# Dementia - different types

**Summary**

- A number of different illnesses can result in dementia and each has its own characteristics.
- Alzheimer's disease is the most common form of dementia and accounts for two thirds of dementia cases.
- Vascular dementia is the second most common form of dementia and it is associated with disease in the blood vessels in the brain.
- Lewy body disease is an umbrella term that describes conditions that include Parkinson's disease and Pakinson's disease dementia.
- Alcohol-related dementia can be avoided by following Australian guidelines for alcohol consumption.

Dementia is a broad term used to describe the symptoms of a large group of illnesses that affect the brain and cause a progressive decline in a person's functioning. It is not one specific disease. Dementia symptoms include memory loss, confusion, and personality and behavioural changes. These symptoms interfere with the person's social and working life.

Dementia is more common in people over 65, but it is not a normal part of ageing. A number of different illnesses can result in dementia and each has its own features. In most cases, the reason people develop these conditions is not known.

## Alzheimer's disease

Alzheimer's disease is the most common form of dementia and accounts for approximately two thirds of cases. It is a progressive degenerative illness that attacks the brain and causes gradual increase in cognitive (memory and thinking) problems.

In Alzheimer's disease, the physical damage in the brain is caused by amyloid plaques and neurofibrillary tangles. The plaques (fibrous patches) form when a protein called beta-amyloid forms abnormal clumps. The tangles are twisted strands of a protein called tau.

Most of the cases of Alzheimer's disease are not caused by known changes to specific genes. This type is called sporadic Alzheimer's disease and mostly occurs in people over the age of 65. Familial Alzheimer's disease is a rare type that is inherited, with symptoms often appearing when the person is between 40 and 60. In familial Alzheimer's disease, genetic changes in three specific genes cause an increase in the production of the protein present in amyloid plaques.

Almost all people with Down syndrome will get Alzheimer's disease and it will occur at a younger age than those without Down syndrome. People with Down syndrome produce more of the protein that forms amyloid plaques in Alzheimer's disease. This is because they have an extra copy of chromosome 21 that contains the gene that produces amyloid protein.

## Vascular dementia

Vascular dementia is the broad term for dementia associated with disease in the blood vessels of the brain. This blood vessel disease affects the circulation of blood to the brain and causes damage.

Vascular dementia may appear similar to Alzheimer's disease. A mixture of Alzheimer's disease and vascular dementia can occur in some people. Vascular dementia is the second most common form of dementia and there

are a number of different types.

#### Strategic infarct dementia

A single large stroke can sometimes cause strategic infarct dementia, depending on the size and location of the stroke. A large stroke can result in sudden onset of symptoms involving behaviour or thinking. The type of symptoms will depend on the area of the brain that was damaged by the stroke.

If no further strokes occur, sometimes the symptoms of dementia can stabilise or even get better over time. If there is other disease in the blood vessels of the brain or if the person has another stroke, the symptoms of dementia may get worse.

#### Multi-infarct dementia

This type of vascular dementia is caused by a number of small strokes, called mini-strokes or transient ischaemic attacks (TIAs). This is caused by disease in the large blood vessels of the brain. The strokes are often ‘silent’, meaning that the person does not know that they are having mini-strokes.

As more strokes occur, the damage in the brain increases, and reasoning and thinking skills are affected. Depression and mood swings can occur, but the symptoms depend on the location of the strokes.

Multi-infarct dementia can have a step-wise progression, where symptoms worsen after a new stroke, then stabilise for a time.

#### Subcortical vascular dementia

Also known as Binswanger’s disease, this type of vascular dementia is caused by disease in the small blood vessels deep within the brain, which damages the subcortical (deep) areas of the brain.

Subcortical vascular dementia can be related to untreated high blood pressure or diabetes that leads to vascular disease. It is caused by high blood pressure, thickening of the arteries and inadequate blood flow.

Symptoms often include deterioration of reasoning and thinking skills, mild memory problems, walking and movement problems, behavioural changes and lack of bladder control.

Subcortical vascular dementia is usually progressive, with symptoms getting worse over time as more vascular damage occurs, although the person’s abilities can fluctuate.

### Lewy body disease

Lewy body disease (LBD) is an umbrella term that describes conditions characterised by the formation of clumps in the brain called Lewy bodies. The clumps build up in brain cells and are made of a protein called alpha-synuclein. These clumps occur in specific areas of the brain, causing changes in movement, thinking and behaviour.

People with LBD may experience large fluctuations in attention and thinking. They can go from almost normal performance to severe confusion within short periods. Visual hallucinations are also a common symptom.

LBD is called a spectrum disease, because three overlapping conditions fall under the umbrella term, including:

- dementia with Lewy bodies
- Parkinson’s disease
- Parkinson’s disease dementia

All of these conditions involve the formation of Lewy bodies, but the timing of the symptoms will determine the

diagnosis.

LBD sometimes occurs along with Alzheimer's disease and vascular dementia. The overlapping of symptoms can make diagnosis of LBD difficult, except for Parkinson's disease, which has established methods for diagnosis. This means that if the movement (Parkinson's) symptoms appear first, then diagnosis is more straightforward than if the symptoms of dementia appear first.

#### Dementia with Lewy bodies

If the first symptoms to appear are changes to the person's thinking and behaviour, the diagnosis will be dementia with Lewy bodies.

#### Parkinson's disease

If the first symptoms to appear are movement symptoms, the diagnosis will be Parkinson's disease. These symptoms include tremors, stiffness in limbs and joints, speech impediments and difficulty initiating physical movements.

#### Parkinson's disease dementia

Most people with Parkinson's disease will develop symptoms of dementia. If the symptoms affecting movement appear first and are followed by symptoms affecting thinking and behaviour, the diagnosis will be Parkinson's disease dementia.

### Frontotemporal dementia

Frontotemporal dementia (FTD) is the name given to a group of dementias that involve degeneration in one or both of the frontal or temporal lobes of the brain. It is sometimes called frontotemporal lobar degeneration or Pick's disease.

The frontal and temporal lobes of the brain are involved in mood, social behaviour, attention, judgement, planning and self-control. Damage to these areas of the brain can lead to a decrease in intellectual abilities and changes in personality, emotion and behaviour. Damage can also cause difficulty in recognising objects, or understanding or expressing language.

Unlike Alzheimer's disease, memory might not be affected, especially in the early stages. The symptoms will depend on which part of the brain is damaged. When the frontal lobes are affected first, the main changes are in personality and behaviour, whereas damage to the temporal lobes affects language skills.

FTD typically affects people at a younger age than Alzheimer's disease, with symptoms beginning between the ages of 50 and 70, and sometimes younger.

#### Behavioural-variant FTD

In the frontal or behavioural variant of FTD, there are changes in the person's behaviour, habits, personality or emotional responses. Symptoms vary from person to person, depending on which areas of the frontal lobes are damaged. Some people with behavioural-variant FTD become very apathetic, while others will lose their inhibitions.

#### Semantic dementia

In the temporal lobe form of FTD, the initial symptom is usually a decline in language abilities. In semantic dementia, the ability to assign meaning to words is gradually lost. Reading, spelling, comprehension and

expression are usually affected.

#### Progressive non-fluent aphasia

Progressive non-fluent aphasia (PNFA) is the least common form of FTD and tends to have a later onset. The ability to speak fluently is gradually lost. People with PNFA have difficulty communicating, due to slow and difficult production of words, distortion of speech and a tendency to produce the wrong word.

#### Inherited FTD

Some forms of FTD are inherited and are caused by specific genetic changes. Familial FTD accounts for only around 10 to 15 per cent of all FTD cases. Two genes account for around 50 per cent of familial FTD cases – the genes for the tau protein and the progranulin protein. Several other less common genetic changes cause FTD. For affected families, genetic testing is available.

#### Frontotemporal dementia with Parkinsonism-17 (FTDP-17)

One form of familial FTD, also known as frontotemporal dementia with Parkinsonism-17 (FTDP-17), is caused by genetic changes in the gene for tau protein, located on chromosome 17. No other risk factors for this condition are known.

FTDP-17 is rare and accounts for only three per cent of all cases of dementia. Symptoms progressively get worse over time and usually appear between the ages of 40 and 60. The condition affects both thinking and behavioural skills and movements such as rigidity, lack of facial expression and problems with balance (like Parkinson's disease).

It can be distressing to be told that you (or a family member) have a genetic disorder or are at risk of having one. Genetic counselling provides the person and their family with information about a genetic disorder and its likely impact on their lives. This can assist a person with FTDP-17 to make informed medical and personal decisions about how to manage their condition and the challenges it presents to their (and their family's) health and wellbeing. Prenatal genetic counselling is also available for parents to help them decide about a pregnancy that may be at risk of FTDP-17.

### Alcohol-related dementia

Too much alcohol, particularly if associated with a diet deficient in thiamine (vitamin B1), can lead to irreversible brain damage. Many doctors prefer the terms 'alcohol-related brain injury' or 'alcohol-related brain impairment', rather than alcohol-related dementia, because alcohol abuse can cause impairments in many different brain functions.

The most vulnerable parts of the brain are those used for memory and for planning, organising and judgement, social skills and balance.

This type of dementia is preventable. The National Health and Medical Research Council of Australia recommends that men and women drink no more than two standard drinks per day to reduce the risk of health problems associated with alcohol.

Wernicke-Korsakoff syndrome is sometimes referred to as alcoholic dementia or alcohol-related dementia, but it is caused by thiamine deficiency rather than being a direct result of alcohol abuse.

#### Wernicke's encephalopathy

Alcohol damages the lining of the stomach and affects absorption of vitamins. The resulting lack of thiamine can

cause Wernicke's encephalopathy.

Symptoms of this condition include:

- jerky eye movements or paralysis of muscles moving the eyes, or double vision
- loss of muscle coordination, poor balance, staggering or an inability to walk
- confusion.

High doses of thiamine can be used to treat the condition and most symptoms should be reversed. If left untreated, permanent brain damage and death can occur.

#### Korsakoff's syndrome

Wernicke's encephalopathy that is untreated or not treated soon enough can lead to Korsakoff's syndrome. Korsakoff's syndrome can also develop on its own. It usually develops gradually and the damage is mainly to the area of the brain that is important for short-term memory.

Symptoms of Korsakoff's syndrome include:

- short-term (and sometimes long-term) memory loss
- inability to form new memories or learn new information
- personality changes
- making up stories to fill gaps in memory (confabulation)
- seeing or hearing things that aren't really there (hallucinations)
- lack of insight into the condition.

The progress of Korsakoff's syndrome can be stopped if the person completely abstains from alcohol, adopts a healthy diet and takes vitamin supplements. Thiamine supplementation may help prevent further brain damage from occurring.

### Human immunodeficiency virus-associated dementia

Human immunodeficiency virus (HIV)-associated dementia (HAD) is a complication that affects some people with HIV and acquired immune deficiency syndrome (AIDS). This condition was known as AIDS-related dementia or AIDS dementia complex (ADC).

HAD is associated with severe cognitive, motor and behavioural problems that impair day-to-day functioning, and reduce independence and quality of life. It is uncommon in people in the early stages of HIV/AIDS, but may increase as the disease advances.

Not everyone who has HIV/AIDS will develop HAD. It is thought to affect around seven per cent of people with HIV/AIDS who are not taking anti-HIV medication.

HAD is the most severe form of HIV-associated neurocognitive disorder (HAND). Milder forms affect cognitive functions (thinking skills such as memory, language, attention and planning), but not to the extent that a diagnosis of dementia is warranted.

In Australia, where most people who are HIV-positive receive treatment with combination antiretroviral therapy, HAD is fortunately uncommon. However, despite effective treatment, the milder forms of HAD affect many HIV-positive people.

### Younger-onset dementia

The term younger-onset dementia is usually used to describe any form of dementia diagnosed in people under the

age of 65 years. It is also sometimes called early-onset dementia.

Dementia in younger people is much less common than dementia occurring after the age of 65, but it is sometimes diagnosed in people between the ages of 30 and 60. Younger-onset dementia can be difficult to diagnose and it is not clear how widespread it is. Particular forms of dementia are more likely in younger people, including familial Alzheimer's disease and frontotemporal dementia.

### Dementia caused by Huntington's disease

Huntington's disease is an inherited degenerative brain disease that affects the mind and body. It usually appears between the ages of 30 and 50, and is characterised by intellectual decline and irregular involuntary movement of the limbs or facial muscles. Other symptoms include personality change, memory disturbance, slurred speech, impaired judgement and psychiatric problems.

There is no treatment available to stop the progression of this disease, but medication can control movement disorders and psychiatric symptoms. Dementia occurs in the majority of people with Huntington's disease.

### Creutzfeldt-Jakob disease

Creutzfeldt-Jakob disease (CJD) is an extremely rare and fatal brain disorder caused by a protein particle called a prion. It occurs in one in every million people. The two types of CJD are:

- classic CJD – includes sporadic and familial (very rare) forms
- variant CJD – related to 'mad cow disease'.

Early symptoms include dementia, failing memory, changes of behaviour and lack of coordination. As the disease progresses – usually very rapidly – mental deterioration becomes pronounced, involuntary movements appear and the person may become blind, develop weakness in the arms or legs and, finally, lapse into a coma.

### Where to get help

- Your doctor
- Your local community health service
- Your local council
- National Dementia Helpline – Dementia Australia Tel. 1800 100 500
- Aged Care Assessment Services Tel. 1300 135 090
- My Aged Care (Australian Government information line) Tel. 1800 200 422
- Cognitive Dementia and Memory Service (CDAMS) clinics Tel. 1300 135 090
- Carers Victoria Tel. 1800 242 636 (also known as Carers Advisory and Counselling Service)
- Commonwealth Respite and Carelink Centres Tel 1800 052 222
- Dementia Behaviour Management Advisory Service (DBMAS) Tel. 1800 699 799 – for 24-hour telephone advice for carers and care workers

### Things to remember

- A number of different illnesses can result in dementia and each has its own characteristics.
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Dementia Australia