How is it diagnosed?

A definite diagnosis of Creutzfeldt-Jakob disease can only be made by autopsy of the brain tissue; this occurs after the patient has died. Once a diagnosis of CJD has been confirmed by autopsy a DNA test on tissue can be done if the family wish to know if there is a genetic cause. This is a separate test that needs to be consented to by the family and genetic services can be helpful in assisting families through the decision process. This specialised test for people with typical signs and symptoms can help to make a provisional diagnosis, but do not confirm the diagnosis. There is no screening test for CJD prior to the onset of symptoms.

How is it treated?

There is no specific treatment for Creutzfeldt-Jakob disease, however support care will be offered.

Can I still have visitors?

Yes – you can still have visitors.

How will my care change whilst in hospital?

Your care won't change in the ward or in any other area of the hospital, standard precautions will be used as for all patients. There will be review of the risks of any surgery to be performed and processes for reprocessing reusable instruments and equipment in the operating theatre and where appropriate (base on risk) single use items will be used if you are undergoing high CJD risk surgery. Whilst in the ward and recovery, your care will be no different if you had not CJD alert risk.

Good hand hygiene practices

Hand Hygiene is the most effective way to prevent ALL infections, including the flu and the common cold. Encourage your family and friends to maintain good hand hygiene practices every day. Hand Washing Alternative - Alcohol based hand rub / gel. During your stay in hospital you may have seen the staff using an alcohol-based hand rub, as an alternative to soap and water. Alcohol-based hand rubs or gels can be used for hand hygiene as long as your hands are not visibly soiled / dirty.

Where can I get further information?

CJD Support Network
National Toll Free Number 1800 052466or email:
contactus@cjdsupport.org.au

References

National CJD support group network
Australian CJD guidelines, 16th Jan 2013
NSW health CJD patient fact sheet. March 2019

Ramsay Health Care

ramsayhealth.com.au

People caring for people.

MR0024462 RHC CGU Patient Information - CJD DL 0221

Creutzfeldt-Jakob disease

Patient Information

Ramsay Health Care

About Creutzfeldt-Jakob disease

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

CJD – two different types

1. Classical CJD (eCJD), the term 'Classical CJD' is used in the National Infection Control Guidelines (CJD 2013) to describe all forms (inherited, sporadic & acquired) of human CJD except Variant CJD (vCJD). Classical CJD occurs in Australia and about one in one million people per year develops the disease. Approximately 90% of CJD cases occur by chance (sporadic CJD) and 10% of cases are hereditary (familial or inherited CJD). Extremely rare cases of the disease being transferred between patients following medical procedures is known as iatrogenic CJD.
2. Variant CJD, also commonly known as 'Mad Cow' disease is a disease that emerged from the UK in the 1990's. Variant CJD is related to the consumption of beef where the cow has been affected by bovine spongiform encephalopathy (BSE) and to date we have had no reported cases of vCJD in Australia.

What are the symptoms?

People with classical CJD have progressive neurological symptoms that may include behavioral 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 Creutzfeldt-Jakob disease spread?

Most cases of CJD occur because of changes to proteins (referred to a as a prion) within 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.

The national infection control guidelines (CJD 2013) recommend special precautions be followed for instruments used on these patients but only when surgery involves high infectivity tissue such as brain, spine and posterior section of the eye. Screening procedures and improvements to standards of infection prevention and 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 meat and meat products from cattle with BSE, via transfusion of blood and blood products, and some other operations if the instruments are not re-processed properly.

Who is at risk?

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 TSE 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. Most forms of familial CJD are impossible to differentiate from sporadic CJD and it is not until a gene test is done that a genetic cause can be established or ruled out. Patients with GSS often survive for several years and patients with FFI suffer from a progressive and untreatable form of insomnia

The risk of transmission in the health care setting (iatrogenic) is extremely low. CJD is not contagious and cannot be transmitted from person to person by normal contact. Nursing a CJD patient uses standard precautions and personal contact e.g. kissing a loved one with CJD does not pose any risk of transmission.

How is it prevented?

Special infection control precautions are used for handling of instruments and equipment for patients thought to at risk of CJD. Prions are remarkably resistant to conventional sterilisation and disinfectant techniques, 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.