

Variant Creutzfeldt-Jakob Disease (vCJD) Fact Sheet

What is “variant CJD” (vCJD)?

Variant CJD (vCJD) is a recently identified type of Creutzfeldt-Jakob disease (CJD), a rare, fatal brain disease caused by abnormal prions (microscopic protein particles). (Information on the classic types of CJD is available in the [Creutzfeldt-Jakob Disease Fact Sheet](#)). vCJD was first reported in 1995 in the United Kingdom (UK) following an epidemic of bovine spongiform encephalopathy (BSE) that was occurring among cattle in that country in the 1980s into the 1990s. vCJD in humans and BSE in cattle belong to a class of prion diseases known as transmissible spongiform encephalopathies.

What is bovine spongiform encephalopathy (BSE)?

BSE is a progressive neurological disorder of cattle that results from infection by prions. BSE was first recognized in the UK in the 1980s in cattle that were fed meat and bone meal made from leftovers of cattle and other livestock. Cattle with BSE progress from having no symptoms to worsening behavioral and neurological signs including lethargy, unsteadiness, and aggression.

How do cattle get BSE?

Cattle can get BSE through consumption of meat products from BSE-infected cattle.

How do people get vCJD?

People can get vCJD from eating meat products from BSE-infected cattle or from receiving transfused blood that came from a person with vCJD.

Where have cases of vCJD been reported?

Most cases of vCJD have been reported in the UK and Western Europe, and the number of vCJD cases has decreased since its peak in 2000. In the US, only three cases of vCJD have been identified and all three patients have been determined to have been exposed outside the country. No case of vCJD has been reported in California.

What are the signs and symptoms of vCJD?

Unlike sporadic CJD, vCJD occurs in younger persons (median age 28), has a longer duration of illness, and has different initial signs and symptoms. Symptoms of vCJD usually do not develop until years after exposure. vCJD patients have psychiatric symptoms such as depression, aggression, or anxiety in the beginning of the illness. As the illness progresses, dementia and neurological signs including unsteadiness in walking and sudden jerking movements develop. Death occurs a median of 13-14 months after the onset of symptoms.

How is vCJD diagnosed?

It is hard to diagnose any kind of CJD. Clinical expertise from a neurologist (a medical specialist dealing with the nervous system) is usually needed, and often it is through the process of elimination of other diseases. Examination by a neurologist, laboratory tests, and other medical evaluations may help a doctor to suspect vCJD, but the diagnosis can be confirmed only through a brain biopsy or autopsy.

How is vCJD treated?

There are no effective treatments for any kind of CJD and the disease is invariably fatal. The only medications available for CJD patients focus on easing their symptoms and discomfort.

How is vCJD prevented?

Following the detection of BSE in the UK, the US Department of Agriculture implemented measures to ensure that the supply of beef from US cattle was safe to eat. These ongoing measures include: 1) prohibiting use of rendered cattle as feed for other cattle; 2) prohibiting the meat or products from “downer” cattle with neurologic illness from entering the human food market; 3) eliminating high-risk tissues (e.g., nervous tissue) from entering the human food market; and 4) routine surveillance and testing of cattle. The US Food and Drug Administration instituted a policy that excludes from blood donation anyone who spent at least three months in the UK between 1980 and 1996.

Where can I get more information on vCJD?

To get more information regarding vCJD, please visit:

The U.S. Centers for Disease Control and Prevention website

<http://www.cdc.gov/ncidod/dvrd/prions/index.htm> and

http://www.cdc.gov/ncidod/dvrd/vcjd/factsheet_nvcjd.htm

California Department of Public Health
Veterinary Public Health Section
1616 Capitol Avenue, MS 7308
Sacramento CA 95814
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