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Surgical treatment of moyamoya disease in children: which is more effective procedure, EDAS or EMS?

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Abstract. At present, encephaloduroarteriosynangiosis (EDAS) and encephalomyosynangiosis (EMS) are the treatments of choice for moyamoya disease in children, but no attempts have been made to determine which is the more effective procedure, for the ischemic lesions in moyamoya disease. Ten patients (seven children and three adults) underwent EDAS and/or EMS: three patients EDAS on both sides; seven patients EDAS on one side and EMS on the other. These ten patients were followed up with a neurological examination and r-CBF and angiographic studies. Postoperative angiograms and r-CBF studies demonstrated more revascularization from the external carotid artery in sides treated with EDAS than with sides treated with EMS. From these results, it is concluded that the EDAS surgical procedure is superior to that of EMS for moyamoya disease.

Key words: Moyamoya disease – MRI – r-CBF – Surgical treatment – Chronic cerebrovascular occlusive disease – Cerebrovascular anastomosis.

Moyamoya disease is an unusal form of chronic cerebrovascular occlusive disease. It is characterized by bilateral stenosis of distal internal carotid arteries (ICA) and their area and by a hazy network of collateral circulation at the base of the brain. It is a rare condition and is most often found among the Japanese [4, 5, 6, 7]. Many reports and studies of the treatment of this disease have been published in Japan. The surgical procedures used at present for moyamoya disease are: superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis; encephalomyosynangiosis (EMS); and encephaloduroarteriosynangiosis (EDAS). As both the donor scalp artery and the recipient cortical artery are quite small in children, STA-MCA anastomosis is not easily applicable and sometimes hazardous. At present, EDAS and EMS are the treatments of choice in children, but no attempts have been made to determine in the same patients which is the more effective procedure for the ischemic lesions of moyamoya disease. In this report, evaluation of EDAS and EMS as operative methods is discussed from the point of view of the postoperative angiographic and r-CBF studies.

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Materials and methods

In the past 10 years, we have treated 21 cases of moyamoya disease. Ten patients (seven children and three adults) have undergone both EDAS and EMS; three patients had bilateral EDAS and seven EDAS on one side and EMS on the other. EMS was performed according to Henschen's [1] and Karasawa's reports [3] and EDAS according to the report by Matsushima et al. [5]. The EDAS or EMS sides were arbitrarily chosen, without considering the preoperative angiographic or neurological findings. The operation was performed in both sides on the same day. These ten patients were followed up by EEG, r-CBF, CT, and MRI, angiography, and neurological examination after operation. The follow-up periods ranged from 4 months to 3 years after operation. Regional cerebral blood flow was measured by the ^{133}Xe inhalation method and was calculated according to Obrist as an initial slow index.

Results

Clinical findings

The age and sex distribution of the 21 cases are shown in Fig. 1. The 13 females and 8 males with moyamoya disease ranged in age from 3 to 52 years. Their clinical features are summarized in Table 1. Of the juvenile patients, 6 had transient ischemic attacks (TIAs), 5 had TIAs followed by completed stroke, and 3 had convulsions. The adult patients presented with completed strokes in 3 cases, TIA in 2 cases, and convulsions in 3 cases. Intracerebral hematoma was observed in 4 cases: 1 child and 3 adults.

The use of CT and MRI in moyamoya disease were classified into four types according to the degree of the ischemic lesions, as shown in Fig. 2. The CT studies revealed that ischemic lesions in moyamoya disease most often occur in the watershed area in the frontotemporal area supplied by the anterior cerebral artery (ACA) and the middle cerebral artery (MCA), or in the temporo-occipital area supplied by MCA and the posterior cerebral

artery (PCA). These ischemic lesions most often occurred in the frontotemporal area as a watershed infarct because of ICA occlusion in moyamoya disease, as shown in Table 2. Severe forms of moyamoya disease (caused by cerebral ischemia) are more often seen in children. Eighteen patients with moyamoya disease, studied with CT or MRI, were classified according to the quality of activity in daily life and according to CT grades, as shown in Table 3. As the patients in CT grade IV had a poor prognosis, moyamoya disease should be treated in its early stages, before large ischemic lesions occur.

Postoperative angiographic findings

Preoperative angiography showed either occlusion or stenosis of the bilateral ICAs and basal moyamoya vessels in all cases. Postoperative angiography was repeated in ten cases. The latest angiography was performed more than 1 year after surgery in three cases, and in seven cases between 2 months and 6 months. Preoperative and postoperative angiographic findings were compared to study the degree of revascularization via the external carotid artery (ECA) and the change of the moyamoya vessels' size.

These angiographic findings were classified into three types according to the degree of the revascularization via ECA as shown in Table 4. Spontaneous revascularization between ECA and MCA was observed in 19 sides. Good filling of the entire MCA territory via ECA with decrease in size of moyamoya vessels was observed in 8 sides with EDAS (Fig. 3) and in 2 sides with EMS (Fig. 4). Good filling of the localized MCA territory via ECA was observed in 5 sides with EDAS and in 4 sides with EMS. The younger the patients and the more advanced the stage in moyamoya disease, the better the revascularization via ECA was.

Table 1. Clinical summary of 10 operative cases with moyamoya disease

Case	Age (years)	Sex	Symptoms	Surgery method	CT type
1	11	F	r-Hemiparesis; convulsion; mental retardation;	r-EDAS l-EDAS	IV
2	13	F	Convulsion; TIA	r-EDAS l-EMS	II
3	3	M	r-Hemiparesis; mental retardation	r-EMS l-EDAS	IV
4	7	F	TIA	r-EMS l-EDAS	II
5	10	F	TIA; hemianopia	r-EDAS l-EMS	II
6	10	F	TIA; hemianopia	r-EDAS l-EDAS	II
7	13	F	TIA	r-EDAS l-EMS	I
8	47	F	Hemiparesis	r-EDAS l-EMS	II
9	35	M	TIA	r-EMS l-EDAS	II
10	45	F	r-Hemiparesis; dementia	r-EDAS l-EDAS	IV

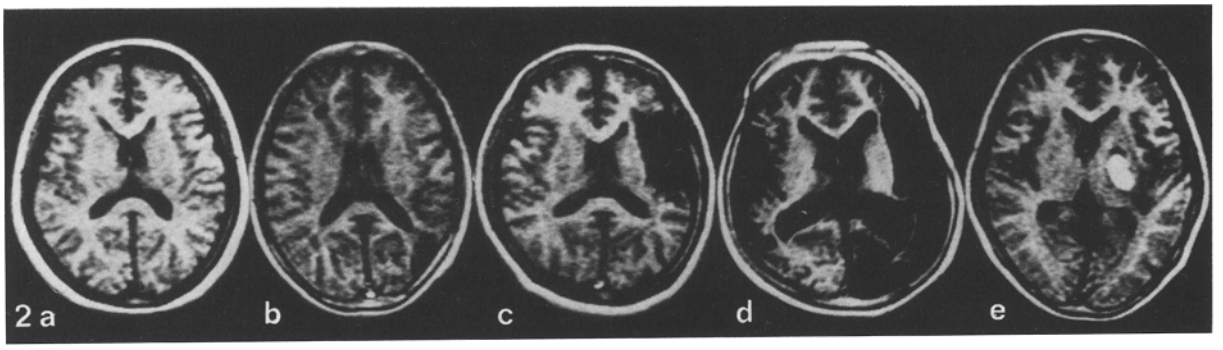
Fig. 1. Age and sex distribution of the 21 cases with moyamoya disease

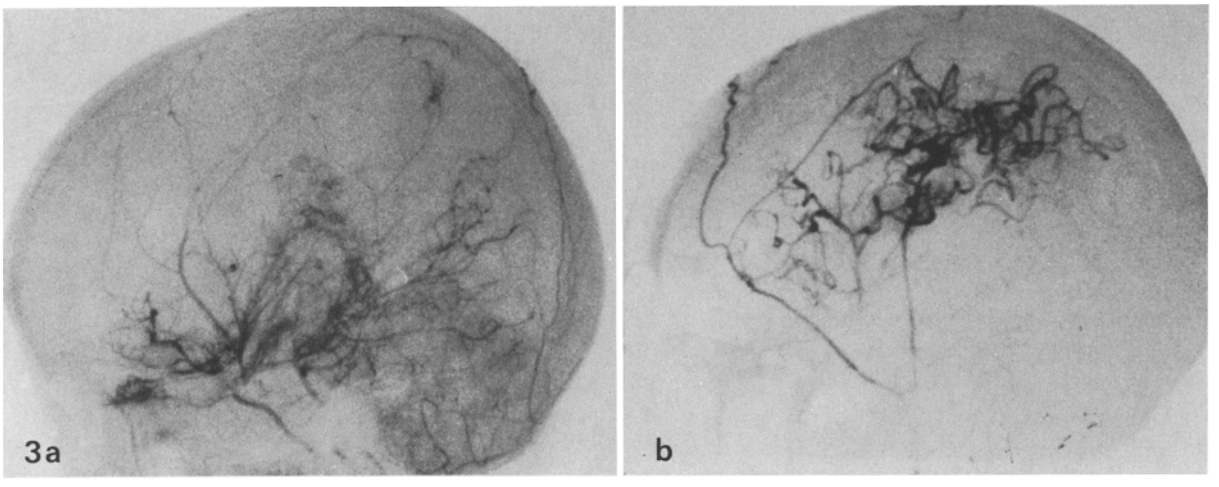
Table 3. Relationship between type of CT and outcome of moyamoya patients. E, full social recovery; G, limited social recovery; F, useful domestic life; P, incapable of self-care; D, dead

CT type	Cases	Child	Adult	Outcome				
				E	G	F	P	D
I	1	1	0	1				
II	6	4	2	6				
III	2	1	1	1	1			
IV	5	4	1			3	2	
V	4	1	3	1	1	2		

Table 2. CT findings in moyamoya disease

	Site of infarction							Cortical atrophy			Hemorrhage
	Frontal	Temporal	Parietal	Occipital	Frontal + temporal	Frontal + occipital	Temporal + occipital	Marked	Mild	Normal	
Child	4		1		1	2	5	4	8	2	1
Adult	4					1		1	2	1	2
Total	8		1		1	3	5	5	10	3	3





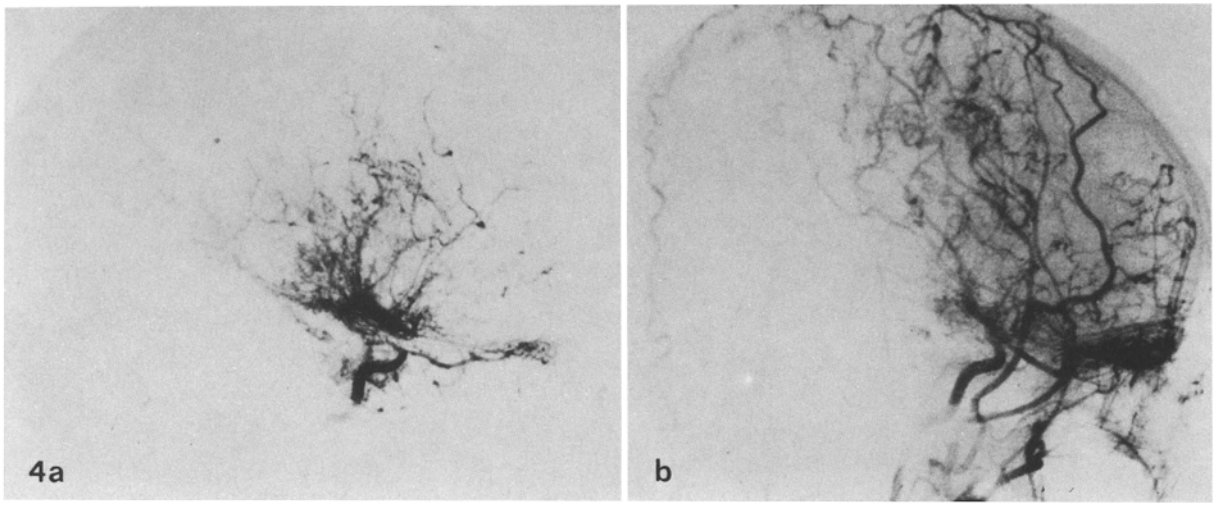


Fig. 2a–e. Classification of CT in moyamoya disease (ischemic type). **a** Type I, normal; **b** type II, small watershed infarct with localized cortical atrophy; **c** type III, large watershed infarct in frontal or occipital area with localized cortical atrophy; **d** type IV, large cerebral infarcts in both cerebral hemispheres with diffuse cortical atrophy and large ventricles. Hemorrhagic type: **e** type V, intracerebral hemorrhage

Fig. 3a, b. Change of angiographic findings after EDAS. Good filling of the entire MCA territory via ECA with decrease in size of moyamoya vessels was observed on EDAS side. **a** Before surgery, **b** after surgery

Fig. 4a, b. Change of angiographic findings after EMS. Good filling of the MCA territory in the frontal and temporal area, via ECA, with decrease in size of moyamoya vessels was also observed on EMS side. **a** Before surgery, **b** after surgery

Table 4. Angiographic changes after operation

	EMS	EDAS	Total
No change	1 side	0 side	1 side
Slight improvement of revascularization	4 sides	5 sides	9 sides
Marked improvement of revascularization	2 sides	8 sides	10 sides
Total	7 sides	13 sides	

Table 5. Change of m-CBF after surgery

	Preoperative (ml/100 g per min)	Postoperative (ml/100 g per min)
EMS	54.2±9.0	61.3±8.9
EDAS	53.8±9.1	64.2±9.2

Obrist, ISI

Table 6. Results of surgical treatment in 10 cases

Symptoms	Before surgery	After surgery	
Episode of TIA	6	improvement	6
		stationary	0
Motor disturbance	3	improvement	2
		stationary	1
Visual disturbance	2	improvement	1
		stationary	1
Mental handicap, ineducable (IQ < 50)	2	improvement	0
		stationary	2

The time required for revascularization in EDAS is 2 months, but in EMS it is 3 months on average, which means that revascularization occurs earlier in EDAS sides than in EMS sides. These results show that both EDAS and EMS are effective procedures to improve the ischemic condition in moyamoya disease, but EDAS is superior to EMS as a surgical procedure in moyamoya disease.

Postoperative r-CBF study

Preoperative and postoperative r-CBF were measured in five juvenile patients. All patients showed low values of mean hemispheric blood flow (m-CBF) preoperatively, but m-CBF was increased in all sides postoperatively, as shown in Table 5. In EMS sides, m-CBF was increased from 54.2±9.0 ml/100 g per min to 58.3±8.9 ml/100 g per min after surgery and in EDAS sides from 53.8±9.1 ml/100 g per min to 64.2±9.2 ml/100 g per min after surgery. These results also support the superiority of EDAS to EMS as a surgical procedure for moyamoya disease.

Operative results

Six patients with TIAs became free of TIAs 2 months after surgery, and two patients with hemiparesis improved to the point where they could walk without assistance. The intellectual deficits remained unchanged, as shown in Table 6. None of the cases showed neurological deterioration after surgery.

Discussion

Since the pathogenesis of moyamoya disease is still unknown, no effective treatment has been found for this disease. At present, the most reasonable approach to improve the ischemic condition in this disease appears to be reconstruction of new anastomotic channels via the external carotid system to increase blood supply to the brain. For this purpose, STA-MCA anastomosis, EMS, and EDAS seem to be the most effective and are, therefore, the treatments of choice at present. However, as both the donor scalp artery and the recipient cortical artery are quite small in children, STA-MCA anastomosis is not easily applicable and is sometimes hazardous because of temporary occlusion of the recipient cortical artery during the procedure. Thus, EDAS and EMS are the treatments of choice in pediatric patients. In this investigation, we have attempted to determine in the same patients which is the more effective procedure for the ischemic lesions of moyamoya disease – EMS or EDAS?

Karasawa et al. [3] reported satisfactory clinical improvement as seen in nine of ten moyamoya patients who underwent EMS. Postoperative, superselective deep temporal angiograms demonstrated the development of rich vascular blood supply from the temporal muscle to the brain surface. Matsushima et al. [5] developed a new operative method, EDAS, for the surgical treatment of pediatric moyamoya disease. They stated that this procedure was performed on five patients with satisfactory results and that more abundant blood supply to the brain was obtained from the scalp arteries of the EDAS than from an EMS muscle flap.

We treated moyamoya patients with EDAS on one side and EMS on the other, in the same patients, to determine which is the more effective procedure. In our cases, postoperative angiograms revealed more abundant revascularization from ECA in EDAS sides than EMS sides. R-CBF studies also confirmed that m-CBF was more increased on the EDAS than the EMS sides. The outcome in the cases operated upon showed that TIAs disappeared in all patients, but intellectual faculties were not greatly improved. From these results, it may be concluded that the EDAS surgical procedure is superior to that of EMS for brain revascularization in moyamoya disease and that the operation should be performed before irreversible changes occur.

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