Outcome of repeat revascularization surgery for moyamoya disease after an unsuccessful indirect revascularization

Clinical article

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Object. Revascularization for moyamoya disease, either by direct anastomosis or indirect procedures, is an accepted and effective form of treatment for prevention of future ischemic events. Indirect procedures do not provide sufficient collateral vessels in a subset of patients, who then have persistent or new symptoms. Repeat revascularization procedures may be recommended for these patients.

Methods. Sixteen patients underwent repeat revascularization after undergoing an indirect procedure in the same hemisphere. These patients were included in the study, and a retrospective review of their clinical details, neuroimaging results, surgical details, and outcome was performed. Direct revascularization was the procedure of choice; however, in patients with no acceptable recipient vessel (> 0.6 mm) the authors added a second indirect procedure for further revascularization.

Results. Over the last 19 years, 16 patients (8 male and 8 female patients, age range 5–48 years, mean 16.7 years, 10 pediatric and 6 adult patients) underwent repeat revascularization for moyamoya disease. Initially all patients presented with ischemic symptoms (4 transient ischemic attacks [TIAs] and 12 strokes; 2 patients had bilateral symptoms). Angiography revealed that 13 patients had bilateral disease, and 3 had unilateral disease. Initial surgery was bilateral encephaloduroarteriosynangiosis (EDAS) in 9, unilateral EDAS alone in 3, unilateral EDAS with contralateral superficial temporal artery–middle cerebral artery (STA-MCA) bypass in 2, bilateral encephalomyosynangiosis (EMS) in 1, and unilateral EMS in 1. Thirteen of the 16 patients continued to have TIAs in the hemisphere ipsilateral to surgery, whereas 1 patient had seizures and cognitive deficit, 1 had asymptomatic infarct on MR imaging, and 1 had visual symptoms. Poor revascularization was seen on angiography studies in all patients. The median duration between the surgeries was 24 months (3 months–10 years).

Repeat revascularization was performed in 23 hemispheres (16 patients). Direct revascularization was performed in 14 hemispheres (60.9%): STA-MCA bypass in 10, external carotid artery–MCA vein bypass in 2, occipital artery (OA)–MCA in 1, and OA–posterior cerebral artery in 1 hemisphere. Indirect revascularization was performed for patients without an acceptable recipient vessel, and was done in 9 hemispheres. The procedures included EMS (4 hemispheres), repeat EDAS (2), and omental transposition (3). There was 1 postoperative death in a patient undergoing a high-flow vein graft implantation. None of the other patients experienced any neurological worsening after surgery.

Follow-up was available in all patients, ranging from 3 to 144 months (mean 34 months, median 12 months). Of the 15 patients who survived repeat revascularization surgery, 12 (80%) were free from any TIA, stroke, or any other neurological symptoms. Two patients had occasional TIAs, less frequent than before, whereas 1 patient had frequent TIAs and underwent revision of the revascularization. Angiographic studies were available in 11 patients, and showed improved flow in the hemispheres in 10 patients. Follow-up MR imaging performed at 6 months did not reveal a new infarct in any patient.

Conclusions. Repeat revascularization procedures are effective for patients who are clinically symptomatic and have inadequate collateral vessels following indirect procedures. Although direct procedures are preferred, the choice of procedure depends on the operative findings and the status of donor and recipient vessels.

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Key Words • moyamoya disease • repeat revascularization • superficial temporal artery–middle cerebral artery bypass • vascular disorders

Abbreviations used in this paper: ACA = anterior cerebral artery; CBF = cerebral blood flow; DS = digital subtraction; ECA = external carotid artery; EDAS = encephaloduroarteriosynangiosis; EEG = electroencephalography; EMAS = encephalomyoarteriosynangiosis; EMS = encephalomyosynangiosis; ICA = internal carotid artery; MCA = middle cerebral artery; OA = occipital artery; PCA = posterior cerebral artery; SSEP = somatosensory evoked potential; STA = superficial temporal artery; TIA = transient ischemic attack.

Moyamoya disease is characterized by idiopathic, chronic, usually bilateral stenosis or occlusion of intracranial ICAs, proximal MCAs, and ACAs, and is a cause of stroke or TIAs in children and adults. Both direct and indirect methods of revascularization are described in the literature, and are aimed at providing collateral blood supply through the ECAs to the ischemic hemisphere.
Repeat revascularization surgery for moyamoya disease

A variety of indirect procedures have been described, including EDAS, EMS, omental transpositions, placement of multiple bur holes, and a combination of these procedures. Although they are successful in the majority of patients, especially in the pediatric population, a potential disadvantage of these procedures is that sometimes they do not provide sufficient collateral vessels to prevent symptoms. Repeat revascularization procedures are required for these patients with persistent or new symptoms. Depending on the donor and recipient vessels available, these might be either direct or indirect procedures. There have been a few reports describing the outcomes of repeat revascularization procedures, mostly from Japan. We present our experience and outcomes in patients undergoing repeat revascularization for moyamoya disease after an unsuccessful indirect procedure. This is the first report from North America describing repeat revascularization in moyamoya disease.

Methods

A prospectively maintained database (MD Analyze; Medtamic, Inc.) of patients with vascular malformations treated at Stanford University Medical Center was used to identify all patients with moyamoya disease. Over the last 19 years, we have performed revascularization procedures in 502 patients with moyamoya disease. Of these patients, 16 had repeat revascularization after undergoing indirect procedures in the same hemisphere. These patients were included in this study, and were analyzed retrospectively. A total of 23 hemispheres were revascularized in these 16 patients. Fifteen underwent initial revascularization at another institution, whereas 1 had the first revascularization at our institution. One other patient had a third revascularization procedure at another institution after undergoing the repeat revascularization procedure at our institution, as mentioned later in the paper. Thus, of the 502 patients undergoing revascularization surgery at our institution, the percentage of patients requiring a repeat surgery was 0.4%

The clinical presentation, neuroimaging, and surgical details of the patients previous to the first procedure were obtained from the initial institutions. After the referral to our institution, all 16 patients were evaluated with a clinical history and examination, MR imaging of the brain with perfusion, a 6-vessel angiogram, and CBF studies with either SPECT or Xe-CT scanning before and after administration of acetazolamide. The clinical findings and the radiological data were discussed in a multidisciplinary conference comprising the neurosurgeon, the neuroradiologist, and sometimes the stroke neurologist. The adequacy of the previous revascularization was assessed using the results from all the investigations, as well as from clinical examination. A patient was considered for repeat revascularization if he/she was symptomatic, and if the angiographic and/or CBF data suggested incomplete revascularization. A second surgery for revascularization was also considered if the patient was asymptomatic but had new infarcts on MR imaging of the brain, with angiographic and/or CBF studies suggesting incomplete revascularization. All these data were analyzed retrospectively by the authors with the neuroradiologist, and the charts, radiology films, and records were reviewed.

Choice of Procedures

In most of the patients, direct revascularization with STA-MCA bypass was our procedure of choice. The technique of STA-MCA bypass for moyamoya disease has been described before. In most of the patients, the branch used for EDAS was left intact, and another branch of the STA (or in 1 case, the OA) was used for direct revascularization. This was done to avoid disrupting the collateral vessels from the previous indirect surgery, and the potential ischemic sequelae. In only 2 instances, we used the same branch that was previously used for EDAS for direct revascularization, because the other branch was very small and the revascularization from the indirect procedure was extremely limited. In these patients the dissection of the STA was difficult, and there were adhesions to the brain from the previous indirect procedure. However, with careful dissection, it was possible to separate the vessel from the underlying brain. Although exact measurements were not performed, the craniotomies in the previous surgeries were not any different from those in the other patients in whom the indirect revascularization was successful. In patients in whom there was no acceptable (> 0.6 mm) recipient vessel available, we added a second indirect procedure, such as EMS, omental transpositions, or EDAS performed using another branch of the STA, for further revascularization. The techniques for these procedures have been described before in the literature.

Follow-Up Data

Follow-up was obtained from the clinic visit notes or through telephone communication when there was no clinic visit following the surgery. Imaging studies, including MR images of the brain, SPECT scans with and without acetazolamide, and a 6-vessel angiogram, were obtained at 6 months and 3 years following surgery.

Results

Patient Population

Over the last 19 years, 16 patients underwent repeat revascularization for moyamoya disease. There were 8 male and 8 female patients, whose ages ranged from 5 to 48 years (16.7 ± 12.57 years; mean ± SD). There were 10 pediatric (< 18 years of age) and 6 adult patients. Table 1 lists the demographic details of the patients.

Initial Surgery

Initial presentation was with ischemic symptoms in all patients; 4 presented with TIA, whereas 12 had stroke with deficits. Two patients had bilateral symptoms. Of the 12 patients presenting with stroke, 5 had major hemispheric stroke with residual deficits, whereas the other 7 had small cortical/subcortical infarcts with minor deficits, which improved prior to surgery. None of the patients had hemorrhage as the clinical presentation of the
disease. Brain MR imaging showed watershed infarcts in 6 patients, small hemispheric infarcts in 2, and large cortical infarcts in 8. Angiographic studies were done in all the patients and revealed bilateral moyamoya disease in 13 patients, whereas 3 had unilateral disease.

The predominant form of indirect surgery was EDAS, with bilateral procedures in 9 patients and unilateral in 5 (2 of these latter patients also underwent contralateral STA-MCA bypass). One patient underwent bilateral EMS and 1 had a unilateral EMS (Table 1). One patient had a major postoperative hemispheric infarct, and 1 patient had a major stroke 1 month after the indirect surgery. Two other patients had mild worsening of power in the contralateral hand, which recovered over a period of 1 month.

Indications for Repeat Surgery

Thirteen of the 16 patients continued to have TIAs in the hemisphere ipsilateral to their surgery after their indirect revascularization procedures. All of these patients had had recent TIAs (within the preceding month) when they presented to our clinic. One of these patients had a mild stenosis of the M1 segment, which progressed and caused severe M2 stenosis at follow-up. Of 3 other patients, 1 had seizures and also complained of increasing cognitive deficits. Another patient was clinically asymptomatic, but had evidence of recent infarcts on follow-up MR imaging. One patient who had a major hemispheric infarct with homonymous hemianopia had contralateral visual symptoms, which were thought to be posterior circulation TIAs. The median duration between the two surgeries was 24 months (3 months–10 years). New (or recent) infarcts on MR imaging on the side ipsilateral to indirect surgery were documented in 5 patients. Details of CBF studies were available in 12 patients. Imaging using HMPAO-SPECT was done in 8 patients, whereas 4 patients underwent Xe-CT with and without the administration of acetazolamide. All of them had perfusion deficits on the baseline CBF studies, with 7 patients showing acetazolamide reactivity, whereas 5 patients did not show expected augmentation after acetazolamide administration. Details of flow studies were not available in 4 patients. Angiography was performed in all patients, and showed poor revascularization in all of them, characterized by filling of fewer than 2 cortical branches of the MCA.

Repeat Surgery

Repeat revascularization was performed in 23 hemispheres, in 16 patients (Table 2). Direct revascularization with STA-MCA bypass was performed in 10 hemispheres, whereas an ECA-MCA anastomosis was done using a vein graft in 2 hemispheres. Direct revascularization in the form of OA-PCA and OA-MCA anastomosis was done in 1 hemisphere each. Thus, a total of 14 hemispheres (60.9%) underwent direct revascularization. In most of the STA-MCA anastomosis procedures, the branch used for initial EDAS was left intact, and another branch of the STA was used for contralateral the STA was used for anastomosis. In only 2 patients was the same branch of the STA used for direct anastomosis because the collateral vessels supplied by the EDAS were extremely limited on the preoperative angiogram, and the other branch of the STA was unavailable. This procedure was done after temporary clipping of the donor vessel, and waiting for 10 minutes to observe any changes in the EEG findings or SSEPs. Indirect revascularization was performed in 9 hemispheres, including EMS (4 hemispheres), repeat EDAS using another branch of the STA (2), and omental transposition (3). All of these patients underwent indirect revascularization because there were no acceptable recipient vessels available. One patient undergoing ECA-MCA bypass suffered reperfusion hemorrhage and a large intracerebral hematoma and

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Initial Presentation</th>
<th>Angio Findings Preop</th>
<th>Indirect Op</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>5, M</td>
<td>lt hemi TIA</td>
<td>bilat</td>
<td>bilat EDAS</td>
</tr>
<tr>
<td>2</td>
<td>5, M</td>
<td>rt hemi stroke</td>
<td>unilat</td>
<td>rt EMS</td>
</tr>
<tr>
<td>3</td>
<td>6, M</td>
<td>lt hemi stroke</td>
<td>bilat</td>
<td>bilat EDAS</td>
</tr>
<tr>
<td>4</td>
<td>7, M</td>
<td>rt hemi stroke</td>
<td>unilat</td>
<td>lt EDAS</td>
</tr>
<tr>
<td>5</td>
<td>8, F</td>
<td>rt hemi stroke</td>
<td>bilat</td>
<td>rt EDAS w/ multiple bur holes</td>
</tr>
<tr>
<td>6</td>
<td>8, M</td>
<td>bilat TIAs, lt hemi stroke</td>
<td>bilat</td>
<td>bilat EDAS</td>
</tr>
<tr>
<td>7</td>
<td>8, M</td>
<td>lt hemi stroke</td>
<td>bilat</td>
<td>bilat EMS</td>
</tr>
<tr>
<td>8</td>
<td>10, F</td>
<td>rt hemi stroke</td>
<td>bilat</td>
<td>bilat EDAS</td>
</tr>
<tr>
<td>9</td>
<td>11, F</td>
<td>lt hemi TIA</td>
<td>bilat</td>
<td>lt EDAS, rt STA-MCA bypass</td>
</tr>
<tr>
<td>10</td>
<td>14, F</td>
<td>lt hemi stroke/TIA</td>
<td>bilat</td>
<td>bilat EDAS</td>
</tr>
<tr>
<td>11</td>
<td>24, F</td>
<td>lt hemi stroke</td>
<td>bilat</td>
<td>lt STA-MCA bypass, rt EDAS</td>
</tr>
<tr>
<td>12</td>
<td>26, M</td>
<td>lt hemi stroke/TIA</td>
<td>bilat</td>
<td>bilat EDAS</td>
</tr>
<tr>
<td>13</td>
<td>28, F</td>
<td>lt hemi stroke</td>
<td>bilat</td>
<td>bilat EDAS</td>
</tr>
<tr>
<td>14</td>
<td>28, M</td>
<td>rt hemi stroke/bilat TIAs</td>
<td>bilat</td>
<td>bilat EDAS</td>
</tr>
<tr>
<td>15</td>
<td>31, F</td>
<td>lt hemi TIA</td>
<td>unilat</td>
<td>rt EDAS</td>
</tr>
<tr>
<td>16</td>
<td>48, F</td>
<td>lt hemi TIA</td>
<td>bilat</td>
<td>bilat EDAS</td>
</tr>
</tbody>
</table>

* Angio = angiography; hemi = hemisphere.
Repeat revascularization surgery for moyamoya disease

**TABLE 2: Repeat surgery and outcomes in 16 patients with moyamoya disease**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>1st Op</th>
<th>Sx After 1st Op</th>
<th>Duration Btwn 2 Ops (mos)</th>
<th>Op in Previously Revascularized Hemi(s)</th>
<th>No. of Hemis Re-revascularized</th>
<th>Outcome at FU</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>bilat EDAS</td>
<td>bilat TIA</td>
<td>36</td>
<td>bilat STA-MCA bypass</td>
<td>2</td>
<td>occasional TIAs, flow normal, robust collaterals on angio</td>
</tr>
<tr>
<td>2</td>
<td>rt EMS</td>
<td>rt hemi TIA</td>
<td>10</td>
<td>rt STA-MCA bypass</td>
<td>1</td>
<td>no TIA/stroke</td>
</tr>
<tr>
<td>3</td>
<td>bilat EDAS</td>
<td>seizures, cognitive decline</td>
<td>18</td>
<td>lt STA-MCA bypass</td>
<td>1</td>
<td>no TIA/stroke</td>
</tr>
<tr>
<td>4</td>
<td>lt EDAS</td>
<td>lt hemi TIAIs</td>
<td>48</td>
<td>lt STA-MCA bypass</td>
<td>1</td>
<td>TIA; possible graft occlusion, repeat EMS performed elsewhere</td>
</tr>
<tr>
<td>5</td>
<td>rt EDAS w/ multiple bur holes</td>
<td>rt hemi ischemic stroke w/ sequelae</td>
<td>3</td>
<td>rt EMS</td>
<td>1</td>
<td>no TIA/stroke</td>
</tr>
<tr>
<td>6</td>
<td>bilat EDAS</td>
<td>bilat TIA, fresh infarct on MRI</td>
<td>36</td>
<td>rt STA-MCA bypass, lt omental transposition w/ STA anastomosis</td>
<td>2</td>
<td>no TIA/stroke</td>
</tr>
<tr>
<td>7</td>
<td>bilat EMS</td>
<td>lt hemi TIA, fresh infarct on MRI</td>
<td>60</td>
<td>rt OA-MCA bypass, lt omental transposition</td>
<td>2</td>
<td>no TIA/stroke</td>
</tr>
<tr>
<td>8</td>
<td>bilat EDAS</td>
<td>visual</td>
<td>30</td>
<td>lt OA-PCA bypass</td>
<td>1</td>
<td>occasional TIAs, flow normal, robust collaterals on angio</td>
</tr>
<tr>
<td>9</td>
<td>lt EDAS, rt STA-MCA bypass</td>
<td>lt hemi TIAIs</td>
<td>6</td>
<td>lt STA-MCA bypass</td>
<td>1</td>
<td>no TIA/stroke</td>
</tr>
<tr>
<td>10</td>
<td>bilat EDAS</td>
<td>lt hemi TIAIs</td>
<td>120</td>
<td>bilat EDAS, frontal branch</td>
<td>2</td>
<td>no TIA/stroke</td>
</tr>
<tr>
<td>11</td>
<td>lt STA-MCA bypass, rt EDAS</td>
<td>lt hemi TIAIs</td>
<td>6</td>
<td>rt EMS</td>
<td>1</td>
<td>no TIA/stroke</td>
</tr>
<tr>
<td>12</td>
<td>bilat EDAS</td>
<td>rt hemi ischemic stroke</td>
<td>84</td>
<td>rt STA-MCA bypass, lt omental transposition</td>
<td>2</td>
<td>no TIA/stroke</td>
</tr>
<tr>
<td>13</td>
<td>bilat EDAS</td>
<td>lt hemi TIAIs</td>
<td>96</td>
<td>bilat STA-MCA bypass</td>
<td>2</td>
<td>no TIA/stroke</td>
</tr>
<tr>
<td>14</td>
<td>bilat EDAS</td>
<td>asymptomatic infarct on MRI</td>
<td>14</td>
<td>rt EMS</td>
<td>1</td>
<td>no TIA/stroke</td>
</tr>
<tr>
<td>15</td>
<td>rt EDAS</td>
<td>rt hemi TIA</td>
<td>9</td>
<td>rt EMS</td>
<td>1</td>
<td>no TIA/stroke</td>
</tr>
<tr>
<td>16</td>
<td>bilat EDAS</td>
<td>bilat TIA, fresh infarct on MRI</td>
<td>6</td>
<td>bilat ECA-MCA bypass (w/ vein graft)</td>
<td>2</td>
<td>died</td>
</tr>
</tbody>
</table>

* FU = follow-up.

Follow-Up Findings

Clinical follow-up was available in all patients, with the range of follow-up duration being 3–144 months (34 ± 37.9 months [mean ± SD]; median 12 months). Twelve (80%) of the 15 patients who survived repeat revascularization surgery were well and free from any TIA, stroke, or new symptoms. Two patients had occasional TIAs, the frequency and severity of which were significantly less than prior to their repeat surgery. One patient did well for 1 year, when he developed frequent TIAs. He was evaluated at another institution, and his MR imaging studies showed no new ischemic lesions. However, the angiogram revealed nonvisualization of the graft/anastomosis. Hence, he underwent repeat EMS and multiple bur holes, after which he did well, with no further ischemic episodes.

Studies of CBF obtained at 6 months were available in 12 patients, and showed normalization of flow, with good augmentation following acetazolamide in all patients, except in the areas of prior infarct. An angiogram was available in 11 patients. There was improved flow in the hemispheres in 10 patients, and the graft was not visualized in 1 patient (only the report was available from the other institution). Figures 1–3 are representative MR imaging and angiography studies from the patients showing inadequate revascularization after the first procedure and robust revascularization following repeat surgery. The MR imaging studies obtained at 6 months revealed no new infarcts in any patients.

Discussion

Moyamoya disease is characterized by idiopathic, chronic occlusion of bilateral supraclinoid ICAs, proximal MCAs, and ACAs, along with the formation of moyamoya-like collateral vessels. Surgery for moyamoya disease typically involves revascularization procedures to improve cerebral perfusion and prevent or minimize stroke. Repeat revascularization surgery may be considered for patients who continue to experience neurological symptoms despite initial successful surgery. The outcomes in the presented cases demonstrate varying degrees of success, with some patients achieving complete resolution of symptoms and others experiencing recurrent TIAs or ischemic events. Further studies are needed to optimize surgical outcomes and management strategies for patients with moyamoya disease.
Fig. 1. Digital subtraction angiography studies (lateral projection) of the right ECA (A) and left ECA (B) showing inadequate revascularization following bilateral EDAS. Bilateral revascularization was performed with the frontal branch of the STA, leading to robust revascularization in the right ECA (C) and left ECA (D). The branch of the STA used for EDAS is marked with a black arrowhead, and the branch used for the STA-MCA bypass is marked with a black arrow.

Fig. 2. Left: A DS angiography study (lateral projection) of the left ECA injection showing inadequate revascularization from EDAS performed on the left side. Right: A DS angiography study (lateral projection) of the left ECA injection showing better filling of two-thirds of the hemisphere following addition of an STA-MCA bypass. The branch of the STA used for EDAS is marked with a black arrowhead, and the branch used for the STA-MCA bypass is marked with a black arrow.
moya vessels at the base of the brain, and transdural collateral vessels attempting to compensate for the ischemia resulting from the occlusion.\textsuperscript{3,37} The treatment of moyamoya disease is to augment the blood supply of the brain from the ECA system. This is achieved either through direct anastomosis to a cortical MCA branch (or in some cases, an ACA or PCA branch), or through stimulation of collateral formation through a variety of indirect meth-

**Fig. 3.** A: Axial T2-weighted MR imaging study of the brain showing left-sided watershed infarcts. B and C: Digital subtraction angiography studies, right ICA and left ICA injections, lateral projection, showing occlusion of bilateral ICAs with moyamoya vessels. D and E: Digital subtraction angiography studies, right ECA and left ECA injections, lateral projection, showing very small areas of revascularization following bilateral EDAS. F–I: Left ECA injection, anteroposterior (F) and lateral (G) projections, and right ECA injection, anteroposterior (H) and lateral (I) projections, showing excellent revascularization, predominantly from meningeal collateral vessels following bilateral STA-MCA anastomosis and overlaying of the artery on the brain.
odds. This is achieved by placing either an ECA branch (typically the STA) or muscle and dura mater over the ischemic brain to promote angiogenesis. The choice of procedure in both the pediatric and adult population is a matter of debate, with proponents of both procedures publishing their findings in the literature.

Direct revascularization for moyamoya disease is an established technique, and a number of authors including our group have described good results, both in adult and pediatric populations. The main advantages of the direct anastomosis are the establishment of augmented flow immediately following the surgery and more consistent formation of collateral vessels. The results of direct revascularization in adults have been well described by many authors. Long-term studies following delayed revascularization describe patency rates as high as 91%, with 91.8% of patients free of any ischemic event at 1-year follow-up. Other authors have published equally good results in smaller series. The results of direct revascularization in the pediatric population are published less often. However, small case series have described good results in preventing ischemic events in children. Some authors have combined STA-MCA with the indirect procedure in an attempt to improve long-term revascularization. The disadvantages of the direct procedures are as follows: the requirement of technical expertise for this procedure; the nonavailability of appropriate donor or recipient vessels, particularly in the pediatric population; and a higher incidence of complications compared with the indirect procedures. Some authors have noted the occurrence of a hyperperfusion syndrome and transient neurological deficits due to the sudden increase in the blood flow to an ischemic brain. On the other hand, indirect procedures have become very popular, especially in the pediatric population, because of the ease of the surgery, its effectiveness, and lower rates of complications. Although the efficacy of various indirect procedures has been well established in children, the efficacy in adults is still controversial. Although a recent publication suggests that indirect revascularization is also effective in reducing the risk of future ischemic events for adults with moyamoya disease, many authors have published superior results after performing direct procedures.

One of the disadvantages of the indirect procedures, especially the EDAS, is the inconsistent and sometimes incomplete development of adequate collateral vessels. In a study comparing various forms of revascularization, Matsushima et al. found that collateral formation in more than two-thirds of the hemisphere occurred in only 44% of the hemispheres after a single indirect procedure, compared with 52% after multiple combined indirect procedures and 74% after STA-MCA anastomosis. Other authors have also reported on failed EDAS procedures, in which the collateral formation is inadequate, the patients continue to be clinically symptomatic, or both. Various procedures have been described to augment the collateral formation with EDAS, but still there is a subset of patients who have inadequate collateral formation, and who remain clinically symptomatic. The treatment options for this subset of patients have been poorly described in the literature. Most of the reports of reconstruction after failed indirect anastomosis are from the Japanese literature. Ours is the first series from North America describing repeat revascularization of failed indirect anastomosis.

Miyamoto et al. reported a series of 11 patients with failed indirect anastomosis and documented good results, with newly formed collateral vessels on repeat angiography studies after additional STA-MCA bypass or omental transposition. Matsushima et al. described the addition of an EMS or EMAS procedure in 3 patients with failed EDAS, and obtained better collateral vessels. Touho et al. performed STA-MCA anastomosis in all patients, and additional omental transposition in 1 individual, leading to excellent recovery in all of them. The investigators used the same branch of the STA used in the EDAS procedures, and reported no new deficits. They thus concluded that the small amount of collaterals around the EDAS vessel was clinically insignificant.

On the other hand, in our series, we performed direct anastomosis for 14 (60.9%) of the 23 hemispheres revascularized. In the rest of the hemispheres, we could not find an adequate recipient vessel, or there was extensive adherence as a result of the first operation, and we performed indirect procedures. In most of the cases, we left the branch used for EDAS intact, to avoid disrupting the collaterals formed around it, and because of dense adhesions associated with EDAS. In only 2 cases did we use the same branch of the STA; in those cases we monitored any changes in the SSEP and EEG findings following clip occlusion of the STA, and only when there were no significant changes in the electrophysiological potentials did we sacrifice the minimal collateral vessels from the previous procedure. Other authors have also reported on small series of patients with failed indirect procedures, and the reconstruction with further addition of indirect or direct procedures. In the series by Touho et al., all patients were symptomatic, with recurrent ischemic events. In our patients, all except 3 had TIA’s; of those 3, 1 had recent infarcts, 1 had seizures and cognitive decline, and 1 had progression of posterior circulation ischemic symptoms.

In our opinion, the clinical symptoms, radiological and angiographic findings, and the results of CBF studies should be analyzed and correlated before subjecting the patient to a repeat procedure. In the present series, we obtained excellent results in most of the patients. However, there was 1 death secondary to reperfusion hemor-
Repeat revascularization surgery for moyamoya disease

rhage; this occurred in a patient who was having multiple frequent TIAs despite previous EDAS procedures, who had recent infarcts on repeat MR imaging, and received a high-flow vein graft. Since the experience of this adverse outcome after using a saphenous vein interposition graft from the ECA to the M2 artery, we have abandoned the use of higher-flow bypasses for patients with moyamoya disease.

Choice of Procedures

Various authors have described the use of both direct and indirect procedures for reconstruction of an inadequate indirect procedure. In our experience, judicious use of either procedure gives good results. As is our policy for initial revascularization in most patients with moyamoya disease, we prefer direct anastomosis with STA-MCA bypass to provide immediate and reliable augmentation of blood supply to the ischemic hemisphere. However, in cases in which the recipient vessels are small or there are multiple adhesions, we use indirect procedures, especially EMS, or occasionally omental transpositions, if adequate muscle is not available. The choice of the procedure is tailored to the operative findings and availability of donor and recipient vessels.

Conclusions

Repeat revascularization procedures are clinically effective in preventing future ischemic events, and can be safely performed for patients who are clinically symptomatic and have inadequate collateral vessels following indirect procedures, especially EDAS. Although direct procedures are preferred, the choice depends on the operative findings and the status of donor and recipient vessels.

Disclosure

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