High-Resolution Turbo Magnetic Resonance Angiography for Diagnosis of Moyamoya Disease

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Background and Purpose—High-resolution turbo MR angiography with zero-filling interpolation (ZFI) technique is a new vascular imaging method with reduced scan time. The purpose of the present study was to evaluate high-resolution turbo MR angiography for the diagnosis and assessment of moyamoya disease.

Methods—Forty-six patients suspected of having moyamoya disease were examined with high-resolution turbo MR angiography with the ZFI technique, MRI, and conventional angiography. Moyamoya disease was diagnosed in 42 of these patients. Blind, separate interpretation of the images was performed.

Results—High-resolution turbo MR angiography and MRI accurately evaluated 349 (95%) and 325 (88%) of 368 arteries, respectively, but the degree of stenosis was overestimated in the other arteries. MR angiography and MRI depicted basal cerebral moyamoya vessels in 82 (98%) and 82 (98%) of 84 hemispheres, respectively. MR angiography also depicted leptomeningeal and transdural collateral vessels in 51 (100%) of 51 hemispheres and in 38 (88%) of 43 hemispheres, respectively. The sensitivity and specificity of high-resolution turbo MR angiography for the diagnosis of moyamoya disease were 98% and 100%, respectively.

Conclusions—High-resolution turbo MR angiography in reduced scan time is highly accurate in the assessment of both steno-occlusive lesions and collateral vessels in moyamoya disease, thus providing a highly accurate (98%) diagnosis and assessment of moyamoya disease.

Key Words: angiography ■ angiography, magnetic resonance ■ magnetic resonance imaging ■ moyamoya disease

Moyamoya disease is a rare cerebrovascular occlusive disease of unknown cause.1,2 Although this disease occurs predominantly in the Japanese, its occurrence has also been reported in other countries.3,4 The angiographic features of moyamoya disease include (1) bilateral stenosis or occlusion of the supraclinoid portion of the internal carotid artery (ICA) that extends to the proximal portions of the anterior cerebral artery (ACA) and the middle cerebral artery (MCA) and (2) the presence of parenchymal collateral vessels, or moyamoya vessels (MMVs), from the suprasellar cistern to the cerebral base.5,6

Recently, turbo MR angiography with zero-filling interpolation (ZFI) technique has been proposed as a new technique to reduce scan time for the evaluation of cerebrovascular diseases.7,8 The ZFI along the slice direction generates thinner slice sections, and the ZFI along the in-plane phase-encoding direction increases the matrix size of the direction.7–10 Thus, the ZFI technique allows for high-resolution turbo MR angiography in less scan time. To our knowledge, however, no study has described the application of high-resolution turbo MR angiography with the ZFI technique to patients with moyamoya disease, and no attempt has been made to compare the sensitivity and specificity of high-resolution turbo MR angiography with those of conventional angiography. We present the results of a prospective study that was undertaken to determine the clinical efficacy of high-resolution turbo MR angiography, compared with that of conventional angiography, in the diagnosis of moyamoya disease.

Subjects and Methods

Patients

During a 3-year period of 1997 to 1999, 46 consecutive patients (26 females and 20 males, age range 3 to 50 years, mean age 21 years) who were clinically suspected of having moyamoya disease were prospectively examined with high-resolution turbo MR angiography, MRI, and conventional angiography at our department of radiology. The suspicion was based on clinical symptoms suggestive of moyamoya disease, including TIAs that were provoked by hyperventilation. In 42 of these patients, the diagnosis of moyamoya disease was made on the basis of findings at conventional angiography. Twenty-four of the 42 patients were female and 18 were male, with an age range of 3 to 50 years (mean age 21 years). We considered the entire spectrum of moyamoya disease, including cases of mild moyamoya disease. None of the 42 patients had an underlying disease, thereby confirming the diagnosis of idiopathic moyamoya disease.

On the basis of findings at conventional angiography in the remaining 4 patients, 2 patients had unilateral steno-occlusive lesions...
TABLE 1. Comparison of Evaluation of 368 Arteries With Turbo MR Angiography, MRI, and Conventional Angiography

<table>
<thead>
<tr>
<th>MR Study</th>
<th>Normal (n=96)</th>
<th>Stenosis (n=107)</th>
<th>Occlusion (n=165)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Turbo MR angiography</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Normal</td>
<td>95</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Stenosis</td>
<td>1</td>
<td>89</td>
<td>0</td>
</tr>
<tr>
<td>Occlusion</td>
<td>0</td>
<td>18</td>
<td>165</td>
</tr>
<tr>
<td>MRI</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Normal</td>
<td>85</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Stenosis</td>
<td>11</td>
<td>75</td>
<td>0</td>
</tr>
<tr>
<td>Occlusion</td>
<td>0</td>
<td>32</td>
<td>165</td>
</tr>
</tbody>
</table>

Values are given as number of ICAs, ACAs, MCAs, and PCAs in 46 patients.

Image Analysis

MR accuracy was studied by means of blinded, separate interpretations of MR angiograms, MR images, and conventional angiograms made by 2 independent observers (I.Y., T.N.). The conventional angiograms were used as the gold standard in analysis of the MR findings, and the MR angiograms and MR images were independently interpreted, without knowledge of the angiographic findings. In interpretation of the MR images, we used both T1- and T2-weighted images for image analysis. The observers were unaware of the frequency of disease in the patient group. When the observers did not fully agree on the findings, the diagnosis was achieved through discussion.

In a lesion-by-lesion analysis, MR angiographic and MRI findings were reviewed for signs of steno-occlusive lesions and for the presence of basal cerebral MMVs, leptomeningeal collateral vessels, and transdural collateral vessels. These MR findings were also compared on a lesion-by-lesion basis with the findings of conventional angiography. The standard scoring scheme used for steno-occlusive lesions has 3 grades: normal, stenotic, and occluded. “Stenotic” indicated a focal narrowing of the lumen, whereas “occluded” indicated the absence or discontinuity of the lumen. Four arteries were scored bilaterally: the supraclinoid ICA, the proximal ACA, the proximal MCA, and the proximal PCA. Furthermore, based on their presence and appearance, abnormal basal cerebral MMVs were graded as none, mild, or marked. “Mild” indicated that the MMVs were a small vascular network localized in the basal ganglia and the medullary arteries were not depicted, whereas “marked”: indicated that the MMVs were a large vascular network that extended above the basal ganglia and the medullary arteries were depicted. These criteria for steno-occlusive lesions and MMVs were commonly used to describe findings at MR angiography, MRI, and conventional angiography.

In a patient-by-patient analysis, conventional angiographic criteria were used to determine the diagnosis of moyamoya disease: (1) the presence of bilateral stenosis or occlusion involving the supraclinoid portion of the ICA and the proximal portions of the ACA and MCA and (2) the presence of bilateral abnormal basal cerebral MMVs. MR angiographic and MRI criteria for moyamoya disease were the same as for those of conventional angiography: (1) depiction of bilateral narrowing or absence of lumen in the supraclinoid ICA and in the proximal ACA and MCA and (2) depiction of bilateral abnormal basal cerebral MMVs.

TABLE 2. Comparison of Evaluation of Basal Cerebral MMVs in 92 Cerebral Hemispheres With Turbo MR Angiography, MRI, and Conventional Angiography

<table>
<thead>
<tr>
<th>MR Study</th>
<th>None (n=8)</th>
<th>Mild (n=59)</th>
<th>Marked (n=25)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Turbo MR angiography</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>8</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Mild</td>
<td>0</td>
<td>57</td>
<td>0</td>
</tr>
<tr>
<td>Marked</td>
<td>0</td>
<td>0</td>
<td>25</td>
</tr>
<tr>
<td>MRI</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>8</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Mild</td>
<td>0</td>
<td>57</td>
<td>0</td>
</tr>
<tr>
<td>Marked</td>
<td>0</td>
<td>0</td>
<td>25</td>
</tr>
</tbody>
</table>

Values are given as number of cerebral hemispheres in 46 patients.

All patients also underwent conventional angiography that included bilateral internal and external carotid arteriography and unilateral or bilateral vertebral arteriography, with the use of the transfemoral catheterization technique. Conventional angiography was performed within 1 month before or after MR angiography and MRI.

Imaging Examinations

A 1.5-T superconducting system with a 25-mT/m maximum gradient capability (Magnetom Vision; Siemens) and a circularly polarized head coil was used to obtain all MR images. High-resolution turbo MR angiography was performed with a 3-dimensional fast imaging technique (MTC), tilted optimized nonsaturated excitation (TONE), and multislab techniques were also used. The field of view was 35.0/6.4 milliseconds (repetition time/echo time), flip angle of 20 degrees, and 1 signal acquired. Magnetization transfer contrast imaging examinations were obtained by means of a maximum intensity projection (MIP) algorithm. To interpret MR angiograms, both the MIP and section images were reviewed.

In all patients, axial T1-weighted spin-echo images (600/14) were obtained with 2 signals averaged. The T1-weighted images were acquired with a field of view of 165×220 mm, a matrix of 192×256, and a section thickness of 5 mm with a 1-mm intersection gap. Axial T2-weighted turbo spin-echo images (4000/96, echo train length 7) were obtained with 2 signals averaged. The T2-weighted images were acquired with a field of view of 165×220 mm, a matrix of 210×312, and a section thickness of 5 mm with a 1-mm intersection gap.
Finally, statistical analysis was performed with the $\chi^2$ test or the Fisher exact test between groups. $P<0.05$ was considered statistically significant.

**Results**

The findings at high-resolution turbo MR angiography and MRI in the 46 patients were reviewed for steno-occlusive lesions, basal cerebral MMVs, and other collateral vessels, and the findings were compared with those at conventional angiography (Tables 1 to 3). Thus, the sensitivity and specificity of each method for detection of these lesions were evaluated on a lesion-by-lesion basis (Table 4), and those for the diagnosis of moyamoya disease were evaluated on a patient-by-patient basis (Table 5).

**Steno-Occlusive Lesions**

Of a total of 368 arteries that were studied, high-resolution turbo MR angiography and MRI accurately evaluated 349 (95%) and 325 (88%) arteries, respectively (Table 1 and Figure 1). All 165 occlusions were correctly identified with MR angiography and MRI. However, on the basis of findings at MR angiography and MRI, respectively, the degree of stenosis was overestimated as occluded in 18 and 32 of 107 stenotic arteries and as stenotic in 1 and 11 of 96 normal arteries (Figure 2).

The sensitivity and specificity, respectively, for the detection of steno-occlusive lesions were 100% and 99% for high-resolution turbo MR angiography and 100% and 89% for MRI (Table 4). The specificity of high-resolution turbo MR angiography was significantly higher than that of MRI ($P<0.01$). In determining the sensitivity and specificity for the detection of steno-occlusive lesions, we combined the results for stenosis and occlusion, because this was necessary to determine the efficacy for the detection of stenotic and occlusive disease.

Of the 92 supraclinoid ICAs evaluated with MR angiography and MRI, respectively, 87 (95%) and 83 (90%) were correctly evaluated, and 5 and 9 stenotic ICAs were diagnosed as occluded. Of the 92 ACAs evaluated with MR angiography and MRI, respectively, 86 (93%) and 78 (85%) were correctly evaluated, and 6 and 14 stenotic ACAs were diagnosed as occluded. Of the 92 MCAs evaluated with MR angiography and MRI, respectively, 84 (91%) and 83 (90%) were correctly evaluated, 7 and 8 stenotic MCAs were diagnosed as occluded, and 1 normal MCA was diagnosed as stenotic with each modality. Of the 92 PCAs evaluated with MR angiography and MRI, respectively, 92 (100%) and 81 (88%) were correctly evaluated, 1 stenotic PCA was diagnosed as occluded.

**Table 3. Comparison of Evaluation of Leptomeningeal and Transdural Collateral Vessels in 92 Cerebral Hemispheres With Turbo MR Angiography, MRI, and Conventional Angiography**

<table>
<thead>
<tr>
<th>MR Study</th>
<th>Leptomeningeal Collateral Vessels</th>
<th>Transdural Collateral Vessels</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Absent (n=41)</td>
<td>Present (n=51)</td>
</tr>
<tr>
<td>Turbo MR angiography</td>
<td>41</td>
<td>0</td>
</tr>
<tr>
<td>MRI</td>
<td>0</td>
<td>51</td>
</tr>
</tbody>
</table>

Values are given as number of cerebral hemispheres in 46 patients.

**Figure 1.** Images from a 15-year-old girl with moyamoya disease. A, Axial projected MR angiogram shows that the ICA is bilaterally occluded in the supraclinoid portion and that the ACA and MCA are also occluded in the proximal portions. Mild MMVs (arrows) are seen in bilateral basal cerebral regions. B, Lateral projected MR angiogram shows leptomeningeal collateral vessels (arrow) from the PCA branches to the ACA territory, as well as the ICA occlusion and basal cerebral MMVs. C, Right internal carotid arteriogram in frontal projection shows that the ICA is occluded in the distal supraclinoid portion and that the ACA and MCA are also occluded in the proximal portions. Mild MMVs are seen in the basal cerebral region. D, Left internal carotid arteriogram in frontal projection shows that the ICA is occluded in the distal supraclinoid portion and that the ACA and MCA are also occluded in the proximal portions. Mild MMVs are seen in the basal cerebral region. E, Left vertebral arteriogram in lateral projection shows that the enlarged branches of the PCA have sent leptomeningeal collateral vessels (arrow) to the ACA territory.
nosed as occluded only with MRI, and 10 normal PCAs were diagnosed as stenotic only with MRI.

Basal Cerebral MMVs
Of a total of 92 cerebral hemispheres that were studied, high-resolution turbo MR angiography and MRI accurately graded basal cerebral MMVs in 90 (98%) and 90 (98%) hemispheres, respectively (Table 2 and Figure 1). In 84 cerebral hemispheres in which moyamoya disease was manifest, MMVs were not detected in only 2 hemispheres (2%) with high-resolution turbo MR angiography. Similarly, MMVs were not detected in only 2 hemispheres (2%) with MRI.

Therefore, the sensitivity and specificity, respectively, for the detection of MMVs were 98% and 100% for high-resolution turbo MR angiography and 98% and 100% for MRI (Table 4). The accuracy of high-resolution turbo MR angiography was comparable to that of MRI ($P>0.05$).

Leptomeningeal and Transdural Collateral Vessels
On the basis of findings at conventional angiography, leptomeningeal collateral vessels from the posterior to the anterior circulation were present in 51 cerebral hemispheres (Table 3). High-resolution turbo MR angiography depicted leptomeningeal collateral vessels in all 51 cerebral hemispheres (100%) (Figure 1). MRI depicted leptomeningeal collateral vessels in 38 cerebral hemispheres (75%) but failed to identify leptomeningeal collateral vessels in 13 other hemispheres. Thus, the respective sensitivity and specificity for the detection of leptomeningeal collateral vessels were 100% and 100% for high-resolution turbo MR angiography and 75% and 100% for MRI. The sensitivity of high-resolution turbo MR angiography was significantly higher than that of MRI (Table 4) ($P<0.001$).

On the basis of findings at conventional angiography, transdural collateral vessels were present in 43 cerebral hemispheres (Table 3). High-resolution turbo MR angiography depicted transdural collateral vessels in 38 hemispheres (88%) (Figure 3), although it failed to identify transdural collateral vessels in 5 hemispheres. However, MRI detected no transdural collateral vessels in the 43 cerebral hemi-
spheres. Thus, the respective sensitivity and specificity for the detection of transdural collateral vessels were 88% and 100% for high-resolution turbo MR angiography and 0% and 100% for MRI. The sensitivity of high-resolution turbo MR angiography was significantly higher than that of MRI (P<0.001) (Table 4).

**Patient-by-Patient Analysis**

Bilateral steno-occlusive lesions were depicted with both high-resolution turbo MR angiography and MRI in all 42 patients with moyamoya disease and were not detected in 4 patients without moyamoya disease. Thus, the respective sensitivity and specificity for the detection of bilateral steno-occlusive lesions were 100% for high-resolution turbo MR angiography and 0% for MRI. The sensitivity of high-resolution turbo MR angiography was consistent with that determined by conventional angiography. This accuracy was comparable in limited accuracy (79%) of conventional MR angiography and has several limitations, including a long scan time, lower spatial resolution, and thicker section thickness, which result in limited accuracy (79%) of conventional MR angiography for the diagnosis of moyamoya disease. Therefore, a more accurate, noninvasive method in reduced scan time is needed for the definitive diagnosis of moyamoya disease.

The results of a patient-by-patient analysis showed that high-resolution turbo MR angiography had a sensitivity of 100%, a specificity of 99%, and an accuracy of 100% for the detection of steno-occlusive lesions. Furthermore, in 349 (95%) of the 368 arteries that were studied, the grade of steno-occlusive lesions determined by high-resolution turbo MR angiography was consistent with that determined by conventional angiography. It must be noted that severely stenotic arteries may be overestimated as occluded with MR angiography. However, given an accuracy of 100%, high-resolution turbo MR angiography seems to be an alternative method to detect steno-occlusive lesions in patients with moyamoya disease.

In the lesion-by-lesion evaluation, high-resolution turbo MR angiography was found to have a sensitivity of 98%, a specificity of 100%, and an accuracy of 98% for the detection of basal cerebral MMVs. Furthermore, in 90 (98%) of the 92 cerebral hemispheres, the grade of MMVs determined with MR angiography was consistent with that determined with conventional angiography. This accuracy was comparable with that of MRI. It must be noted that very mild MMVs may be underestimated with MR angiography. However, given an accuracy of 98%, high-resolution turbo MR angiography seems to be an alternative method for the detection of basal cerebral MMVs in patients with moyamoya disease.

The results of a patient-by-patient analysis also substantiated the validity of high-resolution turbo MR angiography in known as MMVs, that supply the ischemic brain. Cerebral ischemia tends to occur more often in children, whereas intracranial hemorrhages are more often seen in adults. The usefulness of conventional MR angiography in the diagnosis of moyamoya disease has already been reported, but it has several limitations, including a long scan time, lower spatial resolution, and thicker section thickness, which result in limited accuracy (79%) of conventional MR angiography for the diagnosis of moyamoya disease. Therefore, a more accurate, noninvasive method in reduced scan time is needed for the definitive diagnosis of moyamoya disease.

**Discussion**

Moyamoya disease is characterized by progressive narrowing of the bilateral internal carotid bifurcation, followed by the development of extensive parenchymal collateral vessels,
the diagnosis of moyamoya disease. High-resolution turbo MR angiography for bilateral steno-occlusive lesions of the internal carotid bifurcation had a 100% sensitivity and 100% specificity. High-resolution turbo MR angiography for bilateral basal cerebral MMVs had 98% sensitivity and 100% specificity. Thus, high-resolution turbo MR angiography for the diagnosis of moyamoya disease was found to be 98% sensitive and 100% specific. Given an accuracy of 98%, high-resolution turbo MR angiography appears to be a valid alternative method with which to achieve a diagnosis of moyamoya disease. We compared the sensitivity and specificity of high-resolution turbo MR angiography with those of conventional angiography for the diagnosis of moyamoya disease. We did not perform conventional MR angiography, due to limited examination times, and we compared high-resolution turbo MR angiography, MRI, and conventional angiography. The accuracy (98%) of high-resolution turbo MR angiography in the present study was higher than that (79%) of conventional MR angiography in the previous reports.14,15

Turbo MR angiography measures less data than conventional MR angiography, and the residual spaces in the k-space are filled with zeros, thus reducing the scan time.7,8 The ZFI technique generates thinner slice sections and increases matrix size along the in-plane phase-encoding direction. Thus, the ZFI technique allows for high-resolution turbo MR angiography in less scan time. A prominent limitation of conventional MR angiography is that the spatial resolution is much lower than that of conventional angiography.11,14,15 However, by using high-resolution turbo MR angiography with the ZFI technique, thinner slice sections with increased matrix size can be obtained in reduced scan time.7–10 This probably accounts for why high-resolution turbo MR angiography in reduced scan time had a high degree of accuracy for the evaluation of both steno-occlusive lesions and basal cerebral MMVs compared with that of conventional MR angiography in previous reports.11–16 Furthermore, we used other new imaging techniques, including MTC, TONE, and multislab, in the pulse sequence of high-resolution turbo MR angiography.17–20 These techniques also appear to contribute to a higher accuracy of high-resolution turbo MR angiography in reduced scan time than that of conventional MR angiography in earlier reports that did not use these techniques.11–16

The present results also demonstrated that high-resolution turbo MR angiography had a high level of accuracy (100%) for the detection of leptomeningeal collateral vessels from the PCA to the anterior circulation. Recent reports have shown that the presence of leptomeningeal collateral vessels is significantly correlated with regional cerebral blood flow and perfusion in moyamoya disease.21,22 Thus, high-resolution turbo MR angiography appears to be highly reliable for the assessment of cerebral ischemia in moyamoya disease. Furthermore, results of the present study have shown a high level of accuracy (95%) for the detection of transdural collateral vessels by high-resolution turbo MR angiography. Recently, surgical revascularization has been 1 treatment option for moyamoya disease, so postoperative vascular imaging has been needed to evaluate bypass patency.23 In this regard, our data demonstrate that high-resolution turbo MR angiography in reduced scan time is also suitable for the postoperative evaluation of bypass surgery in patients with moyamoya disease.

Another important aspect of moyamoya disease is its hereditary tendency, showing occasional familial occurrence.11,12 Therefore, high-resolution turbo MR angiography might have a role in the screening of asymptomatic relatives of patients with moyamoya disease. The regional cerebral blood flow in moyamoya disease is decreased in areas with severe steno-occlusive lesions and no collateral vessels.21,22 Thus, high-resolution turbo MR angiography, which accurately evaluates steno-occlusive lesions and collateral vessels, appears to be highly reliable for the detection of low flow. Recently, contrast-enhanced MR angiography has been reported for intracranial arterial disease.24 Compared with the time-of-flight technique, contrast-enhanced MR angiography may have some gain in efficacy because the arterial signal is less dependent on flow effects.

In conclusion, high-resolution turbo MR angiography in reduced scan time was found to have a high degree of accuracy (98% to 100%) for the assessment of both the steno-occlusive lesions and collateral vessels in patients with moyamoya disease. Furthermore, it also depicted leptomeningeal and transdural collateral vessels with a high degree of accuracy (95% to 100%). Therefore, high-resolution turbo MR angiography in reduced scan time is a valid alternative method to provide a highly accurate (98%), definitive diagnosis and assessment of moyamoya disease.

References


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