

Constructive Interference in Steady State Imaging of Moyamoya Disease

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Abstract

The diagnostic accuracy of three-dimensional constructive interference in steady state (CISS) magnetic resonance (MR) imaging was evaluated for the assessment of idiopathic moyamoya disease. Six consecutive patients underwent MR angiography, CISS imaging, and digital subtraction angiography. MR angiography and CISS imaging visualization of the steno-occlusive changes in the distal internal carotid arteries and the development of moyamoya vessels in the basal cistern were compared to the results obtained by digital subtraction angiography. MR angiography revealed the steno-occlusive changes correctly in nine and overestimated the changes in three of 12 hemispheres examined. CISS imaging showed the steno-occlusive changes defined as decreased caliber of the internal carotid artery correctly in two, underestimated the changes in nine, and overestimated the changes in one of the 12 hemispheres. MR angiography detected moyamoya vessels correctly in five and underestimated the vessels in seven of the 12 hemispheres. CISS imaging revealed the moyamoya vessels correctly in 10, underestimated the vessels in one, and overestimated the vessels in one of the 12 hemispheres. CISS imaging can supplement MR angiography in the non-invasive diagnosis of moyamoya disease, especially for the evaluation of moyamoya vessels.

Key words: constructive interference in steady state magnetic resonance imaging, moyamoya disease, magnetic resonance angiography

Introduction

Moyamoya disease is characterized by progressive steno-occlusive changes within the terminal portions of bilateral internal carotid arteries and by the development of so-called 'moyamoya' vessels at the base of the brain.^{11,15)} The clinical manifestation of moyamoya disease is typically brain ischemia in children and brain hemorrhage in adults.

The diagnosis of moyamoya disease is based upon the clinical manifestation and angiographic findings. Repeated angiographic studies are frequently required to evaluate the chronological progression of both steno-occlusive changes in the cerebral arteries and the collateral blood flow. Conventional or digital subtraction angiography is invasive and not without risk, so should be avoided if other diagnostic methods are available, especially in pediatric patients. Magnetic resonance (MR) imaging has developed an increased importance in the diagnosis of moyamoya disease due to its inherent non-invasive-

ness.^{1,5-7,9,12-14,16-18)} Although MR angiography is useful in the diagnosis of moyamoya disease, this technology may overestimate the steno-occlusive changes in the internal carotid arteries and underestimate the development of moyamoya vessels.^{9,13,16-18)}

Three-dimensional constructive interference in steady state (CISS) MR imaging, a steady-state free precession sequence, provides high spatial resolution of the brain structure and high contrast between the cerebrospinal fluid and extramedullary structures.^{2,3,8)} The high spatial resolution of CISS images may provide supplementary information to that obtained with MR angiography concerning the steno-occlusive changes in the cerebral arteries and moyamoya vessels. This study evaluated the usefulness of CISS images for the diagnosis of moyamoya disease.

Materials and Methods

Six consecutive patients (3 females and 3 males) aged 8 to 67 years old (mean 28.3 years) underwent

Table 1 Correlation between digital subtraction angiographical (DSA) and magnetic resonance (MR) angiographical evaluations of the terminal portions of the internal carotid arteries (ICAs)

ICA on MR angiography	ICA on DSA			
	No stenosis	Mild stenosis	Marked stenosis	Occlusion
No stenosis	0	0	0	0
Mild stenosis	0	0	0	0
Marked stenosis	0	2	3	0
Occlusion	0	1	0	6

both digital subtraction angiography and MR imaging examination. All patients had definite moyamoya disease based on the clinical presentation and angiographic findings. The initial clinical presentation was during childhood in four patients and as adults in two patients. The initial clinical symptom was ischemia in all six patients. Four patients were preoperative or had not received surgery, and two patients were postoperative.

The MR system was a 1.0-Tesla superconductive system (Harmony; Siemens, Erlangen, Germany). MR imaging parameters for MR angiography were 0.90 mm thickness, repetition time (TR) 39 msec, echo time (TE) 10 msec, flip angle 25°, voxel size 0.78 × 0.39 × 0.90 mm, and one excitation. MR imaging parameters for three-dimensional CISS images were 0.86 mm thickness, TR 17 msec, TE 8.1 msec, flip angle 70°, voxel size 0.70 × 0.70 × 0.86 mm, and one excitation. The gray scale of the CISS images was reversed to better visualize the fine structures in the cisterns.

MR angiography and CISS images, as well as digital subtraction angiography, were performed on all six patients within a week. Maximum intensity projection images were used to evaluate MR angiograms in all six patients, and the source images were used in five patients. Steno-occlusive change of the vessels on CISS images was defined as decreased caliber of the vessels.

Visualization of both steno-occlusive changes within the terminal portions of the intracranial internal carotid arteries and the moyamoya vessels in the basal cisterns were evaluated in 12 cerebral hemispheres based on the consenting view of two observers (M.K. and M.N.). Digital subtraction angiography (1024 × 1024 matrices) was used as a gold standard for the other imaging modalities.

The following scoring system was applied for visualization of the terminal portion of the internal carotid arteries on MR angiograms and CISS images: 0 = no stenosis or normal caliber of the vessel, 1 = moderate stenosis or moderate decrease of the vessel caliber (<70%), 2 = marked stenosis or

marked decrease of the vessel caliber (>70%), and 3 = occlusion or non-visualization of the vessel. The scoring system for visualization of moyamoya vessels was as follows: 0 = no visualization, 1 = minimal visualization, 2 = moderate visualization, and 3 = marked visualization.

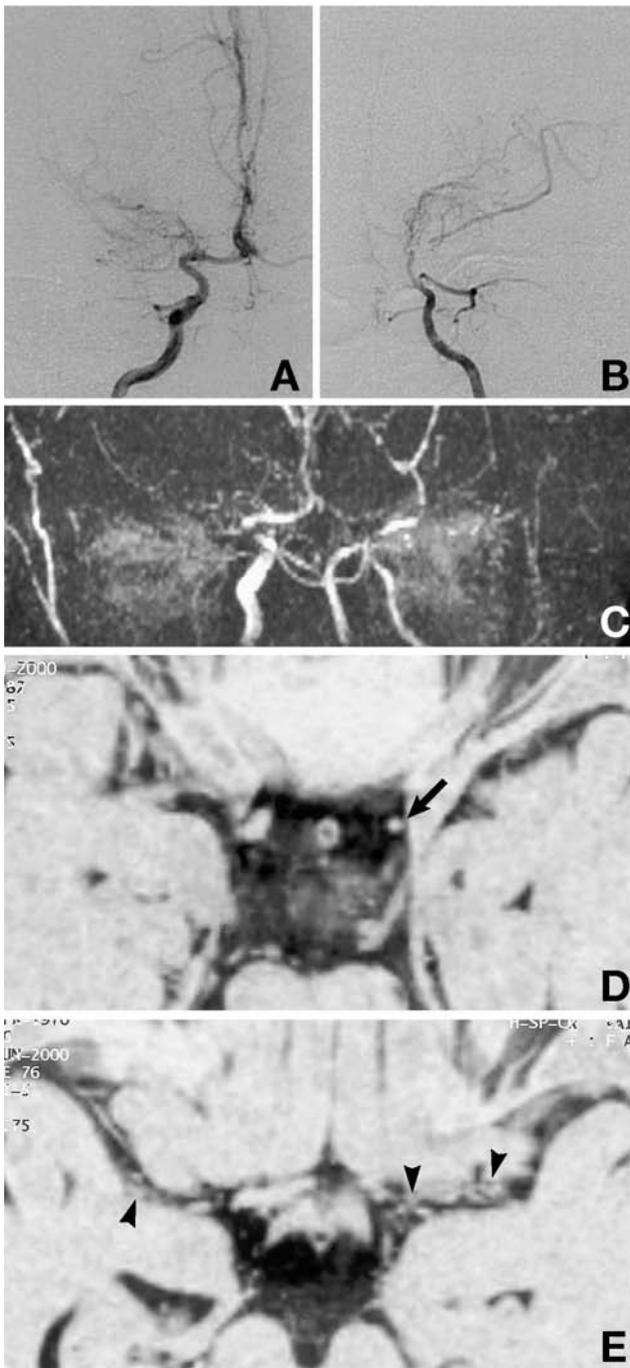
Results

MR angiography revealed steno-occlusive changes in the terminal portion of the internal carotid arteries correctly in nine and overestimated the changes in three of the 12 hemispheres examined (Table 1, Figs. 1C and 2C). The CISS images showed the steno-occlusive changes correctly in two, underestimated in nine, and overestimated the changes in one of the 12 hemispheres (Table 2, Figs. 1D, 1E, and 2D-F). The CISS images disclosed angiographically occluded internal carotid arteries in six hemispheres as distinct structures in the basal cisterns, leading to underestimation of occlusion of the internal carotid arteries.

MR angiography detected moyamoya vessels in the basal cisterns correctly in five and underestimated in seven of the 12 hemispheres (Table 3, Figs. 1C and 2C). The CISS images displayed the moyamoya vessels correctly in 10, underestimated the vessels in one, and overestimated the vessels in one of the 12 hemispheres (Table 4, Figs. 1D, 1E, and 2D-F).

Discussion

MR imaging may show moyamoya disease as any of the following: a) infarction, hemorrhage, brain atrophy, and ventricular dilation; b) absence of flow void in the distal internal carotid artery and/or in the proximal portion of the anterior and/or middle cerebral arteries; and c) a fine flow void in the dilated perforating arteries in the base of the brain (moyamoya vessels).^{1,5-7,9,14} In addition to these classic features, leptomeningeal contrast enhancement in the cerebral cortices may be observed.^{10,12}



←

Fig. 1 A 30-year-old man who underwent bilateral bypass surgery approximately 20 years ago after suffering repeated transient ischemic attacks. **A:** Right internal carotid angiogram (frontal view) demonstrating mild stenosis of the distal internal carotid artery and occlusion of the middle cerebral artery. Moyamoya vessels are moderately apparent. **B:** Left internal carotid angiogram (frontal view) showing occlusion of the distal internal carotid artery and moderate moyamoya vessels. **C:** Magnetic resonance (MR) angiogram revealing severe stenosis of the right internal carotid artery and occlusion of the left internal carotid artery. Moyamoya vessels are minimally apparent bilaterally. **D, E:** Constructive interference in steady state MR images demonstrating moderate stenosis (decrease of the vessel caliber) of the right internal carotid artery and severe stenosis of the left internal carotid artery (arrow). Moyamoya vessels are moderately apparent bilaterally (arrowheads).

derestimation of moyamoya vessels particularly occurs in adults because moyamoya vessels in adults are less apparent than in children.^{9,18)}

MR angiography correctly detects the steno-occlusive changes in the internal carotid arteries in 73–87.5% and overestimates these changes in 12.5–27% of hemispheres.^{9,13,16–18)} In addition, MR angiography demonstrates moyamoya vessels correctly in 65–76.9%, underestimates these vessels in 8.3–31%, and fails to detect them in 15.1–16.7% of hemispheres.^{13,16–18)} MR angiography showed moyamoya vessels correctly in 92.3% of pediatric cases. However, moyamoya vessels were only shown correctly in 69.0%, and were not detected in 31.0% of hemispheres in adult cases.⁹⁾

CISS imaging is useful in the diagnosis of cerebellopontine angle pathologies because of the high spatial resolution, high contrast between soft tissue and cerebrospinal fluid, and fewer motion artifacts resulting from cerebrospinal fluid motion.^{2,3,8)} CISS images underestimated the steno-occlusive changes defined as decreased caliber in the distal internal carotid arteries in 75% of the hemispheres in our series. This high rate of underestimation by CISS imaging may be due to the method of visualization of the vascular structure. CISS imaging detected moyamoya vessels more correctly than MR angiography (83.3% vs. 41.7%) in our series due to its high sensitivity to the small vessels in the cisterns. Although CISS imaging does not demonstrate the actual blood flow in the moyamoya vessels for CISS imag-

MR angiography can demonstrate the steno-occlusive changes in the distal internal carotid arteries and/or proximal anterior and/or middle cerebral arteries, and moyamoya vessels at the base of the brain.^{9,13,16,18)} However, MR angiography may overestimate the steno-occlusive changes in the distal internal carotid arteries due to phase dispersion in the turbulent flow, and underestimate moyamoya vessels due to low spatial resolution.^{9,13,16–18)} Un-

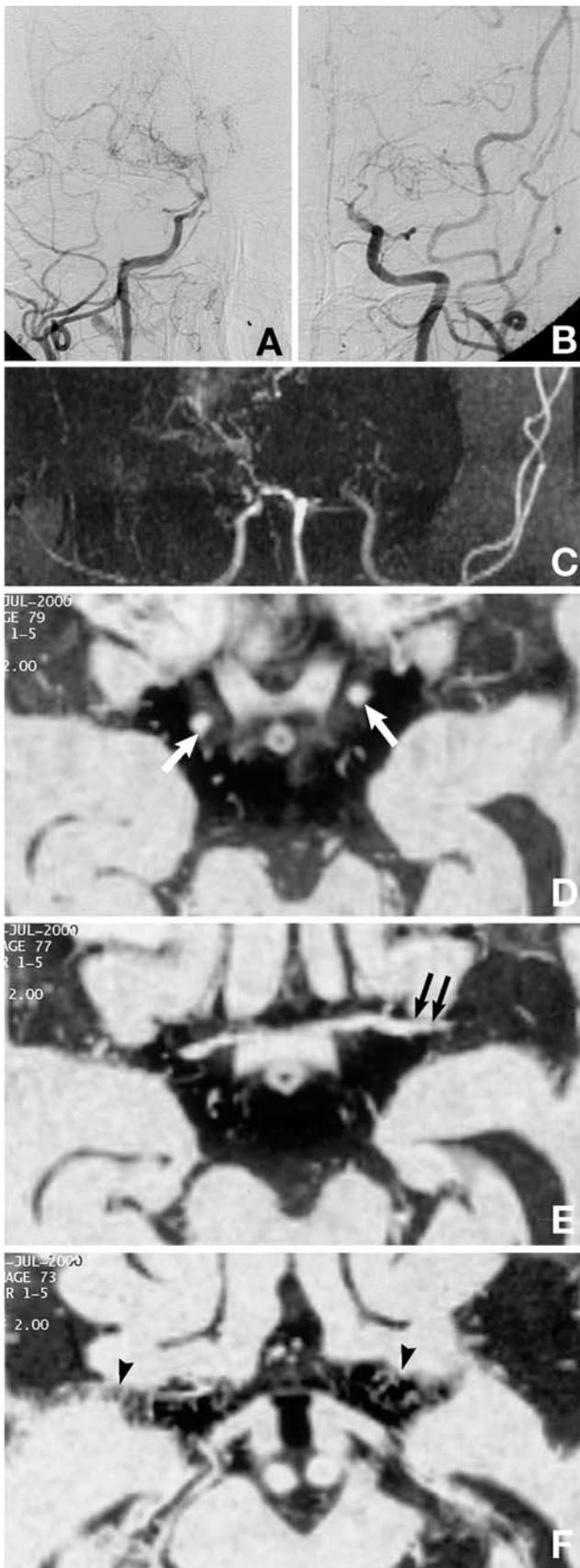


Fig. 2 A 67-year-old woman who developed cerebral infarction in the left frontal lobe 8 years ago. She did not undergo bypass surgery. At the most recent admission, she had developed hemorrhagic infarction in the right basal ganglia. **A, B:** Right (A) and left (B) internal carotid angiograms (frontal views) revealing occlusion of bilateral internal carotid arteries. Moyamoya vessels are moderately apparent bilaterally. **C:** Magnetic resonance (MR) angiogram demonstrating occlusion of the bilateral internal carotid arteries. Moyamoya vessels are moderately apparent on the right side, but only minimally apparent on the left. **D–F:** Constructive interference in steady state MR images demonstrating moderate stenoses (decrease of the vessel caliber) of the bilateral internal carotid arteries (arrows) and anterior and middle cerebral arteries (double arrow). Moyamoya vessels are moderately apparent bilaterally (arrowheads).

ing to detect distinct structures. If the blood flow in moyamoya vessels is reduced, CISS imaging may fail to depict them because the moyamoya vessels are compromised, as seen by digital subtraction angiography.

The noninvasive diagnosis of moyamoya disease is impossible if MR angiography fails to demonstrate moyamoya vessels, which occurs in approximately 15% of patients. Since most errors in diagnosis using MR angiography result from the underestimation of moyamoya vessels,¹⁷⁾ we believe that CISS imaging may become important in the delineation of moyamoya vessels. CISS imaging can detect moyamoya vessels in the cisterns, but not intraparenchymal moyamoya vessels. Intraparenchymal moyamoya vessels are usually observed in patients with well-developed moyamoya vessels, especially in pediatric moyamoya disease patients. Thus, CISS imaging is most useful for detecting moderately developed moyamoya vessels in the cisterns, especially in adults.

High-field (3.0 Tesla) T₂-weighted MR imaging can visualize the detailed anatomy of the brain, especially of the extraaxial structures, due to its high signal-to-noise ratio and heavy T₂ contrast. MR imaging clearly reveals moyamoya vessels in the basal cistern.⁴⁾ However, high-field MR imaging technology is considered investigative equipment and is not cost-effective at present. The spatial resolution of the CISS images was less than 0.9 mm in any orthogonal plane in the present series, which is one of the highest spatial resolutions obtained using com-

Table 2 Correlation between digital subtraction angiographical (DSA) and constructive interference in steady state (CISS) imaging evaluations of the terminal portions of the internal carotid arteries (ICAs)

ICA on CISS images	ICA on DSA			
	No stenosis	Mild stenosis	Marked stenosis	Occlusion
Normal caliber	0	0	0	0
Mild decrease of vessel caliber	0	2	3	0
Marked decrease of vessel caliber	0	1	0	6
Non-visualization of the vessel	0	0	0	0

Table 3 Correlation between digital subtraction angiographical (DSA) and magnetic resonance (MR) angiographical evaluations of moyamoya vessels

Moyamoya vessels on MR angiography	Moyamoya vessels on DSA			
	Not visualized	Minimally visualized	Moderately visualized	Markedly visualized
Not visualized	0	0	0	0
Minimally visualized	0	1	5	1
Moderately visualized	0	0	4	1
Markedly visualized	0	0	0	0

Table 4 Correlation between digital subtraction angiographical (DSA) and constructive interference in steady state (CISS) imaging evaluations of moyamoya vessels

Moyamoya vessels on CISS images	Moyamoya vessels on DSA			
	Not visualized	Minimally visualized	Moderately visualized	Markedly visualized
Not visualized	0	0	0	0
Minimally visualized	0	1	0	0
Moderately visualized	0	0	8	1
Markedly visualized	0	0	1	1

mercially available MR systems at 1.0 or 1.5 Tesla.

The present study indicates that CISS imaging may have a supplementary role to MR angiography in the diagnosis of moyamoya disease, especially for the assessment of moyamoya vessels in the basal cistern.

References

- 1) Brown WD, Graves VB, Chun RWM, Turski PA: Moyamoya disease: MR findings. *J Comput Assist Tomogr* 13: 720-721, 1989
- 2) Casselman JW, Kuhweide R, Ampe W, Meeus L, Steyaert L: Pathology of membranous labyrinth: comparison of T1- and T2-weighted and gadolinium-enhanced spin-echo and 3DFT-CISS imaging. *AJNR Am J Neuroradiol* 14: 59-69, 1993
- 3) Casselman JW, Kuhweide R, Deimling M, Ampe W, Dehaene I, Meeus L: Constructive interference in steady state-3DFT MR imaging of the inner ear and cerebellopontine angle. *AJNR Am J Neuroradiol* 14: 47-57, 1993
- 4) Fujii Y, Nakayama N, Nakada T: High-resolution T2-reversed magnetic resonance imaging on a high magnetic field system. *J Neurosurg* 89: 492-495, 1998
- 5) Fujisawa I, Asato R, Nishimura K, Togashi K, Itoh K, Noma S, Sagoh T, Minami S, Nakao Y, Yonekawa Y, Torizuka K: Moyamoya disease: MR imaging. *Radiology* 164: 103-105, 1987
- 6) Fujita K, Shirakuni T, Nojima N, Tamaki N, Matsumoto S: [Magnetic resonance imaging in moyamoya disease]. *No Shinkei Geka* 14: 324-330, 1986 (Jpn, with Eng abstract)
- 7) Hasuo K, Yasumori K, Yoshida K, Hirakata R, Kuroiwa T, Mizushima A, Matsushima T, Fukui M, Masuda K: Magnetic resonance imaging compared with computed tomography and angiography in moyamoya disease. *Acta Radiol* 31: 191-195, 1990
- 8) Held P, Fellner C, Fellner F, Seitz J, Graf S, Hilbert M, Strutz J: MRI of inner ear and facial nerve pathol-

- ogy using 3D MP-RAGE and 3D CISS sequences. *Br J Radiol* 70: 558–566, 1997
- 9) Houkin K, Aoki T, Takahashi A, Abe H: Diagnosis of moyamoya disease with magnetic resonance angiography. *Stroke* 25: 2159–2164, 1994
 - 10) Komiyama M, Nakajima H, Nishikawa M, Yasui T, Kitano S, Sakamoto H: Leptomeningeal contrast enhancement in moyamoya: its potential role in postoperative assessment of circulation through the bypass. *Neuroradiology* 43: 17–23, 2001
 - 11) Kudo T: Spontaneous occlusion of the circle of Willis: a disease apparently confined to Japanese. *Neurology* 18: 485–496, 1968
 - 12) Ohta T, Tanaka H, Kuroiwa T: Diffuse leptomeningeal enhancement, “ivy sign,” in magnetic resonance images of moyamoya disease in childhood: Case report. *Neurosurgery* 37: 1009–1012, 1995
 - 13) Saeki N, Silva MN, Kubota M, Takanashi J, Sugita K, Nakazaki S, Yamaura A: Comparative performance of magnetic resonance angiography and conventional angiography in moyamoya disease. *J Clin Neurosci* 7: 112–115, 2000
 - 14) Suto Y, Caner BE, Nakatsugawa S, Katsube Y, Ishii Y, Torizuka K: Evaluation of MRI in moyamoya disease. *Radiat Med* 8: 92–95, 1990
 - 15) Suzuki J, Takaku A: Cerebrovascular ‘moyamoya’ disease: disease showing abnormal net-like vessels in base of brain. *Arch Neurol* 20: 288–299, 1969
 - 16) Yamada I, Matsushima Y, Suzuki S: Moyamoya disease: diagnosis with three-dimensional time of flight MR angiography. *Radiology* 184: 773–778, 1992
 - 17) Yamada I, Suzuki S, Matsushima Y: Moyamoya disease: comparison of assessment with MR angiography and MR imaging versus conventional angiography. *Radiology* 196: 211–218, 1995
 - 18) Yoon HK, Shin HJ, Lee M, Byun HS, Na DG, Han BK: MR angiography of moyamoya disease before and after encephaloduroarteriosynangiosis. *AJR Am J Roentgenol* 174: 195–200, 2000

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Commentary on this paper appears on the next page.

Commentary

The authors showed that moyamoya vessels are nicely visualized by CISS in patients with moyamoya disease. Although moyamoya vessels can be detected more frequently by CISS than by MRA, the detection is limited within the basal cisterns and the whole image of the moyamoya vessels cannot be obtained by CISS. They also showed that MRA is more sensitive for the detection of steno-occlusive changes than CISS. Moyamoya disease is identified by the findings of both steno-occlusive changes of the terminal portion of the internal carotid artery and development of moyamoya vessels. Thus, at present, we do not think that this diagnostic imaging method can supplement the conventional diagnostic procedures for moyamoya disease, including MRA and MRI of the brain. We hope that the authors will investigate this interesting imaging technique for clinically more valuable applications.

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The authors evaluated the value of CISS imaging for the diagnosis of moyamoya disease in 12 hemispheres of 6 patients. They compared CISS imaging with 3D-TOF MRA for the visualization of the steno-occlusive changes in the distal carotid artery and the development of moyamoya vessels in the basal cistern. The results indicated that MRA is superior to CISS imaging for the demonstration of the steno-occlusive

changes, but CISS imaging may have a supplementary role to MRA in the diagnosis of moyamoya disease, especially for the assessment of moyamoya vessels. These two methods are completely different for the visualization of the vessels, because 3D-TOF MRA demonstrates the flowing blood itself within the vessels by the inflow effect, whereas CISS imaging demonstrates the shape of the outside wall of the vessels. So I agree with the conclusion of this paper, but it seems to be a natural result.

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In this article, authors stressed the usefulness of CISS (constructive interference in steady state) imaging for the diagnosis of moyamoya disease without invasion. MRA often overestimates intracranial steno-occlusive lesions because the MRA image is constructed from flow information. In contrast, CISS image is essentially morphological, resulting in higher spatial resolution. However, the CISS image shows occluded vessels if the vessels still have some outer caliber. We can evaluate intracranial steno-occlusive lesions in detail by the combination of usual MRA and CISS images. Wall thickness can also be estimated. These methods may be valuable, especially for the follow-up study of moyamoya disease.

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