

Surgical Treatment for Paediatric Patients with Moyamoya Disease by Indirect Revascularization Procedures (EDAS, EMS, EMAS)

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Summary

Surgical results of paediatric patients with Moyamoya disease who were treated by indirect revascularization procedures are reported. Encephalo-duro-arterio-synangiosis (EDAS), encephalo-myo-arterio-synangiosis (EMAS), and/or encephalo-myo-synangiosis (EMS) were performed on 47 sides of 29 children with Moyamoya disease. The results of those non-anastomotic EC-IC bypass procedures were evaluated clinically, angiographically, and by computed tomography (CT). Postoperative external carotid angiograms showed a good collateral formation through EDAS, EMAS, or EMS in about 70–80 percent of all surgically treated sides. The symptoms such as TIA, RIND, and/or involuntary movements disappeared in the cases with a good collateral formation but not in those with insufficient development of the collateral circulation. The indirect EC-IC bypass surgery seems effective for most of the children with Moyamoya disease who present with ischaemic signs.

Keywords: Moyamoya disease; paediatric case; surgical treatment; indirect non-anastomotic EC-IC bypass.

Introduction

The surgical treatment for patients with Moyamoya disease has become popular^{4–9, 16, 19, 21, 22}. It aims at establishing an adequate collateral circulation for the ischaemic brain. On the other hand, a few reports²³ told that the disease could be treated conservatively and the prognosis of the TIA type of the disease was good. Therefore, it is still worthwhile to clarify its natural course and to compare the surgical and nonsurgical results. There are also a few controversies concerning the surgical procedures among neurosurgeons. Different results were obtained by different procedures^{4, 18, 19}.

We operated on paediatric patients with Moyamoya disease employing mainly EDAS, EMAS and/or EMS. The surgical results in those cases are presented in

comparison with the results of conservative treatment which were already reported from the Department of Paediatrics in our hospital^{11, 12}. Advantages and disadvantages of the indirect non-anastomotic EC-IC bypass procedures such as EDAS, EMS, or EMAS are also discussed.

Patients and Methods

All of the 33 Japanese children were diagnosed and treated at the Department of Neurosurgery, Kyushu University Hospital. Two cases of unilateral type (probable case) of Moyamoya disease were also included. The initial symptoms were transient ischaemic attack (TIA) in 24 cases, reversible ischaemic neurologic deficit (RIND) in three, infarction in three, involuntary movement in two, and epilepsy in one. Although no patients developed bleeding as an initial symptom, one patient with the onset at the age of 4 years had a bleeding later at the age of 30 years.

Twenty-nine of the 33 patients received surgical treatment on 47 sides (Table 1).

The EDAS alone following Y. Matsushima *et al.*^{14, 15} was done in the parietal region on 19 sides. In the EDAS, a linear skin incision is done over the posterior branch of the superficial temporal artery (STA). The arterial branch is freed from the periosteum and fascia without dividing distally, and a strip of the galea including the freed arterial branch is made. After parieto-temporal craniotomy and a linear dural incision, the edges of the strip of the galea are sutured to the incisional edges of the dura. Subsequently the mobilized posterior branch comes to run on the cortical surface and under the bone flap.

The EDAS in the parietal region and the EMAS or EMS in the frontal region were performed at the same time on 17 sides. In the EMAS, the anterior branch of the STA, the frontal muscle, and the galea are used. The anterior branch is exposed and divided distally. After a horseshoe-shaped skin incision and craniotomy the dura is incised, and the divided arterial branch placed on the frontal cortical surface. The branch and the cortical surface are covered with a flap of the frontal muscle and galea. In cases with a poorly developed anterior branch, only the flap of the frontal muscle and the galea are placed on the exposed cortex. This procedure is called EMS.

Table 1. *Surgical Procedures in Paediatric Cases of Moyamoya Disease*

EDAS alone in the parietal region	19 sides
EMS alone in the parietal region	5 sides
EMAS or EMS in the frontal region and	
EDAS in the parietal region	17 sides
EDAS and EMS in the parietal region	5 sides
STA-MCA anastomosis and EMS	1 side
Total	47 sides

The EDAS in the parietal region together with the EMS in the parieto-occipital region was performed on five sides. The latter was carried out just posteriorly to the former.

The EMS alone was done on five sides of the patients with poorly developed STA. STA-MCA anastomosis was done only on one side of one patient.

Postoperative angiography was performed from 6 months to 1 year after operation, and the postoperative CT scan was examined once a year. To evaluate the surgical results, attention was directed to the following three points: 1. postoperative outcome of clinical symptoms, 2. collateral formation through the bypasses on the postoperative angiograms and, 3. new development of low density lesions on the postoperative follow-up CT scans. The postoperative collateral circulation was evaluated also for each of the different surgical procedure such as EDAS, EMS, and EMAS.

Surgical Results

1) Clinical Results (Table 2)

The postoperative outcome with regard to the symptoms, which had been observed for more than one year, was evaluated for 35 operated sides in 21 cases. The follow-up period ranged from 1 year to 4 years and 10 months (with an average of 2 years and 8 months). The postoperative outcome was classified into four grades. In grade I category, the symptoms totally disappeared. In grade II, they markedly decreased. In grade III, they persisted but less frequently. In grade IV, they remained unchanged or worsened. Of the 35 operated sides, grade I improvement was observed on 26 sides (74%), grade II on five sides (14%), grade III on two sides (6%) and grade IV on two sides (6%). Both sides in one case without postoperative collateral circulation through EMAS and EDAS showed the grade IV result.

2) Angiographic Results (Table 3)

Collateral circulation was evaluated by postoperative angiography on 36 sides in 22 patients. In cases in which multiple bypass procedures were done in different places on one side, the collateral circulation was judged as (+) when well developed collaterals were seen through at least one of them. Good collateral

Table 2. *Postoperative Outcome of Clinical Symptoms*

Completely disappeared	26 sides (74%)
Markedly decreased	5 sides (14%)
Decreased but persisted	2 sides (6%)
Unchanged and continued	2 sides (6%)
Total	35 sides (100%)

Table 3. *Postoperative Angiographic Results*

i) Cases (36 sides in 22 cases)			
collateral circulation (+)		30 sides (83%)	
collateral circulation (±) or (-)		6 sides (17%)	
ii) Procedures			
	collateral circulation (+)	collateral circulation (-)	circulation total
EDAS in the parietal region	26 (84%)	5	31 sides
EMS in the parietal region	5 (71%)	2	7 sides
EMAS or EMS in the frontal region	7 (64%)	4	11 sides

circulation was present on 30 out of the 36 sides (83%). On the remaining 6 sides the collateral circulation was poor or not present. Among the surgical procedures, the EDAS in the parietal region caused good collaterals on 26 of 31 sides (84%), the EMAS or EMS in the frontal region on 7 of 11 sides (64%) and the EMS in the parietal region on 5 of 7 sides (71%).

3) Results of Follow-up CT Scan

Postoperative CT scans were checked in 22 patients who could be followed for more than one year after surgery. In 18 patients new low density lesions suggesting infarcts were not demonstrated postoperatively. Some small infarcts appeared on five sides of the remaining four patients. Two of the four patients presented with a minor stroke and an infarction within one week after surgery as a postoperative complication.

4) Surgical Complications

Surgical complications included minor stroke in two cases, RIND in one, frequent TIAs in six, seizure in one, and osteomyelitis in one. The strokes and RIND developed within one week after surgery, and frequent TIAs and seizure had continued for 3 or 4 months after surgery. The osteomyelitis was first discovered as a microabscess in the operative skin scar 2 months after surgery and the infected bone flap was removed 7 months later. The minor stroke was the worst complication. However, the two patients with minor stroke

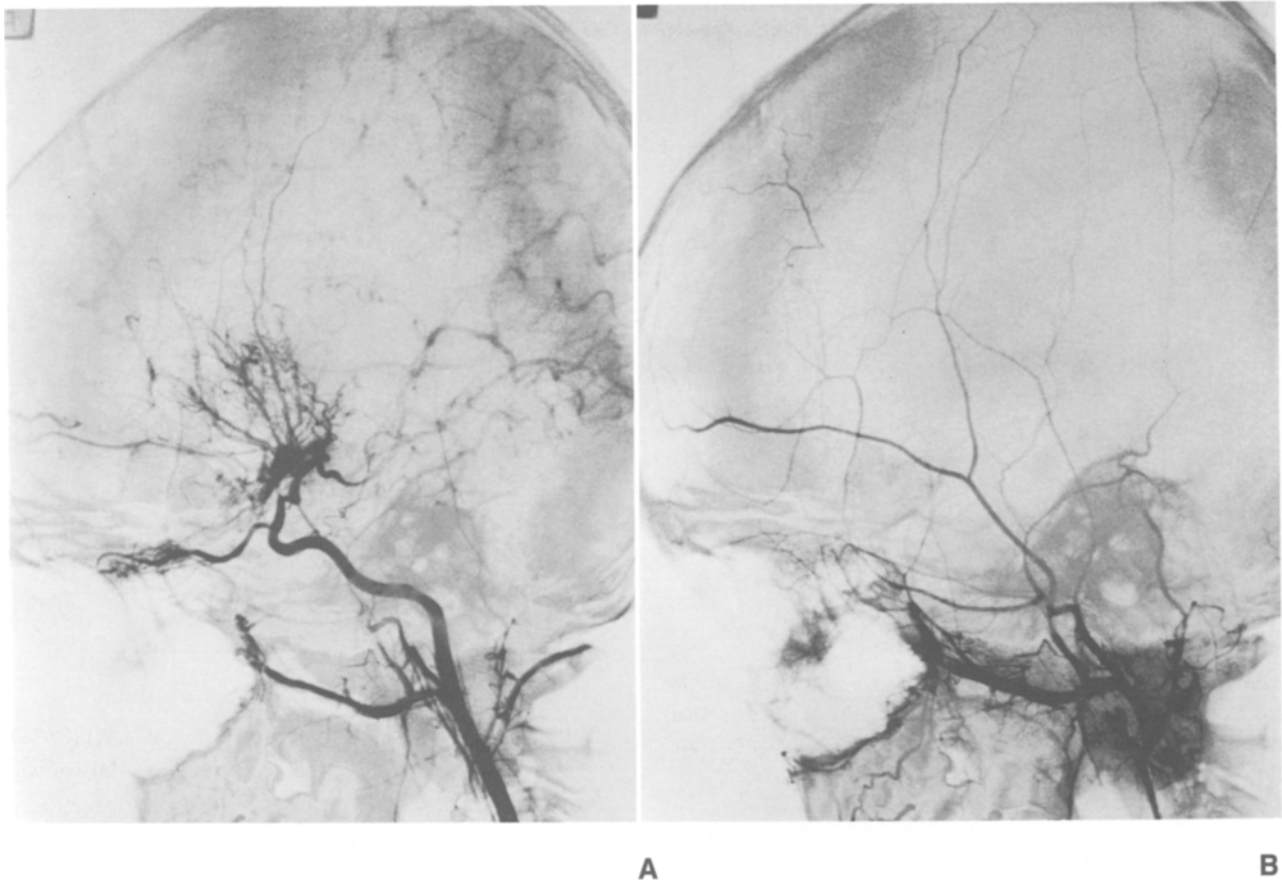


Fig. 1. Preoperative angiograms in case 1. A) Right internal carotid angiogram, lateral view: The supraclinoid portion of the right internal carotid artery was occluded, and the ACA and MCA group were poorly visualized. So-called Moyamoya vessels were seen in the basal ganglia and the frontal base. B) Right external carotid angiogram, lateral view: The superficial temporal and middle meningeal arteries were present but no dural anastomoses were seen

presented only with mild neurological deficits such as slight disturbances of speech and fine movement of the fingers. They are attending at the elementary school at present, having no major difficulties in their daily lives.

Two representative cases in which the indirect EC-IC bypass procedures were performed are described below.

Case 1, Treated by EDAS and EMAS

An 8-year-old boy was admitted because of TIA, which started 3 years previously and always occurred in the left upper and lower limbs. Neurological examination on admission was negative, and there was no low density lesion on CT scans. Hyperventilation load in EEG caused so-called “rebuild-up phenomenon”. Cerebral angiograms demonstrated occlusions of the right ICA and of the A1 portion of the left ACA associated with abnormal vascular network of Moyamoya vessels, the findings being compatible with those of Moyamoya disease (Figs. 1 A, B). The occlusive findings on the right side were striking, while those on the left side were milder. Since the TIAs occurred always on the left side of the body and the angio-

graphic findings were severer on the right side, EDAS and EMAS were performed in the right parietal and frontal regions respectively. After surgery the TIAs completely disappeared. Postoperative right external carotid angiograms demonstrated well developed collateral circulations in both the parietal and frontal regions, the area supplied by the MCA and ACA (Figs. 2 A, B).

Case 2, Treated by EMS

A 17-year-old boy developed the first transient motor weakness of both lower limbs at the age of 3 years. Since then he has had TIAs of the same type once or twice a month, which lasted to the age of 14 years. Since the age of 14 years severe headache persisted and amaurosis fugax on both sides developed. Neurological examination revealed poor mental activities. CT scans showed multiple cerebral infarctions on both sides, and angiograms demonstrated findings compatible with Moyamoya disease. EDAS was performed on the left side. EMS alone was done on the right side because an adequate STA was not available. Postoperative angiograms demonstrated good collaterals formed on both sides (Figs. 3 A, B). Amaurosis fugax gradually subsided and the headache improved post-operatively.

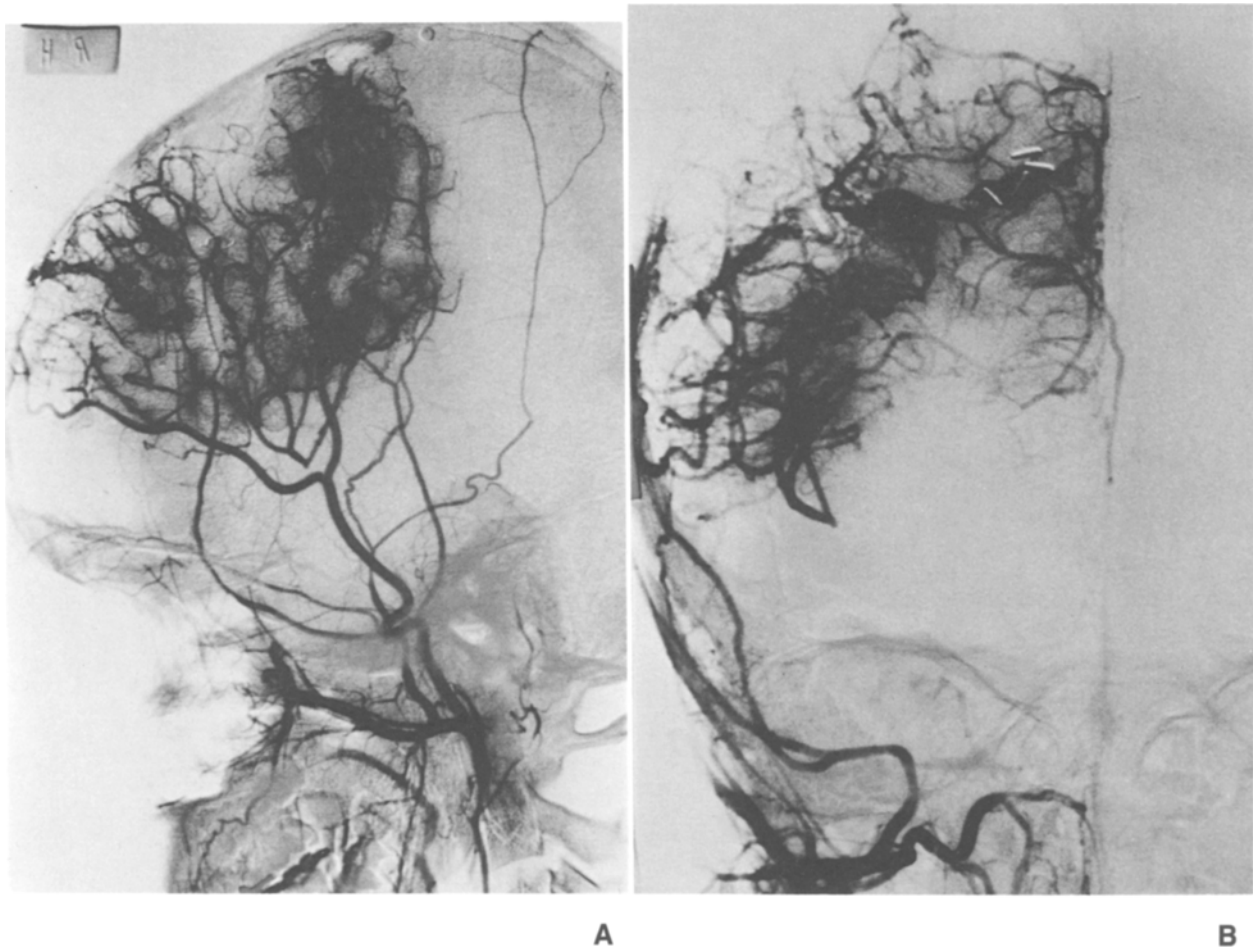


Fig. 2. Postoperative external carotid angiograms in case 1, 7 months after surgery. A) Right external carotid angiogram, lateral view: The superficial temporal artery has developed and dilated. It formed good collaterals in the fronto-parietal region. The anterior branch supplied the ACA area where the EMAS had been done. The posterior branch supplied the MCA area where the EDAS had been performed. B) Right external carotid angiogram, A-P view: Several cortical arteries of the ACA and MCA were demonstrated through the collateral circulation

Discussion

Since the proposals of Krayenbühl¹⁰, and of Karasawa and Kikuchi⁵⁻⁷, vascular reconstructive surgery for Moyamoya disease has become popular. However, it still seems necessary to compare the surgical results with the conservative medical results and to clarify the surgical indications, especially because the EC-IC bypass study group revealed failure of the EC-IC bypass surgery in reducing the risk of ischaemic stroke in arteriosclerotic occlusive disease^{1, 2}. With regard to the natural history of Moyamoya disease, Umezu²³ reported that the prognosis of the paediatric cases of TIA type was not bad, but long-term observations by others^{11, 12, 20} revealed that in general it was not good. Kurokawa *et al.* searched for the results in 27 paediatric patients who had been treated only conservatively in

the Department of Paediatrics of our hospital^{11, 12}. They concluded that in cases of children with Moyamoya disease the frequency of TIA gradually decreased during the course while intellectual deterioration and persistent motor disturbance gradually progressed. The two-thirds of the children, who could be followed for 10 to 15 years, were almost totally dependent on the support of their family in their daily lives, and were enrolled in low grade classes at school.

In our surgical results the clinical symptoms completely disappeared in 74% of the patients and markedly decreased in 14%. Although the follow-up period is not yet long enough, no patients developed any clinical strokes postoperatively or presented new large cerebral infarctions on CT scan except for two children, who had minor strokes within one week after surgery as a surgical complication. Most of the children whose

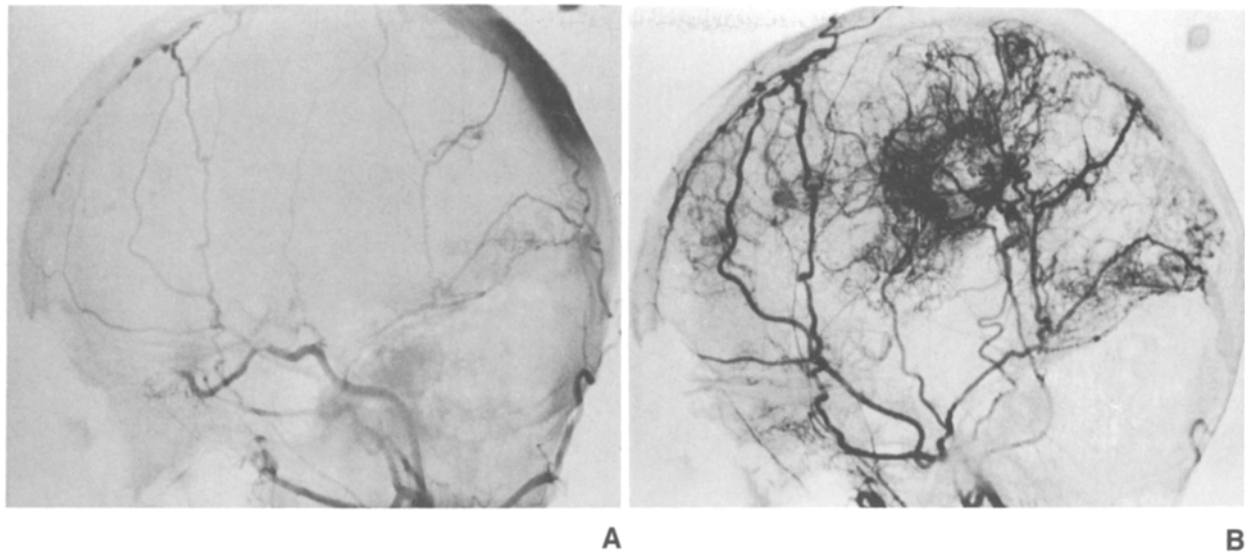


Fig. 3. Pre- and postoperative right external carotid angiograms in case 2, 7 months after surgery. A) Preoperative angiogram, lateral view: The anterior and posterior branches of the middle meningeal artery were present, but as for the superficial temporal artery only the anterior branch was seen. The dural anastomosis was formed by the anterior and posterior branches of the middle meningeal artery. B) Postoperative angiogram, lateral view: Abundant collateral vessels were seen in the fronto-parietal region where the EMS was performed. A few branches of the external carotid artery were dilated and formed the anastomoses

symptoms did not improve postoperatively showed poorly developed or absent collateral circulation through the bypasses. All but one of our patients were independent in active daily life. The facts suggest that the reconstructive surgery is effective for paediatric Moyamoya patients of the ischaemic type. The previous papers on the surgical results in Moyamoya disease also reported similar results, particularly in paediatric cases^{3, 5-7, 17, 22}. It is, however, necessary to follow up for a longer period in a large number of cases, so that the results of surgical and nonsurgical treatments could be reasonably compared.

One of the controversies is on the differing effects of the different surgical procedures. Although various kinds of reconstructive surgical procedures have been used in Moyamoya disease, they are basically of three kinds: STA-MCA anastomosis^{7, 10}, EMS^{5, 6, 9}, and EDAS¹⁴⁻¹⁶. The others are combinations or modifications of the three. The authors have applied mainly the indirect revascularization procedures including EDAS, EMS, and/or EMAS to paediatric Moyamoya cases for the past 5 years. It is because most of our patients were small children and indirect non-anastomotic EC-IC bypass procedures are technically easy.

By these procedures collateral circulations were well formed in about 70–80% of the surgical cases, in which the clinical symptoms subsided. However, in the re-

maining 20–30% of the cases collateral circulation was poor or absent and the clinical symptoms were not improved. Two causes for poor development of the collateral circulation can be considered. One is a technical problem such as excessive coagulation of the STA- or muscle-flap or no contact of the flap with the atrophic brain, and the other is no demand of the brain area, located under the course of the STA or the temporal muscle, for a new collateral circulation. Preoperative clinical examination of cerebral blood flow and metabolism using positron emission tomography may help to give a better indication for EC-IC bypass surgery^{3, 13}.

When the symptoms persist in cases without collateral circulation, another operation of a different procedure might be attempted, as reported by Miyamoto *et al.*¹⁸. If technically possible, the direct anastomotic EC-IC bypass may then be the procedure of choice. Such procedures should be done at the appropriate locations of the ischaemic brain before the brain succumbs infarction. It is stressed that the indirect non-anastomotic EC-IC bypass surgery is effective in most of the paediatric patients with Moyamoya disease.

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