Creutzfeldt-Jakob disease

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 is characterised by 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 human form of prion diseases. CJD 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 approximately 13 per cent of all cases of CJD worldwide.
- Health care acquired CJD.
- Variant CJD.
- Kuru.

Sporadic CJD
Sporadic CJD accounts for the greatest number of human deaths from this group of diseases (approximately 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 in one year. CJD most often affects people between the ages of 50 to 70 years.

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.

Health care acquired CJD
Health care acquired CJD has occurred worldwide as a result of a number of medical treatments. Treatments shown to have transmitted CJD include:

- The use of human pituitary extract hormone for infertility or short stature (five cases in Australia).
- 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 first two of these treatments used products developed from human tissues. Once CJD was identified as a hazard, the use of these products was immediately discontinued, synthetic alternatives developed, sterilising procedures increased. Stronger guidelines have also been established for the use of organ transplants and for the sterilisation of surgical equipment to reduce the risk of CJD acquired through organ donation or through contaminated neurosurgical equipment.

Australian cases of health care acquired CJD
There have been ten cases of health care acquired CJD in Australia. They consist of five deaths following treatment with pituitary extract hormone for either infertility or short stature, with no

further cases since 1991. The five other deaths were caused by dura grafting during brain surgery, where the covering of the brain was repaired. There have been no other health care acquired CJD deaths in Australia.

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 correspondingly 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 firm diagnostic tests to confirm exposure, or diagnose CJD, until symptoms are well advanced. As the disease progresses, extensive investigations are necessary to exclude the possibility of other treatable diseases. Diagnosis is made only as the illness progresses and with results of tests suggesting 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.

It is essential to remember that most people with these symptoms do NOT have CJD, but other causes of the 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.

Where to get help

- If the question relates to a patient of the Royal Melbourne Hospital The direct phone number is 1800 246 211 which is available Monday to Friday from 8 am to 8 pm.
- Public Health, Department of Human Services 03 96374126
- Office of Chief Clinical Advisor, Department of Human Services 03 9616 7324