

STUDENT GUIDELINES FOR DIAGNOSIS OF DEMENTIA

What is dementia?

Dementia is characterised by a slow, sometimes stepwise, deterioration in the elderly person's mental, physical and social functioning. It can be accurately diagnosed through clinical evaluation, cognitive assessment, basic laboratory evaluation and structural imaging.

Diagnosis of Dementia

Effective diagnosis of dementia involves:

1. History and Functional Assessment
2. Physical Examination
3. Investigations
4. Cognitive Assessment

Aim

When the possibility of dementia has been raised, your role is to

1. Determine a differential diagnosis
2. Eliminate alternative causes for altered cognitive state
3. Consider dementia sub-type
4. Determine the severity and extent of disability
5. Evaluate any co-morbidity
6. Assess family and social support and environment

Importance of Early Diagnosis

The ability to diagnose dementia early is important because:

1. A small subset may be treatable and reversible
2. It is possible to focus on improving lifestyle and reducing risks
3. Symptomatic therapy can help in the early stages of disease
4. It is possible to provide information and support to the carer and family
5. The patient is still able to provide consent and is competent to make decisions regarding their treatment and management

1. Steps in Diagnosis

1.1 History

This should include assessment of:

- full clinical history
- Interviews with patient and family, conducted together and separately if possible
- ability to undertake daily activities

If possible, obtain the history from a reliable informant as well as from the patient. Key points are the areas of cognition affected, especially compared with previous function and skills, and the mode of onset and progression. Vascular risk factors, any recent head trauma and family history of dementia should also be ascertained. Examples of cognitive impairment are useful eg. becoming lost in a familiar area; forgetting how to use familiar household objects or appliance (suggests apraxia); not recognising objects for what they are (suggests agnosia); not being able to plan and prepare a meal (suggests a disorder of executive function).

Speaking with a family member may be necessary to ascertain whether the memory complaint represents a consistent change from the previous level of function. A family member will also be able to determine whether there has been any early personality or behavioural changes. Functional

impairment and the patient's ability to perform activities of daily living are also best assessed by speaking to the family. This interview is also an opportunity to get an idea about family support systems, which may prove crucial in the future management of the patient.

1.2 Physical Examination

A complete and thorough physical examination should be conducted. This should be directed towards finding evidence for:

- specific conditions which may cause dementia (stroke, Parkinson's disease, cerebrovascular disease, hypothyroidism)
- Underlying chronic conditions which may aggravate dementia (hypertension, cardiac failure, renal failure, diabetes, amnesia)
- conditions which may cause delirium (respiratory or renal infection)

It is also important to assess the patient's level of consciousness as, if impaired, this may be suggestive of delirium.

1.3 Investigations

Potentially reversible or partly reversible, causes of dementia do exist and it is important to investigate these in the diagnostic process. Such investigations can also aid in ruling out other co-existing pathology.

1.3.1 Laboratory tests

- FBP (to rule out anaemia)
- U&E
- TFT (to rule out hyperthyroidism)
- B12 and folate levels (low levels may impair cognitive function)
- Syphilis serology (only if specific indicators)
- ESR (to rule out hyponatremia)
- Urinalysis (WBC, protein, sugar). Culture if delirium.
- Calcium (to rule out hypercalcaemia)
- BSL (to rule out hyperglycemia)
- +/- lumbar puncture
- EEG
- CXR (if delirium)

1.3.2 Imaging

The use of structural imaging tests such as CT or MRI, can rule out other causes of cognitive decline such as subdural haematoma, normal pressure hydrocephalus or neoplasm. These tests may be normal in people with dementia, may show focal or general atrophy or may show other relevant abnormalities.

CT or MRI is recommended if one or more of the following criteria are present:

- Age < 60 years
- Rapid unexplained decline in cognition or function
- Short duration of dementia (< 2 years)
- Recent significant head trauma
- Unexplained neurologic symptoms (eg., new onset of severe headaches or seizures)
- History of cancer (especially types that metastasise to the brain)
- Use of anticoagulants or history of bleeding disorder
- History of urinary incontinence and gait disorder early in the course of dementia (as may be found in normal pressure hydrocephalus)
- Any new localising sign (eg., hemiparesis or a Babinski reflex)
- Unusual or atypical cognitive symptoms or presentation (eg., progressive aphasia)
- Gait disturbance

Functional imaging such as SPECT or PET can be helpful in investigating early or atypical cases

Both laboratory tests and structural imaging can rule out treatable causes of dementia and also help determine dementia sub-type.

Adapted from Eastly R., Assessment of Dementia, in *Dementia*, Eds Burns, O'Brien and Ames.
and
Watson JD. 2001. Disorders of Memory and Intellect. *Medical Journal of Australia*. 175; 433-437.

1.4 Cognitive Assessment

The Mini-Mental State Examination (MMSE), Clock Drawing Test and similar short instruments are screening tests only. With MMSE, the lower the score the more likely it is that dementia is present. The opposite is true for the Clock Drawing Test. Early dementia is sometimes not picked up by MMSE, but with addition of the Clock Drawing Test, detection is improved.

MMSE

- is well known for reliability and validity
- does not measure some of the early signs of dementia

Clock Drawing Test

- a valid and reliable measure of Executive Function Deficit (cognitive processes used to carry out everyday tasks requiring a correct sequence of events and self-monitoring behaviour)
- EFD may precede or be concurrent with short term memory difficulties, which are often assumed to be the primary indicator of early dementia.
- EFD is included in the DSM IV diagnostic criteria for dementia

It is ideal to conduct both tests in conjunction as this offers the most efficient assessment of early dementia.

When conducting early cognitive tests it is important to take into account other factors that may affect performance, including educational level, skills, prior level of functioning and attainment, language barriers, advanced age, sensory impairment, cultural factors, psychiatric illness and physical or neurological problems. These issues should be considered when interpreting the results.

Neuropsychological Tests

Dementia present for some time may not need specialised neuropsychological assessment. However, formal neuropsychological testing is valuable early in the course of dementia, or if there are unusual features, with dementia in a younger person, or where there is the possibility of depression or similar.

2. Aims of Diagnosis

2.1. Establish a Differential Diagnosis

Using the available evidence from your assessment, dementia must be distinguished from other conditions, particularly normal ageing, delirium, depression and drug effects. The table below gives markers for differentiating between dementia, delirium and depression.

Feature	Delirium	Dementia	Depression
Onset	Acute	Insidious	Acute or subacute
Time Course	Fluctuates during single day	Worsens over lengthy time; cognitive loss precedes depression; behaviour worse at night	Depression precedes cognitive loss
Duration	Hours to weeks	Months to years	Weeks to months; often prior history
Attention	Impaired	Little affected; impaired in later stages	Unimpaired
Orientation	Usually impaired in time, often place, sometimes person	Impaired in later stages	Unimpaired
Alertness	Abnormal: low or high	Normal, at least initially	Normal or low
Consistency of performance	Variable	Consistent	Variable and slow
Sleep-wake cycle	Always affected	Affected in later stages	Difficulty sleeping, early morning wakening
Short-term memory	Always impaired	Not affected initially	Poor effort may lead to variable impairment
Long-term episodic memory	Impaired	Impaired; recent loss exceeds remote	Equal recent and remote loss
Perception	Often visual illusions and hallucinations	Commonly impaired in later stages	Normal, unless psychotic depression
Cognitive Loss	Unaware or denied	Minimised by patient	Maximised by patient
Speech and thought	Disorganised, deluded, slow or rapid	Word-finding difficulty, thoughts of death and dying	Suicidal thoughts
Answers given	Rambling, impersistent	"Near miss" frequent	"Don't know" frequent

Partly adapted from Feinberg and Goodman, 1984.

2.2. Eliminate other causes for cognitive impairment

Based on the results of the investigations, you should be able to eliminate other potential causes of cognitive impairment, establish a differential diagnosis and establish dementia sub-type.

2.3. Establishing Dementia Sub-Type

2.3.1. Alzheimer's Disease

DSM-IV Diagnostic Criteria For Alzheimer's Disease (1994)

- Multiple Cognitive Deficits
- Short and long term memory impairment
- One or more of the following:
 - Aphasia (language difficulty, eg. finding the right words)
 - Apraxia (difficulty performing simple tasks)
 - Agnosia (trouble recognising ordinary objects)
 - Disturbance of executive functioning (abstraction, judgement, reasoning, insight. Planning, initiating, sequencing, monitoring and stopping complex behaviour)
- Gradual onset, functional impairment, continuous decline
- Exclusion of other possible causes
- No evidence for delirium

Clinical course of Alzheimer's Disease

- Slow, progressive disease
- Mild (MMSE 21-30), Moderate (MMSE 11-20), Severe (MMSE 10 or below)
- Pathology begins years before any symptoms or functional compromise manifests
- Characterised in early stages by memory impairment, first personality changes, normal gait and posture, anomia, impaired abstraction, behaviour indifferent or delusional
- Characterised in later stages by evidence of global cognitive impairment, gait impairment, agitated behaviour, stupor or delusional states.

2.3.2. Vascular Dementia

- Cognitive impairment is due to cerebrovascular disease
- Evidence of vascular changes on CT/MRI
- Vascular risk factors present – HTN, cholesterol, diabetes, smoking, existing cardiac disease
- Typically evolves in a stepwise fashion but can also progress insidiously
- Often early onset gait disturbance, falls and urinary incontinence
- Often frontal lobe features, such as emotional lability, pseudobulbar palsy with speech/swallowing difficulties
- Focal neurologic findings frequently found in the early disease course

2.3.3. Lewy Body Disease

- Lewy Bodies evident (neuronal inclusions of neurofilament protein (alpha synuclein))
- Found in basal ganglia in Pick's Disease and diffusely through cerebral cortex in Diffuse Lewy Body Disease.
- Classical features of cognitive impairment plus
 - fluctuation in cognition, alertness and attention
 - Visual hallucinations
 - Parkinsonianism
- Responds to cholinesterase inhibitors
- Very sensitive to antipsychotics (causing rigidity)
- Disease course fluctuates

2.3.4. Fronto-temporal Dementia

- Degeneration on one or both of the frontal or temporal lobes of the brain
- Includes progressive aphasia, semantic dementia and Pick's Disease
- Early loss of personal and social awareness (early personality and behavioural changes)
- Disinhibition often prominent
- Mental rigidity, inflexibility, "concrete"
- Depression and anxiety prominent

- Speech and language disturbance
 - reduced in complexity
 - echolalia, stereotypy
- Early primitive reflexes and urinary incontinence
- Late rigidity, tremor
- Often younger age of onset

(Adapted from Heck J., Dementia and Alzheimer's Disease in *A Practical Guide to Geriatric Medicine*. Ratnaik R.

2.4. Determine severity and extent of disability

Functional status should be assessed in terms of the patient's ability to perform activities of daily living; personal safety; communication ability; nutrition, hygiene and medications; ability to drive; and legal capacity for decision making. Specific issues that need to be considered include:

- safety issues in the home and on the road
- personal hygiene
- financial competency
- self-monitoring of medications
- ability to attend to adequate nutrition
- present legal and future capacity regarding advanced care directives, Enduring Guardianship or Enduring Power of Attorney.

Legal Capacity for Decision Making

Determination of a patient's capacity to make decisions may be an important role of the doctor.

This may apply in one of three situations:

- consent for medical treatment
- giving an advance care directive
- making a will

It may also apply to other tasks such as managing financial affairs or arranging living circumstances.

2.5. Manage Co-morbidity

It is important to correctly identify any existing co-morbidities in order that they may be adequately treated to avoid physical decline. Conditions which may aggravate dementia, such as cardiac or renal failure, nutritional deficiencies and visual and hearing impairments should also be given special attention. In addition to diagnosing and treating current co-morbidities, it is equally important to consider preventing future illness, by ensuring the patient is adequately assessed for functional status and has access to necessary care and support services.

2.6. Assess Family and social support and environment

You should assess carer and family stress and support. Your on-going assessment of the needs of the carer and level of support required is an essential component in your role of the management of the person with dementia. Carer support is as important as patient support.

The stress associated with caring for a person with dementia should never be underestimated. It places an extraordinary burden on those who undertake the caring role. Carers are often elderly, or stressed by other family responsibilities. Higher levels of depression, psychological morbidity and use of psychotropic medications are seen in carers of those with dementia. Difficulties experienced with caring can be enough to produce sufficient stress to place either the person with dementia or the carer at risk, or jeopardise the success of community care.

An assessment of the person's home environment is also important. The home should be assessed for safety of floor coverings, cooking facilities, bathroom, toxic substance storage and heating. The home assessment can also include measuring the person's ability to function safely and optimally within this environment, or whether they will need help performing specific activities of daily living.

Care plans should address activities of daily living (ADL) that maximise independent activity, enhance function, adapt and develop skills, and minimise need for support. They should also address the varying needs of people with different types of dementia. The aim is to support people with dementia so that they may remain living in the community for as long as possible.

3. Communicating a diagnosis of dementia

3.1. What to communicate regarding the diagnosis

- what the diagnosis is, and its prognosis
- how this may affect the person's personality, behaviour and functioning
- when and how to ask for help
- what services are available and how to access them
- legal and financial matters, eg enduring power of attorney, operation of bank accounts
- emotional support systems available
- support and respite care available
- financial assistance available
- how to deal with challenging behaviours and difficult issues such as giving up driving
- residential care options and how to access and evaluate these
- Enduring Power of Attorney or Guardianship
- making of will.

3.2. How to communicate the diagnosis

Listed below are ways to help minimise the distress that breaking the news of dementia may cause:

- Allow adequate time and ensure privacy
- Let the patient decide how much they want to know
- Tell the patient and carer separately, if necessary
- Be empathetic and encourage expressions of feelings
- Break the news in stages over several consultations
- Assess patient's understanding frequently
- Be aware that both patients and carers may suffer reactive depression or anxiety after hearing the diagnosis
- It is perfectly acceptable to refer the patient to a specialist to hear the diagnosis if you feel that passing on the diagnosis will damage your relationship with the patient and/or family

Patients and families should be encouraged to contact the Alzheimer's Australia, which can provide information and support.

4. Management

It is important to explain to the patient and family the features of the dementia prognosis, and to discuss the major cognitive problems. Review of medications that may worsen matters is needed in addition to advice on minimising alcohol intake. Treatment of co-existing conditions, such as depression or poor hearing can help.

Management of behavioural and psychiatric manifestations can be challenging, especially agitation and wandering. Simple behavioural techniques, environmental manipulation and a strongly structured routine may help.

As more basic activities of daily living are lost, the level of care needed increases, eventually requiring nursing home care. This can be discussed early with the patient and family to identify options and costs.

Early contact with Alzheimer's Association and discussion of financial, estate and guardianship issues is important while consent and competency are still intact.

Counselling and support for carers is important at all stages.

The following is a list of recommended actions to assist patients with mild to moderate dementia and their families after a diagnosis has been made. You are not expected to be able to complete such a detailed management plan at this stage of your medical training, but it is important for you to be aware of the necessary steps as it is likely you will need to manage people with dementia as a medical graduate.

1. Inform the patient and their family of the diagnosis
2. Identify what support the primary caregiver can offer and determine caregiver needs – assess regularly
3. Decide on the need for referral for further diagnostic and management assistance - assess regularly
4. Assess for safety risks (eg. driving, financial management, medication management, home safety risks or potentially dangerous behaviours) - assess regularly
5. Determine the presence of any advance planning documents (eg. will, enduring power of attorney, personal directive). If none exist, advise that they be drafted.
6. Assess the patient's decision making capacity - assess regularly
7. Refer the patient and family to local Alzheimer's Australia branch.
8. Provide information and advice about pharmacologic and non-pharmacologic treatment options
9. Develop and implement a treatment plan with defined goals - assess and update regularly
10. Monitor response to initiated therapy
11. Monitor and manage functional problems (eg., incontinence) as they arise
12. Assess and manage behavioural and psychological symptoms of dementia as they arise
13. Monitor nutritional status and intervene as needed
14. Deal with medical conditions and provide on-going medical care
15. Mobilise community based and facility based resources as needed

Encourage caregivers to participate in caregiver educational programs and support groups.

Diversity and Equality

- Always treat people with dementia and their carers with respect.
- Ensure people with dementia are not excluded from services because of their diagnosis, age, or any learning disability.
- If there is a language barrier, offer:
 - written information in the preferred language and/or an accessible format
 - independent interpreters
 - psychological interventions in the preferred language.
- Ensure that people suspected of having dementia because of cognitive and functional deterioration, but who do not have sufficient memory impairment for diagnosis, are not denied access to support services.

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