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CDC HEALTH UPDATE

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CDC and Florida Department of Health investigate a likely case of new variant Creutzfeldt Jakob disease in a U.K. citizen residing in the U.S.

Atlanta: The Florida Department of Health and the CDC are investigating a likely case of new variant Creutzfeldt Jakob disease (vCJD) in a 22-year-old citizen of the United Kingdom living in Florida. The clinical diagnosis was made at a hospital in the U.K and she has since returned to the U.S. Preliminary analysis of information provided by the U.K. indicates that the patient's clinical condition and history are consistent with vCJD acquired in the U.K. However, the only way to confirm a diagnosis of vCJD is through study of brain tissue obtained by a brain biopsy or at autopsy.

New variant CJD is a rare, degenerative, fatal brain disorder that emerged in the U.K. in the mid-1990s. Although experience with this new disease is limited, evidence to date indicates that there has never been a case transmitted from person to person. Rather, the disease is thought to result from consumption of cattle products contaminated with an agent that causes a disease called bovine spongiform encephalopathy (BSE, commonly known as mad cow disease). To date, no case of this cattle disease has been identified in the United States by the USDA

If confirmed, this would be the first case of vCJD reported in a U.S. resident. However, because the disease is thought to have a long incubation period, CDC believes the patient acquired the disease while living in the U.K.

While very tragic, it was not unexpected that a case of vCJD would be identified in the United States in a person who had lived in countries experiencing BSE. Of the 125 vCJD patients worldwide, almost all had multiple-year exposures in the United Kingdom between 1980 and 1996 during the occurrence of a large UK outbreak of BSE among cattle. The risk of being exposed to BSE as a traveler in Europe is extremely small.

CDC received information yesterday, April 17, that this patient's illness was a likely case of vCJD and dispatched a medical epidemiologist who is working with the Florida Department of Health to gather more information.

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New Variant CJD: Fact Sheet

New variant CJD (vCJD) is a rare, degenerative, fatal brain disorder.

Although experience with this new disease is limited, evidence to date indicates that there has never been a case transmitted person to person.

As of April 2, 2002, a total of 125 cases of vCJD had been reported in the world: 117 from the United Kingdom, six from France, and one each from Ireland and Italy.

Almost all the 125 vCJD patients had multiple-year exposures in the United Kingdom between 1980 and 1996 during the occurrence of a large UK outbreak of bovine spongiform encephalopathy (BSE, commonly known as mad cow disease) among cattle.

There has never been a case of vCJD that did not have a history of exposure within a country where this cattle disease, BSE, was occurring.

To date, despite an active USDA surveillance program, no case of this cattle disease has been identified in the United States.

It is believed that the persons who have developed vCJD became infected through their consumption of cattle products contaminated with the agent of BSE. There is no known treatment of vCJD and it is invariably fatal.

The vCJD should not be confused with the classic form of CJD that is endemic throughout the world, including the United States. The median age at death of patients with classic CJD in the United States, for example, is 68 years, and very few cases occur in persons under 30 years of age. In contrast, the median age at death of patients with vCJD in the United Kingdom is 28 years.

The vCJD can be confirmed only through examination of brain tissue obtained by biopsy or at autopsy, but a " probable case" of vCJD can be diagnosed on the basis of clinical criteria developed in the United Kingdom.

The incubation period for vCJD is unknown because it is a new disease. However, it is likely that ultimately this incubation period will be measured in terms of many years or decades. In other words, whenever a person develops vCJD from consuming a BSE-contaminated product, he or she likely would have consumed that product many years or a decade or more earlier.

In contrast to classic CJD, vCJD in the United Kingdom predominantly affects younger people, has atypical clinical features, with prominent psychiatric or sensory symptoms at the time of clinical presentation and delayed onset of neurologic abnormalities, including ataxia within weeks or months, dementia and myoclonus late in the illness, a duration of illness of at least 6 months, and a diffusely abnormal non-diagnostic electroencephalogram.

The BSE epidemic in the United Kingdom reached its peak incidence in January 1993 at almost 1,000 new cases per week. The outbreak may have resulted from the feeding of scrapie-containing sheep meat-and-bone meal to cattle. There is strong evidence and general agreement that the outbreak was amplified by feeding rendered bovine meat-and-bone meal to young calves.

The Centers for Disease Control and Prevention (CDC) monitors the trends and current incidence of CJD in the United States by analyzing death certificate information from U.S. multiple cause-of-death data, compiled by the National Center for Health Statistics, CDC. By 4-year periods from 1987 through 1998, the average annual death rates of CJD (not vCJD) have remained relatively constant, ranging from 0.98 cases per 1 million in 1987-1990 to 1.03 cases per 1 million in 1995-1998. In addition, CJD deaths in persons aged <30 years in the United States remain extremely rare (<5 cases per 1 billion per year). In contrast, in the United Kingdom, over half of the patients who died with vCJD were in this young age group.

In addition, CDC collects, reviews and when indicated, actively investigates reports by health care personnel or institutions of possible CJD or vCJD cases. Also, in 1996-97, CDC established, in collaboration with the American

Association of Neuropathologists, the National Prion Disease Pathology Surveillance Center at Case Western Reserve University, which performs special diagnostic tests for prion diseases including post-mortem tests for vCJD.

Prevention measures

According to the Animal and Plant Health Inspection Service of the U.S. Department of Agriculture, BSE has not been detected in the United States, despite active surveillance efforts since May 1990. To prevent BSE from entering the United States, severe restrictions were placed on the importation of live ruminants, such as cattle, sheep and goats, and certain ruminant products from countries where BSE was known to exist. These restrictions were later extended to include importation of ruminants and certain ruminant products from all European countries.

It is extremely unlikely that BSE would be a foodborne hazard in this country. Because the use of ruminant tissue in ruminant feed was probably a necessary factor responsible for the BSE outbreak in the United Kingdom and because of the current evidence for possible transmission of BSE to humans, the U.S. Food and Drug Administration instituted a ruminant feed ban in June 1997 that became fully effective as of October 1997.

In late 2001, the Harvard Center for Risk Assessment concluded in its study of various scenarios involving BSE in the U.S., that the FDA ruminant feed rule provides a major defense against this disease.

BSE/TSE Action Plan of the Department of Health and Human Services (DHHS)

The BSE/TSE Action Plan of DHHS has four major components:

Surveillance for human disease is primarily the responsibility of CDC.

Protection is primarily the responsibility of the Food and Drug Administration (FDA).

Research **is primarily the responsibility of the National Institutes of Health (NIH).**

Oversight is primarily the responsibility of the Office of the Secretary of DHHS.