

Rarer causes of dementia

Factsheet 442LP
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Alzheimer's disease is the most common cause of dementia, but there are many rarer diseases and conditions that can lead to dementia, dementia-like symptoms or mild cognitive impairment. Rarer forms of dementia account for only around 5% of all dementia cases in the UK.

This factsheet outlines some of these rarer causes and gives some suggestions for where to go for more specialist advice and information. It covers a number of rarer causes of dementia – see 'Contents' for a full list.

Contents

- Corticobasal degeneration (CBD)
- Creutzfeldt-Jakob disease (CJD)
- HIV-associated neurocognitive disorder (HAND)
- Huntington's disease
- Multiple sclerosis (MS)
- Niemann-Pick disease type C
- Normal pressure hydrocephalus (NPH)
- Parkinson's disease dementia (PDD)
- Posterior cortical atrophy (PCA)
- Progressive supranuclear palsy (PSP)
- Other useful organisations.

Rarer causes of dementia

Corticobasal degeneration (CBD)

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 both affected. It is not yet known what causes CBD but producing too much of an abnormal form 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 and a failure to control hand movement on one side (known as ‘alien hand syndrome’). As the disease progresses, these problems will often spread to other limbs. Many people experience symptoms of dementia, including problems with memory and thinking. A small proportion of people with frontotemporal dementia also develop CBD as an ‘overlapping’ condition. Other symptoms of CBD include loss of balance and co-ordination, and difficulties speaking and swallowing.

Currently, 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.

For further information and support contact Rare Dementia Support (see ‘Other useful organisations’).

Creutzfeldt-Jakob disease (CJD)

Creutzfeldt-Jakob disease (CJD) is caused by an abnormally shaped protein called a prion infecting the brain. 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 each year. It is not known what triggers sporadic CJD, but it is not known to be inherited or otherwise transmitted from person to person.

3 Rarer causes of dementia

A more recently identified form of CJD, called new variant CJD, was caused by eating meat from cattle infected with bovine spongiform encephalopathy (BSE). This typically affected younger adults. In new variant CJD, there may be many years between a person being infected and the development of symptoms.

In sporadic CJD, the disease usually progresses within a few months. 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 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 for further assessment (see 'Other useful organisations').

There is no known cure for CJD, however it is an area which is being actively researched.

HIV-associated neurocognitive disorder (HAND)

HIV (human immunodeficiency virus) causes 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 in the brain, which affect up to half of people with HIV. This is known as HIV-associated neurocognitive disorder (HAND).

4 Rarer causes of dementia

Difficulties with memory, thinking and reasoning (aspects of cognition) are common with HIV, but they are usually mild and dementia is much rarer. Before the use of antiretroviral drugs (medication that helps to control HIV), around 20–30% of people with advanced HIV infection previously developed dementia. This figure has now decreased to around 2%.

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

Symptoms may include problems with short-term memory, learning, speed of thinking, difficulties with concentration and decision making, unsteadiness and mood changes. People may also have problems with their sense of smell.

Some people with HIV 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. Even when someone is known to have HIV infection, cognitive impairment can sometimes be difficult to diagnose. This is 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 HAND to relearn skills.

The Mildmay Hospital provides holistic inpatient and day care services for people with HAND (see ‘Other useful organisations’).

Huntington’s disease

Huntington’s disease is an inherited disease causing abnormal movements and problems with coordination. Other symptoms include mood problems and cognitive impairment that gets progressively worse over time. The age of onset and 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 mainly include difficulties with concentration, planning and organisational skills. A loss of short-term memory may also occur. People with Huntington's 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 disease, but research is being carried out to try to find treatments for the future.

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

Multiple sclerosis (MS)

Some people with multiple sclerosis (MS) experience a loss of some of their mental abilities. This happens 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. They may also experience emotional problems, such as mood swings and personality changes.

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 'Other 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 frontotemporal 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 an inherited inability to deal with cholesterol and other fats, causing them to accumulate in cells, including those in the brain. This can lead 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 for the disease, and life expectancy varies. However, researchers have identified the responsible gene and there is continuing research into this area. For further information and support contact the Niemann-Pick Disease Group (see 'Other useful organisations').

Normal pressure hydrocephalus (NPH)

Normal pressure hydrocephalus occurs when excess fluid accumulates in the brain, but without causing pressure to build up in the brain tissue. Symptoms include difficulties with walking, dementia and urinary incontinence. In most cases the cause is unknown, but it sometimes develops after recovery from a head injury, brain haemorrhage (a bleed in the brain) or severe meningitis (an infection of the tissue surrounding the brain).

Treatment involves surgery to drain excess fluid. The success of this treatment varies depending on how early the condition is diagnosed, but symptoms may improve after surgery and some people make an almost complete recovery.

For further information and support, contact Shine (see 'Other useful organisations').

Parkinson's disease dementia (PDD)

People with Parkinson's disease have a higher-than-average risk of developing dementia, although around two thirds of people are unaffected. When dementia does occur, it is typically not until late in the course of the illness. Parkinson's disease dementia accounts for around 2% 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 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 – seeing things which are not really there – may also occur. The person's symptoms vary and can seem better or worse at different times.

It is thought that Parkinson's disease dementia results from microscopic deposits known as Lewy bodies, located in nerve cells in the brain stem (the lower part of the brain just above the spinal cord). As Parkinson's disease progresses, Lewy bodies spread beyond the brain stem into other parts of the brain, causing dementia.

Lewy bodies are also seen in dementia with Lewy bodies (DLB). Parkinson's disease dementia appears very similar to dementia with Lewy bodies. The main difference is that problems with movement occur before cognitive symptoms 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. For more information see factsheet 403,

What is dementia with Lewy bodies (DLB)?

The side-effects of certain drugs for Parkinson's may make symptoms of dementia worse, so adjusting a person's medication accordingly can sometimes be 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 'Other useful organisations').

Posterior cortical atrophy (PCA)

Posterior cortical atrophy (PCA), also known as Benson's syndrome, is a rare degenerative condition in which damage occurs 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 more 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 Rare Dementia Support (see 'Other useful organisations').

Progressive supranuclear palsy (PSP)

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 people typically have symptoms similar to those of Parkinson's disease. As with Parkinson's disease, there may be a tremor (involuntary shaking of the body and limbs), but this is much less prominent in PSP. The 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 that occurs in PSP is unknown, but is linked to abnormal deposits of a protein called tau.

PSP mainly occurs in people over the age of 60, although it occasionally affects younger people. One striking symptom is paralysis of 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'. However, a small proportion of people with frontotemporal dementia also develop PSP as an

‘overlapping’ condition. There is no cure for PSP but there are medications available to help control some symptoms.

For further information and support contact the PSP Association (see ‘Other useful organisations’).

Other useful organisations

Huntington’s Disease Association

0151 331 5444
info@hda.org.uk
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

020 7613 6300
info@mildmay.org
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

0808 800 8000 (free helpline, weekdays 9am–9pm)
helpline@mssociety.org.uk
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 Prion Clinic

020 3448 4037

help.prion@uclh.org

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, diseases caused by prions such as CJD.

Niemann-Pick UK

0191 415 0693 (helpline)

info@niemann-pick.org.uk

www.npuk.org

Charitable organisation dedicated to making a positive difference to the lives of those affected by Niemann-Pick diseases. Raises awareness, provides practical and emotional support, advice and information and facilitates research into potential therapies.

Parkinson's UK

0808 800 0303 (helpline 9am–7pm Monday to Friday,
10am–2pm Saturday)

hello@parkinsons.org.uk

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

0300 0110 122

helpline@pspassociation.org.uk

www.pspassociation.org.uk

Charity that provides information and support and information to people living with PSP and CBD, while funding research into treatments and ultimately a cure.

Rare Dementia Support

07341 776 317

contact@raredementiasupport.org

<http://www.raredementiasupport.org/ftd/>

Runs specialist support services for individuals 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

www.shinecharity.org.uk

Charity that provides information and advice for people with hydrocephalus (and spina bifida).

Factsheet 442LP

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

A list of sources is available on request.

Alzheimer's Society National Dementia Helpline England, Wales and Northern Ireland:

0300 222 1122

9am–8pm Monday–Wednesday

9am–5pm Thursday–Friday

10am–4pm Saturday–Sunday

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