Enlarged Encephalo-Duro-Myo-Synangiosis Treatment for Moyamoya Disease in Young Children

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■ OBJECTIVE: **To retrospectively evaluate the midterm therapeutic effect of enlarged encephalo-duro-myo-synangiosis (EDMS) for moyamoya disease (MMD) in young children.**

■ METHODS: **Seventy-seven children diagnosed with MMD by digital subtraction angiography (DSA) or magnetic resonance angiography (MRA) were treated between January 2011 and December 2014 in our center. Their clinical features, imaging, and operative reports were analyzed.**

■ RESULTS: **Four patients presented with intracerebral hemorrhage, whereas 73 presented with ischemic symptoms. Revascularization procedures were performed unilaterally on 11 left hemispheres and 9 right hemispheres, and 57 had bilateral surgeries. The average length of surgery was 143 ± 24 minutes, with 28 ± 9 mL of blood loss. The mean period of follow-up was 28.43 ± 15.31 months. Cerebral blood flow increased 3 months after the operations in the previously affected regions. Collateralization from the deep temporal artery, superficial temporal artery, and the middle meningeal artery was found by DSA or MRA. In cases where single-photon emission computed tomography was obtained, it demonstrated better perfusion postoperatively. The ischemic symptoms were relieved in 118 (88.06%) hemispheres, and remained stable in 5 (3.73%) hemispheres. There were 12 radiographic cerebral infarctions (8.96%) within 1 month postoperatively.**

■ CONCLUSIONS: **Enlarged EDMS is safe and effective for MMD in young children. Extensive and multilayered revascularization could significantly preserve neurologic function. The long-term effect on posterior circulation disease development needs further investigation.**

INTRODUCTION

Moyamoya disease (MMD) is a spontaneous stenoocclusive process involving the internal carotid artery and its branches, resulting in the formation of disorganized compensatory vascular collaterals.[1] As an acquired occlusive disease of the cerebral vessels, the pathogenesis of MMD remains poorly understood.[2] MMD has been reported in many countries, including China, with a high prevalence in areas of Eastern Asia and a predilection for children.[3-5] In recent years, the incidence of MMD has increased with the increasing use of noninvasive vascular techniques, including computed tomography (CT) angiography and magnetic resonance angiography (MRA), and improved clinical understanding and treatment of MMD. A variety of procedures for treatment of MMD have been performed in different medical centers throughout the world. For the first time in the literature, to our knowledge, we introduce enlarged encephalo-duro-myo-synangiosis (EDMS) with large craniotomy for revascularization of 77 MMD children, together with a midterm follow-up.

METHODS

Clinical Data

We reviewed 77 patients who were treated for MMD at Children's Hospital of Fudan University between January 2011 and December

Key words
- Encephalo-duro-myo-synangiosis
- Indirect anastomosis
- Moyamoya disease
- Pediatric stroke
- Revascularization

Abbreviations and Acronyms
CT: Computed tomography
DSA: Digital subtraction angiography
EDMS: Encephalo-duro-myo-synangiosis
MCA: Middle cerebral artery
MMD: Moyamoya disease
MRA: Magnetic resonance angiography
SPECT: Single-photon emission computed tomography
TIA: Transient ischemic attack

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2014. These patients were diagnosed with MMD by digital subtraction angiography (DSA) or MRA, and according to the guidelines for the diagnosis and treatment of MMD as described in 2012 by Hashimoto et al.[6] Cases with moyamoya syndrome with autoimmune disease, Down syndrome, hyperthyroidism, neurofibromatosis, vasculitis, or other associated diseases were excluded from our cohort. Preoperative evaluation included CT scan and/or magnetic resonance imaging, single-photon emission computed tomography (SPECT), DSA, and neuropsychologic testing.

Patients were categorized according to the classification described by Matsushima et al.[7] There were 26 type I cases (transient ischemic attack [TIA]), 10 type II cases (frequent TIA), 6 type III cases (TIA-infarction), 4 type IV cases (infarction-TIA), 27 type V cases (infarction), and 4 type VI cases (hemorrhage). DSA was performed preoperatively in 53 patients, demonstrating 15 Suzuki stage II, 30 stage III, and 8 stage IV.[1]

Side of Operation

After detailed clinical and radiographic assessment, including evaluation of the bilateral internal carotid arteries and vertebral arteries with MRA or DSA and 99mTc-ECD SPECT perfusion, we performed revascularization procedures on the more severely ischemic hemisphere. If the patient presented with a hemorrhage, we operated on the affected hemisphere initially. If the degree of ischemia was similar on both sides, the dominant hemisphere was treated first, and the contralateral hemisphere was operated on approximately 2 months after the initial surgery. Seventy-seven patients underwent 134 enlarged EDMS totally.

Operation Technique

In positioning for a modified pterional approach, the head was turned 45° and the ipsilateral shoulder was elevated with a shoulder roll. Given cosmetic considerations, we planned the trailing extent of incision behind the upper pinna, and the superior aspect approximately 1–1.5 cm above the superior temporal line. This incision allows for the superficial temporal artery to be more easily protected during dissection. Also, the larger craniotomy allows for an increased surface area to attach the temporalis muscle (Figure 1). After mobilizing the scalp, the temporalis muscle and periosteum were dissected from the skull, forming a temporalis muscle flap, which contains the deep temporal artery network.

A frontotemporal craniotomy was elevated surrounding the pterion extending 1 cm above the superior temporal line. The bone at the sphenoid ridge was thinned. The whole length of the middle meningeal artery was dissected out its boney canal, while protecting the anastomosis between the dura and cortical artery formed in stage V and stage VI patients. The dura was then tacked up to the skull, and the trunk and branches of the middle

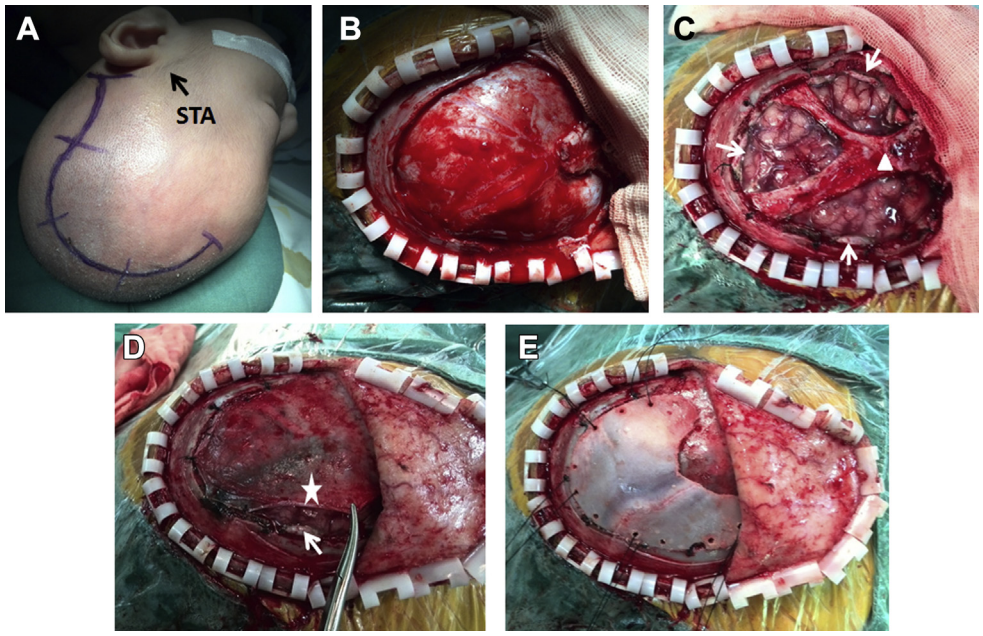

Figure 1. Surgical procedure pictures from a 2-month-old patient. (**A**) The modified pterional incision curved posteriorly to protect the superficial temporal artery. (**B**) The temporalis muscle and periosteum were dissected from the skull, and the bone flap, which was larger than regular encephalo-duro-myo-synangiosis can expose the middle cerebral artery distribution. (**C**) Maintenance of the trunk and branches of the middle meningeal artery (*white triangle*), with reflection of the left dura (*white arrows*) onto the brain surface. (**D**) Free temporalis muscle (*white asterisk*) was placed on the brain surface and sutured to the marginal dura (*white arrow*). (**E**) Because young children have a thin temporalis muscle with minimal mass effect, we fixed the bone flap in situ. STA, superficial temporal artery.