

# 2

## THE EVALUATION AND FORMULATION OF DEMENTIA

This chapter will discuss the evaluation of a patient with memory complaints or suspected dementia. The first part of the chapter addresses the following questions: (1) When should a comprehensive dementia evaluation be performed? (2) What is the purpose of such an evaluation? (3) Who is involved in performing such an evaluation? (4) What is the process of this evaluation?

The second part of the chapter discusses how to perform an assessment of a person with suspected dementia. Chapter 5 discusses how to use the information from the assessment in the differential diagnosis and workup of dementia.

### BACKGROUND

#### **When should a comprehensive dementia evaluation be performed?**

In most cases, the recognition that an evaluation is needed does not come from the patient. Typically, a family member, such as a spouse or a child, notices forgetfulness, communication difficulty, problems in functioning, or a personality change and persuades the patient to be evaluated. Primary care physicians, neurologists, psychiatrists, geriatricians, and specialists in dementia are often the first professionals to see patients with such complaints.

This reliance on family members and patients to recognize dementia

often leads to delays in diagnosis. Of the patients seen for an initial evaluation at the Johns Hopkins Comprehensive Alzheimer Program, only 15%–20% are in the early stages of the disease and one-third are in the late stages of dementia. More often than not, evaluations are sought when crises occur such as dangerous behavior, forgetting to pay bills, having a car accident, withdrawing from social activities, or stopping activities such as cooking and yard work.

There are several benefits of early diagnosis. First, the functional decline due to dementia and its consequences can be better managed if anticipated and addressed early. For example, financial catastrophes and injuries from car accidents or falls can be prevented by the knowledge that the patient is impaired. Second, early identification helps the family and patient understand changes in behavior and judgment that are often early symptoms. Behavioral disorders such as depression, delusions, and aggression are more likely to respond to treatment if caught early and treated appropriately. Third, early diagnosis allows patients and families more time for long-range planning to manage the consequences of dementia. This includes the ability to do estate planning, appoint power of attorney (Chapter 13), and so forth. Fourth, early diagnosis may improve the response to treatment for the cognitive symptoms and delay progression in some diseases.

Despite this, we do not believe that widespread screening of asymptomatic individuals can be justified at present. In the future, when more effective therapies are available and preventive treatments are developed, screening evaluations of at-risk individuals will be warranted.

To improve recognition and early diagnosis of dementia, we recommend that an evaluation be considered for elderly persons and other persons with neurologic disease or head injury who develop any of the signs or symptoms listed in Table 2.1.

Of the problems listed in this table, memory impairment and impaired functioning are most likely to be ascribed to normal aging and to be explained away or ignored. Since there are slight declines in cognition and functioning associated with aging, awareness of the usual changes associated with aging is necessary. For example, difficulty remembering names or coming up with the right word without any of the other symptoms in Table 2.1 is unlikely to be due to dementia. One piece of information that is especially useful in the primary care setting is a standardized cognitive assessment done during routine medical checkups. Tests such as the Mini-Mental State Examination (MMSE) can be administered annually or biannually in less than 10 minutes by a physician or an allied health professional. A decline of more than 3 points on the MMSE from a stable baseline should trigger an evaluation.

TABLE 2.1. Signs and Symptoms That Should Trigger Consideration of a Dementia Evaluation

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<p>1. <i>Cognitive changes</i></p> <ul style="list-style-type: none"> <li>Worsening new forgetfulness</li> <li>Excessive repetition of questions and statements</li> <li>Trouble understanding spoken and written communication</li> <li>Difficulty finding words</li> <li>Not knowing previously known information</li> <li>Disorientation as to time, place, or person</li> </ul>	<p>3. <i>Personality change</i></p> <ul style="list-style-type: none"> <li>Inappropriate friendliness</li> <li>Blunting and disinterest</li> <li>Social withdrawal</li> <li>Excessive flirtatiousness</li> <li>Easy frustration</li> <li>Explosive spells</li> </ul>
<p>2. <i>Psychiatric symptoms</i></p> <ul style="list-style-type: none"> <li>Withdrawal or apathy</li> <li>Depression</li> <li>Suspiciousness</li> <li>Anxiety</li> <li>Insomnia</li> <li>Fearfulness</li> <li>Paranoia</li> <li>Abnormal beliefs</li> <li>Hallucinations</li> </ul>	<p>4. <i>Problem behaviors</i></p> <ul style="list-style-type: none"> <li>Wandering</li> <li>Agitation</li> <li>Noisiness</li> <li>Restlessness</li> <li>Being out of bed at night</li> </ul>
	<p>5. <i>Changes in day-to-day functioning</i></p> <ul style="list-style-type: none"> <li>Difficulty driving</li> <li>Getting lost</li> <li>Forgetting recipes in cooking</li> <li>Neglecting self-care</li> <li>Neglecting household chores</li> <li>Difficulty handling money</li> <li>Making mistakes at work</li> <li>Trouble with shopping</li> </ul>

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## What are the purposes of a dementia evaluation?

The primary purpose of the dementia evaluation is to determine whether dementia is present or absent. Dementia is a clinical diagnosis that depends on the demonstration of multiple declines in cognitive capacity and clear consciousness. The evaluation may demonstrate that dementia is not present and that the complaints or concerns that initiated the evaluation can be attributed to some other cause, such as usual aging, depression, a previously unrecognized neurologic or medical condition such as Parkinson disease or hypothyroidism, or an offending factor in the environment such as alcohol or medication.

Another purpose of the dementia evaluation is to ascertain the cause of the dementia syndrome. This is a necessary step in determining the most appropriate treatment and the prognosis, that is, the likely course over time. An important aspect of assessment is the identification of both disabilities that result from the dementia and remaining abilities, as both should be addressed in the treatment plan. Finally, the evaluation lays the groundwork

for developing a family support plan. It determines the kinds of information, guidance, and emotional support that the patient and family require to deal with a chronic and usually progressive illness.

### **Who is involved in performing a dementia evaluation?**

Most dementia assessments can be accomplished in the community in primary care settings. Specialists are best used when the diagnosis is in question, the case is atypical, the symptoms are complex, or initial management strategies here failed. Specific examples of when specialist input should be sought include the following: the diagnosis of dementia is uncertain, the patient is young (<65), the dementia is rapidly progressive, motor symptoms are prominent, behavioral disorder is pronounced, the dementia is potentially reversible, or the care needs are beyond those usually required.

The assessment of dementia uses medical skills that are within the capabilities of all physicians. Some elements of the evaluation, such as history taking, simple cognitive testing, and psychosocial assessment, can be performed by allied professionals (nurses, psychologists, or social workers) who are specially trained.

In many settings, an interdisciplinary team can carry out such an evaluation. In this model, an allied health professional takes the history from the family and caregiver and performs a mental status exam; a neuropsychologist performs the neuropsychological assessment; and a physician performs a physical examination and a comprehensive mental status exam and reviews the case with the other professionals. We believe such a model can be applied to any practice setting with the appropriate training and experience.

The physician who does not have such a team available should take the history, perform a physical examination and mental status examination of cognitive and noncognitive realms, and order appropriate laboratory studies. Indications for laboratory studies and referrals to a neuropsychologist and other professionals are discussed later in this chapter.

### **What processes are involved in a dementia assessment?**

A comprehensive assessment is typically done in stages (Table 2.2). The first stage involves the patient and one or several informants, requires 1–2 hours, and consists of a complete neuropsychiatric assessment (discussed below). The second stage consists of a family evaluation and is done only if the initial assessment confirms the diagnosis of dementia. It can be performed by a social worker or nurse and requires approximately 1 hour to complete. This, too, will be discussed below. A series of diagnostic tests described later in

TABLE 2.2. Stages of a Comprehensive Dementia Evaluation

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<i>Stage 1:</i> Neuropsychiatric assessment (directed by any trained physician)
<i>Stage 2:</i> Family assessment
<i>Stage 3:</i> Diagnostic tests
<i>Stage 4:</i> Conference discussion, diagnosis and recommendations to patient, family, and others as appropriate

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this chapter should be obtained. These include laboratory studies, brain imaging studies, and neuropsychological tests. They are almost always done on an outpatient basis, but an inpatient assessment may be necessary if severe medical or behavioral problems are present. Finally, the whole picture is pulled together at a Diagnostic and Recommendation Conference where the interdisciplinary team meets with the patient and care providers to review the history and the results of the assessment, to explain the diagnosis, and to develop a treatment plan.

Each of these stages, including the initial assessment, has several purposes. The involvement of family members and/or other informants is crucial at several points, but the patient and family should be evaluated separately to diminish patient embarrassment and allow family members to answer freely questions about the patient's history and current symptoms. Performing the entire assessment with the patient and family together can be awkward and uncomfortable since the patient is being talked about as if he or she were not there. On rare occasions, patients will refuse to be seen alone. We sometimes call the family by telephone at another time to collect information and address concerns.

We typically start an assessment by meeting briefly with the patient and all family members who are present. We begin by stating that we will first meet briefly with everyone, then talk with the patient and family separately, and conclude by meeting together to discuss the findings. Before separating the patient and family, we ask whether there are particular issues that should be discussed with everyone present. We specifically ask whether there are questions that the patient and family want addressed by the end of the evaluation. Occasionally, the family will begin to give a full history at this point. If this happens, we ask them to wait until later. The purpose of this brief joint meeting is to raise general issues that can be explored separately with both care provider and patient. Sometimes patients will say that they do not know why they are coming for an evaluation and do not want to be there. In this case, they can often be reassured that the evaluation will be relatively brief and that they will be returning home in an hour or two.

## THE EVALUATION

### The history

The cornerstone of a comprehensive dementia evaluation is the neuropsychiatric assessment, which is outlined in Table 2.3. The goal of this assessment is to obtain information that will enable the clinician to determine an initial impression, develop a differential diagnosis, and plan treatment. Selected sections of this evaluation are highlighted here.

#### *Family history*

A detailed family history of the grandparents, parents, siblings, and children is taken. This is best recorded as a pedigree on a genogram. An example is provided in Figure 2.1. A genogram helps the clinician and family ask questions about each relative and increases accuracy. This helps the clinician focus on the family's health history, which will aid in the differential diagnosis, and on the current status of the family, which will identify potential and actual human resources available for the patient's care.

#### *Personal history*

The patient's personal background provides the data by which the whole case can be understood. Specifically, information should be obtained about early life cognitive and behavioral difficulties, educational achievement, work history, marital history, quality of relationships among family members, and religious background. In addition to allowing an estimation of premorbid functioning and cognitive reserve, the personal history illustrates the patient's life in a way that allows a clearer understanding of this individual and his or her response to illness. It also identifies interests and wishes, which in turn guide treatment planning. A sexual history should be obtained to assess for possible exposure to human immunodeficiency virus (HIV) risk factors.

#### *Substance abuse history*

It is important to be complete here because several dependency-producing substances, including alcohol and benzodiazepines, impair cognition even at low doses.

#### *Medical history and review of systems*

This is essential. All patients should be asked about hypertension, diabetes, heart disease, cancer, lung disease, surgery, and blood transfusions. Careful attention should be given to determining all medications taken in the prior

TABLE 2.3. Outline of a Neuropsychiatric Assessment

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1. *Identifying Data*: age, marital status, race, sex, referral source
  2. *Chief Complaint*: including the reason for referral and questions to be answered
  3. *Family History*: vital status of parents, grandparents, siblings, and children; if deceased, age at death and cause; any members with psychiatric or neurologic illness; pedigree
  4. *Personal and Social History*: where born, summary of early life experience, education, work history, marital state, living situation, leisure practices, religious faith, typical daily activities
  5. *Substance Abuse*: use of cigarettes, alcohol, prescription and over-the-counter medications; history of abuse or dependency on any of these substances
  6. *Medical History*: medical and surgical problems, active problems and their severity, review of systems, current medications, physicians and other health care providers involved in providing medical care
  7. *Premorbid Personality*: traits, predispositions, affect, activity, reactivity
  8. *Neuropsychiatric History*: psychiatric symptoms or disorders, psychiatric assessments or treatments, seizures, head trauma, stroke, other neurologic disorders
  9. *History of Present Illness*: onset date, course, features, rapidity, and pattern of change; systematic review of systems to include information on cognitive capacity, mental syndromes, unusual experiences, functional status, and behaviors
  10. *Current Psychosocial Environment*: living environment at present, care providers, financial issues, legal issues, use of community resources
  11. *Examinations*: physical, neurologic, cognitive, and mental status exams
  12. *Laboratory Evaluations*: brain imaging, laboratory studies, and other tests
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6 months. Over-the-counter drugs such as aspirin, vitamins, nutraceuticals, and sleeping potions are important to note. Possible exposure, including occupational exposure, to toxins such as heavy metals (lead), organic solvents, and other chemicals should be determined.

### *Premorbid personality*

This will provide a good picture of the patient's predispositions, character, temperament, and interests. It is important in understanding symptoms and in planning treatment and supportive care.

### *Past neuropsychiatric history*

Special attention is paid to a history suggestive of brain injury including trauma, transient ischemic attacks, stroke, paralysis, sensory loss, speech or

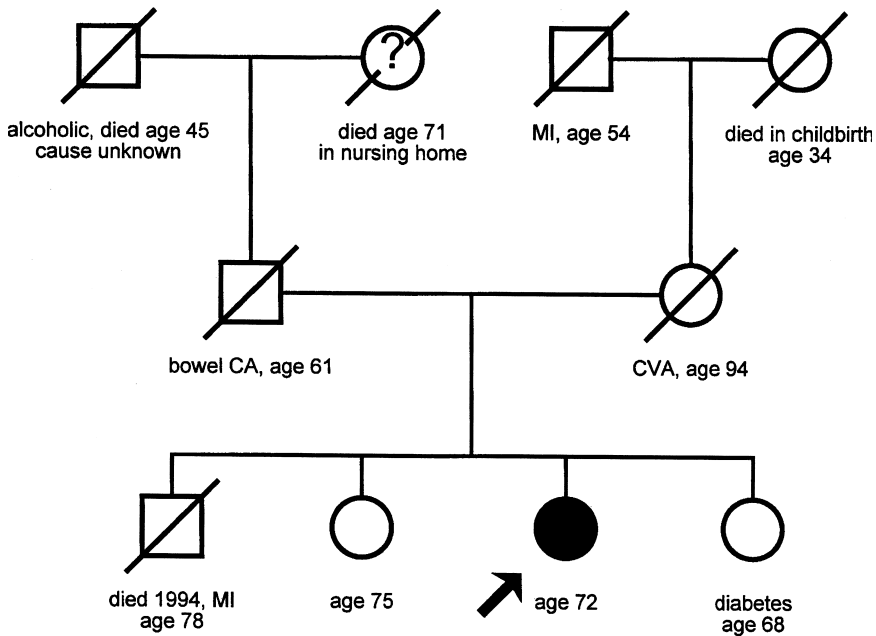


FIGURE 2.1 Example of a pedigree drawing. CA, cancer; CVA, cerebrovascular accident; MI, myocardial infarction. Box, male; circle, female; arrow, patient (proband).

language impairment, and tremor. A full psychiatric history is also taken, including a history of mental symptoms such as depression, prior evaluation, prior treatment, or hospitalization.

*History of present illness*

The examiner should identify the earliest symptoms and provide a chronological history up to the present using a detailed symptom checklist (Table 2.4). Family members and other informants play a major role in obtaining this part of the history. They should be asked specific questions about the functions and impairment listed in Table 2.4. In addition to helping with the differential diagnosis, this symptom checklist aids in the identification of target symptoms requiring treatment.

**The mental status examination**

Since the primary symptoms of dementia are impairments in cognition, behavior, and function, a thorough mental status examination is a necessary part of the evaluation. Some clinicians believe that the mental status examination is intrusive, while others are concerned that it is insulting or too

TABLE 2.4. Symptom Checklist in the Evaluation of Dementia

<i>Impaired Cognition</i>		<i>Problem Behaviors</i>	
Memory	Concentration	Verbal abuse	Catastrophic reactions
Language	Planning and organization	Uncooperative	Noisy
Orientation	Personality change	Physically aggressive	Wandering
Writing	Executing	Sundowning	Hoarding/rummaging
Reading	Loss of social rules	Demands interaction	Intrusive
Calculating		Outbursts	
Recognizing			
Attention			
		<i>Disturbances in Drives</i>	
<i>Abnormal Mental Phenomena</i>		Poor appetite	Sleeps a lot
Depression	Death, suicidal	Weight loss	Out of bed at night
Self-deprecating	Disinterested	Poor Sleep	Sexual aggression
Somatic complaint	Anhedonic	Hypersexual	Hyposexual
Crying spells	low energy level	Excessive appetite	
Diurnal variation		<i>Impaired Function</i>	
Socially withdrawn	Apathetic	Cooking	Mobility impairments or falls
Anxiety	Panicky	Finances	Bathing/grooming
Irritability	Labile	Housekeeping	Feeding
Euphoria	Rapid speech	Driving	Continence
Delusions	Hallucinations	Shopping	
Illusions	Acute confusion	Dressing	
Fatigue		Hearing/sight	

medical. Similar reluctance was reported years ago about taking a sexual history. Refusal is rare if the examiner believes it is an important part of the examination and so informs the patient. It is helpful to assure the patient that this is a routine part of the assessment for every person. In our experience, those who resist are almost always impaired. When individuals refuse to answer questions by saying “That’s a silly thing to ask” or “Of course I can do that,” it is best to turn temporarily to another area of questioning such as the medical history and to ask the questions again later.

It is not uncommon for cognitively impaired patients to be reluctant to answer direct questions. Individuals who will not or cannot answer direct questions about cognition are more likely to answer questions asked in the course of a general conversation. For example, orientation to year can be determined during the life history review. “Where were you born? What year was that? Do you know what year it is now? How old does that make you?”

Resistance to the mental status examination can sometimes be overcome by the examiner’s emphasizing that the assessment is being carried out to

identify remaining abilities as well as impairments (“Let’s see how well you do with this one”) and by acknowledging that some questions are difficult (“I’m going to ask you a more difficult one now. Let’s see how good you are at math. Take 7 away from 100”). Sometimes it is useful to say that the information is being gathered for the benefit of the patient (“I know this is hard, but if I know what you have problems with, I’ll be better able to help you”). There is a fine line between being supportive and being condescending, but helping a person over a difficult question is often reassuring. For example, if a person answers the question “Do you know where we are now?” with “I can’t remember” and is upset, the examiner can be supportive by responding, “Well, let me help you. Do you know what city we are in?” Even in the best of hands, however, some individuals (perhaps 1%–2%) are unwilling to undergo this part of the assessment.

The mental status examination we recommend has seven major headings. An outline is presented in Table 2.5. It is not necessary to ask all the questions together. For example, some aspects of the mental status examination can be assessed during the general patient interview. Examples include appearance and behavior, talk, and mood. However, focused questions pertaining to these and other areas are usually necessary, and we believe it is useful for the examiner to have a specific outline in mind. This allows the clinician to check at the end of the evaluation and determine if all appropriate questions have been asked. It is also helpful to record the information obtained in a specific order because this allows the clinician to check for completeness and, more importantly, put the information together in a meaningful fashion. In addition, having the data in a specific order enables the clinician to present them to others in a comprehensive fashion.

## Elements of the mental status and cognitive examinations

### *Appearance and behavior*

First and foremost, practitioners are nonjudgmental observers. Training and experience guide what the clinician considers relevant. Issues of note include whether patients recognize the clinician as a professional, whether they act in a manner consonant with their background, and whether they are neat in appearance. In general, how patients approach the examination reflects how they react to situations that are unusual or stressful.

The predominant demeanor of patients during the assessment should be noted. Are they relaxed and calm, tense, or distressed? Does the examiner have to work hard to put them at ease? Are they restless, slow-moving, fidgety, or tremulous? Are there frequent, easily induced changes in mood or behavior? None of these actions is necessarily abnormal, but each can be relevant for diagnosis and treatment.

TABLE 2.5. Parts of the Mental Status Examination

1. <i>Appearance/Behavior</i>	5. <i>Content of Thought</i>
2. <i>Talk</i>	Delusions
Rate	Obsessions
Rhythm	Compulsions
Fluidity	Phobias
Spontaneity	6. <i>Insight and Judgment</i>
Latency	7. <i>Cognition</i>
Thought disorder	Consciousness
3. <i>Mood and Affect</i>	Orientation
Observed and reported stability, reactivity, and appropriateness	Memory
Vital sense	Praxis
Self-attitude	Language
Thoughts of death, suicide, homicide	Abstraction
4. <i>Perception</i>	Gnosis
Hallucinations	Knowledge
Illusions	Attention
	Calculation
	Executive function

Observing how patients are dressed is important. Are their clothes neat or do they seem disheveled, mismatched, misbuttoned, or dirty? Are they appropriate for the weather? Individuals who are reported to have always been neat and who come in with messy or stained clothes are probably having significant trouble not only with dressing but also with other complex activities such as preparing meals and keeping house. These clues should raise concern about safety.

The description of behavior should also state whether patients are able to walk into the room unaided, require a wheelchair, use a cane, or are lying in bed. Do they have the stooped posture and flexed body habitus of a person with parkinsonism? Is there obvious weakness or does the person seem to neglect one side of the body? Do they have a tremor or rapid, jerking movements? Do they appear fearful—for example, glancing around the room as if hearing someone talking? Do they seem uninterested, sad, or resistant? Do they become more or less cooperative as the assessment evolves?

### *Talk*

Several aspects of the patients' talk (speech) should be assessed. First, speech should flow naturally (*fluency*) and spontaneously, that is, its speed should follow the examiner's. Speech should demonstrate an appropriate rate, rhythm, and prosody (smoothness). The pragmatics of speech (appropriate use of facial expression and gestures in conversation) should be assessed.

Hesitant speech, in which word-finding difficulty is prominent, speech is telegraphic or without the usual connecting words, and talking is frustrating to the patient suggests a nonfluent (Broca) aphasia. Speaking a great deal but saying little that makes sense indicates a fluent (Wernicke) aphasia. Substituting one word for another—for example, calling a watch a tie, saying words that are combinations of other words (time teller for watch), or saying words that are not in the lexicon (walm for watch)—all called *paraphasic errors*—also indicate a language disorder. Occasional difficulty finding words in the course of a conversation can be normal but repeated instances are not. Therefore, it is important to notice that such difficulties are occurring and to keep track of their frequency.

Second, the patient should be able to *comprehend* questions and *follow instructions*. An inability to do so in response to normally spoken statement or question might be due to a hearing deficit but often indicates difficulty in comprehending spoken language (receptive aphasia). Problems in this realm should be suspected if patients ask for questions and instructions to be repeated, or if they seem to understand simple and straightforward questions but have difficulty with more complicated ones, even if the examiner asks them with the same loudness of voice.

If words are *slurred* or incomprehensible, a disorder of the motor or production aspects of speech is likely. This points to damage of the neurologic or oropharyngeal control mechanisms responsible for producing speech. In contrast, word-finding problems are indicative of an impairment of language processing.

The ability to control speech can be impaired. Hesitancy before answering questions and speaking very softly, slowly, and deliberately can indicate depression. Rapid speech that dominates the conversation and prevents the examiner from interrupting (push of speech) suggests hypomania or mania. If the content of speech does not flow logically, there might be evidence of tangentiality (answering questions well off the mark), circumstantiality (giving excessive detail), or derailment (having no clear *string* or line of thought). *Pallilalic* speech, consisting of repetitive sounds (e.g., *la la la*) or words (e.g., *go go go*), or frequent perseveration (repetition) of words or phrases indicates a severe language disorder.

### *Mood and affect*

Clinicians sometimes distinguish between the terms *mood* and *affect*, although there is not universal agreement on their definition. It is best to describe both how patients feel in their own words (subjective mood) and how the examiner perceives their predominant mood to be (observed mood). It is always helpful to use quotation marks specifying patients' exact words. Vari-

ations in mood during the course of the interview and examination should be described if they occur. The examiner should note if patients cry or laugh easily and if they are emotionally labile. The sudden starting and stopping of crying or laughter following a minor stimulus or no stimulus, referred to as *emotional incontinence*, indicates damage to specific neural fiber tracts and can aid in diagnosis. Mood should reflect what the conversation is about; for example, when patients talk about someone's death, they should appear sad. A lack of reactivity and evidence of unresponsiveness in mood should be noted. Irritability, anxiety, and emotional explosiveness or anger when confronted with a difficult task (a catastrophic reaction) is also important to report.

### *Vital sense*

This refers to patients' assessment of their energy level and whether it differs from their usual self-perception. Patients should be asked if they feel interested and energetic in performing their usual activities, whether they derive enjoyment and pleasure from usually enjoyed activities, whether their body feels well or sick, and whether they feel they will be able to sustain activity over time.

### *Self-attitude*

This is a complicated construct that assesses how patients perceive their own capabilities, and whether they believe these are different from their usual self-concept. Self-attitudes can be elevated, with better than usual self-confidence and self-esteem, or lower than usual, with guilt, remorse, self-deprecation, self-blame, and feelings of incompetence and failure. Low self-attitude is often accompanied by hopelessness, while elevated self-attitude often coexists with an inappropriately elated mood and grandiosity. Fluctuations in self-attitude, especially those not linked to environmental events, may be indicative of a mood disorder. Even when dementia is present, the identification of a *change* in self-attitude is a particularly important indicator of the presence of a mood disorder.

### *Assessment for thoughts of death, suicide, or homicide*

Patients who can understand should be asked if they have thoughts about death, catastrophe, or disaster. Do they wish death upon themselves and, if so, why? Do they think they would be better off dead? Would they consider hurting themselves? If they would rather be dead but are not thinking of hurting themselves, what is stopping them? Are they angry with someone else? Are they thinking of hurting someone? What is the reason for this? What has prevented them from doing it thus far?

### *Perception*

The examiner should investigate the presence of both hallucinations and illusions. *Hallucinations* are sensory perceptions without actual stimuli. They can occur in any of the five sensory modalities (hearing, sight, smell, taste, and touch). The examiner should ask patients if they hear sounds or people, or see things when nobody else is around or that others do not hear or see, whether they smell peculiar or odd odors that others cannot, or if there has been a specific repetitive taste or sensation that has been upsetting. Hallucinations are distinguished from *illusions*. In the latter, there is an actual stimulus that is misinterpreted or distorted—for example, seeing a face in the folds of curtains, believing a lamp is an animal, or looking into a mirror and seeing the face of a stranger.

### *Thought content*

This refers to ideas, beliefs, and explanations a person reports. Included under this heading is the presence of *delusions*. These are defined as ideas that appear to the examiner to be false, fixed (unshakable), and idiosyncratic (unique to the specific patient). Delusions are common in persons with dementia, are often a source of distress to them and others, and can lead to behavior problems. Common delusions in dementia include the belief that someone is coming into the house and stealing, that family members are taking money, or that a spouse is unfaithful. It is important to distinguish between delusions and ideas based on culture and background. One helpful way is to ask family, friends, and acquaintances whether they also believe what the patient believes. For example, the patient may have a religious belief that the examiner does not share. If relatives have the same belief, then it is unlikely to be a delusion and the most appropriate conclusion is that the difference between the examiner's and patient's perspective rests on culture rather than disordered content of thought.

Obsessions, compulsions, and phobias are also surveyed. *Obsessions* are recurrent, intrusive *thoughts* that the patient perceives as his or her own and attempts to resist. They often concern matters described by the patient as trivial or foolish. Over time, resistance may fade. Typical obsessions include preoccupation with cleanliness, orderliness, infestation, and disaster. *Compulsions* are repetitive *behaviors* that the person feels driven to perform, such as handwashing or touching the wall, but that the person perceives as unreasonable and attempts to resist. Compulsions often occur in response to obsessions and are followed by a reduction in anxiety. *Phobias* are disproportionate fears of specific objects or situations. They should be distinguished from the fearfulness that arises in response to a delusion or depression. Although obsessions, compulsions, and phobias occur in patients with

dementia, they are usually manifestations of a psychiatric disorder that preceded the dementia.

### *Insight and judgment*

*Insight* refers to a patient's awareness of cognitive or other deficits or of certain abnormal mental states. Insight is often impaired or lacking in dementia patients, especially those with one of the cortical dementias. This lack of awareness is a consequence of the underlying dementia rather than a psychological denial since it is uncommon in the subcortical dementias. The inability to perceive a deficit is called an *anosagnosia*. Insight is assessed by asking questions such as "Do you think there is something wrong?" or specific questions relating to the individual's function such as "Is your memory functioning well or are you having difficulty with it?" Lack of insight can explain what seem to be foolish, dangerous, or unusual behaviors and, more importantly, can determine how clinicians and caregivers should best relate to the patient. For example, a severely impaired patient with poor insight should not be confronted with a diagnosis of a degenerative disorder such as Alzheimer disease because this could lead to brief distress without a clear benefit (since the patient cannot become aware that there is a deficit).

*Judgment* refers to a person's ability to assess a situation, consider the facts and issues and draw an appropriate conclusion. It can be assessed by asking questions about a health-related situation, for example, "If you had a serious health problem, who would you talk to?" Judgment is also assessed from the history provided by the family and throughout the course of the interview by observing the way the patient approaches the examiner.

### *The cognitive examination*

Since cognitive impairment is the core feature of all dementias, every patient should undergo a thorough assessment of cognition. The extent of the cognitive assessment can vary, however, depending upon the purposes of the examiner and the setting in which the examination is being carried out. A neuropsychologist would be expected to carry out an in-depth, extensive inventory of a patient's cognitive abilities that would take several hours. A social worker, nurse, or physician reassessing a person with Alzheimer disease might use a brief global assessment to monitor the patient's course.

The cognitive assessment should also vary based on the patient's background. Individuals who have always been very bright or have depended on intellectual functions for their livelihood often need to be asked more complex questions to identify and assess cognitive deficits. For example, a

bookkeeper should be able to do more complex math. Patients with other strengths should be tested in areas about which they are especially knowledgeable. Given the wide variability in premorbid ability and exposure, there is no single assessment instrument that is appropriate for all situations or all patients.

Nonetheless, it is useful to have a standard method of cognitive examination with which one starts and then modifies as appropriate, for an individual patient. The most widely used standard cognitive examination is the The Mini Mental State Examination (MMSE) developed by Folstein, Folstein, and McHugh. The major strengths of this examination are its brevity and its broad coverage of cognitive functions. The chief limitations are its inability to identify very early dementia (called the *ceiling effect*), its dependence on language (resulting in very low scores in persons who have primary aphasia), and its inability to discriminate the degree of impairment in severely impaired individuals (called the *basement effect*). As with all cognitive tests, persons with little education do less well. Despite these limitations, the MMSE is a useful tool for assessing and following most individuals with dementia. It can also be used by the individual practitioner to follow normal individuals over time since a *sustained* drop of 3–4 points indicates a high likelihood of dementia. An occasional patient will remember items from previous testing, but this is rarely a problem in clinical settings. Several methods are used to score the MMSE. What is presented below is the version we most often use. Consistency of scoring is important because it allows an individual's performances to be compared over time and the comparison of the capacities of different individuals.

*The Mini Mental State Examination.* The first half of the MMSE consists of items related to memory, attention, and concentration. The second half measures cortical functions. Items 1 and 2 measure *orientation* to time and place. Questions include “Can you tell me where we are now?” and “What city and state are we in?” One point is given for each correct answer. When testing orientation to time (knowledge of the year, month, season, day of the week, and date), the first question asked may depend on whether the person appears, based on the initial conversation, to have a significant impairment. If disorientation is likely, we often first ask if the person knows the month and introduce the questioning in a nonthreatening fashion, for example, “Have you been keeping up with the date? Do you know what month it is?” When a person is doing well, every question should be asked, even the year, since mildly impaired patients sometimes know the day and date but not the year.

Item 3 tests *registration*, that is, the ability to immediately repeat back items being committed to memory. This is the first part of *memory testing*. Three words are given to remember in the following manner: “I’d like to test

your memory by asking you to remember three words. Please listen carefully and repeat these three words after me.” We always choose the same three words for new patients. This not only has the benefit of preventing embarrassment should the examiner forget the words (or forget to write them down) but also teaches the examiner that normal individuals are able to repeat that words without difficulty unless they have a marked hearing deficit. This item is scored by counting the number of words the person is able to repeat correctly the *first* time. If a person misstates a word, he or she does not receive a point for it. If the person asks for the words to be repeated, the examiner should first ask him or her to repeat as many words as are remembered since the score for registration measures how many words an individual reports on the first try. The three words are repeated until the patient is able to say all three or it is clear that they cannot be said all at once.

Difficulty in registering the words can indicate a hearing problem or a language problem. If not previously alerted to the possibility of a hearing problem, the examiner should note this as a possibility and perform a hearing assessment at some point. Sometimes the examiner can raise his or her voice and find that the patient is still unable to respond to a command or question appropriately. Whispering words or commands in the patient’s ear and having the patient listen to a watch or tuning fork are other forms of brief auditory testing.

The next item serves two functions. First, it distracts the patient from reciting the three words just asked; second, it is a test of *attention* and *concentration*. An individual with an eighth-grade or higher education is asked to subtract 7 from 100 and then to continue subtracting 7 from the answer. This is called *serial sevens*. If it is clear that respondents have memorized answers from prior examinations, the subtraction is altered and they are asked instead to subtract beginning from 101 or 103. This is one item that many practitioners do not expect older persons to perform correctly. It is a good example of why it is important to have experience testing the normal elderly. Experience and research demonstrate that individuals with an eighth-grade education or better can perform serial sevens. The speed of performance may slow down with age, so the examiner should be patient. An individual who is able to subtract 7 from 100 correctly should be able to do all the subtractions. One point is scored for each correct subtraction even if the previous subtraction was incorrect (so that “93, 87, 80, 73, 66” is given 4 points).

When individuals do not attempt the first subtraction (note: we do not say whether they *cannot* or *will* not) or when they have less than an eighth-grade education, they are asked to spell backwards a five-letter word with three consonants in a row (usually with *world* or *spray* as a backup). To determine whether persons have the ability to spell the word, it is best to ask them to spell *world* or *spray* forward and then, when they are finished, to

spell it backward. For the occasional person who misspells the word forward, the incorrect spelling in reverse is used as the correct sequence. When scoring backward spelling, a point is given for each response that matches the correct position in the sequence *d-l-r-o-w*. For example, *d-l-o-r-w* would score 3 points, while *d-r-o-l-w* and *l-r-d-o-w* would score 2.

After the distraction task, patients are asked if they can remember any of the three words that they were asked to remember (this tests recall or working memory). It is important to give individuals adequate time. Those in their eighties may take 30 seconds to recall all three words. One point is given for each word correctly recalled. The words must be *spontaneously* remembered to receive a point. For words that cannot be recalled (and thus scored as no points), the examiner may want to determine whether giving a cue or hint or asking the patient to choose the correct word from a list of words, some of which were not in the original three, improves performance. These questions can provide useful information but are *not* scored on the MMSE. For example, persons who cannot benefit from cues will need more direct help in remembering than individuals whose memory benefits from cuing. In giving cues, the clinician might start with a category. For example, “One was an animal.” Persons who are still unable to remember can then be given a choice such as “Was it a puppy, pony, or kitten?” Because the MMSE does not have a set time interval after which items are recalled, it is sometimes best to attempt a recall task with a 5-minute interval, particularly if the patient seems to have memory difficulty but recalls the items correctly the first time.

Several aspects of language are assessed in the MMSE. *Naming* is tested by asking the person to name two simple objects, such as a pen and a watch. A point is given for each correct response. Visually impaired individuals can be asked to name a pencil and a key placed in their palm. Examiners may also want to assess naming in more depth by asking the patient to name less common objects, for example, a button, an eraser, a lapel, the stem of a watch, shoelaces, or the buckle of a belt. Points are not given on the examination for these words, but repeated failures suggest a naming deficit. *Repetition* is assessed by testing the ability to repeat a specific phrase. The suggested MMSE phrase is “No ifs, ands, or buts.” The phrase must be repeated exactly, including all the *s*'s at the end of the words. The patient is allowed only one attempt. One point is given for a correct repetition. An alternate phrase is “Methodist, Protestant, Episcopal.” These phrases are difficult for individuals of some ethnic backgrounds or for those for whom English is not native. If there is a question about this being a problem, the sentence “Today is a (sunny) day in the month of (April—substituting the current weather and month)” can be used in its place. It is necessary that the patient say every word correctly. Repetition is an excellent screen for determining whether a

person has any language problem. It requires intact comprehension, intact registration, and intact expression of language. Repetition can be adversely affected by a hearing impairment. If this is present it should be noted, since this can have important clinical and therapeutic implications, but the item is still scored as not performed correctly. The next item addresses the ability to *read* a sentence *and carry out* the action. Reading the sentence “Close your eyes” is included in the MMSE. Patients are asked to read the sentence to themselves and then carry out the action. The print should be large so that it can be easily read by those with visual problems. Some patients are able to say the sentence but not carry out the action. A point is not given in this case.

Following a *three-step command* requires that people comprehend that the examiner wants them to do something, that they can hear what is said, and that they are able to carry out the three distinct steps. This tests several cognitive abilities but is most indicative of *praxis*, defined as the ability to carry out learned motor movements. The three-step command on the MMSE asks the subject to take a piece of paper in the right hand, fold it in half, and then place it on the floor. A point is given for each step done correctly. The reliability of this item is surprising to some people. Patients who are able to do only one or two steps when first asked will usually be able to do only the same number of steps when asked to do them a second time.

Next, a person is asked to *write* a sentence spontaneously. One point is given if it is a complete sentence (with a subject and a verb), if it is grammatically correct, and if it does not contain language errors. Some patients say that they do not know what to write. In this instance, we encourage them to “write anything that comes to mind.” When an individual is still not able to write a simple sentence, the examiner might suggest one, for example, “Today is a (warm or cold) day,” changing the adjective depending on the temperature. However, a point is given only if the patient *spontaneously* writes a complete, grammatically correct sentence. Finally, the patient is asked to *copy* a design with 2 five-sided interlocking shapes. A point is given if each figure has five sides and five angles and if the overlap is a four-sided figure. This assesses *visuospatial function and praxis*.

The interpretation of an MMSE total score depends on the presence or absence of non-cognitive impairments (e.g., blindness, dominant arm weakness) that might account for the loss of certain points, as well as on the person’s estimated premorbid cognitive abilities, based among other things on his or her education and occupation. An MMSE score below 24 is indicative of significant impairment. For blind individuals, a score of 27 is probably normal, as they would be unable to complete three items due to blindness. In contrast, a score of 25 might be abnormal for a person with a high premorbid ability, such as an attorney or a nuclear physicist.

*The expanded cognitive examination.* The MMSE adequately tests orientation, memory, praxis, language, attention, and calculation. However, there are other aspects of cognition (see Table 4.5) such as consciousness, fund of knowledge, and executive function that are not assessed well by the MMSE. Since successful performance on the MMSE does not necessarily indicate the absence of a dementia, particularly in persons who premorbidly were quite high-functioning intellectually, a more in-depth cognitive examination is sometimes indicated. For these reasons, more sophisticated clinicians may choose to use in their day-to-day practice the Modified MMSE (3MS), which quantifies cognitive functioning on a broader 100-point scale and overcomes many of the limitations of the MMSE, such as the ceiling effect. The 3MS was developed by Teng and Chui in California and has been used in many studies that provide norms against which to compare the performance of specific patients. The scale can be obtained from the original publication (Teng EL, Chui HC. The Modified Mini-Mental State (3MS) examination. *J Clin Psychiatry* 1987; 48(8):314–18).

Alternatively, clinicians may choose to conduct an in-depth assessment of a particular aspect of cognition. The discussion below gives examples of other important parts of the examination.

*Level of consciousness* is assessed by evaluating the patient's ability to engage in and focus on the examination without fluctuation or *waxing and waning*. The presence of any limitation or fluctuation in attention might be indicative of delirium. Attention can be tested by having the patient repeat a sequence of numbers. Normally, seven numbers can be repeated forward and five backward at a minimum.

A more in-depth examination of recent *memory* (or memorizing or memory for newly learned material) includes determining whether the patient can recall lists of more than three words or recall the second of a pair of words when given the first word of the pair. Testing for current events (e.g., what is in the news) is also a memory test. Almost every intact individual knows who the current president is, and the ability to name the previous four presidents also suggests intact memory. An inability to name previous presidents or to name the current vice-president should be interpreted in light of the patient's estimated premorbid abilities. *Remote memory* (or memory for previously learned material) refers to previously learned material such as events from one's personal life, important dates, names of prior acquaintances, historical events, or personalities. It is tested by asking patients about dates, people, and places they would be expected to know. Failure to know the names and ages of grandchildren is a likely indication of impairment, for example.

A more detailed assessment of *praxis*, *planning*, and *visuospatial* function includes asking the patient to draw a flower pot or to draw a clock with

the numbers in the correct place and the hands pointing at “10 minutes past 11.” Assessments of ideomotor (cannot pretend that they are using a tool or instrument) and ideational (can imitate an action only if they copy someone else) praxis include asking patients to demonstrate how people brush their teeth, comb their hair, button a button, or salute.

Testing the patient’s *fund of knowledge* provides information about pre-morbid intellectual ability and current intellectual ability. Questions asked include knowledge of recent news events or the functioning of certain equipment such as a barometer, the color of a ruby, or the capitals of certain states or countries.

*Abstraction* is an element of cognitive function that can be tested through proverb interpretation and the ability to identify similarities and differences. The interpretation of proverbs requires intact language skills but is primarily a measure of abstraction. Abstraction is strongly influenced by cultural factors. Thus, proverbs must be in the cultural repertoire of the patient to be appropriate. When testing this, it is common to start with an easily interpretable proverb such as “Don’t cry over spilled milk” (“What’s done is done”). A second, more difficult proverb is “People who live in glass houses shouldn’t throw stones” (“Don’t criticize others if you also have faults”). Another often used proverb is “A stitch in time saves nine” (“Don’t put things off”). A difficult proverb that is unfamiliar to most individuals but that usually can be interpreted by individuals with intact normal intelligence is “The tongue is the enemy of the neck” (“Don’t talk too much”).

The ability to abstract can also be tested by asking individuals how an apple and an orange are alike. The correct response is “fruit.” If an incorrect answer such as “They are both round” is given, the examiner should say, “And they’re both fruit. Now try . . .” Subsequently, more difficult pairs of items are given, such a hammer and a saw (tools), a table and a chair (furniture), a bicycle and an airplane (means of transportation), or a bird and a tree (both alive). The ability to abstract is also tested by asking the difference between a river and a canal (natural vs. man-made) or a dwarf and a child (a child will grow tall).

Tests that require the ability to focus attention and switch sets (concepts) in rapid succession, sometimes referred to as *executive function*, should be considered, particularly when frontal lobe impairment is possible. Three tests are commonly used in this regard. The first is a verbal form of the Trail Making Test. The test is introduced by saying, “I’m going to ask you to alternate numbers with letters. Please complete the sequence. 1-A, 2-B; keep going.” This requires the ability to recite the alphabet and to count to 15. The examiner keeps track of the patient’s responses and corrects them as needed. For example, the patient might say “1-A, 2-B, 3-C, 4-D, 5-F,” in which case the examiner would say, “5E; keep going.” If the patient reverses

the sequence, as in “1-A, 2-B, C-3, 4-D,” no correction or penalty is applied. Two minutes are allowed for the patient to go all the way to 13-M. The examiner keeps track of time. Taking more than 60 seconds or making three or more errors indicates impairment.

A second test of executive ability is the Luria Hand-Sequencing Test, in which the patient is asked to mimic or copy a sequence of hand movements done by the examiner. The examiner demonstrates a series of three hand positions, which might be an open palm, a fist, or scissor fingers. After each set of three hand positions the patient is asked to copy the exact sequence. Five sets of three positions are given. Most well elderly patients are able to successfully copy four of five sets in the absence of impairment. Younger persons can do better.

The final test is referred to as the Go-No Go Test, in which the examiner gives the instruction, “I am going to tap on the table with my fist. If I tap once, I want you to respond by tapping twice. If I tap twice, I want you to respond by not tapping at all.” Ten trials are given. Most well elderly respondents can successfully complete 6 or 7 of 10; younger persons can do better, often a perfect 10.

### **Use of standardized scales to supplement assessment**

The process of assessment is often complemented by the use of a limited number of standardized scales that assess different domains. The rating of these scales allows the clinician to summarize and organize a complex case by reference to a set of numbers. This is useful in communicating information about severity to other caregivers, provides an objective means of charting the course of illness over time, and helps assess the response to interventions.

Many scales have been developed for rating different features of dementia. We favor the use of scales that are simple to administer, have been shown to be reliable, and have broad coverage. The domains that are important to consider are cognition, mood, behavioral disturbances, and ADLs (both instrumental and basic).

To rate cognition in early and moderately advanced stages, we recommend the MMSE or the 3MS. For late-stage patients we recommend the Severe Impairment Rating Scale (SIRS). To rate depression, we recommend the Cornell Scale for Depression in Dementia (CSDD). To rate dependency on caregivers and to assist in making level-of-care decisions, the Psychogeriatric Dependency Rating Scale-Behavior subscale is preferred (PGDRS). To rate IaDLs we recommend the IADL, and to rate ADLs we recommend the PGDRS-physical subscale. These scales can be copied for individual use and can be obtained from the original references. To rate noncognitive neuropsychiatric disturbances we favor the Neuropsychiatric Inventory (NPI) or

its derivative caregiver questionnaire. Both can be obtained by writing to Jeffrey L. Cummings, M.D., Department of Neurology, Reed Neurology Center, UCLA School of Medicine, 710 Westwood Plaza, Box 951769, Los Angeles, CA 90095-1769.

### The family assessment

The importance of evaluating the patient's family and personal environment cannot be overemphasized. The family has been described as "the lifeline of the patient." Their well-being is essential to the patient's status, and the family is appropriately considered a partner in care. As professionals embark on the care of the patient and family, it must be remembered that the provision of a diagnosis and treatment recommendations will aid the family in the years ahead but must be supplemented by the many other approaches discussed in this book. Data on the family that are collected at around the time of the initial contact can be crucial for understanding the family's needs.

Table 2.6 lists the elements important to the family and their assessment. The family's well-being is best monitored over time by revisiting these areas at intervals of no longer than 6 months. Although family assessment is frequently provided by social workers, it can be accomplished by other health care providers as long as the information is systematically collected. Construction of a family genogram is one effective way to gather these data. An example is in Figure 2.1.

The assessment of the patient's functioning within the family should cover the following topics: knowledge of how the patient's needs and wants are provided for; the way in which the patient spends his or her time; the extent to which family and/or care providers have insight into the patient's condition and its prognosis; the extent to which care providers require help in caring for the patient; and the resources family caregivers have available to provide help.

Through the psychosocial assessment, the clinician will develop an understanding of the patient's immediate environment, day-to-day functioning, and resources. This will add to the intervention problem list and will lay the groundwork for developing appropriate long-term supportive care for the patient and family (see Chapters 7, 8 and 9).

## DIFFERENTIAL DIAGNOSIS AND WORKUP OF DEMENTIA

After completing the assessment discussed at the beginning of this chapter, the clinician is faced with the task of making sense of the information. Doing so has two purposes: diagnosing the cause of the dementia and developing a

TABLE 2.6. Assessment Domains for the Family Evaluation

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1. Family members, their roles, frequency of interaction
2. Health status of care providers
3. Financial status of care providers
4. Spiritual beliefs
5. Knowledge about dementing illnesses
6. Other responsibilities, including work and other dependents, such as children or other ill relatives

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care plan. A list of possible diagnoses, called the *differential diagnosis*, is made, and this clarifies what other information is needed.

The goals of the differential diagnosis are the formulation, classification, and determination of the cause. This process is best understood in a series of sequential steps (outlined in Fig. 2.2) to which we refer throughout this chapter.

In the *formulation*, the clinician organizes the history, physical examination, mental status examination, and laboratory studies in a coherent and systematic fashion. The first step is to decide whether dementia is present or absent. If cognitive impairment is absent (in which case dementia is absent), then the differential diagnosis typically involves disorders such as depression, schizophrenia, factitious disorder, or a neurologic disorder that spares cognition. If cognitive impairment is present, then the clinician must decide if the *clinical* definition of dementia is met (i.e., a decline in multiple cognitive capacities occurring in clear consciousness). If cognitive impairment is present but does not represent a decline, then a diagnosis of mental retardation, borderline intellectual functioning, or development delay (DD) is appropriate. If cognitive decline is evident but is either not global or not sufficiently severe to affect functioning (i.e., does not meet criteria for dementia presented in Table 1.1), then another diagnosis such as cognitive impairment, no dementia (CIND), mild cognitive impairment (MCI), amnesic disorder, aphasia, mild cognitive disorder, or age-associated memory impairment should be considered (see also Chapter 11). If the clinician is still uncertain about the presence or absence of dementia after the initial assessment, then long-term follow-up will be necessary to determine whether dementia is present. Uncertainty is most common when the patient was highly functioning premorbidly and is only mildly impaired, when the clinician is not persuaded that the patient's current functioning represents a decline, or when the patient is very old. Neuropsychological testing, a series of standardized and normed tests of cognitive function performed by a specially trained clinician, can be an invaluable tool in the evaluation of dementia. Referral for testing is indicated

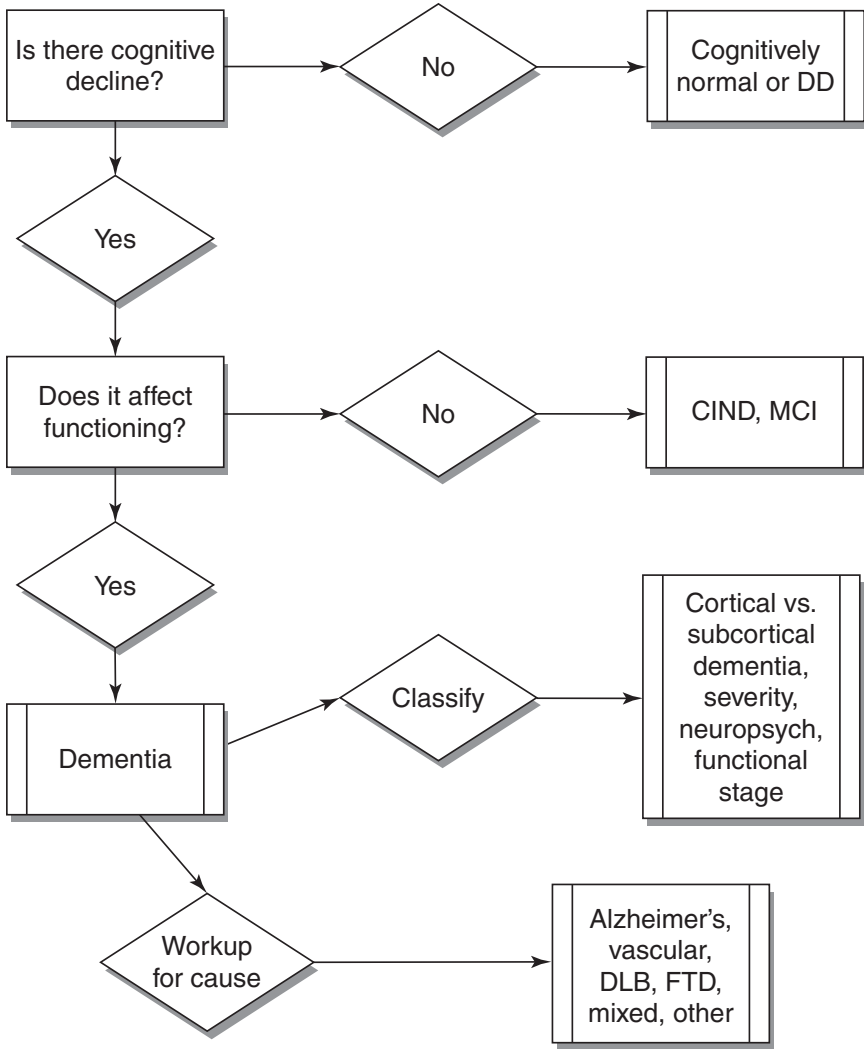


FIGURE 2.2 Differential diagnosis of dementia. CIND, cognitive impairment, no dementia; DD, development delay; DLB, dementia with Lewy bodies; FTD, fronto-temporal dementia; MCI, mild cognitive impairment.

when there is cognitive impairment but it is not clear if it is severe enough to be a dementia, when the impairment could be accounted for by advanced age, when there is dementia but there is uncertainty about its cause, or when there is a need to differentiate dementia from depression or schizophrenia.

If the clinician decides that dementia is present, the next step is the *classification* of the syndrome according to its cognitive features (cortical,

TABLE 2.7. Laboratory Investigations for the Evaluation of Dementia Syndromes

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Urinalysis and microscopy
Blood tests
Complete blood count
Serum electrolytes, including magnesium
Serum chemistries, including liver tests
Thyroid testing
Vitamin B <sub>12</sub>
*Erythrocyte sedimentation rate
*Serologic tests for syphilis (or similar conditions)
*Chest X-rays
*Electrocardiogram
*Toxicology screens
Urine toxicology
Serum toxicology (alcohol, salicylates, other)

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\*Consider; not universally needed.

subcortical, mixed), noncognitive features (behavior, mood, function, motor), and functional impairments. This classification lays the foundation for determining etiology and directing treatment. Tables 1.2, 1.4, and 1.5 will assist in this classification.

The *differential diagnosis* is a list of potential causes (Table 1.3). It is constructed by matching the patient's history, signs, and symptoms with known disorders. Ideally, further clinical and historical data, as well as laboratory testing, imaging, or neuropsychological testing, will identify a single likely cause.

Table 2.7 lists laboratory evaluations for all patients with dementia. The recommendations here are consistent with those developed by the relevant guidelines of the American Psychiatric Association and the American Academy of Neurology. As noted in the table, certain laboratory studies should be ordered in all cases mostly to rule out rare causes of dementia. Note that the table does not include laboratory studies that are being marketed for the diagnosis of Alzheimer disease, such as apolipoprotein E genotyping, presenilin-1 genotyping, or cerebrospinal fluid testing for neurofil protein or different forms of tau protein. At this point, there is not sufficient evidence to support the use of the latter in the differential diagnosis of dementia.

Brain imaging should be considered in all cases but is not always necessary, so clinical judgment should be exercised. If the dementia is long-standing (more than 3 years) or very advanced, we sometimes forgo imaging because a treatable cause is very unlikely and the identification of less common dementias is unlikely to affect care. While a brain computed tomogram (CT)

TABLE 2.8. Second-Stage Laboratory Investigations

<i>Test</i>	<i>Indication</i>
Electroencephalogram (EEG)	Possible seizures; Creutzfeld-Jakob disease
Lumbar puncture	Onset of dementia of <6 months to rule out rare infectious causes Obtain cerebrospinal fluid to assess for Creutzfeld-Jakob disease proteins (G-14) or Whipple disease proteins High-volume “tap” or continuous pressure monitoring to assess for normal pressure hydrocephalus
Heavy metal screen for mercury, arsenic, and lead (urine or serum)	History of potential exposure
Human immunodeficiency virus (HIV) test	History of potential exposure
Lyme disease titer	History of exposure and compatible clinical picture
Ceruloplasmin, arylsulfatase electrophoresis, Slit lamp exam	History and exam suggest Wilson disease
Apolipoprotein E testing	Need to increase the likelihood that the diagnosis of Alzheimer disease is correct
Genetic testing for presenilin-1 Alzheimer gene, CAG repeat determination in Huntington gene or other dementia genes	Family history is strong and confirmation is clinically necessary

is sometimes adequate, we usually order a brain magnetic resonance imaging (MRI) scan because it offers more information. Single photon emission tomography (SPECT) or positron emission tomography (PET) should be ordered to rule specific causes in or out. For example, SPECT/PET can identify specific patterns of blood flow or metabolic abnormality and confirm or rule out specific disorders, such as frontotemporal dementia or Alzheimer disease.

When the clinical circumstances, history, or examination indicate that a *specific* cause might be present for which a specific diagnostic test is available (Table 2.8), it should be ordered. Table 2.8 also contains genetic testing for Alzheimer-associated genes that may be of value in specific clinical circumstances.

Differential diagnosis is a top-down process in which the clinical phenomena, both signs and symptoms, first indicate the presence or absence of the syndrome of dementia and then direct the classification of the syndrome

into recognizable patterns. Because it is assumed that in all cases of dementia there is an underlying disease of the brain, and since in most cases the clinical pattern correlates well with findings at autopsy, knowledge and understanding of the clinical syndrome is important. However, clinicians should not lose sight of the fact that in a sizable number of cases, perhaps 15%–20%, the clinical presentation of dementia is atypical—that is, it does not follow a recognizable pattern and therefore the pathologic cause is less certain.

We do not favor classifying dementia as reversible or irreversible, however. Even when a dementia for which there is a specific treatment is diagnosed, it often does not fully respond to treatment, particularly if it has been present for more than several months. Also, many of the treatments that are available now and others that will become available in the near future will lead to partial or transient improvement but not full recovery.