CREUTZFELDT-JAKOB DISEASE FACT SHEET

1. **What is Creutzfeldt-Jakob Disease (CJD)?** – CJD is a fatal prion disease affecting the brain and spinal cord of humans that has been known since the 1920's. The word *"prion"* is an abbreviation for *"infectious protein."*

a. Specifically, a prion is a malformed protein capable of causing other normal prion proteins to also miss-fold and, thereby, self-replicate. The key to understanding prion disease is an understanding that the three-dimensional shape of all proteins is critical to their normal cellular function. The abnormally shaped prion serves no function and is not easily removed.

b. In CJD prion proteins accumulate exponentially causing progressive brain damage, gait and balance disturbances, difficulty swallowing, weight loss, behavioral changes, and death. When viewed at autopsy, stained tissue slides from the brain are filled with characteristic microscopic holes created by the excess accumulation of the abnormal prion protein. CJD incidence increases with age with most disease being diagnosed in persons over 50 years of age.

c. The two important forms of CJD are "classic CJD" which occurs worldwide, and a rare CJD variant disease (vCJD) occurring in young people and associated with eating beef in Europe. Classic CJD is not related to *"mad cow"* disease in cattle, or to vCJD in humans.

d. The most common form of classic CJD occurs in a scattered and infrequent pattern caused by the spontaneous development of an abnormal prion that then proceeds to change other normal prion proteins into the abnormal shape. Spontaneous CJD occurs worldwide at an annual rate of one to two cases per 1,000,000 human population. About 12 cases of classic CJD would be expected to occur in Pennsylvania each year.

2. **What are the symptoms of CJD?** – CJD results in abnormal behavior, memory loss, gait and balance disturbances, and difficulty swallowing.

3. **Is there a treatment for CJD?** – There is no specific treatment. CJD is rapidly progressive and always fatal. Death usually occurs within 1 year following onset of illness.

4. **What is vCJD disease?**

a. In contrast to the classic form of CJD, the vCJD form mostly affects younger persons and has a very different clinical presentation and subsequent laboratory test results.

b. Bovine Spongiform Encephalopathy (BSE) is a fatal prion brain disorder of cattle. It is also known as mad-cow disease. Scientific evidence suggests that the vCJD outbreak began when humans ate BSE infected beef. In the United Kingdom (U.K.), where over 1 million cattle have been infected with BSE, a substantial species transmission barrier appears to protect humans from more widespread illness. As of August 2005, a total of 179 cases of vCJD had been reported worldwide; of these, 156 had occurred in the U.K. Based on this data, the increased risk to human health from BSE in the U.S. is considered to be extremely low, but not zero.

5. **Is there evidence directly linking vCJD to BSE exposure?**

a. There is strong epidemiologic and laboratory evidence for a causal association between vCJD and BSE. The absence of confirmed cases of vCJD in other geographic areas free of BSE supports a causal association. In addition, the interval between the most likely period for human exposure to potentially BSE-contaminated food (1984-1986) and onset of the first vCJD cases (1994-1996) is consistent with known incubation periods for CJD.

b. Several experimental studies with mice and monkeys support this causal link.

6. **Is there any monitoring of the incidence of classic CJD in the United States?** -

a. Yes. The Centers for Disease Control and Prevention (CDC) monitors trends and the current incidence of CJD in the U.S. by analyzing death certificate information from multiple cause-of-death data.

b. The average annual CJD death rate in the U.S. has remained relatively stable at about one case per million population per year.

c. In addition, CJD deaths in persons aged <30 years in the U.S. remain extremely rare (<1 case per 100 million per year). In contrast, in the U.K., over half of the patients who died with vCJD were in this young age group.

7. **Are increased surveillance efforts in place to determine whether the newly recognized variant of CJD occurs in the United States?**

a. Yes. In addition to ongoing review of national CJD mortality data, the Centers for Disease Control and Prevention (CDC) have conducted active CJD surveillance in its Emerging Infections Program Monitoring Sites.

b. Additionally, with the support of all state health departments and the Council of State and Territorial Epidemiologists, CDC conducts follow-up review of clinical and neuropathology records of people who died of CJD aged <55 years who are identified through the national mortality data analysis.

c. In 1997, CDC, in collaboration with the American Association of Neuropathologists, established the National Prion Disease Pathology Surveillance Center at Case Western Reserve University, Cleveland, Ohio in 1996-1997. This pathology center provides free, state-of-the-art diagnostic services to U.S. physicians. It also helps to monitor the possible occurrence of emerging forms of prion diseases, such as vCJD, in the U.S. See its website at: http://www.cjdsurveillance.com.

8. **Has there ever been a human vCJD case in the U.S.?** - As of June 2008, the total number of vCJD cases identified in residents of the United States is three; all of which were epidemiologically linked to likely exposures to cattle products contaminated with bovine spongiform encephalopathy (BSE, commonly known as "mad cow disease")while residing in the United Kingdom (2 cases) or Saudi Arabia (1 case).

9. **Is BSE a food borne hazard for travelers to Europe?** - The current risk for infection with the BSE agent among travelers to Europe is extremely small, if it exists at all.

10. **Are there other human forms of CJD?** - Whereas the majority of cases of classic CJD (about 85%) occur as sporadic (random) disease, a smaller proportion of patients (5-15%) develop CJD because of inherited mutations. These inherited forms include Gerstmann-Straussler-Scheinker syndrome and fatal familial insomnia.

11. **For more information:** http://www.cdc.gov/ncidod/dvrd/cjd/index.htm

This fact sheet provides general information. Please contact your physician and/or veterinarian for specific clinical information related to you or your animal.

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