



Unit 494 May 2013

# Dementia







## **Dementia**

Unit 494 May 2013

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The five domains of general practice **©** Communication skills and the patient–doctor relationship

- Applied professional knowledge and skills Population health and the context of general practice
- Professional and ethical role 44 Organisational and legal dimensions



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FROM THE EDITORS check Dementia

A diagnosis of dementia can be devastating for the patient and their family. Dementia is the second most feared disease, behind cancer. We fear no longer being in control of our mind. As well as this fear there is, unfortunately, a stigma attached to the diagnosis of dementia. While many people consider it a natural part of ageing, it is in fact a chronic disease process.

An early diagnosis of dementia has many benefits. It can potentially delay progress of the disease and treat reversible causes. Support systems can be put into place early, and planning can be organised before the patient's competency is compromised.

Dementia is a progressive disease that requires a multidisciplinary approach. It involves not only the patient but their family and carers as well. Comorbidities such as hypertension and diabetes need to be managed, and the health of carers needs to be monitored. The family GP is ideally placed to provide this support.

Alzheimer's Australia coordinated the original manuscripts, and further information on early intervention for people with dementia can be found at www.detectearly.org.au.

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We would also like to thank Henry Brodaty, Diana Fayle, Carmelle Peisah and Lee-Fay Low for their valuable contributions to this issue of *check*.

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### The learning objectives of this unit are to:

- display increased knowledge of the different types of dementias and their presentations
- understand the importance of treating comorbidities in patients diagnosed with dementia to improve health outcomes and patient wellbeing
- become familiar with cognitive testing tools that can be used in culturally and linguistically diverse (CALD) populations
- identify the difference between delirium and dementia
- display an increased understanding of the testamentary issues involved with dementia
- advise patients of protective lifestyle factors such as exercise and mental stimulation, to decrease the rate of disease progression.

We hope you find this edition of *check* useful in the diagnosis and management of dementia.

Kind regards

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GUIDE TO	GUIDE TO ABBREVIATIONS AND ACRONYMS IN THIS UNIT OF CHECK				
ACAT ACD AD APOE £4 AMTS BADL BP BPSD bvFTD CALD CASS CT CXR DBMAS	Aged Care Assessment Team advanced care directive Alzheimer disease	DSM IV  ECG eGFR EUC FBE	Diagnostic and Statistical Manual of Mental Disorders IV electrocardiogram estimated glomerular filtration rate urea and creatinine full blood count fluorodeoxyglucose positron emission tomography frontotemporal dementia frontotemporal lobar degeneration General Practitioner assessment of Cognition Kimberley Indigenous Cognitive Assessment tool Lewy body dementia liver function test mild cognitive impairment Mini-Mental State Examination magnetic resonance imaging	MSU NDD PBS PET RBG REM RSBD RUDAS SPECT TC TFT UTI VaD WACHA	midstream urine neurodegenerative disease Pharmaceutical Benefits Scheme positron emission tomography random blood glucose rapid eye movement REM sleep behaviour disorder Rowland Universal Dementia Assessment Scale single photon emission computed tomography testamentary capacity thyroid function test urinary tract infection vascular dementia Western Australian Centre for Health and Ageing
		TVII II	-magnotio robonance imaging		

### CASE 1

# DAISY IS FORGETTING HOW TO COOK HER FAVOURITE MEALS

Daisy, aged 78 years, migrated from Hong Kong 10 years ago with her husband Wu under Family Reunion status to be with their two children in Sydney. They both live with their married daughter Ling in her home, and speak Chinese and limited English.

Daisy assists Ling, who runs a small business, by doing the household chores, shopping and looking after her three teenage grandchildren. Daisy's son lives in another part of town. He is not always around, but he and his family keep in touch regularly.

Ling brings her mother to see you for regular checkups because Daisy has moderate hypertension and has recently developed type 2 diabetes. You manage both these conditions with oral medication. Daisy also takes ginseng tea, Ginkgo biloba and a Chinese medicinal tea for general wellbeing.

Daisy occasionally attends a social club to meet with other Chinese friends for lunch, but she is not otherwise physically active.

Recently, Ling has noticed that Daisy is more anxious, is making mistakes when she cooks her favourite meals and is becoming more forgetful, particularly when doing more than one preparation task for dinner. Ling is concerned about her mother's memory and has brought her to see you with a specific request to review this.

QUESTION 1 ( )
What are the key features in your history and examination of Daisy?
QUESTION 2 (C)
What is the most likely diagnosis?
QUESTION 3 (C)
What criteria need to be satisfied in order to make a diagnosis of dementia?

CASE 1 check Dementia

### **FURTHER INFORMATION**

On examination, Daisy's weight is stable and her neurological examination is normal. Her blood pressure (BP) is 130/70 mmHg, her pulse rate is 72 beats per minute and her random blood glucose (RBG) is 5.5 mmol/L.

You use the online Chinese version of the General Practitioner assessment of Cognition (GPCOG), with the assistance of Ling. Daisy scores 5/9. This assessment only takes a few minutes to complete.1

You request that Daisy returns a week later with Ling and Wu to discuss the results of investigations you order today. You ask Daisy to complete a Geriatric Depression Scale form<sup>2</sup> before her next visit to exclude depression.

Investigations show haemoglobin of 13.2 g/L with a normal full blood count (FBE). Daisy's urea is 6.1 mmol/L (3-8.00 mmol/L) and her estimated glomerular filtration rate (eGFR) is 65 (>60 mL/min/1.72 m<sup>2</sup>). All other electrolytes are normal. There is no evidence of a urinary tract infection (UTI) and Daisy has a normal chest X-ray (CXR) and a normal electrocardiogram (ECG).

### QUESTION 4 (1) (2)

With Daisy's permission, what further office-based assessment tool
could you use to evaluate her memory in the presence of Wu and Ling?

### **FURTHER INFORMATION**

You explain that Daisy has some form of memory loss or mild cognitive impairment, or even early dementia, and that you feel a computerised tomography (CT) scan of Daisy's brain would be useful. You advise Daisy's family that she may need to be referred to a memory clinic.

Ling is relieved by this explanation, but Wu is initially adamant that there is nothing wrong with his wife. He doesn't think Daisy needs any further testing and he says he will watch over her when she cooks. You explain that further testing may help Daisy and referral to a memory clinic could assist with providing medication, support and advice for Daisy and her family.

### QUESTION 5 (1)

What factors	can affect th	e diagnosi	s of demer	ntia?	

### **FURTHER INFORMATION**

Wu is reassured by your explanation and has agreed to Daisy having a CT scan of her brain, as well as being referred to a memory clinic.

The CT scan shows generalised global atrophy with small lacunar infarcts consistent with vascular dementia (VaD).

### DIJECTION C O TO CO







Vhat are you	ur expectations of Daisy's memory clinic visit?

What else can be done to improve Daisy's health?

### **CASE 1 ANSWERS**

### **ANSWER 1**

It is important to take a complete medical and family history.

Specifically ask about forgetfulness, orientation, problem solving, coping with everyday life, mood, alcohol consumption, depression and new physical symptoms.

Ask about a time frame. When were the changes first noticed? Were they sudden or gradual? Are the difficulties getting worse?<sup>3</sup>

With Daisy's consent, ask other family members what they have noticed. Ask them to complete the 'informants questionnaire' from the General Practitioner assessment of Cognition (GPCOG). It is a useful resource for GPs that is available in several languages, including Chinese.

Conduct a general and neurological examination.

### **ANSWER 2**

The most likely diagnosis is mild cognitive impairment or early dementia requiring further investigation.

It is also important to consider that Daisy may not have dementia. It is important to rule out renal failure, chronic low-grade infection (e.g. UTI), depression, drugs, traditional Chinese medicines, cerebrovascular disease and other cerebral causes (e.g. tumour).

### **ANSWER 3**

For a diagnosis of dementia there must be multiple cognitive deficits that commonly, but not always, involve memory loss. In some forms of dementia, memory may be preserved in the early stages.

- As well as having some form of cognitive impairment, there must also be impairment in one of the following domains.<sup>4</sup>
  - Aphasia: problems with language. This is often heard during the
    history more than elicited directly as a sign (e.g. being able to
    identify a watch but being unable to describe what it does, such
    as tell the time).
  - Agnosia: failure to recognise what objects are used for. Failure to recognise family members or well-known friends is a late feature of agnosia. This is not so much struggling to remember a name as it is struggling to recall that the person is known at all.
  - Apraxia: inability to carry out purposeful movements even though there is no sensory or motor impairment. This can present as difficulty with dressing in which there is a loss of detail.
  - Executive dysfunction: impaired planning, sequential organisation and attention. This may present as poor initiation, difficulty with problem solving and a reluctance to change routines.

Cognitive deficits must be severe enough to interfere with occupational and/or social functioning. They must also represent a decline from a previously higher level of function, as well as not occurring exclusively during the course of delirium.<sup>4</sup>

There needs to be a variation from the normal 'score' in one of the following standard assessment tools, all of which have Chinese translations available.

- Mini-Mental State Examination (MMSE)<sup>5</sup>
- Rowland Universal Dementia Assessment Scale (RUDAS)<sup>5,6</sup>
- GPCOG

In this particular case, RUDAS would be the tool of choice because it is particularly appropriate for patients who are from CALD backgrounds (see *Resources*).

Investigations to include as part of a dementia screen are: FBE, electrolytes, urea and creatinine (EUC), liver function tests (LFTs), calcium, RBG, thyroid function tests (TFTs), B12 and folate levels, midstream urine (MSU), ECG, CT brain and CXR.<sup>4</sup>

### **ANSWER 4**

A significant proportion of elderly people in Australia are born overseas with English as their second language. Standard assessment tools, such as the MMSE, are not easy to translate and administer to this demographic. Culturally appropriate dementia assessment tools are now available and include RUDAS and the Kimberley Indigenous Cognitive Assessment tool (KICA) for the rural and remote Indigenous population.<sup>7,8</sup> Daisy's RUDAS score is 23/30.

### **ANSWER 5**

There are several barriers to the diagnosis of dementia, which include differentiating between normal ageing and dementia, a perceived lack of need to make a specific diagnosis and the lack of perceived treatment options. There can be a negative stigma attached to a diagnosis of dementia and the patient may have impaired ability, which can hinder an accurate history and participation in self care. There may also be denial by the patient or their family.<sup>3</sup>

There are many benefits to making an early diagnosis of dementia. These include treating any reversible causes and modifying risk factors. Education and safety issues can be addressed, and legal planning can be undertaken while the patient is still competent. The diagnosis is often a relief to carers. They can be supported in their role and their own health needs can be addressed by their GP.

CASE 1 check Dementia

### **ANSWER 6**

Your expectation of Daisy's memory clinic visit should be to confirm the diagnosis of dementia, and to provide an overall management plan for Daisy. This would facilitate her staying at home for as long as possible.

The memory clinic can reassess Daisy's memory at a later stage, as well as referring her to a local geriatrician.

Daisy's family should be provided with education about dementia and ongoing support in caring for her.

### **ANSWER 7**

Daisy's diabetes and hypertension need to be controlled and monitored regularly.

You can encourage Daisy to increase her exercise by walking or doing resistance exercise. Regular resistance exercise may help reduce her anxiety and improve her overall wellbeing. It has the added benefit that it can be done with her husband Wu.<sup>9,10</sup>

You can also discuss support services such as Alzheimer's Australia, (which offers resources for families and carers of culturally diverse backgrounds), the Chinese Australian Services Society (CASS) and the Dementia Behaviour Management Advisory Service (DBMAS) (see *Resources*).

An assessment from the Aged Care Assessment Team (ACAT) may also be helpful.

### CASE 2

### HARRY HAS BECOME MORE FORGETFUL

Harry, aged 75 years, is brought to see you by his wife Doris, who reports a 6-month history of forgetfulness. This includes Harry leaving taps running in the bathroom and losing his wallet and keys. Harry now has difficulty balancing his chequebook and has lost interest in watching sport on television. Harry is an irregular attendee at your practice. He has a history of hypertension and tends to be noncompliant with his medication. Doris has to consistently remind him to take his medication, which he resists at times.

On examination Harry's BP is 170/90 mmHg. After a thorough assessment you perform an MMSE. Harry's MMSE score is 21/30. You make a provisional diagnosis of dementia.

QUESTION 3 🔱		
What drugs are available for the treatment of AD?		
QUESTION 4 🖎		
What are the indications for prescribing an acetyl cholinesterase inhibitor? What are the contraindications and the possible side effects?		

### QUESTION 1 (1)



What are the main causes of cognitive decline in the elderly?

QUESTION 2 ( ) ( )

When would you refer patients with suspected dementia to a specialist or memory clinic?

### **FURTHER INFORMATION**

You recommend that Harry be seen by an appropriate specialist or attends a memory clinic. He is assessed and a diagnosis of mixed Alzheimer disease (AD) and VaD is made. Harry is given a prescription for an acetyl cholinesterase inhibitor.

### **FURTHER INFORMATION**

Doris asks what she can do in terms of slowing the progression of Harry's dementia, and asks specifically about activities to keep Harry healthy. She also asks about the expected progression of the disease, and is concerned about what will happen if and when Harry deteriorates and needs more care than she can offer.

### QUESTION 5 ( )



What are the key features in the management of Harry's dementia?

### 

What is the role of exercise and recreational pursuits in dementia care, and what activities would you recommend for Harry?

CASE 2 check Dementia

### **CASE 2 ANSWERS**

### **ANSWER 1**

The main causes of cognitive decline in the elderly include the four Ds – delirium, depression, dementia and drugs. It can also be caused by normal ageing as well as mild cognitive impairment (MCI). <sup>11,12</sup>

### **ANSWER 2**

Referral to a specialist is indicated if you are unsure of the diagnosis, or if the patient is relatively young. You would refer if multiple complex comorbidities were present and if the presentation was atypical. A psychotic episode or severe behavioural disturbance would also be indications for referral.

### **ANSWER 3**

There are currently four drugs approved in Australia for the treatment of AD to slow cognitive decline. Donepezil (Aricept®), rivastigmine (Exelon®) and galantamine (e.g. Reminyl®) are cholinesterase inhibitors that make more of the neurotransmitter acetylcholine available at brain synapses and help to enhance memory function. Memantine (e.g. Ebixa®) acts on the neurotransmitter glutamate and can reduce the rate of symptom progression in the middle and later stages of AD.³ A diagnosis of AD must be confirmed or made in consultation with a specialist for the first 6 months of treatment with a cholinesterase inhibitor¹2.¹³ and/or memantine. These drugs can reduce the rate of decline of cognitive function for a period of time, but the effect tends to wear off.

### **ANSWER 4**

Cholinesterase inhibitors such as donepezil, rivastigmine and galantamine can be prescribed on an authority prescription under the Pharmaceutical Benefits Scheme (PBS). A diagnosis of AD needs to be made and confirmed by a specialist and cognitive testing results with the MMSE must be greater than 10/30. Memantine has similar restrictions, but the MMSE needs to be between 10–14/30. Memantine can also be used with cholinesterase inhibitors, but is only subsidised as monotherapy. However, GPs can prescribe these drugs with no prior testing if the patient wants to pay for the nonsubsidised medication. <sup>13</sup> For patients with a diagnosis of AD, the restrictions and re-assessment process after the initial 6 months of treatment have been removed, as of 1 May 2013, for the use of those medications subsidised by the PBS.

The relative contraindications to cholinesterase inhibitors include bradyarrhythmias, active peptic ulcer disease and severe asthma.

Common side effects of cholinesterase inhibitors include gastrointestinal disturbances such as anorexia, nausea, diarrhoea and vomiting. Other side effects include the urge to defaecate or urinate, sleep disturbances, nightmares and muscle cramps. There is also an increased risk of epileptic fits.

### **ANSWER 5**

While there is currently no cure for dementia<sup>3</sup> there is much that can be done to maintain Harry's health and wellbeing, as well as delay his cognitive decline. It is important to reduce his cardiovascular risk factors, in particular his hypertension, <sup>14,15</sup> as well as managing any comorbidities such as hyperlipidaemia and diabetes. In addition to the medication he has been prescribed to delay cognitive decline, it is important to promote exercise, socialisation and some form of cognitive stimulation.

Preventative management includes immunisation and ensuring adequate hygiene, rest/sleep, hydration, nutrition and dental care.

Psychosocial treatments and support such as attention to safety, activities of daily living, mental activity and stimulation, and medication supervision are important.

Carer education and support are essential. This may include a referral to a support organisation such as Alzheimer's Australia that can offer resources for Harry and his family. <sup>16</sup> As his functional impairment increases, Doris may need to be directed to support services in the community to help with his care.

Harry will need to be assessed for his fitness to drive and legal issues will need to be raised while he is still competent. These include having an up-to-date will, an enduring power of attorney (for financial matters) and appointing an enduring guardian for health decisions, as well as completing advanced care directives (s).<sup>17</sup>

It is also important to consider Doris' health, because the stress and added burden of care at an older age can affect her overall health and mental wellbeing.

### **ANSWER 6**

Exercise has a positive effect on memory and health. 9,10,18 Consider sports interests and activities that were part of Harry's premorbid lifestyle (e.g. Harry was a keen lawn bowler and gave it up because he could not remember the score or the names of visiting bowlers). Encourage Harry to resume this activity with another member who can act as his 'minder' by keeping the score and knowing the names of the other players.

You may like to suggest that Harry and Doris join a local community exercise group. This may include activities such as hydrotherapy, walking and strength training.

Encourage Harry to be physically active, and encourage ongoing mental stimulation (e.g. choirs, art classes or joining a men's shed group). You may also suggest that Harry and Doris join a seniors' group for ongoing social support, group activities and outings.

### CASE 3

### ANNE IS GETTING MORE AGGRESSIVE

Anne, aged 80 years, presents at your surgery with her daughter Jane, who is worried that her mother is becoming more aggressive. Jane tells you that Anne is becoming irritable and shouts at her and her children for no reason. Anne is also wandering around at night and waking the family.

Anne was diagnosed with AD 3 years ago. Until then, she had lived in her own apartment, even after the death of her husband Bruce. After her diagnosis she moved into a purpose-built 'granny flat' attached to Jane's home.

Jane is married and has two teenage children. Jane brought Anne to the initial consultation with you 3 years ago, as she had been concerned about her becoming more forgetful. Jane has attended all subsequent consultations with Anne, both at your practice and at the memory clinic.

When Anne was diagnosed with AD, Jane expressed relief - 'Finally we have an answer. I knew something was wrong with mum.'

At the time of diagnosis, Jane discussed with you her plan to care for Anne for as long as possible.

Jane asked you how AD progresses and what features might present at a later stage, as well as asking about a time frame for the development of these features.

### **FURTHER INFORMATION**

QUESTION 3 (1)

Jane is aware that her role as Anne's carer will, at various stages, include many different responsibilities. She knows she will be responsible for managing medications, therapies and medical emergencies. She will also provide supervision and emotional support, and assist with personal care, mobility and household tasks.

At this consultation with you, Jane said she is concerned that she may not be coping as her mother's condition progresses.

What are the issues that may affect carers as a result of their role?

QUESTION 4 👄
What issues do carers generally feel they would like GPs to manage
QUESTION 5 👄
What tool can be used to assess the caregiver burden?

### QUESTION 1 (1)



What were you able to tell Anne and Jane about the symptom progression in AD?

### QUESTION 2 ( )



What were the safety features that Jane needed to consider in terms of the physical environment inside the home for Anne?

CASE 3 check Dementia

QUESTION 6 (L)
What term is used collectively to describe irritability, aggression and wandering as they present in dementia? List other features that may also occur.
QUESTION 7 👄
What are the differential diagnoses of behavioural and psychological symptoms of dementia (BPSD)?
QUESTION 8 ( )
What advice would you give Jane to help her deal with Anne's aggressive incidents?

### **CASE 3 ANSWERS**

### **ANSWER 1**

It was important to address Anne as well as Jane when answering questions. You told Jane and Anne that the symptoms and time frame of AD are variable. You may have used *Figure 1* to explain the general trends of AD using scores on the MMSE.

The basic activities of daily living (BADL) are bathing, continence, dressing, eating, toileting and transferring (walking). The degree to which the person with AD loses control over any two or more of these activities will affect the level of care required by that person, and the possibility of obtaining subsidised community 'packages' of care for them.

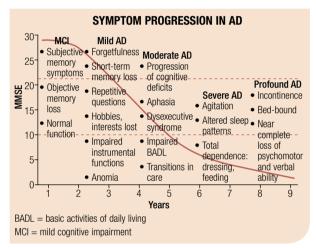


Figure 1. Symptom progression in AD From Feldman HH, 2007. 19 Reproduced with permission.

### **ANSWER 2**

The home environment needed to be tailored to assist Anne to be independent. It needed to be familiar. Changes to the environment could produce or exacerbate Anne's confusion. <sup>20,21</sup>

Safety in the home required consideration of the physical features of the building. These include those listed below.

- Access to the home: Is it level, or does it need a ramp?
- The house itself: Is each room safe?
- Falls prevention: Are there grab rails in the bathroom and shower, and no 'scatter rugs'?
- Safety in the kitchen: Is there an electric, rather than gas, stove, and is a fire extinguisher easily accessible?

Jane may also have considered using local social work and occupational health services for assessment of the home. The ACAT can help to assess the needs of the person with dementia and their carers, as well as the home environment.

Jane also needed to consider safety outside the home (e.g. did Anne need an identity bracelet, or should there be bells on the doors?) and whether Anne had access to social and leisure activities (e.g. clubs, bowls, shopping mall).

### **ANSWER 3**

Jane's roles, at different times, might include managing medications, therapies and medical emergencies; providing supervision and emotional support; and assisting with personal care, mobility and household tasks.<sup>22</sup>

Carers may feel considerable satisfaction when caring for relatives; however, they often feel exhausted, isolated and burdened by their responsibilities. In a survey of carers, 22 more than half reported that their physical health had been adversely affected, a third said they had sustained a physical injury, and over half reported depression, anxiety, high levels of stress and other effects on their mental health.

Carers often used words such as tiring, demanding, depressing and frustrating. Half the caregivers surveyed said they sometimes suffered depression. The losses and sacrifices they listed included having no social life or holidays, having less time available for children or other dependents, having a greater financial burden and needing to give up or limit their employment.<sup>23</sup>

#### **ANSWER 4**

Carers want GPs to be proactive in addressing their needs. Ways in which this can be done are shown in *Table 1*. In addition to the issues in the table, consider whether the carer is eligible for federal government funded assistance — a carer's allowance and/or a carer's pension.

# Table 1. What carers would like general practitioners to do

- Recognise their carer status and care responsibilities and include them in care planning and decision-making.
- Avoid assumptions about carer's capacity, confidence and willingness to provide home care.
- Provide plain-language information to the carer on the patient's condition, prognosis, treatment, care needs and management (including behaviour management).
- Provide information and referrals relevant to carers (e.g. in-home and residential respite care options, counselling, peer support groups, financial entitlements, self care and coping strategies).
- Give referrals to carer associations and state-wide condition specific bodies as a starting point.
- Discuss and, where appropriate, assess the carer's own physical and psychosocial health needs.
- Engage other family members in understanding and sharing care responsibilities.
- · Recognise grief and loss on cessation of caring.

From Nankervis JM, Waxman PJ, O'Hara DA, Burbidge M. Caring for family carers in general practice. Med J Aust 2002;177(8):408–10. © Copyright 2002 *The Medical Journal of Australia* – reproduced with permission.

### **ANSWER 5**

Counselling and psychosocial interventions for caregivers can have a positive effect on patients with AD, and other dementias, as well as their caregivers. However, one study showed that a semi-tailored program of counselling, education and support for patients with mild AD and their caregivers may not improve outcomes.<sup>24</sup>

The burden of care may have a significant impact on the carer and should be assessed sporadically, where possible, as the person with dementia progresses in their illness. The Caregiver Burden Scale (see *Resources*) is a 22-item self-administered questionnaire that can be used to assess the 'experience of burden'.

### **ANSWER 6**

Irritability, aggression and wandering when present in dementia are called BPSD. They may also be referred to as neuropsychiatric symptoms.

Other BPSD include:25

- symptoms of disturbed perception (hallucinations), thought content (delusions), mood (depression) and anxiety
- behavioural changes such as wandering, aggression, sexual disinhibition, screaming and hoarding
- aberrant motor behaviours including pacing, rummaging, wandering and pointless hyperactivity
- apathy, sleep disturbance and agitation.

The most common BPSD are apathy, wandering, aggression and agitation.

### **ANSWER 7**

Differential diagnoses of BPSD include:<sup>25</sup>

- delirium
  - from infection, pain or medication
- drugs
  - anticholinergic medications
  - anti-Parkinson medication
- changes to the environment or routine.

### **ANSWER 8**

There are several ways to assess and manage BPSD, and there is no consensus on the optimum management process. One helpful model is the ABC model, which is described below.<sup>26</sup>

- Antecedent or triggering event that preceded the behaviour
- Behaviour itself
- Consequence of that behaviour

This is a way of characterising events and resultant behaviour.

When Anne is aggressive, if Jane could intervene using non-aggressive words in a calming voice with gentle body language, it would be more likely that Anne's behaviour might be modified.

CASE 4 check Dementia

QUESTION 3

What investigations might be helpful?

### CASE 4

### ROBERT IS CONFUSED

Robert, aged 76 years, is brought in to see you by his wife, Mavis. She is concerned that over the past 6 months Robert has begun to move more slowly and is prone to bouts of confusion. Last month he underwent an elective cholecystectomy, which was complicated by post-operative confusion and behavioural disturbance. He was diagnosed with delirium and given haloperidol to reduce his symptoms. Mavis tells you that his memory has not returned to its pre-operative level.  On further questioning, Mavis says Robert's memory may have been poor for up to 18 months before his admission to hospital. Despite this, he has remained at home and she had attributed his decline to 'old age'.  On examination, Robert is bradykinetic, with moderate cogwheeling in his upper limbs. Tremor is absent. You notice that his thought processes appear to be slow (bradyphrenia) and that it takes him time to answer your questions. His MMSE score is 23/30. Robert loses 2 points on orientation to time, 4 points on the serial sevens task, and 1 point on the picture copying task.	QUESTION 4
QUESTION 1	QUESTION 5
QUESTION 2	QUESTION 6

QUESTION 7 👄
How would you treat a psychotic episode in a patient with LBD?
QUESTION 8 👄
How is LBD different from Parkinson dementia?
now to LBB difform form familiation domontal.

### **CASE 4 ANSWERS**

### **ANSWER 1**

It is important to consider persisting delirium, Parkinson disease, dementia (including AD, VaD, LBD and Parkinson-related dementia), medication and stroke.

### **ANSWER 2**

Obtaining a time frame of Robert's symptoms is vital. If parkinsonism has been present only since the use of haloperidol then it is most likely a side effect of this medication. However, 'physical slowing' was reported by Mavis several months prior to Robert's operation. It is important to clarify what she means by this as Robert may have had undiagnosed Parkinson disease. The use of haloperidol may have exacerbated a pre-existing parkinsonism and raises the possibility of underlying Parkinson disease or LBD. If Robert's cognitive decline came before his physical slowing then LBD becomes more likely than Parkinson disease. A dramatic decline in his cognition post-operatively may suggest an ongoing delirium or stroke. A history of smoking, hypertension, diabetes or hypercholesterolaemia is more suggestive of VaD or AD.

### **ANSWER 3**

It is important to exclude reversible conditions that might be causing Robert's symptoms. A standard 'dementia screen' includes FBE, EUC, RBG, LFT, TFT, B12 and folate levels, and CT brain. Ongoing delirium is a possible cause of Robert's symptoms, so excluding infection with urine microscopy and culture, wound swab and CXR is important. If a vascular cause is suspected (i.e. the patient is found to be in atrial fibrillation or has carotid bruits) consider an ECG and carotid ultrasound.

### **ANSWER 4**

The role of standard neuroimaging in the diagnosis of dementing illnesses is controversial. However, some form of structural neuroimaging is recommended by the *American Academy of Neurology Guidelines*. CT scanning is useful for eliminating the possibility of the presence of mass lesions and stroke, and for detecting the presence of significant atrophy. Atrophy of the hippocampus is a classic finding in AD (best seen in the coronal view). The presence of lacunar infarcts and extensive deep white matter ischaemic changes suggest a vascular cause. Low resolution is a limitation of CT scanning.

The improved resolution that magnetic resonance imaging (MRI) can offer is a significant advantage for the detection of vascular changes (especially if fluid attenuated inversion recovery imaging is requested) and provides more accurate visualisation of the medial temporal lobe structures. Functional imaging, such as single photon emission computed tomography (SPECT) and positron emission tomography (PET) scanning can aid in the differential diagnosis of AD, LBD and VaD, but SPECT is limited by false negative findings<sup>29</sup> and PET is hard to access outside of tertiary care centres.

CASE 4 check Dementia

### **ANSWER 5**

LBD accounts for 10–15% of all cases of dementia. There are three key diagnostic features for LBD. These are visual hallucinations, parkinsonism and significant fluctuation in cognition. The presence of two or more features in someone with dementia indicates probable LBD  $^{30}$ 

Visual hallucinations, when present, are classically described as 'cinematic', in that they convey a striking sense of realism. Interestingly, they most commonly take the form of small animals, small children or adult figures, and they are often paradoxically non-distressing to the patient.

The parkinsonism seen in LBD tends to be akinetic in nature, with tremor often being absent. It is often dopamine non-responsive, and blunting of affect, bradykinesia and bradyphrenia are commonly observed.

While cognitive fluctuation can occur in a number of different dementing illnesses, it is particularly striking in LBD, occurring not only from day to day but throughout the day as well.

# Table 2. Consensus criteria for the clinical diagnosis of probable and possible LBD<sup>31</sup>

- I. The central feature required for a diagnosis of LBD is progressive cognitive decline of sufficient magnitude to interfere with normal social or occupational function. Prominent or persistent memory impairment may not necessarily occur in the early stages, but is usually evident with progression. Deficits on tests of attention and of fronto-subcortical skills and visuospatial ability may be especially prominent.
- II. Two of the following core features are essential for a diagnosis of probable LBD:
  - fluctuating cognition with pronounced variations in attention and alertness
  - recurrent visual hallucinations that are typically well formed and detailed
  - spontaneous motor features of parkinsonism.
- III. Features supportive of the diagnosis are:
  - repeated falls
  - syncope
  - · transient loss of consciousness
  - · neuroleptic sensitivity
  - systemised delusions
  - · hallucinations in other modalities.
- IV. A diagnosis of LBD is less likely in the presence of:
  - stroke disease, evident as focal neurologic signs or on brain images
  - evidence on physical examination and investigation of any physical illness or other brain disorder sufficient to account for the clinical picture.

From McKeith IG, O'Brien JT, Burn DJ, et al. Dementia with Lewy bodies. In Davis KL, Charney D, Coyle, JT(eds) et al. Neuropsychopharmacology: the fifth generation of progress. Reproduced with permission.

### **ANSWER 6**

There are a number of other symptoms that support a diagnosis of LBD even though they do not form part of the diagnostic criteria.

These include those listed below.

- Autonomic instability this tends to manifest either as falls (resulting from postural hypotension) or as urinary incontinence.
- Daytime somnolence spouses will often describe their partners falling asleep as soon as they sit down to watch television.
- Increased tendency to develop delirium this could reflect the relatively greater cholinergic deficiency that is present in LBD compared to AD.<sup>31</sup>
- Exquisite vulnerability to neuroleptic agents such as haloperidol even relatively small doses can cause dramatic extrapyramidal sideeffects.
- REM sleep behaviour disorder (RSBD) this condition is characterised by a loss of the paralysis that normally accompanies stage IV REM sleep. The key to diagnosis is the presence of vivid dreams that are often violently acted out. This can result in bedclothes being in disarray when a patient awakes and a patient's partner needing to sleep in a separate bed.

### **ANSWER 7**

If an antipsychotic must be given to a patient with LBD due to behavioural disturbance, low-dose quetiapine is generally the agent of choice. Quetiapine has minimal dopamine-blocking properties, and is therefore less likely to precipitate parkinsonian side effects than other agents. Doses in the range of 12.5–25 mg nocte should be trialled initially, and only rarely is a dose in excess of 100 mg nocte required. Quetiapine itself is not without risk. Its main side effects are sedation and postural hypotension in this patient group, who may already suffer from autonomic instability and daytime somnolence. Clozapine is also effective for psychotic symptoms in LBD, but its use is best restricted to specialist services.

### **ANSWER 8**

The distinction is largely arbitrary. They can be viewed as a continuum, with Parkinson disease at one end and LBD at the other end. Parkinson dementia is diagnosed if motor symptoms precede the onset of cognitive decline by at least 12 months. Vice versa, LBD is diagnosed if cognitive symptoms either precede, or occur within 12 months of the onset of motor parkinsonism.<sup>32</sup> Lewy bodies are cellular inclusions composed of ubiquitin and alpha-synuclein. They are the pathological hallmark of Parkinson disease, where they are found in the deep cerebral striatal structures of the caudate and putamen. At autopsy, however, scattered Lewy bodies are also present in the cerebral cortex. Patients who have suffered from Parkinson disease for at least 20 years almost invariably show signs of dementia. This reflects Lewy body<sup>33</sup> spread to the cortical regions. LBD is characterised by the presence of Lewy bodies predominantly in the cortex with relatively few found in the deeper striatal structures. It is likely that Lewy bodies within these deeper structures can explain both the parkinsonism and extreme neuroleptic sensitivity that LBD patients demonstrate.

### CASE 5

### **DIANNE PRESENTS WITH ODD BEHAVIOUR**

Dianne, aged 64 years, presents with her son Peter. Dianne appears nonplussed when entering the room, in stark contrast to the concern evident on Peter's face. Dianne wonders why she has been brought to the doctor, as she feels well. Peter's body language shows that he has concerns, so you politely ask Dianne if she would mind if you spoke with Peter alone for 5 minutes while she returns to the waiting room. You reassure her that this is common practice.

Peter expresses his concern regarding changes in his mother's behaviour, personality and memory over the past 3 years. The initial changes were in the social context. Dianne began to show poor judgement and display inappropriate behaviour. Once a reserved lady, Peter says his mother has become overfamiliar and offensive at times. Once an empathic mother and friend, she has become more distant, and will sometimes look through a person rather than at them. At the same time, his mother's personal hygiene has deteriorated, and she has lost nearly all social contacts because 'they all annoy her'. Peter mentions that he thinks his mother's social isolation might stem from 'paranoia'. She has accused friends of stealing items from her house, and of later returning them.

Dianne returns to the room and you take a history from her while Peter is present. There is no personal or family history of neurological or psychiatric illness. Dianne's developmental history was unremarkable, and there is no clinically significant alcohol or drug history. There is no history of head

On examination, you find no signs of neurological or movement disorders.

Dianne's MMSE is 28/30. Her orientation is mostly intact; her attention and concentration are poor. On the clock-drawing test she shows poor planning in inserting the numbers, and finds it difficult to conceptualise where to place the hands to indicate 10 minutes past 11 o'clock.

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υι	JEST	IUN	



What diagnoses could explain Dianne's presentation?

QUESTION	2				
How would	you	differentiate	between	these	diagnoses?

QUESTION 3 ( )
What investigations could help confirm the diagnosis?
That intodegations sould holp solimin als diagnosis.
QUESTION 4 🖎
What are the frontotemporal dementias (FTDs)?
-
OHECTION E
QUESTION 5 🗅
What treatment is available for FTDs?

CASE 5 check Dementia

### **CASE 5 ANSWERS**

### **ANSWER 1**

The differential diagnosis for Dianne includes a neurodegenerative disease (NDD) or a psychiatric disorder.

NDDs to consider would be frontotemporal dementia (FTD), a frontal variant of AD or VaD.

Psychiatric disorders to consider would be depression, late-onset schizophrenia, schizoaffective disorder and bipolar affective disorder.

The gradual changes in personality and behaviour in addition to cognitive decline and psychosis favour a diagnosis of FTD for Dianne.

### **ANSWER 2**

A focused history, mental-state examination and physical examination can help to form a diagnostic hierarchy. Dianne's presentation, with behaviour and personality changes along with memory deficits, suggests a process involving the frontal and temporal lobes. Other features to look for are further manifestations of executive dysfunction, language problems (aphasias) and new onset movement disorders.

FTD is often difficult to diagnose. It is one of the dementias that can occur in younger people and needs to be considered in those aged under 60 years. There are numerous areas of clinical overlap between NDDs and psychiatric disorders such as schizophrenia and depression.

- NDDs are commonly associated with neuropsychiatric phenomena such as delusions, hallucinations and mood disorders. FTD can be associated with all of these phenomena.<sup>34</sup>
- Chronic psychotic illnesses such as schizophrenia and NDDs may both present with altered personality, behaviour and cognition, and may both be associated with neuropsychiatric phenomena.
- Schizophrenia (although usually presenting at an earlier age) can be thought of as having multiple symptom domains positive (e.g. delusions, hallucinations), negative (e.g. flat affect, alogia, amotivation, anhedonia, asociality), cognitive and disorganised.
   The negative features of schizophrenia are most likely deficits in frontal lobe function (pre-frontal areas).<sup>35</sup> Hence, there is clinical overlap between negative symptoms of schizophrenia and NDDs affecting the pre-frontal cortices, which commonly manifest as apathy.

Having reviewed the areas of clinical overlap, you may now explore the presenting features and look for supporting information.

You need to elicit more detail about Dianne's altered behaviour and personality. Why is she behaving as she is, and why do her friends 'annoy' her? The behaviour may be due to either delusions regarding her friends (such as fixed, false persecutory ideas) or impulsivity. If impulsivity is present, its extent should be sought, as it can manifest in multiple ways (e.g. a loss of manners, rash decisions, hypersexuality, complex behaviours such as gambling). You must also exclude psychosocial issues.

It is important to explore Dianne's apparent paranoia. The distinction between true paranoid delusions and 'delusions of theft' is a useful one. Although boundaries may blur, delusions of theft are common in those with short-term memory deficits and reflect a primary memory deficit rather

than a psychotic process. The patient merely forgets that they have moved an item in their house, and rationalises the absence of the item by accusing others of pilfering. Such 'delusions' are thus theoretically unlikely to respond to antipsychotics, although there is limited evidence that risperidone is effective.<sup>36</sup>

### **ANSWER 3**

Dianne has pronounced behaviour and personality change with cognitive impairment. There is also an absence of marked positive symptoms of schizophrenia. Hence, if investigations fail to reveal features of a dementia, an NDD would still remain a provisional diagnosis.

Blood tests will help determine whether a reversible cause for cognitive decline and behaviour change is present. A standard dementia screen would be appropriate, comprising FBE, EUC, RBG, LFTs, TFTs, C-reactive protein, and vitamin B12 and folate levels.

Structural imaging in dementia, once used mainly to exclude surgical lesions, can now aid in confirming a particular diagnosis. Structural neuroimaging is an appropriate starting point, looking for frontotemporal atrophy, often asymmetrical with preservation of posterior structures. CT would be the investigation of choice in the primary care setting. This can be followed by referral to a specialist or to a memory clinic for ongoing investigation.

### **FEEDBACK**

Other tests include MRI, which has greater sensitivity and specificity than CT.<sup>37</sup> A negative scan does not exclude an FTD, particularly in the earlier stages of the disease.<sup>38</sup> Functional scans such as SPECT and fluorodeoxyglucose positron emission tomography (FDG-PET) have greater sensitivity than structural scans<sup>39</sup> and may show a characteristic pattern of frontal hypoperfusion.<sup>39</sup>

### **ANSWER 4**

The FTDs are a group of dementias that primarily affect the frontal and temporal lobes. As neurones die in this area, the frontal and temporal lobes atrophy and shrink. With time, this causes behavioural change, problems with cognition, difficulty with walking and other movements, and problems with communication. FTD disorders are complex because they have a number of different features that do not necessarily correlate well with each other.

Clinically, FTDs can present in three different ways.

- Behaviour and personality changes can predominate. In this case, the entity is called behavioural variant frontotemporal dementia (bvFTD) (see *Table 3*).
   FTDs can also present with symptoms consistent with a functional psychiatric disorder such as mania or schizophrenia.<sup>40</sup>
- FTDs can present with primary language dysfunction. These clinical syndromes
  are subsumed under the name 'primary progressive aphasia'. Each syndrome
  has its characteristic aphasia, associated anatomical underpinnings and
  neuropathology. They are a non-fluent/agrammatic type and a semantic type.
- FTDs can present clinically with neurological signs. These variants include motor neurone disease, progressive supranuclear palsy and corticobasal degeneration.<sup>41</sup>

### **ANSWER 5**

Management of FTDs may be very challenging for patients, carers and doctors. As with most dementias, there is no disease-modifying treatment available. Furthermore, some experts recommend strongly against the use of cholinesterase inhibitors, as studies have revealed a deterioration in

behaviour associated with treatment. However, medications are available to ameliorate specific symptoms. Liaison with a specialist in the field and a multidisciplinary team approach to support the patient and carers are of

paramount importance. Guidance and support for managing behavioural problems can be accessed through support groups such as Alzheimer's Australia and FRONTIER (see *Resources*).

### Table 3. International consensus criteria for bvFTD

### I. Neurodegenerative disease

The following symptom must be present to meet criteria for bvFTD.

A. Shows progressive deterioration of behaviour and/or cognition by observation or history (as provided by a knowledgeable informant).

#### II. Possible byFTD

Three of the following behavioural/cognitive symptoms (A–F) must be present to meet criteria. Ascertainment requires that symptoms be persistent or recurrent, rather than single or rare events.

- A. Early\* behavioural disinhibition [one of the following symptoms (A.1-A.3) must be present]:
  - A.1. Socially inappropriate behaviour
  - A.2.Loss of manners or decorum
  - A.3.Impulsive, rash or careless actions
- B. Early apathy or inertia [one of the following symptoms (B.1-B.2) must be present]:
  - B.1.Apathy
  - B.2.Inertia
- C. Early loss of sympathy or empathy [one of the following symptoms (C.1–C.2) must be present]:
  - C.1.Diminished response to other people's needs and feelings
  - C.2.Diminished social interest, interrelatedness or personal warmth
- D. Early perseverative, stereotyped or compulsive/ritualistic behaviour [one of the following symptoms (D.1-D.3) must be present]:
  - D.1.Simple repetitive movements
  - D.2.Complex, compulsive or ritualistic behaviours
  - D.3. Stereotypy of speech
- E. Hyperorality and dietary changes [one of the following symptoms (E.1-E.3) must be present]:
  - E.1.Altered food preferences
  - E.2.Binge eating, increased consumption of alcohol or cigarettes
  - E.3. Oral exploration or consumption of inedible objects
- F. Neuropsychological profile: executive/generation deficits with relative sparing of memory and visuospatial functions [all of the following symptoms (F.1–F.3) must be present]:
  - F.1.Deficits in executive tasks
  - F.2. Relative sparing of episodic memory
  - F.3. Relative sparing of visuospatial skills

### III. Probable bvFTD

All of the following symptoms (A-C) must be present to meet criteria.

- A. Meets criteria for possible bvFTD
- B. Exhibits significant functional decline (by caregiver report or as evidenced by Clinical Dementia Rating Scale or Functional Activities Questionnaire scores)
- C. Imaging results consistent with bvFTD [one of the following (C.1–C.2) must be present]:
  - C.1.Frontal and/or anterior temporal atrophy on MRI or CT
  - C.2.Frontal and/or anterior temporal hypoperfusion or hypometabolism on PET or SPECT

### IV. bvFTD with definite frontotemporal lobar degeneration (FTLD) pathology

Criterion A and either criterion B or C must be present to meet criteria.

- A. Meets criteria for possible or probable bvFTD
- B. Histopathological evidence of FTLD on biopsy or at post-mortem
- C. Presence of a known pathogenic mutation

### V. Exclusionary criteria for bvFTD

Criteria A and B must be answered negatively for any bvFTD diagnosis. Criterion C can be positive for possible bvFTD but must be negative for probable bvFTD.

- A. Pattern of deficits is better accounted for by other non-degenerative nervous system or medical disorders
- B. Behavioural disturbance is better accounted for by a psychiatric diagnosis
- C. Biomarkers strongly indicative of AD or other neurodegenerative process

\*As a general guideline 'early' refers to symptom presentation within the first 3 years.

From Rascovsky K, Hodges JR, Knopman D, et al. Sensitivity of revised diagnostic criteria for the behavioural variant of frontotemporal dementia. Brain 2011;134(9):2456–77. Reproduced by permission of Oxford University Press.

CASE 6 check Dementia

QUESTION 3 🕮

### CASE 6

### MARGARET PRESENTS WITH ONGOING CONFUSION

Margaret, aged 75 years, is brought in to see you by her daughter, Rose. Margaret is happily married and until recently volunteered at the local soup kitchen. You have known her for many years, and immediately you notice a distinct change. Usually an impeccably dressed lady, Margaret appears somewhat dishevelled. Her gait is slowed and her usually warm smile lacks depth.

Margaret has a history of hypertension, insulin resistance and urinary incontinence, and has had a tendency towards anxiety and depression in the past. Her current medications are perindopril 4 mg mane, oxybutynin 5 mg bd and amitriptyline 150 mg nocte. Three months ago Margaret suffered a UTI, for which she required hospitalisation due to associated delirium (acute organic brain syndrome). Due to the severity of the delirium a CT scan of the brain was performed, which showed 'age appropriate generalised atrophy'.

Rose is worried about her mother. At the hospital, the doctors assured her the delirium would pass. However, Margaret has only partially improved. The 'strange thoughts' have gone, but she remains more confused and forgetful than before the UTI. Rose wants to know if the confusion will resolve.

QUESTION 1 (C)	
What are the features of delirium?	
QUESTION 2 💭	
Which cognitive feature will be seen on MMSE in delirium?	
which cognitive leature will be seen on wiwse in definiting	

What is the neuropathogenesis of delirium?

### **FURTHER INFORMATION**

You reassure Rose as best you can, telling her that delirium can take some time to resolve. You explain that you would first like to rule out any ongoing medical problems. You perform an MMSE and Margaret scores 21/30. Six months ago her score was 30/30 on routine screening. Margaret lost 2 points on orientation, and although she was once a bookkeeper, she only scored 1/5 on the serial sevens task. She also scored 0/3 on the recall task. To confirm that the serial sevens task was performed poorly due to poor attention, rather than a limitation in mathematics, you try another test requiring attention. Margaret has great difficulty saying the months in reverse order, and is frequently distracted from the task.

A reneat MSII and delirium screen are ordered, and the

following week Rose and Margaret return for the results. They are all essentially normal.
Rose turns to you and asks, 'Will my mum get better?'
QUESTION 4 😂 🥪
Is delirium reversible?
·
·

QUESTION 5				
Vhat do you	tell Rose?			
QUESTION 6 What can be				

### **CASE 6 ANSWERS**

### **ANSWER 1**

Delirium is an acute confusional state that is also known as an 'acute brain syndrome' or an 'acute organic reaction'.

It is characterised by acute onset with inattention the most common cognitive deficit.<sup>43</sup> It is also characterised by disorganised thinking and an altered level of consciousness.

### Table 4. Delirium – DSM IV definition

- Disturbed consciousness
- · Reduced ability to focus, sustain or shift attention
- Change in cognition or development of perpetual disturbance
- · Rapid onset (hours to days), fluctuates during the course of the day
- Evidence that it is a result of illness

From Diagnostic and Statistical Manual of Mental Disorders DSM-IV-TR Fourth Edition, American Psychiatric Association.

It is important to consider two main presentations of delirium — hyperactive and hypoactive delirium. Hyperactive patients may be loud, restless, excitable and hyper-vigilant. They are more prone to falls. Hypoactive delirium can easily be overlooked as the patient lies passively in bed and can appear to be the 'ideal patient'. They are quiet and tend to drift off to sleep, and have slow, incoherent speech. Mixed, fluctuating presentations are common. Dementia and depression are common differential diagnoses.

### **ANSWER 2**

Patients with delirium tend to perform poorly on MMSE due to their limited attention span. In particular, patients will perform poorly on the serial sevens task, which requires focused thought. Short-term memory is also impaired, probably as a direct consequence of inattention.<sup>44</sup> Executive functions such as planning, organisation and abstraction are affected by delirium, but this will not be picked up on an MMSE.

### **ANSWER 3**

The underlying mechanisms of delirium remain elusive. However, the most robust evidence to date supports a relative cholinergic deficit and dopaminergic excess in the brain.<sup>45</sup>

### **ANSWER 4**

The question of prognosis is a challenge. It is clear that delirium is associated with adverse outcomes such as increased hospital stay, cognitive decline, functional decline, institutionalisation and mortality.  $^{46}$ 

There is a common association between dementia and delirium and evidence shows that many people who appear cognitively

CASE 6 check Dementia

well, but who develop delirum, have cognitive problems long-term. This is supported by a recent study published in the *Archives of Internal Medicine*, <sup>47</sup> which showed that controlling for pre-existing dementia and medical disease burden, patients with delirium had a significant and sustained deterioration in cognition for up to 5 years compared to the control group. Although the study was not without its shortcomings<sup>48</sup>, the authors concluded that delirium per-se is neurotoxic and causes long-term cognitive decline. While particular aspects of delirium, such as psychosis and severe attentional deficits, seem to be readily reversible, there is a non-reversible, negative impact on cognition.

### **ANSWER 5**

It would be appropriate to explain to Rose that the understanding of delirium is evolving. While we have thought for many years that delirium is reversible, it is becoming clear that this is not always the case. While the majority of cognitive and neuropsychiatric phenomena of a delirium are reversible in most cases, it may be that Margaret will not improve further. In fact, Margaret may continue to deteriorate.

### **ANSWER 6**

A review of Margaret's current medication is appropriate. It is important to reduce or eliminate any medication with an anticholinergic effect. Margaret is currently prescribed two strongly anticholinergic medications — oxybutynin for bladder instability and amitriptyline for depression. Anticholinergic burden has a clear association with cognitive decline, which is explained by the current cholinergic deficit theory mentioned in *Answer 3*.

The clinical need for the oxybutynin and its current dose need to be re-evaluated. As a first step you decide to halve the dose. Similarly, the indication for amitriptyline requires revision. Until now you have been loath to wean the amitriptyline as Margaret has had long-term issues with anxiety, insomnia and depression. In discussion with Margaret and Rose a decision is made to wean and cease the antidepressant. Should Margaret's mental state deteriorate you will try another, less anticholinergic medication.

If Margaret's cognition does not improve significantly after altering her medication, it would be appropriate to refer Margaret for assessment of her cognitive state, in particular for dementia. She may be prescribed a cholinesterase inhibitor if she has AD.

### **FEEDBACK**

The presence of delirium does complicate and make the treatment of serious illnesses more difficult, but it can also result in permanent, irreversible brain damage. It is wise to show equal concern for the brain as we do for the other organs of the body.<sup>50</sup>

### CASE 7

### ALBERT IS WORRIED HE MAY DEVELOP DEMENTIA

Albert, a secondary school teacher aged 49 years, presents for a check-up and prescription renewal. He tells you he is very worried about getting AD, which his mother has. He and his 54-year-old sister had been caring for their mother for several years until she moved into residential aged care a month ago. Albert wants to know if he and his sister can be tested to determine whether they are destined to develop dementia, and what he can do to avoid his mother's fate. Albert lives with his wife and two teenage children and is relatively healthy. However, he has been taking medication for hypertension for 2 years and has mild hypercholesterolaemia.

Two months ago Albert's fasting lipids were total cholesterol 6.0 mmol/L, HDL 1.2 mmol/L (normal >1.0 mmol/L), LDL 4.3 mmol/L (normal <2.5 mmol/L), TG 2.3 mmol/L (normal <1.7 mmol/L). You told him at that time to lower his cholesterol intake and to return in 2 months for repeat blood tests.

On this visit, Albert's BP is 147/95 mmHg.

OUESTION 1 ( )					
	N 1 @ T	11	CION	IF ST	n

What do you ask Albert about his family history to assess whether he may be at risk of a genetic form of dementia?

### **FURTHER INFORMATION**

Albert believes his mother first displayed symptoms of dementia at age 74 years. She was diagnosed with AD 3 years later. To his knowledge, no other family member has been affected. His mother's father and sister lived to around 75 years of age, but her mother and brother died 'young'.

QUESTION	2	G

isk factors affect his dementia risk?
about managing his vascular risk factors in
sk?

CASE 7 check Dementia

What other potential dementia risk factors should you ask Albert about?	CASE
	ANSWER
	In the rare
	chromoso
	transmiss
	These ger
	accounts
	genetic fo dementia
	Ask Alber
	mother's
	affected.
FURTHER INFORMATION	ANSWER
Albert's work is intellectually demanding and he is learning	There is r
Spanish. His diet is relatively healthy and his body mass	dominant
index is in the normal range. He has never smoked and only	indication
occasionally drinks alcohol. He is on his feet a few hours a day	For famili
at work, but does no regular exercise.	dementia available
QUESTION 6 ( )	In sporad
What do you advise Albert about lifestyle strategies to reduce his risk of dementia?	has been genes hav cerebrova not currer
	will or will Albert car
	cases are
	test availa
	However,
	risk beca
	can't cha somethin
	Comouning
QUESTION 7 ( )	ANSWER Hypertens
	Hypertens
Albert wants to know if he will definitely avoid dementia if he does all these things. What do you tell him?	assessing
	cumulativ
	High midl
	of any de reduced r
	Albert's h of later de
	disease a

### **ANSWERS**

familial form of AD, genetic mutations on three mes have been identified that cause autosomal dominant on and younger onset (typically before 60 years of age). nes account for perhaps 1% of AD cases.51 Familial FTD for around 10-15% of cases of FTD.52 There are also rare rms of LBD, cerebrovascular disease and other causes of

about his family history and the age of onset of his symptoms, and whether other family members have been

### 2

othing in Albert's family history to suggest autosomal transmission or younger onset dementia, so there is no his family is affected by a genetic cause of AD.

es who are affected by single gene mutations that cause genetic counselling and predictive genetic testing are hrough state-based genetics services (see Resources).

c AD, the apolipoprotein E epsilon 4 allele (APOE ε4) identified as a major risk factor, and other susceptibility re been identified. APOE ε4 may also increase risk of scular disease and possibly of LBD. Clinical APOE testing is itly recommended because it is not possible to predict who not develop AD.

be reassured that only a small proportion of dementia thought to be inherited. In his case there is no genetic ble to determine whether or not he will develop AD. epidemiological studies suggest he is at 2-3 times higher ise he has an affected first-degree relative. 53 While he nge this, there are many other risk factors that he can do about.

### 3

ion is a risk factor for cerebrovascular disease and VaD. ion in midlife can also increase the risk of AD. Studies long-term use of antihypertensives from midlife suggest a e reduction in risk of dementia for each year of treatment.54 fe total serum cholesterol is associated with increased risk mentia and of AD, and statin use may be associated with sk.<sup>55</sup>

pertension and high cholesterol potentially increase his risk veloping dementia. They may contribute to cerebrovascular and may also exacerbate AD pathology.

### **FEEDBACK**

Diabetes and pre-diabetes syndromes are also risk factors for all dementia, AD and VaD. Few studies have examined the effect of treatment of diabetes on dementia risk, and for those that have, the results are mixed.<sup>56</sup>

### **ANSWER 4**

Albert should be advised to control his high blood pressure and cholesterol using evidence-based strategies. Vascular risk factors require treatment for a range of reasons, and potentially reducing the risk of dementia may be an added benefit and provide motivation for Albert to adhere to medication and lifestyle recommendations.

### **ANSWER 5**

Higher participation in cognitively stimulating activities is associated with reduced dementia risk. Mental challenge and learning contribute to increased brain volume and efficient cognitive function. Regular physical exercise is associated with lower risk of developing dementia. The mechanisms by which physical activity protects against dementia likely include reduction of vascular risk factors, neurogenesis and neuroprotection.

Diet may play a role in dementia risk, but the evidence for specific nutrients is inconclusive. Diets low in saturated fat and high in vegetable consumption may be beneficial. Obesity and underweight in midlife are associated with increased dementia risk. 59

Moderate alcohol consumption is associated with reduced dementia risk, but there is no evidence that non-drinkers should take up alcohol.<sup>60</sup> Smoking is a risk factor for dementia.<sup>61</sup>

### **ANSWER 6**

Albert should be encouraged to maintain regular participation in cognitively challenging activities. He should be advised to be more physically active. The Australian Physical Activity Guidelines (see *Resources*) recommend adults perform at least 30 minutes per day of moderate-intensity physical activity to reduce their risk for a range of conditions.

Serious head injury is associated with increased dementia risk. It is therefore appropriate to encourage Albert to wear head protection should he engage in sports or activities that could result in head injuries, such as bicycling.

Albert should be advised to follow the Australian Dietary Guidelines (see *Resources*), limit his saturated fat intake and eat plenty of fruit and vegetables. He should maintain his drinking and smoking status.

### **ANSWER 7**

Albert cannot be guaranteed that these interventions will decrease his risk of AD, despite their other health benefits. The recommendations are based on epidemiological research evidence of what, on average, puts people at lower risk, rather than clinical trials that prove interventions can reduce risk. *Table 5* is a summary of factors associated with dementia risk.

	nary of factors associated with dementia risk
Health or lifestyle factor	Risk for cognitive decline and dementia
Brain	
Mental activity	Higher mental stimulation through education, occupation or leisure is associated with lower risk.
Social activity	Higher social interaction in late life is associated with lower risk.
Body	
Alcohol	Moderate alcohol consumption is associated with lower risk, but comes with a warning — alcohol consumption can cause other health problems.
Diet	Findings for individual nutrients are inconsistent. Higher intakes of fruit, vegetables and fish seem to be associated with lower risk.
Physical activity	Regular physical exercise at all ages is associated with lower risk.
Heart	
Blood pressure	Untreated midlife high blood pressure is associated with increased risk.
Cholesterol	Untreated midlife high cholesterol is associated with increased risk.
Diabetes	Type 2 diabetes is associated with increased risk.
Smoking	Current smoking is associated with increased risk, former smoking is not.
Weight	Midlife obesity and underweight are associated with increased risk.
body and heart for	permission from Farrow M, O'Connor E. Targeting brain, cognitive health and dementia prevention. Alzheimer's . 2012. Alzheimer's Australia: Canberra.
	Alzheimer's Australia's Your Brain Matters program, which stic approach to looking after your brain, body and heart at

CASE 8 check Dementia

### CASE 8

### MARK IS HAVING DIFFICULTY MAKING DECISIONS

Mark, aged 75 years, was diagnosed with VaD 5 years ago. His wife Lillian is his primary carer and they have continued to live independently in their unit. They have two adult children who live close by. Their daughter Sarah is more involved in her parent's care, assisting with driving to the shops and appointments, and helping with household chores. Sarah's role has expanded over the past 3 years as Mark's dementia has progressed and Lillian's ability to manage their household has decreased. The family is now considering placing Mark in a residential care facility.

When Mark was diagnosed with VaD, you, as the longstanding family doctor, were invited and took part in a family meeting called by their son Travis. The family wished to discuss longer-term planning, ongoing care and what Travis called 'legal issues' with you. The family wanted to make sure that Mark's wishes were respected, and that he participated in the decision-making process while he still had capacity.

### **FURTHER INFORMATION**

QUESTION 3 ( ) ( )

About 1 year after Mark's diagnosis of VaD, Sarah had a baby. You received a call from Mark's attorney asking if you were able to assess Mark's testamentary capacity (TC) because Mark and Lillian wanted to update their will, as they had done with the birth of each new grandchild.

testator, and	uired to assess TC in terms of the understanding of a d what are the pitfalls related to the use of the MMSE you water in this area?
mignt encol	unter in this area?
Shortly after and asked concerned at the shop regularly a Mark was a intended to his licence.	
	4 (

### QUESTION 1 ( ) ( )

What issues may have been appropriate to discuss at this meeting? How should legal issues be addressed?

### QUESTION 2 ( )

What key features should you cover in a meeting to deliver bad news, and what system may assist you with delivering these messages?

QUESTION 5 (C)
What are the key features to elicit in the assessment of fitness to drive?
-
FURTHER INFORMATION:
Mark has progressed further and has now been placed in a
local residential care facility. He no longer recognises Lillian
or his children, and he has lost considerable weight. The staff
call you to tell you that they have noticed some blood in Mark's
stools. You order a series of investigations, and on examination

QUESTION 6 😂 🚳

lesion next to his caecum on CT abdomen.

What features differentiate an ACD from a plan of care?

of the results notice that he has significant anaemia and a large

### **CASE 8 ANSWERS**

### **ANSWER 1**

Issues that might have been discussed at an initial meeting with the family include:

- current living arrangements and the need for and role of a carer
- education and training for carers to assist with managing both the carer and the patient, and to limit the impact on the carer and to delay placement of the patient into a facility<sup>63,64,65,66</sup>
- · legal issues that facilitate planning.

Legal issues (e.g. enduring power of attorney for financial issues, enduring guardianship for accommodation and healthcare decisions, ACDs for future treatment, including end-of-life care) should be addressed at an early stage. Decisions about these issues may have already been made.

Assessment of decision-making capacity should focus only on the decisions that need to be made or the documents that need to be signed. Patients should only be encouraged to make the decisions they are capable of making, and their ability to do so will depend on the complexity of the decision and their situation in light of the nature and severity of their cognitive impairment (see *Figure 2*). Even people with early dementia may be unable to make some complex decisions, whereas people with severe dementia may still be able to make simple decisions.

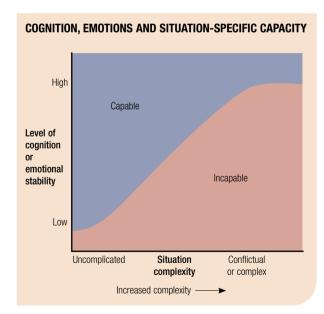


Figure 2. Assessment of testamentary capacity and vulnerability to undue influence From Shulman, 2007.  $^{\rm 62}$ 

Decision-making capacity depends on the decision to be made. It may be different for every decision made, even within one domain. It depends on the complexity of the decision.  $^{67}$ 

CASE 8 check Dementia

In this case, the family's agenda was clearly to facilitate planning and to give voice to Mark's decisions and wishes. However, it is important to be aware of the vulnerability of people with dementia to undue influence. Be wary when people with dementia are encouraged, by others seeking to gain from them, to gratuitously change documents such as wills and powers of attorney that have been properly executed in the past.

Finally, it is important to include comprehensive documentation of discussions with the patient and family members, including telephone conversations, in the patient's medical record. The patient assessment, including the tools relied upon for the assessment, also need to be documented in the records.

### **ANSWER 2**

The four main objectives of an interview to disclose bad news are to:

- gather information from the patient
- · transmit medical information
- provide support to the patient
- develop a collaborative strategy and management plan for the future.

The SPIKES system is a system to help with delivering bad news to patients. It may be modified to assist carers and families, as the patient may not have the cognition to fully participate. However, the patient has the right to information about their condition, and you need to skilfully ascertain how much information the patient would like to know.

The SPIKES system consists of six steps. 68,69

- 1. **S**etting up the interview
- 2. Assessing the Patient's Perception
- 3. Obtaining the patient's Invitation (for information)
- 4. Giving Knowledge and information to the patient
- 5. Addressing the patient's **E**motions with **E**mpathic responses
- 6. **S**trategy and **S**ummary

Other important principles include the following.

- · Break bad news gradually.
- Titrate the amount of information according to the reaction and expressed wishes of the patient and carers.
- Hold out some hope (i.e. research into possible cures).
- Ask the patient if they would like to see you alone or with family member/s.
- You may need more than one session with the patient to discuss diagnosis and consequences.
- Strong emotions will preclude the patient attending to information, so it may be best to schedule another visit.
- Follow-up with another interview. The patient and their family will
  think of many questions when they go away. They may want to
  come back with other family members.

### **ANSWER 3**

The assessment of TC in someone with dementia depends on an assessment of the complexity of the task and the person's situation, in the context of the severity of their cognitive impairment (see *Figure 2*).

TC, as defined by the Banks v. Goodfellow Criteria, requires that the testator has: $^{62,70}$ 

- understanding of the nature of a will
- knowledge of the nature and extent of one's assets
- knowledge of persons who have a reasonable claim to be beneficiaries
- understanding of the impact of the distribution of the assets of the estate
- a confirmation that the testator is free of any delusions that influence the disposition of assets
- ability to express wishes clearly and consistently in an orderly plan of disposition.

Pitfalls related to the use of the MMSE and the clock-drawing test to assess TC are that they assess higher-level brain functions and are screening tests for cognitive impairment and used to show changes over time. They are not diagnostic of dementia and cannot be used as a measure of  $TC.^{62}$ 

It is not recommended that GPs, or any other healthcare professional, assess or comment on TC unless they are trained to do so. You might refer to an expert in the field of assessing TC, such as a neurologist, psychiatrist, psychologist or geriatrician.<sup>67</sup>

Again, thorough documentation of all discussions in the patient's medical record is essential.

### **ANSWER 4**

Dementia may affect the ability to drive in many ways, including:<sup>71</sup>

- navigational errors (e.g. forgetting familiar routes or becoming lost in a familiar environment)
- limited attention span or breaks in concentration (e.g. not seeing road signs or not responding to them)
- judgement errors (e.g. misjudging distances when parking or stopping, misjudging speed)
- confusion in a stressful situation (e.g. hitting the accelerator instead of the brake)
- impaired decision-making and problem-solving (e.g. not giving way or stopping at an inappropriate time)
- · lack of insight and denial of limited ability
- · slow reaction time and not responding to directions
- inadequate eye-hand coordination.

### **ANSWER 5**

Issues that might be helpful in assessing a patient's ability to drive include the following. 71

- Be aware of different state and territory requirements.
- Use a combination of medical/specialist and practical assessment.
- · Ask about and assess:
  - driving history, including accidents or referral by police or other authority for assessment
  - vision, both front and side
  - hearing
  - reaction time
  - problem solving
  - coordination
  - praxis
  - alertness and perception
  - insight
  - other aspects of driving performance such as telling left from right, confusion on familiar routes, confusion at roundabouts, staying in the correct lane, 'stop' and 'go' at the lights, reading a map or directions, mood and confidence while on the road
  - if in doubt, send the patient for an on-road assessment, preferably with an occupational therapist.
- · Cognitive tests correlate poorly with driving ability.
- Patients will need to tell their insurance company when they are given a diagnosis of dementia, as their policy may no longer be valid once they obtain this diagnosis.

In New South Wales, nobody with dementia can have an unconditional driving licence.

### **ANSWER 6**

### **ACD**

An ACD is a written or oral statement made by a capable adult regarding wishes, preferences, values and beliefs about future treatment decisions, including end-of-life treatment. It may include instructions about future use or restriction of particular medical treatments ('treatment directive') and/or the details of a preferred substitute decision-maker ('proxy directive'). It is only used when the person loses the capacity to make decisions about their healthcare.<sup>72</sup>

### Plan of care

A plan of care is a consensus-based discussion involving the patient (who, regardless of not having capacity, may want to have some input into this discussion), carer (usually the person responsible) and medical staff around best interests, as the patient is no longer able to provide informed consent about their future treatment. The carer or person responsible can state their wishes for the patient's healthcare, based on what they believe is in the patient's best interest and reflecting what the patient would have wanted.<sup>72</sup>

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RESOURCES check Dementia

### **RESOURCES FOR DOCTORS**

- GPCOG is available at www.gpcog.com.au, and is a useful, short cognitive assessment tool.
- The Geriatric Depression Scale is a useful screening tool for depression and is available at www.stanford.edu/~yesavage/GDS. english.short.score.html [accessed 27 March 2013].
- KICA is available at www.wacha.org.au/docs/misc/KICA-Tool.pdf [accessed 27 March 2013].
- Alzheimer's Australia provides culturally appropriate assessment tools, which are available at www.fightdementia.org.au/understandingdementia/culturally-appropriate-dementia-assessment-tools-1.aspx [accessed 27 March 2013].
- The Dementia Collaborative Research Centre provides advice and copies of assessment tools for cognition, function and BPSD. It is available at www.dementia-assessment.com.au [accessed 27 March 2013].
- RUDAS is available at www.fightdementia.org.au/understandingdementia/rowland-universal-dementia-assessment-scale.aspx [accessed 27 March 2013].
- PBS information on drugs can be found at www.pbs.gov.au/browse/ medicine-listing [accessed 27 March 2013].
- The Caregiver Burden Scale can be found at www.dementiaassessment.com.au or at www.scfmresidency.com/SCFM\_ Curriculum/Neurology/Alzheimers\_AAFP\_2004.pdf [both accessed 27 March 2013].
- Alzheimer's Australia's brain health and dementia risk reduction program, Your Brain Matters at http://yourbrainmatters.org.au provides evidence-based information about strategies associated with better brain health, better cognitive function and reduced dementia risk.
- For a comprehensive summary of the state of the evidence for dementia risk reduction, see Targeting brain, body and heart for cognitive health and dementia prevention: current evidence and future directions. It can be found on the Alzheimer's Australia website, available at http://yourbrainmatters.org.au [accessed 27 March 2013].
- For a summary of the evidence, references and resources for GPs and patients for 12 dementia risk factors, see Dementia risk reduction: a practical guide for general practitioners. It can be found on the Alzheimer's Australia website, available at http:// yourbrainmatters.org.au [accessed 27 March 2013].
- The AlzRisk database at www.alzrisk.org provides a comprehensive, publicly available collection of epidemiologic reports that evaluate environmental (i.e. non-genetic) risk factors for Alzheimer disease.
- Each state and territory offers state-based genetics services. These
  can be found by entering your state or territory and 'genetics
  services' into an internet browser search.

- The Australian Physical Activity Guidelines offer advice about appropriate levels of exercise for all Australians, and are available at www.health.gov.au/internet/main/publishing.nsf/Content/healthpubhlth-strateg-phys-act-guidelines [accessed 27 March 2013].
- The Australian Dietary Guidelines offer advice about diet and nutrition for Australians, and are available at www.nhmrc.gov.au/guidelines/ publications/n55 [accessed 27 March 2013].
- WACHA e-ageing is a self-directed learning package for health professionals in rural and remote settings. It has 10 interactive modules on age-related care, including delirium. It is available at http://e-ageing.wacha.org.au/index.php?id=1521 [accessed 27 March 2013].
- The Clinical Practice Guidelines for the Management of Delirium in Older People are available at www.health.vic.gov.au/acute-agedcare [accessed 27 March 2013].
- A recent article in the Australian Family Physican, 'Frontotemporal dementia, features, diagnosis and management,' is a useful resource and overview of FTD for GPs.
- The article by P Shelton and K Rockweeod<sup>40</sup>, How golden is the gold standard of neuropathology in dementia?, in the *References* section gives a good overview of dementia.
- See check January/February 2012, units 478/479 'Fitness to drive' and the Alzheimer's Australia website at www.fightdementia.org.au [accessed 27 March 2013].
- For information on advance planning, see The Advance Care
   Directive Association's website, www.advancecaredirectives.org.au,
   or contact them by phone or email on 0423 157 003 and info@
   advancecaredirectives.org.au.
- The Respecting Patient Choices Advanced Care Planning website, available at www.respectingpatientchoices.org.au, provides resources and information on general and state-specific advanced care planning.
- The Office of the Public Advocate or Public Guardian in each state and territory provides a range of fact sheets that can assist patients and their families in understanding legal issues related to capacity, such as the legal requirements for power of attorney. For example, the New South Wales Government has created an excellent Capacity Toolkit, available at www.publicguardian.lawlink.nsw.gov.au/ agdbasev7wr/publicguardian/documents/pdf/capacity\_toolkit0609. pdf [accessed 27 March 2013], which uses case studies to illustrate the meaning and assessment of capacity.
- The Australian Medico-Legal Handbook published by Elsevier Health is an excellent resource for issues surrounding capacity and decision making. Also see AFP Medicolegal handbook for general practice by Sara Bird, published by the RACGP in 2006.

check Dementia RESOURCES

### **RESOURCES FOR PATIENTS**

- The AlzRisk database (www.alzrisk.org) provides a comprehensive, publicly available collection of epidemiologic reports that evaluate environmental (i.e. non-genetic) risk factors for Alzheimer disease.
- Each state and territory offers state-based genetics services. These
  can be found by entering your state or territory and 'genetics
  services' into an internet browser search.
- The Australian Physical Activity Guidelines offer advice about appropriate levels of exercise for all Australians, and are available at www.health.gov.au/internet/main/publishing.nsf/Content/health-publith-strateg-phys-act-guidelines [accessed 27 March 2013].
- The Australian Dietary Guidelines offer advice about diet and nutrition for Australians, and are available at www.nhmrc.gov.au/guidelines/ publications/n55 [accessed 27 March 2013].
- Alzheimer's Australia's website provides resources and information about services, support and education to assist families and carers.
   It includes cultural diversity resources. It is available at www. fightdementia.org.au [accessed 27 March 2013].
- The Alzheimer's Australia National Dementia Helpline (1800 100 500) provides carer support, living with memory loss programs and other support services, including a Chinese helpline. Further information on the services provided by the helpline it is available at www.fightdementia.org.au/services/national-dementia-helpline.aspx [accessed 27 March 2013].
- Information about dementia-related safety issues can be found at www.fightdementia.org.au/services/safety-issues.aspx [accessed 27 March 2013].
- Culturally specific Chinese dementia resources can be found at the CASS. Their head office is located in New South Wales and they can be contacted via their website (www.cass.org.au) or by telephone 02 9789 4587.
- DBMAS provides a helpline (1800 699 799) and a website (www. dbmas.org.au) with information for carers and families of people with dementia.
- Carers Australia (www.carersaustralia.com.au) provides support and resources for carers.
- The Better Health Channel provides a good basic overview of dementia, covering many frequently asked questions. It is available at www.betterhealth.vic.gov.au/bhcv2/bhcarticles.nsf/pages/Dementia\_ explained [accessed 27 March 2013].
- Independent Living Centres in each state and territory provide information and resources to enable people to continue to live independently. Independent Living Centres Australia also provides a national helpline (1300 885 886).
- FRONTIER is a clinical research group in Sydney dedicated to the study of FTD and related disorders. They have a useful link for carers, available at www.neura.edu.au/frontier/carer-support [accessed 27 March 2013].

- The National Institute of Health website gives an excellent overview of FTD and is a useful resource for patients. It is available at www.nia. nih.gov/alzheimers/publication/frontotemporal-disorders-resource-list [accessed 27 March 2013].
- The Respecting Patient Choices Advanced Care Planning website, available at www.respectingpatientchoices.org.au, provides resources and information on general and state-specific advanced care planning.
- The Office of the Public Advocate or Public Guardian in each state and territory provides a range of fact sheets that can assist patients and their families in understanding legal issues related to capacity, such as the legal requirements for power of attorney. For example, the New South Wales Government has created an excellent Capacity Toolkit, available at www.publicguardian.lawlink.nsw.gov.au/ agdbasev7wr/publicguardian/documents/pdf/capacity\_toolkit0609. pdf [accessed 27 March 2013], which uses case studies to illustrate the meaning and assessment of capacity.
- The Australian Institute of Health and Welfare released *Dementia* in Australia in 2012, which provides a comprehensive overview of dementia and the issues that may arise for both patients and carers. It is available for free download at www.aihw.gov.au/publication-detail/?id=10737422958 [accessed 27 March 2013].
- The Department of Health and Ageing website provides a list of resources (available at www.health.gov.au/internet/main/publishing. nsf/content/dementia-resources) and information (available at www. health.gov.au/internet/main/publishing.nsf/Content/dementiainformation) about dementia for patients, families and carers [both accessed 27 March 2013].
- Aged Care Australia's website provides excellent information on aged care assessment teams, resources and support programs for people with dementia, their families and their carers. It is available at www. agedcareaustralia.gov.au/internet/agedcare/publishing.nsf/Content/ Streaming+page [accessed 27 March 2013].

### Dementia

In order to qualify for 6 Category 2 points for the QI&CPD activity associated with this unit:

- read and complete the unit of check in hard copy or online at the gplearning website at www.gplearning. com.au. and
- log onto the *gplearning* website at www.gplearning. com.au and answer the following 10 multiple choice questions (MCQs) online, and
- · complete the online evaluation.

If you are not an RACGP member, please contact the *gplearning* helpdesk on 1800 284 789 to register in the first instance. You will be provided with a username and password that will enable you access to the test.

The expected time to complete this activity is 3 hours.

Do not send answers to the MCQs into the *check* office. This activity can only be completed online at www. gplearning.com.au.

If you have any queries or technical issues accessing the test online, please contact the *gplearning* helpdesk on 1800 284 789.

FOR A FULL LIST OF ABBREVIATIONS AND ACRONYMS USED IN THESE QUESTIONS PLEASE GO TO PAGE 3.
FOR EACH OUESTION BELOW SELECT ONE OPTION ONLY.

### **QUESTION 1**

Jarli, an Indigenous man from a rural and remote community aged 67 years, is brought in to see you by his daughter Kirra. He has a history of hypertension, type 2 diabetes and cataracts. Kirra is concerned that Jarli seems increasingly forgetful. He is sometimes unable to name things and no longer enjoys family gatherings. You suspect Jarli may have dementia. What is the most appropriate assessment tool to screen for dementia in this situation?

- A. MMSE
- B. RUDAS
- C. GPCOG
- D. Abbreviated Mental Test Score (AMTS)
- E. KICA.

### **QUESTION 2**

Amy, aged 80 years, comes in for her annual influenza vaccine with her husband, Bob. He tells you that Amy has been forgetting the names of her grandchildren, she sometimes leaves the gas on the stove on and is having difficulty with dressing. Amy's MMSE last year was 27/30 when she had her annual health assessment. Her MMSE today is 24/30. You make a provisional diagnosis of dementia. What is currently the most common type of dementia?

- A. Mixed dementia
- B. AD
- C. VaD
- D. FTD
- E. LBD.

### **QUESTION 3**

Amy's husband Bob from *Question 2* is reluctant to accept a diagnosis of dementia and suggests that Amy may just be 'getting old'. What features in her history and examination need to be included to make a diagnosis of dementia?

- A. A decline in multiple areas of cognition
- B. Significant impairment in occupational functioning
- C. Frequent falls
- D. Daytime somnolence
- E. Both A and B.

### **OUESTION 4**

Bruce, aged 78 years, is admitted to hospital with respiratory distress. He is a smoker and has a history of hypertension and gout. His wife, Bev, calls you because she is worried about her husband. He looks flushed and seems agitated and distressed. Bev can't always understand what he is saying and his attention moves from one subject to another. He has already pulled out his IV twice. He is usually a keen reader and plays golf regularly. You suspect Bruce may have delirium. What are the features of delirium?

- A. Acute onset with inattention, disorganised thinking and an altered level of consciousness
- B. Progressive memory loss
- C. Visual hallucinations, parkinsonism and fluctuations in cognition
- D. Autonomic instability and sensitivity to neuroleptic agents such as haloperidol
- E. RSBD.

### **QUESTION 5**

Lynette, aged 50 years, comes to see you. Her widowed mother, Mavis, was diagnosed with mixed AD and VaD 6 months ago. Mavis is no longer able to live independently and has just moved into a nursing home. Lynette has found it distressing to see her mother lose her memory. She asks if there is anything she can do to avoid developing dementia when she gets older. What are some lifestyle factors that appear to be protective against dementia?

- A. A varied and healthy diet
- B. Physical activity
- C. Mental and social activity
- D. Control of vascular risk factors (diabetes and hypertension)
- E. All of the above.

### **QUESTION 6**

Mary, aged 75 years, is brought to your clinic by her daughter Jane, who wants to know whether her mother is able to change her will. Mary was diagnosed with AD 4 years ago, and stopped driving her car 2 years ago. She has been in a residential aged-care facility for 12 months. Six months ago, Mary gave power of attorney to Jane. Which of the following is correct regarding Mary's capacity to change her will?

- A. Mary cannot change her will because she has AD.
- B. Mary's capacity to change her will depends on her MMSE score.
- Mary's capacity to change her will depends on her clock-drawing ability.
- Mary's capacity to change her will depends on her ability to understand the issues involved.
- E. Mary cannot change her will because Jane has power of attorney.

### **OUESTION 7**

Which of the following is NOT true of ACDs and plans of care?

- A. An ACD about future treatment decisions is only used when a person loses capacity to make their own decisions about healthcare.
- An ACD about future treatment decisions is made while the person has decision-making capacity.
- C. A plan of care about future treatment decisions can only be made while the person has decision-making capacity.
- D. A plan of care is made by relatives, carers and medical staff of a person who does not have decision-making capacity. It may involve the person.
- E. An ACD is useful to have in place for people with dementia.

### **QUESTION 8**

Joan, aged 80 years, is brought to see you by her daughter Cathy. Cathy says that Joan has been growing more forgetful over the past 3 years. Joan lives at home with her husband, John, and they have meals delivered. Joan used to be a keen gardener, but has lost interest. She also does not read as much as she previously did. She repeatedly asks the same questions, despite seeming to listen to the replies each time. She has trouble recalling names of objects and people. You refer Joan to a memory clinic where a diagnosis of AD is made. What stage of AD does Joan have?

- A. MCI
- B. Mild AD
- C. Moderate AD
- D. Severe AD
- E. Profound AD.

### **QUESTION 9**

Rob comes to see you to discuss whether he and his siblings should have genetic testing for suspected familial AD. His mother and aunt both developed AD in their 50s, although their two brothers are still living, aged 75 and 78 years, with no symptoms of dementia. Rob's grandmother on his mother's side had dementia early in life. He is unsure of other relatives. What is the mode of transmission of familial AD? Of Rob and his three siblings, how many would be expected to develop the disease?

- A. Autosomal dominant; 2 of 4
- B. Autosomal dominant; 1 of 4
- C. Autosomal recessive; 2 of 4
- D. Autosomal recessive; 1 of 4
- E. X-linked recessive; males only.

### **OUESTION 10**

Julie, aged 48 years, comes into your clinic. She has had testing overseas and is carrying one APOE  $\epsilon 4$  allele. Julie wants to know what this means in relation to her risk of developing AD. Which of the following is true?

- A. Those with one APOE  $\varepsilon 4$  allele will definitely develop AD.
- B. APOE ε4 is not a risk factor for AD.
- C. Some people with APOE ε4 will not develop AD.
- D. Those with APOE  $\epsilon 4$  do not have a greater risk of developing AD at a younger age.
- E. APOE ε4 is not a AD susceptibility gene.