Dural inversion procedure for moyamoya disease

Technical note

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Moyamoya disease is a chronic occlusive cerebrovascular disease characterized by the progressive stenosis of the proximal internal carotid artery and its branches with the gradual development of moyamoya vessels or basilar collateral branches. When it occurs in childhood, the patient generally presents with cerebral transient ischemic attacks, seizures, headache, or completed strokes; when it occurs in the adult, the patient usually experiences intracranial hemorrhage. Although many idiopathic cases are seen, associated conditions may include neurofibromatosis, Down’s syndrome, sickle cell anemia, meningitis, prior radiation to the brain, craniopharyngioma, optic glioma, and connective tissue diseases. A strong familial tendency seems to exist in Japan. The disease progresses in six stages, with the development of external carotid circulation collateral branches as the internal carotid stenosis proceeds. It is believed that ischemic symptoms develop if collateral vessel formation is not able to keep up with the progressive reduction in internal carotid blood flow.

Surgical attempts at treatment have centered around using the unaffected external carotid system to provide blood flow to the ischemic brain during this tenuous phase of collateral vessel development. Direct anastomosis between the superficial temporal artery (STA) and a branch of the middle cerebral artery (MCA) has been used with limited acceptance. Other indirect techniques have relied on the tendency of the ischemic brain to attract collateral vessels from any available source. Probably the most accepted technique in the Western hemisphere is the encephaloduroarteriosynangiosis (EDAS), in which the STA is dissected from the scalp and directly apposed to the surface of the brain. Reports concerning the success of this procedure are mixed, however, with some patients developing relatively poor collateralization.

Although the middle meningeal artery (MMA) and its branches are in proximity to the ischemic cortex, collateral vessels from this source are not seen to as great a degree as might be expected in moyamoya disease. It has been recognized that the inner meningeal layer of the dura must serve as an impediment to collateral vessel ingrowth from the extremely vascular outer periosteal dura into the brain. If this outer layer could be directly applied to the cortical surface while leaving its blood supply intact, vascular ingrowth could possibly be enhanced. The current report describes a technique in which the MMA is used to promote revascularization over a large surface area of the brain.

Illustrative Case

This 6-year-old right-handed girl presented with an acute onset of severe left-sided hemiparesis without speech involvement. The diagnostic workup included magnetic resonance (MR) imaging, which showed evidence of cerebral ischemic changes involving a large por-
tion of the right hemisphere in the MCA distribution (Fig. 1). Cerebral angiography demonstrated stenosis of the supraclinoid internal carotid artery on the right side as well as stenosis of the proximal left anterior cerebral artery and MCA just beyond the internal carotid bifurcation (Fig. 2). No significant collateral vessels from the external carotid circulation were seen on either side, although deep basilar collateral branches were noted bilaterally. The child slowly improved over the subsequent week with mild residual hemiparesis and was taken to surgery for treatment of presumed moyamoya disease.

Operative Technique

Attention was first directed to the right hemisphere, which was the side of the ischemic injury. Endotracheal induction of general anesthesia without hyperventilation was used and the patient was placed supine with the head turned full lateral. The entire right frontoparietotemporal area was shaved. The posterior branch of the STA was located with Doppler ultrasonography and its location was carefully marked by lightly scratching the scalp. After preparation, the scalp was incised only through the dermis and, using blunt spreading with a hemostat, we were able to identify the vessel. The dissection over the artery was continued distally toward the vertex, incising the skin superficial to the hemostat. After several centimeters of vessel had been thus exposed, the skin edges were retracted and the galea was incised 5 mm on either side of the artery, thereby separating the vessel with its galeal cuff from the scalp. This tissue was then retracted to the side and a full-thickness scalp incision was made that extend-
Dural inversion for moyamoya disease

Results of Illustrative Case

Postoperative angiography had not been planned until 6 months after the second procedure. However, the child had a subtle episode of slight right-sided hemiparesis 4 months after left hemisphere revascularization, which cleared completely after several minutes. For this reason, angiography was performed at that time.

Internal carotid angiography revealed progression of the proximal stenosis bilaterally. External carotid angiography demonstrated rather extensive revascularization of the MCA distribution bilaterally, originating almost exclusively from the MMA (dural inversion procedure) rather than from the STA (EDAS procedure) (Figs. 5 and 6). No problems arose after the examination and the child is presently asymptomatic.

Discussion

Many different surgical procedures have been proposed for the treatment of moyamoya disease. Direct STA–MCA anastomosis was first used by Yaşargil to treat a 4-year-old child with this disorder.9 This technique has not gained wide popularity, however, because the small size of the vessels in children make this procedure technically difficult. In addition, the temporary occlusion of the MCA branch that is required during the anastomosis can interrupt the leptomeningeal collateral branches seen in these patients and result in a stroke.15,22 Attention has therefore turned mainly to indirect methods of revascularization.

Encephalomyosynangiosis (EMS), described by Henschel,9 involves placement of the inner surface of the temporalis muscle directly against the brain under a temporal craniotomy. This has not been widely used in the Western hemisphere, but some series from Japan report good results.12,14 Complications include postoperative seizures, which are possibly caused by mechanical traction on the brain from muscle contraction or even from direct spread of myoelectric activity to the cortex.7,18 Subdural fluid accumulations can also be a problem following this procedure, and a large cosmetic defect is left in the temporal fossa. Other techniques have therefore become more popular.
The unexpected observation by both Ausman and colleagues\(^2\) and Spetzler, et al.,\(^3\) that spontaneous collateral vessels would grow into the brain from an STA placed directly on the cortex without direct anastomosis led to the development of the EDAS procedure for moyamoya disease by Matsushima, et al.\(^4\) This has been the most popular technique because of its simplicity, the lack of a need for temporary occlusion of an MCA branch, the ability to plan the craniotomy so as to avoid disrupting already present collateral vessels, and because several scalp arteries may be available for use, including the occipital branches. Results, however, have proved to be less than ideal, with recurrent symptoms and poor postoperative angiographic results in close to 50\% of children treated.\(^11,12,16,21\) Adelson and Scott\(^4\) modified the EDAS procedure by opening the arachnoid membrane over the cortex using microtechnique and suturing the STA adventitia directly to the pia (pial synangiosis). This resulted in excellent postoperative angiographic results in 85\% of cases, although two unstable patients among the 34 treated suffered perioperative strokes.

Other attempts at achieving better revascularization have used combined techniques. Matsushima and associates\(^12\) have reported improved results after performing STA–MCA anastomosis in conjunction with the EMS procedure. However, the problems associated with these techniques, described previously, are not avoided. Kinugasa and colleagues\(^6\) have combined EDAS and EMS while opening the dura in multiple locations and have demonstrated collateral ingrowth from multiple sources. Once again, however, the complications associated with EMS must be considered.

It is apparent that the MMA has a powerful potential to offer collateral blood flow to an ischemic cortex. It has been noted under these circumstances that placement of a simple burr hole with opening of the dura will allow ingrowth of vessels from the dural edges.\(^4\) Placement of multiple burr holes has, therefore, been used in the treatment of moyamoya disease with some success.\(^7\) A related technique has recently been described by Kashiwagi, et al.\(^8\) After separation of the layers of the dura, reapproximation of the outer layer over small areas of the cortex results in the ingrowth of a moderate number of collateral branches derived from middle meningeal sources.

Although the dura clearly has the capacity to revascularize the ischemic brain, observations in patients with moyamoya disease reveal that this occurs to a very limited extent in the natural course of the disease.\(^8\) However, if one examines the histological structure of the dura, an explanation is suggested. The dura of the spinal canal is composed of only one layer, which is poorly vascularized. At the foramen magnum, the dura is joined by the inner periosteal layer of the skull, which fuses with the inner or meningeal dura and becomes the outer periosteal layer of the dura. This outer layer is richly vascularized by multiple arterial branches and is considered by some authors not to represent actual dura.\(^9\) We propose that the inner layer of the dura acts as a natural anatomical barrier between the internal carotid territory and the external carotid circulation, thus explaining the relative lack of natural collateralization between the MMA circulation and the ischemic brain in this disease. The technique described in this report, therefore, circumvents this barrier through inversion of the dural layers.

The procedure illustrated here has a number of possible advantages over previously described techniques. It is extremely simple, adding only a few minutes to a standard EDAS procedure. The EDAS can be performed at the same time along one edge of the exposure. The complications of EMS are not a concern and the problems associated with a direct vascular anastomosis are avoided. Because a large dural flap can be fashioned, collateral branch ingrowth over a very large surface area of the ischemic cortex can be achieved. In addition, dural flaps could be placed anywhere over the brain to provide revascularization. For example, dural flaps near the midline could be constructed, based near the sagittal sinus, and then inserted into the interhemispheric fissure to supply vasa vasorum to the anterior cerebral artery territory. Recurrent symptoms in this distribution have been reported following a standard EDAS procedure, with angiograms showing poor flow to this region from the graft.\(^4\) Finally,
because the technique does not require sacrifice of large meningeal branches, preexisting collateral branches are not disrupted.

**Conclusions**

We propose that this technique be added to the standard EDAS or pial synangiosis procedures in the treatment of moyamoya disease. Initial results are promising, the theoretical basis seems sound, and the risks are minimal. Further study, including many more procedures and years of follow-up review, will, of course, be required before its effectiveness can be accurately determined. Finally, in view of the extensive degree of revascularization seen with this procedure, one might consider investigating its use in the treatment of patients with other disease processes, such as internal carotid occlusion from atherosclerosis that is not amenable to carotid endarterectomy.

**References**


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