Journal of Korean Medical Science
Vol. 5, No. 2 85-90 June 1990

MR Imaging Findings of Moyamoya Disease

Kee Hyun Chang, M.D., Jeong Geun Yi, M.D., Moon Hee Han, M.D.,
In One Kim, M.D.

*Department of Diagnostic Radiology, Seoul National University College of Medicine
Seoul, Korea*

The brain MR images of 23 patients with angiographically proved moyamoya disease were reviewed to evaluate the capability of MR to demonstrate vascular and parenchymal abnormalities. All the MR images were obtained on a 2.0 T superconducting system and included T1-weighted sagittal and T2-weighted axial images without implementation of flow compensation (FC). The vascular abnormalities demonstrated on MR images were narrowing of the cavernous internal carotid artery (ICA) (73%), narrowing or occlusion of the supraclinoid ICA (87%) and proximal middle cerebral artery (MCA) (91%), and multiple collateral vessels in the basal ganglia and/or thalamus (96%). The parenchymal abnormalities included ischemic infarctions (74%), predominantly located in watershed areas, hemorrhagic infarctions (26%), intracerebral hematomas (13%), and intraventricular hemorrhage (13%). In conclusion, MR imaging was a useful diagnostic modality for detecting both vascular and parenchymal abnormalities associated with moyamoya disease. This may obviate the need for invasive angiography as far as the diagnosis is wanted at the non-quantitative level.

Key Words: *Moyamoya disease, Brain, MRI, carotid artery stenosis*

INTRODUCTION

The imaging diagnosis of moyamoya disease has been greatly facilitated by the advent of magnetic resonance (MR) imaging, because of its high contrast-making and demonstrability of arterial blood flow (Fujisawa et al., 1987; Bruno et al., 1988; Brown et al., 1989; Brooks et al., 1987; Katz et al., 1989; Wilms et al., 1989). MR diagnosis of moyamoya disease relies on the observation of the signal void collateral vessels as well as of narrowing or occlusion of intracranial internal carotid artery (ICA). However, the demonstrability of vascular abnormalities are variable on the routine MR images, depending upon the imaging system and techniques used (Fujisawa et al., 1987; Bruno et al., 1988). In this study, we analyzed the MR imaging findings of moyamoya disease to determine how often the vascular and parenchymal abnormalities could be detected.

Corresponding Author: *Kee Hyun Chang, M.D., Department of Diagnostic Radiology. Seoul National University Hospital 28 Yongon-dong, Chongno-gu, Seoul 110-744, Korea Tel: (02) 7601-2516*

This study was supported in part by the grant of clinical research fund of Seoul National University Hospital (1990).

PATIENTS AND METHODS

Twenty-three patients with angiographically proved moyamoya disease were studied with MR imaging. Twelve patients were men and 11 were women with the age ranging from 6 to 43 (mean=16.3 years). MR imaging was performed on a 2.0 T superconducting unit (Spectro-20000, Goldstar, Seoul, Korea) with multislice spin echo (SE) sequences. T1-weighted images

(SE 500-600/30/2: TR/TE/Excitation No) were obtained in sagittal plane, covering the middle two thirds of the brain from the right sylvian fissure through the left one. The proton density-and T2-weighted images without using flow compensation (FC) (SE 2500/30, 80/1) were obtained in axial plane. The slice-thickness was 5 mm with the interslice gap being 2 mm. The number of acquisition matrices varied from 180x256 to 256x256.

The MR images were assessed with respect to parenchymal and vascular abnormalities including narrowing of the cavernous and supraclinoid ICA and the proximal horizontal portion (M-1) of the middle cerebral artery (MCA), and dilated collateral vessels at various locations. The cavernous and supraclinoid ICA was designated to be "narrowed", when the diameter was smaller than or equal to that of the basilar artery on the axial images. MCA was described to be "narrowed" or "occluded", when it was not identified on the axial image as a smooth, curvilinear, signalvoid area that corresponded to the course of the M-1 segment of the artery. Absence or presence of the collateral vessels was determined on the sagittal and axial images by subjective observation of two neuroradiologists.

RESULTS

1) Vascular abnormalities

The vascular abnormalities observed are as summarized in Table 1. On cerebral angiography, 22 patients had severe stenosis or occlusion of the supraclinoid ICA bilaterally. In one patient the ICA involvement was unilateral.

On the axial MR images, narrowing of the cavernous ICA was demonstrated in 33 out of 45 involved ICA of 23 patients (73%) (Fig. 1). The supraclinoid portion of ICA appeared narrowed or occluded in the 39 arteries (87%). The M-1 portion of MCA appeared narrowed or occluded in 41 arteries (91%) (Fig. 2-a) and equivocal in the remaining four among 45 involved MCA. Collateral vessels shown as multiple, small, round or tortuous signal void areas were most commonly noted in the basal ganglia and/or thalami (96%). They were also seen around the circle of Willis, inferior frontal lobes, around or within the lateral ventricles, around ambient or quadrigeminal cisterns, and in areas of high convexity of brain on the axial images (Figs. 2-b and 2-c). such findings were well demonstrated on both the first echo (proton-density weighted) images and the second echo (T2-weighted) images, but the demonstration was more conspicuous in the former, because of greater contrast between the signal void vessels and the brain parenchyma.

On T1-weighted sagittal images, narrowing or occlusion of the ICA and MCA was identified in none of the patients, and collateral vessels were less fre-

Table 1. Vascular Abnormalities Demonstrated on MR Images

MR Findings	Case No. (%)
Narrowing or occlusion (n = 45 arteries)	
Cavernous ICA	33 (73)
Supraclinoid ICA	39 (87)
MCA (M-1 segment)	41 (91)
Collateral vessels (n = 23 patients)	
Basal ganglia/thalamus	22 (96)
Suprasellar/sylvian cistern	19 (83)
Inferior frontal lobe	19 (83)
Intra-or periventricular area	17 (74)
Perimesencephalic cistern	13 (57)
High convexity cortical area	12 (52)

ICA = internal carotid artery, MCA = middle cerebral artery
M-1 = proximal horizontal portion of middle cerebral artery

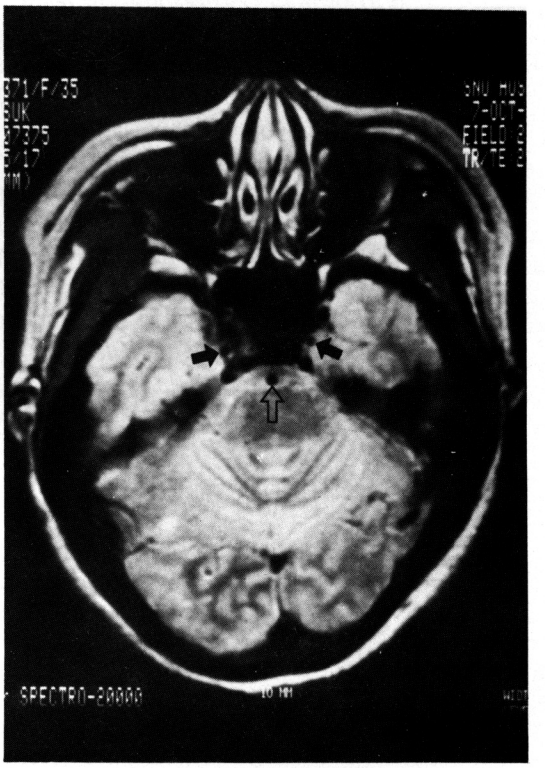

Fig. 1. Axial MR image (SE 2500/30/1) of narrowing of cavernous internal carotid arteries. The cavernous portions of both internal carotid arteries (arrows) appear slightly smaller than the basilar artery (open arrow), suggesting narrowing of the artery.

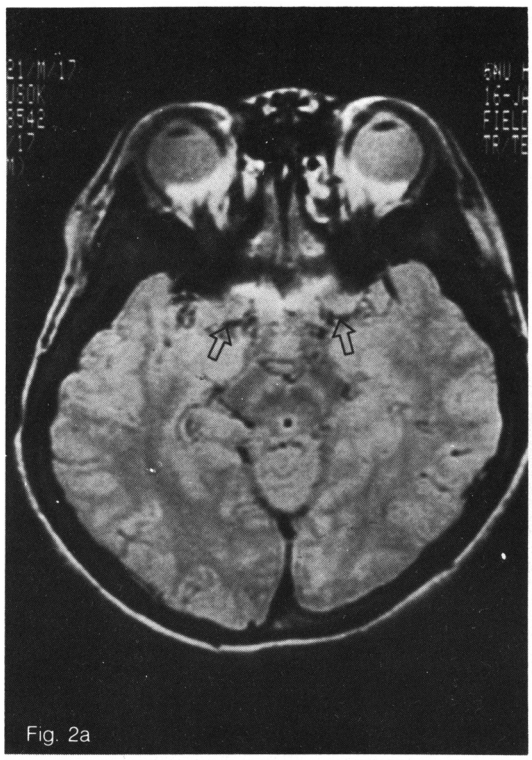

Fig. 2a

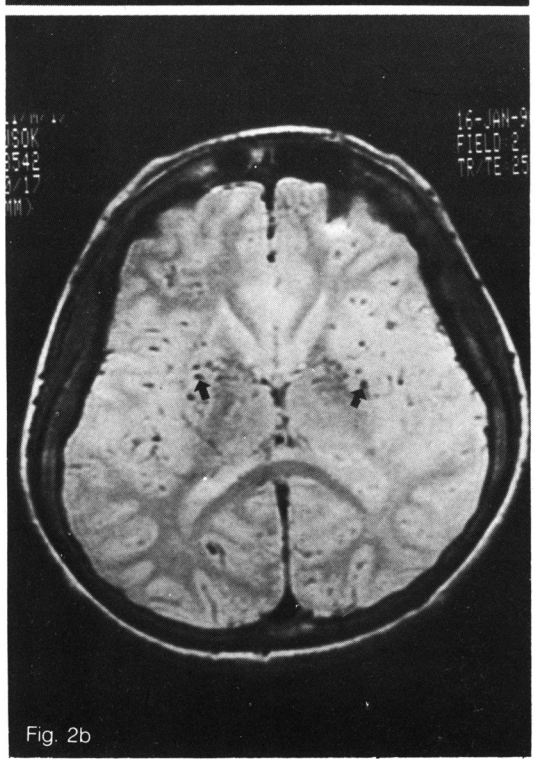

Fig. 2b

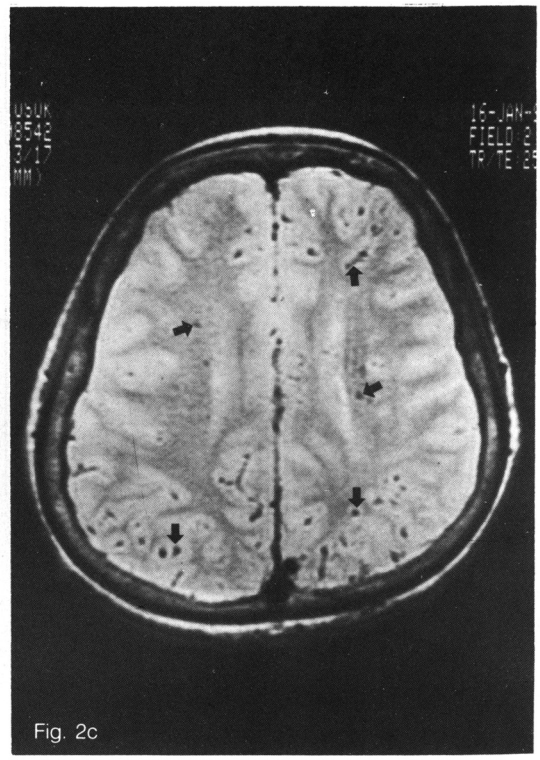

Fig. 2c

Fig. 2. Axial MR images (SE 2500/30/1) of a 17-year-old male showing abnormalities of horizontal portions of both middle cerebral arteries and collateral vessels. Only short segments of the horizontal portions (open arrows) of both middle cerebral arteries are visualized (a). Multiple signal void collateral vessels (arrows) are well demonstrated in the basal ganglia (b), parietal cortical areas and periventricular white matter bilaterally and in the frontal cortex on the left (c).

quently demonstrated than on the axial images. The distributions of collateral vessels were in the basal ganglia (Fig. 3), around the lateral ventricle, and around ambient or quadrigeminal cisterns in 19 patients (83%), 14 patients (61%), and 10 patients (43%), respectively. Collateral vessels in the other areas were difficult to evaluate on the T1-weighted sagittal images.

2) Parenchymal abnormalities

The parenchymal abnormalities observed are as summarized in Table 2. Ischemic infarctions were found in 17 out of 23 patients (74%), and multiple in 16 patients, and varied from a few milimeter to seven centimeter in size. The lesions were observed predominantly in such hemodynamically vulnerable areas as the periventricular white matter and centrum semi-

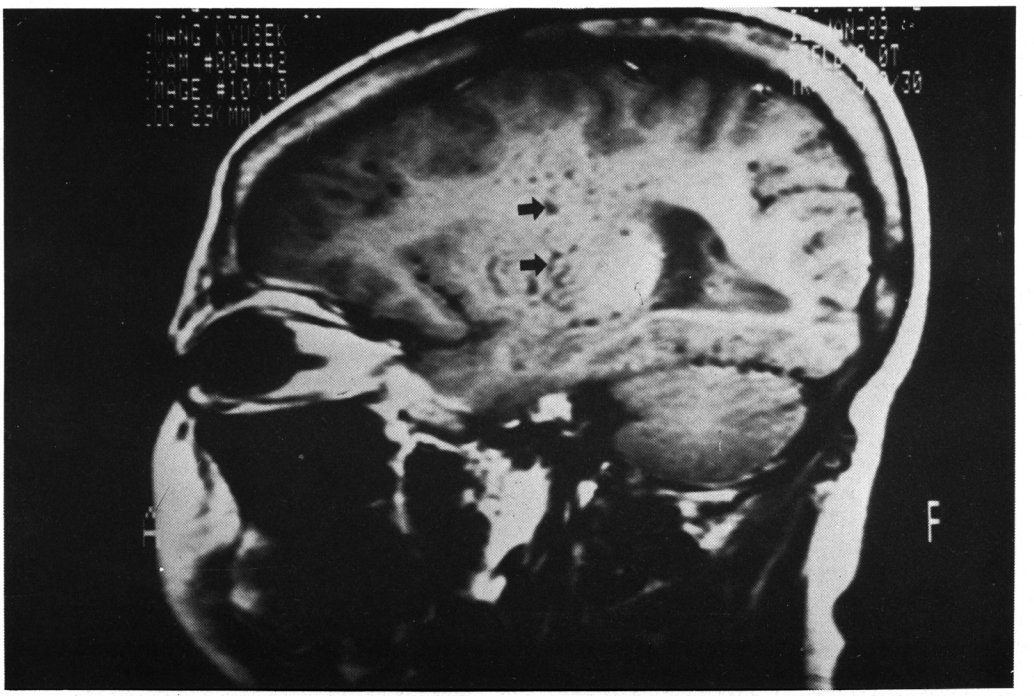

Fig. 3. T1-weighted sagittal MR image (SE 500/30) of collateral vessels. Multiple, small, round or tortuous signal void collateral vessels (arrows) are well seen in the basal ganglia and periventricular white matter.

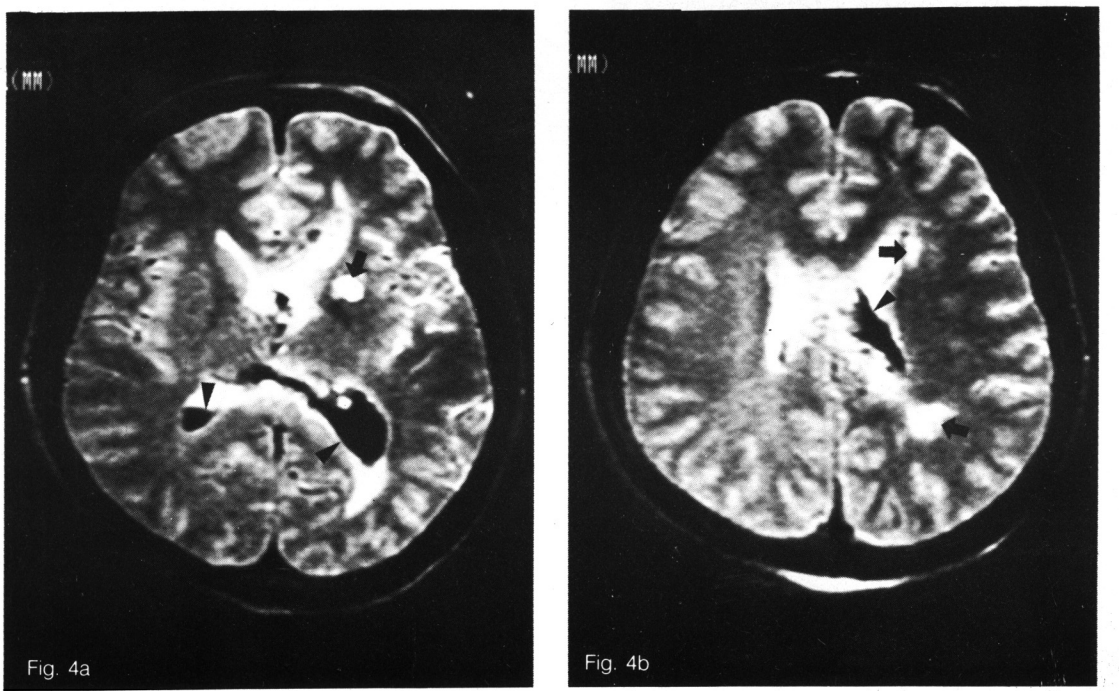

Fig. 4a

Fig. 4b

Fig. 4. Axial MR images (SE 2500/80/1) of intraventricular hemorrhage and ischemic infarcts in watershed areas. There are multiple areas of high intensity in periventricular white matter on left side, indicating ischemic infarcts (arrows), and low signal intensity within the lateral ventricle representing acute hematoma (arrowheads).

ovale, basal ganglia, and borderzone areas between the anterior, middle, and posterior cerebral arteries. The ischemic infarcts corresponding to the major vascular territories were also seen. Hemorrhagic infarctions were noted in six patients (26%), three in the basal ganglia and the other three in hemispheric cortices. Intracerebral hematomas (ICH) were seen in three patients and located in the thalamus and the parietal cortex. Intraventricular hemorrhage (IVH) was found in the lateral ventricle in three patients: two solely in IVH and the other with ICH in the thalamus.

Table 2. Parenchymal Abnormalities Demonstrated on Conventional MR Images

	n = 23
MR findings	Case No. (%)
Ischemic infarction	17 (74)
Multiple	16 (70)
PVWM/centrum ovale	12 (52)
Basal ganglia	9 (39)
Borderzone between ACA, MCA, and PCA	6 (26)
Major vessel territory	7 (30)
Hemorrhagic infarction	6 (26)
ICH	3 (13)
IVH	3 (13)
Normal	2 (9)

PVWM = periventricular white matter
ACA = anterior cerebral artery
MCA = middle cerebral artery
PCA = posterior cerebral artery
ICH = intracerebral hematoma
IVH = intraventricular hemorrhage

DISCUSSION

Moyamoya disease is a rare cerebrovascular disease of unknown etiology that consists of progressive narrowing or occlusion of the supraclinoid portion of ICA and proximal portions of ACA and MCA in association with extensive parenchymal, transdural, and leptomeningeal collateral vessels, most often reported from Japan (Nishimoto et al., 1968; Suzuki et al., 1969). CT scan usually demonstrates only non-specific findings including ischemic infarctions, hemorrhage, or brain atrophy (Handa et al., 1977), although high resolution and dynamic CT scan can depict the specific findings of moyamoya disease (Takahashi et al., 1980; Asari et al., 1982). Because of its ability to demonstrate arterial blood flow, MR imaging is capable of detecting abnormal vascularities including narrowing of ICA and MCA and collateral vessels that are suggestive of moyamoya disease (Fujisawa et al., 1987; Bruno et al., 1988).

With our routine MR imaging technique, the the cavernous and supraclinoid portions of ICA can easily be seen as signal void that is consistently larger in size than the basilar artery on the axial images in the subjects without known intracranial vasculopathy. Likewise, the whole segment or at least more than one half of M-1 segment of MCA is also identified as a smooth, linear or curvilinear signal void in the areas corresponding to MCA in all "normal" patients. Therefore, findings of the intracranial ICA smaller in size than the basilar artery and nonvisulalization of normal M-1 of MCA are considered to be highly suggestive of moyamoya disease, even though its accuracy depends upon the imaging system and technique used, particularly upon slice thickness/gap. These vascular findings are, however, not demonstrated in every patient with moyamoya disease (Fujisawa et al., 1987; Bruno et al., 1988), as in the present series. The demonstration of collateral vessels appears more important and conclusive in the diagnosis of moyamoya disease. Currently, the FC soft ware has been almost routinely implemented in the cranial MR imaging for reducing flow artifacts. Nevertheless, the imaging with FC has some drawbacks, including suppression of signal void from slowly flowing blood such as cortical veins or from CSF passing through the aqueduct (Haacke et al., 1987; Elster, 1988; Quencer et al., 1988; Atlas et al., 9188). On the angiography of moyamoya disease, many of the collateral vessels proved to have slow flow that persists up to the venous phase. Such collateral vessels appear either isointense or hyperintense on the images with FC due to signal refocussing.

Concerning the imaging plane and pulse sequence in the evaluation of moyamoya disease, the axial T2-weighted imaging without using FC is considered better than the imaging using FC because of its better demonstration of the vascular abnormalities. But the former frequently produces flow artifacts which may obscure the parenchymal abnormalities. In the present series, the flow artifacts of variable degree were seen in almost all patients, but they did not actually prevent from the correct diagnosis. T2-weighted imaging using FC produces little flow artifacts, but it may show much less conspicuous collateral vessels. T1-weighted sagittal images can also demonstrate the collateral vessels, but they have limitation in covering the whole brain and in demonstrating ICA and MCA abnormalities as in the present series. Axial or coronal T1-weighted images may well visualize the ICA and MCA abnormalities and the collateral vessels with thin slice thickness, but again the whole brain can not be included in single acquisition imaging. T1-weighted im-

ages (axial, sagittal or coronal images) are necessary for the evaluation of possible hemorrhage in the subacute stage.

The MR findings of parenchymal abnormalities in the moyamoya disease have been described in the literature (Fujisawa et al., 1987; Bruno et al., 1988; Brown et al., 1989; Brooks et al., 1987; Wilms et al., 1989). In keeping with the location of vascular involvement, parenchymal lesions were found in the region of the carotid artery distribution bilaterally. The majority of the ischemic infarctions have been reported to be in the watershed regions (Fujisawa et al., 1987; Bruno et al., 1988), and this is the case in our series. As compared with CT in respect to depicting parenchymal abnormalities, MR appeared nearly identical or slightly superior to CT.

In conclusion, the finding on MR images of cavernous and supraclinoid ICA that was smaller than the basilar artery, nonvisualization of the M-1 segment of MCA, and multiple signal void collateral vessels in the basal ganglia and other areas, with or without multiple ischemia/infarcts in the watershed areas and/or hemorrhages were considered to be pathognomonic of moyamoya disease. We believe that MR imaging is the diagnostic modality of choice for detecting both vascular and parenchymal abnormalities associated with moyamoya disease. However, cerebral angiography can be called in for planning of surgical treatment.

REFERENCES

Asari S, Satoh T, Sakurai M, Yamamoto Y, Sadamoto K: *The advantage of coronal scanning in cerebral computed angiotomography for diagnosis of moyamoya disease. Radiology 1982;145:709-11.*

Atlas SW, Mark AS, Fram EK. Aqueduct stenosis: *Gradient-echo rapid MR imaging. Radiology 1988;169:449-53.*

Brooks BS, Gammal TE, Adams RJ, Hartlage PL, Smith WB: *MR imaging of moyamoya disease in neurofibromatosis. AJNR 1987;8:178-9.*

Brown WD, Graves VB, Chun RWM, Turski PA: *Moyamoya disease: MR findings. J Comput Assist Tomogr 1989;13(4):720-3.*

Bruno A, Yuh WTC, Biller J, Adams HP, Cornell SH: *Magnetic resonance imaging in young adults with cerebral infarction due to moyamoya. Arch Neurol 1988;45:303-6.*

Elster AD: *Motion artifact suppression technique (MAST) for cranial MR imaging: Superiority over cardiac gating for reducing phase-shift artifacts. AJNR 1988;9:671-4.*

Fujisawa I, Asato R, Nishimura K, et al: *Moyamoya disease: MR imaging. Radiology 1987;164:103-5.*

Haacke EM, Lenz GW: *Improving MR image quality in the presence of motion by using rephasing gradients. AJR 1987;148:1251-8.*

Handa J, Handa H, Nakano Y, et al: *Computed tomography in moyamoya: Analysis of 16 cases. Comput Axial Tomogr 1977;1:165-74.*

Katz BH, Quencer RM, Kaplan JO, Hinks RS, Post MJD: *MR imaging of intracranial carotid occlusion. AJNR 1989;10:345-50.*

Nishimoto A, Takeuchi S: *Abnormal cerebrovascular network related to the internal carotid arteries. J Neurosurg 1968;29:255-60.*

Quencer RM, Hinks RS, Pattany PH, Horen M, Post MJD: *Improved MR imaging of the brain by using compensating gradients to suppress motion-induced artifacts. AJR 1988;151:163-70.*

Suzuki J, Takaku A: *Cerebrovascular "moyamoya" disease. Disease showing abnormal net-like vessels in base of brain. Arch Neurol 1969;20:288-99.*

Takahashi M, Miyauchi T, Kowada M: *Computed tomography of moyamoya disease: Demonstration of occluded arteries and collateral vessels as important diagnostic signs. Radiology 1980;134:671-6.*

Wilms G, Marchal G, Van Fraeyenhoven L, et al: *Unilateral moyamoya disease: MRI findings. Neuroradiology 1989;31:442.*