

Blood Safety

The American Red Cross (ARC) is denying blood donations from individuals who have spent six months or more in Europe since 1980, as well as that of any blood relative of a CJD victim. Sporadic CJD is not believed to be transmissible through blood, however, the ARC is taking strict precautions. The first case of suspected blood transfusion transmission occurred in the U.K in 2003 through blood donated from a pre-symptomatic variant CJD victim. Even though scientists can't provide positive proof that this case was transfusion-transmitted, in order to protect public health this should be considered the first case. There have been two other cases of suspected transfusion-transmission in the U.K. In 2009, the U.K. authorities also reported possible transmission of vCJD through a treatment for hemophilia produced from blood plasma

Other Concerns

The public is at risk because the federal surveillance system for monitoring CJD and BSE incidence is poorly funded and fragmented. There is no uniform reporting mechanism in place and each state sets its own requirements. The public is at risk because so little is known about how to detect and treat CJD. Until the gap in that continuum is closed, the public will remain at risk. The linkage of CJD, BSE, the blood supply, and the lack of

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knowledge and reporting, unfortunately, combines to fuel those fears.

From a public policy perspective, a deeper understanding of CJD - both its causes and the rate of incidence, would provide direction for scientists in their goals to better understand prion diseases and the ultimate goal to find the cure.

Chronic Wasting Disease (CWD)

Chronic Wasting Disease was first diagnosed in 1967 in Colorado, and it has subsequently been found in captive herds in several other areas. The source of infection for wild and captive deer and elk in new geographical areas is unknown in many instances. While it is not known exactly how CWD is transmitted, lateral spread from animal to animal through shedding of the infectious agent from the digestive tract appears to be important, and indirect transmission through environmental contamination with infective material likely. CWD has not been proven to cross the species barrier.

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Creutzfeldt-Jakob Disease
Foundation, Inc.

CJD FACT SHEET



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What is CJD?

Creutzfeldt-Jakob Disease (CJD) is a rare, fatal brain disorder. The incidence of CJD cases worldwide is one case, per million, per population. In the United States this statistic translates to approximately 300 new cases per year. Eighty-five percent of the cases are sporadic, meaning there is no known cause at present.

There are three types of CJD:

- Sporadic (sCJD)
- Familial (fCJD)
- Acquired (vCJD or iCJD)

In the early stages of the disease, CJD patients may exhibit failing memory, behavior changes, impaired coordination and/or visual disturbances. As the illness progresses, mental deterioration becomes more pronounced, and involuntary movements, blindness, weakness of extremities, and, ultimately, coma may occur. sCJD usually occurs later in life and typically leads to death within a few weeks or months to one year following the onset of symptoms.

HelpLine 1.800.659.1991

Causes of CJD

Sporadic CJD

- Unknown

Familial CJD

- Genetic mutation inherited from a parent

Iatrogenic CJD

- Contaminated surgical instruments
- Contaminated dura mater transplant
- Contaminated corneal transplant
- Contaminated human growth hormone

Variant CJD

- Contaminated beef
- Contaminated blood or blood plasma transfusion

The sporadic form of CJD is the most prevalent form of CJD affecting approximately 300 new people in the United States each year. The familial form of CJD accounts for 10-15% of the cases. Acquired CJD, which includes iatrogenic and variant CJD, accounts for less than 1% of all cases. More information can be found on www.cjdfoundation.org. As of May 2009, there are no known cases of endemic vCJD in the United States.

Diagnosis of CJD is very difficult and is often made from clinical observation and/or process of elimination of other diseases. The diagnosis of CJD can only be confirmed through a brain biopsy or autopsy. Cerebral spinal fluid testing positive for a 14-3-3 or tau protein is often used to confirm a possible diagnosis, this test, however, can be ambiguous.

Public Health Concerns

CJD is NOT “Mad Cow Disease.” Bovine Spongiform Encephalopathy (BSE), the technical term for Mad Cow Disease, occurs only in cows.

The first documented case of BSE found in the United States occurred in Washington State in December 2003 in a cow imported from Canada. The first endemic case was found in Texas and was announced in 2005. Eating infected beef is widely believed to be the cause of the variant form of CJD (**vCJD**) in humans. vCJD usually affects young people. An endemic case has not yet been documented as originating in the United States.

Is the public at risk of exposure to CJD?

Although most Americans have never heard of Creutzfeldt-Jakob Disease, they have heard of “Mad Cow Disease” and fear it. They do not know what it is, but know it is catastrophic. The confirmation of a case of vCJD originating in the U.S. would likely lead to a chain reaction of panic-driven decisions and policymaking.

www.cjdfoundation.org