National Scrapie Surveillance Plan

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# Table of Contents

- Executive Summary .................................................................................................................. 3
- 1. Disease Description ................................................................................................................. 5
- 2. Purpose and Rationale for Surveillance .................................................................................... 10
- 3. Surveillance Objectives ........................................................................................................... 12
- 4. Expected Outcomes ................................................................................................................. 13
- 5. Stakeholders and Responsible Parties ..................................................................................... 13
- 6. Population Description and Characteristics ............................................................................. 14
- 7. Case Definition ......................................................................................................................... 16
- 8. U.S. Surveillance for Scrapie: National Scrapie Eradication Program (NSEP) ....................... 18
- 9. Data Presentation and Reports .................................................................................................. 29
- 10. Surveillance System Implementation ....................................................................................... 30
- 11. Surveillance Plan Performance Metrics .................................................................................. 31
- 12. Surveillance System Evaluation ............................................................................................... 32
- References ..................................................................................................................................... 32
- Acronyms Used In This Document .............................................................................................. 37
Executive Summary

Scrapie is a transmissible spongiform encephalopathy (TSE) affecting sheep and goats. Classical scrapie is transmitted to other sheep or goats under natural conditions primarily through exposure to the birth tissues and fluids of an infected animal. In contrast, Nor98-like scrapie is thought to occur spontaneously in a small proportion of older sheep and goats. Nor-98-like scrapie is believed to be either not transmitted or rarely transmitted under natural conditions.

The presence of classical scrapie in the U.S. sheep and goat population affects industry economically through production losses, lost exports, and increased production and disposal costs. Potential public health concerns related to the transmission of bovine spongiform encephalopathy (BSE) to humans have resulted in efforts to eradicate all TSEs in food-producing animals.

Surveillance for scrapie in the United States is conducted through the National Scrapie Eradication Program (NSEP), a cooperative State-Federal-industry program. The current surveillance components of the NSEP include:

1. Regulatory Scrapie Slaughter Surveillance (RSSS);
2. Non-slaughter surveillance (e.g., trace investigations, on-farm testing); and
3. The Scrapie Flock Certification Program (SFCP).

The program’s goals are to eradicate classical scrapie in the United States and to meet World Organization for Animal Health (OIE) criteria for disease freedom. Since 2002, the prevalence of scrapie has decreased significantly through existing eradication efforts, largely a result of effective slaughter surveillance.

In order to achieve the goal of eradication, efforts must focus on improving the flock-level sensitivity and increasing surveillance to find the remaining cases. This will be accomplished by sampling apparently healthy and clinical sheep and goats at slaughter and enhancing on-farm surveillance efforts, including specifically targeting underrepresented flocks/herds and geographic regions. This effort will require enhancing the following surveillance activities:

1. Passive observation and reporting, by enhancing disease awareness of producers and veterinarians;
2. Laboratory surveillance, by ensuring appropriate samples from targeted and clinical animals are forwarded to Veterinary Services’ National Veterinary Services Laboratories (NVSL) or an approved contract laboratory;
3. Active surveillance, by expanding existing slaughter surveillance to include new collection sites, enhancing on-farm surveillance, conducting trace investigations, and increased testing of mature animals that die in SFCP participating flocks;
4. Focusing efforts to reach under-sampled flocks and geographic areas; and
5. Increased compliance with identification requirements.
VS regional and field staff will continue to work together to develop effective implementation plans to achieve State-level targets.

In the past 3 years, surveillance initiatives were implemented to address existing gaps in surveillance. These included surveillance to establish the prevalence of scrapie in goats sent to slaughter, a spatial evaluation to improve representativeness of surveillance sampling, and surveillance of cull ewes likely to be exported. A new initiative implemented in FY 2009 included limiting sampling from animals whose official identification tags could be linked to a flock that had been excessively sampled in the previous 36 months through RSSS. Evaluation of historical data to identify sampling requirements to establish a flock as low risk is underway; upon completion, this will allow refinement of sampling protocols and flock certification programs. These efforts help reduce unnecessary sampling from flocks, while still retaining confidence in flock disease status, and allow redirection of funds to the under-sampled flocks and regions.

This surveillance plan focuses on the eradication of classical scrapie from the U.S. sheep and goat population. Surveillance for classical scrapie has led to incidental discovery of Nor98-like scrapie in a small number of sheep in the United States. In line with the OIE 2009 policy change on Nor98 and Nor98-like scrapie in which OIE determined it to be a distinct disease, APHIS revised its policy on Nor98-like scrapie by removing the requirement to depopulate or permanently restrict Nor98-like scrapie exposed sheep and goats.

Currently, surveillance efforts are focused on sampling higher risk populations such as black-faced sheep and meeting State-level sampling targets. As the number of cases in the United States continues to decline, efforts will shift appropriately to group States regionally and to establish region-level targets. Once no case has been identified for a period of 7 years, there will be a shift toward meeting a national-level goal and high-level monitoring that will rely less on slaughter-based surveillance and more on passive reporting and investigation of suspect cases.
1. Disease Description

Scrapie is a progressive disease affecting the central nervous system (CNS) of sheep and goats and belongs to a group of diseases called transmissible spongiform encephalopathies (TSEs).

A. Etiologic Agent

The agent responsible for scrapie and other TSEs is smaller than the smallest known virus and has not been completely characterized. There are a variety of theories regarding the nature of the agent. The most widely accepted is that disease is caused by an infectious protein, or prion, that causes the normal cellular version of the protein to change shape such that it can no longer be degraded by the cell, causing the protein to accumulate and damage the cell. The agent is extremely resistant to heat and to normal sterilization processes and does not evoke any detectable immune response or inflammatory reaction in sheep and goats.

Scrapie cases are classified as either classical or Nor98-like scrapie. Classical scrapie and Nor98-like scrapie differ in immunohistochemistry (IHC) staining pattern, Western blot (WB) band pattern, genetic susceptibility, and epidemiology. Classical scrapie isolates have been further classified into “strains” by various researchers based on differences in the incubation period, brain pathology, clinical manifestations, interspecies transmission capability, and biochemical characteristics (Morales et al. 2007). It remains unclear whether the grouping of classical scrapie isolates into the different strains based on these characteristics is significant for disease eradication. Another way to group scrapie isolates or cases into types is by their epidemiology in sheep and goats. Only two epidemiologically distinct types of classical scrapie are known to exist in the United States: valine-dependent and valine-independent. The more prevalent type is valine-independent scrapie. When a classical scrapie case is detected, the United States Department of Agriculture (USDA) Animal and Plant Health Inspection Service (APHIS) Veterinary Services (VS) uses the genotype of the positive sheep associated with an outbreak to determine regulatory action; this is because genotype is an indicator of the epidemiological type rather than strain classification. When discussing classical scrapie, this document will refer only to those two epidemiological types of scrapie that have been observed in the United States.

B. History and Distribution

Scrapie is the oldest known TSE. It was first recognized in sheep in Great Britain and other countries of Western Europe over 250 years ago. Scrapie has been reported in sheep throughout most of the world, affecting many sheep-producing regions. Naturally occurring scrapie has also been reported in goats in the United States, Canada, Cyprus, Finland, France, Greece, Italy, Switzerland, and the United
Kingdom. Only Australia and New Zealand are recognized by the United States as being free of classical scrapie.

Scrapie was first discovered in the United States in 1947 in a flock that had imported sheep of British origin from Canada. Since 1952, VS has worked to control and eradicate scrapie in the United States. In the United States, scrapie has primarily been reported in the Suffolk breed, although it has also been identified in a number of other breeds and in crossbreeds (Wineland et al. 1998; USDA, unpublished data). The national prevalence of scrapie in sheep in 2003 was estimated to be 0.2 percent (National Animal Health Monitoring System, 2004). Since that time, the national prevalence in sheep has decreased to an estimated 0.05 percent in FY 2009, primarily through active slaughter-based surveillance (USDA, unpublished data).

VS conducted a study from May 2007 through March 2008 to estimate the prevalence of scrapie in goats in the United States. VS collected samples from mature goats with potentially higher likelihood of infection, (i.e., those residing in States where scrapie-infected flocks or herds originated), those processed in slaughter establishments where scrapie has previously been detected or those that process animals from States with infected flocks or herds, and those processed through smaller slaughter establishments likely to slaughter goats from local populations. The study samples were combined with samples collected through RSSS, and the prevalence of classical scrapie in goats was determined to be less than 0.1 percent (USDA, unpublished data).

C. Epidemiology

Under natural conditions, only sheep and goats are known to be affected by scrapie. However, experimentally, classical scrapie can be transmitted to other ruminants, primates, cats, and a variety of rodents. Once infected, the animal remains infected for life. The disease is always fatal; however, it is common for infected animals to be slaughtered or to die first of other diseases or trauma. The scrapie agent may be found in some lymphoid tissues by the age of 4 months and in the brain by 2.5 years, approximately 6 months before the onset of clinical signs (Detwiler and Baylis, 2003).

Transmission of the classical scrapie agent is not completely understood, and apparently healthy sheep infected with the agent can transmit disease. Susceptibility to infection and the incubation period in sheep have been shown to vary according to sheep genetics and breed (Baylis and Goldman 2004; Baylis et al. 2002; Belt et al. 1995; Hunter et al. 1996; Hunter et al. 1997). The long incubation period between exposure and clinical disease allows animals to shed the agent for an extended period. The scrapie agent is thought to spread most commonly from the ewe to her offspring and other lambs through contact with the placenta and placental fluids. Sheep and goats are typically infected as young lambs or kids. Placental infectivity occurs in the
incubation/preclinical stage of disease, but is not constant with every pregnancy. This is likely the result of placental genotype and stage of disease. Genetically susceptible lambs born to dams that develop clinical scrapie have a higher risk of developing the disease. Ram genetics will contribute to scrapie susceptibility in their offspring.

Oral exposure likely accounts for most scrapie cases; however, infection may also occur via ocular exposure or contact with abraded skin or mucous membranes (Detwiler and Baylis 2003). Transmission to lambs through milk from infected ewes has been reported, as well as subsequent horizontal transmission among lambs (Konold et al. 2008). Other infectious tissues have also been found, including: central nervous tissue, lymphoid tissue, peripheral nerves, blood, muscle, liver, nasal mucosa, and salivary glands (Detwiler and Baylis 2003). Infected rams are not known to transmit scrapie.

Environmental contamination with the disease agent also likely plays a role in the recurrence of scrapie and the introduction of scrapie into new flocks. Scrapie has recurred on farms after all exposed animals are culled and new stock introduced; studies show the agent may survive in the environment for years, with the longest reported duration at least 16 years (Georgsson et al. 2006).

Risk factors for introduction and maintenance of scrapie in a flock are related to movement of animals, flock size, breeding practices, and lambing management. Specifically, these include: purchasing infected breeding animals, sharing pastures, breeding and raising home-bred replacements, ewes lambing in group pens (vs. unconfined on pasture or in individual pens), and the number/proportion of genetically susceptible sheep within a flock (Detwiler and Baylis 2003).

**Genetics**
For classical scrapie found in the United States, the codons at positions 136 and 171 in the gene that codes for amino acids in the prion protein (PrP) have been associated with scrapie susceptibility. Codon 171 is thought to be the major determinant of susceptibility, with glutamine (Q) and histidine (H) conferring susceptibility and arginine (R) resistance. The effect of lysine (K) at codon 171 on scrapie susceptibility is unknown due to its infrequent occurrence. Codon 136 affects susceptibility to the less common valine-dependent classical scrapie, with alanine (A) and valine (V) conferring resistance and susceptibility, respectively.

All QQ sheep are susceptible to the more common valine-independent classical scrapie and can transmit the disease to susceptible flock mates. Conversely, AARR sheep are nearly completely resistant to classical scrapie. These sheep are highly unlikely to carry or transmit scrapie. AAQR are rarely infected, and it is unknown whether infected AAQR sheep can transmit scrapie.

Like the valine-independent scrapie, there is a genetic difference in an animal’s susceptibility to the valine-dependent scrapie. AVQQ, VVQQ, and AVQR sheep are
susceptible to valine-dependent scrapie. AAQQ and AAQR, sheep appear to be resistant to valine-dependent scrapie. AARR sheep appear to be resistant to both valine-dependent and valine-independent scrapie (Evoniuk et al. 2007).

Dam-offspring transmission likely occurs perinatally. While transplacental infection could occur, it has never been demonstrated. Placental infectivity is not constant with every pregnancy, occurs during the incubation/preclinical and clinical stages of disease, and requires a QQ fetus or an adjacent QQ fetus for scrapie prion protein (PrPsc) accumulation (Androletti et al. 2002; Tuo et al. 2002; Alverson et al. 2006).

At present, there is an insufficient understanding of genetically-based scrapie resistance in goats to reliably assign goat risk categorically based on genetics. Therefore, all goats are considered to be genetically susceptible for program purposes.

**Nonclassical scrapie**

Nonclassical scrapie includes those cases of scrapie reported in the literature as the Nor98, Nor98-like, or “atypical” scrapie. Nonclassical scrapie was first detected in Norway in 1998 and was designated Nor98. With increased surveillance, other countries have since detected nonclassical scrapie. Nor98-like scrapie has been detected in the United States since 2006.

Nonclassical scrapie differs pathologically and biochemically from classical scrapie and can be differentiated from classical scrapie and BSE using histopathologic patterns of lesions in the lymphoreticular and nervous systems, WB, and IHC. Clinical signs in nonclassical scrapie cases are often absent; when present, the signs are indistinguishable from those of classical scrapie, although pruritus has not been reported in nonclassical cases.

In addition to the pathologic and biochemical differences, there are epidemiological differences between classical and nonclassical scrapie (Benestad et al. 2008; Lühken et al. 2007). In the European Union (EU), where most of the nonclassical cases have been detected, additional positive animals were found infrequently when exposed animals in the flock of origin were euthanized and tested. Additional cases were found to be associated with flocks larger than 500 sheep, and it was uncommon to detect more than one additional case per flock. In contrast, more than 10 percent of genetically susceptible animals were identified as infected following the depopulation of flocks infected with classical scrapie. Nonclassical scrapie cases have been reported to be more widely distributed geographically and to affect mostly sheep older than 5 years, whereas classical scrapie cases were somewhat geographically limited and generally affected sheep between 2 and 4 years of age. There is some evidence that nonclassical scrapie may occur through a spontaneous process – possibly not transmissible under natural conditions or not transmissible at a rate that could be sustained naturally (Fediaevsky et al. 2010; Lühken et al. 2007; McIntyre et al. 2008).
At this time, it is unknown whether infectivity accumulates in the placenta of nonclassical scrapie-infected animals.

**Genetics**

PrP genotypes phenylalanine (F) at codon 141 or H at codon 154 are associated with nonclassical scrapie cases. In addition, sheep with QR or RR at codon 171, known to be less susceptible or resistant to classical scrapie, have been affected by nonclassical scrapie.

**D. Clinical Signs**

Clinical signs of classical scrapie typically appear between 2 to 5 years after infection; therefore, infected animals rarely show clinical signs of infection before the age of 2 years, with the average age of clinical onset being 3-4 years. Nonclassical scrapie may appear with similar clinical signs, although the age of onset has generally been older sheep (older than 5 years). The prolonged incubation period, the subclinical nature of the infection during its early stages, and the fact that the only diagnostic tests currently available require brain or lymphoid tissue make detection of scrapie difficult. Sheep typically live 1 to 6 months after the onset of clinical signs, but some will die earlier or later. Duration of clinical signs may depend on the observational abilities of the producer. Some sheep may simply be found dead.

On the farm, veterinarians suspect scrapie based on the clinical signs combined with knowledge of the animal’s signalment and history. However, the clinical presentation of scrapie includes a wide range of nonspecific signs that develop very slowly. Due to damage to nerve cells, affected animals often show behavior changes, such as nervousness or aggression, intense rubbing, and locomotor incoordination that progresses to recumbency and death. Other clinical signs may include tremors (especially of head and neck), head pressing or “star gazing,” significant weight loss with no decrease in appetite, wool pulling, and hyperesthesia. Additional signs in affected goats may include difficulty milking, premature kidding, and pica.

**E. Control and Prevention**

The best method for preventing scrapie from occurring in a flock or herd is to maintain a closed flock/herd, particularly with regard to breeding females. Any replacement females or breeding males should originate from flocks/herds not known to be affected with scrapie and under management practices precluding the introduction of scrapie. In the case of sheep, replacement females or breeding males should be of resistant PrP genotypes. Susceptible ewes of unknown or questionable disease status should be bred to RR rams or separated from the rest of the flock prior to and following lambing until there is no vaginal discharge to minimize spread to other animals. Another method used by some producers is selective breeding to reduce overall flock susceptibility based on PrP genotype. This method consists of breeding only with rams that are RR or QR.
Once an infected animal is detected, eradication of the disease from the flock or herd may consist of either selective depopulation of certain higher-risk exposed animals (e.g., only those that are genetically susceptible, heavily exposed, test positive or inconclusive, and/or showing clinical signs) or, less commonly, complete flock depopulation, as well as cleaning and disinfection of the premises. Owners may opt to restock with rams that are RR and ewes of resistant genotype.

The use of selective breeding and culling to increase genetic resistance to scrapie infection raises concern regarding the practices’ effect on the genetic diversity of the domestic sheep population and on production traits. A number of studies have been completed evaluating effect of PrP genotype selection and production traits (e.g., meat, milk production, litter sizes), with some studies providing limited evidence of associations between PrP genotype and traits (e.g., Isler et al. 2006; Man et al. 2007; Sawalha et al. 2007) but not all (Alvarez et al. 2006; Alexander et al. 2006; de Vries et al. 2004; Sweeney et al. 2007). Overall, when observed, associations between PrP genotype and performance traits tended to be neither strong nor consistent across populations, and there was no tendency for associations between scrapie-resistant PrP alleles and performance traits to be adverse (Dawson et al. 2008; Sweeney and Hanrahan 2008). A study did find that producer perception of animal quality (based on the physical characteristics of hardiness, wool quality, conformation, and body size) was not influenced by animal susceptibility to scrapie (as determined by PrP genotype). In other words, farmers were not able to predict the genotype of their sheep based on their performance, and farmers’ assessments of their best-performing animals were not biased toward scrapie-susceptible genotypes (Nicholls et al. 2006).

2. Purpose and Rationale for Surveillance

The purpose of the National Scrapie Surveillance Plan is to meet the goals of the National Scrapie Eradication Program (NSEP), which are:

1. To eradicate classical scrapie from the sheep and goat population in the United States, then
2. To document the eradication of classical scrapie, and
3. To achieve scrapie-free status, as described by the World Organization for Animal Health (OIE), in the United States. Currently, the OIE requires that a country or region be able to provide, for at least 7 years, 95 percent confidence of detecting scrapie at a prevalence of 0.1 percent of the target population and that no case of scrapie has been reported during this period (OIE 2009).

This surveillance plan focuses on the eradication of classical scrapie. Nor98-like scrapie has its own unique epidemiology, and in 2009 the World Animal Health Organization (OIE) concluded that it is a separate disease from classical scrapie. Because it is suspected that nonclassical scrapie occurs through a spontaneous
process and either is not transmitted or is transmitted at an unsustainable rate in natural conditions, it is believed that eradication of nonclassical scrapie from the United States is neither necessary nor feasible.

The rationale for conducting surveillance and scrapie eradication are as follows:

**Economic Impact on Industry:** Scrapie is a non-febrile and insidious disease. Infected flocks with a high percentage of susceptible animals can experience significant production losses. Over several years, the number of infected animals in a flock increases and onset of clinical signs occurs in younger animals, making these flocks economically unviable. Female animals sold from infected flocks can spread scrapie to other flocks. The presence of scrapie in the United States is estimated to cost American sheep producers $10-20 million per year, principally in lost exports of sheep products and breeding stock, semen and embryos; decreased value of and, in some cases, increased expenditures for offal and carcass disposal; and increased production costs. It has been estimated that the eradication of scrapie would offer U.S. producers an increase in revenue of at least $10.8 million annually, particularly in export markets (Seitzinger, Paarlberg and Lee 2006).

**Potential Public Health Concerns:** The apparent transmission of bovine spongiform encephalopathy (BSE), another TSE, to humans in the United Kingdom has resulted in a call for the eradication of all TSEs in food-producing animals. In 2005, BSE in a goat was confirmed at the Community Reference Laboratory in Weybridge. The goat was slaughtered in 2002 in France and was tested as part of a slaughter surveillance program. An epidemiologic investigation conducted at the time of the initial TSE diagnosis did not detect any additional cases in the herd (Eloit et al. 2005; OIE 2009). There have been no other naturally occurring cases of BSE reported in ovines or caprines. Previous research has demonstrated that BSE could be successfully transmitted to sheep and goats orally, and that sheep genotypes traditionally resistant to scrapie were susceptible to BSE (Foster et al. 1993; Foster et al. 1994; Foster et al. 2001). This has resulted in increased public concern. There is no scientific evidence to indicate that scrapie poses a risk to human health or that scrapie of sheep and goats is transmitted to humans. Because BSE in sheep and goats is detected using the same tests as scrapie, eradication of scrapie would protect human health from the risk of BSE in sheep or goats, should it occur, by eliminating the risk of BSE being masked by scrapie. Also, the surveillance conducted to facilitate scrapie eradication and to document freedom from scrapie following eradication will allow earlier detection of BSE in sheep or goats should it occur. This would increase consumer confidence both domestically and internationally.

This scrapie surveillance plan for the United States calls for transitioning to demographic-based surveillance, and ultimately to flock-level surveillance, at a critical period in scrapie eradication efforts. The United States has been involved in scrapie control efforts for more than 50 years. Since 2002, the prevalence of scrapie has decreased significantly through existing eradication efforts, largely a result of
effective slaughter surveillance. In order to achieve the goal of eradication, efforts must focus on improving flock-level sensitivity and increasing surveillance efforts to find the remaining cases.

3. Surveillance Objectives

In general, scrapie surveillance is conducted to:
1. Detect infected sheep and goats,
2. Trace infected animals to their flocks or herds of origin, and
3. Locate exposed and potentially exposed animals for testing and monitoring.

This will be done in three stages: eradication of the disease in the U.S. sheep and goat population by finding the remaining cases (expected timeline: FY 2008-2016), high-level monitoring to ensure that no cases remain (FY 2017-2020), and ongoing monitoring to meet OIE requirements (FY 2021 and beyond). This surveillance plan document addresses the first stage (disease eradication by detecting remaining cases). This will be accomplished by increasing sampling of apparently healthy and clinical sheep and goats from underrepresented flocks and herds and underrepresented geographic regions that are being missed through current surveillance efforts.

Scrapie eradication will be accomplished by increasing efforts in the following areas:

1. **Passive observation and reporting**: This effort requires enhancing awareness of the disease and associated clinical signs through educational programs/materials. The objective is to increase the sensitivity of producers and veterinarians to detecting clinical cases in the field and submitting appropriate samples to diagnostic laboratories.

2. **Laboratory surveillance**: This requires ensuring that diagnostic laboratories forward appropriate tissues from all mature sheep and goats presented for necropsy and diagnostic testing to NVSL or an approved contract laboratory for scrapie testing, regardless of whether another diagnosis is made. Additionally, this requires ensuring that public health laboratories forward appropriate tissues from mature sheep and goats that test negative for rabies to the NVSL or an approved contract laboratory for scrapie testing.

3. **Active surveillance**: This will be enhanced using the following seven strategies:
   a. Expansion of existing slaughter surveillance sampling into additional federally- and State-inspected plants, custom and ethnic slaughter facilities, and other concentration sites where targeted sheep and goats may be found, such as markets, cull ewe feedlots and exporters.
   b. Introduction and possible future expansion of a “Do Not Collect List” that removes over-sampled flocks from surveillance. This will allow
more efficient use of limited resources for activities such as targeting under-sampled flocks and geographic areas.
c. Continued use and refinement of targeting criteria for sampling high-risk animals. This requires annual evaluation to insure that these high-risk animals are being sampled.
d. Fill in surveillance gaps through enhanced on-farm surveillance in targeted geographic areas and/or flocks.
e. Expansion of active surveillance to include apparently healthy goats.
f. Continued active trace investigations of exposed animals and potentially exposed flocks that are initiated upon identification of an infected animal.
g. Incorporation of the SFCP program participants. SFCP flocks that may not be tested through slaughter channels will also be incorporated, as participating flocks are required to be inspected annually by an official animal health representative or accredited veterinarian. SFCP flocks are also required to follow reporting requirements for scrapie-suspect animals, animals suspected of other neurologic and chronic debilitation (prolonged wasting) illnesses, and any mature animals found dead.

4. Expected Outcomes

Detection of infected sheep will result in actions that promote eradication of the disease agent; these actions are described in the Scrapie Eradication Uniform Methods and Rules (UM&R). The expected outcome of the comprehensive surveillance program outlined in this document is the progressive reduction of scrapie prevalence, resulting in the eradication of disease in the U.S. sheep and goat populations. The data and information generated from the surveillance program will also inform decision-makers about future surveillance needs and trade-related issues.

5. Stakeholders and Responsible Parties

- National Surveillance Unit (NSU): Surveillance planning and evaluation
- National Center for Animal Health Programs (NCAHP), VS regional staff and NVSL: Surveillance planning, implementation, and oversight; training; communication;
- Regional staff, VS Area Veterinarians-in-Charge (AVICs), State animal health authorities: Surveillance implementation and communication with local producers and industry;
- VS Office of the Chief Information Officer (OCIO): Development, training, deployment, and maintenance of scrapie Veterinary Services Laboratory Submission (VSLS) and Mobile Information Management (MIM) modules, including integration of Animal Identification Management (AIM) system, scrapie mapping module and the Animal Health and Surveillance
Management (AHSM) System to allow rapid data analysis at the flock level; and
- Sheep and goat organizations: communication with producers and industry, including:
  - American Sheep Industry Association (ASI)
  - American Goat Federation
  - American Goat Society
  - American Meat Goat Association
  - American Dairy Goat Association
  - American Boer Goat Association
  - U.S. Boer Goat Association
  - National Institute for Animal Agriculture (NIAA)

6. Population Description and Characteristics

Sheep and goat production occurs throughout the United States, but the top sheep-producing states are Texas, California, Wyoming, Colorado, and South Dakota (Table 1; NASS Sheep and Goats, January 2010). In Texas and other Western States, sheep production is largely in range flocks that are grazed on large, open rangeland. In the East, sheep production tends to be in concentrated farm settings. Nationally, 28.1 percent of operations have Suffolk sheep comprising the majority of their flocks. Suffolk is the breed with the most reported scrapie cases in the United States.

The value of the sheep industry is related to the two major uses for sheep: meat production (lamb) and pelts and wool production, with meat production being the primary source of income. The total commercial slaughter of sheep and lambs was 2.52 million head in 2009, with approximately 93.2 percent of that slaughter being lambs and yearlings (NASS Livestock Slaughter, April 2010). Colorado slaughtered the largest volume of sheep (Table 2). In 2009, 92.3 percent of commercial lamb and sheep slaughter was federally inspected. The total U.S. wool production in 2009 was 30.9 million pounds with a total value of $24.4 million (NASS Sheep and Goats, January 2010).

The U.S. sheep inventory on January 1, 2010, was 5.63 million head, with 4.19 million head breeding stock. The breeding stock includes over 3.34 million ewes and 195,500 rams 1 year of age or older and 655,000 replacement lambs (NASS Sheep and Goats, January 2010). There were a total of 82,000 sheep operations in the United States in 2007. Most sheep operations (93.7 percent) had less than 100 head, and these operations accounted for 36.2 percent of the U.S. sheep inventory. Only 1.1 percent of the operations had over 500 head, but these operations accounted for 43 percent of the U.S. inventory (NASS Farms, Land in Farms and Livestock Operations, February 2010). In 2000, approximately 16.6 percent of the ewes died or
were culled. Of those ewes, 10.4 percent were culled or dead with progressive weight loss, despite a normal appetite and no respiratory problems (NAHMS 2001).

The two major uses for goats are meat production and mohair production. The total U.S. mohair production in 2009 was 1.01 million pounds from 160,500 goats (total value of $2.692 million). On January 1, 2010, there were approximately 2.53 million breeding goats, 518,000 market goats and kids, 2.54 million head of meat and other goats, 355,000 milk goats, 150,000 angora goats, and 1.94 million kids (NASS Sheep and Goats, January 2010). In 2009, over 659,290 goats were commercially slaughtered, with the largest volume slaughtered in New Jersey (Table 2; NASS Livestock Slaughter, April 2010). There were a total of 152,000 goat operations in
the United States in 2009; these included 30,000 milk goat operations and 130,000 meat goat operations (NASS Farms, Land in Farms and Livestock Operations, February 2010).

The United States is not a major exporter of live sheep, historically accounting for less than 1 percent of the total world trade in live sheep. In 2009, the U.S. exported 157,337 mature sheep, the majority to Mexico (78,754), but also to Canada, Trinidad, and Tobago. In addition, the United States exported 8,820 goats, again the majority to Mexico (4,072), but also to Canada, Trinidad and Tobago, the Cayman Islands, Germany, Panama, and Suriname (Global Trade Atlas, accessed June 28, 2010).

7. Case Definition

For the purpose of classical scrapie surveillance and disease eradication, only those sheep and goats tested using an official test that are confirmed positive for classical scrapie by NVSL (or a laboratory to which the NVSL has referred a case for such testing) are designated as classical scrapie cases. The identification of suspect cases leads to further investigation, but these cases are not considered to be classical scrapie cases until confirmatory test results have been reported.

Case Description
Sheep and goats of many breeds have been affected. Most cases of clinical classical scrapie in sheep occur when the animal is between 2 and 5 years of age. Although rare, clinical signs may arise in sheep under 1 year. In some instances, the commercial life span of sheep and goats may be too short to allow clinical signs to develop. Progression of the clinical disease is variable, from weeks to months, with a fatal outcome. Since signs may not appear until months or years after transmission to other sheep or goats in the flock, relying on clinical signs to detect cases is not ideal. Therefore, the primary focus of scrapie surveillance is the identification of subclinical classical scrapie cases.

Clinical Description
Some sheep and goats infected with the scrapie agent may not develop clinical signs before death or culling. Clinical disease only develops when the infection enters the CNS. Due to the influence of host genotype and scrapie agent type, clinical signs vary among individual animals. In general, due to damage to nerve cells, affected animals often show behavior changes, such as nervousness or aggression, rubbing, and locomotor incoordination, that progress to recumbency and death. Other clinical signs may include tremors (especially of head and neck), head pressing or “star gazing,” significant weight loss with no decreased appetite, wool pulling, and hyperesthesia. Additional signs in affected goats may include difficulty milking, premature kidding, and pica. Because of the variability in clinical presentation, clinical diagnosis of scrapie can be difficult.
Epidemiologic criteria and restrictions
The case definition pertains to all captive and/or domestic sheep and goat populations in the United States. Animals of the genus *Ovis* are considered to be sheep, and animals of the genus *Capra* are considered to be goats.

Laboratory criteria
Infection with the scrapie agent is determined by the detection of the abnormal prion protein accumulation in nervous tissue and/or lymphoreticular tissues and/or histopathologic lesions in central nervous tissue in susceptible species. The abnormal prion protein can be detected by the use of an approved screening enzyme-linked immunosorbent assay (ELISA), WB, and/or by performing IHC on CNS and/or lymphoid tissues (i.e., third eyelid or anorectal lymphoid tissue, tonsil or lymph node, obex, cerebellum or other brain tissue). The characteristic histopathologic change of nervous tissue is vacuolation of neurons, producing a distinctive appearance of spongiform change. The vacuolar changes may be accompanied by other microscopic features, such as neuronal degeneration, neuronal loss, gliosis, and cerebrovascular amyloidosis. Typically, the histopathologic lesions have bilaterally symmetrical distribution, although the distribution pattern and changes may vary between type of agent and host genetics. The scrapie type may be further characterized as classical or nonclassical scrapie by performing IHC and/or WB assay. All cases that produce positive, suggestive, or inconclusive results or that show any unusual staining when initially tested by an approved laboratory are submitted to the NVSL for further evaluation. Confirmatory testing may include any of the following methods, used alone or in combination: IHC, WB, histopathology, enzyme immunosorbent assay (EIA), ELISA, or animal inoculation studies (i.e., bioassay). A case must be confirmed positive by NVSL or a laboratory to which the NVSL has referred a case for such testing to be designated a scrapie case.

Case Classification
*Suspect*
A sheep or goat meeting *at least one* of the following criteria is considered a scrapie suspect:
1. Has been condemned by the Food Safety and Inspection Service (FSIS) or a State inspection authority for CNS signs or rabies, or
2. Exhibits any of the clinical signs compatible with scrapie and has been determined to be suspicious for scrapie by an accredited veterinarian or a State or USDA representative. Compatible clinical signs may include, but are not limited to:
   • Weakness of any kind, not including those with visible traumatic injuries and no other signs of scrapie. Signs of weakness may include:
     ▪ stumbling,
     ▪ falling down, or
     ▪ having difficulty rising;
• Significant weight loss, despite retention of appetite in an animal with adequate dentition;
• Increased sensitivity to noise and sudden movement;
• Tremors;
• Star gazing;
• Head pressing;
• Bilateral gait abnormalities, not including abnormalities involving only one leg or one front and one back leg. Signs of gait abnormalities may include:
  ▪ incoordination,
  ▪ ataxia,
  ▪ high-stepping gait of forelimbs,
  ▪ bunny-hop movement of rear legs, or
  ▪ swaying of back end.
• Repeated intense rubbing with bare areas or damaged wool in similar locations on both sides of the body or, if on the head, both sides of the poll;
• Abraded, rough, thickened, or hyperpigmented areas of skin in areas of wool/hair loss in similar locations on both sides of the animal’s body or, if on the head, both sides of the poll; or
• Other signs of CNS disease;
3. Has a positive test result for scrapie or for a protease-resistant protein associated with scrapie on an unofficial test or screening test; or
4. Has a suspect, inconclusive, or suggestive test result on an official test.

**Confirmed**
Laboratory testing of a submitted sample(s) by the NVSL or a laboratory to which the NVSL has referred a case for such testing has returned positive result(s).

**Reporting Requirements**
Per 9 CFR parts 54, 79, and 161.3 and the Scrapie Eradication Uniform Methods and Rules (UM&R), all suspect and presumptive cases require reporting to State animal health officials and the VS AVIC for follow-up investigation and sampling.

8. U.S. Surveillance for Scrapie: National Scrapie Eradication Program (NSEP)

The NSEP is a cooperative State-Federal-industry program administered by APHIS and consistent States to control and eradicate scrapie. Its components consist of: producer/industry education and compliance; identification monitoring and compliance; slaughter- and nonslaughter-based surveillance; trace investigations; monitoring flocks for occurrence or recurrence of scrapie; flock cleanup through genetic testing and indemnification of susceptible exposed animals; and the SFCP.
**State-level Sampling Targets**

State-level sampling targets are established based on the population demographics of mature sheep and goats in each State. Population estimates were based on NASS Census of Agriculture information and annual sheep and goat farm information (2007 and 2009, respectively). The OIE requires demonstrating freedom from disease by adequate sampling of targeted sheep and goats for 95 percent confidence of detection at less than 0.1 percent prevalence nationally on an annual basis. Tables 3 and 4 present the statistically-based sampling target levels required for each state to detect scrapie at this level in their flock (Cannon 2001).

Since the OIE sampling level is designed to establish the presence or absence of disease and not for eradication, APHIS will continue to support sample collection of targeted sheep and goats until all States have not had a case in the preceding 5 years and have collected sufficient samples to detect scrapie at 0.1 percent prevalence with 95 percent confidence or 5 percent of each State’s breeding sheep population since their last case, whichever is less. As contiguous States meet sampling target criteria, they may be treated as a region for establishing sampling goals, which will significantly reduce sampling within the region. APHIS may also consider the percent of flocks sampled within a State when establishing lower risk regions. Once all States have met these criteria, total sampling nationally will be approximately 3,000 sheep and 3,000 goats annually to document ongoing freedom. After cases are no longer detected, surveillance will enter the 7-year monitoring period required by OIE to document disease freedom.

The following assumptions were made to calculate these minimum sample numbers:

1. The sampling of mature sheep at slaughter establishments is representative of the national sheep population. It is known that there are flock owners that home-slaughter or sell animals for noncommercial slaughter. There is insufficient knowledge of these practices within the United States to validate this assumption.
2. The prevalence of scrapie in the sheep sampled at participating slaughter establishments is the same as the prevalence in all of the mature sheep going to slaughter, including the non-traceable white or mottle-faced sheep not sampled at participating establishments.
3. All sheep in a State have an equal likelihood of contact with an infected animal, regardless of separations (e.g., fences, rivers) and geographic location and landscape. Limited information is available regarding distribution of sheep at a local level. NASS census data is most readily available at the State level. NASS records census data at the ZIP code level, but even at this level, there is limited ability to consider manmade and natural barriers of contact between populations within an area or State. This would require knowing the specific geographic location of all flocks in a State in order to use landscape and other geographic information systems (GIS) data and recording of all
barriers preventing contact between adjacent flocks (as well as recording of those flocks that may share pasture or otherwise have contact). Thus, based on the limited information available, we can only assume that all animals in a State have an equal likelihood of contacting a scrapie-infected animal in that State, that contact occurs at a time when the scrapie agent is being shed, and that the duration of contact is sufficient for transmission to occur.

Spatial analysis of scrapie surveillance data

The majority of sampling for surveillance will continue to occur through RSSS, supplemented by nonslaughter surveillance routes. Enhancement of current RSSS and nonslaughter surveillance efforts has been an ongoing and evolving process. Most recently, an analysis of scrapie surveillance data was conducted on samples from slaughtered sheep from the following States: Iowa, Illinois, Indiana, Ohio, Texas, and Wyoming. The purpose of the analysis was to identify geographic areas and flocks that are not being reached through current surveillance efforts. This utilized official identification tag information and the flocks listed in the scrapie national generic

<table>
<thead>
<tr>
<th>State (of Tag Origin)</th>
<th>Surveillance Sampling Target</th>
<th>State (of Tag Origin)</th>
<th>Surveillance Sampling Target</th>
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</table>
National Scrapie Surveillance Plan

September 2010

21

database (SNGD). This effort allowed visualization of sampling gaps on maps (at the ZIP code level), and illustrated that these findings of the analysis were similar across the States:

1. The ZIP codes where none of the listed flocks had been sampled for scrapie surveillance are those ZIP codes with fewer than three flocks;
2. The ZIP codes where none of the listed flocks had been sampled for scrapie surveillance are those ZIP codes with the smaller sheep populations; and
3. Most of the listed flocks that had never been sampled for scrapie surveillance are located in those ZIP codes with at least three flocks.

The findings of these analyses are likely applicable to the other States. Thus, for the remainder of the States, using scrapie surveillance data for FY 2003 through FY 2008, maps have been generated for each State identifying ZIP codes with listed flocks and where scrapie surveillance samples have and have not been collected from flocks. Identification of these ZIP codes and flocks will enable field personnel to increase surveillance efforts (e.g., recruit smaller slaughter establishments or establish on-farm surveillance) to increase the coverage of scrapie surveillance in the United States.

A. Regulatory Scrapie Slaughter Surveillance (RSSS)

Implemented in 2003, the RSSS is a targeted surveillance program consisting of sample collection from mature sheep and goats sent to participating cull ewe slaughter facilities, as well as dead, disabled, or suspect animals found at concentration points for mature ewes, including markets and cull ewe feedlots.

Sampling method:
The RSSS targets mature sheep and goats that meet specific criteria based on age, face color, and/or clinical signs at slaughter. A study found that among cull sheep entering slaughter in 2002-2003, the prevalence of scrapie in apparently healthy black- and mottled-faced sheep was greater than that in white-faced sheep (0.85 percent, 0.12 percent and less than 0.01 percent, respectively) and that black-faced sheep were 38 times more likely to be infected than white- or mottled-faced sheep (NAHMS 2004). Additionally, analysis of 2005 RSSS data showed that the majority of positive sheep sampled at slaughter were at least 2 years of age (NSU 2006). Therefore, sampling of the apparently healthy sheep at slaughter has primarily been targeted based on age (those at least 14 months of age) and face color (black- and mottled-faced). Since the implementation of RSSS, the apparent prevalence of scrapie in black- and mottled-faced sheep (those with more than 1 percent black on their face) at slaughter has steadily decreased to 0.18 percent and 0.05 percent, respectively, in FY 2009 (USDA, unpublished data).

In FY 2009 the targeting criteria was adjusted due to the decreasing prevalence in mottled-faced sheep and the failure to detect cases in white-
faced sheep at slaughter, despite white-faced cases being detected through testing of scrapie-exposed sheep. Analysis of RSSS data in FY 2008 showed that the apparent prevalence is higher in sheep 2, 3, or 4 years of age than in sheep 1 year of age or 5 years or older. Evaluation of nonslaughter surveillance data supports this finding. Currently, all white- and mottled-face sheep age 2, 3, 4, or 5 are being sampled through RSSS; white- and mottled-faced older sheep as evidenced by broken, missing, blunt, elongated, or splayed teeth; or other significant signs of dental wear are not sampled.

Analysis of RSSS data of mottled-faced sheep sampled at slaughter shows that the prevalence of scrapie in sheep with minimal mottling (i.e., less than 1 percent black on their face) is closer to that of white-faced sheep than to mottled-faced sheep with more black. Also, minimally mottled are considered white-face by producers. Therefore, apparently healthy mottled-faced sheep with minimal mottling will be treated the same as white-faced sheep for RSSS sampling and reporting.

In addition to the apparently healthy sheep sampled at slaughter for scrapie surveillance, all animals presenting with suspect clinical signs at slaughter are tested for scrapie under RSSS. Signs used to target clinical suspect animals are nonspecific (i.e., wool loss, rubbing, unthrifty appearance, weakness, non-ambulatory and/or other evidence of central nervous system disorder). Analysis of RSSS data of clinical sheep sampled at slaughter FY 2003-2007 showed very few of these animals with clinical signs identified as scrapie-positive, and there was insufficient data to support any change of targeting criteria for clinical suspect animals (USDA, unpublished data).

Based on these analyses, the target population for slaughter surveillance is mature sheep or goats, as evidenced by complete eruption of at least one permanent incisor (first permanent incisor is level with or extends above the remaining deciduous teeth). All mature black-faced sheep and clinical suspects should be sampled. All other mature sheep (i.e., not black-faced) and mature goats should only be sampled if they meet at least one of the following criteria:

1. Southdown or Montadale sheep;
2. Dead prior to slaughter or condemned on antemortem inspection;
3. Nonambulatory:
   a. Down and unable to rise; or
   b. Able to rise but only for brief periods of time.
   Nonambulatory animals can include those with broken appendages, severed tendons or ligaments, nerve paralysis, fractured vertebral columns, or metabolic conditions;
4. Signs of CNS disease, including:
a. Weakness of any kind, including stumbling, falling down, or having difficulty rising, not including those with visible traumatic injuries that should be recorded as nonambulatory animals; or
b. Behavioral abnormalities; or
c. Increased sensitivity to noise and sudden movement; or
d. Tremors; or
e. Star gazing; or
f. Head pressing; or
g. Bilateral gait abnormalities, including incoordination, ataxia, high stepping gait of forelimbs, bunny-hop movement of rear legs, or swaying of back end, but not including abnormalities involving only one leg or one front and one back leg; or
h. Other CNS signs.

5. Intense rubbing, abrasions, or rough, thickened, and/or hyperpigmented skin:
   a. Repeated intense rubbing with bare areas in similar locations on both sides of the animal’s body or, if on the head, both sides of the poll; or
   b. Abraded, rough, thickened, or hyperpigmented areas of skin in areas of wool/hair loss in similar locations on both sides of the animal’s body or, if on the head, both sides of the poll.

6. Scrapie-exposed animals that are not in source or infected flocks (i.e., those that are identified by yellow “exposed” ear tag or that are received on a VS 1-27 permit).

7. Any other sheep that is 2, 3, 4, or 5 years of age as evidenced by examination of the teeth. A sheep will be considered 5 years of age if the fourth permanent incisor is fully erupted and shows some evidence of wear, and there are no broken, missing, blunt, elongated, splayed teeth, or other significant signs of dental wear present.

8. Goats 2, 3, 4, or 5 years of age as evidenced by examination of the teeth, that originated from historically high prevalence States or States with goat cases in the preceding 5 years. For FY 2011, this will include Michigan, Ohio, Indiana, and Illinois.

9. Condemned postmortem for emaciation.

10. Animals of higher risk than the general population or needed to achieve State sampling targets as determined by the regional scrapie epidemiologist.

11. Sheep or goats identified with a Scrapie Flock Certification Program tag (up to 30 animals cumulative per flock or herd); after 30 animals are collected, only animals meeting the targeting criteria listed above should be collected.

Gaps in existing slaughter surveillance
Scrapie sampling does not occur at all slaughter facilities in the country. Participation in the RSSS is voluntary for slaughter establishments that do not engage in interstate commerce. Furthermore, due to limited resources, sampling has not been implemented at all establishments that engage in
interstate commerce. In FY 2009, 108 slaughter facilities collected samples as part of the RSSS; these include 71 Federal establishments that processed about 82 percent of the volume of mature sheep going through Federal inspection. Approximately 89 percent of the RSSS surveillance samples collected in FY 2009 was from these federally-inspected establishments.

Gaps in existing slaughter surveillance currently being addressed include:

- Apparently healthy goats,
- Export cull ewes,
- Mature sheep slaughtered at non-participating slaughter plants, including some Federal and many non-Federal slaughter plants (such as State-inspected plants, live animal markets, or custom-exempt plants), and
- Animals from producers who slaughter at home or sell animals only for noncommercial slaughter.

To fill these gaps, two surveillance initiatives have been or will be implemented. These include the following:

1. **Apparently healthy goats**
   Starting in FY 2011, the surveillance strategy includes slaughter surveillance of apparently healthy goats, with emphasis on those in higher-risk States (i.e., those residing in States with a history of goat cases or a disproportionate number of scrapie-infected flocks or herds). We will implement the prorated share of the national OIE goal for goats the fiscal year following publication of the revised scrapie rule, which will require the identification of all mature goats in interstate commerce.

2. **Small-volume slaughter establishments**
   Some non-Federal facilities already participate in RSSS and collected approximately 11 percent of the samples in FY 2009. The scrapie program encourages new and continued RSSS participation of these smaller-volume plants to reach new sheep and goat populations.

Currently, the animal’s State of origin is not part of the targeted selection criteria. This can only be accurately surmised for animals identified with official flock identification tags; animals identified with serial tags are often tagged at livestock markets and may have originated in another State. Therefore, with the implementation of demographic-based surveillance, compliance with the required identification rule of sheep and goats (9 CFR Part 79) and use of flock-ID tags or attribution of serial tags to flocks are critical in determining whether a State is meeting its sampling target. These efforts will allow the surveillance program to match a sampled animal back to a flock and a State. Although we recognize that this identification may not
necessarily reflect the State of birth, it is currently the best data available to provide information on where the sheep has moved geographically.

It is recognized that some flocks have been repeatedly sampled through the RSSS and on-farm surveillance activities. The number of samples needed to designate a flock as low-risk for scrapie is unknown, as is the number of samples needed to subsequently continue monitoring these low-risk flocks. However, even if those parameters were known, no nationwide mechanism is currently in place to prevent multiple animals from a single flock from being tested at slaughter over time, regardless of past history of multiple negative results from the flock. To address this issue, VS in 2009 introduced a pilot program, a “Do Not Collect” list. The purpose of the list is to remove historically heavily-sampled, scrapie-negative flocks from the sampling pool. VS will assess the effectiveness of this pilot program at the end of 2010, and if successful, will eventually expand it to remove such flocks nationwide. In parallel to this implementation, an evaluation of historical surveillance data is underway to determine sampling requirements for defining low-risk flocks; upon completion, this will allow refinement of the list.

Data Sources
RSSS summary data collected on the day of submission includes the number of mature animals with official identification, the number of goats slaughtered, the number of sheep slaughtered, the number of black- and mottled-faced sheep slaughtered, and the total number of heads sampled, in addition to collector and collection site information. Data collected for each individual animal sampled include the following: animal identification, age, gender, face color (sheep), and designation (i.e., non-clinical, clinical, suspect, known exposed, SFCP or tested at discretion of the scrapie regional epidemiologist).

Data from RSSS surveillance is captured via the Veterinary Services Laboratory System (VSLS). VSLS is an electronic laboratory submission system intended as a common entry vehicle for all future APHIS laboratory data. Data entered via VSLS and subsequent test results are stored in the AHSM database to support analysis and investigations.

B. Nonslaughter Surveillance
Nonslaughter surveillance is targeted toward groups considered to be at higher risk for scrapie and those not being seen at participating slaughter facilities. Currently, nonslaughter surveillance primarily includes the following: exposed and potentially exposed animals identified through trace investigations from infected animals; source and exposed flocks often identified through RSSS; clinically suspect, mature animals submitted for necropsy to diagnostic laboratories; animals submitted to public health laboratories that test negative for rabies; animals tested for scrapie as part of
the SFCP (see [c] below); and QQ sheep in flocks with risk factors for scrapie (antemortem testing). In addition, a small number of samples are collected through voluntary on-farm surveillance.

**Sampling Methods**

Non-slaughter surveillance will be increasingly important as the prevalence of scrapie decreases and scrapie surveillance transitions to a more demographic-based and eventually flock-level approach.

Specifically, on-farm testing will increasingly be utilized to help a State meet its targets. In addition, as fewer trace investigations are initiated because of the decreasing prevalence, on-farm surveillance will be essential as we move toward more population-based strategies for surveillance and documenting disease freedom.

1. **Targeted flock-level sampling (on-farm)**

This increasingly important component of scrapie surveillance consists of sampling of apparently healthy sheep and goats based on certain criteria, including (but not limited to): geographic location; prior testing of animals through other surveillance routes; and other demographic or management practices. The purpose of this component is to specifically target sampling of flocks or herds that have had limited or no scrapie surveillance.

NCAHP and CEAH have analyzed scrapie surveillance data to identify flocks and areas not sampled through current slaughter and nonslaughter surveillance efforts, using official identification tags to designate the flock of origin. It is recognized that these tags may not necessarily reflect flock of birth, but provide the best available information to identify where the animal may have originated. The analysis identified which flocks have not been sampled, factors associated with not being sampled (i.e., number of flocks and/or sheep in a ZIP code), and associations of positive flocks with these findings. This will allow regional and local field personnel to focus and prioritize efforts to fill in the gaps either by slaughter or nonslaughter surveillance routes.

2. **Laboratories**

This component requires enhancing awareness of the importance of scrapie testing in mature sheep and goats by ensuring that diagnostic laboratories forward appropriate tissues from all mature sheep and goats presented for necropsy and diagnostic testing to the NVSL or an approved contract laboratory for scrapie testing, regardless of another diagnosis. This also includes ensuring that public health laboratories forward appropriate tissues from sheep and goats of any age that test negative for rabies to the NVSL or an approved contract laboratory when appropriate samples are available. Information reminders stressing the importance of submitting these samples for scrapie testing are being disseminated to these laboratories in 2010.
3. Epidemiologic Investigations
This group of surveillance activities consists of surveillance information collected on-farm from clinical suspect sheep and goats or as part of trace investigations of animals from source and infected flocks.

a. Trace Investigations
When a positive sheep or goat is identified through slaughter or other routes, trace investigations (forward and backward) are initiated to identify additional infected and source flocks. These investigations involve identifying flocks that may have been exposed to an infected animal or were the source of infection for that animal. Samples collected from exposed and potentially exposed animals residing in flocks/herds not already found to be infected or source flocks/herds are included as part of scrapie surveillance. As fewer trace investigations are initiated because of the decreasing prevalence of scrapie, this source of on-farm surveillance is expected to decrease.

b. Reporting of clinically suspect sheep and goats (on-farm)
This component of surveillance is based on reporting of clinical suspects and depends on disease awareness of farmers/producers and veterinary practitioners and their willingness to report a suspect case. According to the 2001 National Animal Health Monitoring System (NAHMS) Sheep Study, awareness of disease is high, with 92.6 percent of the operations reporting they had heard of scrapie. Only 1.2 percent of operations had suspected or confirmed scrapie in the previous 3 years. However, most operations, when they suspected disease in their flocks, did not have the disease diagnosed by either a veterinarian or a diagnostic laboratory. Only 26.7 percent of operations that reported presence of scrapie on their operations had the disease diagnosed by a veterinarian or laboratory. Educational campaigns have been implemented through NIAA and ASI to inform producers and veterinarians about scrapie and the importance of reporting and testing clinical suspects.

Data Sources
A number of separate applications and spreadsheets make it difficult to obtain and analyze a complete testing history of animals or flocks/herds using the multiple nonslaughter surveillance approaches. Laboratory submissions and test results of these surveillance samples are currently not entered into VSLS. These data are currently stored in spreadsheets maintained separately from RSSS data, with reporting occurring through NVSL and the regional scrapie epidemiologists. Genotype data, tag distribution data, and trace investigation information are collected through VSLS, Animal Identification Number Management (AINM) system, and the current AHSM and stored in the VS
C. Scrapie Flock Certification Program (SFCP)

Implemented in 1992, the SFCP is a voluntary cooperative effort among producers, industry representatives, accredited veterinarians, State animal health officials, and APHIS. Currently, approximately 1,650 flocks are enrolled in the program. This represents only a small proportion (approximately 1 percent) of the total sheep flocks and goat herds in the United States.

The objectives of SFCP are to reduce the occurrence and spread of scrapie, to identify flocks that have been free of evidence of scrapie over specified time periods, to contribute to the eventual eradication of scrapie, and to enhance the marketability of enrolled animals. Flock certification is based solely on absence of disease, not on genetics. Certification status categories include: complete monitored (enrolled or certified), selective monitored, and export monitored (enrolled or certified). In order to meet and maintain certification status, participants must meet identification, record-keeping, and reporting and testing requirements, and they must follow restrictions on flock additions. Participants also are required to record scrapie susceptibility genotype, if known. If genotypes are known, SFCP participants must sample the susceptible animals.

Data Sources
Sheep and goat producers participating in the SFCP are required to report and submit for testing any animals that exhibit clinical signs and some mature animals that die on the farm while enrolled in the program. In addition, regulatory personnel conduct annual inspections to assess flock/herd health and assure animal identification requirements are being met. The flock’s SFCP status and summary of annual inspections are entered into the AHSM database. Some States enter animal inventories for each SFCP flock into AHSM, but this is not a mandatory requirement.
9. Data Presentation and Reports

Internal Reports for Program Monitoring

RSSS
The NSU provides monthly and annual RSSS summary reports that are reviewed in detail by the VS scrapie program staff and regional scrapie epidemiologists to monitor progress toward national sampling targets and program performance measures. These reports are also made available to VS Area Offices and State agencies on the “Scrapie Quickplace” Web site.

The monthly and annual RSSS summary reports present cumulative (since April 2003) and current fiscal year summaries of RSSS specimen collections and test results in tabular format. The reports also include tables that stratify these counts by FY, month of specimen collection, State of specimen collection, collection site, State of animal identification, animal species/face color, and animal age as well as detailed line listings for samples with positive test results.

Non-Slaughter Surveillance
A similar reporting and review process also occurs for surveillance testing conducted on animals at locations other than slaughter. However, this process is currently done manually using spreadsheets. NVSL provides spreadsheets that list epidemiologic information and test results for third eyelid testing and regulatory scrapie testing submitted by field personnel to scrapie program staff on a monthly basis. These spreadsheets are collated and distributed to the designated scrapie epidemiologist (DSEs) on a quarterly basis. The DSEs identify those samples that meet the criteria to be considered as non-slaughter surveillance samples, and return the completed information to the scrapie program staff. Nonslaughter surveillance summary reports are then prepared by scrapie program staff.

VS is developing a module for the Web-based submission of all samples collected through field activities using the VSLS system. Once this module is fully implemented, a reporting process similar to that described for RSSS above will be implemented and these summaries will be incorporated into a single report. This will improve the ability to integrate, monitor, and analyze the scrapie eradication program data from the various surveillance components with respect to this demographic-based surveillance plan.

External Reports to Stakeholders
Scrapie program staff prepares and posts monthly and annual reports on the USDA VS Web site. These reports summarize both regulatory and surveillance activities for interested stakeholders using tables, graphs, figures, and explanatory text. These reports can be accessed by the public at:
Scrapie program staff and regional scrapie epidemiologists regularly present these same program data to stakeholders at national animal health meetings including the United States Animal Health Association (USAHA), ASI, the American Dairy Goat Association, and NIAA.

10. Surveillance System Implementation

Available resources will prevent implementation of optimal sampling targets, locally and nationally, that would minimize the time required to achieve eradication. The budget allocation for scrapie surveillance includes sample collection at slaughter, and also field investigations of clinical suspects and trace investigations to identify new infected or source flocks associated with a confirmed scrapie case. Optimally, targeted populations would include sampling at slaughter all black-faced sheep and all sheep and goats 2, 3, 4, or 5 years of age that originate from flocks that have not been adequately sampled. Optimal targeted populations would also include all sheep and goats in high-risk groups, such as clinical suspects; exposed and potentially exposed animals; dead or disabled mature sheep and goats on farm, at laboratories or at other sites; and apparently healthy sheep and goats on farm if required to meet flock-level goals.

To ensure that a sufficient percentage of flocks/herds are sampled, it will be necessary to reach under-sampled flocks. The number of samples that can be tested within the budget depends on a number of factors, most importantly the cost to collect each sample. With efforts focused on reaching into new slaughter facilities, such as small-volume and custom-exempt plants, the cost to collect each sample will increase. In addition, it will be necessary to conduct on-farm surveillance of apparently healthy sheep and goats to reach some undersampled populations; this will increase the cost to collect each sample. Thus, overall, scrapie surveillance and eradication efforts will be limited by available resources, and meeting optimal targets for sheep and goats will not be possible in most States.

Due to the resource limitations, NSU and national and regional scrapie staff set targeting criteria and modify State-level targets to establish sampling levels within the allocated budget to efficiently detect remaining cases while meeting or exceeding the OIE-recommended detection level. Thus, the minimum annual collection goal for all States will be separate for sheep and goats and will be prorated to meet the OIE detection level nationally. If more resources become available, the NSEP can increase its sampling efforts by developing more inclusive targeting criteria and
higher State-level targets to decrease the time required to achieve eradication. State-level targets will be developed annually, based on sheep and goat demographics, scrapie prevalence, the evolving epidemiology of scrapie in the United States, previous area performance (of meeting targets), and resource availability. Regional, area, and State scrapie staff are responsible for establishing the local RSSS and nonslaughter surveillance routes best able to meet the targets, based on local demographics and available resources.

In addition, implementation of the surveillance plan as described is limited by the current version of VSLS. Surveillance plan components that are impacted include:

1. Regular evaluation of targeting criteria to identify high-risk animals;
2. Assignment of serial tags to flocks;
3. Establishment and implementation of annual sampling targets by State;
4. Electronic identification of sufficiently sampled flock and notification of collector on-site of status (i.e., a ‘Do Not Collect’ notification upon scanning of official identification tag); and
5. Regular identification of geographic areas and/or flocks that are undersampled.

To effectively implement these components, significant modifications to the VSLS beyond that currently planned are required.

Due to this limitation, the above activities are conducted less frequently and information is made available to field personnel by less efficient modes. The “Do Not Collect” is currently updated quarterly and the list distributed to field personnel. Targeting criteria are evaluated every 1-2 years, with changes made to program documents and shared with field personnel via conference call and e-mail. Identification of unsampled flocks and/or geographic areas will occur even less frequently; dissemination of this information is limited to e-mail notification and by access to VS server. With the ongoing updates to VSLS, processes such as these should improve with time, given resource availability, but will not be fully realized for several years.

11. Surveillance Plan Performance Metrics

The following metrics will be considered annually to assess the surveillance program and its ability to meet the objectives stated in this surveillance plan:

- Evaluation of whether States met the annual sampling target for sheep and goats originating in their State;
- Evaluation of number and quality of surveillance samples collected nationally – specifically, meeting the annual target and whether sampling criteria were followed;
- Evaluation of the prevalence of scrapie in the U.S. sheep and goat population. The goal is to see a consistent decline in the prevalence of cases until cases are no longer detected. Upon achievement of this goal, it will be necessary to
continue to conduct surveillance at a level well above the OIE minimum of 95 percent confidence of detection at 0.1 percent prevalence to ensure that eradication has actually been achieved. After all States have been free of scrapie cases for 7 years and have met their cumulative sampling goal following their last case, the surveillance program will need to be appropriately revised to ensure adequate monitoring to document ongoing disease freedom, as defined by the OIE;

- Evaluation of geographic or flock level gaps in surveillance sampling and the targeting of the geographic areas with sheep and/or goat populations or flocks that are missed;
- Evaluation of percentage of flocks in the United States sampled through slaughter or on-farm testing or monitored and tested through SFCP participation. The goal is to see a consistent increase, with an ideal goal of 100 percent of flocks tested or monitored.

12. Surveillance System Evaluation

Annual evaluation of the surveillance system will include: sample collection in new or under-represented geographic areas and flocks, expansion of sampling into new slaughter facilities and other RSSS collection sites, and a documented decline in national scrapie prevalence. The goal will be to re-evaluate and refine sampling plans, and ultimately to allow the development of a revised surveillance plan to document freedom of disease.

References


### Acronyms Used In This Document

<table>
<thead>
<tr>
<th>Acronym</th>
<th>Description</th>
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<tbody>
<tr>
<td>AHSM</td>
<td>Animal Health and Surveillance Management</td>
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<td>AINM</td>
<td>Animal Identification Number Management</td>
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<td>APHIS</td>
<td>Animal and Plant Health Inspection Service</td>
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<td>ASI</td>
<td>American Sheep Industry Association</td>
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<td>AVIC</td>
<td>Area Veterinarian-in-Charge</td>
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<tr>
<td>BSE</td>
<td>Bovine Spongiform Encephalopathy</td>
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<td>CEAH</td>
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<td>CNS</td>
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<td>DSE</td>
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<td>EIA</td>
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<tr>
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<tr>
<td>NSEP</td>
<td>National Scrapie Eradication Program</td>
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<td>Prion Protein</td>
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