

Clinical Practice Guideline for Dementia 2017

Publication of the English Version of “Clinical Practice Guideline for Dementia 2017”

The first “Treatment Guideline for Dementia” was released in 2002. This was followed by publication of the “Treatment Guideline for Dementia 2010” using the Clinical Question (CQ) format in 2010. In 2014, revision of the “Treatment Guideline for Dementia 2010” was decided. Around this time, preparation of the English version of the “Clinical Practice Guideline for Chronic Headache 2013” was in progress, aiming to be published in 2015. Furthermore, the XXIII World Congress of Neurology was scheduled to be held in Kyoto in September 2017. These events prompted a plan to publish the English version of the revised treatment guideline for dementia.

In August 2017, the “Clinical Practice Guideline for Dementia 2017” in Japanese language was published. However, the English version was not ready in time for the XXIII World Congress of Neurology. Nevertheless, discussions were continued toward publication of the English version as originally planned.

After the publication of the “Clinical Practice Guideline for Dementia 2017” (Japanese version), some questions and opinions were received. The Committee addressed these issues and made some revisions, and at the same time simplified the contents to some extent to prepare the text for the English version of the Guideline.

The policies of developing clinical practice guidelines are gradually evolving, and the current guideline is expected to be revised in the future. For the development of future guideline, some issues in the “Clinical Practice Guideline for Dementia 2017” have to be addressed. These will be discussed in the next revision. With anticipation to develop further improved and more refined clinical practice guideline for dementia in the future, we embarked on preparing the English version of “Clinical Practice Guideline for Dementia 2017”.

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Preface:

Clinical Practice Guideline for Dementia 2017, Japanese version

July 2017

The first “Treatment Guideline for Dementia” was published in 2002. Then in 2010, six academic societies; namely, the Japanese Society of Neurology, Japanese Society of Neurological Therapeutics, Japanese Society of Psychiatry and Neurology, Japan Society for Dementia Research, Japan Geriatrics Society and Japanese Psychogeriatric Society, jointly developed the “Treatment Guideline for Dementia 2010” using the Clinical Question (CQ) format. Thereafter, with the addition of some new knowledge, the “Treatment Guideline for Dementia 2010, Compact Edition” was published in 2012. Subsequent to this publication, revision of the guideline was discussed at the Japanese Society of Neurology Guideline Executive Committee, and revision of the guideline was decided in 2014.

[Target Readers of this Guideline]

Along with the 2002 and 2010 guidelines, the current guideline was prepared assuming that the readers are general doctors in principle. However, the preparation work was done with the intention that this guideline will also be read by people other than doctors.

[Flow of Guideline Revision]

In 2014, after revision of the guideline was decided, chairman of the committee was elected and the office of guideline preparation was placed in the chairman’s affiliated facility. In the same manner as the previous guideline, the revision task was to be undertaken jointly by the six dementia-related societies mentioned above. Another five societies were invited to participate or collaborate. Committee members, research collaborators, and evaluation/coordination committee members were elected from the six societies. The method for the revision work was discussed and decided at the committee, and it was decided that each committee member would provide feedback to his/her academic society as the work progressed. Moreover, although the previous guidelines were called “treatment guidelines”, the contents also included diagnosis. Like the previous guidelines, the revised guideline is not limited to treatment but covers the entire clinical practice from diagnosis to treatment of dementia. Therefore, the revised guideline was developed as a “clinical practice guideline”

The Japanese Society of Neurology Guideline Executive Committee considered that although the current revision was in principle to be implemented according to the Minds Handbook for Clinical Practice Guideline Development 2014, the system was still not fully developed in Japan. As a policy to respond appropriately to the actual situation, it was decided that the specific methods for guideline development should be decided through discussions at the Dementia Clinical Practice Guideline Development Committee.

Based on the above guideline development policy, the guideline: (1) used the CQ format; (2) confirmed the sources of funding for guideline preparation and managed the conflict of interest (COI) of committee members; (3) conducted literature search using a uniform method; (4) discussed and decided evidence level and strength of recommendation based on the GRADE system recommended by Minds 2014; (5) listened to opinions from patient groups; (6) for CQs in which “recommendation” statement was difficult to compose because the strength of recommendation could not be decided, an “Answer” statement was prepared; (7) the draft guideline was reviewed by the evaluation/coordination committee and external committees, and (8) the draft guideline was opened to the public, and comments were invited. Thereafter, the “Clinical Practice Guideline for Dementia” was finalized.

[Funding Sources and Conflict of Interest (COI)]

Funds necessary for preparation of this guideline were borne by the Japanese Society of Neurology. The funds provided payment of expenses such as meeting room charges for committee meetings and transportation expenses for attending the committee, but did not provide remuneration for committee members and research collaborators for drafting manuscripts and participation in meetings.

This guideline was prepared based on appropriate COI management according to the “Regulations related to preparation of Japanese Society of Neurology clinical practice guideline”, “Guide to preparation of Japanese Society of Neurology clinical practice guideline” and “Rules for establishment and operation of Japanese Society of Neurology conflict of interest committee”.

Every year, the chairman, vice chairman, committee members, research collaborators, and evaluation/coordination committee members declared COI to the Chairman of Board of Directors of the Japanese Society of Neurology. Declaration was based on the following criteria: board member remuneration, etc. (1 million yen or more); stocks, etc. (1 million yen or more, or 5% or more of the total stocks); patent royalty (1 million yen or more); lecture fees, etc. (500,000 yen or more); manuscript fee, etc. (500,000 yen or more); contract research fee, joint research fees, etc. (2 million yen or more, 1 million yen or more for the 2015 declaration); scholarship (incentive) donation, etc. (2 million yen or more, 1 million yen or more for the 2015 declaration); endowed chair; provision of travel and gifts (50,000 yen or more).

Enterprises that declared COI are as follows:

Ajinomoto Co., Inc.; Astellas Pharma Inc.; AstraZeneca Co., Ltd.; Igaku-Shoin, Ltd.; Iset Co.; Eisai Co., Ltd.; MSD K.K.; Otsuka Pharmaceutical Co., Ltd.; Ono Pharmaceutical Co., Ltd.; Kyowa Hakko Kirin Co., Ltd.; GlaxoSmithKline K.K.; Kowa Pharmaceutical Co., Ltd.; Social and Medical Corporation Kowakai Sapporo Shirakabadai Hospital; Medical Corporation Seijinkai; Daiichi Sankyo Co., Ltd.; Sumitomo Dainippon Pharma Co., Ltd.; Takeda Pharmaceutical Co., Ltd.; Chugai Pharmaceutical Co., Ltd.; Nippon Boehringer Ingelheim Co., Ltd.; Nippon Medi-Physics Co., Ltd.; Novartis Pharma K.K.; Bayer Yakuhin Ltd.; Pfizer Japan, Inc.; Mochida Pharmaceutical Co., Ltd.; Morimoto Pharma Co., Ltd.; Janssen Pharmaceutical K.K.

[Process of Guideline Revision, Determination of Level of Evidence and Recommended Grade]

The first committee meeting was held in September 2014, and guideline development work began. First, we invited an instructor from Minds to confirm the Minds policy for guideline development. We also invited external committee members to participate, and decided the committee composition, guideline development policy, schedule, and items, and also decided work sharing. The scope was discussed. The scope of this guideline was decided to cover from diagnosis to treatment and long-term care for dementia. CQs would be prepared taking into consideration that the guideline is a reference material for doctors to decide the best methods of clinical care. Comments would be written as general items according to the scope. CQs were to be formulated with reference to PICO (P: patients, problem, population; I: interventions; C: comparisons, controls, comparators; O: outcomes).

Thereafter, the Guideline Executive Committee decided that this guideline is expected to be written concisely with contents that support routine clinical care. To address this point, it was necessary to consider the user-friendliness of guidelines; i.e., users can easily access the content they want to read from the table of contents. Initially, the plan was to limit CQs to important clinical issues, while general clinical and epidemiological features are described under the preface of each section. However, the general features also would have to be presented in a way such that readers can easily find what they want to read. Eventually, the plan was changed: all the contents are presented based on CQs. This format is similar to that used in the previous version. For this reason, items concerning specific clinical issues are mixed with items describing general clinical and epidemiological features, all under CQs (without the initially planned preface). Despite the adoption of this format in the present guideline, which is also partially due to consideration of continuity with the previous version, continuous efforts have to be made to find the optimal format for use in the next revision.

We determined key words (KW) for the CQs and requested Professor Shinichi Abe at the Academic Information Center, the Jikei University School of Medicine to perform search of literature. Since literature search for the previous guideline was conducted until 2008, the search period for this guideline was in principle from 2009. Therefore, literature search from 2009 until April 2015 was conducted during May to July 2015. From the reference list obtained from the search, the references were evaluated using evaluation sheets, not only by subcommittee members and research collaborators, but also by collaborators recommended by each committee member. The level of evidence for each outcome and the body of evidence were evaluated. For CQs that did not have sufficient references from this search process, the key words were changed as necessary, and literature search was repeated. Furthermore, additional manual search was conducted when it was deemed necessary by the committee.

Regarding systematic reviews, the system for conducting quantitative systematic reviews including meta-analysis is not yet adequately established. Therefore, following the policy of the Japanese Society of Neurology Guideline Executive Committee, quantitative systematic review was a non-binding target, and would be conducted if feasible based on the judgment of each committee member, while the main work was to conduct systematic literature search followed by qualitative systematic review, as was done in previous guideline development. All the collaborators of this work will be acknowledged with gratitude at the end of this message.

The level of evidence was evaluated not for individual reference, but by each outcome for studies grouped by study design, such as randomized controlled trial and observational study. The body of evidence was evaluated for risk of bias, indirectness,

inconsistency, imprecision, and publication bias. In the next revision, the methods of conducting systematic review including quantitative systematic review will be thoroughly examined before the work begins. Next, the responsible area for each committee member and research coordinator was decided. Texts for CQs, recommendation statements, and comments and evidence were drafted, and the draft was discussed and decided by all members of the committee.

Table 1. Recommendation grade and level of confidence.

Recommendation grade:

1 (Strong): Recommend to “perform” or “not to perform”

2 (Weak): Propose to “perform” or “not to perform”

Strength of body of evidence:

A: Strong

B: Moderate

C: Weak

D: Very weak

The manuscript thus prepared was reviewed by the evaluation/coordination committee. In addition, external committee members and persons related to patient organizations were invited to review the manuscript, and this guideline development committee also participated to listen to their opinions.

Public comments were invited during August 1–21, 2016. The comments received were reviewed and corrections were made. There were opinions pointing out the need for including more recent references, especially reports from Japan, after April 2015, which exceeded the period of literature search set for this guideline. This was discussed at the guideline development committee and additional manual search as needed was adopted. Since the public comments also included a request to conduct another round of public comment invitation, public comments were again invited during November 16–30, 2016.

[Contents and Items in this Guideline]

This guideline also places importance on continuity, and the contents almost conform to the previous “Treatment Guideline for Dementia 2010”. The guideline covers general items including definition, epidemiology, symptoms, assessment scales, diagnosis, examinations, non-pharmacological treatments and pharmacotherapy, interventions for delirium and coexisting diseases, risk factors, prevention, mild cognitive impairment, severity and interventions by severity, long-term care, social resources, community liaison, ethics, and legal issues. In addition, for diseases of dementia by etiology, which are dealt with in the section of “Specific Diseases”, Alzheimer’s disease dementia, Lewy body disease, frontotemporal lobar degeneration (FTLD), progressive supranuclear palsy (PSP), corticobasal degeneration (CBD), argyrophilic grain dementia, senile dementia of the neurofibrillary tangle type, vascular dementia, prion disease, and medical diseases are included.

For PSP, CBD, and Huntington’s disease, although management of motor symptoms is important, including these discussions will further expand the volume of the guideline. Therefore, it was decided to limit discussions to cognitive impairment only, which is in line with the previous guideline. Incidentally, a guideline for these diseases has been planned, which includes motor symptoms and all other disorders except cognitive impairment. The guideline was in the process of development as of November 2016.

Although molecular pathological classification shows that FTLD includes PSP and CBD, clinical differentiation of these diseases is possible based on clinical symptoms and other tests. Furthermore, since FTLD, PSP, and CBD are specified separately as designated intractable disease in Japan, these three diseases are handled as related diseases and described in separate chapters in this clinical practice guideline. For differential use of terms between FTLD and frontotemporal dementia (FTD), and between CBD and corticobasal syndrome (CBS), please refer to the descriptions in the corresponding chapters.

On the other hand, since a separate guideline for prion disease has been developed (Clinical Practice Guideline for Prion Disease 2014), prion disease is described briefly in this guideline while citing the Clinical Practice Guideline for Prion Disease 2014.

In an evaluation questionnaire after publication of the previous guideline, there was opinion that medical diseases caused by vitamin deficiency should also be included. In this guideline, therefore, vitamin deficiency, hypothyroidism, neurosyphilis, hepatic encephalopathy, and idiopathic normal pressure hydrocephalus (iNPH) are described. In the next revision, while reviewing the target diseases to be included in the guideline, it seems necessary to discuss whether the above diseases/items should be included.

[Names of therapeutic drugs and terms]

In principle, the terms used in the previous “Treatment Guideline for Dementia 2010” are adopted in this guideline.

<Names of therapeutic drugs>

Drug that are approved for dementia treatment in Japan and those that are used clinically even though they are not covered by health insurance for dementia treatment in Japan are written in katakana. Drugs that are used only overseas but not used in Japan are written in English.

<“Cognitive impairment” and “behavioral and psychological symptoms of dementia (BPSD)”>

“Core symptom” of dementia is described as “cognitive impairment”. The term “peripheral symptoms” is not used, and is replaced by “behavioral and psychological symptoms of dementia (BPSD)”. A combination of cognitive impairment and BPSD is described as “dementia symptoms”.

<Description of severity>

Regarding the severity of dementia, the terms “severe” and “high level” are used almost synonymously for the advanced stage. “Severe” was used in the Treatment Guideline for Dementia 2002 and 2010. Following these previous guidelines, “severe” is used in this guideline.

<Classification related to time of onset: “juvenile dementia”>

Depending on the time of onset, terms such as “juvenile dementia”, “pre-senile dementia”, “senile dementia”, “early-onset dementia”, and “late-onset dementia” are used. There is opinion that use of the term “juvenile dementia” may not be desirable, because the same term may denote different age groups [Japan Society for Dementia Research (Ed.): Textbook of Dementia, 2008]. However, the term “early-onset dementia” is used in administrative documents such as “Measures for early-onset dementia” (2009) (Ministry of Health, Labor and Welfare), as well as the Orange Plan and the New Orange Plan. In response to these developments, a patient aged under 65 years with dementia is referred to as “early-onset dementia” in this guideline (see “Chapter 5: Various Systems and Social Resources for Supporting Persons with Dementia and Their Families; C. Early-onset Dementia”).

<“Alzheimer’s disease” and “Alzheimer’s disease dementia”>

The term “Alzheimer’s disease” may refer to the pathological state of the disease or may be used to describe the clinical syndrome when dementia symptoms due to “Alzheimer’s disease” have become evident. According to the National Institute on Aging and Alzheimer’s Association (2011), “Alzheimer’s disease” is defined as a term encompassing the underlying pathophysiological processes, and “Alzheimer’s disease” is distinguished from “Alzheimer’s disease dementia” which describes the state of dementia caused by “Alzheimer’s disease”. In Japan, “Alzheimer’s disease dementia” has been used from the past as a term to describe the state of dementia considered to be caused by “Alzheimer’s disease”. Therefore, in this guideline, the term “Alzheimer disease dementia” is used for dementia that is considered to have occurred based on the pathological background of “Alzheimer’s disease”. However, this guideline does not differentiate between “Alzheimer disease dementia” (diagnosed from clinical features) and “Alzheimer disease dementia caused by Alzheimer’s disease” (evidence that dementia is caused by Alzheimer’s disease has been confirmed), because this differentiation has little utility in clinical practice.

<Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5)>

In DSM-5 published by the American Psychiatric Association in 2013, the term “neurocognitive disorder” was introduced. This term includes “delirium” and “major and mild neurocognitive disorder”. The Japanese translations for “major and mild neurocognitive disorder” are “dementia (DSM-5)” and “mild cognitive impairment (DSM-5)”, respectively.

<“Mild cognitive impairment” and “MCI”>

In this guideline, “mild cognitive impairment” and “MCI” are described as “mild cognitive impairment (MCI)”.

<Others>

Terms including “senile dementia of the neurofibrillary tangle type (SD-NFT)”, “tangle-predominant senile dementia/NFT-predominant form of senile dementia”, “tangle only dementia”, and “primary age-related tauopathy (PART)” are used uniformly in the guideline.

[Using this clinical practice guideline in clinical care for dementia]

This guideline provides reference materials to support the clinical practice and care for dementia with the purpose to improve clinical practice for dementia, and does not intend to constrain clinical practice for dementia in the clinical setting. With future developments and changes in medical care and research, as well as diversification of persons with dementia and the environment surrounding these persons, guidelines are expected to change over time. This guideline does not restrict the decision of clinicians over treatment, neither does it stipulate medical treatment in response to changes in the clinical setting. Sometimes the recommendations in this guideline may not apply. There are cases where the physician in charge may perform treatments that deviate from the descriptions in this guideline, and cases where the deviation may be appropriate. In actual clinical practice, it is important to formulate an individualized treatment plan that responds to the dementia of each patient, without being restricted by the contents of this guideline. In addition, the contents described in this guideline do not serve as grounds for medical lawsuits.

[Promotion of utilization of the guideline, future plans including the next revision, and evaluation]

In order to promote the utilization of this guideline, the guideline is scheduled to be introduced and publicized by posting on the websites of the academic societies that participated in guideline development, presented at scientific conferences and other meetings of the academic societies, and published in scientific journals.

For the convenience of readers, a compact edition has been published both for the Treatment Guideline for Dementia 2002 and 2010 editions. For this guideline also, publication of a compact version will be considered. Furthermore, the Japanese Society of Neurology has published the English version of other clinical practice guideline, and we hope that the possibility of publishing the English version of the Clinical Practice Guideline for Dementia will be discussed at the guideline development committee.

In the case that addition or correction is necessary due to new knowledge obtained after publication of this guideline, additional or revised CQ will be prepared as a supplement and posted on the society website. In addition, the Japanese Society of Neurology has a policy of revising clinical practice guidelines every five years. This guideline development committee will also consider the next revision and composition of the next guideline development committee, and the results will be submitted to the Japanese Society of Neurology Guideline Executive Committee for deliberation.

The previous two guidelines for the treatment of dementia were evaluated by an evaluation committee set up at the Japanese Society of Neurology. It is anticipated that the current Clinical Practice Guideline for Dementia 2017 will also be evaluated by an evaluation committee.

[Acknowledgment to committee members and research collaborators]

Revision and development of the present guideline were undertaken with the collaboration of six dementia-related associations. I would like to express my gratitude to all members of the committee and the research collaborators for their participation in the committee and for their great efforts despite their busy schedules. This guideline was developed with the support of many colleagues such as evaluation/coordinating committee members, external committee members, and collaborators. I would like to thank them all for their collaboration. I would also like to thank Ms. Noriko Ono and Ms. Yoriko Matsushita from the Tokyo Branch of the Alzheimer's Association Japan for their opinions in preparing the guideline. Furthermore, I would like to express my sincere appreciation to those who have provided many valuable comments in the invitation of public comments. I would also like to thank staff members of Minds for their advice.

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