MoyaMoya is a progressive arteriopathy of unknown origin affecting the branches of the internal carotid artery (ICA). The arteriopathy can present as an isolated medical condition, affecting both sides of the brain (“moyamoya disease”) or can be unilateral or found in association with systemic disorders (“moyamoya syndrome”). The ischemia resulting from luminal narrowing predispenses children to transient ischemic attacks and stroke—the primary presentations of affected patients. Although it is rare—afflicting 1 in 1 million children in the US—moyamoya is implicated in 6% of all childhood strokes. Diagnosis is defined by characteristic findings on arteriograms, including stenosis of the branches of the ICA and a pathognomonic spray of small collateral vessels in this region, descriptively likened to a “puff of smoke” (“moyamoya” in Japanese). Treatment is predicated on restoration of cerebral blood flow by surgical revascularization. The rarity of this disorder has limited research and the development of evidence-based clinical management. While acknowledging these limitations, in this article the authors aim to summarize current studies of pediatric moyamoya, with the objective of providing a framework for construction of evidence-based guidelines for treatment. The compilation of current data in these guidelines should serve as a resource to aid pediatric neurosurgeons in their role as advocates for providing appropriate care to affected children.

Spontaneous occlusion of the circle of Willis in children: pediatric moyamoya summary with proposed evidence-based practice guidelines

A review

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Moyamoya is a progressive arteriopathy of unknown origin affecting the branches of the internal carotid artery (ICA). The arteriopathy can present as an isolated medical condition, affecting both sides of the brain (“moyamoya disease”) or can be unilateral or found in association with systemic disorders (“moyamoya syndrome”). The ischemia resulting from luminal narrowing predispenses children to transient ischemic attacks and stroke—the primary presentations of affected patients. Although it is rare—afflicting 1 in 1 million children in the US—moyamoya is implicated in 6% of all childhood strokes. Diagnosis is defined by characteristic findings on arteriograms, including stenosis of the branches of the ICA and a pathognomonic spray of small collateral vessels in this region, descriptively likened to a “puff of smoke” (“moyamoya” in Japanese). Treatment is predicated on restoration of cerebral blood flow by surgical revascularization. The rarity of this disorder has limited research and the development of evidence-based clinical management. While acknowledging these limitations, in this article the authors aim to summarize current studies of pediatric moyamoya, with the objective of providing a framework for construction of evidence-based guidelines for treatment. The compilation of current data in these guidelines should serve as a resource to aid pediatric neurosurgeons in their role as advocates for providing appropriate care to affected children.

Key Words • pediatric moyamoya • stroke • arteriopathy • pial synangiosis • revascularization

Abbreviations used in this paper: ACA = anterior cerebral artery; AHA = American Heart Association; CCA = common carotid artery; CTA = CT angiography; ECA = external carotid artery; ICA = internal carotid artery; MCA = middle cerebral artery; MR = MR angiography; STA = superficial temporal artery; TCD = transcranial Doppler; VA = vertebral artery.

Controversy surrounds a great deal of the literature on moyamoya, including the name itself and the question of how to define the condition. Some groups have proposed “spontaneous occlusion of the circle of Willis” as an alternative to moyamoya (the term currently recognized by the ICD). Terminology can be unclear, with “moyamoya,” “moyamoya disease,” and “moyamoya syndrome” sometimes used interchangeably. Each is distinct, with moyamoya being the most general term and defined as the characteristic findings on angiography, independent of any clinical qualifiers. Moyamoya disease is the presence of the arteriopathy bilaterally—either equally or to differing degrees on each side—in the absence of any other associated systemic disorder. In contrast, all unilateral cases are defined as moyamoya syndrome, as are any bilateral cases that are found in conjunction with the presence of well-recognized systemic disorders (see below).

The underlying cause of the arteriopathy described as moyamoya remains elusive. It is likely that many different factors—both genetic and environmental—contribute
to the disorder.\textsuperscript{14,39} As detailed in Table 1, moyamoya has been found in children with congenital malformations, genetic syndromes, and following the environmental stressor of cranial radiation.\textsuperscript{1,2,8,19,22,33,38,39,54} Independent of cause, evidence suggests that affected children are at risk for ischemic damage from progressive arteriopathy. Overall, the condition worsens in nearly all individuals, and more than two-thirds of patients will have clear symptomatic progression within 5 years, resulting in permanent neurological deficits or death without treatment.\textsuperscript{7,11,21,29,47}

Data indicate that a patient’s neurological status at the time of treatment is the most important predictor of long-term outcome.\textsuperscript{39} The inconsistent results of attempted medical therapy do not match the marked success of surgical treatment, which reduces symptomatic progression from >66% down to <4%.\textsuperscript{15} However, the rare nature of the disorder, particularly in North America, has resulted in the evolution of markedly individualized methods of evaluation and treatment of moyamoya among physicians caring for affected children.

Although we appreciate that previous publications have offered guidance on the management of moyamoya in children, including the recently published \textit{Management of Stroke in Infants and Children: A Scientific Statement From a Special Writing Group of the American Heart Association Stroke Council and the Council on Cardiovascular Disease in the Young},\textsuperscript{34} there remains a need for solely surgeon-authored guidelines in North America to complement these works, given the unique perspective offered by the physicians who directly treat the disorder. The compelling evidence supporting the use of surgery as a primary treatment for moyamoya mandates the need for pediatric neurosurgeons to advocate for its use on behalf of affected individuals.\textsuperscript{15,34}

It is important to note that the objective of this work is to unite those involved in the surgical care of children affected with moyamoya. While many groups—including the authors—may prefer specific surgical techniques or management strategies, the goal of this article is to present data in a neutral manner to serve all surgeons. In addition, it is worth explicitly noting that these guidelines are neither endorsed by any governing body nor serve as a substitute for individual physician judgment or expertise in any specific case. Rather, the authors seek solely to provide a summary of information as a (hopefully) useful resource to surgeons caring for children with moyamoya.

What follows is a summary of recent literature relevant to the screening, diagnostic evaluation, indications for surgery, and perioperative and operative management of the disease, along with follow-up, in children with moyamoya. These data are summarized into discrete guidelines for each section, with the level of evidence supporting their use. The scale used is directly derived from the AHA levels of evidence grading algorithm (Table 2). Information was derived from a search of all MEDLINE articles citing moyamoya that were written in the English language and dated up to the time of submission of this article.

\textbf{Definition and Screening}

The diagnosis of moyamoya is defined by 3 angiographic criteria, based on the Japanese Ministry of Health and Welfare guidelines.\textsuperscript{13} First, there must be stenosis of the distal (intracranial) ICAs, up to and including the bifurcation, along with segments of the proximal ACA and MCA. Second, dilated basal collateral vessels must be present (to varying degrees, depending on stage). Third, the findings must be bilateral (Fig. 1).

\begin{table}[h]
\centering
\begin{tabular}{|l|}
\hline
sickle cell disease \\
NF1 \\
previous cranial therapeutic radiation \\
Down syndrome \\
primary dwarfism \\
congenital cardiac anomaly \\
renal artery stenosis \\
giant cervicofacial hemangiomas & PHACE syndrome \\
hyperthyroidism \\
Alagille syndrome \\
\hline
\end{tabular}
\caption{Moyamoya syndrome—associated conditions*}
\end{table}

* NF1 = neurofibromatosis Type 1; PHACE = posterior fossa abnormalities, hemangioma, arterial lesions, cardiac abnormalities and/or aortic coarctation, and eye abnormalities.
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**TABLE 2: Definition of classes and levels of evidence used in AHA Stroke Council recommendations**

| Class I | conditions for which there is evidence for and/or general agreement that the procedure or treatment is useful and effective |
| Class II | conditions for which there is conflicting evidence and/or a divergence of opinion about the usefulness/efficacy of a procedure or treatment |
| Class IIa | the weight of evidence or opinion is in favor of the procedure or treatment |
| Class IIb | usefulness/efficacy is less well established by evidence or opinion |
| Class III | conditions for which there is evidence and/or general agreement that the procedure or treatment is not useful/effective and in some cases may be harmful |

**therapeutic recommendations**
- level of evidence A: data derived from multiple randomized clinical trials
- level of evidence B: data derived from a single randomized trial or nonrandomized studies
- level of evidence C: consensus opinion of experts

**diagnostic recommendations**
- level of evidence A: data derived from multiple prospective cohort studies using a reference standard applied by a masked evaluator
- level of evidence B: data derived from a single grade A study, ≥1 case-control studies, or studies using a reference standard applied by an unmasked evaluator
- level of evidence C: consensus opinion of experts


There are many situations in which the diagnosis of moyamoya is highly probable but may not meet these strict criteria. Moyamoya syndrome is unilateral and may not involve all the segments described above, although anterior circulation involvement is mandatory. The condition may be determined with the use of other imaging modalities, such as CTA or MRA, although care must be taken to account for technical issues that may be related to these techniques (such as the hematocrit in patients with sickle cell disease or other anemias).

There are no broad-based initiatives nor any Class I data supporting general screening protocols for moyamoya syndrome. Current AHA guidelines do not support indiscriminate screening, and state that imaging should be reserved for individuals with specific symptoms to suggest the presence of moyamoya in selected populations (see Table 1). This practice is supported by data demonstrating a lower Suzuki stage and decreased stroke burden—both clinically and radiographically—when screening is performed in these at-risk populations.

Special note should be made regarding patients with sickle cell disease. In this population, 10% of patients will have a stroke before the age of 20 years, with many of these children (approximately 40%) demonstrating “moyamoya-like” changes on imaging. Data indicate that in this subset of patients medical therapy is likely to fail, and there is some evidence to support the premise that strokes can be reduced or abrogated with cerebral revascularization.

The evidence supporting screening first-degree relatives of patients with moyamoya is less compelling. The familial incidence of affected first-degree relatives in Japan is 7%–12%, and a similar rate of approximately 6% was found in a North American series.

**Guidelines**

1. Screening for moyamoya may be considered in select populations (Table 1), particularly in the setting of symptoms suggestive of cerebral ischemia (Class IIb, diagnostic recommendation C).
2. Screening first-degree relatives of patients diagnosed with moyamoya may be of markedly limited utility in the absence of symptoms or multiple affected family members (Class IIb, diagnostic recommendation C).
3. Patients with sickle cell disease who manifest persistently elevated TCD velocities (> 200 cm/second) unresponsive to exchange transfusions (with sickle cell hemoglobin fraction < 30%) should be offered noninvasive imaging (such as MRA) to screen for moyamoya angiopathy (Class IIa, diagnostic recommendation B).

**Diagnostic Investigations**

Moyamoya syndrome should be considered and diagnostic evaluation begun in any child who presents with symptoms of cerebral ischemia (for example, a transient ischemic attack manifesting as episodes of hemiparesis, speech disturbance, sensory impairment, involuntary movement, and/or visual disturbance), especially if the symptoms are precipitated by physical exertion, hyperventilation, or crying. Although immediate evaluation commonly includes the use of CT scanning to assess for the presence of hemorrhage or large infarction, MRI and MRA have greater sensitivity for detecting acute stroke (with diffusion-weighted imaging) and the presence of the characteristic vessel changes associated with moyamoya. The formal diagnosis of moyamoya requires the use of catheter angiography, although this is not clinically necessary in all cases.
On CT scans, small areas of hypodensity suggestive of stroke are commonly observed in cortical watershed zones, basal ganglia, deep white matter, or periventricular regions. Although rare in children, hemorrhage from moyamoya vessels can be readily diagnosed on head CT scans, with the most common sites of hemorrhage being the basal ganglia, ventricular system, medial temporal lobes, and thalamus. The use of CT scanning alone is not adequate to confirm the diagnosis of moyamoya, although the use of CTA has the capacity to identify arterial narrowing and—in more advanced cases—the presence of collateral vessels at the base of the brain.

For most moyamoya cases, MRI will be performed. Acute infarcts are well seen using diffusion-weighted imaging, chronic infarcts are better delineated with T1- and T2-weighted imaging, and cortical ischemia may be inferred from FLAIR sequences that demonstrate a linear high signal following a sulcal pattern, which is believed to represent slow flow in poorly perfused cortical circulation. Most suggestive of moyamoya on MRI is the finding of diminished flow voids in the ICA, MCA, and ACA, coupled with prominent collateral flow voids in the basal ganglia and thalamus. These imaging findings are virtually diagnostic of moyamoya syndrome.

Because of the excellent diagnostic yield and noninvasive nature of MRI, it has been proposed that MRA be used as the primary diagnostic imaging modality for moyamoya syndrome instead of conventional cerebral angiography. Although MRA affords the ability to detect stenosis of the major intracranial vessels, visualization of basal moyamoya collateral vessels and smaller-vessel occlusions is frequently subject to artifact. Therefore, to confirm the diagnosis of moyamoya syndrome and to visualize the anatomy of the vessels involved and the patterns of flow through the hemispheres, conventional catheter-based cerebral angiography is typically required.

Catheter angiography offers the advantages of providing a gold standard for diagnosis, while also confirming the Suzuki stage and revealing blood supply derived from collateral vessels—including those from the ECAs. This information can be useful in planning operative approaches, allowing the surgeon to minimize disruption of existing collateral networks. The risk of complications from performing angiography in children with moyamoya syndrome has been demonstrated to be no higher than the risk of performing angiography in nonmoyamoya populations being evaluated for cerebrovascular disease.

Cerebral blood flow studies, using techniques such as TCD ultrasonography, Xe-enhanced CT, PET, and SPECT with acetazolamide challenge also can be helpful in the diagnostic evaluation of patients with moyamoya syndrome as well as assisting in treatment decisions. The Xe-CT, PET, and SPECT methods can be used both to detect regional perfusion instability prior to treatment and to determine the extent of improvement of functional perfusion after therapy.

There are scarce data supporting the use of other diagnostic methods—such as electroencephalography or TCD studies—as the primary means by which to confirm the presence of moyamoya. To date, there are no established genetic tests that identify moyamoya.

Indications for Surgery

Currently, there are no Class I or II data to support specific determinants of indications for medical versus surgical therapy. The quality of evidence compiled in a recent meta-analysis of 1448 patients reported in 57 studies in the English language led to recommendations that were graded “D” (on a scale from A to D), meaning that they are based completely on Class III data (nonanalytic studies) and expert opinions. The review noted multiple difficulties with objectively assessing the efficacy of surgery, including the limited data on the natural history of moyamoya as a comparison, the absence of head-to-head trials comparing treatments, the lack of universally accepted indications for surgery, the wide variation in surgical techniques, the potentially subjective nature of clinical outcome measures, the paucity of more objective outcome data (such as stringent demonstration of postsurgical improvement in vascularization or perfusion), and the small number of studies with long-term follow-up. With these caveats, the analysis concluded that “the data from the medical literature suggest that surgical revascularization is a safe intervention for pediatric moyamoya syndrome and most treated patients derive some symptomatic benefit.”

Indications for surgery were noted in < 15% of studies and varied between centers. General indications and timing of surgery remain controversial. The meta-analysis noted that the following statements had been used as indications for surgery: “neurological signs and symptoms likely to be related to cerebral ischemia and angiographic documentation of moyamoya disease,” “repeated ischemic attacks or progressive mental retardation,” “low cerebral blood flow in the frontal or occipital area, frontal or occipital atrophy on CT,” “transient weakness or hypoperfusion,” “inexorable progression of symptoms,” “symptomatic/decreased hemodynamic reserve on cerebral blood flow study,” “recurrent TIA [transient ischemic attack] and/or stroke after first operation,” and “ischemic symptoms.”

Recent guidelines from Japan’s Ministry of Health and Welfare regarding indications for surgical treatment of moyamoya state the following: “In the cases with (1) repeated clinical symptoms due to apparent cerebral ischemia, or (2) a decreased regional cerebral blood flow, vas-
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Cerebral response and perfusion reserve, based on the findings of a cerebral circulation and metabolism study, surgery is indicated. However, the guidelines are unclear regarding the timing of surgery, except to state that patients who present with acutely symptomatic hemorrhage may require emergency operative decompression or CSF diversion. In the US, the recent AHA guidelines are similarly broad, suggesting that indications for revascularization surgery include “progressive ischemic symptoms or evidence of inadequate blood flow or cerebral perfusion reserve in an individual without a contraindication to surgery.”

Guidelines

1. Revascularization should be offered to children with evidence of moyamoya, including ongoing ischemic symptoms and/or evidence of compromised blood flow or cerebral perfusion reserve, barring other medical contraindications to surgery. Specifically, children who are clinically asymptomatic but who demonstrate radiographic or functional evidence of impaired cerebral perfusion in the setting of moyamoya arteriopathy should be considered as surgical candidates (Class I, therapeutic recommendation B).

2. No firm recommendations can be made on specific timing for surgery, although the general principle of minimizing the time between diagnosis and revascularization is supported. Delays may be reasonable—or required—to allow scheduling of experienced anesthetic and ICU staff to maximize the safety of the procedure. There may also be medical contraindications to surgery that mandate delays prior to revascularization (such as recent infarction, infection, or hemorrhage)—(Class IIb, therapeutic recommendation C).

Perioperative and Operative Management

Treatment of moyamoya is predicated on restoring blood flow to the affected hemisphere, because there are no known methods of arresting the underlying arteriopathy. Operative goals focus on reducing the risk of ischemic and hemorrhagic injury through improved cerebral perfusion.

Perioperative Management

Medical therapy has 3 major components: 1) prevention of thrombosis; 2) maintenance of intravascular volume; and 3) mitigation of nonischemic symptoms (such as headache and seizure). Antithrombotic agents are used in many moyamoya centers to prevent microthrombi at sites of arterial stenosis. Aspirin is most commonly used and is dosed according to weight (usually 81 mg daily in children), with some clinicians preferring the use of low-molecular-weight heparin. Maintenance of intravascular volume often does not require administration of medication, but does involve careful monitoring of fluid balance in children. The primary focus is on avoidance of dehydration than supplementation of overall intake. Care must be taken in children at risk for fluid loss from illness (diarrhea, vomiting, and so on), exercise, or activity in hot weather. Particular attention to fluid status must be paid in the perioperative period (see below). Mitigation of nonischemic symptoms usually involves treatment of seizures in affected patients with antiepileptic medication and management of headache with analgesics. Particular note should be made of the practice of using calcium channel blockers to ameliorate headache, because they can be very effective, but they can also potentially increase the risk of stroke through their propensity to induce hypotension.

Perioperative management of moyamoya in children requires coordinated care between the anesthesia, ICU, nursing, neurosurgery, and affiliated medical teams (such as hematology in patients with sickle cell disease). Evidence indicates that hyperventilation—as occurs in crying—can increase the risk of stroke in the perioperative period, supporting the use of measures to decrease pain (appropriate analgesia, use of absorbable sutures, sedation for potentially painful procedures) and reduce psychological stressors in children. These efforts are particularly important in select populations, such as the very young, children with developmental delays (such as Down syndrome), and children who may have preexisting high tolerances to pain medications (such as sickle cell patients).

One of the greatest concerns regarding surgery is the risk of perioperative stroke, reported to occur in 4%—10% of operations. There is evidence that the use of pre- and postoperative hydration may reduce the risk of stroke by minimizing blood pressure fluctuations. Selective use of neuroprotective anesthetic agents (such as propofol) to reduce cerebral metabolism during periods of intraoperative instability (such as electroencephalographic slowing), coupled with tight blood pressure control, may reduce the risk of stroke. Postoperative management of blood pressure and oxygenation, coupled with systemized monitoring of the neurological examination, as can be done in the ICU, may also improve outcomes.

Operative Management

Surgery for moyamoya is predicated on the finding that the branches of the ECA are not affected in the disorder, and thus can be used as a source of blood supply to the ischemic brain. Two major approaches are commonly used (direct and indirect), with some surgeons opting to combine them in single procedures. Direct approaches use a branch of the ECA (usually the STA) as a graft, anastomosed to a cortical artery (usually a branch of the MCA). Indirect approaches use vascularized tissue (such as the dura mater, muscle, or an artery with a cuff of adventitia) to stimulate the growth of a new vascular network when placed in contact with the brain. Both types of approaches have been used with great success in children, markedly reducing the risk of stroke when performed at high-volume centers.

Considerable debate exists over the selection of an individual operative approach, and centers differ in their philosophy of treating bilateral disease with single or staged craniotomies. However, all surgical treatments, when performed at high-volume centers on carefully selected patients, yield excellent results when contrasted to the expected natural history. Long-term follow-up of patients treated with pial synangiosis revealed that 67%
of them had strokes preoperatively, but with those who had 5 years of follow-up, the rate was only 4.3% after surgery.\textsuperscript{19} This mirrors data from other centers, including a meta-analysis of \textgreater 1100 treated individuals, supporting the premise that surgical treatment of moyamoya confers durable, marked reductions in stroke.\textsuperscript{15,17,39}

One area of controversy in the treatment of moyamoya is selection of the surgical approach. Overall, indirect techniques are used in approximately 75% of all pediatric moyamoya cases, with the remainder composed of direct or combined approaches.\textsuperscript{15} Most US centers will initially revascularize the territory of the MCA, although it is the practice in some Asian hospitals to also include ACA revascularization as part of the initial surgery.\textsuperscript{27} Although each center often has a preferred technique, data would suggest that outcomes are very similar regardless of specific approach. Rather, the key factor in successful outcome appears to be the experience of the surgeon and institution with the care of patients with moyamoya.\textsuperscript{15,38,39}

An evolving field in moyamoya is the application of endovascular techniques to the treatment of both the primary disease and also associated complications. Recent reports have described attempts to open stenosed vessels with endovascular tools such as stents, with poor results.\textsuperscript{9,25} With current technology, it appears that sustained patency of vessels is not possible, probably due to the progressive nature of moyamoya, involving long stretches of multiple vessels. More data are needed to assess the possibility of using endovascular treatments to manage acute ischemic events more effectively, with direct application of angioplasty and intraarterial vasodilators and/or thrombolytic agents.\textsuperscript{10}

Guidelines

1. Use of techniques to reduce pain and crying in children with moyamoya during the perioperative period may reduce the risk of stroke (Class IIb, therapeutic recommendation C).
2. Administration of perioperative intravenous hydration, coupled with intraoperative control of blood pressure at normal or slightly elevated levels, use of neuroprotective agents, and maintenance of oxygenation is associated with reduced stroke rates in children with moyamoya (Class IIb, therapeutic recommendation C).
3. Long-term aspirin use may reduce the risk of stroke in children with moyamoya. Only patients unable to take aspirin or who exhibit refractory symptoms should be primarily considered as candidates for other anticoagulation agents (Class IIb, therapeutic recommendation C).
4. Surgical revascularization provides long-term, durable, and marked reductions in stroke risk for children with moyamoya. These results are achievable with both direct and indirect techniques, with the major predictor of outcome centered primarily on surgeon experience (Class I, therapeutic recommendation B).
5. Current evidence does not support the use of endovascular stent placement as a primary treatment for moyamoya in children (Class III).

Follow-Up

After the diagnosis of moyamoya, patients often receive lifetime follow-up care by neurosurgeons or neurologists, because the primary arteriopathy is not arrested—even with surgery—and there remains a lifelong risk of stroke. This risk is markedly reduced with surgery, however, and there is a 96% probability of remaining stroke free over the subsequent 5 years.\textsuperscript{7,38,59}

One challenging area in the follow-up of patients with moyamoya involves unilateral arteriopathy. It is unclear which patients will go on to develop bilateral moyamoya and which will remain stable, although data suggest that approximately one-third will need surgery within 5 years, with risk factors for progression including young age and the presence of any arteriographic anomalies on the initially unaffected side.\textsuperscript{24,41}

Guidelines

1. Given the possibility of disease progression and further stroke even after successful surgery, periodic follow-up of surgically treated patients with moyamoya by clinicians familiar with moyamoya is supported, with initial visits ideally annually for 3–5 years (Class IIb, therapeutic recommendation C).
2. Patients with unilateral moyamoya should obtain annual imaging with MRA or CTA to assess for disease progression for a period of at least 3–5 years (Class IIb, therapeutic recommendation C).

Conclusions

Moyamoya remains a complex entity to diagnose and treat. Evidence supports the use of dedicated imaging studies to confirm the diagnosis, including MRI and catheter angiography. Medical and perioperative management is predicated on the reduction of stroke risk through avoidance of thrombosis and minimizing fluctuations in cerebral blood flow and oxygenation. There are strong data supporting the effectiveness of cerebral revascularization as a method to reduce ischemic injury substantially and durably when it is performed in high-volume centers. Long-term follow-up is important, particularly in patients with unilateral arteriopathy.

The goal of these guidelines is to provide recommendations for the diagnosis and treatment of children with moyamoya. The preponderance of evidence supports the use of surgical revascularization as an effective treatment for this condition. The compilation of current data in these guidelines should serve as a resource to aid pediatric neurosurgeons in their role as advocates for providing appropriate care to affected children.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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