AUSTRALIAN VETERINARY EMERGENCY PLAN

AUSVETPLAN

Disease Strategy

Bovine spongiform encephalopathy

Version 3.1, 2005

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

Primary Industries Ministerial Council

This disease strategy forms part of:

AUSVETPLAN Edition 3

This strategy will be reviewed regularly. Suggestions and recommendations for amendments should be forwarded to:

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Approved citation: Animal Health Australia (2005). Disease Strategy: Bovine spongiform encephalopathy (Version 3.1). Australian Veterinary Emergency Plan (AUSVETPLAN), Edition 3, Primary Industries Ministerial Council, Canberra, ACT.

Publication record:
Edition 1: BSE was not included in this edition
Edition 2:
Version 2.0, 1996 (new manual)
Version 2.1, 1998 (update to include new disease information from the UK etc)
Edition 3:
Version 3.0, January 2003 (major update of text and policy, and inclusion of new cost-sharing arrangements)
Version 3.1, January 2005 (update)

AUSVETPLAN is available on the internet at:
http://www.animalhealthaustralia.com.au

ISBN 0 642 24506 1 (printed version)
ISBN 1 876 714387 (electronic version)

IMPORTANT NOTE: Important regulatory information is contained in the OIE Terrestrial Animal Health Code for bovine spongiform encephalopathy, which is updated annually and is available on the internet at the OIE website: http://www.oie.int/eng/normes/en_mcode.htm. Further details are given in Appendix 3 of this manual).

DISEASE WATCH HOTLINE

1800 675 888

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

Preface

This disease strategy for the control and eradication of bovine spongiform encephalopathy (BSE) is an integral part of the **Australian Veterinary Emergency Plan**, or **AUSVETPLAN (Edition 3)**. AUSVETPLAN structures and functions are described in the **AUSVETPLAN Summary Document**.

This strategy sets out the revised disease control principles that were approved by the Animal Health Committee of the Primary Industries Ministerial Council (PIMC) out-of-session in January 2003 for use in an animal health emergency caused by the occurrence of BSE in Australia. Relevant livestock industries have also been involved in the consultation and approval process (see below).

BSE is a disease listed by the World Organisation for Animal Health (OIE, formerly Office International des Epizooties). Listed diseases are 'communicable diseases which are considered to be of socioeconomic and/or public health importance within countries and which are significant in the international trade of animals and animal products'. The principles contained in this document for the diagnosis and management of an outbreak of BSE conform with the *OIE Terrestrial Animal Health Code* (see Appendix 3).

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).[1]

Category 2 diseases are emergency animal diseases that have the potential to cause major national socioeconomic consequences through very serious international trade losses, national market disruptions and very severe production losses in the livestock industries that are involved. Category 2 also includes diseases that may have slightly lower national socioeconomic consequences, but also have significant public health and/or environmental consequences. For this category, the costs will be shared 80% by governments and 20% by the relevant industries (refer to the EAD Response Agreement for details).

Detailed instructions for the field implementation of AUSVETPLAN are contained in the disease strategies, operational procedures manuals, management manuals and wild animal manual. Industry-specific information is given in the relevant enterprise manuals. The full list of AUSVETPLAN manuals that may need to be accessed in an emergency is:

[1]Some information on the EAD Response Agreement can be found at:
http://www.animalhealthaustralia.com.au/programs/eadp/eadra.cfm

Disease strategies
- Individual strategies for each disease

Operational procedures manuals
- Decontamination
- Destruction of animals
- Disposal procedures
- Public relations
- Valuation and compensation

Management manuals
- Control centres management (Volumes 1 and 2)
- Animal Health Emergency Information System
- Laboratory preparedness

Enterprise manuals
- Animal quarantine stations
- Artificial breeding centres
- Aviaries and pet shops
- Feedlots
- Meat processing
- Poultry industry
- Saleyards and transport
- Veterinary practices
- Zoos

Wild animal manual
- Wild animal response strategy

Summary document

In addition, *Exotic Diseases of Animals: A Field Guide for Australian Veterinarians* by WA Geering, AJ Forman and MJ Nunn, Australian Government Publishing Service, Canberra, 1995 is a source for some of the information about the aetiology, diagnosis and epidemiology of the disease.

Earlier versions of this manual were prepared by writing groups with representatives from the Australian national, state and territory governments and industry. For Version 3.1, the document was reviewed by Reg Butler, Australian Government Department of Agriculture, Fisheries and Forestry. Scientific editing was by Dr Janet Salisbury of Biotext, Canberra.

The revised manual has been reviewed and approved by Animal Health Australia and its relevant members:

Government
- Commonwealth of Australia
- State of New South Wales
- State of Queensland
- State of South Australia
- State of Tasmania
- State of Victoria
- State of Western Australia
- Northern Territory
- Australian Capital Territory

Industry
- Cattle Council of Australia
- Australian Lot Feeders Association
- Australian Dairy Farmers' Federation

The National Health and Medical Research Council Special Expert Committee on Transmissible Spongiform Encephalopathies has also reviewed this manual.

The complete series of AUSVETPLAN documents is available on the internet at: http://www.animalhealthaustralia.com.au

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1 Nature of the disease

Bovine spongiform encephalopathy (BSE) is a disease of cattle, strongly linked to a similar disease of people. It is therefore of concern not only for the welfare of animals, but also as a food safety issue. As a consequence, any outbreak will involve veterinary authorities, health authorities and food safety agencies.

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. The disease is one of the transmissible spongiform encephalopathies (TSEs) or 'prion' diseases. These diseases 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 transmissible forms (Prusiner 1998).

1.1 Aetiology

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

Although the prion hypothesis is compelling, other aetiological possibilities are still under consideration. They include a robust virus or a virino, which is a nucleic acid protected by host protein. Evidence that environmental factors or toxic chemicals cause BSE is unconvincing.

It is apparent that the BSE epidemic in the UK resulted from the feeding of meat-and-bone meal (MBM) contaminated with the BSE agent to cattle. However, the ultimate origin of the BSE agent itself is uncertain (Collee and Bradley 1997ab, Brown et al 2001). Hypotheses under consideration include a cross-species transmission of the prion responsible for scrapie in sheep or a novel prion arising in cattle or some other mammalian species (Horn Committee Report, UK DEFRA 2001[2]).

A particular feature of prions that accumulate in the bodies of animals with prion diseases is their resistance to inactivation by physical or chemical procedures. These include freezing, desiccation, ultraviolet (UV) radiation, burial, the usual methods for chemical and heat disinfection, and degradation by certain proteolytic enzymes (Taylor DM 1996ab; Taylor K, 1996).

[2] http://www.defra.gov.uk/animalh/bse/publications/bseorigin.pdf

1.2 Susceptible species

Domestic cattle

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

Wild bovids and cats

During the BSE epidemic in cattle in the UK, a spongiform encephalopathy was also identified in various zoo species such as antelopes, 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. Bioassay studies in mice in several of these cases produced a characteristic incubation period and profile of neuropathological changes indicating that the aetiological agent was the BSE strain. Affected bovid species had received MBM as a dietary supplement, and the exotic felid species were fed bovine carcases including spinal cord.

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

Small ruminants

Sheep have been experimentally infected with BSE. The disease agent had a tissue distribution in the infected animals similar to scrapie (the sheep TSE). 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 first BSE case confirmed in a naturally infected small ruminant animal was in a French goat in 2005. If BSE does occur in sheep in the UK, it is likely to occur at a very low prevalence (Kao et al 2002).

Pigs

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

Chickens

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

Dogs and horses

No cases have been reported in dogs or horses.

Primates

Various primate species, including common marmosets, macaques and lemurs, have proved susceptible to BSE in experiments.

Humans

Creutzfeldt–Jakob disease (CJD) is a TSE that affects humans. Most cases arise spontaneously with no known cause (*sporadic CJD*), with an even annual incidence worldwide of one case per million. 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 by means of 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 central nervous system and is always fatal. It is believed to be associated with the consumption of 'specified risk materials' (like brain and spinal cord) from BSE-affected cattle. In subsequent laboratory studies, the pathological agents isolated from BSE-infected 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, further cases have occurred in the UK, France, Italy and the Republic of Ireland. Cases have occurred in Canada, the United States and some non-European countries in individuals who lived in Europe for extended periods. This has led to concerns about an impending epidemic of the disease. However, it appears likely that the number of cases will be smaller than originally predicted (Brown 2001, Ghani et al 2003). Up-to-date information on the incidence of vCJD can be obtained from the internet site of the United Kingdom CJD Surveillance Unit.[3]

1.3 Worldwide distribution and occurrence in Australia

BSE was first diagnosed in England in 1986, and its annual case incidence in the UK peaked in 1992. Although the great majority of cases have occurred in cattle in the UK, smaller-scale epidemics, linked to the export of live cattle and MBM from that country and subsequently from other BSE-infected countries, have occurred elsewhere in Europe. Increasing numbers of cases have been detected in other countries of Europe since 1999, when more sensitive surveillance programs were introduced. In 2001, the first cases in cattle born outside Europe were confirmed in Japan. In 2003, Canada reported its first indigenous case of BSE. Table 1 shows the distribution of worldwide cases up to 21 November 2004. Up-to-date information can be obtained from the OIE website.[4]

[3] http://www.cjd.ed.ac.uk
[4] http://www.oie.int/eng/info/en_esb.htm

Table 1 Number of reported cases of BSE worldwide until the end of 2004

Country	2001 and before	2002	2003	2004	Total
Austria	1	0	0	–	1
Belgium	65	38	15	11	129
Canada[a]	1	0	2[b]	1	4
Czech Republic	2	2	4	7	15
Denmark[a]	8	3	2	1	14
Finland	1	0	0	–	1
France[a]	515	239	137	54	945
Germany[a]	138	106	54	59	357
Greece	1	0	0	–	1
Ireland (Rep)[a]	833	333	183	126	1475
Israel	0	1	0	0	1
Italy[a]	50	38	29	7	124
Japan	3	2	4	5	14
Liechtenstein	2	0	–	–	2
Luxembourg	1	1	0	0	2
Netherlands	28	24	19	6	77
Poland	0	4	5	11	20
Portugal[a]	640	86	133	92	951
Slovakia	5	6	2	7	20
Slovenia	1	1	1	2	5
Spain	84	127	167	131	509
Switzerland	403	24	21	3	451
United Kingdom including Northern Ireland	182,044	1144	612	338	184,138
Total	**184,826**	**2179**	**1390**	**861**	**189,256**

– = data not available

a Includes imported animals

b One case diagnosed in Canada in May 2003 + 1 case diagnosed in the United States in December 2003 and confirmed as having been imported from Canada.

Note: Cases have also been reported, in imported animals only, in the Falkland Islands (1 case in 1989) and Oman (2 cases in 1989).

Source: OIE website (http://www.oie.int/eng/info/en_esb.htm), March 2005

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 BSE strain. 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. 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. The carcase was disposed of in an approved manner.

1.4 Diagnostic criteria

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

A diagnosis of BSE in cattle, sufficient to initiate an emergency response, should include the following:

- a suggestive history, including disease history, management history and fixed history or signalment (breed, age etc) of the animal;
- consistent clinical signs;
- evidence for opportunity of exposure to the BSE agent;
- neurohistological changes characteristic of a spongiform encephalopathy; and
- immunohistochemical and immunochemical results showing the accumulation of abnormal prion protein (PrP^{Sc}).

A complete diagnosis of the TSE as BSE of the type observed in the UK and continental Europe would result from a mouse bioassay demonstrating infectivity associated with an incubation period and lesion profile consistent with the BSE agent. Less time-consuming assays may arise from further research on prion diseases.

The *Australian and New Zealand Standard Diagnostic Protocols – TSEs* (ANZSDP–TSEs 2000)[5] provide the authoritative guide to laboratory diagnosis and contain methods that are consistent with the current edition of the OIE *Manual of Standards for Diagnostic Tests and Vaccines for Terrestrial Animals* (OIE Manual; see Appendix 3). Laboratory examination of brain and spinal cord samples collected at postmortem examination is essential to confirm a suspected case of BSE.

1.4.1 Clinical signs

Due to the long incubation period, signs usually appear when animals are between 2 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; they represent 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 hind limbs. Other abnormalities of posture and movement 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.

[5] http://www.aahc.com.au/surveillance/ntsesp/asdt00.pdf

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. It is noteworthy that a higher incidence of BSE has been found in Europe in emergency slaughter cattle than in animals passing preslaughter inspection; when BSE has been diagnosed in either circumstance, there is often a history of overlooked clinical signs of BSE.[6]

1.4.2 Pathology

Gross lesions

There are no gross changes in BSE.

Microscopic lesions (histopathology)

The characteristic histological TSE changes in the central nervous system (CNS) are vacuolation of grey matter neuropil (spongiform change) and/or vacuolation of neurons, astrocytosis and neuronal degeneration. In cattle with BSE, these changes have a predilection for 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 prion protein (PrP) can be demonstrated within these lesions. Further details are contained in the *Australian and New Zealand Standard Diagnostic Protocols – TSEs* (ANZSDP–TSEs 2000) and in the OIE Manual (see Appendix 3).

1.4.3 Laboratory tests

Specimens required

The range of samples and the methods of sample collection, preservation and submission are described in the *National Guidelines for Field Operations,* first published in 2000 and updated in February 2004.[7] 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, by detection of abnormal fibrils in the brain using transmission electron microscopy, or by bioassay in mice (see Table 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

[6] The video, *A Tale of Transmission* (see Video and training resources in References), clearly demonstrates the clinical signs of BSE.
[7] http://www.aahc.com.au/tsefap/NTSESP%20FIELD%20OPS04.pdf

specimens. However, in the case of strong clinical suspicion of BSE, every effort should be made to collect undamaged brain and cord samples. The videos, *Prionics Test Trial Program – Methods for removing brains for testing with the Prionics rapid BSE test* and *National TSE Surveillance Program – Methods for removing brains for TSE testing,* show how to remove the appropriate specimens (see Video and training resources in References 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 must be packed according to transportation regulations, and the laboratory advised well in advance of specimen arrival times and conditions. Unfixed samples of cervical spinal cord and/or medulla caudal to the obex can be transferred chilled (packed with sufficient cooler bricks) to the laboratory, where they will be held frozen pending forwarding to the CSIRO Australian Animal Health Laboratory (CSIRO-AAHL) in Geelong. Formalin-fixed tissues including brain should be securely packed in leak-proof containers.

Specimens should initially be sent to the state or territory diagnostic laboratory, from where they will be forwarded to CSIRO-AAHL for emergency disease testing after obtaining the necessary clearance from the chief veterinary officer (CVO) of the state or territory of the disease outbreak and informing the CVO of Victoria about the transport of the specimens to Geelong.

Laboratory diagnosis

Laboratory examination of brain is necessary to confirm a diagnosis of BSE. Histological examination to detect the characteristic changes in the CNS mentioned in Section 1.4.2 is the first step because it may also provide an alternative diagnosis and thus conclude an investigation. According to the OIE Manual (see Appendix 3), 'the correlation between the clinical diagnosis and the neurohistological diagnosis in BSE can, with appropriate experience, be greater than 90%'.

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

- Detection of so-called 'scrapie-associated fibrils' (SAFs) by electron microscopy. SAF detection gives results similar to histological examination and can be used on autolysed tissue (Cooley et al 2001).
- Immunohistochemistry on formalin-fixed sections of CNS. This uses specific antibody 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 and more are under development. Western blotting, also known as immunoblotting, is available in Australia. Antibody specific for PrP is employed and PrP and PrP^{Sc} are distinguished through an enzyme digestion step designed to remove PrP. Western blotting is based on electrophoresis and has the capacity to distinguish the molecular weight and the pattern of glycosylation of PrP^{Sc}.

Although confirmation of a TSE in cattle by immunohistochemistry or immunochemistry is sufficient to initiate an emergency response, at the current stage of test development an absolutely definitive diagnosis of BSE would require mouse transmission tests. These tests 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 (OIE Manual 2000; see Appendix 3). Table 2 shows the tests for BSE that are currently available in Australia. Additional tests will be evaluated progressively in Australia following their evaluation overseas.

Table 2 Laboratory tests currently available at CSIRO–AAHL for the diagnosis of BSE

Test	Specimen required	Test detects	Time taken to obtain result
Histopathology	Formalin-fixed brain tissue	Vacuolation of grey matter neuropil (spongiform change) and/or vacuolation of neurons; astrocytosis; neuronal degeneration	2 days
Immunohisto-chemistry	Formalin-fixed brain tissue or cervical spinal cord	Excessive accumulation of PrP	1 day
Immunochemistry – Prionics® Immunoblot[a]	Unfixed brain tissue containing obex or cervical spinal cord	Accumulation of abnormal PrP (PrP^{Sc})	1 day
Electron microscopy[b]	Unfixed brain tissue or cervical spinal cord	Scrapie-associated fibrils (SAFs)	2 days
Isolation of agent by intracerebral inoculation into mice[c]	Unfixed brain tissue or cervical spinal cord	BSE agent	Up to and beyond 1 year

a This test may be of value in preclinical diagnosis late in the incubation period. Currently under evaluation for use at AAHL. Requires confirmation by immunohistochemistry in the case of positive or doubtful results.
b This test is almost obsolete.
c This test is performed to confirm transmissibility of a spongiform encephalopathy and to strain-type the causative agent.
Source: Information provided by CSIRO-AAHL, 2002 (refer to AAHL for most up-to-date information).

1.4.4 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 Australian 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/selenium deficiency)
- metabolic diseases
  - hypomagnesaemia/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 (BHV1.3/BEHV)
  - sporadic bovine encephalomyelitis (SBE)
  - bovine malignant catarrhal fever (BMCF)
  - bovine ephemeral fever
  - focal symmetrical encephalomalacia (*Clostridium perfringens*)
- toxicoses
  - lead toxicosis
  - plant toxicoses
    - perennial rye grass staggers (*Acremonium* sp, endophyte on *Lolium perenne*)
    - annual rye grass staggers, blown grass staggers/flood plain staggers (*Clavibacter* sp 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)
  - cerebellar abiotrophy (Angus)
  - progressive ataxia (Charolais)
  - progressive spinal myelinopathy (Murray Grey)
  - neuronal ceroid-lipofuscinosis (Devon)
  - tomaculous-like neuropathy (Santa Gertrudis)
- neoplasia.

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

Also, in cases of progressive chronic wasting and/or an inability to stand in mature cattle, the possibility of the BSE agent being implicated should be considered.

1.4.5 Treatment of infected animals

There is no treatment for animals with TSEs.

1.5 Resistance and immunity

1.5.1 Innate and passive immunity

There is no evidence for passive immunity playing any part in resistance to BSE. 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. Ram selection can be used to control the incidence of clinical scrapie. In cattle, no such genetic factors affecting susceptibility to BSE have yet been identified. Up-to-date information on innate resistance to TSEs is given in *Review of the Origin of BSE* (UK DEFRA 2001).[8]

1.5.2 Active immunity

The disease is fatal in all cases, affected animals do not develop immunity, and no protective immunological response has been detected.

1.5.3 Vaccination

There is no vaccine for BSE.

1.6 Epidemiology

The epidemiology of BSE in cattle is determined principally by its long incubation period and its mode of transmission, which so far has been attributed solely to ingestion at a young age of BSE-contaminated MBM (Collee and Bradley 1997ab, Wilesmith 1998, Brown et al 2001). While BSE does not have a known sporadic incidence in cattle as CJD does in humans (see Section 1.2), the occurrence of a TSE in either a sporadic or familial form in cattle is a reasonable hypothesis. However, there is no evidence to date of sporadic cases of TSE occurring outside the UK in the large cattle populations of the world. All BSE cases in countries other than the UK have origins in the importation and feeding to young cattle of MBM, or the importation of live cattle that entered that the animal feed chain, originating from the UK. The likelihood of a sporadic case arising in the Australian cattle population is extremely low.

1.6.1 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 six months of life, and the incubation period of BSE is long (the average incubation period is cited as five years). However, in the UK dataset from 1987 to 1997, 90% of cases occurred in animals from 3 to 8 years of age and 10% of cases occurred in animals aged 9 years and above. A small percentage of the BSE cases in the UK were recorded in animals older than 12 years. Only three clinically identified cases of BSE were recorded in cattle aged 30 months or younger in the two years before the 'over 30-month' rule was introduced in the UK

[8] http://www.defra.gov.uk/animalh/bse/publications/bseorigin.pdf

in 1996 (UK Food Standards Agency 2000). Under the rule, 'beef from cattle aged over thirty months at slaughter is banned for sale for human consumption in the UK', with the exception that 'meat from cattle on registered very low BSE risk assurance scheme herds may be sold for human consumption if the animal was no more than 42 months at slaughter'.

1.6.2 Persistence of agent

General properties

Horizontal transmission was not regarded as a major factor in the UK epidemic of BSE. However, residues of contaminated MBM stored on farm and fed to cattle after 1996 may be responsible for the continuing trickle of BSE cases in animals born after the feed ban. Because of their peculiar protein structure, prions are resistant to freezing, desiccation, ultraviolet radiation, most disinfectants and burial. Postmortem, CJD infectivity can persist for 28 months at room temperature. On the other hand, prions are susceptible to extremes of pH and some organic acids, which inactivate their infectivity. Failure to demonstrate horizontal transmission of BSE to date indicates that environmental contamination is not a major factor affecting the spread of disease.

Live animals

The movement of clinically normal but infected cattle is a risk factor for the introduction of BSE into new areas if there is no prohibition on the feeding of ruminant-derived tissues to ruminants and if rendered material from such animals enters the cattle feed supply. This risk applies during the period of infectivity of tissues from such animals, which begins shortly before the appearance of clinical signs (Wells et al 1998). Experimental studies have shown a close temporal (time) association of the onset of infectivity, detection of abnormal PrP and typical histological lesions in CNS tissue at a late stage of the incubation period.

The distribution of infectivity in the tissues of BSE-affected cattle has been reviewed in a number of publications, and a detailed account is included in a report and opinion of the Scientific Steering Committee (SSC) of the European Commission (EC), January 2002 (SSC 2002).[9] Table 3 shows estimates of the doses needed to orally infect 50% of exposed cattle (Co ID50) for each tissue at the height of infectivity for that tissue (SSC 1999b).[10]

[9] http://europa.eu.int/comm/food/fs/sc/ssc/out241_en.pdf
[10] http://europa.eu.int/comm/food/fs/sc/ssc/out67_en.html

Table 3 European Commission SSC estimate of cattle oral infective dose (Co ID50) with each tissue at the height of infectivity for that tissue type.

Tissue	Co ID50 per BSE case	Total infective load per animal (%)
Brain	5000	64.1
Spinal cord	2000	25.6
Trigeminal ganglia	200	2.6
Dorsal root ganglia	300	3.8
Ileum	260	3.3
Spleen[a]	26	0.3
Eyes	3	0.04

a Some data suggest that the extrapolation from scrapie to BSE is not valid and that spleen is unlikely to be infective (see Section 1.6.2, Live animals).

Source: Opinion of the European Commission SSC: Human exposure risk (HER) via food with respect to BSE, 10 December 1999, page 11. (Also reproduced in the Joint WHO/FAO/OIE *Technical Consultation on BSE: Public Health, Animal Health and Trade*, 2001, page 7 (see Further reading and links in References).

Cattle with BSE differ from other animals with other TSEs in that infectivity in the lymphoreticular system is slight and located in Peyers patches and perhaps bone marrow and spleen. Unpublished data quoted by the European Commission SCC (SSC 2002) suggests that infectivity of splenic tissue is at least less than 1, and possibly as low as 0.1, cattle intracerebral LD50 per gram (lethal dose for 50% of cattle inoculated). 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. Trace infectivity was found in sternal bone marrow at 38 months after exposure.

In naturally occurring clinical cases of BSE, infectivity is concentrated in neural tissue such as brain, retina, spinal cord, and dorsal root and trigeminal ganglia (Wells et al 1998, 1999). Abnormal PrP first appeared in the CNS at 32 months after exposure, which coincided with the earliest detection of infectivity by bioassay in mice and preceded evidence of typical histological BSE changes in the brain and clinical disease at 36, 38 and 40 months after exposure. Infectivity was detected in the dorsal root and trigeminal ganglia at 32–40 months after exposure.

Up-to-date information on the above and other BSE research can be obtained from the UK Department of Environment, Food and Rural Affairs website.[11]

Animal products and byproducts

The BSE agent survives for long periods in carcases and withstands many of the procedures currently involved in the processing of product. Decontamination is discussed in Section 2.2.9. Preliminary evidence demonstrates that TSE agents can be effectively inactivated by alkaline hydrolysis (Taylor et al 1999).

[11] http://www.defra.gov.uk/animalh/bse/index.html

Veterinary instruments

As an aberrant protein, the BSE agent is very resistant to 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. Methods of decontamination are described in Section 2.2.9.

1.6.3 Modes of transmission

Live animals

Cattle

BSE is not a contagious disease in the usual sense and there is no convincing evidence for horizontal spread of BSE between cattle, either directly or indirectly. This is consistent with the restriction of its infectivity to CNS tissue and is further 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.

There is only circumstantial evidence of vertical transmission of BSE from dam to calf, and only at a level that is not sufficient to perpetuate the disease. The European Commission SSC (SSC 1999a) drew the following conclusion:

> There is an enhanced risk of approximately 10% of BSE in offspring born to BSE-affected dams. The results of all epidemiological studies undertaken to date have been consistent with a rate of direct maternal transmission of approximately 10% in calves born to dams within 12 months of onset of clinical signs of BSE, with lower rates up to 24 months before the onset of clinical signs in the dam. Enhanced genetic susceptibility cannot be excluded on the basis of these data but such genetic susceptibility at present is only speculative. On the basis of these data the UK SEAC [Spongiform Encephalopathy Advisory Committee] concluded that there is some evidence of direct maternal transmission at a low level but they cannot rule out variation in genetic susceptibility to feedborne infection as an additional factor [UK SEAC 1996]. It is thus still unclear if maternal transmission of BSE in cattle in the traditional sense occurs or not and if it does the mechanism involved.

Wrathall et al (2002) have completed an extensive study in which 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.

Further information on maternal transmission is included in the DEFRA website BSE: Science and Research — Epidemiology at:
http://www.defra.gov.uk/animalh/bse/science-research/epidem.html#spread

Most of the epidemiological evidence indicates that cattle become infected with BSE when they are calves (Donnelly and Ferguson 2000). On the basis of a computer simulation model, Wilesmith et al (1988) demonstrated that the risk of

exposure for calves was 30 times greater 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 1995–96 peak followed the feed ban in 1988, taking into consideration the 5-year incubation period. Wilesmith et al (1988) also demonstrated that most animals were infected in the first 6 months of life.

Sheep

The question of BSE in sheep arises because sheep and cattle in the UK were fed the same contaminated MBM that drove the BSE epidemic in cattle. There is concern that if BSE is present in sheep, it may behave like scrapie and spread from infected ewes around the time of lambing. There are, however, significant differences in the clinical expression of BSE and scrapie in sheep that may well preclude this spread.

At the time of writing, insufficient samples have been tested to rule out the possibility of BSE in the UK sheep flock. However, if BSE does occur in sheep in the UK, it is likely to occur at a very low prevalence (Kao et al 2002).

Animal products and byproducts

The potential transmission of BSE by animal products and byproducts has been reviewed and documented by Food Standards Australia New Zealand (FSANZ).

Good information is available on the BSE infectivity of animal products and byproducts. The starting point is the distribution of infectivity in the tissues and excretions of cattle as demonstrated by mouse transmission studies with bovine material obtained from natural and experimental infections. Infectivity of tissues from cattle with natural BSE was found in brain, spinal cord and retina. It was not found in fractions of blood, bone marrow, milk, cerebrospinal fluid, fat, alimentary tract, heart, kidney, pancreas, liver, lung, spleen, tonsil, lymph nodes, muscles, peripheral nerves, skin, trachea or reproductive tracts, including embryos and semen (Fraser et al 1988, Dawson et al 1990, Fraser et al 1992, Middleton and Barlow 1993, Taylor et al 1995a, Wrathall 1997). A study of 44 tissues from experimentally infected cattle showed a distinct anatomical distribution of BSE infectivity in cattle depending upon whether the disease was in the incubation stage or was clinically expressed (Wells et al 1994, 1998, 1999). Infectivity was found in the distal ileum during the first 6–18 months after experimental infection. During the period of clinical onset and disease, infectivity was found in brain, spinal cord and trigeminal and dorsal root ganglia. At this time, low or questionable infectivity was found in bone marrow but not in leucocyte fractions or thymus.

Information on the infectivity of various bovine tissues determines the controls on specified risk materials (SRMs) that operate in BSE-affected countries to prevent those parts of cattle likely to contain the BSE agent from entering the human food or animal feed chains. Internationally, SRMs are defined differently according to both cattle age and tissue type.

SRMs as defined in the UK are the tonsils, intestine from the duodenum to the rectum and the mesentery in animals of all ages. The entire head (excluding the tongue, but including the brain, eyes, trigeminal ganglia), thymus, spleen and

spinal cord in animals over 6 months of age at slaughter. The vertebral column, excluding the vertebrae of the tail, the spinous and transverse processes of the cervical, thoracic and lumbar vertebrae and the median sacral crest, the wings of the sacrum, but including the dorsal root ganglia in animals aged over 30 months of age at slaughter.
http://www.defra.gov.uk/animalh/bse/general/qa/section6.html#q6

SRMs are defined by FSANZ in its 18 July 2001 document *Bovine Spongiform Encephalopathy (BSE): Human health requirements for the importation of beef and beef products* as: the skull, brains, eyes, the tonsils, vertebral column and spinal cord, including dorsal root ganglia, of bovine animals aged over 12 months; and the intestines from the duodenum to the rectum of bovine animals of all ages.
http://www.foodstandards.gov.au/whatsinfood/bovinespongiformencephalopathybse/bovinespongiformence713.cfm

The European Commission SSC has produced a set of considered and authoritative opinions on bovine products and byproducts in countries where BSE occurs, as follows.

- There is no reason to restrict the use of milk. However, milk from BSE-affected cows should be kept out of the human food supply as a precautionary measure (SSC 1999a).
- BSE risk can be considered negligible from dicalcium phosphate if it has been sourced from animals fit for human consumption, after SRM removal and provided production has been carried out to an appropriate standard. This production involves several steps, an example of which includes crushing and degreasing in hot water, submitting over 4–5 days to increasing concentrations of hydrochloric acid, alkaline treatment with lime, and storing and drying at specific temperatures (SSC 2003a). For more details, see:
http://europa.eu.int/comm/food/fs/sc/ssc/out322_en.pdf
- Gelatine acquired from ruminant hides does not present a risk with regard to BSE, provided contamination with potentially infected materials is avoided. BSE risk is higher with gelatine sourced from ruminant bones than from hides. Negligible risk can be attained for gelatine from bones via sourcing and production conditions related to those required for the production of dicalcium phosphate (SSC 2003b). For more details, see:
http://europa.eu.int/comm/food/fs/sc/ssc/out321_en.pdf
- There is no evidence that tallow derived from ruminant animals constitutes a BSE risk. Tallow derived from discrete fat tissues in animals fit for human consumption can be used for all applications. Tallow from other tissues in such animals can be safely used for feed and petfood if it is purified to less than 0.15% insoluble impurities (SSC 2001a). For more details, see:
http://europa.eu.int/comm/food/fs/sc/ssc/out219_en.pdf
- Purified tallow derivatives that do not contain proteins or peptides can be considered safe, provided a) the raw material is safe for human or animal consumption or b) the production process uses the appropriate, validated and scientifically most up-to-date methods of inactivating the BSE agent. Risk is modulated according to the source of the tallow derivatives and the BSE risk categorisation of the relevant country (SSC 2003c). For more details, see:
http://europa.eu.int/comm/food/fs/sc/ssc/out359_en.pdf

- Those parts of ruminant hides used for the production of collagen do not present a BSE risk if contamination with potentially infective material is avoided (SSC 2001a).
- Hydrolysed protein can be considered safe if the raw material from which it is obtained does not contain BSE infectivity (SSC 2000).

Due to the lack of evidence for horizontal transmission of the BSE agent, there is no reason to consider that bovine faeces or urine pose a risk for BSE transmission. A risk assessment on CJD in humans indicates that faeces, urine and other body fluids do not transmit the disease (Brown et al 1994).

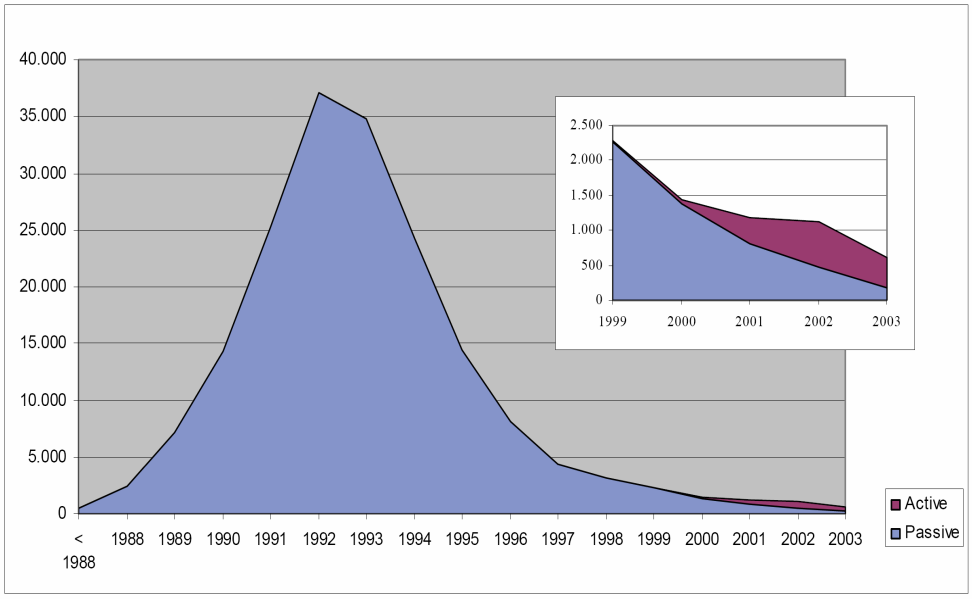

Figure 1 Evolution of BSE detected by passive surveillance and active monitoring in the United Kingdom

From *Report on the monitoring and testing of ruminants for the presence of transmissible spongiform encephalopathy (TSE) in the EU in 2003, including the results of the survey of prion protein genotypes in sheep breeds*, European Commission, Brussels, May 2004, pp 12–13.
http://europa.eu.int/comm/food/food/biosafety/bse/annual_report_tse2003_en.pdf

After the 1988 ban on feeding ruminant-derived MBM to ruminants, the 1990 ban on specified bovine offal for use in animal nutrition and the more comprehensive 1996 ban on feeding mammalian-origin MBM to any farmed animal species, the annual incidence of BSE in the UK peaked in 1992 (37 280 cases) and has declined steeply since, as shown in Figure l. Since 2001, the number of BSE cases has continued to fall sharply in the UK, with 1144 cases recorded in 2002, 612 in 2003 and only 338 in 2004.

Artificial breeding

Early evidence suggested that the BSE agent is not transmitted in semen or embryos (Wrathall 1997). The studies are now complete and the results of extensive testing demonstrate no transmission (Wrathall et al 2002).

Biological products

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 BSE-affected cattle therefore provide a possible route of transmission of this disease and must be considered during disease investigations. While they pose a theoretical threat, there is no epidemiological evidence to suggest that they have been a source of BSE in the UK or elsewhere. Quarantine controls are in place in Australia against the importation of such biological products. Examples are veterinary vaccines and catgut sutures.

Veterinary instruments

Surgical and veterinary instruments have not been a recognised method of BSE spread to cattle in the UK outbreak. 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. Surgical instruments used for procedures with CNS exposure (eg eye ablation) may also be contaminated if the animal is incubating BSE, but such procedures are rare and this form of transmission is very unlikely. Methods of decontamination are described in Section 2.2.9.

Other equipment and materials

There is no evidence that other equipment or materials have a role in spreading BSE.

Vectors

There is no documented evidence for transmission of BSE by arthropod vectors. However, there is ongoing research in the UK to determine if transmission by external or internal parasites is possible.

1.6.4 Factors influencing transmission

The principal factor influencing the transmission of BSE to date is the feeding of MBM contaminated with the BSE agent. Australia has prohibited the feeding of MBM and other compounded feeds containing vertebrate materials to ruminants, and this is audited. Indeed, most cattle in Australia are raised in grass-fed production systems, which greatly reduces any chance of transmission of the BSE agent in feed.

1.7 Manner and risk of introduction to Australia

Key factors in the epidemiology of BSE are well established. They point to three pathways for the introduction of the disease agent into Australia from BSE-affected countries.

1. **Importation of animals of the family Bovidae from BSE-affected countries.** Importation of bovids into Australia from the UK ceased in 1988 and from continental Europe in 1991. The cattle and buffalo that are still alive from the limited number of previous imports have been placed under lifetime quarantine. They have been permanently identified with rumen boluses so that they will never enter the human food or animal feed chains and will be disposed of safely at the ends of their lives. There is a very slight risk that these animals were exposed to the BSE agent. Given knowledge of the incubation period of BSE, there remains a low and diminishing risk that they may be infected. Similar action has been taken for cattle imported from non-European countries that were subsequently BSE affected.

2. **Importation of contaminated feedstuff originating from BSE-affected countries**. The importation of animal-derived MBM (except for fishmeal) was banned in Australia in 1966 as a measure against the importation of anthrax spores. Although other stockfeeds have been imported over time, the theoretical risk of their cross-contamination with residual imported MBM is extremely low for Australia. In March 2001, the then Agricultural and Resource Management Council of Australia and New Zealand agreed to extend the ruminant feed ban to include material from all vertebrate animals (including fish). This was enacted in legislation between March 2001 and March 2002.

3. **Importation of biologicals contaminated with the BSE agent.** Quarantine risk assessments have been made of vaccines and other biologicals that involve bovine products in their manufacture. Restrictions on the importation of these products have been extended in line with emerging knowledge of the BSE status of countries throughout the world. This risk is considered to be extremely low for Australia.

Stringent controls are in place against the introduction of BSE through these three pathways. Furthermore, the legislated bans in Australia on the feeding to ruminant animals of MBM derived from mammals, birds or fish will prevent any BSE being propagated and amplified into an epidemic.

If a case of TSE is diagnosed in Australia, the possibility that it is a sporadic TSE and not BSE itself is a justifiable consideration. The possibility of a TSE in cattle appearing sporadically is indicated by the molecular biology involved in the prion hypothesis, and the behaviour of the TSEs as a disease class. Sporadic CJD in humans is a suitable analogy. However, the frequency of its occurrence (about one in a million in the human population worldwide) does not easily translate to cattle because of species differences, such as the different age structures of cattle and human populations. There is no assurance that a sporadic TSE would involve the same strain as that associated with BSE in the UK and Europe.

2 Principles of control and eradication

2.1 Introduction

Some clear principles for the control and eradication of BSE derive from consolidated experience with the disease and its control in the UK and Europe, as follows.

- BSE arose in the UK and was propagated through the recycling of bovine tissues into animal feed. Later, the export of infected animals and contaminated feed spread the BSE agent to other countries, where it was again recycled and propagated through the feed chain (SSC 2000).
- BSE has only been seen in a transmitted form spread through the feeding of MBM. BARB cattle ('born after the real feed ban') in the UK have been investigated and do not challenge the view that ingestion of infective agent is the key means of transmission (SSC 2003d, EFSA 2005).
- Regarding vertical transmission, semen and embryos have been evaluated and found to be ineffective vectors (SSC 1999a, Wrathall et al 2002).
- The possibility of sporadic or familial TSE is indicated by the molecular biology of prions, and the behaviour of the TSEs as a disease class. This theoretical possibility must be considered in Australia's contingency planning.
- The dose of infective agent required to initiate BSE in cattle is not known with certainty. It depends upon the infectivity titre in feed, the uneven distribution of infectivity as 'packets' throughout the feed and the impact of cumulative doses of the infective agent (UK BSE Inquiry, 2000; vol 2:101). Results of an attack-rate study begun in 1992 showed that infection could be transmitted by mouth with a dose as small as one gram of infected brain. The brain and spinal cord from an infected bovid may contain 5000 and 2000 cattle oral ID50s (doses that infect 50% of exposed cattle), respectively, whereas the ileum and spleen may contain 256 and 3 cattle oral ID50s, respectively (SSC 1998).

The BSE agent, like all TSE agents, has unusual resistance to the usual physical and chemical methods for disinfection and may persist in the environment. However, the BSE agent is not absolutely resistant and appropriate methods are available for disinfection when this is required.

2.2 Methods to prevent spread and eliminate pathogens

2.2.1 Veterinary investigations

To aid emergency disease management, the following four categories of cases and in-contact animals can be identified by laboratory and epidemiological investigations.

Clinically consistent animal	An animal that is found with clinical signs considered consistent with BSE.
Suspected case	An animal of the genus *Bos* (cattle) or genus *Bubalus* (buffalo) with history, clinical signs and histological changes consistent with BSE as described in Section 1.4, until an alternative diagnosis is substantiated. OR An animal with a positive result from a sensitive and specific screening test such as the Prionics test (see Section 1.4.3).
Confirmed case	A suspected case, or clinically consistent animal, that has been confirmed on postmortem by positive results from immunohistochemistry, immunochemistry, demonstration of characteristic fibrils by electron microscopy or by a test not previously performed on this case.
At-risk animal	An animal that has had a relevant association with suspected or confirmed cases of BSE (see below).

As BSE can only be definitively diagnosed at postmortem, clinically consistent cases must be killed as soon as possible in order for appropriate specimens to be taken and tested at an approved laboratory. Suspected and confirmed cases are, by definition, already dead. Material from a confirmed case would be sent to the recognised world reference laboratory (Veterinary Laboratory Agency, Weybridge, UK) for corroboration of the result.

The above definitions are based on the assumption that any TSE diagnosed in cattle in Australia will be a concern for animal health, human health and food safety regardless of whether it is the same as the BSE observed in the UK and Europe. Mouse transmission assays, which take a year or more to complete, are required to determine whether the diagnosed TSE is a novel disease or is classical BSE. These assays identify the pattern of distribution of lesions in the CNS that is distinctive for a particular strain of prion. It is an untested assumption that a sporadic or familial TSE would behave the same as classical BSE in the mouse transmission assay.

Identification of at-risk animals (epidemiological investigation)

Once BSE has been confirmed, animals that have an association with a confirmed case need to be identified and classified as *at-risk animals*. These are:

- any imported cattle originating from the same foreign property as a confirmed case;
- cohorts — animals born in the same calving season and raised with a confirmed case, or other cattle that may have been brought into contact with material containing the BSE agent;
- all progeny born from a confirmed BSE case within two years before or after the onset of clinical signs;
- the dam of a confirmed case;
- other ruminant animals, including sheep, goats, deer, camelids and elk, that have resided on the same property as a confirmed case at a time when they may have come into contact with material containing the BSE agent; and
- animals that may have had parenteral administration of a product containing the BSE agent (eg vaccine).

A recipient of semen, or an embryo derived from a confirmed male or female BSE case, is not considered to be at an increased risk of contracting the disease (Wrathall 1997, Wrathall et al 2002).

The disease control measures required for each of these four categories are described in Sections 2.2.2 to 2.2.8.

2.2.2 Quarantine and movement controls

Clinically consistent animals

Clinically consistent animals (as defined in Section 2.2.1) should be notified to a government veterinarian or animal health officer. BSE is a notifiable disease in all Australian states and territories.

The animal should be euthanased and the relevant brain and spinal cord samples sent for diagnosis. It is unlikely that movement controls will be placed on the property of origin.

Suspected cases

Suspected cases (as defined in Section 2.2.1) should be notified to a government veterinarian or animal health officer. BSE is a notifiable disease in all Australian states and territories.

To prevent further spread of possibly infected animals, movement of susceptible animals should be restricted until the results of clinical and epidemiological investigations are known or until confirmatory testing of the brain and other tissues sent to an approved laboratory returns negative results.

Confirmed cases

If the laboratory result is positive, the premises with the infected animal should be declared an infected premises (IP).

Other premises that may contain any potentially infected animals (see Section 2.2.1), should be identified as suspect premises (SPs). All IPs and SPs should be placed under quarantine to prevent movement of all susceptible animals, animal products and materials associated with the confirmed case. Any private veterinary practitioner servicing the premises should be notified and kept informed.

At-risk animals

Further action depends on the outcome of veterinary investigations to identify the risk status of animals and materials associated with a confirmed case (See Section 2.2.1). This would begin with obtaining a complete history of feeding practices and identification of all premises where the confirmed cases had lived from birth to diagnosis. Subsequently, any potentially infected animals, animal products, feedstuffs and biological materials would be traced (see Section 3.2).

The recommended approach to possible actions is outlined in Table 4 (Section 3.2). Long-term or lifelong quarantine of some animals identified by these investigations will be considered.

Zoning

Zoning, such as is implemented for control of other emergency diseases, is unlikely to be appropriate for BSE because, when first detected, the disease would be expected to be confined to one or, at most, a few properties.

2.2.3 Tracing

Tracing must be undertaken to:

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

In the event of the occurrence of one or more cases, there will be tracing and isolation of:

- any suspected or confirmed case, back to all premises where it has resided since birth;
- all at-risk animals associated with a confirmed case (see Section 2.2.1);
- all carcases and feedstuffs associated with the confirmed case, including rendered products; and
- all suspected feedstuff and biological materials.

The current manager and the manager responsible during the year of birth of the affected animals should be interviewed. The standard questionnaire in Appendix 6 can be used as a guide for these interviews.

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 in cattle observed there during at least the previous five years.

2.2.4 Surveillance

A systematic program of inspection and examination of at-risk animals will be required to determine the presence or absence of BSE and will build on the National TSE Surveillance Program. It will extend the intensity of surveillance beyond that required to meet a '99% probability of detecting BSE or scrapie if they accounted for 1% of the cases of neurological disease in cattle and sheep in Australia'. The program might need to be maintained for a prolonged period, probably for the life of these animals.

A basic program will include:

- regular, careful examination of all susceptible animals to detect the development of characteristic clinical signs, until the animals are killed;
- identification, by the National Livestock Identification System (NLIS) of all animals under long-term surveillance, including approved RFID ear tags or rumen boluses with details entered in the NLIS database;
- sampling and testing of each at-risk animal for BSE when it is killed.

Information on specimen collection and diagnosis is given in Section 1.4.3.

2.2.5 Treatment of infected animals

Treatment of BSE-infected animals is not effective.

2.2.6 Destruction of animals

Clinically consistent animals must be killed as soon as possible in order for appropriate specimens to be taken and tested at an approved laboratory. Such cases must not be sent for slaughter at abattoirs, knackeries or anywhere else where infected material could enter the human food or animal feed chains.

Killing of cases on farm is preferred to transporting them for slaughter at another site. Killing on farm reduces the risk of the possible spread of the BSE agent from a knackery or abattoir and leads to a focusing of control measures in one place, rather than extending the focus to other premises.

Animals should be photographed or filmed to confirm their identification for tracing purposes. The photographs and film should include close-up images of eartag, tail tag, brand, tattoo identification or other features that will assist the verification of each animal's identity.

As brain material is required for diagnosis, animals should not be shot through the head. Shooting will also increase the risk of dissemination of the agent in the environment.

See the **Destruction Manual** for further information.

2.2.7 Treatment of meat and other animal products

(See also Section 2.2.8 Disposal of animal products and byproducts and Section 2.2.9 Decontamination)

Meat and animal products assessed as being a risk because of their association with suspect cases, confirmed cases or at-risk animals must be destroyed by incineration or other acceptable method. Because of the difficulty of ensuring complete inactivation of the BSE agent, it is largely impractical to treat meat and other animal products in order to decontaminate them.

Specified risk materials

A protocol for inspection of carcases and removal and disposal of specified risk materials (SRMs) would need to be agreed upon without delay. The tissues and organs recognised as SRMs in cattle are described in Section 1.6.3.

2.2.8 Disposal of animal products and byproducts

It is imperative that infected carcases are *not* processed in any way that may permit infected material to enter the human food or animal feed chains.

See the **Disposal Manual** for detailed information.

The following points should be kept in mind in relation to carcase disposal.

- Wherever possible, carcases should be burned.
- The burning of carcases must be under the supervision of disease control authorities to ensure that it is performed appropriately and that all contaminated material is completely burned.
- The ash should be collected, mixed with agricultural lime to create suitably alkaline conditions, and buried deeply at a suitable site.
- Where burning is not practical, carcases and other materials that cannot be adequately decontaminated should be buried deeply in a suitable site and preferably with caustic materials that will create an alkaline environment.
- Consideration must be given to the future use of the burial site, and any associated water sources, as the agent may remain in a transmissible state in the soil for many months.
- Dogs, cats and other potential scavengers should be kept away from destruction and disposal sites.
- New methods of destruction of carcases and contaminated material based on alkaline hydrolysis can be considered.
- The location of burial sites for carcases, products or ash should be recorded and marked.
- Rendering is not an acceptable means of disposal of confirmed cases because the temperatures and pressures currently used would not be high enough to guarantee complete inactivation of the disease agent (see Appendix 5 for more details on the effectiveness of various rendering processes against the BSE agent).

2.2.9 Decontamination

Decontamination precautions should be taken in proportion with risk and for areas, fixtures and fittings that may have been contaminated with the tissues from confirmed cases. Decontamination may be required for premises with the potential for heavy contamination, such as field necropsy sites, knackeries and laboratory postmortem rooms, but decontamination of a property with confirmed or suspected cases is not necessary, other than at the site of destruction and disposal.

See the **Decontamination Manual** for further general information on decontamination procedures. Because many of the standard methods of decontamination cannot ensure complete inactivation of the BSE agent, the emphasis must be on removal of the agent by thorough cleaning. This should be followed by an appropriate steam sterilisation or liquid chemical treatment as described below (Taylor 2001, NHMRC 1996, ACDP 1998[12]).

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. The recommended methods of chemical decontamination for TSE agents are as follows.

- Sodium hypochlorite solution containing 2% (20 000 ppm) available chlorine for more than 1 hour at 20°C. For the BSE agent, the OIE Manual recommends overnight chemical disinfection of equipment.
- 2 M (80 g per litre) sodium hydroxide for more than 1 hour at 20°C. This method is known not to be completely effective unless alkali-to-tissue ratio is high enough (Taylor 2001).
- For histological samples only, 96% formic acid for 1 hour. However, formalin fixation of infected tissues stabilises the scrapie agent so that it cannot then be inactivated by steam sterilisation. Residues of formalin-fixed tissues should therefore be disposed of by incineration.

Decontamination of infected premises

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. The theoretical risk arises because the BSE agent has been found in various cattle tissues, including brain, eye, spinal cord, tonsils, thymus, spleen, intestines, dorsal root ganglia, trigeminal ganglia, skull and vertebral column. There is no evidence for spread of infection by urine, faeces or any discharges.

Entry of other animals to IPs should be prevented until decontamination is complete. However, it is not considered necessary to impose ongoing farmgate disinfection at IPs or SPs (see Appendix 1).

[12] http://www.official-documents.co.uk/document/doh/spongifm/report.htm

Decontamination of veterinary instruments

Instruments used for postmortem removal of brain or other potentially infected tissue from suspected cases or at-risk animals should either be discarded after a single use, or decontaminated using one of the methods described above and by the OIE before re-use on live animals. If BSE were confirmed in a native-born Australian animal, equivalent controls on instruments used on animals that are not considered at risk (eg for eye ablation or routine postmortem) might be warranted, depending on the circumstances.

2.2.10 Vaccination

Not applicable.

2.2.11 Grazing management

Epidemiological studies of the relatively small number of cases of BSE that have been reported in cattle born in the UK after the introduction of the comprehensive feed ban on 1 August 1996 suggest that the risk of infection from pasture is inconsequential. Any risk would apply to places such as field necropsy sites where infective material may have been present. However, epidemiological studies of these cases are continuing, and the issue of grazing management should be kept under review.

2.2.12 Wild and stray animal control

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

See the **Wild Animal Response Strategy** for further information.

2.2.13 Vector control

Not applicable.

2.2.14 Public awareness

One of the most important elements of a public health response will be the communication strategy to ensure that accurate and timely information is provided to the media and the community. Unsubstantiated reports of BSE could have serious ramifications for the livestock industry, its communities, the Australian economy and international relations. This needs to be addressed by the provision of accurate information, appointment of key spokespeople and clear coordination between the relevant organisations, especially human health authorities.

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 on the disease (eg 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, such as the pre-existing bans on feeding vertebrate protein to ruminants and longstanding restrictions on imports from countries with BSE;
- trade implications; and
- comparison with the UK epidemic and the situation in other countries.

See the **Public Relations Manual** and Section 3.2.9 of this manual for further information on provision of public information about emergency animal diseases.

2.3 Occupational safety guidelines

As the BSE agent is confined to the CNS in clinically affected cattle, there is no risk of human or animal exposure to the BSE agent from live cattle.

However, persons handling potentially infected material (CNS tissue from suspected BSE cases) must take adequate precautions to avoid exposure to these agents, as pointed out by the UK health authorities (ACDP 1998).

Veterinarians, laboratory workers and slaughterhouse workers should wear appropriate face and eye protection devices and gloves when handling tissues suspected of containing high levels of the agent.

Care should be taken to minimise environmental contamination during necropsy procedures. Carcases should be disposed of carefully and instruments disposed of by incineration or thoroughly decontaminated.

The use of single-use surgical and necropsy instruments and materials is preferred.

Extensive decontamination of the environment should not be necessary.

2.4 Feasibility of control in Australia

BSE is a notifiable disease in all states and territories of Australia, and owners and veterinarians are obliged to notify animal health authorities of any illness, death or impending movement of any *possible* case of the disease. BSE is a target disease under the National Animal Health Information System (NAHIS).[13] Australia currently has a National TSE Surveillance Program (NTSESP) in place to examine those cattle and sheep reported with clinical signs where a TSE is a reasonable differential diagnosis (NTSESP 2000).[14]

If an occurrence of BSE in Australia can be linked to an imported animal, and if tracing can identify all potentially infected stock, there is a high probability that the disease will be eradicated rapidly.

[13] http://www.aahc.com.au/nahis
[14] http://www.aahc.com.au/surveillance/ntsesp

In the unlikely event that BSE is found to be widespread, eradication of the disease will be more difficult and take longer. For example, in the unlikely event of iatrogenic transmission via a contaminated biological product as the source of the disease, it may be difficult to confirm the contamination of a particular product batch because of the long incubation period of the disease. Considerable industry cooperation would be required to determine the extent of the problem and to identify at-risk animals and products. A prolonged program would probably be necessary to achieve eradication.

3 Policy and rationale

3.1 Overall policy

Bovine spongiform encephalopathy (BSE) is an OIE listed disease that is significant in the international trade in cattle and cattle products. BSE is also a zoonosis and a safety hazard in human food.

The disease response policy is to find all cases, to determine the extent and origin of the disease in the Australian cattle herd and its mode of spread, and then to eradicate the disease as quickly as possible. A number of strategies to achieve and then verify eradication will be used, including:

- ***initial quarantine*** **of all cattle and any other at-risk animals on affected and suspect premises; as BSE is spread through contaminated feed, only in exceptional circumstances would it be necessary to establish a restricted or control area;**
- ***epidemiological investigations*** **to identify the source of infection and then identify cattle and other at-risk animals that might have acquired infection because of a similar exposure;**
- ***destruction and disposal*** **of the confirmed case or cases and part or all of their herds, depending on the findings from veterinary investigations;**
- ***surveillance and monitoring*** **by means of postmortem tests on brain and spinal cord material to the extent defined by veterinary investigations, in order to determine the limits of the outbreak and provide evidence of eradication and ongoing freedom from disease following the response;**
- **after eradication,** ***permanent quarantine of at-risk animals*** **with clinical examination every three months for life; and**
- **a** ***public awareness campaign.***

BSE is an Animal Health Australia Category 2 disease under the government–industry EAD Response Agreement for cost-sharing arrangements. Category 2 diseases are those for which costs will be shared 80% by government and 20% by industry.

The chief veterinary officer (CVO) in the state or territory in which the outbreak occurs will be responsible for developing an Emergency Animal Disease (EAD) Response Plan. This plan will be approved for technical soundness and consistency with AUSVETPLAN by governments and technical representatives of the affected livestock industry on the Consultative Committee on Emergency Animal Diseases (CCEAD). The plan would ultimately be approved for cost-sharing arrangements by government and industry through the national management group (NMG) of government and industry representatives established for the incident.

CVOs will implement disease control measures as agreed in the EAD Response Plan and in accordance with relevant legislation. They will make ongoing decisions on follow-up disease control measures in consultation with the CCEAD and the

NMG. The detailed control measures adopted will be determined using the principles of control and eradication (Section 2) and epidemiological information about the outbreak.

For further information on the responsibilities of the state or territory disease control headquarters and local disease control centres, see the **Control Centres Management Manual**, Part 1.

3.2 Strategy for control and eradication

This policy will apply once one or more confirmed cases of BSE are diagnosed, as defined in Section 2.2.1. Details of the policy will depend on the type of incident that initiates an emergency response (ie BSE in an imported animal or native-born Australian animal or a human case of vCJD) and the categorisation of the animals involved after the epidemiological investigation has been carried out (see Section 2.2.1). Table 4 shows details of the actions that will be required.

Australia should be able to continue to meet the conditions for BSE-free status under the *OIE Terrestrial Animal Health Code* (OIE Terrestrial Code; see Appendix 3) if all cases of BSE are clearly shown to occur in imported cattle and not to be the result of transmission within the country. The occurrence of BSE in native-born animals may indicate either a cycle of transmission in Australia, an iatrogenic event or a realisation of the theoretical possibility of a sporadic or familial TSE. The distinction between the two situations is important and will require an enhanced program of surveillance and monitoring to an extent and intensity determined by the epidemiological and other veterinary investigations.

Table 4 Actions required in Australia for different categories of infected or potentially infected animals and for different potential presentations

Animal category[a]	ACTIONS	
	Animals	**Other measures**
BSE in an imported animal		
– Clinical case – Cohorts residing in Australia – Progeny (if the case is in a cow) – Dam (if also imported)	Review animal identification to confirm that the animal was imported. Slaughter, test and destroy carcase (of clinical case).	Trace edible and inedible products derived from recently slaughtered cohorts as far as possible. Implement a tracing and surveillance program (additional to the existing program). Quarantine affected premises and stop movement of animals until a full history, epidemiological studies and identification of at-risk animals have been completed. Immediately suspend all imports of live animals and specified products from the country concerned, pending a revised epidemiological investigation of that country. Assist source country in epidemiological investigation to identify at-risk animals. Introduce, in conjunction with health authorities, an enhanced public awareness program that describes measures taken to protect human and animal health.
– Other animals	Depending on epidemiology findings: • slaughter, test, destroy and dispose of carcase; • quarantine; or • take no action. (Decision based on assessment of potential exposure to the BSE agent.)	Implement an enhanced surveillance program, which could involve more screening tests at slaughter and tests of all fallen stock and those with nervous signs. Seek support from health authorities in epidemiological investigations and a whole-of-government approach.
vCJD in an Australian resident		
– Not applicable	Not applicable	Support health authorities in epidemiological investigations and a whole-of-government approach.[b] Improve surveillance and testing of livestock (if investigations show no evidence of patient exposure in a BSE-affected country). Institute, in conjunction with the Department of Health and Ageing, an enhanced public awareness program, especially outlining the country's previous surveillance and prevention programs.

contd…

Table 4 Actions required in Australia for different categories of infected or potentially infected animals and for different potential presentations (continued)

Animal category[a]	ACTIONS	
	Animals	**Other measures**
BSE in an animal of Australian origin[c]		
– Clinical case	Review animal identification to confirm that the animal is of Australian origin. Slaughter, test and destroy carcase.	Review case diagnosis, confirmatory testing, parallel testing at the Australian Animal Health Laboratory and the world reference laboratory (VLA, Weybridge). Trace the animal to property of birth. Quarantine premises until epidemiological investigation complete. Conduct thorough epidemiological investigation including herd health history. Apply the same controls to farms to which cohort animals have moved.
– Animal with a history of association with confirmed case	Depending on epidemiology findings: • slaughter, test and destroy carcase; • quarantine; or • take no action. (Decision based on assessment of potential exposure to the BSE agent.)	Implement an enhanced surveillance program.
Common risk factor but no clinical disease[d]		
– Not applicable	Not applicable	Implement a thorough assessment of the imported product, including its use in Australia. Trace animals exposed to the contaminated product. Identify, quarantine and monitor any animals considered to be at risk. Institute, in conjunction with the Department of Health and Ageing, an enhanced public awareness program, especially outlining the country's previous surveillance and prevention programs. Delineate/zone risk premises. Trace product from exposed animals as far as possible.

a For details of animal categories, see Section 2.2.1.
b The Australian Government Department of Health and Ageing has prepared a vCJD response plan.
c Whether identified through overseas or domestic testing. The possibility exists that the case could be sporadic or familial and not have originated from BSE in the EU or UK.
d BSE arising in another country as a result of use of an imported veterinary therapeutic (including vaccine), where that same veterinary therapeutic has been imported into Australia

3.2.1 Stamping out

In the case of BSE, stamping out as defined in the OIE Terrestrial Code is not applicable.

Slaughter of animals and destruction of carcases will be based on the categories defined in Section 2.2 and the policies in Table 4.

3.2.2 Quarantine and movement controls

Quarantine will be imposed on infected premises (IPs) and suspect premises (SPs) and movement controls on animals and products will be introduced in the short term until the situation is defined. Further decisions can then be made based on data from veterinary investigations (see Table 4). There will be few ongoing restrictions for most animals or products. The declaration of restricted and control areas for BSE is not envisaged unless exceptional circumstances occur, such as there being suspicion of the widespread presence of disease in a defined geographical area.

See Appendix 1 for further details on declared areas.

See Appendix 2 for further details on quarantine and movement controls.

Zoning

It is most unlikely that zoning of geographic areas would be appropriate for BSE, unless a widespread but geographically defined outbreak of BSE were discovered. However, certain classes of animals could be exempt from BSE controls. In the UK, for example, cattle from specialist beef herds at very low risk of BSE and registered under the beef assurance scheme in that country were allowed to be slaughtered for sale for human consumption up to 42 months of age.

3.2.3 Tracing and surveillance

Trace-forward of animals and animal products will decide SPs and will be based on previous relationships with confirmed cases and the possibility of exposure to the BSE agent. At-risk animals will be examined every three months to detect the development of characteristic clinical signs until they die or are slaughtered. All animals under surveillance will be identified by National Livestock Identification System embedded microchip devices and will be tested for BSE when they are killed.

Trace-back will attempt to locate the possible source of exposure. In the case of an imported animal, exposure is most likely to have occurred overseas. Although very unlikely because of Australia's quarantine controls, exposure in Australia could be through the introduction of an animal product, the use of a contaminated biological product, or some other iatrogenic source.

If the disease is due to administration of a contaminated biological product, it is likely that there may be more affected animals at a number of different locations, all at a similar stage of disease. In such a situation, information about the attack rates in known infected herds may assist with investigations in suspect herds possibly exposed to the same product.

See Appendix 4 for further details on surveillance.

3.2.4 Vaccination

There is no vaccine for BSE and therefore vaccination is not applicable.

3.2.5 Treatment of infected animals

There is no treatment that is effective and cases must be slaughtered, sampled and carefully disposed of.

3.2.6 Treatment of animal products and byproducts

As described in Section 2.2.7, there is no treatment for animal products that is guaranteed to be effective in inactivating the BSE agent under normal commercial operations. Meat or animal products from confirmed cases of BSE must not be rendered for meat-and-bonemeal (MBM) or for other products and must be disposed of by incineration.

The OIE Terrestrial Code for BSE (see Appendix 3) outlines the minimum procedures that should be adopted for international trade in animals and animal products. These same procedures should be adopted for domestic trade.

3.2.7 Carcase disposal

The carcases of all animals that are killed to eradicate BSE will be completely destroyed in accordance with procedures described in Section 2.2.8.

3.2.8 Decontamination

There will be a clean-up of locations with confirmed cases or contaminated feed, or where material from confirmed cases was handled, in accordance with procedures described in Section 2.2.9.

Veterinary (surgical) instruments used for postmortem removal of brain tissue for diagnosis, or which have otherwise contacted CNS tissues of potentially infected animals, should be either discarded after a single use or decontaminated using one of the methods described in Section 2.2.9.

3.2.9 Public awareness and media

The public, especially those in the livestock industries, must be informed as soon as possible after any disease is confirmed. Accurate information must be provided to help prevent public alarm. There should be clear coordination of information among the relevant organisations, including human health authorities, industry organisations, food safety authorities and the National Health and Medical Research Council (NHMRC) Special Expert Committee on TSEs (see Section 3.2.10).

Section 2.2.14 gives further information on the approach required for providing information to the public.

3.2.10 Preventive measures and response plan for vCJD in Australia

As described in Section 1.2, a variant form of Creutzfeldt–Jakob disease (vCJD) was identified in the UK in the mid-1990s, and found to be associated with the consumption of beef products from BSE-affected cattle. An Australian National CJD Registry has been maintained in the Department of Pathology, University of

Melbourne since 1993 and all cases of CJD are investigated. No cases of vCJD have yet been reported in Australia. However, cases could conceivably occur in the future (for example, in people who lived in the UK before the removal of potentially BSE-infected beef products from the human food chain).

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

Australian regulatory agencies, such as the Therapeutic Goods Administration (TGA) and Food Standards Australia New Zealand (FSANZ), have undertaken extensive reviews of all products under their control to identify constituents of bovine origin and have taken measures to prevent exposure of the Australian population. Donor deferral procedures, among other safeguards, have been put into place to protect the safety of the Australian blood supply, although there is no evidence that vCJD is transmitted by blood.

As part of Australia's preparedness in addressing the potential public health, medico-legal, social, community, political, trade and international relations impacts of vCJD, the Department of Health and Ageing has prepared a draft response plan in the event that a case of vCJD occurs. The plan is based on a risk management approach for biological emergencies that recognises that such an event will occur very infrequently, the evidence base for decision making may be limited and evolving, and the community reaction may be disproportionate to the level of physical risk.

3.3 Social and economic effects

The occurrence of even a single indigenous case of BSE in Australia would carry a high risk of major social and economic impacts. There may possibly be an immediate reduction in domestic consumption of beef, and there would be restrictions on Australia's international trade in livestock and livestock products. It is therefore most important that all aspects of the response be activated immediately to address public concerns and facilitate rapid restoration of market access.

3.4 Criteria for proof of freedom

Proof of freedom begins with the emergency response, which should be thoroughly documented. The level of activity undertaken in verifying freedom will depend on the extent of the outbreak and the results of tracings of at-risk animals and their products. At all times, the OIE Terrestrial Code for BSE, including the appendix on surveillance and monitoring systems for BSE (see Appendixes 3 and 4), should be used as a guide.

The OIE Terrestrial Code recognises a number of levels of freedom from BSE, which vary according to the source and extent of the outbreak, and thus determine the actions required:

- BSE-free country or zone
- provisionally BSE-free country or zone
- country or zone with a minimal BSE risk.

In the event of a BSE detection, it will be necessary to show that there are no high-risk products remaining in the human food and animal feed chains and that the Australian cattle population and any other high-risk groups of other species are again free of BSE infection.

To verify that there is no chance of high-risk products entering food chains, extensive auditing of existing arrangements will be maintained, and revision of relevant risk analyses that comply with the BSE chapter in the OIE Terrestrial Code will be undertaken. Food safety authorities will have a lead role in the protection of the human food supply.

The National TSE Surveillance Program (NTSESP)[15] managed by Animal Health Australia will be used to gather evidence of freedom from BSE, based on examination of native-born animals displaying clinical signs consistent with a differential diagnosis that includes BSE. The number of animals to be examined will be determined at the time, based on the size and nature of the detection and the epidemiological study.

The TSE surveillance program is currently structured to comply with the May 2003 revision of the OIE Terrestrial Code, so that annually there is a 99% probability of detecting BSE or scrapie if they account for 1% of the cases of neurological disease in adult cattle and sheep in Australia. By targeting neurological cases, the detection limit under the assumptions made in the code is one in a million of the adult population.

In addition, it may be necessary to examine additional selected subpopulations in accordance with the OIE requirements for surveillance and monitoring systems for BSE (see Appendix 3).

See Appendix 4 for further details on proof of freedom.

3.5 Funding and compensation

BSE is classified as a Category 2 emergency animal disease under the EAD Response Agreement between the governments of Australia and the livestock industries.

Category 2 diseases are emergency animal diseases that have the potential to cause major national socioeconomic consequences through very serious international trade losses, national market disruptions and very severe production losses in the livestock industries that are involved. Category 2 also includes diseases that may have slightly lower national socioeconomic consequences, but also have significant public health and/or environmental consequences. For this category, the costs will be shared 80% by governments and 20% by the relevant industries (refer to the EAD Response Agreement for details).[16] Further information on the cost-sharing arrangements can be found in the AUSVETPLAN **Summary Document** and in the **Valuation and Compensation Manual**.

[15] http://www.aahc.com.au/surveillance/ntsesp
[16] Some information on the EAD Response Agreement can be found at: http://www.animalhealthaustralia.com.au/programs/eadp/eadra.cfm

3.6 Strategy if the disease becomes established

The ban on the feeding of MBM to ruminants in Australia will prevent the establishment of BSE in Australia. If spread 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; and
- eradication programs for identified infected herds, as determined by veterinary investigations.

In addition, an accreditation program for cattle herds might be necessary.

Appendix 1 Guidelines for classifying declared areas

Premises

Infected premises (IP)

A premises classified as an IP will be a premises (which may be a paddock or part of a property) on which a case of BSE has been confirmed or is suspected and/or a premises that contains contaminated animal products or byproducts.

Dangerous contact premises (DCP)

Not applicable.

Suspect premises (SP)

Premises classified as SPs will be premises containing at-risk animals of the following categories:

- any imported cattle originating from the same foreign property as a confirmed case;
- cohorts — animals born in the same calving season and raised with a confirmed case, or other cattle that may have been brought into contact with the source of the BSE agent;
- all progeny born from a confirmed BSE female case within two years before or after the onset of clinical signs;
- the dam of a confirmed case;
- other ruminant animals, including sheep, goats, deer, camelids and elk, that have resided on an IP at a time when they may have come into contact with material containing the BSE agent; or
- animals that may have had parenteral administration of a product containing the BSE agent (eg vaccine).

'Suspect premises' is a temporary classification for premises containing animals that are at risk of having the disease. High priority should be given to clarifying the status of the suspect animals so that the SP can be reclassified as either an infected premises (IP) and appropriate quarantine and movement controls implemented, or as free from disease, in which case no further disease control measures are required.

Restricted areas and control areas

Not applicable.

Appendix 2 Recommended quarantine and movement controls

Premises

Quarantine and movement controls	Infected premises	Suspect premises
Movement out of:		
– *susceptible animals*	Movement by permit dependent on epidemiological investigation	Movement by permit dependent on epidemiological investigation
– *specified products*	Movement by permit dependent on epidemiological investigation	
– *other animals*	Control movement of other ruminant species	
Movement in of:		
– *susceptible animals*	Movement by permit dependent on epidemiological investigation	No restriction, but advise owner of implications
Movement in and out of:		
– *people*	No restriction	
– *vehicles and equipment*	No restriction, except for equipment that has been in contact with dead animals. Such equipment (especially surgical equipment) will require decontamination before movement out of infected premises	

Areas

The declaration of restricted areas and control areas is very unlikely during a BSE outbreak.

Appendix 3 OIE animal health code and diagnostic manual for terrestrial animals

OIE Terrestrial Code

The objective of the *OIE Terrestrial Animal Health Code* is to prevent the spread of animal diseases, while facilitating international trade in live animals, semen, embryos and animal products. This annually updated volume is a reference document for use by veterinary departments, import/export services, epidemiologists and all those involved in international trade.

The OIE Terrestrial Code is amended in May each year. The current edition is published on the OIE website at:

http://www.oie.int/eng/normes/mcode/A_summry.htm

Chapters of the code relevant to this disease strategy are:

Chapter 2.3.13 Bovine spongiform encephalopathy

Chapter 1.3.5 Zoning, regionalisation and compartmentalisation

Chapter 1.3.6 Surveillance and monitoring of animal health

Appendix 3.8.4 Surveillance systems for bovine spongiform encephalopathy

OIE Terrestrial Manual

The purpose of the *OIE Manual of Standards for Diagnostic Tests and Vaccines for Terrestrial Animals* is to contribute to the international harmonisation of methods for the surveillance and control of the most important animal diseases. Standards are described for laboratory diagnostic tests and the production and control of biological products (principally vaccines) for veterinary use across the globe.

The OIE Terrestrial Manual is updated approximately every four years. The 5th edition was published in 2004 and is available on the OIE website at:

http://www.oie.int/eng/normes/mmanual/A_summry.htm

The chapter of the manual relevant to this disease strategy is:

Chapter 2.3.13 Bovine spongiform encephalopathy

Appendix 4 Procedures for surveillance and proof of freedom

Information on Australia's current National Transmissible Spongiform Encephalopathy Surveillance Program (NTSESP) is available on the Animal Health Australia website at http://www.aahc.com.au/surveillance/ntsesp/index.htm.

In the event of an outbreak of BSE in Australia, the policy for surveillance and proof of freedom from disease will be to follow the OIE standard as set out in Appendix 3.8.4 of the current edition of the *OIE Terrestrial Animal Health Code*, which is available on the OIE website at http://www.oie.int/eng/normes/mcode/en_chapitre_3.8.4.htm

For ease of reference, the text of Appendix 3.8.4 from the 2004 edition of the OIE Terrestrial Code is shown below. However, as indicated in Appendix 3, the code is reviewed annually in May, and readers should ensure that they consult the most recent version.

APPENDIX 3.8.4. (2004)

SURVEILLANCE SYSTEMS FOR BOVINE SPONGIFORM ENCEPHALOPATHY

Article 3.8.4.1.

Surveillance for bovine spongiform encephalopathy (BSE) has at least two goals: to determine whether BSE is present in the country, and, if present, to monitor the extent and evolution of the epizootic, thus aiding control measures and monitoring their effectiveness.

The cattle population of a country or zone not free from BSE, will comprise the following sub-populations in order of decreasing size:

1. cattle not exposed to the infective agent;
2. cattle exposed but not infected;
3. infected cattle, which may lie within one of three stages in the progress of BSE:
   a. the majority will die or be killed before reaching a stage at which BSE is detectable by current methods;
   b. some will progress to a stage at which BSE is detectable by testing before clinical signs of disease appear;
   c. the smallest number will show clinical signs of disease.

Surveillance programmes should be determined by, and commensurate with the outcome of the risk assessment referred to in Article 2.3.13.2. and should take into account the diagnostic limitations associated with the above sub-populations and the relative distributions of infected animals among them.

Surveillance programmes developed before the advent of rapid diagnostic tests focused on the population containing cattle displaying clinical signs compatible with BSE as described in Article 3.8.4.2. While surveillance should focus on this sub-population, investigations of other sub-populations using the new diagnostic

techniques may provide a more accurate picture of the BSE situation in the country or zone. A surveillance strategy may therefore need to combine several strategies. Recommended strategies for surveying the various sub-populations are described below.

Available data suggest the possibility that a gradient might be established to describe the relative value of surveillance applied to each sub-population. All countries should sample sub-populations identified in Article 3.8.4.2. and Article 3.8.4.3. In countries where surveillance of cattle identified in Article 3.8.4.2. is unable to generate the numbers recommended in Table 1, surveillance should be enhanced by testing larger numbers of cattle identified in Article 3.8.4.3. Any shortfall in the first two sub-populations should be addressed by the sampling of normal cattle over 30 months of age at slaughter. Exclusive dependence on random sampling from normal cattle is not recommended, unless the number of samples examined annually is statistically sufficient to detect a disease prevalence of of 1 in 1,000,000.

Surveillance for BSE requires laboratory examination of samples in accordance with the methods described in the Terrestrial Manual.

For surveillance purposes, testing a part of the population is consistent with Chapter 1.3.6. on surveillance and monitoring of animal health.

Article 3.8.4.2.

Examination of cattle displaying clinical signs consistent with BSE

Cattle affected by illnesses that are refractory to treatment, and displaying progressive behavioural changes such as excitability, persistent kicking when milked, changes in herd hierarchical status, hesitation at doors, gates and barriers, as well as those displaying progressive neurological signs without signs of infectious illness, are candidates for examination. Since BSE causes no pathognomonic clinical signs, all countries with cattle populations will observe individual animals with compatible clinical signs. It should be recognised that cases may display only some of these signs, which may also vary in severity, and such animals should still be investigated as potential BSE affected animals.

Table 1 indicates the minimum number of animals exhibiting one or more clinical signs of BSE that should be subjected to diagnostic tests according to the total cattle population over 30 months of age. As this sampling is not random, the numbers indicated in this table are a subjective interpretation rather than a strict statistical deduction.

Table 1 Minimum number of annual investigations of cattle showing clinical signs consistent with BSE required for effective surveillance according to the total cattle population over 30 months of age

Total cattle population over 30 months of age	Minimum number of samples to examine
500,000	50
700,000	69
1,000,000	99
2,500,000	195
5,000,000	300
7,000,000	336
10,000,000	367
20,000,000	409
30,000,000	425
40,000,000	433

Article 3.8.4.3.

Examination of targeted cattle displaying clinical signs not necessarily indicative of BSE

Cattle that have died or have been killed for reasons other than routine slaughter should be examined. This population will include cattle which have died on farm or in transit, 'fallen stock', and stock sent for emergency slaughter.

Many of these cattle may have exhibited some of the clinical signs listed in Article 3.8.4.2. which were not recognised as being compatible with BSE. Experience in countries where BSE has been identified indicates that this population is the second most appropriate population to target in order to detect BSE.

Article 3.8.4.4.

Examination of cattle subject to normal slaughter

In countries not free from BSE, sampling at routine slaughter is a means of monitoring the progress of the epizootic and the efficacy of control measures applied, because it offers continuous access to a cattle population of known class, age structure and geographical origin.

Within each of the above sub-populations, countries may wish to target cattle identifiable as imported from countries or zones not free from BSE, cattle which have consumed potentially contaminated feedstuffs from countries or zones not free from BSE, offspring of BSE affected cows and cattle which have consumed feedstuffs potentially contaminated with other TSE agents.

Appendix 5 Experimental protocols used to mimic commercial rendering processes

Industrial pilot-scale facsimiles of rendering practices used within the European Community were spiked with the BSE agent (Taylor et al 1995b) and inactivation was assessed by mouse bioassay. Six types of rendering system were tested, including a batch pressure cooker system which, to achieve 133°C, would have reached absolute pressure of 3 bar (gauge pressure of 2 bar plus atmospheric pressure).

BSE inactivation under different conditions of pressure, temperature and time

Process	Particle diameter (mm)	End temperature (°C)		Time (min)	Inactivation of BSE[a]
		Planned	Achieved		
Batch atmospheric	150	120	121	150	YES
Continuous atmospheric (natural fat)	30	100–125	112	50	NO
	30	125	123	125	YES
	30	100–140	122	50	NO
	30	140	139	125	YES
Continuous atmospheric (high fat)	30	140	136	30	YES
	30	140	137	120	YES
Continuous vacuum (high fat)	10	125	120	20	NO
	10	125	121	57	NO
Continuous wet rendering (natural fat)	20	100–120	101	120	YES
	20	120	119	240	YES
	20	70	72	240	YES
Batch pressure[b] (natural fat)	50	133	133	30	YES
	30	136	135	28	YES
	30	145	145	28	YES

a Mouse bioassay
b Batch pressure cooker system which, to achieve 133°C, would have reached absolute pressure of 3 bar (gauge pressure of 2 bar plus atmospheric pressure).
Source: Taylor et al (1995b), Table 1.

The results for BSE inactivation support the recommended minimum acceptable temperature for rendering to inactivate spongiform encephalopathy agents being not less than 133°C for a minimum of 20 minutes at an absolute pressure of 3 bar ('133°C/20 minutes/3 bar'). The four rendering processes that failed are characterised by lower temperatures (112–121°C) and relatively shorter times.

Appendix 6 BSE tracing questionnaire

(adapted from New Zealand MAF Questionnaire; suitable for intensively run properties only and would need adaptation for use with extensive holdings)

Name of owner / manager (specify which)

_____

Address of herd (Property name, road name, local town, district, region, state, area code)

_____

_____

_____

Tail tag number

_____

Phone (day, night, mobile) _____

Postal address

_____

_____

_____

Herd details / Farm type Dairy ❑ **Beef** ❑ **Other** _____

Age structure (please insert the number of animals)

Age (years)	<1	2	3	4	5	6	7	8	9	10+
Beef										
Dairy										

Other animals on farm (totals)

Deer	_____	**Sheep**	_____	**Goats**	_____	**Horses**	_____
Camelids	_____	**Dogs**	_____	**Cats**	_____	**Other**	_____

Other properties owned/managed by this person

_____

_____

Case details

Animal identification_____

Ear tag number_____

Animal name (if any)_____

Other ID/description

_____

_____

Date of birth of animal

_____

How was date of birth confirmed?
(available records or dentition)_____

Breed_____**Sex (M/F/S)**_____

Dam_____

Sire_____

At the time of birth of this case, where were cows calved?
(on a calving pad, in housing or at pasture)

_____

Are placenta removed from the calving sites?_____

How long would calf remain in calving accommodation?_____

Has this animal ever calved? _____**Number** _____

Location of offspring_____

_____

_____

Are there any siblings of this animal (born within two years by the same dam) and were these animals retained in the adult herd (if sold or culled, give dates and reasons).

_____

_____

_____

Case details (continued)

Other calves born the same year as this animal (particularly those reared/fed the same)

Case details (continued)

Proprietary concentrates fed to animal. Please indicate the rations fed to this animal throughout its life, if known. This includes rations fed to calves and in-feed medication. Please specify where the feed was supplied from, whether it was supplied in bulk or bagged, the frequency of purchase, how feed is stored (bagged, bulk bins, floor etc) and whether storage bins are emptied between deliveries.

Product	Supplier	Supplied in	Frequency	Storage

Are storage bins emptied between deliveries? **Yes** ❑ **No** ❑

Case details (continued)

Clinical history. Please inquire about nervous symptoms, attitude and gait changes, abnormal behaviour, bodyweight, condition and milk yield changes, and note any progression of symptoms.

Which veterinarian/veterinary practice is/was used?

Phone _____

Case details (continued)

Details of dam of case

ID_____ **Date of birth** _____

Is she still in the herd?_____ **Date of departure**_____

Reason for departure_____

Destination_____

Herd history/health. Clinical signs (eg nervous signs), treatment history (eg vaccinations), herd movements (eg overseas introductions)

Comments

Date of visit			
Forms completed by			
Signature			
Phone number		**Email**	

Glossary

Animal byproducts	Products of animal origin that are not for consumption but are destined for industrial use (eg hides and skins, fur, wool, hair, feathers, hooves, bones, fertiliser).
Animal Health Committee	A committee comprising the CVOs of Australia and New Zealand, Australian state and territory CVOs, Animal Health Australia, and a CSIRO representative. The committee provides advice to PIMC on animal health matters, focusing on technical issues and regulatory policy (formerly called the Veterinary Committee). *See also* Primary Industries Ministerial Council (PIMC)
Animal products	Meat, meat products and other products of animal origin (eg eggs, milk) for human consumption or for use in animal feedstuff.
At-risk animals (BSE)	Animals that have had a relevant association with suspected or confirmed cases of BSE. S*ee* Section 2.2.1 for further details.
Australian Chief Veterinary Officer	The nominated senior Australian government veterinarian in the Department of Agriculture, Fisheries and Forestry who manages international animal health commitments and the Australian Government's response to an animal disease outbreak. *See also* Chief veterinary officer
AUSVETPLAN	*Aus*tralian *Vet*erinary Emergency *Plan*. A series of technical response plans that describe the proposed Australian approach to an emergency animal disease incident. The documents provide guidance based on sound analysis, linking policy, strategies, implementation, coordination and emergency-management plans.
Biological products	Agents of biological origin (eg sera, hormones) for therapeutic use in the diagnosis or treatment of certain diseases.
Bonemeal	*See* Meatmeal/bonemeal
Chief veterinary officer (CVO)	The senior veterinarian of the animal health authority in each jurisdiction (national state, State or territory) who has responsibility for animal disease control in that jurisdiction. *See also* Australian Chief Veterinary Officer
Clinically consistent animal (BSE)	An animal that is found with clinical signs considered consistent with BSE. S*ee* Section 2.2.1 for further details
Cohort	A group of animals (of the same species) sharing a common temporal experience (eg the calving season or supplementary feeding) that is observed through time.

Compensation	The sum of money paid by government to an owner for stock that are destroyed and property that is compulsorily destroyed because of an emergency animal disease.
Confirmed case (BSE)	A suspected case, or clinically consistent animal, that has been confirmed BSE positive on postmortem by positive results from immunohistochemistry, immunochemistry, demonstration of characteristic fibrils by electron microscopy or by a test not previously performed on this case. *See* Section 2.2.1 for further details
Consultative Committee on Emergency Animal Diseases (CCEAD)	A committee of state and territory CVOs, representatives of CSIRO Livestock Industries and the relevant industries, and chaired by the Australian CVO. CCEAD convenes and consults when there is an animal disease emergency due to the introduction of an emergency animal disease of livestock, or other serious epizootic of Australian origin.
Control area	A declared area in which the conditions applying are of lesser intensity than those in a restricted area (the limits of a control area and the conditions applying to it can be varied during an outbreak according to need). Not applicable to BSE; *See* Appendix 1
Dangerous contact animal	Susceptible animals that have been designated as being exposed to other infected animals or potentially infectious products following tracing and epidemiological investigation. Not applicable to BSE; *See* Section 2.2.1
Dangerous contact premises	Premises that contain dangerous contact animals or other serious contacts. Not applicable to BSE; *See* Appendix 1
Declared area	A defined tract of land that is subjected to disease control restrictions under emergency animal disease legislation. Types of declared areas include *restricted area*, *control area*, *infected premises*, *dangerous contact premises* and *suspect premises*. *See* Appendix 1 for further details
Decontamination	Includes all stages of cleaning and disinfection.
Destroy (animals)	To slaughter animals humanely.
Disease agent	A general term for a transmissible organism or other factor that causes an infectious disease.
Disease Watch Hotline	24-hour freecall service for reporting suspected incidences of emergency diseases — **1800 675 888**
Disinfectant	A chemical used to destroy disease agents outside a living animal.

Disinfection	The application, after thorough cleansing, of procedures intended to destroy the infectious or parasitic agents of animal diseases, including zoonoses; applies to premises, vehicles and different objects that may have been directly or indirectly contaminated.
Disposal	Sanitary removal of animal carcases, animal products, materials and wastes by burial, burning or some other process so as to prevent the spread of disease.
Emergency animal disease	A disease that is (a) exotic to Australia or (b) a variant of an endemic disease or (c) a serious infectious disease of unknown or uncertain cause or (d) a severe outbreak of a known endemic disease, and that is considered to be of national significance with serious social or trade implications. *See also* Endemic animal disease, Exotic animal disease
Emergency Animal Disease (EAD) Response Agreement	Agreement between the Australian and state/territory governments and livestock industries on the management of emergency animal disease responses. Provisions include funding mechanisms, the use of appropriately trained personnel and existing standards such as AUSVETPLAN.
Endemic animal disease	A disease affecting animals (which may include humans) that is known to occur in Australia. *See also* Emergency animal disease, Exotic animal disease
Enterprise	*See* Risk enterprise
Epidemiology	The study of disease in populations and of factors that determine its occurrence.
Epidemiological investigation	An investigation to identify and qualify the risk factors associated with the disease. *See also* Veterinary investigation
Exotic animal disease	A disease affecting animals (which may include humans) that does not normally occur in Australia. *See also* Emergency animal disease, Endemic animal disease
Exotic fauna/feral animals	*See* Wild animals
Fomites	Inanimate objects (eg boots, clothing, equipment, instruments, vehicles, crates, packaging) that can carry an infectious disease agent and may spread the disease through mechanical transmission.
Iatrogenic disease	A case of disease caused by medical or veterinary procedures (eg an infection spread by surgical procedures).
Immunochemistry	The branch of immunology, or a diagnostic test, concerned with chemical substances and reactions of the immune system, specifically antigens and antibodies and their interactions with one another.

Immunohistochemistry	Immunochemistry applied to the study, or testing, of cells and tissues.
In-contact animals	Animals that have had close contact with infected animals, such as non-infected animals in the same group as infected animals.
Incubation period	The period that elapses between the introduction of the pathogen into the animal and the first clinical signs of the disease.
Index case	The first or original case of the disease to be diagnosed in a disease outbreak on the index property.
Index herd	The first or original herd in which a case of the disease has been diagnosed. *See also* Index case, Index property
Index property	The property on which the first or original case (index case) in a disease outbreak is identified to have occurred.
Infected premises	A defined area (which may be all or part of a property) in which an emergency disease exists, is believed to exist, or in which the infective agent of that emergency disease exists or is believed to exist. An infected premises is subject to quarantine served by notice and to eradication or control procedures. *See* Appendix 1 for further details
Local disease control centre (LDCC)	An emergency operations centre responsible for the command and control of field operations in a defined area.
Meatmeal/bonemeal	The solid protein products obtained when animal tissues are rendered. *See also* Rendering (of carcases)
Monitoring	Routine collection of data for assessing the health status of a population. *See also* Surveillance
Movement control	Restrictions placed on the movement of animals, people and other things to prevent the spread of disease.
National management group	A group established to direct and coordinate an animal disease emergency. NMGs may include the chief executive officers of the Australian Government and state or territory governments where the emergency occurs, industry representatives, the Australian CVO (and chief medical officer, if applicable) and the chairman of Animal Health Australia.
Native wildlife	*See* Wild animals

OIE Terrestrial Code	*OIE Terrestrial Animal Health Code.* Reviewed annually at the OIE meeting in May and published on the internet at: **http://www.oie.int/eng/normes/mcode/a_summry.htm** *See* Appendix 3 for further details
OIE Terrestrial Manual	*OIE Manual of Standards for Diagnostic Tests and Vaccines for Terrestrial Animals.* Describes standards for laboratory diagnostic tests and the production and control of biological products (principally vaccines). The current edition is published on the internet at: **http://www.oie.int/eng/normes/mmanual/a_summry.htm** *See* Appendix 3 for further details
Operational procedures	Detailed instructions for carrying out specific disease control activities, such as disposal, destruction, decontamination and valuation.
Owner	Person responsible for a premises (includes an agent of the owner, such as a manager or other controlling officer).
Premises	A tract of land, including its buildings, or a separate farm or facility that is maintained by a single set of services and personnel.
Prevalence	Proportion (or percentage) of animals in a particular population affected by a particular disease (or infection or positive antibody titre) at a given point in time.
Primary Industries Ministerial Council (PIMC)	The council of Australian national, state and territory and New Zealand ministers of agriculture that sets Australian and New Zealand agricultural policy (formerly the Agriculture and Resource Management Council of Australia and New Zealand). *See also* Animal Health Committee
Prion	Word coined in the 1980s for 'proteinaceous infectious particle'. Prion protein (PrP^{Sc}) is an abnormal form of a common cellular membrane protein (PrP^{c}). Unlike PrP^{c}, PrP^{Sc} is resistant to protein-digesting enzymes (proteases) 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 in the case of BSE. *See also* Scrapie-associated fibrils
Quarantine	Legal restrictions imposed on a place or a tract of land by the serving of a notice limiting access or egress of specified animals, persons or things.
Rendering (of carcases)	Processing by heat to inactivate infective agents. Rendered material may be used in various products according to particular disease circumstances.

Restricted area	A relatively small declared area (compared to a control area) around an infected premises, which is subject to intense surveillance and movement controls. Not applicable to BSE; *See* Appendix 1
Risk enterprise	A defined livestock or related enterprise that is potentially a major source of infection for many other premises. Includes intensive piggeries, feedlots, abattoirs, knackeries, saleyards, calf scales, milk factories, tanneries, skin sheds, game meat establishments, cold stores, AI centres, veterinary laboratories and hospitals, road and rail freight depots, showgrounds, field days, weighbridges, garbage depots.
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 UK and many other parts of the world (but not in Australia). It can be transmitted naturally or experimentally to other animal species, including mice, and has been the experimental model for much TSE research.
Scrapie-associated fibrils	Abnormal fibrils caused by an accumulation of protease-resistant prion protein (PrP^{Sc}) and identified by electron microscopy. First identified in scrapie-infected mice but now recognised as a characteristic of all TSEs. *See also* Prion
Sensitivity	The proportion of truly positive units that are correctly identified as positive by a test. *See also* Specificity
Serotype	A subgroup of microorganisms identified by the antigens carried (as determined by a serology test).
Sibling	For BSE, animals that share a common dam.
Signalment	The fixed history of an animal, including its species, breed, age and sex.
Specified risk materials (BSE)	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. Also called specified bovine material (SBM) or (formerly) specified bovine offal (SBO) in the UK.
Specificity	The proportion of truly negative units that are correctly identified as a negative by a test. *See also* Sensitivity

Stamping out	Disease eradication strategy based on the quarantine and slaughter of all susceptible animals that are infected or exposed to the disease.
State or territory disease control headquarters	The emergency operations centre that directs the disease control operations to be undertaken in a particular state or territory.
Surveillance	A systematic program of investigation designed to establish the presence, extent or absence of a disease, or of infection or contamination with the causative organism. Includes the examination of animals for clinical signs, antibodies or the causative organism.
Susceptible animals	Animals that can be infected with a particular disease (for BSE — mainly cattle; *see* Section 1.2).
Suspect animal (general)	An animal that may have been exposed to an emergency disease such that its quarantine and intensive surveillance, but not pre-emptive slaughter, is warranted. OR An animal not known to have been exposed to a disease agent but showing clinical signs requiring differential diagnosis. *See also* At-risk animals (BSE), Suspected case (BSE)
Suspect premises	Temporary classification of premises containing suspect animals. After rapid resolution of the status of the suspect animals contained on it (for BSE, this is at-risk animals), a suspect premises is reclassified either as an infected premises (and appropriate disease-control measures taken) or as free from disease. *See* Appendix 1 for further details for BSE
Suspected case (BSE)	An animal of the genus *Bos* (cattle) or genus *Bubalus* (buffalo) with history, clinical signs and histological changes consistent with BSE as described in Section 1.4, until an alternative diagnosis is substantiated. OR An animal with a positive result from a sensitive and specific screening test, such as the Prionics test. *See* Section 2.2.1 for further details
Tracing	The process of locating animals, persons or other items that may be implicated in the spread of disease, so that appropriate action can be taken.
Transmissible spongiform encephalopathies	A group of diseases affecting various animal species, all of which diseases involve non-inflammatory vacuolated (spongiform) degeneration of the grey matter areas of the brain and spinal cord.

Vaccination	Inoculation of healthy individuals with weakened or attenuated strains of disease-causing agents to provide protection from disease.
Vaccine	Modified strains of disease-causing agents that, when inoculated, stimulate an immune response and provide protection from disease.
Vector	A living organism (frequently an arthropod) that transmits an infectious agent from one host to another. A *biological* vector is one in which the infectious agent must develop or multiply before becoming infective to a recipient host. A *mechanical* vector is one that transmits an infectious agent from one host to another but is not essential to the life cycle of the agent.
Veterinary investigation	An investigation of the diagnosis, pathology and epidemiology of the disease. *See also* Epidemiological investigation
Wild animals	
– native wildlife	Animals that are indigenous to Australia and may be susceptible to emergency animal diseases (eg bats, dingos and marsupials).
– feral animals	Domestic animals that have become wild (eg cats, horses, pigs).
– exotic fauna	Nondomestic animal species that are not indigenous to Australia (eg foxes).
Zoning	The process of defining disease-free and infected areas in accord with OIE guidelines, based on geopolitical boundaries and surveillance, in order to facilitate trade.
Zoonosis	A disease of animals that can be transmitted to humans.

Abbreviations

AAHL	Australian Animal Health Laboratory
ACDP	Advisory Committee on Dangerous Pathogens (UK)
ANZSDP	Australia and New Zealand Standard Diagnostic Protocols
AUSVETPLAN	Australian Veterinary Emergency Plan
BARB	'born after the real feed ban' (of cattle in the UK)
BSE	bovine spongiform encephalopathy
CCEAD	Consultative Committee on Emergency Animal Diseases
CJD	Creutzfeldt–Jakob disease
CNS	central nervous system
CSIRO	Commonwealth Scientific and Industrial Research Organisation
CVO	chief veterinary officer
DAFF	Department of Agriculture Fisheries and Forestry (Australian Government)
DCP	dangerous contact premises
FSANZ	Foods Standards Australia New Zealand
IP	infected premises
MBM	meat-and-bone meal
NAHIS	National Animal Health Information System
NHMRC	National Health and Medical Research Council
NLIS	National Livestock Identification System
NMG	national management group
NTSESP	National TSE Surveillance Program
OIE	World Organisation for Animal Health (Office International des Epizooties)
PIMC	Primary Industries Ministerial Council
PrP	host-encoded, highly conserved, normal cell membrane protein of unknown function
PrP^{Sc}	abnormal, protease-resistant isoform of PrP
SAFs	scrapie-associated fibrils
SP	suspect premises
SSC	Scientific Steering Committee (of the European Commission)
SRM	specified risk material
TSE	transmissible spongiform encephalopathy
vCJD	variant Creutzfeldt–Jakob disease
VLA	Veterinary Laboratory Agency of the UK

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Further reading and internet links

Transmissible Spongiform Encephalopathies Freedom Assurance Program (TSEFAP): Animal Health Australia
http://www.aahc.com.au/tsefap/index.htm

Numbers of BSE cases (worldwide)
http://www.oie.int/eng/info/en_esbmonde.htm

Office International des Epizooties (World Organisation for Animal Health, OIE)
http://www.oie.int/eng/info/en_statesb.htm

World Health Organization (WHO)
http://www.who.int/csr/disease/bse/en/

Joint WHO/FAO/OIE Technical Consultation on BSE: Public health, animal health and trade
http://www.oie.int/eng/publicat/ouvrages/a_110.htm

Information Concerning BSE for the Scientific World
http://sparc.airtime.co.uk/bse/welcome.htm (to January 2002)
http://www.priondata.org (since January 2002)

UK Department for Environment, Food and Rural Affairs (DEFRA)
http://www.defra.gov.uk/animalh/bse/directory.html

UK Creutzfeldt–Jakob Disease Surveillance Unit
http://www.cjd.ed.ac.uk

UK BSE Inquiry (2000)
http://www.bseinquiry.gov.uk

European Commission BSE website
http://europa.eu.int/comm/food/fs/bse/index_en.html

Scientific opinions

European Scientific Steering Committee
http://europa.eu.int/comm/food/fs/bse/scientific_advice_en.html

European Food Safety Authority
http://www.efsa.eu.int/science/tse_assessments/bse_tse/catindex_en.html

Video and training resources

A Tale of Transmission — Scrapie and BSE, Australian Animal Health Laboratory. Available from AAHL.

Brain Removal Techniques in Cattle and Sheep for TSE Surveillance. Available from Sedgwick Video Productions, 24 Broadbent Rd, Sedgwick, Vic 3551. Ph: 03 5439 6338; Fax: 03 5439 6464. Cost in 2001: $15 plus postage.

Clinical Findings in Cows with Bovine Spongiform Encephalopathy (BSE). Produced by the Clinic of Ruminant and Equine Medicine. Available from the Office of the Chief Veterinary Officer, Department of Agriculture, Fisheries and Forestry, GPO Box 858, Canberra, ACT 2601.

National TSE Surveillance Program — Methods for removing brains for TSE testing. Available from the Office of the Chief Veterinary Officer, Department of Agriculture, Fisheries and Forestry, GPO Box 858, Canberra, ACT 2601.

Prionics Test Trial Program — Methods for removing brains for testing with the Prionics rapid BSE test. Available from the Office of the Chief Veterinary Officer, Department of Agriculture, Fisheries and Forestry, GPO Box 858, Canberra, ACT 2601.

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