

Creutzfeldt-Jakob disease (CJD)

Creutzfeldt-Jakob disease (CJD) is a rare brain disease that is caused by abnormal brain proteins called prions. Classical CJD usually occurs by chance. Variant CJD, which is linked to consumption of contaminated meat products, does not occur in Australia.

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What is Creutzfeldt-Jakob disease?

Creutzfeldt-Jakob disease, also known as CJD, is a rare degenerative disease of the brain that is fatal. It is one of a group of diseases known as the transmissible spongiform encephalopathies.

In CJD, the structure of a normal brain protein changes slightly forming prions. The build-up of prions damages brain cells and causes the neurological symptoms of CJD. Unlike bacteria or viruses, prions resist normal methods of heat and chemical sterilization and, very rarely, prions can be transmitted to others.

There are two different types of Creutzfeldt-Jakob disease:

- Classical CJD occurs in Australia and about one in one million people per year develops the disease. There are three types of classical CJD. About 90% of classical CJD cases occur by chance (sporadic CJD) and 10% of cases are hereditary (familial or inherited CJD). The disease has very rarely been transferred between patients following medical procedures such as brain surgery or the use of dura mater grafts or contaminated human pituitary hormones (iatrogenic CJD).
- Variant CJD, is a disease that emerged in the UK in the 1990s. Variant CJD is linked to the consumption of meat products from cattle infected with bovine spongiform encephalopathy (BSE, or "mad cow disease"). Variant CJD is a separate disease to classical CJD, although some of the symptoms are similar. No cases of variant CJD have been identified in Australia to date. Australian cattle remain free of BSE.

What are the symptoms?

People with classical CJD have progressive neurological symptoms that may include behavioural changes, blindness, weakness, loss of balance and incoordination, difficulty walking or speaking and muscle spasm. Confusion in the early stages usually progresses to dementia.

The disease is fatal, usually weeks to months after onset of symptoms.

People with variant CJD tend to be younger, have a slower rate of deterioration and tend to have more psychiatric symptoms or personality changes than patients with classical CJD.

How is it spread?

Most cases of CJD occur because of mutations with a person's brain and are not spread from other people.

Some medical procedures carried out on people with CJD have very rarely resulted in the disease being transmitted to other people. For example:

- Human pituitary hormones derived from people who died with CJD resulted in transmission in the past and five of these cases occurred in Australia. Therapeutic pituitary hormones are no longer obtained from this source and no high-risk treatments occurred in Australia after 1985.
- Neurosurgical instruments contaminated following an operation on someone with CJD has resulted in five cases worldwide, and none since the 1970s.
- Dura mater grafts taken from donors who had CJD and used to patch holes in the lining outside the brain; corneal grafts taken from donors who had CJD and used in others have resulted in three cases of transmission worldwide and none in Australia.

Screening procedures and improvements to standards of infection control have made transmission of CJD in the modern health care setting extremely unlikely.

Variant CJD may be spread more easily from person-to-person than classical CJD. In contrast to classical CJD, variant CJD may also be transmitted to humans after eating contaminated meat and meat products from cattle with BSE, via transfusion of blood and blood products, and some other operations if the instruments are not processed properly.

Who is at risk?

Each year, about one in every million Australians develops sporadic CJD and most have no risk factors for the disease. The average age of onset is about 65 years.

Familial or inherited CJD includes familial CJD, Gerstmann-Sträussler-Scheinker (GSS) syndrome and fatal familial insomnia (FFI) and is carried from one generation of a family to the next by abnormal genes.

The risk of transmission in the health care setting is extremely low.

How is it prevented?

Special infection control precautions are used for patients thought to at risk of CJD. Products or instruments potentially contaminated with prions are removed from use.

In Australia, there is a very low risk of variant CJD; to safeguard the blood supply, people are excluded from donating blood if they have lived in the United Kingdom for more than 6 months between 1980 and 1996.

How is it diagnosed?

A definite diagnosis of Creutzfeldt-Jakob disease can only be made by special tests of the brain tissue; this almost always occurs after the patient has died. Other specialized tests for people with typical signs and symptoms can help to make a diagnosis, but do not confirm the diagnosis.

To confirm the diagnosis after death an autopsy is usually recommended. The RPA Hospital's Department of Neuropathology provides specialised CJD diagnostic services in NSW.

There is no screening test for CJD.

How is it treated?

There is no specific treatment for Creutzfeldt-Jakob disease.

What is the public health response?

Hospitals and laboratories are required to notify patients with Creutzfeldt-Jakob disease to the local public health unit (under the NSW Public Health Act 2010). The [Australian National Creutzfeldt-Jakob Disease Registry](#) coordinates surveillance and testing for CJD cases in Australia. The Registry works with NSW Health and local public health units to identify if cases have had high-risk procedures and to provide specialist advice for clinicians.

Further information

[CJD Support Group Network Australia](#) - the CJDSGN offers support, information and assistance for family members and friends of patients suffering with suspected CJD and other prion disease and for those at increased risk of developing CJD.

Further information for clinicians on testing for CJD, see the RPA Hospital [Department of Neuropathology CJD testing information](#).

[Australian National Creutzfeldt-Jakob Disease Registry](#).

For further information please call your local Public Health Unit on **1300 066 055** or visit the New South Wales Health website www.health.nsw.gov.au.