doi: 10.2176/jns-nmc.2022-0072

Neurol Med Chir (Tokyo) 62, 307-312, 2022

Online May 25, 2022

Diagnostic Criteria for Moyamoya Disease - 2021 Revised Version

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Abstract

In this report, we, the Research Committee on Moyamoya Disease (Spontaneous Occlusion of the circle of Willis), describe in detail the changes in the new "Diagnostic Criteria 2021" for moyamoya disease and its scientific basis to make it widely known to the world. The revised criteria cover all aspects of the disease, including a definition of the disease concept, diagnostic imaging, and the concept of quasi-moyamoya disease (moyamoya syndrome).

Keywords: moyamoya disease, diagnostic criteria, cerebral angiography, MRI

Introduction

Moyamoya disease has been reported sporadically in Japan since the 1950's, and it was recognized as an independent disease entity in the late 1960's.[1-3] It has been more than 50 years since Suzuki and Takaku first reported it as "moyamoya disease" in 1969 in an English-written journal.[4] Currently, moyamoya disease is widely known around the world, and many research results and treatment outcomes have been reported. However, it is needless to say that an accurate diagnosis of moyamoya disease is essential for the basis of these results.[5]

We, the Research Committee on Moyamoya Disease (Spontaneous Occlusion of the circle of Willis), were established in Japan in 1974, and we have been studying the diagnosis and treatment of moyamoya disease for approximately 45 years. In 1978, this committee first established the diagnostic criteria for moyamoya disease. Since then, the criteria have been revised four times, in 1987, 1995, 2009, and 2015, in accordance with the changes in disease concepts and advances in diagnostic imaging. In 2021, we refined the diagnostic criteria for moyamoya disease again to further improve the accuracy of the diagnosis.

The diagnostic criteria for this worldwide known moyamoya disease should be shared widely across the world as a common language. We have published an English version of the 1995 diagnostic criteria in 1997[6] and then an English version of the 2009 diagnostic criteria in 2012.[7] However, an English version of the 2015 diagnostic criteria has not been published yet.[8] In this report, therefore, we disclose the English version of the 2015 and 2021 diagnostic criteria together for the first time in English and discuss the reasons for the revisions.

Definition of Moyamoya Disease on Cerebral Angiography

The 2009 English version of the diagnostic criteria is

Received March 3, 2022; Accepted March 22, 2022

Table 1 Diagnostic criteria 2009 of moyamoya disease (English version published in 2012[17])

Diagnostic Criteria 2009

(1) Cerebral angiography is considered essential for the diagnosis and must show at least the following findings:
  (i) Stenosis or occlusion of the terminal portion of the internal carotid artery or the proximal portion of the anterior and/or the middle cerebral artery.
  (ii) Abnormal vascular networks in the vicinity of the occlusive or stenotic lesions in the arterial phase.
  (iii) Bilaterality of findings (i) and (ii).

(2) However, when magnetic resonance imaging (MRI) and magnetic resonance angiographic (MRA) findings meet all of following criteria, cerebral angiography can be omitted. See the "Guideline for Diagnostic Imaging by MRI and MRA."
  (i) MRA shows stenosis or occlusion of the intracranial internal carotid artery or the proximal portions of the anterior and/or middle cerebral artery.
  (ii) MRA shows abnormal vascular networks in the basal ganglia on MRA.
  Note: When two or more visible flow voids are present in the basal ganglia on MRI, at least unilaterally, they can be deemed as representing an abnormal vascular network.
  (iii) Bilaterality of findings (i) and (ii).

(3) Moyamoya disease is an illness of unknown etiology. The differential diagnosis of this disease includes similar cerebrovascular lesions associated with the following underlying diseases, which should therefore be excluded:
  (i) Atherosclerosis,
  (ii) Autoimmune disease,
  (iii) Meningitis,
  (iv) Brain tumors,
  (v) Down's syndrome,
  (vi) von Recklinghausen's disease,
  (vii) Head injury,
  (viii) Cerebrovascular lesions after head irradiation,
  (ix) Others.

(4) Pathological findings that can be used as references for the diagnosis
  (i) Thickening of the arterial intima, mainly in the terminal portion of the internal carotid arteries, and narrowing or blockade of the lumen caused by this change, usually bilateral. Occasionally, lipid deposits are also present in the thickened intima.
  (ii) Arteries such as the anterior, middle, and posterior cerebral arteries forming the circle of Willis occasionally show varying degrees of stenosis or occlusion associated with fibrocellular thickening of the intima, waviness of the internal elastic lamina, and thinning of the media.
  (iii) Numerous small vascular channels (perforating and anastomotic branches) can be seen around the circle of Willis.
  (iv) Pia mater may also show reticular conglomerates of small vessels.

Diagnostic Assessment

Moyamoya disease should be classified as definite or probable based on the abovementioned items (1) to (4). When autopsy is performed in the absence of cerebral angiography, the condition should be diagnosed based on the criteria in item (4).

Definite moyamoya disease:

All criteria listed in (1) or (2) and in (3) should be met. In children, however, the criteria in item (1) or (2) (i) and (ii) on one side and visible stenosis around the terminal portion of the internal carotid artery on the other side are sufficient for a definite diagnosis.

Probable moyamoya disease:

All criteria are fulfilled, except item (1) (iii) and/or item (2) (iii) among the criteria of (1) or (2) and (3).

shown in Table 1.[7] As stated therein, the localization of the main vascular lesion in moyamoya disease has long been defined since 1978 as "the terminal portion of the internal carotid artery and the proximal portion of the anterior and middle cerebral arteries." In the 2015 revision,[8] this part of the phrase was changed to "the arteries centered on the terminal portion of the intracranial internal carotid artery" to further emphasize that a stenotic lesion at the terminal portion of the internal carotid artery is the most fundamental feature of moyamoya disease (Table 2).

In addition, the 2015 revision eliminated the distinction between "definite case" and "probable case," which had been used for a long time since 1978.[8] When moyamoya disease was first discovered, it was unclear whether the pathogenesis differed between "definite cases" with bilateral stenotic lesions and "probable cases" with unilateral stenotic lesions. Subsequently, however, many reports revealed that unilateral moyamoya disease is not rare to pro-

Table 2 Diagnostic criteria 2021 of moyamoya disease

Diagnostic Criteria 2021

A. Radiological Findings

Radiological examination such as cerebral angiography is essentially mandatory for diagnosis, and at least, the following findings must be present.

Especially in the case of unilateral lesions or lesions complicated by atherosclerosis, it is essential to perform cerebral angiography to exclude other diseases.

1. Cerebral angiography
   (1) Stenosis or occlusion in the arteries centered on the terminal portion of the intracranial internal carotid artery.
   (2) Moyamoya vessels (abnormal vascular networks) in the vicinity of the occlusive or stenotic lesions in the arterial phase.
   Note: Both bilateral and unilateral cases can be diagnosed as moyamoya disease.
2. MRI and MRA
   Moyamoya disease can be diagnosed when all of the following findings are found on MRI and MRA (time-of-flight; TOF) using a scanner with a static magnetic field strength of 1.5 Tesla (T) or higher (3.0 T is even more useful).
   (1) Stenosis or occlusion of the terminal portion of the intracranial internal carotid artery.
   (2) Decrease in the outer diameter of the terminal portion of the internal carotid artery and the horizontal portion of the middle cerebral artery bilaterally on heavy T2-weighted MRI.
   (3) Abnormal vascular networks in the basal ganglia and/or periventricular white matter on MRA.
   Note: When two or more visible flow voids are present in the basal ganglia and/or periventricular white matter at least unilaterally on MRI, they can be judged as representing abnormal vascular networks.
   Note: It is important to confirm the presence of a decrease in the outer diameter of the involved arteries on heavy T2-weighted MRI in order to differentiate atherosclerotic lesions.

B. Differential Diagnosis

Moyamoya disease is a disease of unknown etiology, and similar cerebrovascular lesions associated with the following should be excluded as quasi-moyamoya disease or moyamoya syndrome.

(1) Autoimmune disease (SLE, antiphospholipid syndrome, polyarteritis nodosa, Sjögren syndrome, etc.),
(2) Meningitis,
(3) Brain tumors,
(4) Down's syndrome,
(5) Neurofibromatosis type 1,
(6) Cerebrovascular lesions after head irradiation.

Note: Cases with hyperthyroidism can be diagnosed as moyamoya disease.

Diagnostic Assessment

Moyamoya disease is diagnosed when (1) and (2) of A-1 or (1) to (3) of A-2 are met and B is excluded.

The terms "definite case" and "probable case" were abolished in the 2015 revision of the diagnostic criteria for moyamoya disease.

gress to bilateral moyamoya disease. Since *RNF213* gene mutations were shown to play a major role in the pathogenesis of moyamoya disease in 2011, the knowledge regarding genetic mutations in moyamoya disease has expanded dramatically.[9,10] As the results, the genetic background of unilateral moyamoya disease and bilateral moyamoya disease has been shown to be similar.[11-14] More importantly, the term "probable" used to refer to unilateral moyamoya disease has given the impression to patients and medical professionals that unilateral moyamoya disease is not moyamoya disease. On the basis of these facts, when we revised the diagnostic criteria in 2015, we decided to remove the distinction between "definite case" and "probable case" to make patients and medical professionals aware that there is no longer any need to distinguish between bilateral and unilateral moyamoya cases.[8] In fact, in a recent multicenter study of unilateral moyamoya disease, RNF213 R4810K mutation was shown as one of the risk factors for contralateral progression in patients with unilateral moyamoya disease.[15]

Even after the revision in 2015, however, the fact that the distinction between "definite case" and "probable case" was removed has not widely been recognized in clinical settings, so we decided to include the sentence "Both bilateral and unilateral cases can be diagnosed as moyamoya disease" as a note in the 2021 version of the diagnostic criteria (Table 2).

Role of Magnetic Resonance Imaging (MRI) and Magnetic Resonance Angiography (MRA) in the Diagnosis of Moyamoya Disease

Since the diagnostic criteria were revised in 1995, MRI and MRA have played an important role in the diagnosis of moyamoya disease.[6] In fact, the English version of the 2009 diagnostic criteria published in 2012 states that when MRI and MRA findings meet all of the following findings, cerebral angiography can be omitted (Table 1):[7]

(i) MRA shows stenosis or occlusion of the intracranial internal carotid artery or proximal portions of the anterior and/or middle cerebral artery.
(ii) MRA shows abnormal vascular networks in the basal ganglia.
Note: When two or more visible flow voids are present in the basal ganglia on MRI, at least unilaterally, they can be deemed as representing an abnormal vascular network.
(iii) Bilaterality of findings (i) and (ii).

As has been repeatedly pointed out before, however, there are still many patients for whom it is difficult to differentiate moyamoya disease from other diseases such as atherosclerosis by MRI and MRA alone. In particular, this is apparent in adults with unilateral lesions and elderly patients. Cerebral angiography should be performed to provide final differentiation from atherosclerosis in such cases. In the 2021 version of the diagnostic criteria, therefore, we decided to clearly state as follows: "Especially in the case of unilateral lesions or lesions complicated by atherosclerosis, it is essential to perform cerebral angiography to exclude other diseases." Thus, the 2021 version of the diagnostic criteria emphasizes the importance of cerebral angiography more than ever.

Recent studies have shown that the involved arteries in moyamoya disease have a narrowing of the lumen due to intimal thickening and at the same time a reduction in their outer diameter on heavy T2-weighted MRI such as three-dimensional constructive interference in steady state (3D-CISS).[16,17] This finding becomes more pronounced as the disease progresses and occurs not only in the internal carotid artery system but also in the posterior cerebral artery.[18,19] On the other hand, in the case of middle cerebral arterial stenosis caused by atherosclerosis, the lumen is only narrowed and the outer diameter of the vessel does not shrink.[16,17] Similar findings specific for moyamoya disease have been reported by other investigators.[20,21] Therefore, in adults, especially the elderly, this imaging technique is considered useful to differentiate moyamoya disease from intracranial artery stenosis caused by atherosclerosis. Therefore, the above items (i) and (ii) remain almost the same as before, but for this 2021 revision, the finding of heavy T2-weighted images has been added as an essential item for diagnosis by MRI and MRA alone as follows:

(1) Stenosis or occlusion of the terminal portion of the intracranial internal carotid artery.
(2) Decrease in the outer diameter of the terminal portion of the internal carotid artery and the horizontal portion of the middle cerebral artery bilaterally on heavy T2-weighted MRI.
(3) Abnormal vascular networks in the basal ganglia and/or periventricular white matter on MRA.

Note: When two or more visible flow voids are present in the basal ganglia and/or periventricular white matter at least unilaterally on MRI, they can be judged as representing abnormal vascular networks.

Note: It is important to confirm the presence of a decrease in the outer diameter of the involved arteries on heavy T2-weighted MRI to differentiate atherosclerotic lesions.

However, it is important to note that in many cases, the reduction in the outer diameter of the involved artery does not become apparent until after Suzuki's Stage 3.[17] In other words, in the early stages of the disease (Suzuki's Stages 1-2), arterial shrinkage is often not apparent. In fact, it is well known that the diagnosis of moyamoya disease in the early stage of the disease is not easy by MRA alone. In early-stage cases where the above three findings cannot be obtained by MRI and MRA, we recommend definitive diagnosis by cerebral angiography.

Concept of Quasi-Moyamoya Disease (Moyamoya Syndrome)

When the diagnostic criteria for moyamoya disease were first established in 1978, moyamoya disease was defined as a disease of unknown cause. Therefore, when a patient meets the diagnostic criteria for moyamoya disease on cerebral angiography but has other comorbidities, the disease is defined as a "quasi-moyamoya disease" and is distinguished from moyamoya disease without any comorbidity. Quasi-moyamoya disease is also often referred to as moyamoya syndrome. In the 1987 revision, the specific diseases that may cause quasi-moyamoya disease (moyamoya syndrome) were listed up, and the list of the comorbidities has remained almost unchanged to date (Table 1).

However, some of them are considered inconsistent from the current perspective on moyamoya disease, so we decided to critically review the above comorbidities and to exclude atherosclerosis, hyperthyroidism, head trauma, and others from the underlying comorbidities of quasi-moyamoya disease in the 2021 version of the diagnostic criteria because of the reasons described below (Table 2).

First, as aforementioned, moyamoya disease and arteriosclerosis should be completely different in terms of their etiology and pathophysiology. The reason that atherosclerosis was considered one of the comorbidities of quasi-moyamoya disease is now unclear. However, many medical experts and researchers feel strange regarding calling intracranial arterial stenosis due to "atherosclerosis" as a

moyamoya disease, even if the word "moyamoya disease" is preceded with the prefix "quasi-." This opinion is supported by several reports. For example, Komotar et al. (2009) reported that indirect bypass did not promote spontaneous collateral formation in patients with symptomatic ICA or MCA steno-occlusion and hemodynamic failure,[22] although indirect bypass can develop in approximately 80% of adult patients with moyamoya disease.[23] As described above, the arterial shrinkage of the involved arteries can be observed in moyamoya disease but not in the intracranial arterial stenosis due to atherosclerosis.[16,17]

According to previous reports, most "autoimmune diseases" are hyperthyroidism. In the past, the majority of hyperthyroidism was thought to develop by Basedow's disease (Graves' disease) and was considered a very rare disease in Japan.[24] Because both moyamoya disease and hyperthyroidism were considered very rare previously, hyperthyroidism was considered the cause of quasi-moyamoya disease. However, recent studies have revealed that hyperthyroidism is more prevalent than previously thought, being as high as 16%.[25] In fact, Li et al. (2011) evaluated thyroid function in children with moyamoya disease (n = 114) and normal children (n = 114) and found that the frequency of hyperthyroidism in normal children was 0.9%, being much lower than 10.5% in moyamoya children ($p = 0.003$).[26] On the basis of their systematic review, Ahn et al. (2018) concluded that elevated anti-thyroid peroxidase antibody (TPOAb) and hyperthyroidism were significantly more frequently associated with moyamoya disease in adults than in healthy subjects (OR = 7.7 and 10.9, respectively).[27] Therefore, we decided to exclude hyperthyroidism from "autoimmune diseases" in the 2021 version of the diagnostic criteria. Autoimmune diseases other than hyperthyroidism have not been reported as frequently as hyperthyroidism in cases of comorbidities with moyamoya disease, so at present, other autoimmune diseases remain as the causes of "quasi-moyamoya disease" as before.

In addition, there are only two reports of moyamoya disease associated with head trauma even after searching PubMed.[28,29] In both reports, the causal relationship between head trauma and moyamoya disease is unclear. Finally, "others" are excluded from the comorbidities of quasi-moyamoya disease because the term "others" itself is quite unclear.

Conclusion

In this article, we have described in detail the changes in the new "Diagnostic Criteria 2015/2021" for moyamoya disease and its scientific basis to make it widely known to the world (Table 2). The revised criteria cover all aspects of the disease, including a definition of the disease concept, diagnostic imaging, and the concept of quasi-moyamoya disease (moyamoya syndrome). The "Diagnostic Criteria 2015/2021" would be the best solution for the current situation. At the same time, however, we would like to remind the readers that the diagnostic criteria may need to be reconsidered in the near future in view of the fact that novel findings and evidence on moyamoya disease are emerging one after another every year.

Acknowledgments

The authors thank the Japan Stroke Society, the Japan Neurosurgical Society, and the Japanese Society on Surgery for Cerebral Stroke for their critical review and approval of the 2021 revision of the diagnostic criteria. This work was supported by the Ministry of Health, Labor, and Welfare, Japan.

Conflicts of Interest Disclosure

There is no COI to be disclosed. All authors have registered online self-reported COI Disclosure Statement Form through the website for JNS members.

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