

Scrapie – Advisory notes for farmers



Llywodraeth Cynulliad Cymru
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SCOTTISH EXECUTIVE
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Department for Environment
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Introduction

Scrapie is a fatal brain disease of sheep and goats. Disease develops due to changes in a protein present in the brain known as prion protein. It has been present in British sheep for over 200 years. By law, any animals suspected of having scrapie must be reported to your local Animal Health Divisional Office (AHDO). It is important that every single suspected case of scrapie is reported, not just the first case on your farm. We need more research into scrapie to establish how common the disease is, and to find out the similarities and differences between scrapie and other similar diseases such as BSE, a disease found in cattle, and CJD, a rare disease that is found in humans. To do this, we need farmers to look out for any signs of this disease, and to report it immediately.

This leaflet will tell you what signs to look out for in your sheep and goats, and what to do if you think they may have scrapie.

How do I know if my animals have scrapie?

There is often a general change in temperament or behaviour weeks before more specific signs develop. In most cases animals will show a combination of signs, but none of the signs alone can be regarded as a definite indication of scrapie. Scrapie should be considered in any sheep or goat showing nervous signs or changes in behaviour. If there is no obvious alternative diagnosis, the case must be reported to your Divisional Veterinary Manager (DVM) for further investigation.

When does scrapie occur?

Age

Most cases of scrapie occur in sheep between two and five years of age. Although rarely seen in sheep less than one year of age or over five years old, age alone cannot be used to rule out the presence of scrapie.

It is rarely reported in goats in the UK but, again, the possibility of the disease cannot be ruled out.

Numbers affected

Because clinical signs of disease appear a long time after initial infection, cases generally appear singly in a flock. This is very different from diseases such as sheep scab (psoroptic mange), which can spread quickly through a flock. Occasionally, several cases can occur over a short period of time, mostly in animals born at about the same time and possibly to the same sire.

Season

Cases can occur at any time of year, but stress can cause the clinical signs to appear. Cases are often more easily identified at times when sheep and goats are collected for management purposes, such as when sheep are brought in for lambing, or in the autumn, at tupping.

Development of clinical signs

Most sheep show a gradual development of clinical signs over a period of several weeks or even months, although in some cases an animal's condition may worsen rapidly. Some sheep and goats affected with scrapie may just be found dead without showing any clinical signs beforehand.

Clinical signs of scrapie

Irritation

- repeated rubbing of flanks and hindquarters against objects such as fences, posts or hay racks
- repeated scratching of the flanks
- nibbling or grinding teeth when rubbing themselves or when rubbed firmly on the back

- continued scratching of the shoulder or ear with a hind foot
- unusual or agitated nibbling of the feet, legs or other parts of the body
- excessive wool loss or damage to the skin

Changes in behaviour

- becoming excitable
- drooping ears
- increased nervousness or fear response
- lagging behind
- aggression
- depression or vacant stare

Changes in posture and movement

- trembling (mainly of the head)
- unusual high stepping trot in early stages
- severe incoordination
- stumbling
- standing awkwardly
- weak hind legs
- unable to stand

Later clinical signs

- weight loss
- death

How can I tell whether these clinical signs are caused by scrapie or another disease?

When a combination of the signs on pages 3–4 has been noticed, it is possible your animal has scrapie. You or your vet must report the suspicion to the Divisional Veterinary Manager (DVM) at your local Animal Health Divisional Office. A Veterinary Officer will visit (free of charge) to examine the suspect animal. **IF IN DOUBT, CALL US OUT. WE WOULD RATHER VISIT A CASE WHICH TURNS OUT NOT TO BE SCRAPIE THAN MISS A GENUINE CASE.**

What else could these clinical signs indicate?

If **single signs** occur it is possible that other causes may be to blame:

- **Irritation:** can also be caused by skin parasites such as sheep scab mites or lice.
- **Changes in posture and movement:** can also be seen in other infectious nervous diseases (such as 'Gid', listeriosis, or Maedi-Visna and non-infectious diseases (such as hypomagnesaemia and pregnancy toxaemia).
- **Excessive weight loss:** can also be seen in many other illnesses.

Could I definitely recognise scrapie straightaway?

In individual sheep and goats, scrapie takes a long time to develop, and in the early stages of the disease process there may be no overt clinical signs of the disease. Clinical signs usually appear months or even years after the animal has become infected. The only certain way to find out if an animal has scrapie is by doing laboratory tests on tissues taken from the animal after its death. There is no test available yet which can reliably identify infected live animals before they develop clinical signs although we are optimistic that one will become available within the next few years.

What would happen if I did not report a suspected scrapie animal?

The penalties for not reporting suspected cases of scrapie are severe. Under the TSE Regulations 2002, failure to comply can result in heavy fines or imprisonment, or both. In addition the scrapie compensation payments will not be made.

What will happen when I report that I suspect an animal has scrapie?

As explained earlier, once suspicion of scrapie has been notified to the DVM at your local Animal Health Divisional Office, a Veterinary Officer will visit to examine the animal to confirm or rule out the suspicion of scrapie. If scrapie is suspected, the animal will be humanely slaughtered by lethal injection. This may take place on the farm or holding, but the animal may be transported to a veterinary diagnostic laboratory (at Government expense) if welfare conditions allow.

Transport of live animals to the laboratory means that fresh tissues can be collected; this is very important to enable further research into the disease. Samples of brain and other tissues will then be submitted to a specialist laboratory within the Veterinary Laboratories Agency for diagnosis and research and the carcass will be incinerated. When the tests are complete you will be informed whether or not scrapie was confirmed by the laboratory examination.

If the *clinical* examination of the suspect animal by a Veterinary Officer is inconclusive, the suspect animal will be placed under a movement restriction on the farm where it is examined. These restrictions will remain in place for up to 28 days. The suspect animal is not required to be isolated from other animals unless it is likely to give birth while under restriction. The animal will be seen again by the Veterinary Officer within a few days.

What about the rest of my flock or herd?

If scrapie is confirmed in a sheep or goat on a holding, there can be implications for all of the sheep and goats on that holding.

New or further cases of confirmed scrapie

From Summer 2004, new European legislation will be implemented whereby owners have to join the Compulsory Scrapie Flocks Scheme.

In most cases, all of the sheep on the holding will be genotyped. Restrictions will then be applied on the sheep that can be used for breeding on the holding, or sold off for breeding or for slaughter for human consumption or that have to be destroyed, based on their relative resistance or susceptibility to scrapie. In exceptional circumstances it may be agreed with the owner that all of the sheep should be culled. Goats on the affected holding **will** be culled, as they are known to be uniformly susceptible to scrapie.

For flocks that have had scrapie in the past

Those flocks which have had a confirmed case of scrapie between July 1998 and Spring 2004 will be invited to join the Voluntary Scrapie Flocks Scheme which has similar but less onerous rules than that for the Compulsory Scrapie Flocks Scheme. Note – if you have a confirmed case of scrapie after the Compulsory Scrapie Flocks Scheme (CSFS) is introduced, the provisions of the CSFS will be implemented on your holding. If you are already a member of any other National Scrapie Plan scheme, that membership will be cancelled. Please see the National Scrapie Plan web-pages for the most up to date position on both schemes (www.defra.gov.uk/nsp) or phone the NSP Helpline – 0845 601 4858.

Trading with the EU

Trading of breeding sheep and goats within the EU is subject to regulatory requirements designed to control, among other things, the spread of scrapie. The Scrapie Monitoring Scheme is designed to assist producers who wish to export breeding animals under these arrangements by offering a system that confirms that their flock/herd has been monitored for scrapie. The scheme is currently under review and therefore its requirements may change, but further details about the current arrangements can be obtained from your local Animal Health Divisional Office.

Since 1 October 2003, breeding sheep of the most resistant ARR/ARR genotype (see National Scrapie Plan Genotypes Table on page 14) can be exported even if they have come from a holding that has had scrapie within the previous three years. Nevertheless, they must have been genotyped by the Veterinary Laboratories Agency (VLA) or a laboratory authorised by the Government under the National Scrapie Plan (NSP).

All other genotypes must continue to comply with the regulatory requirements, including the stipulation that no case of scrapie has been confirmed on the holding for at least three years.

However, it should be noted that, regardless of genotype, sheep and goats can only be exported to Denmark and/or Sweden if their flocks have not had a case of scrapie confirmed in the previous seven years.

What happens if the Veterinary Officer does not diagnose scrapie?

If the Veterinary Officer considers that the signs are not caused by scrapie, no restrictions will be served on the animal. You may then wish to contact your vet to discuss an alternative diagnosis, or treatment. If you retain the animal

and signs of scrapie recur, you should report your suspicions again to the Divisional Veterinary Manager.

Why is it vital to report scrapie?

A continuing scrapie problem in a flock would probably severely affect a farmer's business. An additional factor is that, whilst this disease does not appear to pose any direct risk to human health, there is a theoretical possibility that BSE is present in sheep in the UK. Although, so far, it has not been found naturally, laboratory research has shown that sheep can be artificially infected with BSE. Because the external signs of scrapie and experimentally induced BSE are similar, if BSE is in sheep, scrapie may be "masking" it. Therefore, to demonstrate that everything is being done to protect public and animal health, the Government needs to check any suspicion of a scrapie-like illness in sheep and goats.

Reporting any suspect animals is therefore vital in this regard and at the same time provides much needed materials to support the Government's research programme which aims to:

- improve knowledge of the disease
- improve control of the disease
- protect animal welfare
- prevent financial difficulties for the farmer by preventing loss of animals, including valuable breeding stock

As we improve our knowledge about the disease, where it is, how it spreads, and how it can be detected, we may have an opportunity to develop an even more effective long-term control programme. All farmers are therefore encouraged to report their suspicion of the disease, not just because it is a legal requirement, but in order to safeguard the future of the sheep industry. Any evidence that scrapie cases are not being reported also

undermines Government efforts to improve or maintain export trade in sheep and sheep products.

Compensation

Compensation is paid for reported scrapie suspects that are compulsorily slaughtered for diagnosis. For sheep or goats that are confirmed as having scrapie (following a laboratory examination), the compensation to be paid is £30 for a cull animal at the end of its productive life and £90 for any other animal. When the tests on the brain tissues are complete you will be informed as to whether or not scrapie has been confirmed by the laboratory examination. A higher level of compensation (linked to the true replacement cost, but limited to a maximum of £400) may be paid where a suspect animal is compulsorily slaughtered, but the laboratory diagnosis is unable to confirm scrapie. In addition to this, we will pay the diagnosis and carcase disposal costs. (Compensation costs correct at time of printing.)

Compensation will also be paid for animals slaughtered under the Voluntary and Compulsory Scrapie Flocks Schemes (see page 7).

Can I do anything to stop my animals contracting scrapie?

The disease is caused by an infectious agent, which appears to be transmitted from a ewe to her lamb, and to other sheep. The way it is transmitted from sheep to sheep is not fully understood, but infected birth fluids, membranes and cleansings (placenta or afterbirth) probably spread the agent around pastures or buildings during lambing. Sheep should not be permitted to eat placenta.

Regular cleaning of buildings used for lambing may reduce the risk of exposure of the sheep to the agent. Timely and complete disposal of cleansings is considered to be particularly important. These methods may

help to control the spread of scrapie within a flock, but they are unlikely to be 100% effective.

In some cases scrapie has been successfully controlled by culling the relatives of scrapie cases from the flock/herd. To be successful this requires clear identification of animals and adequate record keeping. You can also breed increased natural resistance into sheep flocks through the use of scrapie genotyping (see later section on genotyping). You should also ensure that sheep are purchased from verified sources.

What records do I need to keep?

It is very important that your records are kept up to date. It is particularly important that records of a flock's breeding and management history are held. Records kept under the rules of the Sheep Annual Premium Scheme, or those required under the Sheep and Goats Identification and Movement (Interim Measures) (England) (No.2) Order 2002 (as amended) – or under the



equivalent legislation for Wales and Scotland (see below) – are not likely to be adequate to effectively manage an outbreak of scrapie by selective breeding from unaffected, possibly scrapie resistant, animals. (Equivalent legislation for Wales and Scotland is The Sheep and Goats Identification and Movement (Interim measures) (Wales) (No.2) Order 2002 (as amended) and The Sheep and Goats Identification (Scotland) Amendment Regulations 2002.) All movements on and off the farm require recording in the on-farm record book. This includes movements to tack, common grazing and within Sole Occupancy Authorities (SOAs).

The combination of movement records and the use of earmarks in breeding records does help to tackle the problem when disease is found. All lambs and kids must be individually identified; there is, however, no corresponding requirement to record individual identification numbers.

‘Genotyping’: What is it, and how can it help me?

Studies have shown that there is a gene in sheep that determines whether an animal is likely to develop scrapie if exposed to infection. Certain sheep, if exposed to scrapie, are therefore more likely to get the disease than others. Goats appear to be more uniformly susceptible, but the same gene may control the time it takes for clinical signs to show. So, by careful record keeping and a strict breeding plan, it is possible to reduce the number of cases in a flock by selectively breeding to produce sheep which are least likely to get the disease. It is possible to identify resistant or susceptible sheep by a blood test. This test, on a small blood or tissue sample, is called ‘genotyping’ and the result is known as the ‘genotype’ of the animal.

Genotyping and breeding for resistance were recommended by the European Union Scientific Steering Committee and the Spongiform Encephalopathy Advisory Committee and is also endorsed by the Food Standards Agency as a valuable tool in the control of scrapie.

National Scrapie Plan

The National Scrapie Plan (NSP) is a long-term plan which consists of a number of genotyping schemes and initiatives aimed at increasing the genetic resistance to scrapie in the national sheep flock.

The NSP Genotypes Table on the next page shows the 15 known genotypes which occur in sheep and their relative resistance or susceptibility to scrapie. The prevalence and frequency of each genotype differs between breeds. This table is subject to continuous scientific review.

NSP genotypes table

Genotype result	NSP Type	Degree of resistance/susceptibility
ARR / ARR	1	Sheep that are genetically most resistant to scrapie.
ARR / AHQ ARR / ARH ARR / ARQ	2	Sheep that are genetically resistant to scrapie, but will need careful selection when used for further breeding.
AHQ / AHQ AHQ / ARH AHQ / ARQ ARH / ARH ARH / ARQ ARQ / ARQ	3	Sheep that genetically have little resistance to scrapie. Sale and breeding restrictions apply in NSP flocks, which are breed specific.
ARR / VRQ	4	Sheep that are genetically susceptible to scrapie and should not be used for breeding unless in the context of an approved and controlled breeding programme.
AHQ / VRQ ARH / VRQ ARQ / VRQ VRQ / VRQ	5	Sheep that are highly susceptible to scrapie and should not be used for breeding.

Current science indicates that as a result of the NSP we should, in due course, see a very significant reduction in the incidence of scrapie and eventually its disappearance from the national flock. It is hoped that as many eligible breeders as possible will want to play their part in the NSP.

All genotyping, electronic identification and certification are free in NSP flocks. For the foreseeable future it is likely that NSP certified resistant rams may realise higher prices at some breed sales.

European Commission Decision 2003/100 EC requires Member States to introduce genotype based sheep breeding programmes for resistance to Transmissible Spongiform Encephalopathies (TSEs), including scrapie, on a compulsory basis from 1 April 2005.

NSP semen archive

The Semen Archive is being established to store semen from rams of those genotypes that over time will be removed through Ram Genotyping Schemes. This archive should enable the re-establishment of viable populations of these sheep in the future if it is thought necessary. It will also help to protect against the possible loss of beneficial breed characteristics such as hardiness.

Operations will be overseen by a joint Industry-Government Management Board, who will approve the nomination of rams, which must be via breed societies. If you wish to find out more about the Semen Archive please contact your breed society, or the National Scrapie Plan Administration Centre (see below for contact details). The Semen Archive Management Board Secretariat can also provide information:

Tel: 01684 892661

E-mail: samb@nationalsheep.org.uk

For more information on the NSP, or genotyping, please contact:

NSP Helpline: 0845 601 4858 (calls charged at local rate)

Or write to:

National Scrapie Plan Administration Centre

Defra

Whittington Road

Worcester

WR5 2SU

E-mail: nspac@defra.gsi.gov.uk

Web pages: <http://www.defra.gov.uk/nsp>

A three year research project has been commissioned to see if there may be associations between scrapie **genotyping and performance, survival and health characteristics**.

For more information about the project, or to register an interest in participating, please contact the Veterinary Laboratories Agency (VLA) (see below for contact details).

In addition, both the VLA and the Institute for Animal Health (IAH) are running studies on the spread and control of scrapie.

For further information on enrolling in one of the scrapie studies at VLA or IAH contact:

Rachel Eglin

Veterinary Laboratories Agency, Addlestone,

Surrey KT15 3NB

Tel: 01932 357775

E-mail: r.d.eglin@vla.defra.gsi.gov.uk

Matthew Baylis

Institute for Animal Health, Compton, Newbury, Berkshire
RG20 7NN

Tel: 01635 578411

E-mail: matthew.baylis@bbsrc.ac.uk

TSE testing – active surveillance

Under EU Regulation 999/2001 (as amended) the UK is currently required to test a prescribed number of sheep aged over 18 months and slaughtered for human consumption (abattoir survey). These samples will be taken at the abattoirs that process this category of sheep.

The Regulation also requires the testing of a prescribed number of sheep and goats aged over 18 months and not destined for human consumption (fallen stock survey). These samples will be collected directly from farms. Farmers offering fallen stock should in the first instance contact their local Animal Health Divisional Office.

For any queries on fallen stock active surveillance please contact:

TSE Scrapie Testing Section

Defra

(020) 7904 6538

Further information

If you have any queries or concerns regarding scrapie, please first contact the Divisional Veterinary Manager at your local Animal Health Divisional Office. Contact details are at:

<http://www.defra.gov.uk/corporate/contacts/ahdo.htm>

or in your local telephone directory. You may, of course, wish to discuss any suspicions of scrapie in your flock/herd with your own vet.

Alternatively you can direct any questions to officials at the main offices of the relevant Agriculture and Rural Affairs Departments.

For England:

Department for Environment, Food and Rural Affairs (Defra)
Area 2/04
1a Page Street
London SW1P 4PQ

Tel: (020) 7904 6175

For Scotland:

Scottish Executive Environment and Rural Affairs Department (SEERAD)
Room 349, Pentland House
47 Robb's Loan
Edinburgh EH14 1TY

Tel: (0131) 244 6413

E-mail: scrapie@scotland.gsi.gov.uk

For Wales:

Welsh Assembly Government, Department of Environment, Planning and Countryside (DEPC)
Animal Health Division 1, Cathays Park
Cardiff CF10 3NQ

Tel: (029) 2082 5996

You may also wish to find out more about scrapie through the following sources:

Defra website

If you have access to the Internet, as well as the information on the NSP website you can view the latest general scrapie information on Defra's Internet website, at the following location:

<http://www.defra.gov.uk/animalh/bse/bse-science/level-4-scrapie.html>

This website provides further details on areas such as research, symptoms, genetics, incidence, surveillance and controls, as well as specific information on goats and many other subjects.

Scrapie video

A video on the subject of scrapie in sheep and goats is also available from Defra entitled:

Scrapie: clinical signs in sheep and goats
(1995, 19 minutes, V785)

The video is available at the cost of £5.00 excluding VAT from:

Defra Publicity Branch, Room 221,
Whitehall Place (East Block),
London SW1A 2HH

For enquiries, please telephone: 020 7270 8725

Fax enquiries: 020 7270 8717

or by E-mail to: peter.millard@defra.gsi.gov.uk

Prices include postage and packing to all destinations.

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