



vCJD (Variant Creutzfeldt-Jakob Disease)

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Risk for Travelers

Notice to readers: The information below will be published in the forthcoming *Health Information for International Travel: 2007*.

Description

Since 1996, strong evidence has accumulated for a causal relationship between ongoing outbreaks primarily in Europe of a disease in cattle called bovine spongiform encephalopathy (BSE, or "mad cow disease") and a disease in humans called variant Creutzfeldt-Jakob disease (vCJD). Both disorders, which are caused by an unconventional transmissible agent, are invariably fatal brain diseases with incubation periods typically measured in years (1). Transmission of the BSE agent to humans, leading to vCJD, is believed to occur via ingestion of cattle products contaminated with the BSE agent; the specific foods associated with this transmission are unknown. However, a recently published case-control study involving 132 vCJD cases in the United Kingdom showed evidence of an increased risk for vCJD associated with the frequency of consuming beef products likely to contain mechanically recovered meat and head meat (such as burgers, meat pies, and sausages) (2). Bioassays and molecular tests have enabled identification of what World Health Organization consultants have classified as "high-infectivity" and "lower infectivity" tissues of cattle with BSE (3). The high-infectivity tissues include the brain, spinal cord, retina, optic nerve, and dorsal root and trigeminal ganglia, suggesting that these tissues can pose a relatively high risk of transmission. The lower infectivity tissues include peripheral nerves (e.g., sciatic and facial nerves), tonsils, nictitating membrane (third eye lid), distal ileum, bone marrow and possibly thigh muscle. The latter tissue from one cow with BSE transmitted disease to highly BSE-sensitive transgenic mice at a rate indicative of trace levels of infectivity.

Occurrence

From 1995 through mid August 2006, a total of 195 human cases of vCJD were reported worldwide, 162 in the United Kingdom (UK), 20 in France, 4 in Ireland, 2 in the United States, and 1 each in Canada, Italy, Japan, Netherlands, Portugal, Saudi Arabia and Spain. Seven of the non-UK case-patients were most likely exposed to the BSE agent in the UK because of their having resided there during a key exposure period of the UK population to the BSE agent. These latter case-patients were those from Canada, Japan, the United States, 1 of the 20 from France, and 2 of the 4 from Ireland. The median age at death from vCJD in the United Kingdom has been 28 years and almost all cases have been in persons under age 55 years. The reasons for this age distribution are not well understood but it suggests that through the oral route of exposure, older adults are much less susceptible to vCJD than children and young adults. By year of onset, the incidence of vCJD in the UK appears to have peaked in 1999 and to have been declining thereafter. In contrast, the number of reported cases in France has been increasing since the beginning of 2005. However, the future pattern of these ongoing epidemics remains uncertain. In 2004, a prevalence study of asymptomatic vCJD infections in the UK identified three positive appendices out of a sample of 12,674 surgically removed tonsils and appendices that were satisfactory for analysis. Genetic studies completed on two of the appendices regarded as positive for vCJD revealed that both had a different polymorphism at codon 129 of the prion protein gene than any of the patients

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with clinical vCJD tested to date, indicating that more people are genetically susceptible to vCJD infection, although not necessarily to the disease, than had been previously determined (4).

From 1986 through July 2006, >97% of BSE cases worldwide were reported from the UK, where the disease was first described. From 2003 through 2005, however, for the first time, Portugal rather than the United Kingdom reported the highest total country incidence of indigenous cases of BSE per million bovines aged over 24 months, reflecting the relatively more rapid decline of BSE cases in the United Kingdom. As of July 2006, the number of European countries that had ever reported an indigenous BSE case increased to 21. During 2001 through July 2006, four countries outside Europe (Canada, Israel, Japan, and the United States) reported their first indigenous BSE cases, and, except for Israel, other BSE cases in these countries followed.

The reported BSE incidence rates, by country and year, are available on the Internet website of the Office International des Epizooties and new information is being generated on a regular basis. (http://www.oie.int/eng/info/en_esbincidence.htm). As of July 2006, of the countries with at least six reported BSE cases, only four reported that the incidence of BSE had risen in recent years, Canada, Czech Republic, Japan, and Poland.

The identification in 2003 of a BSE case in Canada, and the subsequent identification later that year of a BSE case in the United States that had been imported from Canada led to the concern that indigenous transmission of BSE may be occurring in North America. During 2004 through August 2006, the evidence for such transmission in North America was strengthened by the confirmation of nine additional indigenous North American BSE cases (seven in Canada and two in the United States) (see <http://www.cdc.gov/ncidod/dvrd/bse>). In 2004, both countries had implemented new safeguards to reduce the risk for human exposure to BSE.

(see http://www.hc-sc.gc.ca/ahc-asc/media/nr-cp/2003/bse-esb_e.html and <http://www.cdc.gov/mmwr/preview/mmwrhtml/mm5253a2.htm>). In 2006, Canada also banned cattle tissues capable of transmitting BSE from all animal feeds, pet foods and fertilizers to enhance its BSE-related feed controls; at least four of the seven Canadian BSE cases reported in 2004 through August 2006 had been born after the 1997 US and Canadian feed bans (see <http://www.inspection.gc.ca/english/corpaffr/newcom/2006/20060626e.shtml>).

Transfusion of blood contaminated with the vCJD agent is believed to be responsible for at least three vCJD infections reported in the UK, including two blood recipients with clinical vCJD and one infected recipient who died without signs of neurologic disease. These three recipients indicate that the blood of asymptomatic, infected donors can contain infectivity 18 months to 3.5 years before the onset of vCJD disease. The possibility of transfusion transmission of vCJD had prompted the US Food and Drug Administration to publish guidance in 1999 and 2002 outlining a geography-based donor deferral policy to reduce the risk of such transmission in the United States. This guidance document included an appendix that listed European countries with BSE or a possible increased risk of BSE for use in determining blood donor deferrals (see <http://www.aphis.usda.gov/NCIE/country.html#BSE>).

Risk for Travelers

The current risk of acquiring vCJD from eating beef (muscle meat) and beef products produced from cattle in countries with at least a possibly increased risk of BSE cannot be determined precisely. If public health measures are being well implemented the current risk of acquiring vCJD from eating beef and beef products from these countries appears to be extremely small, although probably not zero. A rough estimate of this risk for the UK in the recent past, for example, was about 1 case per 10 billion servings. Among many uncertainties affecting such risk determinations are 1) the incubation period between exposure to the infective agent and onset of illness, 2) the appropriate interpretation and public health significance of the prevalence estimates of asymptomatic human vCJD infections, 3) the sensitivities of each country's surveillance for BSE and vCJD, 4) the compliance with and effectiveness of public health measures instituted in each country to prevent BSE contamination of human food, and 5) details about cattle products from one country distributed and consumed elsewhere. As of August 2006, despite the apparent exceedingly low risk of contracting vCJD through consumption of food in Europe, the US blood

donor deferral criteria focuses on the time (cumulatively 5 years or more) that a person lived in continental Europe from 1980 through the present. In addition, these deferral criteria apply to persons who lived in the United Kingdom from 1980 through 1996.

Prevention

Public health control measures, such as surveillance, culling sick animals, or banning specified risk materials, have been instituted in many countries, particularly in those with indigenous cases of confirmed BSE, in order to prevent potentially BSE-infected tissues from entering the human food supply. The most stringent of these control measures, including a program that excluded all animals >30 months of age from the human food and animal feed supplies [the Over Thirty Month (OTM) rule], was applied in the UK and appeared to be highly effective. With the decrease in British BSE cases, the OTM rule was replaced in 2005 with a BSE testing regime, and in 2006, the ban on exports of British beef to other members of the European Union was lifted. In June 2000, the European Union Commission on Food Safety and Animal Welfare had strengthened the European Union's BSE control measures by requiring all member states to remove specified risk materials from animal feed and human food chains as of October 1, 2000; such bans had already been instituted in most member states.

To reduce any risk of acquiring vCJD from food, concerned travelers to Europe or other areas with indigenous cases of BSE may consider either avoiding beef and beef products altogether or selecting beef or beef products, such as solid pieces of muscle meat (rather than brains or beef products like burgers and sausages), that might have a reduced opportunity for contamination with tissues that may harbor the BSE agent. These measures, however, should be taken with the knowledge of the very low risk of disease transmission, particularly to older persons, as discussed above. Milk and milk products from cows are not believed to pose any risk for transmitting the BSE agent.

Treatment

As of August 2006, treatment of prion diseases remains supportive; no specific therapy has been shown to stop the progression of these diseases.

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