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527 Surgical Treatment of Moyamoya Disease: Operative Technique for Encephalo-duro-arterio-myo-synangiosis, Its Follow-up, Clinical Results, and Angiograms
Clinical Study

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ABSTRACT: MOYAMOYA SYNDROME IS defined as the development of collateral anastomosis pathways at the base of the brain, associated with chronic progressive stenosis of the carotid fork. Both reconstructive vascular surgery and conservative strategies are used to treat this syndrome, but the latter cannot prevent the disease from progressing. We describe the procedure of encephalo-duro-arterio-myo-synangiosis (EDAMS), and report the results in 17 patients (28 sides) who underwent EDAMS. The clinical symptoms of moyamoya disease include transient ischemic attacks, reversible ischemic neurological deficits, stroke, seizures, Gerstmann's syndrome, involuntary movements, or mental retardation resulting from the lack of cerebral blood flow. The clinical results of EDAMS were poor in one patient, fair in two, good in five, excellent in eight, and fair on one side and excellent on the other side in one patient. Postoperative angiograms showed widespread collateral circulation on the ischemic brain surface in patients undergoing EDAMS.

KEY WORDS: Encephalo-duro-arterio-myo-synangiosis; Encephalo-duro-arterio-synangiosis; Middle meningeal artery; Moyamoya disease; Revascularization

Moyamoya disease often can be diagnosed clinically, but an accurate diagnosis must include angiographic images. Specific radiological features appearing on angiograms include the following: 1) either marked stenosis or complete occlusion of the distal internal carotid arteries; 2) poorly visualized proximal portions of the anterior and middle cerebral arteries; 3) well-developed vascular networks at the base of the brain (basal moyamoya); and 4) well-developed collaterals between leptomeningeal and cortical arteries as well as among cortical arteries.

The typical clinical symptoms of moyamoya disease include repeated ischemic attacks in children and subarachnoid or intracerebral hemorrhage in adults. Symptoms appear most often in the first and third decades of life.

A variety of operative procedures have been described to surgically treat pediatric patients with moyamoya disease. Anastomosis between the superficial temporal and middle cerebral arteries

(STA-MCA) and encephalo-duro-arterio-synangiosis (EDAS) have been performed at our hospital with favorable results. In our experience, however, some patients were found to lack an arterial branch suitable for anastomosis or required more abundant cerebral blood flow to avoid persistent ischemic attacks even after STA-MCA anastomosis or EDAS.

To deal with these situations, we developed a new procedure, encephalo-duro-arterio-myo-synangiosis (EDAMS), which appears to have more satisfactory results. In this report, we describe the operative method of EDAMS and evaluate the postoperative angiographic and clinical findings in 17 patients undergoing EDAMS.

CASE SUMMARIES

Reconstructive surgery by EDAMS has been indicated for the symptomatic side of moyamoya disease since 1984. Between 1984 and 1990, 17 patients (13 children, 4 adults, Patients 1-17) with moyamoya disease underwent EDAMS. EDAMS was performed on 28 sides in 17 patients (average age, 6 yr and 9 mo; range, 11 mo-46 yr). Among the 17 patients undergoing EDAMS, 10 had transient ischemic attacks, 2 had transient ischemic attacks followed by a minor complete stroke, 2 had strokes after a seizure, 2 had strokes alone, 1 had a stroke after reversible ischemic neurological deficit, and 1 had a seizure (Patient 2, right side). The interval between the onset of clinical symptoms and surgery averaged 2 years and 2 months (range, 2 mo-14 yr).

Operative technique of EDAMS and intraoperative findings

After anesthesia is administered, the patient is placed supine, with the head slightly elevated and turned 90 degrees along the horizontal plane. A small pad is placed under a shoulder of patients with limited neck mobility. The superficial temporal artery is palpated and marked with a Doppler ultrasound probe, and the patient's head is placed in a three-point Mayfield head rest.

An incision is made just behind the hairline, proceeding in a curvilinear fashion from a point directly above the pupil to a point just above the zygoma in front of the tragus. The incision should lie posterior to the parietal branch of the superficial temporal artery, allowing this artery to be preserved with the skin flap (Fig. 1). The incision continues posteriorly to reach the temporal fascia. Particular care must be taken to avoid injuring the superficial temporal artery.

Hemostasis is obtained by applying curved hemostats to the margins of the incision, and a pair of scissors is inserted into the plane between the galea and the temporal fascia. The galea is cut parallel to the artery at a distance of 3 to 5 mm, so that a strip of galea is attached to the artery over its entire exposed length. The parietal branch of the superficial temporal artery is carefully dissected and isolated from the inner surface of the skin and the fascia below, especially at the point where the superficial temporal artery crosses the incision. The skin flap is reflected laterally, and the residual temporal fascia and muscle

are opened with cautery. With a periosteal elevator, the periosteum is swept off the skull to the edge of the orbit. The muscle, including the area attached to the superior temporal line, is reflected from the skull. After coagulation of all bleeding points, the soft tissue flap is folded over a sponge to prevent ischemic compression. This flap is held in position with silk sutures placed in the muscle and attached by rubber bands to the drapes.

Two burr holes are made in the inferior region of the superior temporal line and the temporal bone. A craniotomy should be performed without injuring the middle meningeal artery. Five or six dural stay sutures are made through the same drill holes used to fasten the bone flap. Some of these sutures are tied to the temporal muscle over the inferior margin.

The dura is incised with two or three linear cuts, and the branches of the middle meningeal artery are preserved. Leaf-like cuts of the dura, about 5 mm in length, are then made, and the arachnoid membrane is incised along the cortical sulci. The outer surfaces of the leaf-like cuts are folded in and attached to the surface of the brain. The stripped superficial temporal artery with its galea is laid on the surface of the brain, and its edges are fixed with surgical strings to the margin of the dural incision (Fig. 2). The temporal muscle is pulled over the brain surface and roughly sutured to the opened dural edges (Fig. 3).

To prevent leakage of cerebrospinal fluid after surgery, a drainage tube is placed between the temporal muscle and the surface of the brain. Additional pieces of bone are removed where the inlaid arterial pedicle enters and exits the skull. The bone flap is repositioned and fastened into place, and the scalp wound is sutured in layers.

In our series, thickened arachnoid membranes and edema were found during EDAMS. Because of the well-developed pial vessels, the surface of the cortex was red. In most cases, the middle cerebral artery had few branches and constricted arteries or only white strings were frequently observed over the cortical surface or in the sulci.

RESULTS

The patients undergoing EDAMS were followed both clinically and angiographically. For the 17 patients undergoing EDAMS, the clinical follow-up period averaged 3 years and 2 months (range, 10 mo-5 yr, 10 mo). The patients' postoperative neurological status was divided into four categories: poor, fair, good, and excellent.

The poor category included patients who had neurological deficits after surgery. Patient 10 (both sides) had a poor outcome that worsened after surgery.

The fair category included three patients whose preoperative neurological symptoms were unchanged after EDAMS. (In Patient 5, there was an excellent outcome on the left side.)

The good category included five patients whose minor neurological deficits were improved to a certain degree or were reduced in severity. The occurrence of transient ischemic attacks was markedly reduced after surgery in these patients.

The excellent category included eight patients in whom neurological deficits recovered completely and transient ischemic attacks disappeared.

Postoperative angiograms were obtained from 16 sides in 10 patients undergoing EDAMS. The features of the postoperative angiogram were classified into two groups: extensive and localized. Extensive collaterals showed abundant collateral circulation from the external carotid system to the cortical brain surface, with retrograde refilling of the proximal segment of the middle cerebral artery through the collaterals to a point immediately distal to the occlusion. The abnormal fine vascular networks were also observed through these vascular channels, and other branches of the middle cerebral artery were visualized. Localized collaterals showed the part of the territory of the middle cerebral artery that was perfused from the external carotid system. In this series, all sides but one were extensive.

At our institute, before 1984, EDAS as reconstructive surgery for moyamoya disease was performed. Angiograms performed after surgery showed that the ratio of extensive collateral vessels in patients undergoing EDAMS was higher than that of patients undergoing EDAS. The clinical results of the patients undergoing EDAMS were also superior in comparison with those of EDAS.

CASE REPORT

Patient 6

A 3-year-old boy suddenly developed hemiplegia on the right side, lasting a few minutes. After the initial event, transient ischemic attacks consisting of motor weakness in the right extremities and left upper extremity occurred 10 times a day while he was eating hot food. At his admission 2 years after the first event, his neurological examination disclosed no deficit except for the transient ischemic attacks. Cerebral angiography revealed the peculiar features of typical moyamoya disease (Fig. 4). EDAMS was performed bilaterally--on the right side at first and on the left 1 month later. The transient ischemic attacks decreased gradually and did not recur 1 year after EDAMS. Since then, no additional neurological symptoms have occurred. Cerebral angiography taken 8 months after surgery showed abundant collateral flow on the brain's surface after EDAMS (Fig. 5).

DISCUSSION

Several surgical approaches to reconstruct collateral circulation have been suggested as successful in treating moyamoya disease. These include STA-MCA anastomosis^(1,2,5,11,17), cervical carotid sympathectomy⁽¹²⁾, superior cervical ganglionectomy⁽¹²⁾, omental transplantation^(6,16,18), encephalo-myo-synangiosis (EMS)^(3-5,14), and EDAS^(7,8). These approaches are generally divided into two groups: direct anastomotic bypass, such as STA-MCA anastomosis, and indirect nonanastomotic bypass, such as EMS, EDAS, and other procedures.

Recently, STA-MCA anastomosis, EMS, and EDAS have become more popular treatments for moyamoya disease. Although STA-MCA anastomosis has resulted in satisfactory outcomes in

some patients with moyamoya disease, this procedure involves some potential disadvantages. First, the arteries of some children may be too small for anastomosis. Secondly, some patients have worsened after anastomosis because of the temporary stasis of blood needed in the cortical artery for direct anastomosis^(10,18).

Because of these disadvantages, EMS and EDAS were developed to promote the spontaneous development of anastomoses between vessels of the internal and external carotid circulation. As to the middle meningeal artery as a donor artery, one report describes the successful direct anastomosis of the middle meningeal artery to a cortical branch of the middle cerebral artery⁽⁹⁾. Another describes a new surgical procedure in which the dura mater containing the meningeal artery is inserted into the ischemic brain of a patient with arterial thrombosis⁽¹⁵⁾.

However, a technique to promote circulation from the middle meningeal artery to the brain's surface has not yet been reported for moyamoya disease. To our knowledge, the EDAMS procedure is the first report of making the most use of the middle meningeal artery to promote collateral circulation in moyamoya disease.

The development of EDAMS was an attempt to increase collateral circulation from both the middle meningeal artery by folding in the outer surface of the dura and the stripped superficial temporal artery, with surrounding galea applying directly to the surface of the brain. We expected this maneuver to create EDAS. Besides these two arteries, the temporal muscles including deep temporal arteries were placed over the surface of the brain. This maneuver was expected to create EMS. The distal end of the superficial temporal artery was left intact, not only to secure abundant and constant flow through the artery, but also to preserve the extracranial collaterals fed at a distance by this artery. The arachnoid membrane was opened to promote collateral channels between the donor arteries and the cortical arteries, which was thought to be better when the pia mater was widely exposed and had direct contact with the donor arteries. After surgery, many anastomoses may develop between the external carotid system and the middle cerebral artery over the wide surface of the brain where the frontotemporal craniotomy was performed.

One child who had a poor outcome (Patient 10) had reversible ischemic neurological deficit followed by complete minor strokes a few weeks after surgery as a result of precipitating factors, such as crying or swimming. The hyperventilation that occurs in these activities decreases PaCO₂, subsequently reducing cerebral blood flow and triggering transient ischemic attacks or minor strokes.

Tagawa and associates⁽¹³⁾ reported that mean PaCO₂ during hyperventilation or crying was 29.0 ± 3.5 mm Hg in young patients with moyamoya disease and that a reduction of cerebral blood flow would play the most important role in ischemic manifestations in these patients with a sensitive vasoconstrictive response to hyperventilation. Karasawa and colleagues⁽⁵⁾ reported that, in patients

undergoing EMS, the branches of the middle cerebral artery were seen through the muscle flaps on angiograms taken 1 week after surgery. They also reported that a considerably larger number of the branches of the middle cerebral artery were seen through the muscle flaps 3 months after surgery. In some patients in our series, it has been confirmed angiographically that the anastomotic channels begin to develop 2 weeks after EDAMS. In the majority of cases, well-developed anastomotic channels were observed on angiograms 3 months postoperatively when the transient ischemic attacks tended to cease and neurological deficit improved gradually. Therefore, patients with moyamoya disease should avoid behavior such as crying or swimming for a few months after surgery until collateral circulation between the external carotid system and the brain's surface is accomplished.

A definite conclusion about whether to choose EDAMS or another procedure to treat moyamoya disease is unclear. We believe, however, that the clinical results of EDAMS show a greater reduction of the ischemic attacks in moyamoya disease, and angiograms taken after surgery show extensive collateral vessels on the ischemic brain. Therefore, EDAMS seems to be one of the more effective treatments for moyamoya disease.

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COMMENTS

The authors present a new technique for

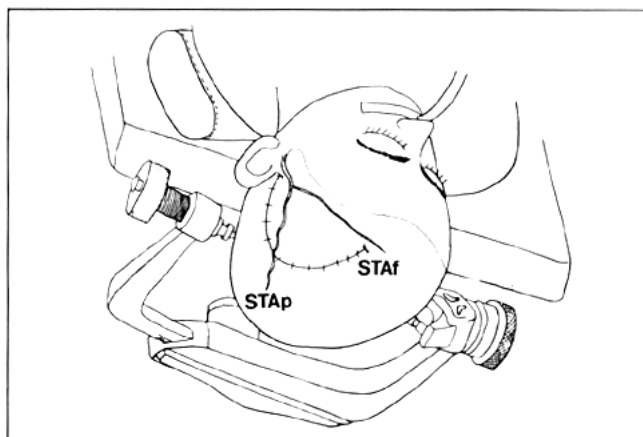


Figure 1. Illustration showing patient's position and the skin incision. *STAf*, frontal branch of the superficial temporal artery; *STAp*, parietal branch of the superficial temporal artery.

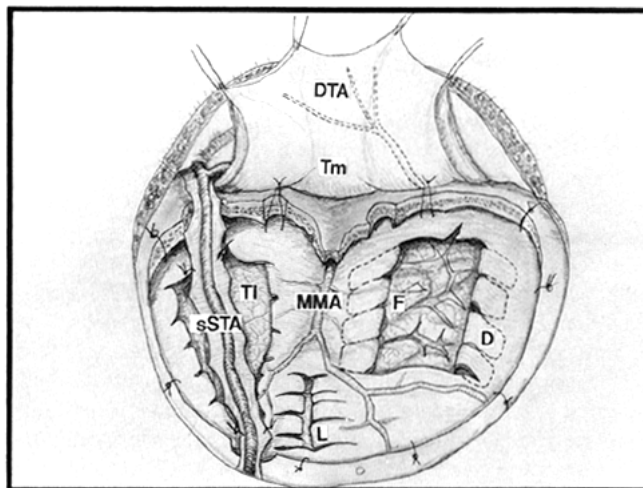


Figure 2. Operative illustration. The skin flap and temporal muscle are reflected temporally, and a frontotemporal craniotomy is performed. *DTA*, deep temporal artery; *D*, folded in dura mater; *F*, frontal lobe; *L*, leaf-like cuts of dura mater; *MMA*, middle meningeal artery; *sSTA*, stripped superficial temporal artery; *Tl*, temporal lobe; *Tm*, temporal muscle.

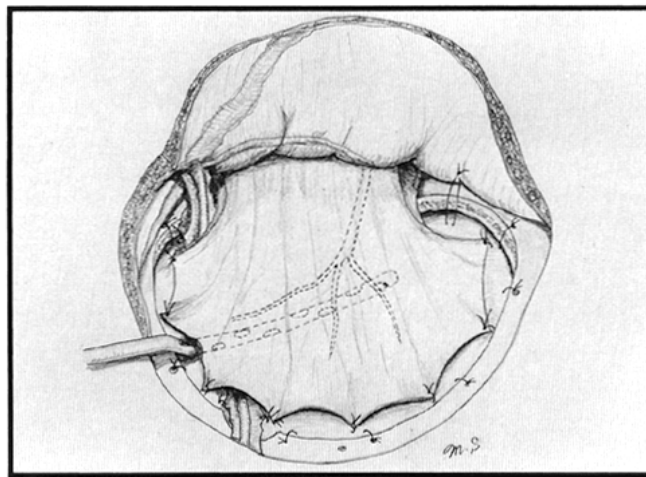


Figure 3. Operative illustration. The brain's surface is covered with the temporal muscle.

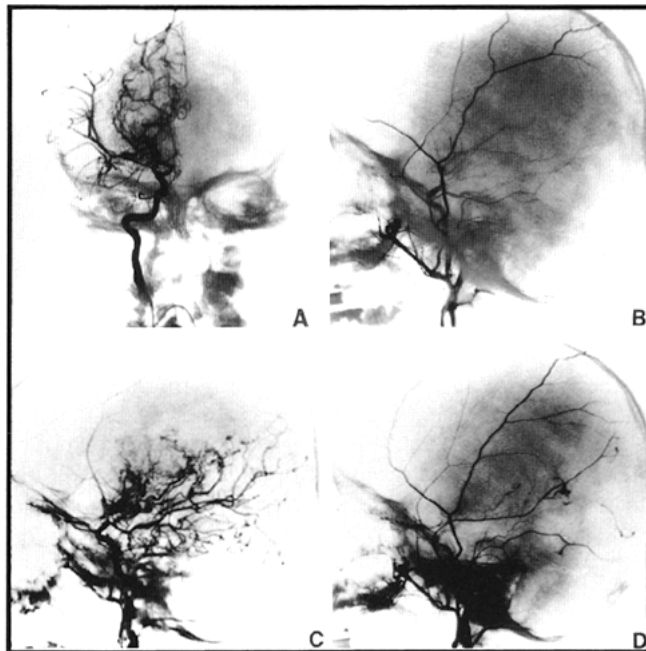


Figure 4. *A*, angiogram of the right internal carotid artery, antero-posterior view. Note the narrowing of the artery at the supraclinoid portion, the stenosis of the proximal segment of the middle and anterior cerebral arteries, and the abnormal vascular networks in the region of the basal ganglia. *B*, angiogram of the right external carotid artery. Note the absence of leptomeningeal collaterals from the external carotid artery. *C*, angiogram of the left common carotid artery, lateral view. Note the complete occlusion of the artery at its supraclinoid portion, the filling of the anterior cerebral artery through abnormal vascular networks (basal moyamoya), and the nonfilling of the middle cerebral artery. *D*, angiogram of the left external carotid artery shows a small leptomeningeal anastomosis in the territory of the middle cerebral artery from the external carotid artery.



Figure 5. External carotid angiograms after EDAMS show numerous collaterals on the surface of the brain through the dilated superficial temporal artery, middle meningeal artery, and deep temporal artery. The patency of the distal superficial temporal artery is seen in the angiogram of the right external carotid artery. *A*, right side; *B*, left side.