

ATYPICAL SCRAPIE IN SMALL RUMINANTS: CONSIDERATION OF THE CURRENT PRECAUTIONARY RISK MANAGEMENT MEASURES

Executive Summary

1. This paper provides information on atypical scrapie (a transmissible spongiform encephalopathy (TSE)) in sheep and goats and the precautionary measures currently in place to protect consumers from the possible risks from TSEs in these species. There are a great many unknowns about atypical scrapie, including the potential implications, if any, for human health.
2. It also reports on the views of stakeholders and consumer focus groups who were asked whether, in the light of this uncertainty, additional precautionary measures were needed and for their views on the Agency's advice on this subject.
3. The Board is asked to:
 - **note** that the Agency's advice has been reworded to take account of the views of stakeholders and the consumer focus groups and will be tested further
 - **note** that the background information on sheep TSEs on the Agency's website will be reviewed
 - **note** that the agricultural departments are planning to review the Ram Genotyping Scheme
 - **note** that surveillance for atypical scrapie will be maintained in order to detect any changes in prevalence.
 - **agree** that the Agency's advice and recommendations on precautionary measures should be kept under review and be brought back to the Board if there are significant changes in the understanding of the risk.
 - **agree** that developments on atypical scrapie be kept under review to enable contingency policy to be refined as new information emerges.
 - **agree** that the Agency should open discussions with the European Commission on the issue of the identification of meat from older sheep or goats and natural sausage casings made from sheep intestines to enable consumer choice.

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ATYPICAL SCRAPIE IN SHEEP AND GOATS: CONSIDERATION OF THE CURRENT PRECAUTIONARY RISK MANAGEMENT MEASURES**Issue**

1. To consider whether the Agency should recommend, on the basis of current evidence, that additional precautionary measures are needed to reduce the possible risk to consumers from atypical scrapie.

Strategic Aims

2. This paper links to the Agency's aim to ensure that BSE controls in the food chain are based on the latest scientific knowledge.

Background

3. Cattle, sheep and goats are known to suffer from a group of transmissible neurological diseases known as TSEs. The best known of these is BSE in cattle. Sheep can suffer from a related neurological disease, scrapie. In a recent EFSA opinion,¹ (endorsed by the Spongiform Encephalopathy Advisory Committee (SEAC)) it was recognised that, using the currently available tests, scrapie could now be divided into two categories – scrapie (now referred to as “classical scrapie”) and “atypical scrapie”. Despite its name (which many now deem inappropriate) atypical scrapie does not appear to be a simple variant of classical scrapie, but is different from both classical scrapie and BSE. Classical scrapie has been recognised for over 200 years. What is now known as atypical scrapie was only identified in 2002/3 upon the introduction of new, more sophisticated testing methods, although it may have been present in the flock for a long time.
4. The Board has recently considered two papers on atypical scrapie in small ruminants (sheep and goats), at their March and April 2006 meetings. There have been no new scientific developments since these papers.
5. In March, the Board discussed a statement on atypical scrapie from the SEAC Sheep Subgroup which gave an opinion on (a) the best interpretation of the current data, (b) the potential risks for human and animal health and (c) the additional information necessary to improve assessment of the risk for human and animal health.
6. The statement notes that atypical scrapie (as defined in the EFSA opinion¹) has been found in most European countries and that modelling suggests that 82,000

¹ EFSA Opinion 26/10/2005

sheep in Great Britain may be infected² with atypical scrapie as compared to 56,000 with classical scrapie. The Subgroup agreed that it was important to try to establish whether atypical scrapie has been in existence for some time – by applying the new tests to any appropriate historical samples, and that surveillance should be maintained to establish whether the prevalence is changing.

7. The March Board paper also reported that it had recently been shown that atypical scrapie can be experimentally transmitted to mice and to sheep. The Subgroup agreed that, since atypical scrapie is experimentally transmissible, the possibility that it is transmissible to humans must be considered. Experiments in mice that have been altered to express the human prion protein should provide further information on this question, but these are still in the early stages and may take several years to complete. Such studies have begun in other European countries and it is possible that data may become available earlier. However the Subgroup also noted that the current specified risk material (SRM) controls may be more effective in reducing any human risk from atypical scrapie than they are for BSE in sheep.

Science and Research

8. In addition, the SEAC sub group identified a number of other areas on which further data was urgently needed. For the Agency the protection of consumers is paramount and therefore the critical areas for research are the human health risk and the tissue distribution of infectivity in sheep.

Human Health Risk

9. The sub group agreed that ‘There is no evidence of a risk to human health, but a theoretical risk cannot be excluded.’ The Agency has called for proposals for research using different experimental methods to assess the risk to humans from TSEs, including atypical scrapie, relative to that from BSE in cattle. A number of proposals have been received and are currently being appraised. Additional research on the human risk may need to be commissioned to ensure that the data for risk assessment is robust.

Tissue Distribution in Sheep

10. Infectivity for classical scrapie and BSE in sheep is more widely spread in the body of the sheep than is BSE in cattle. At this time there is no evidence that the distribution of infectivity in an animal with atypical scrapie extends beyond that determined for ‘classical’ scrapie or experimental BSE in sheep. Indeed, as

² TSE infection builds up in the animal over time and many infected animals will not show any clinical signs of disease. Many animals will die or be slaughtered before they reach an age where clinical signs appear, therefore the number of cases (with clinical disease) of a TSE will be much less than the number of infected animals.

reported by SEAC, atypical scrapie may not be present in the lymph nodes³ (although one study suggests that this may not be true for all genotypes). The Subgroup agreed that it was urgent to clarify whether atypical scrapie was present in the lymphoreticular system of sheep, and whether there were differences between the different sheep genotypes. These tissues are not covered by the current SRM requirements and therefore pose a potential risk to humans from consuming sheep meat. The Agency will be issuing a call for proposals to look at tissue distribution in the near future. However there is a shortage of infected material to use for such work and some preliminary work to generate this material may be necessary. Wherever possible the opportunity for joint funding will be investigated.

11. It is important to recognise that such research cannot provide rapid answers (see Annex 7 for details). It is also unlikely that a definitive answer will be obtained, especially to the question of human risk, and therefore the assessment of the risk will be based on expert opinion of the evidence (e.g. from SEAC). Any emerging evidence and the associated risk assessment from SEAC would be brought back to the Board to enable a review of the recommendations arising from this meeting.

The National Scrapie Plans (NSP/NISP)

12. There are known sequences in the sheep prion protein gene that affect the animal's susceptibility to scrapie. A critical result from scrapie surveillance is that atypical scrapie shows a higher prevalence in those animals considered to be more resistant to classical scrapie, including the most resistant 'ARR/ARR' genotype. The voluntary Ram Genotyping Scheme (RGS), which is part of the NSP⁴, uses selective breeding to remove the genotypes that are most susceptible to classical scrapie, thereby increasing the proportion of resistant genotypes in the flock. The SEAC Sheep Subgroup concluded that the current information on atypical scrapie is insufficient to alter the risk basis of the NSP, but that emerging findings should be kept under constant review. The Board is invited to note that Defra and the devolved GB agricultural departments are now undertaking a full review of the RGS. DARD will also be reviewing their breeding program run under the NISP.

Precautionary Measures Currently in Place

13. Under EC legislation (Regulation EC999/2001 and amendments) a number of precautionary measures that apply to small ruminants were put in place to protect the consumer from the risk of BSE as follows:

- The feed ban

³ For experimental BSE in sheep it has been estimated that the lymph nodes constitute approximately half the infectivity which would enter the food chain from a clinical sheep, under current SRM.

⁴ It should be noted that there are differing views on the benefits of the NSP – see Annex 8

- Specified risk material (SRM)
- Compulsory reporting of TSE suspects
- Differential TSE testing for BSE and scrapie
- Breeding/culling programs applied following a finding of TSE

(Further details are given in Annex 2)

Possible Additional Precautionary Measures

14. A number of strategies for additional precautionary measures to protect consumers from the possible risk of atypical scrapie were put to stakeholders for their views on proportionality. These approaches are essentially extensions of the existing precautionary measures:

- A complete ban on older animals entering the food supply (similar to the OTM rule for cattle).
- Increasing the list of SRM to cover all tissues known to carry scrapie infectivity, with the exception of some lymph nodes associated with edible tissue that would be difficult to remove.
- TSE testing all older animals, with only those with a negative result being allowed into the food supply.
- Continue providing consumers with advice on the possible risk and associated uncertainty from atypical scrapie to facilitate consumer choice.

Impact

Stakeholder workshops

15. Given the limited knowledge of the transmissibility and tissue distribution of atypical scrapie it is impossible to provide any form of quantitative risk assessment at the present time. Therefore the public health benefits to consumers of recommending any of these options are extremely uncertain.

16. The economic costs and the practicalities, particularly their effects on the relevant industries were considered. Full details are given in Annex 1, from which it can be seen that there is a high economic cost for options (I – III) and major practical problems that would impact negatively on the sheep industry, particularly for options (I) and (II).

17. Stakeholders representing consumers (including the Human BSE Foundation and CJD Support Network) and all aspects of the sheep industry were invited to facilitated workshops held in London, Cardiff, Edinburgh and Belfast. A SEAC representative presented their report and stakeholders were provided with details of the options, their costs and practical considerations (see Annex 1). Stakeholders discussed the advantages and disadvantages of each option and came to a consensus view at each workshop on whether additional precautionary measures were needed and, if so, which of the options they would favour. Whilst all stakeholders reached a consensus view, attendance was heavily weighted towards industry representatives. Details of the conclusions are given in Annex 3.
18. At all the workshops there was a concern about the lack of evidence and underlying science and full support for the need for further research to provide this evidence as soon as possible. Coupled with this, was the general view that, given the uncertainties, it was too soon to recommend any additional risk management measures, but that these would be necessary if the science demonstrated that atypical scrapie did represent a risk to human health.
19. The consensus of all groups was that applying an age cut-off was seen as inappropriate because it unnecessarily removed healthy animals from the food supply and would be very expensive with a high impact on industry. If precautionary measures were to be put in place some stakeholders favoured increased SRM removal whilst others preferred testing of older animals. However a number of stakeholders supported some increase in surveillance testing at this time to provide better data on prevalence.
20. One point emphasised in the discussions was that this is not a problem that is unique to the UK and that nothing should be done on a unilateral basis.
21. Given that the conclusion of all the workshops was that at this point in time the Agency focus should be on providing advice to consumers, there was some discussion as to the tone and content that should be used and how the Agency would engage consumers.
22. There was general support for continuing to provide the available information for consumers and letting them make a choice as to what to eat, although some viewed this as impractical given the lack of data on this issue. It was agreed that the advice (see Annex 5) was of no benefit to the consumer if there was no way for them to apply it. Currently there is no legal definition of mutton and therefore no labelling requirement, making it difficult to apply the advice that refers to meat from older animals. The Agency also advises that risk can be reduced by avoiding sausage casings made from sheep intestines. Sausage casings are considered an ingredient when in pre-packed foods and, as such, are covered by the food labelling regulations. As a result they should be listed separately. Agency guidance is that there should be an indication of the species from which they are derived, however it is not clear how extensively this is applied.

23. There were a number of key messages from stakeholders on the advice and information put out by the Agency:

- That it was important that the background information put out by the Agency reflected the current science and all the associated uncertainty in a balanced and measured way.
- That it should include the precautionary measures that are in place
- That it should be made very clear that atypical scrapie is not unique to the UK, and what is known about both EU and third country imports.
- That the Agency should consider if anything could be done to allow consumers to follow the Agency's advice on avoiding older meat and casings made from sheep intestines, should they wish to do so.
- That the Agency should be pro-active in taking their advice to consumers.

Consumer Focus Groups

24. Given the limited presence of consumers at these workshops and the emphasis placed on the Agency's advice the views of a cross-section of consumers were gathered at a series of focus groups (see Annex 10 for details). The primary objectives and conclusions of this research were:

- **Objective:** to explore how consumers feel about the potential risk to human health from BSE in sheep and atypical scrapie, once it has been explained to them.
Conclusions: the majority of respondents were not overly concerned about what they learned about the possible risks. Others either dismissed the topic out-of-hand or took the issue a little more seriously, but did not think there is a major cause for concern.
- **Objective:** to gauge how consumers think the risk should be managed, and who should take responsibility for doing this.
Conclusions: there was a consensus that the Agency is fulfilling its obligations at the moment by maintaining a position of 'honest uncertainty' – by publishing the latest information and warning consumers that there might be a risk, but leaving it up to consumers to choose what they eat.
- **Objective:** to gauge understanding of, and response to, the Agency's advice and information on atypical scrapie and BSE in sheep.
Conclusions: the current advice was seen as woolly, contradictory and unhelpful and it was recommended it should be reworded. Most attendees had no clear understanding of the subject even when provided with additional background information from the Agency's website. However, they also

indicated that they would only react to an issue if there were a major trigger (e.g. intense media coverage or withdrawal of products).

The Agency

25. The difficulty for the Agency is in getting the balance right in the face of such limited knowledge of the risk to human health. At one extreme the Agency could recommend additional precautionary measures that result in high financial costs to the UK government and severe damage to the UK sheep industry with the science eventually demonstrating that there never was a significant risk to human health. At the other end of the scale is the scenario where the Agency does not recommend any additional measures now and the science subsequently demonstrates a significant risk. Both scenarios would be unacceptable and could result in the loss of confidence in the Agency. This is particularly the case, given that the handling of the BSE crisis was a major driver for setting up the Agency and the Phillips report on the lessons to be learnt from the BSE crisis. To aid in the application of the conclusions of the Phillips report the Agency developed a checklist of points that should be considered when developing policy (see Annex 9). However, it should be noted that this situation does not provide a direct parallel because precautionary measures are already in place and the focus of this paper is whether additional measures to further reduce any possible risk should be recommended.

26. It is anticipated that any research or staffing costs for the FSA can be met from within existing budgets.

Conclusions

27. Atypical scrapie is definitely present in the UK flock, and in the flocks of other Member States (MS), and animals with atypical scrapie have, and will be, entering the food supply. However it is not known if this constitutes any risk to human health. Unlike the situation when BSE was first discovered in cattle, precautionary measures are already in place. Based on the limited knowledge of the distribution of infectivity in atypical scrapie, the SEAC Subgroup concluded that the SRM requirements that were put in place on a precautionary basis for BSE in sheep may provide at least a similar level of protection against the possible risk from atypical scrapie.

28. The consideration of the proportionality of any additional precautionary measures is very difficult when the human health risk is unknown, and, as reported by SEAC, there is insufficient data to carry out a risk assessment.

29. Any additional precautionary measures that could be put in place have a high economic cost, are currently highly impractical (see Annex 1 for details) and would impose a cost on industry that would, according to industry stakeholders, be likely to bring into question the economic viability of sheep farming.

30. The least costly option is adding other tissues to the list of SRM, although it is clear that these could generate losses for upstream and downstream industries. For example, the Meat and Livestock Commission estimate of the total turn over of the casing industry, related to sheep intestines, was £26 million in 2002. However, it is not known if increasing the list of SRM would add any real risk reduction, even if atypical scrapie is a human health risk, thus making it a disproportionate option at this time.
31. The highest priority for the Agency is consumer protection. However, it is impossible with the current limited data on atypical scrapie to carry out a full risk assessment, and thereby to assess the proportionality of any particular risk reduction strategy. Therefore, the recommendation, which is supported by the conclusions of the stakeholder workshops, is that no further precautionary risk reduction measures should be recommended at this time, but that measures should be considered on a contingency basis.
32. We do know that atypical scrapie is present in the UK flock and infected animals will be entering the food supply. Therefore, if evidence emerged that atypical scrapie could be a human health risk, the requirement for additional measures to protect consumers would need urgently to be reconsidered. Given the lack of information about the disease the best that could be recommended on present knowledge would be a combination of the exclusion of older sheep from the food supply and removal of more SRM from the remaining sheep. Such action could, as it would for BSE, provide a high level of risk reduction, although the lack of data means that, at present, no estimate of its effectiveness can be made. It is therefore essential that developments on atypical scrapie are kept under review to enable contingency policy to be refined as new information emerges.
33. At the March 2006 Board meeting it was agreed that there should be no immediate change in the Agency's advice on sheep and goat meat (see Annex 5) as a result of the emerging evidence in relation to atypical scrapie. That position remains unchanged. However the Executive has considered the wording and presentation of that advice, particularly as regards addressing the issues of both BSE and atypical scrapie within overarching advice. As discussed in paragraphs 22-24 and Annexes 3 and 10 there were a number of clear messages from stakeholders and consumer research about the wording of the advice that help inform how best to communicate uncertain but possible risk. Taking these views into account the executive will refresh its communications, notably the website content, and proposes to use the following for its current advice:

“On the basis of current evidence, the Agency is not advising people to stop eating sheep or goat meat or products.

Sheep and goats can get brain diseases similar to BSE in cattle, but BSE has never been found in UK sheep. Emerging evidence has identified another brain

disease in sheep known as ‘atypical scrapie’. It is not possible at the present time to determine what risk, if any, atypical scrapie may present to people.

Because the possibility of a risk cannot be entirely ruled out, a number of precautionary controls are in place. These include controls on animal feed and removing certain parts of the animal before the meat goes into the food chain.

Whilst the Agency is not advising anyone to stop eating sheep or goat meat or products, any possible risk could be reduced further by not eating meat from older animals. This is because if there were a risk it would be greater in older animals. Meat from older sheep is known as mutton. In addition, some sausages are contained in natural sheep casings made from sheep intestines which are more likely to carry the disease agent and therefore could present a greater risk.”

Board Action Required

34. The Board is asked to:

- **note** that the Agency’s advice has been reworded to take account of the views of stakeholders and the consumer focus groups and will be tested further
- **note** that the background information on sheep TSEs on the Agency’s website will be reviewed
- **note** that the agricultural departments are planning to review the Ram Genotyping Scheme
- **note** that surveillance for atypical scrapie will be maintained in order to detect any changes in prevalence.
- **agree** that the Agency’s advice and recommendations on precautionary measures should be kept under review and be brought back to the Board if there are significant changes in the understanding of the risk.
- **agree** that developments on atypical scrapie be kept under review to enable contingency policy to be refined as new information emerges.
- **agree** that the Agency should open discussions with the European Commission on the issue of the identification of meat from older sheep or goats and natural sausage casings made from sheep intestines to enable consumer choice.

POSSIBLE OPTIONS FOR ADDITIONAL PRECAUTIONARY MEASURES TO MANAGE ANY RISK FROM ATYPICAL SCRAPIE

(I) Keeping animals over a certain age out of the food chain

Effect	Practicalities	Estimated average annual economic cost
Eliminates any risk from animals above the designated age, as identified by the number of permanent incisors that have erupted.	<ul style="list-style-type: none"> • farmer would need to select route for slaughter based on age • ageing of animals would rely on dentition checks in the abattoir • route for disposal of over-age animals would be needed 	Either 12 or 36 month limit ⁵ £67 million

⁵ see paragraph 2 of accompanying notes for details

(II) TSE testing all animals over a certain age

Effect	Practicalities	Estimated average annual economic cost
<p>Would reduce the risk by the disposal of those animals that test positive i.e. those in the latter stages of the incubation period</p>	<ul style="list-style-type: none"> • number of sheep to be tested would increase to up to 1.9 million (over 18 months) • adequate laboratory testing capacity needed • ageing of animals would rely on dentition checks in the abattoir, in the absence of sheep identification and registration. • abattoir systems for testing would be needed requiring <ul style="list-style-type: none"> - a training program across UK - sampling facilities and program - program for carcass and offal retention and traceability - system for handling test results - supervision 	<p>Testing all sheep over 18 months at slaughter</p> <p>£48⁶ million</p>

⁶ Cost updated from £45 million to cover the increase in the cost of a TSE test.

(III) Increasing specified risk material (SRM)

Effect	Practicalities	Estimated average annual economic cost
<p>Would reduce risk if any infectivity were present in the tissue removed. Additional tissues that could be classed as SRM⁷ from animals above a selected age are:</p> <ul style="list-style-type: none"> - liver - intestine - accessible lymph nodes - stomach - thymus 	<ul style="list-style-type: none"> • not a practical problem to add those tissues that can be removed at the abattoir, but increase in labour required • removal of lymph nodes situated within muscles likely to be impractical (line speed would be decreased and checking removal extremely difficult) • increased abattoir supervision 	<p>Adding all proposed tissues to the SRM list for sheep</p> <p>over 12 months £7⁸ million</p> <p>over 36 months £4 million</p>

⁷ The spleen and ileum from sheep and goats of all ages and the skull (including brain and eyes), spinal cord and tonsils of sheep and goats aged over 12 months are currently removed as SRM

⁸ Increases over that presented to stakeholders because of changes in the allowed number of sheep in this age group

(IV) Providing consumers with advice on the possible risk and associated uncertainty from atypical scrapie to allow better informed consumer choice.

Effect	Practicalities	Estimated average annual economic cost
<p>Would raise awareness of the possible risk from atypical scrapie and enable consumers to make a more informed choice. Could possibly result in decreased market for named items (meat from older animals, sheep sausage casings).</p>	<ul style="list-style-type: none"> • information could be kept under review and updated in the light of any emerging information • would need to ensure that advice reaches consumers – particularly those groups with higher than average consumption of sheep and goat meat (particularly from older animals) • possible need for a labelling requirement if consumers are advised that any risk from atypical scrapie would be reduced by avoiding consumption of meat from older animals 	<p>none</p>

NOTE ON THE COSTING OF THE POLICY OPTIONS CONSIDERED FOR MANAGING THE RISK RELATED TO ATYPICAL SCRAPIE – ECONOMICS 2, 25 MAY 2006

1. This summary presents the assumptions underlying the derivation of the economic costs of the policy options considered by the Board and explains how those costs would arise. Economic costs measure the amount of real resources, or production factors, that would be consumed by a policy and therefore could not be used productively elsewhere in the economy, ultimately leading to a loss of economic output and welfare⁹. All figures are derived for the UK assuming that 1.83 million ewes and two million hoggets are slaughtered annually.

Removal of sheep older than 12/36 months from the food chain

2. The lost value of the culled sheep represents the main cost of this option and is calculated assuming that cull ewes are worth £36 per head based on prices reported by the Meat and Livestock Commission (MLC)¹⁰. There are also incremental¹¹ disposal costs (£4.39 per animal), which are derived from DEFRA's analysis of the effect of the policy options considered by the FSA in its BSE in sheep contingency plan. Importantly, for the 12 months option, it is assumed that farmers previously producing hoggets would adjust to the policy change by bringing the slaughtering age of all sheep raised for meat under one year, hence ensuring that no hogget would need to be disposed of. This explains why the calculated cost of the policy is the same whether the age limit is set at 12 months or 36 months.

Testing

3. The unit cost of the test used in the calculation is based on the amount that the Veterinary Laboratories Agency currently charges DEFRA for TSE testing. There are also incremental disposal costs and a loss of value for the animals testing positive (assuming an incidence rate of TSEs of 1/500 tested), but the cost of the test itself represents more than 99% of the total cost of the policy. Note that this policy option would also have potentially large set up costs (e.g., training of abattoir staff, modification of abattoir premises) that are not included in the estimate presented in the table.

⁹ This is different from an analysis of financial costs that would focus on expenditures incurred as a result of the policy.

¹⁰ A pocketful of meat facts (2005), MLC, page 31.

¹¹ This means that we only consider the costs that are created by the policy over those normally incurred under the 'do-nothing' option. For instance, for the option preventing sheep older than 12/36 months from entering the food chain, slaughtering costs are not included in the calculations as, even in absence of that policy, the animals would have to be slaughtered. By contrast, the costs of disposing of the animals no longer allowed to enter the food chain result directly from the policy and are therefore included in the calculations.

Extra SRM Removal

4. The estimates rely primarily on a MLC report prepared for the FSA in February 2002 and entitled 'BSE and sheep risk management options'. The extra SRM for sheep aged over one year include all thoracic viscera excluding the heart; all abdominal viscera; stomach contents; pre-crural fat; and all accessible lymph nodes. The removal of extra SRM creates direct costs associated with increased labour and supervision requirements in abattoirs and cutting plants, industry costs generated by the loss of revenues for tissues previously sold but now considered SRM, and disposal costs. Altogether, the disposal costs dominate, while the only substantial direct costs result from the removal of lymph nodes. The main industry costs relate to the classification of intestines as SRM, which would prevent their sale to the casing industry.

Other Impacts

5. Subsequent to the stakeholder workshops some additional calculations have been carried to assess the impact of these policy options on other industries.
6. The analysis does not cover the full impact of the policy options on upstream¹² and downstream¹³ industries, which means that the costs are likely to be slightly underestimated. More importantly, the three policy options would have very different impacts on industries related to the sheep sector:
 - The testing regime would have almost no impact.
 - Extra-SRM removal would have limited impact. This would arise mainly from the effect of the policy on the casing industry¹⁴, which would no longer be able to procure intestines from UK sheep older than one year, but could still process the raw material provided by the 13 million lambs slaughtered annually in the UK. The cost estimates presented above take into account the loss of casing sales by abattoirs but do not capture the potential losses of value-added and employment in the casing industry as a result of the policy. This loss, however, would be mitigated by the fact that the industry could source supply from abroad, or use alternative types of casings (e. g., from pigs or synthetic).
 - Preventing sheep older than 12/36 months from entering the food chain would have the largest impact on industries linked to the sheep sector. It is likely that

¹² Those include agricultural contractors, veterinary service providers, animal feed and agricultural machinery manufacturers, and fertiliser manufacturers/compounders.

¹³ Those include auctions, abattoirs, processors, wholesalers, and renderers.

¹⁴ The MLC estimated the total turnover of the casing industry from processing sheep intestine at £26 million in 2002.

the policy would have little effect on the size of the flock¹⁵, and consequently no adverse impact on upstream industries. However, the effect on downstream industries would be significant:

- The activity levels of auctions, processors and wholesalers would contract due to fewer sheep being commercially traded to enter the food chain.
- Abattoirs would not be adversely affected as the slaughtering of sheep older than 12/36months would continue.
- The effect on the rendering industry would be complex. On the one hand, turnover would increase from the processing of cull ewes. On the other hand, tallow derived from those ewes would fetch a relatively low price as it could only be used as fuel oil.

7. A spreadsheet model was developed in order to estimate the turnover and employment losses in downstream industries if sheep older than 12/36 months were prevented from entering the food chain. The results, summarised below, show that the policy option would potentially result in the loss of almost 700 jobs in sheep related industries. The decrease in employment is driven by a reduction in turnover in sheep related industries of nearly £150m. Note, however, that those changes should not be interpreted as additional economic losses as the resources released from the decrease in activity in downstream industries could find alternative productive uses in the economy.

Impact on downstream industries of preventing sheep older than 12/36 months from entering the food chain.

	Magnitude of change			
	Annual Turnover		Employment	
	Value	Share of sheep-related turnover	Number of workers	Share of sheep-related employment
Auctions	-£ 11,376,273	-11%	-11	-3%
Abattoirs	£ -	0%	0	0%
Processors	-£ 63,024,131	-10%	-567	-8%
Wholesalers	-£ 77,280,992	-10%	-137	-5%
Renderers	£ 3,952,519	19%	27	13%
Total	-£ 147,728,878	-7%	-688	-5%

¹⁵ This seems reasonable if some compensation was paid to farmers for the loss of revenue from the sale of cull ewes.

CURRENT PRECAUTIONARY RISK MANAGEMENT MEASURES

- Feed ban – prevents the spread of scrapie or BSE via feed by banning the inclusion of any animal protein
- Specified risk material (SRM) – requires that certain tissues, that have been shown to carry scrapie or BSE infectivity, are removed from the food supply and destroyed

SRM for small ruminants – all MS

All ages: spleen; ileum

Over 12 months (or with a permanent incisor erupted): Skull including the brains and eyes; Tonsils; Spinal cord.

- Compulsory reporting – any cases of suspected TSE must be reported, these are then subjected to confirmatory testing and, if positive, to differential testing.
- Differential TSE testing – for all sheep and goats over 18 months a small random sample of animals slaughtered at abattoirs and of fallen stock (dead on farm) are tested for TSE. The minimum sample sizes are set within the TSE legislation and have changed a number of times to reflect the changing circumstances e.g. goat sample sizes were increased following the finding of a goat with suspected BSE in France in October 2004. All samples that test positive for TSE are further tested to determine whether they should be classified as BSE, classical scrapie or atypical scrapie. (see Annex 4 for the UK results) This testing is essentially used to provide data on prevalence.
- Breeding/culling programs applied following a finding of TSE – an epidemiological investigation is undertaken and for scrapie, under current legislation, the animals from the identified flock(s) are either all culled or they are genotyped and those of non-resistant genotypes are culled. The EC TSE Road Map includes a review of the culling policies for small ruminants. In the UK compulsory culling or genotyping controls are not being applied to atypical scrapie flocks for the time being, pending the outcome of this review, but these flocks are involved in a monitoring study on a voluntary basis. The Agency would expect to ask SEAC for risk assessment advice in relation to any proposals arising from this review.

ATYPICAL SCRAPIE STAKEHOLDER WORKSHOPS MARCH 2006

Attendance

	London	Edinburgh	Cardiff	Belfast
Food advisory committee members	n/a	1	2	1
Consumers	5	1	0	0
Farming industry	2	3	6	6
Abattoirs etc	1	1	1	2
Meat promotion	1	1	1	1
Casings industry	2	1		
Butchers, wholesalers		2		
veterinarians	2	2	0	1
Other government depts	2	2	1	2
Others	2	2		
Total	18	16	11	13

Summary of views

1. There was a general concern about the current lack of scientific evidence and full support for the need for further research to provide this evidence as soon as possible. Coupled with this was the view that, given the uncertainties, it was too soon to recommend any additional risk management measures, but that these would be necessary if the science demonstrated that atypical scrapie did represent a risk to human health.
2. The consensus of all groups was that applying an age cut-off was seen as inappropriate. If precautionary measures were to be put in place some stakeholders favoured increased SRM whilst others preferred testing of older animals. However a number of stakeholders supported some increase in surveillance testing at this time to provide better data on prevalence.
3. The difficulty in determining the age at which any risk management measure should be applied was also recognised. On the one hand, the older the age selected the less the impact on industry and the economic cost, at the same time the older the age selected, the less the potential risk reduction achieved. Without the scientific data to inform this discussion it was recognised that it was essentially impossible to assess the proportionality of any particular age. A

further point of concern was that ageing would largely be based on dentition, which is recognised as having a wide variability.

4. One point that was emphasised in the discussions by industry representatives was that this is not a problem that is unique to the UK and that nothing should be done on a unilateral basis. There was also concern that any actions should not, even indirectly, prejudice trade – for example by careless wording of advice that implied a disproportionate problem with UK sheep as compared to imports.

(I) Keeping animals over a certain age out of the food chain

5. Stakeholders in general considered this the least appropriate option. It was recognised that this would remove the animals that presented the highest risk, but it was seen as indiscriminate in that it would also remove healthy animals that presented no risk. The overwhelming view was that this was disproportionate given the current evidence – that it is not known whether there is a human health risk, the high financial cost and the substantial impact on the industry.

(II) TSE testing all animals over a certain age

6. The major advantages of this were seen as, firstly, that it was more targeted, in that only animals shown to be infected are removed, and all others are able to enter the food supply as now. Secondly, that it was more scientifically based, although there was uncertainty about the most appropriate age limit.
7. It was considered that in removing the theoretically highest risk animals, this approach was providing protection for consumers, which would be recognised as such and therefore increase consumer confidence in the product. It was also noted that increased testing would have the advantage of providing more data on prevalence.
8. However, there were issues of proportionality in respect of the high economic cost and the impact on production. All abattoirs slaughtering older animals would be affected, particularly high throughput abattoirs that would not be able to maintain their current line speeds. Given the large numbers of sheep slaughtered compared to cattle, some felt that the infrastructure that has been set up for cattle could not be applied to sheep.
9. There was some support for this as an appropriate measure, should there be evidence that atypical scrapie did present a risk to human health, but only if the cost to the industry could be reduced to an economic level, the target being a cheap test that could be used in the abattoir. There was also support for some increase in testing at this time to improve knowledge of prevalence, whilst recognising that there would be little benefit in terms of consumer protection.

(III) Increasing specified risk material (SRM)

10. It was recognised that for experimental BSE in sheep the current precautionary SRM would not remove all the infectivity. However, stakeholders were concerned that, at this point in time, it was not known if this was true for atypical scrapie. Research to address this question was seen as a priority. There was also concern by some stakeholders that the removal of all lymph nodes would reduce the value of the carcass below an acceptable level and that the alternative of leaving some lymph nodes in the edible tissue could still present a potential risk to the consumer.
11. This approach was noted as having the advantage of a lower economic cost than other measures, but that there was a potential loss of high value products such as sausage casings.
12. Additional SRM was seen as having potential as a risk reduction measure should there be evidence that atypical scrapie did present a risk to human health and there was scientific data on what tissues/age should be targeted. Although some industry stakeholders expressed a concern that consumers lacked knowledge of SRM measures and would not be reassured by increasing the list of tissues.

(IV) Providing consumers with advice on the possible risk and associated uncertainty from atypical scrapie to allow better informed consumer choice.

13. Given the lack of knowledge and the high cost of other measures this was seen as the only action that was proportionate at this time. It was not viewed as 'doing nothing', but as acting in a proportionate manner. However, there were definite industry concerns about the effect that advice could have on consumer confidence, particularly since the UK might be more open than other countries, with some stakeholders suggesting that any advice should be at the EU level. Some recalled the early days of BSE and the parallels that might be drawn, between the advice on atypical scrapie and that given for BSE.
14. Concerns were expressed about the wording that should be used, the context/underlying message of the advice and how the Agency would engage consumers. It was seen as a positive point that advice could be amended in line with the changing science, particularly if the possible human risk decreases. In the meantime, it was important that research addressed the uncertainties, and that it was made clear what the time scale was likely to be to obtain informative results.
15. There was general support for allowing the consumer to make an informed choice, although some viewed this as impractical given the lack of data on this issue. However, where Agency advice refers to meat from older animals it was agreed that this was of no benefit to the consumer if there was no way they could apply the advice. Therefore the issue of labelling should be investigated.

16. Clarity was seen to be the key to provide a message that is meaningful to consumers, it should include the precautionary measures that are already in place, it must be science based and the uncertainty must be explained. The advice must cover both EU and third country imports. Concerns were expressed about consumer apathy towards the advice (food scare fatigue) and the need to actively target the appropriate audience.

UK RESULTS FOR SURVEILLANCE IN SHEEP AND GOATS

Active surveillance

Year	Survey type	Total tested		Classical scrapie positive		Atypical scrapie positive	
		GB	NI	GB	NI	GB	NI
2002	Sheep abattoir	30115 ¹⁶	1483	34	0	18	0
	Sheep fallen stock	913 ¹³	519	7	0	0	0
	Goat abattoir	9	n/a	0	-	0	-
	Goat fallen stock	3	3	0	0	0	0
2003	Sheep abattoir	71250	1268	46	0	45	0
	Sheep fallen stock	4059	1067	12	1	7	0
	Goat abattoir	191	n/a	1	-	0	-
	Goat fallen stock	53	1	0	0	0	0
2004	Sheep abattoir	10589	715	9	0	12	0
	Sheep fallen stock	4371	725	12	0	4	0
	Goat abattoir	90	n/a	0	-	0	-
	Goat fallen stock	51	1	0	0	0	0
2005	Sheep abattoir	11106	710	12	2	16	0
	Sheep fallen stock	8279	1489	26	1	6	0
	Scrapie monitored flocks		56		0		0
	Monitoring infected flocks		115		5		0
	Goat abattoir	1282	n/a	0	-	0	-
	Goat fallen stock	1329	5	4	0	0	0
2006	Sheep abattoir	4680	396	1	1	1	0
	Sheep fallen stock	12294	1786	20	2	5	0
	Scrapie monitored flocks		86		0		0
	Monitoring infected flocks		65		6		0
	Goat abattoir	815	n/a	0	-	0	-
	Goat fallen stock	1054	2	2	0	0	0
All	Totals	162533	10489	186	18	114	0

Results to May 2006

¹⁶ Of the total 31,028 sheep tested in GB in 2002 only 15,798 were subject to a test capable of detecting atypical scrapie.

PASSIVE SURVEILLANCE: REPORTED SUSPECTS FOUND POSITIVE

Year	Classical scrapie positive		Atypical scrapie positive	
	GB	NI	GB	NI
2002	404	0	-	0
2003	378	1	-	0
2004	309	2	0 ¹⁷	0
2005	229	0	3	0
2006	58	3	1	0
Totals	1378	6	4	0

¹⁷ A test capable of detecting atypical scrapie was first used for passive surveillance in July 2004

CURRENT FSA ADVICE ON THE CONSUMPTION OF SHEEP AND GOAT MEAT AND DAIRY PRODUCTS

The Agency is not advising people to stop eating sheep or goat meat or dairy products.

If BSE were present in sheep or goats or atypical scrapie can be transmitted to humans, consumers could significantly reduce the risk by avoiding meat from older animals (e.g. mutton) and sausages with casings made from sheep intestines.

Revised FSA advice on the consumption of sheep and goat meat and dairy products

“ On the basis of current evidence, the Agency is not advising people to stop eating sheep or goat meat or products.

Sheep and goats can get brain diseases similar to BSE in cattle, but BSE has never been found in UK sheep. Emerging evidence has identified another brain disease in sheep known as ‘atypical scrapie’. It is not possible at the present time to determine what risk, if any, atypical scrapie may present to people.

Because the possibility of a risk cannot be entirely ruled out, a number of precautionary controls are in place. These include controls on animal feed and removing certain parts of the animal before the meat goes into the food chain.

Whilst the Agency is not advising anyone to stop eating sheep or goat meat or products, any possible risk could be reduced further by not eating meat from older animals. This is because if there were a risk it would be greater in older animals. Meat from older sheep is known as mutton. In addition, some sausages are contained in natural sheep casings made from sheep intestines which are more likely to carry the disease agent and therefore could present a greater risk.”

IMPORTS

Data has been obtained from Defra on the import into the UK of sheep and goat meat and edible offal. From the totals given in the table below it is clear that the major exporter to the UK is New Zealand, with only Australia and Ireland also making a significant contribution to these imports.

Countries contributing at least 1% of sheep and goat imports to the UK

Country	Tonnes, 2005	% total imports	Total number of atypical scrapie (reported as at 30/06/05)
New Zealand	91,940	72.3	0
Australia	14,565	11.5	0
Ireland	10,509	8.3	4
Spain	2,460	1.9	17
Argentina	1,889	1.5	0
Germany	1,223	1.0	64
Uruguay	1,302	1.0	0

More detail on TSE testing has been gathered for these three countries.

	New Zealand (data for 2004)	Australia	Ireland (data for 2005)	UK (2005 data for comparison)
National sheep flock	39 million	94 million	3.7 million adults	16.8 million adults
National goat herd	186,000	230,000 (in 1994)	5,276 adults	88,000 adults
TSE tests on fallen stock/casualties	3,075 sheep 12 goats	200 sheep	10,374 sheep 79 goats (over 18 months)	34,112 sheep 2,502 goats (over 18 months)
TSE tests on suspects with neurological or other signs associated with TSEs	105 (sheep over 2 years of age 5 goats (over 18 months) with no other definite diagnosis)	466 sheep (over 18 months)	6 sheep (any age) 0 goats	398 sheep (any age) 0 goats
TSE tests on healthy slaughter animals ¹⁸	No indication that this is in place	None	10,689 sheep 0 goats (over 18 months)	132,634 sheep 2,387 goats (over 18 months)
TSE tests on imported animals	All are tested, irrespective of age.	All are tested?	n/a	n/a
Rapid test used	Prionics Western blot	Prionics Western blot and Bio-Rad	Predominantly Enfer TSE kit version 2	Bio-Rad TeSeE
Test method for suspects	Histopathology (IHC ¹⁹ or SAF ²⁰ if not negative)	Histopathology (Enfer and Bio-Rad rapid tests may be	Histopathology and IHC	Histopathology, IHC, Western blot and Bio-Rad TeSeE

¹⁸ NB A total of over 1000 sheep brains have been supplied to the EU as negative test controls and live sheep have been supplied as negative experimental controls for the UK. None of these have given positive TSE results.

		used if needed for confirmation)		
Is testing to differentiate atypical scrapie established	No indication that this is in place	No	Yes	Yes
Number of scrapie positives	Nil	Nil	Sheep 2002-2006 Classical: Abattoir: 28 Fallen stock:121 Suspects: 153 Atypical: Abattoir: 0 Fallen stock:1 Suspects: 3	Sheep 2002-2006 Classical: Abattoir: 105 Fallen stock:81 Suspects: 1384 Atypical: Abattoir: 92 Fallen stock:22 Suspects: 4

New Zealand and Australia have testing programs in place to look for scrapie in sheep (and also for BSE in cattle). These have been set up to comply with the OIE requirements for international trade. There is no evidence that scrapie is present in either of these countries. However, the tests being used on both neurological suspects and fallen stock are not considered to be capable of detecting atypical scrapie with the exception of those to which the Bio-Rad test is applied in Australia (no numbers are available). Ireland as a member of the EU has to meet the small ruminant testing requirements in the EU TSE legislation and reports the results to the EC.

¹⁹ IHC immunohistochemistry

²⁰ SAF scrapie associated fibrils – the SAF immunoblot should detect atypical scrapie, this has not been determined for SAF microscopy

RESEARCH UNDERWAY OR PLANNED***DEFRA OR FSA FUNDED RESEARCH ON ATYPICAL SCRAPIE
(Update of paper presented to SEAC on April 28 2006)***

1. An Expert Panel was convened by SEAC on 17th September 2003 to discuss the unusual results arising from the introduction of rapid testing methods into the scrapie surveillance programme in Great Britain. This panel recommended research that should be undertaken to investigate the nature and biology of atypical scrapie.
2. In response to these recommendations, Defra has funded several research studies on atypical scrapie. The studies, which are all in progress, include:
 - Intracerebral and oral challenges in sheep of various genotypes (SE1847²¹)
 - Intracerebral challenges of conventional and transgenic mice carrying sheep VRQ and ARQ PrP genes (SE1850)
 - Biochemical characterisation of the prion protein (SE1789)
 - Full sequencing of the PrP gene in sheep with atypical scrapie (SE0240)
 - Estimation of the prevalence of infection (part of SE0243)
 - Investigations on the flocks of origin (funded by Defra's Sheep TSEs Policy Group)
3. The Expert Panel also recommended that the rapid diagnostic tests should be applied to samples from sheep of genotypes associated with atypical scrapie from a source guaranteed to be scrapie free. This has not been carried out because the scrapie-free flock in the UK is too small to design a meaningful study²².
4. Defra is funding studies to investigate the relative transmissibility of atypical scrapie across species by challenges in transgenic mice carrying human and bovine PrP genes. In addition, the Food Standards Agency has requested applications to develop or apply relevant experimental models to assess the risk to humans from other TSEs, including atypical scrapie, relative to BSE in cattle. Proposals have been received and are currently being appraised.

²¹ SE1847 is the Defra code for the research project. This code can be used to find further details of the project on Defra's public web-site (http://www2.defra.gov.uk/research/project_data/Default.asp).

²² It should be noted that the negative controls in the 2004 evaluation of rapid tests for TSEs in small ruminants were 1000 sheep samples sourced from New Zealand, which is free from classical scrapie.

5. Research on atypical scrapie throughout Europe has been limited by the availability of material for study. Nevertheless, there is an increased understanding of atypical scrapie, which is now recognised as a distinct TSE of small ruminants that occurs in sheep that are relatively resistant to classical scrapie. Atypical scrapie has been experimentally transmitted to sheep and ovinised transgenic mice by the intracerebral route.
6. Information from the Defra-funded research programme and from other European laboratories was presented at the SEAC Sheep Subgroup Workshop on 24th January 2006 and summarised in a position statement²³.
7. The Sheep Subgroup recommended that further information about atypical scrapie was needed, including:
 - the historical prevalence
 - the distribution of infectivity within sheep of different genotypes
 - comparative transmission studies with other species
 - the incidence and nature of clinical disease
 - the route(s) of natural transmission
8. To investigate the past prevalence of atypical scrapie in sheep in GB, Defra is determining the availability of suitable archived material from sheep collected since the 1960's to which current tests for the differentiation of atypical scrapie can be applied.
9. Comparative transmission studies in humanised transgenic mice are planned and studies in different sheep genotypes have been put in place previously (2 and 4 above). The latter will also provide some information on the tissue distribution of infectivity and possibly the potential for a carrier state. These studies may be expanded as enough suitable atypical scrapie material becomes available for further challenges.
10. Knowledge of the tissue distribution is obviously fundamental to ensuring that effective risk reduction measures, such as the SRM controls are in place. The FSA plans to call for proposals for further work in this area. However the shortage of infected material to use as inocula for these experiments may necessitate a preliminary experiment primarily aimed at generating infected tissue for these studies and for use in test evaluation. Ideally this would be subject to joint funding.

²³ <http://www.seac.gov.uk/pdf/positionstatement-sheep-subgroup.pdf>

11. A small number of natural cases of atypical scrapie known to have expressed clinical disease have been found. Monitoring for further clinical cases and the experimental challenges in sheep will provide a profile of the disease phenotype over time.
12. Natural transmission and the behaviour of atypical scrapie in sheep populations will be further investigated in an epidemiological case control study on farms which have had cases detected by abattoir surveillance.
13. The FSA and Defra are also funding projects that, whilst not specifically targeted at atypical scrapie, are working on improved diagnostics, with potential for tests that can be applied to live animals or improved strain differentiation.
14. Annex 7A provides a time-line for research studies on atypical scrapie that are in progress or planned.

TIMING OF DEFRA-FUNDED RESEARCH TO INVESTIGATE ATYPICAL SCRAPIE

ANNEX 7A

	Experiment start date ²⁴	Status in April 2006	Results expected/delivered	Project end date
TRANSMISSION TO SHEEP (SE1847)				
Intracerebral challenges	Sheep challenged Nov 2004–Feb 2005 and Jan 2006	Challenges completed	2006 onwards	2011
Oral challenges	Sheep to be challenged in Jul 2006	Planned	2008 onwards	2011
TRANSMISSION TO MICE (SE1850)				
To C57BL/6 and VM mice	Mice challenged July - Dec 2005	In progress	Late 2006 onwards	Oct 2007
To transgenic lines carrying <ul style="list-style-type: none"> • sheep VRQ (tg338) • sheep ARQ (TgShpXI) 	Mice challenged July - Dec 2005	In progress	Early 2006 onwards	Oct 2007
To transgenic lines carrying human or bovine PrP (SE1439)	July 2006	planned	Late 2008 onwards	
BIOCHEMICAL STUDIES (SE1789)				
PrP PK digestion kinetics, Western Blots	Jan 2006	In progress	May - Sep 2006	Jul 2007
PrP fragment characterisation	Sep 2006	Planned	Feb 2007	Jul 2007
MOLECULAR GENETICS (SE0240)				
PRNP DNA sequence analysis – ORF, 5'promoter region, 3'UTR	Jan 2005	Sequencing completed; analysis	Apr 2005 onwards	Mar 2006

²⁴ It should be noted that the timing and extent of many projects was often limited by the availability of material.

		in progress		
EPIDEMIOLOGICAL STUDIES				
Estimation of prevalence of infection from all data sources (part of SE0243)	Nov 2005	In progress	Sep 2006 (initial analysis) Sep 2007 (extended analysis)	Sep 2007
Investigations on the flocks of origin/case control study (VLA and Defra's Sheep TSEs Policy Group)	Oct 2005	In progress	Feb 2007 (initial analysis) Feb 2009 (extended analysis)	Mar 2009

TIMING OF FSA-FUNDED RESEARCH TO INVESTIGATE ATYPICAL SCRAPIE

	Experiment start date	Status in April 2006	Results expected/delivered	Project end date
RELATIVE HUMAN RISK				
A number of proposals are being considered for funding	(Subject to approval) Sept 2006	Undergoing appraisal	2008 onwards	
PATHOGENESIS				
Pathogenesis studies and/or generation of tissues for such studies	(Subject to approval) Autumn 2006	Call for proposals to be issued by July 2006		
DIAGNOSTICS				
M03038: Development of a rapid, high throughput, pre-clinical test (may include atypical scrapie, subject to availability)	Oct 2005	In progress	2006 onwards	Sept 2008
Further diagnostic work, with particular focus on identifying the infective agent	(Subject to approval) Autumn 2006	Call for proposals to be issued by July 2006		

RESEARCH IN OTHER MEMBER STATES

In Germany 96 atypical scrapie cases have been reported (to end 2005). Research results on the biochemistry and diagnosis of atypical scrapie and the associated brain pathology have been published. This work is continuing along with bioassays in mice expressing the ovine prion protein.

A Norwegian research group defined the first case of 'Nor98' atypical scrapie in a sheep that died in 1998. Research has been ongoing since this time and currently includes epidemiology, the influence of genotype on susceptibility and tissue distribution of the abnormal protein, and sheep to sheep transmission.

In France 69 atypical scrapie cases have been reported (to end June 2005). Several research groups are working on atypical scrapie. The successful transmission to mice expressing the ovine prion protein has been published, as has work on the different cellular mechanisms associated with different sheep genotypes. Current work is focussing on transmission to mice, including those expressing the human prion protein and on the different genotypes and their associated biochemical characteristics in relation to susceptibility and possible sub-types of atypical scrapie.

Neuroprion is A network of the majority of European TSE researchers (including those referred to above). Within the stated areas of research covered by this network are the differentiation of scrapie strains, the detection of atypical scrapie using both existing and new approaches, the bioassay of identified strains in rodents and the comparison for different models for strain differentiation.

The EC also funds TSE research and as a result of recent calls in the Food Quality and Safety thematic priority of Framework Programme Six, a number of projects on the investigation and characterisation of atypical scrapie strains and the use of TSE diagnostic tests in goats are currently either recently started or undergoing contract negotiation.

THE HOUSE OF LORDS DEBATE ON THE TSE REGULATIONS 2006 (7 MAR 2006)

Extracts from the House of Lords debate in which the Countess of Mar made a number of points in relation to atypical scrapie (Hansard 7 March 2006). Comments on the sections relevant to the Agency are given below each extract.

"We have repeatedly been assured that the Government base all decisions on sound science, where applicable. Since the first regulations came into force, the science has moved on considerably, yet there is no indication that this knowledge has found its way into the regulatory arena. In fact, much recent research raises doubts about the origins of the so-called "infectious agent". These and other regulations are based on the widespread assumption that a rogue isoform of a protein designated PrP—called prion protein by some—causes TSEs. That hypothesis is unsupported by rigorous analysis and substantial data contradict it."

The Agency is fully aware of the debate around the nature of the infectious agent and the policy implications should it be shown that this does not include the abnormal prion protein. Following a discussion of the issue, the SEAC minutes of November 2005 note that 'abnormal prion protein may not always be a good surrogate marker for infectivity' and 'that the precise role of abnormal prion protein in relation to the infectious and neurodegenerative properties of TSE agents remains unclear'. To date all of the TSE tests rely on detection of the abnormal prion. The Agency is planning a research call in the near future for the development of a live animal, pre-clinical diagnostic test that can include work on identification of the infectious agent as the basis for the test. (see Annex 7) All proposals are subject to peer review by a panel of TSE experts to ensure that the latest scientific results have been taken into consideration.

"In recent weeks, it has been announced that another type of scrapie, distinct from what some refer to as classical scrapie, has emerged. Professor Christopher Higgins—chairman of the Spongiform Encephalopathy Advisory Committee, or SEAC—was heard to say, on "Farming Today" on 27 February, that there was a classical scrapie but that there is now a new disease, which can be distinguished from both it and BSE and has been named "atypical scrapie". I am astonished.

In 1986, Dickinson et al presented a paper at the second Paris symposium entitled "Further evidence that scrapie agent has an independent genome". In their summary they state that,

"Many distinct strains of scrapie have now been characterised in terms of their biological and neuropathological properties and a number of them have been re-isolated several times from different breeds of sheep and goats".

I understand that the neuropathogenesis unit in Edinburgh had, at that time, isolated more than 25 strains and characterised two-thirds of them in detail. Throughout the 1970s, Dr. Dickinson's group correctly predicted a variety of aspects of TSE infectious agents and their diseases; for example, that infection with more than one strain of scrapie could involve competition between strains in production of disease. These accurate predictions were summarised in 1998 by Farquhar C.F. et al in Nature. Why is it that TSE researchers in Edinburgh, such as those at Moredun Research Institute, are refused information about which scrapie strains have been used as a basis for the design of the National Scrapie Plan?

A number of researchers, including Hugh Reid, former head of microbiology at the Moredun Research Institute—which I shall call the MRI—Ian Aitken and Bill Martin, both former directors of the MRI, and Alan Dickinson, former director of the ARC and MRC Neuropathogenesis Unit, Edinburgh have been closely involved in TSE research, the last named for longer than anyone else worldwide. One of them recommended over 20 years ago that measures to reduce scrapie should avoid genetical extremes. Since then, many more details have been added to their understanding of TSE diseases, but the principles underlying that recommendation have not changed. They say that,

"In particular, the possibility still exists that some TSE strains may exist, or may arise, that will be able to replicate in the purportedly resistant genotype. In this event, the entire sheep population would then be at high risk".

That prediction is now proving to be correct because the "atypical scrapie" cases are in genotypes being selected by the National Scrapie Plan. In previous debates in your Lordships' House I, too, voiced my concerns that this is what would happen."

As noted by the Countess of Mar the observation that some TSE strains can replicate in the purportedly resistant genotype of sheep were reported in the peer reviewed literature in the 1970s and 1980s. In addition more recent work has also supported the possibility that resistant genotypes may not be fully resistant. This possibility has always been recognised when considering the NSP. However the main driver for the Agency's support for the NSP was the decrease in the possible risk to consumers from BSE in sheep, which was not an issue in the 1970s and is still supported by the research results to date. Sheep breeding and culling programs are the responsibility of agricultural departments, and in paragraph 34 of this paper the Board are invited to note that this policy is to be subject to review in the light of current increase in scientific knowledge of atypical scrapie.

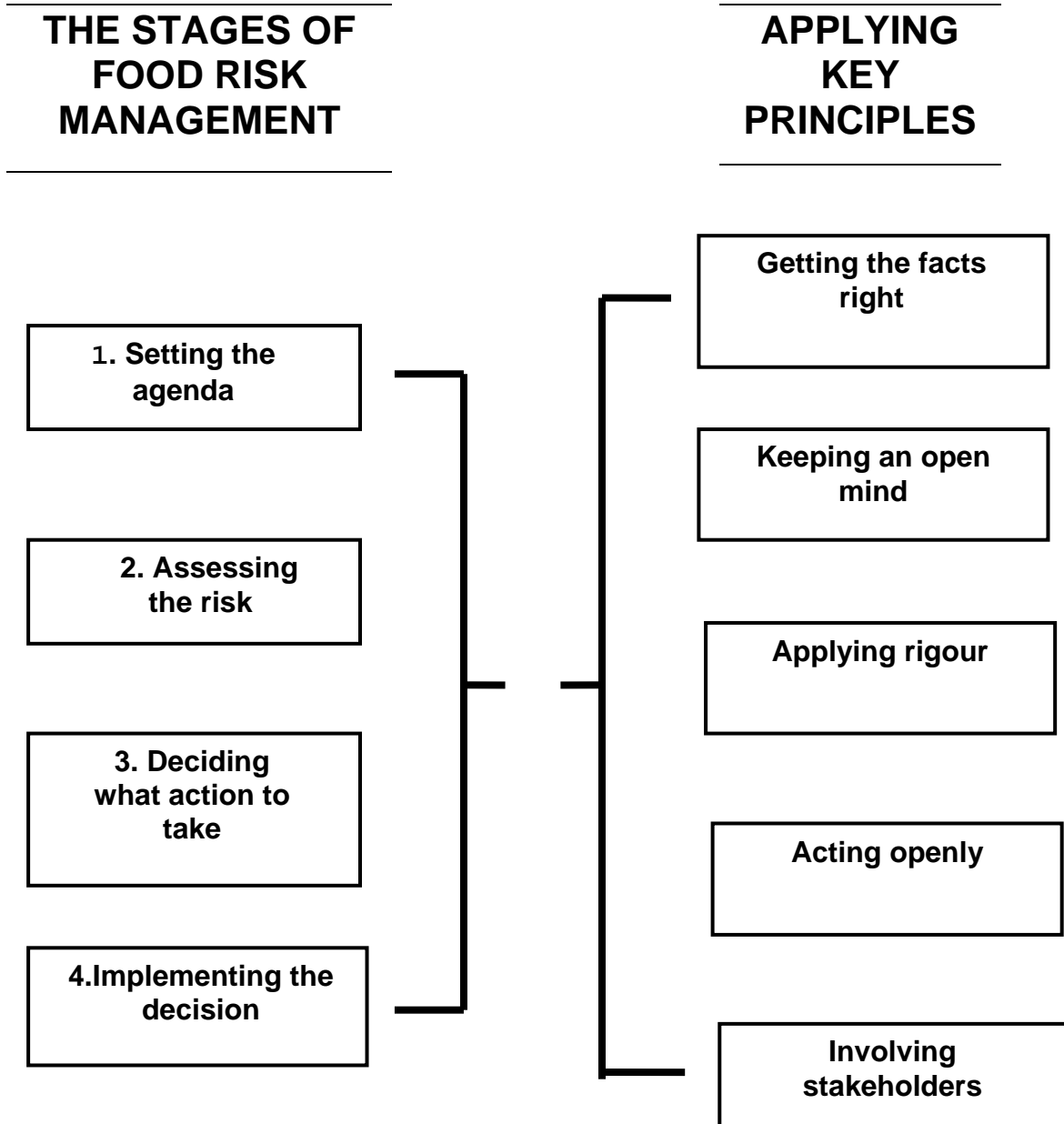
"Most of those currently investigating the nature of TSE agents have chosen to work only with the quickest and highest infectivity titre rodent models and have assumed that extrapolation to other models would be valid. Furthermore, they have almost exclusively used the intracerebral route, which will never be a natural route of infection. That route may simply be the means of transferring amyloid primer and cerebral amyloidosis may be confused with a TSE infection.

There is also concern that the methodology is not always explained in detail, which is essential as, for example, the likelihood of laboratory contamination with TSE agents and cross-contamination between strains is well known. There is now sufficient new and reassessed information to challenge the hypothesis that the proteinase K-resistant PrP present in TSEs is the real infectious agent per se. Can the Minister please explain why, in a situation such as that surrounding scrapie, BSE, CJD or foot and mouth disease, the Government seem to turn to scientists who are not regarded by their peers as experts in the area while ignoring those who are? Is it not in everybody's interests to obtain the very best scientific advice, especially in areas where scientific uncertainties exist? Had we obtained the very best advice from the beginning, I wonder whether we would be in the position of requiring this legislation."

The limitations of using one particular combination of agent strain/host animal as a model for another combination are well known. Models with the closest approximation to those of agricultural or medical importance are used whenever possible. Until the nature of the agent is defined this is the best compromise possible.

Information on research is obtained not only from peer reviewed publications, where it is the responsibility of reviewers and editors to ensure that all necessary information is given, not only for TSE research, but for the entire range of experimental science. Information is also obtained directly from the researchers, often in great detail, for example at scientific conferences, in comprehensive reports on research that has been funded and in presentations to government advisory committees. Therefore there is extensive information from TSE experts on which to base policy decisions. The Agency places great importance on policy being based on sound science and being open about the uncertainties. In so doing the Agency seeks expert opinion and risk assessment advice from SEAC and is open to alternative views. This is underpinned by the Agency's Phillips checklist (annex 9).

APPLYING BSE LESSONS TO FOOD RISK MANAGEMENT



FSA PHILLIPS CHECKLIST

NB Not all questions are applicable to every situation. On each occasion the question “Is this question appropriate to this particular issue/incident?” should be asked.

Key

1. Getting the facts right
2. Keeping an open mind
3. Applying rigour
4. Acting openly
5. Involving stakeholders

Identifying a Risk		actioned
1	Are we being slow in identifying new risks? Are we adequately tracking emerging information on known risks?	√
2	Are we taking account of those with different views to our own? Are we prepared to reopen issues where new facts emerge?	√
3	Are we assessing priorities for attention, and allocating our time sensibly between them?	√
4	Are we being open and honest about our agenda?	√
5	Are we checking with relevant stakeholders what information they have?	√
Assessing the Risk		
1	Are we using our in-house scientific expertise? Are we consulting scientific experts representing all major viewpoints? Are we taking steps to clarify areas of scientific uncertainty?	√
2	Are we taking account of conflicting views?	√
3	Are we undertaking a formal risk assessment? Are we distinguishing adequately between known risk and areas of scientific uncertainty?	Not possible √
4	Are we publishing our risk assessment, and the facts underpinning it?	n/a

	Are we being open and honest about areas of uncertainty?	√
5	Are we asking stakeholders to contribute to the risk assessment?	n/a
Deciding what Action to take		
1	Do we have all the relevant (including economic) facts available?	
2	Are we listening to all interest groups – consumers, enforcement agencies, producers, special interest groups – before deciding what action is appropriate? Is there a case for reconsidering our decision? Do we need to do so?	√
3	Are we considering all the relevant options for action? Are we weighing up their costs and benefits rigorously? Are we considering their practicality and enforceability?	√
4	Are we publishing our decision, and the assessments underpinning it?	√
5	Are we explaining to stakeholders why we have decided on the particular action?	
Implementing the Decision		
1	Are we taking steps to find out about the impact of our decision?	
2	Are we allowing for changes in light of experience?	
3	Are we following through our decision rigorously and effectively? Are we ensuring that it is clearly understood by consumers, producers and enforcers?	
4	Are we publishing our implementation plans?	
5	Are we allowing for feedback from stakeholders?	
Monitoring and Reviewing the Impact of the Decision		
1	Is the decision having the intended effect? Do we need to review it?	
2	If it is not having the intended effect, is that now important? Have the circumstances which led to the decision changed since it was made?	
3	Is the implementation of the decision being rigorously enforced if appropriate?	
4	Are we publishing the results of the enforcement? Are we being open about the impact of the decision?	
5	Are we obtaining feedback from stakeholders on the impact of the decision? Are we consulting stakeholders on any review of the decision?	

DETAILS FROM A REPORT²⁵ ON THE CONSUMER FOCUS GROUPS

BACKGROUND AND OBJECTIVES

1. Objectives

- the primary objectives of the research were threefold:
 - to explore how consumers feel about the potential risk to human health from BSE and atypical scrapie, once it has been explained to them
 - to gauge how consumers think the risk should be managed, and who should take responsibility for doing this
 - to gauge understanding of and response to the FSA's advice and information on atypical scrapie and BSE
 -

METHODOLOGY AND SAMPLE

2. Methodology

- the research comprised seven group discussions with members of the public and two with members of food co-operatives which had won the Agency's Sheila McKechnie award. Each group consisted of seven to nine people.

3. Sample

- the general public sample was configured as follows:
 - G1: 18-25, ABC1, mixed sex, South Wales
 - G2: 26-40, C1C2D, mixed sex, African-Caribbean, Midlands
 - G3: 26-40, C2DE, male, Scotland
 - G4: 41-55, ABC1, female, Scotland
 - G5: 41-55, C1C2D, female, Muslim, London
 - G6: 55+, C2DE, female, South Wales
 - G7: 55+, ABC1, male, Midlands
- all were at least jointly responsible for buying and preparing food for their household and bought and prepared sheep and/or goat products

²⁵ Research conducted and report prepared by Cragg Ross Dawson on behalf of The Food Standards Agency

MAIN FINDINGS

1. Initial response to the issue

1.1 *Prior Awareness*

- there is no unprompted awareness of any health risk associated with eating sheep or goats
 - one or two think they heard about it in connection with the BSE in cattle crisis

Attendees were shown an article from the Daily Mail of 10/03/06

- when they see the Daily Mail article, many are very surprised that they have not heard about this before...
 - so many 'non-stories' make their way into the tabloids, it is surprising that a story with some credible basis was not reported more widely
 - this is doubly surprising given the link to BSE – although BSE is not a current concern, the media are expected to be able to generate considerable mileage out of a 'new BSE crisis'
- particular areas of concern among these respondents are that...
 - the disease is widespread (82,000 sounds like a lot of sheep, though no basis for judging this)
 - the real risk to human health will not be known for several years
- however, only one or two say they might change their behaviour on the basis of this article, and with little conviction
- most think they would need much more evidence to stop them buying sheep and goat products, in the form of...
 - a proven case of harm to human health, especially in their local area
 - much more media noise, particularly on TV and in the broadsheets

- a reaction from the government, particularly a ban on sheep and goat products
- most think that, unless and until these things occur, there is no need to worry too much or to stop buying these foods

1.2 *Response to the FSA's Advice*

- many find the advice confusing, unhelpful and irritatingly vague, in light of the Daily Mail article
- the first para – advising people not to stop eating sheep or goat products – is welcome...
 - seems to be a clear-cut message of reassurance
 - the FSA is quite widely known and generally respected as a trustworthy source
- second para much more problematic...
 - appears to contradict the first para – ‘we don't suggest you do anything, but if you want to do anything...’
 -
 - tone seems weaselly, non-committal – ‘if it were present’, ‘the risk, if any’
- messages about goats, older meat (mutton) and sausages also cause confusion and some concern
- some of those who eat goat (mostly African-Caribbean) and mutton (mostly Asian and African-Caribbean, some white British) wonder whether they should be particularly concerned...
 - African-Caribbean respondents already among the more concerned; this heightens their sense of worry
 - Asian women already worried about the safety and quality of meat from halal butchers; this increases the perceived risk
- reference to ‘older meat’ (e.g. mutton) is puzzling to others
- also considerable confusion about reference to sausages, and some mistakenly assume this is not relevant to them...

- overall, most feel that the tone of the advice suggests that the FSA is not particularly concerned about the risk from BSE or atypical scrapie, and few are more worried than they were before
- however, some Asian and African-Caribbean respondents are now worried about the risk from mutton and goat
- and many are generally irritated by the advice, which they think is woolly

1.3 *Response to Background Information taken from the FSA Website*

- for more articulate respondents, this helps to clear up a number of issues, namely...
 - the relationship between BSE, classical scrapie and atypical scrapie
 - the fact that BSE has not been found in sheep but could theoretically be present, whereas atypical scrapie has been found but is not yet known to cause harm to humans
 - the reason why there is uncertainty as to whether atypical scrapie is a new disease
- but those who find the topic difficult to understand are generally left more confused than before
- among those who take it in, the information on this sheet is less likely to be dismissed than the Daily Mail article, seemingly because it has not come from a tabloid paper

1.4 *Response to other measures the FSA is taking*

- reference to the precautionary measures tends to polarise response
- some (typically those who were not particularly worried about the issue before) feel reassured that measures are being taken to protect them...
 - 'more research' is widely welcomed; seen as an obvious step

- the other measures are not well-understood but suggest that the government is doing what it should be doing to tackle the problem
- for some, this implies that lessons have been learned from the BSE in cattle crisis
- others find the information rather alarming

2. **Considered Response**

- at the end of the discussions, very few think they will change their behaviour as a result of what they have learnt
- the link with the BSE in cattle crisis also appears to blunt potential concern for some respondents...
- among most concerned, only one or two think they might stop eating sheep or goat products altogether, until more is known; a few more think they might cut down the amount they give to their children
- the general consensus is that responsibility for deciding whether or not to keep eating sheep and goat products lies with consumers, as long as they are given sufficient information
- most feel that the FSA/government's role should be to continue researching the issue and publishing information and advice for consumers
- it is generally not thought sufficient for this information and advice to be published on the FSA website...
 - few would think to look there
 - FSA is duty-bound to give people a decent chance of seeing the information