

OBSERVATIONS FROM THE PASTURE

Prions Redux

In the December 2003 issue of the Newsletter my column was entitled *Mad Cow, Prions and Animal Ethics*¹. At the time I indicated that I would be monitoring prion disease² literature and also keeping my legislators informed of my concerns about prion diseases. My intent in this column, with the exception of the introductory paragraph below, is to cover information I have acquired since the initial article.

Disclosure

To the best of my knowledge prion diseases have yet to be diagnosed in camelids and, to the best of my knowledge, there is no evidence indicative of susceptibility of camelids to prion diseases.

Introduction

Prion diseases are a group of fatal neurodegenerative diseases that are found in mammals. Examples of prion diseases, also referred to as transmissible spongiform encephalopathies (TSEs), include scrapie in sheep and goats; chronic wasting disease (CWD) in deer, elk and most recently moose; mad cow disease (bovine spongiform encephalopathy – BSE) found in cattle and other bovines; and Creutzfeldt-Jakob disease (CJD) found in humans. A variant of CJD, vCJD, has been linked to consumption of beef from cattle infected with BSE. A common feature of these and other prion diseases is the development of small cavities (vacuoles) in the gray matter of the brain, giving the brain a sponge-like appearance.

Why this Column was Written Now

My decision to write a follow-up column on prion diseases at this time was based on an accumulation of disturbing events:

- BSE has been transmitted to cattle through the incorporation of diseased tissue from cattle and sheep in prepared feed. To curb this form of transmission the FDA instituted a ban in 1997 of certain animal protein in animal feed for cattle and other ruminants. There have been questions of the efficacy of the administration of this ban. In response to a request from Senators Chambliss, Harkin, Cochran and Durban the United States Government Accountability Office (GAO) prepared a report³ on the effectiveness of the ban. The report essentially concluded that while there has been some improvement, there

are a sufficient number of holes in the administration of the program so as to limit its effectiveness.

- On March 13, 2006, a cow was found in Alabama that was ultimately determined to have BSE. The USDA was unable to trace the cow's herd of origin⁴.
- The USDA denied a request by Creekstone Premium Beef for permission to test every animal it processes for BSE⁵. The reason given for the denial was that the USDA and larger meat packers oppose such a high level of testing because it would not assure food safety since "testing rarely detects BSE in the age group of animals primarily used for meat".
- The Consumers Union, in a July 25, 2005 letter to the Secretary of Agriculture, Mike Johanns, raised serious issues and problems with the testing program for BSE⁶. The issues and problems are serious enough to raise doubts about any conclusions drawn from the testing program.
- On April 28, 2006 the Agriculture Secretary Mike Johanns announced that the USDA wants to cut back on testing for BSE⁷. The justification for cutting back is the small number of animals found with BSE. Critics, including myself, believe that any projections of BSE prevalence have been compromised by the testing procedures and administration.

Time for a Sigh of Relief

Marsharee Wilcox, at Camelid Community 2005, reported that the USDA deems camelids to be at low risk for transmitting BSE⁸. This conclusion is not surprising since BSE appears to be transmitted via consumption of infected tissue. For any possible risk of BSE transmission to occur it would appear that a camelid would first have to be susceptible to cross-species BSE infection, would then have to ingest infected tissue, e.g., feed prepared with infected tissue, and finally would have to enter the food chain.

My Perspective

As an actuary I am naturally suspicious of statistical studies until I understand the methodology and data collection procedures used. I have found that there are at least three impediments to achieving valid results, namely money, zealotry and preconceived notions. I have observed on occasion that the brains of those performing studies turn to mush when there

is a financial reward if the results come out a particular way. A similar situation exists for zealots where the reward is one of “proving” a point. Finally, I have found that individuals with preconceived notions tend to obtain results supporting those notions. With respect to the testing program for BSE I note that there is a very substantial amount of money involved in the cattle industry and that there has been at least one incident where USDA Food Safety Inspection Service officials at a meat processing plant requested a BSE test on a suspect cow and were overruled by an APHIS official⁹. I have concluded that there is little appetite in official Washington for definitively determining the prevalence of BSE, let alone other prion diseases.

Where’s the Beef

My concern for prion diseases and camelids is not with BSE, rather it is with prion diseases in general and the apparent increase in prevalence of these diseases. I have referenced BSE above because, currently, it has gathered the most attention about prion diseases because there is a cross-species linkage involving humans.

Currently it is not known whether Chronic Wasting Disease (CWD), is transmissible cross-species by eating infected tissue. Specially adapted mice have been used to test whether CWD can be transmitted via the consumption of deer meat¹⁰. The researchers, upon reviewing the results of the experiment, concluded that “humans consuming or handling meat from CWD-infected deer are therefore at risk to prion exposure”. A study is now in progress of individuals who consumed venison from a CWD infected deer at a sportsman’s feast in Verona, NY in 2005¹¹.

Concerns about Prion Diseases and Camelids

CWD has been transmitted experimentally by intracerebral injection to a number of other species¹². This is not an indication of natural cross-species infectivity, but it does raise questions of cross-species susceptibility. Of particular concern is that changes in cross-species infectivity can occur when the disease does crossover to another species. Prion changes as a result of that crossover may have enhanced infectivity for other species.

CWD, like scrapie, spreads from animal to animal. Of particular concern is that it has been determined that prions adhere to some soils and retain their infectivity¹³. Consequently reservoirs of infectivity may be created by shedding (urine, feces, saliva and

other bodily fluids) and decomposition of infected carcasses. There is concern that contaminated soil may increase the likelihood of the spread of prion diseases to other species¹⁴. It should be noted that infective prions are virtually indestructible. Incineration and disinfectants have not worked. At the very least camelid owners and prospective camelid owners should avoid housing their four-footed friends on land where sheep or goats had been infected with scrapie. Similarly, in areas where CWD is prevalent, land which has been occupied by deer, elk or moose should be avoided.

What Can/Should We Do

First, we should acknowledge that, to date, there has been no evidence of prion disease in camelids. We should also acknowledge that prion diseases may be becoming more prevalent. If, indeed, there is an increase in prevalence, it may be the result of some environmental stress factor. This leads me to my first observation/recommendation: we should continue to monitor the situation and communicate with our legislators whenever we are not pleased with government actions in this area.

Second, when and if, prion diseases are discovered in camelids it will be the result of a post-mortem exam of the brain of an infected animal. Symptoms of prion disease are similar to other neurodegenerative diseases, e.g., Alzheimer’s and dementia in humans. It will not surprise me if the first diagnosis of prion disease in a camelid occurs as a result of a post-mortem exam of the brain of a camelid which was assumed to have contracted meningeal worm. We advance the understanding and care of our animals when we have a post-mortem exam performed when the death is clearly not due to advanced age. Please consider doing so, particularly if the animal exhibited symptoms similar to meningeal worm infection and did not respond to standard meningeal worm treatment.

¹ An Adobe PDF copy is available at www.greenbriarllamas.com/Observations24.pdf.

² The disease agents in prion diseases are not viruses or bacteria, but rather are proteins. The diseases are both infectious and hereditary. The diseases are also sporadic, i.e., there are cases where there is no known risk factor. For more information you may wish to review *The Prion Diseases* by Stanley Prusiner, www.cyber-dyne.com/~tom/prionSP.html. This article is oriented for the layperson. For more detailed technical information you may wish to review *The*

Prion Diseases by Drs. James Mastrianni and Raymond Roos, www.medscape.com/viewarticle/410863.

³ The report, *Mad Cow Disease - FDA's Management of the Feed Ban Has Improved, but Oversight Weaknesses Continue to Limit Program Effectiveness* may be found at www.gao.gov/new.items/d05101.pdf. A convenient summary of the findings of the report, *GAO faults FDA handling of BSE-linked feed ban* is located at www.cidrap.umn.edu/cidrap/content/other/bse/news/april2605bse.html.

⁴ *US can't find birthplace of latest mad cow*. Reuters, Wednesday, May 3, 2006 11:40 EST.

⁵ *Meatpucker Sparks Mad Cow Testing Fight*, CBS News, March 22, 2006, www.cbsnews.com/stories/2006/03/22/ap/health/printableD8GGT1003.shtml.

⁶ *Mad Cow: Consumers Union calls on USDA to release data*. www.nodowners.org/media_union.htm

⁷ *Mad-Cow Testing in U.S. Likely to be Scaled Back*, The Wall Street Journal Online, April 28, 2006.

⁸ See Camelid Community 2005 Meeting Minutes at www.llamaweb.com/lamas/meetingnotes/CamelidCommunity_2005.html

⁹ *Mad Cow: Consumers Union calls on USDA to release data*. www.nodowners.org/media_union.htm

¹⁰ *Deer muscle transmits prion disease to altered mice*, www.cidram.umn.edu/cidrap/content/other/bse/news/jan3106cwd.html.

¹¹ *Chronic Wasting Disease in deer and humans*, Disease/Infection News, Saturday, 26-Nov-2005, www.news-medical.net.

¹² *Chronic Wasting Disease of Deer and Elk*, Emerging Infectious Diseases, www.cdc.gov/eid, Vol. 10, No. 6, June 2004

¹³ *Prions Adhere to Soil Minerals and Remain Infectious*, by Johnson, Phillips, Schramm, McKenzie and Aiken, PLoS Pathogens, www.plospathogens.org, April 2006, Volume 2, Issue 4.

¹⁴ *Prions Adhere to Soil Minerals and Remain Infectious*, by Johnson, Phillips, Schramm, McKenzie and Aiken, PLoS Pathogens, www.plospathogens.org, April 2006, Volume 2, Issue 4