WORLD TRADE ORGANIZATION

G/SPS/GEN/221
5 December 2000

(00-5249)

Committee on Sanitary and Phytosanitary Measures Original: English

BOVINE SPONGIFORM ENCEPHALOPATHY (BSE)

Submission by the World Health Organization (WHO)[1]

1. Bovine Spongiform Encephalopathy (BSE) is a fatal, neuro-degenerative, transmissible brain disease of cattle. The disease is fatal for cattle within weeks to months of its onset. BSE first came to the attention of the scientific community in November 1986 with the appearance in cattle of a newly-recognized form of neurological disease in the United Kingdom.

Source of the epidemic

2. Epidemiological studies conducted in the United Kingdom suggest that the source of BSE was cattle feed prepared from carcasses of ruminants, and that changes in the process of preparing cattle feed may have been a contributing risk factor.

3. Speculation as to the cause of the appearance of the agent causing the disease has ranged from spontaneous occurrence in cattle, the carcasses of which then entered the cattle food chain, to entry into the cattle food chain from the carcasses of sheep with a similar disease, scrapie.

Cause

4. BSE is associated with a transmissible agent. The agent affects the brain and spinal cord of cattle and lesions are characterized by sponge-like changes visible with an ordinary microscope.

5. The agent is highly stable, resisting freezing, drying and heating at normal cooking temperatures, such as those used for pasteurization and sterilization at usual temperatures and for usual times.

6. Under investigation, the nature of the BSE agent is still a matter of debate. According to the prion theory, the agent is composed largely, if not entirely, of a self-replicating protein, referred to as a prion. Another theory argues that the agent is virus-like and possesses nucleic acids which carry genetic information. Although strong evidence collected over the past decade supports the prion theory, the ability of the BSE agent to form multiple strains is more easily explained by a virus-like agent.

Cases of BSE

7. Between November 1986 and November 2000 approximately 180,000 cases of this newly recognized cattle disease were confirmed in the United Kingdom.

8. Since 1989 when the first BSE case was reported outside of the United Kingdom, relatively small numbers of BSE cases (in total approximately 1,500) have also been reported in native cattle in

[1] Fact Sheet N° 113, revised November 2000.

Belgium, Denmark, France, the Republic of Ireland, Liechtenstein, Luxembourg, Netherlands, Portugal and Switzerland. However, all but a couple of dozen have been reported in four countries – Ireland, Portugal, Switzerland and France. Small numbers of cases have also been reported in Canada, the Falkland Islands (Islas Malvinas), Germany, Italy and Oman, but solely in animals imported from the United Kingdom. The International Office for Epizootic Diseases (OIE) reports these cases on their web site: www.OIE.int

Measures taken to prevent the spread of BSE

9. In July 1988, the United Kingdom banned the use of ruminant proteins in the preparation of animal feed. The use in the food chain of bovine offals considered to pose a potential risk to humans was also banned in the United Kingdom in 1989, and the list was revised and expanded on several occasions as new information became available. In other countries, including European countries, measures taken, the date of implementation and the extent of enforcement vary from country to country.

10. Starting in 1996, bans prevented the sale of food and food products containing beef from the United Kingdom to other countries. Other products (e.g. tallow, gelatin) derived from bovine tissues were also prohibited from sale from the United Kingdom to other countries. However, the European Union recently lifted the ban for meat fulfilling specific requirements; for example, de-boned beef from animals from farms where there have been no cases of BSE and where the animals are less than 30 months of age at slaughter.

11. Cattle are continuously monitored for BSE and BSE is decreasing in the United Kingdom. New monitoring programmes using newly developed tests for the diagnosis of BSE in dead cattle have been introduced in Switzerland and France, and may be expected throughout the European Union.

Transmissible Spongiform Encephalopathies (TSEs) in animals

12. TSEs are diseases characterized by spongy degeneration of the brain with severe and fatal neurological signs and symptoms. BSE is one of several different forms of transmissible brain disease affecting a number of animal species. Scrapie is a common disease in sheep and goats. Mink, as well North American mule deer and elk, can contract TSEs. A neurological disease in household cats and in ruminant and feline species in zoos has been linked to consumption of food contaminated by BSE; most cases in such animals appear to have occurred in the United Kingdom.

Creutzfeldt-Jakob disease

13. While several human TSEs exist, Creutzfeldt-Jakob disease (CJD) is the prototype human TSE. CJD occurs in a form associated with a hereditary predisposition (approximately 5-10 per cent of all cases) and in a more common, sporadic form that accounts for 85-90 per cent of cases.

14. A small percentage of cases (less than 5 per cent) are iatrogenic (resulting from the accidental transmission of the causative agent via contaminated surgical equipment or as a result of cornea or dura mater transplants). It has also been shown that CJD can be transmitted to humans as a result of treatment with natural human growth hormone. Such means of transmission have now been controlled in the industrialized countries where these procedures were practised.

15. A newly recognized form of CJD, variant Creutzfeldt-Jakob disease (vCJD) was first reported in March 1996 in the United Kingdom (cf. WHO Fact Sheet N° 180 on variant Creutzfeldt-Jakob Disease). In contrast to classical forms of CJD, vCJD seems to affect mainly young patients and has a relatively long duration of illness. In contrast to the traditional forms of CJD, vCJD has affected

younger patients (average age 29 years, as opposed to 65 years), has a relatively longer duration of illness (median of 14 months as opposed to 4.5 months) and is strongly linked to exposure, probably through food, to a TSE of cattle called Bovine Spongiform Encephalopathy (BSE).

16. From October 1996 to early November 2000, 85 cases of vCJD have been reported in the United Kingdom, three in France and a single case in the Republic of Ireland. Insufficient information is available at present to make any well-founded prediction about the future number of vCJD cases.

17. Since few countries have surveillance systems, the geographical distribution of the incidence of vCJD needs to be better defined.

18. Recent studies have confirmed that vCJD is due to a strain of agent which is distinct from that of sporadic and acquired CJD. Similarities observed between the strain of the agent responsible for vCJD and those of BSE and closely related agents transmitted naturally and experimentally to different animal species, are consistent with the hypothesis discussed during two 1996 WHO consultations: that the cluster of vCJD cases is due to the same agent that caused BSE in cattle. The route of exposure is unknown, but seems likely to be through food.

19. Further data are urgently required from scientific studies on vCJD cases. More retrospective and prospective monitoring and surveillance studies on all forms of CJD, modelled on current European collaborative studies, are required throughout the world.

World Health Organization (WHO) work on TSEs

20. Since 1991, WHO convened nine scientific consultations on issues related to human and animal TSEs; the ultimate goal of the meetings was to better protect human and animal health. Experts who participated reviewed the possible human public health implications of animal TSEs, with special emphasis on BSE. The consultations also reviewed the evolving state of knowledge on these diseases, evaluated possible means of transmission and identified risk factors for infection.

21. The group of independent experts assembled by WHO is continually updating the state-of-the-art as more scientific information on BSE and vCJD becomes available. WHO provides a neutral scientific forum in which scientific questions related to BSE and vCJD can be reviewed, evaluated and debated.

22. Advancing current knowledge about TSEs through research will permit the best possible decisions to be taken to safeguard public health, while securing consumer confidence so that national economies dependent on the beef industry can be maintained and developed. Therefore, research on all TSEs is promoted by WHO, especially on early diagnostic procedures and epidemiology. WHO has published a comprehensive priority list for new research. One question which needs investigation is whether or not BSE has infected sheep populations.

23. WHO is helping to expand standardized surveillance of CJD and its variants in order to better understand the disease's geographic spread in the world and to better protect public health globally. From 1997-2000, WHO held a series of training courses worldwide, particularly in developing countries, with the intention of helping individual countries establish national surveillance of CJD and its variants.

24. WHO published guidelines for infection control of TSEs in 2000. The full text is available at http://www.who.int/emc-documents/ under the heading "TSE".

WHO conclusions and recommendations to reduce exposure to the BSE agent

25. Recent studies have confirmed that vCJD is due to a strain of agent which is distinct from that of sporadic and acquired CJD. Similarities observed between the strain of the agent responsible for vCJD and those of BSE and closely related agents transmitted naturally and experimentally to different animal species, are consistent with the hypothesis discussed during two 1996 WHO consultations, that the cluster of vCJD cases is due to the same agent that caused BSE in cattle. The route of exposure is unknown, but seems likely to be through food.

26. BSE must be eradicated. All countries must prohibit the use of ruminant tissues in ruminant feed and must exclude tissues that are likely to contain the BSE agent from any animal or human food chain. All countries are encouraged to conduct risk assessments to determine if they are at risk for BSE in sheep and goats. It is advised that any tissue which may come from deer or elk with Chronic Wasting Disease (CWD is a transmissible spongiform disease of mule deer and elk) is not used in animal or human food; however, at this time there is no evidence to suggest that CWD in deer and elk can be transmitted to humans.

27. Human and veterinary vaccines prepared from bovine materials may carry the risk of transmission of animal TSE agents. The pharmaceutical industry should ideally avoid the use of bovine materials and materials from other animal species in which TSEs naturally occur. If absolutely necessary, bovine materials should be obtained from countries which have a surveillance system for BSE in place and which report either zero or only sporadic cases of BSE. These precautions apply to the manufacture of cosmetics as well.

28. Milk and milk products are considered safe. Tallow and gelatin are considered safe if prepared by a manufacturing process which has been shown experimentally to inactivate the transmissible agent. No infectivity has yet been detected in skeletal muscle tissue. Reassurance can be provided by removal of visible nervous and lymphatic tissue from meat (skeletal muscle).

Note: For further information, journalists can contact the Spokesperson's Office, WHO, Geneva Telephone (+41 22) 791 2599. Fax (+41 22) 791 4858. E-mail: inf@who.int. All WHO Press Releases, Fact Sheets and Features can be obtained on Internet on the WHO home page: www.who.int.