

Efficacy of Superficial Temporal Artery-Middle Cerebral Artery Double Anastomoses in a Patient with Rapidly Progressive Moyamoya Disease: Case Report

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Abstract

Moyamoya disease can be associated with a rapidly progressive course in young patients. This report describes a patient with moyamoya disease who experienced rapid disease progression, resulting in cerebral infarction and a wide area of diminished cerebral perfusion. Double superficial temporal artery (STA)-middle cerebral artery (MCA) anastomoses were utilized to immediately increase cerebral perfusion in the affected area. This case involved a 5-year-old girl who had been diagnosed with moyamoya disease and had undergone STA-MCA anastomosis with indirect bypass in the right hemisphere at the age of 3. At the time of presentation, magnetic resonance (MR) imaging showed cerebral infarction at the left frontal lobe, and MR angiography showed rapidly progressive narrowing of the left MCA that had not been present 3 months prior. N-isopropyl-p-[I123] iodoamphetamine single-photon emission computed tomography (IMP-SPECT) showed markedly decreased uptake in the left hemisphere. She underwent emergent STA-MCA double anastomoses with indirect bypass on the left side. IMP-SPECT showed marked increase in uptake in the left hemisphere. The anterior cerebral artery (ACA) territory adjacent to the cerebral infarction also showed increased uptake on the SPECT. Postoperatively, there were no clinical or radiographic indications of ischemic or hemorrhagic complications. Double anastomoses are effective in quickly and significantly increasing blood flow. The postoperative course in this case was uneventful. Double anastomoses are a surgical option for patients with moyamoya disease who show rapid disease progression, even in those in the acute phase of cerebral infarction.

Key words: cerebral infarction, superficial temporal artery-middle cerebral artery double anastomoses, moyamoya disease

Introduction

Moyamoya disease is characterized by progressive arterial stenosis or occlusion of the bilateral intracranial carotid artery (ICA) and the development of extensive collateral vessels (moyamoya vessels).¹⁾

It is a cause of stroke and transient ischemic attack (TIA) in both pediatric and adult patients. Surgical revascularization for moyamoya disease can prevent cerebral ischemic attacks by improving cerebral blood flow (CBF), and indirect bypass, such as encephalo-duro-arterio-synangiosis (EDAS), encephalo-myo-synangiosis (EMS), or encephalo-gareo-synangiosis (EGS) with or without direct bypass, such as superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis, is generally employed as the standard surgical treatment for moyamoya disease.

Indirect bypass surgery is most commonly performed for the pediatric patient with moyamoya disease. However, some pediatric patients exhibit a rapidly progressive course that results in multiple cerebral infarctions. In such cases, indirect bypass does not prevent subsequent cerebral infarction before the growth of a newly formed vascular network by indirect revascularization. Therefore, direct bypass, such as STA-MCA bypass, is required for rapidly progressive cases.

The present report describes a case of a patient with moyamoya disease who showed rapid disease progression that resulted in cerebral infarction in the anterior cerebral artery (ACA) territory and considerably diminished cerebral perfusion within an extensive area in the ACA and MCA territories. We consider that a single STA-MCA anastomosis was not sufficient to increase the cerebral perfusion of such a wide ischemic area; therefore, double STA-MCA anastomoses were used to immediately increase cerebral

perfusion in the affected area. The result was cerebral perfusion improvement without any adverse effects.

Case Report

A 5-year-old girl was referred to our department with right lower extremity paresis and transient motor aphasia. She had a previous history of left-sided hemiconvulsion as an initial symptom of moyamoya disease at the age of 3 years and had undergone STA-MCA single anastomosis with encephalo-gareo-duro-myo-synangiosis (EGDMS) on the right side. Her postoperative course was uneventful, and she had been discharged without neurological deficit. On admission for the present evaluation, she was suffering from repeated episodes of aphasia and right-sided leg weakness.

I. Imaging examination

Magnetic resonance imaging (MRI) demonstrated cerebral infarction at the medial side of the left frontal lobe in the territory perfused by the ACA (Fig. 1A). Magnetic resonance angiography (MRA) revealed stenosis of the terminal portion of the left internal carotid artery that had not been detected on similar studies performed 3 months prior (Fig. 2A, B). N-isopropyl-p-[I123] iodoamphetamine single-photon emission computed tomography (IMP-SPECT) showed markedly decreased uptake in the left fronto-temporal region that was not detected previously (Fig. 3A, B).

II. Operation

The usual strategy, consisting of single STA-MCA anastomosis with EGDMS, was not thought not to be sufficient to address the wide area of hypoperfusion area in a rapid fashion. Thus, double STA-MCA anastomoses were elected. Two days after admission, the patient underwent left STA-MCA double anastomoses with EGDMS in the left hemisphere. The surgery began by exploring the frontal and parietal branch of the left STA under general anesthesia with careful attention to maintain blood pressure and avoid hypocapnia. Then, fronto-temporo-parietal craniotomy was performed. After dural incision, the recipient arteries at the M4 segment of the upper and lower trunk of the MCA were explored, and each artery was anastomosed to the frontal and parietal branch of STA, respectively. Finally, EGDMS was performed (Fig. 4).

III. Postoperative course

The postoperative course was uneventful, and the patient did not experience new neurological deficit or TIA after surgery. The IMP-SPECT study performed 3 days after surgery showed marked increased uptake in the left hemisphere, except for the area of infarction that was detected on MRI (Fig. 3C). MRI studies performed 7 days after surgery demonstrated no additional ischemic lesions (Fig. 1C). Edema or hemorrhage was not detected at the site of anastomosis or at the infarcted area. The patient's lower leg paresis that was present preoperatively gradually

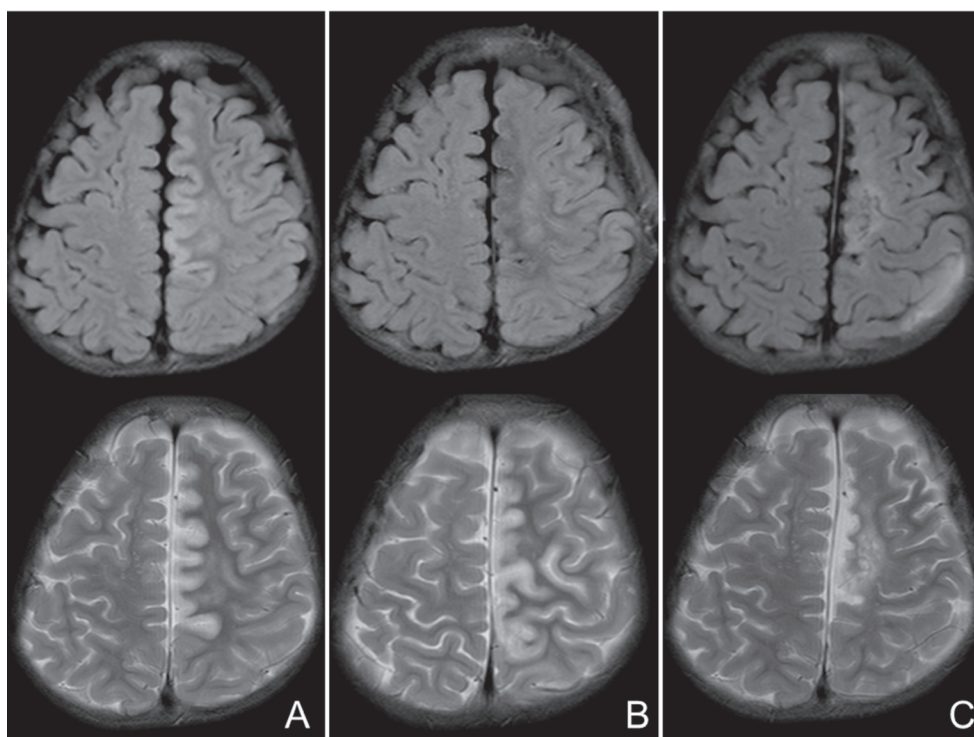


Fig. 1 Magnetic resonance imaging study of the fluid-attenuated inversion recovery (*upper*) and T₂-weighted image (*lower*). A: Images on admission. High signal is present in the territory perfused by the anterior cerebral artery (ACA). B: Postoperative images obtained 7 days after surgery demonstrate disappearance of the high signal. No new ischemic signs are present. C: 3 months after surgery. Infarcted area in the ACA territory has transformed to atrophy.

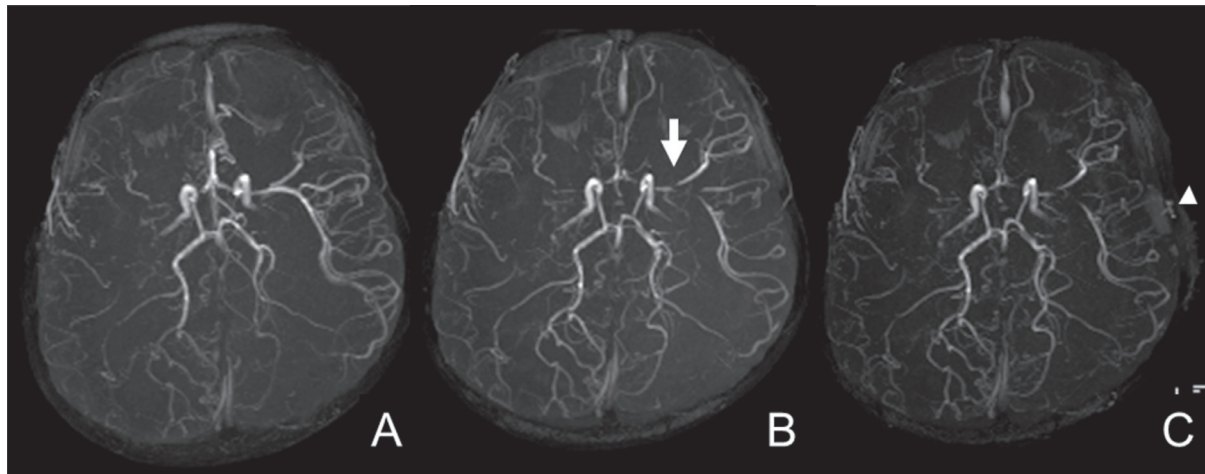


Fig. 2 Axial image of magnetic resonance angiogram. A: 3 months before surgery, the left middle cerebral artery (MCA) is patent. B: Preoperative image reveals immediate progression of the stenosis of the left M1 portion (arrow). C: Postoperative image obtained 7 days after surgery. Superficial temporal artery (STA)-MCA double bypass were patent, and STA did not show very high signal (arrowhead).

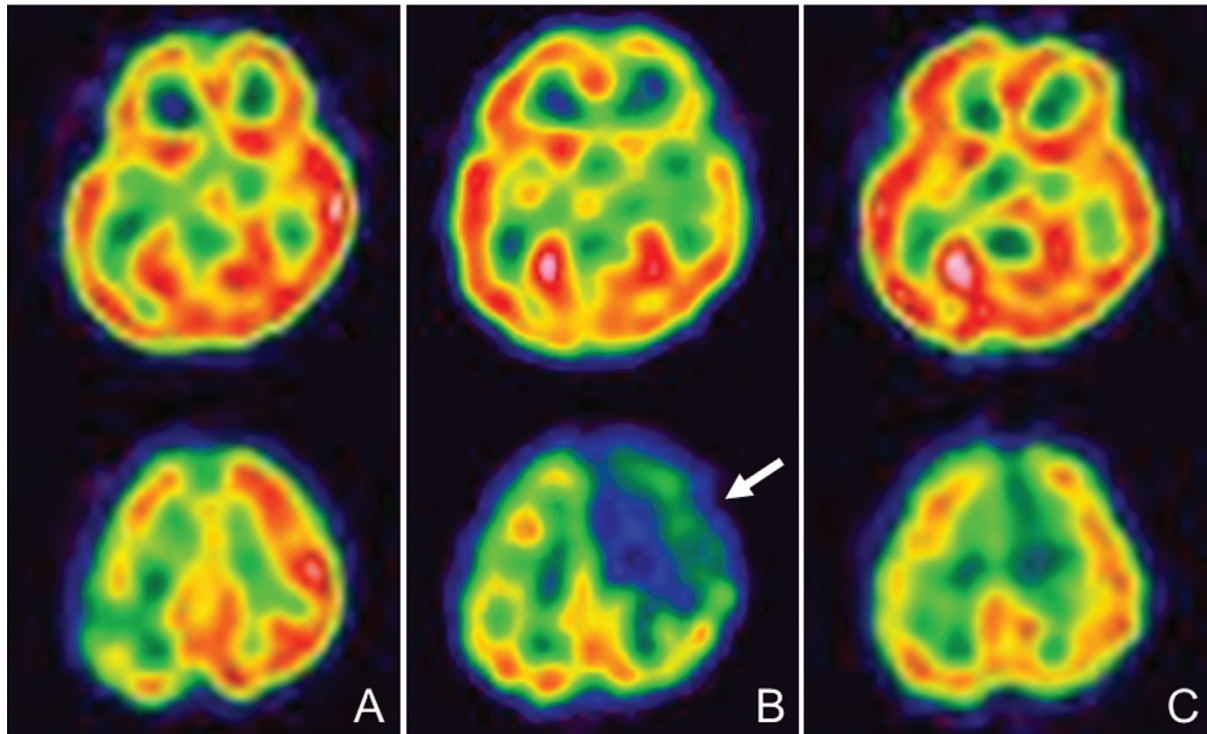


Fig. 3 Cerebral hemodynamic studies with resting states of N-isopropyl-p-[123] iodoamphetamine single-photon emission computed tomography images. A: 2 years before surgery. B: Preoperative images show poor perfusion in the right fronto-temporal lobe (arrow). C: Postoperative images obtained 3 days after surgery demonstrate a wide increase in the perfusion of the left hemisphere, with the exception of the area of infarction.

improved after surgery. The patient was discharged 15 days after surgery with mild gait disturbance.

Discussion

This is the first case report to describe detailed pre- and

postoperative radiological findings of pediatric moyamoya patients who underwent STA-MCA double bypass with indirect bypass in the acute phase of repeated ischemic stroke. Results from the present case support the notion that double STA-MCA anastomoses were effective; postoperative SPECT showed that this strategy resulted in a

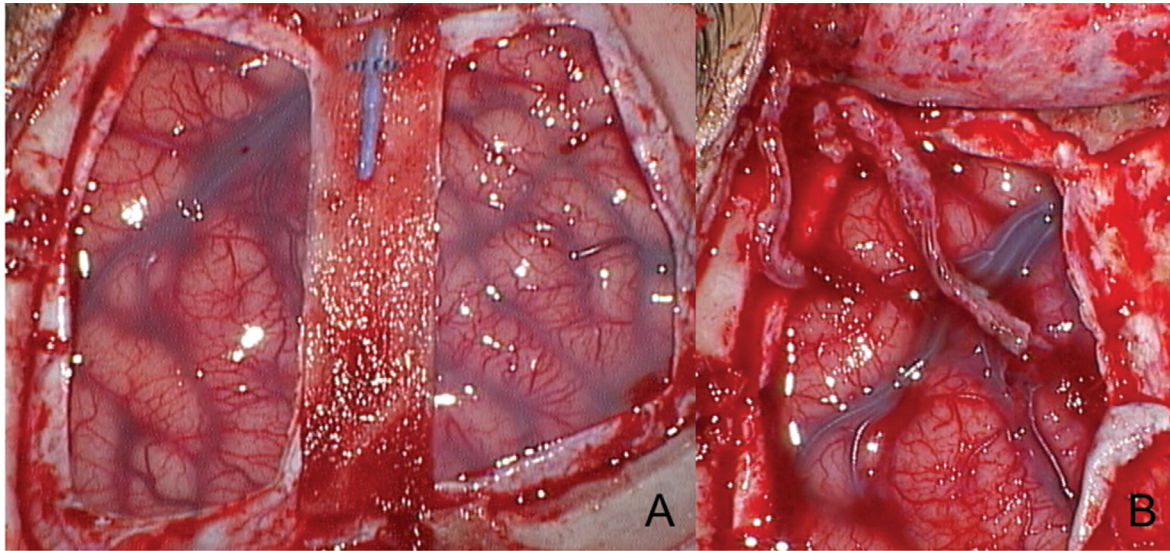


Fig. 4 Operative photographs. A: View after the indirect bypass surgery. B: View of the superficial temporal artery-middle cerebral artery double anastomoses sites on the left cerebral convexity.

wide, uniform, and marked improvement of uptake in the MCA territory and in the ACA territory without adverse postoperative effect.

Most pediatric patients with moyamoya disease develop ischemic symptoms due to TIA and cerebral infarction.²⁾ Several reports have shown that cerebral infarction was more common in younger than in older patients with moyamoya disease.^{3–5)} Therefore, early diagnosis and active surgical treatment are necessary, especially for young patients. Revascularization surgery with direct and/or indirect bypass surgery is the strategy of choice to eliminate the ischemic neurological symptoms.⁶⁾ Various types of surgeries have been proposed, including direct bypass, such as STA-MCA anastomosis; indirect bypass, such as EDAS, EMS, and EGMDS; and a combination of direct and indirect bypasses.^{7,8)} Our usual choice for the initial treatment of the patient with moyamoya disease is a combination of STA-MCA single anastomosis and EGMDS.⁴⁾ Direct bypass, such as STA-MCA anastomosis, is considered to be effective for prompt improvement of cerebral hemodynamics and might contribute to the reduction of the perioperative ischemic symptoms. On the other hand, the beneficial effects of indirect bypass techniques are not immediate because it takes 3–4 months for collaterals to develop and during that time there is a risk of perioperative ischemic stroke. Therefore, direct bypass will be useful if rapid increase in CBF is required.

To date, many clinicians have recommended that bypass surgery should be performed at least 4 to 6 weeks after the last ischemic episode. Therefore, most of the bypass surgeries for pediatric moyamoya disease with cerebral infarction are done in the chronic phase of cerebral infarction. However, when used in cases of

progressive ischemic stroke (as in the present case), this strategy would likely result in poor functional outcome due to further repeated cerebral infarction. Therefore, we believe that early revascularization surgery is indicated in such cases. In order to safely avoid instability during the acute ischemic phase, it is necessary to apply conventional therapy, such as preservation of blood pressure, avoidance of hyperventilation, and prophylaxis against seizure, to ensure stable cerebral blood supply. Nevertheless, the only treatment proven to improve cerebral hemodynamics is bypass surgery, which carries the risk of postoperative cerebral ischemia. Recently, Hayashi et al. described patients who suffered an ischemic event due to an insufficient increase in flow after STA-MCA single bypass in the early postoperative period.⁹⁾ They suggested that an instability state (called “watershed shift”), involving postoperative hemodynamic changes in the CBF, may occur due to disproportion of flow, resulting in a subsequent postoperative ischemic event in an area remote from the bypass. They also stated that patients with frequent preoperative ischemic events are at high risk of “watershed shift” and that a more aggressive procedure should be selected for such patients.

In response to their observation, we selected double STA-MCA anastomoses for our case. STA-MCA double anastomosis resulted in a wide and more uniform improvement in CBF when compared to single bypass.^{10,11)}

While there was a concern that a rapid increase of the CBF due to direct bypass in the acute stage of cerebral infarction might cause cerebral edema, hemorrhage, and other reperfusion injuries, our case showed no significant perioperative complications. We hypothesized that the cerebral infarction in moyamoya disease was not a result

of cessation of CBF but was instead the result of decreased cerebral perfusion. In other words, the level of cerebral perfusion at the affected area was not low enough to result in endothelial injury but was low enough to induce neural death. This notion is supported by the fact that cerebral infarction in patients with moyamoya disease is often seen in the cerebral cortex region but not in the cerebral white matter. The ischemic threshold of the cerebral cortex is higher than that of the cerebral white matter, whereas the ischemic threshold of endothelial cells is lower than that of the white matter.¹²⁾

Another report has described the efficacy of double direct bypass. Ishikawa et al. reported that STA-MCA and STA-ACA double anastomoses with pan-synangiosis resulted in excellent outcome.¹⁰⁾ But they did not describe about the timing of their bypass surgery in their report. Moreover, this procedure requires quite advanced techniques and is not always selected as a general option for pediatric moyamoya patients, mainly because of the difficulty in securing enough length of STA for STA-ACA bypass and anastomosing quite small vessel during STA-ACA bypass in pediatric patients, as they say in their literature. On the other hand, STA-MCA anastomosis is a well-established technique, and to add another STA-MCA anastomosis is not difficult if we invest a little time.

In these terms, we expected that double STA-MCA bypass with indirect bypass surgery in the acute ischemic phase can ensure stable blood supply without causing reperfusion injury.

Conclusion

Based on these observations, we conclude that double STA-MCA bypass is an effective and safe strategy for the management of rapidly progressive pediatric moyamoya patients with a widespread ischemic area on SPECT study that is larger than the infarcted area detected on MRI. However this is only one case report, and further investigation of a larger number of patients is required to define the comparative utility of either double bypass with indirect bypass or single bypass with indirect bypass for cases of rapidly progressive stroke in pediatric patients with moyamoya disease.

Conflicts of Interest Disclosure

The authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices in the article. All authors who are members of The Neurosurgical Society (JNS) have registered online Self-reported COI Disclosure Statement Forms through the website for JNS members.

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