Moyamoya, also known as Spontaneous Occlusion of the Circle of Willis, is an uncommon cerebral vasculopathy. It is associated with progressive occlusion of the terminal internal carotid arteries and the formation of a fine network of neovascularization referred to as moyamoya vessels. The disorder takes its name from the Japanese word for the angiographic appearance, which resembles a wafting puff of smoke. This condition was first described in 1957 in Japanese children. The Asian form of moyamoya disease is an idiopathic condition with a well-defined phenotype that occurs predominately in children of Japanese ancestry. However, a clinically distinct and less well described secondary form of moyamoya occurs in adults of all ethnic backgrounds and is termed moyamoya syndrome. While relatively rare, this condition is potentially treatable and, with increasing awareness, will likely be discovered more frequently.

Adult moyamoya syndrome is strongly associated with cranial radiation exposure, typically occurring two or more years after treatment. Several genetic diseases have been reported as risk factors, including Down syndrome, sickle cell disease, and neurofibromatosis type 1. Severe intracranial atheromatous disease may also lead to the formation of moyamoya vessels. In addition, fibromuscular dysplasia, hyperthyroidism, bacterial meningitis, combined cigarette smoking and oral contraceptive use, Sjogren syndrome, and cocaine abuse have all been implicated.

Diagnosis and Management
Diagnosis of moyamoya requires the presence of the characteristic angiographic findings. The terminal internal carotid arteries initially demonstrate narrowed lumens that progress to complete occlusion in later stages. The occlusive disease is almost always bilateral but may be asymmetric and spread to involve the anterior or middle cerebral and posterior communicating arteries. In response to the reduced blood flow, innumerable tiny penetrating vessels sprout from the affected vessels and act as collateral channels. Branches of the external carotid may also be recruited for collateral circulation. A grading system illustrating the six stages devised by Suzuki and Takaku is shown in Table 1.

This compensatory vascular network, however, is fragile and ultimately unable to sustain the ischemic tissue. The friable moyamoya vessels may rupture leading to hemorrhage. Hemodynamic changes that increase flow through collateral vessels may lead to the formation of aneurysms, most

<table>
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<tr>
<th>Suzuki Stage</th>
<th>Angiographic Finding</th>
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<tr>
<td>I</td>
<td>Narrowing of carotid arteries</td>
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<td>II</td>
<td>Initial appearance of moyamoya vessels</td>
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<td>III</td>
<td>Intensification of moyamoya vessels</td>
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<td>IV</td>
<td>Minimization of moyamoya vessels</td>
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<td>VI</td>
<td>Disappearance of moyamoya vessels</td>
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commonly in the posterior circulation. The vascular territories supplied by the neovasculature are chronically oligemic, which often leads to ischemic infarction.

The typical clinical presentation in the United States is a young woman with either recurrent ischemic or hemorrhagic events, which may coexist in the same patient. The adult syndrome occurs in women two to four times more often than in men and has a peak incidence between 30 and 50 years of age. Ischemic stroke in the vulnerable territory occurs in approximately three-quarters of patients. Approximately one quarter of patients present with hemorrhage, yet a small number of patients are asymptomatic at the time angiographic findings are incidentally discovered.

While the natural history of this syndrome is largely unknown and likely dependent in part on the associated underlying condition, the risk of recurrent events is high: as high as 65 percent in those treated conservatively. In contrast, the rate of progression in surgically treated patients is substantially lower at 2.6 percent, as estimated by a meta-analysis of 1,156 patients. Revascularization may be accomplished surgically through direct or indirect techniques.

In the majority of direct revascularization procedures, the superficial temporal artery is anastomosed to the middle cerebral artery or another large intracranial artery. When the diameter of the STA is too small for direct anastomosis, the occipital artery can be considered as a donor artery. The result is immediate restoration of blood flow with a high rate of long-term patency and normalization of cerebral hemodynamics. But the procedure is not without significant complications. Perioperative ischemia may occur due to clamping of the recipient artery during anastomosis, and one recent study of adult moyamoya patients found the perioperative morbidity and mortality was 12.3 percent and 4.6 percent, respectively. The immediate increase in cerebral blood flow also places the patient at risk for symptomatic hyperperfusion and transient neurologic deterioration.

Indirect techniques involve a wide craniotomy and placement of a vascularized tissue supplied by the external carotid in direct contact with the cortical surface. Collateral vessels then sprout from the graft. These techniques are less invasive, do not require clamping of a recipient vessel, and are technically simpler. A number of indirect procedures have been described.

Encephalomyosynangiosis (EMS), first described in 1977, involves placing a section of temporalis muscle directly over the underlying cerebral cortex after the arachnoid layer has been removed. Encephaloduroarteriosynangiosis (EDAS), described in 1980, entails suturing a galeal cuff with an intact STA onto the cortical surface once the dura has been removed. These procedures or other variations may be used singly or in combina-
tion and are tailored to the patient's anatomy. The main disadvantage of indirect techniques is the extended time period required for neovascularization to occur, which is usually several months.\textsuperscript{34} While indirect techniques have been shown superior in the pediatric population,\textsuperscript{26} the success rate in adults is significantly less. In 40-50 percent of adults receiving indirect revascularization, collateral vessels fail to grow.\textsuperscript{26,35}

**Aggressive Intervention Warranted**

It is important to note that the majority of research published on the treatment of moyamoya in adults relates to the Asian form (moyamoya disease) and not the secondary moyamoya syndromes more commonly encountered in the United States. One fact, however, is indisputable: moyamoya syndrome and disease are associated with considerable morbidity. Aggressive treatment appears to yield the best outcomes.

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7. Koc, F., D. Yerdelen, and Z. Koc, Neurofibromatosis type I association with moyamoya dis-

**Figure 2.** A CT angiogram from a 49-year-old woman presenting with intraventricular hemorrhage demonstrating Suzuki Stage II moyamoya syndrome. The right internal carotid artery is nearly occluded. The anterior and middle cerebral arteries demonstrate moyamoya vessels.

**Figure 3.** A CT perfusion study from the same patient in Figure 2 demonstrating impaired perfusion in the right hemisphere.

**Figure 4.** Antero-posterior and lateral views of a cerebral angiogram demonstrating occlusion of the left ICA terminus with appearance of moyamoya arteries in a typical “puff of smoke” pattern.