AUSTRALIAN VETERINARY EMERGENCY PLAN

AUSVETPLAN

Response strategy

Bovine spongiform encephalopathy

Version 5.1

AUSVETPLAN is a series of 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

Disclaimer

This publication 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, as relevant. Any views and opinions expressed in this document do not necessarily represent the views and opinion of the authors or contributors, Animal Health Australia or the Commonwealth of Australia.

This publication is for use in emergency situations. The strategies and policy guidelines in this work are not applicable to quarantine policies for imported livestock or livestock products.

This publication is not legal or professional advice and should not be taken as a substitute for legal or other professional advice.

This publication is not intended for use by any person who does not have appropriate expertise in the subject matter of the work. Before using this publication, you should read it in full, consider its effects and determine whether it is appropriate for your needs.

This publication was created in December 2025. Laws, practices and regulations may have changed since that time. You should make your own inquiries as to the currency of relevant laws, practices and regulations, as these may have changed since publication of this work.

Warranty

No warranty is given as to the correctness of the information contained in this work, or of its suitability for use by you. To the fullest extent permitted by law, Animal Health Australia is not, and the other contributing parties are not liable for any statement or opinion, or for any error or omission contained in this work, and it and they disclaim all warranties with regard to the information contained in it, including, without limitation, all implied warranties of merchantability and fitness for a specific purpose. Animal Health Australia is not liable for any direct, indirect, special or consequential losses or damages of any kind, or for any loss of profit, loss or corruption of data, business interruption or indirect costs arising out of, or in connection with, the use of this work or the information contained in it, whether such loss or damage arises in contract, negligence, tort, under statute or otherwise.

This manual may contain text under development

In this manual, text in square brackets and greyed out [xxx] indicates that that aspect of the manual remains unresolved or is under development. This 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
PO Box 998
Dickson ACT 2602
ausvetplan@animalhealthaustralia.com.au

EMERGENCY ANIMAL DISEASE HOTLINE: 1800 675 888

The Emergency Animal Disease 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

Approved citation

Animal Health Australia (2025). *Response strategy: Bovine spongiform encephalopathy* (version 5.1). Australian Veterinary Emergency Plan (AUSVETPLAN), edition 5, Canberra, ACT.

Publication record

Edition 1 – none

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

Edition 4 – none

Edition 5

- Version 5.0, 2021 (incorporation into the Edition 5 format)
- Version 5.1, 2025 (updates to scientific information on the disease and for consistency with the latest World Organisation for Animal Health information)

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).

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.

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 (WOAH) *Terrestrial animal health code* (Chapters 1.8 and 11.4) and the WOAH *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.

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). 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[2]), where applicable.

1.3 Training resources

EAD preparedness and response arrangements in Australia

The EAD Foundation Online course[3] 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:

- https://animalhealthaustralia.com.au/maintaining-australias-freedom-from-tses/.

[1] https://animalhealthaustralia.com.au/ausvetplan/

[2] https://animalhealthaustralia.com.au/eadra/

[3] https://animalhealthaustralia.com.au/online-training-courses/

2 Nature of the disease

Three known strains of bovine spongiform encephalopathy (BSE) have been identified in cattle: classical BSE (C-type), 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 several countries for more than 20 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.

BSE is a World Organisation for Animal Health (WOAH)-listed disease.[4]

2.1 Aetiology

A protease-resistant isoform (PrPSc) of a normal cellular prion protein (PrPc) 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).

2.1.1 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).

2.1.2 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 several 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-

[4] WOAH-listed diseases are diseases with the potential for international spread, significant mortality or morbidity within the susceptible species, and/or potential for zoonotic spread to humans. WOAH member countries that have been free from a notifiable disease are obliged to notify WOAH within 24 hours of confirming the presence of the disease.

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

Bovines

BSE is primarily a disease of domestic cattle (genus *Bos*) and buffalo (genus *Bubalus*), i.e. bovines. Other animals may be infected either naturally or experimentally, but they are not recognised as being epidemiologically significant.

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

Experimental transmission of BSE has been reported in pigs (intracerebral, intraperitoneal and intravenous), but pigs have not been shown to be susceptible to oral challenge (Hedman et al 2016).

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). 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 have 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 has been maintained on the website of the United Kingdom CJD Research and Surveillance Unit, however the funding for this initiative ended in March 2025.[5]

2.3 World distribution

For the latest information on the distribution of BSE, refer to the World Organisation for Animal Health (WOAH) 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

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

[6] https://wahis.oie.int/#/home

cattle and MBM from the UK and subsequently from other BSE-affected countries, have occurred in mainland Europe, Canada, Japan and Israel.

Cases of atypical BSE in cattle have been reported in several countries, including in the UK, mainland Europe, Canada, the United States, Japan, and Brazil. However, these cases are extremely rare, with just over 100 cases identified to date. Several countries have detected atypical BSE strains despite having no recorded cases of BSE 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 of these cases, except one in Japan, were 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 of feeds containing BSE contaminated MBM is mainly by young animals (Collee and Bradley 1997ab, Wilesmith 1998, Brown et al 2001). All classical 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.

WOAH 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).

WOAH incubation period

There is no incubation period specified for the purposes of the WOAH Terrestrial Code; however, the WOAH Terrestrial Manual indicates that the incubation period is at least 2 years, but may extend beyond 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 of 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 ID50 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]

Animal products

TSE agents survive for long periods in carcases and withstand many of the procedures currently used to process products.

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 WOAH as safe commodities not representing a risk of transmitting BSE (as per Chapter 11.4 of the WOAH Terrestrial Animal Health Code, Article 11.4.2).

- milk and milk products
- semen and in vivo–derived bovine embryos collected and handled in accordance with the relevant chapters of the WOAH Terrestrial Code
- hides and skins
- gelatine and collagen
- 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)

[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

- foetal blood.

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 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 (e.g. 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. Biosecurity 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 (e.g. 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 Procedures* for TSEs (SCAHLS 2010) is the authoritative guide to laboratory diagnosis. Its methods are consistent with the current edition of the WOAH *Manual of Diagnostic Tests and Vaccines for Terrestrial Animals* (WOAH 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 Procedures* for TSEs (SCAHLS 2010) and the WOAH 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*[10] *2023-24*. 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***.

[10] Latest version available via: Maintaining Australia's freedom from TSEs - Animal Health Australia

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 Procedures for Transmissible Spongiform Encephalopathies* (SCAHLS 2010).

Histological examination to detect the characteristic changes in the CNS mentioned in Section 2.5.2 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, WOAH 2024a).

CSIRO-ACDP tests

Tests for detecting accumulated PrPSc 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
Immunohistochemistry	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 effective treatment available for an animal with a TSE. As the disease is inevitably fatal, humane destruction is the only option to prevent further suffering.

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 that had been imported from Europe, Japan, Canada (1996 onwards) and the United States (1996 onwards) were permanently identified under the National Livestock Identification System (NLIS). They were also in official 'lifetime quarantine' and never entered the human food, or animal feed, chains. None of these imported cattle are still alive. 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 biosecurity 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 (i.e. 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 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

Critical considerations for formulating a policy for the response to an incident of BSE in Australia include the following

- Classical 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 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, 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.
- 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 subsequently reported BSE cases were traced, and those still alive at the time were placed under official, permanent quarantine. None of these cattle are still alive.
- 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.
- Further case-by-case evaluation may be required to enhance traceability data derived from the NLIS database, in support of lifetime traceability.
- 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.

4 Policy and rationale

4.1 Introduction

4.1.1 Summary of policy

Bovine spongiform encephalopathy (BSE) is a World Organisation for Animal Health (WOAH)-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 classical 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 occurrence of classical 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
- *destruction and 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 Sections 2.5.1 and 2.5.2), 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 2.5.5), 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
Cohorts to a BSE case	Cattle which 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

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

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).[11] 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 WOAH categorises countries into three risk levels associated with BSE: negligible, controlled and undetermined (WOAH, 2024b). Beef importing countries, including Australia, either adopt the WOAH's categorisations or conduct their own using similar criteria. Australia is classified by the WOAH as negligible BSE risk.

Negligible BSE risk countries that subsequently report a case of classical BSE in indigenous cattle will be reclassified by the WOAH as controlled risk, unless all cases were born more than 11 years ago.

The WOAH risk classification system (as per the WOAH Terrestrial Animal Health Code, Article 11.4.3) 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 assessment (entry assessment, exposure assessment, consequence assessment and risk estimation) that identifies all potential factors for BSE (classical) occurrence and their historic perspective
- the ongoing implementation of a BSE surveillance program within the bovine population, supported by
    - 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
    - compulsory notification and investigation of all cattle showing clinical signs consistent with BSE
    - appropriate laboratory testing in accordance with the WOAH Terrestrial Manual
    - robust, documented, evaluation procedures and protocols for the definition of the target population for BSE surveillance, the report of bovines displaying suspected clinical signs, the determination of animals to be subjected to laboratory testing, the collection and submission of samples for laboratory testing, and the follow-up epidemiological investigations for BSE positive findings.
- the history of occurrence and management of cases of BSE and bovines affected by atypical BSE.

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 WOAH. 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. 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

[11] Information about the EAD Response Agreement can be found at https://animalhealthaustralia.com.au/eadra/

- 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 WOAH Terrestrial Animal Health 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 WOAH requirements.

In the event of an outbreak, the National TSE Surveillance Program — within the TSE Freedom Assurance Program[12] 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 WOAH 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* (Part 1 and Part 2)**.

4.2 Public health implications

As described in Section 2.2.3, 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 (e.g. 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 National Health and Medical Research Council, established a Special Expert Committee on TSEs, the Transmissible Spongiform Encephalopathies Advisory Committee. The committee's purpose was 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. The committee was in place until 2015.

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, were put in place to protect the safety of the Australian blood supply. The blood donor restrictions were lifted in July 2022, allowing UK residents to now donate blood in Australia.

[12] https://animalhealthaustralia.com.au/maintaining-australias-freedom-from-tses/

Information on CJD, including how it is defined in Australia and how cases are monitored, is available on the Australian Government Department of Health website.[13]

4.3 Control and eradication policy

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

Details of the policy will depend on the type of incident that initiates an emergency response (e.g. 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 WOAH's conditions for a negligible BSE risk country if a case is reported in indigenous cattle and subsequent investigations confirm that any identified source of infection has been controlled, and the risk of BSE agents being recycled within the bovine population has continued to be negligible.

A case of atypical BSE in indigenous cattle is a sporadic event and will not affect Australia's negligible BSE risk status if any bovines affected by atypical BSE have been destroyed and disposed of to ensure that they have not entered the feed or food chain.

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 source of infection
- the incidence and prevalence of infection
- the presence and spatial distribution, or absence of:
    - populations of susceptible species
    - infected and disease-free populations
- pathways of spread and the likely size of the outbreak
- risk factors for the presence of infection and susceptibility to disease.

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 (e.g. 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.

[13] https://www.health.gov.au/diseases/creutzfeldt-jakob-disease-cjd

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 infected premises (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 the NLIS radio frequency identification devices (RFID) 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.

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*.[14]

[14] Latest version available via: Maintaining Australia's freedom from TSEs - Animal Health Australia

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.

4.3.6 Treatment of infected animals

There is no effective treatment available.

Any animal exhibiting clinical signs associated with BSE should be humanely destroyed to alleviate suffering and ensure the welfare of the affected animal is not further compromised.

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 (e.g. alkaline hydrolysis).

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 WOAH Terrestrial Animal Health 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, 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**
confirmed case and 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-ACDP 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-ACDP = Australian Centre for Disease Preparedness (CSIRO); DCP = dangerous contact premises; IP = infected premises; SRM = specified risk material; TP = trace premises

4.3.9 Disposal of animals, animal products and byproducts

Destruction and testing of cattle on farm is preferred to transporting them for slaughter at another site. Destruction 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 WOAH for transmissible spongiform encephalopathies (TSEs), can be considered (e.g. alkaline hydrolysis).
- The location of burial sites for carcasses, products or ash must be marked and comprehensively documented.

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 WOAH 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, 98% 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 should preferably be discarded after a single use. If reuse is necessary, they need to be correctly 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 (e.g. 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 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 and food safety authorities. 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 (particularly the fact that a case of atypical BSE doesn't affect Australia's BSE status)
- comparison with the UK epidemic and the situation in other countries.

See the *Biosecurity Incident Public Information Manual*[15] for further information on provision of public information about emergency animal diseases.

4.3.14 Other strategies

Apart from eradication (the policy described above), a secondary policy option would include 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.

The identification of a sporadic, atypical BSE case does not impact Australia's BSE status, provided the affected bovines are destroyed and disposed of in accordance with the WOAH Terrestrial Animal Health

[15] https://animalhealthaustralia.com.au/bipim/

Code (Article 11.4.4), ensuring they do not enter the feed or food chain. Such cases would require thorough investigation, documentation, and communication of findings.

4.4 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.[16] 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***.

[16] https://animalhealthaustralia.com.au/eadra/

5 Declared areas and premises

Information on declared areas and premises classifications is provided in the **AUSVETPLAN** ***Guidance document: Declared areas and allocation of premises classifications in an emergency animal disease response***.

Declared areas 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. It is also possible that other types of areas which are not legally declared, may be used for disease control purposes in some jurisdictions.

6 Movement controls

6.1 Principles

General principles for quarantine practices and movement controls for managing EADs are provided in the **AUSVETPLAN *Guidance document: Movement controls***.

6.2 Guidelines for issuing permits

Recommended biosecurity and movement controls 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. This response strategy indicates 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.

Guidelines for issuing permits are provided in the **AUSVETPLAN *Guidance document: Movement controls***.

Movements not reflected within this manual may be considered by the relevant jurisdictional CVO on a risk-assessed case-by-case basis.

6.3 Types of permits

Permits are either general or special. Emergency permits are a form of special permit (see also Glossary).

They 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. Both general and special permits may be in addition to documents required for routine movements between or within jurisdictions (e.g. health certificates, waybills, consignment notes, National Vendor Declarations – NVDs).

Details on permit types are provided in the **AUSVETPLAN *Guidance document: Movement controls***.

6.4 Recommended movement controls

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

Table 6.1 shows the recommended movement controls that will apply to IPs, DCPs and TPs in the initial stages of a BSE incident. Movements under permit are subject to risk assessment. 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	Allowed under normal jurisdictional requirements	Prohibited except under SpP1	Allowed under normal jurisdictional requirements	Prohibited except under SpP2
DCP	Prohibited except under SpP1	Prohibited except under SpP2	Allowed under normal jurisdictional requirements	Prohibited except under SpP1	Allowed under normal jurisdictional requirements	Prohibited except under SpP2
TP	Prohibited except under SpP1	Prohibited except under SpP2	Allowed under normal jurisdictional requirements	Prohibited except under SpP1	Allowed under normal jurisdictional requirements	Prohibited except under SpP2
Nondeclared premises	Allowed under normal jurisdictional requirements	Allowed under normal jurisdictional requirements	Allowed under normal jurisdictional requirements	Allowed under normal jurisdictional requirements	Allowed under normal jurisdictional requirements	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): animals must be permanently identified to enable lifetime traceability; GP1 (general permit 1): decontamination of the IP/DCP/TP is required before introduction of animals

Appendix 1 Bovine spongiform encephalopathy (BSE) 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 in several countries for more 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 (WOAH) 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 WOAH 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, gelatin and collagen, 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) and foetal blood.

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

Terms and definitions

Standard AUSVETPLAN terms

For definitions of standard AUSVETPLAN terms, see the **AUSVETPLAN *Glossary***.

Manual-specific terms

Term	Definition
Bonemeal	See Meatmeal/bonemeal
Cohort cattle	Cattle which, during their first year of life, were reared with a BSE case, 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 (e.g. 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.

Term	Definition
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.

Abbreviations

Standard AUSVETPLAN abbreviations

For standard AUSVETPLAN abbreviations, see the **AUSVETPLAN *Glossary***.

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.

Hedman C, Bolea R, Marín B, Cobrière F, Filali H, Vazquez F, Pitarch JL, Vargas A, Acín C, Moreno B, Pumarola M, Andreoletti O, Badiola JJ. Transmission of sheep-bovine spongiform encephalopathy to pigs. Vet Res. 2016 Jan 7;47:14. doi: 10.1186/s13567-015-0295-8. PMID: 26742788; PMCID: PMC4705642.

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.

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, (viewed 12 July 2010)

SEAC (Spongiform Encephalopathy Advisory Committee) (2006). Sheep Subgroup Statement**.**

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.

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.

WOAH (World Organisation for Animal Health) (2024a). Bovine spongiform encephalopathy. In: *Manual of Diagnostic Tests and Vaccines for Terrestrial Animals*, chapter 3.4.5 (adopted in May 2021), www.woah.org/fileadmin/Home/eng/Health_standards/tahm/3.04.05_BSE.pdf

WOAH (World Organisation for Animal Health) (2024b). Bovine spongiform encephalopathy. In: *Terrestrial Animal Health Code*, chapters 1.8 and 11.4, www.woah.org/en/what-we-do/standards/codes-and-manuals/terrestrial-code-online-access/

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.