

## REPORT OF THE COMMITTEE ON SCRAPIE

Chair: Jim Logan, Riverton, WY  
Vice Chair: Charles Palmer, Redding, CA

Deborah L. Brennan, MS; Shane A. Brookshire, GA; Beth W. Carlson, ND; John R. Clifford, DC; Thomas F. Conner, OH; Walter E. Cook, WY; Linda A. Detwiler, NJ; William F. Edmiston Jr. DVM, TX; Anita J. Edmondson, CA; Dee B. Ellis, TX; Dave E. Fly, NM; Keith R. Forbes, Nv; Michael J. Gilsdorf, MD; William L. Hartmann, MN; Burke L. Healey, NC; Susan J. Keller, ND; James W. Leafstedt, SD; Mary J. Lis, CT; Michael R. Marshall, UT; Cheryl A. Miller, In; Alecia L. Naugle, MD; Brian V. Noland, ID; Kristine R. Petrini, MN; Jewell G. Plumley, WV; Michael R. Pruitt, OK; Paul E. Rodgers, CO; Joe D. Ross, TX; Larry A. Schuler, ND; Ben Smith, WA; Diane L. Sutton, MD; Lynn Anne Tesar, SD; Delwin D. Wilmot, NE; Nora E. Wineland, CO; Cindy B. Wolf, MN.

The Committee met on October 28, 2008 at the Sheraton Greensboro Hotel in Greensboro, North Carolina, from 12:30 p.m. to 3:45 p.m. There were 14 members and 26 guests present. The meeting was called to order by Vice-Chair Dr. Charles Palmer. Chair Dr. Jim Logan joined the committee meeting at 1:00 p.m. The Committee heard and discussed the following presentations:

United States Department of Agriculture (USDA), Animal and Plant Health Inspection Service (APHIS) Scrapie Program Update – Epidemiological Update was presented by Dr. Diane Sutton, Veterinary Services (VS), USDA-APHIS.

In Fiscal Year 2008 the Scrapie Eradication Program focused on: 1) cleaning up infected and source flocks utilizing a genetic based approach; 2) tracing and testing exposed animals and animals in exposed flocks; 3) expansion of regulatory slaughter surveillance (RSSS); 4) producer education, 5) ID compliance; 6) finalizing the National Scrapie Surveillance Plan; 7) completion of the Caprine Scrapie Prevalence Study; and 8) implementing the Veterinary Services Laboratory Submission System for regulatory scrapie slaughter surveillance.

### **Scrapie Flock Certification Program**

As of September 30, 2008, there were 1,971 flocks participating in the Scrapie Flock Certification Program (SFCP). Of these flocks 505 were certified flocks, 1,436 were complete monitored flocks, 24 were export monitored and 4 were selective monitored.

### **National Scrapie Surveillance Plan**

The National Scrapie Surveillance has been finalized and posted at [http://www.aphis.usda.gov/vs/nahss/sheep/national\\_scrapie\\_surveillance\\_plan\\_08192008.pdf](http://www.aphis.usda.gov/vs/nahss/sheep/national_scrapie_surveillance_plan_08192008.pdf). The plan provides a comprehensive review of scrapie surveillance in the US, explains the basis for implementing state-of-origin sampling targets and ultimately flock level surveillance, and establishes minimum targets for FY 2009 and 2010.

### **Infected and Source Flocks**

As of September 30, 2008, there were 31 scrapie infected and source flocks, a decrease of 16 percent from September 30, 2007. There were a total of 61 new infected and source flocks reported for FY 2008, a decrease of 15 percent from FY 2007. Chart 1 shows the number of new infected and source flocks by year. The total infected and source flock statuses that were released in FY 2008 was 64. 174 positive scrapie cases were confirmed and reported by the National Veterinary Services Laboratories (NVSL) for FY 2008. Of these, 40 were RSSS cases, (collected in FY 2008), 128 positive field necropsy cases, 4 rectal biopsy and 2 third eyelid tests. Five of the field cases were goats that originated from the same herd. One RSSS case was consistent with Nor98 scrapie. NOTE: Ante-mortem scrapie testing in sheep and goats using rectal biopsy was approved for program use by USDA for in January 2008.

Approximately 2,438 animals were indemnified comprised of 51.4 percent non-registered sheep, 30.5 percent registered sheep, 9.6 percent non-registered goats and 8.5 percent registered goats.

### **Regulatory Scrapie Slaughter Surveillance (RSSS)**

RSSS was designed based on the findings of the Center for Epidemiology and Animal Health (CEAH) Scrapie: Ovine Slaughter Surveillance (SOSS) study. The results of SOSS can be found at <http://www.aphis.usda.gov/vs/ceah/cahm/Sheep/sheep.htm>. RSSS started April 1, 2003. It is a targeted

slaughter surveillance program which is designed to identify infected flocks for clean-up. During FY 2008, collections increased by 6 percent overall and by 13 percent for black and mottled face sheep compared to FY2007. Improvement in the overall program effectiveness and efficiency is demonstrated by the 30 percent decrease in percent positive black faced sheep compared to FY 2007 (.27 to .19 percent, based on test results posted before October 10, 2008). During FY 2007, 43,887 samples were collected (chart 2). There have been 40 NVSL confirmed positive cases collected in FY2008. Face colors of these positives were 39 black and 1 white. The white face case was consistent with Nor98 scrapie. The percent positive by face color is shown in the chart 3 below. One black face case was in an AA<sub>136</sub> QR<sub>171</sub> ewe.

### **Caprine Scrapie Prevalence Study (CSPS)**

CSPS was conducted from May 2007 to March 2008, to estimate the national prevalence of scrapie in adult goats at slaughter. 3,032 goats were sampled for scrapie testing. None tested positive for scrapie; from this we are able to conclude that the prevalence is less than 0.1 percent.

### **Scrapie Testing**

As of September 30, 2008, 48,269 animals have been sampled for scrapie testing: 43,887 RSSS, 1,517 goats for the CSPS, 2,277 regulatory field cases, 139 necropsy validations, and 282 and 306 regulatory third eyelid and rectal biopsies respectively.

### **Animal ID**

As of September 30, 2008, 145,343 sheep and goat premises had been assigned identification numbers in the Scrapie National Generic Database and 113,656 premises had received official ear tags.

*Note: report based on data available as of October 10, 2008*

Nor98-like Scrapie in the United States of America was presented by Drs. Christina M. Loiacono, S. Mark Hall, and Bruce V. Thomsen, National Veterinary Services Laboratory, USDA-APHIS-VS.

This paper describes the first six sheep diagnosed with Nor98-like disease in the United States and serves to acknowledge the increased efforts of diagnosticians and the USDA program to control and eradicate scrapie disease. Classical scrapie, a fatal neurodegenerative disease affecting the central nervous system of sheep and goats, is among a number of diseases classified as transmissible spongiform encephalopathies (TSEs). Recently, a distinct strain of scrapie was diagnosed in sheep in Norway<sup>1</sup> and has been identified in numerous countries of the European Union (EU). The disease has been identified, among other names, as Nor98 or Nor98-like scrapie. Distinctions between classical scrapie and Nor98-like scrapie are made based on signalment, clinical signs, histopathology and immunodiagnostic results. In the past, the classical scrapie disease was confirmed by examination of the brain tissue for a triad of histopathological signs – vacuolation, loss of neurons and gliosis – and, more recently, by immunohistochemical (IHC) or biochemical detection of abnormal prion protein (PrP<sup>Sc</sup>) in the brain, or lymphoid tissues. In the case of Nor98-like scrapie there is generally little or no vacuolation in the brain and, to date, no lymphoid accumulation of PrP<sup>Sc</sup> has been detected. Classical scrapie typically has the most intense PrP<sup>Sc</sup> immunostaining at the obex (motor nucleus of the vagus), while this area is spared in Nor98-like scrapie. Alternatively, Nor98-like scrapie consistently has PrP<sup>Sc</sup> immunostaining in the spinal nucleus of the trigeminal nerve and variable, but often an intense immunostaining for PrP<sup>Sc</sup> in the cerebellum. Thus the diagnosis of Nor98 and Nor98-like disease can be based on immunohistochemistry identifying abnormal prion protein in regions of the brain not typically associated with classical scrapie. Additionally there is a distinct diagnostic western blot pattern for Nor98 and Nor-98 like disease consisting of three or more protein bands with the unglycosylated band being less than 15 kd, compared to classical scrapie in which the unglycosylated band is greater than 15 kd. Nor98 and Nor-98 like disease is associated with older sheep, usually greater than four years of age, while sheep in the range of three to five years of age are more commonly affected by classical scrapie. Clinical signs are uncommon with Nor98 and Nor98-like disease but when present most often include ataxia without pruritis. Genotypes known to provide sheep with resistance to classical scrapie are not spared from Nor98 and Nor98-like disease.

The six U.S. cases had no clinical signs reported. Three cases were detected during slaughter surveillance, two were detected as a result of classical scrapie being found in the flock, one found during testing associated with diagnostic necropsy. Five of the 6 cases had genotypes that are susceptible to classical scrapie and one was AARR. Only one Nor98-like scrapie case was found per flock.

1. Benestad SL, Sarradin P, Thu B, Schönheit J, Tranulis MA, Bratberg B., Cases of scrapie with unusual features in Norway and designation of a new type, Nor98. Vet. Rec.

The Role of Economics in Scrapie Regulations was presented by Mr. Paul Rodgers, American Sheep Industry Association.

Why Eradicate?

- The presence of scrapie in the U.S. restricts trade.
- The presence of scrapie in flocks costs producers money.
- It is costly to prevent if the disease is present in other flocks (still no early preclinical test).
- Eliminating any/all TSE's from U.S. animal populations makes good sense.
- The cost to the sheep industry is estimated to be at least \$25 million annually (death loss, loss of markets).
- Scrapie prevention costs to producers is high (closed flocks is the safest practice and that limits genetic improvement opportunities).

The National Accelerated Scrapie Eradication Program began in 2001. Many millions of dollars were spent trying to eradicate scrapie between 1952 and 1992. From 1992-2001 approximately \$3 million per year was spent on scrapie control by USDA APHIS through the Scrapie Flock Certification program. Ten million dollars in CCC funds was added in 2001 when the accelerated program began. Current spending is approximately \$18 million annually.

Scrapie costs the United States' sheep industry an estimated \$25 million annually in death loss and loss of marketability, and scrapie prevention costs are also significant. The economic value of the sheep industry was estimated to be \$767.5 million in 2007. The current federal investment in scrapie eradication is \$3 per head of sheep in the U.S., making the value 43 times the cost at current spending.

Rapid, aggressive disease eradication programs are efficient. Federal program disease eradication history proves that low or slow investments over time ends up costing the public sector significantly. Strong investment in diagnostic tools, animal identification and tracking, and enforcement of compliance with program requirements will increase the return on investment to both private industry and government agencies.

Goat Genetics – Update on Progress was given by Dr. Stephen White, USDA-ARS. There are at least two PRNP genotypes that, to our knowledge, have never been observed in goats with scrapie despite the presence of such animals in positive flocks. Oral challenge experiments began this spring to investigate both genotypes. Additional numbers will be added to boost statistical power. And it will take some time for incubation before any assessments can be made.

Scrapie: Information From Canada Regarding Import Requirements for Sheep and Goats Imported from the United States was presented by Dr. Maria Koller-Jones, Canadian Food Inspection Agency.

A report from the Canadian Food Inspection Agency informed the committee that currently, Canada requires that the flock/herd of origin must be enrolled in either the export certified pathway or the complete monitored pathway (as long as the flock/herd is testing all on-farm deads) of the U.S. scrapie flock/herd certification program.

Canada intends to amend its import conditions to require that the flock/herd of origin:

- has been enrolled in the U.S. scrapie flock/herd certification program for a specified period of time, and;
- has been in compliance with the requirements of the export pathway or the complete monitored pathway plus testing of all on-farm deads, for a specified period of time.

It is expected that these new conditions for the importation of female breeding sheep and goats will come into effect in the fall of 2009.

The Committee passed one resolution encouraging USDA to increase funding for scrapie eradication.