

Scrapie

**USDA APHIS VS Career Services Program
Program Diseases Training Module**

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Scrapie

This course is designed to provide updated information on the major domestic diseases for which Veterinary Services (VS) has program responsibility. It will provide information on surveillance, disease control and eradication for these diseases. It will also give an overview of the duties of a field Veterinary Medical Officer (VMO) as a support worker of VS animal disease programs and how they interact with other units in APHIS.

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1. DISEASE INFORMATION

a. Agent

Scrapie is a naturally occurring infectious neurodegenerative disease of sheep and goats characterized by vacuolar or spongy changes in the central nervous system.¹ Scrapie is believed to be caused by an abnormal protein called a prion. Several other theories regarding the infectious agent have been proposed. Prions are an abnormal form of a normal prion protein found in the brain and other tissues. The abnormal prion protein is resistant to digestion with enzymes that breakdown normal protein. The prion is also highly resistant to environmental conditions and substantially resistant to procedures such as cooking or rendering, as well as many disinfectants. The abnormal prion protein appears to cause the normal cellular prion protein to be converted to an abnormal form that cannot be removed by the cell, causing it to accumulate in the brain. Prions do not evoke any detectable immune response or inflammatory reaction in host animals, so formation of scrapie-specific antibodies does not result.^{1,2}

b. Transmission

Scrapie behaves as an infectious and contagious disease, but the means of transmission is not fully understood.⁵ It is traditionally recognized that the scrapie disease agent is transmitted maternally and horizontally. The scrapie agent in sheep appears to be most commonly passed from ewe to lamb in the period from parturition to weaning and also, possibly, prenatally. The evidence supporting prenatal infection is weak. Infection can also pass laterally to unrelated sheep or goats, especially when parturition occurs in confined areas. Fetal membranes and fluids are thought to be a source of infection.^{2,3} The oral route appears to be the most important route of natural infection.⁵ The ram does not seem to play a significant role in transmission as the agent has not been found in the seminal fluids or sperm.⁴ It is not likely that scrapie is transmitted through urine, feces or milk.² The genetics of the sheep affects their susceptibility to scrapie.³

There is presently no evidence that scrapie poses a risk to humans. In the laboratory, the scrapie agent has been transmitted to hamsters, mice, rats, voles, gerbils, mink, cattle, and some species of monkeys by inoculation.³

The scrapie agent is resistant to freezing, thawing, boiling for 30 minutes, storage at room temperature for 42 days, exposure to ultraviolet light and incubation with 0.35% formalin. The scrapie agent is not inactivated by acetyleneimine, a compound that inactivates viral nucleic acids.²

c. Clinical signs

Signs of scrapie usually appear 2 to 5 years of age in sheep exposed at or near birth with the majority being four years of age. Sheep exposed post-weaning are less likely to become infected and when infected typically show signs at seven years of age or older. Signs of scrapie vary widely among individual animals and develop very slowly. Due to damage to nerve cells, affected animals usually show behavioral changes, tremor (especially of head and neck), pruritus, and locomotor incoordination that progress to recumbency and death.^{1,3}

Early signs include subtle changes in behavior or temperament. These changes may be followed by scratching and rubbing against fixed objects, apparently to relieve itching.³ The name

scrapie comes from this observation.⁴ Other signs are loss of coordination, weight loss despite retention of appetite, biting of feet and limbs, lip smacking, and gait abnormalities, including high-stepping of the forelegs, hopping like a rabbit, and swaying of the back end.³

An infected animal may appear normal if left undisturbed at rest. However, when stimulated by a sudden noise, excessive movement or the stress of handling, the animal may tremble or fall down in a convulsive-like state.³

Onset of clinical disease may exceed the commercial life span of the sheep.¹ Sheep may live one to six months or longer after the onset of clinical signs, but death is inevitable.³

d. Disease differentials

The clinical signs may, especially in the early phase of the disease, resemble those of some other conditions of adult sheep. These include: ectoparasitism, pseudorabies (Aujeszky's disease), rabies, encephalic listeriosis, ovine progressive pneumonia (maedie-visna), pregnancy toxemia (ketosis), hypomagnesaemia and chemical and plant intoxications.¹

e. Epidemiology

Scrapie is widespread and can be found in the United Kingdom, Ireland, France Belgium, Iceland, Norway, Cyprus, Israel, Japan, Canada, parts of Asia, and the United States. Outbreaks have occurred in Australia and New Zealand but the disease was eradicated.^{2,3,5} In flocks where scrapie is endemic the number of affected animals may reach 40%, but it is usually much less. Case fatality rate is 100%.²

While sheep are the natural host for the disease, infection can also be maintained in goats. Scrapie can be propagated in goat herds that have no direct contact with sheep. The incubation period in goats is slightly shorter than in sheep. At this time no resistant genotypes of goats have been identified, therefore, all goats are assumed to be susceptible.⁴

While there is no demonstrated breed susceptibility to scrapie, it has occurred most commonly in the Suffolk breed in the United States. It has also been diagnosed in a Border Leicester, Cheviots, Corriedales, a Cotswold, Dorsets, Finn sheep, Hampshires, Merinos, Montadales, Rambouellets, Shropshires, Southdowns, Texel and a number of crossbreeds. It is presumed that some flocks are infected but remain undetected due to the disease's long incubation period and difficulty in making a diagnosis.

While the interaction between the scrapie agent and host genetics is not fully understood many important discoveries have come to light in recent years. All animals have a gene that codes for a normal prion protein (PrP) which is present in all tissues. The function of the normally occurring cellular PrP is unknown. Scrapie is believed to be caused by an abnormally conformed prion protein PrP_{scrapie}, which serves as a template to influence a geometrical conformation change in the normal PrP. This abnormal PrP scrapie accumulates and after a period of years, it causes nervous system dysfunction and eventually death. The PrP scrapie proteins are found in the nervous system, the spleen, lymph nodes, placenta, intestine, blood, pancreas, ovary and liver of infected sheep.

The gene that encodes the normal PrP has polymorphisms at codons 136, 154 and 171. Polymorphisms at codon 171 consist of Glutamine (Q), Arginine (R), Histidine (H) or Lysine (K)

(over 99% of U.S. cases of scrapie have been QQ at codon 171). These polymorphisms influence the ability of the normal PrP to be altered by the abnormal PrP_{scrapie} template when the animal is exposed to it. Sheep with genotype RR are considered resistant to scrapie, sheep with QR genotype are considered rarely or much less susceptible and sheep with the genotype QQ are considered highly susceptible to scrapie.^{4,6}

f. Diagnosis

There are no specific gross pathological changes seen in scrapie. A serological immune response to the scrapie agent in infected sheep and goats has not been detected. Currently, a diagnosis is based on histopathological changes in the brain, detection of the abnormal prion protein in brain, lymph nodes, or tonsils by immunohistochemistry or immunoblotting, or third eyelid lymphoid tissue by immunohistochemistry.¹

Histological lesions are confined to the CNS, for the most part in the brain. Neuropathology is bilaterally symmetrical and is characterized by spongiform change and neuronal vacuolation. There may also be reactive astrogliosis and amyloid plaque formation.¹ Because incidents of scrapie are recorded in which neuronal vacuolation was undetectable, diagnosis of suspected scrapie cannot be refuted by a failure to find significant vacuolar changes in the brain.¹

A more sensitive diagnosis can be made by detecting the abnormal prion protein in the central nervous system or lymphoid tissue. Accumulations of the prion can be found in unfixed brain extracts by immunoblotting and in fixed tissues by immunohistochemistry. The diagnosis can also be confirmed by finding characteristic fibrils of the prion protein with electron microscopy in brain extracts. Some of these tests can be used on frozen or autolyzed brains.⁵

Immunohistochemistry on third eyelid lymphoid tissue biopsies was approved by APHIS in October 2001 for scrapie program use. Immunohistochemistry on lymph nodes and tonsils was approved by APHIS for program use in November 2003. New tests to diagnose scrapie in live sheep are being investigated. These include fluorescent labeled peptides to detect abnormal prion protein in the blood and immunoblotting to detect abnormal prion protein in the blood, cerebrospinal fluid or tissues.⁵

g. Prevention and control

Since there is no effective method for treating sheep with scrapie, control measures to prevent the spread of the disease are especially important. Prevention for the herd/flock owner is based on maintaining a closed flock. New animals (especially females) introduced into the flock must come from reliable sources known to be free of scrapie.

The scrapie agent is highly resistant to many standard disinfection procedures such as heating, ultraviolet and ionizing radiation (including levels normally used to treat foodstuffs), and exposure to formalin. Wet heat is much more effective than dry heat. A single porous load autoclave cycle of 274-281 °F for 60 minutes is recommended for disinfection of instruments. The approved chemical disinfectant is sodium hypochlorite (bleach), at a concentration of 2.0% available chlorine for one hour at room temperature (at least 18.3 °C) or a 1-2 molar solution of sodium hydroxide (5-10 ounces of lye dissolved in one gallon of water).⁴ Both of these solutions are corrosive. Their material safety data sheets should be reviewed before use and appropriate protective equipment used.

Contaminated pastures or paddocks should be left empty of goats and susceptible sheep. The precise time required for natural inactivation of the scrapie agent in the environment is unknown. Stalls, corrals, and contaminated sheds should be thoroughly cleaned and then disinfected with one of the approved chemical disinfectants.

h. Related diseases

The Transmissible Spongiform Encephalopathies (TSE) family of diseases includes bovine spongiform encephalopathy (BSE); transmissible mink encephalopathy; feline spongiform encephalopathy (FSE); chronic wasting disease of deer and elk (CWD); kuru; both classical and variant Creutzfeldt-Jakob disease (CJD or vCJD); Gerstmann-Straussler-Scheinker syndrome; and fatal familial insomnia. TSE's have also been reported in Europe in captive wild or exotic animals including ruminants in the bovid family, cats, and monkeys. The occurrence of TSE's in captive wild animals is believed to have resulted from BSE-contaminated feed.^{2,3}

i. Public health consequences

There is no scientific evidence to indicate that scrapie poses a risk to human health. There is no epidemiologic evidence that scrapie of sheep and goats can be transmitted to humans, such as through contact on the farm, at slaughter plants, or butcher shops.³

Creutzfeldt-Jakob disease and kuru in humans have pathologic and clinical similarities to scrapie in sheep. The relationship, if any, between these etiologic agents is as yet unknown. Scrapie has been transmitted to cynomolgus monkeys by inoculation of brains from naturally infected sheep, goats and mice. Monkey species more closely related to humans have not become infected when experimentally inoculated with scrapie. The disease produced in the monkeys was clinically indistinguishable from that of Creutzfeldt-Jakob disease. An epidemiologic study performed in France where the prevalence rate of scrapie is high showed no relationship between the occurrence of scrapie in sheep, the rate of lamb consumption, and the occurrence of Creutzfeldt-Jakob disease in human beings. As with any infectious agent, it may be wise to employ precautions to minimize the potential for exposure to the agent.² The World Health Organization recommends the exclusion of small ruminants showing signs of a TSE from slaughter to address a theoretical risk.³ USDA regulations preclude the slaughter of sheep and goats showing neurological signs.

j. Economic Impact

It is estimated that U.S. producers incur more than \$20 million year in losses and millions of dollars more in lost potential markets and flock productivity due to scrapie.⁷ The presence of scrapie prevents the export of breeding stock, semen, and embryos to many other countries. As New Zealand and Australia are recognized as scrapie free, they are currently the only nations that can freely sell breeding stock.

2. HISTORY OF DISEASE AND CONTROL PROGRAM

First recognized as a disease of sheep in Great Britain and other countries of Western Europe more than 250 years ago, scrapie has been reported throughout the world. Only two countries Australia and New Zealand are recognized by the United States as being free of scrapie.

The first case of scrapie in the United States was diagnosed in 1947 in a Michigan flock. The flock owner had imported sheep of British origin through Canada for several years. In the United States, scrapie has primarily been reported in the Suffolk breed. It also has been diagnosed in a Border Leicester, Cheviots, Corriedales, a Cotswold, Dorsets, Finn sheep, Hampshires, Merinos, Montadales, Rambouillets, Shropshires, Southdowns, and a number of crossbreeds. Through September 2003, approximately 2,360 cases in sheep and 12 cases in goats have been reported in the United States.^{3,14}

The U.S. has had a scrapie eradication or control program in place since 1952. Over the years it has been changed several times. The program was established to help increase animal health status and decrease production losses for producers. Infected flocks that contain a high percentage of susceptible animals can experience significant production losses. In flocks where scrapie is endemic the number of infected animals increases and the age at onset of clinical signs decreases over a period of several years making these flocks economically unviable.

More recently, increased attention and concern is being paid to all TSE's, including scrapie, as a result of the discovery of BSE in cattle, feline spongiform encephalopathy (FSE) in cats and variant Creutzfeldt-Jakob disease (vCJD) in people in Europe. This increased concern has led to packers and producers having difficulty in disposing of sheep offal and dead sheep causing them to incur significant increases in disposal costs. Other countries have expressed concerns and have indicated that they may prohibit or restrict certain ruminant products because the U.S. has scrapie.

Our domestic and international markets for sheep-derived meat and bone meal have also been adversely affected, and the American Sheep Industry Association identified scrapie as a major impediment to the well-being of the U.S. sheep industry. The combination of all these factors has led to the decision to develop a strong scrapie eradication program.^{3,8}

3. CURRENT CONTROL PROGRAMS

a. Scrapie Flock Certification Program

Since 1952 the U.S. Department of Agriculture and the sheep industry have attempted to eradicate scrapie through various approaches. The purpose of all previous USDA scrapie programs was to identify scrapie and eradicate it. That approach changed by implementation of the Scrapie Flock Certification Program (formerly Voluntary Scrapie Flock Certification Program) on October 1, 1992. The certification program is a voluntary, cooperative effort among producers, allied industry representatives, accredited veterinarians, State animal health officials and APHIS. The program provides participating producers with the opportunity to protect their sheep from scrapie and to enhance the marketability of their animals through certifying their origin in scrapie-free flocks. Any sheep or goat owner or manager may apply to participate in the Program.⁹

1) Classification of flocks for the program.

a) Complete monitored (Enrolled or Certified). Once a flock is approved to participate in the Program, that flock will be considered an Enrolled flock with a status date based on the date the application is approved by the Board. Advancement to Certified status is based on an Enrolled flock's compliance with the program standards and maintenance of a status date that is longer than five years.¹⁰

b) Selective monitored (Select). This category is open to any flock and is mainly intended to help slaughter-lamb producers who wish to have an additional method of scrapie surveillance in large production flocks.¹⁰

c) Exposed. Any flock in which a scrapie-positive or suspect animal was born or lambed. Any flock containing a female high-risk or suspect animal or that once contained such an animal that lambed in the flock and from which obex and lymphoid tissues either were not submitted for official testing or were not found negative.⁹

d) Infected. Any flock in which it is determined that a scrapie-positive female animal has resided unless an epidemiological investigation conducted by a State or APHIS representative shows that the animal did not lamb or abort in the flock.⁹

e) Source. A flock in which at least one animal was born that was diagnosed as a scrapie-positive animal at an age of 72 months or less, or in which a scrapie-positive animal has resided throughout its life.⁹

2) Oversight. Oversight of this program is through the National Scrapie Oversight Committee, a national committee composed of industry, state and federal representatives. Priority is given to producers enrolled in the program. The committee reviews the Scrapie Flock Certification program and makes individual recommendations regarding policy and technical improvements in the program.¹¹

3) State Scrapie Certification Boards. A State Scrapie Certification Board will be formed within each State. Composed of the APHIS Area Veterinarian-in-Charge, animal producers, accredited veterinarians, and state animal health officials, members are appointed by the AVIC in cooperation with the state animal health official and flock owners. They will meet yearly, be chaired by an enrolled producer and encourage board members who own sheep or goats to be enrolled in the program, and support the Program at local sheep and goat events and in the sheep and goat producing community. Boards have flexibility to delegate authority or enact more stringent requirements to fit their state's scrapie situation. Duties will include: 1) administer the SFCP. 2) Review Program enrollment applications and status advancement 3) Review situations that may result in a reduction of certification status or dismissal from the program and 4) Educate producers regarding scrapie.¹¹

4) Duties of program participants. Duties of APHIS with regard to the Scrapie Flock Certification Program are to perform inspections, provide guidance and education, and collect and submit diagnostic samples in cooperation with State animal health agencies and accredited veterinarians as well as maintain a records database for use in the program.

Inspections for complete monitored and selective monitored flocks which occur every 11-13 months with animals inspected for official identification, signs of scrapie and records examined for completeness, accuracy and all acquisitions departures, births and deaths

The Centers for Epidemiology and Animal Health (CEAH) maintain a national information database, the Scrapie National Generic Database (SNGD) which includes information about enrolled and certified flocks in the SFCP and which generates a public web page with SFCP flock status information. As of January 31, 2004, there were 1,843 flocks participating in the Scrapie Flock Certification Program (SFCP).

Producers are responsible for establishing and maintaining records, making animals and records available for inspection by APHIS representatives or State animal health officials and State Scrapie Certification Board representatives. They will authorize access to records maintained by breed associations, registries, livestock markets, and packers. Producers will identify animals with official identification as specified by the Program standards, and allow State, Federal, or State Scrapie Certification Board representatives to enter their premises to carry out Program procedures. For these inspections producers will have the necessary facilities and personnel available to assist in the inspection of animals and animal records. It is also the producer's responsibility to report scrapie-suspect animals to a State animal health official, APHIS representative, or an accredited veterinarian and ensure that tissue samples are collected and submitted for diagnostic purposes. Producers will report acquisitions of lower status or nonparticipating animals.

Breed registries and associations, livestock markets, and packers will maintain liaison with flock owners and assist in providing flock owners with information on the Scrapie Flock Certification Program. They will facilitate the information exchange regarding transfer of animals; and encourage information reporting between flock owners, State animal health agencies, and APHIS.¹¹

5. Entry into the program. Entry into the program requires completed application by the producer, a list of identifiable animals including breed and sex information, statement by an accredited veterinarian, State or APHIS representative that the flock is scrapie free along with an inspection report verifying all of the information and animal identification.¹¹ State or Federal animal health officials will provide each enrolling Program participant and his or her accredited veterinarian with an educational scrapie review.

Once approved to enter the Program, a flock will be issued an enrollment date. This date will serve as the status date until the flock fails to meet any of the Program standards. For flocks that acquire animals or commingle with animals that do not meet Program standards, the status date will change to the date the non-enrolled animals are acquired or commingled, or to the status date of the flock with the most recent status date for animals from Enrolled flocks. The enrollment date will not change.¹¹

6. Advancement to Certified status. When a flock has obtained a status date that is over 5 years old it may apply to advance to Certified status. The State Scrapie Certification Board will review advancement applications for movement from Enrolled status to Certified status. The application package includes: 1) A completed Program advancement application. 2) A list of animals in the flock, including official identification numbers, breed, and sex

information and additional information required on acquired and natural additions. 3) A statement by an accredited veterinarian declaring that to the best of his or her knowledge there has been no evidence of scrapie in the flock since the status date; and 4) An inspection report by a State or APHIS regulatory official, including verification of the information provided on the application by the flock owner and verification of the official identification of each animal listed in the application.

7. Maintenance of status. During the annual inspection, a flock's status date will be maintained if the minimal requirements for the status currently held by a flock have been fulfilled. Scrapie must not have been diagnosed in the flock and not been deemed a source flock since its enrollment. The flock must also only have acquisitions or commingling of animals approved for the current status.¹¹

8. Downgrading of certification status.

a) Scrapie is found. If an Enrolled or Certified flock has a confirmed diagnosis of scrapie or is revealed through epidemiologic investigation to be a source flock, the flock will be removed from the Program.¹¹

b) Female animals are added. If a Certified flock acquires or commingles with female animals that are not from a Certified flock, the status of the Certified flock will be lowered to Enrolled, and its status date will change. If an Enrolled flock acquires or commingles with female animals from a nonparticipating flock or from a flock with a more recent status date, the receiving flock will continue with Enrolled status, but the status date will change. After acquiring or commingling with female animals from nonparticipating flocks, the receiving flock's status date will become the date that the female animals were acquired or commingled. After acquiring or commingling with female animals from Enrolled flocks with a more recent status date, the receiving flock will have its status date changed to the most recent status date of the flocks involved.¹¹

c) Male animals are added. Current scientific information provides evidence that the use of unknown-status rams poses a negligible risk of transmitting scrapie to breeding flocks. A ram or buck that is officially identified; shown on the flock inventory; not a scrapie suspect, scrapie-positive, affected, exposed, or high risk animal or an animal that has resided in a source, infected, or exposed flock in which the records are either unavailable or inadequate to determine whether an animal is an exposed or high-risk animal, or not currently part of an infected or source flock can be used in or acquired by an SFC-enrolled flock – including certified flocks—without lowering the flock or herd's status. A ram or flock joining a higher status flock will retain the status of its flock of origin. A ram joining a lower-status flock will take on the status of the lower-status flock. A ram or buck residing in a higher-status flock will be identified in the flock records with its individual SFCP status date and will advance in the certification program based on this date. If the flock or herd's status is reduced and the male's individual status is higher than the new status date, the male's status will be lowered to the flock or herd's status date. Lower-status rams and bucks must be kept separate from ewes and nannies during lambing (kidding) and from lambs and kids less than six months of age.

b. National Accelerated Scrapie Eradication Program (NASEP)

USDA has initiated a national accelerated scrapie eradication program. This cooperative State-Federal-Industry program is administered by APHIS and consistent states (a State that conducts an effective State scrapie control program and is in compliance with federal regulations to control and eradicate scrapie).⁷ The National Accelerated Scrapie Eradication Program is supervised by full-time animal health officials employed by the State or the Federal Government. Designated Scrapie Epidemiologists (DSE's) are appointed by APHIS to make decisions about the use and interpretation of diagnostic tests and field investigation data and the management of scrapie affected flocks in their state.⁹ Under the NASEP the Scrapie Ovine Slaughter Surveillance (SOSS) was established to estimate the national and regional prevalence of scrapie in mature ewes The Regulatory Scrapie Slaughter Surveillance (RSSS) was also initiated in April of 2003. It is a targeted slaughter surveillance program which is designed to identify infected flocks for clean-up.⁹

The National Accelerated Scrapie Eradication Program is based on the following key concepts^{7,8,9}

1) Identification of preclinical infected sheep through live animal testing and active slaughter. States may pilot live-animal sampling programs for at-risk populations on farms, at markets, or as a condition to participate in a sale in areas of high scrapie prevalence or in high-prevalence classes within an area or for other appropriate reasons. Movement beyond the pilot stage will be based on the relative cost of identifying new infected flocks through the piloted methods as compared with slaughter surveillance.

2) Effective tracing of infected animals to their flock/herd of origin made possible as a result of the new identification requirements. Sheep or goats must be identified to their flock of origin, and for an animal born after January 1, 2002, to its flock of birth, by the owner of the flock. Records must be maintained and available for inspection and copying for at least five years after the person has sold or otherwise disposed of the sheep or goat to another person. Program-approved means of identification include tamper resistant ear tags approved by APHIS, flank or ear tattoos (or in goats registered with the American Dairy Goat Association tail web tattoos) or electronic identification (radio frequency identification (RFID) and/or premises identification.

3) Clean up strategies. The National Genetic Based Flock Clean-up Plan will provide effective cleanup strategies which will allow producers to stay in business, preserve breeding stock, and remain economically viable. APHIS will do this by providing the following to exposed and infected flocks/herds that participate in cleanup plans.

- a) Indemnity for high-risk, suspect, and scrapie positive sheep and goats, which owners agree to destroy. Indemnification will be based on commercial market prices reported by USDA's Agricultural marketing Service.⁴
- b) Scrapie live-animal testing.
- c) Genetic testing. As of January 1, 2004, the USDA APHIS has approved six laboratories to conduct official scrapie genotyping tests.¹²
- d) Testing of exposed animals that have been sold out of infected and source flocks/herds.³
- e) Assistance with disposal costs⁴

Genetic based flock clean-up and monitoring is a new aspect of the scrapie eradication program. The plan applies only to scrapie infected, source and exposed flocks. The basic steps of the plan include three steps:

- a) When an infected flock has been identified, the sheep are genotyped. The sheep's genotype determines its risk for scrapie infection. Research indicates that QQ sheep and goats are the most susceptible to scrapie infection, whereas QR sheep are much less susceptible and RR sheep are resistant.⁹
- b) Susceptible genotypes are either removed or their movement restricted. If scrapie is diagnosed in a flock, APHIS will either provide genetic testing and removal of genetically susceptible breeding stock, or in some cases, whole flock depopulation. Indemnity will be paid on sheep and goats that are removed as part of the flock plan. The movement of ewes, rams, does and bucks will be restricted until the owner, the State and APHIS agree to a flock clean-up plan and a post exposure management monitoring plan (PEMMP). Exposed animals that are determined not to be susceptible and that are not the female offspring of a scrapie positive female animal will be released from restriction when the flock plan has been completed. If producers elect to retain exposed susceptible animals, restrictions will be placed on these animals and any susceptible animals that are acquired through birth or other means. Sheep and goats that were moved out of the flock after the infection was believed to have been introduced will be traced to their new owners. Genetic testing, scrapie testing and/or removal of exposed animals will be paid for by APHIS.⁴

In most cases producers will be able to keep many more of their sheep with a genetics based plan. It is estimated that, on average, 60% of a flock can be preserved when using a genetics based plan compared to 25% when using a traditional plan.⁸

- c) The flock is placed under surveillance for five years.

4. Interstate movement of scrapie-exposed animals. The use of genetic testing will be used to determine which scrapie-exposed animals can move in interstate commerce. All states are now able to use flock cleanup plans based on genetic testing. The plans will allow owners to retain or sell exposed animals from infected or source flocks without restriction if they have met certain criteria and if genetic testing conforms that these animals are scrapie resistant. The plan calls for all sexually intact sheep not being moved directly to slaughter to be genotyped for scrapie resistance. Genetically susceptible exposed female animals and, in rare cases, genetically less susceptible exposed female sheep will be removed under indemnity or permanently restricted to the premises. All animals in the flock will be officially identified and entered in the scrapie national generic database. Animals that are retained will have their genotype confirmed and the genetically susceptible exposed animals and the genetically less susceptible exposed sheep must be identified with a microchip electronic identification device.

To comply with conditions of the genetics-based cleanup, a post exposure management and monitoring plan will be required. The monitoring plan requires the following: 1) official identification of sexually intact animals that are sold or acquired 2) A record of any persons from whom sexually intact animals are acquired or to whom they are sold. 3) Reporting of any deaths of mature animals and animals showing clinical signs of scrapie and 4) annual inspections. All female genetically susceptible exposed animals, all those who test positive and

the female offspring of positive animals must be removed from the flock. Flocks that remove all susceptible female animals will not be considered exposed flocks once they have completed the flock cleanup plan. Flocks not removing all susceptible female animals will still be considered exposed. Accordingly, until the monitoring plan is completed, these flocks will have restrictions placed on susceptible animals in the flock and such animals born or brought into the flock.¹³

4. CONTROL PROGRAM STATUS

All 50 states have been determined to have active scrapie control programs and are considered consistent states.⁷ As of November 30, 2003, there were 1,830 flocks participating in the Scrapie Flock Certification Program (SFCP). Of these flocks 110 were certified flocks. 1,713 are complete monitored, and 7 are selective monitored flocks.

As of November 30, 2003 there were 59 scrapie infected and source flocks. There were 5 newly infected flocks reported in November 2003 with a total of 10 reported for fiscal year 2004. The total infected and source flocks released in 2004 was eight. Seven scrapie cases have been confirmed and reported by the National Veterinary Services Laboratories. Twelve cases of scrapie in goats have been reported since 1990. No new goat cases have been reported since fiscal year 2002.

As of November 30, 2003, the Center for Epidemiology and Animal Health (CEAH) has released the first results of the Scrapie Ovine Slaughter Surveillance (SOSS) The objective of SOSS was to estimate the national and regional prevalence of scrapie in mature cull ewes. Prior to this study the prevalence of scrapie in the United States was estimated to be 0.07%. The SOSS study estimate is 0.20%. The prevalence phase of the SOSS study started in April 2002 and continued through March 2003. During this time period samples were collected from 12,508 mature sheep at 22 slaughter facilities as well as a major livestock market. Of those tested 33 were found to be scrapie positive. Of these there were 27 black face, three mottled face, one white face and two unknown face color. A complete report and analysis will be available in January 2004.

Regulatory Scrapie Slaughter Surveillance (RSSS) is a targeted slaughter surveillance designed to identify infected flocks for clean up. Samples have been collected from 7,441 sheep since April 1, 2003, of which, results for 5,658 are reported. Samples have been submitted from 21 plants. There have been 20 positive or suspect sheep of which three were white face and 17 black face. During November 2003, 1058 sheep were tested by NVSL and 2 new positives were reported.¹⁴

During fiscal year 2003 a total of 16,803 animals were sampled or tested for scrapie. This included 3,724 regular field necropsy cases, 42 third eyelid biopsies for the test validation project, 244 necropsy test validations, 579 third eyelid biopsies for the regulatory program, and approximately 12,214 animals for SOSS and RSSS.¹⁵

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