Surgical Treatment of Childhood Moyamoya Disease

—Comparison of Reconstructive Surgery Centered on the Frontal Region and the Parietal Region—

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Abstract

Indirect revascularization procedures centered on the parietal region, such as encephalo-myo-arterio-synangiosis (parietal synangiosis) and direct procedures centered on the frontal region using both the anterior and the posterior branches of the superficial temporal artery (STA), such as STA to middle cerebral artery anastomosis combined with encephalo-duro-arterio-synangiosis (frontal anastomosis) were compared in childhood moyamoya disease patients. The parietal synangiosis group consisted of 10 sides in five patients, and the frontal anastomosis group consisted of 30 sides in 15 patients. The development of postoperative collateral circulation was assessed by external carotid angiography, the neurological outcome was monitored for 2 years after surgery, and the intelligence quotient (IQ) was measured at least 6 months after surgery. Frontal anastomosis achieved superior results compared to the parietal synangiosis assessed by development of collateral circulation, in particular to the orbitofrontal artery, the prefrontal artery, and the precentral artery (p < 0.01), and reduction in the incidence of ischemic attacks, such as transient ischemic attacks (p < 0.05). The mean IQ in the frontal anastomosis group was higher than that in the parietal synangiosis group. Vascular reconstruction centered on the frontal region utilizing both the anterior and posterior branches of the STA is more efficacious than only synangiosis centered on the parietal region.

Key words: moyamoya disease, pediatric surgery, superficial temporal artery-middle cerebral artery anastomosis, synangiosis, revascularization

Introduction

Childhood moyamoya disease is characterized by progressive arterial stenosis or occlusion of the major intracranial cerebral arteries and by development of extensive collateral circulation, including a fine network of vessels at the base of the brain referred to as the "moyamoya vessels."12,22] The major symptom is cerebral ischemia. The pathogenesis of this disease is unknown, so no basic treatment has been established. Therefore, the symptom of cerebral ischemia is treated by increasing the blood supply to the ischemic brain from the external carotid system using several types of vascular reconstructive surgery: superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis,6] encephalo-myo-synangiosis (EMS),7] encephalo-duro-arterio-synangiosis (EDAS),15,16] encephalo-myo-arterio-synangiosis (EMAS),18-20] encephalo-duro-arterio-myo-synangiosis (EDAMS),10] omentum implantation,8] and combinations of these procedures.

In childhood moyamoya disease cerebral circulatory insufficiency occurs in the frontal lobe. The regional cerebral blood flow (rCBF) of the frontal lobe is reduced23] and the frontal rCBF decreases as a result of hyperventilation,9] because the MCA and anterior cerebral artery are affected at practically the same time. Therefore, resolution of this frontal lobe ischemia is a very important target of surgical revascularization.4,24] However, most indirect
bypass procedures such as EDAS, EMS, and EMAS achieve vascular reconstruction centered on the parietal region. The anterior branch of the STA is not used because it travels through the forehead area. Only the posterior branch is used, and the craniotomy is centered on the parietal region. Follow-up angiography studies show that the collateral vascular channels linked to the external carotid system are localized in the operative field. The collateral circulation flows through the cortical branches of the MCA peripherally, but proximal flow is retrograde, because moyamoya disease is characterized by multiple stenotic and/or occlusive lesions of the cortical arteries. This contrasts with the findings in atherosclerotic steno-occlusive disease. Therefore, circulation to the central-parietal-temporal area improves angiographically, but collateral formation to the prefrontal area is inevitably inadequate. Another problem is that some cases are refractory to indirect procedures.

To solve these problems, we have created anastomoses between the anterior branch of the STA and cortical branch of the MCA in the frontal region since 1986. Here, we compare the postoperative angiographic and clinical findings of conventional synangiosis centered on the parietal region and anastomosis with synangiosis centered on the frontal region.

**Materials and Methods**

Twenty children with moyamoya disease were surgically treated in the Department of Neurosurgery, Hokkaido University Hospital. A total of 40 cerebral hemisphere operations were performed. From 1982 to 1985, only synangiosis was performed centered on the parietal region. Only the posterior branch of the STA was used, since the anterior branch often naturally forms a collateral channel. A horseshoe-shaped scalp incision was made around the posterior branch of the STA above the ear, and a wide craniotomy performed in the parietal region. The middle meningeal artery (MMA) was preserved as extensively as possible. The posterior branch of the STA was laid on the exposed cortex and then covered with temporal muscle. When the MMA was damaged during the craniotomy, EMAS was performed for indirect revascularization. When both the anterior and the posterior branches of the STA were anastomosed, encephalo-duro-myo-synangiosis (EDMS) or EMS was performed for indirect revascularization.

EDAMS was performed on two sides in one patient, EMAS on six sides in three patients, and EMS on two sides in one patient. STA-MCA anastomosis was performed on 25 sides in 15 patients with EDAMS, on three sides in three patients with EMAS, on one side in one patient with EDMS, and on one side in one patient with EMS.

The 40 sides were divided into two groups according to the surgical procedure; a “parietal synangiosis group” of 10 sides in five patients (EDAMS, EMAS, and EMS), and a “frontal anastomosis group” of 30 sides in 15 patients (STA-MCA anastomosis combined with either EDAMS, EMAS, EDMS, or EMS). Table 1 shows the age on admission, age at onset, type of moyamoya disease, and preoperative angiographic stage according to Suzuki and Takaku for each group. Differences in distribution

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were measured by the $\chi^2$ test, and differences between the mean values were assessed using a Student's $t$-test. There were no significant differences between the two groups.

Collateral formation was reviewed using external carotid angiograms taken 6–12 months after surgery. A transparent plastic template was superimposed on the lateral view of the postoperative external carotid angiograms. The degree of opacification in each cortical branch of the MCA was evaluated for each patient using a four-step scale according to the area of the MCA cortical branch distribution supplied: 3 points, when two-thirds or more of the area was covered; 2 points, when between two-thirds and one-third was covered; 1 point, when less than one-third was covered; and 0 points, when no perfusion was observed.

Complications occurring within 1 month of surgery were assessed on the basis of medical records.

Postoperative follow-up examinations were continued for 2 years after bypass surgery. The neurological outcome was evaluated according to the frequency of ischemic attacks during the year following surgery: excellent when cerebral ischemic attacks had stopped, and good when the frequency of cerebral ischemic attacks had diminished. The surgical effects were evaluated on the 40 operated sides based on the symptoms.

The intelligence quotients (IQs) of five patients in the parietal synangiosis group and 11 patients in the frontal anastomosis group were studied using Wechsler Intelligence Scale for Children-Revised at least 6 months after surgery, to investigate the relationship between the operative method and the IQ. The classification of the IQ was: normal (IQ ≥

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**Table 1 Clinical features of patients**

<table>
<thead>
<tr>
<th></th>
<th>Parietal synangiosis</th>
<th>Frontal anastomosis</th>
<th>Statistical significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age on admission* (yrs)</td>
<td>6.7 ± 1.8</td>
<td>8.2 ± 3.0</td>
<td>NS</td>
</tr>
<tr>
<td>Age at onset* (yrs)</td>
<td>4.7 ± 2.5</td>
<td>6.3 ± 2.6</td>
<td>NS</td>
</tr>
<tr>
<td>Type of moyamoya disease (sides)</td>
<td>NS</td>
<td></td>
<td></td>
</tr>
<tr>
<td>TIA</td>
<td>7</td>
<td>19</td>
<td></td>
</tr>
<tr>
<td>cerebral infarction</td>
<td>3</td>
<td>11</td>
<td></td>
</tr>
<tr>
<td>Angiographic stage**</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>1</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>III</td>
<td>8</td>
<td>26</td>
<td></td>
</tr>
<tr>
<td>IV</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>V</td>
<td>0</td>
<td>3</td>
<td></td>
</tr>
</tbody>
</table>

*Values are means ± SD. **According to Suzuki and Takaku. NS: not significant, TIA: transient ischemic attack.
80), borderline (IQ 70–79), and mental retardation (IQ ≤ 69). Since the IQs of patients who underwent surgery before 1987 and that of patients under 5 years old were studied using other tests, pre- and postoperative IQs could not be compared.

Results

I. Angiographic results

The parietal synangiosis group developed significantly less collateral formation to the orbitofrontal artery, the prefrontal artery, and the precentral artery (p < 0.01; Student’s t-test), which was predominantly from the central artery to cortical arteries of the MCA in the parietal and the temporal lobes (Fig. 2, Table 2). In contrast, the frontal anastomosis group had very good collateral formation to the orbitofrontal, prefrontal, and precentral arteries, and was better than in the parietal synangiosis group even in the parietal and temporal lobes (Fig. 3, Table 2). However, even though the cortex was included in the operative fields in both groups, there were also areas where neovascularization did not occur as a result of indirect revascularization (Figs. 2 and 3). Overall examination of the angiograms in both groups suggested that a wider craniotomy generally resulted in better neovascularization from the MMA and the deep temporal artery.

Table 2  Angiographic comparison of collateral formation

<table>
<thead>
<tr>
<th>MCA cortical branch</th>
<th>Parietal synangiosis</th>
<th>Frontal anastomosis</th>
<th>Statistical significance</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>0 1 2 3 Mean</td>
<td>0 1 2 3 Mean</td>
<td></td>
</tr>
<tr>
<td>OF</td>
<td>9 0 0 1 0.10</td>
<td>1 0 0 29 2.90</td>
<td>p &lt; 0.01</td>
</tr>
<tr>
<td>PF</td>
<td>5 0 2 3 1.40</td>
<td>0 2 0 28 2.87</td>
<td>p &lt; 0.01</td>
</tr>
<tr>
<td>PC</td>
<td>3 0 2 5 1.90</td>
<td>0 0 0 30 3.00</td>
<td>NS</td>
</tr>
<tr>
<td>C</td>
<td>1 0 0 9 2.70</td>
<td>1 1 1 27 2.80</td>
<td>NS</td>
</tr>
<tr>
<td>AP</td>
<td>2 0 0 8 2.40</td>
<td>5 1 1 23 2.40</td>
<td>NS</td>
</tr>
<tr>
<td>PP</td>
<td>2 1 2 5 2.00</td>
<td>4 2 3 21 2.37</td>
<td>NS</td>
</tr>
<tr>
<td>A</td>
<td>3 1 2 4 1.70</td>
<td>2 2 7 19 2.43</td>
<td>NS</td>
</tr>
<tr>
<td>TO</td>
<td>3 1 3 3 1.60</td>
<td>2 3 7 18 2.37</td>
<td>NS</td>
</tr>
<tr>
<td>PT</td>
<td>3 0 2 5 1.90</td>
<td>3 0 5 22 2.53</td>
<td>NS</td>
</tr>
<tr>
<td>MT</td>
<td>3 0 2 5 1.90</td>
<td>5 2 5 18 2.20</td>
<td>NS</td>
</tr>
<tr>
<td>AT</td>
<td>5 0 3 2 1.20</td>
<td>11 2 2 15 1.70</td>
<td>NS</td>
</tr>
<tr>
<td>TP</td>
<td>8 0 1 1 0.40</td>
<td>20 0 3 7 0.90</td>
<td>NS</td>
</tr>
</tbody>
</table>

Opacification of territories supplied by MCA branches was assessed on external carotid angiograms. Opacification score shows area of branch distribution visualized: 3, 2/3 or more; 2, 2/3–1/3; 1, less than 1/3; 0, no perfusion. A: angular artery, AP: anterior parietal artery, AT: anterior temporal artery, C: central artery, MT: middle temporal artery, NS: not significant, OF: orbitofrontal artery, PC: precentral artery, PF: prefrontal artery, PP: posterior parietal artery, PT: posterior temporal artery, TO: temporo-occipital artery, TP: temporo-polar artery.

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tal anastomosis group was lower than in the parietal synangiosis group, but there was no significant difference.

III. Clinical results

In the parietal synangiosis group, six sides (60%) had excellent outcome, while in the frontal anastomosis group, 28 sides (93%) were excellent. The postoperative neurological outcome in the frontal anastomosis group was significantly better than in the parietal synangiosis group (p < 0.05; \( \chi^2 \) test).

IV. IQ

In the parietal synangiosis group, two patients (40%) had a normal IQ, one patient (20%) was borderline, and two patients (40%) were mentally retarded. In the frontal anastomosis group, seven patients (64%) had a normal IQ, two patients (18%) were borderline, and two patients were mentally retarded. The parietal synangiosis group had a mean IQ of 77.4 ± 19.8, while the frontal anastomosis group had a mean IQ of 87.1 ± 20.1 (Fig. 4). There was no significant difference.

Discussion

The present study has shown that development of collateral circulation, especially in the prefrontal area, is better in patients treated by frontal anastomosis than by parietal synangiosis. Subsequent intellectual development apparently reflects this superior collateral circulation.

Three-dimensional hemodynamic changes after vascular reconstruction in childhood moyamoya disease have been investigated using xenon-133 (\(^{133}\text{Xe}\))-inhalation single photon emission computed tomography (SPECT). Inadequate rCBF was observed in the frontal lobe after parietal synangiosis, but the low-perfusion area disappeared after frontal anastomosis. Isobe et al. also examined rCBF after hyperventilation using \(^{133}\text{Xe}\)-inhalation SPECT, and reported that rCBF in the frontal lobe after hyperventilation in patients treated by parietal synangiosis was markedly reduced compared with rCBF in other regions and rCBF in patients treated by frontal anastomosis. These findings show that frontal lobes which are not revascularized remain in a state of circulatory insufficiency. Therefore, the revascularization procedure, i.e. craniotomy, should be extended to the frontal region. In addition, since the STA is the best blood supply source in the external carotid system, the anterior branch of the STA should also be utilized whenever possible.

Another question is whether STA-MCA anastomosis should be performed. Miyamoto et al. suggested that non-anastomotic bypass procedures cannot provide immediate revascularization, and are not suitable for patients with frequent transient ischemic attacks. In addition, the occurrence of moyamoya disease refractory to indirect procedures has been emphasized. In our study, immediate postoperative aggravation was less in the frontal anastomosis group in which STA-MCA anastomosis was performed.

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was performed, and we have also experienced a case refractory to the indirect procedure which needed another anastomosis procedure.^{4)}

Matsushima et al.^{14)} compared surgical results after indirect and direct revascularization procedures, and found that STA-MCA anastomosis with EMS in the parietal region was superior to EDAS in the parietal region in both development of collateral circulation and postoperative clinical improvement, as in the present study. They concluded that direct anastomosis should be the treatment of first choice. We would like to emphasize that the MCA cortical branch of the anastomosis should be selected on the basis of the preoperative CBF study. However, Karasawa et al.,^{9)} based on follow-up carotid angiography, found that the STA-MCA anastomosis became increasingly obstructed even though the cortical branches of the MCA were opacified through the fine network produced in the region of the anastomosis. Therefore, both the anterior and posterior branches of the STA should be utilized as synangioses to extend the potential area for neovascularization as widely as possible over the ischemic brain surface. If possible, the side branches of the STA should be anastomosed to frontal cortical branches of the MCA and other cortical branches in the low-perfusion area. When the lateral branch of the STA cannot be used, the anterior or the posterior branch of the STA should be cut peripherally, and after drawing over the exposed cortex, the cut end of the STA should be anastomosed to the MCA. In addition, all vessels that might serve as future collateral channels, including the MMA and the deep temporal artery, should be utilized.

Some authors have claimed that STA-MCA anastomosis procedures have some considerable disadvantages: the anastomosis is difficult to perform, and some patients deteriorate after anastomosis because of the temporary stasis of blood needed in the cortical artery for direct anastomosis.^{10,20,26)} An STA-MCA anastomosis is difficult to perform in patients with childhood moyamoya disease because the arteries anastomosed are very small, with an outer diameter ranging from 0.5 to 1.0 mm,^{6)} and are thin and fragile like veins.^{14)} In addition, the ostia of arteriotomies become invisible because of their transparency after the blood in the lumen flows out. Staining of the ostium of an arteriotomy with a pyoktanine blue solution can provide visualization, allowing rapid identification of the ostium and performance of the correct anastomosis. In addition, we select the lateral branch of the anterior or the posterior branch of the STA as the donor arterial vessel instead of the anterior or the posterior branch itself, because the lateral branch has a suitable caliber matching the small cortical arteries.

We consider that the combination of a large craniotomy centered on the frontal lobe, utilizing all vessels capable of providing future collateral circulation, and creating STA-MCA anastomoses in the low-perfusion area whenever possible is the most effective treatment for the childhood moyamoya disease. However, even this procedure cannot prevent ischemia in the posterior or anterior cerebral artery territories in some patients, so further studies must investigate the need for more extensive vascular reconstruction, such as omentum implantation.^{9)}

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References

8) Karasawa J, Kikuchi H, Kawamura J, Sakaki T: In-


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