cerebral infarction occurred more frequently in patients found to have cerebral ischemia on medical examination, no cerebrovascular events reportedly occurred in the 6 patients who underwent cerebral revascularization[45]. Therefore, patients with asymptomatic moyamoya disease are also considered to be potentially at risk for cerebrovascular events. When the disease is conservatively followed up, careful long-term observation of the course using MRI/MRA is considered to be necessary.

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Classification of evidence levels and recommendation grades in the Guidelines.

**Table 1 Classification of evidence levels in the Guidelines**

| Evidence level | Contents |
|---|---|
| Ia | A meta-analysis of RCTs (The results of RCTs are practically consistent.) |
| Ib | RCT |
| IIa | Well-designed controlled study (non-randomized) |
| IIb | Well-designed quasi-experimental study |
| III | Well-designed non-experimental, descriptive study (comparison/ correlation/ case study) |
| IV | Report/comments/experience of specialists |

This classification is according to that adopted in the "2004 Guidelines for Stroke Treatment" by the Japan Stroke Society.

**Table 2 Classification of recommendation grades in the Guidelines**

| Recommendation grade | Contents |
|---|---|
| A | Strongly recommended |
| B | Recommended |
| C1 | Can be considered, but adequate scientific rationale lacking |
| C2 | Not recommendable because of absence of scientific rationale |
| D | Not recommended |

Precautions for the use of the Guidelines

(1) The clinical condition needs to be assessed in individual patients, and the Guidelines are not uniformly applicable to all individual patients. Therefore, the judgment of the treating physician who most accurately understands his/her patient's clinical condition should be afforded priority in the management of patients with moyamoya disease.

(2) The Guidelines should not be referred to without careful consideration, used as data for evaluation of medical examinations, or for medical accidents or lawsuits. Attention should be paid to the fact that the Guidelines include drugs not approved or therapies not authorized in Japan and drugs used for purposes other than the original intent.

(3) The number of patients is small and the cause of moyamoya disease is still unknown. Therefore, there are many aspects of the disease that still remain unresolved and for which adequate amount of evidence has not yet been collected. Therefore, it must be borne in mind while using the Guidelines that there may be many inaccuracies and that the contents may not always necessarily be up to date or the best for the time.

2005 to 2008 Health Labour Sciences Research Grant for Research on Measures for Intractable Diseases
Research Committee on the Pathology and Treatment of Spontaneous Occlusion of the Circle of Willis

Chief Researcher:
Nobuo Hashimoto, Department of Neurosurgery, Kyoto University Graduate School of Medicine
(Current) National Cardiovascular Center

Researchers:
Teiji Tominaga, Department of Neurosurgery, Tohoku University Graduate School of Medicine
Susumu Miyamoto, Department of Cerebrovascular Surgery, National Cardiovascular Center
Izumi Nagata, Department of Neurosurgery, Nagasaki University Graduate School of Biomedical Sciences
Kiyohiro Houkin, Department of Neurosurgery, Sapporo Medical University, School of Medicine
Norihiro Suzuki, Department of Neurology, Keio University School of Medicine
Akio Koizumi, Department of Health and Environmental Sciences, Kyoto University, Graduate School of Medicine, School of Public Health
Shigeru Nogawa, Department of Internal Medicine, Tokyo Dental College Ichikawa General Hospital
Jyoji Nakagawara, Department of Neurosurgery, Nakamura Memorial Hospital
Kazuo Kitagawa, Department of Neurology, Osaka University, Graduate School of Medicine
Satoshi Kuroda, Department of Neurosurgery, Hokkaido University Graduate School of Medicine
Kenichiro Kikuta, Department of Neurosurgery, Kyoto University Graduate School of Medicine

Research collaborators:
Miki Fujimura, Department of Neurosurgery, Tohoku University Graduate School of Medicine
Jun Takahashi, Department of Cerebrovascular Surgery, National Cardiovascular Center
Kentaro Hayashi, Department of Neurosurgery, Nagasaki University Graduate School of Biomedical Sciences
Koichi Oki, Department of Neurology, Keio University School of Medicine
Haruhiko Hoshino, Department of Neurology, Keio University School of Medicine

Secretariat:
Yasushi Takagi, Department of Neurosurgery, Kyoto University Graduate School of Medicine