

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. This information sheet outlines some of these rarer causes and provides some ideas on where to go for more specialist advice and information.

Binswanger's disease

Binswanger's disease is a rare form of vascular dementia in which damage occurs to the blood vessels in the deep white matter of the brain.

Symptoms of Binswanger's most often occur in people after the age of 60. Binswanger's mainly affects memory and mental abilities such as thinking and learning. The person may also experience mood swings, tremors, problems in walking and seizures.

The cause of Binswanger's disease is unknown and there is no cure, although the person may be given medication, for example to control blood pressure or to treat depression.

Dementia with Lewy bodies (DLB)

In dementia with Lewy Bodies microscopic deposits in the brain (Lewy bodies) cause damage to nerve cells. Many of the symptoms of DLB, such as loss of memory and reasoning skills, are similar to those in Alzheimer's disease. In addition, many people with DLB develop Parkinson-type symptoms such as slowness, stiffness and tremor. Visual hallucinations and falls are common.

Particular care needs to be taken with certain tranquilizing medicines (neuroleptics) as these can be dangerous for people with DLB.

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- ***Problems with communication may mean that early signs of dementia are missed so it is important for carers and relatives to be alert to any changes in behaviour or capabilities.***

For more information, see the information sheet *What is dementia with Lewy bodies (DLB)?*

Down's syndrome

People with Down's syndrome are at particular risk of developing Alzheimer's disease in adult life and this risk increases with age. Research suggests that over half the people with Down's syndrome in their 60s have Alzheimer's. People with other forms of learning disability also have an increased risk of dementia, though not nearly as high as that of people with Down's syndrome.

For more information, see the information sheet *What is dementia with Lewy bodies (DLB)?*

Problems with communication may mean that early signs of dementia are missed so it is important for carers and relatives to be alert to any changes in behaviour or capabilities. Special tests are being developed to help in diagnosis.

For more information see the information sheet *Learning disabilities and dementia* by Dementia SA.

Fronto-temporal dementia

The term 'fronto-temporal dementia' is used for a range of conditions including Pick's disease, frontal lobe degeneration and the dementia associated with motor neurone disease.

Damage occurs in the frontal or temporal lobe areas of the brain, or both.

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Younger people – those under the age of 65 – are more likely to be affected. Symptoms vary but often include personality and behaviour changes, problems with judgement and planning and loss of language skills.

HIV and dementia

There are a number of different terms used to describe the dementia caused by the human immunodeficiency virus (HIV). These terms include 'Aids dementia complex', 'HIV dementia' and 'HIV related brain impairment'.

Although HIV can have a direct effect on the brain, the majority of people with HIV do not develop dementia or show any marked decline in their mental abilities. In addition, far fewer cases of dementia or cognitive (mental) impairment are being seen in the UK at present owing to the combination of drug therapies that prevent the virus from multiplying.

Symptoms, where they occur, will differ from person to person and even change at different times of the day. They may include forgetfulness, difficulties with concentration and complex thought, apathy, mood swings and hallucinations.

Some people may only experience 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.

Huntington's disease

Huntington's disease is a progressive hereditary disease. It most often becomes apparent in adults in their 30s, although it can occur earlier or later. There is also a juvenile type of Huntington's, which affects children.

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The course of the disease will vary for each person and dementia can occur at any stage in the illness.

Symptoms of dementia associated with Huntington's disease include loss of short term memory and loss of planning and organizational skills. People are likely to lack insight into their condition and other people's needs and be reluctant to accept help.

There may be obsessive behaviour. However, this form of dementia differs from Alzheimer's disease in that those affected continue to recognize people and places.

At present the dementia associated with Huntington's is untreatable, but a great deal of research is going on in this area.

Korsakoff's syndrome

Korsakoff's syndrome may result from continual heavy drinking over a long period. It is caused by lack of thiamine (Vitamin B1). This may be due to poor nutrition or to poor absorption of vitamins resulting from the

effects of alcohol on the stomach lining, or both. It is not strictly a dementia as damage is confined to small areas in the mid-part of the brain. It can be halted if the person stops drinking and eats healthily.

Alcohol may also have a harmful effect on the nerve cells in the outer layer of the brain, affecting a wide range of skills and abilities. This is sometimes known as alcoholic dementia. However, there may be some recovery if people abstain from alcohol.

There is though to be considerable overlap between Korsakoff's syndrome and alcoholic dementia.

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Multiple sclerosis (MS)

Some people with multiple sclerosis may experience a loss of some of their mental abilities as their disease progresses. This will occur if damage owing to MS occurs in certain parts of the brain.

People may be affected in 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 Alzheimer's disease, for example. It is more usual to describe the person as experiencing cognitive difficulties.

Niemann-Pick disease type C

Niemann-Pick disease type C is one of a group of rare inherited disorders. It mainly affects school-age children but it can occur at any time from early infancy to adulthood. It is caused by the inability of the body to deal with cholesterol and leads to progressive loss of movement and difficulties with walking and swallowing.

Dementia is often a particular problem when the disease becomes apparent in late adolescence or early adulthood. The symptoms of dementia include confusion, memory problems, difficulties in concentrating and learning. There is no treatment so far and life expectancy varies. However, research has identified the affected gene.

Normal pressure hydrocephalus (NPH)

Normal pressure hydrocephalus (NPH) occurs when an obstruction in the normal flow of spinal fluid causes pressure to build up in the issues of the brain. Symptoms include difficulties with walking, dementia and urinary incontinence.

People who have had a history of meningitis, encephalitis or head injury are more likely to develop NPH. The condition is sometimes treatable.

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Parkinson's disease

While people with Parkinson's disease have a higher risk of developing dementia than those without Parkinson's disease, the majority will remain unaffected.

How dementia occurs in Parkinson's disease is not yet understood. 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 (see above). In addition, the side-effects of certain drugs for Parkinson's may exacerbate symptoms of dementia. Adjusting medication for Parkinson's can sometimes be helpful.

Symptoms of dementia associated with Parkinson's disease will vary from person to person. The most common are memory loss and the loss of the ability to reason and to carry out normal everyday tasks. The person may become obsession and there may be a loss of emotional control with sudden outbursts of anger or distress. Visual hallucinations may occur. Symptoms often fluctuate so that the person will seem better or worse at different times.

Prion diseases

These are a group of rare diseases in which a transmissible agent, known as prion protein, accumulates in the brain. This causes dementia and neurological symptoms including unsteadiness and jerky movements.

Different prion diseases occur in humans and animals. One of these, Creutzfeldt-Jakob disease (CJD) has been identified for some time in a small number of humans. Recently a new form of (vCJD) known as variant (vCJD), has been identified.

Progressive supranuclear palsy (PSP)

Progressive supranuclear palsy (PSP) is a comparatively rare progressive condition, sometimes known as Steele-Richardson-Osliwski syndrome. It affects the brain stem and adjacent areas and some early features resemble those experienced in Parkinson's disease.

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PSP mainly occurs in people over the age of 50, although it occasionally occurs in younger people. A striking feature is paralysis affecting eye movements and problems with vision.

Although there are likely to be problems with more complex and abstract thought, the person will remain very much aware of what is going on around them. In most cases it is more likely that the person will be described as experiencing cognitive difficulties than dementia.

Thyroid deficiency

An underactive thyroid gland (hypothyroidism) can lead to the symptoms of dementia. Simple tests can detect this condition. The dementia symptoms include loss of interest, apathy, slowing down of mental abilities and poor short term memory.

Treatment involves replacing the naturally occurring thyroid hormones with synthetic hormone preparations. This is more likely to be effective in reversing the dementia if the problem is identified and treated within two years of its onset.

Vitamin deficiency

Vitamin deficiency, particularly a lack of the B vitamins, including thiamine (Vitamin B1), riboflavin (Vitamin B2), niacin (Vitamin B3), pyridoxine (Vitamin B6), folic acid and Vitamin B12, can cause dementia-like symptoms.

Vitamin deficiencies may be due to a poor diet or to problems with the body's absorption of vitamins. Vitamin deficiencies can be detected through blood tests. Treatment may include vitamin supplements and an improved diet, or in some cases, dealing with conditions that lead to poor absorption of vitamins.

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