

Creutzfeldt-Jakob disease (CJD) - *Factsheet*

Dementia web 
Information resource for carers, professionals and you

www.dementiaweb.org.uk



Creutzfeldt-Jakob disease (CJD)

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Dementia is not an illness in itself, but a term for a collection of conditions and diseases which cause disorders in the brain. One of the rarer causes of dementia is prion disease which accounts for 0.1% of all dementias. Prion diseases are caused by an abnormal protein called a prion.

CJD which is a prion disease is a very rare disease of the nervous system which causes damage to the brain. Two German doctors called Creutzfeldt and Jakob first drew attention to the disease in 1920. However, it had been known about in animals for hundreds of years. CJD makes up 80% of cases of prion disease and it usually affects people aged around sixty to sixty-five. However, there are different forms of CJD – see below.

How CJD affects the brain

All the forms of CJD are caused by faulty proteins in the brain called prions. These prions occur naturally in the brain, but in CJD some become distorted, or 'abnormally folded'. These produce the infectious particles which spread throughout the brain like a virus. They form clumps which disrupt the brain cells, and leave large holes in the brain tissue. This action gives the brain a sponge-like appearance, and another name for the prion diseases is transmissible spongiform encephalopathies.

Prions are very tough; much tougher than bacteria or viruses. Antibiotics and antivirals have no effect on them. They will survive extremes of heat and radiation and are not broken down by the enzymes which usually control the protein levels in the body.

The different types of CJD

At present, the reason why people with learning disabilities are more prone to developing dementia is not fully understood. There could be genetic factors, or a particular type of brain damage associated with learning disability may be implicated.

It has been discovered that almost all people with Down's syndrome have the plaques and tangles in their brains which are associated with Alzheimer's disease. However, not all of these people will develop the symptoms of Alzheimer's disease. Research has, however, shown that amyloid protein found in the plaques and tangles is linked to a gene on chromosome 21. As Down's syndrome is caused by an extra copy or copies of chromosome 21, this could explain the increased risk. Raised levels of cholesterol have also been implicated by some researchers.

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There are four types of CJD:

Sporadic (or classical) CJD

This mostly affects people in late middle age. It is the most common form of CJD, accounting for 85% of cases. However, it affects only one in a million people worldwide each year. The cause is unknown. It cannot be caught, it is not related to diet and it is not inherited. 78 people died of it in UK in 2009

Inherited (or familial) CJD

This is passed down through families and is caused by a faulty gene which produces prions. If a parent has the gene, there is a 50% chance that any child will inherit it. Sometimes the symptoms are very similar to sporadic CJD and the only way to establish the genetic link is by the family history or carrying out genetic testing. People who develop this form of the disease tend to do so at a younger age, and the illness tends to last for longer than for sporadic CJD. 3 people died of it in UK in 2009.

Iatrogenic CJD

Iatrogenic means "induced inadvertently by the medical treatment or procedures or activity of a physician". Iatrogenic CJD can result from infection being spread from someone who has the disease through medical or surgical treatment. Increased awareness means that cases are now extremely rare. There were 2 deaths in the UK in 2009.

Variant CJD

This form of the disease was identified only in 1996. It is caused by eating meat from animals infected with bovine spongiform encephalopathy (often known as mad cow disease). It mostly affects people in their twenties, the median age of onset being twenty-eight. This disease peaked in 2000 and is now in decline. In total, 168 people have died from this disease since 1995, 3 people died of it in UK in 2009. Controls are now in place to prevent the spread of this type of CJD from animals to humans. But because the incubation period is unknown, there are conflicting estimates of how many more people might develop this variant.

Symptoms of CJD

Symptoms vary according to the type of CJD. The effects of CJD on the brain result in symptoms of brain dysfunction including memory problems, difficulty with movement and dementia. Early symptoms may include headaches, tiredness, depression, sleep disturbance and loss of appetite. The disease progresses very rapidly, and before long people may report that they feel confused and have become unsteady on their feet. Their speech may be slurred or unusually slow, and they are likely to experience incontinence..

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People may have problems with their vision and may experience hallucinations. Memory problems increase, as do poor balance, jerkiness, and, eventually an inability to mobilise at all. People often lose their sight and their ability to talk. Eventually they will fall into a coma and are likely to die of infections, such as pneumonia. In the later stages of the disease people are not likely to be aware of what is happening to them, and there is no evidence that they are in pain.

It may be that many years pass between someone being infected and the first symptoms becoming evident. Once symptoms are seen, most people affected by CJD die within six to twelve months, but people with genetic or variant CJD may survive for longer.

What treatment can be given?

There is no cure for any of the forms of CJD. Research is ongoing, and some drugs are theoretically promising, but current treatment is limited to making people more comfortable.

Drugs can alleviate some symptoms. Anxiety, agitation and restlessness may be reduced with sedatives and depression treated with antidepressants. Drugs are also available to treat muscle spasms. Speech therapy and occupational therapy may also be helpful, and the support of district nurses and social workers can be invaluable. Many people with CJD will need terminal care in a hospice or hospital, but it is possible for people to be cared for at home with sufficient support.

What to do?

It is important to consult a doctor as soon as worrying symptoms appear. There are a number of reasons for this:

- Other possible causes for the symptoms can be investigated.
- An early diagnosis will allow the person and their family to learn about the disease and to come to terms with the situation.
- The person will be able to state their preferences about future treatment, financial arrangements, care choices etc.
- Carers will be able to seek help and advice for themselves, as well as for the person with CJD.

Where to get help

Contact your GP for support, advice and information. The GP will rule out reversible or temporary causes for symptoms, carry out first line tests, refer to a specialist and assume ongoing responsibility for the person's general health. The GP can be seen as a "gatekeeper" who can provide access to a range of specialist services.

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Many other people may also be able to help, such as social workers, community psychiatric nurses, district nurses, speech therapists, occupational therapists, counsellors, advocates and carers who may come into the home to assist with personal care.

The CJD Support Network is a patient support group established in 1994. It offers help and support for people with all forms of CJD, their carers and professionals. It provides practical and emotional support and can sometimes offer grants to help with the cost of care.

The Network operates a helpline – **01630 673973**, and its website is www.cjdsupport.net

CJD Support Network,
P.O.Box 346,
Market Drayton,
Shropshire,
TF9 4WN

The Brain and Spine Foundation (www.brainandspine.org) offers the following information:

"The Department of Health recognises the difficulties that carers may face when looking after someone with CJD. When a new case is diagnosed, doctors are requested to report it to the National CJD Surveillance Unit based at Edinburgh University, which is also home to the National CJD Care Team.

The Care Team has been set up to ensure that any gaps in local services which leave people affected by CJD with unmet needs or which hinder carers, are addressed, and an appropriate package of care put together. This is likely to involve not only doctors and nurses, but also occupational therapists, physiotherapists, speech therapists, dieticians, specialist incontinence advisors, palliative and terminal care teams and social workers. These diverse professionals will be coordinated by the patients' "keyworker".

This keyworker, who can be from health or social services, such as a GP, neurologist or social worker, is allocated when a new diagnosis of CJD is made. The aim is to identify someone with whom the patient, carer and family feel comfortable and who can work across all the areas from which help and support are received to ensure that these are as good and appropriate as possible. You will be offered the opportunity to help choose who this person should be. The keyworker will play an important role in liaising with the National Care Team if appropriate and also ensuring that those who have variant CJD and their families gain access to the government compensation scheme."

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It may be that the person's illness reaches a stage where their family can no longer cope with their care. If moving into a care home is seen to be in the person's best interests, independent reports and quality ratings on all homes in your local area can be found on the website of the Care Quality Commission which regulates them. www.cqc.org.uk (use its webform)

Or telephone 8.30 am to 5.30 pm Monday to Friday on **03000 616161**

You can also refer to our factsheet: "Moving to a Care Home and Funding Your Care".

Guideposts Trust provides specialist information and care services for people with dementia and their carers. www.dementiaweb.org.uk

Contact the Helpline number: **0845 1204048** available Monday to Friday office hours, answer service at other times or by email at info@dementiaweb.org.uk

Carers UK Adviceline: **0808 808 7777** (Wednesday and Thursday 10am to 12pm and 2pm to 4pm)

Email: adviceline@carersuk.org

Website: www.carersuk.org

Dementia Information Service for Carers

Helpline Number **0845 1204048**

Call in normal office hours. Answer phone at other times.

Email: info@dementiawebgloucestershire.org.uk

Web: www.dementiawebgloucestershire.org.uk

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