

Complication Incidence and Management of Adult and Pediatric Moyamoya Disease after Bypass Surgeries

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Abstract

Background: Multiple literatures have shown that extra-intracranial vascular direct bypass provides good outcome for patients diagnosed as moyamoya disease (MMD), but complications occurred frequently after surgeries. This study aims to discuss the postoperative complications, occurrence rate and postoperative management of MMD patients who underwent surgeries.

Materials and Methods: We retrospectively collected data of consecutive patients diagnosed with MMD and then underwent combination surgery of direct bypass and EDMS in our department between Jan 2013 to Dec 2015. Postoperative complications include acute brain infarction, intracranial bleeding and CHS. All patients were strictly monitored to avoid hypovolemia, low blood pressure and anemia after surgeries. Edaravone was the main medicine used for postoperative treatment.

Results: Totally 368 consecutive patients (408 hemispheres) are included. The most frequent complication was CHS-related neurological deficits which were observed in 73 patients (19.8%). The most frequently occurred deficits was aphasia (88.5%). There were more left sided surgeries in the CHS plus brain infarction group (76.9% vs. 49.7%, $p < 0.05$). Furthermore, CHS and brain infarction seemed to occur more frequently in adult patients (21.3%) when compared to pediatric patients (16.7%), but not significantly ($p > 0.05$).

Discussion: Incidence of CHS is higher in left lateralization surgeries than right lateralization, and aphasia is the most frequent CHS symptom. Adult patients seem at higher risk for postoperative hyper-perfusion than pediatric patients, but not statistically significant in our study. It is important to observe CHS correctly and timely for neurosurgical nurses, in order to manage it properly.

Keywords: Moyamoya disease; STA-MCA bypass; Encephalo-Duro-Arterio-Myo-Synangiosis (EDMS); Post-operative complications; Post-operative management

Introduction

Moyamoya disease (MMD) is a type of cerebrovascular disease due to progressive stenosis to occlusion of the internal carotid artery system with compensatory hyperplasia of the basilar vascular network. It is more common in eastern Asia with high incidence of disability. Surgical revascularization like bypass surgery is the main treatment, which can effectively improve cerebral blood perfusion. Multiple literatures have shown that extra-intracranial vascular direct bypass provides good outcome for patients diagnosed as moyamoya disease (MMD) [1,2]. In our center, we usually prefer a combination surgery of direct bypass and Encephalo-Duro-Arterio-Myo-Synangiosis (EDMS) to treat MMD. If a bypass fails during surgery, we usually choose EDMS, i.e., indirect bypass. Although bypass surgeries have been improved significantly since the first report, in clinical practice, postoperative complications are commonly seen, within which cerebral hyper-perfusion syndrome (CHS) most frequently occurred, presenting with a variety of symptoms. It is important to understand

these complications and know how to deal with them for neurosurgical nurses. This study aims to discuss the postoperative complications, occurrence rate and postoperative management of MMD patients who underwent surgeries.

Materials and Methods

In this study we retrospectively collected data of consecutive patients diagnosed with moyamoya disease and then underwent combination surgery of direct bypass and EDMS in department of neurosurgery, Huashan hospital, between Jan 2013 to Dec 2015.

We reviewed paper and electronic chart documentations from both nurses and doctors to collect patient's demographic data and other information. Computed Tomography (CT), Magnetic Resonance Imaging (MRI), Single-Photon Emission Computed Tomography (SPECT), digital subtraction angiography (DSA) reports from radiological and neurosurgical department are also included in the charts. Neurosurgeons in our department usually used intraoperative indocyanine green (ICG) to confirm the patency of the anastomosis. Postoperative observation by neurosurgical nurses includes patients' vital signs, consciousness scoring, and signs of neurological deficits.

Abnormality found by observation will be discussed with patients' neurosurgeons.

Postoperative complications include acute brain infarction (confirmed by head MRI), intracranial bleeding (observed by head CT) and cerebral hyper-perfusion syndrome (CHS). CHS related neurological deficits includes aphasia, motor weakness, sensory disturbance. Symptom onset time and recovery time were also assessed. Postoperative CT scans were acquired in every patient at least twice, instantly and first day after surgeries. For cases who suffered neurological deficits, MRI scans would also be given to exclude acute brain infarction. Some patients may accept SPECT to evaluate brain perfusion.

Postoperatively, all patients were strictly monitored to avoid hypovolemia, low blood pressure and anemia, especially during the first 3 post-operative days. If lab tests showed hemoglobin level below 8.0 gram per deciliter (g/dl), blood transfusion would be considered to correct anemia. Systolic blood pressure was stabilized to be not below 100 mmHg, or above 140 mmHg. Patients are suggested to be on bed in the first 2 postoperative days. The surgical side shouldn't be compressed too much (e.g. elastic bandage net) to avoid occlusion of the bypass vessels. Edaravone was the main medicine used for postoperative treatment, especially when CHS symptom was observed.

Data Collection and Analysis

Totally 368 consecutive patients (408 hemispheres, mean age, 41.0 ± 8.4 years; range, 11.0–64.0 years) who underwent combined direct and indirect revascularization for MMD in our center are included. There were 356 adult patients and 12 pediatric patients (under 14 years old). Divided by onset type, there were 236 ischemic onset cases and 132 hemorrhagic-onset cases.

Only 1 case (0.3%) suffered postoperative cerebral hemorrhage, while 5 cases (1.4%) were found acute brain infarction by head MRI. The most frequent complication was CHS. CHS symptom onset varied from 1 to 10 days after surgeries (mean, 4.4 days). A mean duration of hyper-perfusion-related symptoms was 8.7 days post-surgically. CHS-related neurological deficits were observed in 73 patients (19.8%). The deficits included aphasia (most frequently occurred, 88.5%), motor weakness, sensory disturbance of the limbs. Some patients suffered clinical seizures (5 cases, 1.4%). Comparing patients group suffering neurological deficits (CHS plus brain infarction) and the others, we found there were more left sided surgeries in the former (76.9% vs. 49.7%, $p < 0.05$, χ^2 test), indicating that the left sided surgeries tended to cause the deficits. CHS and brain infarction seemed to occur more frequently in adult patients (21.3%) when compared to pediatric patients (16.7%), but not significantly ($p > 0.05$, χ^2 test).

Results

Bypass surgery has become the choice surgical strategy for treating MMD and been proven safe [3]. A combination of direct and indirect bypass surgery is suggested recently and has become the surgical treatment for most adult patients as well as some pediatric patients. However, postoperative complications happen and it's better for medical staff especially neurosurgical nurses to know more about and further deal with them. Among multiple types of complications, CHS is the most common one, especially after the direct bypass surgeries and the symptoms appear quite various across cases [3]. Left sided bypass surgeries tend to cause CHS more frequently [4]. In our study, we found aphasia is the most frequent type of CHS related neurological

deficits, and the left side surgeries tend to cause CHS. Several hemodynamic factors could be reasons to cause such complications in MMD, including a pressure gradient caused by autoregulatory vasodilation of recipient vessels in response to cerebral ischemia, a large superficial temporal artery size relative to the recipient artery, poor distribution of bypass flow and free radical damage [4-6].

Fortunately, most of these neurological deficits caused by CHS disappear with time, on average in 8.7 days after surgery in our study. Medication of free radical scavenger edaravone is broadly used for postoperative situation of MMD and is believed to be useful for CHS and perioperative application of it is suggested [4]. Postoperative observation by well-trained neurological nurses is important to discover the deficits and beneficial for further medical treatment and physical/ psychological recovery for patients. Usually all neurological symptoms are transient and disappear within several days by necessary medication, strictly controlling blood pressure and properly correcting anemia. In our experience, for the first 3 days after surgery, systolic blood pressure needs to be controlled strictly above 100 mmHg but below 140 mmHg. Anemia below 8.0 g/dl needs to be corrected by blood transfusion which is beneficial for patient's recovery.

Discussion

It is stated in literature that adult patients with moyamoya disease are at high risk for symptomatic hyper-perfusion after surgery when compared to pediatric patients [2,7]. Our study showed similar results, but the significance is weak since only a few pediatric cases (12 cases) were included. The incidence of symptomatic hyper-perfusion is around 20% in adult patients with moyamoya disease [2,4,8], our study showed concordant results. Incidence of radiological hyper-perfusion could be even higher [2]. Thus, it is important to observe CHS correctly and timely, in order to manage it properly [9,10].

Conclusion

Our study showed incidence of CHS is higher in left lateralization surgery than right lateralization, and aphasia is the most frequent CHS symptom, which showed concordant results to literature. Adult patients seem at higher risk for postoperative hyper-perfusion than pediatric patients, but not statistically significant in our study. Larger sample studies including more pediatric patients are required to evaluate the difference between them.

Compliance with Ethical Standards

This manuscript has been read and approved by all authors, who acknowledge due care in ensuring the integrity of the work. All authors have made substantial contributions to the design, collection, analysis and/or interpretation of the data, and many have contributed to the writing and intellectual content of the article.

Ethical Approval

This study was approved by the Administration Committee of Human, Shanghai, China.

Statement of Human and Animal Rights

All of the experimental procedures involving patients were conducted in accordance with the Institutional Human Care guidelines

of Huashan Hospital, Fudan University, China and approved by the Administration Committee of Human, Shanghai, China.

Statement of Informed Consent

Verbal informed consent was obtained from the patients for their anonymized information to be published in this article.

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Conflicts of Interest

The authors declare that they have no conflicts of interest. Equal contribution on this work as the first authors by Zhang Q, Feng R and Zhang X.

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