

Recognizing Dementia: The Importance of an Accurate Diagnosis

BY JOE MAIER, JR.

D ementia is a syndrome in which the progressive deterioration of cognitive abilities is so severe that it interferes with an individual's normal social and occupational functioning. Although the prevalence of dementia increases with advancing years, dementia *is not a part of normal aging.* Because the diagnosis of dementia has significant implications for the patient and his or her family, it is critical for the physician to use available resources to assure an accurate diagnosis. The essential features of dementia are the development of multiple cognitive deficits that include memory impairment and at least one of the following cognitive disturbances: aphasia, apraxia, agnosia, or an alteration in executive functioning (see below).¹

Memory Loss and Cognitive Disturbances

Memory is the ability to perceive, interpret, store, and, when necessary, retrieve information for appropriate use. Typically, patients with dementia first exhibit changes associated with their short-term memory—forgetting newly acquired instructions or recently scheduled appointments. As dementia progresses, long-term and sensory memory are also impaired such that, for instance, someone married for 50 years may no longer recognize his or her spouse. Physicians must be aware that complaints about memory normally increase with age, but that in isolation



The typical US caregiver is a female in her mid-50s, often a wife caring for a spouse.

these complaints should not be used to form a diagnosis. Rather, they are an indication that further evaluation is necessary.

Aphasia—the inability to use and appropriately comprehend spoken language-manifests itself first in a patient's periodic inability to find a word. This may progress to the patient's use of nonsensical utterances or the loss of speech altogether. Apraxia is an impairment that interferes with a patient's ability to perform previously mastered skills. Normally apraxia is first seen in reduced capacity to carry out instrumental activities of daily living-for example, a career carpenter who is no longer able to hit a nail with a hammer or use a ruler to measure a length of wood. Ultimately, these patients become unable to carry out the basic activities of daily living (ADLs) such as bathing, grooming, and dressing. Agnosia is related to changes associated with visual and spatial perception. A patient exhibiting symptoms of this cognitive disturbance may enter an office and accidentally lower him- or herself on the arm of the chair instead of the seat. Finally, the process of executive functioning enables an individual to use information in successful decision-making endeavors and abstract reasoning, such as when driving. In dementia cases, physicians and caregivers report that removing driving privileges is the most difficult decision to make because, despite evidence to the contrary, the patient believes he or she is still able to drive and may become quite agitated when no longer allowed to do so. Additionally, as basic ADL losses mount, a patient may become resistant to the caregiver's efforts to help. Imagine a 70-yearold woman attempting to change the soiled diaper of her incontinent husband, all the while he is fighting her efforts because he is incapable of interpreting the intent or necessity of her actions.

Types of Dementia

Alzheimer's disease (AD) is the most common form of dementia, comprising approximately 60 percent of all cases, with cerebrovascular disease and Parkinson's disease making up roughly 19 percent of all dementia cases and mixed dementia accounting for about 16 percent. The remaining 5 percent is associated with Pick's, Huntington's, Lewy body, and Creutzfeldt-Jakob diseases, as well as HIV and head trauma. Due to the prevalence of Alzheimer's, unless otherwise stated, this article focuses on AD.

All dementias have common traits, specifically in how they affect the family—especially the primary caregiver. This person needs to be ever-vigilant, 24 hours a day, seven days a week, in many cases for as long as 10 to 20 years. As the disease and length of time progress, it becomes increasingly difficult for

Case Study A

Patient History

DS was a 67-year-old female diagnosed with AD by her primary care physician in June. The diagnosis was confirmed in a second opinion by a psychiatrist. Three months later, DS was brought to Crozer's Adult Day Center (ADC)—a dementia-specific day care program—by her daughter for an intake assessment.

At intake, the daughter explained that the physician and the psychiatrist told her that her mother could no longer live alone for safety reasons and so she had placed her on a waiting list for a local nursing home. In the meantime the daughter had taken a leave of absence from work and moved her mother into her home. At the time of intake, the daughter had been out of work for two months and was in jeopardy of losing her job. Being a single mother, she needed to return to work as soon as possible and was seeking ADC admission for her mother only until a bed in a nursing home became available.

Evaluation

During the intake process the medical report from the primary care physician was reviewed. The information was limited, but an MMSE score of 15—moderate dementia was identified. The ADC screening MMSE was 20, still in the range of dementia, but significantly improved over a period of time when one would normally expect a decline. As mentioned above, the only change in DS's life had been moving to her daughter's house, so a recent social history was conducted and identified:

- daily alcohol (ETOH) use for years, which stopped two months before the evaluation because of the move;
- daily use of over-the-counter sleep medication, which continued after the move;
- significant social isolation—limited to visits by her daughter—for 18 months prior to the move;
- weight loss of 30-40 pounds prior to the move and a gain of 10 pounds since; and
- feelings of depression and loneliness expressed by the patient.

Treatment

DS was admitted to the ADC program on a five-day-per-week schedule—allowing the daughter to return to work. DS's treatment plan included counseling for depression and nutrition, a non-alcohol formulation of sleep medication, and structured therapeutic activities within a social milieu. The patient was subsequently weaned off the sleep medication.

Within a month, DS showed significant signs of improvement. Her demeanor was appropriate, vegetative signs had disappeared, and she demonstrated retention of new information, such as names of staff and other clients. DS's daughter also recognized signs of improvement at home and therefore withdrew her mother's name from the nursing home waiting list.

A consultation with a geropsychiatrist resulted in a diagnosis of depression and ETOH abuse, and ruled out AD. DS's MMSE score was 26-low normal-she remained in the ADC program and continued to demonstrate improvement. DS especially appeared to benefit from and enjoy assisting other clients. Although she no longer displayed the symptoms appropriate to be a client, she did not want to leave the program. At a meeting with ADC staff and her daughter, DS agreed to become a volunteer in the program. She flourished in this role and eventually was hired as a part time ADC aide, a position she held for seven years until her death due to respiratory failure. Over the course of that time she had remained cognitively healthy.

Outcome

It is clear that depression, isolation, nutrition, and ETOH use were the causative factors for the presenting symptoms. Apparent reliance on a single element in the diagnostic process (MMSE) was the cause for the initial misdiagnosis. As these causative issues were addressed and corrected, the symptoms vanished. Subsequently, DS never entered the nursing home, she was able to be productive and feel good about her contribution to others, and was able to remain with her daughter, adding to the quality of her life and the lives of those she touched. DS's case stresses the importance of conducting a comprehensive diagnosis process and screening for depression and ETOH.

primary caregivers to maintain normal social outlets. They frequently neglect their own health needs and the disease often eats away at the family finances. It is estimated that 35 percent of primary caregivers in the United States work outside the home and therefore pay for professional care while working. The typical US caregiver is a female in her mid-50s, usually a daughter or wife caring for a parent or spouse.

Early and accurate diagnosis allows time for the patient and

his or her family to learn about the disease and begin planning for the future while the patient is still capable of participating in the decision-making process. It also provides an opportunity for long-range planning and the making of legal, financial, and medical arrangements. Early and ongoing support and education—such as that provided by organizations like the Alzheimer's Association—can empower caregivers to face current and future challenges. Because AD is a diagnosis of exclusion (see below), it is often difficult to determine whether a patient is suffering from it or from other disorders that manifest themselves with similar symptoms. To illustrate the difficulty of this process, two case studies are provided. It is suggested that you read these now as they will be referred to throughout the rest of the article.

The Early and Middle Stages of Dementia

The Alzheimer's Association estimates that symptoms of dementia may be present three to five years before a diagnosis occurs. The reasons for this are simple. Typically a patient with AD does not recognize changes that are taking place and automatically compensates through the use of long-term social skills. Family members tend to notice gradual rather than abrupt changes, and likewise are inclined to rationalize or deny their persistence. Note when a caregiver reports abrupt cognitive changes, this might indicate a delirium, vascular, or another type of dementia rather than AD. Early symptoms of AD may include:

- more-than-normal forgetfulness;
- periodic disorientation;
- word-finding difficulty (aphasia);
- difficulty in completing instrumental activities of daily living, such as writing checks or looking up telephone numbers (apraxia); and
- mild difficulties in abstract processes such as mathematical calculations.

As the dementia progresses into the middle stage of the disease, the severity of symptoms becomes more pronounced and may include:

- increased and persistent memory loss;
- greater difficulty finding words or the inappropriate use of words;
- an inability to perform ADLs, such as dressing, grooming, bathing;

Case Study B

Patient History

WK was a 76-year-old married male who, along with his wife, owned and operated an active and successful business in the community for more than forty years. WK took care of all the finances for the business, while his wife and daughter attended to the day-to-day operations. WK's wife noticed that her husband was taking much longer to complete the end-of-day accounts. She also discovered that some bills had not been paid, which had never happened before (apraxia). She also had found that WK had been placing soiled garments back into drawers with clean clothes (executive functioning). When confronted, WK denied any problems and felt that someone else must had placed the soiled clothes in the wrong place. WK's wife reported her concerns to WK's primary care physician.

Evaluation

The primary care physician completed a routine physical examination along with blood chemistries and an ECG. These results were all within normal limits for someone WK's age, and the physician found him to be in excellent physical health. There was no evidence of hypertension, cardiovascular disease, diabetes, pulmonary diseases, or other major disease processes. Although the primary care physician did not do an MMSE, he did interview WK and asked him questions about recent events, the current date, and to do some simple calculations. WK did not do as well as the physician thought was appropriate given WK's education and experience. During his interview with WK, the primary care physician found some discrepancies between the report from the wife and WK. He found that WK's explanations did not make sense (aphasia). The physician was concerned about the mild cognitive changes reported by the wife and those in evidence during the interview so he referred WK to Crozer's Geriatric Assessment Program for a differential diagnosis of the apparent cognitive changes.

During the assessment process WK was evaluated by a social worker, geriatrician, and geropsychiatrist, who also interviewed his wife. It was determined that WK rarely used alcohol, took only prescribed medications, and that he currently was not on any medication. The geriatrician reviewed the report and laboratory work forwarded by the primary care physician, completed a physical examination, and ordered a CT scan of the brain (without contrast). All findings were within normal limits. The CT scan showed no evidence of subdural hematoma, tumors, or ischemic disease. There was mild cerebral atrophy, but again within normal limits. The geropsychiatrist evaluated for current psychiatric symptoms and depression, which resulted in negative findings. An MMSE was conducted with WK scoring 20 (mild dementia). The MMSE demonstrated that WK had deficits in memory loss (registration and recall) and executive functioning (calculation). Reports from WK's wife indicated a number of activities WK could no longer complete or did poorly (apraxia). During the interview WK was found to have fabricated some information when he stated he had served in the Army (confabulation). Having evaluated WK and, finding no evidence to suggest another cause for the cognitive changes, the team rendered a diagnosis of dementia, probable Alzheimer's type.

Treatment

The team recommended a re-evaluation in one year and provided educational material

confusion and/or agitation when others try to help.

Normally it is during the transition from the early to the middle stage of the illness that families present their concerns to a primary care physician.

Diagnosing Dementia

Due to the AD patient's normal tendency to deny or mask his or her losses, the primary care physician's first indication of cognitive changes may come from reports from the caregiver, as can be seen in Case B. However physicians should be advised to periodically test for cognitive changes as part of their routine examinations of elderly patients. There are many short and easy-to-use mental state exams; one such exam is the Flostein Mini-Mental State Examination (MMSE).² An example of this exam is provided in Figure 1, page 32. The MMSE measures orientation, registration, attention, calculation, recall, and lan-

to WK's wife, scheduled her for a "New Caregiver's Orientation Workshop" hosted by the Crozer's Adult Day Center and the local Alzheimer's Association, and suggested she attend the monthly caregiver support group. They also advised her how she could keep her husband active in the business while avoiding the mistakes in financing. For example, they had WK review the accounts and books with the adult son on a weekly basis to "teach his son the business." In reality the son was overseeing the accuracy and making necessary corrections. (Note: At the time of WK's diagnosis there were no drugs approved in United States for the use in cases of AD; currently there are three: Cognex, Aricept, and Excelon. Had these drugs been available, WK would have been an appropriate candidate for their use.)

At the follow-up appointment a year later it was determined that WK's physical health had not changed but there was increased cognitive decline. WK's MMSE was now 15 and he wandered away from work. The assessment team concurred with the previous diagnosis of AD and recommended that WK attend the Adult Day Center to provide the supervision and structured activities that would reinforce remaining strengths and provide his wife the reassurance that he would be safe during the day while she was at work.

Outcome

WK attended Crozer's Adult Day Center five days a week for five years until his death. He participated in exercise groups, cognitive stimulation activities, arts and crafts, and other group activities. His wife continued to attend support groups and educational offerings. WK's MMSE declined to 0 a year before his death. He required others to feed him and he had become incontinent (loss of basic ADLs). During the last two years of his life, WK was no longer able to join the family on vacations or business trips. WK's wife put her husband in 24-hour respite services (shortterm stays in dementia-specific units within local nursing homes) so she could continue to spend time with her family and go on business trips. In his last year at the Center, WK most enjoyed walking in the courtyard looking at the trees and flowers. He especially enjoyed the time in the afternoon when his wife would come to take him home, but before doing so, they would walk around the courtyard arm in arm.

guage. However, as can be seen in Case A, reliance on a mental state exam alone may lead to a misdiagnosis. Instead, tests such as the MMSE provide the physician with evidence that further investigation is warranted.

Once the physician identifies evidence that warrants an assessment for dementia, thorough and comprehensive guidelines, such as the NINCDS-ADRDA,³ criteria should be followed (see Fig. 2, page 33). In the United States, the primary care physician may complete this process alone or in coordination with a geriatric assessment team, or with specialists such as neurologists, geropsychiatrists, or neuropsychologists. Physicians need to be aware that there are numerous problems, such as depression, that can cause dementia-like symptoms. In some instances, once identified, these problems can be treated and the symptoms may be reversed. DS's situation (see Case Study A) indicates that had her physician done a complete medical and social history, symptoms such as significant weight loss, daily use of sleep med-

ication, daily alcohol (ETOH) use, and feelings of loneliness and depression would have potentially raised a red flag that her condition was caused, at least in part, by depression. As we can see, subsequent identification and treatment reversed the presenting cognitive symptoms. In WK's case, the assessment team evaluated for and ruled out depression, ETOH use, and other potential medical causes for the presenting cognitive symptoms. Subsequently, there was greater reliability on the resultant diagnosis of AD.

For other illnesses, such as a stroke, identification and treatment can halt or slow the progression of symptoms. To date, there is no definitive test to determine AD and we are dependent on an autopsy to confirm a diagnosis. For these reasons the diagnosis of AD is a diagnosis of exclusion.

The diagnostic process should be viewed as a jigsaw puzzle with the answers to the different components being pieces of that puzzle. As the case studies illustrate, the more pieces of the puzzle that come together, the clearer or more accurate the diagnostic picture becomes.



Medical History

As part of the diagnosis process, a thorough medical history should be completed with the patient and then confirmed with a reliable informant, such as the primary caregiver. Remember that the patient may have gaps in his or her memory or may fabricate (confabulation) imaginary experiences to fill in gaps. Using the caregiver as a resource will prove valuable in gaining insight into the patient's problem. The medical history screening should include a review of familial history for dementia, stroke, vascular disease, thyroid conditions, and depression.

Additionally, a review of ALL medications—including over-the-counter drugs—is critical. The case of one woman emphasizes the importance of a thorough medication review. This woman was brought to the Crozer's Geriatric Assessment

Mini-Mental State Examination (MMSE) Max. Score Score Orientation 5 What is the (year) (season) (day of the week) (month) (date)? 5 Where are we: (state) (county) (town) (hospital) (floor)? Registration 3 Name 3 unrelated objects, allow 1 second to say each. Then ask the patient to repeat all 3. Give 1 point for each correct answer. Repeat them until he learns all 3. Count trials and record. Trials: Attention and Calculation Ask patient to count backwards from 100 by sevens. 1 point for 5 each correct answer. Stop after 5 answers. Recall Ask patient to recall the 3 objects previously stated. Give 1 point 3 for each correct.answer. Language · Show patient a wristwatch; ask patient what it is. Repeat for 9 a pencil (2 points). • Ask patient to repeat the following: "No ifs, ands, or buts" (1 point). · Ask patient to "take a piece of paper in your right hand, fold it in half, and put it on the floor" (3 points). · Ask patient to read and obey the following sentence which you have written on a piece of paper. "Close your eyes" (1 point). • Ask patient to write a sentence (1 point). Ask patient to copy a design (1 point) Total Score Suggested guideline for determining the severity of cognitive impairment: Mild: MMSE \geq 21 Moderate: MMSE 10-20 Severe: MMSE \leq 9 Expected decline in MMSE scores in untreated mild to moderate Alzheimer's patient is 2 to 4 points per year.*

Figure 1. Example of an MMSE

Program by her husband who wanted a second opinion about an Alzheimer's diagnosis. During the geriatrician's review of medications, it was discovered that the woman was using a certain type of eye drop for glaucoma that was known to have potential cognitive side effects. The geriatrician consulted with the woman's ophthalmologist who agreed to try another eye drop formulation. Within weeks the woman's cognition—as measured by the MMSE—improved to normal levels.

Along with prescribed medications, it is also important to determine a patient's ETOH (alcohol) use, taking into consideration all sources of ETOH use, including nighttime sleep remedies, cough and cold preparations, and alcoholic beverages. ETOH use/abuse will exacerbate a co-morbid dementia or could be a factor in some forms of dementia, such as Korsakoff's Dementia.

> Finally, because depression has been identified as a principal cause for the misdiagnosis of AD (Case A), it is imperative to screen a patient for it. It is widely believed that patients with dementia suffer disproportionately from depression, and depression superimposed on dementia exacerbates the symptoms of dementia. There are many simple tools that the primary care physician may use to screen for depression (see "Depression as a General Medical Problem," page 19). The primary care physician may also want to enlist the support of a geropsychiatrist to screen for depression and possible other behavioral manifestations. If discovered, depression should be aggressively treated. Successful treatment may reverse the presenting symptoms or reduce the depressive component to give a more accurate picture of the dementia.

Treatment Program

Once the diagnosis of AD or other dementia is made, the primary care physician and other healthcare providers need to identify the primary caregiver (spouse, daughter, etc.) who must become a principal ally in the treatment to ensure the best outcome for the patient. Open communication between the primary care physician and the primary caregiver is critical if the physician wants to track the course of the illness and the effectiveness of treatment. Over time, the demented patient will develop other illnesses and diseases, and these should be aggressively treated so as not to exacerbate the dementia symptoms. The physician should be knowledgeable about current approved medications for the treatment of dementia as well as those treatments that are in experimental stages of development. (*Note:* Current approved medications have demonstrated the potential to slow the progression of dementia symptoms. To date no medication has been proven to halt and/or reverse the dementia itself.)

Patients with dementia benefit from daily structured activities that focus on reinforcing remaining strengths while de-emphasizing their losses. Sustaining simple routines will help the demented patient maintain skills like ADLs for as long as possible. Finally, the primary care physician should be aware that even experts in dementia are still learning about the disease and potential treatments. The primary care physician should not be concerned if he or she feels overwhelmed by the process and the enormity of need presented by the patient and the family. The primary care physician should form alliances with other professionals to develop a team approach to the care of their demented patients.

The primary caregiver will be responsible for carrying out the treatments as prescribed and will take the brunt of the frustration and anxiety of the family as they witness the steady decline in their loved one. For this reason it is imperative that the caregiver be educated as to the disease process and the course it will take. A greater understanding of the changes that have taken and will take place prepares the caregiver for the inevitable challenges to

be faced. Caregiver support groups can be very valuable as they not only provide sound information about coping with a demented loved one, but also provide reassurances that the caregiver is not alone. The primary care physician can be a strong advocate to assure that the healthcare system addresses these needs either directly or in conjunction with Alzheimer's associations and societies around the world.

Primary care physicians should also be aware that primary caregivers often neglect their own healthcare needs and thus should advise caregivers to avail themselves of preventive care such as flu shots and mammograms, and to seek regular medical care for themselves. The primary care physician should also advise the caregiver to obtain help from other family members and friends and to use available resources such as day care programs that provide respite for the caregiver. Although caring for a loved one who is suffering from dementia

Diagnostic Process

Here are some topics to consider when assessing a patient for AD. This is meant to be a sampling of the types of information and topics that should be covered rather than an exhaustive list.

Social History

This should include questions designed to determine:

- the patient's occupation and any exposure to head trauma or toxins;
- whether the patient engages/engaged in sports, such as boxing, football, gymnastics, that might cause head trauma; and
- current social and family support.

Medical History and Physical Examination This should include:

- a family history, especially of parents and grandparents and whether they suffered from AD, stroke, vascular disease, thyroid disorders, diabetes, etc.;
- the mode of onset for the patient of cognitive changes; whether the onset was abrupt or gradual;
- a thorough medication review, including all over-the-counter drugs, ETOH, and other substance use/abuse;
- results from any laboratory or radiologic tests, including CBC, B12, serology, folate, and HIV tests; ECGs (to rule out Creutzfeldt-Jakob's Disease); and CTs and MRIs (to check normal pressure hydrocephalus)
- a neurologic evaluation;
- a geropsychiatric evaluation that looks for depression;
- an MMSE and/or other cognitive screening tools;
- possible neuropsychological battery of tests (especially in complex presentations); and
- tests or evaluation as deemed appropriate.

Figure 2: Physicians should extensively evaluate a patient's medical and social history

is difficult at best, it is important to note that early diagnosis, an increasing variety of treatment options, and counseling and support groups can all help make the situation more bearable for all those concerned.

References

- American Psychiatric Association: Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition, Washington, DC, American Psychiatric Association (1994).
- M.F. Folstein *et al.*, "Mini-Mental State: A practical method for grading the cognitive state of patients for the clinician," J. of Psychiatric Research 12 (3), 189-98 (1975).
- M.C. Tierney *et al.*, "The NINCDS-ADRDA Work Group criteria for the clinical diagnosis of probable Alzheimer's disease: A clinico-pathological study of 57 cases," Neurology **38**, 359-364 (1998).

Joe Maier, MS, is a gerontologist and director of Crozer's Adult Day Centers (dementia-specific day care programs) at Crozer-Chester Medical Center in Pennsylvania.