Mad Cow Disease

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Bovine spongiform encephalopathy (BSE, also known as mad cow disease) has surfaced in Great Britain, France, Switzerland, Canada, Spain, Germany, Japan, Russia, and, as of December 22, 2003, in the United States as well. Health authorities consider it to be the most likely cause of a new variant of Creutzfeldt-Jakob disease (vCJD), a fatal brain disease that, as of May 2003, has affected 139 people worldwide.[1]

Americans are now being alerted to the fact that BSE presents a health threat in the United States. Herein, we present scientific evidence that supports growing concern about BSE and vCJD and the need for corrective action to protect the health of humans and animals in the United States:

- The conditions that led to the emergence of BSE in Britain have also been present in the United States. Although some changes have been made, many U.S. livestock rendering and feeding practices are similar to those present in Britain at the onset of the BSE epidemic.
- The agent that causes BSE has already spread to at least one cow and some other species of animals in the United States. The extent to which BSE and other encephalopathies have entered the human food supply is unknown.
- Between 1979 and 1998, 4,751 Americans died of Creutzfeldt-Jakob disease, and the possibility that BSE played a role in some of those deaths cannot be ruled out.[2]

Brain Disease in Cows

Bovine spongiform encephalopathy is a fatal central nervous system disease first identified in the UK in 1986. Affected cows show increased apprehension, poor coordination, difficulties in walking, and weight loss. The infections that cause BSE apparently existed for several years before the disease was recognized in England.[3-5]

BSE is not limited to Britain. It has been found in native cattle in France, Switzerland, Northern Ireland, the Republic of Ireland, the Channel Isles, and the Isle of Man. Cattle exports carried the disease to Canada, Denmark, Oman, and the Falkland Islands.[3,6] And the first case in the United States was reported in December 2003. Moreover, at least 100,000 cattle whose BSE status is unknown have been shipped from the UK to other countries. Most of these animals are unaccounted for.

Related Brain Disease in Humans

VCJD, like other transmissible encephalopathies, robs an affected individual of mental faculties and muscle coordination, eventually leading to coma and death. This category of illnesses is caused by prions, proteins that are normal in their molecular makeup but abnormal in their shape, like springs that have been bent out of configuration. It is believed that vCJD results from contact with prions in tissues of cattle with bovine spongiform encephalopathy (BSE), or "mad cow disease," which, in turn, distort normal proteins in human brain and nerve cells. Only minuscule amounts of prion-tainted tissues are required in order to transmit the disease.[7] Prions concentrate in the brain and spinal cord, but also have been found in blood and muscle tissue. Prions are very difficult to destroy, even by the chemical or heat disinfectant methods used in hospitals. Heating to 134 degrees Celsius (273 degrees Fahrenheit) does not reduce their infectivity.[8]

Similar Diseases in Other Species

Transmissible encephalopathies have long been documented in animals in the United States, indicating the need to protect human populations. Scrapie, the encephalopathy analogous to BSE occurring in sheep and which may have been the origin of BSE, was first reported in the United States in 1947, and infected sheep flocks have been identified in virtually all parts of the United States where sheep are raised.[9,10] Similar diseases have been found in other species, including chronic wasting disease in deer and elk, and transmissible mink encephalopathy.[8] While scrapie is not known to cause human disease, passage of the prions that cause scrapie through other animals may potentially alter their infectivity and disease presentation.

Indirect evidence from mink on fur farms who were routinely fed cattle-derived remains suggests that BSE may have been present as early as 1985 in at least some U.S. cattle. In at least five separate outbreaks, mink fed in this way have developed a disease called transmissible mink encephalopathy (TME) which is remarkably like BSE.[4,11] After a 1985 TME outbreak in Stetsonville, Wisconsin, experimenters injected brain tissue from diseased mink into Holstein cattle, finding that the cattle developed spongiform encephalopathy. They then fed remains of these cattle to healthy mink, who soon developed TME.[12] Reviewing evidence that the

mink disease came from cattle-based feeds, University of Wisconsin researchers concluded, "If this is true, there must exist an unrecognized bovine spongiform encephalopathy (BSE)-like infection in American cattle."[12] In other words, U.S. cattle have been sporadically infected with BSE, which has shown its pathological effects when passed on to other species.

The Failure of U.S. Government Policies

The fact is that USDA officials have not been seriously looking for BSE cases. Data from the National Veterinary Sciences Laboratories BSE Surveillance program from 1990 to 2000 show that, of approximately 900 million cattle slaughtered, only 11,954 brains (approximately 1 in 75,000) were examined for BSE. In fiscal year 2002, the USDA tested only slightly more-19,990-cattle for BSE.[13] Further, brain examinations have generally been prompted by the presence of neurological symptoms. However, the symptoms of BSE do not commonly manifest in cattle until about five years of age, which is after the usual age of slaughter. For example, most U.S. dairy cows are slaughtered before four years of age, when even a prion-infected cow is likely to appear healthy. In the UK, 70 percent of dairy cows remain alive past this point, making identification of infected animals much easier.[3] Despite all this, the first official U.S. case was recently identified in a cow raised in Washington State. The USDA has been slow to take steps against the potential spread of encephalopathies from one animal to another, or from animals to humans. Since 1989, the U.S. government has banned the import of live ruminants (cattle, sheep, goats) and most ruminant products from countries where BSE has been reported, and, in December 2000, the USDA prohibited imports of animal-derived livestock feeds from Europe, regardless of species of origin, after determining that feed derived from non-ruminant species was potentially cross-contaminated with BSE. However, given that BSE is already present in the United States, appropriate steps are needed to protect against its spread. The measures instituted so far by the U.S. government are grossly inadequate, as evidenced by the following examples:

- U.S. feed producers are blatantly violating restrictions on feed production. Despite a 1997 Food and Drug Administration (FDA) ban on the feeding of most mammalian remains to ruminants-which unfortunately includes significant exceptions impairing the protective intent of the law-a January 2001 FDA report showed that, of 180 renderers, 16 percent lacked warning labels on feeds designed to differentiate those intended for ruminants from those for nonruminants and 28 percent had no system to prevent the actual mixing of these feeds.
- There is no restriction on the use of animal by-products, including blood and blood products, gelatin, milk, and milk products, in feeds through which prions may be transmitted.
- There are no limits on the use of non-ruminant, such as pig or horse, remains in feeds, due to an exemption in the 1997 ban. Because prions are so difficult to destroy, if the remains of BSE infected cow are fed to a pig or horse and then the pig or horse remains are fed to cows, the cows may subsequently be infected. Similarly, ruminant remains can be fed to poultry and, in turn, poultry feces are routinely used in cattle feed.
- There are no limits on the "recycling" of beef or other meat products in the form of garbage from restaurants or other institutions for use in animal feeds.

Monitoring for human illness is even more haphazard. Transmissible encephalopathies are not yet reportable diseases for the Centers for Disease Control and Prevention. Individuals showing signs of dementia due to such a condition may be misdiagnosed as suffering from Alzheimer's disease or stroke, and most dying with neurological illnesses are never autopsied, so their brains are never examined.

There is simply no way of knowing whether vCJD has begun in the United States or not. Death certificates from 1979 to 1998 show that 4,751 people were identified with CJD in the United States. While the presumption is that they had the "classical" form of the disease, rather than the new variant form which is believed to come from animal tissues, this remains uncertain. While most victims were older (a sign of classical CJD), a small number were surprisingly young.[2] The reported cases are probably underestimates due to the problems of misdiagnosis and underreporting.

On a more optimistic note, restrictions on blood products are becoming more stringent, which is appropriate given that they may also be vectors of disease.[14] Currently, people who have spent a cumulative total of three months or more in the United Kingdom since 1980 or a total of six months or more in any European country or combination of countries are not permitted to donate blood in the United States. Nonetheless, it is clear that diseases closely related to vCJD exist in the United States, both in animals and humans, that these diseases should be considered highly infectious, that monitoring programs are too spotty to track the extent of these diseases, and that current preventive steps are far too lax.

Recommendations

The following are PCRM's recommendations to the government for protecting the public against vCJD:

- Ban the use of animal-derived livestock feeds for any species, given the likelihood that animal byproducts will, in turn, be recycled to ruminants (that is, cows, sheep, and goats).
- Ban the slaughter of downed animals, animals too sick to stand, for human food. The Washington state cow that tested positive for mad cow disease in December 2003 was a downed animal.
- Prohibit animal byproducts in all medications, supplements, or cosmetics.
- Label all foods containing animal byproducts (such

as gelatin or "natural flavorings"), indicating both the presence of animal byproducts and the species of origin.

- Provide warning labels on all foods that carry a risk of vCJD, using standards similar to those for tobacco and alcohol products.
- Institute comprehensive monitoring programs to check for diseased animals and humans in the United States. Monitoring programs for BSE and other encephalopathies in animals should include but not be limited to testing all suspect animals (rather than a fraction of them) and holding back the carcasses of tested animals from the food supply until the test results are known. For humans, monitoring programs should be implemented that require all states to report CJD cases and dementia of unknown cause (especially in young individuals) to the Centers for Disease Control so that any cases where vCJD is suspected can be confirmed or dismissed by autopsy.

It should be recognized that the consumption of livestock products is clearly linked to a much higher risk of serious and sometimes fatal diseases, apart from the risk of transmissible encephalopathies. These diseases include coronary artery disease, colon and possibly other forms of cancer, diabetes, hypertension, obesity, and infection with salmonella, campylobacter, and E.coli O157:H7, among others. Making meat "safe" is not a realistic or attainable goal. Ironically, while the feeding of animal remains to other animals is now acknowledged as a dangerous practice that is restricted in some countries, the feeding of animal remains to humans is encouraged by government programs and massive industry efforts.

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