Encephaloduroarteriosynangiosis (EDAS) for the Treatment of Childhood Moyamoya Disease

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Moyamoya disease is defined by the angiographic demonstration of stenosis or occlusion of the vessels of the anterior circulation at the base of the brain and the concomitant development of collateral blood supply. Untreated, the disease is often clinically progressive, resulting in significant neurologic sequelae. Encephaloduroarteriosynangiosis (EDAS), which involves the transposition of a segment of a scalp artery onto the surface of the brain, is a surgical treatment aimed at improving collateral blood flow. Six children underwent 8 EDAS procedures and were followed from 6 months to 9 years after surgery. No patient experienced further deterioration in neurologic status. Postoperative angiography demonstrated cerebral revascularization from the donor scalp artery on 3 of the 6 EDASs that were studied. The 2 patients who did not revascularize after EDAS demonstrated angiographic regression of their disease. The data suggest that EDAS is a safe procedure for the treatment of childhood moyamoya disease. Given the potential severity of the sequelae, early operative intervention is recommended in all children with this disease.

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Introduction

Moyamoya disease is an entity with which the neurologic community is now familiar. Suzuki first used the term "moyamoya" to describe this cerebrovascular disorder [1]. The angiographic appearance of the collateral blood vessels that develop in response to stenosis and then occlusion of the main vessels at the base of the brain reminded Suzuki of "puffs of cigarette smoke," for which the Japanese vernacular term for misty, "moyamoya," seemed most appropriate. The initial presentation of the child with moyamoya disease is usually an ischemic one, with either stroke or transient ischemic attack (TIA), whereas in the adult population, subarachnoid hemorrhage secondary to rupture of fragile collateral vessels or associated aneurysms may also occur [2]. Seizures are a common clinical manifestation of childhood moyamoya disease. The computed tomographic (CT) scan usually demonstrates appropriate hypodensities or hyperdensities in the presence of ischemic stroke or hemorrhage. Serial angiography establishes that the vascular occlusion is progressive, the stages of which have been described by Suzuki [1].

Medical treatment, including antiplatelet agents and vasodilators, has not met with much success in preventing the development of neurologic deficits. Early surgical efforts with superior cervical ganglionectomy and perivascular sympathectomy of the internal carotid artery (ICA) were found to result in short-term improvement, but did not halt the progression of the disease [3]. Thus surgical management has been directed at improving the collateral blood flow, bypassing the stenotic or occluded segments of blood vessels. Options for revascularization include superficial temporal artery to middle cerebral artery (STA-MCA) anastomosis [4], omental transplantation onto the cortical surface [5], encephalomyosynangiosis (EMS) [6], and encephaloduroarteriosynangiosis (EDAS) [7]. EDAS was first described in the English literature by Matsushima in 1981 [8]. Borrowing on the concept of increasing the collateral blood flow to the cerebral hemisphere by transplanting omentum or muscle onto the cortex, Matsushima suggested transposing a portion of a branch of the external carotid artery, usually the posterior branch of the superficial temporal artery (STA), onto the cortex. The elegance and simplicity of this procedure make it an attractive choice in the treatment of childhood moyamoya disease.

The North American and European experience with

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childhood moyamoya disease has not been as extensive as that of the Japanese. Because many pediatric neurologists are not aware of the current opinion that aggressive early surgical management is recommended, we present our experience with 6 consecutive children treated with EDAS and review the literature on the subject.

Methods

Operative Technique. Briefly, the procedure of EDAS was performed as follows. The suitable vessel, usually the posterior branch of the STA, was traced on the scalp with the aid of a Doppler probe. Using magnification, an incision was then made along this line and the vessel found. As long a segment of the vessel as possible, between 5 and 10 cm, was isolated with galea on either side to a width of about 10 to 14 mm. The vessel was then gently retracted as a linear craniotomy was fashioned at least 20 mm wide. The dura mater was opened in a straight line and then to it were sutured the galeal edges of the isolated segment of artery. Some authors have also opened the arachnoid, but we did not elect to do this. The bone flap was usually not replaced, although when this was done, careful attention was paid to the vascular pedicle to ensure that it was not compressed by the bone. The donor vessel was not divided at any time, so that flow was sustained in it throughout the procedure. The wound was closed in the usual fashion. Special attention was paid to anesthetic management during these cases, with maintenance of P_{CO_2} between 35 and 45 mm Hg [9,10].

Patients. Six children, 2 male and 4 female, were treated with EDAS for moyamoya disease at our institution between June, 1982 and June, 1992. First symptoms appeared between the ages of 18 months and 7 years, and all patients eventually presented with single or recurrent episodes of sudden hemiplegia (Table 1). One patient had neurofibromatosis type 1 (Patient 3) and another coarctation of the aorta (Patient 5). All but 1 patient (Patient 3) were first seen at our institution within 1 year of the first symptom. Investigations included CT scan, which demonstrated hypodensities in all patients, and electroencephalography (EEG) which showed abnormal slow waves on the more affected sides. Diagnosis was made by conventional angiography, and the findings are summarized in Table 2. EDAS was performed bilaterally in 2 patients (Patients 1 and 4), and ipsilateral to the more affected hemisphere in the other 4 patients. Five patients underwent postoperative conventional angiography, and 6 EDASs were evaluated (Table 2). Three of 6 EDASs were found to contribute blood flow to the brain. Donor vessels were angiographically patent in all 6 patients. The 2 children who did not develop any collateral circulation from the EDAS manifested angiographic regression of their disease, with diminished stenosis or recanalization of previously occluded vessels. Intravenous digital subtraction angiography was performed postoperatively in 1 patient and 1 patient underwent postoperative magnetic resonance angiography. These latter 2 examinations were judged to be of inadequate resolution to determine the patency of and collateral blood supply to the brain via the EDAS. In 2 patients preoperative positron-emission tomography (PET) scanning was performed, one of whom (Patient 1) was also subjected to postoperative regional cerebral blood flow (rCBF) and regional cerebral metabolic rate of oxygen utilization ($rCMRO_2$) measurements.

All patients tolerated surgery well, with no observed incident of neurologic deterioration. During the follow-up period, which ranged from 6 months to 9 years, there have been no instances of neurologic deterioration. Outcome was judged to be good in 4 of the 6 patients. Patient 1 continues to have mild spastic quadriparesis, dysarthria, and behavioral problems at 13 years of age. Patient 3 is globally delayed, requiring special schooling at age 17 years.

Case Reports

Two representative cases have been chosen for discussion. The first case (Patient 1) had advanced disease at the time of intervention. Surgery resulted in successful revascularization of 1 hemisphere and the patient remained clinically stable with no further strokes. No improvement in CBF, however, was found postoperatively and the ultimate outcome was judged to be poor. In the second case (Patient 5) surgical intervention was at a much earlier disease stage and the angiographic and clinical outcomes were much better.

Patient 1. A previously well 19-month-old boy presented with an acute onset of right hemiparesis and epilepsia partialis continua of the right side of the face. CT scan showed hypodensity in the left mid-parietal area and

Table 1. Summary of clinical presentation, treatment, and outcome in six children with moyamoya disease

Pt. No.	Presentation			Treatment		Outcome
	Age	Sex	Symptoms	Surgery Age	Medical Rx.	
1	1.5 yrs	M	L stroke, then R stroke	L. EDAS, 2.5 yrs R. EDAS, 3 yrs	ASA, CBZ	Clinically stable for 9 yrs. Mild spastic quadriparesis and developmental delay. Remains on medication.
2	2 yrs	F	L stroke, then alternating hemiplegia	L. EDAS, 12 yrs	ASA, propranolol, PHT, phenobarbital	Clinically stable for 7 yrs. Very minimal hemiparesis. Gave birth at age 19 yrs. All medication discontinued.
3	3 yrs	M	R stroke, seizures, NF1	R. EDAS, 9 yrs	PHT, CBZ	Clinically stable for 9 yrs. Seizures and developmental delay. Remains on medication.
4	6 yrs	F	L stroke	L. EDAS, 6 yrs R. EDAS, 6 yrs	ASA	Clinically stable for 5 yrs. Neurologically intact. All medication discontinued.
5	3 yrs	F	L stroke, coarctation of aorta	L. EDAS, 3 yrs	Antihypertensives	Clinically stable for 3 yrs. Neurologically intact. Remains on medication.
6	7 yrs	F	L stroke	L. EDAS, 7 yrs	ASA	Clinically stable for 6 mos. Mild hemiparesis, improving. Remains on medication.

Abbreviations:
ASA = Acetylsalicylic acid
CBZ = Carbamazepine
NF1 = Neurofibromatosis type 1
PHT = Phenytoin

Table 2. Summary of pre- and post-EDAS angiograms in six children with moyamoya disease

Pt. No.	Preoperative Angiogram	Surgery	Postoperative Angiogram
1	Occluded L ICA. Stenosis R ICA (C4), A1 and M1. Bilateral basal moyamoya. Transdural collaterals. Posterior circulation not assessed.	L EDAS then R EDAS	L EDAS patent but not contributing to intracranial circulation. R EDAS contributing to R MCA circulation. Angiogram otherwise unchanged.
2	Occluded R and L ICA (C4). Bilateral basal moyamoya. Transdural collaterals. Blood supply via posterior circulation and patent R anterior choroidal artery.	L EDAS	Not done
3	Occluded R and L ICA (C4). Bilateral basal moyamoya. Transdural collaterals. Overall, better collateral circulation to L hemisphere. Posterior circulation not assessed.	R EDAS	R EDAS patent and contributing to R hemisphere circulation via R middle meningeal artery and collaterals. Angiogram otherwise unchanged.
4	Stenosis L ICA (C4) and M1. Occlusion L A1. Mild stenosis R ICA (C4). L basal moyamoya. Normal posterior circulation.	L EDAS then R EDAS	L EDAS patent but not contributing to intracranial circulation. R EDAS not assessed. Angiogram demonstrated marked regression of stenosis with recanalization of L A1.
5	Occluded L ICA (C4). Stenotic R A1. Most of L hemisphere filled via ophthalmic collaterals and R ICA. Posterior circulation not assessed.	L EDAS (2 vessels)	L EDAS patent (both vessels) and contributing much flow to L MCA circulation. Angiogram otherwise unchanged.
6	Stenotic L ICA (C4), A1 and M1. Mild R ICA (C4) stenosis. No moyamoya or transdural collaterals. Normal posterior circulation.	L EDAS	L EDAS patent but not contributing to intracranial circulation. Decreased stenosis of L ICA and branches evident.

Abbreviations:
ICA = Internal carotid artery
A1 = A1 segment of anterior cerebral artery
M1 = M1 segment of middle cerebral artery
C4 = C4 segment of ICA

EEG was remarkable for moderate slow-wave disturbance over the left posterior quadrant. Angiography revealed complete occlusion of the left ICA with collateral vessels supplying the left hemisphere. While the right ICA was patent, stenoses of the A1 and M1 segments [11] of the anterior cerebral artery (ACA) and middle cerebral artery (MCA) on this side were seen. About 1 year later, the patient presented again; this time with an acute onset of left hemiparesis and aphasia. CT scan revealed a right parieto-temporo-occipital hypodensity and angiography demonstrated occlusion of the right MCA, which filled in a retrograde fashion only, and increased basal moyamoya vessels in the basal ganglia. The patient was referred for surgical treatment. Because the disease was more severe on the left, EDAS was first performed on this side. Four months after the EDAS, he underwent PET scanning with $^{15}O_2$ rCBF and ^{15}O-CO_2 $rCMRO_2$ determinations. The findings were consistent with right MCA and left posterior-frontal infarctions. He then underwent EDAS on the right side. Surgery was well tolerated and the PET scan was repeated 6 months later. This revealed no changes from the previous study. Although an angiogram at this time indicated patency of the donor STA on the left side, it did not contribute blood flow to the cerebral vasculature. Some filling of the right MCA via the right EDAS, however, was seen. The patient remained clinically stable thereafter, suffering no neurologic deterioration. At last follow-up, 9 years after the initial EDAS, he was being treated with carbamazepine for his seizure disorder and acetylsalicylic acid for stroke prevention. His spastic quadriparesis had improved to the point where he was able to ambulate on his own, albeit with a wide-based gait. Speech was dysarthric and there were some behavior problems.

Patient 5. A 3-year-old girl presented to our institution with an acute history of right-sided hemiparesis and dysphasia. She had complained of headaches and had had intermittent fevers for 2 weeks. On the day prior to admission amoxicillin therapy had been initiated for otitis media. A CT scan on the day of admission revealed several left-sided hypodense areas. Cerebral angiography demonstrated complete occlusion of the left supraclinoid ICA (Figure 1). Some left hemispheric blood flow was contributed by collateral vessels that had developed from the ophthalmic artery on this side, and additional flow to the left hemisphere came from the right ICA via the anterior communicating complex. The A1 segment of the left ACA was also stenotic, as was the supraclinoid segment of the right ICA. The patient's speech and hemiparesis rapidly improved while she was in hospital. On the sixth day of admission, left EDAS was performed using both the anterior and posterior branches of the STA. The surgery was well tolerated and the patient was discharged 1 week later. In the 3 years since this procedure the patient has had no further episodes of cerebral ischemia and no seizures. Two months after the EDAS surgery, an incidentally detected coarctation of her aorta was repaired successfully, with no neurologic sequelae. Follow-up cerebral angiography 6 months after EDAS revealed excellent filling of the left MCA territory via the transplanted STA branches (Figure 2). The left ICA remained occluded and the right supraclinoid ICA segment and left A1 ACA segments remained stenotic.

Discussion

The natural history of the childhood form of moyamoya disease is not known as there have been no prospective randomized studies of the disease. Hence there has been some controversy over how to select patients for operative intervention. The high incidence of ischemic abnormalities in children, however, indicates that an aggressive approach to revascularization should be undertaken. Kurokawa et al. [12] retrospectively examined the files of 27 pediatric patients with moyamoya disease. Eleven had

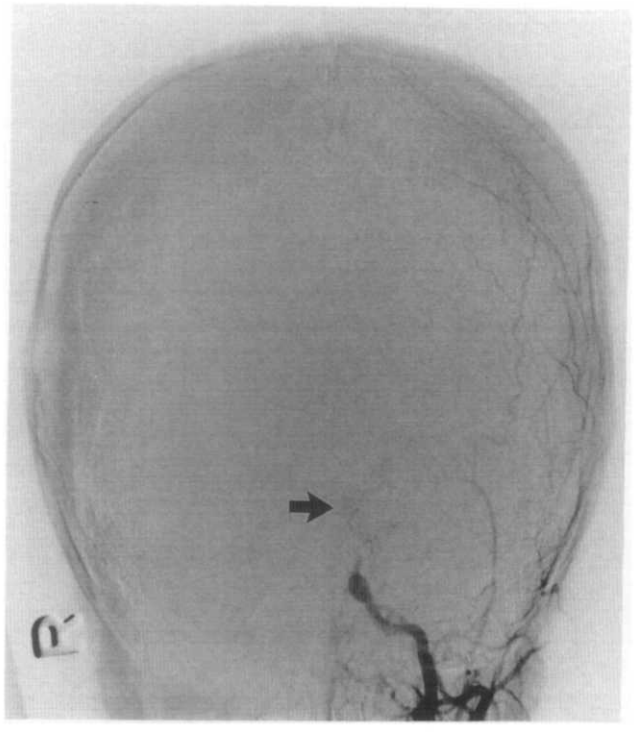

Figure 1. Anterior-posterior (AP) projection of left common carotid artery (CCA) pre-EDAS angiogram (Patient 5). Note occlusion of supraclinoid internal carotid artery (ICA) and early development of basal moyamoya vessels (arrow).

been treated with surgery, and follow-up ranged between "less than" 4 years to 15 years from the time of diagnosis. A poor outcome was seen in 21% of the patients who either died (3%), required continuous 24-hour care (7%), or required special schooling or institutional or parental care after reaching adulthood (11%). In 78% the outcome was believed to be good, with no sequelae in 19%, occasional TIAs or headaches in 33%, and mild intellectual or motor impairment in 26%. These authors suggested that hypertension and early age of onset were the best predictors of poor prognosis. The experience of 23 patients from the Hospital for Sick Children in Toronto [13], 15 of whom were surgically treated, suggested that early treatment offered the best chance to prevent neurologic deficit. All 8 patients who were not surgically treated either died or had fixed neurologic deficits, with or without intellectual impairment. Another study has reviewed intelligence in 20 children who were subjected to cerebral blood flow (CBF) measurements and surgery for moyamoya disease [14]. Preoperative intelligence quotients (IQs) were lower in older patients and those with more severe reductions of CBF. It was found that IQ increased significantly in 10 of the 15 patients who were assessed postoperatively; 3 remained the same and 2 deteriorated. With such information, early intervention aimed at the preservation of blood flow seems indicated.

EDAS is presently considered the surgical procedure of choice for childhood moyamoya disease [7,15], the other procedures have several important drawbacks. STA-MCA anastomosis, although technically feasible even in the small child, is difficult to perform with small donor and recipient vessels and thus prone to failure in children. EMS involves the application of temporalis muscle directly to the cortex. The incidence of seizures has been reported to be higher after this procedure, possibly be-

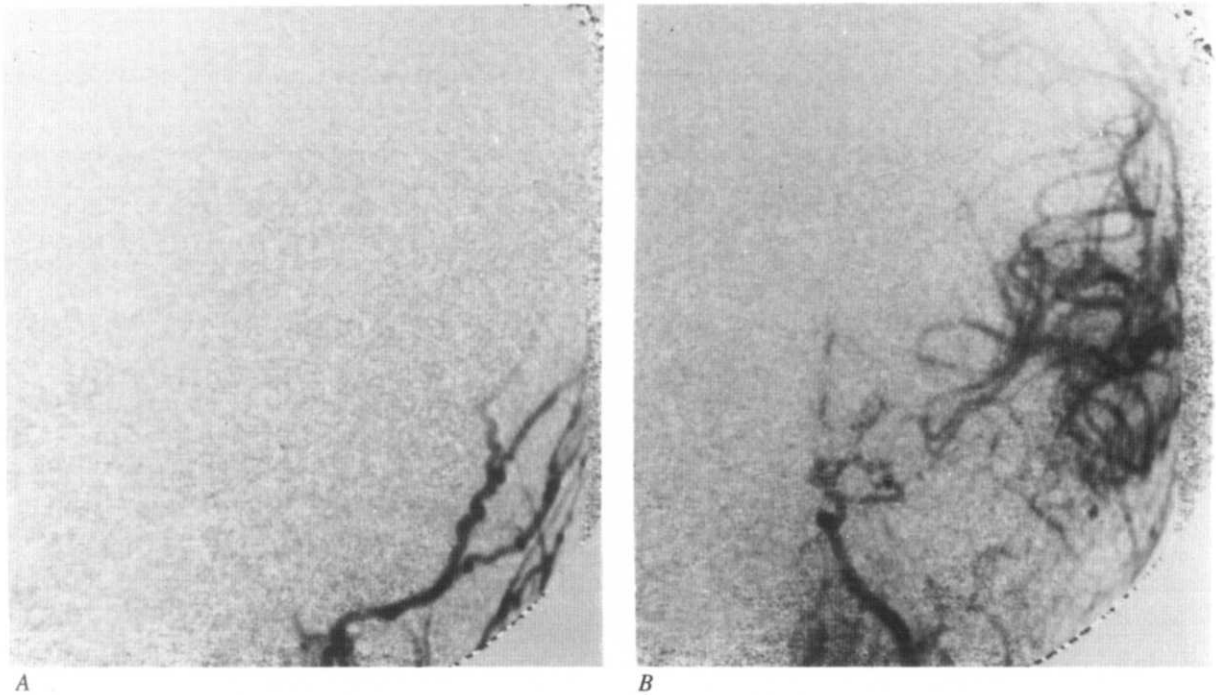

Figure 2. (A) AP projection of left CCA angiogram post-EDAS of the anterior and posterior branches of the left superficial temporal artery (STA) (Patient 5). Early phase. Note preferential, early filling of both branches of the STA which are now quite dilated. (B) Same projection; later phase. Note that the ICA is still occluded. However, there is excellent filling of the middle cerebral artery territory from the EDAS.

cause of electrical activity within the muscle itself. The mass effect of the muscle on the cortex is also potentially deleterious and the cosmetic result is not often very good because a large craniectomy is required. Fugita et al. [16] attempted to determine if there was a difference in outcome between EMS and EDAS for moyamoya disease. In a retrospective study, they were able to conclude that cerebral revascularization was more rapid with EDAS. Also, the angiographic and CBF assessments indicated better results with the EDAS. Combination of EDAS and EMS in the same procedure has recently been reported with good outcome [17]. Transplantation of autogenous omentum onto the surface of the brain to promote the development of collateral blood flow has been successfully used [15,18], but is technically more difficult and perhaps more prone to failure. A recently reported case of omental transposition from the abdomen to the head on a subcutaneously tunneled pedicle was successful in an adult with moyamoya disease [19]. Endo et al. [20] have reported a simple procedure that can be used to augment collateral flow. In 6 patients either at the time of EMS or subsequent to it, they fashioned frontal burr holes and carefully opened the dura and arachnoid. Increased flow to the underlying cortex was documented in 5 patients who were studied postoperatively. All of the alternative procedures, despite their disadvantages, have been used successfully and can be considered when EDAS fails [21,22].

While all of our patients remained clinically stable after the EDAS procedures, the precise role of the surgery in the overall outcome is not certain. Only 3 of the 6 EDAS operations that were evaluated postoperatively by conventional angiography showed definitive filling of the internal cerebral circulation via the EDAS, even though the transposed STAs were all patent. It is of interest why the other patients who did not revascularize via the EDASs did well. In 2 of the 3 studies, there was angiographic evidence of regression of the disease, with either less stenosis or recanalization of occluded vessels at the base of the brain. It is unlikely that the improvement of the angiographic picture of the moyamoya disease in these patients was secondary to the EDAS procedures. Collateral circulation probably did not develop in these patients because the brain was not ischemic enough to elaborate the necessary angiogenic factors that incite such a phenomenon. Rooney et al. [23] recently reported 5 children with moyamoya disease who underwent EDAS. All became neurologically stable thereafter, even though development of increased collateral flow through the EDAS was only demonstrated in 3 of the 5 patients and angiographic progression of the disease in 4 patients. It is not possible to predict which patients will progress clinically and angiographically. Given the possibility of neurologic deterioration in children with this disease, we would recommend early treatment of all symptomatic patients with angiographically demonstrated disease.

In conclusion, we believe that EDAS is a safe, technically simple procedure for the treatment of moyamoya disease in children. Benefit from the procedure is likely greatest early in the course of the disease, when the potential for preventing permanent neurologic deficits is greatest. As it is not possible to detect that subset of patients who require surgery, we recommend early EDAS in all children with angiographic evidence of moyamoya disease. Other procedures, including STA-MCA anastomosis, EMS, omental transfer, and simple burr holes remain available for refractory cases.

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