Mad Cow:
What's the Danger?

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A prion is a protein particle associated with and believed to be the cause of various encephalopathies. The predominant human prion diseases are listed in Table 1, whereas Table 2 lists the predominant animal prion diseases. One of the most talked about animal prion diseases is bovine spongiform encephalopathy (BSE), also known as mad cow disease.

What are the common features?

BSE is a transmissible spongiform encephalopathy (TSE). In most TSEs, the incubation period is measured in years rather than months. Vacuolation in nerve cells always occurs. Once the disease appears, it invariably leads to the death of the patient. Confirmatory diagnosis is made through histopathology, immunohistochemistry (IHC) and Western blot.

The odds of acquiring CJD is 1 in 1 million

How are TSEs transmitted?

The natural transmission of TSEs is believed to be almost exclusively by the oral route. An oral dose of 1 g of infected brain tissue fed to a young calf is sufficient to result in clinical BSE in the animal four years later. Higher doses shorten the incubation period. Iatrogenic transmission within, and across, species barriers is possible via intracerebral inoculation of infected tissue.

Mad cow disease in Canada

BSE is a relatively new disease, first confirmed in cattle in 1986. Many experts believe the practice of recycling infected carcasses into meat and bone meal (MBM), and feeding it back to cattle, amplified the spread of the condition.

In Canada, BSE became a "reportable disease" in 1990, with active surveillance beginning a year later. In 1993, Canada had its first case of BSE. The infected cow, discovered near Red Deer, Alberta, was imported from the U.K. in 1987. At that time, all related and contact animals were disposed of by incineration. In 1997, Canada imposed a ruminant-to-ruminant feed ban.

The second case of BSE in Canada occurred May 2003 in an eight-year-old cow on a commercial cattle operation in northern Alberta. After the animal had been slaughtered, BSE was suspected upon examination by provincial animal health authorities. A brain sample was determined to have lesions consistent with BSE.

The Canadian Food Inspection Agency (CFIA) immediately quarantined farms with

Table 1
Human prion diseases

1. **Gerstmann-Straussler-Scheinker syndrome (GSS)**
   - GSS is a group of illnesses with varying genetic abnormalities
2. **Fatal familial insomnia (FFI)**
   - Linked to a specific genetic mutation
3. **Kuru**
   - A prion-associated disease related to the consumption of dead relatives (ritualistic funeral rites)
4. **Creutzfeldt-Jakob disease (CJD)**
   - Affects humans worldwide at a rate of 1:1 million annually
   - Suspected to be an individual mutation
5. **vCJD ("new variant" CJD)**
   - First appeared in 1996 in the U.K.
   - Occurs in abnormally young age group
   - Thought to be linked to products derived from BSE-infected cattle

any possible link to the index farm. Approximately 2,700 animals were destroyed soon after. The brain tissue of approximately 2,300 animals, those over 24 months of age, was sent for testing.

Histopathology and IHC tests, which use monoclonal antibodies to prion protein, were conducted. Fresh brain tissue was also subjected to testing by Western blot. All samples were found to be negative.

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What's the current situation?

Canada has amended both the Food and Drug and the Health of Animals Regulations to enforce the removal of, and prohibit the sale of, specific risk materials (SRM) from cattle carcasses destined for the human food chain. SRM includes:

- skull,
- brain,
- trigeminal ganglion,
- eyes,
- tonsil,
- spinal cord, and
- dorsal root ganglia

of cattle aged greater than 30 months. The distal ileum of the intestine of cattle of all ages is also included.

The CFIA has also increased its surveillance of the existing cattle population to confirm that BSE is non-existent in the Canadian cattle herd. The surveillance focuses on the classes of livestock most likely to be affected, including:

- those with neurologic signs,
- non-ambulatory cases,
- animals found dead, and
- animals which die during transport.

What does the future hold?

At this time, there are no live animal tests available, no treatments, and no vaccines for these diseases. The search for accurate, rapid, and easily-applied tests continues. The goal is to find a test that can be administered to the live animal. A verification of enzyme-linked immunosorbent assay (ELISA) tests is currently underway. ELISA technology would allow the testing of large numbers of samples, but still requires that brain tissue be used. Genetic resistance to disease has not been confirmed and requires further investigation.

Frequently Asked Questions

1. Can BSE, scrapie, or CWD be transmitted to humans and other animals?

BSE is believed to be linked to cases of "new variant" CJD in humans, and has also been suspected to be related to cases of TSE in cats. Scrapie and CWD have not been known to cross species barriers.

2. What causes mad cow disease?

It is thought to be an abnormal protein particle referred to as a "prion". Prions cannot be killed by normal disinfection procedures, but are susceptible to very high temperature sterilization, and to bleach and lye solutions.

3. Is there a treatment for mad cow disease?

There are no treatments, no live animal tests, and no vaccines for this type of disease.

4. What is Canada doing to ensure that cattle in this country are free of BSE?

The CFIA is continuing its surveillance by collecting brain tissue from older cattle that are being sent to slaughter plants. The brain tissue is sent to a laboratory facility that screens for BSE.

Table 2
Animal prion diseases

1. **Scrapie**
   - Recognized as an entity in sheep for over 250 years
   - Has not been shown to cross species barriers
2. **Chronic wasting disease (CWD)**
   - Has been recognized since 1980
   - Occurs in elk, mule deer, and white-tail deer in the U.S., and in Canada since 1998
3. **Transmissible mink encephalopathy (TME)**
   - Occurs in mink in Finland, Germany, Russia, Canada, and the U.S.
   - The most recent case was recorded in 1985 in Wisconsin
4. **Feline spongiform encephalopathy (FSE)**
   - Occurs in cats, with most cases being reported in the U.K. during the 1990s
   - Assumed to be linked to consumption of food products derived from BSE-infected meat
5. **Exotic ungulate encephalopathy (EUE)**
   - Is reported to occur in ungulates in zoologic parks
   - Has been associated with the larger BSE occurrence
6. **Bovine spongiform encephalopathy (BSE)**
   - Occurs in cattle

Puting risk into perspective

The chance of dying while playing soccer or driving a car has been estimated in the neighbourhood of 1:10,000. The liklihood of being hit by lightning has been placed at less than one in a million. To put the risk into perspective, the odds of acquiring CJD is one in a million.

When considering the steps being taken by the CFIA and Health Canada to remove risk materials from the food chain, Canadians can rest assured that the risk of acquiring vCJD from a contaminated food product will continue to be less than one in 10 million. CME

Suggested readings
1. Brown C, Bolin C: Emerging diseases of animals. ASM Press, Washington,D.C. 2000; 131-59.
2. Rabineau HF, Cinatl J, Doerr HW: Prions—A challenge for science, medicine and public health system. Contributions to Microbiology, 2001.

t Readings

1. Kuru
   www.nature.com/nsu/030407/|030407-13.html
2. Kuru and Genetics
   www.nature.com/nsu/010208/010208-5.html
3. Prions and CJD
   www.nature.com/nsu/030331/030331-12.html
4. Canadian Food Inspection Agency—BSE update and links
   www.inspection.gc.ca/english/anima/heasan/disemala/spong/
5. Health Canada - BSE and vCJD
   www.hc-sc.gc.ca/

Take-ho message

- Mad cow disease is thought to be caused by an abnormal protein particle called a prion.
- The odds of acquiring CJD is estimated to be one in a million.
- Currently, there are no treatments, vaccines, or live animal tests for this disease.

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