

CREUTZFELDT-JAKOB DISEASE Q&A

What is CJD?

Creutzfeldt-Jakob Disease (CJD) is a rare, degenerative, fatal brain disorder. Symptoms include: dementia, loss of motor control, paralysis, and death.

What causes CJD?

CJD is caused by a filterable, self-replicating agent called a prion. A normal version of the protein is found in all people. It is somehow converted to an abnormal form that produces brain lesions resulting in CJD.

How is CJD spread?

CJD is only infectious when infected tissue is passed into the body of someone else. This has happened during surgery, treatment with human growth hormone, certain transplants, and blood transfusion.

How common is CJD?

The Centers for Disease Control and Prevention (CDC) estimates the incidence of CJD to be one case per million population. The highest incidence of CJD occurs in persons over the age of 65 years.

What are the symptoms of CJD?

Symptoms may include depression, memory lapses, and dementia. Symptoms may progress to unsteadiness, lack of coordination, progressive dementia, sudden jerky movement, and eventually the loss of the ability to move or speak.

How soon do symptoms appear after infection?

After infection, symptoms may not appear for fifteen months up to thirty years.

How can a person find out if they have CJD?

Currently, there is no test available to identify the presence of prions causing CJD. Once symptoms begin, diagnosis is made based on clinical symptoms, magnetic resonance imaging



(MRI) to rule out other problems such as tumors, characteristic electroencephalogram (EEG) patterns, the presence of certain proteins in the cerebrospinal fluid, or brain biopsy to detect signs of spongiform change.

What is the treatment for CJD?

There is no known treatment for CJD. There are drugs available to relieve some of the symptoms of CJD.

What is Variant CJD (vCJD)?

Variant CJD is a newly recognized variant of CJD. It is associated with prior exposure to bovine spongiform encephalopathy (BSE), or “mad cow disease.” It predominantly affects younger people compared to CJD. Its initial symptoms differ from other forms of CJD. They include: prominent psychiatric or sensory symptoms and delayed onset of neurologic abnormalities, including unsteadiness and lack of coordination, and dementia. In all cases of CJD, symptoms may progress to unsteadiness, lack of coordination, progressive dementia, sudden jerky movement, and eventually the loss of the ability to move or speak.

What causes vCJD?

Variant CJD (vCJD) is thought to be caused by exposure to bovine spongiform encephalopathy (BSE), or “mad cow disease,” a prion disease found in cattle.

What is “mad cow disease” (BSE)?

Bovine spongiform encephalopathy (BSE), or “mad cow disease,” is a prion disease that occurs in cattle. Exposure to BSE is a risk factor for vCJD.

The United States has been conducting active surveillance for BSE since May 1990. There is no evidence of the presence of BSE in cattle in the United States. To prevent BSE from entering from other countries, the United States placed severe restrictions on the importation of live ruminant and certain ruminant products from countries where BSE is known to exist.

Because of a possible risk of CJD through blood products, the United States and Canada have restricted blood donations coming from donors who spent 3 or more months in the UK or 6 or



more months in BSE endemic countries, or received non-USA licensed bovine insulin or other injectable products from cattle in BSE endemic countries.

What is chronic wasting disease (CWD)?

Chronic wasting disease (CWD) is a prion disease that occurs in deer and elk. CWD typically includes chronic weight loss resulting in death. There is no known relationship between CWD and prion diseases in other animals or people.

Where can I get additional information on CJD, vCJD, BSE, and CWD?

Contact the Georgia Department of Public Health, Epidemiology Branch, by email at dph-gaepinfo@dph.ga.gov. The following web sites may be useful:

- CDC Questions and Answers Regarding Bovine Spongiform Encephalopathy (BSE) and Variant Creutzfeldt-Jakob Disease (vCJD) <http://www.cdc.gov/ncidod/dvrd/vcid/ga.htm>
- CDC vCJD Fact Sheet http://www.cdc.gov/ncidod/dvrd/vcid/factsheet_nvcid.htm
- National Prion Disease Pathology Surveillance Center <http://www.cjdsurveillance.com/>
- USDA BSE News and Information <http://www.usda.gov/wps/portal/usda/usdahome?navtype=SU&navid=BSE>
- USDA Chronic Wasting Disease Information http://www.aphis.usda.gov/animal_health/animal_diseases/cwd/
- American Red Cross Blood Donation Guidelines <http://www.redcrossblood.org/>

