Original Article

Indirect Revascularization Procedures for Surgical Treatment of Moyamoya Disease in Pediatric Patients

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ABSTRACT

Background: Many forms of indirect cerebral revascularization had been adapted to establish neovascularization in moyamoya disease (MMD) affecting pediatric age group. Objective: The aim of this study was to analyze the clinical outcome and different angiographic responses following combined indirect revascularization [encephaloduroarteriosynangiosis (EDAS) with bifrontal encephaloduroarteriosynangiosis (EGS)] in pediatric patients with MMD. Patients and Methods: This study was conducted as a retrospective study in the period between January 2013 and January 2015, in Department of Neurosurgery Ain Shams University, on pediatric patients with MMD who underwent combined EDAS and Bifrontal EGS. Clinical outcomes were assessed. Preoperative and postoperative angiograms from twenty-eight treated hemispheres were collected and analyzed as regard good revascularization signs. Follow up ranged from 6 to 24 month. Results: Twenty-nine hemispheres were treated in fifteen patients, age ranged from 1.5 to 10 years with a mean of 5.57 ±2.96. There were nine males and six females. In our study, 93.3% of the patients presented with ischemic neurological symptoms. Eighty % of the patients showed stabilization and improvement of the symptoms after 3 month of the treatment. One patient 6.7% had postoperative major posterior circulation stroke, two patients 13.7% hand ischemic event between the two stages of treatment with good outcome. Mortality was 6.7%. In the twenty-eight hemispheres with follow up, good revascularization was established in 78.6% in the middle cerebral artery (MCA) territory, and in 71.4% in the anterior cerebral artery (ACA) territory. Superficial temporal artery (STA) showed neovascularization in 72.7% of the improved hemispheres and the middle meningeal artery (MMA) in 90%. STA showed increase in caliber in 42.9% and MMA in 50%. There was a decrease or disappearance of moyamoya vessels in 65% of the followed hemisphere.

Conclusion: Our results demonstrate that indirect revascularization (EDAS and Bifrontal EGS) with arachnoid opening should be considered as a primary line of treatment once diagnosis of MMD is made in pediatric age group. It is technically feasible, well tolerated, safe with good clinical outcome, decrease in the rate of the strokes, and decrease in morbidity and mortality rates .The combination of EDAS and Bifrontal EGS gives a wide cortical surface covering including MCA and ACA territory with good neovascularization rates and this could alter the progressive natural history and the angiographic appearance of MMD. Outcomes of these procedures are best evaluated clinically and not only angiographically. For more adequate results, a longer follow up with a bigger sample is needed and launching of an Egyptian registry for treatment of pediatric MMD, could do this.

INTRODUCTION

MMD is a rare cerebrovascular disease, yet it is the most common pediatric cerebrovascular disease in eastern Asia. It is a chronic, progressive occlusive cerebrovascular disease involving bilateral stenosis or occlusion of the terminal portion of the Internal Carotid arteries (ICAs) and/or the proximal portions of the ACAs and MCAs.

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It is also characterized by irregular perforating vascular networks, called moyamoya vessels, near the occluded or stenotic regions corresponding to the lenticulostriate and thalamoperforate arteries. It is this outgrowth of small vessels that produces the radiological image of a hazy “puff of smoke” giving the disease its name, “moyamoya” in Japanese.
an autosomal dominant inheritance pattern with the gene found in the telomeric region of 17q25.34,29.

Clinically, children with MMD present with ischemic attacks (either strokes or TIAs), and adults present with either ischemic or hemorrhagic events. However, this point has recently undergone debate, depending on the patient’s geographical location.28 In most cases, the disease is progressive and does not respond to medical therapy with vasodilators or antithrombotic agents.31

Since the first reports of abnormal angiograms with hypoplasia of bilateral carotid arteries and subsequent naming and further characterization of this abnormality as MMD by Suzuki and Takaku in 1969,45 several surgical options have been used in the treatment of MMD. Direct revascularization involving extracranial-intracranial anastomoses provides immediate blood flow but is technically difficult, especially in children.46 Several alternative indirect methods that are technically simpler have been developed to promote neovascularity to the ischemic brain. These methods involve applying either vascularized muscle (encephalomyosynangiosis [EMS]) or the intact STA to the surface of the brain, with various modifications (EDAS or pial synangiosis) encephaloduroarteriomyosynangiosis [EDAMS]31, and multiple cranial burr holes with EDAS, all these techniques either single or in combinations are widely used in treating this rare disease in patients for whom direct bypass is unfeasible or controversial (pediatric age group)10,26.

PATIENTS AND METHODS

In our current study, we have a group of fifteen patients, 18 years old or younger with MMD, we will present an analysis of the clinical outcome and angiographic changes that occur after indirect revascularization surgery, with a focus on the observed changes of the STA, and MMA.

This study was conducted in the period between January 2013 and January 2015, in Department of Neurosurgery Ain Shams University, on pediatric patients with MMD. Fifteen pediatric patients with confirmed MMD were included in the study, with twenty-nine treated hemispheres. The age of the patients range was 1.5 to 10 years with a mean of 5.57±2.96, there were nine males (60%) and six females (40%). Median hospital stay was 4 days. All patients underwent almost the same surgical treatment combined EDAS and bifrontal EGS by the same neurosurgeon. We summarized the surgical procedure, which was performed in this study since the results are closely related to the techniques of the surgery.

All patients presenting with symptoms consistent with MMD (transient ischemic attacks [TIAs], cerebral infarction, and/or hemorrhage) received both brain magnetic resonance imaging (MRI) and cerebral angiography, including external carotid artery injections. MRI perfusion studies were not used routinely. This was done according to guidelines (Table 1) previously described in many reports.41 We applied the diagnostic angiographic criteria based on the Japanese Ministry of Health and Welfare guidelines (Table 2).3

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<th>Table 1: Guidelines for Diagnosis of MMD</th>
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<td>1. Diagnostic evaluation of children suspected of having moyamoya should include an MRI study.</td>
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<td>2. Confirmation of the presence of moyamoya arteriopathy is defined by catheter angiography (which is recommended to include all 6 vessels, although the use of a noninvasive arterial imaging study, such as CT Angiography or MRA, may be sufficient to identify the condition with a high likelihood of certainty when catheter angiography may not be feasible.</td>
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<td>3. Techniques to measure cerebral perfusion may be useful in the diagnosis and follow-up of children with moyamoya</td>
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<th>Table 2: Diagnostic Angiographic Criteria of MMD</th>
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<td>1. Stenosis of the distal (intracranial) ICAs, up to and including the bifurcation, along with segments of the proximal ACA and MCA.</td>
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<td>2. Dilated basal collateral vessels to varying grades</td>
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<td>3. The condition is bilateral</td>
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After confirming the diagnosis, MMD was classified according to Suzuki grade.49 In general, all patients with symptomatic MMD are considered for indirect revascularization procedures, because the natural history and predictive value of MRI and angiograms are imprecise.

Guidelines for Surgical Intervention
1. Revascularization should be offered to children with evidence of moyamoya, including ongoing ischemic symptoms and/or evidence of compromised blood flow or cerebral perfusion reserve. Specifically, children who are clinically asymptomatic but who demonstrate radiographic or functional evidence of impaired cerebral perfusion.

2. No firm recommendations can be made on specific timing for surgery, although the general principle of minimizing the time between diagnosis and revascularization is supported. Delays may be reasonable or required to allow scheduling of experienced anesthetic and ICU staff to maximize the safety of the procedure.41

The anesthesia team is instructed to keep strict control of the blood pressure to avoid hypotension. Generally, the systolic blood pressure is kept above 120 mm Hg, throughout the operation. Anesthesia is also instructed to avoid hyperventilation given the risk of vasoconstriction induced by hypocapnia. Additionally, special care should be taken to provide adequate...
perioperative analgesia to avoid crying in the early postoperative period, which can cause hyperventilation. Because there is no temporary occlusion of any intracranial blood vessels (as there would be in a bypass procedure), we didn’t use intraoperative electroencephalographic monitoring, systemic hypothermia or barbiturate neuroprotection during indirect revascularization procedures.

Surgery was usually performed in two stages, initially in the symptomatic and hemodynamically affected hemisphere EDAS with bifrontal EGS. The average interval between the first and second operations was 6 months (range, 3 to 9 month). The general technique used for the EDAS is similar to that described previously in the literature by Matsushima and Inaba. After the STA is identified and dissected out of the scalp, leaving a cuff of adventitia, a craniotomy is performed. The underlying dura is opened in a cruciate fashion. The entire dural layer is folded into the subdural space. And as MMA is important for neovascularization also it may be sharing in already exiting collateral circulation to the MCA, attempts have been made to construct the dural opening in a manner that preserves as many of the MMA branches as possible. The arachnoid membrane over the cortical sulci is opened with the surgical microscope, to promote the in-growth of neovasculature. Although the arachnoid is wide open, the STA is not sutured to the pia as has been described as pial synangiosis by Adelson and Scott and Scott et al. The STA adventitia is then sutured to the edges of the dura before the bone flap is replaced. Meticulous hemostasis of the arachnoid, dura, STA adventitia, and scalp before closure is a must. Given that many patients are on aspirin preoperatively, there is considerable dural dissection, and if the dura is left open, even small amounts of bleeding can accumulate in the subdural space, causing a postoperative subdural hematoma. (Fig. 1)

To obtain collateral neovascularization in the ACA territory, bifrontal EGS was combined with EDAS. The scalp was incised separately for EDAS and EGS. At the EGS site, an S-shaped scalp incision was made 2 cm anterior to the coronal suture. The galea and/or the pericranium was dissected and incised in a zigzag pattern. A 4-8 cm craniotomy, crossing the superior sagittal sinus, was made. The dura was incised separately on both hemispheres, and the arachnoid membrane was incised. The apex of the prepared galeal (periosteal) flap was inserted as deeply as possible into the interhemispheric fissure and was sutured to the dura (Fig. 1).
For closure of the burr holes, titanium plates that completely cover the burr hole are not recommended because both dural and scalp vessels appear to contribute to the intracranial circulation postoperatively. Either the burr hole is left open without a plate, or 1 or 2 linear plates are used as a bridge to support the scalp and thus prevent cosmetic deformity while allowing room for new vessel growth from the scalp. The scalp is then closed over the burr hole in the usual fashion.

Both preoperative and postoperative (3 to 12 month, with an average of 6 month) angiograms were reviewed, angiographic results were analyzed. In preoperative angiograms, the general pattern of MMD, Suzuki grade\(^4\) presence of moyamoya vessels, and collateralization (leptomeningeal) were noted. Additionally, digital measurements on the lateral angiographic projection: of the width of the proximal STA just distal to the bifurcation, and the width of the MMA in its largest intracranial portion, just distal to foramen spinosum.

The postoperative angiograms were performed more than three months after the surgery, when the induction of the neovascularization is thought to have reached its plateau. Angiograms were examined for any change in the MMD obstruction, Suzuki grade\(^4\) decrease of Moyamoya vessels, STA, MMA, and burr holes were examined for the presence of vascular blush, neovascularity, and anastomoses to intracranial vessels. Digital calibers of the MMA, STA were measured at the same locations as on the preoperative angiogram. Good neovascularization was defined as the apparent development of new arteries (anastomosis), presence of an intracranial blush from the donor vessels (MMA, STA) to the cortical branch of the MCA, ACA, and enlargement of the caliber of MMA and/or STA compared to that seen in pre-operative angiograms. Poor revascularization is the absence of these criteria.

**RESULTS**

The most common presentation of MMD in the current study was presence of symptoms related to cerebral ischemia, including both TIAs and infarction. Fourteen patients (93.3%) had ischemic symptoms, nine patients (60%) had prior strokes and five patients (33.3%) had TIAs. Only one patient (6.7%) had subarachnoid hemorrhage with a small internal carotid bifurcation aneurysm that was first coiled before the surgery of the contralateral hemisphere and 90% from the twenty-two hemispheres with good revascularization. While good neovascularization was induced from MMA in twenty out of twenty-eight hemispheres (71.4%)%, and 72.7% from the twenty-two hemispheres with good revascularization. When respect to filling of the ACA territory, the analysis of follow-up angiograms demonstrated good revascularization in ten of the fourteen patients (71.4%).

Good neovascularization via STA (development of new arteries to the cortical branch, blush) was observed on sixteen out of twenty-eight hemispheres (57.1%), and 72.7% from the twenty-two hemispheres with good revascularization. When respect to filling of the ACA territory, the analysis of follow-up angiograms demonstrated good revascularization in ten of the fourteen patients (71.4%).

Moyamoya vessels were found in twenty-six out of the twenty-eight hemispheres before treatments, in the follow up seventeen out of the twenty-six hemispheres (65%) showed significant decrease or disappearance of the moyamoya vessels (Fig. 2,3,4).
Fig. 2 a-j: A 4 year-old boy who presented with episodic hemiparesis and paraparesis. a: preoperative right ICA anteroposterior (AP) angiogram reveals tight MCA and ACA stenosis with moyamoya collaterals. b: preoperative right external carotid artery injection. c: preoperative left ICA (AP) angiogram reveals tight middle and anterior cerebral artery stenosis with moyamoya collaterals. d: preoperative left external carotid artery lateral injection. e: postoperative right ICA lateral injection 6 months after combined EDAS and bifrontal EGS reveal a significant decrease in moyamoya vessels. f: postoperative early arterial-phase right ECA lateral injection reveals proximal STA increased in size (big arrow) and MMA increased in size (small arrow). g: postoperative late arterial-phase right ECA reveals angiographic blush and neovascularity of the MCA territory (small arrows) from both STA and MMA. h: postoperative arterial phase of left ECA injection revealing MMA increase in size (small arrows) and neovascularization of ACA at site of EGS bifrontal craniotomy (big arrow). i,j: postoperative late arterial phase of left ECA lateral and AP injection with extensive intracranial collaterals and blush associated with the frontal craniotomy site in the ACA distribution (small arrows).

Fig. 3 a-i: A 5 year-old girl who presented with right hemiparesis and dysphasia a,b: preoperative left ICA AP and lateral injections reveals tight MCA and ACA stenosis with moyamoya collaterals. c: preoperative left external carotid artery injection. d: postoperative 6 month after (EDAS) early arterial-phase left ECA lateral injection reveals proximal STA and MMA increase in size (big arrow) and significant angiographic blush and neovascularity of the MCA territory (small arrows) from both STA and MMA. e,f: postoperative late arterial-phase lateral left ECA injection reveals angiographic blush and neovascularity of the MCA territory (small arrows) from both STA and MMA and neovascularization and filling of the ACA (big arrows). g,h,i: postoperative late arterial-phase AP left ECA injection reveals angiographic blush and neovascularity of the MCA territory (small arrows) from both STA and MMA and neovascularization and filling of the ACA (big arrows). Curved arrow reveals neoanastomosis between ACA and MCA.
Fig. 4a-i: A 2 year-old boy who presented with episodic attacks of TIAs in the form of left hemiparesis. 

a: preoperative right ICA AP injection reveals tight MCA and ACA stenosis with minimal moyamoya collaterals. 
b: preoperative right ECA lateral injection. 
c: postoperative 3 month after (EDAS) arterial-phase right ICA AP injection. 
d: postoperative early arterial-phase ECA lateral injection reveals proximal STA (small arrow) and MMA increase in size (small arrow) e,f,g: postoperative mid and late arterial-phase right ICA injection reveals significant angiographic blush and neovascularity of the MCA territory (small arrows) from both STA and MMA. 
h,i: postoperative arterial-phase AP right ECA injection reveals angiographic blush and neovascularity of the MCA territory (small arrows) from both STA and MMA.

**DISCUSSION**

Moyamoya disease causes progressive neurological deterioration in affected patients. The morbidity rate for untreated moyamoya disease has been reported to be more than 70%\(^3\)\(^1\). The primary goal of treatment for patients with MMD is to improve blood flow to the brain and, in so doing, to reduce the risk for future ischemic injury. Furthermore, relieving the hemodynamic stress on the moyamoya vessels at the base of the brain may reduce the risk for future hemorrhage from these abnormal vessels\(^5\)\(^6\)\(^7\)\(^8\)\(^9\)\(^10\)\(^11\)\(^12\)\(^13\)\(^14\)\(^15\)\(^16\)\(^17\).

Revascularization by both direct and indirect methods (EDAS, EDAMS, EGS, Multiple burr holes) can improve neurological outcomes\(^14\). However, much debate has centered on which method is preferable to use direct bypass versus indirect revascularization techniques, although some authors have noted an advantage to direct bypass and advocated its use whenever possible\(^11\)\(^12\)\(^13\)\(^14\). Proponents of indirect revascularization emphasize its noninvasiveness, safety, and technical ease, particularly for young children. Surgeons who perform direct anastomoses think that these procedures are more efficacious and are equally safe. Also, previous EDAS or EMS may render the STA no longer suitable as a donor artery in patients who experience failure of such procedures. The technical difficulty of direct anastomosis in children, because of the smaller size of both recipient and donor arteries, has been emphasized\(^16\). It has also been suggested that sectioning of the STA may compromise existing, spontaneous extracranial to intracranial anastomoses. Additionally, temporary clipping of the MCA may impair blood flow to the already compromised brain and increase the risk for perioperative infarction\(^2\)\(^8\). The EDAS and combined indirect revascularization (Multiple burr holes, Bifrontal EGS) may allow vessel ingrowth over a larger area of cortex, eventually revascularizing a larger area than would the direct anastomosis STA-MCA alone only revascularizing MCA territory. In addition, the EDAS provides an alternative route for collateralization, if the primary anastomosis fails\(^18\).
Another theoretical concern is bypass literature for nonmoyamoya cerebrovascular occlusive disease. Bypass to the MCA in patients with tight MCA stenosis caused by intracranial atherosclerosis can prompt further occlusion and thrombosis of the stenotic MCA segment, occasionally causing infarction resulting from obstruction of perforator arteries. In addition as the donor scalp artery is stopped for sometime during anastomosing in case of anastomotic failure occurs the vessel with good blood now maybe lost. Also there is a difficulty in surgical bypass procedure: special training is needed; the time of operation and anesthesia are prolonged; results are unstable: it is not expected that the best skilled surgeon can make anastomosis 100% patent. Finally, bypass surgery to treat childhood MMD should be performed before irreversible ischemic damage occurs, because there is no development of collateral vessels after bypass surgery in already infarcted areas.

Despite these concerns, and that there are multiple reports of successful revascularization using a variety of indirect procedures. Yet there is no general consensus of the superiorities of either direct or indirect revascularization for MMD. As Indirect methods do not immediately provide increased blood flow but instead depend on the ingrowth of vessels, which can take months. Matsushima and Inaba reported that angiographically visible revascularization after EDAS was present by 6 months. Strokes that occur during this time period can be devastating. Other reports stated that reaching a plateau of revascularization could be achieved as early as 3 month. Perhaps most importantly, children sometimes may present with acute exacerbations of stroke or TIAs, in the period between surgery and establishment of revascularization, or between two stages of surgery.

In our current series all patients were treated by indirect revascularization technique, a combination of EDAS and Bifrontal ECS, regardless of the child age, or size of the donor (STA, MMA) and recipient (MCA, ACA) vessels. Fourteen out of the fifteen patients tolerated twenty-eight surgical procedures of indirect revascularization. We only had one patient with an immediate postoperative stroke in the PCA distribution; this was due to posterior circulation affection by moyamoya disease, which was not evident in the preoperative angiograms done one month before the procedure and was not symptomatizing before the surgery. Unfortunately this patient died, with the only mortality in our series (6.7%).

Because impaired cerebral vascular reserve makes patients vulnerable to the hemodynamic stresses of surgery regardless of the surgical technique, several groups have emphasized the importance of close hemodynamic monitoring and aggressive intraoperative management of blood pressure and partial pressure of CO2, to avoid strokes. A number of perioperative events may be unavoidable, and the best strategy may be to perform bilateral procedures in a single session to minimize complications, although published data are lacking.

We have used staged procedures with careful anesthetic management and neuroprotection and to date have observed only one perioperative stroke. This finding is consistent with the reports of perioperative stroke (some occurring up to 10 days postoperatively) among patients undergoing any mode of revascularization. Crying (which can induce hypocapnia and relative vasoconstriction) and dehydration (which can lead to hypoperfusion) have both been associated with perioperative ischemic events. Given these findings, maintenance of normovolemia or mild hypervolemia and special attention to pain control are of particular importance.

In our study, two patients had symptoms due to cerebral ischemia from the previously asymptomatic contralateral hemisphere in the period between the 2 stages of treatment (in the duration between surgery of the symptomizing hemisphere and before surgery of the contralateral one). Both improved with minimal neurological deficits in one of them. Other postoperative complications were insignificant. Twelve patients out of fifteen (80%) experienced stabilization or resolution of their symptoms during the period of clinical follow up from 6 to 24 month. Additionally, no patients had MMD-related hemorrhage after treatment. This suggests that the patients develop enough new circulation to relieve ischemia and to reduce the hemodynamic stress on the abnormal moyamoya vessels significantly. Our clinical outcome goes with the reports that indirect revascularization methods are thought to be safe with good clinical outcome.

Radiographic studies confirmed moyamoya disease and provided evidence of successful revascularization. In the current study preoperative angiography demonstrated the classic appearance of moyamoya disease, with stenosis of the intracranial branches of the internal carotid artery and fine basal ganglion anastomotic vessels. Preoperative collateral filling was generally poor, in five (16.7%) out of thirty hemispheres.

Angiographic neovascularity after indirect revascularization procedures has been demonstrated in the literature. In particular, new STA and MMA branches have been demonstrated to supply the intracranial blood vessels or even form spontaneous direct anastomoses with large intracranial branches. Yamada et al measured the size of the STA and MMA preoperatively and postoperatively in twenty seven pediatric cases treated with EDAS and showed some increase in size after indirect revascularization. Dusick et al reported almost similar results. Rates of neovascularization of more than one-third of the MCA territory have been reported to be 62 to 84% after simple EDAS. Transdural anastomoses gradually develop 6 to 12 months after
EDAS, with diminishing moyamoya vessels, widening of the superficial temporal artery and the middle meningeal artery, and clinical improvements. In the current study, the comparison of preoperative and postoperative angiograms showed a wide range of angiographic responses that demonstrated good revascularization and neo-anastomosis. Good revascularization of the MCA territory after EDAS was radiographically confirmed in 78.6% of patients as early as 3 months after surgery. Good neovascularization via STA occurred in 57.1% of treated hemispheres, and 72.7% of revascularized hemispheres, while good neovascularization via MMA occurred in 71.4% of treated hemispheres, and 90% of revascularized hemispheres.

The finding that MMA contributes considerably to intracranial blood flow indicates the importance of its preservation and protection of its branches during the surgical procedures particularly in planning the dural opening. This may maximize the ability of the dural vessels to revascularize the underlying brain and also protect the already existing anastomosis. We have planned the durotomy in such a way to preserve the major dural arteries.

Patients in the current study demonstrated a significant increase in vessel size postoperatively. The caliber of MMA increased in 50% of treated hemispheres and the caliber of the STA increased in 42.9%. Although some of this change may be due to the physical act of dissecting the artery out of the scalp and its adventitial coverings, part of the change in the STA may be accounted for by hemodynamic alterations secondary to increased demand. However, the MMA, which is not dissected and remains in its fixed dural and bony environment at the skull base, also demonstrated marked increase in size, likely as a result of the increased flow to new intracranial branches. The increase in vessel diameter, the appearance of angiographic blush, and new branches to the intracranial circulation, were all signs of good revascularization. However, presence of large area of encephalomalacia, may cause poor or no surgical collateral vessels; nevertheless, the remaining uninfarcted territory may be well revascularized.

Globy et al reported that patients with increase in moyamoya vessels exhibited relatively worse collateral filling, whereas patients with marked decrease in moyamoya vessels were those with the best surgical collateral vessels. This may be viewed as evidence of decreased ischemia-induced angioproliferative factors, indicating successful revascularization. In our current study preoperative angiograms showed moyamoya vessels were found in 92.9% of the treated hemispheres, in the follow up angiography 65% showed significant decrease or disappearance of the moyamoya vessels.

Most surgical approaches have focused on increasing the blood supply primarily in the middle cerebral artery (MCA) territory and do not directly benefit the anterior cerebral artery (ACA) territory, which is also an important area in the developing brain. Revascularization of the ACA territory should be considered for the treatment of pediatric MMD, for several reasons. First the disease process in children is dynamic and progressive. This progression eventually involves the ACA, therefore, deterioration of blood flow in the ACA territory may continue to progress despite good collateral formation in the MCA territory. Second, impaired circulation in the ACA territory is as frequent as that in the MCA territory. According to a report by Karasawa et al., motor weakness of the lower extremities was the second most common ischemic symptom. Third, the frontal area is essential for the intellectual development of children. Ischemic brain damage in the ACA territory may lead to poor intellectual outcomes, directly affecting the patients’ quality of life. Finally, multiple infarctions represent the main reason for poor clinical outcomes and bad neovascularization. This situation is the same in the ACA territory. Therefore, prophylactic revascularization surgery should also be considered for the ACA territory.

For revascularization of the ACA territory, some operative approaches, including direct bypasses such as STA-ACA bypass, indirect bypasses such as omental transplantation, and the ribbon procedure, and other techniques have been used as supplementary measures for patients with MMD. The results obtained with these procedures have been reported to be excellent. However, it is still unclear which technique is safest and most effective.

In our current study we used bifrontal EGS as a supplementary measure for the ACA territory because all of our patients were children and our policy was to produce more predictable collateral formation with minimal risk. By using EDAS with bifrontal EGS, we thought that we might enhance collateralization of the ACA territory in addition to the MCA territory. With respect to filling of the ACA territory, the analysis of follow-up angiograms demonstrated good revascularization in 71.4% of patients. In a study by Nakashima et al., all patients who underwent simple EDAS exhibited no revascularization of the ACA territory, whereas 89% of the patients who underwent ribbon EDAMS demonstrated filling of the ACA territory. Kim et al. reported higher rates of ACA territory revascularization using the bifrontal EGS.

**CONCLUSION**

Our results demonstrated that indirect revascularization (EDAS and Bifrontal EGS) with
arachnoid opening should be considered as a primary line of treatment once diagnosis of MMD is made in pediatric age group. It is technically feasible, well tolerated, safe with good clinical outcome, decrease in the rate of the strokes, and decrease in morbidity and mortality rates. The combination of EDAS and Bifrontal EGS gives a wide cortical surface covering including MCA and ACA territory with good neovascularization rates and this could alter the progressive natural history and the angiographic appearance of MMD. Outcomes of these procedures are best evaluated clinically and not only angiographically. For more adequate results, a longer follow up with a bigger sample is needed and launching of an Egyptian registry for treatment of pediatric MMD, could do this.

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