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Postoperative Electroencephalogram for Follow up of Pediatric Moyamoya Disease

It is well known that the electroencephalographic finding in patients with moyamoya disease demonstrates the characteristic "re-build up" phenomenon a few minutes after hyperventilation. To evaluate the usefulness of an electroencephalogram (EEG) in the postoperative management of children with moyamoya disease, we studied the presence or absence of improvement in the clinical, single photon emission computed tomography (SPECT) and EEG findings, before and after surgery. Twenty-two patients, who underwent indirect revascularization surgery for moyamoya disease, were included in our study. Clinical improvement was assessed as the disappearance or decrease of a transient ischemic attack or headache. The findings on the EEG and SPECT were considered improved when the re-build up phenomenon was absent and when there was improvement in the cerebrovascular reserve as a result of the acetazolamide challenge test. The statistical correlation analysis for both clinical and EEG improvement were consistent (kappa value=0.409, p<0.05). However, the result from the clinical and SPECT improvement as well as that from EEG and SPECT improvement were not statistically significant. Our results suggest that EEG can be used as a noninvasive and simple follow-up test for moyamoya disease after indirect revascularization surgery if the hyperventilation procedure is effectively performed during EEG recording.

Key Words : *Moyamoya Disease; Electroencephalography; Follow-up Studies*

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INTRODUCTION

Although cerebrovascular diseases are rare in childhood, moyamoya disease is a relatively important and common disease, especially among East Asian children. The pathophysiology of the moyamoya disease is that of progressive narrowing of the major medium- and/or large-sized cerebral arteries; however, the cause of the arterial narrowing is not known. Moyamoya disease was named for the abnormal vascular network at the base of the brain that is typically found on cerebral angiography (1). The disease was first reported by Takeuchi and Shimizu in 1957 and the term "moyamoya" means "something hazy just like a puff of cigarette smoke in the air" (1, 2).

In children, the disease generally presents as either transient ischemic attacks manifesting as a motor disturbance (70 to 80%), or with epileptic seizures (20 to 30%) (3). Once the diagnosis of moyamoya disease is suspected on clinical evaluation, confirmation by neuroradiological studies follows. Use of either invasive angiography or magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) are commonly employed for diagnosis (3, 4). The most consistent finding on the neuroradiological examination is the stenosis or occlusion of the terminal portion of the bilateral internal carotid arteries that is associated with an abnormal vascular network at the base of the brain. Therefore, several test modalities, such as the single photon emission computed tomography (SPECT) or MRA, have been used for diagnosis (5-7).

The electroencephalogram (EEG) is not commonly used as a diagnostic tool for this disorder. The EEG findings associated with moyamoya disease are high-amplitude slow waves, called hemispheric posterior slowing or centrotemporal slowing; these findings often appear in association with cerebral ischemia (8). The characteristic EEG findings associated with moyamoya have been reported to be a "build up" phenomenon, that is, the appearance of monorhythmic generalized and high voltage slow waves during hyperventilation followed by the "re-build up" phenomenon, the reappearance of polymorphous high amplitude slow waves several minutes after the end of hyperventilation (9, 10).

The aim of this study was to evaluate the usefulness of the EEG as a follow-up test for the postoperative status of patients with moyamoya disease. We studied the clinical characteristics of patients who underwent surgical revascularization for moyamoya disease and analyzed the correlationship among clinical, EEG and SPECT findings before and after the surgical procedure.

MATERIALS AND METHODS

A total of one hundred and thirteen patients below the age of fifteen years-old underwent surgical revascularization at the Asan Medical Center, Seoul, Korea from March 1995 to March 2001. We reviewed the medical records and the patients who did not have EEG data before and after the operation were excluded from this study. As well, patients who did not perform the effective hyperventilation during the EEG recording were excluded. We defined effective hyperventilation on the basis of the build up phenomenon or on the technician's electroencephalographic description. The patients with a mild cerebral infarction on MRI or with continuous focal slowing on the EEG were included. We recruited twenty-two patients for this study.

We performed the EEG or SPECT evaluations 6-12 months after the surgery; clinical evaluation for the recurrence of symptoms was done every three months after the surgery. The EEG was done on an international 10-20 system and was recorded 2 to 5 min after hyperventilation for 3 min (Fig. 1). We obtained SPECT images from the Triad XLT 20 (Trionix, OH, U.S.A.) using the Tc-99m ethyl cysteinate dimer with acetazolamide stress.

Clinical improvement was defined as the decrease in frequency or intensity of the TIAs or headache. Improvement of EEG findings was based on the disappearance of the re-build up phenomenon. Improvement on SPECT imaging was defined by improvement in the cerebrovascular reserve with acetazolamide stress test.

In order to evaluate the usefulness of the EEG as a follow-up test for moyamoya disease, we used the kappa value for statistical analysis. After indirect revascularization surgery we compared various sets, which can consist of pairs out of clinical, EEG and SPECT data. A p value less than 0.05 was considered significant.

RESULTS

Clinical characteristics

Patients presented initially with the following symptom and sign: TIA (n=19), seizure (n=1), hemiplegia (n=1) and headache (n=1). The ratio of male to female patients was 1:1. The median age of the patients was six years old. The duration from the onset of symptoms to the operation ranged from one month to nine years (median duration, 16 months). Before the surgery, cerebral infarction was detected in 8 patients and associated with hydrocephalus in one case. One patient had a cerebral infarction after the first operation. The follow-up duration ranged from 7 months to 60 months with a median duration of 26 months (Table 1). Clinical improve-

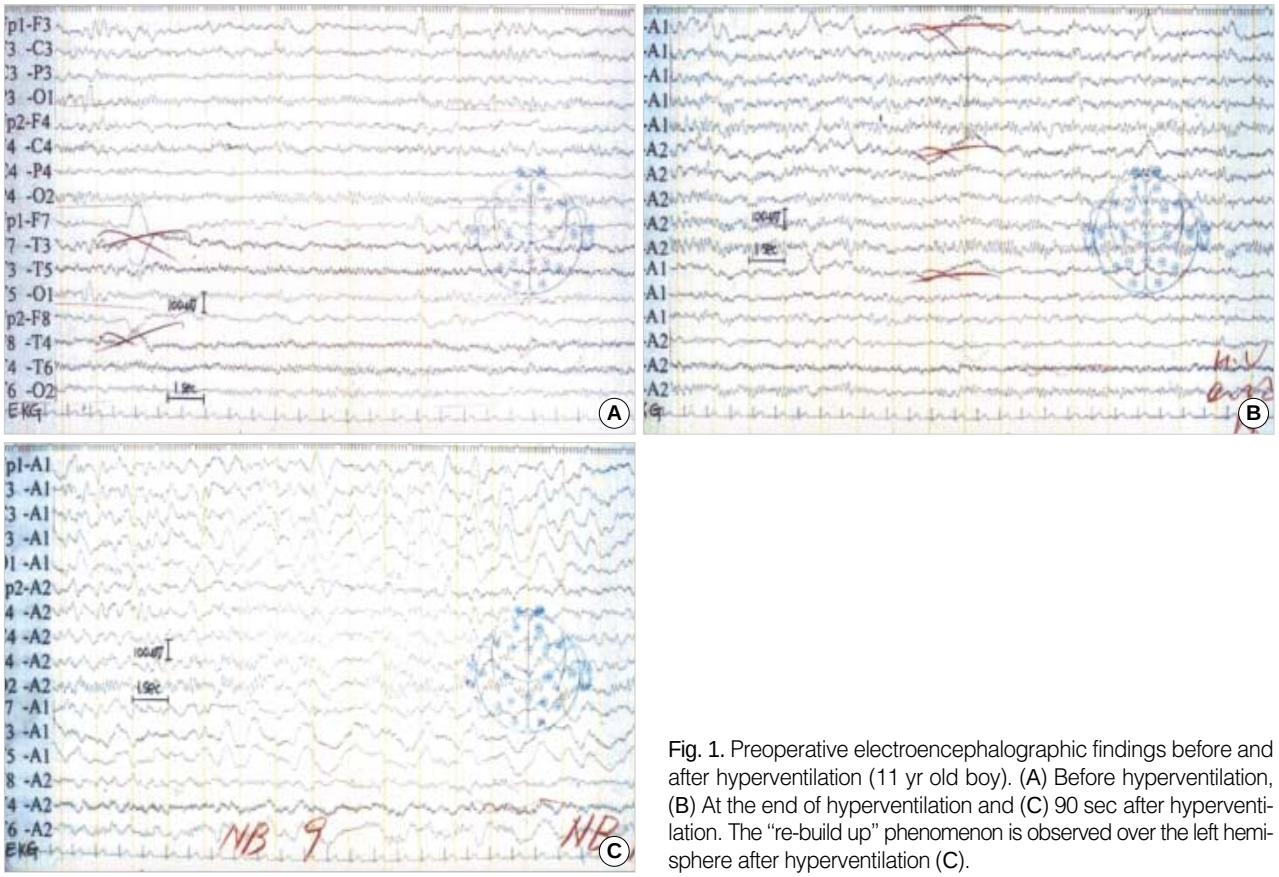

Fig. 1. Preoperative electroencephalographic findings before and after hyperventilation (11 yr old boy). (A) Before hyperventilation, (B) At the end of hyperventilation and (C) 90 sec after hyperventilation. The "re-build up" phenomenon is observed over the left hemisphere after hyperventilation (C).

Table 1. The clinical characteristics of patients with moyamoya disease

Sex	
Male	11 (50.0%)
Female	11 (50.0%)
Age at operation	4-15 yr
Chief complaint at first visit	
Transient ischemic attacks	19
Headache	1
Seizure	1
Hemiplegia	1
Preoperative	
Duration of symptom	1 month-9 yr (median, 16 months)
Cerebral infarction	8 (36.4%)
Abnormal EEG findings*	17 (77.2%)
Postoperative	
Follow-up period	7-60 months (median, 26 months)

EEG, electroencephalogram. *, Abnormal EEG findings included slow wave discharges or re-build up phenomenon.

ment was found in about 82% of patients. Improvement in EEG and SPECT was about 68%, 73% respectively (Table 2).

Agreement among clinical, EEG and SPECT improvement

Based on the statistical analysis for clinical and EEG improvement, the kappa value was 0.409, which signifies good agreement ($p<0.05$). In the case of clinical and SPECT improvement, there were poor agreement and no significant result. In addition, agreement between the postoperative EEG and SPECT improvement did not show statistical significance. Out of four cases with no clinical improvement, three cases showed no improvement in EEG findings, whereas there was no SPECT improvement in one case. In seven cases of no EEG improvement, SPECT improvement did not observed in two cases.

DISCUSSION

Moyamoya disease was first reported by Takeuchi and Shimizu (2). The disease is characterized by hypoplasia of the bilateral internal carotid arteries. The abnormal vascular network is found on angiography (1, 2). The prevalence of the disease is high in East Asian countries, especially in Japan, China and Korea. Although the etiology of moyamoya disease is still not known, it is considered to be multifactorial condition that is affected by elements such as vascular growth factors, cytokines, basic-fibroblast growth factor or genetic abnormality (11-15).

In children, the disease generally presents with either frequent TIAs manifesting as hemiparesis, monoparesis and/or sensory disturbance (70 to 80%), or with epileptic seizures (20 to 30%) (3). In addition, headache or involuntary movements can also be a presenting symptom. In our study, hemiparesis was observed in about 86.4%; the presence of seizure or headache was noted in a small number of patients.

Table 2. The improvement in clinical, EEG and SPECT findings after revascularization surgery

	No improvement	Improvement
Clinical finding	4	18
EEG finding	7	15
SPECT finding	6	16

EEG, electroencephalogram; SPECT, single photon emission computed tomography.

The diagnosis of moyamoya disease can be confirmed by conventional angiography or MRA; the findings from these studies can be useful for planning surgical treatments (3, 4). Cerebral blood flow is evaluated to determine the clinical severity of moyamoya disease, the effectiveness of indirect revascularization surgery and it can be measured by ^{133}xenon computed tomography, SPECT, or positron emission tomography (5-7, 16, 17).

In children, EEG may provide definite information on moyamoya disease even when clinical symptoms are missing (10). The EEG findings associated with moyamoya disease shows high-amplitude slow waves in association with cerebral ischemia and the characteristic "re-build up" phenomenon. A "re-build up" phenomenon is the reappearance of polymorphous slow waves several minutes after cessation of the hyperventilation (8-10). About 95% of all reported cases where a hyperventilation test was performed during EEG recording showed this "re-build up" phenomenon (10). In this study we obtained the re-build up phenomenon in about 80% of the patients who were capable of performing an effective hyperventilation. When a patient presented with seizure and characteristic EEG changes, moyamoya disease was considered as a possible etiology.

The mechanism that is responsible for the EEG re-build up phenomenon is still unclear, but several studies have suggested a possible mechanism. Electrophysiological build-up during hyperventilation is based on a reduction of the arterial partial pressure of carbon dioxide as a result of hyperventilation and the consequent reduction of cerebral perfusion (18). The concentration of oxygenated hemoglobin progressively increases during hyperventilation and subsequently decreases after hyperventilation for 5 to 7 min. The re-build up phenomenon occurs when oxygenated hemoglobin decreases and the deoxygenated hemoglobin increases (19). In a region where cerebrovascular reactivity is disturbed, a reduction of regional cerebral blood flow after hyperventilation results. Therefore, the regional cerebral hypoxia and the disturbance in oxygen metabolism play important roles in the occurrence of the re-build up phenomenon after hyperventilation (20, 21).

Surgical revascularization procedures are used to augment cerebral blood flow and indirect revascularization surgery is

preferred for young children. Within a few months after surgery, the ischemic attacks disappear in most patients (4). Most patients included in this study had received the encephaloduroarteriomyosynangiosis, which is known to be the most effective procedure for preventing ischemic symptoms in the middle- and anterior cerebral arteries in children with moyamoya disease (22, 23).

After surgery for moyamoya disease, it is important to follow patients for possible recurrence of symptoms associated with moyamoya disease because of progressive vascular narrowing and ischemic insults. Moreover, because revascularization surgery may prevent irreversible neuronal damage, early detection of areas with impaired hemodynamics is critical in the management of moyamoya disease. It is also important to evaluate whether there is the improvement of cerebral perfusion after the operation. Improved blood supply to the ischemic brain from the external carotid artery branches has been demonstrated. Determination of the correlation between clinical outcome and the degree of revascularization from surgery, as evaluated by neurological investigation, is difficult (24). Therefore, effective methods to monitor the change of regional cerebral perfusion reserve after surgery is required. SPECT is valuable in assessing the cerebral perfusion reserve and predicting a long-term prognosis, but the procedure is somewhat inconvenient to perform in children. Unlike SPECT, the EEG cannot directly reflect the blood perfusion or vascular reserve in cortical areas. However, the EEG is a noninvasive diagnostic tool that is commonly used, and well tolerated, in children. We performed EEG and SPECT 6-12 months after surgery. The postoperative EEG findings showed the disappearance or the reduction in the duration and distribution of the "re-build up" phenomenon; these findings are consistent with other reports (9). We found that there was good agreement between clinical and EEG improvement using the kappa statistic and that EEG may be useful tool for predicting the disappearance or recurrence of clinical symptoms in children with moyamoya disease. In conclusion, we suggest that an EEG, a simple and noninvasive study, may be useful as the primary follow-up test after surgery for moyamoya disease in children.

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