

Indirect Revascularization Techniques for Treating Moyamoya Disease

Neil N. Patel, DO, MBA^{a,*}, Francesco T. Mangano, DO^a,
Paul Klimo Jr, MD, MPH, Maj, USAF^b

KEYWORDS

- Indirect revascularization techniques
- Moyamoya disease • Omental transplantation
- Multiple burr holes • EMS • EDAS • Pial synangiosis
- Combined

Moyamoya disease (MD), originally described by Takeuchi in the 1950s, is a disease that results in cerebral ischemia from progressive bilateral stenosis of the internal carotid arteries. Children with MD typically present with symptoms of ischemia, while adult patients tend to present with intracranial hemorrhage. Definitive diagnosis originally required cerebral angiography, which demonstrated the stenotic arteries and the classic puff of smoke, the description given to the slowly filling basal perforator angiogenesis. As noninvasive vasculature imaging modalities such as magnetic resonance (MRA) and computed tomography angiography (CTA) have dramatically improved over the last 10 years, this technology has assumed a greater role in both pre- and post-treatment.

MD is a surgical disease and as such, there are a multitude of different techniques. Medical management most often is used to complement surgery but not replace it. The evolution of surgical procedures has been described by Reis and colleagues.¹ None of the surgical procedures described to treat MD are curative, but rather preventative in nature. The true underlying etiology of moyamoya still is not fully understood. The

article by Smith and Scott in this volume of *Neurosurgery Clinics of North America* discussed the epidemiology, presentation, diagnosis and prognosis of moyamoya disease.

Surgical revascularization techniques for moyamoya are divided into direct, indirect and combined approaches. In general, indirect techniques require less time and have a decreased overall risk than direct revascularization approaches.^{2–4} Direct techniques can be difficult to perform because of the small diameter of donor or recipient vessels, and an increased risk of middle cerebral artery (MCA) cerebrovascular accident, or intracerebral hemorrhage.⁵ Direct revascularization techniques, however, appear to provide improved results over a shorter time frame⁶ and may lead to more robust revascularization as suggested by the literature review conducted by Fung and colleagues.⁷

This article reviews numerous indirect revascularization procedures, focusing more on the technical aspects, as there have been numerous more outcome-focused articles published recently. A PubMed and Medline review of the English literature for moyamoya was conducted.

^a Division of Pediatric Neurosurgery, Cincinnati Children's Hospital Medical Center, 3333 Burnet Avenue, MLC 2019, Cincinnati, OH 45229, USA

^b Neurosurgery, 88th Medical Group, SGOS/SGCXN, 4881 Sugar Maple Dr Wright-Patterson Air Force Base, OH 45433, USA

* Corresponding author.

E-mail address: npatel777@aol.com

This yielded 26 articles specifically focused on indirect revascularization techniques. The procedures reviewed include

- Cervical sympathectomy (CS)
- Omental transplantation (OT)
- Multiple burr holes (MBH)
- Encephalo-myo-synangiosis (EMS)
- Encephalo-arterio-synangiosis (EAS)
- Encephalo-duro-synangiosis (EDS)
- Encephalo-myo-arterio-synangiosis (EMAS)
- Encephalo-duro-arterio-synangiosis (EDAS)
- Encephalo-duro-arterio-myo-synangiosis (EDAMS)
- Encephalo-duro-galeo (periosteal)-synangiosis (EDGS)
- Multiple combined indirect procedures (MCI)
- Indirect combined with direct procedures (I+D).

CERVICAL CAROTID SYMPATHECTOMY AND SUPERIOR CERVICAL GANGLIONECTOMY

CS and ganglionectomy were the first surgical procedures used in the treatment of moyamoya by Suzuki and Takaku in 1969.⁸ Previous research had demonstrated loss of adrenergic axons within the walls of arteries and arterioles in dogs after a superior cervical ganglionectomy. The authors theorized that this would promote dilation of cerebral arteries and thus improve the collateral circulation. They performed this procedure in 10 children. Even though clinical symptoms initially improved, the progression of moyamoya was not halted both clinically and angiographically. Although the procedure now can be performed using a thoracoscopic approach, it has fallen out of favor because of the development of alternate revascularization techniques with better results and less surgical morbidity.

OMENTAL TRANSPLANTATION

OT first was described in a case report by Karasawa in 1978.⁹ The patient presented with bilateral motor ischemic events and blindness. The procedure consisted of a fronto-parieto-occipital skin incision, preserving the superficial temporal artery, and associated superficial temporal vein. The antero-inferior border of the craniotomy was used for insertion of the omentum. Via a midline laparotomy, a large 13 cm × 13 cm segment of omentum containing perforating vessels of the gastroepiploic artery and vein was isolated. An end-to-end anastomosis between the superficial temporal artery and vein to the gastroepiploic artery and vein, respectively, was performed. Durotomy and

arachnoid incision permitted spreading of the transplanted omentum to the cortical surface. The patient improved clinically over the next 2 years without any new cerebrovascular events. Havlik and colleagues¹⁰ described a procedure whereby a pedicled graft of omentum was tunneled subcutaneously to the cerebral cortex in a patient who failed a direct (superficial temporal artery [STA]-MCA) bypass. The rate for patency of the omental graft has been reported to be as high as 70% over a 2-year period.¹¹

Touho and colleagues¹² performed OT in five children who failed prior EMS, EDAS, or STA-MCA direct bypass surgeries. All patients experienced resolution of their neurologic deficits after several months. Similar results also were achieved by Karasawa and colleagues.¹³ His study involved 30 children, of whom 19 patients underwent omental transplant using the anterior cerebral artery distribution, and 13 patients underwent omental transplant using posterior cerebral artery distribution. All these patients except two in the posterior artery distribution group demonstrated neurologic improvement and increase in collateral vessels on follow-up angiography.⁵

MULTIPLE BURR HOLES

MBH were developed as a treatment for MD after the discovery of neovascularization around burr holes performed for ventriculostomies. Endo and colleagues¹⁴ first reported this in 1984 in a pediatric patient who required bifrontal ventriculostomies. Using this observation as a starting point, Endo performed the first multiple burr hole procedure for MD in 1986. Angiographically adequate neovascularization at 12 months follow-up was noted with no ischemic events.¹⁴

More recently this procedure was reported on 15 patients and 24 hemispheres by Sainte-Rose and colleagues.¹⁵ The patient is placed supine with head flexed to expose the entire calvarium if bilateral procedures are necessary. The incision is bi-coronal in a zigzag form for cosmesis. The exposure allows access to the frontal, parietal, temporal, and partial occipital regions bilaterally. If a unilateral procedure is indicated, the head is rotated to the contralateral side, and a T-shaped incision is fashioned.

The galea is dissected carefully, in a meticulous fashion to preserve vascularity of the scalp. The periosteum should remain intact, to preserve the vessels that will form future collateral networks. The reason that complete subperiosteal dissection is not preferred is mainly to prevent postoperative collection of cerebrospinal fluid (CSF), and also to minimize blood loss. Many triangular incisions are

made in the periosteum and lifted as small flaps to expose the skull (**Fig. 1**). These openings are placed roughly 3 cm apart, covering the appropriately targeted vascular territories, and 3 cm from the midline to avoid bleeding resulting from injury to the superior sagittal sinus or the associated draining veins. Burr holes then are made at each exposed area (**Fig. 2**). The dura is opened through each burr hole using the microscope to avoid middle meningeal arterial branches. The arachnoid and pia are carefully opened while preventing any bleeding. Hemostasis is obtained by using cottonoid patties and gentle saline irrigation, as cautery is to be avoided to preserve any potential anastomotic vessels. The periosteal flap that elevated was previously is now placed in contact with the exposed parenchyma through each burr hole. The galea is carefully repositioned, with a two-layered skin/galea closure. A compressive dressing is applied for 4 days to facilitate hemostasis and limit facial swelling. Postoperative skull radiographs and a CT scan are obtained to exclude complications and assess placement of burr holes as a baseline, for comparison to follow-up angiograms and to assess for hemorrhage and CSF collections. None of their patients suffered ischemic events postoperatively, and angiography revealed excellent revascularization of the hemisphere by the external carotid system. Subcutaneous CSF collections occurred in 5 of the 18 procedures, but were treated successfully by tapping and wrapping the head; one patient also required a temporary lumbar drain.

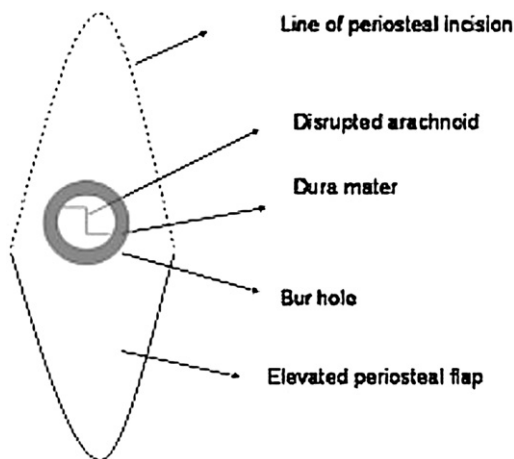


Fig. 1. Multiple burr hole technique (MBH). Schematic diagram depicting the location of the periosteal incision, elevation of the flap, and opening of the dura. (Reprinted from Sainte-Rose C, Oliveira R, Puget S, et al. Multiple burr hole surgery for the treatment of moyamoya disease in children. *J Neurosurg* 2006;105(6):439; with permission.)

The most significant benefit of this technique is the ability to use it anywhere on the cranium. It can be, and often is, combined with other procedures, direct or indirect, and it is technically simpler than other approaches. Another potential benefit as discussed by Baaj and colleagues⁵ is that certain patients can undergo this procedure under local anesthesia, thus avoiding the risks of general anesthesia.

ENCEPHALOMYOSYNANGIOSIS

Encephalomyosynangiosis (EMS) first was described by Karasawa and colleagues¹⁶ in 1975 and represented the first indirect revascularization technique for the treatment of MD. It was developed after reports in 1950 from Henschen, who demonstrated revascularization from muscle flaps after cerebral injuries.¹⁷ EMS requires opening the dura and arachnoid layer of the cerebral cortex, then placing a vascularized section of temporalis muscle directly over it (**Fig. 3**). The muscle then is sutured or tacked up to the superior aspect of the durotomy, to prevent mobility of the muscle, and resultant mass effect. The dural flap is sutured back in place over the muscle, allowing a portion of the temporalis to enter the inferior aspect of the durotomy. A small craniectomy at the site where the temporalis enters the calvarium may be necessary to prevent ischemic compression of the muscle. Over time, collateral angiogenesis will develop between the vascular-rich muscle and the ischemic underlying cerebral tissue. One must be careful to allow the inflow vessels of the transplant to be patent and perfusing through the temporalis muscle by not applying too much pressure on it with the head wrap.⁵ Touho and colleagues¹⁸ have described a gracilis muscle transplantation technique, unilaterally and bilaterally, to revascularize frontal and occipital regions with good results.

Takeuchi and colleagues¹⁹ performed EMS on 13 patients and 24 total hemispheres. Seventy-five percent of these patients achieved revascularization in more than one third of the MCA distribution. In addition, seven of these patients presented with transient ischemic attacks (TIAs) preoperatively, and four of seven had complete resolution of the TIAs, the remainder having significant decreases in the frequency of TIAs postoperatively. Similar good results were reported by Caldarelli and colleagues.²⁰ Disadvantages of EMS include the need for a larger craniotomy, and reported postoperative complications include seizures, mass effect from the muscle, and associated increase in intracranial pressure. A case report by Touho²¹ described calcification of the graft with significant

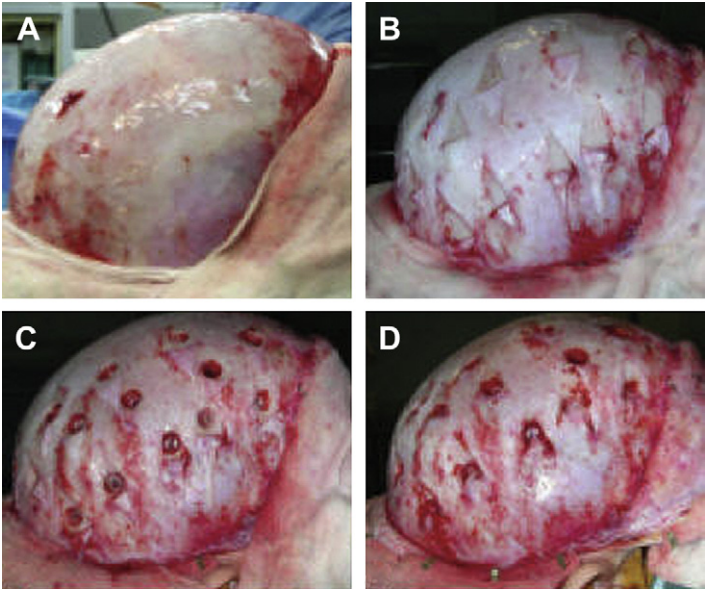


Fig. 2. MBH technique. After the scalp is reflected (A), multiple triangular incisions in the periosteum are made (B). The burr holes then are made in the exposed skull, approximately 3 cm apart (C). The dura is opened, followed by the arachnoid and pia. The triangular flap of periosteum then is placed into the burr hole, making direct contact with the exposed cortex (D). (Reprinted from Sainte-Rose C, Oliveira R, Puget S, et al. Multiple bur hole surgery for the treatment of moyamoya disease in children. *J Neurosurg* 2006;105(6):438; with permission.)

mass effect and ischemia 6 years after EMS. This complication resulted in removal of the graft. The use of EMS now is incorporated more often into a combination of procedures and used less frequently as a single operation.

ENCEPHALOARTERIOSYNGIOSIS

Encephaloarteriosynangiosis (EAS) is mainly an intermediate procedure most often used as part of an EDAS or EDAMS. It, however, has been described as a single procedure in the past when a direct anastomosis between the STA and the MCA cannot be achieved due to MCA insufficiency,

especially in posthemorrhagic presentation of moyamoya disease.²² Because it is commonly used as part of the EDAS or EDAMS, the results on its isolated use are not readily available. The technique involves first carefully dissecting the STA. It then is retracted softly while a temporal craniotomy and durotomy are performed. The STA then simply is placed in contact with the brain. Using this technique, Touho²³ recently reported complete resolution of TIAs in 19 of 21 operative sides, with the remaining two sides having marked decrease in TIA frequency. Houkin and colleagues²⁴ found that neovascularization from the superficial temporal artery in EAS often was insufficient and that the deep temporal artery (temporalis muscle) and middle meningeal artery (dura) were better sources. Thus EAS, like EMS, often is used in combination with other procedures both direct and indirect.

ENCEPHALODUROSYNGIOSIS

Encephalodurosangiosis (EDS) is also an intermediate procedure that is used as part of the EDAS or the EDAMS. EDS involves the direct placement of dura with its blood supply (usually the middle meningeal artery) on the pial surface. The same principle can be applied in a more localized fashion by a burr hole with a dural incision as described with the MBH technique. EDS is used most commonly to generate collaterals to ischemic anterior cerebral artery (ACA) territories.²⁵ This procedure, like the EMS and the EAS mentioned previously, can be used in combination with other procedures both direct and indirect. As

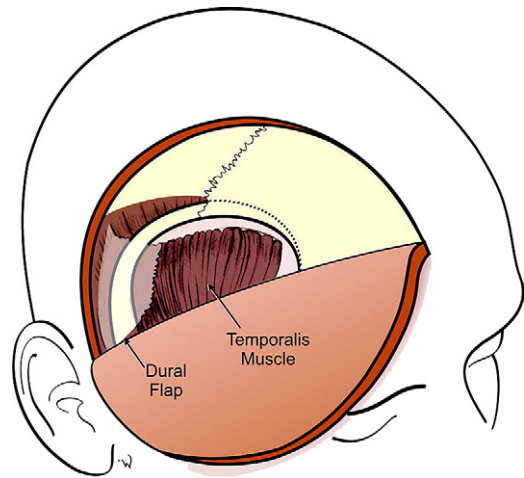


Fig. 3. Encephalomyosynangiosis (EMS). The temporalis muscle is freed from its fascia and placed directly on the exposed surface of the brain. The dura is reflected posteriorly.

an isolated procedure, EDS has not been studied in depth, and results of its isolated use are not available.

ENCEPHALOMYOARTERIOSYNANGIOSIS

Encephalomyoarteriosynangiosis (EMAS) is also basically a sum of its parts. Matsushima and colleagues²⁶ explained the procedure in detail. They described it in variants of a frontal EMAS. In the frontal EMAS, the anterior superficial temporal artery is exposed using a cut-down technique, and then divided distally to make a muscle flap attached to it. The skin incision is extended to create a horseshoe skin flap with the epicenter in the anteroinferior skull, at the origin of the STA. A craniotomy then is performed and the dura resected. The temporalis muscle flap with the STA branch attached is sutured to the dural edge to make contact with the frontal cortical surface.²² This technique can be applied to posterior and middle cerebral circulation as well using the posterior branch of the superficial temporal artery and the posterior auricular or the occipital artery as needed. In Matsushima's study, 10 patients and 16 hemispheres were studied. The results of this technique yielded vascular collateral formation in 88% of the procedures as evidenced by angiogram at 25 months after surgery.²² EMAS was used as a combined technique in this study and therefore clinical outcomes were not directly correlated to revascularization from EMAS alone.

ENCEPHALODUROARTERIOSYNANGIOSIS

Encephalo-duro-arterio-synangiosis (EDAS) is probably the most commonly performed indirect procedure for moyamoya. This being the case, there are many variants and adjuncts put onto the technique by individual surgeons for additional support for revascularization. This includes most commonly the following combinations

- EDAS + pial synangiosis
- EDAS + dural inversion
- EDAS + split dura technique.

The origin of this procedure dates back to 1979.²⁷ At that time, the best current surgical therapy used was STA-MCA bypass with associated high frequency of neurologic decline and seizures in pediatric patients. The first patient was a 9-year-old boy with moyamoya disease. Over the course of 4 years, the authors reported 75% success rates and decreased complication rates compared with direct STA-MCA bypass.^{26,28}

The procedure can be completed in many different ways, but the basics remain the same.

The procedure is best described in detail by Kashiwagi and colleagues.²⁹ The patient is in supine position, with head turned to the side facing anesthesia and away from the surgeon and held on a head rest. The anterior and posterior branches of the STA are palpated and marked with a Doppler ultrasound probe. The skin incision is made along the course of the parietal (posterior) branch of the STA, starting at a point 2 cm above the zygoma in front of the tragus and extending vertically and posteriorly to a point 10 cm above the zygoma, then curving anteriorly to a point 2 cm lateral to the midline at the level of the hairline. The STA is dissected at its proximal portion and separated from the inner surface of the skin.

A plane between the subcutaneous fat and the STA is dissected, and the skin incision is performed above this plane so as not to encroach upon the STA. The galea is incised parallel to the STA to provide a cuff of tissue over the exposed length of the vessel. The STA then is dissected carefully away and isolated from the fascia below, including the point at which the artery crosses the skin incision. The skin incision then is extended to the frontal region just behind the hairline. The skin flap is turned over a moist sponge used to prevent ischemic compression. The temporalis muscle is separated from the bone with a periosteal elevator and retracted posteriorly with the STA. Three burr holes are made. The first one, inferior temporal, is under the proximal portion of the STA, and the second one, superior temporal, under the distal portion of the STA. The third one, anterior frontal, is located 2 cm lateral to the midline, just in front of the coronal suture. The craniotomy is performed by connecting these burr holes, taking care not to injure the dural vasculature. A Penfield dissector is helpful in dissecting dural attachments from the inner aspect of the skull. A linear durotomy is made along the course of the STA, and the galeal cuffs on the STA strip are sutured to the edges of the dural incision to hold the STA in place over the cortex using interrupted silk or nylon sutures to complete the EDAS (**Fig. 4**). Isono and colleagues³⁰ performed EDAS on 16 hemispheres in 10 patients and found that it produced more robust neovascularization than EMS or EDAMS. Fujita and colleagues³¹ found more revascularization from the external carotid artery in sides treated with EDAS than with sides treated with EMS.

In an EDAS with pial synangiosis, the dura is not opened in a linear fashion, but is opened into at least six flaps to increase the surface area of dura exposed to the pial surface while sparing any of the dural vessels, specifically the middle meningeal artery (MMA) and its branches (**Fig. 5A**).³² This is thought to increase the

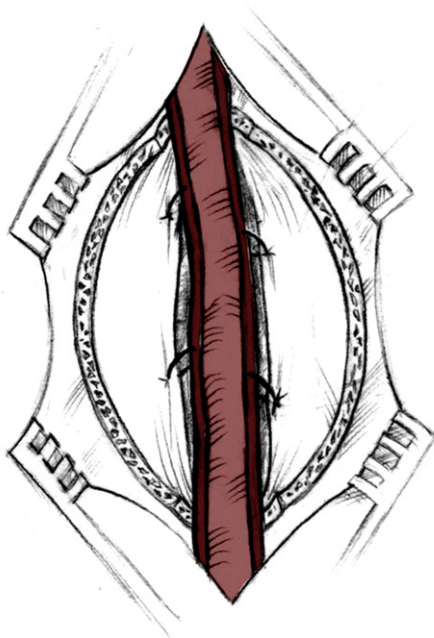


Fig. 4. Encephaloduroarteriosynangiosis (EDAS). The dissected superficial temporal artery is attached to the edges of the dura with interrupted silk or nylon sutures.

formation of collateral vessels from the dural vascular supply. The arachnoid is opened widely over the surface of the brain exposed by the durotomy. Then, the donor STA is sutured directly to the pial surface using four to six interrupted 10–0 nylon sutures placed through the donor vessel adventitia and the underlying pia (see **Fig. 5B**). A large piece of gel foam is used to cover the durotomy defect, and the bone flap is placed softly over it to avoid compression of the donor artery. This procedure

aims at opening the arachnoid, which is believed to be a barrier to the ingrowth of new blood vessels onto the brain parenchyma.²⁰

In 2004, Scott and colleagues³³ reported their extensive experience with pial synangiosis over a 17-year period at the Children’s Hospital Boston. There were 143 patients (89 females and 54 males), with stroke being the presentation in 67.8%, and 43.4% experiencing one or more TIAs preoperatively. Average follow-up was 5.1 years. Within the first 30 days following 271 craniotomies for pial synangiosis, there were 11 episodes of stroke and three severe TIAs. In 126 patients followed for more than 1 year, four suffered a late-onset stroke; one suffered a severe reversible TIA, and two experienced persistent TIAs. In 46 patients followed for more than 5 years in whom the major initial presentation was stroke alone, only two late-onset strokes have occurred.

In an EDAS with dural inversion described by Dausser and colleagues³⁴ the craniotomy is completed after the posterior branch of the STA is dissected and protected. This craniotomy is completed so that the posterior branch of the MMA is visualized in the center of the craniotomy, so planned from the preoperative angiogram. The dura then is cut on either side of the MMA branch, thereby creating two rectangular dural flaps, each of which is situated around the artery itself. Care must be taken not to compromise flow through the vessel. The dural flaps then are inverted so that one passes over the artery and the other beneath it (**Fig. 6**). This maneuver allows the outer, richly vascularized layer of the dura to have direct contact with the surface of the cortex while maintaining both inflow and outflow through the middle meningeal artery supplying the inverted dura. The flaps are held into position loosely with absorbable suture, taking care not to twist and stenose the

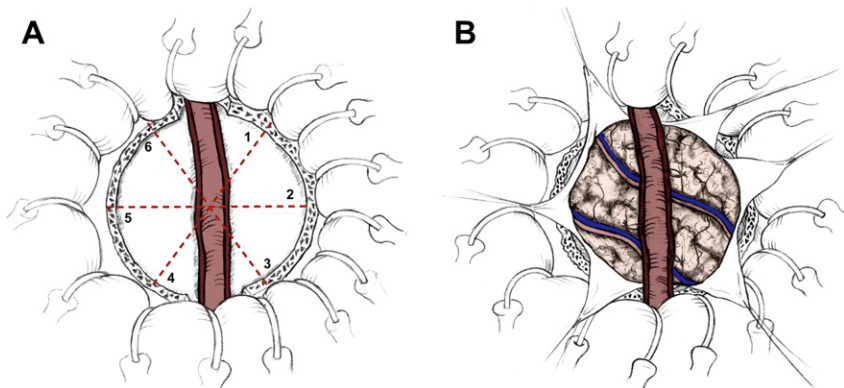


Fig. 5. Pial synangiosis. Rather than a linear opening as performed in EDAS, the dura is opened in 6 leaflets, thus maximizing the potential in growth of blood vessels from the cut edges of the dura (A). With the dural leaflets under fish hook-type retractors and the arachnoid widely opened, the STA is secured to the pia (B).

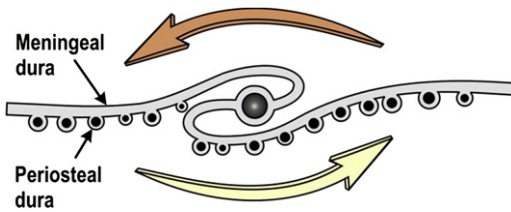


Fig. 6. Dural inversion technique. This cross-sectional schematic diagram shows how the dural flaps on either side of the middle meningeal artery are inverted, thus placing the vascular rich outer surface (periosteal dura) in direct contact with the cortex.

MMA branch. The superficial temporal artery then is sutured to the dura along the posterior margin of the exposed area under the dural flap, allowing this vessel to be in contact with the surface of the brain, as is done in the a standard EDAS procedure.

In an EDAS with the split-dural technique described by Kashiwagi and colleagues,³⁵ a similar approach is taken. After the STA is sutured to the linear durotomy avoiding the MMA, the course of the dural arteries, especially that of the anterior and posterior branches of the MMA, is inspected thoroughly. An H-shaped linear incision is made carefully through the outer layer of the dura adjacent to the MMA and the STA, not encroaching upon either vessel. This is done usually between the parietal STA and the posterior branch of MMA, and just anterior to the anterior branch of the MMA. The outer layer of the dura is separated, or split, from the inner layer and turned over. The inner layer is incised along the same H-shaped configuration as the initial incision and then folded over into the subdural space so that this split surface is attached to the cortical surface. This in-folding of the inner layer also exposes a window of cortical surface. The outer layer is closed with interrupted silk sutures, so that the internal surface of the outer layer is attached to the cortical surface. Bleeding from the dural incision or separation is controlled with oxycellulose and cottonoid patties but with minimal use of bipolar coagulation so as not to lose the blood supply to the dura. The results for this technique demonstrate 85% disappearance in TIAs by 1 year, and complete loss of TIAs by 1.5 years. Muto and Oi recently described a similar dural splitting-type technique called intradural arteriosynangiosis in which the supratemporal artery is anastomosed to the inner layer of the dura mater and surrounded by the outer layer as a sandwich with a blunt procedure of dural layer separation.³⁶

The EDAS has been a widely used and successful technique that allows for the use of multiple variations. For this reason, as well as its relative

ease and safety, it is the prime indirect revascularization technique used for pediatric moyamoya disease.

ENCEPHALODUROARTERIOMYOSYNANGIOSIS

EDAMS is an extended technique from the EDAS and the EMS that uses the temporalis muscle's deep temporal artery (DTA), the STA, and the MMA to act as adjuncts to facilitate neovascularization (**Fig. 7**). The EDAMS technique is one of the most powerful indirect techniques available to create neovascularization but requires all three donor areas to be adequate in size.

It was proposed and developed in 1984 by Kinugasa and colleagues³⁷ to combine aspects of all the indirect revascularization techniques. The procedure is summarized best by the technique described by Kim.³⁸ A skin incision is made along the parietal STA branch and the distal frontal branch of the STA in question mark shape in the fronto-parieto-temporal (FPT) region. After reflecting the skin flap anteriorly, both branches of the STA with attached strip of galea of 10 to 15 mm are carefully dissected from the pericranium and the fascia below. The underlying temporalis fascia and muscle and frontal pericranium are incised in a T-shape and elevated from the skull so that the temporalis has a posterior and an anterior cuff. A wide craniotomy is made in the fronto-parietal-temporal region while protecting the middle meningeal artery and other dural vessels as well as the dissected STA branches. The dura is opened in both the frontal and temporo-parietal regions alongside the MMA, creating two flaps with the MMA free-floating in the center. The stripped frontal STA branch is placed on the surface of the frontal region, and only the proximal and the distal portions of its galea are roughly sutured with one or two stitches to the opened dural margin. The parietal branch of the STA, however, is anastomosed near the largest cortical branch of the MCA on the temporo-parietal region. This posterior opened dural edge is multiply incised and folded inward into the subdural space to lay on the exposed brain surface. The split temporalis muscles are placed on the intact arachnoid membrane of the frontal and temporo-parietal cortex, respectively and sutured to the adjacent dura to facilitate CSF permeation. After rongeur the lower parts of the squamous temporal bone as well as the bone flap itself to prevent compression of the temporalis muscle pedicle, the bone flap is replaced and secured to the cranium and the scalp is closed. The results for the EDAMS technique have demonstrated on average 85% revascularization rate over 2 years.²⁵

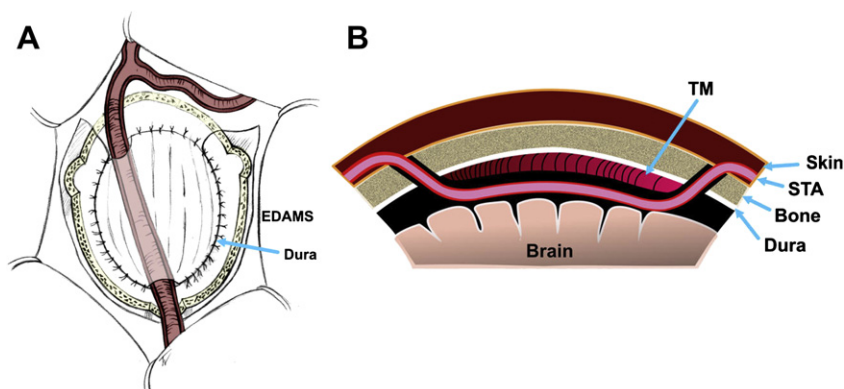


Fig. 7. Encephaloduroarteriomyosynangiosis (EDAMS). This technique takes advantage of all local vascularized structures. The parietal branch of the STA is placed between the parenchyma and the temporalis muscle; suturing of the dura mater to the temporalis muscle has been done (A, B).

In 2006, Ozgur developed a slightly modified version.³⁹ This version involves complete excision of the remaining dural flaps. In addition, Ozgur also recommend incising the pia–arachnoid surface and adding several small incisions overlying and parallel to the sulci, while avoiding vascular structures. After the arachnoid incision, the parenchyma is punctured, and then irrigated to eliminate any micro/macro hemorrhages. The temporalis muscle can be put back sparingly (or just the posterior cuff placed) if the muscle is too bulky to avoid causing mass effect and other complications associated with EMS.

ENCEPHALODUROGALEOSYNANGIOSIS

Encephalodurogaleo (periosteal)-synangiosis (EDGS) has been an adjunct that has been used alongside an EDAS or an EDAMS. It incorporates multiple incisions and has been shown to be beneficial mainly for ACA territory ischemia. Kawamoto and colleagues,⁴⁰ who coined the term galeoduroencephalosynangiosis (GDES), first described it in 2000, but this term later was switched to the reverse anatomic name (EDGS) to correlate with the remainder of indirect techniques. This technique has been described using two different methods. In a unilateral case of moyamoya disease, a small elliptical incision is fashioned just off midline with the apex toward the midline. Two incisions can be made in a curvilinear fashion just paramedian from two burr holes.⁴¹ After a burr hole is made, the galea is dissected off the curvilinear skin flap, and a durotomy is performed. The galea is placed over the medial cortex and into the interhemispheric fissure. The galea then is tacked to the dura, and the skin is closed. Another method described by Kim and colleagues⁴² that can be used for bilateral procedures begins with

a horizontal S-shaped incision, 2 cm anterior to and parallel with the coronal suture crossing over the midline (Fig. 8A). Although the authors use the term galea, it is actually the periosteum that then is incised in a zigzag pattern, creating two rectangular flaps. A 4 × 8 cm craniotomy, crossing the superior sagittal sinus, is made. The dura then is incised separately on both hemispheres, and if preferred, the arachnoid membrane also may be incised for added benefit. The apex of the galeal/periosteal flaps is inserted as deeply as possible into the interhemispheric fissure and sutured to the dura (see Fig. 8B). Revascularization (20-month angiogram) rate for this technique was reported at 83% in a total of 159 patients.

COMBINED INDIRECT PROCEDURES

Multiple combined indirect procedures use many of the previously mentioned techniques to obtain the widest and safest coverage and allow revascularization of oligemic cerebrum. These techniques have included many different combinations, some of which were mentioned in the previous sections, but the more commonly described are EDAS plus bifrontal EGDS,⁴² bilateral or unilateral concomitant EDAS,⁴³ EDAS plus EMS plus EMAS,⁴⁴ EDAS plus EMS only,^{26,45,46} parietal EMAS with a frontotemporal EDAMS,⁴⁷ MBH with EMS, and EDAS and pial synangiosis.³ The results for these procedures vary depending on specific technique combinations and the authors.

COMBINED INDIRECT AND DIRECT PROCEDURES

Indirect combined with direct procedures are performed more commonly in adults with moyamoya disease mainly because of the difficulty in

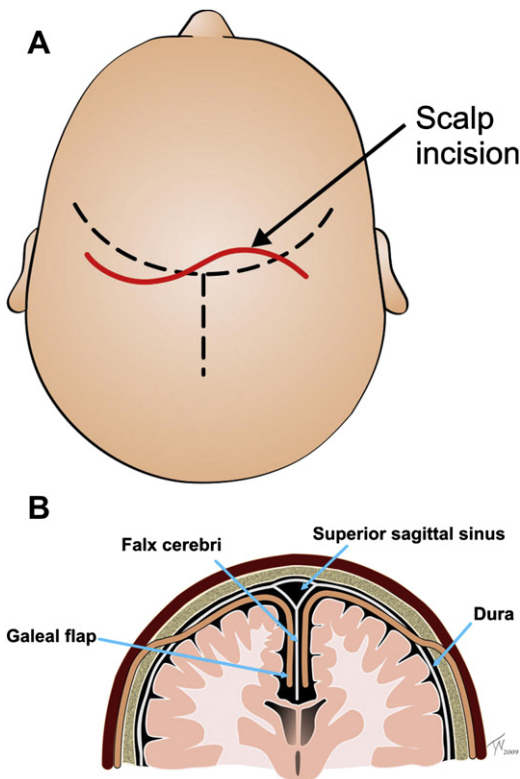


Fig. 8. Encephalodurogaleo (periosteal)-synangiosis (EDGS). An S-shaped scalp incision is made centered in the midline to access the interhemispheric space bilaterally (A). The periosteal (or what Kim and colleagues⁵¹ call the galea) flaps then are advanced as far as possible into the interhemispheric space (B).

obtaining adequate vessel caliber for pediatric patients for direct anastomosis, as well as frequent complications in pediatric patients as opposed to adults. In the most recent review of direct versus indirect bypass, Starke and colleagues⁴ state that there is evidence that both the direct and indirect bypass techniques are effective means of revascularization and reduce the incidence of ischemic events in patients with moyamoya disease. In their experience, they feel that the best collateral formation resulted from EDAMS procedures given the wider coverage to both MCA and ACA distributions. They recommended these procedures primarily for pediatric patients and combined direct and indirect bypass in adult patients. Veeravugu and colleagues⁴³ reviewed outcomes of many indirect and direct procedures and concluded that for children, there is as of yet no compelling evidence to support one technique over another. The authors did find that indirect procedures alone were less efficacious in elderly patients possibly because of an age-associated reduction in angiogenic capability.

Of the indirect/direct procedure combinations available, the most common is the STA-MCA anastomosis with an EMS using the same craniotomy.^{2,48} Other combinations have included STA-MCA anastomosis with EDAS, MMA-MCA anastomosis with EDAS, occipital artery-PCA anastomosis with EGDS,⁴⁹ STA-MCA anastomosis with EDAMS.⁵⁰ The results of these techniques are variable. Specific techniques using direct anastomoses will be discussed in a separate article by Steinberg elsewhere in this issue.

SUMMARY

Revascularization surgery for moyamoya disease in pediatric patients is recommended to prevent ischemia or hemorrhage. For this reason, it is crucial to minimize the perioperative risks. This is accomplished most commonly with indirect techniques. To date, there have been no controlled trials to determine the efficacy of surgical revascularization and to establish specific benefit/risk ratios. Also, no standard surgical approaches for the treatment of moyamoya disease have been established as evidenced by the multitude of techniques that have been described over the last 50 years. Therefore the decision ultimately is based on the surgeon's experience and favor of technique.

Based on the literature review completed, not one of the articles individually discusses all the techniques identified and illustrated in this article. The procedures detailed in this article represent most of the indirect techniques being performed (or that have been performed) for moyamoya disease.

REFERENCES

1. Reis CV, Safavi-Abbasi S, Zabramski JM, et al. The history of neurosurgical procedures for moyamoya disease. *Neurosurg Focus* 2006;20(6):E7.
2. Matsushima T, Inoue T, Ikezaki K, et al. Multiple combined indirect procedure for the surgical treatment of children with moyamoya disease. A comparison with single indirect anastomosis and direct anastomosis. *Neurosurg Focus* 1998;5(5):e4.
3. Smith JL. Understanding and treating moyamoya disease in children. *Neurosurg Focus* 2009;26(4):E4.
4. Starke RM, Komotar RJ, Connolly ES. Optimal surgical treatment for moyamoya disease in adults: direct versus indirect bypass. *Neurosurg Focus* 2009;26(4):E8.
5. Baaj AA, Agazzi S, Sayed ZA, et al. Surgical management of moyamoya disease: a review. *Neurosurg Focus* 2009;26(4):E7.

6. Golby AJ, Marks MP, Thompson RC, et al. Direct and combined revascularization in pediatric moyamoya disease. *Neurosurgery* 1999;45(1):50–8 [discussion: 58–60].
7. Fung LW, Thompson D, Ganesan V. Revascularisation surgery for paediatric moyamoya: a review of the literature. *Childs Nerv Syst* 2005;21(5):358–64.
8. Suzuki J, Takaku A. Cerebrovascular moyamoya disease. Disease showing abnormal net-like vessels in base of brain. *Arch Neurol* 1969;20(3):288–99.
9. Karasawa J, Kikuchi H, Kawamura J, et al. Intracranial transplantation of the omentum for cerebrovascular moyamoya disease: a two-year follow-up study. *Surg Neurol* 1980;14(6):444–9.
10. Havlik RJ, Fried I, Chyatte D, et al. Encephalo-omental synangiosis in the management of moyamoya disease. *Surgery* 1992;111(2):156–62.
11. Gerber M, Spetzler RF. Burr holes for moyamoya disease. *Barrow Quarterly* 2002;18:22–4.
12. Touho H, Karasawa J, Tenjin H, et al. Omental transplantation using a superficial temporal artery previously used for encephaloduroarteriosynangiosis. *Surg Neurol* 1996;45(6):550–8 [discussion 558–9].
13. Karasawa J, Touho H, Ohnishi H, et al. Cerebral revascularization using omental transplantation for childhood moyamoya disease. *J Neurosurg* 1993;79(2):192–6.
14. Endo M, Kawano N, Miyaska Y, et al. Cranial burr hole for revascularization in moyamoya disease. *J Neurosurg* 1989;71(2):180–5.
15. Sainte-Rose C, Oliveira R, Puget S, et al. Multiple burr hole surgery for the treatment of moyamoya disease in children. *J Neurosurg* 2006;105(6):437–43.
16. Karasawa J, Kikuchi H, Furuse S, et al. A surgical treatment of moyamoya disease encephalo-myo synangiosis. *Neurol Med Chir (Tokyo)* 1977;17:29–37.
17. Henschen C. Surgical revascularization of cerebral injury of circulatory origin by means of stratification of pedunculated muscle flaps. *Langenbecks Arch Klin Chir Ver Dtsch Z Chir* 1950;264:392–401.
18. Touho H, Karasawa J, Ohnishi H. Cerebral revascularization using gracilis muscle transplantation for childhood moyamoya disease. *Surg Neurol* 1995;43(2):191–7 [discussion: 197–8].
19. Takeuchi S, Tsuchida T, Kobayashi K, et al. Treatment of moyamoya disease by temporal muscle graft encephalo-myo-synangiosis. *Childs Brain* 1983;10(1):1–15.
20. Caldarelli M, Di Rocco C, Gaglioli P. Surgical treatment of moyamoya disease in pediatric age. *J Neurosurg Sci* 2001;45(2):83–91.
21. Touho H. Cerebral ischemia due to compression of the brain by ossified and hypertrophied muscle used for encephalomyosynangiosis in childhood moyamoya disease. *Surg Neurol* 2009;72:725–7.
22. Matsushima T, Inoue T, Katsuta T, et al. An indirect revascularization method in the surgical treatment of moyamoya disease—various kinds of indirect procedures and a multiple combined indirect procedure. *Neurol Med Chir (Tokyo)* 1998;38(Suppl):297–302.
23. Touho H. Subcutaneous tissue graft including a scalp artery and a relevant vein for the treatment of cerebral ischemia in childhood moyamoya disease. *Surg Neurol* 2007;68(6):639–45.
24. Houkin K, Kuroda S, Ishikawa T, et al. Neovascularization (angiogenesis) after revascularization in moyamoya disease. Which technique is most useful for moyamoya disease? *Acta Neurochir (Wien)* 2000;142(3):269–76.
25. Nissim O, Bakon M, Ben Zeev B, et al. Moyamoya disease—diagnosis and treatment: indirect cerebral revascularization at the Sheba Medical Center. *Isr Med Assoc J* 2005;7(10):661–6.
26. Matsushima T, Inoue TK, Suzuki SO, et al. Surgical techniques and the results of a fronto-temporo-parietal combined indirect bypass procedure for children with moyamoya disease: a comparison with the results of encephalo-duro-arterio-synangiosis alone. *Clin Neurol Neurosurg* 1997;99(Suppl 2):S123–7.
27. Matsushima Y, Fukai N, Tanaka K, et al. A new surgical treatment of moyamoya disease in children: a preliminary report. *Surg Neurol* 1981;15(4):313–20.
28. Matsushima Y, Suzuki R, Ohno K, et al. Angiographic revascularization of the brain after encephaloduroarteriosynangiosis: a case report. *Neurosurgery* 1987;21(6):928–34.
29. Kashiwagi S, Kato S, Yasuhara S, et al. Use of a split dura for revascularization of ischemic hemispheres in moyamoya disease. *J Neurosurg* 1996;85(3):380–3.
30. Isono M, Ishii K, Kamida T, et al. Long-term outcomes of pediatric moyamoya disease treated by encephaloduro-arterio-synangiosis. *Pediatr Neurosurg* 2002;36(1):14–21.
31. Fujita K, Tamaki N, Matsumoto S. Surgical treatment of moyamoya disease in children: which is more effective procedure, EDAS or EMS? *Childs Nerv Syst* 1986;2(3):134–8.
32. Hannon KE. Pial synangiosis for treatment of moyamoya syndrome in children. *AORN J* 1996;64(4):540–54 [quiz 557–60].
33. Scott RM, Smith JL, Robertson RL, et al. Long-term outcome in children with moyamoya syndrome after cranial revascularization by pial synangiosis. *J Neurosurg* 2004;100(Suppl 2):142–9.
34. Dauser RC, Tuite GF, McCluggage CW. Dural inversion procedure for moyamoya disease. Technical note. *J Neurosurg* 1997;86(4):719–23.
35. Kashiwagi S, Kato S, Yamashita K, et al. Revascularization with split duro-encephalo-synangiosis in the pediatric moyamoya disease—surgical result and clinical outcome. *Clin Neurol Neurosurg* 1997;99(Suppl 2):S115–7.

36. Muto J, Oi S. Intradural arteriosynangiosis in pediatric moyamoya disease: modified technique of encephalo-duro-arterio-synangiosis with reduced operative damage to already growing revascularization. *Childs Nerv Syst* 2009;25(5):607–12.
37. Kinugasa K, Mandai S, Kamata I, et al. Surgical treatment of moyamoya disease: operative technique for encephalo-duro-arterio-myo-synangiosis, its follow-up, clinical results, and angiograms. *Neurosurgery* 1993;32(4):527–31.
38. Kim DS, Kye DK, Cho KS, et al. Combined direct and indirect reconstructive vascular surgery on the fronto-parieto-occipital region in moyamoya disease. *Clin Neurol Neurosurg* 1997;99(Suppl 2):S137–41.
39. Ozgur BM, Aryan HE, Levy ML. Indirect revascularisation for paediatric moyamoya disease: the EDAMS technique. *J Clin Neurosci* 2006;13(1):105–8.
40. Kawamoto H, Kiya K, Mizoue T, et al. A modified burr hole method galeoduroencephalosynangiosis in a young child with moyamoya disease. A preliminary report and surgical technique. *Pediatr Neurosurg* 2000;32(5):272–5.
41. Kawamoto H, Inagawa T, Ikawa F, et al. A modified burr hole method in galeoduroencephalosynangiosis for an adult patient with probable moyamoya disease—case report and review of the literature. *Neurosurg Rev* 2001;24(2–3):147–50.
42. Kim SK, Wang KC, Kim IO, et al. Combined encephaloduroarteriosynangiosis and bifrontal encephalogaleo (periosteal)synangiosis in pediatric moyamoya disease. *Neurosurgery* 2002;50(1):88–96.
43. Veeravagu A, Guzman R, Patil CG, et al. Moyamoya disease in pediatric patients: outcomes of neurosurgical interventions. *Neurosurg Focus* 2008;24(2):E16.
44. Miyamoto S, Kikuchi H, Karasawa J, et al. Pitfalls in the surgical treatment of moyamoya disease. Operative techniques for refractory cases. *J Neurosurg* 1988;68(4):537–43.
45. Ishii K, Fujiki M, Kobayashi H. Invited article: surgical management of Moyamoya disease. *Turk Neurosurg* 2008;18(2):107–13.
46. Matsushima T, Inoue T, Suzuki SO, et al. Surgical treatment of moyamoya disease in pediatric patients—comparison between the results of indirect and direct revascularization procedures. *Neurosurgery* 1992;31(3):401–5.
47. Takahashi A, Kamiyama H, Houkin K, et al. Surgical treatment of childhood moyamoya disease—comparison of reconstructive surgery centered on the frontal region and the parietal region. *Neurol Med Chir (Tokyo)* 1995;35(4):231–7.
48. Shrestha P, Sakamoto S, Ohba S, et al. Multiple concurrent anastomotic procedures in the management of moyamoya disease: a case report with review of literature. *Hiroshima J Med Sci* 2008;57(1):47–51.
49. Hayashi T, Shirane R, Tominaga T. Additional surgery for postoperative ischemic symptoms in patients with moyamoya disease: the effectiveness of occipital artery–posterior cerebral artery bypass with an indirect procedure: technical case report. *Neurosurgery* 2009;64(1):E195–6 [discussion E196].
50. Houkin K, Kuroda S, Nakayama N. Cerebral revascularization for moyamoya disease in children. *Neurosurg Clin N Am* 2001;12(3):575–84, ix.
51. Kim CY, Wang KC, Kim SK, et al. Encephaloduroarteriosynangiosis with bifrontal encephalogaleo (periosteal)synangiosis in the pediatric moyamoya disease: the surgical technique and its outcomes. *Childs Nerv Syst* 2003;19:316–24.