Optimal surgical treatment for moyamoya disease in adults: direct versus indirect bypass

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Moyamoya disease is a chronic cerebrovascular occlusive disorder that results in severe morbidity and death. There is much controversy surrounding the optimal treatment for adult patients with the disorder. There have been no randomized trials to assess the efficacy of any single surgical treatment, and existing case series suffer from inadequate power, selection bias, and inherent differences in patient characteristics. In this article the authors review the literature concerning the optimal surgical treatment of adult patients with moyamoya disease. (DOI: 10.3171.2009.01.FOCUS08309)

KEY WORDS • indirect bypass • encephaloduroarteriosynangiosis • moyamoya disease • outcome • stroke • hemorrhage

MOYAMOYA disease is a chronic cerebrovascular disorder defined by progressive occlusion of the intracranial vessels. The stenosis begins with the intracranial carotid arteries and can progress to involve the anterior, middle, and posterior cerebral arteries. As these arteries gradually stenose, a collateral network of capillaries develops at the base of the brain, producing the characteristic reticulate appearance (“puff of smoke”) on angiography.

In Asian populations, moyamoya disease has a well-defined phenotype. The disease has a bimodal age of presentation, with ischemia developing in children because of inadequate collateral vessels and adults presenting with intracranial hemorrhage due to the rupture of fragile collateral vessels.^{35,39} A number of studies have provided evidence that moyamoya disease outside of Asia can represent a different phenomenon.^{4,7,21,36,38} Patient ethnicities are in proportion to the ethnicities of the people in the region of diagnosis, and adult patients present with ischemic symptoms rather than intracranial hemorrhage.

Although no completed randomized clinical trials have addressed the benefit of surgical revascularization, the dismal prognosis of moyamoya disease^{2–4,7,8,21,23,31,33,41–43} and a large number of case series demonstrating an improved outcome in surgically treated patients have provided evidence in support of revascularization procedures in a select group of patients.^{4,7–11,16,17,25,29,30,32,34,40,43}

Currently, the Ministry of Health and Welfare of Japan reports that bypass surgery is indicated when there are 1) repeated clinical symptoms due to apparent cerebral ischemia and (2) decreased regional cerebral blood flow, vascular response, and perfusion reserve.⁶ Some authors have reported a benefit from revascularization in patients presenting with intracranial hemorrhage.^{8,10,15,40} Although patients with nonatherosclerotic moyamoya disease and hemodynamic failure appear to benefit from surgical revascularization procedures, the optimal bypass technique has yet to be elucidated.

Direct Bypass for Moyamoya Disease in Adults

Direct STA-MCA bypass surgery has been successfully used to augment collateral blood flow in patients with moyamoya disease for > 30 years.^{11,13,22,25,26} Many case series have demonstrated a decrease in ischemic symptoms and/or the maintenance of cognitive abilities in patients treated with direct bypass, but these series have been composed chiefly of children and often have included patients treated with indirect bypass or a combination of direct and indirect bypasses.^{4,11,13,25,34,37,43} Thus far, no study has documented overall long-term rates of ischemic events in adult patients treated solely with direct bypass.

Indirect Bypass for Moyamoya Disease in Adults

Encephalomyosynangiosis was first proposed for the treatment of indirect bypass by Karasawa et al.¹² in 1977, and EDAS by Matsushima et al. in 1981.²⁸ Since

Abbreviations used in this paper: EDAMS = EMS and EDAS; EDAS = encephaloduroarteriosynangiosis; EMS = encephalomyosynangiosis; MCA = middle cerebral artery; STA = superficial temporal artery.

then a number of other procedures have been suggested, including a combined EDAS and EMS approach (EDAMS),¹⁹ pial synangiosis,¹ and a combined direct and indirect approach.³⁰ Indirect bypass has also been reported to be beneficial in the treatment of moyamoya disease.^{4,8,11,19,20,24–26,34,37,40,43} In one of the largest series of pediatric patients with moyamoya syndrome (143 cases), the majority of patients stopped having strokes and transient ischemic attacks and experienced excellent functional outcomes.⁴⁰ However, most series have been composed chiefly of children and often have included patients treated with direct bypass or a combination of direct and indirect bypass. A case series of 4 adult patients treated with indirect bypass in Japan documented adequate collateral vessel formation and decreased moyamoya vessels.²⁹ In a series of adult patients (range 16–60 years) presenting in Korea with primarily intracranial hemorrhage, the 17 patients treated with EDAS had significantly better clinical outcomes than the 9 patients in the nonoperative group after a median 12-month follow-up period.⁸ In a case series of 20 adult patients (≥ 21 years old, median 30 years) treated with pial synangiosis in North America, 9 of 11 patients had radiographic evidence of collateral formation (Smith et al., unpublished data, 2008). Patients had low rates of periprocedural complications and long-term ischemic events. In a series of 43 adult patients (mean $\pm$ SD, 40 ± 11 years, range 18–69 years) presenting with ischemic symptoms and treated with pial synangiosis, we found that indirect bypass promotes adequate pial collateral development and increased perfusion. After surgery, patients had a low incidence of transient ischemic attacks, infarction, and hemorrhage, with most patients having preserved or improved functional independence (R.M. Starke et al., unpublished data, 2008).

Comparison of Direct and Indirect Bypass for Moyamoya Disease

Some consider direct STA-MCA bypass to be the optimal procedure for patients with moyamoya disease, but there is little evidence to reveal the best overall treatment. Currently, there have been no randomized trials to assess the efficacy of any single surgical treatment, and case series suffer from inadequate power, selection bias, and inherent differences in patient characteristics. Frequently, authors have reported the outcomes of both children and adults, and differences in the severity of the disease as well as in the timing and mode of presentation have made it difficult to assess the best overall treatment option. Currently, only relatively small case series have demonstrated the natural history and long-term outcomes of surgically treated patients with moyamoya disease.

A number of researchers have noted that it is particularly difficult to judge outcomes following surgery in patients with moyamoya disease, as surgery is a form of palliative care in a disease with a poor natural history.⁴³ Researchers have not used standardized outcome measures to assess patients on presentation and on long-term follow-up. Furthermore, neuropsychological testing after surgical treatment is critical to accurate outcome assessment and should be integrated into future clinical trials.¹⁴

Direct STA-MCA bypass can be difficult in children because of both the size and progressive occlusion of the MCAs. Indirect bypass is also considered easier and safer in patients with serious medical comorbidities and more feasible in patients with inadequate recipient or donor artery grafts. Rates of periprocedural ischemia have ranged from 4–31% in patients treated with bypass for moyamoya disease.^{4,7–9,11,25,27,34} Higher rates of periprocedural ischemia are often a composite of short-term reversible and permanent deficits. Although not addressed by most authors, indirect bypass is safer and leads to fewer periprocedural complications.^{4,43} Furthermore, up to 28% of patients can experience symptomatic cerebral hyperperfusion after direct bypass, although these deficits are usually transient.^{5,18}

Authors have suggested that direct bypass is more beneficial because it provides immediate revascularization as compared with indirect bypass, which can require several weeks to form collateral vessels.^{11,43} Although patients were not matched or randomized in 2 pediatric studies (10 patients²⁵ and 34 patients¹¹), the authors have reported significantly improved outcomes in patients treated with a combination of direct and indirect versus indirect bypass alone. Another series of 56 pediatric and 15 adult patients has revealed similar low rates of ischemic events in those treated with STA-MCA bypass, EDAS, EDAMS, ribbon EDAMS, and combination procedures.³⁴ The best rates of collateral formation occurred with EDAMS and ribbon EDAMS procedures because of wider distribution to both the MCA and anterior cerebral artery distributions. The authors have recommended these procedures primarily for pediatric patients and a combined direct and indirect bypass for adult patients. In another series of 16 adults and 7 children in which each patient was treated with both direct and indirect bypass, 2 children experienced minor transient ischemic events and the adults experienced no ischemic event. The authors of this study reported better collateral formation with direct bypass in the older patients and better collateral formation through the indirect bypass in younger patients.³⁰ Additional studies are necessary to better address which type of bypass provides the best collateral formation, the lowest rates of periprocedural complications, the lowest rates of long-term ischemic events, and the best preservation of functional outcome.

Conclusions

Currently, the most effective treatment in adult patients with moyamoya disease is unknown. There is some evidence that both direct and indirect bypasses are effective means of revascularization and reduce the incidence of ischemic events in adults with moyamoya disease. A randomized clinical trial is necessary to determine the best treatment modality in patients with this disorder.

Disclaimer

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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