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Multiple EDAS (encephalo-duro-arterio-synangiosis). Additional EDAS using the frontal branch of STA and the occipital artery for pediatric Moya-Moya patients in whom EDAS using the parietal branch of STA was insufficient

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Introduction: Although parietal EDAS or STA-MCA anastomosis are effective in pediatric Moya-Moya disease, they do not adequately prevent ischemia in the frontal and occipital lobes. Some additional methods that can prevent ischemia in the frontal and occipital lobes are sometimes needed. We investigated whether EDAS using a frontal branch of the superficial temporal artery (frontal EDAS) or EDAS using the occipital artery (occipital EDAS) is preferable.

Patients and Methods: Frontal or occipital EDAS was performed at 15 sites in seven patients with pediatric Moya-Moya disease. The outcome was evaluated by angiography 3 months later, CT findings 3 months later, neurological findings during the follow up period and perioperative complications. The mean follow up period was 15 ± 6 months after frontal or occipital EDAS.

Results: Good revascularization from frontal or occipital EDAS was shown in eleven of fifteen surgical sites (73%) in angiography. None of the patients showed deterioration of symptoms after frontal or occipital EDAS during the follow up period. None of the patients developed surgical complications.

Conclusion: EDAS using the frontal branch of STA and the occipital artery is an effective and safe method for preventing ischemia in the frontal and occipital lobe on pediatric Moya-Moya disease.

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Efficacy of direct revascularization to prevent intracranial rebleeding in Moya-Moya disease

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Introduction: Moya-Moya disease is a clinical entity with spontaneous occlusion of the circle of Willis and most patients with this disease present with cerebral ischemia and/or intracranial bleeding. An intracranial bleeding is one of the major catastrophic events in the course of Moya-Moya disease. Intracranial bleeding in Moya-Moya disease is considered to be a result of ruptured weak Moya-Moya vessels which are forced to act as collateral pathways and are unusually under increased hemodynamic stress. But the mechanism underlying intracranial bleeding and the therapeutic modalities to prevent bleeding are not well known. With a hope to prevent intracranial bleeding as well as cerebral ischemia, we have performed direct revascularization (superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis) in the patients who presented with bleeding. To clarify the efficacy of direct revascularization to prevent rebleeding of Moya-Moya disease, we analyzed the clinical results of the patients who underwent STA-MCA anastomosis.

Methods: We retrospectively analyzed 48 patients with Moya-Moya disease who had an episode of intracranial bleeding and were treated in our institute. Occurrence of rebleeding, angiography findings, postoperative regression of Moya-Moya vessels and correlation of these factors were reviewed.

Results: Intracranial bleeding was counted 73 times in the 48 patients. The 96 hemispheres of these 48 patients, 53 had one or more bleeding and 43 had no bleeding. There was no difference in the Suzuki and Takaku's angiographic staging between the hemispheres with and without bleeding. We performed STA-MCA anastomosis on 74 hemispheres in 46 patients. Postoperative regression of the Moya-Moya vessels was confirmed in 38 of 55 hemispheres which had been studied by angiography after surgery. Rebleeding was counted 12 times in 9 patients before treatment and 9 times in 7 patients after revascularization surgery. Of these 9 postoperative rebleeding, 4 occurred in the operated hemispheres and 5 were in the non-operated ones. All of 4 hemispheres which had been revasculized and had rebleeding did not show regression of Moya-Moya vessels.

Conclusion: Direct revascularization has a potential to reduce hemodynamic stress on weak Moya-Moya vessels and is considered to be effective for the prevention of rebleeding, but in limited cases.

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Omental transplantation for paraparetic transient ischemic attacks in childhood Moya-Moya disease. Hemodynamic evaluation

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Introduction: The hemodynamic mechanisms responsible for the appearance of parapateric transient ischemic attacks in ten patients with childhood Moya-Moya disease who subsequently underwent bifrontal omental transplantation were investigated.

Materials and Methods: Cerebral perfusion (CP) was measured with 99mTc-hexamethyl-propyleneamine oxime single photon computed tomography prior to and after administration of 10 mg/kg of acetazolarnide. Cerebral perfusion was obtained by dividing radioisotope uptake per pixel in regions of interest by that in cerebellum. Hemodynamic reserve was defined as [CP after acetazolarnide – CP before acetazolarnide/CP before acetazolarnide × 100.

Results: Amounts of CP in the anterior portion of the frontal lobe and in the paracentral lobule were 0.70 ± 0.04 and 0.74 ± 0.03 , respectively, before appearance of the transient ischaemic attacks. The latter was significantly higher than the former (p < 0.0001). Hemodynamic reserves were -11.1 ± 2.8 and 9.6 ± 3.0 , respectively, at that time. These two parameters were significantly decreased just after paraparetic transient ischemic attacks and two parameters in the paracentral lobule were more decreased than those in the anterior portion of the frontal lobe. But these increased again after bifrontal omental transplantation in these two regions.

Conclusions: The watershed region was located anterior to the paracentral lobule before appearance of the transient ischaemic attacks, and widened and moved backward to include the paracentral lobule just before appearance.

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Changes in cortical CBF and vascular response after vascular reconstruction in patients with adult onset Moya-Moya disease

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Introduction: In adult onset Moya-Moya disease patients who underwent vascular reconstructive surgery, we evaluated changes in regional cerebral blood flow (rCBF) and vascular reserve (Δ CBF) and assessed the newly perfused cerebral area.

Methods: In 7 patients who underwent vascular reconstructive procedures in 12 hemispheres, rCBF and ΔCBF were measured by ¹³³Xe SPECT with acetazolamide loading before and 6 months after surgery. Newly perfused cerebral areas were determined by the intra-arterial injection of ^{99m}Tc-HMPAO into the bypass artery.

Result: (1) Average preoperative rCBF at the postoperatively positive perfusion regions from the bypass artery (57 regions) was 41.6 \pm 12.0 which was slightly below the lower limit of normal range and Δ CBF at this regions was 3.6 \pm 19.8% showing significantly lower than the normal value. However, the average preoperative rCBF and Δ CBF in post-operatively negative perfusion regions (115 regions) were 55.4 \pm 9.6 and 30.6 \pm 24.1%, respectively, where both values were normal. (2) After surgery, in the positive perfusion regions rCBF was significantly increased to 57.0 \pm 13.6 up to the normal range and Δ CBF had slightly increased to 12.3 \pm 13.5%, but the normal range and Δ CBF had slightly increased to 12.3 \pm 13.5%, but was still significantly below the normal value. In the negative perfusion regions, no significantly changes were noted.

Conclusion: Cerebral perfusion from the bypass arteries mainly developed in the low rCBF and low \triangle CBF territory after the vascular reconstruction in the patients with adult onset Moya-Moya disease. Although rCBF was significantly improved after surgery, the restoration of vascular reserve was not significantly attained even in the regions perfused by bypass arteries.

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Follow-up study of Moya-Moya and Moya-Moya-like diseases in adults

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Introduction: Surgical indication for Moya-Moya disease with hemorrhagic episodes is still controversial. In this report, the authors studied postoperative follow-up results to determine the benefit of direct or indirect bypass surgery for patients with Moya-Moya and Moya-Moya like disease.

Patients and Methods: This series included 14 patients with Moya-Moya disease (Group M) and 9 patients with Moya-Moya like Disease (Group O). Nine of Group M and six of Group O had direct or indirect bypass surgery. 6 patients had follow-up SPECT study with Diamox challenge. The follow-up period ranged from 6–312 months with mean of 96 months.

Results: Group M: Five of 9 patients with bypass surgery had episodes of rebleeding postoperatively, while two of 5 patients without surgery had rebleeding. Group O: Three of 6 patients with bypass surgery had rebleeding postoperatively, while none of 3 patients without surgery had rebleeding.

The time interval to rebleeding ranged from 1–252 months with mean of 114 m. All 3 patients with rebleeding showed poor neo-vascularization and poor response to Diamox challenge in the postoperative follow-up studies. In the three patients without rebleeding, good neo-vascularization and cerebrovascular reserve were found.

Conclusion: The benefit of surgical procedure to prevent rebleeding is not conclusive, but the risk of rebleeding could be reduced in the selected patient