focal fixed or worsening neurologic defect. MRA head without IV contrast provides complementary information and should be performed in conjunction with MRI head without IV contrast in this clinical scenario. These procedures are otherwise equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care).

Supporting Documents

The evidence table, literature search, and appendix for this topic are available at <https://acsearch.acr.org/list>. The appendix includes the strength of evidence assessment and the final rating round tabulations for each recommendation.

For additional information on the Appropriateness Criteria methodology and other supporting documents, please go to the ACR website at <https://www.acr.org/Clinical-Resources/Clinical-Tools-and-Reference/Appropriateness-Criteria>.

Appropriateness Category Names and Definitions

Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate	7, 8, or 9	The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk-benefit ratio for patients.
May Be Appropriate	4, 5, or 6	The imaging procedure or treatment may be indicated in the specified clinical scenarios as an alternative to imaging procedures or treatments with a more favorable risk-benefit ratio, or the risk-benefit ratio for patients is equivocal.
May Be Appropriate (Disagreement)	5	The individual ratings are too dispersed from the panel median. The different label provides transparency regarding the panel's recommendation. "May be appropriate" is the rating category and a rating of 5 is assigned.
Usually Not Appropriate	1, 2, or 3	The imaging procedure or treatment is unlikely to be indicated in the specified clinical scenarios, or the risk-benefit ratio for patients is likely to be unfavorable.

Relative Radiation Level Information

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures

associated with different diagnostic procedures, a relative radiation level (RRL) indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, because of both organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as compared with those specified for adults (see Table below). Additional information regarding radiation dose assessment for imaging examinations can be found in the ACR Appropriateness Criteria® [Radiation Dose Assessment Introduction](#) document.

Relative Radiation Level Designations

Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range
0	0 mSv	0 mSv
●	<0.1 mSv	<0.03 mSv
● ●	0.1-1 mSv	0.03-0.3 mSv
● ● ●	1-10 mSv	0.3-3 mSv
● ● ● ●	10-30 mSv	3-10 mSv
● ● ● ● ●	30-100 mSv	10-30 mSv

*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (e.g., region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as “Varies.”

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Disclaimer

The ACR Committee on Appropriateness Criteria and its expert panels have developed criteria for determining appropriate imaging examinations for diagnosis and treatment of specified medical condition(s). These criteria are intended to guide radiologists, radiation oncologists and referring physicians in making decisions regarding radiologic imaging and treatment. Generally, the complexity and

severity of a patient's clinical condition should dictate the selection of appropriate imaging procedures or treatments. Only those examinations generally used for evaluation of the patient's condition are ranked. Other imaging studies necessary to evaluate other co-existent diseases or other medical consequences of this condition are not considered in this document. The availability of equipment or personnel may influence the selection of appropriate imaging procedures or treatments. Imaging techniques classified as investigational by the FDA have not been considered in developing these criteria; however, study of new equipment and applications should be encouraged. The ultimate decision regarding the appropriateness of any specific radiologic examination or treatment must be made by the referring physician and radiologist in light of all the circumstances presented in an individual examination.

^aBoston Children's Hospital, Boston, Massachusetts. ^bEmory University and Children's Healthcare of Atlanta, Atlanta, Georgia. ^cBoston Children's Hospital, Boston, Massachusetts; American Academy of Neurology. ^dPanel Vice-Chair, Vanderbilt Children's Hospital, Nashville, Tennessee. ^eGlobal Advanced Imaging, PLLC, Little Rock, Arkansas; Commission on Nuclear Medicine and Molecular Imaging. ^fTexas Children's Hospital, Houston, Texas. ^gEmory University and Children's of Atlanta (Egleston), Atlanta, Georgia. ^hHospital for Sick Children, Toronto, Ontario, Canada; American Association of Neurological Surgeons/Congress of Neurological Surgeons. ⁱSutter Medical Center Sacramento, Sacramento, California; American College of Emergency Physicians. ^jChildren's Hospital of Wisconsin, Milwaukee, Wisconsin. ^kEmory University and Children's Healthcare of Atlanta, Atlanta, Georgia. ^lChildren's Hospital Colorado, Aurora, Colorado. ^mChildren's National Health System, Washington, District of Columbia; American Association of Neurological Surgeons/Congress of Neurological Surgeons. ⁿStanford University, Stanford, California; American Academy of Pediatrics. ^oIndiana University Health, Indianapolis, Indiana. ^pThe University of Vermont Medical Center, Burlington, Vermont. ^qCincinnati Children's Hospital Medical Center, Cincinnati, Ohio; Commission on Nuclear Medicine and Molecular Imaging. ^rUniversity of Arizona College of Medicine, Tucson, Arizona. ^sChildren's National Health System, Washington, District of Columbia. ^tSpecialty Chair, Riley Hospital for Children Indiana University, Indianapolis, Indiana.