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# Complications following Encephalo-Duro-Arterio-Myo-Synangiosis in a case of Moyamoya disease

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## Abstract:

We report a case of an 18-year-old girl diagnosed to have Moyamoya disease (MMD), who underwent bilateral encephalo-duro-arterio-myo-synangiosis. Literature search has clearly inferred that in comparison to an adult patient, children with MMD can have a good prognosis if early diagnosis and active surgical intervention are achieved. Evidence has demonstrated that active surgical management, including indirect bypass surgery, may improve the cerebral circulation on a relatively larger scale than direct bypass surgeries, which can only improve the cerebral circulation in the area of the vascular reconstruction.

## Keywords:

Encephalo-duro-arterio-myo-synangiosis, moyamoya, puff of smoke, stroke in encephalo-duro-arterio-myo-synangiosis

## Introduction

Moyamoya disease (MMD) is a rare idiopathic cause of cerebrovascular ischemia in children, which is characterized by irreversible vaso-occlusion of the cerebral vessels. A patient usually presents with symptoms of headache, neurofocal deficits, and seizures that are refractory to medical treatment. This explains why surgical revascularization techniques are gaining importance as the primary mode of treatment. Nevertheless, the outcome is based on the neurological status of the patient at the time of diagnosis followed by active surgical intervention. This case report highlights the possible complications following surgical intervention in the case of MMD, of which controlling immediate postoperative stroke is the most important factor.

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## Case Report

An 18-year-old girl presented with complaints of intermittent headache, generalized weakness and intermittent tremors of the left upper and lower limbs for about a month. Neurologically, she was conscious and well oriented; however, on the left side, she had spastic ataxic paresis, decreased motor power 4/5, exaggerated deep tendon reflexes, and pronator drift. There was no evidence of neurocutaneous markers, asymmetry of the face, and signs of meningeal irritation. Cranial nerve examination was within the normal limits.

Laboratory investigations including complete blood count, liver function tests, kidney function tests, coagulation profile, and immunological workup (Anti nuclear antibody, lupus anticoagulant, pANCA, cANCA and APLA) were normal. Magnetic resonance angiography of the brain and digital subtraction angiogram [Figure 1 and Video 1] confirmed the diagnosis of MMD. Bilateral

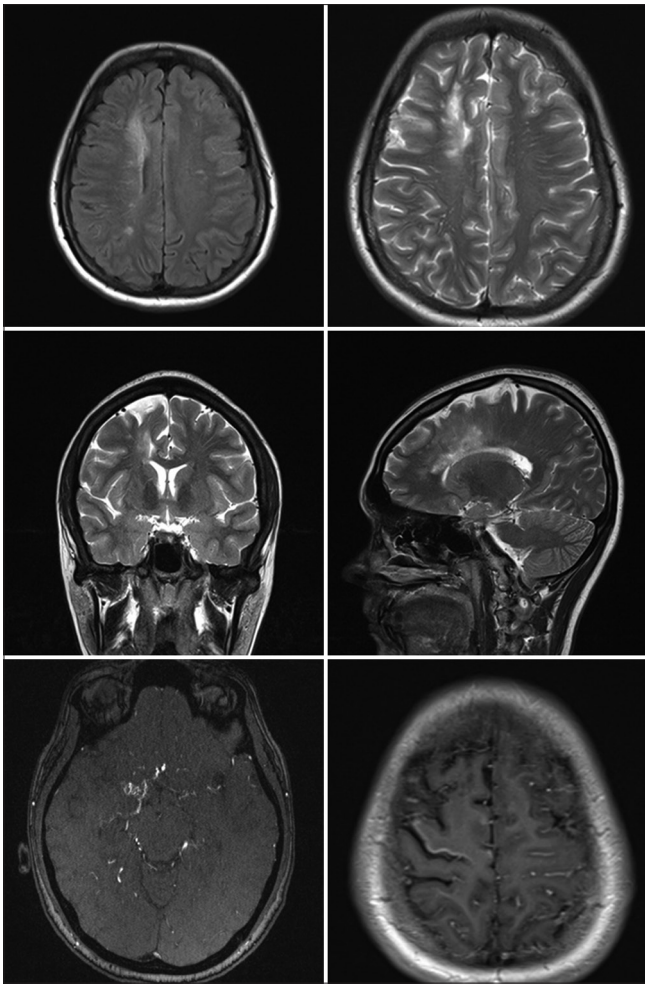
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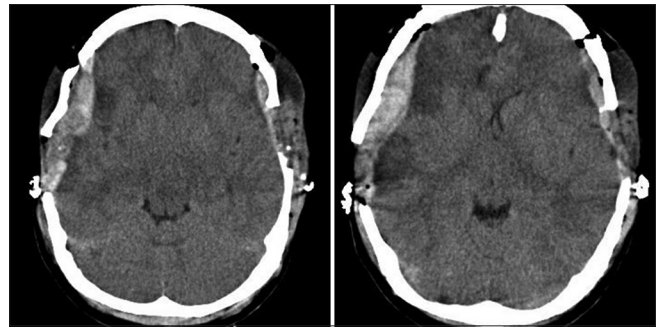
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**Figure 1:** Magnetic resonance imaging of the brain showed chronic infarction in the right superior and middle frontal gyri with acute infarction in the right parietal lobe. Perfusion-weighted images were suggestive of vascular compromise involving both the cerebral hemispheres, which was more on the right side with sparing of the left occipital and posterior temporal lobes. Magnetic resonance angiography revealed multiple collateral vessels in both the posterior cerebral artery territories (more on left side), with prominent lenticulostriate vessels, features that are suggestive of Moyamoya disease

encephalo-duro-arterio-myo-synangiosis (EDAMS) was performed on the patient. During the immediate postoperative period, she developed flaccid dysarthria and worsening of left-sided weakness (motor power 1/5) and oropharyngeal dysphagia due to the delayed laryngeal elevation. Noncontrast computed tomography of the brain revealed scattered infarcts in the right frontal, temporal, and parietal lobes associated with patchy gliosis in the right frontal lobe [Figure 2]. She was initiated on a single antiplatelet and a calcium channel blocker along with rehabilitation therapy (speech/swallow/limb movements) and supportive care to which she responded well. At discharge (postoperative day – 14), she was fully conscious, oriented with improved left hemiparesis (motor power 4/5), and was tolerating soft/semi-solid oral diet and was able to speak. On follow-up after 1 year, she has no neurological deficits.



**Figure 2:** Noncontrast computed tomography of the brain revealed scattered infarcts in the right frontal, temporal, and parietal lobes associated with patchy gliosis in the right frontal lobe

## Discussion

MMD is a rare and interesting entity that typically presents as cerebrovascular accident (s) in children. The term “Moyamoya” is coined on the basis of cerebral angiogram appearance that literally means “a puff of smoke” due to the abnormal collateral formation. Screening high-risk groups, such as those with a family history of MMD, would improve prognosis due to early diagnosis and intervention. This case report highlights the probable complications following surgical intervention in the case of MMD, of which controlling immediate postoperative stroke is the most important factor. Several studies reveal indirect revascularization techniques to be superior to direct revascularization techniques for pediatric patients. EDAMS procedure was proposed in the year 1984 to combine aspects of all the indirect revascularization procedures. The literature review revealed that surgically treated pediatric patients with MMD showed a good social adaptation. Houkin *et al.*<sup>[1]</sup> demonstrated that, in comparison with direct microanastomosis, EDAMS may be better suited for pediatric patients with MMD.

The potential complications following surgical intervention in a review article by Kim in 2016 stated the average annual incidence of stroke after the indirect revascularization procedure to be 1.6% in pediatric cases and 0%–14.3% in adult cases. R. Guzman *et al.*<sup>[2]</sup> reported that significant neurological deficits occurred in 15 patients undergoing both direct and indirect revascularization procedures, of which 1 patient had undergone indirect bypass and developed stroke in the right temporal region and had right-sided hemiparesis and aphasia resembling our case.<sup>[3]</sup> Other complications included hemorrhagic stroke (0.7%–8.0%), hyperperfusion syndrome (21.5%–50.0%), and epidural hematoma<sup>[4]</sup> in 4.8% of cases. Hankinson *et al.*<sup>[5]</sup> performed bilateral indirect bypass revascularization in 7 patients and unilateral in 5 patients, of which 2 patients (16.7%) suffered cerebrovascular events, 3 weeks postprocedure. Other studies by Ishikawa *et al.*<sup>[6]</sup> reported perioperative

ischemic events that occurred in 31% of patients undergoing EDAMS and 13% undergoing a combination of surgery with superficial temporal artery to middle cerebral artery bypass and EDAMS. Different studies by Karasawa *et al.*,<sup>[7]</sup> Kinugasa *et al.*,<sup>[8]</sup> and Miyamoto *et al.*<sup>[9]</sup> showed similar results. Miyamoto *et al.*<sup>[9]</sup> also stated that persistent ischemic attacks after receiving indirect bypass surgery needed to have an additional direct bypass surgery. Sakamoto *et al.*<sup>[10]</sup> demonstrated that the incidence of postoperative ischemic events was related more to the severity of MMD and the type of surgical procedure than to other factors, including anesthetic management. They retrospectively reviewed the perioperative course of 368 cases of revascularization. Ischemic events were noted in 14 cases (3.8%), 4 of which were strokes and the others were transient ischemic attack. After the diagnosis of ischemic stroke, patients should be started on the best medical therapy, including antiplatelet agents (aspirin) and calcium channel blockers (nimodipine), with other supportive measures, depending on the size of the infarct. The efficacy of these treatment modalities has not yet been proven to be of benefit. The use of anticoagulation should be carefully thought through before administration because of the concern of hemorrhagic stroke.<sup>[4-6]</sup>

To reduce the incidence of postoperative ischemic complications, most of these studies suggest maintaining euvolemic status and normal blood pressure, as hypotension and hypercapnia can aggravate hypoperfusion during or after surgery.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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