Advancing practice in the care of people with dementia

3rd edition

Module 3:

Diagnosing Dementia

Dementia Training Australia

Contents

Module 3: Diagnosing dementia

Introduction

Dementia is a clinical diagnosis and there is no single step to diagnosing the syndrome. Differential diagnoses must be considered and excluded before a diagnosis of dementia can be made. This module discusses the importance of accurate diagnosis as well as the process by which a diagnosis of dementia is determined. The module also includes information about some of the available screening and assessment tools.

Objectives

On successful completion of this module you will be able to:

- Understand the differential diagnoses of dementia
- Understand the importance of differentiating between the various types of dementia
- Explain the importance of early diagnosis of dementia
- Describe the diagnostic criteria for dementia
- Understand the steps involved in diagnosing dementia
- Have an understanding of the various screening instruments and assessment tools which can be applied in the diagnostic process
- Debate issues relating to informing the client and family of the diagnosis
- Understand the importance of referral and follow-up

Module topics

Diagnostic criteria

Differential diagnoses of dementia

- Mild cognitive impairment
- Delirium
- Depression
- Other differential diagnoses

Differential diagnoses of sub-types of dementia

Early diagnosis

Diagnostic tools, steps and tests

Overview of tools and tests

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References

Suggested readings for this module

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http://faculty.ksu.edu.sa/daif/Neurology%20for%20interneal%20medicine/care_dementia_guide%20for%20IM.pdf

Victorian Geriatric Medicine Training Program Dementia Module
http://www.anzsgm.org/vgmtp/Dementia/

Diagnostic criteria

A diagnosis of dementia can only be made if the cognitive impairment is progressive and not due to drugs, a medical condition or delirium.

Until 2013 it was accepted that dementia is diagnosed when other causes of cognitive decline are excluded and there is development of multiple cognitive deficits manifested by **both memory impairment and one or more** of the following:

- Aphasia – or language disturbance
- Apraxia – impairment in carrying out skilled motor activities despite intact motor function
- Agnosia – impaired ability to recognise familiar objects or people despite intact sensory function
- Disturbance in executive function – planning, initiating, organising and abstract reasoning.

(AIHW, 2007, p.22)

However, the fifth edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-V) presented a different approach by replacing the term 'Dementia' with 'Minor Neurocognitive Disorder' and 'Major Neurocognitive Disorder' and proffering new diagnostic criteria.

Minor Neurocognitive Disorder diagnostic criteria

A. *Evidence of modest cognitive decline from a previous level of performance in one or more cognitive domains (complex attention, executive function, learning and memory, language, perceptual-motor, or social cognition) based on:*
    - *Concern of the individual, a knowledgeable informant, or the clinician that there has been a mild decline in cognitive function; and*
    - *A modest impairment in cognitive performance, preferably documented by standardized neuropsychological testing, or, in its absence, another quantified clinical assessment.*

B. *The cognitive deficits do not interfere with capacity for independence in everyday activities (i.e., complex instrumental activities of daily living such as paying bills or managing medications are preserved, but greater effort, compensatory strategies, or accommodation may be required).*

C. *The cognitive deficits do not occur exclusively in the context of a delirium.*

D. *The cognitive deficits are not better explained by another*

mental disorder (e.g., major depressive disorder, schizophrenia).

(American Psychiatric Association [DSM-V], 2013)

Major Neurocognitive Disorder diagnostic criteria

A. *Evidence of significant cognitive decline from a previous level of performance in one or more cognitive domains (complex attention, executive function, learning and memory, language, perceptual-motor, or social cognition) based on:*
    - *Concern of the individual, a knowledgeable informant, or the clinician that there has been a significant decline in cognitive function; and*
    - *A substantial impairment in cognitive performance, preferably documented by standardized neuropsychological testing or, in its absence, another quantified clinical assessment.*

B. *The cognitive deficits interfere with independence in everyday activities (i.e., at a minimum, requiring assistance with complex instrumental activities of daily living such as paying bills or managing medications).*

C. *The cognitive deficits do not occur exclusively in the context of a delirium.*

D. *The cognitive deficits are not better explained by another mental disorder (e.g., major depressive disorder, schizophrenia).*

(American Psychiatric Association [DSM-V], 2013)

Despite this change in terminology it is anticipated that the term "dementia" will continue to be used.

Other published diagnostic criteria include:

- The International Statistical Classification of Diseases (ICD 10) definition of dementia states that there must be:
    - A six-month minimum timeframe
    - Progressive intellectual decline with impaired activities of daily living
    - Multiple cognitive domains affected
    - Memory involvement is not mandatory. (National Centre for Classification in Health, 1998)
- The National Institute of Neurological and Communicative Disorders and Stroke (NINCDS) and Disease and Related Disorders Association (ADRDA – now known as the Alzheimer's Association) Alzheimer's criteria (McKhann et al., 1984). These criteria require that the presence of cognitive impairment and a suspected dementia syndrome be confirmed by neuropsychological testing for a clinical diagnosis of possible or probable Alzheimer's disease, while they need

histopathologic confirmation (microscopic examination of brain tissue) for the definitive diagnosis. As well, they specify eight cognitive domains that may be impaired in Alzheimer's disease.

Criteria for all-cause Dementia

Dementia is diagnosed when cognitive and behavioural symptoms:

- Interfere with work or usual social activities; and
- Represent a decline from prior levels of functioning and performing; and
- Are not explained by delirium or a major psychiatric disorder.

Cognitive impairment is detected and diagnosed through a combination of history-taking from the patient, a knowledgeable informant and an objective cognitive assessment, either a 'bedside' mental status examination or neuropsychological testing, and involves at least two of the following domains:

- Impaired ability to acquire and remember new information – symptoms: repetitive questions or conversations, misplacing personal belongings, forgetting events or appointments, getting lost on a familiar route
- Impaired reasoning and handling of complex tasks, poor judgment – symptoms: poor understanding of safety risks, inability to manage finances, poor decision-making ability, inability to plan complex or sequential activities
- Impaired visual and spatial abilities – symptoms: inability to recognise faces or common objects or to find objects in direct view despite good acuity, inability to operate simple implements or orient clothing to the body
- Impaired language functions (speaking, reading, writing) – symptoms: difficulty thinking of common words while speaking, hesitations, speech, spelling and writing errors
- Changes in personality/usual character, impaired motivation, initiative – symptoms: increasing apathy, loss of drive; social withdrawal, decreased interest in previous activities.

Criteria for the Diagnosis of Alzheimer's Disease Dementia

- Insidious onset. Symptoms have a gradual onset over months to years, and the onset was not sudden over hours or days
- Clear-cut history of worsening of cognition by report or observation
- Cognitive deficits are evident on history and examination in one of the two categories:

1. Amnestic presentation: The most common syndromic presentation of Alzheimer's disease dementia. The deficits should include impairment in learning and recall of recently learnt information. There should also be evidence of cognitive dysfunction in other cognitive domains as defined above.
2. Non-amnestic presentations:
    - Language presentation – the most prominent deficits

are in word-finding, but dysfunction in other cognitive domains should be present.

- Visual presentation – the most prominent deficits are in spatial cognition, including object agnosia, impaired face recognition, simultanagnosia and alexia. Deficits in other cognitive domains should be present.
- Executive dysfunction – the most prominent deficits are in impaired reasoning, judgment and problem solving. Deficits in other cognitive domains should be present. http://www.alz.org/research/diagnostic_criteria/dementia_recommendations.pdf

The steps required to have the above criteria widely accepted and used in practice are publication in a peer-reviewed journal followed by systematic validation through incorporation of the criteria into clinical trials. For further information visit: http://www.alz.org/research/diagnostic_criteria/

Cognitive impairment and dementia

Differential diagnoses of dementia

A differential diagnosis of cognitive impairment is the first step towards diagnosing dementia, as many causes of cognitive impairment are reversible and should be identified or excluded before dementia is diagnosed. Cognitive impairment is often mistakenly labelled as dementia. Further, it is important to be aware that cognitive impairment may be due to a combination of dementia and another disease.

Therefore, always consider and exclude Mild Cognitive Impairment (MCI) in early presentations and differentiate the other big Ds:

- Delirium
- Depressive symptoms have been reported to occur in approximately 40 to 50 per cent of people with Alzheimer's disease (Victorian Department of Health, 2010) and can give rise to some symptoms similar to dementia

Mild cognitive impairment (MCI)

Mild cognitive impairment (MCI) is an important clinical syndrome which has been given at least ten names and presents as cognitive deficits with preserved function.

Given results from a number of studies in the United States as well as results from the Sydney Memory and Ageing Study, there is an increased risk for dementia or Alzheimer's Disease with the rate of progression anywhere between 6 to 25% (Petersen et al., 2001; Tsang et al., 2013).

Further

- There is currently no indication to use cognitive enhancers to treat people with MCI with the goal of delaying or preventing further functional or mobility decline (Odasso et al., 2009)
- It is unclear if MCI is a precursor to developing dementia; therefore, misdiagnosis of MCI as dementia has substantial psychological, social and legal ramifications for the patient and the practitioner
- MCI is a major issue in primary care settings
- The value of any intervention for MCI remains uncertain
- Donepezil: No benefit at three years (Petersen et al., 2005)
- Galantamine: No benefit; in fact there were increased deaths
- Antioxidants, Vitamin E: No benefit

Delirium

Delirium is characterised by a disturbance of consciousness and a change in cognition that develops over a short period of time. The disorder has a tendency to fluctuate during the course of the day, and there is evidence from the history, examination or investigations that the delirium is a direct consequence of a general medical condition, drug withdrawal or intoxication.

(American Psychiatric Association [DSM-V], 2013)

Causes of delirium

- General medical conditions such as hypoxia, fluid or electrolyte imbalance, hepatic or renal disease, systemic infection, thiamine deficiency, post-surgery
- Unrecognised medical illnesses such as hypothyroidism
- Substance-induced; for example, alcohol , digoxin toxicity

Depression

The precise rates of depression and anxiety in older people are not yet known. Estimates of between 10 to 15% of older people experience depression and approximately 10% experience anxiety. Rates of depression among people living in residential aged-care facilities are believed to be much higher at, 52%, according to a report released by AIHW in 2013 and reported by Beyond Blue http://www.beyondblue.org.au/media/media-releases/media-releases/new-report-highlights-the-need-for-depression-awareness-training-for-those-working-with-older-people.

Symptoms of Depression

Common behaviour associated with depression includes:

- General slowing down or restlessness
- Neglect of responsibilities and self-care
- Withdrawing from family and friends
- Decline in day-to-day ability to function, with confusion, worry and agitation
- Inability to find pleasure in any activity
- Difficulty getting motivated in the morning
- Behaviour that is out of character
- Denial of depressive feelings – this can be used as a defence mechanism

(Beyond Blue – Depression in Older People, Fact Sheet No. 17; Hay et al., 1998)

Underlying cognitive status cannot be accurately assessed in the presence of depression.

The table below provides a guide to differentiating between dementia, delirium and depression

Table 3.1: Differentiating between dementia, delirium and depression

Features	Delirium	Depression	Dementia
Onset	Acute	Variable	Insidious
Duration	Hours, days or weeks	Weeks, months, possibly years	Months to years
Course	Fluctuates	Variable	Progressive Progression dependent on type of dementia
Alertness	Fluctuates	Normal	Generally normal

Orientation	Variable: frequently impaired Difficult to engage	Generally normal	Initially normal, deteriorates as dementia progresses
Memory	Variable; often impaired short-term memory	Short-term memory may be impaired	Initially short-term memory impaired then long-term memory as dementia progresses
Thinking	Confused Suspiciousness is common	May be preoccupation with negative ideation Slowing of thoughts may be evident	Often delusions with behavioural and psychological symptoms common
Perception	Hallucinations Misinterpretation of environmental stimuli common	Delusions may be present	Usually normal until later stages
Emotions	Withdrawn Agitated	Flat, unresponsive or sad May be irritable	Usually blunted Irritability as condition advances
Sleeping patterns	Nocturnal confusion	Early morning awakening	Often disturbed Nocturnal wandering and confusion Disorientation to time

Other differential diagnoses

The testing for dementia presumes that the person being assessed has the ability to engage at a certain level of interaction; however, there are states/conditions which may impact on the person's ability to function that must be taken into account. Always consider the person's lifetime level of function before embarking on testing for dementia. Important states/conditions to consider include:

- Low levels of education and literacy
- Superior levels of intellect
- Developmental disability
- Unrecognised medical or psychiatric illness
- Sensory deficits (especially relevant to testing / screening)
- Known presence of mild cognitive impairment
- Culture and language
- The setting in which the test is being conducted.

Differential diagnoses of sub-types of dementia

A formal diagnosis of the sub-type of dementia is important in order to inform treatment and management options. It is especially critical for people who are young, those who may benefit or be harmed by therapeutic interventions and where a detailed prognosis is required.

In future, as better diagnostic tests and therapeutic options increase and become more specific, clinicians will need to be able to diagnose sub-types. Formal specific diagnosis of the sub-types usually requires referral to and evaluation by a specialist service.

It is important to note that:

- Many patients will have a mixed form of dementia, the most common being a combination of vascular dementia and Alzheimer's disease
- A differential diagnosis (DDx) assists in determining the range and extent of investigations required
- The DDx may be divided according to incidence, aetiology, and whether the condition is deemed reversible/treatable

Mixed types of dementia are common. This fact is poorly understood by clinicians and patients.

Early diagnosis

The availability of pharmaceutical treatments and recognition of the need for advanced planning brought about a move towards seeking early diagnosis of dementia. Early diagnosis is possible. Caution is necessary when considering an early diagnosis as there are drawbacks as well as benefits.

The benefits of early diagnosis include:

- Allowing legal and life decisions to be made and advance care plans to be put in place whilst the person is able to actively participate
- Allowing planning for and the implementation of strategies for person and community safety (e.g., implications for driving and other work, lifestyle issues)
- Earlier provision of information and implementation of support services can minimise early stresses on carers and family.

The drawbacks to early diagnosis might include:

- Fear of being labelled and stigmatised, which in turn may increase stress and risk of depression
- Implications for life insurance and or work once a diagnosis has been made
- A risk that early diagnosis of dementia may be a misdiagnosis of mild cognitive impairment (MCI). See information above on MCI.

Evidence suggests that in spite of some reluctance on the part of carers and health professionals, those with dementia want to know their diagnosis.

Early diagnosis in younger people

Specialist multi-disciplinary services such as Cognitive, Dementia and Memory Services (CDAMS) should do the assessment and early diagnosis of younger people.

The assessment and formulation of the diagnosis should not be made in isolation as this may increase the likelihood of misdiagnosis.

It must be noted that diagnosis has significant implications for patient and family. See Module 10, *Younger onset dementia* for more information on the issues facing younger people with dementia.

Diagnostic tools, steps and tests

No single test will diagnose dementia—it is a clinical diagnosis. The ideal clinical approach is to obtain the patient's history directly from the patient and a collateral history from their family, followed by a detailed physical examination, before proceeding to simple non-invasive investigations. The following must be considered:

- Dementia is difficult to diagnose early (ceiling effect of some screening tests; e.g., MMSE)
- Key informant/carer reports are critical in providing evidence of deterioration over time; the first key marker for diagnosis
- A detailed medical history is vital to exclude depression or other conditions
- Physical examination including neurological examination may assist in identifying sub-types of dementia and identifying other diseases (e.g., strokes)
- Pathology tests:
  - Blood tests – full blood count (FBC), electrolytes, urea and creatinine (EUC), liver function tests (LFTs), B12, folate, calcium, thyroid stimulating hormone
  - Urine examination – mid-stream urine sample, microscopy, culture and sensitivity
  - In patients who are at high risk of infections – syphilis serology, HIV testing
- Imaging
  - Computerised tomography (CT) is routine, but in the absence of focal neurological signs is helpful in only a small number of cases
  - Magnetic resonance imaging (MRI) – assessment of medial temporal structures, can be helpful in MCI
- Neuropsychology assessment for complex cases or to support management.

Overview of tools and tests

A wide range of tools and tests exist to assess cognitive impairment in different patient populations. However, they all have their limitations. For a *summary of benefits and limitations of selected cognitive assessment and screening tools*, see Table 2.3, p. 14, AIHW, 2006) http://www.aihw.gov.au/WorkArea/DownloadAsset.aspx?id=6442454183

A recent update of cognitive screening tools authored by Ismail et al.

(2010) and published in the International Journal of Geriatric Psychiatry is available for download at:
http://onlinelibrary.wiley.com/doi/10.1002/gps.2306/abstract

Green et al. (2006). *Towards a measure of function for home and community care services in Australia: Part 2 – Evaluation of the screening tool and assessment instruments*.

It is important to note that these tools are not a diagnostic test and that no assessment tool used in isolation will confirm a diagnosis of dementia.

Dementia assessment tools:

- Are useful for monitoring patient progress in the clinical setting and are used by specialist services to assist diagnosis of dementia sub-types
- Are necessary to access prescribed PBS drugs
- May assist in substantiating a subjective impression for legal requirements such as appointing a medical power of attorney or guardian.

All available tools have limitations. Clinicians using any of the available tools have a responsibility and must be aware of these limitations and how these influence the interpretation of test results.

We recommend that clinicians :

- Be familiar with a small range of tools considered most useful in their area of practice
- Understand the utility and limitations of tools

Especially

- When to administer
- What results mean

Mini-Mental State Examination

Mini-Mental State Examination (MMSE)

The MMSE was developed by Folstein and Folstein in 1975 primarily to assess cognitive functioning in psychiatric patients; it is now the most widely used cognitive screening tool (Folstein et al., 1975).

The instrument is administered by asking a series of questions and giving instructions that enable assessment of orientation, registration, attention and calculation, recall, language and visual construction.

The total possible score is 30 points and a score of less than 23–25 is suggestive of cognitive impairment.

The MMSE has recently been copyrighted by Psychological Assessment Resources (PAR; www.parinc.com) and permission and payment are now required to use the MMSE. Please see the *Tool and Resource Evaluation Template*, adapted by the National Ageing Research Institute (NARI) from an evaluation template created by Melbourne Health, under 'References'.

Advantages of the MMSE

- It is simple and quick to administer; taking ten to fifteen minutes to complete
- It can be used to monitor progression over time and is one of the most commonly used tests
- It is quick and simple test to use in an office setting
- It is required for the prescription of cognitive enhancers on the PBS in Australia

Limitations of the MMSE

- Does not allow for sensory losses
- Incorrect interpretations may create harm
- It is a screening tool not a diagnostic tool
- It does not diagnose dementia
- It requires a minimum level of education
- It is culturally and linguistically specific; as such there is some indication that it is of limited value in culturally and linguistically diverse (CALD) populations
- It is less sensitive than other tests
- Now subject to copyright costs

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General Practitioner test of cognition (GP-cog)

General Practitioner test of cognition (GP- cog)

- One of the most commonly used tests
- Quick and simple test to use
- Involves carers in the assessment
- Not subject to copyright restrictions

A copy of and more information about the GPCog can be found on the GPCog website http://www.gpcog.com.au/info.php

The following reviews of cognitive assessment tools are available at:

Stein et al. (2010). ***The Assessment of Changes in Cognitive Functioning: Reliable Change Indices for Neuropsychological Instruments in the Elderly – A Systematic Review.***

Woodford and George (2007). ***Cognitive assessment in the elderly: a review of clinical methods.***

A number of other instruments may be used in specific circumstances, predominantly by specialists / specialist centres or where educational or sensory limitations are marked (e.g. Clinicians Global Impression).

Rowland universal

Rowland Universal Dementia Assessment Scale (RUDAS)

Developed in Sydney, New South Wales, the RUDAS is suitable for

dementia assessment scale

people from culturally and linguistically diverse (CALD) backgrounds as its reliability does not appear to be affected when translated into languages other than English. It is also not affected by years of education as with the MMSE. The RUDAS is a 6-item test which is easy to administer (Storey, Rowland, Conforti, & Dickson, 2004).

More information on the RUDAS can be found at: http://journals.cambridge.org/action/displayAbstract?fromPage=online&aid=213687&fulltextType=RA&fileId=S1041610204000043

Montreal cognitive assessment

Montreal Cognitive Assessment

The Montreal Cognitive Assessment (MoCA) was designed as a rapid screening instrument for mild cognitive dysfunction (http://www.mocatest.org/) and is available in a number of languages.

Cognitive screening and assessment in Aboriginal and Torres Strait Islander populations

As discussed above, most of the commonly used screening and diagnostic tools are culturally specific and not suitable for culturally and linguistically diverse and Aboriginal and Torres Strait Islander populations–one exception being the RUDAS. Much research is being undertaken to develop culturally appropriate tools.

KICA-cog

Kimberley Indigenous Cognitive Assessment tool (KICA-Cog)

The KICA-Cog is designed specifically to assess cognition in older Indigenous people in the Kimberley region of Western Australia (LoGiudice et al., 2006). The authors suggest the tool could be used in other Indigenous populations in Australia. More information on the KICA-Cog can be found on the KICA-Cog website http://www.wacha.org.au/kica.html

A background paper and guidelines for Screening and diagnostic assessment of non-English speaking people with dementia by Grypma et al. (2007) are available on the Alzheimer's Australia website http://www.alzheimers.org.au/understanding-dementia/culturally-appropriate-dementia-assessment-tools-1.aspx

The decision to disclose the diagnosis

Informing the person and their family

Health professionals are in the position of informing the patient and their family of the diagnosis. A diagnosis of dementia has enormous implications for the person concerned and their family.

It remains essentially a diagnosis of a degenerative condition for which there is no available curative treatment; as such informing the person and the family causes great anxiety to health professionals and carers.

Pinner & Bouman (2003) suggested that in mild dementia 92% (46/50) of patients wish to be informed, 98% (48/50) of carers would wish to

be informed.

Despite recognition of the right of the individual to be informed of the diagnosis there is evidence across a broad range of diagnosis that this is variably delivered. Studies indicate that 30–60% of clinicians across all specialities found disclosure difficult (Wolff et al., 1995). Iliffe et al. (1999) and Rae et al. (2001) noted that health professionals found telling carers only slightly easier than informing patients.

The literature suggests that health professionals and carers are fearful that disclosure of the diagnosis will have a negative impact on the person, although a study by Pinner (2003) did not identify any major incidents in the year following disclosure. This study did find that 6% of patients developed depression within the year post-disclosure. Bamford et al. (2004) suggested that the consequences of disclosure are difficult to measure due to the difficulties involved in differentiating these from the normal course of dementia. The majority of studies report that where carers did not support disclosure initially, most approved of that disclosure once it had occurred, despite some initial difficulty.

Breaking the news

Informing the person and the family requires a sensitive approach. What is said and how to say it are important considerations in sharing results of diagnostic tests with the person tested and their family. No two situations are the same and each situation, as far as is possible, requires careful consideration of the needs of all those who will be affected by the diagnosis. See section on *Current controversies* below, Module 6, *Philosophy of care* and Module 7, *Communication for further information on this topic.*

See Alzheimer's Australia Help sheet *1.3 Diagnosis: Informing the person with dementia* can be found at: http://www.alzheimers.org.au/understanding-dementia/section-1-about-dementia.aspx

Referral and follow up

Dementia is a complex, chronic syndrome and as such planning for care requires an interdisciplinary approach that includes:

- Optimal management of co-morbid conditions to improve quality of life and delay need for relocation to residential care
- Access to services designed for specific patient needs, including a range of specialty clinics (e.g., CADMS, ACAS)
- Consideration of treatment options for any underlying condition or behavioural manifestations
- Incorporation of rapidly changing knowledge in speciality areas
- Contact with Alzheimer's Australia for information, support and education.

Referral and follow-up

- Optimises health and social wellbeing of patient and carers
- Patients want formal diagnosis and prognostic information
- Support groups.

Carers require ongoing support and follow-up as dementia has a high impact on carers—including higher rates of depression—even after the patient has been admitted to residential care.

Resources

The Alzheimer's Australia website has a section on *Understanding Dementia and Memory Loss* – A fact sheet on *Tests used in diagnosing dementia*, Update Sheet No. 8, June 2008 is available at http://www.alzheimers.org.au/understanding-dementia/update-sheets.aspx (Retrieved July 2014.)

Alzheimer's Australia Help Sheet on early diagnosis http://www.fightdementia.org.au/understanding-dementia/early-diagnosis-of-dementia.aspx

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National depression helpline www.beyondblue.org.au

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