Chapter 1

Dementia
The Clinical and Diagnostic Context

1.1 Introduction

The purpose of this chapter is to give a brief overview of some of the clinical features of dementia. A level of knowledge is assumed and there are other texts that comprehensively examine general aspects of dementia diagnosis, neurobiology, current interventions, etc.[1] We will provide an overview of the range of cognitive systems and how impairments in these systems relate to the clinical presentation. While decision-making capacity and cognitive abilities are related, they are not the same thing. The first refers to the patient's ability to make a particular decision (at a particular point in time), and the latter encompasses a broad range of processes, including a person's memory, problem-solving, language, attention and visuospatial abilities. Understanding different cognitive domains and how they interact is necessary for accurate clinical assessment and in determining disease severity. Also, in relation to legal considerations, characterising specific deficits may also help to explain why decision-making capacity is impaired or, just as importantly, in identifying how to apply measures to support an individual's decision-making.

1.2 Definition and Causes of Dementia

The term 'dementia' embraces a wide range of diseases of which Alzheimer's disease (AD) is the most common. The International Classification of Diseases revision 11 (ICD-11)[2] gives a general description of dementia in the following way:

> Dementia is characterized by the presence of marked impairment in two or more cognitive domains relative to that expected given the individual's age and general premorbid level of cognitive functioning, which represents a decline from the individual's previous level of functioning. Memory impairment is present in most forms of dementia, but cognitive impairment is not restricted to memory (i.e., there is impairment in other areas such as executive functions, attention, language, social cognition and judgment, psychomotor speed, visuoperceptual or visuospatial abilities). Neurobehavioural changes may also be present and, in some forms of dementia, may be the presenting symptom. Cognitive impairment is not attributable to normal aging and is severe enough to significantly interfere with independence in an individual's performance of activities of daily living.

The definition of dementia is therefore necessarily clinical and pragmatic, saying nothing about the challenges, complexity and diversity of symptoms of these diseases. While typically progressive, dementia in some people may be a relatively static – for example, following cerebral infarctions[3] or head injury.

https://doi.org/10.1017/9781911623250.002 Published online by Cambridge University Press

There are many other diagnosable cerebral and systemic diseases that may result in dementia and are discussed elsewhere.[4] Each subtype of dementia has its own underlying neuropathological findings and constellation of signs, symptoms and clinical course. Consensus guidelines exist for the clinical and pathological diagnosis of many different forms of dementia (e.g. Alzheimer's disease[5, 6]). After Alzheimer's disease, the second most prevalent cause of dementia is vascular dementia (VaD), which itself may be caused by several disease processes in the brain.[7] The term 'mixed dementia' generally refers to the co-existence of Alzheimer's and vascular pathology, but the distinction between both diseases is controversial.[8] There are many other causes of dementia, including frontotemporal lobar dementia[9] (FTLD) and dementia with Lewy Bodies[10] (DLB). Frontotemporal lobar dementia is a descriptive term that encompasses a heterogeneous group of clinical syndromes associated with non-Alzheimer pathology and arises from degeneration of the frontal and temporal lobes. The most commonly occurring syndrome is behavioural-variant frontotemporal dementia, characterised by changes in behaviour and personality in association with frontal-predominant cortical degeneration.[11] Memory disturbance may be less prominent and appear later in the course of the illness. The characteristic clinical history of dementia with Lewy Bodies (in addition to diagnostic requirements for dementia) are two or more features of: parkinsonism (e.g. resting tremor); recurrent visual hallucinations (typically well formed); episodic confusion and rapid eye movement sleep behaviour disorder.[12]

Classification of alcohol-related brain damage (ARBD) is problematic (discussed in Royal College of Psychiatrists, College report CR185[13]) and refers to a variety of different conditions. Chronic alcohol consumption may result in discrete neurological syndromes such as Wernicke encephalopathy or, its sequel, Korsakoff syndrome. Personality changes may also occur, as well as long-term cognitive deterioration sometimes manifesting as a 'frontal dysexecutive syndrome' (see section 1.7.2). The use of the term 'dementia' may be moot as many people with alcohol-related brain damage can improve with appropriate care. Although in general we will not separate the different pathologies when we discuss legal aspects, complications of 'alcohol abuse' appear to be a significant diagnostic factor in welfare applications to the Court of Protection (CoP).[14] Not only will we discuss the role of the CoP in detail in Chapter 13, but we will also refer to it throughout the book.

1.3 Epidemiology and Demographics of Dementia

There were an estimated 850,000 people with dementia in the UK in 2015.[15] Alongside the increase in life expectancy, the UK population is growing. If the prevalence of dementia remains the same, the total number of people with dementia in the UK is forecast to increase to over 1 million by 2025 and over 2 million by 2051. Estimates of frequency of dementia, and other diseases, in a population are important as they highlight the extent of the healthcare and public health challenges. While there are some difficulties in methodology and interpretation, studies of prevalence (the actual number of cases alive, with the disease at a point in time) and incidence (the rate of new or newly diagnosed cases) of dementia and Alzheimer's disease have consistently shown an almost exponential increase with advancing age.[16]

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Dementia may also affect younger people. In a London-based survey, the prevalence of dementia in those aged 30 to 64 was 54 per 100,000, with an estimate of more than 18,000 people in the UK under the age of 65 with dementia.[17] Dementia also occurs at a much higher rate among older people with learning disabilities than it does among the general population.[18] In people with Down's syndrome, there is an increased risk of developing Alzheimer's disease in middle age.[19]

As the most frequent cause of dementia, studies on risk factors for dementia have mainly focused on Alzheimer's disease. Mostly, Alzheimer's disease is a sporadic, age-dependent, late-onset disease.[20] A positive family history is found in approximately 20% of diagnoses and there is a higher prevalence in women. Several genetic mutations have been identified in early-onset autosomal dominant familial Alzheimer's disease, involving genes for amyloid precursor protein (*APP*), presenilin-1 (*PS-1*) and presenilin-2 (*PS-2*).[21] The major genetic risk associated with sporadic late-onset Alzheimer's disease is conferred by a positive family history of dementia[22] and by the e4 variant of Apolipoprotein (*APOE*) genotype.[23]

1.4 The Cost of Dementia

Dementia is a critically important issue in terms of both the high personal and social costs related to the disease and the wider impact on other parts of the health and care system. The overall cost of dementia care in the UK is currently £26 billion a year[24] (including £4.3 billion in NHS care and £10.3 billion in social care costs). Driven by demographic changes in the population, unsurprisingly, this is set to rise as the number of people with dementia grows. This works out at an average annual cost of £32,250 per person with dementia. Alarmingly, either directly in terms of charges for services or indirectly through providing personal and other interventions, carers and families currently shoulder an enormous amount of the cost themselves (£11.6 billion). The Department of Health has indicated that the cost effectiveness of dementia care could be significantly improved and point to the requirement for improvements in diagnosis, treatment, and care and support for people with dementia and their carers.[25]

1.5 Symptoms and Signs of Dementia

The clinical spectrum of cognitive symptoms, or observable deficits, is wide and is shown in Table 1.1.

1.6 Diagnosis of Dementia

The neuropsychological components of the dementia syndrome consists of a range of cognitive impairments, as shown in Box 1.1.

The effects of dementia, therefore, result in a decline in intellectual functioning and interference with activities of daily living (ADLs). ADLs are categorised as basic or instrumental and both types are valuable for evaluating the disease severity and in assessing the person's level of independence. ADLs are therefore an important marker for diagnosis, progression and prognosis of dementia.

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Note
Basic and Instrumental Activities of Daily Living

***Basic* ADLs** are a limited set of tasks of everyday life, such as eating, bathing, dressing and toileting, which are fundamental to caring for oneself and maintaining independence.

***Instrumental* ADLs** are more complex skills which maintain a person's independence and include activities such as shopping, cooking, managing medications, housework and managing finances.

Table 1.1 The clinical spectrum of cognitive symptoms

Effects of aging	Healthy older people usually retain the ability to use compensatory strategies, such as keeping written prompts, lists and calendars, and are capable of new learning and adaptive skills.
Mild cognitive impairments (MCI)	A syndrome defined as: • Cognitive decline greater than expected for an individual's educational level, but that does not interfere notably with activities of daily life.[26] • MCI has been proposed as a condition of impairment intermediate between what is considered normal for aging and that which is insufficient for a diagnosis of dementia. • Criteria for MCI require the presence of a subjective memory complaint with objective evidence of memory impairment by cognitive testing in the setting of generally preserved activities of daily living. • However, not all MCI subjects progress to Alzheimer's disease; some remain stable and others progress to non-Alzheimer's dementias. This relative sparing of functional impairment represents the fundamental distinction from dementia.
Diagnosable dementia syndrome	Symptoms that signal the onset of dementia may be subtle and not necessarily noticeable for several years. The earlier signs may only be identified in retrospect and often misattributed to 'aging' rather than the early manifestations of a neurodegenerative condition. Although memory loss is often thought of as the core symptom in dementia, other cognitive domains may be affected and include executive functioning, attention, language, social cognition and judgment, perceptual skills and personality.

Therefore, to make the diagnosis with a view to clarifying the severity and subtype of dementia, the following may need to be conducted:

- a medical history, neurological examination and mental state examination;
- standardised memory or other neuropsychological testing, involving an analysis of a range of cognitive processes utilising a set of cognitive tests which may be supplemented if there is a requirement for a more detailed or individualised approach;

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Box 1.1 Neuropsychological features of dementia

- memory impairments (e.g. declarative and procedural memory problems; see section 1.7.1);
- aphasias (an impairment of language, affecting the production or comprehension of speech and the ability to read or write);
- apraxias (inability to carry out directed coordination of movements despite intact sensory and motor nervous systems);
- agnosias (inability to recognise specific elements of an individual's environment or self);
- attentional difficulties (including sustained and divided attention);
- executive functioning impairments (including difficulties with abstraction, cognitive flexibility, inhibition, planning, organising and adaptation to novel stimuli; see section 1.7.2).

- basic blood tests may be used to help exclude other causes of memory difficulties, including potentially reversible causes;
- assessments of functional abilities;
- brain scans.

Biomarkers are increasingly employed in the research setting to detect onset of the disease[27] and to track progression, but cannot yet be used routinely in clinical diagnosis without further testing and validation. If available, the history and the perspective of a reliable informant is often invaluable.

1.7 Cognitive Domains

The following represents a brief account of a range of cognitive domains in order to provide a conceptual model of how they may operate.

1.7.1 Memory Systems

Although it is common to conceptualise memory into long- and short-term components, a more useful framework for analysing memory divides it into broad domains or systems, illustrated in Figure 1.1 and Table 1.2.

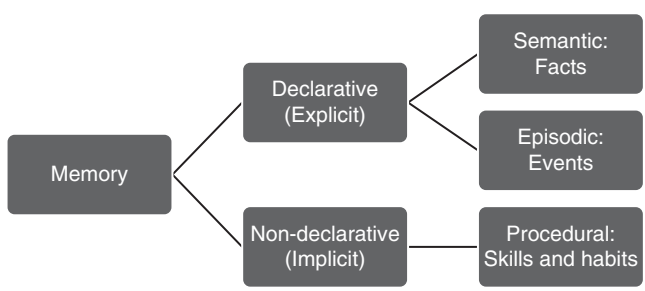

Figure 1.1 Memory systems

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Table 1.2 Memory systems

Memory system	Material	Clinical effects of impairment
Episodic memory	Knowledge of personal (autobiographical) facts and information	Impaired episodic memory, referred to as amnesia, has two components: **anterograde amnesia** (inability to create new memories) suggested by forgetting recent personal or family events; losing items around the home; repetitive questioning or increasing reliance on lists; **retrograde amnesia** (loss of previously created memories) suggested by the loss of memory of past events (jobs, past homes, major news items) or getting lost.
Semantic memory	Refers to the store of general world knowledge, basic facts and vocabulary acquired over a lifetime	Inability to recall facts and concepts, reduced vocabulary and word-finding difficulties

- **Declarative (explicit) memory** is available to conscious access and has **episodic** and **semantic** components (see Table 1.2), but the memory systems are connected.
- The unconscious counterpart of declarative memory is **non-declarative** or implicit memory, one component of which forms our procedural memory. Procedural memory lets us perform some actions automatically (such as writing or riding a bike) without necessarily consciously thinking about it and may be acquired without explicit awareness. Declarative and non-declarative memory functions are consolidated in different areas of the brain.

While memory impairment is a major and early feature of Alzheimer's disease and some other dementias, differentiating these memory systems can sometimes have diagnostic relevance. For example, the disproportionate severity in anterograde episodic memory in contrast to other cognitive domains distinguishes Korsakoff syndrome from other dementias. Selective disorders of semantic memory may arise after viral encephalitis or vascular lesions. Semantic dementia (SD) is a progressive neurodegenerative disorder characterised by loss of semantic memory and may present with word-finding difficulties and other language deficits.

Working memory is a further cognitive system with limited capacity that allows us to retain and manipulate information for a few seconds. This is what, in neuropsychological terms, would be considered 'short-term' memory. It is typically assessed by determining the individual's digit span and is characteristically preserved in Korsakoff syndrome, but may become impaired as Alzheimer's disease progresses in severity. The ability to retain information may also be influenced by *attentional deficits*. Attentional disturbances may leave a person disoriented or easily distracted and can be seen in patients with psychosis or delirium (sometimes called acute confusional state).

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1.7.2 Executive Functions

The frontal lobes are crucial to many aspects of 'higher-order' cognitive functions, including personality. As a corollary, other important areas for cognitive testing are the executive functions which are dependent on the frontal cortex. These are complex cognitive processes necessary for goal-directed behaviour and are commonly impaired early in the course of many dementias. Executive functions allow us to anticipate outcomes, form concepts and adapt to novel situations. Executive *dysfunction* therefore results in deficits in problem-solving, abstraction, reasoning, decision-making and judgment. It can be difficult to characterise these domains clinically and, similarly, it can be a challenge to determine the effects of executive dysfunction on decision-making capacity. Disorders of frontal-lobe function may occur in frontal type dementias, following cerebral infarctions, major closed head injury and other causes of dementia.

1.7.3 Language and Other Functions

Language deficits manifesting during the course of dementia may result in impairments in language comprehension (receptive aphasia), the verbal expression of language (expressive aphasia), difficulties in reading and writing, or otherwise communicating effectively. These can be important considerations as some individuals who manifest with expressive aphasia may experience difficulties in word finding or making themselves clearly understood, but other cognitive processes may be comparatively well preserved. This highlights the importance of utilising appropriate skills and aids to support communication and in providing information for decisions to be made. Listening to the patient and utilising some relatively simple bedside tests will reveal many language deficits. Visuo-spatial dysfunction (which relate to one's perception of and relation with the environment) is common in Alzheimer's disease and may manifest as impaired driving ability, getting lost and difficulty producing drawings of figures.

1.7.4 Cognitive Assessment

A wide variety of clinical measures are available for the evaluation of cognitive and behavioural performance of individuals with suspected dementia. Aspects of cognitive functioning which are assessed typically include orientation, new-learning/memory, language, visuo-spatial and executive function. These measures provide useful information to aid in clinical diagnosis and monitoring of disease progression.

For example, screening tools for cognitive impairment include:

- the Folstein Mini-Mental State Examination (MMSE);[28]
- the Abbreviated Mental Test Score (AMTS);[29]
- the Montreal Cognitive Assessment (MoCA);[30] and
- the Addenbrooke's Cognitive examination revision III (ACE-III)[31] – a further brief screening tool that takes longer to administer than these other tests, but is useful in both identifying and differentiating dementia subtypes.

Neuropsychological assessment is the standardised administration of cognitive tests and the individualised interpretation of the results. This may complement history taking and neuroimaging and can be helpful diagnostically or in contributing to management decisions.

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Note

Although cognitive abilities and decision-making capacity are to some degree correlated, cognitive tests should not be used as a substitute for a specific assessment of decision-making capacity.

1.8 Conclusion

Dementia exacts a huge personal and societal toll, with the numbers of people with dementia predicted to double and the costs to treble. Early diagnosis enables people to gain access to treatment, plan ahead and make important decisions while they are still able and also allows families to receive practical information and guidance. Sadly, diagnosis and post-diagnostic dementia care and support appear to have suffered during the Covid-19 pandemic and its aftermath. In the next chapter, we will look at the range of settings that provide care for people with dementia and to some degree how this correlates with disease severity.

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