BSE
BOVINE SPONGIFORM ENCEPHALOPATHY

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**Definition:** Bovine Spongiform Encephalopathy (BSE) is a fatal, transmissible neurodegenerative disease of dairy and beef cattle. It is characterized by a long incubation period (years) followed by a clinical period lasting days to months in duration.

**Etiology:** The agent responsible for BSE and the other spongiform encephalopathies has not been characterized yet. The agent has been historically called a prion (infectious protein particle), a virino (subviral particle), or an unconventional virus. The main reason the agent remains un-characterized is that researchers have been unable to replicate it outside a host animal fulfilling Koch's postulate. It is known that the agent produces insoluble strands of protein in the brains of affected cattle.

Although the origins of BSE and the molecular characteristics are not established, epidemiologic analyses suggest that the current epidemic in the United Kingdom was due to the introduction and recycling of the agent in cattle feedstuffs. Specifically in the rendered meat and bone meal protein supplements. Factors that aided in the introduction and spread of the agent include reduction of rendering temperatures, reduced use of hydrocarbon solvents during rendering, and an increased use of animal derived protein supplements in cattle rations.

**History:** The first documented case of BSE occurred in, Weybridge, United Kingdom, November 1986. Retrospective studies suggest that the first cases appeared in 1985 but were misdiagnosed. The incidence of BSE increased from 1-2 cases per month in 1988, to over 2,500 cases per month in January 1992.

Northern Ireland, Republic of Ireland, Switzerland and France have had native cattle affected with BSE. Canada, Portugal, Oman, Denmark, And the Falkland Islands have had imported cases.

**Incubation:** Cattle have developed BSE in a range of 22 months to 15 years of age. The age of most affected cattle is 3.5 to 5 years old.

**Transmission:** By extension from the study of other related transmissible encephalopathies it is presumed that natural infection occurs primarily in neonatal animals. Contaminated feedstuffs such as milk replacers / creep feeds and bypass protein supplements are implicated. (feedstuffs
containing meat and bone meal) Calves less than one year of age have the highest risk factors. Maternal transmission has been documented, but it occurs at a low rate. (At a level too low to maintain the epidemic) BSE does not appear to spread by casual cattle to cattle contact, via aerosols or through an intermediate vector. Studies to find the infectivity of tissues and excretions (saliva, urine, feces, semen, placenta) have not been concluded. Risk assessment and dose needed to affect adult cattle is not known due to the long incubation and limited lifetime of cattle in most commercial production systems.

**Signs:** Initial signs
- Apprehension and reluctance to move through doorways
- Nervousness or appearance of fear
- Tactile, ocular (light) or auditory hyperesthesia
- Kicking during handling
- Aggressive behavior toward other animals and handlers
- Locomotor difficulties or mild incoordination
- Loss of body condition and reduced milk production
- Cattle continue to eat
- Difficult to differentiate clinically from hypomagnesemia or nervous ketosis
- Lack of response to treatment & slow progression help differentiate these diseases from BSE.

**Progression**
- continued weight loss
- moaning
- Excessive salivation
- Limited puritis with licking of nose and other areas of body
- Grinding of teeth
- Severe incoordination leading to hypermetria, falling, and frenzy
- Terminal recumbency

**Rarely seen signs**
- Circling, blindness (2%) 
- & Knuckling at the fetlock (10%)
- Head pressing/rubbing

**Rule-Outs**
- Listeriosis
- Rabies
- CNS tumors, abscesses, trauma, parasitic infection
- Mineral or plant toxicity
- Injuries and lameness
- Hypomagnesemia
- Nervous Ketosis

**Diagnosis:** No preclinical or clinical test is available. Laboratory tests are 1) Histopathology, 2) Immunobiochemical staining - Western Blot Test, 3) Electron microscope looking for Scrapie Associated Fibrils - SAF.
Treatment: None

What to do?? Should any cow show signs of BSE CALL your local reportable disease authority. This could be the State Veterinarian's office, APHIS - Veterinary Services office, or your local district veterinarian. The best time to report suspected Foreign Animal Diseases (FAD) is when you are out on the farm just after examining the animal. Be ready to give a short history and the current signs the animal is exhibiting.

LOCAL RESPONSE - FAD diagnostician and on farm investigation

Can a group of samples be sent to a local diagnostic lab for domestic disease rule - outs? Yes. We would still prefer you report the suspected FAD at the time you suspect it. The local response has quite a few options. One of those options may be to wait for local lab confirmation of domestic diseases before an FAD diagnostician is sent to talk with the farmer. It is not always a knee jerk response.

IF A CONFIRMED POSITIVE BSE IS FOUND....

farm, state, national actions/reactions

Note that we are already being accused of not looking hard enough for BSE by researchers in this country and by many industry people from other countries.

Control Measures: United Kingdom -

1. Make the disease reportable
2. Ban on feeding ruminant derived protein supplements to other ruminants.
Compulsory slaughter and incineration of suspect cattle.
3. Ban on the human consumption of specific bovine offal, including brain, spinal cord, spleen, thymus, tonsils and intestines.
4. Ban on bovine offal or their products being fed to pets or farm animals.
5. Prohibiting consumption of milk from BSE affected or suspect cows by either animals or humans (except dam to its calf)

Miscellaneous: Host range: Cattle natural transmission

Mice experimentally
Sheep Goats Pig

Wild Animals Zoo (five ungulate and three cheetahs)
Bovine Spongiform Encephalopathy

Bovine spongiform encephalopathy (BSE) is a chronic degenerative disease affecting the central nervous system of cattle. The disease was first diagnosed in 1986 in Great Britain. The British sometimes call BSE "mad cow disease," a term that could be confused with rabies in cattle if used in the United States.

BSE has had a substantial impact on the British livestock industry. The disease also has been confirmed in domestic cattle in Ireland, France, Portugal, Switzerland, and in cattle exported from England to Oman, the Falkland Islands, Germany, Denmark, Canada, and Italy. The U.S. Department of Agriculture's (USDA) Animal and Plant Health Inspection Service (APHIS) is enforcing import restrictions and is conducting surveillance for BSE to ensure that this serious disease does not become established in the United States.

Clinical Signs

Cattle affected by BSE experience a progressive degeneration of the nervous system. Affected animals may display changes in temperament, such as nervousness or aggression; abnormal posture; incoordination and difficulty in rising; decreased milk production; or loss of body weight despite continued appetite. Affected cattle die. The causative agent of the disease is not completely characterized, and there is no treatment.

The incubation period (the time from when an animal becomes infected until it first shows disease signs) is thought to be from 2 to 8 years. Following the onset of clinical signs, the animal's condition deteriorates until it either dies or is destroyed. This usually takes from 2 weeks to 6 months. Most cases in England have occurred in dairy cows between 3 and 5 years of age.

Currently, there is no test to detect the disease in a live animal; veterinary pathologists confirm BSE by postmortem microscopic examination of brain tissue. BSE is so named because of the spongy appearance of the brain tissue of infected cattle when sections are examined under a microscope.

Related Diseases

BSE belongs to a group of related diseases known as the transmissible spongiform encephalopathies, which are all caused by uncharacterized agents that produce spongiform changes in the brain. The group includes scrapie, which affects sheep and goats; transmissible mink encephalopathy; feline spongiform encephalopathy; chronic wasting disease of mule deer and elk; and kuru, Creutzfeldt-Jakob disease, and Gerstmann-Straussler syndrome, three rare diseases in humans. Other cases of spongiform encephalopathies have been reported in Great Britain in kudus, an eland, a nyala, and a gemsbok.

No scientific evidence indicates that BSE can be transmitted from infected cattle to humans through contact or consumption of beef or dairy products. The World Health Organization does not consider BSE to be a human health hazard based on current scientific evidence.

Epidemiology

There are different scientific hypotheses concerning the origins of BSE. One theory is that BSE had existed in undetectable levels in the British cattle population prior to 1986. Another theory stems from epidemiologic data that suggest that BSE in England may have been caused by feeding cattle rendered protein produced from the carcasses of scrapie-infected sheep. The practice of using products such as meat and bone meal in cattle rations as a source of protein has been common for several decades. Scrapie has a long incubation period—up to 60 months—and has been endemic in Great Britain for centuries. Changes in rendering operations in the early 1980's—particularly the removal of a solvent-extraction process and the elimination of a second steam-heat treatment—may have played a part in the appearance of the disease and the large number of cases that developed.

There is no evidence that BSE spreads from unrelated cattle to cattle or from cattle to other species by contact. Moreover, researchers have not gathered sufficient epidemiologic evidence or experimental data to determine if maternal transmission of BSE occurs.
The extremely small infectious agent responsible for BSE and scrapie, although not completely characterized, has been theoretically classified as a "slow virus," a "prion," or a "virino." This agent is extremely resistant to heat and to normal sterilization processes. It also does not evoke a detectable immune response or inflammatory reaction in host animals.

The BSE agent has been found only in brain tissue and the spinal cords of cattle naturally affected with BSE. In an experimental study of the disease's biological route of development in cattle, traces of the BSE agent were detected in the small intestines of calves that had been fed large doses of material from BSE-infected animals. Evaluation of the presence of the BSE agent in tissues is complicated by the lack of a definitive laboratory test. Failure to identify the agent in tissues may indicate either a true absence of the agent or simply a decreased sensitivity of current diagnostic methods.

History
During the period from November 1986 (when BSE was first identified as a separate disease entity) until September 1994, an estimated 134,000 head of cattle in more than 30,000 herds were diagnosed with BSE in Great Britain. The epidemic peaked at almost 1,000 cases per week. Agricultural officials in Great Britain have taken a number of actions to eradicate BSE, including (1) making BSE a notifiable disease, (2) prohibiting the inclusion of ruminant-derived proteins in ruminant feed, (3) destroying all animals showing signs of BSE, (4) prohibiting the consumption of milk from affected or suspect cows by either animals or humans (except for milk from a dam to its calf), and (5) stopping human and animal consumption of certain bovine organs, including brain, spinal cord, spleen, thymus, tonsils, and intestines.

As a result of these actions, the rate of newly reported cases of BSE is decreasing. Currently, less than 500 cases are occurring per week. (There are approximately 10 million cattle and 38 million sheep in Great Britain.)

USDA Actions in Response to BSE
To prevent BSE from entering the United States, APHIS has restricted the importation of live ruminants and ruminant products from countries where BSE is known to exist. Other products derived from ruminants, such as fetal bovine serum, bone meal, meat and bone meal, blood meal, offal, fats, and glands, also cannot be imported into the United States from these countries, except under a special permit for scientific research purposes.

In addition to international importation restrictions, APHIS has increased surveillance efforts to detect BSE if it is accidentally introduced into the United States. More than 250 APHIS and State veterinarians specially trained to diagnose foreign animal diseases regularly conduct field investigations of suspicious disease conditions.

APHIS veterinary pathologists and field investigators also have received training from their British counterparts for diagnosing BSE. These pathologists examine brain tissue from cattle over 2 years of age that show signs of neurological disease.

More than 60 veterinary diagnostic laboratories throughout the United States are participating in the BSE Surveillance Program along with the National Veterinary Services Laboratories in Ames, IA. As of December 1994, nearly 2,000 specimens from 42 States had been received, and no evidence of BSE had been seen.

As part of increased surveillance for BSE, APHIS veterinarians are tracing 499 head of cattle imported from Great Britain between 1981 and 1989 (before the ban on imports went into effect) to check their health status. As of December 1994, 452 of the animals had been accounted for, and no signs of BSE had been found. Efforts continue to trace the remaining cattle.

APHIS leads the interagency effort to coordinate surveillance for BSE. Officials of USDA's Food Safety and Inspection Service notify APHIS of cattle having neurological signs at slaughter. State diagnostic laboratories and public health officials also submit the brains of rabies-negative cattle to NVSL for testing.

Getting the Word Out
As part of the increased surveillance activities, APHIS is continuing an education effort to inform U.S. cattle producers and veterinarians about this new disease. Numerous briefings have been held for industry groups. In addition to press releases and factsheets, a British videotape on BSE and an information packet were distributed to all APHIS field offices, State veterinarians, extension veterinarians, colleges of veterinary medicine, and industry groups.

For additional information, contact
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Telephone: (301) 734-8073.
For information about importing animals or animal products, contact
USDA, APHIS, VS
National Center for Import/Export Animals Program
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