

HOSPITAL PHYSICIAN®

NEUROLOGY BOARD REVIEW MANUAL

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The *Hospital Physician Neurology Board Review Manual* is a peer-reviewed study guide for residents and practicing physicians preparing for board examinations in neurology. Each quarterly manual reviews a topic essential to the current practice of neurology.

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Neurodegenerative Disorders: Dementia

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Table of Contents

Introduction	2
Mild Cognitive Impairment	2
Alzheimer's Disease	3
Frontotemporal Lobar Degeneration	7
Vascular Dementia	8
Dementia of Lewy Body Type	9
Creutzfeldt-Jacob Disease	10
Case Discussions	12
Summary Points	14
References	14

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Neurodegenerative Disorders: Dementias

Malgorzata B. Franczak, MD, and Rama Maganti, MD

INTRODUCTION

The word *dementia* is an ancient term used by the Roman poet and philosopher Lucretius to describe the state of “being out of one’s mind.” Today, the term *dementia* describes a clinical syndrome characterized by acquired impairment in multiple neuropsychological and behavioral domains, including memory, cognition, visuospatial skills, and language. Most definitions of dementia refer to intellectual or cognitive deficits of severity sufficient to interfere with social or occupational functioning; however, the term *dementia* does not imply a specific cause or pathologic process.

Dementia is one of the most disabling and costly diseases associated with aging. More than 70 illnesses, some of which are nonprogressive, can cause dementia. All types of dementia are treatable—at least with psychosocial interventions—therefore, accurate diagnosis is essential to initiate appropriate treatment and provide information about prognosis and factors affecting the cause of illness. Although Alzheimer’s disease is the most common form of dementia, less common causes of dementia must also be considered when evaluating a patient with cognitive decline. For example, focal neurologic findings or progressive deterioration raise the suspicion of vascular dementia, whereas early onset of language dysfunction or personality changes suggests frontotemporal dementia; rapid progression of neurologic impairment should initiate evaluation for treatable/partially reversible conditions and prion disease.

Although dementia has often been considered a global disorder, no dementing illness involves all areas of the brain equally. The specific topography of involvement is manifested by two basic patterns of neuropsychological impairment: (1) *cortical dementias* such as Alzheimer’s disease and frontotemporal dementia, and (2) *subcortical dementias* such as vascular dementia, extrapyramidal syndromes, and normal pressure hydrocephalus. The clinical features of cortical and subcortical dementias differ as noted in **Table 1**.

This manual reviews the common and uncommon dementias. Three brief cases vignettes are discussed at the end of this review.

MILD COGNITIVE IMPAIRMENT

DIAGNOSIS

In general terms, mild cognitive impairment (MCI) is an intermediate stage between normality and dementia. Although the general concept is easy to grasp, the development of precise diagnostic criteria for MCI has been slow. Use of cut-off scores on neuropsychological tests ignores the possibility that individual patients may have always performed poorly and thus may not have a true decline in intellectual capacity. In contrast, patients who have deteriorated significantly might still score above the cut-off if their pre-morbid level of performance was high. These considerations have led to development of both research and practical guidelines for diagnosis of MCI. Practical guidelines for a diagnosis of amnesic MCI (adapted from the Petersen criteria¹) include: (1) memory complaint by patient, confirmed by family or physician, (2) normal activities of daily living, (3) normal general cognitive function, (4) objective memory impairment for age and education, and (5) no dementia present.

The research definition of MCI uses global staging scales such as the Global Deterioration Scale (score of 2 or 3) and the Clinical Dementia Rating Scale (score of 0.5) to define MCI. The Mini-Mental State Examination (MMSE) score for MCI is greater than 24 and usually in the range of 26 to 28. Because different cut-offs are used depending on the demographic group to maintain the sensitivity and specificity of the MMSE, this test is more useful to track the degree of impairment in a given individual than to make the diagnosis. Rather than the MMSE, neuropsychological testing using one or several memory tests with published norms is more appropriate for determining objective memory impairment. Memory function is usually assessed by learning over trials or delayed recall on a multiple-trial free-recall task, such as the Auditory Verbal Learning Test or the Wechsler Memory Scale-Revised or Wechsler Memory Scale-III, Logical Memory II, or Visual Reproduction II tests. Generally, scores of patients with MCI tend to fall 1.5 standard deviations below age- and education-matched controls.