

Creutzfeldt-Jakob disease (CJD) – the facts

What is Creutzfeldt-Jakob disease?

Creutzfeldt-Jakob disease (CJD) is a rapidly progressive disease causing damage to the brain. It is one of a group of rare diseases that affects humans and animals, known as the transmissible spongiform encephalopathies or prion diseases. Creutzfeldt-Jakob disease causes dementia and walking difficulties. Death can occur up to two years after the first symptoms, however, most people die within six months. There is no treatment or cure.

CJD is the main prion disease affecting humans. It was first recognised in humans in the 1920s. The human prion diseases include:

- sporadic CJD, which causes 85 to 90 per cent of cases
- genetic CJD, which causes around 13 per cent of cases
- healthcare acquired CJD
- variant CJD
- kuru.

Sporadic CJD

Sporadic CJD accounts for the greatest number of human deaths from this group of diseases (around 85 per cent). CJD affects approximately one in every million people each year. So, in the Australian population of about 20 million, there are likely to be approximately 20 cases each year. CJD most often affects people between 50 and 70 years of age.

Genetic disease

Genetic prion disease is extremely rare and is usually recognised from a family history of the illness in brothers, sisters or parents. It is an inherited disease, passed from a parent to child at conception through the body's genetic material (DNA). The disease is not always passed on; each child born from a parent carrying genetic CJD has a 50 per cent chance of inheriting the disease-causing mutation.

Healthcare acquired CJD

Healthcare acquired CJD has occurred worldwide as a result of some medical treatments. Treatments that have transmitted CJD include:

- the use of human pituitary extract hormone for infertility or short stature (five cases in Australia – most recently in 1991)
- dura mater grafts used in brain surgery to repair damage to the membrane covering the brain (five cases in Australia)
- corneal grafts (three cases worldwide)
- exposure to contaminated neurosurgical equipment (five cases worldwide).

The risk of healthcare acquired CJD has been reduced in Australia by ceasing the use of human tissue in growth hormone treatments and dura mater grafts, and by establishing stronger guidelines for organ donation and for sterilising neurosurgical equipment.

Is CJD the same as mad cow disease?

No, while variant CJD sounds like CJD in name, they are different diseases. Variant CJD, the human form of bovine spongiform encephalopathy (BSE), sometimes called 'mad cow disease', was first recognised in 1996 in the United Kingdom. It has not been found in Australia. BSE is a prion disease that occurs in cattle. The British epidemic of BSE in the 1980s was most likely caused by the transmission of a disease of sheep, scrapie, to cattle through the food chain. Scrapie and BSE have not been found in Australian livestock.

Kuru

Kuru is a human prion disease found only in the central highlands of New Guinea. It was caused by the practice of ritualised cannibalism of deceased relatives. The practice has been discontinued and the number of Kuru cases has therefore declined over time. Kuru has never been found outside of Papua New Guinea.

Symptoms of CJD

CJD is difficult to diagnose. The early symptoms can be vague and there are no tests that can tell whether someone has been exposed or to diagnose CJD until symptoms are well advanced. As the disease progresses, extensive investigations are needed to check if the person has something else that may be treatable. Diagnosis is only made as the illness progresses and tests suggest the disease. Examination of brain tissue after death is the only way to confirm CJD. Symptoms may include:

- confusion or disorientation, which rapidly advances to a dementia
- personality changes
- behavioural changes
- weakness, or loss of balance and muscle control, causing difficulty walking
- muscle spasms
- visual symptoms such as double vision or blindness.

Most people with these symptoms do not have CJD, but other causes of their symptoms.

CJD is not transmitted by casual contact

CJD is not transmitted by casual contact like drinking from the same cup, kissing or close physical contact with an individual suffering from CJD.

For further information, contact the Communicable Disease Prevention and Control Unit at the Department of Health on 1300 651 160 or visit www.betterhealth.vic.gov.au.

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