Medicine Science 016;5(Supp):86-95 *Anesthesia, Moyamoya Disease*

Case Report *doi: 10.5455/medscience.2016.05.8413*

Anesthetic Management of Children with Moyamoya Disease: A Case Report

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Abstract

Moyamoya disease is a cerebrovascular disease named after its angiographic image diagnosis, the etiology of which is not known and which is characterized by the chronic progressive stenosis of the main internal cerebral arteries that make up the Willis polygon. Ischemic symptoms are at the forefront in this disease which is more frequently observed in children. The definitive treatment of this disease is surgery and cerebral ischemia is a complication that can develop frequently during surgery. That is why, care during the perioperative period is very important. Perioperative anesthetic goal is to ensure the balance between the delivery of oxygen to the brain and the consumption of oxygen. The objective of this presentation is to discuss the anesthesia application in a child case who underwent encephalo-dura-arterio-cynangiozis (EDAS) surgery because of a 15 month moyamoya disease.

Keywords: Anesthesia, moyamoya disease, children

(Rec.Date: Dec 14, 2015 Accept Date:Dec 29, 2015)

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Introduction

Moyamoya disease is a rare cerebral circulation anomaly in which generally bilateral and compensatory collaterals develop and which is characterized with chronic progressive stenosis of the main internal cerebral arteries that constitute the Circle of Willis [1,2]. As a result of slow-growing occlusion and stenosis at distal part of bilateral internal carotid artery, an anastomosis is made between internal and external carotid artery [2,3]. Moyamoya means " crumbling cigarette smoke in the air" in Japanese and it s a name which is given for angiographic appearance of anormal collateral vascularization in the basal ganglia [3,4]. Angiography is the gold standard diagnostic method. Moyamoya is the most common pediatric cerebrovascular pathology in Japan and its incidence is 1/1.000.000 per year [3,5]. Moyamoya can be seen at any age but makes two peak points at age intervals of 1-5 and 36-40. Approximately half of the patients are under 10 years of age [2,3].

The clinical appearance of the disease can vary between children and adults.3 In juvenile form; ischemic symptoms are more common and in adult form bleeding and ischemic symptoms are observed with equal frequency [2,3,5]. The aim of the mediacal treatment is to make vasodilatation or increasing blood flow by reducing blood viscosity but medical treatment can not stop the progression of the disease [2,5]. Medical treatment is a palliative treatment which includes anticoagulants, antiplatelets, antiepileptics and calcium channel blockers. Morbidity is 70% in patients who did not have treatment. The main treatment is surgery [5].

In this report we aimed to discuss anesthesia application in a pediatric patient who underwent (by taking "informed consent" from parent) encephaloduroarteriosynangiosis (EDAS) surgery because of moyamoya disease which was diagnosed 15 months ago.

Case Report

16 month old male patient applied to our hospital due to falling from a height of one meter about 5 months ago followed by an epileptic seizure which started one week after the fall. Ischemic areas were identified in the basal ganglia during computerized brain tomography and magnetic resonance imaging. About 80-90% stenosis of the proximal portion of bilateral anterior and middle cerebral artery was detected in four-vessel cerebral arterial digital

subtraction angiography (DSA) and it was observed that mainly distal areas were stained with leptomeningeal collaterals and as a result of these observations moyamoya disease was diagnosed.

EDAS procedure was planned fot this patient by neurosurgery clinic. In pre-operative evaluation; complete blood count and blood chemistry were normal. After applying pre-medication with 0.1 mg/kg i.v. midazolam preoperatively, patient was taken to operation room. He had 3-4 points in Ramsey Sedation Scale while was taking to operation room.

Electrocardiography (EGC), pulse oximetry and noninvasive arterial blood pressure monitoring were carried ou in the operation room. The measurements recorded were: heart rate 144 beats/min, percutaneous oxygen saturation (SpO2): 99% and Blood pressure: 95/48 mmHg. For anesthesia induction; 5 mg/kg thiopental, 1 mg/kg lidocaine, 2 µg/kg of fentanyl and 0.1 mg/kg vecuronium bromide were used. After intubation with 4.0 spiraled endotracheal tube, 50%/50% O2 and air mixture and 1.5-2.5% concentration of sevoflurane were used for maintenance of anesthesia. From 10 µg/mL remifentanil infusion, dose of 0.1 µg/kg/min was used for preoperative analgesia. For invasive blood pressure monitoring, left radial artery modified Allen test was applied to the patient and after that arterial cannulation was made with 22 G catheter. A second peripheral venous route was opened from right cubital fossa region with 22 G catheter. For replacement fluid, 0.45% NaCl and 5% dextrose was used. Peri-operative blood sugar (glucose) was kept between 90-150 mg/dl, systemic arterial pressure was kept between 90-110 mmHg, and end tital carbon dioxide (ETCO2) values were kept betweeb 35-40 mmHg. Heart rate was between 80-120 beats / min and the esophageal body temperature was between 35.5-36.5 ℃. For prevention of body temperature decrease, external heater blanket was used and operation room temperature was elevated to 23-24 ℃. Operation lasted for about 6 hours. Total 700 ml of liquid was given. There was 170 ml urine output. The patient did not need blood transfusions. After operation, patient was taken to intensive care unit as intubated. was Patient was extubated in the intensive care unit at post-operative day 1.

Discussion

MMD was defined in Japan for the first time and it is a chronic occlusive cerebrovascular disease which is characterized with bilateral spontaneous stenosis of intracranial arteries at the base of the brain and widespread collateral vascularization (moyamoya vessels) [2,6]. The disease was first reported in 1957 by Takeuchi and Shimizu as Bilateral ICA hypogenesis and in 1969 it was named as moyamoya by Suzuki and Takaku. Diagnosis of the disease is made by cerebral angiography [1]. In our case diagnosis was ensured with digital subtraction angiography (Figure 1-2).

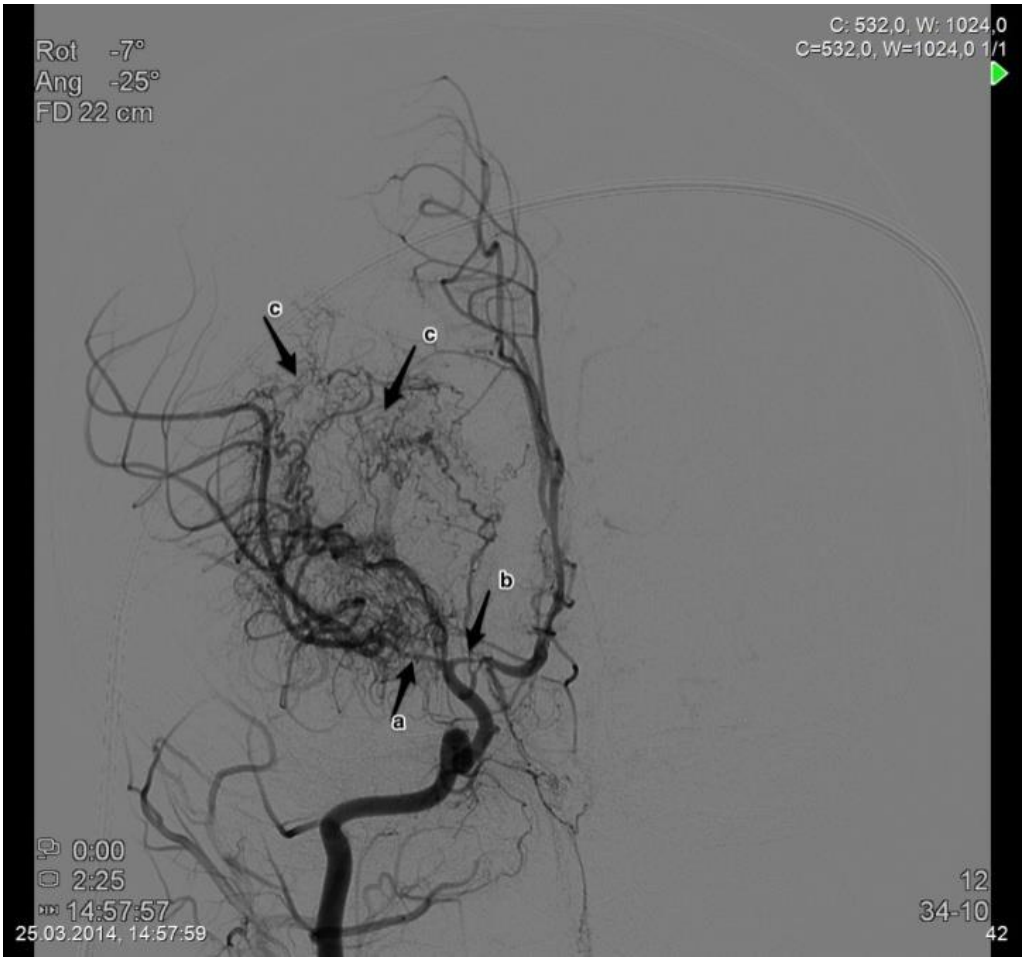

Figure 1. In the imaging of right internal carotid artery (ICA) with digital subtraction angiography; occlusion of the front and middle cerebral artery (MCA) and little collateral branches (moyamoya vessels) were observed, Towne view. a- ACA, b-MCA proximal stenosis, c- Leptomeningeal collateral

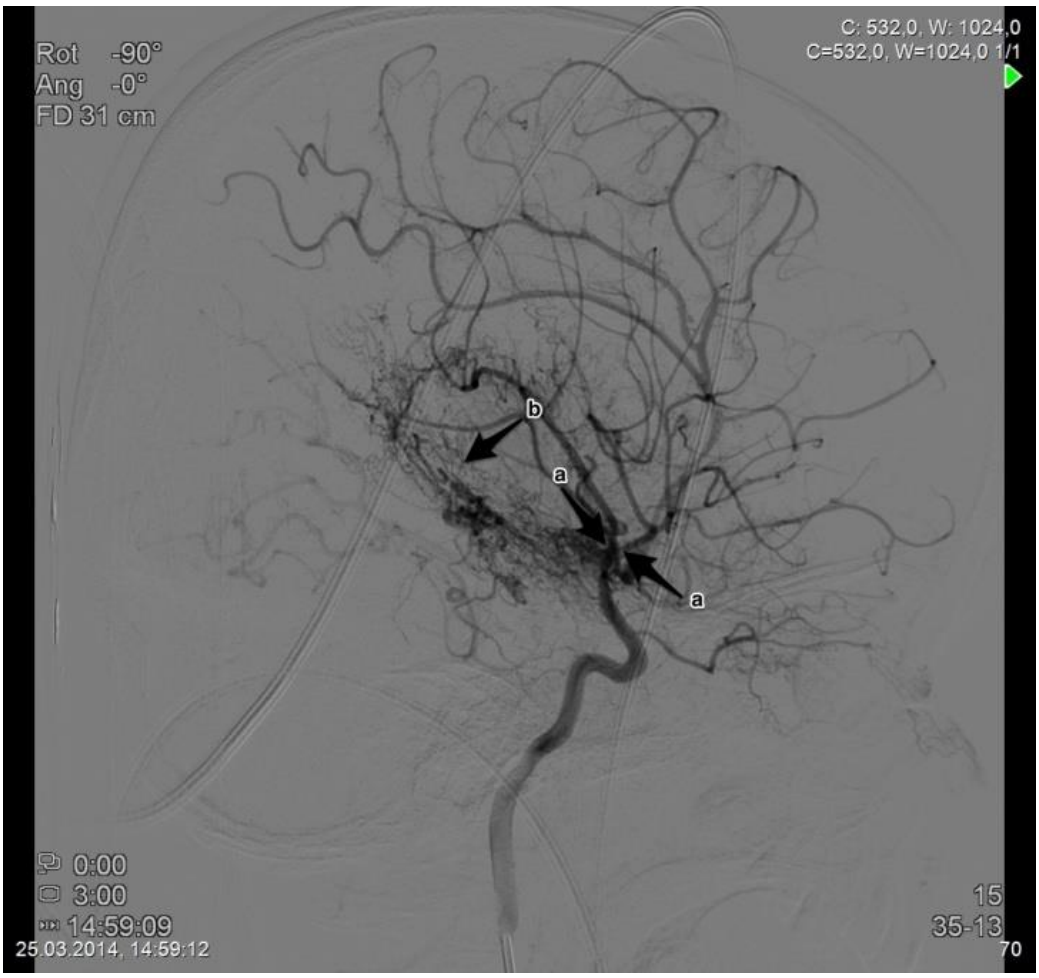

Figure 2. In the imaging of right ICA with digital subtraction angiography; occlusion of the front and MCA and little collateral branches (moyamoya vessels) were observed, side view. a- ACA, b- MCA proximal stenosis, c- Leptomeningeal collateral

The history and physiological status of the patient should be focused on during the pre-operative evaluation of patients; cerebral perfusion disorder and the presence of any neurological deficits should be disclosed. In patients with Moyamoya disease; observation of signs and symptoms of transient ischemic attack or ischemic stroke is typical. Less often subarachnoid hemorrhage (is common in adults) symptoms, headaches and seizures can be seen. Transient ischemic attack history of patient causes increased risk for intra-operative and post-operative ischemic injury. Most of these patients might have mental retardation as a result of segmental motor deficit, epilepsy and chronic ischemia [7]. An experienced neuroanesthesia team is important for the success of surgery. Patients should continue to take aspirin or other antiplatelet agents before and after the surgery [8]. In the preoperative

anesthetic evaluation of our case, another vascular disorder was not seen but our patient had epilepsy and using antiepileptics for this disease.

If anxiolytic premedication is not given to children with Moyamoya disease preoperatively, hyperventilation due to crying might cause cerebral vasoconstriction. Cerebral vasoconstriction can cause reduced cerebral blood flow and finally cerebral ischemia. Because of this reason, using anxiolytic premedication for children with Moyamoya disease in preoperative period might be useful.[8] But it should be kept in mind that excessive sedation can cause hypercapnia which is a risk factor for ischemic complications [9]. If an intravenous route is going to be opened before anesthesia induction, topical anesthetic cream (EMLA) application can be used for preventing pain and pain-related crying [4]. In a study of Tagawa et al., they observed that transient hemiparesis occurs in crying children because of reduction in regional cerebral blood flow [10]. Effects of diazepam and midazolam on brain blood flow is minimal. Midazolam can be used in oral, nasal and intravenous routes. Dose of oral midazolam is 0.5-0.75 mg/kg (maximum 20 mg), nasal dose is 0.2-0.5 mg/kg, and intravenous dose for children under 5 years is 0.05-0.1 mg/kg and over 5 years is 0.025-0.5 mg/kg [6]. In our case; there was a previous vascular access but intravenous premedication was applied because of the reaction for leaving family in that age group.

There are several opinions for anesthetic management in surgery of Moyamoya disease. For the choice of anesthesia, total intravenous anesthesia (TIVA) in which propofol and opioids are used can be prefered or other anesthesia methods in which volatile agents and opioids are used can be selected [5]. The main objective in the management of anesthesia is protecting the balance between blood flow and oxygen consumption.

The aim of surgery is to increase cerebral blood flow. The general approach method in Moyamoya disease patients is using of revascularization methods in surgical procedures for increasing cerebral blood flow [5]. Surgical revascularization can be grouped under three main headings: direct, indirect and combined techniques [2,3,5]. Generally indirect technique is used in children and direct technique is used in adults. The reason for choosing indirect techniques in children is their safety and efficiency and also their applicability in patients to whom other by-pass techniques were applied. Only disadvantage is that the effective

collateral blood flow will develop after a few months [1]. In our case, a indirect technique, namely EDAS, was used.

In pediatric cases, recommended monitoring during surgery are ECG, non invasive blood pressure, if needed invasive blood pressure (if frequent monitoring of the cerebral perfusion pressure is needed), pulse oximetry, capnography, body temperature, urine output and partial pressure of carbon dioxide (PCO_2) monitoring [9]. Besides electrophysiological techniques (electroencephalography, somatosensory evoked potentials and motor evoked potentials) are also recommended in monitoring [8].

Anesthesia induction can be made via inhalation or intravenous route [7]. Induction of anesthesia by inhalation in children can be made with sevoflurane. But during induction with sevoflurane, it should be kept in mind that hypercapnia due to hyperventilation can occur. For intravenous induction in children; sodium thiopental, propofol or etomidate can be used. But thiopental and propofol can reduce cerebral perfusion pressure by causing hypotension. If hypotension occurs during induction or maintenance; blood pressure should be adjusted with a vasoconstrictor agent [9]. Non-depolarizing muscle relaxant should not cause histamine release and/or hemodynamic changes. For this purpose best option is vecuronium bromide in maintenance of anesthesia [7,9]. Intravenously given opioids and Lidocaine; may reduce response to laryngoscopy and tracheal intubation. Hypoxemia, hypercapnia, cough and jerking motion sould be avoided during laryngoscopy and tracheal intubation. Induction of anesthesia should not elevate intracranial pressure and reduce cerebral perfusion pressure [9]. Surgery of Moyamoya disease is a lengthy operation and because of this endotracheal tube should be well fixed for securing the airway [6]. In our case; sodium thiopental, lidocaine, fentanyl and vecuronium bromide as muscle relaxant were used for the induction of anesthesia.

For the maintenance of anesthesia; inhalation anesthesia or TIVA in which propofol and opioids or low dose volatile agents are used can be preffered. Besides in patients with moyamoya; general anesthesia or neuraxial techniques can be used for interventions which are not related with disease [9]. The main objective in the management of anesthesia is protecting the balance between blood flow and oxygen consumption. All volatile anesthetics cause cerebral dilatation and increase of cerebral blood flow with decrease of cerebral oxygen

consumption [7]. Isoflurane makes slight cerebral vasodilatation, it reduce cerebral metabolism or it can be protective against cerebral ischemia. Potent cerebral vasodilators such as halothane can cause steal phenomenon [2,4,7]. It was shown that cerebral vasodilators can cause cerebral steal in Moyamoya disease [9]. The dynamic autoregulation can be protected better in sevoflurane anesthesia when compared with isoflurane anesthesia [7]. In low doses, Sevoflurane can cause indirect vasoconstriction by reducing metabolic needs and in high doses it can cause vasodilatation due to its intrinsic effects. In doses with 1.3 MAC (minimal alveolar concentration) and more, brain blood flow clearly increases but intracranial pressure increase will not be seen due to autoregulation. Pharmacokinetic and pharmacodynamic properties of sevoflurane are similar with isoflurane and its vasodilatation effect is directly dose-dependent. In 1.5 MAC sevofurane; the brain autoregulation is protected but this is not applicable for the same concentration of isoflurane [11].

When making cerebral reconstruction in Moyamoya disease, normocarbia is recommended for optimum cerebral circulation [9]. Carbon dioxide is a powerful regulator of cerebrovascular tone. Changes in ventilation can effect cerebral blood flow and can be a major factor for determination of neurological complications [7]. Kurehara et al., showed in their study that hypercapnia during anesthesia could decrease cortical blood flow [12]. In the study of Chiu et al., it was showed that hypocapnia (level of $PaCO_2$ was 29 mmHg and lower) could decrease regional cerebral blood flow [13].

Maintaining blood pressure at or above the preoperative values perioperatively is important. A decrease of middle arterial pressure can cause cerebral blood flow resulting with cerebral ischemia and infarction and a decrease of cerebral perfusion pressure. Perioperative hypotension can occur due to antihypertensive therapy, dehydration, lack of resuscitation of fluids during surgery, blood loss and anesthetic agents. Hypotensive anesthetic technique should be avoided. Hyperosmotics drugs should be avoided due to dehydration and hypotension. Dopamine, phenylephrine and ephedrine should be used when hypotension evolves. If intracranial pressure increases, ventricular drainage can be used instead of hyperosmolar drug application. Many anesthetics and potential agents lower blood pressure therefore they should be used with caution and anesthetics should be titrated. When middle arterial pressure is between 60-160 mmHg, cerebral blood flow remains constant [2,7,8]. In

our case, anesthesia was maintained with sevoflurane and middle arterial pressure was maintained between 85-100 mmHg.

Although ideal hematocrit value is not known exactly, it is recommended to be in the range of 30-42%. In polycythemia; the risk of cerebral infarction associated with increased viscosity is available. Sato et al., showed in their study that in hematocrit values of 29% and below; cerebral ischemia can be provoked by decrease in blood oxygen carrying capacity despite the increase in cerebral blood flow [14]. In our case hematocrit values were between 30-33%.

Normothermia is recommended during surgery. Hypothermia causes of cerebral vasospasm and hyperthermia can cause an increase of metabolica rate by increasing oxygen consumption and possible cerebral ischemia [5,7]. In our case we kept the measuredesophageal body temperature between 35.5-36.5 ℃.

Urine output in patients under general anesthesia generally reflects the volume status. Low urine output has been associated with complications in Moyamoya disease [7]. Sato et al defined adequate urine output as 2,2 ml/kg/hour [14]. In our case urine output was 2 ml/kg/hour.

After surgery, imaging in these patients is usually done with computed tomography. They should be followed-up in postoperative intesive care unit. Postoperative pain, predisposes cerebral infarction and ischemic attacks. Therefore there is a need for good pain management. Another postoperative problem is nausea and vomiting. Nausea and vomiting should be avoided in order to prevent raised intracranial pressure. For this purpose; 4 or 8 mg of ondansetron can be applied intraveously during dura closure [8]. Kim et al., investigated effect of dexamethasone in combination with low dose propofol and dexamethasone alone on nausea and vomiting in children with Moyamoya disease. They did not find any difference in terms of nausea and vomiting between two groups [15].

In conclusion, anesthesia management of Moyamoya disease is as important as surgical management. For prevention of the development of neurological complications; intraoperative normocapnia, normotension, normothermia and adequate fluid resuscitation is needed.

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