Australia’s response to the first case of variant Creutzfeldt-Jakob disease (vCJD)- A Guide for doctors and health care workers

The vCJD Response Plans

The Australian Government has developed a contingency plan to deal with the possibility of vCJD being detected in Australia. The vCJD Response Plan for Australia outlines how the health system may respond should a case of vCJD be diagnosed in Australia and includes an infection control guideline for vCJD. The Response Plan can be accessed through the Australian Government Department of Health and Ageing web site (www.health.gov.au).

What is vCJD?

vCJD is the human form of bovine spongiform encephalopathy (BSE, ‘mad cow disease’). It is believed that vCJD is acquired by humans through the consumption of beef infected with the BSE agent. vCJD has distinct clinical-pathological features compared to other forms of human CJD, which are referred to as classical CJD (cCJD). All forms of CJD are transmissible spongiform encephalopathies (TSEs), where the infectious agent is an abnormal, protease-resistant prion protein, relatively resistant to routine instrument processing and sterilisation methods currently used in health care establishments.

It is important to recognise that CJD and vCJD are different diseases. CJD usually occurs randomly in the general community; around 20 cases are diagnosed each year in Australia. In 10-15% of cases, CJD is an inherited disease and rarely, transmission of CJD occurs iatrogenically, through medical procedures or therapies involving infectious human tissues or material (such as through dura mater grafts or from receiving human pituitary growth hormones).

Once the abnormal prion protein has entered the body it is believed that it infects lymphoreticular tissue, such as tonsil and spleen, where levels build up before travelling to the brain. The brain is unable to clear the abnormal form and it starts to accumulate, causing the spongy appearance of an affected brain.

To date, over 200 people have been infected with vCJD, the majority in the United Kingdom (UK), with a small number also occurring in France, Ireland, Italy, Canada and the United States of America.
What are the symptoms of vCJD?

The severity and course of symptoms described below will differ slightly between individuals affected with vCJD. In the early stages of the disease, individuals affected usually experience psychiatric symptoms, such as depression and many become emotionless and withdrawn. Less often, fleeting delusions may develop. Sensory disturbance, such as foot pain and loss of feeling in hands and face may also be experienced. Usually within 6 months of disease onset, neurological signs begin to develop, mainly in the form of muscular incoordination with unsteadiness, difficulty walking and involuntary movements. Those individuals affected will eventually become increasingly dependent, unresponsive and unable to speak. vCJD is always fatal; patients usually die within 18 months of the onset of the disease.

A sub-committee of the Communicable Disease Network Australia (CDNA) has reviewed the Australian case definition for vCJD. The case definition is included in the vCJD Response Plan for Australia. This case definition is published on the CDNA website (www.health.gov.au/CDNA) and the website of the National Notifiable Diseases Surveillance System (www.health.gov.au/internet/main/publishing.nsf/content/cda_surveil-nndss-dislist.htm).

Definitive diagnosis of all forms of CJD can only be obtained by brain autopsy or biopsy, however, the latter is not recommended by the World Health Organization (WHO). Although a definite diagnosis of vCJD cannot be achieved using clinical criteria alone, it should be considered in the differential diagnosis of a patient presenting with the clinical criteria presented in the case definition.

When would the vCJD Response Plan be used?

The vCJD response plan would be used when a physician suspects that an individual may be presenting with vCJD. The physician may ask about the individual's travel history—especially travel to the UK and Europe. As the early symptoms of vCJD are similar to a number of other diseases, the physician may advise that a range of tests be carried out to exclude other options. The Australian National CJD Registry can advise on diagnostic testing procedures if vCJD is considered in the differential diagnosis. These tests may include:

**MRI:** A characteristic distribution of symmetrical hyperintensity of the pulvinar nucleus of the thalamus, known as the pulvinar sign, is seen in over 90% of vCJD cases. Although it is not possible to conclusively diagnose vCJD from MRI, the pulvinar sign is currently the best non-invasive diagnostic test of vCJD.
EEG: An EEG can be performed to rule out other illnesses, such as sporadic CJD.

Tonsil biopsy: Tonsil tissue can be examined for evidence of abnormal prion protein. Because of the risk of false-positive and false-negative results, tonsil biopsy is recommended in the classification of Probable, but not Definite vCJD.

Lumbar puncture: In most cases a lumbar puncture is conducted to ensure there is no infection of the brain, such as meningitis, which can be treated. However, in some cases a tentative diagnosis of vCJD may be reached when high levels of specific cerebrospinal fluid (CSF) proteins are detected.

CT scan: It is not possible to conclusively diagnose vCJD from a CT scan of the brain.

What treatment is available if vCJD is diagnosed?

At present there is no known treatment that can halt or slow down the progress of the disease. Individuals diagnosed with vCJD may wish to consider participating in clinical trials, which are currently researching potential treatments for vCJD.

There are many services available to support affected individuals and their families as the disease progresses. A representative from the State/Territory Health Department and/or the treating physician can help organise counselling services, home care and hospital admission if and when needed. It is important that affected individuals and their families are supported through all stages of the disease.

What else should be expected during this process?

If vCJD is suspected, the physician should contact the State health authorities and the Australian National CJD Registry, to alert them to the possible diagnosis. The individual and their family may be contacted by the State health authority to seek additional information. This might include:

- Their travel history;
- Questions about foods eaten, both overseas and in Australia;
- Whether there is any history of blood transfusion or donation;
- Questions about past surgical or dental procedures -- particularly any within the previous 12 months; and
- Details of medication/supplements used that were not prescribed by a physician, particularly any supplements ordered over the internet or acquired overseas.
This information will be used by health authorities to trace the source of the infection.

Definitive diagnosis aids prevention of spread of vCJD through blood and organ donation and identification of surgical instruments used on that patient that need to be destroyed. Clinicians should discuss autopsy with the patient and their next-of-kin to obtain permission.


**Where can I obtain further information?**

**State Health Department** - Please refer to your State/Territory Public Health Authorities  
**Australian National CJD Registry** - [www.path.unimelb.edu.au](http://www.path.unimelb.edu.au)  
**Australian Government Department of Health and Ageing** - [www.health.gov.au](http://www.health.gov.au) -  