

## Creutzfeldt-Jakob Disease Information

Creutzfeldt-Jakob Disease is a degenerative brain disease that has been recognized since the 1920s. It is a human prion (infectious protein) disease that progresses rapidly and is always fatal, usually within one year from onset of illness.

CJD is not “Mad Cow disease” in people, which is caused by a different prion and is technically called variant CJD or vCJD. CJD is not transmissible from person-to-person through ordinary contact, but it can be transmitted from one person to another by contaminated instruments used for brain surgery or by transplant of infected tissue. There is no known environmental trigger for development of CJD. For more information on CJD, see <http://www.cdc.gov/ncidod/dvrd/cjd/> and the links under “Where can I get more information about CJD?”

### Creutzfeldt-Jakob Disease (CJD) in Idaho: Historical

CJD was not reportable in Idaho until 2004. Prior to that, information on the number of cases of CJD was based on death certificates. Among Idaho residents in the last 20 years, there have been 25 deaths reported on death certificates listing CJD as the cause.

The annual number of reported CJD deaths in Idaho during the last 20 years ranged from zero to three. CJD deaths were reported from 17 Idaho counties. From 1985 to 2004, only 10 of 25 (40%) of persons with CJD on their death certificate had received an autopsy. Autopsy is the only way to confirm the diagnosis of CJD.

### Creutzfeldt-Jakob Disease (CJD) Deaths among Idaho Residents: 1985-2003, and 2004 Preliminary

Year	Number of Deaths	Counties
1985	0	
1986	3	Bonneville, Gem, Washington
1987	0	
1988	3	Ada, Canyon, Twin Falls
1989	2	Minidoka, Nez Perce
1990	0	
1991	0	
1992	0	
1993	3	Bonner, Canyon, Minidoka
1994	2	Ada, Gooding
1995	2	Ada, Twin Falls
1996	2	Bannock, Canyon
1997	1	Payette
1998	1	Idaho
1999	1	Ada
2000	2	Canyon, Owyhee
2001	1	Elmore
2002	0	
2003	1	Kootenai
2004 Preliminary	1	Franklin
Total 1985-2003, and 2004 Preliminary	25	

*\*Number of deaths reported on death certificate as CJD, 1985-2004 (2004 preliminary). Idaho resident deaths may have occurred in Idaho or out of state. ICD-9 code used from 1985-1998: 046.1; ICD-10 code used from 1999-2003: A81.0 (Creutzfeldt-Jakob Disease) and B94.8 (Sequelae of other specified infectious and parasitic diseases and Creutzfeldt-Jakob Disease is specified). Data for 2004 are preliminary and are based on records filed as of August 12, 2005.*

*Source: Bureau of Health Policy and Vital Statistics, Idaho Department of Health and Welfare (8/2005).*

### **CJD in Idaho: 2005**

**South Central Idaho.** In late July 2005, South Central District Health began investigating reports of possible CJD cases. Five possible cases reported between February and July 2005 were investigated by South Central District Health and the Idaho Department of Health and Welfare, Office of Epidemiology and Food Protection. The Centers for Disease Control and Prevention (CDC) was also consulted.

The investigation included a review of medical records, including autopsy results if an autopsy was performed, and interviews with treating physicians. A survey for information on dietary habits, residence, travel, occupation, surgeries, and other life experiences also was conducted through interviews with family members.

#### ***South Central Idaho Investigation Findings***

Autopsies are the only method to verify CJD as the cause of death. Of the five people who were reported as possibly having CJD, autopsies were conducted on three. Laboratory results provide an initial test result on whether the disease was a prion disease, which includes CJD, and may be followed by additional testing for more in-depth information on the type of prion disease.

To date, initial autopsy results show a prion disease in two people, with tests indicating one person was not infected with a prion disease. Final lab results indicate neither person died from variant CJD. Molecular lab tests also indicate no correlation between the two confirmed CJD cases.

Review of the information on the two non-autopsied cases by a CDC neuroepidemiologist determined that the evidence was not compelling enough to classify the cases as possible or probable CJD according to World Health Organization (WHO) case definitions for CJD.

#### ***Additional reports of possible CJD***

The Idaho local public health districts and State health department monitor additional reports of possible cases of CJD. With each report, medical records are examined and interviews conducted with the physicians and families, if necessary. Local and state health officials investigate and evaluate each report. Four additional reports were investigated in 2005:

- A report on an Elmore County man in August was investigated with the Central District Health Department. Autopsy results indicate the person did not have a prion disease.

- A report on a woman in her 50s from Benewah County was investigated with Panhandle Health District. Final autopsy results confirm she had CJD, not variant CJD.
- A report on a woman over the age of 55 from Bear Lake County was investigated with Southeastern Health District. The person was not autopsied. Review of information on this case by a CDC neuroepidemiologist resulted in its classification as probable CJD according to WHO case definitions.
- A report on a man over the age of 55 from Caribou County was also investigated with the Southeastern Health District. The person also was not autopsied. Review of information on this case by a CDC neuroepidemiologist resulted in its classification as probable CJD according to WHO case definitions.

### ***2005 Statewide Summary***

In summary, nine reports of persons in whom CJD was being considered as the cause of their illness were brought to the attention of IDHW in 2005. Of these persons, five were autopsied and four were not. Of the five autopsied persons, two did not have prion disease and three had CJD. Of the four cases in which an autopsy was not performed, two met the WHO case definition for probable CJD and in two the evidence was not compelling enough to classify the cases as either probable or possible CJD. None of the reported cases were compatible with variant CJD and there was no evidence of iatrogenic transmission. It appears that the apparent increase in the number of reported CJD cases in Idaho in 2005, of which three were confirmed and two were probable, was likely due to enhanced surveillance activities, although chance occurrence cannot be ruled out.

**For frequently asked questions about CJD and additional resources concerning CJD, click [here](#).**

**For the Idaho Disease Bulletin concerning CJD, dated August 2004, click [here](#).**

**For the Idaho Disease Bulletin concerning CJD, dated October 2005, click [here](#).**