

Long-term results of surgically treated Moyamoya disease

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Abstract

Various surgical procedures have been tried for patients with Moyamoya disease. The most effective treatment, however, is still controversial. We retrospectively evaluated the long-term results of 71 patients (26 men and 45 women) with Moyamoya disease surgically treated in our institute. They consisted of 56 pediatric patients (younger than 15 years) and 15 adult patients. Symptoms in all patients were due to cerebral ischemia. We did 123 operations on 119 hemispheres: 18 superficial temporal artery–middle cerebral artery (STA–MCA) anastomoses, six STA–MCA anastomoses with indirect bypass (IB), 41 encephalo-duro-arterio-synangiosis (EDAS), 29 encephalo-duro-arterio-myo-synangiosis (EDAMS) and 29 ribbon EDAMS. Average follow-up periods for each procedure were: 7 years for STA–MCA anastomosis, 6.2 years for STA–MCA anastomosis with indirect bypass, 11 years for EDAS, 5.6 years for EDAMS and 2.6 years for ribbon EDAMS, respectively. The results of each procedure were satisfactory because the preoperative transient ischemic attacks disappeared. Analysis of follow-up angiograms shows excellent filling of the ACA and MCA territory in the patients undergoing ribbon EDAMS. However, long-term follow-up study shows that about 10% of the patients had severe difficulty in social or school life because of intellectual impairment. © 1997 Elsevier Science B.V.

Keywords: Moyamoya disease; Surgical treatment; Long-term follow-up; Clinical result; Angiography; Complication

1. Introduction

Surgical treatment for patients with Moyamoya disease has become popular, although some controversy still surrounds the surgical indications and operative procedure. The benefit of surgery for the ischemic type of Moyamoya disease has been established. In contrast, it is still unclear whether a bypass operation can prevent future bleeding. Various surgical procedures have been used, superficial temporal artery–middle cerebral artery (STA–MCA) anastomosis [1], encephalo-duro-arterio-synangiosis (EDAS) [2], encephalo-duro-arterio-myo-synangiosis (EDAMS) [3], and omental transplantation [4]. These procedures can be divided into two groups, direct anastomosis and indirect anastomosis. However, it is still controversial which is most effective. We followed 71 surgically treated patients who had an ischemic onset and retrospectively analyzed

their clinical results, postoperative collateral flow on angiograms, and cerebral blood flow (CBF).

2. Materials and methods

We have treated 144 patients with Moyamoya disease medically or surgically since 1967. Of these, 71 patients surgically treated between 1976 and 1996 were enrolled in this study. Their symptoms were due to cerebral ischemia. They included 45 females and 26 males, consisting of 56 pediatric patients (younger than 15 years) and 15 adults. Fourteen patients were younger than 5 years and 30 patients were from 5 to 10 years old. As for the clinical type, 43 patients had transient ischemic attacks (TIA), 24 had infarction and three patients had the epileptic type. In one patient, the major symptom was severe headache. In this study, we operated on 119 hemispheres with 123 procedures: 18 STA–MCA anastomoses, six STA–MCA anastomoses with indirect by-

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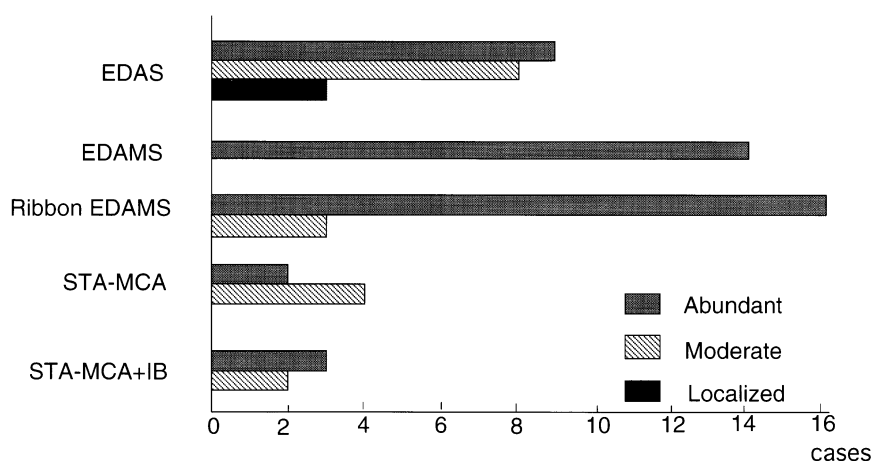


Fig. 1. Collateral formation from the ECA in the MCA territory on the postoperative follow-up angiogram. Abundant, more than two-thirds of cortical branches of the MCA were filled by the external carotid artery system. Moderate, two-thirds to one-third of cortical branches of the MCA were filled. Localized, slight visualization of a few cortical branches through the bypass.

pass (IB), 41 EDAS, 29 EDAMS and 29 ribbon EDAMS [5]. Average follow-up periods for each procedure were: 7 years for STA–MCA anastomosis, 6.2 years for STA–MCA anastomosis with indirect bypass, 11 years for EDAS, 5.6 years for EDAMS and 2.6 years for ribbon EDAMS, respectively. Postoperative clinical results were reviewed retrospectively in the hospital record. Follow-up angiograms were done principally 6 to 12 months after the surgery. Postoperative cerebral blood flow (CBF) studies were examined periodically in general. The patient's status long after surgical treatment was obtained either through interview or questionnaire.

3. Indications for surgical treatment

Patients with ischemic symptoms and misery perfusion on the CBF study are good candidates for surgical treatment. However, it is unknown whether bypass surgery prevents intracranial bleeding. The indications for surgical treatment for patients with Moyamoya disease in our institute are as follows: (1) symptomatic patients, such as those with TIA, ischemic stroke or epilepsy; (2) patients with decreased CBF and hemodynamic reserve on a CBF study; and (3) patients with cerebral aneurysm. We have not operated on the asymptomatic hemisphere with a normal CBF pattern. Patients operated on because of cerebral aneurysm were excluded from this study.

4. Surgical treatment

In the 1970s, we initially used STA–MCA anastomosis. Thereafter, an indirect bypass was done for pediatric patients, and a direct bypass was done for adults.

Among the various indirect bypass procedures, EDAS was mainly used in early 1980s. Since 1984, we have used EDAMS, and recently have begun to use the ribbon EDAMS procedure. For direct bypass, we prefer an STA–MCA anastomosis combined with indirect bypass. Our present choice of surgical procedure is as follows. For children, we recommend the ribbon EDAMS. For patients with definite bilateral involvement seen on the CBF study, we usually operate on both hemispheres at one session. In children and adults, however, when we can find a good recipient artery on the brain surface and the STA has no collaterals, we usually choose STA–MCA anastomosis with indirect bypass.

5. Operative techniques

In general, the craniotomy site is determined according to the preoperative CBF pattern. A CBF pattern indicating misery perfusion is a prerequisite for excellent revascularization in indirect bypass surgery. In EDAMS [3], the STA is followed with Doppler, and a question-mark incision is made to avoid injuring the STA. The frontal and parietal branches of the STA are carefully dissected and protected. A large craniotomy is done, and the meningeal arteries are preserved. Special care is necessary near the pterion to prevent injury of the middle meningeal artery. The dura is cut in several sleeves and reflected onto the surface of the brain. The arachnoid membrane is opened wide. This procedure effectively places the arterial surface of the outer layer of the dura tightly against the brain. The temporalis muscle and the stripped branches of the STA are laid on the brain surface and sutured to the dura. The drainage tube is usually placed beneath the temporalis muscle for 12 h. The bone flap is replaced to prevent

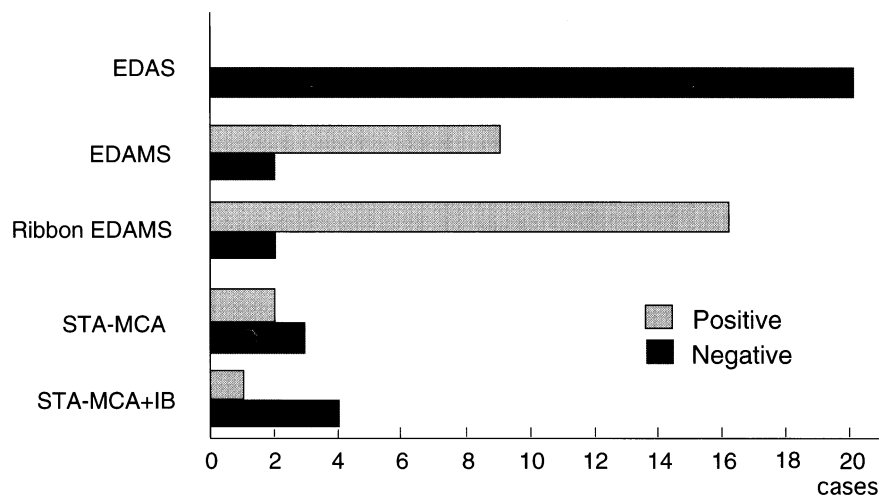


Fig. 2. Collateral formation from the ECA on the ACA territory in the postoperative follow-up angiogram. Positive, visible collateral flow in the midline ACA territory from the ECA on postoperative angiograms. Negative, no visible collateral flow in the midline ACA territory from the ECA.

compression of the STA. We recently combined the ribbon procedure with EDAMS, which we call ribbon EDAMS [5], to develop collaterals from the frontal branch of the STA to the midline territory of the anterior cerebral artery (ACA). A linear incision is made 2 cm anterior and parallel to the coronal suture and the galea containing the pericranium is dissected and incised in a zigzag fashion. A small craniotomy is made bilaterally along the superior sagittal sinus. A small portion of the dura is incised, and the arachnoid membrane is also incised. The apex of the cut galeal flap is inserted into the interhemispheric fissure as deeply as possible, and sutured to the falx.

6. Results

6.1. Follow-up angiographic examination

The development of collateral circulation of the MCA territory through the bypass was evaluated as follows: (1) abundant: more than two-thirds of the cortical branches of the MCA were filled by the external carotid artery system; (2) moderate: two-thirds to one-third of the cortical branches were filled; (3) localized: slight visualization of a few cortical branches through the bypass. Evaluation of the collateral flow in the MCA territory appears in Fig. 1. Most of the patients undergoing EDAMS or ribbon EDAMS showed abundant revascularization of the MCA territory. After EDAS, the number of patients showing abundant collateral and those showing moderate collateral were almost equal, and the collateral formation in 15% of these patients were considered localized. The collaterals from direct anastomosis seemed inferior to those from a combined indirect anastomosis such as

EDAMS. The poor patency of direct bypass in the early series from the 1970s unfavorably affected this angiographical evaluation. Collateral flow of the ACA territory through the bypass was divided into two groups: positive and negative. Patients in the positive category had visible collateral flow in the midline ACA territory from the ECA on postoperative angiograms. Patients in the negative category had no visible collateral flow in the midline ACA territory. These results are shown in Fig. 2. In the patients undergoing ribbon EDAMS, almost all showed the filling of the ACA territory, in striking contrast to the patients undergoing EDAS.

6.2. CBF study

Regional CBF was measured with the ^{133}Xe inhalation method, iodine – 123 -IMP single photon emission computed tomography (SPECT), HM-PAO SPECT and, recently, stable Xe CT. Hemodynamic reserve was evaluated through the acetazolamide challenge test. Postoperative CBF studies showed that regional CBF of the MCA territory increased and the response to acetazolamide challenge improved in the patients with abundant collateral formation. The postoperative blood flow pattern of the ACA territory seemed to improve in patients undergoing ribbon EDAMS.

6.3. Clinical results

The postoperative neurologic status of patients was divided into four categories: excellent, good, unchanged and worse. The excellent category included patients whose preoperative symptoms, such as TIAs or seizures, disappeared. The good category included patients whose preoperative symptoms diminished sub-

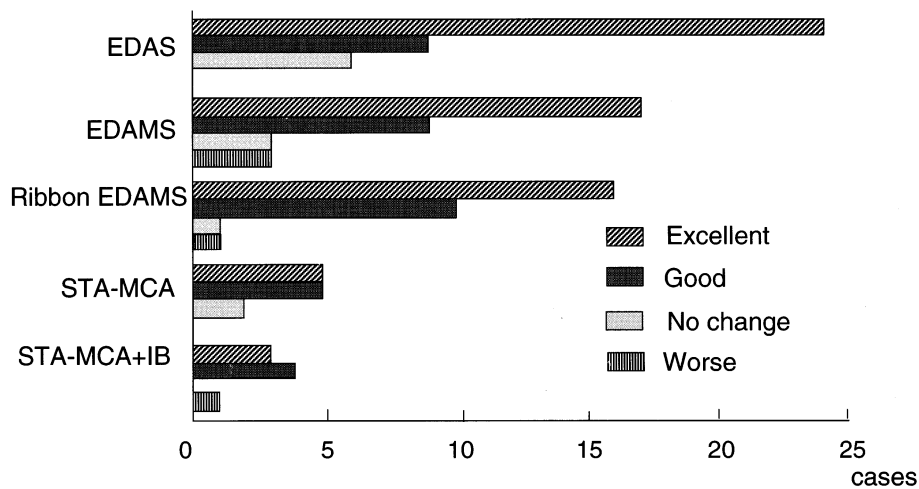


Fig. 3. Result of each surgical procedure on preoperative ischemic symptoms such as TIAs or seizure.

stantially, but still remained. The results of each procedure are shown in Fig. 3. In general, all procedures had satisfactory results and most of the preoperative ischemic symptoms disappeared within 1 year in this series. A few patients, however, were in the unchanged or worse category.

Five patients showed new neurologic deficits in the perioperative period. Four of these five patients showed motor weakness and aphasia from cerebral infarction. These signs were elicited through intraoperative hyperventilation or during episodes of crying postoperatively. One of these patients suffered an asthma attack and another had severe anemia postoperatively. One patient had a postoperative subdural hematoma; his symptoms resolved after the hematoma was removed.

Four patients required additional bypass operations in the ipsilateral hemisphere because of recurrent TIAs. In two, the first operation was EDAS and the recurrent TIAs were from decreased blood flow in the ACA territory. TIAs disappeared after an additional ribbon EDAMS procedures. In the other two patients, the initial operation was EDAMS. The symptoms in these patients improved after encephalo-galeo-synangiosis at the parieto-occipital region.

The latest functional status of the patients after long-term follow up were categorized as follows: (1) normal; can carry out the activities of daily living in good health; (2) moderately disabled; are independent but with minor deficits; and (3) severely disabled; are dependent, or have severe difficulties in school or social performance. The results of each procedure after the follow-up period are shown in Fig. 4. More than 70% of the patients undergoing EDAMS or ribbon EDAMS and 54% of the patients undergoing EDAS were in the normal category. A substantial number of patients, however, were in the moderately or severely disabled category. The primary factor responsible for the severe disability was mental retardation. Although some pa-

tients had various degrees of motor weakness or other focal deficits, except for severe visual impairment, these deficits were not a severe problem in patients who had normal intelligence. The main factor responsible for severe disability was multiple cerebral infarcts which had completed preoperatively. Additional factors appeared to be early onset in a young child and perioperative complications. A 24-year old female patient died suddenly probably because of a cerebrovascular attack 5 years after she underwent STA–MCA anastomosis.

7. Discussion

It is generally accepted that patients with ischemic symptoms from Moyamoya disease are good candidates for surgical treatment, and a variety of surgical procedures have been used to develop collateral circulation from the ECA [1–5]. The best procedure to use, however, remains controversial. Some authors conclude that direct bypass surgery is more effective than indirect bypass procedures, mainly EDAS [6]. We evaluated the clinical results of STA–MCA anastomosis, STA–MCA anastomosis with indirect bypass, EDAS, EDAMS and ribbon EDAMS performed in our institute. This study is retrospective and is not randomized, and there is a bias of selection of surgical procedure. Our choice of surgical procedure is generally indirect bypass for pediatric patients and direct bypass for adult patients. This choice is supported by data showing that advancing age apparently affects the development of collateral formation through the indirect bypass [7]. Our data demonstrate that indirect bypass procedures have satisfactory results in preventing further ischemic symptoms, at least in young patients. Because of the poor patency of direct bypass in the early series from the 1970s and the small number of patients with direct bypass included in this study, our evaluation of direct bypass is not con-

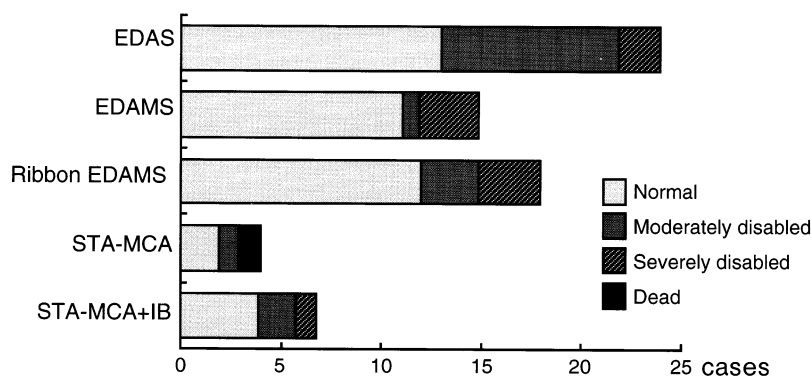


Fig. 4. Functional outcome after long-term follow-up for each surgical procedure.

clusive. Angiographic follow-up studies show that the combined indirect bypass procedure, i.e. ribbon EDAMS, develops more prominent collateral circulation, especially in the ACA territory, than the simple indirect bypass procedure, i.e. EDAS. A large craniotomy tailored according to the preoperative CBF pattern and the active use of all tissues capable of providing future collaterals seem to be key points of successful collateral formation in the indirect bypass technique. We open the arachnoid membrane widely to reduce obstacles for revascularization, although this technique is still controversial.

Because of the special vasoreactivities peculiar to Moyamoya disease, surgical intervention for this disease has a higher risk of perioperative ischemic events than it does for other cerebrovascular occlusive disease. This study shows that the association of perioperative complications clearly influences the clinical outcome. To avoid ischemic complications, the patient must be kept normocapneic for several weeks after surgery. Preventing children from crying, however, is not always easy. The administration of analgesics and sedatives may be necessary in this period. Greater attention is necessary for patients with frequent TIAs or a history of asthma. Other important points are to maintain normotension and avoid postoperative hypovolemia and anemia. Blood transfusion is sometimes necessary. In patients with definite bilateral involvement, we have recently begun to operate on both hemispheres during one session to reduce the risk.

Another important problem relating to the operative technique is the revascularization of the ACA and the posterior cerebral artery (PCA) territory. Recurrent TIAs of the lower limbs and visual symptoms several years after the initial bypass surgery resulting from impaired circulation of the territory of the ACA and PCA have been reported [4]. There are some reports of additional operation with omental transplantation [4] or other methods [8,9] for patients whose symptoms are refractory to the initial operation. The ribbon procedure ameliorates the circulation of the ACA territory.

The problem of revascularizing the PCA territory has not been solved, although the number of patients complaining of visual symptoms is relatively small. Based on the results of the CBF study, we add the encephalo-galeo-synangiosis procedure for such patients.

The beneficial effects of surgery for patients with preoperative TIAs are established. However, there are many questions about the long course of this disease. The long-term outcome of surgically treated patients in this series shows that approximately 70% treated with the combined indirect bypass procedure live normal daily lives. Focal deficits, such as motor weakness, usually improve with the rehabilitation. However, many patients have difficulty in social or school life because of mental retardation. The primary factor for intellectual impairment seems to be a major stroke completed preoperatively, especially in patients with early onset. Early diagnosis and surgical treatment are essential in preventing mental retardation in the young patient, although it is not easy to find symptoms or signs in an infant. Another important factor is perioperative complications.

8. Conclusion

(1) EDAMS combined with the ribbon procedure appears to be the most effective surgical procedure, for preventing ischemic symptoms in the MCA and ACA territories in children with Moyamoya disease. (2) Preventing perioperative complications is mandatory. (3) Mental retardation is the main factor influencing disability in long-term follow-up in young children.

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