

Scrapie and Genotyping in Goats

Information Brief

January 2022

INTRODUCTION

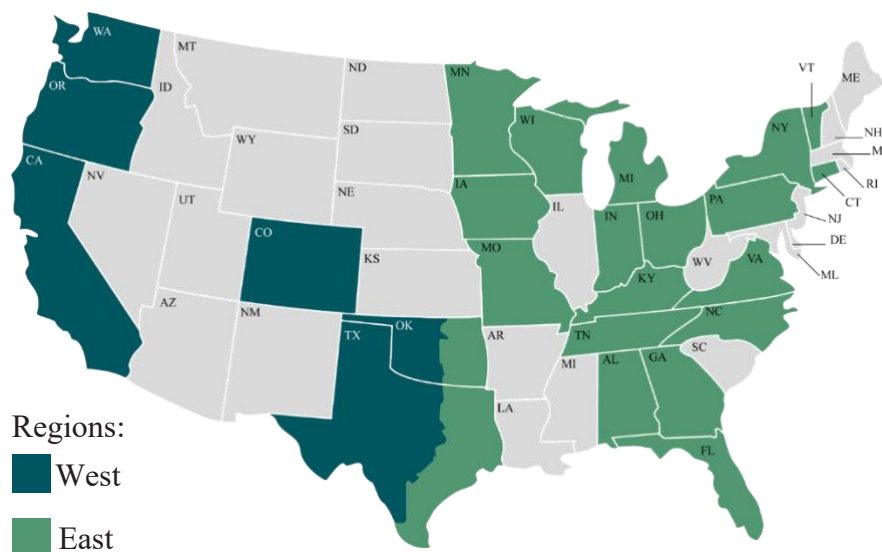
Scrapie is a fatal, degenerative disease of sheep and goats that mainly affects the animals' brain and spinal cord. Scrapie belongs to a group of diseases called transmissible spongiform encephalopathies (TSEs). TSEs are caused by an infectious agent known as a prion. Unlike bacteria and viruses, prions are a misfolding of normal proteins found in the body and are infectious when passed to a person or animal. Scrapie economically impacts sheep and goat industries through production losses, lost exports, and increased production and disposal costs. In the United States, the economic impact of scrapie is estimated at \$10 to \$20 million per year. Therefore, eliminating scrapie in the United States is beneficial for the sheep and goat industries. Scrapie genotype testing is one mechanism to help breed goats that are less susceptible to Scrapie.

NAHMS GOAT 2019 STUDY

U.S. Department of Agriculture's National Animal Health Monitoring System (NAHMS), in collaboration with the National Agricultural Statistics Service, conducted its second national study of the U.S. goat industry in 2019. The NAHMS Goat 2019 study gathered information on goat health and management practices on U.S. goat operations through two phases and biologic sampling. The study was conducted in 24 of the nation's major goat-producing states, on selected operations with 5 or more adult goats (figure 1).

Goat producers who completed the study's phase I and II questionnaires were eligible to participate in biologic testing. Whole blood samples were collected from up to 15 goats, aged 15 months or more, on operations that elected to participate in the scrapie genotype testing. All blood samples were collected by Federal veterinarians or animal health technicians and sent to the National Veterinary Services Laboratory for genetic testing. Farms with more than one breed of goat were eligible to have an additional 5 samples taken from unrelated bucks or does of the other breed(s). Goats were considered unrelated if they did not share a sire or dam. The goals for scrapie susceptibility genotyping were to determine the prevalence of resistant genotypes in U.S. goats and discern variations in prevalence among goat breeds, gender, region (West, East), and goat production types (dairy, meat, other).

Figure 1. States/Regions that participated in the 2019 NAHMS Goat 2019 Study



*Texas and Oklahoma were divided on a line corresponding to north-south Interstate 35. The western halves of the States were included in the West region, and the eastern halves were included in the East region.

KEY TERMS



- **Dairy** refers to operations that primarily produce milk.
- **Meat** refers to operations that primarily raised goats marketed for consumption.
- **Other** refers to operations that primarily raised goats for other reasons.

Operation Size (by Head of Goats)

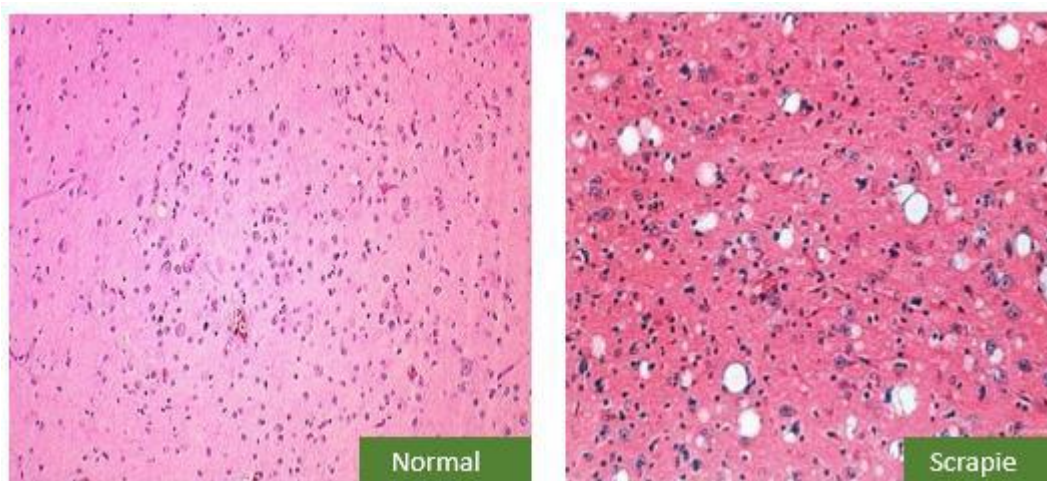


SCRAPIE OVERVIEW

Scrapie is the oldest known TSE— signs of the disease were recorded in sheep in England in the 1700s. The first known case of scrapie in the United States was confirmed in 1947. Today the disease is found worldwide, with the exceptions of Australia and New Zealand. While scrapie is not known to infect humans, public health concerns about TSEs have increased because CJD in humans has been linked to eating meat from BSE-infected cattle. These concerns have resulted in efforts to eliminate all TSEs in food producing animals, including scrapie in sheep and goats.

Clinical signs of scrapie include weakness, significant weight loss without loss of appetite, increased sensitivity to noise, sudden movement, tremors, staring (also known as “star gazing”), head pressing, and repeated intense rubbing against fixed objects (apparently to relieve itching). These clinical signs are caused by the misfolded protein accumulating in the brain and spinal cord, killing nerve cells, and causing tiny holes in the brain (figure 2). The body does not recognize prions as a problem and lacks the enzymes to remove them. The incubation period of scrapie can range from 18 months to 5 years. Once signs begin, the animal’s condition slowly gets worse, which usually occurs over several months until the animal dies.

Figure 2. Brain tissue from a normal sheep and a sheep with scrapie



NATIONAL SCRAPIE ERADICATION PROGRAM

The National Scrapie Eradication Program (NSEP) focuses on eliminating scrapie from the United States to reduce scrapie-associated economic losses, increase international marketing opportunities, and ensure the health of the Nation's sheep and goat herds. Samples for scrapie testing are collected from animals at slaughter via the Regulatory Scrapie Slaughter Surveillance program (RSSS), on farms, and from animals exposed or potentially exposed to scrapie. The national goal is to test at least 40,000 sheep and goats for scrapie every year. For adult goats tested under RSSS from 2017–2019, about 1 in 10,000 goats were found to be infected.

Animal identification is an important part of the NSEP. If a sheep or goat is diagnosed with scrapie, official identification, and other rules under the NSEP allow for tracing. Tracing is used to find the likely source of infection as well as other sheep or goats potentially exposed to the disease. There are several USDA-approved official identification devices for use in sheep and goats (e.g. official eartags, implants, tattoos). To learn more about identification requirements, visit <https://www.aphis.usda.gov/aphis/ourfocus/animalhealth/animal-disease-information/sheep-and-goat-health/scrapie-tags/id> or call 1-866-USDA-TAG.

Producers have used genotyping to detect resistance to scrapie in sheep since 2003. Information gleaned through genotyping allows producers to selectively breed their animals for scrapie resistance, which has helped to decrease the number of sheep in the United States with scrapie. Currently, it appears that 65 to 70 percent of sheep slaughtered in the United States have genotypes resistant to scrapie. Until recently, scrapie resistant genotypes in goats were not well defined. Research on genetic resistance in goats, however, has now advanced to the point where genotype testing in goats can be used to identify scrapie resistant or less susceptible goats.

SCRAPIE GENOTYPE TESTING

Specific segments of DNA coding for the scrapie prion are associated with susceptibility to scrapie in sheep and goats. These segments, or codons, provide templates for the building blocks of proteins produced by the body. These building blocks are known as amino acids. Each codon is assigned a specific number. A single change in an amino acid in two specific codons is associated with susceptibility to scrapie. Codons 146 and 222 appear to be the most important codons impacting susceptibility to scrapie in goats.

Based on current knowledge, goats that have an S (amino acid serine) or a D (amino acid aspartic acid) present at codon 146, or goats that have a K (amino acid lysine) present at codon 222, appear to be less susceptible to scrapie.^{1,2,3} Research has shown that goats with codons with an S or D present at 146 or a K present at 222 increases the disease incubation period by four to five times when compared with goats with more susceptible codons. Because genotypes at these codons are passed down from bucks and does to their kids, breeding goats that are less susceptible to scrapie is possible (figure 3). To determine if breeding to decrease genetic susceptibility could be used in goats, the USDA needed to determine what percentage of U.S. goats naturally carry the resistant genotypes.

Figure 3. Offspring's genetic risk of susceptibility to scrapie based on parents' genotype at codon 146

Doe at Codon 146	Buck at codon 146	
	N	S
N	NN- Susceptible	NS- Less susceptible
S	NS- Less susceptible	SS- Less susceptible

SCRAPIE GENOTYPING RESULTS

Overall, 84.0 percent of eligible operations (n=654), accounting for 6,029 goats, participated in scrapie genotyping. The primary production types of sampled operations were 45.8 percent dairy, 34.0 percent meat, and 20.2 percent other, which included angora/fiber operations. Over one third, of sampled operations (37.8 percent) were in the West region and 62.2% were in the East region (see figure 3). Sampled goats represented 19 different breeds, with the most common being crossbred (17.8 percent) Boer (16.8 percent), and Nubian (11.9 percent). A higher percentage of does than bucks were sampled (82.4 and 17.6 percent, respectively).

CODON 146

Overall, 33.3 percent of goats sampled had a genotype associated with lower susceptibility to scrapie at codon 146 (S or D). A higher percentage of Savannah, Spanish, Boer, and Nubian goats (65.5, 42.6, 47.0, and 38.6 percent, respectively) had a less susceptible scrapie genotype at codon 146. A higher percentage of does (33.3 percent) had a less susceptible genotype at codon 146 compared with bucks (23.6 percent). A higher percentage of meat operations (88.9 percent) had any goats with genotypes associated with lower susceptibility scrapie at codon 146 compared with dairy and other operations (51.0 and 57.6 percent, respectively).

Figure 4. Percentage of operations that had goats with genotypes less susceptible to scrapie at codon 146

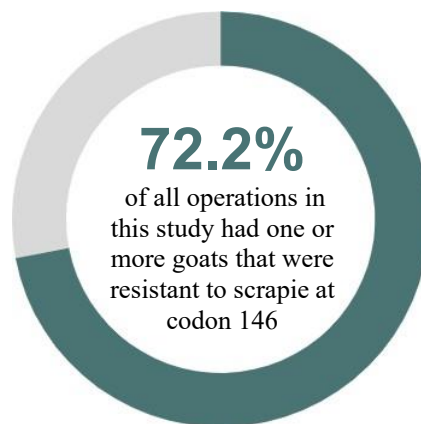
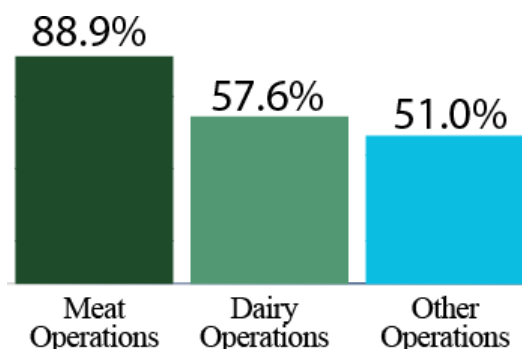


Figure 5. Percentage of operations with goats that had genotypes less susceptible to scrapie at codon 146 by primary production



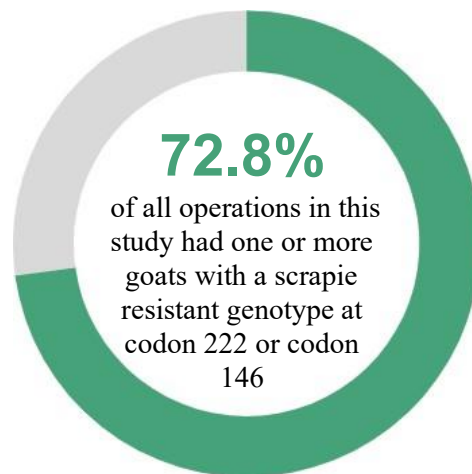
CODON 222

Goats that had a genotype associated with lower susceptibility to scrapie at codon 222 (K) were rare in this study. Only 0.6 percent of sampled goats had a genotype less susceptible to scrapie at codon 222. A higher percentage of sampled goats in the East region (1.0 percent) had a genotype less susceptible to scrapie at codon 222 compared with sampled goats in the West region (0.1 percent).

ANY RESISTANCE

When combining the results of codons 146 and 222, 33.8 percent of goats had a genotype associated with lower susceptibility to scrapie at codons 146 and 222. Overall, 34.7 percent of sampled does and 24.4 percent of sampled bucks had any genotypes less susceptible to scrapie at codons 146 and 222. However, these goats were spread across 72.8 percent of U.S. goat operations. Does with genotypes less susceptible to scrapie were found on 69.5 percent of sampled operations and bucks with scrapie resistant genotypes were found on 21.3 percent (figure 6).

Figure 6. Percentage of operations with any goats that had genotypes less susceptible to scrapie at codons 146 and 222



CONCLUSION

Goats with genotypes less susceptible to scrapie are found on many goat operations across the United States, but only one-third of all U.S. goats carry the genotypes that are less susceptible to scrapie. Scrapie in U.S. sheep has been greatly reduced, in part because of breeding for genetic resistance. Sheep resistant to scrapie develop clinical signs much later in life, if ever, and shed fewer scrapie prions, reducing the amount of the infectious agent on the farm. Actions that helped reduce scrapie in sheep are now being advocated for in goats. These actions include official identification, testing, and breeding for resistant genotypes. Biosecurity actions such as limiting outside herd additions and purchasing breeding animals from scrapie free herds, as well as breeding for scrapie genetic resistance, are important ways to prevent scrapie in goat herds.

An efficient method to increase the prevalence of scrapie resistant genotypes in a herd is breed only bucks that carry genotypes less susceptible to scrapie. Goat producers should discuss genotype testing for scrapie susceptibility with their veterinarian.

The cooperation of goat and sheep producers is vital to the elimination of scrapie in the United States. Producers are urged to work with their veterinarian regarding rules for official identification, test their animals on farm and at slaughter, and consider breeding scrapie resistance into flocks and herds. More information on scrapie, the NSEP, and genetic resistance is available at:

https://www.aphis.usda.gov/aphis/ourfocus/animalhealth/SA_Animal_Disease_Information/sheep-goat

REFERENCES

1. Papasavva-Stylianou P, Simmons MM, Ortiz-Pelaez A, Windl O, Spiropoulos J, Georgiadou S. Effect of Polymorphisms at Codon 146 of the Goat PRNP Gene on Susceptibility to Challenge with Classical
2. Fast C, Goldmann W, Berthon P, Tauscher K, Andreoletti O, Lantier I, et al. Protecting effect of PrP codons M142 and K222 in goats orally challenged with bovine spongiform encephalopathy prions. *VetRes.* 2017;48(1):52. Epub 2017/09/21. doi: 10.1186/s13567-017-0455-0. PubMed PMID: 28927447; PubMed Central PMCID: PMC5606029.
3. White SN, Reynolds JO, Waldron DF, Schneider DA, O'Rourke KI. Extended scrapie incubation time in goats singly heterozygous for PRNP S146 or K222. *Gene.* 2012;501(1):49-51. Epub 2012/04/21. doi: 10.1016/j.gene.2012.03.068. PubMed PMID: 22516690.

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