

Chapter 1

What Is Dementia?

All Alzheimer’s disease is dementia, but not all dementia is Alzheimer’s disease. Despite this crucial distinction, even experienced clinicians often confuse these terms. It is important to understand and differentiate the specific type of dementia affecting a patient as the specific diagnosis may determine both treatment and prognosis. And these are usually the overarching concerns of loved ones from the very start of the dementing illness and the clinical evaluation. So—what is dementia? And—what are the signs?

Basic Clinical Definitions

First, let us define some commonly used clinical terms. The clinical term prognosis refers to the expected course of a disease, including the duration or how long it will last. This is usually one of the most important concerns of a patient and his or her family regarding any clinical diagnosis (including dementia). When physicians use the term “progressive or progressing,” we are usually referring to the progressive *worsening* of a disease state, unless specifically stated otherwise. We also use the term “onset” to indicate the way a disease starts. An acute onset refers to a disease that presents suddenly over seconds, minutes, hours, or perhaps even a few days. A subacute onset means that a disease presents over days to a few weeks. Illnesses that occur over weeks, months, and years are considered to have a “gradual” onset. The progression of a disease may also be acute, subacute, or gradual. Dementia is therefore usually a disease with gradual onset and progression. Because the symptoms of dementia often begin so subtly and surreptitiously, “sneaking up” on a patient, dementia is often described as having an “insidious” onset.

Table 1.1 Types of onset and progression

| | Onset of symptoms | Disease progression |
|----------|---------------------|--|
| Acute | Days or less | Days or less |
| Subacute | Days to a few weeks | Days to a few weeks |
| Gradual | Months or years | Months or years |
| Stepwise | Acute or gradual | Months or years interspersed with time points of acute worsening which may be followed by stabilization or improvement of acute symptoms |

In some cases, fluctuations in the progression may be seen, such that occasional acute worsening or even transient mild improvements may be seen, but a dementia will continue to gradually worsen (progress) over a period of years. Stepwise progression refers to a type of progression that is gradual overall but with time points of acute worsening, sometimes followed by improvement and/or stabilization of the acute symptoms. This may be seen in Vascular dementia in which a patient has acute worsening of cognitive and/or behavioral symptoms each time he or she has a stroke, perhaps followed by some stabilization or even improvement in the new symptoms (but not of the problems preceding the stroke) (Table 1.1).

If symptoms begin or progress (worsen) more suddenly, then an unusual type of dementia or an alternate diagnosis should be considered. Dementia is a terminal illness in that it may eventually result in death in the absence of any other factors. However, many patients with dementia have cardiovascular and other associated disease states. Therefore, patients with dementia may die of cardiovascular (e.g., strokes or heart attacks) or other causes before succumbing to dementia [1].

Dementia: Definitions

Dementia refers to a gradually progressive brain illness that affects cognition (thinking) and/or behavior to such an extent that daily function is impaired. We therefore use the terms dementing illness and dementia interchangeably throughout our text.

Alzheimer's disease (AD) refers to a specific type of dementia in which memory "leads the way" and problems with recall are the "first and worst" issue. Although researchers and medical publications often use the term "Alzheimer disease" for this specific type of dementia, our usage of the possessive term, Alzheimer's disease, reflects the common usage of the general public, clinicians, and residential and support organizations, all of whom comprise the target audience for this book. We also use the abbreviation AD, which is a commonly accepted shorthand for Alzheimer's disease (or Alzheimer disease).

Clinicians use the terms cognition or mental status function, when referring thought processes such as attention and concentration, memory, language, visuospatial skills (which include hand-eye coordination), and complex thought processes known as executive functions. Each of these categories constitutes the

so-called cognitive domain. The cognitive domain of *executive functioning* includes those complex thought processes that help us learn in school and perform at work, including sustained attention, planning, organization, judgment, anticipation, ability to alternate between tasks (mental set-shifting), and response inhibition. Synonyms for cognition used in this text include thinking, mentation, mental processes, and mental status function. Formal assessment of these cognitive domains may be done by physicians, neuropsychologists, or others and referred to as mental status testing, neurocognitive evaluation, or neuropsychological testing. Neuropsychological evaluations tend to be the most comprehensive of all of these.

Mood is used clinically to indicate how someone describes his or her own emotional state. Affect is the clinical term referring to how the emotional state of a patient is perceived by the professional evaluator. Psychosis refers to an affective condition in which a patient has lost touch with reality and has hallucinations, delusions, or both. Hallucinations refer to abnormal sensory experiences in which one perceives something that isn't really there. These may be visual (seeing things), auditory (hearing something), or tactile (sensation of feeling something), or affect taste (gustatory) or smell (olfactory). In dementia, visual hallucinations are the most common, but auditory hallucinations may also occur. Tactile hallucinations suggest a different diagnosis than dementia. Delusions designate a mistaken belief, such as believing that another person is present when they are not. Delusions may involve paranoia (in which a patient mistakenly believes that others are trying to inflict harm in some way). The presence of psychosis in the initial months or years of a dementia suggests a non-Alzheimer's dementia.

Onset of Dementia

As noted above, dementia starts gradually and often insidiously, such that the initial symptoms do not cause much concern. Behavioral or cognitive problems that interfere with daily function are not normal. Gradually progressive memory loss is the most common, but by no means the only way in which dementia may begin (see Chap. 2). The initial symptoms should be noted carefully as they often help in making a specific diagnosis. A useful mnemonic (memory device) to recall the major symptom areas involved is to remember the "ABCs" of dementia: Activities, Behaviors, and Cognition.

Progression of Dementia

As specifically applied to dementia, "gradual progression" refers to worsening of the clinical condition over years. A few rare types of dementia can result in more rapid deterioration. However, even in these unusual cases, the initial onset of symptoms usually occurs over months to years (and not over minutes or hours or even

days or weeks). By definition, symptoms of dementia must be present for at least 6 months to make a diagnosis. There is no such thing as “rapidly progressive” AD and an alternative or additional diagnosis should be sought if a patient has been diagnosed with AD and the progression of symptoms occurs over fewer than 6 months.

Stages of Care

Different stages of dementing illness have distinctive care requirements. Staging schemes can be useful constructs but serve as guidelines only since dementia progresses uniquely for each individual.

We find the classification of dementia into mild, moderate, severe, and late stages [2] to be the most clinically useful construct and the most practical paradigm for caregivers. A person with a mild dementia may have memory loss (and/or other mild cognitive and/or behavioral deficits) with a few problems in more complex daily functions (or if using the Mild Cognitive Impairment rubric, no such functional deficits). Someone with moderate-stage dementia would have more serious cognitive and/or behavioral deficits along with significant deficits in daily activities and might need some assistance with simple Activities of Daily Living (ADLs), such as eating, dressing, and behavior. A patient with severe dementia has severe cognitive and/or behavioral impairments and needs significant or total assistance with basic tasks. Late-stage dementia denotes the terminal phase of this illness and is discussed in Chap. 16.

We recommend that caregivers retain a general understanding of and plan of care for each of these stages, but focus their attention and efforts in meeting the current and ongoing needs of a loved one with dementia.

References

1. Kukull WA, Brenner DE, Speck CE, Nochlin D, Bowen J, McCormick W, Teri L, Pfanschmidt ML, Larson EB. Causes of death associated with Alzheimer disease: variation by level of cognitive impairment before death. *J Am Geriatr Soc.* 1994;42(7):723–6.
2. Morris JC. Clinical dementia rating: a reliable and valid diagnostic and staging measure for dementia of the Alzheimer type. *Int Psychogeriatr.* 1997;9 Suppl 1:173–6. discussion 177–8.