

# What is Creutzfeldt-Jakob disease (CJD)?

Prions are infectious agents that attack the central nervous system and then invade the brain, causing dementia. The best-known prion disease is Creutzfeldt-Jakob disease, or CJD. This information sheet provides an overview of the symptoms and different types of CJD.

## Prion disease

Prions are abnormal forms of protein that are extremely hardy and cannot be eradicated by normal sterilisation procedures. Their presence in the brain results in spongiform encephalopathy, so-called because areas of the brain where cells have died take on a sponge-like appearance when viewed under the microscope.

There are a number of prion diseases that affect humans. These include Creutzfeldt-Jakob disease, as well as two extremely rare inherited diseases, Gerstmann-Straussler-Scheinker disease (GSS) and fatal familial insomnia (FFI).

## Creutzfeldt-Jakob disease (CJD)

Creutzfeldt-Jakob disease was first reported by two German doctors (Creutzfeldt and Jakob) in 1920.

## What are the symptoms of CJD?

- Early symptoms include minor lapses of memory, mood changes and loss of interest.
- Within weeks, an infected person may complain of clumsiness and feeling muddled, become unsteady in walking and exhibit slow or slurred speech.
- The disease progresses to jerky movements, shakiness, stiffness of limbs, incontinence and the loss of the ability to move or speak. It is likely that the person is no longer aware of their surroundings or disabilities at this stage. Eventually they will need full nursing care.
- People affected by CJD usually die within six months of early symptoms, often from pneumonia. In a minority of patients, the disease may take two years to run its course. Very rarely, the disease can last for many years.

There is no evidence that someone with CJD is in pain, provided they are well nursed and comfortable. Drugs can alleviate symptoms such as shakiness.

## Types of CJD

There are four forms of CJD: sporadic, familial, iatrogenic and variant.

## Sporadic CJD

The cause of sporadic CJD (also called classical CJD) remains unknown, but the disease mainly affects those over the age of 50. Sporadic CJD is marked by rapid onset of dementia. The course of the disease is usually measured in months.

## Familial CJD

This is an inherited form of CJD. Those affected appear to be genetically predisposed to produce the abnormal form of prion proteins. People usually develop familial CJD at an earlier age than the sporadic form and the course of the illness is usually longer.

## Iatrogenic CJD

This form of CJD arises from contamination with tissue from an infected person, usually as a result of a medical procedure such as corneal transplants, grafts or the use of growth hormone. A few people have been infected through the use of neurosurgical instruments.

Today, no transplants are taken from infected people, growth hormone is made artificially and surgical instruments used on people with CJD are never used on other patients

## Variant CJD and the link with BSE

In 1996, a new type of CJD, variant CJD, was reported. Variant CJD appears to affect younger people than the other forms of the disease. The average age of death is 29 years. (However, one case of a 74-year-old man with variant CJD has been reported.)

There is now evidence that variant CJD is caused by bovine spongiform encephalopathy (BSE), a form of prion disease affecting cattle. Prion diseases have been found in several other animal species, including sheep (scrapie), deer, cats, mink and zoo animals. The consumption of infected beef products appears to have led to the development of BSE in humans.

Since 1989, action has been taken to remove those parts of cattle where the greatest concentrations of infective agent are found, including brains and spinal cords, from the human food chain.

There is some evidence that the number of deaths from variant CJD has reached a peak and is now in decline. However, there is currently no firm scientific basis for predicting future numbers of cases of variant CJD and there could be more than one peak to the epidemic. One problem is that the length of the incubation period for the disease remains uncertain. Prions may exist in the body for many years before symptoms begin.

## Helpful organisations

## **Creutzfeldt-Jakob Disease Support Network**

Gillian Turner  
PO Box 346  
Market Drayton  
Shropshire  
TF9 4WN

Helpline: 01630 673 993  
Email: [info@cjdsupport.net](mailto:info@cjdsupport.net)  
Website: <http://www.cjdsupport.net/>

The CJD Support Network can provide more detailed fact sheets about the different forms of CJD, as well as advice, information and support for carers and professionals.

## **Institute of Child Health**

30 Guildford Street  
London  
WC1N 1EH

Telephone: 020 7242 9789  
Website: <http://www.ich.ucl.ac.uk/>

For information about iatrogenic CJD, contact Leah Davidson at the above address.

## **National CJD Surveillance Unit**

Western General Hospital  
Crewe Road  
Edinburgh  
EH4 2XU

Telephone: 013 1537 1980  
Website: <http://www.cjd.ed.ac.uk/>

## **National Prion Clinic**

Box 98  
National Hospital for Neurology and Neurosurgery  
Queen Square  
London  
WC1N 3BG

Telephone: 020 7886 6883  
Email: [help.prion@st-marys.nhs.uk](mailto:help.prion@st-marys.nhs.uk)

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