

# Long Term Magnetic Resonance Angiography Follow-up in Moyamoya Disease

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## ABSTRACT

**Objective** : Revascularization is an effective treatment for the ischemic symptom of moyamoya disease. Indirect revascularization is also effective. Magnetic resonance angiography (MRA) has the ability for collateral formation that is equivalent to conventional angiography. This study analyzed the results of indirect revascularization by MRA. **Methods** : A total of 25 patients underwent bilateral EDAS for the management of moyamoya disease. All patients underwent MRA after surgery more than 24 months later. The collateral formation was graded as Good, Fair, and Poor. The clinical outcome was assessed as Excellent, Good, Fair, and Poor. **Results** : Good collateral formation was 32 sides of the EDAS, and fair was 18. An excellent clinical outcome was obtained in 15 patients, Good in 8, Fair in 1, and Poor in 1. There was a significant correlation between the preoperative symptom, gender, and the clinical outcome. **Conclusion** : In the management of ischemic moyamoya disease, indirect revascularization has been the golden standard with remarkably low morbidity and mortality. Moreover, and MRA can replace conventional angiography in the follow-up of moyamoya patients. (*Kor J Cerebrovascular Surgery* 9(3):188-92, 2007)

**KEY WORDS** : Moyamoya · MRA · Revascularization

## Introduction

Natural history of moyamoya disease has been known to be progressive repeated ischemic symptom of young age and hemorrhagic symptom of adult.<sup>2)</sup> Revascularization is the main treatment, with both direct and indirect revascularization. They increase blood flow and perfusion of cerebral cortex and decrease moyamoya vessels. Finally ischemic symptom decreases. Its angiogenesis of revascularization has been studied, by conventional angiography. However, conventional angiography needs tolerance of patient and puncture of large

artery, so risk of complication has existed. The quality of Magnetic Resonance Angiography (MRA) has been improved and study for angiogenesis of revascularization by MRA has been reported.<sup>4)</sup> Some studies have been reported regarding the value of MRA in diagnosis and follow up of moyamoya disease.<sup>6)</sup> But most of them had some limitations, such as small cases or short term follow-up period. The aim of this study is to show that MRA is useful for evaluation of postoperative result of indirect revascularization in moyamoya disease.

## Patients and Methods

### Patients

From January 2002 to January 2005, patients who were diagnosed as moyamoya disease and underwent indirect anastomosis, encephaloduroarteriosynangiosis(EDAS), were reviewed. Preoperative cerebral infarction was excluded for removal of influence of parenchymal structural changes on

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collateral vessel formation. All patients had been followed up at least 24 months. Preoperative Suzuki grade was checked by conventional angiography and MRA.

### Techniques of encephaloduroarteriosynangiosis (EDAS)

Our modality of indirect revascularization was EDAS. Surgery was performed according to the Suzuki stage and Matsushima grade. If the patient needed bilateral revascularization, surgery was continued after 4 or 6 weeks. With endotracheal general anesthesia, patients were placed on a table in supine position. Head was rotated to contralateral side to operation side. Superficial temporal artery (STA) was palpated and hair was shaved along the course of STA with about 2cm width. Ultrasonic Doppler might be used in some cases. Skin was incised along the STA path, from bifurcation of frontal and temporal branches. For protection of STA, blunt dissection between subcutaneous and galeal layer exposed STA and its branches. Bleeding control was done with very careful attention. Using the monopolar coagulator a galeal flap which contained STA was made. It was preserved during craniotomy. One burr hole was drilled at superior hinge point of the flap, than a craniotome drill made round shaped bone flap. Dura was incised straightly parallel to the meningeal arteries. Dura was split in inner and outer layer. Inner dura layer was excised, and then the galeal flap was sutured to outer dura margin. Bone flap was ground at flap insertion points, and fixed on the cranium. Drainage catheter was placed on the skull and wound was closed layer by layer.

### MRI follow up and outcome evaluation

All patients were followed at out-patient clinic. Collateral grade was assessed according to the collateral vessel formation and visualization;

Good: collateral vessels supplied more than two-thirds of the MCA territory.

Fair: between one-thirds and two-thirds of the MCA territory.

Poor: less than one-third of the MCA territory.

Clinical outcome was categorized as follows;

Excellent: Patients has been free from symptoms.

Good: Decreased frequency of symptoms.

Fair: Symptoms were slightly improved or persistent same as preoperative status.

Poor: Symptoms were aggravated. Frequency or intensity increased.

### Statistical analysis

Independent T-test and ANOVA test were used for comparison between preoperative clinical symptom, preoperative angiographic grading, and clinical outcome. Multivariate regression analysis was performed to analyze the prognostic impact of the following parameters on outcome grade: age, gender, and preoperative grade. The level of significance was  $p < 0.05$ .

## Results

Total 25 patients all underwent bilateral EDAS and total 50 EDAS were done. Male patients were 11 and female were 14. Mean age on operation is 8.5 years old (range from 4 to 15 years old). Mean follow up period is 35.8 months after last operation (range from 24 to 60 months). Other preoperative data was listed in Table 1. There was no significant difference in sex, age, preoperative symptom, and preoperative angiographic grading.

Collateral vessels were identified at final MRA(Fig. 1). Good collateral revascularization was observed in 32 cases of EDAS. Fair was 18 cases. There were no poor collaterals.

Postoperative outcome was like followings. Excellent outcome was observed in 15 patients(60%). Eight patients had bilateral good collaterals and others had mixed grade of

**Table 1.** Clinical data of 25 patients who underwent bilateral EADS due to moyamoya disease

Age at operation	8.52 (4~15) years old
Gender	
Male	11
Female	14
Clinical Symptoms	
TIA <sup>a</sup>	6
Frequent TIA	19
Angiographic grading <sup>b</sup>	
II	7
III	34
IV	9

a TIA : Transient ischemic attack

b Angiographic stage was evaluated according to the method of Suzuki and Kodama.<sup>17)</sup> Grade I, V and VI were not detected in this study

good and fair collaterals. Good outcome patients were 8(32%). Bilateral good collateral were 2, and bilateral fair collateral were 2. Others had that one side was good and other side was fair. There was 1 fair grade patients who had right side good collateral and left side fair collateral(4%). Only one patient had poor outcome, whose collaterals were bilateral fair grades(4%)(Table 2). Collateral formation was assessed with preoperative angiographic grades and clinical symptoms(Table 3). There was no statistical significance in each grade.

Statistically analysis showed significant correlation

**Table 2.** Clinical outcome of 25 patients and collateral vessel formation grading by MRA

Clinical outcome	N	MRA collaterals	Grade
Excellent	15	Good	23
		Fair	7
Good	8	Good	8
		Fair	8
Fair	1	Good	1
		Fair	1
Poor	1	Good	0
		Fair	2
Total	25		50

between preoperative symptom and postoperative outcome (Table 4). Other factors did not show significance, but sex and collateral grades had significance( $p < 0.05$ ). Female patients showed more favorable clinical outcome.

## Discussion

Indirect revascularization is one of effective methods for treatment of ischemic symptom of Moyamoya disease.<sup>5,12</sup> Indirect revascularization is easier and simpler procedure than direct anastomosis which is more appropriate if possible.<sup>11</sup> In

**Table 3.** Collateral grade and preoperative Suzuki grade and preoperative clinical symptoms

	Collateral grade		
	Good	Fair	Poor
Suzuki Grade			
II	4	3	0
III	23	11	0
IV	6	3	0
Clinical Symptoms			
TIA	11	1	0
Frequent TIA	22	16	0

**Table 4.** Multivariate regression analysis for outcome grade in patients who underwent EDAS

Variables	Regression coefficient	Standard error	P
Sex	0.539	0.249	0.006*
Age	0.021	0.187	0.575
Matsushima grade	0.171	0.038	0.494
Suzuki grade	-0.299	0.178	0.101
Collateral grade	0.621	0.202	0.003*

\* statistically significant ( $p < 0.05$ ).



**Fig. 1.** Collateral grading on MRA. (A) Good collateral formation (B) Fair collateral (C) Poor collateral.

our study, improvement of symptom was observed in 92% of patients. This result is same or superior to previous reported studies about result of indirect or direct revascularization of Moyamoya disease.<sup>8)13)</sup> It also has less complication compared with direct one. Mild headache and nausea occurred in some cases, but it disappeared soon. There was no morbidity and mortality in our study.

Postoperative image study is essential to evaluate the result and outcome of moyamoya surgery. MRA has become the choice of study for follow up and to check the result of revascularization. In this study, collateral vessel formation on postoperative MRA showed statistically significant correlation with clinical outcome. Development of collateral had been resulted in blood flow increase, and finally perfusion of cerebral hemisphere increased. Other factors did not have any statistical significance except gender. Female patients had more favorable outcome than male. No study about gender which was affecting the disease progression and severity of moyamoya disease was found. Further study may be needed. About other prognostic factor, recently a study reported that postoperative blood flow on SPECT is statistically prediction factor of clinical outcome.<sup>16)</sup>

MRA has ability to evaluate the collateral formation. In the past, MR study had been considered as an additional and supportive tool to angiography.<sup>1)14)18)</sup> Conventional angiography has been used as a golden tool of evaluation of cerebral vascular structure in moyamoya disease. But, patients were exposed to radiation and had to bear the pain during examination. Technology has improved and MR image became more detailed. MRA have many advantages over conventional angiography. There is no radiation exposure and pain. MR study is possible to evaluate brain parenchyma and vascular structures at the same time. Younger patients may need sedation or general anesthesia during angiography. Repeated anesthesia might increase risk of ischemic attack and cerebral infarction in moyamoya disease.<sup>9)</sup> Usefulness of MRA that was compared with conventional angiography had been proved by several authors.<sup>3)7)15)</sup> Fine moyamoya vessels could be visualized at the high resolution MR image, and also collateral vessels from implanted STA. MR Perfusion image has been available and it may substitute the conventional SPECT images.<sup>10)</sup>

The limitation of this study is retrospective and relatively small patients group. With more cases, the correlation

between development of collateral vessels and symptom improvement will be studied.

## Conclusions

In the management of ischemic moyamoya disease, indirect revascularization has been a golden standard with remarkably low morbidity and mortality. However the patient has been followed with conventional angiography with comparable risk. Fortunately the techniques of MRA have been remarkably improved to see fine collateral vessels. Therefore we suggest that the MRA can replace the conventional angiography in follow-up of moyamoya patient.

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