

**AUSTRALIAN VETERINARY EMERGENCY PLAN**

# **AUSVETPLAN**

**1998**

## **Disease Strategy**

### **Scrapie**

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

**Agriculture and Resource Management Council of Australia and New Zealand**

**This Disease Strategy forms part of:  
AUSVETPLAN Edition 2, 1996**

[AUSVETPLAN Edition 1, was published in 1991]

**This strategy will be reviewed regularly. Suggestions and recommendations for amendments should be forwarded to the AUSVETPLAN Coordinator (see Preface).**

**Record of amendments to this manual:**

[Insert record of amendments as necessary]

[AUSVETPLAN Edition 2 Edition was published in 1996]

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## PREFACE

This **Disease Strategy** for the control and eradication of **scrapie** is an integral part of the **Australian Veterinary Emergency Plan**, or AUSVETPLAN (Edition 2). AUSVETPLAN structures and functions are described in the **Summary Document**.

The strategy sets out the disease control principles that were approved by the Agriculture Resource Management Council of Australia and New Zealand (ARMCANZ) out-of-session in January 1996, for use in a veterinary emergency caused by the introduction of scrapie to Australia. This document has been amended to take into account developments that have occurred over the past few years.

Scrapie is designated as a List B disease by the Office International des Epizooties (OIE). List B 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 scrapie conform with the **OIE International Animal Health Code 1992** (OIE Code; see Appendix 3)

Scrapie is not included in the Commonwealth/States cost-sharing agreement for eradication of certain emergency animal diseases.

Detailed instructions for field implementation of the strategies are contained in the **AUSVETPLAN Operational Procedures Manuals** and **Management Manuals**. Cross-reference to strategies, manuals and other AUSVETPLAN documents are expressed in the form:

Document Name, Section no.

For example, **Decontamination Manual, Section 3**.

In addition, *Exotic Diseases of Animals: A Field Guide for Australian Veterinarians* by W.A. Geering, A.J. Forman and M.J. Nunn, Australian Government Publishing Service, Canberra, 1995 (**Exotic Diseases Field Guide**) is a source for some of the information about the aetiology, diagnosis and epidemiology of the disease and should be read in conjunction with this strategy.

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

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*The writing group was responsible for drafting this strategy. However, the text may have been amended at various stages of the consultation/approval process and the policies expressed in this version do not necessarily represent the views of all members of the writing group. Contributions may also have been made by other people not listed above and the assistance of all involved is gratefully acknowledged.*

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# 1 NATURE OF THE DISEASE

Scrapie is a transmissible spongiform encephalopathy of sheep and goats, characterised by a long incubation period followed by progressive degenerative disease of the nervous system and death. The disease has been known for over 200 years.

## 1.1 Aetiology

Scrapie is caused by an unconventional agent, that consists of an arrangements of an altered host protein, *prion*, which can cause modification of the same protein when it is produced by the host thus increasing the amount of the pathogenic protein present in the host cells.

Characteristic of these diseases is an accumulation of a protease-resistant form of a host cell membrane amyloid protein called PrP. Different strains of scrapie have been identified by studies in laboratory animals. The agent is extremely resistant to heat, irradiation and many chemical disinfectants.

No agent can be detected in any tissues from lambs up to 8 months of age. At 10–14 months of age low infectivity can be detected in the large masses of lymphoreticular tissue in the intestines (Peyer's patches), lymph nodes associated with the gastrointestinal tract and elsewhere, spleen and tonsil. The titres in these tissues increase subsequently and, before clinical signs appear, infectivity can be detected in the spinal cord, medulla and some other areas of the brain. By the time animals show clinical disease, levels of infectivity in the central nervous system, including the spinal cord, have risen above those in the lymphoreticular system (OIE 1991).

## 1.2 Susceptible species

Sheep and goats are susceptible to scrapie. However, while breeds of sheep vary significantly in their susceptibility to the disease, goat breeds appear to be universally susceptible.

Scrapie-free Merino and Poll Dorset sheep from Australia and New Zealand have normal frequencies of the scrapie-susceptible PrP genotypes. Also Cheviot and Suffolk sheep of scrapie-susceptible genotypes have been found in Australia and New Zealand.(Hunter)

While spongiform encephalopathies occur naturally in humans, there is no evidence that the scrapie agent is transmissible to humans.

## 1.3 World distribution and occurrence in Australia

Scrapie is present in several European countries, especially the United Kingdom; in Canada and the United States; and in Iceland, India, Japan and Brazil. There have been isolated reports of scrapie from a number of countries, including Australia (1952), New Zealand (1954) and the Republic of South Africa (1972). In these instances the disease was confined to imported sheep and was eradicated by destruction of the affected group.

## 1.4 Diagnostic criteria

[See Glossary for terms not defined in the text]

Laboratory examination of tissues collected at postmortem examination is essential to confirm a diagnosis. However, affected animals have characteristic clinical signs. The long incubation period results in clinical signs usually appearing when animals are between two and five years of age.

**Note:** It is advised that persons handling suspect animals or tissues take precautions to minimise the risks of exposure (see Section 2.2.5).

### 1.4.1 Clinical signs

The earliest signs of disease are reduced exercise tolerance, followed by the development of an unsteady gait. Animals go to water frequently but drink little and begin to rub, especially the poll, the buttocks and the rump. After about two months, animals start to lose condition, are ataxic and become rapidly fatigued. They are excitable and signs of localised rubbing are obvious from loss of wool or hair. A nibbling response can be elicited by rubbing alongside the spine over the rump. Often a papular rash appears on haired parts of the skin. By three or four months after the first signs, animals are severely affected showing marked muscle wastage, and are confused and agitated. Finally, during the next two to four weeks they become unable to stand and die.

The Australian Animal Health Laboratory (AAHL) video, *A Tale of Transmission*, provides a clear demonstration of the clinical signs of scrapie.

### 1.4.2 Pathology

There are no characteristic gross pathological changes but histopathology reveals spongiform changes in the brain and characteristic structures called scrapie-associated fibrils (SAFs) can be identified by electron microscopy.

### 1.4.3 Laboratory tests

Tests to detect scrapie in the live animal have been developed by dutch researchers (*New Scientist* - tonsil tissue, 1996) and US researchers (*Vet. Record* - the third eyelid, 1998).. The third eyelid test involves the biopsy of the lymphoid tissue of the third eyelid and detection of abnormal prion material using immunohistochemistry techniques. Prions have been detected well in advance of development of clinical disease in the animal in this lymphoid tissue.

Animal specimens collected should initially be sent to the State or Territory diagnostic laboratory from where they will be forwarded to the Australian Animal Health Laboratory (AAHL), Geelong for 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 (for transport of the specimens to Geelong).

### Specimens required/transport

The whole brain with the brainstem intact is removed from the skull of animals killed by intravenous barbiturate injection as soon as possible after death. A sample (3-10gm) of cervical spinal cord and/or medulla caudal to the obex, is frozen for possible detection of PrP<sup>Sc</sup> by Western blotting or as scrapie-associated fibrils (SAFs) by transmission electronmicroscopy. The rest of the brain, after appropriate microbiological sampling, is fixed in neutral buffered 10% formol saline for histological examination.

For information on the collection of specimens see Geering et al 1995. For further information see the **Laboratory Preparedness Manual, Section 6 and Appendix 3.**

### Laboratory diagnosis

*AAHL tests.* Currently laboratory diagnosis is based on histological changes in the brain (see Section 1.4.1, above). These tests can only be undertaken on tissues taken after death or euthanasia. Table 1 shows the tests for scrapie that are currently available at AAHL.



**Table 1 Diagnostic tests currently available at AAHL for scrapie**

Test	Specimen required	Test detects	Time taken to obtain result
Histopathology	brain tissue	characteristic spongiform changes	2 days
Electron microscopy	brain tissue	scrapie-associated fibrils	2 days
Isolation of agent by intracerebral inoculation into mice	brain tissue	scrapie agent	upwards to and beyond 1 year

**Notes:**

As brain material is required for diagnosis, animals should preferably not be shot through the head. Brain material should still be submitted, however, even if the animal has been shot through the head.

Source: Information provided by AAHL, 1995 [refer to AAHL for the most up-to-date information].

*Other tests.* The protease-resistant form of the host PrP protein can also be detected by immunological methods but this test is not currently available in Australia.

#### 1.4.4 Differential diagnosis

The following diseases must be considered in the differential diagnosis of scrapie:

- external parasites such as lice, mites, mange and sheep scab
- chronic enterotoxaemia (focal symmetrical encephalomalacia)
- maedi-visna
- polioencephalomalacia
- Aujeszky's disease
- louping ill
- listeriosis
- hepatic encephalopathy due to plant toxicity
- rabies

It is expected that an occurrence of scrapie would be associated with imported livestock. However contaminated veterinary therapeutics must also be considered a potential source of infection, along with contaminated surgical instruments. Where a contaminated therapeutic agent is the source of an outbreak, the disease may initially appear much more widespread than if the source was imported livestock.

## 1.5 Resistance and immunity

### 1.5.1 Innate and passive immunity

There is well-documented evidence of genetic control of susceptibility to scrapie in sheep. The incubation period in sheep and laboratory animals is under the control of a single genetic locus with alleles for short and long incubation periods. It is not clear whether this confers resistance to infection, or the animals have such a long incubation period that signs do not become evident during the lifespan of the animal. Flocks of sheep can vary in susceptibility and selection can lead to resistant flocks.

There is no evidence for passive immunity playing any part in resistance to scrapie.

### 1.5.2 Active immunity

There is no acquired immunity to the scrapie agent.

### 1.5.3 Vaccination

Vaccination is not applicable to this disease.

## 1.6 Epidemiology

The epidemiology of scrapie is determined principally by the long incubation period, host resistance factors and the mode of transmission. It is important to recognise that exposed animals, irrespective of their genetic susceptibility (see Section 1.5.1, above) may be infected.

### 1.6.1 Incubation period

The incubation period is prolonged. Following natural perinatal exposure, scrapie occurs most frequently between 2 and 5 years later, with a peak incidence at 3.5 years in sheep and somewhat less for goats.

### 1.6.2 Persistence of agent

#### Environment

The scrapie agent is highly resistant. It has been known to survive in a desiccated state for at least 30 months. Some infectivity remains after exposure to dry heat for 24 hours at 160°C. See Detwiler (1992) for further details.

The agent is resistant to most common disinfectants, including ethanol, formaldehyde, iodophors and phenolics (see Section 2.2.8). Absolute decontamination might be almost impossible but this may not be critical because it is extremely unlikely that a sufficient quantity of the agent would be ingested from environmental contamination compared to the higher concentration encountered during maternal transmission or by injection of veterinary product.

#### Live animals

Sheep infected with scrapie have infective material principally in the central nervous system tissues, lymphoreticular system and placenta. There is no immune response to eliminate the agent and, because the disease has a long incubation period, live clinically normal animals in the incubation period have been the principal source of introduction of disease into countries. Goats may have a more limited distribution of the agent in tissues.

#### Animal products and by-products

The scrapie agent survives carcass decomposition and many of the procedures involved in the processing of product. Thermal processing requires steam heating at 134–136°C for 18 minutes to inactivate the agent (Detwiler 1992).

### 1.6.3 Modes of transmission

#### Live animals

Scrapie is primarily transmitted from ewe (or doe) to offspring. This occurs either prenatally or else shortly after birth due to close contact between dam and offspring, probably via contaminated uterine fluids. Other horizontal spread occurs but is thought to be a relatively infrequent event, at least for sheep at pasture. Where lambing takes place under housed

conditions, horizontal spread to other adults and lambs is more common and there is even a possibility of spread through fomites. However, housing of ewes is not common in Australia. There is no evidence that cattle are infected by sheep at pasture (see Section 1.7 on risk of introduction to Australia.).

### **Artificial breeding**

The disease agent is not present in semen. Work is still being carried out on transmission via embryo transfer.

### **Biological products**

Spongiform encephalopathy agents can be spread by inoculation of biologically-derived therapeutic products (iatrogenic spread) such as:

- biological products derived from central nervous system extracts (in the same way as human pituitary gland extracts were contaminated with the agent for a human spongiform encephalopathy known as Creutzfeldt-Jakob disease); it is unlikely such a product would be legally imported and used
- a therapeutic agent that has incorporated a contaminated ingredient in manufacture (for example contaminated brain–heart infusion broth used as a substrate for a bacterial vaccine). While this poses a theoretical threat, there is no epidemiological evidence to suggest that this has been a source of scrapie elsewhere.

### **Fomites**

Transmission by fomites should not be a real concern. However, consideration should be given in particular to avoiding transmission on instruments used for veterinary applications and surgery.

### **Vectors**

There is no documented evidence for transmission by insect or arthropod vectors.

## **1.6.4 Factors influencing transmission**

The following factors could influence the transmission of scrapie:

- stocking densities
- housing of sheep during lambing
- breed
- feeding meatmeal

Natural spread of scrapie under Australian conditions is likely to be inefficient because ewe to offspring transmission is uncommon when lambing does not take place in housed conditions. However, spread of the agent by inoculation of a contaminated veterinary product would be much more efficient and an outbreak originating from such a source would affect many more animals in the flock at the one time.

## **1.7 Manner and risk of introduction**

Any occurrence of scrapie in Australia can be reasonably expected to be an isolated event associated with imported livestock. With the stringent quarantine requirements placed on sheep and goats imported into Australia and the previous use of the Scrapie Freedom Assurance Program, it is most unlikely that this disease will be introduced with legally-imported livestock.

Transmission from contaminated biologicals would require the source of the product to be outside Australia (unlikely to be approved), or for some contaminated material to be inadvertently included in a formulation (possible if the ingredient has passed through several suppliers).

## 2 PRINCIPLES OF CONTROL AND ERADICATION

The principles of control are:

- to slaughter and completely destroy all clinically affected-animals; and
- conduct a thorough risk assessment.

The key consideration is whether the disease is associated with imported animals or has been introduced with biological products (iatrogenic transmission). In conjunction with an outbreak control program, there would need to be an increased level of surveillance of the national flock. Such a surveillance program would be designed to accommodate probabilities of the agent infecting other properties on a local, regional, state and national level.

Difficulty in achieving complete inactivation of the agent raises some particular concerns. It is necessary to carry out intensive clean-up of areas identified as potentially heavily contaminated, eg necropsy sites and laboratories, because of the persistence of the agent in the environment. Efforts should be restricted to these areas.

### 2.1 Risk assessment

A risk assessment process aims to:

- determine the source of the outbreak;
- identify animals of equivalent risk status to the confirmed case(s); and
- classify the risk of infection in other groups of stock.

The following classifications are used.

- *Affected* animals — those showing clinical signs of scrapie.
- *Equivalent risk* animals — any imported sheep or goats originating from the same property as affected animals, the dams, progeny and litter mates of affected animals.
- *Exposed* animals — sheep or goats that have been in close physical contact with affected animals, especially around parturition; animals imported with the same group as affected animals; and animals exposed to invasive surgical equipment used on affected animals.
- *Low risk* animals — are sheep or goats on the same property as affected animals that are not derived from and that have not been in direct or indirect physical contact with those animals.

The risk assessment process will be dynamic and adjusted according to the results of monitoring of equivalent risk, exposed and low risk groups(suspect animals). Different classes of animals would need to be individually identified.

Based on this risk assessment, the eradication strategy would include:

- establishing adequate security on identified premises (see Section 2.2.1); and
- monitoring all deaths and maintenance of regular surveillance (see Sections 2.2.2 and 2.2.3);
- selective slaughter of stock (see Section 2.2.5); and
- developing a grazing management plan (see Section 2.2.10).

## **2.2 Methods to prevent spread and eliminate pathogens**

### **2.2.1 Quarantine and movement controls**

Infected premises (IPs; containing affected animals) and suspect premises (SPs; containing equivalent risk, exposed and low risk animals) should be placed under quarantine in the first instance. Further quarantine and movement controls depend on the outcome of risk assessment. Where property security and management is unsatisfactory consider controlled herd depopulation over a period of six to twelve months. Long-term quarantine of some groups of animals may be necessary (see Appendixes 1 and 2).

### **Zoning**

It is not anticipated that zoning is likely to be appropriate for scrapie. It would be expected that, when first detected, disease would be confined to one property or a few foci that could be readily isolated. In the event that the disease occurred in a geographically well defined area, zoning may be considered.

### **2.2.2 Tracing**

Tracing must be undertaken to establish the source of infection (especially associations with imported livestock) and determine the presence of other potentially-infected flocks. The order of priority should reflect the order of the risk categories listed above.

A major problem could arise in the event of implication of a contaminated therapeutic agent as the source of the disease. It is virtually impossible to confirm the contamination of a particular batch of the therapeutic agent, especially because of the long incubation period of the disease. Considerable industry cooperation would be required to determine the extent of the problem and to trace risks. The occurrence of widespread outbreaks may result in the disease being considered endemic.

### **2.2.3 Surveillance**

Suspect animals should be carefully examined at regular intervals to determine the presence of any characteristic clinical signs. Animals that develop clinical signs suggestive of scrapie must be subject to laboratory examination of the central nervous system. When animals over 2 years old are slaughtered, a statistically valid sample must be monitored for evidence of scrapie. Surveillance should be maintained for a prolonged period, bearing in mind the average incubation period of four years, and may need to be life-long. Heightened monitoring of the national flock should occur. Active surveillance will be necessary to instil confidence in overseas markets. Owners must be obliged to notify animal health authorities of any illness, death or impending movement of suspect animals.

### **2.2.4 Treatment of affected animals**

Treatment of animals is not possible.

### 2.2.5 Destruction of animals

Affected animals should be promptly slaughtered. This will remove real or perceived disease risks and allow a definitive diagnosis. Other risk categories could also be slaughtered, or placed under quarantine and close surveillance.

As brain material is required for diagnosis, animals should not be shot through the head. In addition, shooting will increase the risk of dissemination of the agent in the environment. It is recommended that any animal being destroyed for diagnosis be killed by the administration of intravenous barbiturate. Animals should be necropsied as close to the site of disposal as possible.

### 2.2.6 Treatment of animal products

Some experimental work has been conducted in the United Kingdom, to determine criteria for rendering carcasses that would ensure inactivation of the scrapie agent. The experiments are protracted and expensive and may never reach a satisfactory conclusion due to lack of sufficient infective material. It is unlikely that rendering would be an acceptable means of disposal in Australia because the temperatures and pressures used would not be high enough to completely inactivate the disease agent (see Section 1.6.2 and the **Red Meat Enterprises Manual**). Products assessed as being a significant risk should be disposed of by incineration (see Section 2.2.7).

Offal from suspect animals must not be used for production of meatmeal and bonemeal.

### 2.2.7 Disposal

Wherever possible, carcasses should be burned. The burning of carcasses must be under supervision of disease control authorities to ensure that it is performed appropriately and that all contaminated material is completely burned. In selecting a site, consideration should be given to the future use of the area.

Where burning is not practical, carcasses and other materials that cannot be adequately decontaminated should be buried in a site that will never be used for agricultural purposes (see the **Disposal Procedures Manual**). Consideration has to be given to the future use of the burial site, as the agent will remain in the soil for many months.

### 2.2.8 Decontamination

Decontamination procedures should be undertaken on any materials that are contaminated through close contact with infected carcasses, or exposed stock at parturition. It is not necessary, to impose farm gate disinfection of materials leaving an IP/SP.

Because of the difficulty of ensuring complete inactivation of the scrapie agent, it is largely impractical to treat products in order to decontaminate them. Products assessed as being a significant risk should be disposed of by incineration.

For items that will withstand steam sterilisation, and whose value justifies it, autoclaving at 134–136°C for at least 18 minutes is recommended (see Section 1.6.2). Steam sterilising at 121°C in the presence of 1 molar (M) sodium hydroxide is also effective (Taguchi et al 1991). Sodium hypochlorite at a concentration of 1.4% will achieve surface inactivation in 30 minutes (see also Geering et al 1995). The only other chemical decontamination that is

acceptable is exposure to 1M sodium hydroxide for at least 1 hour<sup>1</sup>. This will not guarantee absolute inactivation but has been shown to reduce infectivity by at least 6 logs (Brown et al 1986). Recent data from the United States (Ernst et al 1993) suggested that an acid phenolic compound called LpH is also very effective for surface decontamination. This product is no longer available but a similar product is presently under evaluation.

*Note: sodium hydroxide at 1M concentration is highly caustic and may cause damage to items being treated. It is also a hazard for operators who should wear eye protection. Any splashing on the skin should be followed by immediate rinsing with clean water. In the case of splashing in the eye, it should be irrigated with saline and medical attention sought immediately. However 1 M sodium hydroxide has been suggested as a skin wash and disinfectant for persons working with the materials derived from Creutzfeldt-Jakob disease cases. It has been stated that unbroken skin will tolerate this concentration for about 5 minutes (Brown et al, 1984, 1986).*

Formalin fixation of infected tissues stabilises the scrapie agent so that it cannot then be inactivated by steam sterilisation under the conditions described above. Exposure of formalin fixed tissues to 100% formic acid for 18 hours will allow safe processing for histopathology examination. Residues of formalin-fixed tissues should be disposed of by incineration. For further information see the **Decontamination Manual, Tables 2.6 and 3.15**.

### **2.2.9 Vaccination**

Not applicable.

### **2.2.10 Grazing management**

There is no evidence to support excretion of the scrapie agent into the environment other than via the placenta of infected ewes, and there is minimal risk of horizontal spread of scrapie other than under intensive husbandry systems. Confined areas associated with lambing ewes should be considered significantly contaminated. A grazing management plan should be developed to keep breeding and young stock away from high-risk areas. The appropriate strategy would be to identify grazing and management practices to minimise the risk of further transmission of the agent, for example grazing adult wethers, grazing cattle on the area or using crop rotations for a period. Where property security and management are unsatisfactory, controlled flock depopulation should be considered.

### **2.2.11 Wild animal control**

Carcases must be disposed of in such a way as to prevent access by wild carnivores and omnivores and by osteophagic cattle and sheep.

It is possible in some parts of Australia that scrapie could become endemic in the feral goat population. In the case of scrapie occurring in goats or sheep that have contact with feral goats, it would be important to establish some form of surveillance of feral goat populations to determine whether these animals are a reservoir of the agent.

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<sup>1</sup>Refer to Laboratory preparedness manual, Appendix 6.



### **2.2.12 Vector control**

Not applicable.

### **2.2.13 Public awareness**

Advice to the media must be carefully considered. It is necessary to inform the public, especially those in the livestock industries, of the circumstances of the outbreak and any trade implications. Despite scrapie being recognised for over 200 years, there is absolutely no evidence of transmission to humans. It is possible that fear could be raised about human health issues related to consumption of meat, however, and media sources could draw parallels with concerns raised in the United Kingdom because of BSE.

Early discussions with human health authorities is advisable, in order to ensure that a consistent public health position is developed. In addition, advice should be given that products regarded as a significant risk of containing the agent would not be allowed to enter the food chain. These assurances cannot be provided with surety if the source of the disease is a therapeutic agent that has been widely used for some time. There may be some additional concern if the ingredient implicated as the source of the disease has been used to prepare human therapeutics.

There is also likely to be a considerable level of public concern if major trading partners decline to accept Australian exports. A perception that the domestic market is being supplied with an unsafe product is likely to arise. Media liaison officers need to be prepared for this issue.

For further information see the **Public Relations Manual**.

## **2.3 Safety precautions**

There has been no evidence of transmission of scrapie to humans (see Kimberlin 1992 for a discussion of this issue). However, persons involved in handling potentially infected material must take adequate precautions to avoid exposure to these agents. Veterinarians, laboratory workers and slaughterhouse workers should wear gloves and eye protection when handling tissues suspected of containing high levels of the agent.

Care should be taken to minimise environmental contamination during necropsy procedures. Carcasses should be disposed of carefully and instruments thoroughly decontaminated. More extensive decontamination of the environment should not be necessary and must be assessed on a case-by-case basis.

Meat and meat products from affected animals must not enter the animal or human food chain. ARMCANZ agreed in 1997 that mammalian-derived proteins should not be fed to ruminants.

## **2.4 Feasibility of control in Australia**

Where there is an occurrence of scrapie in Australia that can be linked to an introduced animal, and where forward tracing can identify all other potentially infected stock, eradication of the disease will almost certainly be achievable. If scrapie was found to be widespread and occurrences could not be linked to each other, control of the disease could be very difficult with current diagnostic technology. A specific policy and prolonged program of surveillance would then be necessary to achieve control and eradication.

If cases of scrapie occur in imported animals located in a quarantine station (ie a 'quarantine incident'), there is the option of using either the Commonwealth *Quarantine Act 1908* or the relevant State disease control legislation to effect control measures.

## 3 POLICY AND RATIONALE

### 3.1 Overall policy for scrapie

Scrapie is an OIE List B disease that is invariably fatal in clinically-affected sheep and goats, and is significant in the international trade in sheep and goats, and their products.

The policy is to eradicate the disease as quickly as possible using a combination of strategies including:

- ☞ *a total management plan* to focus the action on risk animals and to maximise the efficiency of the eradication program;
- ☞ *slaughter and sanitary disposal* of all clinically-affected, exposed and equivalent-risk stock (note – these terms have been specifically defined in Section 2.1);
- ☞ *depopulation* (partial or complete) should be undertaken if doubt exists concerning the status of animals in the herd;
- ☞ *quarantine* of the stock (sheep or goats) on infected and suspect premises;
- ☞ *tracing and surveillance* to define the limits of the incident and provide additional evidence to re-establish free status;
- ☞ *risk assessment* to identify stock (sheep and goats) that have had any opportunity to acquire infection because of contact or lineage with confirmed cases; and
- ☞ *a public awareness campaign* to facilitate cooperation from industry and the community.

As any infected or suspect premises should be placed in individual quarantine, it is not necessary to establish a restricted or control area.

There will be some international restrictions on the export trade of live sheep and goats, particularly breeding animals, and possible disruption to the sheep and goat meat trade may occur.

Scrapie is not included in the Commonwealth/States cost-sharing agreement.

The CVO(s) in the State(s)/Territory(s) in which the outbreak(s) occurs will be responsible for implementing disease control measures (in accordance with relevant legislation), and will make ongoing decisions on follow-up disease control measures in consultation with the Consultative Committee on Emergency Animal Diseases (CCEAD), the State/Territory and Commonwealth governments, and representatives of the affected industries. 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/Territory disease control headquarters and local disease control centre(s), see the **Control Centres Management Manual, Part 1, Sections 3 and 4**.

## 3.2 Strategy for control and eradication

The general strategy will be to destroy and dispose of affected animals immediately, quarantine the infected premises, and undertake tracing and a risk assessment of the information obtained so that more detailed eradication planning decisions can be implemented. As scrapie is spread slowly between animals it is not essential for plans to be made in haste and strategies should be carefully planned, based on risk assessment and surveillance. Suspect properties identified by tracing will need to be quarantined and an appropriate level of movement controls placed on animals and products.

The strategy of slaughter of affected animals; quarantine and movement controls; tracing and risk assessment; laboratory evaluation; and decontamination, will be used regardless of the source of infection. When the source of the introduction has been determined the extent of the actions required will be decided. Hygiene and good farm management practices, and progressive destocking will be employed where the disease may be present in specific groups on particular farms.

As the presence of the disease will lead to national and international disruption to trade, regular and ongoing liaison with industry, the media, public health officials and the public is an integral part of the strategy for eradication.

### 3.2.1 Stamping out

Stamping out may be adopted for some groups to overcome long and costly ongoing surveillance and quarantine, or due to international and/or domestic trade issues.

The occurrence of disease in one or a few flocks/herds should be addressed by slaughter of all affected sheep and goats. Risk assessment will then define further actions and strategies. This may include slaughter and laboratory assessment of high risk groups (that is equivalent risk, and exposed animals; see Section 2.1). Further actions would be defined by findings from these animals.

In the event of an outbreak of scrapie not traceable to imported animals, the source of the infection should be determined and investigations conducted (including the slaughter of sufficient animals) with a view to establishing the distribution and prevalence of infection.

Management options for disposal should be left as flexible as possible in order that disease control authorities, industry bodies and owners can reach the most cost-effective option for the industry and the community as a whole. The best option will, to some extent, be determined by the source and extent of the outbreak.

It might also be more expedient to slaughter a small number of suspect animals rather than maintain them under costly and disruptive prolonged surveillance and quarantine or when property management practices and security cannot be maintained at a level that can ensure eradication.

### **3.2.2 Quarantine and movement controls**

The initial infected premises and any suspect premises will be placed into immediate quarantine while tracing and the epidemiological investigations are being completed and movement controls introduced. The ongoing level of quarantine and controls on movement will be determined by epidemiological investigations and the risk classification of the animals involved on the various premises. The declaration of restricted and control areas for scrapie is unwarranted.

It is possible that on some premises there may be different risk categories of animals that will be subject to different management practices and controls. For further information see Appendixes 1 and 2.

#### **Zoning**

It is most unlikely that zoning of geographic areas would be appropriate, unless a widespread but geographically-defined outbreak of scrapie was discovered.

### **3.2.3 Treatment of affected animals**

Infected animals do not respond to treatment.

### **3.2.4 Treatment of animal products and by-products**

Because of the high resistance of the scrapie agent to treatments it is not appropriate to attempt to treat edible animal products.

By-products also cannot be satisfactorily treated under most methods of commercial production and products from infected animals must be destroyed by incineration or burial. The distribution of products from other animals on declared premises will be dependent on the risk classification.

### **3.2.5 Vaccination**

Vaccination is not appropriate to this disease.

### **3.2.6 Decontamination**

Decontamination of the scrapie agent is difficult but is required for items or materials that have become contaminated through close contact with infected animals or carcasses. Effective surface disinfectants are available but should be used with caution. Good hygiene and farm management practices should be introduced for contaminated areas.

### **3.2.7 Tracing and surveillance**

Tracing and surveillance will be used to attempt to define the source of the disease and the extent of spread. The priority given to both tracing and surveillance will depend to a large extent on the risk classification of the affected animals. Even in the event of the infection

being introduced through importation of an animal or genetic material it is possible that, because of its long incubation period, the disease could be widespread by the time it is detected.

It is likely that if a single contaminated therapeutic administration has been the source of the disease, most infected animals will have similar degenerative neurological changes. Information about the attack rate will assist in risk assessment of the index flock and surveillance of other flocks exposed to the same product.

Failure to identify a source would preclude immediate risk assessment of the index flock and necessitate more widespread investigation and surveillance before developing a control/eradication strategy.

### **3.2.8 Media and public relations**

Special attention needs to be paid to media and public relations. It is highly likely that the disease will be linked to bovine spongiform encephalopathy and be widely publicised by the media as a potential human health risk, leading to unnecessary public alarm, with significant effect on domestic and international trade. This needs to be countered with a clear, easily understood explanation and information on the disease.

## **3.3 Social and economic effects**

A major implication of an occurrence of scrapie in Australia would be the costs associated with restrictions on Australia's international trade in livestock and livestock products. The selected strategy must address the concerns of major trading partners. It may be necessary to take actions and impose restrictions that are not technically justifiable in order to satisfy the requirements of trading partners.

Domestic consumer markets could also have concerns about product safety because of the link to BSE. This could affect local sales of sheep and goat meat and dairy products, and programs to counter these concerns will need to be in place. Processing establishments could be affected in the short term while consumer confidence in sheep and goat products is being re-established.

An isolated occurrence of the disease in an imported flock to which we responded promptly should have minimal impact.

## **3.4 Criteria for proof of freedom**

It is necessary to take rapid and decisive action in disposing of affected animals, and possibly high risk animals, immediately implementing a thorough investigation to determine the source of infection and other suspect animals and impose strict quarantine and movement controls in the first instance.

Programs, to ensure producers and other agricultural workers are aware of the signs of the disease, are reporting suspicious cases, and that these are fully investigated, must be in place.

Proof of freedom after an outbreak can only be achieved by absence of clinical disease for 5 years, with routine laboratory examination of any suspect cases to exclude this disease. Active surveillance and monitoring based on histological examination of brains should be carried out to support a declaration of freedom.

### **3.5 Funding and compensation**

As Scrapie is not included in the Commonwealth/States cost-sharing agreement for the eradication of certain emergency animal diseases, funds to pay the costs of eradication, including compensation, will have to be found from other sources. Possible sources are:

- State government funds;
- State disease compensation funds;
- Commonwealth government funds; and
- special industry levies;
- other agreed arrangements.

Alternatively, the costs and losses might have to be borne by individual owners.

### **3.6 Strategy if the disease becomes established**

If scrapie becomes established in Australia the strategy for controlling it may vary, depending on the distribution (geographic, species, breed and even breeding line) and the perceived mode of transmission. Establishment of scrapie within a breed or breeding line of sheep could be addressed by gradual elimination of that line from the national flock, or by breeding for resistant offspring. Due to the nature of scrapie, there will be adequate time to plan the strategies in the event that scrapie becomes endemic.

Any strategy to eradicate the disease in the event of it becoming established will require drastic action in relation to specific risk categories of animals with possible wider stamping out and restrictive controls on animal and product movements.

A widespread dissemination of scrapie by inoculation of a contaminated therapeutic agent would require a protracted program of monitoring and a high level of industry cooperation to eliminate the disease. It is possible that the nature of sheep farming in Australia might reduce the likelihood of transmission of the disease in the field. Certainly eradication of the disease would require extensive record keeping by commercial farmers, and a program of accreditation for studs might assist the process.

## APPENDIX 1 Guidelines for classifying declared areas

### Infected premises (IP)

A premises classified as an IP will be a defined area (which may be a part of a property) on which a case of scrapie has been confirmed or is believed to exist.

### Dangerous contact premises (DCP)

Not applicable.

### Suspect premises (SP)

Premises classified as SPs will be those containing (see Section 2.1):

- *Equivalent risk* animals — any imported sheep or goats originating from the same property as affected animals, the dams, progeny and litter mates of affected animals.
- *Exposed* animals — sheep or goats that have been in close physical contact with affected animals, especially around parturition; animals imported with the same group as affected animals; and animals exposed to invasive surgical equipment used on affected animals.
- *Low risk animals* — sheep or goats on the same property as affected animals that are not derived from and that have not been in close physical contact with those animals.

### Restricted area (RA)

Not applicable.

### Control area (CA)

Not applicable.



## APPENDIX 2 Recommended quarantine and movement controls

### Infected premises

#### *Movement out of susceptible animals:*

Movement dependent on risk assessment

#### *Movement in of susceptible animals:*

No restriction but advise owner of implications

#### *Movement out of specified products:*

May need to restrict — depending on circumstances and as deemed necessary

#### *Movement out of other animals:*

Control movement of sheep and goats.

#### *Movement in and out of people:*

No restriction

#### *Movement in and out of vehicles and equipment:*

No restriction apart from equipment that has been in contact with slaughtered animals. Equipment will require decontamination first.

#### *Movement out of crops and grains:*

No restriction.

### Suspect premises

Movement dependent on risk assessment.

No restriction but advise owner of implications

## **APPENDIX 3 OIE International Animal Health Code for scrapie**

Scrapie is a List B disease but has no specific chapter in the code.

## **APPENDIX 4 Procedures for surveillance and proof of freedom**

All animals regarded as at risk of significant exposure must be examined at the time of disposal for the presence of changes in the central nervous system. Decisions on the status of the affected group can then be updated. The procedure for sampling the central nervous system is that published in Geering et al (1995). A structured survey or surveillance by abattoir sampling may be necessary to assure trading partners of the limited nature of the outbreak. The numbers of brains required for examination would need to be determined at the time. Such brains could be processed through regional veterinary laboratories and examined by specifically-certified veterinary pathologists. Demonstration of protease-resistant fibrils to confirm a histological diagnosis could be carried out at AAHL, or at the central veterinary laboratory of each state, depending on the extent of the problem.

## GLOSSARY

Allele	One of the alternative forms of a specified gene. Different alleles usually have different affects on the phenotype.
Animal by-products	Products of animal origin destined for industrial use, eg raw hides and skins, fur, wool, hair, feathers, hooves, bones, fertiliser.
Animal products	Meat products and products of animal origin (eg eggs, milk) for human consumption or for use in animal feeding.
AUSVETPLAN	A series of documents that describe the Australian response to emergency animal diseases linking policy, strategies, implementation, coordination and counter-disaster plans.
Biological products	Reagents of biological origin (eg sera, hormones) for therapeutic use in the diagnosis or treatment of certain diseases.
Consultative Committee on Emergency Animal Diseases	A committee of State/Territory CVOs, AAHL and CSIRO, chaired by the CVO of Australia (Cwlth DPIE), to consult in emergencies due to the introduction of an exotic disease of livestock, or serious epizootics of Australian origin.
Control area	A larger area (bigger than a restricted area) where movement restrictions will reduce the chance of the disease spreading further afield ( <i>see</i> Appendix 1).
Declared area	A defined tract of land for the time being subject to disease control restrictions under emergency disease legislation. Types of declared areas include <i>restricted area</i> ; <i>control area</i> ; <i>infected premises</i> ; and <i>dangerous contact premises</i> .
Decontamination	Includes all stages of cleaning and disinfection.
Disposal	Sanitary removal of animal carcasses and things by burial, burning or some other process so as to prevent the spread of disease.
Emergency animal disease	Includes emergency animal diseases and endemic diseases that warrant a national emergency response.
Fomites	Inanimate objects (eg surgical equipment) that can carry the emergency agent and spread the disease through mechanical transmission.
Genetic locus	The genetic position (on a chromosome) occupied by the alleles of a specified gene.
Iatrogenic disease	A disease caused by medical/veterinary procedures, usually occurring as a side effect of pharmacological agents
Incubation period	The period which elapses between the introduction of the pathogen into the animal and the occurrence of the first clinical signs of the disease.
Index flock	The first or original flock in which a case of the disease has been diagnosed (also, index case; index property).
Infected premises	<i>see</i> Appendix 1
Local disease control centre	An emergency operations centre responsible for the command and control of field operations in a defined area.
Movement controls	Restrictions placed on movement of animals, people and things to prevent spread of disease.
Premises	A defined area or structure, which may include part or all of a farm, enterprise or other private or public land, building or property.
Quarantine	Legal restrictions imposed on a place, animal, vehicle or other things limiting movement.
Rendering (of carcasses)	Processing by heat to inactivate infective agents. Rendered material may be used in various products according to particular disease circumstances.

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Restricted area	A relatively small declared area (compared to a control area) around an infected premises that is subject to intense surveillance and movement controls ( <i>see</i> Appendix 1).
Risk assessment	Assessment of the relative likelihood of an event, taking into consideration all relevant available and unavailable information.
Sentinel animals	Animals of known health status used for detecting the presence of a specific emergency disease agent.
Spongiform encephalopathies	A group of diseases affecting various animal species all of which involve non-inflammatory vacuolated (spongiform) degeneration of the grey matter areas of the brain and spinal cord.
Stamping out	Eradication procedures based on quarantine and slaughter of all animals infected or exposed to infections.
State/Territory disease control headquarters	The emergency operations centre that directs the disease control operations to be undertaken in the State/Territory.
Surveillance	A systematic program of inspection and examination of animals or things to determine the presence or absence of an emergency disease.
Susceptible animals	Animals that can be infected with the disease (for scrapie—sheep and goats)
Suspect premises	<i>see</i> Appendix 1
Tracing	The process of locating animals, persons or things that may be implicated in the spread of disease, so that appropriate action be taken.
Viraemia	The presence of viruses in the blood.
Zoning	The process of defining disease-free and infected areas in accord with OIE guidelines, in order to facilitate trade.
Zoonosis	Disease transmissible between animals and people.

## Abbreviations

AAHL	CSIRO Australian Animal Health Laboratory, Geelong
ARMCANZ	Agriculture and Resource Management Council of Australia and New Zealand
BSE	Bovine spongiform encephalopathy
CVO	Chief veterinary officer
IP	Infected premises
OIE	World Organisation for Animal Health [Office International des Epizooties]
PrP	Protease-resistant form of host cell membrane protein
SAF	Scrapie-associated fibrils
SP	Suspect premises

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## Video/training resources

*A Tale of Transmission — Scrapie and BSE*, AAHL available from the Emergency Disease Strategies Section (formerly the Foreign Diseases Unit), DPIE, Canberra; or AAHL

## OIE publications

- OIE Code (1992). *International Animal Health Code* (6th edition), OIE, Paris, France.
- OIE Manual (1992). *Manual of Standards for Diagnostic Tests and Vaccines* (2nd edition), OIE, Paris, France.
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