

Rarer causes of dementia

Factsheet 442LP
January 2020



Most people who are living with dementia have Alzheimer's disease or vascular dementia. However, many other diseases and conditions can also cause dementia. This factsheet explains some of these rarer types.

In the UK, about 1 in 20 people living with dementia have a rarer type. Alzheimer's Society provides support and information for anyone affected by dementia. This factsheet includes information about how to get in touch with specialist organisations that specifically help people with rarer causes of dementia.

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Rarer causes of dementia

Dementia is caused when the brain is damaged by diseases, such as Alzheimer's disease, or a series of strokes. Rarer types of dementia can involve these same changes, but they may start in a different part of the brain to more common types. For this reason, a person with a rarer type of dementia might have different symptoms, particularly during the early stages of the condition. For more information see factsheet 400, **What is dementia?** and factsheet 456, **Dementia and the brain.**

Rarer types of dementia may also be caused by something different, such as an infection that affects the brain. Others, such as CADASIL and Huntingdon's disease, are caused by a person having a faulty gene. It is not fully understood why a person might develop a rarer type of dementia rather than a common type.

Atypical Alzheimer's disease

Problems with memory are the most common symptom of Alzheimer's disease, but around 1 in 20 people with Alzheimer's have different early symptoms. This is called atypical Alzheimer's disease. It is caused by the same kinds of changes in the brain as typical Alzheimer's, but these changes start in a different part of the brain and so cause different symptoms.

The two most common types of atypical Alzheimer's are frontal variant Alzheimer's disease (fvAD) and posterior cortical atrophy (PCA).

Frontal variant Alzheimer's disease (fvAD)

For about 1 in 50 people who have Alzheimer's disease, the condition affects the frontal lobes of their brain during the earlier stages – something that wouldn't happen until much later in most typical cases. When this happens, the condition is known as 'frontal variant Alzheimer's disease' (fvAD). For more information on the different lobes of the brain and their functions see factsheet 456, **Dementia and the brain**.

The damage to the frontal lobes can cause:

- behavioural symptoms (behavioural Alzheimer's disease) or
- problems with a type of thinking known as 'executive function' (dysexecutive Alzheimer's disease).

However, most people who have fvAD will show some of both types of symptoms.

A person who has fvAD is likely to show changes in their personality and behaviour. They may:

- lose their inhibitions – behave in socially inappropriate ways or without thinking. This could include making insensitive or inappropriate comments or invading someone's personal space, acting in a sexually inappropriate way, staring at strangers, or being verbally or physically aggressive
- lose motivation to do things that they used to enjoy
- lose the ability to understand what others might be thinking or feeling, become less sympathetic to the needs of others and show less social interest or personal warmth. They may also show reduced humour and their reactions might be inappropriate, for example laughing at another person's misfortune
- show repetitive, compulsive or ritualised behaviours – this can include repeated use of phrases or gestures, hoarding and obsessions with timekeeping. It may also include new interests, such as music or spirituality
- crave sweet, fatty foods or carbohydrates and forget their table manners. They may also no longer know when to stop eating and drinking.

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A person who has fvAD is also likely to have symptoms related to 'executive function' and may:

- struggle with tasks which follow a series of steps – for example making a cup of tea
- find it hard to think about more than one piece of information at a time when making a decision
- be easily distracted and find it hard to concentrate
- sometimes fixate on a specific thought or on something in the room
- copy other people's behaviour.

Some people who have the condition may also have memory problems, but these are generally much less severe during the earlier stages than in typical Alzheimer's disease.

Sometimes fvAD is misdiagnosed as a more common type of dementia that affects the frontal lobes of a person's brain. This can include vascular dementia or behavioural variant frontotemporal dementia.

As with other types of Alzheimer's disease, there is no treatment that can slow the progression of fvAD. Drugs such as donepezil, rivastigmine and galantamine may improve a person's symptoms. In people with other types of dementia, such as typical Alzheimer's disease, they can help a person to think more clearly. However, there haven't been any clinical trials to test how well these drugs work for people who have fvAD. This is because it is a rare type of dementia and is very difficult to diagnose correctly.

People who have frontal variant Alzheimer's disease are often given similar treatment to people with behavioural variant frontotemporal dementia. For more information see factsheet 404, **What is frontotemporal dementia (FTD)?**

Posterior cortical atrophy (PCA)

Posterior cortical atrophy (PCA) is most often caused by the same changes in a person's brain that cause Alzheimer's disease. However, in PCA the disease starts in a different part of the brain. At first it mainly affects the back of the brain – a region called the 'visual cortex'. This part of the brain processes information that comes from a person's eyes and turns it into what the person sees. For more information see factsheet 456, **Dementia and the brain**. PCA can also be caused by other types of disease that affect a person's visual cortex, but this is less common.

A person who has PCA will usually start to have symptoms between the ages of about 55 and 65. The first signs are often subtle problems with their vision. It can take a long time to confirm that sight problems are caused by damage to the brain rather than by eye problems. This means there may be a delay of several months or even years before the person gets an accurate diagnosis of PCA. This can cause a lot of frustration and worry.

In the early stages of PCA, a person will have problems with their vision that become more serious over time. These may include:

- difficulty recognising faces and objects in pictures
- finding it hard to judge distances
- having problems with spatial awareness.

A person who has PCA may be able to think clearly for several years after they are diagnosed. However, they may have increasing problems with reading, writing, spelling and arithmetic. They may also find it difficult to use digital technology, such as computers or mobile devices. PCA can cause problems with vision which may mean a person can no longer drive safely. A person's vision is likely to become increasingly distorted and they are likely to need support with most visual activities. In the later stages of PCA many people are registered blind due to them becoming increasingly visually impaired as the disease progresses.

Over time the disease that causes PCA can start to affect other parts of the brain. This causes symptoms that are more common in people with typical Alzheimer's disease, such as:

- memory loss
- confusion
- difficulties with communication.

During the late stages of PCA, a person is likely to have care needs that are similar to people with late-stage Alzheimer's disease.

There are no specific drugs to treat PCA. However, some of the symptoms to do with thinking and memory may improve slightly if they take the drugs donepezil, rivastigmine or galantamine. If the person has depression or other symptoms related to their mood, these are often treated with antidepressant medication.

Rare Dementia Support has a website with detailed information for people affected by PCA. They also have a PCA Support Group Adviser who can be contacted by phone or email. For more information see 'Other useful organisations' on page 17.

CADASIL

CADASIL is a rare, inherited type of vascular disease (a disease of the blood vessels such as arteries and veins) that can cause dementia. CADASIL stands for 'Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy'.

CADASIL is caused by a faulty gene. If a person has one parent who carries the faulty gene they have a 1 in 2 chance of developing CADASIL. People who are worried that they may have inherited the faulty gene can have genetic counselling and testing. They can ask their GP to refer them to these services. For more information see factsheet 405, **Genetics of dementia**.

A person who has CADASIL is likely to have a series of mild strokes that can damage their brain tissue, particularly the 'white matter' that connects different parts of their brain together.

CADASIL can cause a range of symptoms that develop slowly over many years. These include problems with memory and thinking as well as physical and emotional problems. They often start when a person is between about 30 and 50. Common symptoms include:

- migraines
- slurred speech
- weakness down one side of the body.

Women who have CADASIL may first notice symptoms when they become pregnant.

Around 2 in 3 people who have CADASIL will develop dementia at some point in their lives. The age that this happens can vary a lot from person to person. The symptoms of dementia are usually very similar to the symptoms of more common types of vascular dementia, particularly subcortical vascular dementia. For more information see factsheet 402, **What is vascular dementia?** and factsheet 456, **Dementia and the brain**.

Symptoms related to dementia include thinking more slowly, and problems with:

- paying attention
- planning
- organising
- reasoning
- problem-solving
- decision-making.

CADASIL may also affect the person's memory and their ability to work out how the things they see are arranged in space (visuospatial skills). They may have emotional changes that begin as apathy where the person has a general lack of motivation. They may also feel anxious and depressed.

If a person experiences these symptoms they can have a range of interviews, tests and scans to see what is causing the problems. People with CADASIL are likely to have a family history of strokes and dementia. They may also have a particular pattern of damage to their brain that shows up on a brain scan. The diagnosis will then be confirmed by a genetic test for the condition.

There is currently no cure for CADASIL. Some people can successfully treat some of their symptoms, such as migraines. They are usually advised to take medication, such as drugs to manage high blood pressure and cholesterol levels, and to make changes to their lifestyle to prevent them having more strokes in the future. There is not enough evidence from clinical trials to suggest that people with CADASIL should take drugs used for other types of dementia to treat their dementia symptoms.

CADASIL Support UK has a website with information about CADASIL and links to more support. The Cambridge Stroke website also has more information. See 'Other useful organisations' on page 17.

Corticobasal syndrome (CBS)

The symptoms of corticobasal syndrome (CBS) include problems with movement, language, memory and visual perception (how the brain interprets information that comes from the eyes). CBS usually affects people aged between 60 and 80.

Problems with movement may include:

- being stiff or slow
- having jerky movements
- problems with balance
- problems with coordination (usually on one side of the body).

As a result, CBS is often diagnosed by a doctor who specialises in movement disorders (usually a neurologist), rather than a health professional at a memory service who specialises in dementia. However, a person who has CBS may also have problems with their thinking and perception. This can include problems with their memory, concentration and decision-making.

CBS is sometimes referred to as ‘corticobasal degeneration’ or CBD. The terms CBS and CBD describe the same condition.

It can be difficult to tell that a person has CBS rather than a more common type of dementia. Something that can make CBS stand out is how it affects a person’s language skills. They may:

- find it difficult to read words and letters
- stutter or find it difficult to speak fluently
- use the wrong word – for example they may say ‘I taken the dog for a walk’ rather than ‘I took the dog for a walk’.

CBS may affect a person’s ability to work out how the things they see are arranged in space (visuospatial skills). This can make it difficult for them to judge how far away objects are, which may mean they fall or bump into things.

There is currently no cure for CBS or a way to slow down the disease that causes it. However, drugs may help to reduce some of the symptoms, for example donepezil or memantine to help with symptoms related to memory and thinking. Other drugs can reduce the physical symptoms, such as muscle stiffness, jerky movements and bladder problems. A person with CBS might also benefit from speech and language therapy, physiotherapy and occupational therapy.

PSP Association has a website and a helpline that give people affected by CBS information and support. See 'Other useful organisations' on page 17 for more information.

Creutzfeldt-Jakob disease (CJD)

Creutzfeldt-Jakob disease (CJD) is caused by an abnormally shaped protein infecting the brain. This protein is called a 'prion'. It is not known what causes prions to build up in the brain, but in most cases CJD is not thought to be inherited or transmitted from person to person.

CJD affects about 1 in every million people each year. The most common type is sporadic CJD. This normally affects people aged over 40. If a person has sporadic CJD, their symptoms of dementia usually progress very quickly (within just a few weeks or months). Early symptoms include minor memory loss, mood changes and apathy.

Within weeks the person may become clumsy and confused. They may also become unsteady when they walk and have slow or slurred speech. Their symptoms are likely to progress to:

- jerky movements
- shakiness
- stiff limbs
- incontinence
- loss of the ability to move or speak.

By this stage the person is unlikely to be aware of their surroundings or disabilities.

There is currently no cure for CJD, although some of the physical symptoms can be managed better with drugs, such as clonazepam for stiff limbs and seizures.

For sporadic CJD to be diagnosed, a person will have a medical procedure known as a lumbar puncture. In this, a thin needle is inserted between two bones in the lower back to remove some spinal cord fluid and look for specific changes in it. This normally involves a referral to the National CJD Research & Surveillance Unit. See 'Other useful organisations' on page 17 for contact details.

A form of CJD called 'new variant CJD' was identified more recently. It may be caused by eating meat from cattle that have bovine spongiform encephalopathy (BSE). This is why new variant CJD is also known as 'mad cow disease'. In the past, it caused a small number of people to develop new variant CJD. It is now very rare for a person in the UK to develop this condition.

HIV-associated neurocognitive disorder (HAND)

Most people in the UK who are living with human immunodeficiency virus (HIV) can take very effective drugs called 'combination antiretroviral therapy' (cART). These drugs keep the levels of the virus in the person's system down to very low levels.

Before cART some people with HIV would go on to develop dementia, but this is now uncommon. However, many people who have HIV still experience a milder type of cognitive impairment, such as problems with their memory and thinking. This is called HIV-associated neurocognitive disorder (HAND).

People with HAND may have difficulties with their:

- concentration
- memory
- planning
- organising
- decision-making.

These symptoms usually remain stable over time, rather than progressing to dementia. The person may also have problems with their mood, such as depression or irritability.

If someone has recently been diagnosed with HIV they should have their cognitive and emotional wellbeing assessed within three months. They should then be re-assessed at least once a year.

HIV causes an infection that weakens a person's immune system. This makes it harder for their body to fight infections and disease. There is ongoing research to find out whether HIV causes Alzheimer's disease to develop more quickly in a person's brain. Until more research is done it is difficult to know how a person's risk of developing dementia might increase if they live with HIV for many years. Older people who have HIV should visit their GP or HIV specialist if they develop problems with their thinking or mood.

The Mildmay Hospital in London gives people with HAND inpatient and day care services. For more information see 'Other useful organisations' on page 17.

Huntington's disease

Huntington's disease is an inherited genetic condition that causes dementia. It causes a slow, progressive decline in a person's movement, memory, thinking and emotional state. Huntington's affects about 8 in every 100,000 people in the UK. It usually affects people aged between about 35 and 45, but symptoms can appear in younger adults and children.

Huntington's disease is normally diagnosed when a person starts to have problems with controlling their movements. Many people with Huntington's disease may have been having emotional and behavioural symptoms for years before this, such as:

- severe depression
- apathy
- irritability
- obsessive-compulsive behaviours.

They are also more likely to misuse alcohol or drugs, or to self-harm.

In the early stages of Huntington's, some people may develop symptoms of dementia such as problems with their thinking and perception. They may be less able to recognise other people's emotions. They may also find it difficult to concentrate, plan and remember things. Because of these symptoms and emotional problems, the person may have difficulties with their relationships and work.

The memory problems that people with Huntington's disease have are often different to the memory problems that people with Alzheimer's disease have. For example, people with Huntington's may have a good memory of recent events but often forget how to do things (known as 'procedural memory'). Those affected may continue to recognise people and places until the very late stages of the disease, unlike people with Alzheimer's disease.

During the later stages of Huntington's disease, the person may have a lot of difficulty with moving, eating or speaking. They will be likely to need more personal care and support.

Huntington's is a genetic condition. If a person has one parent who carries the faulty gene that causes Huntington's disease, they will have about a 1 in 2 chance of inheriting the condition. If people are worried that they may carry the gene, they can ask their GP to refer them for genetic testing and counselling. For more information see factsheet 405, **Genetics of dementia**.

There is no cure for Huntington's disease. Research is currently being carried out to try to switch off the gene that causes the condition. It is hoped that this will reduce the symptoms of Huntington's disease, including dementia.

The Huntington's Disease Association has a website and a helpline that provide information and support for people affected by the condition. For more information see 'Other useful organisations' on page 17.

Normal pressure hydrocephalus (NPH)

Normal pressure hydrocephalus (NPH) happens when too much fluid builds up in a person's brain without increasing pressure in their brain tissue. People who have NPH are usually aged over 60.

A person who has NPH may have symptoms such as:

- difficulty staying focused on one thing or switching between tasks
- problems with organising and planning tasks
- problems with memory, particularly of recent events
- poor awareness of their problems with memory and thinking
- increased confusion
- difficulty responding to questions, particularly long or complicated ones
- loss of bladder control
- difficulty with walking – this can include shuffling or a 'magnetic' gait, where the person's feet appear stuck to the floor.

Unlike more common causes of dementia, the symptoms of NPH usually progress quickly – over just a few months.

For most people who have NPH, the cause of the condition is not known. It sometimes develops after a person has recovered from a head injury, a brain haemorrhage (bleeding in the brain) or severe meningitis (an infection of the tissue that surrounds the brain).

NPH may be treated with surgery. A thin tube is put into one of the spaces in the brain where fluid is building up. This 'shunt' will allow the excess fluid to drain into another part of the body, where it can be safely absorbed into the blood. Surgery can help with the movement symptoms of NPH, but isn't always effective for treating the symptoms of dementia (related to memory and thinking). NPH can be hard to diagnose, and health professionals often mistakenly think the symptoms are being caused by more common health conditions.

The charity Shine has a website that provides information and support for people who are affected by NPH. See 'Other useful organisations' on page 17 for contact details.

Progressive supranuclear palsy (PSP)

Progressive supranuclear palsy (PSP) is a condition that causes both dementia and problems with movement. It mainly affects people aged over 60.

About 1 in 10 people who have PSP have symptoms related to thinking and perception when they are diagnosed. However, about 7 in 10 people who have PSP are likely to develop dementia at some point. Although memory is not often badly affected by the condition, PSP can affect other parts of a person's thinking. This includes having problems with:

- concentration
- decision-making
- problem-solving
- organising
- planning.

People who have PSP can also have difficulties with their ability to work out how the things they see are arranged in space (visuospatial skills). This can make it difficult for them to judge how far away objects are, which may mean they fall or bump into things.

People who have PSP can also have problems with language. They may:

- find it difficult to read words and letters
- stutter or find it difficult to speak fluently
- use the wrong word – for example they may say 'I taken the dog for a walk' rather than 'I took the dog for a walk'.

Over time they may also develop behavioural symptoms, such as becoming apathetic, stubborn or impulsive.

PSP can cause symptoms related to movement, which may include:

- being stiff or slow
- having a jerky tremor (shaking)
- problems with balance
- problems with coordination (usually on one side of the body).

The word ‘supranuclear’ refers to the parts of the brain just above the nerve cells that control eye movement. When a person has PSP these areas become damaged. ‘Palsy’ means a person is unable to move a part of their body – in this case their eye. This means:

- they may find it difficult to move their eyes in the direction they want to look – particularly up or down
- their eyes may not co-ordinate with each other, which causes blurred or double vision
- their eyelids may not open normally and may become stuck shut.

There is currently no cure for PSP. However, there are drugs and other therapies that can help to manage some of the symptoms. The drugs that are used depend on the particular type of PSP the person has. In some types, they may be offered a drug known as levodopa. This boosts levels of dopamine in the brain and can help with slowness, stiffness and balance. If the person is struggling to open their eyelids this can sometimes be improved with injections of botulinum toxin (commonly known as ‘botox’).

PSP Association has a website and a helpline that give people affected by PSP information and support. For more information see ‘Other useful organisations’ on page 17.

Other useful organisations

CADASIL Support UK

info@cadasilsupportuk.co.uk
www.cadasilsupportuk.co.uk
www.facebook.com/groups/cadasilsupport

CADASIL Support UK is a charity that provides support and advice. It also raises awareness of the condition. It provides mutual support through its Facebook group.

Cambridge Stroke

info@cambridgestroke.com
www.cambridgestroke.com/cadasil.php

This website is run by clinicians and researchers at the University of Cambridge, which has a CADASIL clinic at Addenbrooke's Hospital in Cambridge. The website has information about CADASIL and links to other organisations that provide support.

Huntington's Disease Association

0151 331 5444 (9am–5pm Monday–Friday)
info@hda.org.uk
www.hda.org.uk

Huntington's Disease Association provides information, advice, support and publications for families affected by Huntington's disease in England and Wales. The Association can help people contact a regional adviser and their nearest branch or support group.

Mildmay Hospital

020 7613 6300
info@mildmay.org
www.mildmay.org

Mildmay Hospital in east London provides inpatient and day care services for people who are affected by HIV-related cognitive impairment. Hospital or community services can refer people to Mildmay Hospital.

National CJD Research & Surveillance Unit

0131 537 1980

contact.cjd@ed.ac.uk

www.cjd.ed.ac.uk

National CJD Research & Surveillance Unit researches prion diseases such as CJD. A person may be referred to the unit in Edinburgh if they have sporadic CJD.

PSP Association

0300 0110 122

helpline@pspassociation.org.uk

www.pspassociation.org.uk

PSP Association is a charity that provides information and support to people living with PSP and CBS. It also funds research into treatments and finding a cure.

Rare Dementia Support

020 3325 0828

contact@raredementiasupport.org

www.raredementiasupport.org

Rare Dementia Support runs specialist support services for people who are living with (or affected by) one of five rare dementia diagnoses:

- familial Alzheimer's disease (fAD)
- frontotemporal dementia (FTD)
- familial frontotemporal dementia (fFTD)
- posterior cortical atrophy (PCA)
- primary progressive aphasia (PPA).

Shine

01733 555988 (9am–5pm Monday–Friday)

firstcontact@shinecharity.org.uk

www.shinecharity.org.uk

Shine is a charity that provides information and advice for people with hydrocephalus (and spina bifida).

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Our information is based on evidence and need, and is regularly updated using quality-controlled processes. It is reviewed by experts in health and social care and people affected by dementia.

Reviewed by: Dr Tim Rittman, Senior Clinical Research Fellow, Department of Clinical Neurosciences, University of Cambridge and Dr Lucette Cysique, Senior Lecturer, Faculty of Medicine, University of New South Wales, Sydney, Australia

This factsheet has also been reviewed by people affected by dementia.

To give feedback on this factsheet, or for a list of sources, please email publications@alzheimers.org.uk

People affected by dementia need our support more than ever. With your help we can continue to provide the vital services, information and advice they need.

To make a single or monthly donation, please call us on **0330 333 0804** or go to alzheimers.org.uk/donate

Alzheimer's Society is the UK's leading dementia charity. We provide information and support, improve care, fund research, and create lasting change for people affected by dementia.

For support and advice, call us on **0333 150 3456** or visit alzheimers.org.uk



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