Monograph

Mon, 01/24/2011 - 16:56 — Anonyme **definition:**
Bovine spongiform encephalopathy (BSE) commonly known as "mad cow disease" is a disease which affects the central nervous system and is characterized by the development of cavities within brain nerve cells, giving the brain a sponge-like appearance.

BSE is a transmissible, inoculable disease but it is not contagious. It affects adult bovines following a long incubation period. The disease leads inevitably to death.

Situation in America:
The disease was declared to the OIE by Canada in 2004 and the USA in 2003.

Susceptible species:
• Domestic and wild bovidae: bovines, bisons, etc.

• Domestic and wild felines: cats, tigers, pumas, ocelots, etc.

• Man: the BSE causes a rare form of the Creutzfeld Jakob (CJD) disease in man, the new variant CJD.

• Possibility of experimental transmission to ovines, caprines, pigs, mice, etc.

Etiological agent:
The "prion" hypothesis:

BSE is caused by an infectious protein particle, or pathogenic prion, which is currently the only infectivity-related macromolecule that we are able to detect. This non-conventional transmissible agent (NCTA) is strictly similar to the agent responsible for scrapie in small ruminants.

The pathogenic prion protein is a partially protease-resistant isoform from a host cell prion protein, expressed normally in the central nervous system. Compared to the cell prion protein, the pathogenic prion protein can not be physiologically eliminated by the proteinase K enzyme and instead accumulates in brain cell cytoplasms causing their degeneration.

The normal prion protein becomes pathogenic through conformation modification. The pathogenic prion protein, abnormally conformed, also has the ability to irreversibly modify normal protein conformation, causing it to become pathogenic.

Non-Conventional Transmissible Agent characteristics:

Can be distinguished from other pathogenic agents (bacteria, viruses, etc.) by the following properties:

• Absence of detectable nucleic acid.

• Absence of any detectable immunological reaction.

• High resistance to principal physical and chemical agents.

• Preserved through refrigeration and freezing.

• Inactivation (but not sterilisation):

- Through heat: 136°C, at 3 bars for 20min.

- In 2N sodium hydroxide solutions for 1h at 20°C.

NB: decontamination measures reduce pathogenic agent titres but their efficacy is often limited. The infectious agent survives especially in cadaveric tissue even after thorough quartering processes.

Methods of transmission **Source:**
BSE occurs through ingestion of feed containing contaminated meat and bone meal.

Virulent matter: the central nervous system as a whole: eyes, brain, spinal cord of clinically affected animals.

Age: adult bovines mainly between 4 and 7 years.

Breed: dairy (exposed to industrially processed feed, longer life expectancy, compatible with the disease incubation period).

Transmission:
Direct: through ingestion of contaminated meat and bone meal. Risk of cross-contamination in industrial food production chains intended for cattle.

- Vertical: risk of infection for calves born to affected cows. No transmission mechanism has been demonstrated.

- Horizontal: no proof to support transmission between bovines.

Indirect: no proof to support iatrogenic transmission or transmission through pasture.

Human contamination: the appearance of a new variant of Creutzfeldt-Jakob disease suggests transmission to man through food.

Method of contamination:
By oral route (ingestion of contaminated meat and bone meal).

Symptoms:
Incubation period: very long, on average 3 to 5 years.

Great individual variability as to the expression of symptoms. Central nervous signs dominate clinical presentation.

• **Behaviour disorders:** nervousness, fear, aggressiveness, stereotypical behaviour (licking, pruritis), depression. Affected animals graze apart from the rest of the herd.

• **Sensory modifications:** hyperaesthesia originating from overreaction to noise, light and touch (trembling, panic, falls).

• **Locomotor disorders:** unsteady gait, falls, difficulty in rising and in changing direction. Hypermetria and ataxia of the hind limbs may occur as motor lack of coordination.

• **Neurovegetative disorders:** suspended rumination, bradycardia, cardiac arrhythmia.

• **Other symptoms:** drop in milk production, occasionally pruritis.

• **Terminal phase:** general deterioration of health, recumbence. Progression towards death in several weeks.

Comments:

- Absence of fever. Absence of any detectable immune response.

- Cows presenting BSE symptoms at the end of the gestation period will fully express the disease after farrowing, which is considered a "stress" that reveals the disease.

Lesions**Macroscopic lesions:**
Absence of macroscopic lesions except where the animal may have fallen (traumatic lesions).

Microscopic lesions:
The only specific lesions observed in affected animals are situated in the central nervous system and particularly in the brain stem basal ganglia. The latter present symetrically distributed vacuolation of the grey matter, giving the characteristic "sponge-like" appearance to the brain.

Diagnostics**Clinical diagnosis:**
During the clinical examination of an adult bovine, any behavioural disorders associated with hypersensitivity, hyper reactivity and locomotor disorders worsening over time should arouse suspicion of BSE.

Differential diagnosis:
• Hypomagnesemia (grass tetany)

• Hypocalcaemia (milk fever)

• Silage-related Listeriosis (1/3 of BSE unconfirmed suspicions)

• Nervous forms of acetonaemia, gangrenous coryza

• Rabies

• Abscesses, brain and spinal cord tumours

• Severe copper deficiency

• Botulism

Laboratory diagnosis:
This must be carried out in a specialist laboratory

Laboratory diagnosis requires access to the brain and therefore euthanasia of the suspect animal and a rapid brain tissue sample (< 24h *post mortem*). Spongiosis lesions or the pathogenic prion protein must be present in the brain stem to confirm the diagnosis.

- First-line rapid test

Detection (in the obex and brain stem) of the pathogenic prion protein using a WesternBlot or ELISA test. This gives a reliable result available in less than 24h. It is implemented in those countries where the disease is recognised as present.

If the result is positive, it must be confirmed by a reference laboratory. If the result is negative, the test does not provide any information as to the disease responsible for the symptoms observed.

- Histopathological examination:

Whole brain sample. Characterization of the vacuolation symetrical lesions in the brain stem basal ganglia neuronal bodies. If BSE is not confirmed any other disease process relating to another disease can be detected.

Treatment:
No effective cure is known either for man or for animals.

Prophylaxis:
Sanitary prophylaxis

Disease-free countries:

• Targeted monitoring: detecting symptoms evocative of BSE

• Importation-related protection measures

• Ban on the use of meat and bone meal in animal feed

Infected countries:

• Slaughter rewarded by financial compensation for confirmation of cases by the laboratory before complete carcass destruction.

• Cohort slaughter (ancestors and offspring of the infected animal, animals of the same age having received the same feed).

• Ban on the use of meat and bone meal in animal feed.

• Animal protein recycling controls.

• Cattle identification and traceability.

• Withdrawal and systematic destruction of specified risk material (head, spinal cord, ileum, spleen, thymus, intestines, etc.) in carcasses for consumption.

Vaccines:
No human or animal vaccine exists.

- BSE [1]

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[1] http://www-old.caribvet.net/en/diseases/bse