# Chronic Wasting Disease: Summary of papers at the NADVet Conference, Austin, Texas, 19-20 Feb, 2002

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#### **Abstract**

Chronic wasting disease (CWD) is a transmissible spongiform encephalopathy (TSE) that is characterised by accumulations of abnormal proteinaceous or prion material in the CNS. CWD, which occurs mostly in adults, is typified by progressive chronic weight loss and is always fatal. Behavioural changes also occur in the majority of cases, including decreased interactions with other animals, listlessness, hyperexcitability, nervousness, lowering of the head, blank facial expression, and repetitive walking in set patterns. Affected animals continue to eat grain but may show decreased interest in hay. Excessive salivation and grinding of the teeth also are observed. Most deer show increased drinking and urination. Diagnosis is based on necropsy findings, histopathological changes and immunohistochemical testing for the abnormal prion. Gross lesions include emaciation and often aspiration pneumonia, which may be the actual cause of death.

CWD has been found only in elk, mule deer, white-tailed deer, and black-tailed deer in North America. First recognized as a clinical "wasting" syndrome in 1967 in mule deer in a wildlife research facility in northern Colorado, it was identified as a TSE in 1978. Cases from wild cervids of these species were first reported in 1981. In the mid-1980s, CWD was detected in free-ranging deer and elk in NE Colorado and SE Wyoming. It has subsequently been found in free-ranging deer in Nebraska, Wisconsin and the Canadian province of Saskatchewan. It also has been documented in captive herds in Colorado, Montana, South Dakota, Oklahoma, Kansas, and Nebraska and the Canadian provinces of Saskatchewan and Alberta.

Transmission of CWD is thought to be primarily horizontal and the youngest animal diagnosed with CWD to date has been 17 months of age. There is ongoing research to further explore the possibility of transmission of CWD to other species. To date there is no evidence of transmission to humans, which make it similar to scrapie and unlike BSE.

## Introduction

Chronic wasting disease (CWD) is one of a group of fatal mammalian neurodegenerative diseases known as transmissible spongiform encephalopathies (TSEs) that is characterised by accumulations of abnormal proteinaceous material that is a protease resistant (PrPres) form of cellular protein (PrPc) or prion normally produced in the central nervous system. PrPres is a transmissible agent and catalyzes the conversion of PrPc to PrPres in susceptible hosts.

CWD has been found only in elk, mule deer, white-tailed deer, and black-tailed deer in North America. First recognized as a clinical "wasting" syndrome in 1967 in mule deer in a wildlife research facility in northern Colorado, it was identified as a TSE in 1978. Cases from wild cervids of these species were first reported in 1981 but it is likely that the disease was present before then. In the mid-1980s, CWD was detected in free-ranging deer and elk in contiguous portions of north-eastern Colorado and south-eastern Wyoming. It has subsequently also been documented in free-ranging deer herds in Nebraska, Wisconsin and the Canadian province of Saskatchewan. It also has been documented in captive herds in Colorado, Montana, South Dakota, Oklahoma, Kansas, and Nebraska and the Canadian provinces of Saskatchewan and Alberta. It very likely to be diagnosed in other states as surveillance involving surveys of hunter-shot animals and material submitted from farmed animals is undertaken.

Transmission of CWD is thought to be primarily horizontal. The youngest animal diagnosed with CWD to date has been 17 months of age, but the date of infection was not known, so that a true incubation period in natural infections is undetermined. There is ongoing research to further explore the possibility of transmission of CWD to other species. To date there is no evidence of transmission to humans, which makes it similar to scrapie and unlike BSE.

Free-ranging deer herds	NE Colorado SE Wyoming Nebraska Wisconsin	Saskatchewan
Captive herds	Colorado Montana South Dakota Oklahoma Kansas Nebraska	Saskatchewan Alberta

# **State programs on Chronic Wasting Disease**

Ken Waldrup (Texas Animal Health Commission, Austin, Texas)

USDA sets standards for interstate movement of animals, while individual states may set additional requirements for importation or "entry requirements". Currently many states are banning the importation of wild or farmed deer from states that have reported CWD in wild or farmed deer. Some states require a minimum of 5 years CWD-clear status before importation is allowed. Over 10 states now have CWD monitoring programmes, which may be voluntary, and typically these will require the examination of heads from all deer >16 mo that die. There is a preliminary screen of sections of medulla oblongata by a local (TVNDL) laboratory and suspicious material sent to NVSL, Ames, Iowa for definitive prion tests.

# **CWD** in Saskatchewan

Jerry Haigh (University of Saskatchewan, Saskatoon, Canada)

1995 - First case diagnosed in farmed elk in NW Saskatchewan and whole herd destroyed. Affected animal believed to have been imported from South Dakota.

1998 - Second case diagnosed.

*Up to Dec 31, 2001* - 7500 deer destroyed on 38 properties, with 190 positive cases of which 163 had clinical signs. This involved very complicated trace-back and all contacts are now cleared.

Jan 2002 - New case identified, which has not yet been traced back to previous cases.

After extensive monitoring of hunter killed deer (1536 mule, 2614 WTD and 371 elk in 2001-2), two cases have been confirmed in mule deer, one in 2000 and one in 2001, both from the same area and possibly siblings.

Now all elk farms in the Saskatchewan Agriculture and Food (SAF) monitoring programme. All animals tagged and records kept of all births, deaths and movements. All animals showing suspicious clinical signs are killed and necropsied. Deaths of all animals > 12 months must be reported within 24 hours and brain tissue examined. Herd status classified as "low risk" if all samples are clear after 3 years and "free" if clear after 5 years.

The effect on elk farmers in Saskatchewan has been dramatic ("pariah status"). There have been bans on the export of velvet, breeding stock, semen and embryos. Meat sales have fallen and the anti-game ranching lobby are having a field-day. The affect on farmers has been immense.

Affected farms that have been destocked now faced with SAF program to decontaminate the farm and restock. See: http://www.agr.gov.sk.ca

Comment on early clinical signs; there are often swallowing problems, which can lead to pneumonia and some cases can look like "hard-ware" disease.

More information on Murray Woodbury's website:

http://www.usask.ca/wcvm/herdmed/specialstock/

# **US Federal programs on CWD**

Lyn Creekmore

(Animal & Plant Health Inspection Service, Fort Collins, Colorado, USA)

This paper presented the latest information on the USDA efforts to control CWD.

To date the following farmed elk herds have been depopulated: 7 in South Dakota, 1 in Colorado, 1 in Montana, 1 in Nebraska and 1 in Kansas. Infected herds have resulted in 218 interstate traces and 193 intrastate traces. This is an ongoing process and so far 186 of 305 traced animals have been purchased and euthanased and one positive case has been identified. Compensation is 95% market value with US\$3000 maximum. So far 1900 elk from 6 Colorado, 1 Kansas and 1 Nebraska farms have been depopulated at a cost of US\$12.15 million.

In Nebraska they have found a high prevalence of CWD (>30%) in a herd of WTD on a hunting ranch, which was associated with an infected elk herd. In the area around the pen they have found 9/114 positive wild WTD and mule deer.

USDA and NAEBA are developing a programme for controlling and eliminating CWD in captive/farmed elk. This will include minimum fencing requirements, animal ID and inventory, surveillance of all deaths >16 months and examination of brains, etc. It appears to be very similar to the Saskatchewan programme.

There are suggestions of a genetic susceptibility/resistance to CWD, which is similar to the situation with scrapie in sheep. Sequence analysis of the PrP gene suggest that different amino acids (leucine and methionine) at the polymorphic site at codon 132 may affect susceptibility to CWD. Infected and uninfected elk are being tested for LL, LM and MM genotypes.

# **CWD: international implication**

Peter Wilson (Massey University, Palmerston North, New Zealand)

This paper discussed CWD from the perspectives of international producers, vets, industry, regulatory authorities, markets and the general public. The following is an abbreviated version of Peter's paper.

#### Introduction

CWD of deer has been confirmed only in the United States of America, Canada and South Korea. New Zealand has the world's largest deer industry and there is heightened awareness and growing concern amongst farmers, veterinarians and regulatory authorities of the potential impact of this disease on market access, and the effect the disease would have on their industry if it were diagnosed.

This disease possesses many of the least desirable attributes one could wish for from an animal disease: it results in death, there is no live animal test, transmission is vertical and horizontal, it survives in the environment, the incubation period is long, allowing it to spread

insidiously between herds, it has wildlife reservoir hosts, it is related to serious, high-profile spongiform encephalopathies of other animals including man, the disease has spread insidiously from the country of origin to a second and now a third country, it is a difficult disease to research.

## Stakeholders potentially affected

**Producers:** Wapiti imported from North America to NZ in the last 10 years are causing real concern. Farmers have been advised by MAF officials that there is currently no evidence that CWD is present in New Zealand. However, there is still a concern, based on the long incubation period, plus the knowledge that trace-backs from infected herds in Canada suggest that infection might have entered one herd supplying live wapiti to NZ in 1996. It appears that 14 live deer still exist from those shipments. The only deer from the suspect origin farm died a number of years ago, and a post-mortem with laboratory diagnostic confirmation excluded CWD. The current location of the remaining 14 live deer is known and their owners have been advised of the need for surveillance and veterinary attention in the event of animal health problems. Thus, because of the insidious nature of the disease, there is a certain anxiety amongst deer producers in New Zealand for their own herds. MAF and the veterinary profession have heightened the awareness of producers about this disease and its clinical manifestation. Farmers are actively encouraged to contact their veterinarians if they see deer showing nervous or chronic wasting signs. However, there is a dilemma for farmers in reporting suspect cases, with some probably preferring to ignore or dispose of suspect cases rather than risking diagnosis.

**Veterinary practitioners:** MAF and the Veterinary Association have heightened awareness about CWD to the practising veterinary profession, including the fact that pneumonia may be the presenting sign of terminal cases. Veterinarians have an important role in informing farmers of the need for surveillance, and encouraging them to seek veterinary attention to sick animals, particularly those with neurological signs and wasting. Veterinarians are currently paid \$100 by MAF for specimens submitted specifically for investigation of spongiform encephalopathies, including those suspected in deer.

Deer Farmers' Association: The NZDFA, MAF and NZVA have been liaising and discussing the need to maintain awareness, and to heighten surveillance so NZ can meet criteria for disease status declaration according to the OIE International Animal Health Code, when the chapter on CWD is completed. Amongst the issues discussed has been the risk of international movement of deer producers. Deer farmers frequently host producers from other countries, and it is important for them to know the risk of transmission of this disease through clothing and footwear, despite that border Biosecurity should detect these risks before they reach the farm. However, farmers should be advised to provide alternative clothing and footwear, particularly for the visitor known to be from an infected farm in North America or South Korea.

**Regulatory Authorities:** Regulatory authorities in all countries, which have imported wapiti and/or deer from Canada or the USA should be preparing contingency response plans. Biosecurity authorities would be advised to trace back herds of origin of any deer or wapiti imported from North America, and to attempt to locate those deer and identify herds that have contained those deer at any stage during their time in their new country, as carried out in New Zealand. Clear identification of all imported animals should be mandatory. Similarly, semen and embryos that have been imported from North America, and resultant offspring, should undergo trace-back.

In the event of a diagnosis of CWD, whole herd slaughter is likely to be necessary and Government agencies would need to fund this. Lobbying of politicians should be undertaken in advance.

*Disease status declaration:* International trade in deer products depends on declaring its disease status, based on valid sampling criteria applied to various diseases in the OIE

International Animal Health Code. A code chapter on CWD is currently in preparation. Lack of clinical evidence of disease may not be sufficient to convince importing countries of disease status. Thus, there is a need for regulatory authorities to implement targeted surveillance.

The New Zealand MAF was quick to prohibit importation of live deer, semen and embryos from Canada after the implications of CWD were confirmed. In addition, in February 2000 the importation of velvet antler from Canada was banned

## The dilemma of public awareness

There is a balance between public awareness and causing distress amongst the farming fraternity and the deer product-consuming public at large. CWD, as a spongiform encephalopathy, creates particular difficulties in convincing consumers about food safety, given the experience in the United Kingdom with BSE and variant CJD.

### The market for velvet

The ban of importation of North American velvet to Korea is, on the surface, good news for other velvet producers, including New Zealand that currently exports velvet antler worth more than \$40 million. However, awareness of CWD in the Korean and North American velvet markets has potential implications:

- 1. Industries must be cautious about criticising competitors because those industries or sectors within them may choose to denigrate the suppliers' industry aimed at farming methods, welfare or environmental concerns.
- 2. Branding of velvet by origin to differentiate it from product from other countries is important.
- 3. There is a risk of consumer resistance. The possible link with variant CJD could frighten the consumers despite technical statements that this is likely to be a very low risk. Any risk is often considered to be too high.
- 4. The occurrence of CWD in Korea will heighten the awareness of this disease in that country, and is likely to have a negative consequence amongst consumers and possibly even the traditional medical profession on all velvet consumption regardless of the status in countries of origin.
- 5. Common commercial practice is for buyers to use all methods possible to reduce their purchase price. Thus, CWD could be used as an excuse for driving down the price paid to the producer.

Thus, CWD currently has implications for all velvet industries, regardless of CWD status. This is a factor that farmers must consider when reviewing or deciding farming strategy. Some may conclude that the combined risks of consumer concern about disease and welfare are too great, and choose not to farm stags for velvet.

#### The market for venison

Of greater concern to the New Zealand deer industry is protection of venison exports, which currently exceed \$240 million annually and are predicted to grow substantially into the future. The predominant reason is the association between this disease and BSE, and the link between BSE and variant CJD. Food safety concerns go beyond the boundary of official disease status declaration. They are a direct personal concern to the consumer. Potentially more seriously for the NZ deer industry, is selective buying, on claimed food safety grounds (but really based upon pressure to support local suppliers), by large retail chains to achieve competitive advantage in the market. These retail chains have significant buying power, and can act independently of officialdom. They can respond to local producer's pressure to market local produce, even if the cost of production and therefore price is higher. They can respond extremely quickly to changing circumstances, without the need for technical and scientifically

robust justification for their decisions, unlike state veterinary authorities. Thus, they may discriminate against outside suppliers who are only even remotely associated with the disease.

Food safety is the number one concern amongst consumers of animal products. The BSE debacle has done irreparable damage to the image of red meat. Paradoxically, venison is currently selling strongly on the European market in particular, because it is not associated with BSE. However, heightened public awareness of CWD in consumer countries could substitute concerns about BSE, and put demand for venison at equivalent risk to beef. It is understood that there is little or no export of venison from North America. However, the long-term future for any deer industry, even if markets are entirely domestic, must be predominantly for venison if that industry is to grow in the future. Thus, internal markets in North America for venison could well be threatened by this disease, if not already the case. In the short term this is more likely to affect the internal North American market than the European market because of high profile publicity in the national media about this disease, and the emotive issues related to depopulation of deer herds, fuelled by media interest in expenditure of tax-payer funds. Overseas suppliers of venison to the North American market will be affected by generic association of the product with deer, regardless of country of origin.

#### Conclusion

It is tempting for deer industries in countries that do not have CWD to be relieved and use that status for competitive advantage in the marketplace. However, it may not be prudent to allow complacency or smugness to overcome the reality, which is that CWD has implications to all deer industries throughout the world. It can potentially affect the economic viability of deer farming directly through the need to depopulate if the disease is diagnosed. But further, the impact that this disease could have on safety perceptions of the principal products from the deer industry, velvet and venison, could severely erode consumer confidence, and therefore threaten deer industries that do not even have the disease.

It is therefore essential for deer industries worldwide to work collectively in prompting awareness and research and supporting appropriate management of this disease where it exists, so that the longer-term sustainability of the global deer industry can be secured.