

Chronic Wasting Disease
Thomas J. Roffe, PhD, DVM

Presentation to Midwest Association of Fish and Wildlife Agencies
15 July 2002; All numbers and data as of 1 July 2002.

Introduction

1. Chronic Wasting Disease (CWD) is a transmissible spongiform encephalopathy (TSE) of deer and elk.
2. Several TSEs are known in other species including Bovine Spongiform Encephalopathy (BSE) (or “mad cow disease”) in cattle. Scrapie in sheep and goats is one of the oldest, and may have been the origin for BSE.
3. BSE has been linked to disease in other species and humans. In humans, the disease is called variant Creutzfeld-Jakob Disease (vCJD).
4. CWD is not known to affect humans, however because of interspecies transmission of BSE and multiple cervidae species affected the effectiveness of “species barriers” is unknown. Research is underway to assess CWD transmission to non-human primates.

What is it?

1. TSEs are diseases of the central nervous system that result in neuron death. CWD follows this pattern.
2. The diseases are associated with accumulation of abnormal, protease-resistant prion protein. All mammals have cellular prion protein, but the agent associated with TSEs causes a change in cellular prion protein that changes this normal protein to the abnormal, resistant form.
3. The transmissible agent is unknown, however the leading theory is that the abnormal protein itself is responsible for the transmission. Others believe that a bacterium, virus or virus-like particle is responsible for transmission but it has yet to be discovered.
4. Characteristics of the transmissible agent are that it apparently lacks genetic material, provokes no host immune response, and certain host genotypes are more susceptible to infection.
5. In CWD transmission is horizontal. The agent resists environmental degradation and survives for prolonged periods.
6. Incubation is unknown but prolonged. 3 year incubation periods have been documented. Shorter incubation periods also found, especially in white-tailed deer. The mechanism of shedding is unknown but appears to occur before overt disease is expressed.
7. Clinically animals may appear “wasted”, have abnormal behavior (depressed, dull, aimless wandering, or excitability) and excessive salivation.
8. Gross pathology is not specific and may include poor nutritional condition and aspiration pneumonia. Presumably the aspiration pneumonia results from neurologic disease and failure to properly ingest food.
9. Species affected include mule deer, elk, white tailed deer. Black tailed deer and deer hybrids are also reported to be susceptible.

Diagnosis

1. Several diagnostic techniques are available, but the standard is histopathology of specific areas of the brain stem (the obex).
2. Histopathological appearance under standard stains shows holes (overall spongy appearance) where neurons once were.
3. Immunohistochemistry, which stains for the abnormal protein, is a frequent adjunct to histopathology and confirmatory.
4. Other tests, such as western blot and ELISA, are also used or being tested for their validity at detecting CWD.
5. Only one antemortem test, based on tonsillar biopsy subjected to the immunohistochemistry, exists. This test works only in deer.

History and Progression

1. First discovered in Colorado research facility in mule deer in 1967.
2. Not known to be a TSE until 1978.
3. 1981 first case in free-ranging wildlife in Wyoming discovered.
4. 1996 first case in captive elk in Saskatchewan. Several other states and provinces followed shortly thereafter:
 - a) 1997 South Dakota
 - b) 1998 Nebraska and Oklahoma
 - c) 1999 Montana and Colorado
 - d) 2001 Kansas
 - e) 2002 Alberta
5. Epidemiologic links among these game farms demonstrate disease movement from location to location by animal relocations.

Current Status

1. Saskatchewan
 - a) 1996 – 2002 41 game farms found affected
 - b) Almost 8000 animals killed
 - c) About 200 found affected
 - d) Over \$19M US spent on indemnity and cleanup
 - e) 3 wild, free-ranging mule deer found affected
2. Colorado
 - a) Endemic since 1967
 - b) Prevalences as high as 15% in mule deer and 1% in elk in some hunting districts
 - c) Shipments from affected premises went to many states
 - d) Last fall declaration of a national disease emergency permitted indemnity payments. 9 affected game farms depopulated winter 2001-2002.
 - e) 16 additional game farms in endemic area bought out. 2 found affected.
 - f) 2 others remained in operation, 1 recently broke with CWD.
 - g) Recent extensions to the west slope (Routt Co.) and southward into Boulder and east of Denver.
 - h) Mandatory testing of hunted deer and elk taken in the endemic area.
3. Nebraska

- a) Game farms depopulated
- b) Extension into Kimball Co (3 mule deer) seen as natural progression from endemic area. Further east and north progression to Cheyenne Co and Scott's Bluff Co in white-tailed deer.
- c) Northern Sioux Co game farm outbreak. 51% of white tailed deer inside the fence were positive, almost 8% outside the fence positive.
- 4. Wisconsin
 - a) New outbreak in 2002, in southern Wisconsin.
 - b) White tailed deer, total of 18 positive.
 - c) No game farm connection known.
 - d) Depopulation is being attempted. Depopulation and adjacent herd reductions will result in removal of up to 120,000 deer.
- 5. Kansas, Oklahoma and Montana
 - a) Each with a single affected game farm. All depopulated.
- 6. South Dakota
 - a) Depopulated 7 game farms
 - b) Single free-ranging white tailed deer positive. An additional animal inside a game farm positive.
- 7. New Mexico
 - a) Most recent new episode, reported June 2002.
 - b) Single mule deer
 - c) Over 500 miles from the endemic area
 - d) No known game farm connection
 - e) Animal Health Emergency declared.

What to do

- 1. National Task Force
 - a) USDA: APHIS, FS, CREES, ARS
 - b) USDO: FWS, NPS, USGS, BLM, BIA
 - c) States: AZ, CO, IA, LA, MI, NE, SD, WI, WY
 - d) Developing national plan address communication, technical information, diagnostics, disease management, research and surveillance.
 - e) Budget is being re-worked but is in the millions.
 - f) Number one state effort – prevention!
 - g) Regulatory efforts of states
 - I. Joint regulatory control by agriculture and natural resources in majority of states.
 - II. 49 states have CWD specific regulations
 - III. 26 states regulate movements from endemic areas while 21 states totally forbid interstate movement of cervidae.
 - IV. 31 states developing new or additional CWD regulations
 - V. 23 states test captive cervidae, 11 states developing such regulations
 - VI. 37 states test wild cervidae, 6 others developing such programs
 - h) State actions
 - I. Cooperation among state agencies within state and regionally
 - II. Cooperation with other land managers in adjoining districts

- III. Focus on protection of resources rather than eradicating disease
- IV. Develop internal expertise and point of contact for agency people. This POC maintains currency in CWD issues.
- V. Develop both passive and active monitoring and surveillance programs
- VI. Strictly manage wildlife relocations and cervid movements
- VII. Develop field protocols for agency personnel to handle public, press and disease monitoring.