AUSTRALIAN VETERINARY EMERGENCY PLAN

AUSVETPLAN

Response strategy

Bovine spongiform encephalopathy

Version 5.0

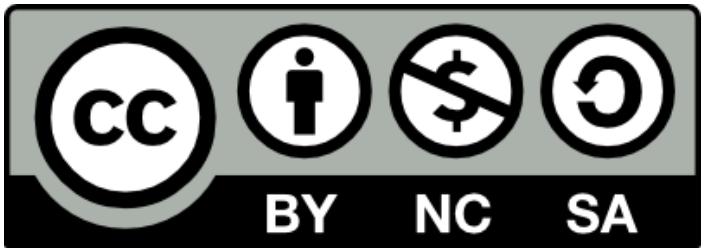
AUSVETPLAN is a series of technical response plans that describe the proposed Australian approach to an emergency animal disease incident. The documents provide guidance based on sound analysis, linking policy, strategies, implementation, coordination and emergency-management plans.

National Biosecurity Committee

ISBN 0 642 24506 1 (printed version)

ISBN 1 876 71438 7 (electronic version)

Licence



Moral rights

Disclaimer and warranty

Text under development

In this manual, text placed in square brackets [xxx] indicates that that aspect of the manual remains unresolved or is under development; such text is not part of the official manual. The issues will be further worked on by experts and relevant text included at a future date.

Contact information

If you have any requests or inquiries concerning reproduction and rights, or suggestions or recommendations, you should address these to:

AUSVETPLAN – Animal Health Australia
Executive Manager, Emergency Preparedness and Response
PO Box 5116
Braddon ACT 2612
Tel: 02 6232 5522
email: aha@animalhealthaustralia.com.au

Approved citation

Animal Health Australia (2021). *Response strategy: Bovine spongiform encephalopathy* (version 5.0). Australian Veterinary Emergency Plan (AUSVETPLAN), edition 5, Canberra, ACT.

DISEASE WATCH HOTLINE: 1800 675 888

The Disease Watch Hotline is a toll-free telephone number that connects callers to the relevant state or territory officer to report concerns about any potential emergency disease situation. Anyone suspecting an emergency disease outbreak should use this number to get immediate advice and assistance.

Edition 2:
Version 2.0, 1996 (new manual)
Version 2.1, 1998 (update to include new disease information from the United Kingdom, etc.)

Edition 3:
Version 3.0, January 2003 (major update of text and policy, and inclusion of new cost-sharing arrangements)
Version 3.1, January 2005 (update)
Version 3.2, February 2012 (update of scientific information on the disease, refinements to the control policy and inclusion of a movement controls matrix)

Edition 5:
Version 5.0, 2021 (incorporation into the Edition 5 format)

Contents

Tables

1 Introduction

1.1 This manual

1.1.1 Purpose

As part of AUSVETPLAN (the Australian Veterinary Emergency Plan), this response strategy contains the nationally agreed approach for the response to an incident – or suspected incident – of bovine spongiform encephalopathy (BSE) in Australia. It has been developed to guide decision making to ensure that a fast, efficient and effective response can be implemented consistently across Australia with minimal delay.

1.1.2 Scope

This response strategy covers BSE caused by the BSE prion.

This response strategy provides information about:

- the disease (Section 2)
- the implications for Australia, including potential pathways of introduction, social, environmental, human health and economic effects, and the critical factors for a response to the disease (Section 3)
- the agreed policy and guidelines for agencies and organisations involved in a response to an outbreak (Section 4)
- declared areas and premises classifications (Section 5)
- biosecurity controls, including quarantine and movement controls (Section 6)
- response surveillance and establishing proof of freedom (Section 7).

The key features of BSE are described in the **Bovine spongiform encephalopathy Fact Sheet** (Appendix 1).

1.1.3 Development

The strategies in this document for the diagnosis and management of an outbreak of BSE are based on risk assessment. They are informed by the recommendations in the World Organisation for Animal Health (OIE) *Terrestrial animal health code* (Chapter 11.4) and the OIE *Manual of diagnostic tests and vaccines for terrestrial animals* (Chapter 3.4.5). The strategies and policy guidelines are for emergency situations and are not applicable to policies for imported animals or animal products.

This manual has been produced in accordance with the procedures described in the **AUSVETPLAN *Overview***, and in consultation with Australian national, state and territory governments; the relevant livestock industries; nongovernment agencies; and public health authorities, where relevant.

In this manual, text placed in square brackets [xxx] indicates that that aspect of the manual remains unresolved or is under development; such text is not part of the official manual. The issues will be worked on by experts and relevant text included at a future date.

1.2 Other documentation

This response strategy should be read and implemented in conjunction with:

- other AUSVETPLAN documents, including the operational, enterprise and management manuals; and any relevant guidance and resource documents. The complete series of manuals is available on the Animal Health Australia website[1]
- relevant nationally agreed standard operating procedures (NASOPs).[2] These procedures complement AUSVETPLAN and describe in detail specific actions undertaken during a response to an incident. NASOPs have been developed for use by jurisdictions during responses to emergency animal disease (EAD) incidents and emergencies
- relevant jurisdictional or industry policies, response plans, standard operating procedures and work instructions
- relevant Commonwealth and jurisdictional legislation and legal agreements (such as the Emergency Animal Disease Response Agreement – EADRA[3]), where applicable.

1.3 Training resources

EAD preparedness and response arrangements in Australia

The EAD Foundation Online course[4] provides livestock producers, veterinarians, veterinary students, government personnel and emergency workers with foundation knowledge for further training in EAD preparedness and response in Australia.

1.3.1 Disease-specific training

National Transmissible Spongiform Encephalopathies Surveillance Program (NTSESP) Training Guide (2008). Clinical signs and brain removal techniques for TSE surveillance in Australia, Animal Health Australia. Available via:

- compact disc prepared and distributed by Animal Health Australia, June 2008
- www.animalhealthaustralia.com.au/what-we-do/disease-surveillance/tse-freedom-assurance-program/surveillance-of-tses/.

[1] www.animalhealthaustralia.com.au/our-publications/ausvetplan-manuals-and-documents
[2] www.animalhealthaustralia.com.au/what-we-do/emergency-animal-disease/nationally-agreed-standard-operating-procedures
[3] https://animalhealthaustralia.com.au/what-we-do/emergency-animal-disease/ead-response-agreement
[4] www.animalhealthaustralia.com.au/emergency-animal-disease-training-program

2 Nature of the disease

Three known strains of bovine spongiform encephalopathy (BSE) have been identified in cattle: classical BSE, low-type (L-type) BSE and high-type (H-type) BSE. L-type BSE and H-type BSE are also collectively called 'atypical BSE'.

BSE is a progressive neurodegenerative disease of adult cattle. It was first recognised in the United Kingdom (UK) in 1986 (Wells et al 1987, Kimberlin 1992, OIE 1996) and became a serious epidemic in that country. Atypical BSE is a very rare disease that has been recognised in a number of countries for less than 10 years. All three strains of the disease are transmissible spongiform encephalopathies (TSEs) or 'prion' diseases. TSEs are characterised by long incubation periods, the accumulation in the central nervous system (CNS) of an abnormal isoform of a host-encoded prion protein (PrP), and a possible manifestation in sporadic, inherited or acquired forms (Prusiner 1998).

The BSE agent causes a disease in people similar to that in cattle. BSE is therefore of concern not only for the welfare of cattle, but also for food safety. An outbreak due to any of these agents will involve veterinary authorities, health authorities and food safety agencies.

2.1 Aetiology

A protease-resistant isoform (PrP^{Sc}) of a normal cellular prion protein (PrP^{c}) has a pivotal role in the pathogenesis of TSEs and, according to the prion hypothesis, is the sole TSE transmissible agent (Prusiner 1998).

Other aetiological possibilities have largely been discounted. They include a robust virus, a virino (a nucleic acid protected by host protein), environmental factors and toxic chemicals.

A particular feature of the abnormal isoform of prion protein is resistance to inactivation by physical or chemical procedures, including freezing, desiccation, ultraviolet radiation, burial, common methods for chemical and heat disinfection, and degradation by certain proteolytic enzymes (Taylor DM 1996ab, Taylor K 1996).

BSE

The BSE epidemic in the UK resulted from feeding cattle meat-and-bone meal (MBM) contaminated with the BSE agent. However, the origin of the BSE agent itself is uncertain (Collee and Bradley 1997ab, Brown et al 2001). Hypotheses include a cross-species transmission of the prion responsible for scrapie in sheep, and a novel prion arising in cattle or another mammalian species (UK DEFRA 2001, Capobianco et al 2007).

Atypical BSE

Atypical BSE is characterised by either a lower (L-type) or higher (H-type) molecular mass of the unglycosylated abnormal form of prion protein, determined using western blot analyses. These strains have been detected in a number of countries during large-scale surveillance for BSE in cattle. The origin of these rare conditions is not yet known, but a spontaneous, noncontagious origin cannot be excluded. One case of H-type BSE identified in the United States was attributed to a heritable polymorphism in the prion gene (Nicholson et al 2008).

2.2 Susceptible species

2.2.1 BSE

Species that have been experimentally infected with the BSE agent, both parenterally (by injection) and orally, include mice, cattle, sheep, goats, nonhuman primates and mink.

Domestic cattle

BSE is primarily a disease of domestic cattle (genus *Bos*), but it also affects other bovine animals, including buffalo (genus *Bubalus*). In this manual, any references to cattle may also refer to buffalo.

Wild bovids and cats

During the BSE epidemic in cattle in the UK, a spongiform encephalopathy was also identified in various zoo species — including antelopes and cattle (Bovidae) and cats (Felidae) — as well as in domestic cats. Affected exotic species included ankole cattle, Arabian oryx, eland, gemsbok, kudu, nyala, scimitar-horned oryx, bison, cheetah, puma, ocelot and lion. In several of these cases, bioassay studies in mice produced a characteristic incubation period and profile of neuropathological changes, indicating that the aetiological agent was the BSE agent. Affected bovid species had received MBM as a dietary supplement, and the exotic felid species were fed bovine carcases, including spinal cord.

Small ruminants

Sheep have been experimentally infected with the BSE agent, and the disease agent had a tissue distribution that also involved the lymphoreticular system, similar to that seen with classical scrapie; BSE was naturally transmitted between sheep in an experimental flock (Bellworthy et al 2005ab). BSE challenge of sheep that have a PrP genotype resistant to classical scrapie has resulted in subclinical infection (Bencsik and Baron 2007). The question of BSE in sheep arises because sheep in the UK were fed the same contaminated MBM that drove the BSE epidemic in cattle.

The European Union has had an extensive surveillance program in place for some years in an attempt to identify whether BSE exists in small ruminants. Despite many hundreds of thousands of tests on brains from sheep and goats, the only cases of BSE confirmed (retrospectively) in naturally infected small ruminants have been in a goat that died in 2002 (France) and in a goat that died in 1990 (UK). Risk assessments have concluded that the prevalence of BSE in the UK sheep flock was zero or very low, if it was present at all (SEAC 2006).

Pigs

Pigs are susceptible to BSE infection, and developed a TSE disease following multiple injections with BSE brain homogenate, but have not been shown to be susceptible to oral challenge.

Chickens

Chickens have not developed BSE following either injection or oral exposure.

Dogs and horses

No cases have been reported in dogs or horses.

Primates

Various nonhuman primate species are susceptible to BSE, both naturally and in experiments.

2.2.2 Atypical BSE

Natural cases of atypical BSE have only been found in cattle. Cattle have been experimentally infected with both strains (L-type and H-type) by intracerebral injection, and the same route has been used to infect nonhuman primates with the L-type BSE agent (Comoy et al 2008, Kong et al 2008, Lombardi et al 2008). Oral challenge studies in cattle and nonhuman primates will further assess the potential for intraspecies and interspecies transmission of these strains and their zoonotic risk. The known epidemiology of these strains indicates that it is highly unlikely that they are spread horizontally or vertically from cattle.

2.2.3 Zoonotic potential

Creutzfeldt–Jakob disease (CJD) is a TSE that affects humans. Most cases arise spontaneously with no known cause (sporadic CJD) — the annual incidence in countries worldwide is approximately one case per million people. Some cases of CJD have also occurred because of health-care related procedures in which the infection has been transmitted from an infected individual to another individual through infected biological products or instruments (iatrogenic CJD). Some families also have a predisposition to the disease (familial CJD).

In addition to these known forms of the disease (sporadic, iatrogenic and familial), in March 1996, the UK reported 10 cases of a new clinicopathological variant of CJD (variant CJD or vCJD) in adolescents and adults under the age of 40 years with unusual neuropathological findings (Will et al 1996).

Like BSE, vCJD is a degenerative disease affecting the CNS and is always fatal. Primary cases are caused by the consumption of foods containing specified risk materials (SRMs) — such as brain and spinal cord — from BSE-affected cattle. In laboratory studies, the pathological agents isolated from BSE-affected cattle and human cases of vCJD have shown similar distinctive biological and molecular-biological features (Collinge et al 1996, Lasmezas et al 1996, Bruce et al 1997, Hill et al 1997).

Since vCJD was first identified in the UK, further cases have occurred there and in mainland Europe. There have been a few cases in some non-European countries, in individuals who lived in the UK or may have consumed foods from the UK that contained SRMs. A small number of secondary vCJD cases has been reported in the UK, due to blood transfusion from asymptomatic, infected donors. This initially led to concerns about an impending epidemic of the disease. However, it appears likely that the number of cases will be much smaller than originally predicted (Clarke and Ghani 2005). Up-to-date information on the incidence of vCJD can be obtained from the website of the United Kingdom CJD Research and Surveillance Unit.[5]

[5] www.cjd.ed.ac.uk

2.3 World distribution

For the latest information on the distribution of BSE, refer to the World Organisation for Animal Health (OIE) World Animal Health Information System.[6]

2.3.1 Distribution outside Australia

BSE was first diagnosed in the UK in 1986, and its annual case incidence there peaked in 1992. Although the great majority of cases have occurred in cattle in the UK, smaller scale epidemics, linked to the export of live cattle and MBM from the UK and subsequently from other BSE-affected countries, have occurred in mainland Europe, Canada, Japan and Israel.

Around 50 cases of atypical BSE had been identified in cattle by 2009 from the UK, mainland Europe, Canada, the United States and Japan. Several countries have reported these atypical strains in the absence of BSE cases in indigenous cattle.

Two cases of feline TSE have been diagnosed in imported animals in Australian zoos. In 1992, a case was seen in a cheetah imported from the UK to a zoo in Western Australia, and the agent was subsequently typed as the classical BSE strain (Peet and Curran 1992). This animal and two littermates imported at the same time were destroyed and incinerated. The source of infection was traced to a zoo in the UK. In July 2002, a second case was diagnosed in an Asiatic golden cat imported from the Netherlands (Young and Slocombe 2003). The cat, which was born in Germany, died suddenly of a pancreatic condition, and the TSE was detected as an incidental finding on routine histopathology of the brain.

2.3.2 Occurrence in Australia

No strain of BSE has been identified in cattle in Australia to date.

2.4 Epidemiology

The epidemiology of atypical BSE is not well understood. Millions of cattle worldwide have been screened for BSE strains, but, from 1986 to 2009, only around 50 cases of these rare diseases had been diagnosed in Europe, Canada, the United States and Japan. All cases, except one in Japan, have been reported in cattle 8 years of age or older (Dobly et al 2010). They may have a spontaneous, noncontagious origin (Biacabe et al 2008). A heritable polymorphism in the PrP gene responsible for one case of H-type BSE in the United States is rare (Heaton et al 2008). Based on this information, the likelihood of these rare conditions arising in the indigenous cattle population is extremely low.

The rest of this section concerns the epidemiology of classical BSE in cattle, which is determined principally by its long incubation period and its mode of transmission — in the natural setting, ingestion mainly by young animals of feeds containing BSE-contaminated MBM (Collee and Bradley 1997ab, Wilesmith 1998, Brown et al 2001). All BSE cases in countries other than the UK have origins in the importation and feeding to young cattle of MBM, or the importation from the UK of live cattle that entered the animal feed chain.

[6] www.oie.int/animal-health-in-the-world/the-world-animal-health-information-system/the-oie-data-system

The OIE has assessed that there is a negligible risk that BSE is present in Australia or that it has been introduced to cattle in Australia through the importation of commodities potentially contaminated with the disease agent.

2.4.1 Incubation period

The age-specific incidence of BSE in the UK has provided insight into the incubation period of the disease and its distribution (Wilesmith 1998). Most cattle became infected in the first 6 months of life, and the incubation period is long (the average is cited as 5 years). In the UK dataset from 1987 to 1997, 90% of cases occurred in cattle from 3 to 8 years of age, and 10% occurred in cattle aged 9 years and over. The age profile of infected cattle has steadily increased in European countries as strict controls on animal feeds have minimised the number of cattle being infected — 125 cases were reported in the European Union in 2008, compared with 2167 in 2001 (EC 2009).

OIE incubation period

For the purposes of the OIE, the incubation period for BSE is a minimum of 2 years, but can be greater than 10 years.

2.4.2 Persistence of agent and modes of transmission

General properties

Residues of contaminated MBM stored on farm and fed to cattle after 1996 may be responsible for the continuing trickle of BSE cases in cattle born after the feed ban in some countries. Because of their peculiar protein structure, prions are resistant to freezing, desiccation, ultraviolet radiation, most disinfectants and burial. The CJD agent can remain infectious for 28 months at room temperature after the infected person's death. On the other hand, pH extremes and some organic acids can inactivate prions.

Live animals

BSE is not a contagious disease of cattle in the usual sense, and there is no evidence for horizontal or vertical spread of BSE between animals. This is consistent with the restriction of its infectivity largely to CNS tissue, and is supported by the fact that very few cases of BSE have been reported in cattle in the UK born after the introduction of the comprehensive feed ban on 1 August 1996 (referred to as 'born after the real feed ban', or BARB, cattle). The continuing appearance of BSE in BARB cattle in the UK can be attributed to residues of contaminated MBM on farms (Hill 2005).

Most cattle become infected with BSE when they are calves (Donnelly and Ferguson 2000). Using a computer simulation model, Wilesmith et al (1988) demonstrated that the risk of exposure was 30 times greater for calves than for adult cattle. The most compelling evidence for infection occurring mainly during calfhood is the peak age incidence of BSE and the feeding patterns of the dairy industry in the UK (Wilesmith 1998). Cattle usually present with the disease at about 5–7 years old, and the peak in 1995–96 seen in the UK after the 1988 feed ban is consistent with a 5-year incubation period. Wilesmith et al (1988) also demonstrated that most cattle were infected in the first 6 months of life.

The movement of clinically normal but infected cattle is a risk factor for the introduction of BSE into new countries if rendered material from such cattle enters the cattle feed supply. This risk applies

during the period of infectivity of tissues from such cattle, which begins shortly before the appearance of clinical signs (Wells et al 1998, Wells et al 2007).

After ingestion in contaminated feed, the BSE agent spreads in an infected animal via the neural route to the CNS. Experimental data point to simultaneous spread of infection via the vagus nerve and splanchnic nerves to the spinal cord, from where infection ascends to the brain (EFSA 2007).

Attack rate studies in the UK have demonstrated that high doses can decrease the incubation period, but very low doses have more influence on lowering attack rates than on increasing the incubation period. Epidemiological and experimental data suggest that most natural BSE cases were exposed to low doses. The oral ID50 (that is, the dose needed to orally infect 50% of exposed cattle) for clinical BSE cases is around 0.2 g of BSE brain tissue (Wells et al 2007), and one of 15 orally challenged calves became infected at a dose of 0.001 g of BSE brain tissue (SEAC 2003). When cattle are orally challenged with 1 g of BSE brain tissue, the shortest incubation period seen is 45 months. Experimental data also show that BSE infectivity in the CNS is below detectable levels or absent until 75% of the average incubation period has passed (EFSA 2007, Wells et al 2007). End-point titration of the pool of brainstem homogenate used in these studies in RIII mice gave a titre of 103.5 mouse parenteral ID50/g.

Several publications have reviewed the infected tissue distribution of BSE-affected cattle. Table 2.1 shows estimates of the levels of infectivity of each tissue (expressed as ID50 units) at the height of infectivity for that tissue.

Table 2.1. Estimate of cattle oral ID_{50} with each tissue at the height of infectivity

Tissue	**Weight of tissue**	**Infectivity**		**% of total infectivity**
	(g/animal)	**ID_{50}/g**	**ID_{50}/animal**	
Brain	500	50	25 000	60.2
Spinal cord	200	50	10 000	24.1
Distal ileum	800	5	4 000	9.6
Dorsal root ganglia	30	50	1 500	3.6
Trigeminal ganglia	20	50	1 000	2.4
Tonsil	50	0.005	0.25	0.0
Total	**1 600**		**41 500**	

Source: Comer and Huntly (2003)

BSE in cattle differs from some other TSEs in that infectivity in the lymphoreticular system is slight, and located in Peyers patches and tonsils. Infectivity appears in the Peyers patches in the distal ileum between 6 and 18 months after exposure, and reappears between 36 and 40 months after exposure.

Up-to-date information on this and other BSE research can be obtained from the websites of the UK Department of Environment, Food and Rural Affairs,[7] the Advisory Committee on Dangerous Pathogens[8] and the European Food Safety Authority.[9]

[7] www.gov.uk/government/publications/active-tse-surveillance-statistics
[8] www.gov.uk/government/groups/advisory-committee-on-dangerous-pathogens
[9] www.efsa.europa.eu/en/topics/topic/bovine-spongiform-encephalopathy-bse

Animal products

TSE agents survive for long periods in carcases and withstand many of the procedures currently used to process products. The OIE has made recommendations regarding the destruction of the BSE agent in Chapter 11.4 of the OIE Terrestrial Code[10].

Data from studies on the infectivity of cattle tissues have enabled international standards to be established for tissues that transmit BSE and tissues that can be safely traded. Irrespective of the BSE risk status of a country, the following commodities are recognised by the OIE as not representing a risk of transmitting BSE[11]:

- milk and milk products
- semen and in vivo–derived cattle embryos collected and handled in accordance with the recommendations of the International Embryo Technology Society[12]
- hides and skins
- gelatine and collagen prepared exclusively from hides and skins
- tallow with maximum level of insoluble impurities of 0.15% in weight, and derivatives made from this tallow
- dicalcium phosphate (with no trace of protein or fat)
- deboned skeletal muscle meat (excluding mechanically separated meat) from cattle that were not subjected to a stunning process before slaughter, with a device injecting compressed air or gas into the cranial cavity, or to a pithing process, and that passed antemortem and postmortem inspections and was prepared in a manner to avoid contamination with SRMs (see below)
- blood and blood byproducts from cattle that were not subjected to a stunning process before slaughter, with a device injecting compressed air or gas into the cranial cavity, or to a pithing process.

The OIE recognises that:

- tonsils and distal ileum from cattle of any age represent a BSE risk for controlled and undetermined BSE risk countries (see Section 7 for further information on OIE country categories)
- brain, eyes, spinal cord, skull and vertebral column represent a BSE risk if they are derived from cattle over 30 months of age in a controlled BSE risk country, or over 12 months of age in an undetermined BSE risk country
- cattle tissues from a negligible BSE risk country (the status given to Australia) do not represent a BSE risk.

Decontamination is discussed in Section 4.3.10.

Semen and embryos from live susceptible animals

In an extensive study by Wrathall et al (2002), embryos from cattle clinically affected with BSE were implanted into New Zealand–born, BSE-free cattle. The embryos did not transmit BSE to the recipient cattle. In addition, when more than 1000 nonviable embryos were inoculated intracerebrally into

[10] www.oie.int/index.php?id=169&L=0&htmfile=chapitre_bse.htm
[11] www.oie.int/index.php?id=169&L=0&htmfile=chapitre_bse.htm
[12] Based on available research and field information, the Research Subcommittee of the Health and Safety Advisory Committee of the International Embryo Transfer Society has categorised some diseases based on their relative risk of dissemination by properly processed and handled in vivo–derived embryos.

susceptible mice, no lesions were demonstrated after 2 years. It is important to note, however, that the embryos were washed according to internationally accepted standards.

Biological products (eg vaccines)

TSEs can be spread iatrogenically. For example, CJD has been transmitted between people through extracts of human pituitary gland that were contaminated with the disease agent. Biological products derived from the tissues of cattle affected with BSE therefore provide a possible route of transmission of the disease and must be considered during disease investigations.

Although TSEs must be considered in risk assessments for biological products, there is no epidemiological evidence that such products have been a source of BSE cases in the UK or elsewhere. Quarantine controls are in place in Australia for the importation of biological products such as veterinary vaccines.

Equipment, including personal items

Surgical and veterinary instruments are not recognised as a route of BSE spread to cattle. The potential for transmission of BSE by fomites is limited, because contamination requires exposure to CNS tissue from affected cattle. However, care is required in the disposal or decontamination of equipment used for the postmortem removal of brain tissue from suspected BSE cases. As an aberrant protein, TSE agents are very resistant to the physicochemical conditions that inactivate conventional viruses and bacteria. Prions may persist on veterinary instruments that have been steam sterilised at 121 °C or decontaminated by most commonly applied chemical procedures. Surgical instruments used for procedures with CNS exposure (eg eye ablation) may be contaminated if the animal is incubating BSE, but such procedures are rare. This form of transmission is therefore extremely unlikely.

Other equipment, vectors and materials do not have a role in spreading BSE.

Arthropod vectors

Vectors do not play a role in the transmission of BSE.

2.4.3 Factors influencing transmission

The most significant risk factor for the transmission of BSE to cattle is the feeding of MBM contaminated with the BSE agent. Global eradication of BSE is expected, following the implementation since 1996 of measures to prevent the feeding of ruminant-derived MBM to cattle.

2.5 Diagnostic criteria

There is no validated diagnostic test currently available for the BSE agent in live animals. Laboratory tests on brain and spinal cord tissue obtained at postmortem examination are therefore required for confirmation of this disease.

The *Australian and New Zealand Standard Diagnostic Protocols* for TSEs (SCAHLS 2010) is the authoritative guide to laboratory diagnosis. Its methods are consistent with the current edition of the OIE *Manual of Diagnostic Tests and Vaccines for Terrestrial Animals* (OIE Terrestrial Manual). Submission of samples to an international reference laboratory may also be required.

2.5.1 Clinical signs

Due to the long incubation period of BSE, signs in cattle that were exposed as calves usually appear when the animals are between 3 and 7 years of age. BSE usually has an insidious onset and a slowly progressive clinical course extending over weeks to months. The following three signs are most frequently seen in affected animals:

- apprehension (mental status)
- hyperaesthesia (sensation)
- ataxia (posture and movement).

At least one of these signs is present in most BSE cases.

Changes in mental status affect behaviour and temperament; the first sign of BSE may be when a normally placid animal becomes aggressive and kicks in the milking shed. Hypersensitivity can be to touch, sound and light. Ataxia affects mainly the hind limbs. Other posture and movement abnormalities include falling, tremor and abnormal head carriage. In advanced cases, generalised weakness and loss of condition can cause recumbency, and signs of altered mental status and hyperaesthesia may no longer be obvious. The clinical history of any recumbent or chronically wasted animal should be sought, especially in an abattoir situation. Loss of bodyweight and reduced milk yield often accompany the nervous signs as the disease progresses.

In Europe, BSE is also considered in the differential diagnosis of 'sudden' death or cases of purported misadventure. A higher incidence of BSE has been found in Europe in emergency slaughter cattle than in cattle passing preslaughter inspection; when BSE has been diagnosed in either circumstance, there is often a history of overlooked subtle, early clinical signs of BSE.

All natural cases of atypical BSE, except one in Japan, have been reported in cattle that are at least 8 years of age (Dobly et al 2010). Clinical signs of atypical BSE (when present) can be similar to those of classical BSE; experimentally, they have included mental dullness and amyotrophy (Lombardi et al 2008).

2.5.2 Pathology

Gross lesions

There are no gross lesions with any strain of BSE.

Microscopic lesions

In clinical TSE cases, the characteristic histological changes in the CNS are vacuolation of grey matter neuropil (spongiform change), and/or vacuolation of neurons, astrocytosis and neuronal degeneration. In cattle with BSE, these changes are more common in certain neuroanatomical nuclei, particularly within the brainstem, and are bilateral and usually symmetrical. The characteristic lesion profile in cattle is the basis for routine histological screening for BSE. Accumulation of PrP can be demonstrated within these lesions. The *Australian and New Zealand Standard Diagnostic Protocols* for TSEs (SCAHLS 2010) and the OIE Terrestrial Manual contain further details.

In preclinical TSE cases, the characteristic histological changes may be absent.

2.5.3 Differential diagnosis

BSE is a progressive disease of the nervous system and should be considered in the differential diagnosis of locomotory and neurological disorders in cattle over 30 months of age. The following disorders of the nervous and locomotory systems are known to occur in Australia and provide a background guide for the differential diagnosis of BSE:

- trauma
    - brain and spinal cord
- musculoskeletal diseases
- nutritional myopathy (vitamin E or selenium deficiency)
- metabolic diseases
    - hypomagnesaemia or hypocalcaemia
    - nervous acetonaemia
    - polioencephalomalacia
    - hepatic and renal encephalopathy
    - heat stress
- infectious diseases
    - brain or spinal abscess (including cranial or vertebral osteomyelitis)
    - listeriosis
    - thromboembolic meningoencephalomyelitis
    - cerebral babesiosis
    - bovine herpesvirus encephalitis (type 1.3 — BHV1.3)
    - sporadic bovine encephalomyelitis
    - bovine malignant catarrhal fever
    - bovine ephemeral fever
    - focal symmetrical encephalomalacia (*Clostridium perfringens*)
- toxicoses
    - lead toxicosis
    - plant toxicoses
        - perennial ryegrass staggers (*Acremonium lolii*, endophyte on *Lolium perenne*)
        - annual ryegrass staggers, blown grass staggers/floodplain staggers (*Clavibacter toxicus* on seedheads)
        - paspalum staggers (ergotism: *Claviceps paspali* on *Paspalum dilatatum*)
        - phalaris staggers
        - *Swainsona* toxicosis
        - *Xanthorrhoea* toxicity
        - pyrrolizidine alkaloidosis
    - botulism
    - urea toxicosis
    - snakebite
- genetic diseases
    - cerebellar hypoplasia (Shorthorn, Brahman cattle)
    - cerebellar abiotrophy (Angus cattle)

- progressive ataxia (Charolais cattle)
- progressive spinal myelinopathy (Murray Grey cattle)
- neuronal ceroid-lipofuscinosis (Devon cattle)
- tomaculous-like neuropathy (Santa Gertrudis cattle)

- neoplasia.

BSE should also be differentiated from other diseases exotic to Australia, including rabies.

2.5.4 Laboratory tests

Samples required

The range of samples and the methods of sample collection, preservation and submission are described in the *National Guidelines for Field Operations*[13], first published in 2000 and updated in 2008. The preferred specimen is the whole brain with the brainstem intact, removed from the skull immediately after the animal is killed by intravenous barbiturate injection. A 3–10 g sample (1–2 cm) of unfixed cervical spinal cord and/or medulla from the back of the head (obex) should be collected and stored frozen, preferably at –80 °C. This specimen is suitable for detection of PrP^{Sc} by western blotting and rapid immunodiagnostic methods (see Table 2.2). After appropriate microbiological sampling, the brain should be fixed, without longitudinal sectioning or distortion, in 10% neutral buffered formalin for histological and possible immunohistological examination.

If mechanical injury to the brain has occurred — for example, following euthanasia by captive bolt, an attempt should still be made to submit samples as described above, as it may be possible to salvage diagnostically useful material from less than ideal specimens. However, in the case of strong clinical suspicion of BSE, every effort should be made to collect undamaged brain and cord samples. The National Transmissible Spongiform Encephalopathies Surveillance Program (NTSESP) Training Guide shows how to remove the appropriate specimens (see Section 1.3 for details).

Anticoagulated blood samples (lithium heparin) and fresh and fixed tissues should be collected and stored for genetic predisposition studies and parentage typing, which may be required for legal or epidemiological reasons at a later stage.

Transport of specimens

Specimens should be submitted in accordance with agreed state or territory protocols. Specimens should initially be forwarded to the state or territory laboratory for appropriate analysis, and assessment of whether further analysis will be required by the CSIRO Australian Centre for Disease Preparedness (CSIRO-ACDP), Geelong.

If the state or territory laboratory deems it necessary, duplicate samples of the specimens should be forwarded to CSIRO-ACDP for emergency disease testing, after the necessary clearance has been obtained from the chief veterinary officer (CVO) of the state or territory of the suspect case, and after the CVOs of Victoria and Australia have been informed about the case and the transport of the specimens to Geelong (for the first case). Sample packaging and consignment for delivery to CSIRO-ACDP should be coordinated by the relevant state or territory laboratory.

For further information, see the **AUSVETPLAN management manual *Laboratory preparedness***.

[13] www.animalhealthaustralia.com.au/wp-content/uploads/NTSESP-Field-Guidelines-2017-18_final.pdf

2.5.5 Laboratory diagnosis

Laboratory examination of brain is necessary to confirm a diagnosis of BSE. Test methods are discussed in greater detail in the *Australian and New Zealand Standard Diagnostic Procedure for Transmissible Spongiform Encephalopathies* (SCAHLS 2010).

Histological examination to detect the characteristic changes in the CNS mentioned in Section 1.4.2 is the first step because it may also provide an alternative diagnosis and thus conclude an investigation. Appropriately targeted histopathological examination of the brain in clinically affected animals can detect characteristic lesions with high sensitivity and specificity (Wells et al 1989, OIE 2011).

CSIRO-ACDP tests

Tests for detecting accumulated PrP^{Sc} in CNS tissue provide a more definitive diagnosis of BSE. Three sets of methods are available:

- Immunohistochemistry on formalin-fixed sections of CNS.
    - This uses specific antibodies to detect accumulated PrP^{Sc} in situ, and has similar sensitivity to immunochemical methods.
- Immunochemical detection of PrP^{Sc} in homogenates of unfixed CNS tissue.
    - Various tests are available. Western blotting, also known as immunoblotting, is available in Australia. Tissue homogenates are processed through a variety of digestion and concentration steps before specific antibody is used to detect PrP^{Sc}. Western blotting is based on electrophoresis and has the capacity to distinguish the molecular weight and the pattern of glycosylation of PrP^{Sc}. A number of enzyme-linked immunosorbent assay (ELISA) and rapid western blot techniques are also available and are generally used as screening tests in surveillance programs.
- Detection of scrapie-associated fibrils (SAFs) by electron microscopy.
    - This assay detects disease-specific ultrastructural elements by negative staining electron microscopy. SAF detection is less sensitive than immunodetection, but can be used on autolysed tissue (OIE 2011).

Confirmation of a clinical diagnosis of BSE in cattle is based on recognition of distinctive histopathological changes in the CNS, with confirmation by immunohistochemistry on the fixed tissues, by immunochemistry (western blot, ELISA) on unfixed CNS tissue, or by detection of SAFs. There are no serological assays for BSE, as no specific immune response is recognised as part of the disease process. Table 2.2 shows the tests for BSE that are currently used for diagnosis in Australia.

Table 2.2 Laboratory tests currently available at CSIRO–ACDP for the diagnosis of BSE

Test	Specimen required	Test detects	Time taken to obtain result
Histopathology	Formalin-fixed brain	Characteristic lesions	2 days
Immunohisto-chemistry	Formalin-fixed brain	Accumulation of PrP^{Sc}	3 days
Western blot	Unfixed brain tissue or cervical spinal cord	PrP^{Sc}	1 day

ELISA	Unfixed brain tissue or cervical spinal cord	PrP^{Sc}	1 day
Electron microscopy	Unfixed brain tissue or cervical spinal cord	Scrapie-associated fibrils (SAFs)	2 days

ELISA = enzyme-linked immunosorbent assay

Source: Information provided by the then CSIRO-AAHL, 2011 (refer to CSIRO-ACDP for most up-to-date information).

Other tests

Other tests are also available but are not in routine use and are essentially research tools. These include bioassays, most often mouse transmission tests, which involve intracerebral inoculation and take a year or more to complete. They identify patterns of distribution of brain lesions that are distinctive for different prion strains. However, the long incubation period precludes the routine use of this type of assay (SCAHLS 2010). Serial protein misfolding cyclic amplification (sPMCA) is a test under development that has potential for screening tissues and secretions, including milk and urine (Maddison et al 2009), as well as environmental samples (Nichols et al 2009).

2.6 Resistance and immunity

Innate and passive immunity

There is no evidence for passive immunity playing any part in resistance to TSEs. In both scrapie in sheep (Hunter et al 1997) and vCJD in humans (Brown et al 2001), susceptibility or resistance to disease is associated with polymorphisms within the PrP gene. In cattle, some genetic risk factors affecting susceptibility to BSE have been identified (Murdoch et al 2010).

Active immunity

The disease is fatal in all cases, and no protective immunological response has been detected.

2.7 Vaccination

There is no vaccine for any TSE.

2.8 Treatment of infected animals

There is no treatment for an animal with a TSE.

3 Implications for Australia

3.1 Potential pathways of introduction

Key factors in the epidemiology of BSE are well established. They point to three pathways for the introduction of BSE into Australia:

1. **Importation of cattle from BSE-affected countries.**
   Importation of cattle into Australia from the UK ceased in 1988, and importation from continental Europe ceased in 1991. Live cattle cannot now be imported into Australia from any BSE-affected country. The small number of cattle still alive that had been imported from Europe, Japan, Canada (1996 onwards) and the United States (1996 onwards) have been permanently identified under the National Livestock Identification System. They are also in official 'lifetime quarantine' and will never enter the human food, or animal feed, chains. Risk assessments have shown that there is a negligible risk that BSE has been introduced into Australia by importation of these cattle.

2. **Importation of contaminated feedstuff originating from BSE-affected countries.**
   Importation into Australia of animal-derived MBM (except for fishmeal) from all countries except New Zealand was banned in 1966 as a measure against the importation of anthrax spores. Risk-based import controls minimise the chance that other imported stockfeeds or stockfeed ingredients have been contaminated with MBM. Risk assessments have shown that there is a negligible risk of introduction of BSE into Australia by importation of these commodities.

3. **Importation of biologicals contaminated with the BSE agent.**
   Vaccines and other biologicals that involve bovine products in their manufacture have been subjected to quarantine risk assessments. Restrictions on the importation of these products have been extended, in line with emerging knowledge of the BSE status of countries throughout the world. The risk of introduction of BSE into Australia in such products is considered to be negligible.

Stringent controls are in place to prevent the introduction of BSE through these three pathways. In the unlikely event that the BSE agent is introduced, the legislated bans in Australia on feeding ruminant animals MBM derived from mammals, birds or fish (ie restricted animal material) would prevent BSE being propagated and amplified. It has been illegal to feed ruminant MBM to ruminants in all Australian states and territories since 1997. The ban is enforced by state and territory authorities, with support from quality assurance programs in the farming, feedlot, rendering and stockfeed manufacturing industries.

3.2 Social, economic and environmental effects

The economic effects of a temporary loss of market access as a result of a case of BSE in cattle in Australia have been modelled for three hypothetical scenarios, involving a midrange, low-end and high-end reduction in exports, with the high-end scenario also including a reduction in domestic consumption (Yainshet et al 2006). The study found that:

A case of BSE in Australia is likely to impose significant costs, not just to the beef industry but across the broader economy. The results indicate that these impacts may be greater than that observed in countries such as the United States, Canada and Japan that have already experienced isolated cases of BSE. This reflects the highly export oriented nature of the Australian beef industry and the concentration of beef exports in a few key markets that are highly sensitive to BSE. The three scenarios highlight the importance of quickly regaining export markets, with costs escalating rapidly as the closure period lengthens. The high end scenario also highlights the importance of managing consumer reactions in Australia to limit the impact a BSE case could have on the domestic market for beef.

3.3 Critical factors for an Australian response

Features of the disease

- Classical bovine spongiform encephalopathy (BSE) is primarily a disease of domestic cattle (genus *Bos*) but also affects other bovine animals, including buffalo (genus *Bubalus*).
- BSE has an insidious onset and a slowly progressive clinical course.
- BSE results from the ingestion, primarily by young animals, of feed containing meat-and-bone meal (MBM) contaminated with the BSE agent.
- BSE arose in the United Kingdom and was propagated through the recycling of bovine tissues into animal feed. Later, the export of infected cattle and contaminated feed spread the BSE agent to other countries, where it was again recycled and propagated through the feed chain.
- The ID_{50} of BSE brain material that causes clinical disease in cattle is less than 1 g, and the average incubation period is 5 years. The distribution of tissue infectivity in BSE cases is well known. Very small oral doses result in low attack rates of clinical disease.
- The agent for the three known strains of BSE, like all transmissible spongiform encephalopathy agents, is extremely resistant to the usual physical and chemical methods of disinfection. However, it is not absolutely resistant, and appropriate methods may be available for decontamination.
- There is no validated diagnostic test for the BSE agent in live animals.
- BSE is not a contagious disease in the usual sense, and there is no evidence of horizontal or vertical transmission of BSE between cattle. Bovine embryos and semen, dairy products, beef and some other bovine products do not appear to transmit BSE.
- BSE can be spread iatrogenically.
- Atypical BSE (L-type and H-type BSE) is an extremely rare disease of cattle over 8 years of age. It may have a sporadic aetiology and theoretically could rarely arise in Australian cattle. The tissue distribution of infectivity of these agents outside the central nervous system is not known. They are highly unlikely to spread horizontally or vertically, and their ability to infect cattle through feeding of MBM is not yet known.
- A variant form of Creutzfeldt–Jakob disease (vCJD) in humans is caused by the consumption of foods containing specified risk materials (such as brain and spinal cord) from BSE-affected cattle.

Features of susceptible populations

- Australia does not import live cattle from BSE-affected countries, nor animal-derived MBM (except for fishmeal) from any country except New Zealand.

- Australia suspended the importation of cattle from the United Kingdom in 1988, from other European countries in 1991, and from other BSE-affected countries from the date the disease was first reported.
- Cattle imported from countries that have subsequently reported BSE cases were traced, and those still alive at the time were placed under official, permanent quarantine. Measures in place allow the normal commercial management of these animals, but prohibit their use for the production of human or animal food.
- There is a negligible risk that Australian cattle have been, or will be, infected with the BSE agent.
- In 1997, Australia banned feeding of ruminant MBM to ruminants.
- In 2008–09, Australia had approximately 27 million cattle and buffalo; of these, about 2.5 million were dairy cattle and 700 000 were in feedlots. Around 7 million animals are estimated to have been less than 1 year of age. No definitive data are available on the number of cattle that are over 8 years of age (the theoretical risk group for atypical BSE) at a particular time.
- Fear of repercussions may deter producers from reporting disease.
- The expected severe market disruption associated with an outbreak will reduce the value of all related industries.

Options for control or eradication

Based on an assessment of the above factors, managing an emergency response to a finding of BSE in Australian cattle may require the use of some or all of the following strategies:

- registration of all commercial and noncommercial livestock holdings; compulsory biosecurity programs; awareness and rapid resolution of ownership of animals and animal products on farms
- epidemiological investigations of the case(s) and cohorts
- swift declaration and effective policing of movement controls on live cattle and certain animal products
- modified stamping out
- increased targeted active surveillance
- strengthening of current BSE risk reduction measures
- close liaison with affected industries and public health agencies
- a public awareness campaign to improve early detection, support consumer confidence in beef, and maintain access to export markets for Australian cattle and their products
- individual animal identification; and tracing of animals, and products and byproducts from the case(s) and cohorts.

The policy options for management of a case of any strain of BSE in Australian cattle, based on consultation and cooperation between government and the livestock industries, are:

- investigation and communication of findings only
- containment, with a view to eventual eradication
- eradication.

The policy to be implemented is described in Section 4.

4 Policy and rationale

4.1 Introduction

4.1.1 Summary of policy

Bovine spongiform encephalopathy (BSE) is a World Organisation for Animal Health (OIE)-listed disease that is significant in the international trade of cattle and cattle products. Classical BSE is also a foodborne zoonosis. A confirmed case of any strain of BSE in Australia could result in serious economic loss within the livestock industries, due to loss of export markets and disruption to business continuity from falls in domestic consumption of beef.

The default policy is to eradicate any strain of BSE as quickly as possible using *modified stamping out*, supported by a combination of strategies including:

- *timely recognition* and laboratory confirmation of a case(s)
- *initial quarantine* of infected and dangerous contact premises
- *quarantine and movement controls* over animals and animal products
- *improved risk reduction measures*, such as revisions to the ruminant feeding ban
- *tracing and increased surveillance* (based on epidemiological assessment) to identify cohort cattle and the source and extent of infection, and subsequently to establish proof of freedom from the disease
- *zoning/compartmentalisation* (if applicable) to define infected and disease-free premises and industry sectors
- *disposal* of confirmed case(s)
- *destruction and disposal* of all cohort cattle, depending on the findings of veterinary investigations
- *recall* of animal products likely to be contaminated
- a *public awareness campaign* that describes measures taken to protect human and animal health.

4.1.2 Case definition

Table 4.1. Case definitions for BSE

Case type	Definition
Suspect BSE case	An animal of the genus *Bos* (cattle) or *Bubalus* (buffalo) with history, clinical signs and histological changes consistent with BSE (as described in Section 1.4), until BSE is confirmed or excluded OR An animal with a positive result from a sensitive and specific screening test such as an ELISA for TSEs (see Section 1.4), until BSE is confirmed or excluded
Confirmed BSE case	A suspect case with positive results from a TSE-specific immunohistochemistry, immunochemistry or other validated confirmatory test

Case type	Definition
Cohorts to a BSE case[a]	Cattle which, during their first year of life, were reared with the BSE cases during their first year of life, and which investigation showed consumed the same potentially contaminated feed during that period OR If the results of the investigation are inconclusive, all cattle born in the same herd as, and within 12 months of the birth of, the BSE cases

BSE = bovine spongiform encephalopathy; ELISA = enzyme-linked immunosorbent assay;
TSE = transmissible spongiform encephalopathy
a OIE *Terrestrial Animal Health Code*[14]

In this response strategy, references to cattle also apply to buffalo.

This response strategy does not cover the response to BSE agents causing disease in small ruminants, domestic cats or zoo felids. The experience overseas is that these cases only arise as spillover events from a significant BSE epidemic in indigenous cattle.

This response strategy does not cover a BSE outbreak caused by veterinary vaccines or other veterinary therapeutics. Although it is important that BSE is considered in risk assessments for biological products, there is no evidence to suggest that they have been a source of BSE cases overseas.

4.1.3 Cost-sharing arrangement

In Australia, BSE is included as a Category 2 emergency animal disease in the Government and Livestock Industry Cost Sharing Deed in Respect of Emergency Animal Disease Responses (EAD Response Agreement – EADRA).[15] When cost sharing of the eligible response costs of an incident is agreed, Category 2 diseases are those for which costs will be shared 80% by government and 20% by industry.

4.1.4 Criteria for proof of freedom

The OIE categorises countries into three risk levels associated with BSE: negligible, controlled and undetermined[16]. Beef importing countries, including Australia, either adopt the OIE's categorisations or conduct their own using similar criteria. Australia is classified by the OIE as negligible BSE risk. Negligible BSE-risk countries that subsequently report a case of any strain of BSE in indigenous cattle will be reclassified by the OIE as controlled risk, unless all cases were born more than 11 years ago.

The OIE risk classification system requires that the BSE risk status of the cattle population of a country, zone or compartment be determined on the basis of the following criteria:

- the outcome of a risk (release and exposure) assessment that identifies all potential factors for BSE occurrence and their historic perspective
- an ongoing awareness program for veterinarians, farmers and workers involved in transportation, marketing and slaughter of cattle, to encourage reporting of all cases showing clinical signs consistent with BSE

[14] www.oie.int/index.php?id=169&L=0&htmfile=chapitre_bse.htm
[15] Information about the EAD Response Agreement can be found at www.animalhealthaustralia.com.au/what-we-do/emergency-animal-disease/ead-response-agreement.
[16] www.oie.int/index.php?id=169&L=0&htmfile=chapitre_bse.htm

- compulsory notification and investigation of all cattle showing clinical signs consistent with BSE
- examinations of brain or other tissues collected within the framework of a surveillance and monitoring system.

Commodities from the cattle population of a country, zone or compartment may pose a negligible risk or a controlled risk of transmitting the BSE agent, depending on the extent to which the country meets conditions listed by the OIE. The cattle population of a country, zone or compartment is considered to pose an undetermined BSE risk if it cannot be demonstrated that it meets the requirements of another category.

Proof of freedom is difficult because of the long incubation period for BSE and the possible sporadic nature of atypical BSE. The ultimate aim of an emergency response is to verify either that Australian cattle and their products represent a negligible risk of transmitting the BSE agent, or that Australia has controlled this risk through appropriate risk reduction measures. How this aim is achieved will depend on:

- the age of the index case and any subsequent cases
- the strain of BSE
- the putative source of the BSE agent
- the extent of the outbreak
- the results of tracings of cohort cattle and their products.

If a BSE strain is detected in Australian cattle less than 11 years of age, it will be necessary to demonstrate that Australia has minimised the likelihood of BSE risk materials entering the human food, and animal feed, chains. This will require increased auditing of existing arrangements and completion of the risk analyses specified in the BSE chapter in the OIE Terrestrial Code.

Food safety authorities will have a lead role in the protection of the human food supply — for example, removal of SRMs from the human food chain if this is indicated by the risk assessment.

Australia's BSE surveillance program is structured to comply with the OIE requirements, which will allow the detection of BSE around a design prevalence of at least one case per 100 000 adult cattle at a confidence level of 95%.

In the event of an outbreak, the National TSE Surveillance Program — within the TSE Freedom Assurance Program[17] managed by Animal Health Australia — would be used as the basis for an improved surveillance program. This will comprise the examination of native-born cattle over 30 months of age that either display clinical signs consistent with a differential diagnosis that includes BSE, or are in other OIE at-risk subpopulations — such as downer, emergency slaughter and fallen cattle. The age, target subpopulations and number of cattle to be examined will be determined at the time, based on the results of investigation and the need to support domestic and export markets for Australian cattle and their products.

4.1.5 Governance

Governance arrangements for the response to EADs are outlined in the **AUSVETPLAN *Overview***.

Information on the responsibilities of a state coordination centre and local control centre is available in the **AUSVETPLAN management manual *Control centres management* (Parts 1 and 2)**.

[17] www.animalhealthaustralia.com.au/what-we-do/disease-surveillance/tse-freedom-assurance-program/

4.2 Public health implications

As described in Section 2.2, variant Creutzfeldt–Jakob disease (vCJD) was identified in the UK in the mid-1990s and found to be associated with consumption of beef products contaminated with certain tissues from BSE-affected cattle. The zoonotic potential of atypical BSE is not yet fully understood.

An Australian National CJD Registry has been maintained in the Department of Pathology, University of Melbourne, since 1993, and cases of possible CJD are investigated by medical neurologists. No case of vCJD has yet been reported in Australia. However, cases could conceivably occur in the future (eg in people who lived in the UK before BSE-contaminated beef products were removed from the human food chain).

In 2000, Australia's peak public health advisory and medical research body, the NHMRC, established a Special Expert Committee on TSEs, which is now the Transmissible Spongiform Encephalopathies Advisory Committee. The committee's purpose is to provide expert and timely advice to Australian governments on all matters necessary to prevent the occurrence and spread of vCJD and other TSEs in Australia.

Australian health regulatory agencies, such as the Therapeutic Goods Administration and Food Standards Australia New Zealand, have extensively reviewed all products under their control to identify constituents of bovine origin and have taken measures to prevent exposure of the Australian population. Blood-donor deferral procedures, among other safeguards, have been put in place to protect the safety of the Australian blood supply.

The Department of Health has prepared a response plan for vCJD as part of Australia's preparedness to address the potential impacts of the disease, including public health, medico-legal, social, community, political, trade and international relations impacts[18]. The plan is based on a risk management approach for biological emergencies. It recognises that such an event will occur very infrequently, the evidence base for decision making may be limited and evolving, and the community reaction could be disproportionate to the level of physical risk.

There is no risk of human or animal exposure to the BSE agent from live cattle. However, people handling potentially infected material (eg central nervous system tissue from suspected BSE cases) must take adequate precautions to avoid exposure to these agents. Veterinarians, laboratory workers and slaughterhouse workers need to wear appropriate face and eye protection, and gloves when handling tissues suspected of containing high levels of the agent. Care should be taken to minimise environmental contamination during necropsy procedures and to decontaminate.

4.3 Control and eradication policy

This policy will apply once a confirmed case of any strain of BSE is diagnosed in cattle in Australia.

Details of the policy will depend on the type of incident that initiates an emergency response (eg imported or indigenous case; age of animal; strain involved, if indigenous) and the results of the epidemiological investigation. Table 4.2 shows the actions that may be required.

Australia will continue to meet the OIE's conditions for a negligible BSE-risk country if a case is in imported cattle or if the case is in indigenous cattle aged 11 years or older, and the assessed BSE risk of Australia's imports has not changed. The occurrence of BSE in indigenous cattle will indicate a cycle of transmission in Australia. A case of atypical BSE in indigenous cattle will probably be a sporadic

[18] www.health.gov.au/internet/main/publishing.nsf/Content/ohp-vcjd-response-plan

event. The distinction between these situations is important because different response measures will be indicated. An improved surveillance and monitoring program may be required, to an extent and intensity determined by the epidemiological and other veterinary investigations.

4.3.1 Epidemiological assessment

Epidemiological investigation or assessment draws on multiple sources of information to build understanding of the disease and how it is behaving in an outbreak. This helps inform response decision making.

The key objectives for an epidemiological assessment will be to identify:

- the spatial distribution of infected and free animal populations
- potential vectors involved, including as potential amplifying hosts
- the source of infection
- the prevalence of infection
- pathways of spread and the likely size of the outbreak
- risk factors for the presence of infection and susceptibility to disease (including weather and insect populations).

Epidemiological assessment, and tracing and surveillance activities (see Section 4.3.3) in an EAD response are interrelated activities. Early findings from tracing and surveillance will be inputs into the initial epidemiological assessment (eg considering spatial distribution of infection). The outcomes of the initial epidemiological assessment will then guide decisions on subsequent tracing and surveillance priorities.

The outcomes of the epidemiological assessment will also be used initially to determine the feasibility of eradication versus long-term control and to guide the selection of other appropriate response measures (including the application of movement controls) and assess the progress of disease control measures.

Ongoing epidemiological assessment is important for any EAD response to aid evaluation of the continued effectiveness and value of response measures, and assessment of the progress of disease control measures. Ongoing epidemiological assessment will consider the outcomes of tracing and surveillance activities, and will contribute evidence to support any later claims of disease freedom.

4.3.2 Quarantine and movement controls

Quarantine will be imposed on IPs and dangerous contact premises (DCPs). After investigation, it may be possible to reduce quarantine restrictions on cohort cattle or their potentially contaminated products. Further decisions can then be made based on data from veterinary investigations (see Table 4.2).

Declaration of restricted and control areas for BSE will not be required, due to the nature of the disease.

4.3.3 Tracing and surveillance

Tracing will be undertaken to:

- identify, as accurately as possible, the age of the case(s)

- assist in establishing the source of infection
- determine the presence of other potentially infected herds
- find risk materials that might enter the human food, or animal feed, chains.

Once BSE has been confirmed, cohort cattle and their potentially contaminated products will be traced.

If possible, feedstuffs that a confirmed BSE case consumed during its first year of life will also be traced. This will be very difficult, given that the average age of BSE cases is 5 years.

The current manager of the cattle and the manager responsible during the year of birth of the case and cohort cattle will be interviewed. Any private veterinary practitioner who has serviced the premises involved or the property of origin of the confirmed cases should also be interviewed to discuss the range of clinical presentations observed in cattle at those properties during at least the previous 9 years.

Trace-forward of cohort cattle and products will be used to declare DCPs and trace premises (TPs). The cattle will be examined periodically until death to detect any development of characteristic clinical signs. All such cattle will be identified by National Livestock Identification System microchip devices and will be tested for BSE at death, if possible. In response to a BSE case, investigations will attempt to include the tracing of cohort ruminants other than cattle. However, based on overseas experience, measures would usually not be required for these animals.

Trace-back for BSE cases will attempt to determine, as accurately as possible, the age of the case(s) and to locate the possible source of exposure. In the case of BSE in an imported animal, although exposure will almost certainly have occurred overseas, the possibility that exposure was via feed in Australia still needs to be assessed.

A systematic program of testing of cohort and other risk subpopulations of cattle will be required to determine the extent of BSE cases in Australia, and to help provide proof that Australia's negligible BSE risk status can be retained or that the disease has been controlled. In response to a BSE case(s), the program might need to be maintained for a prolonged period.

Information on specimen collection and diagnosis is given in Section 2.5 and the *NTSESP National Guidelines for Field Operations*.[19]

4.3.4 Zoning and compartmentalisation for international trade

Because of the nature of BSE, zoning has not been appropriate overseas, and the same would probably apply in Australia. However, in exceptional circumstances, certain classes of cattle may be able to be compartmentalised and made exempt from some BSE controls. In the United Kingdom (UK), for example, cattle from specialist beef herds at very low risk of BSE and registered under the UK's beef assurance scheme were allowed to be slaughtered for sale for human consumption up to 42 months of age.

4.3.5 Vaccination

Vaccination is not applicable.

[19] www.animalhealthaustralia.com.au/wp-content/uploads/NTSESP-Field-Guidelines-2017-18_final.pdf

4.3.6 Treatment of infected animals

There is no effective treatment.

4.3.7 Treatment of animal products and byproducts

No treatment for animal products is guaranteed to be effective in inactivating the BSE agent under normal commercial operations. Meat and animal products from confirmed cases of BSE and from cohort cattle will not be rendered for meat-and-bone meal or for other products, but will be disposed of by incineration or another acceptable method.

Depending on the outcome of epidemiological investigations and risk assessment, risk reduction measures could be strengthened — for example, through revisions to ruminant feeding restrictions. Specified risk materials may need to be removed from human food and animal feed, and then disposed of. Cattle tissues and organs recognised as specified risk materials are described in Section 2.4.2.

4.3.8 Destruction of animals

Stamping out

Modified stamping out will be undertaken, as defined in the OIE Terrestrial Code. BSE is a notifiable disease in all Australian states and territories, and suspect cases must be notified to a government veterinarian or animal health officer.

The premises with the index case will be declared an infected premises (IP), and part or all of the premises will be placed under quarantine. Movement controls will be imposed on all cattle on the premises until the full results of epidemiological investigations are known.

Subsequent strategies will depend on the outcome of veterinary investigations to identify the risk status of cohort cattle and relevant materials associated with a confirmed case (see Table 4.2). The investigation will begin with a complete history of feeding practices and identification of all premises where the confirmed case had resided from birth to diagnosis. Subsequently, any cohort cattle, potentially contaminated products from the case or cohort cattle, and potentially contaminated feedstuffs and biological materials will be traced.

In response to a BSE case, investigations will include assessment of the risk posed by cohort ruminants other than cattle. However, based on overseas experience, measures would be required for these animals only in exceptional circumstances.

Table 4.2. Potential actions required for categories of infected or potentially infected cattle

Animal category	**Actions**	
	Animals	**Other measures**
Imported cattle		
• Confirmed case	Review animal identification to confirm that the case was imported Dispose of carcase appropriately	Quarantine IPs, DCPs and TPs until epidemiological studies and identification of cohort cattle have been completed

Animal category	Actions	
	Animals	**Other measures**
• Cohort cattle residing in Australia	Assess potential for feedborne exposure in Australia Trace cohort cattle to establish their fate and, if they are alive, place them in lifetime quarantine (if not already in lifetime quarantine)	Trace and isolate potentially contaminated products (edible and inedible) derived from the case and cohort cattle if they were not in lifetime quarantine Advise source country Introduce, in conjunction with health authorities, a public awareness program that describes measures taken to protect human and animal health
Indigenous cattle		
• Confirmed case • Cohort cattle	Review animal identification to confirm that the case is of Australian origin Trace the case to property of birth and other properties where it has resided Depending on epidemiology findings and risk assessment: • kill and test cattle on declared premises, and dispose of carcases appropriately • quarantine premises or • take no action (decision based on assessment of potential exposure to the BSE agent)	Review case diagnosis, strain of BSE, confirmatory testing, parallel testing at CSIRO-AAHL and the world reference laboratory (Veterinary Laboratory Agency, Weybridge) Quarantine IPs, DCPs and TPs until epidemiological studies and identification of cohort cattle have been completed Trace and isolate potentially contaminated products (edible and inedible) derived from case and cohort cattle Introduce, in conjunction with health authorities, a public awareness program that describes measures taken to protect human and animal health
• Other indigenous cattle		Depending on epidemiology findings and risk assessment, consider implementing: • an increased BSE surveillance program • a protocol for removal and disposal of SRMs from human food and animal feed

BSE = bovine spongiform encephalopathy; CSIRO-AAHL = Australian Animal Health Laboratory of the Commonwealth Scientific and Industrial Research Organisation; DCP = dangerous contact premises; IP = infected premises; SRM = specified risk material; TP = trace premises

4.3.9 Disposal of animals, and animal products and byproducts

The carcasses of all animals that are killed to eradicate BSE will be completely destroyed in accordance with procedures described below.

Destruction and testing of cattle on farm is preferred to transporting them for slaughter at another site. Killing on farm reduces the risk of spread of the BSE agent from a knackery or abattoir, and focuses control measures in one place.

Care is required in collecting postmortem samples of brain (which is required for diagnosis), using methods described in the *NTSESP National Guidelines for Field Operations*.

The following points are relevant to carcass disposal (see the **AUSVETPLAN operational manual *Disposal*** for detailed information):

- Wherever possible, carcasses will be burned or incinerated.
- Burning or incineration of carcasses will be supervised by disease control authorities to ensure that appropriate methods are used and that all contaminated material is completely burned.
- The ash will be collected, mixed with agricultural lime to create alkaline conditions, and buried deeply at a suitable site.
- Where burning is not practical, carcasses and other materials that cannot be adequately decontaminated will be mixed with caustic materials that will create an alkaline environment and buried deeply at a suitable site.
- Consideration will be given to the future use of the burial site, and to any associated water sources, as the agent may remain in a transmissible state in the soil for long periods (however, environmental sources have not been definitively implicated in BSE transmission).
- Dogs, cats and other potential scavengers should be kept away from destruction and disposal sites.
- Other methods of destruction of carcasses and contaminated material, as specified by the OIE for transmissible spongiform encephalopathies (TSEs), can be considered (ie alkaline hydrolysis).
- The location of burial sites for carcasses, products or ash will be recorded and marked.

Rendering will not be used to dispose of confirmed cases or cohort cattle, because the temperatures and pressures currently used would not be high enough to guarantee complete inactivation of the disease agent.

4.3.10 Decontamination

Areas, fixtures and fittings that might have been contaminated with the tissues from confirmed cases during a postmortem examination will be decontaminated. The decontamination measures used will be in proportion to risk. Decontamination may be required for premises with the potential for heavy contamination, such as field necropsy sites and laboratory postmortem rooms, but decontamination of a property with confirmed or suspected cases is not necessary except as outlined above.

The **AUSVETPLAN operational manual *Decontamination*** contains general information on decontamination procedures. Because many of the standard methods of decontamination cannot ensure complete inactivation of the BSE agent, the emphasis will be on removal of the agent by thorough cleaning, followed by an appropriate steam sterilisation or liquid chemical treatment, as described below.

Most common disinfectants, including ethanol, formalin, hydrogen peroxide, iodophors and phenolics, and gases such as ethylene oxide and formaldehyde, are *not* effective against the agent. One of the following methods of chemical decontamination for TSE agents will be used:

- Sodium hypochlorite solution containing 2% (20 000 ppm) available chlorine for more than 1 hour at 20 °C. For the BSE agent, the OIE Terrestrial Manual recommends overnight chemical disinfection of equipment.
- 2 M (80 g/L) sodium hydroxide for more than 1 hour at 20 °C. This method is not completely effective unless the alkali-to-tissue ratio is high enough.
- For histological samples only, 96% formic acid for 1 hour. However, formalin fixation of infected tissues stabilises the scrapie agent so that it cannot then be inactivated by steam sterilisation. Residues of formalin-fixed tissues should therefore be disposed of by incineration.

The risk of horizontal transmission of BSE through environmental contamination with infected tissues is theoretical only and is not supported by overseas experience with the disease. However, entry of ruminants to necropsy sites on IPs will be prevented until decontamination is complete. It is not necessary to impose ongoing farm-gate disinfection at IPs.

Instruments used for postmortem removal of brain or other potentially infected tissue from suspect cases, confirmed cases and cohort cattle will either be discarded after a single use, or decontaminated using one of the methods described above before they are reused on live ruminants. If BSE is confirmed in indigenous cattle, equivalent controls on instruments used on cattle that are not considered at risk (eg for eye ablation or routine postmortem) are not warranted.

4.3.11 Wild animal management

Carcasses will be disposed of in such a way that ingestion by wild and stray animals, including dogs, pigs, cattle and sheep, is prevented.

See the **AUSVETPLAN operational manual *Wild Animal Response Strategy*** for further information.

4.3.12 Vector management

Vector control is not applicable.

4.3.13 Public awareness and media

One of the most important elements of a public health response will be the communication strategy. Unsubstantiated reports of BSE could have serious ramifications for the livestock industry, its communities, the Australian economy and international relations. The public, especially those in the livestock industries, need to be provided with accurate information to support domestic beef consumption after any strain of BSE is confirmed. There should be clear coordination of information among the relevant organisations, including human health authorities, industry organisations, food safety authorities and the Transmissible Spongiform Encephalopathies Advisory Committee of the National Health and Medical Research Council (NHMRC). Communications with countries that import Australian cattle and their products will be critical to maintaining or regaining market access.

Information provided to the public after confirmation of a case of BSE should cover:

- the circumstances of the outbreak, and exactly what is known and not known
- facts about the disease (including fact sheets)
- the planned response to the outbreak, with regular updates
- issues related to the consumption of meat, with a clear explanation of how the food chain is being protected
- arrangements to prevent spread of the disease, such as the pre-existing bans on feeding vertebrate protein to ruminants and longstanding restrictions on imports from countries with BSE
- trade implications
- comparison with the UK epidemic and the situation in other countries.

See the **Public Relations Manual** for further information on provision of public information about emergency animal diseases.

4.4 Other control and eradication options

Apart from eradication (the policy described above), other policy options are:

- investigation and communication of findings only — for example, if a sporadic atypical case were found in an older animal
- containment, with a view to eventual eradication — for example, if spread of the disease were to occur due to iatrogenic transmission through a contaminated biological product, a program with a high level of industry cooperation would be required to achieve eradication. The eradication program would comprise
    - extensive surveillance using rapid diagnostic tests on nervous tissue obtained postmortem at abattoirs (supported by confirmatory testing of positives)
    - trace-back and other veterinary investigations
    - interim quarantine, where required
    - eradication programs for identified infected herds, as determined by veterinary investigations.

4.5 Funding and compensation

Details of the cost-sharing arrangements can be found in the Government and Livestock Industry Cost Sharing Deed in Respect of Emergency Animal Disease Responses.[20] Details of the approach to the valuation of, and compensation for, livestock and property in disease responses can be found in the **AUSVETPLAN operational manual *Valuation and compensation***.

[20] www.animalhealthaustralia.com.au/what-we-do/emergency-animal-disease/ead-response-agreement

5 Declared areas and premises

When an emergency animal disease (EAD) is first suspected, the premises involved would undergo a clinical and/or epidemiological investigation. If the case definition, as defined in the relevant AUSVETPLAN response strategy, is met (ie the index case[21]), the relevant chief veterinary officer (CVO) or their delegate will determine the premises classification and may declare the premises an infected premises (IP).

After the identification of the first IP, a restricted area (RA) and a control area (CA) may be declared.[22] A transmission area (TA) may also be defined, if appropriate. All premises within these areas will be classified.

At the beginning of an EAD incident, the initial premises classifications would be IP, at-risk premises (ARP), premises of relevance (POR), unknown status premises (UP) and zero susceptible species premises (ZP).

Any premises within the RA or CA will have only one classification at any one time. After an epidemiological investigation, clinical assessment, risk assessment or completion of control measures, a premises may be reclassified.

Once the first IP has been identified, intelligence gathering through veterinary epidemiological investigations would quickly lead to the identification of suspect premises (SPs) and trace premises (TPs). These would be high priorities for follow-up investigation by the relevant state or territory authorities. In a worst-case scenario, an SP could become an IP; therefore, SPs need to be investigated as a matter of very high priority. Similarly, investigation and risk assessment of a TP might identify it as an IP, dangerous contact premises (DCP) or dangerous contact processing facility (DCPF). An SP or TP might also be assessed as negative and qualified as SP-AN or TP-AN, and eventually reclassified as an ARP, POR or ZP.

All premises classifications are subject to change as a result of a modification in the case definition(s) or investigation(s) as the incident response proceeds.

Classifications should be applied with information needs of managers in mind. They should assist managers to monitor and report progress. Premises classifications to be used should be agreed early in a response, so that control centre personnel can apply the correct and consistent classifications and definitions from the outset of the investigation and response.

5.1 Declared areas

Declared areas are not applicable to BSE in Australia.

5.1.1 Restricted area (RA)

RAs are not applicable to BSE in Australia.

[21] The first case to come to the attention of investigators

[22] This is invariably the case with highly contagious diseases (eg foot-and-mouth disease, equine/avian/swine influenza, classical swine fever) but may not apply to less contagious diseases (eg Hendra virus, anthrax, Australian bat lyssavirus).

5.1.2 Control area (CA)

CAs are not applicable to BSE in Australia. In exceptional circumstances, compartments may be created to address different risks presented by different sectors of the cattle industry.

5.2 Other areas

It is possible that other types of areas (eg vaccination area or surveillance area), which are not legally declared, may be used for disease control purposes in some jurisdictions.

5.3 Premises classifications

Detailed guidelines for classifying premises statuses are provided in the **AUSVETPLAN guidance document *Declared areas and application of premises classifications in an EAD response***, and the definitions are in the Glossary.

5.3.1 Premises status classifications

For bovine spongiform encephalopathy (BSE), the premises classifications to be used are:

- infected premises (IP)
- suspect premises (SP)
- trace premises (TP)
- dangerous contact premises (DCP)
- dangerous contact processing facility (DCPF)
- approved processing facility (APF)
- approved disposal site (ADS)
- at-risk premises (ARP)
- premises of relevance (POR)
- resolved premises (RP)
- unknown status premises (UP)
- zero susceptible species premises (ZP).

5.3.2 Qualifiers

Please also refer to the **AUSVETPLAN guidance document *Declared areas and premises classifications*** for more detail on qualifiers.

For bovine spongiform encephalopathy (BSE), the qualifier to be used is:

- assessed negative (AN).

6 Movement controls

6.1 Principles

The principles for the recommended quarantine practices and movement controls are as follows:

- Containment and eradication of bovine spongiform encephalopathy (BSE) is the highest priority. Therefore, 'normal business movements' are not allowed.
- Live animals pose the greatest risk of disease spread; therefore, their movements from all premises within the restricted area (RA) and control area (CA) must be strictly controlled.
- The outside area (OA) should remain as 'clean' as possible. Therefore, movement of animals from the RA to the OA is prohibited, and movement of products is generally prohibited. Movement of animals and products from the CA to the OA will also be restricted.
- Trace premises (TP) and suspect premises (SP) are temporary classifications, and every effort should be made to resolve the status of these premises as soon as possible.
- The numbers of susceptible animals within the RA should be minimised. Therefore, movements of animals into the RA will be limited and usually for slaughter only.
- Movement restrictions are more stringent within the RA than within the CA, and will be more stringent in the early stages of the response.
- Movement controls may be varied during a response from those listed here. However, this will involve a variation to the agreed Emergency Animal Disease Response Plan, with endorsement by the Consultative Committee on Emergency Animal Diseases (CCEAD) and the National Management Group (NMG).
- Recommended movement controls apply to any movement off a premises, whether on foot or by vehicle, that involves either public or private land.
- All movement control matrixes and narratives are for guidance.
- Application for a movement permit does not automatically mean that one will be granted.
- In emergency or exceptional circumstances, any proposed movement may be considered by the jurisdictional chief veterinary officer (CVO) on a risk-assessed case-by-case basis.
- Interstate movements will need to meet the import requirements of the receiving jurisdiction.

6.2 Guidelines for issuing permits

In an emergency animal disease (EAD) event, quarantine and movement controls must strike a balance between quick and effective disease control and business continuity. Therefore, it is not appropriate to simply prohibit all movement of animals and products. On the other hand, diligence needs to be applied to minimise the risk of further spread of the disease.

Recommended biosecurity and movement controls in each AUSVETPLAN response strategy provide guidance on which movements can be allowed and under what conditions. This is based on an analysis of the disease risks that are presented by a specific movement, of a specific commodity, at a specific time during the EAD response phase. Each disease strategy will indicate whether a proposed movement is:

- allowed (under normal jurisdictional, including interstate, requirements)
- prohibited – except under the conditions of a general, special or emergency permit
- prohibited.

Permits may not be available until the relevant CVO provides approval for movements, and this may not be available in the early stages of a response. When assessing risk for the purposes of issuing a permit, the elements to consider may include:

- sources of risk
    - risk material such as live or dead susceptible animals, semen, embryos, meat, meat products, waster products, offal, paunch screenings, manure, render material, fertiliser, biological specimens, casings, used wrappers and cartons, effluent, fomites (vehicle, people, nonsusceptible animals, crops, grains, hay silage and mixed feeds)
    - presence of disease agent on both the originating and destination premises, and uncertainty
    - location of source and destination premises
    - fate at destination premises (eg for slaughter vs for growing out)
    - current vector activity, if relevant
    - organisation and management issues (ie confidence in animal tracing and surveillance, biosecurity)
    - proposed use of the animals or products
    - proposed transport route
    - vaccination status of the animals, if relevant
    - treatment of animals and vehicles to prevent concurrent movement of vectors, if relevant
    - security of transport
    - security and monitoring at the destination
    - environment and natural events
    - community and human behaviour
    - risk of sabotage
    - technology
    - regulations and standards
    - available resources for compliance and enforcement
- areas of impact
    - livestock health (health of affected species, including animal welfare)
    - human health (including work health and safety)
    - trade and economic impacts (including commercial and legal impacts)
    - environmental impacts
    - organisational capacity
    - political impacts
    - reputation and image
- proposed risk treatment measures
    - vaccination
    - destruction of animals
    - processing of product
    - disinfection or other treatment of animals, vehicles and fomites
    - vector control, if relevant
    - security
    - communication.

6.3 Types of permits

Permits are either general or special. Emergency permits are a form of special permit. Permits are legal documents that describe the animal(s), commodities or things to be moved, the origin and destination, and the conditions to be met for the movement. Either type of permit may include conditions. Once permit conditions have been agreed from an operational perspective, all permit conditions must be met for every permit. Both general and special permits may be in addition to documents required for routine movements between or within jurisdictions (eg health certificates, waybills, consignment notes, National Vendor Declarations – NVDs).

General permit

General permits (GPs) are used for lower-risk movements, and create a record of each movement to which they apply. They are granted without the need for direct interaction between the person moving the animal(s), commodity or thing and a government veterinarian or gazetted inspector of stock. The permit may be completed via a webpage or in an approved place (such as a government office or commercial premises). A printed version of the permit must accompany the movement. The permit may impose preconditions and/or restrictions on movements. GPs may not be available until the relevant CVO gives approval for general movements, and this may not be available in the early stages of a response.

Special permit

Special permits (SpPs) are issued by the relevant government veterinarian or gazetted inspector of stock. They are used for higher-risk movements, and therefore require formal application and individual risk assessment. SpPs describe the requirements for movement of an animal (or group of animals), commodity or thing, for which a specific assessment has been conducted by the relevant government veterinarian or gazetted inspector of stock. A printed version of the permit must accompany the movement. The permit may impose preconditions and/or restrictions on movements.

Emergency permit

An emergency permit is an SpP that specifies strict legal requirements for an otherwise high-risk movement of an animal, to enable emergency veterinary treatment to be delivered, to enable animals to be moved for animal welfare reasons, or to enable any other emergency movement under exceptional circumstances. These permits are issued on a case-by-case basis under the authorisation of the relevant CVO.

Other movement requests

Movements not reflected within any of the movement control matrixes or narratives may be considered by the relevant jurisdictional CVO on a risk-assessed case-by-case basis.

6.4 Recommended movement controls

Table 6.1 shows the recommended movement controls that will apply to IPs, DCPs and TPs in the initial stages of a BSE incident. Subsequently, movement restrictions may be amended to apply, for example, to only part of a premises or to cohort cattle only until they can be destroyed and tested.

Table 6.1. Recommended movement controls for declared premises

To **From**	**Nondeclared premises**					**IP/DCP/TP**
	Cattle/ buffalo	Other ruminants	Other animals	Specified products[a]	Equipment	Cattle/ buffalo
IP	Prohibited except under SpP1	Prohibited except under SpP2	No restrictions	Prohibited except under SpP1	No restrictions	Prohibited except under SpP2
DCP	Prohibited except under SpP1	Prohibited except under SpP2	No restrictions	Prohibited except under SpP1	No restrictions	Prohibited except under SpP2
TP	Prohibited except under SpP1	Prohibited except under SpP2	No restrictions	Prohibited except under SpP1	No restrictions	Prohibited except under SpP2
Nondeclared premises	No restrictions	No restrictions	No restrictions	No restrictions	No restrictions	Prohibited except under GP1

a For example, cattle carcases, ruminant meat and bone meal
DCP = dangerous contact premises; IP = infected premises; TP = trace premises
Notes:
SpP1 (specific permit 1): for destruction and disposal only
SpP2 (specific permit 2): for animal identification
GP1 (general permit 1): decontamination of the IP/DCP/TP is required before introduction of animals

Appendix 1

BOVINE SPONGIFORM ENCEPHALOPATHY FACT SHEET

Disease and cause

Three known strains of bovine spongiform encephalopathy (BSE) have been identified in cattle: classical BSE and two strains of 'atypical' BSE. As the BSE agent causes a similar disease in humans, BSE is important not only for the welfare of cattle, but also as a food safety issue.

BSE is one of the transmissible spongiform encephalopathies (TSEs) or 'prion' diseases, and causes progressive neurodegenerative disease. TSEs are characterised by long incubation periods and the accumulation in the central nervous system of an abnormal form of a prion protein.

Distribution

BSE was first recognised in the United Kingdom (UK) in 1986 and became a serious epidemic in that country. Atypical BSE is a very rare disease in older cattle that has been recognised for less than 10 years; the origin of atypical BSE is not yet known, but a spontaneous, noncontagious origin cannot be excluded.

BSE aetiology involves feeding cattle (particularly young cattle) meat-and-bone meal (MBM) contaminated with the BSE agent. All BSE cases in countries other than the UK have origins in the importation and feeding of MBM to young cattle, or the importation from the UK of live cattle that entered the animal feed chain.

The assessment of the World Organisation for Animal Health (OIE) is that there is a negligible risk that BSE is present in Australia or that it has been introduced to cattle in Australia through the importation of commodities potentially contaminated with the BSE agent.

Species affected

BSE is primarily a disease of domestic cattle (genus *Bos*), but also affects other bovine animals, including buffalo (genus *Bubalus*).

Creutzfeldt–Jakob disease (CJD) is a TSE that affects humans. In March 1996, the UK reported 10 cases of a new clinicopathological variant of CJD (variant CJD or vCJD). Primary cases of vCJD are caused by the consumption of foods containing specified risk materials (such as brain and spinal cord) from BSE-affected cattle.

Clinical signs

Due to the long incubation period of BSE after exposure of calves, signs usually appear when cattle are between 3 and 7 years of age. BSE usually has an insidious onset and a slowly progressive clinical course, extending over weeks to months. Apprehension, hyperaesthesia and ataxia are the main signs, and at least one of these signs is present in most BSE cases; these three signs are the most frequent changes in mental status, sensation, and posture and movement, respectively.

Changes in mental status affect behaviour and temperament; the first sign of BSE may be when a normally placid animal becomes aggressive and kicks in the milking shed. Hypersensitivity can be to touch, sound and light. Ataxia affects mainly the hind limbs. Other abnormalities of posture and movement include falling, tremor and abnormal head carriage.

Diagnosis

There is currently no validated diagnostic test for the BSE agent in live animals. Laboratory tests on brain and spinal cord tissue obtained at postmortem examination are therefore required to confirm this disease.

Persistence of the agent

A particular feature of TSE agents, including BSE, is resistance to inactivation by physical or chemical procedures such as freezing, desiccation, ultraviolet radiation, and the usual methods of chemical and heat disinfection. TSE agents survive for long periods in carcases and withstand many of the procedures currently used in the commercial processing of bovine products.

Irrespective of the BSE risk status of a country, the following commodities are recognised by the OIE as *not* representing a risk of transmitting BSE: milk and milk products, semen and in vivo–derived cattle embryos that are collected and handled correctly, hides and skins, and deboned skeletal muscle meat.

Impacts for Australia

A case of BSE in Australia is likely to impose significant costs, not just to the beef industry but across the broader economy. There may also be significant public health impacts, with Creutzfeldt–Jakob disease affecting people who have consumed foods containing specified risk materials such as brain and spinal cord from BSE-affected cattle.

Glossary

Disease-specific terms

Bonemeal	See Meatmeal/bonemeal
Cohort cattle	Cattle which, during their first year of life, were reared with a BSE case during their first year of life, and which investigation showed consumed the same potentially contaminated feed during that period or If the results of the investigation are inconclusive, all cattle born in the same herd as, and within 12 months of the birth of, a BSE case.
Confirmed case	A suspect case with positive results from a TSE-specific immunohistochemistry, immunochemistry or other confirmatory test.
Iatrogenic disease	A case of disease caused by medical or veterinary procedures (eg an infection spread by surgical procedures).
Immunochemistry	The branch of immunology, or a diagnostic test, concerned with chemical substances and reactions of the immune system, specifically antigens and antibodies and their interactions with one another.
Immunohistochemistry	Immunochemistry applied to the study, or testing, of cells and tissues.
Meatmeal/bonemeal	The solid protein products obtained when animal tissues are rendered. See also Rendering (of carcasses)
Prion	Word coined in the 1980s for 'proteinaceous infectious particle'. Prion protein (PrPSc) is an abnormal form of a common cellular membrane protein (PrPc). PrPSc is more resistant to protein-digesting enzymes (proteases) than PrPc and is the major constituent of scrapie-associated fibrils. Prion proteins are thought to be involved in the transmission of TSEs and to be the sole disease agent for BSE. See also Scrapie-associated fibrils
Rendering (of carcasses)	Processing by heat to inactivate infective agents. Rendered material may be used in various products according to particular disease circumstances.
Ruminant	Any of various cud-chewing, cloven-hoofed quadrupeds, such as cattle, deer or camels, that usually have a stomach divided into three or four compartments.
Scrapie	A TSE found in sheep and goats. Scrapie is endemic in the United Kingdom and many other parts of the world (but not in Australia). It can be transmitted naturally or experimentally to other animal species, including mice, and has been the experimental model for much TSE research.

Scrapie-associated fibrils	Abnormal fibrils caused by an accumulation of protease resistant prion protein (PrPSc) and identified by electron microscopy. First identified in scrapie-infected mice but now recognised as a characteristic of all TSEs. See also Prion
Specified risk materials (SRM)	Those parts of infected cattle considered likely to contain the BSE agent and therefore prevented by regulations from entering the human food or animal feed chains. Definitions vary between countries in terms of both cattle age and anatomy.
Transmissible spongiform encephalopathies (TSEs)	A group of diseases, affecting various animal species, that involve noninflammatory vacuolated (spongiform) degeneration of the grey matter areas of the brain and spinal cord.

Standard AUSVETPLAN terms

Animal byproducts	Products of animal origin that are not for consumption but are destined for industrial use (eg hides and skins, fur, wool, hair, feathers, hoofs, bones, fertiliser).
Animal Health Committee	A committee whose members are the chief veterinary officers of the Commonwealth, states and territories, along with representatives from the CSIRO Australian Centre for Disease Preparedness (CSIRO-ACDP) and the Australian Government Department of Agriculture, Water and the Environment. There are also observers from Animal Health Australia, Wildlife Health Australia, and the New Zealand Ministry for Primary Industries. The committee provides advice to the National Biosecurity Committee on animal health matters, focusing on technical issues and regulatory policy. *See also* National Biosecurity Committee
Animal products	Meat, meat products and other products of animal origin (eg eggs, milk) for human consumption or for use in animal feedstuff.
Approved disposal site	A premises that has zero susceptible livestock and has been approved as a disposal site for animal carcasses, or potentially contaminated animal products, wastes or things.
Approved processing facility	An abattoir, knackery, milk processing plant or other such facility that maintains increased biosecurity standards. Such a facility could have animals or animal products introduced from lower-risk premises under a permit for processing to an approved standard.
At-risk premises	A premises in a restricted area that contains a live susceptible animal(s) but is not considered at the time of classification to be an infected premises, dangerous contact premises, dangerous contact processing facility, suspect premises or trace premises.
Australian Chief Veterinary Officer	The nominated senior veterinarian in the Australian Government Department of Agriculture, Water and the Environment who manages international animal health commitments and the Australian Government's response to an animal disease outbreak. *See also* Chief veterinary officer
AUSVETPLAN	*Aus*tralian *Vet*erinary Emergency *Plan*. Nationally agreed resources that guide decision making in the response to emergency animal

	diseases (EADs). It outlines Australia's preferred approach to responding to EADs of national significance, and supports efficient, effective and coherent responses to these diseases.
Carcase	The body of an animal slaughtered for food.
Carcass	The body of an animal that died in the field.
Chief veterinary officer (CVO)	The senior veterinarian of the animal health authority in each jurisdiction (national, state or territory) who has responsibility for animal disease control in that jurisdiction. *See also* Australian Chief Veterinary Officer
Compartmentalisation	The process of defining, implementing and maintaining one or more disease-free establishments under a common biosecurity management system in accordance with OIE guidelines, based on applied biosecurity measures and surveillance, to facilitate disease control and/or trade.
Compensation	The sum of money paid by government to an owner for livestock or property that are destroyed for the purpose of eradication or prevention of the spread of an emergency animal disease, and livestock that have died of the emergency animal disease. *See also* Cost-sharing arrangements, Emergency Animal Disease Response Agreement
Consultative Committee on Emergency Animal Diseases (CCEAD)	The key technical coordinating body for animal health emergencies. Members are state and territory chief veterinary officers, representatives of CSIRO-ACDP and the relevant industries, and the Australian Chief Veterinary Officer as chair.
Control area (CA)	A legally declared area where the disease controls, including surveillance and movement controls, applied are of lesser intensity than those in a restricted area (the limits of a control area and the conditions applying to it can be varied during an incident according to need).
Cost-sharing arrangements	Arrangements agreed between governments (national and state/territory) and livestock industries for sharing the costs of emergency animal disease responses. *See also* Compensation, Emergency Animal Disease Response Agreement
Dangerous contact animal	A susceptible animal that has been designated as being exposed to other infected animals or potentially infectious products following tracing and epidemiological investigation.
Dangerous contact premises (DCP)	A premises, apart from an abattoir, knackery or milk processing plant (or other such facility) that, after investigation and based on a risk assessment, is considered to contain a susceptible animal(s) not showing clinical signs, but considered highly likely to contain an infected animal(s) and/or contaminated animal products, wastes or things that present an unacceptable risk to the response if the risk is not addressed, and that therefore requires action to address the risk.
Dangerous contact processing facility (DCPF)	An abattoir, knackery, milk processing plant or other such facility that, based on a risk assessment, appears highly likely to have

	received infected animals, or contaminated animal products, wastes or things, and that requires action to address the risk.
Declared area	A defined tract of land that is subjected to disease control restrictions under emergency animal disease legislation. There are two types of declared areas: restricted area and control area.
Decontamination	Includes all stages of cleaning and disinfection.
Depopulation	The removal of a host population from a particular area to control or prevent the spread of disease.
Destroy (animals)	To kill animals humanely.
Disease agent	A general term for a transmissible organism or other factor that causes an infectious disease.
Disease Watch Hotline	24-hour freecall service for reporting suspected incidences of exotic diseases – 1800 675 888.
Disinfectant	A chemical used to destroy disease agents outside a living animal.
Disinfection	The application, after thorough cleansing, of procedures intended to destroy the infectious or parasitic agents of animal diseases, including zoonoses; applies to premises, vehicles and different objects that may have been directly or indirectly contaminated.
Disinsectation	The destruction of insect pests, usually with a chemical agent.
Disposal	Sanitary removal of animal carcasses, animal products, materials and wastes by burial, burning or some other process so as to prevent the spread of disease.
Emergency animal disease	A disease that is (a) exotic to Australia or (b) a variant of an endemic disease or (c) a serious infectious disease of unknown or uncertain cause or (d) a severe outbreak of a known endemic disease, and that is considered to be of national significance with serious social or trade implications. *See also* Endemic animal disease, Exotic animal disease
Emergency Animal Disease Response Agreement	Agreement between the Australian and state/territory governments and livestock industries on the management of emergency animal disease responses. Provisions include participatory decision making, risk management, cost sharing, the use of appropriately trained personnel and existing standards such as AUSVETPLAN. *See also* Compensation, Cost-sharing arrangements
Endemic animal disease	A disease affecting animals (which may include humans) that is known to occur in Australia. *See also* Emergency animal disease, Exotic animal disease
Enterprise	*See* Risk enterprise
Enzyme-linked immunosorbent assay (ELISA)	A serological test designed to detect and measure the presence of antibody or antigen in a sample. The test uses an enzyme reaction with a substrate to produce a colour change when antigen–antibody binding occurs.

Epidemiological investigation	An investigation to identify and qualify the risk factors associated with the disease. *See also* Veterinary investigation
Epidemiology	The study of disease in populations and of factors that determine its occurrence.
Exotic animal disease	A disease affecting animals (which may include humans) that does not normally occur in Australia. *See also* Emergency animal disease, Endemic animal disease
Exotic fauna/feral animals	*See* Wild animals
Fomites	Inanimate objects (eg boots, clothing, equipment, instruments, vehicles, crates, packaging) that can carry an infectious disease agent and may spread the disease through mechanical transmission.
General permit	A legal document that describes the requirements for movement of an animal (or group of animals), commodity or thing, for which permission may be granted without the need for direct interaction between the person moving the animal(s), commodity or thing and a government veterinarian or inspector. The permit may be completed via a webpage or in an approved place (such as a government office or commercial premises). A printed version of the permit must accompany the movement. The permit may impose preconditions and/or restrictions on movements. *See also* Special permit
In-contact animals	Animals that have had close contact with infected animals, such as noninfected animals in the same group as infected animals.
Incubation period	The period that elapses between the introduction of a pathogen into an animal and the first clinical signs of the disease.
Index case	The first case of the disease to be diagnosed in a disease outbreak. *See also* Index property
Index property	The property on which the index case is found. *See also* Index case
Infected premises (IP)	A defined area (which may be all or part of a property) on which animals meeting the case definition are or were present, or the causative agent of the emergency animal disease is present, or there is a reasonable suspicion that either is present, and that the relevant chief veterinary officer or their delegate has declared to be an infected premises.
Local control centre	An emergency operations centre responsible for the command and control of field operations in a defined area.
Monitoring	Routine collection of data for assessing the health status of a population or the level of contamination of a site for remediation purposes. *See also* Surveillance
Movement control	Restrictions placed on the movement of animals, people and other things to prevent the spread of disease.

National Biosecurity Committee	A committee that was formally established under the Intergovernmental Agreement on Biosecurity (IGAB). The IGAB was signed on 13 January 2012, and signatories include all states and territories except Tasmania. The committee provides advice to the Agriculture Senior Officials Committee and the Agriculture Ministers' Forum on national biosecurity issues, and on the IGAB.
National Management Group (NMG)	A group established to approve (or not approve) the invoking of cost sharing under the Emergency Animal Disease Response Agreement. NMG members are the Secretary of the Australian Government Department of Agriculture, Water and the Environment as chair, the chief executive officers of the state and territory government parties, and the president (or analogous officer) of each of the relevant industry parties.
Native wildlife	*See* Wild animals
OIE Terrestrial Code	OIE *Terrestrial animal health code*. Describes standards for safe international trade in animals and animal products. Revised annually and published on the internet at: www.oie.int/international-standard-setting/terrestrial-code/access-online.
OIE Terrestrial Manual	OIE *Manual of diagnostic tests and vaccines for terrestrial animals*. Describes standards for laboratory diagnostic tests, and the production and control of biological products (principally vaccines). The current edition is published on the internet at: www.oie.int/en/standard-setting/terrestrial-manual/access-online.
Operational procedures	Detailed instructions for carrying out specific disease control activities, such as disposal, destruction, decontamination and valuation.
Outside area (OA)	The area of Australia outside the declared (control and restricted) areas.
Owner	Person responsible for a premises (includes an agent of the owner, such as a manager or other controlling officer).
Polymerase chain reaction (PCR)	A method of amplifying and analysing DNA sequences that can be used to detect the presence of viral DNA.
Premises	A tract of land including its buildings, or a separate farm or facility that is maintained by a single set of services and personnel.
Premises of relevance (POR)	A premises in a control area that contains a live susceptible animal(s) but is not considered at the time of classification to be an infected premises, suspect premises, trace premises, dangerous contact premises or dangerous contact processing facility.
Prevalence	The proportion (or percentage) of animals in a particular population affected by a particular disease (or infection or positive antibody titre) at a given point in time.
Proof of freedom	Reaching a point following an outbreak and post-outbreak surveillance when freedom from the disease can be claimed with a reasonable level of statistical confidence.

Qualifiers	
– assessed negative	Assessed negative (AN) is a qualifier that may be applied to ARPs, PORs, SPs, TPs, DCPs or DCPFs. The qualifier may be applied following surveillance, epidemiological investigation, and/or laboratory assessment/diagnostic testing and indicates that the premises is assessed as negative at the time of classification.
– sentinels on site	Sentinels on site (SN) is a qualifier that may be applied to IPs and DCPs to indicate that sentinel animals are present on the premises as part of response activities (ie before it can be assessed as an RP).
– vaccinated	The vaccinated (VN) qualifier can be applied in a number of different ways. At its most basic level, it can be used to identify premises that contain susceptible animals that have been vaccinated against the EAD in question. However, depending on the legislation, objectives and processes within a jurisdiction, the VN qualifier may be used to track a range of criteria and parameters.
Quarantine	Legally enforceable requirement that prevents or minimises spread of pests and disease agents by controlling the movement of animals, persons or things.
Resolved premises (RP)	An infected premises, dangerous contact premises or dangerous contact processing facility that has completed the required control measures, and is subject to the procedures and restrictions appropriate to the area in which it is located.
Restricted area (RA)	A relatively small legally declared area around infected premises and dangerous contact premises that is subject to disease controls, including intense surveillance and movement controls.
Risk enterprise	A defined livestock or related enterprise that is potentially a major source of infection for many other premises. Includes intensive piggeries, feedlots, abattoirs, knackeries, saleyards, calf scales, milk factories, tanneries, skin sheds, game meat establishments, cold stores, artificial insemination centres, veterinary laboratories and hospitals, road and rail freight depots, showgrounds, field days, weighbridges and garbage depots.
Sensitivity	The proportion of truly positive units that are correctly identified as positive by a test. *See also* Specificity
Sentinel animal	Animal of known health status that is monitored to detect the presence of a specific disease agent.
Seroconversion	The appearance in the blood serum of antibodies (as determined by a serology test) following vaccination or natural exposure to a disease agent.
Serosurveillance	Surveillance of an animal population by testing serum samples for the presence of antibodies to disease agents.
Serotype	A subgroup of microorganisms identified by the antigens carried (as determined by a serology test).
Serum neutralisation test	A serological test to detect and measure the presence of antibody in a sample. Antibody in serum is serially diluted to detect the highest

	dilution that neutralises a standard amount of antigen. The neutralising antibody titre is given as the reciprocal of this dilution.
Slaughter	The humane killing of an animal for meat for human consumption.
Special permit	A legal document that describes the requirements for movement of an animal (or group of animals), commodity or thing, for which the person moving the animal(s), commodity or thing must obtain prior written permission from the relevant government veterinarian or inspector. A printed version of the permit must accompany the movement. The permit may impose preconditions and/or restrictions on movements. *See also* General permit
Specificity	The proportion of truly negative units that are correctly identified as negative by a test. *See also* Sensitivity
Stamping out	The strategy of eliminating infection from premises through the destruction of animals in accordance with the particular AUSVETPLAN manual, and in a manner that permits appropriate disposal of carcasses and decontamination of the site.
State coordination centre	The emergency operations centre that directs the disease control operations to be undertaken in a state or territory.
Surveillance	A systematic program of investigation designed to establish the presence, extent or absence of a disease, or of infection or contamination with the causative organism. It includes the examination of animals for clinical signs, antibodies or the causative organism.
Susceptible animals	Animals that can be infected with a particular disease.
Suspect animal	An animal that may have been exposed to an emergency disease such that its quarantine and intensive surveillance, but not pre-emptive slaughter, is warranted. or An animal not known to have been exposed to a disease agent but showing clinical signs requiring differential diagnosis.
Suspect premises (SP)	Temporary classification of a premises that contains a susceptible animal(s) not known to have been exposed to the disease agent but showing clinical signs similar to the case definition, and that therefore requires investigation(s).
Swill	Also known as 'prohibited pig feed', means material of mammalian origin, or any substance that has come in contact with this material, but does not include: (i) Milk, milk products or milk by-products either of Australian provenance or legally imported for stockfeed use into Australia. (ii) Material containing flesh, bones, blood, offal or mammal carcases which is treated by an approved process.[1] (iii) A carcass or part of a domestic pig, born and raised on the property on which the pig or pigs that are administered the part are held, that is administered for therapeutic purposes in accordance with the written instructions of a veterinary practitioner.

	(iv) Material used under an individual and defined-period permit issued by a jurisdiction for the purposes of research or baiting. [1] In terms of (ii), approved processes are: 1. rendering in accordance with the 'Australian Standard for the Hygienic Rendering of Animal Products' 2. under jurisdictional permit, cooking processes subject to compliance verification that ensure that a core temperature of at least 100 °C for a minimum of 30 minutes, or equivalent, has been reached. 3. treatment of cooking oil, which has been used for cooking in Australia, in accordance with the 'National Standard for Recycling of Used Cooking Fats and Oils intended for Animal Feeds' 4. under jurisdictional permit, any other nationally agreed process approved by AHC for which an acceptable risk assessment has been undertaken and that is subject to compliance verification. The national definition is a minimum standard. Some jurisdictions have additional conditions for swill feeding that pig producers in those jurisdictions must comply with, over and above the requirements of the national definition.
Swill feeding	Also known as 'feeding prohibited pig feed', it includes: • feeding, or allowing or directing another person to feed, prohibited pig feed to a pig • allowing a pig to have access to prohibited pig feed • the collection and storage or possession of prohibited pig feed on a premises where one or more pigs are kept • supplying to another person prohibited pig feed that the supplier knows is for feeding to any pig. This definition was endorsed by the Agriculture Ministers' Council through AGMIN OOS 04/2014.
Trace premises (TP)	Temporary classification of a premises that contains susceptible animal(s) that tracing indicates may have been exposed to the disease agent, or contains contaminated animal products, wastes or things, and that requires investigation(s).
Tracing	The process of locating animals, people or other items that may be implicated in the spread of disease, so that appropriate action can be taken.
Unknown status premises (UP)	A premises within a declared area where the current presence of susceptible animals and/or risk products, wastes or things is unknown.
Vaccination	Inoculation of individuals with a vaccine to provide active immunity.

Vaccine	A substance used to stimulate immunity against one or several disease-causing agents to provide protection or to reduce the effects of the disease. A vaccine is prepared from the causative agent of a disease, its products or a synthetic substitute, which is treated to act as an antigen without inducing the disease.
– adjuvanted	A vaccine in which one or several disease-causing agents are combined with an adjuvant (a substance that increases the immune response).
– attenuated	A vaccine prepared from infective or 'live' microbes that are less pathogenic but retain their ability to induce protective immunity.
– gene deleted	An attenuated or inactivated vaccine in which genes for non-essential surface glycoproteins have been removed by genetic engineering. This provides a useful immunological marker for the vaccine virus compared with the wild virus.
– inactivated	A vaccine prepared from a virus that has been inactivated ('killed') by chemical or physical treatment.
– recombinant	A vaccine produced from virus that has been genetically engineered to contain only selected genes, including those causing the immunogenic effect.
Vector	A living organism (frequently an arthropod) that transmits an infectious agent from one host to another. A *biological* vector is one in which the infectious agent must develop or multiply before becoming infective to a recipient host. A *mechanical* vector is one that transmits an infectious agent from one host to another but is not essential to the life cycle of the agent.
Veterinary investigation	An investigation of the diagnosis, pathology and epidemiology of the disease. *See also* Epidemiological investigation
Viraemia	The presence of viruses in the blood.
Wild animals	
– native wildlife	Animals that are indigenous to Australia and may be susceptible to emergency animal diseases (eg bats, dingoes, marsupials).
– feral animals	Animals of domestic species that are not confined or under control (eg cats, horses, pigs).
– exotic fauna	Nondomestic animal species that are not indigenous to Australia (eg foxes).
Wool	Sheep wool.
Zero susceptible species premises (ZP)	A premises that does not contain any susceptible animals or risk products, wastes or things.
Zoning	The process of defining, implementing and maintaining a disease-free or infected area in accordance with OIE guidelines, based on geopolitical and/or physical boundaries and surveillance, to facilitate disease control and/or trade.
Zoonosis	A disease of animals that can be transmitted to humans.

Abbreviations

Standard AUSVETPLAN abbreviations

Abbreviation	Full title
ACDP	Australian Centre for Disease Preparedness
AN	assessed negative
ARP	at-risk premises
AUSVETPLAN	Australian Veterinary Emergency Plan
CA	control area
CCEAD	Consultative Committee on Emergency Animal Diseases
CSIRO	Commonwealth Scientific and Industrial Research Organisation
CVO	chief veterinary officer
DCP	dangerous contact premises
DCPF	dangerous contact processing facility
EAD	emergency animal disease
EADRA	Emergency Animal Disease Response Agreement
EADRP	Emergency Animal Disease Response Plan
EDTA	ethylenediaminetetraacetic acid (anticoagulant for whole blood)
ELISA	enzyme-linked immunosorbent assay
GP	general permit
IETS	International Embryo Technology Society
IP	infected premises
LCC	local control centre
NMG	National Management Group
OA	outside area
OIE	World Organisation for Animal Health
PCR	polymerase chain reaction
POR	premises of relevance
RA	restricted area
RP	resolved premises
SCC	state coordination centre
SP	suspect premises

Abbreviation	Full title
SpP	special permit
TP	trace premises
UP	unknown status premises
ZP	zero susceptible stock premises

References

Bencsik A and Baron T (2007). Bovine spongiform encephalopathy agent in a prion protein $(PrP)^{ARR/ARR}$ genotype sheep after peripheral challenge: complete immunohistochemical analysis of disease-associated PrP and transmission studies to ovine-transgenic mice. *Journal of Infectious Diseases* 195:989–996.

Bellworthy SJ, Dexter G, Stack M, Chaplin M, Hawkins SAC, Simmons MM, Jeffrey M, Martin S, Gonzalez L and Hill P (2005a). Natural transmission of BSE between sheep within an experimental flock. *Veterinary Record* 157:206.

Bellworthy SJ, Hawkins SAC, Green RB, Blamire I, Dexter G, Dexter I, Lockey R, Jeffrey M, Ryder S, Berthelin-Baker C and Simmons MM (2005b). Tissue distribution of bovine spongiform encephalopathy infectivity in Romney sheep up to the onset of clinical disease after oral challenge. *Veterinary Record* 156:197–202.

Biacabe AG, Morignat E, Vulin J, Calavas D and Baron T (2008). Atypical bovine spongiform encephalopathies, France, 2001–2007. *Emerging Infectious Diseases* 14:298–300.

Brown P, Will RG, Bradley R, Asher DM and Detwiler L (2001). Bovine spongiform encephalopathy and variant Creutzfeldt–Jakob disease: background, evolution, and current concerns. *Emerging Infectious Diseases* 2001:7(1).

Bruce ME, Will RG, Ironside JW, McConnell I, Drummond D, Suttie A, McCardle L, Chree A, Hope J, Birkett C, Cousens S, Fraser H and Bostock CJ (1997). Transmissions to mice indicate that 'new variant' CJD is caused by the BSE agent. *Nature* 388:498–501.

Capobianco R, Casalone C, Suardi C, Mangieri M, Miccolo M, Limido L, Catania M, Rossi G, Di Fede G, Giaccone G, Grazia Bruzzone M, Minati L, Corona C, Acutis P, Gelmettis D, Lombardis G, Groschup M, Buschmann A, Zanusso G, Monaco S, Caramelli M and Tagliavini F (2007). Conversion of the BASE prion strain into the BSE strain: the origin of BSE? *PLoS Pathogens* 3:3.

Clarke P and Ghani A (2005). Projections of the future course of the primary vCJD epidemic in the UK: inclusion of subclinical infection and the possibility of wider genetic susceptibility. *Journal of the Royal Society Interface* 2(2):19–31.

Collee JG and Bradley R (1997a). BSE: a decade on — part 1. *Lancet* 349:636–641.

Collee JG and Bradley R (1997b). BSE: a decade on — part 2. *Lancet* 349:715–721.

Collinge J, Sidle KCL, Meads J, Ironside J and Hill AF (1996). Molecular analysis of prion strain variation and the aetiology of 'new variant' CJD. *Nature* 383:685–690.

Comer PJ and Huntly J (2003). Exposure of the human population to BSE infectivity over the course of the BSE epidemic in Great Britain and the impact

of changes to the Over Thirty Month Rule. www.food.gov.uk/multimedia/pdfs/otmcomer.pdf

Comoy EE, Casalone C, Lescoutra-Etchegaray N, Zanusso G, Freire S, Marce D, et al (2008). Atypical BSE (BASE) transmitted from asymptomatic aging cattle to a primate. *PLoS One* 2008;3:e3017. DOI: 10.1371/journal.pone.0003017.

Dobly A, Langeveld J, van Keulen L, Rodeghiero C, Durand S, Geeroms R, Van Muylem P, De Sloovere J, Vanopdenbosch E and Roels S (2010). No H- and L-type cases in Belgium in cattle diagnosed with

bovine spongiform encephalopathy (1999–2008) aging seven years and older. *BMC Veterinary Research* 6:26 doi:10.1186/1746-6148-6-26.

Donnelly CA and Ferguson M (2000). Population models: results and sensitivity analyses. In: *Statistical Aspects of BSE and vCJD Models for Epidemics*, Donnelly CA and Ferguson M (eds), Chapman & Hall/CRC, London, 59–84.

EC (European Commission) (2009). Report on the monitoring and testing of ruminants for the presence of transmissible spongiform encephalopathy (TSE) in the EU in 2008. http://ec.europa.eu/food/food/biosafety/tse_bse/monitoring_annual_reports_en.htm

EFSA (European Food Safety Authority) (2007). Opinion of the Scientific Panel on Biological Hazards on a request from the European Commission on the infectivity in SRM derived from cattle at different age groups estimated by back calculation modelling. *The EFSA Journal* 476:1–47.

Heaton MP, Keele JW, Harhay GP, Richt JA, Koohmaraie M, Wheeler TL, et al (2008). Prevalence of the prion gene E211K variant in US cattle. *BMC Veterinary Research* 4:25.

Hill AF, Desbruslais M, Joiner S, Sidle KCL, Gowland I, Collinge J, Doey LJ and Lantos P (1997). The same prion strain causes vCJD and BSE. *Nature* 389:448–450.

Hill G (2005). *Review of the Evidence for the Occurrence of 'BARB' BSE Cases in Cattle*, report to the Animal Health and Welfare Directorate General of the United Kingdom Department for Environment, Food and Rural Affairs, London.

Hunter N, Goldmann W, Foster JD, Cairns D and Smith G (1997). Natural scrapie and PrP genotype: case–control studies in British sheep. *Veterinary Record* 141:137–140.

Kimberlin RH (1992). Bovine spongiform encephalopathy. *Scientific and Technical Review*, *Office International des Epizooties* 11:347–390.

Kong Q, Zheng M, Casalone C, Qing L, Huang S, Chakraborty B, et al (2008). Evaluation of the human transmission risk of an atypical bovine spongiform encephalopathy prion strain. *Journal of Virology* 82:3697–3701 doi: 10.1128/JVI.02561-07.

Lasmezas CI, Deslys JP, Demaimay R, Adjou KT, Lamoury F, Dormont D, Robain O, Ironside J and Hauw JJ (1996). BSE transmission to macaques. *Nature* 381:743–744.

Lombardi G, Casalone C, D'Angelo A, Gelmetti D, Torcoli G, Barbieri I, et al (2008). Intraspecies transmission of BASE induces clinical dullness and amyotrophic changes. *PLoS Pathogens* 4:e1000075.

Maddison BC, Baker CA, Rees HC, Terry LA, Thorne L, Bellworthy SJ, Whitelam GC and Gough KC (2009). Prions are secreted in milk from clinically normal scrapie-exposed sheep. *Journal of Virology* 83:8293–8296.

Murdoch B, Clawson M, Laegreid W, Stothard P, Settles M, McKay S, Prasad A, Wang Z, Moore S and Williams J (2010). A 2cM genome-wide scan of European Holstein cattle affected by classical BSE. *BMC Genetics* 11:20 doi:10.1186/1471-2156-11-20.

Nichols TA, Pulford B, Wyckoff AC, Meyerett C, Michel B, Gertig K, Hoover EA, Jewell JE, Telling GC and Zabel MD (2009). Detection of protease-resistant cervid prion protein in water from a CWD-endemic area. *Prion* 3:171–183.

Nicholson E, Brunelle B, Richt J, Kehrli M and Greenlee J (2008). Identification of a heritable polymorphism in bovine PRNP associated with genetic transmissible spongiform encephalopathy: evidence of heritable BSE. *PLoS ONE* 3:8.

OIE (World Organisation for Animal Health) (1996). Bovine spongiform encephalopathy: an update. *Scientific and Technical Review, Office International des Epizooties* 15(3):1087–1118.

OIE (World Organisation for Animal Health) (2011). Bovine spongiform encephalopathy. In: *Manual of Diagnostic Tests and Vaccines for Terrestrial Animals*, article 2.4.6. www.oie.int/fileadmin/Home/eng/Health_standards/tahm/2.04.06_BSE.pdf

Peet RL and Curran JM (1992). Spongiform encephalopathy in an imported cheetah. *Australian Veterinary Journal* 69(7):171.

Prusiner SB (1998). Prions. *Proceedings of the National Academy of Sciences of the United States of America* 95:13363–13383.

SCAHLS (Sub-Committee on Animal Health Laboratory Standards) (2010). *Australia and New Zealand Standard Diagnostic Procedure — Transmissible Spongiform Encephalopathies*.

www.scahls.org.au/__data/assets/pdf_file/0006/1864752/ANZSDP_TSE_FINAL.pdf

SEAC (Spongiform Encephalopathy Advisory Committee) (2003). The effect of oral dose on attack rate and incubation period of BSE in cattle. Reserved business SEAC/INF/80/7. www.seac.gov.uk/papers/80-7-reserved.pdf (viewed 12 July 2010)

SEAC (Spongiform Encephalopathy Advisory Committee) (2006). Sheep Subgroup Statement. www.seac.gov.uk/statements/sheepsubgrp-statement131006.pdf

Taylor DM (1996a). Exposure to, and inactivation of, the unconventional agents that cause transmissible degenerative encephalopathies. In: *Methods in Molecular Medicine: Prion Diseases*, Baker H and Ridley RM (eds), Humana Press Inc., Totowa, New Jersey, 105–118.

Taylor DM (1996b). Inactivation studies on BSE agent. *British Food Journal* 98:36–39.

Taylor K (1996). Review of the UK/EU BSE and scrapie situation. In: *Proceedings of the Workshop on the Implications of International Disease Emergencies. BSE: a case study*, Office of the Australian Chief Veterinary Officer, Sydney, 15–23.

UK DEFRA (United Kingdom Department for Environment, Food and Rural Affairs) (2001). *Review of the Origin of BSE (Horn Committee Report)*, DEFRA, London. www.defra.gov.uk/animalh/bse/publications/bseorigin.pdf

Wells GAH, Scott AC and Johnson CT (1987). A novel progressive spongiform encephalopathy in cattle. *Veterinary Record* 121:419–420.

Wells GA, Hancock RD, Cooley WA, Richards MS, Higgins RJ and David GP (1989). Bovine spongiform encephalopathy: diagnostic significance of vacuolar changes in selected nuclei of the medulla oblongata. *Veterinary Record* 125:521–524.

Wells GA, Hawkins SA, Green RB, Austin AR, Dexter I, Spencer YI, Chaplin MJ, Stack MJ and Dawson M (1998). Preliminary observations on the pathogenesis of experimental bovine spongiform encephalopathy (BSE): an update. *Veterinary Record* 142:103–106.

Wells GAH, Konold T, Arnold ME, Austin AR, Hawkins SAC, Stack M, Simmons MM, Lee YH, Gavier-Widén D, Dawson M and Wilesmith JW (2007). Bovine spongiform encephalopathy: the effect of oral exposure dose on attack rate and incubation period in cattle. *Journal of General Virology* 88(4):1363–1373.

Wilesmith JW (1998). *Manual on Bovine Spongiform Encephalopathy. FAO Animal Health Manual – 2*, Food and Agriculture Organization of the United Nations, Rome.

Wilesmith JW, Wells GAH, Cranwell MP and Ryan JMB (1988). Bovine spongiform encephalopathy: epidemiological studies. *Veterinary Record* 123:638–644.

Will RG, Ironside JW, Zeidler M, Cousens SN, Estibeiro K, Alperovitch A, Poser S, Pocchiari M, Hofman A and Smith PG (1996). A new variant of Creutzfeldt–Jakob disease in the UK. *Lancet* 347:921–925.

Wrathall AE, Brown KDF, Sayers AR, Wells GAH, Simmons MM, Farrelly SSJ, Bellerby P, Squirrell J, Spencer YI, Wells M, Stack MJ, Bastiman B, Pullar D, Scatcherd J, Heasman L, Parker J, Hannam DAR, Helliwell DW, Chree A and Fraser H (2002). Studies of embryo transfer from cattle affected by BSE. *Veterinary Record* 150:365–378.

Yainshet A, Cao L and Elliston L (2006). A hypothetical case of BSE in Australia: economic impact of a temporary loss of market access. ABARE report

prepared for Product Integrity, Animal and Plant Health, Australian Government Department of Agriculture, Fisheries and Forestry, Canberra.

Young S and Slocombe RF (2003). Prion-associated spongiform encephalopathy in an imported Asiatic golden cat (*Catopuma temmincki*). *Australian Veterinary Journal* 81:295–296.

Further reading and internet links

Advisory Committee on Dangerous Pathogens (2003). *Transmissible spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection*, Stationery Office, Norwich, UK. www.dh.gov.uk/ab/ACDP/TSEguidance/index.htm

Australia's Transmissible Spongiform Encephalopathies Freedom Assurance Program (TSEFAP).

www.animalhealthaustralia.com.au/programs/biosecurity/tse-freedom-assurance-program

Brown P, Gibbs CJ Jr, Rodgers-Johnson P, Asher DM, Sulima MP, Bacote A, Goldfarb LG and Gajdusek DC (1994). Human spongiform encephalopathy: the National Institutes of Health series of 300 cases of experimentally transmitted disease. *Annals of Neurology* 35:513–529.

Cohen JT, Duggar K, Gray GM, Kreindel S, Abdelrahman H, HabteMariam T, Oryang D and Tameru B (2001). *Evaluation of the Potential for Bovine Spongiform Encephalopathy in the United States*. Harvard School of Public Health/ College of Veterinary Medicine, Tuskegee University.

Cooley WA, Clark JK, Ryder SJ, Davis LA, Farrelly SSJ and Stack MJ (2001). Evaluation of a rapid western immunoblotting procedure for the diagnosis of bovine spongiform encephalopathy (BSE) in the UK. *Journal of Comparative Pathology* 125:64–70.

Dawson M, Wells GAH and Parker BNJ (1990). Preliminary evidence of the experimental transmissibility of bovine spongiform encephalopathy to cattle. *Veterinary Record* 126:112–113.

European Commission (2002). Updated opinion and report on: A treatment of animal waste by means of high temperature (150°C, 3 hours) and high pressure alkaline hydrolysis. EC Health & Consumer Protection Directorate-General, Scientific Steering Committee. http://ec.europa.eu/food/fs/sc/ssc/out297_en.pdf>

European Food Safety Authority (2004). Annex to The EFSA Journal (2004) 56. Report of the Working Group on the scientific justification for proposing amendments to the United Kingdom Date Based Export Scheme and the Over Thirty Months rule. European Food Safety Authority, opinion published 21 April 2004. www.efsa.eu.int/science/biohaz/biohaz_opinions/429/opinion_biohaz_10_dbesotm_ef56_report_en1.pdf

Fraser H, Bruce ME, Chree A, McConnell I and Wells GAH (1992). Transmission of bovine spongiform encephalopathy and scrapie to mice. *Journal of General Virology* 73:1891–1897.

Fraser H, McConnell I, Wells GAH and Dawson M (1988). Transmission of bovine spongiform encephalopathy to mice. *Veterinary Record* 123:472.

Geering WA, Forman AJ and Nunn MJ (1995). *Exotic Diseases of Animals. A Field Guide for Australian Veterinarians*, Department of Primary Industries and Energy, Australian Government Publishing Service, Canberra.

Kao RR, Gravenor MB, Baylis M, Bostock CJ, Chihota CM, Evans JC, Goldmann W, Smith AJ and McLean AR (2002). The potential size and duration of an epidemic of bovine spongiform encephalopathy in British sheep. *Science* 295:332–335.

Middleton DJ and Barlow RM (1993). Failure to transmit bovine spongiform encephalopathy to mice by feeding them with extraneural tissues of affected cattle. *Veterinary Record* 132:545–547.

NHMRC (National Health and Medical Research Council) (1996). *Creutzfeldt–Jakob Disease and Other Human Transmissible Spongiform Encephalopathies: Guidelines on Patient Management and Infection Control*, Australian Government Publishing Service, Canberra, 1–46.

Ombardi G, Casalone C, D'Angelo A, Gelmetti D, Torcoli G, Barbieri I, Corona C, Fasoli E, Farinazzo A, Fiorini M, Gelati M, Iulini B, Tagliavini F, Ferrari S, Caramelli M, Monaco S, Capucci L and Zanusso G (2008). Intraspecies transmission of BASE induces clinical dullness and amyotrophic changes. *PLoS Pathogens* 4:5.

SSC (Scientific Steering Committee of the European Commission) (1998). Opinion on BSE risk, adopted by the Scientific Steering Committee at its meeting of 26–27 March 1998, following a public consultation on the preliminary opinion adopted on 19–20 February 1998, European Commission, Health and Consumer Protection Directorate-General. http://ec.europa.eu/food/fs/sc/ssc/outcome_en.html

SSC (Scientific Steering Committee of the European Commission) (1999a). *The Possible Vertical Transmission of Bovine Spongiform Encephalopathy (BSE).* Report of the Working Group, submitted to the Scientific Steering Committee at its meeting of 18–19 March 1999.
http://ec.europa.eu/food/fs/sc/ssc/outcome_en.html

SSC (Scientific Steering Committee of the European Commission) (1999b). Opinion on the human exposure risk (HER) via food with respect to BSE, adopted by the Scientific Steering Committee at its meeting of 10 December 1999. European Commission, Health and Consumer Protection Directorate-General.
http://ec.europa.eu/food/fs/sc/ssc/outcome_en.html

SSC (Scientific Steering Committee of the European Commission) (2000). Final opinion on the geographical risk of bovine spongiform encephalopathy (GBR), adopted by the Scientific Steering Committee on 6 July 2000. European

Commission, Health and Consumer Protection Directorate-General, 5.
http://ec.europa.eu/food/fs/sc/ssc/outcome_en.html

SSC (Scientific Steering Committee of the European Commission) (2001a). Opinion on hypotheses on the origin and transmission of BSE, adopted by the Scientific Steering Committee on 29–30 November 2001. European Commission, Health and Consumer Protection Directorate-General. http://ec.europa.eu/food/fs/sc/ssc/outcome_en.html

SSC (Scientific Steering Committee of the European Commission) (2001b). Opinion on the six BARB BSE cases in the UK since 1 August 1996, adopted by the Scientific Steering Committee at its meeting of 29–30 November 2001. European Commission, Health and Consumer Protection Directorate-General.

SSC (Scientific Steering Committee of the European Commission) (2002). Opinion on TSE infectivity distribution in ruminant tissues (state of knowledge, December 2001), adopted by the Scientific Steering Committee at its meeting of 10–11 January 2002. European Commission, Health and Consumer Protection Directorate-General.
http://ec.europa.eu/food/fs/sc/ssc/outcome_en.html

SSC (Scientific Steering Committee of the European Commission) (2003a). Updated opinion and report on the safety of dicalcium phosphate and tricalcium phosphate from bovine bones, used as animal feed additive or as fertilizer, adopted by the Scientific Steering Committee at its meeting of 6–7 March 2003.
http://ec.europa.eu/food/fs/sc/ssc/outcome_en.html

SSC (Scientific Steering Committee of the European Commission) (2003b). Updated opinion on the safety with regard to BSE risk of gelatine derived from ruminant bones or hides. Adopted by the Scientific Steering Committee at its meeting of 6–7 March 2003. http://ec.europa.eu/food/fs/sc/ssc/outcome_en.html

SSC (Scientific Steering Committee of the European Commission) (2003c). Opinion of the safety of tallow derivatives from cattle tallow, adopted by the Scientific Steering Committee on 10–11 April 2003. http://ec.europa.eu/food/fs/sc/ssc/outcome_en.html

SSC (Scientific Steering Committee of the European Commission) (2003d). Opinion and report on BSE in the UK's cattle born after 31 July 1996 (BARBS), adopted by the Scientific Steering Committee on 10–11 April 2003. http://ec.europa.eu/food/fs/sc/ssc/outcome_en.html

Taylor DM (2001). Resistance of transmissible spongiform encephalopathy agents to decontamination. In: *Prions: A Challenge for Science, Medicine and Public Health System*, Rabenau HF, Cinatl J and Doerr HW (eds), Karger Publishers, Basel, 58–67.

Taylor DM, Fernie K and Steele P (1999). Boiling in sodium hydroxide inactivates mouse-passaged BSE agent. Abstracts of a meeting of the Association of Veterinary Teachers and Research Workers, Scarborough, 29–31 March 1999, 22.

Taylor DM, Ferguson CE, Bostock CJ and Dawson M (1995a). Absence of disease in mice receiving milk from cows with bovine spongiform encephalopathy. *Veterinary Record* 136:592.

Taylor DM, Woodgate SL and Atkinson MJ (1995b). Inactivation of the bovine spongiform encephalopathy agent by rendering procedures. *Veterinary Record* 137:605–610.

UK Food Standards Agency (2000). *Review of BSE Controls*, UK FSA, London.

UK SEAC (United Kingdom Spongiform Encephalopathy Advisory Committee) (1996). Press release: SEAC Statement on Maternal Transmission of BSE, July 1996.

Wells GA, Dawson M, Hawkins SA, Green RB, Dexter I, Francis ME, Simmons MM, Austin AR and Horigan MW (1994). Infectivity in the ileum of cattle challenged orally with bovine spongiform encephalopathy. *Veterinary Record* 135:40–41.

Wells GA, Hawkins SA, Green RB, Spencer YI, Dexter I and Dawson M (1999). Limited detection of sternal bone marrow infectivity in the clinical phase of experimental bovine spongiform encephalopathy (BSE). *Veterinary Record* 144:292–294.

Wilesmith JW, Wells GAH, Ryan JBM, Gavier-Widen D and Simmons MM (1997). A cohort study to examine maternally-associated risk factors for bovine spongiform encephalopathy. *Veterinary Record* 141:239–243.

Wrathall AE (1997). Risks of transmitting scrapie and bovine spongiform encephalopathy by semen and embryos. *Scientific and Technical Review, Office International des Epizooties* 16:240–264.

Numbers of BSE cases (worldwide)

www.oie.int/animal-health-in-the-world/bse-portal

European Commission TSE website

http://ec.europa.eu/food/food/biosafety/tse_bse/index_en.htm

European Food Safety Authority scientific opinions (post-May 2010) http://www.efsa.europa.eu/en/biohaztopics/topic/bse.htm

European Scientific Steering Committee scientific opinions (pre-May 2003) http://ec.europa.eu/food/food/biosafety/tse_bse/scientific_advices_en.htm

UK Advisory Committee on Dangerous Pathogens

www.dh.gov.uk/ab/ACDP/index.htm

UK Creutzfeldt–Jakob Disease Surveillance Unit — vCJD cases worldwide

www.cjd.ed.ac.uk/vcjdworld.htm

UK Department for Environment, Food and Rural Affairs (DEFRA)

http://archive.defra.gov.uk/foodfarm/farmanimal/diseases/atoz/bse/science-research/

World Health Organization (WHO)

www.who.int/csr/disease/bse/en/

World Organisation for Animal Health (OIE) BSE risk status of member countries

www.oie.int/eng/Status/BSE/en_BSE_free.htm