

IMPORTANT FACTS ABOUT CJD AND OTHER FORMS OF PRION DISEASE

- To date there have been no reported cases of vCJD in Australia.
- BSE has not been found in livestock in Australia.
- CJD is unique in that it can be genetic and is transmissible.
- Prions are remarkably resistant to conventional sterilisation and disinfectant techniques.
- CJD is not contagious and cannot be transmitted from person to person by normal contact. Nursing a CJD patient or kissing a loved one with CJD does not pose any risk of transmission.

- CJD is invariably fatal with presently no effective treatments and no cure.
- There is no test available to detect cCJD prior to the onset of symptoms.
- Although a possible or probable diagnosis can be made during the clinical illness the only way to definitely confirm CJD is by autopsy.
- Most forms of genetic 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.
- Family members of a confirmed sporadic CJD patient have the same chance of developing CJD as the general population. One in a million!

ALSO AVAILABLE

- Information packs for families
- Information packs for health care professionals
- DVD 'Understanding CJD'

CJDSGN acknowledges funding provided by the Department of Health & Ageing for the production of this brochure.

The information is provided by the **CJD Support Group Network**

For more detailed information please refer to our fact sheets

CONTACT US

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WHAT IS CJD?

CREUTZFELDT-JAKOB DISEASE

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WHAT IS (CJD)?

CJD is a rare and fatal degenerative brain disease in humans.

CJD is one of a group of diseases that affects humans and animals known as transmissible spongiform encephalopathies (TSE) or prion disease.

In animals, the best known TSE is BSE (bovine spongiform encephalopathy) commonly referred to by the media as 'Mad Cow Disease'.

CLASSICAL CJD (cCJD)

For simplicity, the term 'Classical CJD' is used to describe all forms of human CJD except Variant CJD (vCJD).

In Australia we average about 25 cases of classical CJD a year. Symptoms of CJD include loss of memory, dementia, confusion, patients often become clumsy and lack coordination which is known as ataxia and can develop jerking movements known as myoclonus. The rapid decline, particularly with sporadic CJD is very characteristic.

Classical CJD includes:

- * Sporadic CJD (sCJD)
- * Genetic (gCJD)
- * Iatrogenic (iCJD)

SPORADIC CJD (sCJD)

Sporadic CJD (sCJD) is a rapidly progressive disease that has no known cause. It occurs at random in about one person per million of the population per year and accounts for 85% – 90% of all cases of prion disease. sCJD mainly affects people in the 50 – 70 year age group. The length of illness can vary but sporadic CJD is often recognised for the rapid progression with survival usually only 4– 6 months.

INHERITED FORMS OF PRION DISEASE

Genetic CJD (gCJD) accounts for only between 5% and 15% of cases of classical CJD and includes:

- * Familial CJD (fCJD)
- * Gerstmann Straussler Scheinker Disease (GSS)
- * Fatal Familial Insomnia (FFI)

Genetic CJD is usually recognised from a family history of the illness in two or more blood relatives or can be diagnosed from a positive prion protein gene (PRNP) test. A person carrying a genetic mutation has a 50% chance of passing it onto each of their children. Someone carrying the gene will most likely develop the illness in their lifetime, however there is no test to know when or if they will live long enough to become symptomatic.

IATROGENIC CJD

Iatrogenic CJD is the form associated with medical treatments. Although rare, iatrogenic CJD has occurred due to the use of human derived pituitary hormones for fertility and short stature, dura mater grafts and corneal transplants. There is also a recognised risk of transmission from the use of contaminated instruments used in procedures involving high infectivity tissues. Iatrogenic CJD is an acquired form of classical CJD.

OTHER ACQUIRED FORMS OF PRION DISEASE

VARIANT CJD (vCJD)

Variant CJD first recognised in 1996 relates to the consumption of BSE contaminated products. It is now known that vCJD can be transmitted through blood and blood products. Variant CJD, also referred to by the media as 'Mad Cow Disease' is clinically quite different to classical CJD, has a longer duration of illness, presents often with psychiatric symptoms and affects a much younger age group.

KURU

Kuru is an acquired form of prion disease. In the 1960's it was realised that Kuru, affecting the Fore people in the Eastern Highlands of Papua New Guinea, was transmitted through cannibalism.