

detected in the U.S. despite active surveillance for the disease since May 1990.

In 1996, a newly recognized form of the human disease CJD, called variant CJD (vCJD), was reported in 10 patients in the U.K. vCJD is a chronic, neurodegenerative disease that affects humans. Scientific and epidemiological studies have linked vCJD to exposure to BSE, probably through human consumption of beef products contaminated with the agent that causes BSE.

Until recently, vCJD had not been detected in the U.S. In April 2002, the Florida Department of Health and the Centers for Disease Control and Prevention (CDC) began investigating a likely case of vCJD in a citizen of the U.K. living in Florida. In October 2002, CDC reported the investigation of this case and stated that it represents the first probable vCJD case in a U.S. resident (CDC, *Morbidity and Mortality Weekly Report*, 51(41): 927–929, 2002). CDC believes, however, that the patient was exposed to the BSE agent while living in the U.K. This is likely to be the case, as the disease is thought to have a long incubation period and the appearance of symptoms does not mean that exposure was recent.

Surveillance data from European countries in which BSE has been detected indicate that cattle with clinical signs of a central nervous system (CNS) disorder, “dead” cattle (*i.e.*, died otherwise than by slaughter), and cattle that cannot rise from a recumbent position (*i.e.*, nonambulatory, cattle commonly referred to as “downer” cattle in the U.S.), have a greater incidence of having BSE than other cattle. The FSIS regulations prohibit for use as human food cattle with clinical signs of a CNS disorder or certain infectious or parasitic diseases, or that are in a dying condition or that died otherwise than by slaughter (§§ 309.3, 309.4). All seriously crippled cattle and cattle commonly termed “downers” presented for slaughter are automatically suspected of being affected with a disease or condition that may require condemnation of the animal, in whole or in part, and are identified as “U.S. Suspects” (§ 309.2(b)). Such cattle are examined at ante-mortem inspection by an FSIS veterinarian, and a record of the veterinarian’s clinical findings accompanies the carcass to post-mortem inspection if the animal is not condemned on ante-mortem inspection. Post-mortem inspections on the carcasses of U.S. Suspects cattle are performed by a veterinarian rather than a food inspector, and the results of this

inspection are recorded. U.S. Suspects, unless otherwise released pursuant to § 309.2(p), must be set apart and slaughtered separately (§ 309.2(n)). If, on post-mortem inspection, the meat and meat food products from such cattle are found to be otherwise not adulterated, such products may be used for human food (§ 311.1).

Surveillance for BSE in Europe has shown that the typical clinical signs associated with BSE cannot always be observed in nonambulatory (downer) cattle infected with BSE because the signs of BSE often cannot be differentiated from the typical clinical signs of the many other diseases and conditions affecting downer cattle. Thus, if BSE were present in the U.S., it is possible that downer cattle infected with BSE could be presented for slaughter, and, if the clinical signs of the disease were not obvious, pass ante-mortem inspection. These cattle could then be slaughtered, and, if they pass post-mortem inspection, the meat and meat food products from such cattle could be used for human food. However, the BSE agent has not been detected in muscle tissue of infected cattle. Tissues that have been found to contain high levels of the agent that causes BSE in BSE-infected cattle—such as the brain tissue, the spinal cord, and the retina of the eye—could possibly cross-contaminate muscle tissues with the BSE agent during slaughter and processing.

The U.S. government has implemented a number of measures to prevent BSE from entering the U.S. and to prevent the spread of the disease should it be introduced in the U.S. For example, since 1989, the USDA’s Animal and Plant Health Inspection Service (APHIS) has prohibited the importation of live cattle and certain cattle products, including rendered protein products, from countries where BSE is known to exist. In 1997, because of concerns about widespread risk factors and inadequate surveillance for BSE in many European countries, these importation restrictions were extended to include all of the countries in Europe. On December 7, 2000, APHIS prohibited all imports of rendered animal protein products, regardless of species, from BSE-restricted countries because of concern that feed intended for cattle may have been cross-contaminated with the BSE agent.

In addition, APHIS leads an ongoing, comprehensive, interagency surveillance system for BSE in the U.S. and, in cooperation with FSIS, has drafted an emergency response plan to be used in the event that BSE is identified in the U.S. Other Federal

agencies also have contingency plans that work in concert with the USDA plan.

In 1997, the Food and Drug Administration (FDA) prohibited the use of most mammalian protein in the manufacture of animal feeds given to cattle and other ruminants (21 CFR 589.2000). Firms must keep specified records on the manufacture of their feed, must have processes in place to prohibit co-mingling of ruminant feed with non-ruminant feed, which may contain materials prohibited in ruminant feed, and must ensure that non-ruminant feed containing materials prohibited in ruminant feed is labeled conspicuously with the statement, “*Do not feed to cattle and other ruminants.*” These regulations are intended to prevent the introduction and spread of BSE in U.S. cattle through feed contaminated with the BSE agent.

In addition, the CDC monitors the incidence of CJD in the U.S. by analyzing death certificate information from multiple-cause-of-death data compiled by the National Center for Health Statistics. This information is also used to search for possible cases of vCJD in the U.S.

In 1998, USDA entered into a cooperative agreement with Harvard University’s School of Public Health to conduct an analysis and evaluation of the current measures implemented by the U.S. government to prevent the entry and spread of BSE in U.S. cattle herds and to reduce the potential for exposure of Americans to the BSE agent. The Harvard study identifies three pathways or practices that could contribute the most to the spread of BSE and the amount of potentially dangerous tissue in the human food supply: (1) Noncompliance with the FDA feed ban, including misfeeding on the farm and the mislabeling of feed and feed products prohibited for consumption by cattle; (2) unsafe disposition of cattle that die on the farm; and (3) inclusion of high-risk tissue, such as brain and spinal cord, in edible products. With regard to the second pathway listed, a potential use for cattle that die on the farm otherwise than by slaughter would be for rendering as non-ruminant animal feed since rendered product from animals that die otherwise than by slaughter is prohibited for use as human food but may be used to produce animal feed.

On January 17, 2002, FSIS announced the availability of a paper on its current thinking on possible actions to minimize human exposure to meat products from cattle that could contain the infective agent that causes BSE (67 FR 2399). This paper is available on the