

Rarer causes of dementia

[Alzheimer's disease](#) is the most common cause of [dementia](#), but there are many rarer diseases and syndromes that can lead to dementia, dementia-like symptoms or [mild cognitive impairment](#). Rarer forms of dementia account for around only 5 per cent of all dementia cases in the UK. This factsheet outlines some of these rarer causes and gives some ideas about where to go for more specialist advice and information.

This factsheet covers the following rarer causes of dementia:

- Corticobasal degeneration
- Creutzfeldt-Jakob disease
- HIV-related cognitive impairment
- Huntington's disease
- multiple sclerosis
- Niemann-Pick disease type C
- normal pressure hydrocephalus
- Parkinson's disease
- posterior cortical atrophy
- progressive supranuclear palsy.

For information about other types of dementia, please see the following factsheets:

- [What is Alzheimer's disease? \(401\)](#)
- [What is vascular dementia? \(402\)](#) (this factsheet includes Binswanger's disease)
- [What is dementia with Lewy bodies \(DLB\)? \(403\)](#)
- [What is fronto-temporal dementia \(including Pick's disease\)? \(404\)](#)
- [What is Korsakoff's syndrome? \(438\)](#)

Being told that you or a loved one has dementia can be very difficult and you may experience a range of different emotions as time goes on. Support is available if you need it. [Alzheimer's Society's National Dementia Helpline](#) can provide information, support, guidance and signposting to other

organisations.

Corticobasal degeneration

Corticobasal degeneration (CBD) is a rare disease in which parts of the brain become damaged and begin to shrink. The outer layer of the brain (known as the cortex) and deep parts of the brain (called the basal ganglia) are particularly affected. It is not yet known what causes CBD but an overproduction of a protein called tau is thought to play a role. The disease usually affects people between the ages of 60 and 80.

The first symptoms that people with CBD experience are problems with movement, such as stiffness and jerkiness in one or more of their limbs. As the disease progresses, these problems will often spread to other limbs. Many people experience symptoms of dementia (including problems with memory and thinking). Other symptoms of CBD include loss of balance and co-ordination, and difficulties speaking and swallowing.

There is no cure or treatment available to slow the progression of CBD but drugs may help some symptoms and physiotherapy, occupational therapy and speech and language therapy may be beneficial. On average, people live for around eight years after their symptoms first appear.

Creutzfeldt-Jakob disease

Creutzfeldt-Jakob disease (CJD) is caused by an infectious protein in the brain called a prion. Sporadic CJD, which normally affects people over 40, is the most common form of the disease. It is estimated that the disease affects about one out of every 1 million people every year. It is not known what triggers sporadic CJD. A more recently identified form of CJD, called variant CJD, is caused by eating meat from cattle infected with bovine spongiform encephalopathy (BSE) and typically affects younger adults.

There may be many years between a person being infected and the development of symptoms. Early symptoms include minor lapses of memory, mood changes and loss of interest. Within weeks the person may complain of clumsiness and feeling muddled, become unsteady in walking, and have slow or slurred speech. Symptoms progress to jerky movements, shakiness, stiffness of limbs, incontinence and loss of the ability to move or speak. By this stage the person is unlikely to be aware of their surroundings or disabilities. People affected by CJD usually die within six months of their early symptoms developing. In a small number of patients the disease may take longer to run its course.

Diagnosis can often be difficult as a range of causes including Alzheimer's disease and vascular dementia need to be ruled out. (See factsheets [401, What is Alzheimer's disease?](#) and [402, What is vascular dementia?](#)). If no immediate diagnosis is obvious, the neurologist may conclude that the person has a rare neurodegenerative disease (a type of disease that affects the nervous system and gets worse over time) and may refer them to the National Prion Clinic in London (see 'Useful organisations') for further assessment.

There is no known cure for CJD, however it is an area of active research. For more information contact the CJD Support Network (see 'Useful organisations').

HIV-related cognitive impairment and dementia

HIV (human immunodeficiency virus) is an infection that weakens the immune system, making it harder for the body to fight infections and disease. HIV infection can cause a number of different problems with the brain that affect up to half of people with HIV. These include mild cognitive complaints (called 'neurocognitive impairment') and dementia. Neurocognitive impairment is common

in HIV but dementia is much rarer. Before the use of antiretroviral drugs (medication that helps to control HIV), around 20-30 per cent of people with advanced HIV infection developed dementia. This has now decreased to around 2 per cent.

Neurocognitive disorders in people with HIV may be caused by the virus directly damaging the brain or may be the result of a weakened immune system enabling infections and cancers to attack the brain.

Symptoms may include problems with short-term memory, language and thinking, difficulties with concentration and decision making, unsteadiness, mood changes and hallucinations. People may also have problems with their sense of smell. Some people may experience only a few very mild symptoms, such as a decline in the ability to think quickly or clearly. These mild impairments do not amount to dementia.

HIV is easily overlooked as a possible cause of dementia and, even when someone is known to have HIV infection, cognitive impairment can sometimes be difficult to diagnose because the symptoms are similar to those of other conditions such as depression.

Treatment with a combination of at least three antiretroviral drugs often prevents cognitive impairments worsening and, for many people, can reverse the cognitive damage caused by HIV. Rehabilitation programmes may also help people with HIV-related cognitive impairment to re-learn skills.

[For more information, see factsheet 446, What is HIV-related cognitive impairment?](#)

Huntington's disease

Huntington's disease is an inherited disease causing abnormal movements and problems with coordination, together with cognitive impairment that gets progressively worse over time. The course of the disease varies for each person and dementia can occur at any stage of the illness. Symptoms of dementia associated with Huntington's disease include loss of short-term memory and deterioration of planning and organisational skills. People with the disease may also develop obsessive behaviour. This form of dementia differs from Alzheimer's disease in that those affected continue to recognise people and places until the very late stages of the illness.

At present, there is no cure for dementia associated with Huntington's but research is being carried out to try to find treatments for the future.

For more information contact the Huntington's Disease Association (see 'Useful organisations').

Multiple sclerosis

Some people with multiple sclerosis (MS) experience a loss of some of their mental abilities if damage caused by the MS occurs in certain parts of the brain. People may be affected to different degrees, and in different ways, over a period of time. The mental abilities most likely to be affected are memory, concentration and problem solving. There may also be emotional problems, such as mood swings.

The term '[dementia](#)' is not generally used in association with multiple sclerosis because the decline is not usually as severe as it is in other forms of dementia, such as [Alzheimer's disease](#). It is more usual to describe the person as 'experiencing cognitive difficulties'. For more information contact the MS Society (see 'Useful organisations').

Niemann-Pick disease type C

Niemann-Pick disease type C is one of a group of rare inherited disorders. It is not related to fronto-temporal dementia, which is also sometimes called Pick's disease. It mainly affects school-age children but can occur at any time, from early infancy to adulthood. It is caused by the inability of the body to deal with cholesterol, and this leads to progressive loss of movement and difficulties with walking and swallowing.

People who first show symptoms in late adolescence or early adulthood are more likely to experience dementia as part of the disease. The dementia symptoms include confusion, memory problems and difficulties in concentrating and learning.

There is currently no treatment and life expectancy varies. However, researchers have identified the affected gene and there is continuing research into this area. For further information and support contact the Niemann-Pick Disease Group (see 'Useful organisations').

Normal pressure hydrocephalus

Normal pressure hydrocephalus occurs when excess fluid accumulates in the brain, causing pressure to build up in the brain tissue. Symptoms include difficulties with walking, dementia and urinary incontinence. Treatment involves surgery to drain excess fluid. Success of treatment varies depending on how early the condition is diagnosed, but some people make an almost complete recovery.

Parkinson's disease

People with Parkinson's disease have a higher-than-average risk of developing dementia, although the majority are unaffected and dementia typically does not occur until late in the course of the illness. Parkinson's dementia accounts for around 2 per cent of all cases of dementia in the UK. Symptoms of dementia associated with Parkinson's disease vary from person to person. The most common are memory loss, and loss of the ability to think quickly and to carry out everyday tasks. The person may become obsessive, and there may be a loss of emotional control, with sudden outbursts of anger or distress. [Visual hallucinations](#) may also occur. The person's symptoms can seem better or worse at different times.

It is not yet understood how dementia occurs in Parkinson's disease. It may be that the microscopic deposits known as Lewy bodies, which occur in nerve cells in the brain stem in people with Parkinson's, have a role to play, as they do in dementia with Lewy bodies. Dementia associated with Parkinson's disease is very similar to [dementia with Lewy bodies](#). The main difference is that problems with movement occur before cognitive symptoms appear in dementia associated with Parkinson's disease. In dementia with Lewy bodies, cognitive symptoms occur before (or at the same time as) problems with movement (see [factsheet 403, Dementia with Lewy bodies](#)).

The side-effects of certain drugs for Parkinson's may make symptoms of dementia worse, so adjusting medication is sometimes of benefit. Some of the [drugs used in Alzheimer's disease](#) may also be helpful, especially if the person is experiencing hallucinations or delusions.

For further information contact Parkinson's UK (see 'Useful organisations').

Posterior cortical atrophy

Posterior cortical atrophy (PCA), also known as Benson's syndrome, is a rare degenerative condition in which damage is focused at the back (posterior) region of the brain. In the vast majority of people the cause of PCA is Alzheimer's disease. The first symptoms of PCA tend to occur when people are in their mid-50s or early 60s. However, the first signs are often subtle and so it may be some time before a formal diagnosis is made.

Initially, people with PCA tend to have a relatively well-preserved memory but experience problems with their vision, such as difficulty recognising faces and objects in pictures. They may also have problems with literacy and numeracy. These tasks are controlled by the back part of the brain, where the initial damage in PCA occurs. As damage in the brain spreads and the disease progresses, people develop the typical symptoms of Alzheimer's disease, such as memory loss and confusion. There are no specific medications for the treatment of PCA but some people find medications for Alzheimer's disease helpful.

For further information and support contact the National Hospital for Neurology and Neurosurgery (see 'Useful organisations').

Progressive supranuclear palsy

Progressive supranuclear palsy (PSP) is a rare progressive movement disorder, sometimes known as Steele-Richardson-Olszewski syndrome. It affects many areas of the brain and causes symptoms similar to those of Parkinson's disease. Specific parts of the brain that are damaged include the regions that control eye movements and those that keep a person steady when they are walking, resulting in frequent falls. The cause of the damage in PSP is unknown.

PSP mainly occurs in people over the age of 60, although it occasionally affects younger people. One striking symptom is paralysis affecting eye movements and problems with double vision. Other symptoms include stiff or slow movements, difficulties walking and speaking, swallowing problems and personality changes. Although the person may also have problems with their speed of thinking and memory, they will remain aware of what is going on around them. In most cases, the person is more likely to be described as 'experiencing cognitive difficulties' rather than 'having dementia'. There is no cure for PSP but there are medications available to help control symptoms.

For further information and support contact the PSP Association (see 'Useful organisations').

For details of Alzheimer's Society services in your area visit alzheimers.org.uk/localinfo

For information about a wide range of dementia-related topics, visit alzheimers.org.uk/factsheets

Useful organisations

CJD Support Network

PO Box 346
Market Drayton TF9 4WN
T 01630 673 993 (helpline)
E info@cjdsupport.net
W www.cjdsupport.net

Supports people with prion diseases, including forms of Creutzfeldt-Jakob disease (CJD). Provides a range of information on the various forms of prion disease and works with professionals to improve the level of care provided for people with these conditions.

Frontotemporal Dementia Support Group

Miss Penelope Roques
3 Fairfield Park
Lyme Regis DT7 3DS
T 01297445488

E frontotemp@aol.com

W www.ftdsg.org

Supports people with fronto-temporal dementia, Pick's disease, corticobasal degeneration, frontal lobe degeneration, dementia with Lewy bodies and alcohol-related dementia (formerly the Pick's Disease Support Group). Provides information and advice, publishes a range of booklets, and has a network of local contacts. Also supports professionals involved in the care of people with these forms of dementia.

Huntington's Disease Association

Suite 24 Liverpool Science Park I

Innovation Centre 1

131 Mount Pleasant

Liverpool L3 5TF

T 0151 298 3298

E info@hda.org.uk

W www.hda.org.uk

Association that provides information, advice, support and publications for families affected by Huntington's disease in England and Wales. They can put you in touch with a regional adviser and your nearest branch or support group.

Mildmay Hospital

Austin Street

London E2 7NB

T 020 7613 6300

E info@mildmay.org

W www.mildmay.org

Provides holistic inpatient and day care services for people affected by HIV-related cognitive impairment. Referrals must be made by other hospital or community services.

MS Society

MS National Centre

372 Edgware Road

London NW2 6ND

T 0808 800 8000 (free helpline, weekdays 9am-9pm)

E helpline@mssociety.org.uk

W www.mssociety.org.uk

Charity providing information and support to anyone affected by MS. Has a national helpline and a network of over 350 local branches.

National Hospital for Neurology and Neurosurgery

Queen Square

London WC1N 3BG

T 0845 155 5000 or 020 3456 7890

W www.ucl.ac.uk/ion/nationalhospital

The National Hospital for Neurology and Neurosurgery is the UK's largest dedicated neurological and neurosurgical hospital. It provides comprehensive services for the diagnosis, treatment and care of all

conditions that affect the brain, spinal cord, peripheral nervous system and muscles.

National Prion Clinic

Box 98
National Hospital for Neurology and Neurosurgery
Queen Square
London WC1N 3BG
T 020 7692 2397
E help.prion@uclh.org
W www.nationalprionclinic.org

Clinic based at the National Hospital for Neurology and Neurosurgery. It provides diagnosis, care and support for patients with, or suspected of having, any form of human prion disease.

Niemann-Pick Disease Group (UK)

11 Greenwood Close
The Pastures
Fatfield
Washington NE38 8LR
T 0191 415 0693
E niemann-pick@zetnet.co.uk
W www.niemannpick.org.uk

Not-for-profit organisation that supports and promotes research to find a cure or treatments for all types of Niemann-Pick Disease and provides support services to individuals and families affected by the disease.

Parkinson's UK

215 Vauxhall Bridge Road
London SW1V 1EJ
T 0808 800 0303 (helpline 9am-8pm Monday to Friday, 10am-2pm Saturday)
E hello@parkinsons.org.uk
W www.parkinsons.org.uk

Charity that provides information, advice, support and publications for people with Parkinson's disease. They can put you in touch with your nearest branch, offering information, support and social contact for people with Parkinson's and their families.

PSP Association

PSP House
167 Watling Street West
Towcester NN12 6BX
T 01327 322 410
E psp@pspeur.org
W www.pspeur.org

Charity that provides information and support for families affected by progressive supranuclear palsy.

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Alzheimer's Society National Dementia Helpline

England, Wales and Northern Ireland: 0300 222 11 22

9.00am-5.00pm Monday-Friday

10.00am-4.00pm Saturday-Sunday

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