CHAPTER 11.4.

BOVINE SPONGIFORM ENCEPHALOPATHY

Article 11.4.1.

General provisions

1) Bovine spongiform encephalopathy (BSE) is an invariably fatal neurological prion disease of bovines caused by a misfolded form of the prion protein (Pr^{PSc}), which includes both C-type (classical BSE) and H- and L-type (atypical BSE) agents. Oral exposure to contaminated *feed* is the main route of transmission of classical BSE. Atypical BSE is a condition that occurs at a very low rate and is assumed to occur spontaneously in any bovine population. A bovine has been experimentally infected by the oral route with a low molecular weight type of atypical BSE (L-type BSE) and the potential for recycling of atypical BSE cannot be ruled out, although there is no evidence that it plays a significant role in the epidemiology of BSE.
2) BSE primarily affects bovines. Other animal species may be naturally and experimentally susceptible to BSE, but they are not regarded as being epidemiologically significant, particularly when feeding ruminants with ruminant-derived *protein meal* is not practised. The recommendations in this chapter are intended to mitigate the human and animal health risks associated with BSE in bovines only.
3) For the purposes of the *Terrestrial Code*, the occurrence of a *case* of BSE is defined by the detection of the classical BSE agent in brain tissue of a bovine.
4) For the purposes of this chapter, 'bovine' means an animal of the species *Bos taurus* or *Bos indicus*.
5) When *commodities* are imported in accordance with this chapter, the BSE risk of the *importing country* or *zone* of destination is not affected by the BSE risk of the *exporting country*, *zone* or *compartment* of origin.
6) Standards for diagnostic tests are described in the *Terrestrial Manual*.

Article 11.4.2.

Safe commodities

When authorising the importation or transit of the following *commodities* derived from bovines, *Veterinary Authorities* should not require any conditions related to BSE, regardless of the BSE risk posed by the bovine population of the *exporting country*, *zone* or *compartment*:

1) *milk* and *milk products*;
2) semen and *in vivo* derived bovine embryos collected and handled in accordance with the relevant chapters of the *Terrestrial Code*;
3) hides and skins;
4) gelatine and collagen;
5) tallow with maximum level of insoluble impurities of 0.15% in weight and derivatives made from this tallow;
6) dicalcium phosphate (with no trace of protein or fat);
7) fetal blood.

Other *commodities* of bovines can be traded safely if in accordance with the relevant articles of this chapter.

Article 11.4.3.

General criteria for the determination of the BSE risk of a country, zone or compartment

Owing to its specific etiological and epidemiological features, the BSE risk of a country, *zone* or *compartment* is determined on the basis of the following:

1) A BSE *risk assessment*, in accordance with the provisions of the 'Application for official recognition by WOAH of risk status for bovine spongiform encephalopathy' that evaluates the risk of the classical BSE agent being recycled within the bovine population by identifying all potential factors associated with the occurrence of BSE and their

historic perspective. Member Countries should review the *risk assessment* annually to determine whether the situation has changed.

The *risk assessment* for the purpose of BSE, based on the framework provided by Article 2.1.4., consists of:

a) Entry assessment

The entry assessment evaluates the likelihood that the classical BSE agent has been introduced into the country, *zone* or *compartment* through the importation of the following *commodities* in the preceding eight years:

i) bovines;

ii) ruminant-derived *protein meal*;

iii) *feed* (except packaged and labelled pet food) that contains ruminant-derived *protein meal*;

iv) fertilisers that contain ruminant-derived *protein meal*;

v) any other *commodity* that either is or could be contaminated by *commodities* listed in Article 11.4.15.

b) Exposure assessment

The exposure assessment evaluates the likelihood of bovines being exposed to the classical BSE agent during the preceding eight years, either through imported *commodities* or as a result of the presence of the classical BSE agent within the indigenous bovine population of the country, *zone* or *compartment*.

The first step in the exposure assessment involves an evaluation of livestock industry practices through a consideration of the impact of:

i) Livestock industry practices preventing bovines from being fed ruminant-derived *protein meal*, taking account of:

- demographics of the bovine population and production and farming systems;
- feeding practices, including the use of fertilisers containing ruminant proteins on land for grazing or harvesting forage;
- slaughtering and waste management practices;
- rendering practices;
- *feed* production, labelling, distribution and storage.

Depending on the outcome from this step, an evaluation of risk mitigation measures specifically targeting BSE may also need to be included through consideration of the impact of:

ii) Specific risk mitigation measures preventing bovines from being fed ruminant-derived *protein meal*, taking account of:

- the nature and scope of a *feed* ban on feeding ruminants with *protein meal* derived from ruminants;
- the fate of *commodities* with the greatest BSE infectivity as listed in point 1 of Article 11.4.15.;
- parameters of the rendering process;
- prevention of cross-contamination during rendering, *feed* production, transport, storage and feeding;
- an awareness programme under the scope of the *feed* ban;
- monitoring and enforcement of the *feed* ban.

Depending on the outcome of the exposure assessment, a consequence assessment (in point c) below) may not be required.

c) Consequence assessment

The consequence assessment evaluates the likelihood of bovines becoming infected following exposure to the classical BSE agent together with the likely extent and duration of any subsequent recycling and amplification within the bovine population during the preceding eight years. The factors to be considered in the consequence assessment are:

i) age at exposure;

ii) production type;

iii) the impact of bovine industry practices or the implementation of BSE-specific mitigation measures under a *feed* ban.

d) Risk estimation

The risk estimation combines the results and conclusions arising from the entry, exposure and consequence assessments to provide an overall measure of the risk of the classical BSE agent being recycled within the bovine population.

2) The ongoing implementation of a *surveillance* programme for BSE in the bovine population in accordance with Article 11.4.20.

3) The history of occurrence and management of *cases* of BSE and bovines affected by atypical BSE.

Determination of the date from which the risk of BSE agents being recycled within the bovine population has been negligible is based on points 1 to 3 above.

Article 11.4.4.

Negligible BSE risk

The BSE risk of a country or *zone* can be considered to be negligible if all the following conditions for the bovine population are met for at least the preceding eight years:

1) A *risk assessment* as described in point 1 of Article 11.4.3. that has identified all potential risk factors associated with classical BSE has been conducted, and the Member Country has demonstrated through documented evidence that any identified risk factors have been adequately managed and that the risk of the classical BSE agent being recycled within the bovine population has been negligible as a result of:

   EITHER

   a) livestock industry practices ensuring that *protein meal* derived from ruminants has not been fed to ruminants;

   OR

   b) effective and continuous mitigation of each identified risk ensuring that *protein meal* derived from ruminants has not been fed to ruminants.

2) The *surveillance* provisions as described in Article 11.4.20. have been implemented.

3) EITHER:

   a) there has been no *case* of BSE or, if there has been a *case*, each *case* of BSE has been demonstrated to have been imported;

   OR

   b) if there has been an indigenous *case* of BSE:

      either:

      i) all *cases* were born before the date from which the risk of BSE agents being recycled within the bovine population has been negligible;

      or

      ii) where a *case* was born after that date, subsequent investigations have confirmed that any identified source of *infection* has been controlled and the risk of BSE agents being recycled within the bovine population has continued to be negligible.

4) Any *cases* of BSE or any bovines affected by atypical BSE that have been detected have been completely destroyed or disposed of to ensure that they do not enter the *feed* or food chain.

The country or the *zone* will be included in the list of countries or *zones* posing a negligible risk for BSE in accordance with Chapter 1.6. Retention on the list requires annual confirmation of the conditions in points 1 to 4 above. Documented evidence should be resubmitted annually for points 1 to 4 above.

Any changes in the epidemiological situation or other significant events should be notified to WOAH in accordance with Chapter 1.1.

Article 11.4.5.

Controlled BSE risk

The BSE risk of a country or *zone* can be considered to be controlled provided all of the conditions of Article 11.4.4. are met, but one or more of these conditions has not been met for the preceding eight years.

The country or the *zone* will be included in the list of countries or *zones* posing a controlled risk for BSE in accordance with Chapter 1.6. Retention on the list requires annual confirmation of the conditions in points 1 to 4 of Article 11.4.4. Documented evidence should be resubmitted annually for points 1 to 4 of Article 11.4.4.

Any changes in the epidemiological situation or other significant events should be notified to WOAH in accordance with Chapter 1.1.

Article 11.4.6.

Compartment with negligible or controlled BSE risk

The establishment and bilateral recognition of a *compartment* posing negligible or controlled BSE risk should follow the relevant requirements of this chapter and the principles laid down in Chapters 4.4. and 4.5.

Article 11.4.7.

Undetermined BSE risk

The BSE risk of a country or *zone* is considered to be undetermined if it cannot be demonstrated that it meets the requirements for negligible or controlled BSE risk.

Article 11.4.8.

Maintenance of BSE risk status

The BSE risk status of a country or *zone* is not affected by imported *cases* of BSE or *cases* of BSE born before the date from which the risk of BSE agents being recycled within the bovine population has been negligible, or by any bovine affected by atypical BSE, as long as managed in accordance with point 4 of Article 11.4.4.

Should an indigenous *case* of BSE in a bovine born after the date from which the risk of BSE agents being recycled within the bovine population has been negligible occur in a country or *zone* recognised as posing a negligible or controlled risk for BSE, the status of the country or *zone* is maintained, provided that documented evidence regarding the outcome of subsequent investigations is submitted to WOAH within 90 days demonstrating that any identified source of *infection* has been controlled and the risk of BSE agents being recycled within the bovine population has continued to be negligible.

If no documented evidence is provided or if it is not accepted by WOAH, the provisions of Article 11.4.4. or Article 11.4.5. apply.

Article 11.4.9.

Recommendations for importation of bovines from a country, zone or compartment posing a negligible or controlled BSE risk

Veterinary Authorities should require the presentation of an *international veterinary certificate* attesting that:

1) The bovines selected for export are identified through an *animal identification system* enabling them to be traced throughout their lifetime.

AND EITHER:

2) The bovines selected for export were born and kept in a country, *zone* or *compartment* posing a negligible or controlled BSE risk after the date from which the risk of BSE agents being recycled within the bovine population has been demonstrated to be negligible.

OR

3) It is demonstrated that the bovines selected for export have never been fed *protein meal* derived from ruminants.

Article 11.4.10.

Recommendations for importation of bovines from a country or zone posing an undetermined BSE risk

Veterinary Authorities should require the presentation of an *international veterinary certificate* attesting that:

1) The bovines selected for export are identified through an *animal identification system* enabling them to be traced throughout their lifetime.
2) It is demonstrated that the bovines selected for export have never been fed *protein meal* derived from ruminants.

Article 11.4.11.

Recommendations for importation of fresh meat and meat products from a country, zone or compartment posing a negligible or controlled BSE risk

Veterinary Authorities should require the presentation of an *international veterinary certificate* attesting that:

1) the bovine from which the *fresh meat* and *meat products* were derived are identified through an *animal identification system*;
2) they have been subjected to an ante-mortem inspection with favourable results;
3) they were born and kept in:
   a) a country, *zone* or *compartment* posing a negligible BSE risk; or
   b) a country, *zone* or *compartment* posing a controlled BSE risk after the date from which the risk of the BSE agents being recycled within the bovine population has been demonstrated to be negligible; or
   c) a country, *zone* or *compartment* posing a controlled BSE risk before the date from which the risk of the BSE agents being recycled within the bovine population has been demonstrated to be negligible, and the *fresh meat* and *meat products*:
      i) were derived from bovines not subjected to a *stunning* process with a device injecting compressed air or gas into the cranial cavity, or to a pithing process, or to any other procedure that can contaminate blood with nervous tissue, prior to *slaughter*; and
      ii) were produced and handled in a manner which ensures that such products do not contain and are not contaminated with the *commodities* listed in point 1 of Article 11.4.15. or mechanically separated *meat* from the skull or from the vertebral column of bovines over 30 months of age.

Article 11.4.12.

Recommendations for importation of fresh meat and meat products from a country or zone posing an undetermined BSE risk

Veterinary Authorities should require the presentation of an *international veterinary certificate* attesting that:

1) the bovines from which the *fresh meat* and *meat products* were derived are identified through an *animal identification system*;
2) it is demonstrated that the bovines from which the *fresh meat* and *meat products* were derived have never been fed *protein meal* derived from ruminants;
3) the bovines from which the *fresh meat* and *meat products* were derived:
   a) were subjected to an ante-mortem inspection with favourable results;
   b) were not subjected to a *stunning* process with a device injecting compressed air or gas into the cranial cavity, or to a pithing process, or to any other procedure that can contaminate blood with nervous tissue, prior to *slaughter*;
4) the *fresh meat* and *meat products* were produced and handled in a manner which ensures that such products do not contain and are not contaminated with:
   a) the *commodities* listed in point 1 of Article 11.4.15.;
   b) mechanically separated *meat* from the skull or from the vertebral column of bovines over 30 months of age.

Article 11.4.13.

Recommendations for importation of bovine-derived protein meal from a country, zone or compartment posing a negligible BSE risk

Veterinary Authorities should require the presentation of an *international veterinary certificate* attesting that the bovines from which the *protein meal* was derived were identified through an *animal identification system* and were born and kept in a country, *zone* or *compartment* posing a negligible BSE risk, and

EITHER

1) they were born after the date from which the risk of BSE agents being recycled within the bovine population has been demonstrated to be negligible;

OR

2) the *protein meal* was processed in accordance with Article 11.4.19.

Article 11.4.14.

Recommendations for importation of blood and blood products derived from bovines (except fetal blood)

Veterinary Authorities should require the presentation of an *international veterinary certificate* attesting that:

EITHER

1) the blood and blood products were derived from bovines that were identified through an *animal identification system* and were born and kept in a country, *zone* or *compartment* posing a negligible risk, or a country, *zone* or *compartment* posing a controlled BSE risk after the date from which the risk of BSE agents being recycled within the bovine population has been demonstrated to be negligible;

OR

2) the blood and blood products were:
   a) collected from bovines not subjected to a *stunning* process with a device injecting compressed air or gas into the cranial cavity, or to a pithing process, or to any other procedure that can contaminate the blood with nervous tissue, prior to *slaughter*; and
   b) collected and processed in a manner that ensures they are not contaminated with nervous tissue.

Article 11.4.15.

Recommendations in relation to the trade of the commodities with the greatest BSE infectivity

Unless covered by other articles in this chapter, the following *commodities* should not be traded:

1) Distal ileum from bovines of any age; skull, brain, eyes, vertebral column and spinal cord from bovines that were at the time of *slaughter* over 30 months of age, or any *commodity* contaminated by them, which originate from a country, *zone* or *compartment* posing:
   a) an undetermined BSE risk;
   b) a controlled BSE risk if they are derived from bovines born before the date from which the risk of BSE agents being recycled within the bovine population has been demonstrated to be negligible.
2) Food, *feed*, fertilisers, cosmetics, pharmaceuticals including biologicals, medical devices or any other product containing proteins prepared using *commodities* listed in point 1 above.
3) Bovine-derived *protein meal* or any *commodities* containing such product which originate from a country, *zone* or *compartment* posing a controlled or undetermined BSE risk.

Article 11.4.16.

Recommendations for importation of tallow (other than as defined in Article 11.4.2.)

Veterinary Authorities should require the presentation of an *international veterinary certificate* attesting that the tallow:

1) came from a country, *zone* or *compartment* posing a negligible BSE risk; or
2) is derived from bovines which have been subjected to an ante-mortem inspection with favourable results, and has not been prepared using the *commodities* listed in point 1 of Article 11.4.15.

Article 11.4.17.

Recommendations for importation of tallow derivatives (other than as defined in Article 11.4.2.)

Veterinary Authorities should require the presentation of an *international veterinary certificate* attesting that the tallow derivatives either:

1) originate from a country, *zone* or *compartment* posing a negligible BSE risk; or
2) are derived from tallow that meets the conditions referred to in Article 11.4.16.; or
3) have been produced by hydrolysis, saponification, or transesterification that uses high temperature and pressure.

Article 11.4.18.

Recommendations for importation of dicalcium phosphate (other than as defined in Article 11.4.2.)

Veterinary Authorities should require the presentation of an *international veterinary certificate* attesting that the dicalcium phosphate:

1) came from a country, *zone* or *compartment* posing a negligible BSE risk; or

2) is a co-product of bone gelatine.

Article 11.4.19.

Procedures for reduction of BSE infectivity in bovine protein meal

The following procedure should be used to reduce the infectivity of any BSE agents that may be present during the production of *protein meal* containing bovine proteins:

1) the raw material should be reduced to a maximum particle size of 50 mm before heating and the raw material should be heated under saturated steam conditions to a temperature of not less than 133°C for a minimum of 20 minutes at an absolute pressure of 3 bar; or

2) an alternative procedure that has been demonstrated to achieve at least an equivalent level of reduction in BSE infectivity.

Article 11.4.20.

Surveillance

The objective of BSE *surveillance* is to detect occurrence of BSE within the bovine population.

1) BSE is a progressive, fatal disease of the nervous system of bovines that usually has an insidious onset and that is refractory to treatment. A range of clinical signs that vary in severity and between animals have been described for classical BSE:

   a) progressive behavioural changes that are refractory to treatment such as increased excitability, depression, nervousness, excessive and asymmetrical ear and eye movements, apparent increased salivation, increased licking of the muzzle, teeth grinding, hypersensitivity to touch and/or sound (hyperaesthesia), tremors, excessive vocalisation, panic-stricken response and excessive alertness;

   b) postural and locomotory changes such as abnormal posture (dog sitting), abnormal gait (particularly pelvic limb ataxia), low carriage of the head, head shyness, difficulty avoiding obstacles, inability to stand and recumbency;

   c) generalised non-specific signs such as reduced *milk* yield, loss of body condition, weight loss, bradycardia and other disturbances of cardiac rhythm.

   Some of these signs are also likely to be relevant for atypical BSE, particularly those associated with difficulty in rising and recumbency. A nervous form of atypical BSE resembling classical BSE may be observed with over-reactivity to external stimuli, unexpected startle responses and ataxia. In contrast, a dull form of atypical BSE may be observed, with dullness combined with a low head carriage and compulsive behaviour (licking, chewing, pacing in circles).

   The clinical signs of BSE usually progress on a spectrum over a few weeks to several months, but on rare occasions cases can develop acutely and progress rapidly. The final stages of the disease are characterised by recumbency, coma and death.

   Since these signs are not pathognomonic for either classical or atypical BSE, all Member Countries with bovine populations are likely to observe individual animals displaying clinical signs suggestive of BSE. General statements about the likely frequency of occurrence of such animals cannot be made as they will vary depending on the epidemiological situation in a particular country.

2) *Surveillance* for BSE includes all bovines that show signs of the clinical spectrum of BSE.

   In production and farming systems that allow bovines to be subjected to regular observation, it is likely that animals that display clinical signs suggestive of BSE will be more readily seen. Behavioural changes, which may be very subtle in the early clinical phase, are best identified by those who handle animals on a daily basis and who can monitor them closely for a progression of the signs. In production and farming systems, where bovines are not monitored as closely, situations may arise where an animal might be considered as a clinical suspect, yet if It has

not been observed for a period of time, it may only be initially seen as unable to rise or walk without assistance or found dead (fallen stock).

The *surveillance* programme should take into account that the vast majority of *cases* of BSE arise as single, isolated events. The concurrence of multiple animals with behavioural or neurological signs, or non-ambulatory or fallen stock is most likely associated with other causes.

The animals that lie on the clinical spectrum of BSE should be targeted for BSE *surveillance* and the following animals should be reported and followed up with appropriate laboratory testing in accordance with the *Terrestrial Manual* to accurately confirm or rule out the presence of BSE agents, including discrimination between atypical and classical BSE strains:

a) those displaying progressive clinical signs suggestive of BSE mentioned in point 1 that are refractory to treatment, and where the clinical presentation cannot be attributed to other common causes of behavioural or neurological signs (e.g. infectious, metabolic, traumatic, neoplastic or toxic causes);

b) those showing behavioural or neurological signs at ante-mortem inspection at *slaughterhouses/abattoirs*;

c) those unable to rise or walk without assistance, with an appropriate supporting clinical history (i.e. the clinical presentation cannot be attributed to other common causes of recumbency);

d) those found dead (fallen stock), with an appropriate supporting clinical history (i.e. the clinical presentation cannot be attributed to other common causes of death).

3) The credibility of the *surveillance* programme is supported by:

a) ongoing awareness and training programmes to ensure that all those stakeholders involved in the rearing and production of livestock, including bovine breeders, owners and keepers, *veterinarians*, transporters and *slaughterhouse/abattoir* workers are familiar with the clinical signs suggestive of BSE as well as the statutory reporting requirements;

b) the fact that BSE is a *notifiable disease* throughout the whole territory;

c) appropriate *laboratory* testing in accordance with the *Terrestrial Manual*;

d) robust, documented, evaluation procedures and protocols for:

- the definition of the target population for BSE *surveillance*,
- the reporting of bovines described in points 2 a) to 2 d),
- the determination of animals to be subjected to laboratory testing,
- the collection and submission of samples for laboratory testing,
- the follow-up epidemiological investigations for BSE positive findings.

NB: FIRST ADOPTED IN 1992; MOST RECENT UPDATE ADOPTED IN 2023.