



MAF Biosecurity New Zealand
and New Zealand Food Safety
Authority Policy Statement on
responding to a confirmed case
of 'atypical scrapie' in New
Zealand
(September 2009)

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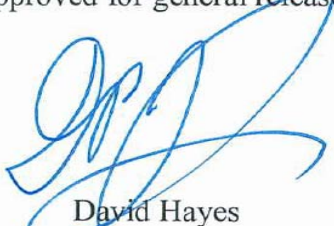
Market Access
New Zealand Food Safety Authority



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Zealand Food Safety Authority Statement
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Approved for general release



David Hayes
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MAF Biosecurity New Zealand

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Tony Zohrab
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Consultation for this response policy statement

A consultation draft precursor to this Response Statement was tabled for endorsement at the TSE Industry Liaison Forum in Wellington on Thursday 24th September 2009. Representation at the Forum comprises:

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Richard Ivess		Ministry of Foreign Affairs & Trade
Pat Helm		Department of Prime Minister & Cabinet
Tony Robinson		Ministry of Research, Science & Technology
Grant Storey		Ministry of Health
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Executive Summary

This document describes how MAF Biosecurity New Zealand and the New Zealand Food Safety Authority will respond in the event of confirmation of a case of atypical scrapie in New Zealand. Development of this updated policy has been guided by the Policy for MAF's Responses to Risk Organisms, the World Organisation for Animal Health's (OIE) Terrestrial Animal Health Code, other information on the global regulatory environment and the latest available scientific evidence.

Atypical scrapie is a transmissible spongiform encephalopathy (TSE) of older sheep and goats which is usually detected without obvious clinical signs in animals sent to slaughter. The OIE's Terrestrial Animal Health Code states that *'atypical' scrapie which is clinically, pathologically, biochemically and epidemiologically unrelated to 'classical' scrapie, may not be contagious and may, in fact, be a spontaneous degenerative condition of older sheep'*. Atypical scrapie has never been detected in animals in New Zealand and the condition is Notifiable and was listed as an Unwanted Organism under the Biosecurity Act (1993) in November 2006.

Atypical scrapie is not considered to be a risk to human health, either via consumption of sheep meat or occupational exposure and has been clearly demonstrated not to be BSE in sheep. This and the observation that atypical scrapie does not present a significant animal health or welfare problem, means that any economic impacts associated with detection of the condition in New Zealand are likely to be limited. However, regulatory actions overseas surrounding BSE and initial ambiguity about the relationship between atypical scrapie and scrapie, has led some countries to undertake regulatory interventions in response to atypical scrapie. For these reasons, developing a specific policy for how MAFBNZ and NZFSA will respond if a case is detected was considered appropriate.

In formulating our policy for responding to a detection of atypical scrapie in New Zealand, MAFBNZ and NZFSA considered three available options, including a range of direct interventions to investigate and manage the condition on farms. Direct intervention is not considered appropriate given that the likely impacts expected to be realised are not substantial and that effective control measures are not available. Rather, MAFBNZ and NZFSA believe that risks arising from any detection can be adequately and most appropriately managed using a suite of communication and liaison focussed activities.

In the absence of any recognised human or animal health issues as a consequence of atypical scrapie, the agreed objectives of any response will include:

- keeping stakeholders and the public informed about the agent and any implications of the find
- allay any public concerns about perceived health impacts of atypical scrapie
- minimising trade impacts
- endeavour to minimise impacts on any property or the owner(s) of the properties where the case is confirmed to have originated from or been present
- identify any research projects that may be required to fill in the gaps in local knowledge

These objectives will be met by undertaking a range of defined activities, in accordance with the *Policy for MAF's Responses to Risk Organisms* and will be implemented using

management procedures described in MAFBNZ's *Biosecurity Response Processes and Procedures*.

The evolving global regulatory environment necessitates that provision for a certain degree of flexibility of response is maintained. Importantly, the magnitude of any economic impacts arising from a detection of atypical scrapie in New Zealand will ultimately be dependent on market reactions. Accordingly, MAFBNZ and NZFSA reserve the right to amend this policy when and how it is considered appropriate to do so.

1. MAF Biosecurity New Zealand and NZFSA – our role

1.1 Our mission

MAF Biosecurity New Zealand's mission is to protect New Zealand's natural advantage by making timely and informed risk management decisions and delivering effective interventions.

The New Zealand Food Safety Authority's mandate is to protect consumers by providing an effective food regulatory programme covering food produced and consumed in New Zealand as well as imports and exports of food and related products.

1.2 MAF's response policy

MAF has responsibility for leading a fully integrated, transparent and efficient biosecurity system, MAF is the lead agency for biosecurity and new organism incursions. The approach is described in *The Policy for MAF's Responses to Risk Organisms¹* released in July 2008 (the *Response Policy*).

MAF will investigate all suspected risk organisms that pose emerging threats to New Zealand's people, environment and/or economic values. We will consider urgent measures where these preserve the range of potential response options. MAF will lead the response where the response has significant public benefit.

We use the generic questions and criteria set out in the Decisions Framework (Appendix Five of the *Response Policy*) and detailed in procedures developed under this policy to evaluate the significance of the risks posed using the best available information.

MAF works closely with the New Zealand Food Safety Authority on biosecurity events that have trade and/or food safety implications. The New Zealand Food Safety Authority protects and promotes public health and safety and provides both the animal health and public health assurances (certification) that facilitates the access to markets for New Zealand food and food related projects. NZFSA also lead the trade response on biosecurity events that have trade and/or food safety implications.

To the extent possible within time and resource constraints, MAF will consult and consider the views of those individuals/organisations, including Māori, or groups representative of these parties, that are:

- a) affected or potentially affected by the risk organism;
- b) affected or potentially affected by the proposed response options; or
- c) able to contribute to the development or execution of the response.

¹ <http://www.biosecurity.govt.nz/biosec/pol/statements/response-policy>

MAF will often make decisions on investigation and urgent measures before being able to consult fully and will have to manage uncertainties such as organism identification. Our principles in this situation are to:

- keep options open;
- be transparent about the decision;
- set up a process and timeframe to engage stakeholders and review decisions;
- reduce overall impact on New Zealand's values; and
- use the Co-ordinated Incident Management System so we can more easily work with other agencies in an emergency.

1.3 The response policy for atypical scrapie

The *Policy for MAF's Responses to Risk Organisms* indicates that in some cases MAF and other agencies may develop readiness plans and / or policies directed at specific biosecurity risks or issues.

In this case, previous events in 2007 surrounding the detection of atypical scrapie in three sheep in the UK, two from parents imported from New Zealand, one imported from New Zealand² prompted MAFBNZ and the NZFSA to consider the issue in detail and prepare a specific, atypical scrapie response policy. This document represents a statement of policy, derived from this earlier analysis, taking into account subsequent advances in scientific understanding of atypical scrapie and developments in the global animal health regulatory arena. Most notable among these is the clear distinction between atypical scrapie and scrapie that has been made by the OIE, based on new scientific information.

² Simmons H. A, Simmons M, Spencer Y.I., Chaplin M. J., Povey G., Davis A., Ortiz-Pelaez A., Hunter N., Matthews D. and Wrathall A. (2009). Atypical scrapie in sheep from a UK research flock which is free from classical scrapie. *BMC Veterinary Research* 5:8

2. Atypical scrapie

2.1 Atypical Scrapie is not scrapie

In 2002, the European Union developed a TSE surveillance programme to establish the prevalence of scrapie and BSE in small ruminants. This surveillance led to the identification of a novel and previously uncharacterized pathology of sheep brains, known as ‘atypical scrapie’. The relationship between atypical scrapie and scrapie was initially uncertain (hence the name ‘atypical’) but analysis of subsequently generated scientific data has led the OIE to conclude that “*‘atypical’ scrapie which is clinically, pathologically, biochemically and epidemiologically unrelated to ‘classical’ scrapie, may not be contagious and may, in fact, be a spontaneous degenerative condition of older sheep*”³. So, if atypical scrapie is very definitely not scrapie – what is it?

2.2 What is atypical scrapie?

Atypical scrapie has been clearly demonstrated not to be ‘BSE in sheep’ and is a condition of older sheep and goats which is usually detected without obvious clinical signs in animals sent to slaughter. In the few reports of cases with clinical signs these have been variable and usually neurological in nature. In most instances there is changed demeanor, tremors, occasionally circling, gait abnormalities and postural changes, ataxia and loss of body condition. Pruritus (intense itchiness) is not a feature of atypical scrapie. The suspected causative agent is a unique form of prion protein (PrP^{Sc}) deposited mostly in the cerebellum but also in other areas of the brain stem and cerebrum.. These PrP^{Sc} deposits in turn are thought to result in degenerative changes, leading to the clinical signs seen in a minority of cases. The variation in clinical signs is believed to be attributable to differences in where the abnormal prions are deposited.

2.3 Where is atypical scrapie?

The majority of the cases have been identified in clinically normal sheep sampled at slaughter. Cases of atypical scrapie have been reported from Norway, Sweden, Finland, the UK, Germany, France, Portugal, Belgium, the Netherlands, Ireland, Denmark, United States, Canada and Falkland Islands. The apparent prevalence of the cases in the EU Member States is very low – for example for the UK it is less than 1 in 1000 tested animals (adult sheep with at least two teeth in wear, i.e. 18 months of age or older).

The diagnosis of cases throughout the EU and in the Falklands suggests that atypical scrapie is a previously unrecognized entity rather than a new condition. Like New Zealand, the Falkland Islands and Denmark are countries that are considered to be free of classical scrapie. The Falkland Islands have a similar sheep importation history as New Zealand with original importations from the UK and, since the mid 1980s, importations just from New Zealand and Australia.- it might therefore, be considered surprising that atypical scrapie has not been seen

³ World Organisation for Animal Health (OIE). Terrestrial Animal Health Code 2009 - Scrapie Chapter. http://www.oie.int/eng/normes/mcode/en_chapitre_1.14.9.htm#rubrique_tremblante_commerce

to occur in New Zealand. It appears that wherever intensive surveillance has been conducted on large numbers of sheep, atypical scrapie has been found.

2.4 Is atypical scrapie transmissible?

Atypical scrapie is not known to be naturally transmissible, i.e. does not spread between animals naturally, in contrast with many other infectious livestock diseases and some other transmissible spongiform encephalopathies like scrapie. Most cases of atypical scrapie are typically found as a single case in a flock. Even when authorities have returned to flocks where a case has been found and slaughtered and tested all the remaining flock members, there are usually no further cases. This and detailed epidemiological investigations in Norway, Great Britain and France have all found no evidence to support an infectious aetiology (cause). This is a particularly important aspect of the biology of atypical scrapie when considering the merits and utility of potential control options.

2.5 Status of atypical scrapie in New Zealand.

Scrapie is an exotic disease in New Zealand and has been notifiable since 1955. It is included in the Biosecurity (Notifiable Organisms) Order 2006 under the category of ‘transmissible spongiform encephalopathy agents’. Atypical scrapie was listed as an Unwanted Organism under the Biosecurity Act 1993 in November 2006.

However, there are two observations that could support the possibility that atypical scrapie may be present in the New Zealand national flock:

- atypical scrapie has been reported in countries that have never had classical scrapie;
- and
- New Zealand sheep have a high prevalence of susceptible genotypes for atypical scrapie.

If cases of atypical scrapie have previously occurred in New Zealand’s sheep flock, there may be several reasons why it has not be previously detected. These include:

- the disease has a very low prevalence and may not have been detected by the routine surveillance processes
- the age structure of the New Zealand flock is young by international standards
- the likelihood of samples being collected is low as it is usually asymptomatic, rare and a sporadic condition
- the condition may not have been previously detected in New Zealand as the histopathological signs are often mild and inconclusive.
- The screening tests used for brains collected at slaughter did not detect this entity.
- Affected sheep usually show no clinical disease or non-specific signs.

2.6 Impacts of atypical scrapie overseas

Impacts associated with atypical scrapie are experienced in countries where direct regulatory intervention is undertaken. These can be seen as impacts of the measures themselves, rather than the condition, and there is no scientific basis for the measures.

Most cases have been detected in animals at slaughter or in fallen stock (found dead) by a small number of countries that have introduced BSE-related testing at the time of slaughter. Animals detected at slaughter were typically healthy and would not have suffered any on farm production loss up to this time. If the carcass was condemned there will be a loss but because of the low prevalence this loss is infrequent.

3. Response to a confirmed case of atypical scrapie in New Zealand

3.1 Case definition for atypical scrapie

The World Organisation for Animal Health (OIE) states that “*atypical’ scrapie which is clinically, pathologically, biochemically and epidemiologically unrelated to ‘classical’ scrapie, may not be contagious and may, in fact, be a spontaneous degenerative condition of older sheep*”.

The following cases definitions will be used to determine when a case of atypical scrapie has been detected:

Suspect animal

- Animal with histopathological lesions that cannot rule out ATS, as assessed by reference pathologist; and / or
- Screening ELISA (BioRad TSE ELISA) positive on brain stem and/or cerebellum sample.

Confirmed infected animal

- Animal with histopathological lesions that cannot rule out ATS, as assessed by reference pathologist; and / or
- Screening ELISA (BioRad TSE ELISA) positive on brain stem and/or cerebellum sample; and
- Confirmatory Western Blot (BioRad) positive on brainstem and/or cerebellum sample; and for the first case in New Zealand
- Immunohistochemistry (IHC) undertaken by world reference laboratory (VLA Weybridge) confirms presence of ATS prion protein

3.2 Risk management policy for a confirmed case of atypical scrapie in New Zealand

The decision about what response is most appropriate following any detection of atypical scrapie in New Zealand has been informed by *Policy for MAF's Responses to Risk Organisms*.

If atypical scrapie is found in New Zealand then it is:

- likely that it has been present for many years, given the highly stringent border measures historically and currently in operation and the low detection probability of current surveillance.

and / or

- likely to have arisen naturally, as the OIE suggests may occur

An analysis of the likely impacts on New Zealand's values indicates that collectively, these are unlikely to be substantial and can be most appropriately managed with communications and liaison activities and appropriate routine epidemiological follow-up. No direct operational intervention would be undertaken by regulatory authorities.

This is the preferred response to the identification of a case of atypical scrapie in (or directly associated with) New Zealand.

MAF Biosecurity New Zealand and the New Zealand Food Safety Authority **will respond** to a confirmed case of atypical scrapie in or closely associated with New Zealand.

The objectives of this response will be to:

- keep stakeholders and the public informed about the agent and any implications of the find (NZFSA and MAFBNZ)
- allay any public concerns about perceived health impacts of atypical scrapie (NZFSA and MAFBNZ)
- minimise trade impacts (NZFSA)
- endeavour to minimise impacts on any property or the owner(s) of the properties where the case is confirmed to have originated from or been present (MAFBNZ in relation to the this policy. NZFSA in relation to any trade impacts).
- identify any research projects that may be required to fill in the gaps in local knowledge (MAFBNZ)

The response will be conducted in accordance with the *Policy for MAF's Responses to Risk Organisms* [2008] and will be implemented using management procedures described in MAFBNZ's *Biosecurity Response Processes and Procedures*⁴

⁴ Available at <http://brkb.biosecurity.govt.nz/response-system/index.htm>

Specific activities that are likely to be undertaken include:

- A Trade team will be established to manage any food safety issues and trade impacts that may result from a confirmed case of atypical scrapie. Liaison with the meat and other industries such as germplasm exporters and pharmaceutical and biological product exporters will be provided.
- NZFSA will keep all New Zealand overseas posts informed (via MFAT) about the event and will ensure key trading partners are informed, both through the posts and through direct contact with key contacts in trading partners where relevant. Should one or more key trading partners initiate any action, it will be necessary to keep other key partners and contacts closely informed to ensure there is no 'cascade' effect.
- The MAFBNZ and NZFSA's Communications Teams will deal with any media or public interest in the event of a confirmed case (see Communications Plan for managing a confirmed case of atypical scrapie in New Zealand). It will also be important to provide timely and clear information for information 'multipliers' such as farming groups, retailers and the media. Information may also need to be gathered, via the trade response team, from authorities such as UK FSA⁵, EFSA and EU Commission.
- MAFBNZ will meet New Zealand's international obligations to the OIE, in particular for timely and transparent reporting in collaboration with MFAT.
- Review of the legal status (unwanted, notifiable etc) of atypical scrapie in New Zealand may be appropriate.
- If required, the MAF Recovery Team will endeavour to support and assist in recovery measures for any affected farmer.⁶

Specific activities that will not be undertaken at this stage include:

- MAFBNZ will not undertake any atypical scrapie eradication or control focused operational activities and will not impose Restricted Place notices or movement restrictions on any property under the Biosecurity Act (1993) where a case of atypical scrapie is confirmed.
- NZFSA will not take steps to require the removal of specified risk materials from the food chain as this is not scientifically justified and provides no known public health benefit.

⁵ Food Standards Agency

⁶ [Farmers are eligible for compensation under the Biosecurity Act 1993 where verifiable losses are incurred as a result of any measures taken using powers under the Biosecurity Act for the diagnosis or investigation of the agent (e.g. the purchase of stock for testing purposes). No such measures are anticipated to form part of any response to atypical scrapie, in line with this policy]

- The imposition of any additional protective measures for workers occupationally exposed to sheep tissues is not supported by available evidence.
- MAFBNZ will not redeploy existing surveillance resources towards enhanced surveillance for atypical scrapie

This policy has been developed and agreed to guide MAFBNZ's and NZFSA's response to a confirmed case of atypical scrapie in or closely associated with New Zealand. However, both agencies recognise that the individual circumstances surrounding any given response scenario may require flexibility of action, as dictated by changes in the available scientific information or other developments in the animal health, trade and public health arenas. As noted above, the potential economic impacts of any case of atypical scrapie in New Zealand will be subject to market reaction, which cannot be predicted with certainty in advance. Accordingly, MAFBNZ and NZFSA reserve the right to amend this policy when and how it is considered appropriate to do so.