

Practice parameter for diagnosis and evaluation of dementia

(Summary statement)

Report of the Quality Standards Subcommittee of the American Academy of Neurology

Overview. The Quality Standards Subcommittee of the American Academy of Neurology is charged with developing practice parameters for neurologists for diagnostic procedures, treatment modalities, and clinical disorders. The subcommittee defines practice parameters as results, in the form of one or more specific recommendations, from scientifically based evidence of a specific clinical problem. The selection of topics for which practice parameters are developed is based on factors such as prevalence, frequency of use, economic impact, membership need, controversy, urgency, external constraints, and resources required. This document will outline what we believe to be the most useful components of the diagnostic evaluation of elderly patients with cognitive complaints suggesting dementia. Recommendations in this paper have been designated as "standards," "guidelines," and "options" based on the strength of available supporting evidence; see Definitions on page 2205 of this article.

Justification. Dementia is a major cause of disability and death in developed countries and accounts for a disproportionate share of medical resource utilization and health care expenditures. Accurate diagnosis of dementia syndromes is important to detect reversible or arrestable dementias. In addition, the impact of a demented individual on his or her family is substantial; accurate diagnosis enables the clinician to provide anticipatory guidance to the patient and family, to more accurately prognosticate, to facilitate legal and financial planning, and to assist with providing access to community resources.

Practice parameter development process. A Medline search of the English-language literature from 1985 to 1993 was used to generate citations, using the key words "dementia," "senile dementia," "Alzheimer's disease," and "vascular dementia" cross-referenced with "diagnosis." Literature published before 1985 was sought by reviewing refer-

ence lists of the articles obtained in the literature search. Of 3,096 citations identified, we reviewed 1,843 abstracts or original articles, based on a preliminary screening of the article titles. Of these, we used 110 articles to prepare this document, those references having clearly described methods, uniformly applied diagnostic definitions, and sufficiently large samples of patients (usually more than 25).

Identification of dementia. Dementia is a clinical state characterized by a significant loss of function in multiple cognitive domains, not due to an impaired level of arousal. The presence of dementia does not necessarily imply irreversibility, a progressive course, or any specific cause.

Diagnosis of dementia requires *either* (1) assessing an individual's current level of cognitive function and documenting a higher level of intellectual function in the past, *or* (2) documenting a decline in intellectual function by serial examinations over time. Cognitive defects due to delirium, restricted brain lesions (eg, aphasia), and psychiatric problems (eg, depression) must be excluded. An initial diagnosis of dementia cannot be made when consciousness is impaired or when conditions exist that prevent adequate evaluation of mental status. If dementia is identified, further evaluation is necessary to determine the etiology of the dementia and to stage its severity.

Individuals who should be evaluated for evidence of dementia include those with memory or other cognitive complaints with or without functional impairment, elderly patients in whom there is a question of incompetency, depressed or anxious patients with cognitive complaints, and patients who arouse physician suspicion of cognitive impairment during their interview despite the absence of complaints (GUIDELINE).

Some patients may not meet criteria for dementia even though they or their families are concerned about changes in intellectual functioning.

Approved by the Quality Standards Subcommittee February 13, 1994. Approved by the Practice Committee February 26, 1994. Approved by the Executive Board May 6, 1994.

Received May 13, 1994. Accepted in final form August 2, 1994.

Address correspondence and reprint requests to Joanne F. Okagaki, American Academy of Neurology, Suite 335, 2221 University Avenue SE, Minneapolis, MN 55414.

This group may include well-educated, high-functioning individuals, patients with psychiatric problems (eg, depression or anxiety), and patients with early or very mild dementia who may be considered to be at risk for dementia. These patients should be encouraged to return for re-evaluation, since observation over time, often 6 to 12 months, may help to document cognitive decline (OPTION). For these patients, neuropsychological testing is often valuable to detect subtle cognitive difficulties (OPTION).

Depending on the severity of the dementia, a skillfully taken history may reveal deficits in several areas of intellectual function. For most patients, this information should be obtained from, or at least substantiated by, an informant. In taking a history, certain functional items, such as difficulty recalling recent events, preparing a meal, playing games of skill, filling out business forms, handling financial records, and shopping alone, are helpful in confirming the presence of a significant intellectual impairment. It is also useful to inquire about a family history of Alzheimer's disease (AD) or other dementia.

Most neurologists gather information regarding cognitive decline, presence of depression, evidence of vascular disease, and social and occupational functioning by history. Many of these elements have been incorporated into instruments that, especially in research settings, may assist the clinician in diagnosis (OPTION).

Cognitive or mental status testing should include assessment of level of arousal, attention, orientation, recent and remote memory, language, praxis, visuospatial function, calculations, and judgment (GUIDELINE). The techniques used to assess these domains are at the discretion of the individual physician (OPTION). Various brief mental status screening instruments are useful adjuncts that may improve recognition of deficits and enhance clinical judgment. However, test scores on such instruments do not, of themselves, establish a diagnosis of dementia nor do they determine the etiology of the dementing illness if one is present. Scores on such screening tests may be abnormal when any form of cognitive impairment exists, and mildly demented patients may score within the normal range. In addition, age, education, ethnicity, and language of the respondent have all been shown to influence responses to mental status test items, and the clinician must make allowances for each of these in assessing patients with cognitive difficulties. Although cut-off points have been recommended for some of the standardized, well-known mental status tests, they are not definitive.

Diagnostic workup. The neurologic history and examination (including mental status examination) are essential components of the diagnostic workup of dementia (STANDARD) and may reveal important clues to the etiology of the patient's dementia. Careful attention should be paid to the existence of focal abnormalities, extrapyramidal signs, and gait

disorders.

Diagnostic tests are also necessary in the differential diagnosis of dementia to rule out metabolic and structural causes (GUIDELINE). The detailed workup depends on the suspected diagnosis, but generally should include the following tests (GUIDELINE): complete blood cell count, serum electrolytes (including calcium), glucose, BUN/creatinine, liver function tests, thyroid function tests (free thyroid index and thyroid-stimulating hormone), serum vitamin B₁₂ level, and syphilis serology. Other tests may be helpful in certain circumstances, but are not recommended as routine studies (OPTION): sedimentation rate, serum folate level, HIV testing (order according to Centers for Disease Control suggestions), chest x-ray, urinalysis, 24-hour urine collection for heavy metals, toxicology screen, neuroimaging study (CT or MRI), neuropsychological testing, lumbar puncture, electroencephalography, positron emission tomography, and single-photon emission computed tomography.

Neuroimaging should be considered in every patient with dementia based on the clinical representation and may facilitate identification of potentially treatable conditions that can otherwise be missed, such as tumors, subdural hematomas, hydrocephalus, and strokes. However, these conditions are uncommon when not anticipated clinically, particularly when clinical evaluations are performed by experienced examiners. In particular, there is no consensus on the need for such studies in the evaluation of patients with the insidious onset of dementia after age 60 without focal signs or symptoms, seizures, or gait disturbances.

Although not generally necessary, *neuropsychological testing* may be helpful in (1) demonstrating cognitive impairment in individuals whose initial evaluation is borderline or suspicious; (2) distinguishing depression from dementia; (3) determining competency for legal purposes; and (4) assisting in the evaluation of early dementia, particularly when major decisions need to be made with regard to a patient's job (eg, disability determination) or other personal affairs (OPTION). Neuropsychological testing is most valuable in making a longitudinal diagnosis of dementia when the presence of a significant cognitive decline is difficult to establish, as it often is in extremely high-functioning individuals, in individuals with mental retardation, or in individuals with very limited educational backgrounds.

Lumbar puncture is *not* recommended as a routine study in the evaluation of dementia (OPTION). However, assuming there are no contraindications, a lumbar puncture should be performed when any of the following are present (GUIDELINE): metastatic cancer, suspicion of CNS infection, reactive serum syphilis serology, hydrocephalus, dementia in a person under age 55, a rapidly progressive or unusual dementia, immunosuppression, and suspicion of CNS vasculitis (particularly in patients with connective tissue diseases).

EEG is not recommended as a routine study but may assist in distinguishing depression or delirium from dementia (OPTION) and in evaluating for suspected encephalitis, Creutzfeldt-Jakob disease, metabolic encephalopathy, or seizures (OPTION).

Differential diagnosis. In a small percentage of cases (fewer than 15% on average), a specific treatable or reversible etiology of the dementing syndrome will be identified, usually with the assistance of the diagnostic studies noted above. The most important examples are medication-induced encephalopathy, depression, thyroid disease, central nervous system infections (eg, neurosyphilis or cryptococcal meningitis), vitamin deficiencies (especially vitamin B₁₂ deficiency), and structural brain lesions (eg, tumors, subdural hematomas, and hydrocephalus). When these conditions have been excluded, the remaining causes of dementia consist largely of AD and vascular dementia.

AD is the most frequent type of dementia in American and most European elderly, afflicting about 50 to 80% of subjects in various clinicopathologic series. Difficulty with memory is the first and most notable complaint of either the patient or the family. As the disease advances, problems with language, arithmetic calculation, visuospatial orientation, and praxis become increasingly apparent. Behavioral alterations such as depression, agitation, delusions, and hallucinations may become evident at any time during the course of the illness. The neurologic examination in AD (excluding mental status testing) is often normal. Associated features may include primitive reflexes (snout, glabellar, or grasp) and impaired graphesthesia. Late in the course, extrapyramidal signs (rigidity and bradykinesia), gait disturbances, and myoclonus can occur.

A diagnosis of AD is supported by the following: (1) the insidious onset and progressive worsening of dementia; (2) prominent difficulty with memory (especially retention and retrieval of new material) early in the course of the illness; (3) onset after age 60; (4) no focal signs or gait difficulties on neurologic examination, especially early in the course; and (5) exclusion of other treatable conditions. The acute or subacute onset of disability or the presence of focal signs, seizures, gait difficulty early in the course of the illness, or the presence of a significant behavioral alteration prior to a memory deficit suggests a dementia etiology other than AD.

A variety of useful and well-validated clinical criteria are available for diagnosis of AD (OPTION). With such criteria and with suitable laboratory and diagnostic studies, positive predictive values of 80 to 85% can be achieved, although lower rates are reported when patients with presenile onset are included. However, the usefulness of such criteria is limited by (1) the difficulty of identifying individuals with mild dementia or atypical presentations; (2) the exclusion of individuals with prevalent and

DEFINITIONS

Classification of evidence

Class I. Evidence provided by one or more well-designed randomized controlled clinical trials.

Class II. Evidence provided by one or more well-designed clinical studies such as case-control studies, cohort studies, and so forth.

Class III. Evidence provided by expert opinion, nonrandomized historical controls, or one or more case reports.

Strength of recommendations

Standards. Generally accepted principles for patient management that reflect a high degree of clinical certainty (ie, based on class I evidence or, when circumstances preclude randomized clinical trials, overwhelming evidence from class II studies that directly addresses the question at hand or from decision analysis that directly addresses all the issues).

Guidelines. Recommendations for patient management that may identify a particular strategy or range of management strategies and that reflect moderate clinical certainty (ie, based on class II evidence that directly addresses the issue, decision analysis that directly addresses the issue, or strong consensus of class III evidence).

Practice options or advisories. Other strategies for patient management for which there is unclear clinical certainty (ie, based on inconclusive or conflicting evidence or opinion). Practice options in certain clinical situations may be considered medically indicated.

Practice parameters. Results, in the form of one or more specific recommendations, from a scientifically based analysis of a specific clinical problem.

potentially overlapping conditions (eg, cerebrovascular disease); (3) the difficulty of applying the extensive psychometric and laboratory evaluations to routine clinical practice; and (4) the availability of considerable latitude for individual interpretation of the criteria, resulting in, at best, modest inter-rater reliability. Furthermore, it is not clear how much incremental gain in validity is achieved with these criteria over non-criterion-based clinical diagnoses, particularly for experienced clinicians.

Vascular dementia, caused by one or more small or large brain infarcts, constitutes about 5 to 10% of cases of dementia. Symptoms appear when a certain volume of infarcted tissue is present or if small strokes are strategically placed, but the size, number, and distribution of vascular lesions necessary to produce dementia remain uncertain.

A diagnosis of vascular dementia is supported by the following: (1) the sudden onset of dysfunction in one or more cognitive domains; (2) a stepwise deteriorating course; (3) focal neurologic signs, including weakness of an extremity, exaggeration of deep tendon reflexes, extensor plantar responses, and

gait abnormalities; (4) history or neuroimaging evidence of previous strokes; and (5) evidence of stroke risk factors and systemic vascular disease. These clinical features have been incorporated into a number of different clinical criteria or "ischemic" scores (OPTION). Unfortunately, there are at present no widely accepted criteria for the diagnosis of vascular dementia. Differentiation of vascular dementia from either AD with superimposed cerebrovascular disease or mixed AD and vascular dementia is especially difficult.

Acknowledgments

The Quality Standards Subcommittee wishes to express particular gratitude to QSS member, Douglas J. Lanksa, MD, for his work in preparing this practice parameter. A background paper written by Jody Corey-Bloom, MD, PhD, David Drachman, MD, Marshall Folstein, MD, Douglas Galasko, MD, Douglas J. Lanksa, MD, Murray Raskind, MD, and Leon Thal, MD, is available upon request from the American Academy of Neurology.

Quality Standards Subcommittee: Milton Alter, MD, PhD; Thomas N. Byrne, MD; Jasper R. Daube, MD; Gary Franklin, MD, MPH; Benjamin M. Frishberg, MD; Michael L. Goldstein, MD; Michael K. Greenberg, MD; Douglas J. Lanksa, MD; Shrikant Mishra, MD; Germaine L. Odenheimer, MD; George Paulson, MD; Richard A. Pearl, MD; Jay H. Rosenberg, MD (Chair); Cathy Sila, MD; and James C. Stevens, MD.

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