The Importance of Encephalo-Myo-Synangiosis in Surgical Revascularization Strategies for Moyamoya Disease in Children and Adults

Hideaki Imai[1], Satoru Miyawaki[1], Hideaki Ono[1], Hirofumi Nakatomi[1], Yuhei Yoshimoto[2], Nobuhito Saito[1]

■ OBJECTIVE: **The optimal surgical procedure (direct, indirect, or combined anastomosis) for management of moyamoya disease is still debated. We evaluated the outcome of our broad area revascularization protocol, the Tokyo Daigaku (The University of Tokyo) (TODAI) protocol, analyzing the relative importance of direct, indirect, and combination revascularization strategies to identify the optimal surgical protocol.**

■ METHODS: **The TODAI protocol was used to treat 65 patients with moyamoya disease (91 hemispheres, including 48 in 29 childhood cases collected during 1996–2012). The TODAI protocol combined direct superficial temporal artery (STA)–middle cerebral artery (MCA) anastomosis with indirect revascularization using encephalo-myo-synangiosis (EMS) for patients ≥10 years old or indirect revascularization using encephalo-duro-arterio-synangiosis (EDAS) with EMS for patients ≤9 years old. Clinical outcome was evaluated retrospectively. Digital subtraction angiography was performed for postoperative evaluation of revascularization in 47 patients (62 hemispheres; 27 adults and 35 children). Based on the relative contribution of additional flow from each revascularization path, 4 revascularization patterns were established.**

■ RESULTS: **The mean follow-up period was 90 months in children and 72 months in adults. Perioperative complications were seen in 4 of 48 operations in children and 1 of 43 operations in adults. Except for 1 child with recurrent transient ischemic attacks and 1 adult with intracerebral hemorrhage, the patients showed excellent clinical outcomes. Postoperative digital subtraction angiography evaluation showed that in STA-MCA anastomosis + EMS cases (34 hemispheres; 25 adults and 9 children), STA-MCA anastomosis provided greater revascularization than EMS (STA-MCA anastomosis > EMS) in 7 hemispheres, the opposite was true (STA-MCA anastomosis < EMS) in 14 hemispheres, an equivalent contribution to revascularization (STA-MCA anastomosis ≈ EMS) was present in 12 hemispheres, and no functioning anastomosis was present in 1 hemisphere. In cases of EDAS + EMS (28 hemispheres; 2 adults and 26 children), all hemispheres showed revascularization: EDAS was dominant to EMS (EDAS > EMS) in 1 hemisphere, the opposite (EMS > EDAS) was true in 14 hemispheres, and EDAS was equivalent to EMS (EDAS ≈ EMS) in 13 hemispheres. EMS plus direct or indirect anastomosis is an effective surgical procedure in adults and children.**

■ CONCLUSIONS: **The TODAI protocol provided efficient revascularization and yielded excellent results in preventing strokes in patients with moyamoya disease with very few complications. EMS had a main role in revascularization in each of the combined techniques.**

INTRODUCTION

Moyamoya disease was first described in the Japanese medical literature in 1957 by Takeuchi and Shimizu (34) as a rare, progressive occlusive cerebrovascular disease of the intracranial internal carotid arteries or their proximal

Key words
- Direct anastomosis
- Encephalo-myo-synangiosis
- Indirect anastomosis
- Moyamoya disease
- Revascularization

Abbreviations and Acronyms
DSA: Digital subtraction angiography
DTA: Deep temporal artery
EDAS: Encephalo-duro-arterio-synangiosis
EMS: Encephalo-myo-synangiosis
MCA: Middle cerebral artery
MMA: Middle meningeal artery
STA: Superficial temporal artery
TODAI: Tokyo Daigaku (The University of Tokyo)

From the [1]Department of Neurosurgery, Faculty of Medicine, The University of Tokyo, Tokyo; and [2]Department of Neurosurgery, Gunma University Graduate School of Medicine, Maebashi, Gunma, Japan

To whom correspondence should be addressed: Nobuhito Saito, M.D., Ph.D.
[E-mail: nsaito-tky@umin.net]

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branches with compensatory development of a fine collateral network at the base of the brain (moyamoya vessels) (33). Moyamoya disease has been regarded as a unique disease with 2 incidence peaks in age distribution: the first in children, at approximately 5 years of age, manifesting mainly as ischemic events, and the second in adults in their mid-40s, manifesting as hemorrhagic events (2, 9, 35). Moyamoya disease was found more recently to have a strong background relationship with a genetic mutation of *RNF213*, and patients with moyamoya disease in Japan share the same variant of *RNF213* in >80% of cases (17, 21, 25-27).

In general, medical treatment has not been effective in preventing clinical events (37). Although a radical surgical treatment to restore the blood flow of the main cerebral arteries in patients with moyamoya disease has not yet been found (3), revascularization surgery is usually performed to compensate for the cerebral blood supply to prevent ischemic or hemorrhagic strokes (6, 8, 10, 16, 24, 30, 31). Vascular anastomoses are created between the extracranial and cerebral arterial systems to supply additional blood flow to hypoperfused areas. There are several surgical procedures for the treatment of moyamoya disease, including direct anastomosis such as superficial temporal artery (STA)—middle cerebral artery (MCA) anastomosis (18), indirect anastomosis such as encephalo-duro-arterio-synangiosis (EDAS) (23) and encephalo-myo-synangiosis (EMS) (19), and combinations of direct and indirect anastomosis (13). The optimal revascularization strategy is still debated (1) and may differ in children and adults.

We proposed and for the last 10 years have applied a surgical procedure of combined EMS with STA-MCA anastomosis for adults with moyamoya disease and a procedure of EMS with EDAS for children with moyamoya disease to accomplish extensive revascularization of the frontal lobe. We evaluated the overall clinical outcome of surgically treated patients with moyamoya disease and examined the contribution of direct and indirect anastomosis in the revascularization pattern after surgical treatment to optimize the choice of surgical procedure for moyamoya disease.

MATERIALS AND METHODS

Patients

Between May 1996 and May 2012, 65 patients (48 hemispheres in 29 childhood cases and 43 hemispheres in 36 adult cases) underwent surgical revascularization for moyamoya disease at Gunma University Hospital and the University of Tokyo Hospital (Table 1). Ischemic events attributed to the hemodynamic stress in the frontal lobe occurred in 26 of 29 children and 33 of 36 adults. All patients developed moyamoya vasculature consistent with Suzuki stage II or III (33) associated with occlusion or stenosis at the terminal portion of the internal carotid artery. The institutional review board of the University of Tokyo Hospital approved the study protocol, and written informed consent was obtained in all patients before participating in this study.

Surgical Strategy and Procedure

Patients with symptomatic moyamoya disease were treated by the Tokyo Daigaku (The University of Tokyo) (TODAI) protocol, which provides extensive revascularization to the frontal lobe (15).

Table 1. Clinical Characteristics of 65 Patients with Moyamoya Disease Under the Tokyo Daigaku (The University of Tokyo) (TODAI) Protocol

	Children (<18 years old at surgery)	Adults (≥18 years old at surgery)
Number of cases (hemispheres)	29 (48)	36 (43)
Quasi-moyamoya disease (*n*)	1	4
Female (*n*)	15 (51.7%)	22 (61.1%)
Age at surgery (years), mean ± SD (range)	8.0 ± 3.4 (2—14)	43.0 ± 15.1 (20—76)
Clinical symptoms (*n*)		
TIA, ischemic infarction	26 (89.6%)	33 (91.6%)
Hemorrhage	1 (3.4%)	2 (5.5%)
Headache	2 (6.8%)	1 (2.7%)

TIA, transient ischemic attack.

Combined direct (STA-MCA anastomosis) and indirect anastomosis (EMS) was performed on patients ≥10 years old, and indirect surgery (EDAS + EMS) was performed on patients ≤9 years old.

Surgical Procedure

Skin Incision and Dissection of STA. The patient's head was fixed by the 3-point Mayfield fixation device under general anesthesia with special attention to maintain normocapnia, normothermia, and normal arterial blood pressure. Before the skin incision was made, the course of the parietal branch of the STA was identified using a Doppler ultrasound probe and marked on the skin. The hair was linearly shaved along the STA parietal branch course. The skin incision was made along this course from the zygoma extending 8—10 cm toward the distal parietal branch segment (Figure 1A). The parietal branch of the STA was carefully dissected from the surrounding tissue avoiding injury to the artery. At this stage, the distal end of the STA was still not divided. The skin incision was directed toward the midline to extend to the hairline. Consequently, the skin incision became L-shaped. The skin flap was lifted dissecting between the galea aponeurotica and temporal fascia.

Temporal Muscle Separation. The temporal muscle covered by its fascia was incised along the parietal branch (Figure 1B) and dissected from the skull. During the procedure, special attention was paid to keeping the deep temporal arteries (DTAs) intact because they are the main source for revascularization in EMS. An osteoplastic craniotomy was done with 6 burr holes avoiding injury to the middle meningeal artery (MMA) (Figure 1C). The extent of the dural opening and the area of brain exposure depended on the available surface of the separated temporal muscle because this dural opening should be covered by the temporal muscle.

Surgical Techniques for Indirect Revascularization: EDAS. The EDAS procedure was performed as previously described (13) with some