

Moyamoya disease

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A 54 year old female patient, with a history of fronto-temporal stroke, presented to our hospital for neurosurgical evaluation.

Angiography was obtained observing fine caliber right internal carotid and occlusion at the birth of the left ophthalmic artery with repermeabilization of the anterior cerebral artery by branches of the anterior choroidal and ophthalmic arteries. Left middle cerebral artery could not be seen (Figure 1A, arrow head) but there was a slight enhancement on the contralateral vessel due to collateral circulation from posterior choroid, temporal and middle meningeal arteries. At the vertebrobasilar territory collateral circulation could be observed with leptomeningeal collateralization (Figure 1B, black arrows). Perfusion magnetic resonance imaging (PWI-MRI) (Figure 2) of the brain was later obtained showing a reduction in volume (CBV) and blood flow (CBF) (A) in right frontal, temporal and occipital areas together with an increase in the mean transit time (MTT) (B) and time to peak (TTP) (C) in the right temporal and left frontal areas. Computer-guided left indirect revascularization surgery was programmed with good results, after which she presented no new ischemic events. Six months later the patient underwent computer-guided right indirect revascularization surgery.

Even though the role of PWI-MRI for pre and postoperative assessment of hemodynamics in Moyamoya disease has not yet been defined, it may be useful to guide treatment and predict its efficacy.



Figure 1. Angiography of intracranial vessels, lateral view. A: Arterial circulation showing a significant caliber reduction at the birth of the left middle cerebral artery. B: Vertebrobasilar territory with important collateral circulation.

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Figure 2. Perfusion magnetic resonance imaging of the brain, axial view. A: It shows a reduction in volume and blood flow in right frontal, temporal and occipital areas. B: increase in the mean transit time in right frontal, temporal and occipital areas. C: increase in time to peak in the right temporal and left frontal areas.

As the angiography gives an accurate image of the vascular anatomy, the PWI-MRI or PWI-CT may offer a better global understanding of the impact those alterations may have on the central nervous system's metabolism, providing data of CBF, CBV and blood transit times.

This study uses water as a tracer and is, therefore, safe to use in patients with kidney injury or with contrast mediated adverse reactions.

It is important for healthcare professionals to have a better understanding of image acquisition; however, further studies are needed to fully understand and determine its indications and value. **Conflicts of interest:** the author declares that he has no conflicts of interest.

REFERENCES

- Ikeda H, Sasaki T, Yoshimoto T, et al. Mapping of a familial moyamoya disease gene to chromosome 3p24.2-p26. Am J Hum Genet 1999; 64:533.
- Inoue TK, Ikezaki K, Sasazuki T, et al. Linkage analysis of moyamoya disease on chromosome 6. J Child Neurol 2000; 15:179.
- Sakurai K, Horiuchi Y, Ikeda H, et al. A novel susceptibility locus for moyamoya disease on chromosome 8q23. J Hum Genet 2004; 49:278.
- Huang S, Guo Z-N, Shi M, et al. Etiology and pathogenesis of Moyamoya Disease: An update on disease prevalence. Int J Stroke. 2017;12(3):246– 253. https://doi.org/10.1177/1747493017694393