

Surveillance for Human Transmissible Spongiform Encephalopathies in a State with Endemic Chronic Wasting Disease

W John Pape¹, Ken Gershman, MD, MPH¹

Neurological Deaths Study Group: Samantha MaWhinney, ScD.²; W John Pape¹; Jeri E Foster²; C Allen Anderson, MD²; Patrick J Bosque, MD³; Michael W Miller, DVM, PhD.⁴

¹Colorado Department of Public Health and Environment, ²University of Colorado Health Sciences Center, ³Denver Health Medical Center, ⁴Colorado Division of Wildlife

BACKGROUND

- Chronic wasting disease (CWD) is a transmissible spongiform encephalopathy (TSE) of cervids (mule and white-tailed deer, elk)
- First described in Colorado as a TSE in 1977, CWD is endemic in defined areas of Colorado with infection rates of <1% for elk and an average of 4-5% for mule deer (range <1 – 15%)
- The 1996 report of variant Cruetzfeldt-Jakob disease (vCJD) associated with exposure to bovine spongiform encephalopathy lead to questions of the human disease risk from exposure to CWD-affected animals.

OBJECTIVES

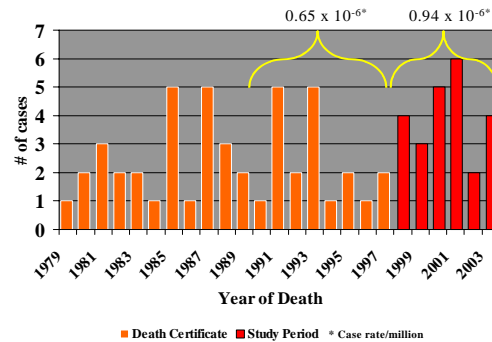
- Document the incidence and pathology of human TSE in Colorado and assess the epidemiological evidence of the potential human health risk associated with exposure to CWD.

METHODS

- Descriptive epidemiology based on human TSE as a physician reportable condition and case investigations.
- Obtain autopsy specimens, with family consent, for examination at the National Prion Disease Pathology Surveillance Center.
- Conduct epidemiological studies
 - Study 1: Analyze the incidence of neurological deaths in 7 counties with endemic CWD versus non-endemic counties (manuscript in preparation).
 - Study 2: Compare hunter license records against state and national death certificate indexes to ascertain TSE cases (study initiated Oct 2003).

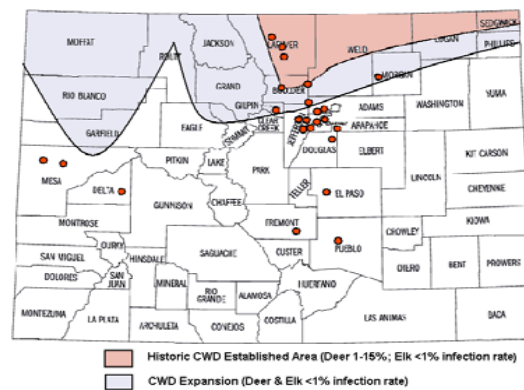
RESULTS

Reported Cases of Cruetzfeldt-Jakob Disease, By Year of Death, Colorado, 1979 - 2003



- Twenty-eight cases identified from 1998 – 2003; 24 Colorado patients and 4 out-of-state (KS, NC, NE, WY).
- Gender: 13 male, 11 female
- Age: median 65.5 yrs (range 40-81 yrs)
- Duration of illness (n=14): median 119 days; range 37 – 511 days

Human TSE Cases by County of Residence at Death, Colorado, 1998-2003 (n=24)



- Atypical Cases
 - 40 year-old with onset 7 years post C1-C2 laminectomy and craniotomy with implantation of a dura mater graft. (Hanna, L, et al. Cruetzfeldt-Jakob disease after receipt of a previously unimplicated brand of dura mater graft. *Neurology*. 2001;56:1080-83)
 - 54 year-old with 511 day illness duration and onset 10 years post L5-S1 laminectomy.
 - 25 year-old with Gerstmann-Straussler-Scheinker syndrome (mutation at codon P102L).
- Cases with Potential CWD Exposure
 - 61 year-old with lifelong consumption of deer from endemic area. Pathology consistent with sporadic CJD (M/M type 1).
 - 64 year-old hunter who moved to Colorado 6 years prior to onset. Harvested two deer during this period; both tested negative for CWD. Brain biopsy consistent with sporadic CJD (M/M type 1), family declined autopsy.
 - 53 year-old native Colorado hunter. Hunting limited to southern Colorado in areas where CWD has not been found. Family declined autopsy.

Study 1: Neurological Death Study

- Assessed death certificates from 1979 –2002 (n=506,335) for CJD and 52 other neurological diagnosis codes that could represent TSE cases.
- Local residents represented 74% (31,377/42,665) of deer hunters in the seven CWD endemic counties based on Division of Wildlife records (1999-01).

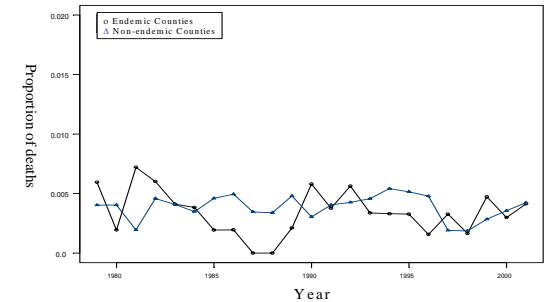
Risk Adjusted* Odds Ratio for CWD Endemic County

Covariant	CJD Diagnosis N=65
CWD Endemic County	p=0.5870
Yes	1.0 (reference group)
No	0.82 (0.41, 1.66)

* covariates: age, sex, marital status, death year, ICD-10, season

- Analysis of restricted (n=29) and broad (n=53) neurological diagnoses codes were also not statistically significant.

Proportion of Neurological Deaths by CWD Endemic County Over Time, Restricted Diagnosis Code and Age (12 –55)



LIMITATIONS

- Conducting surveillance for what would be an extremely rare subset of cases of a rare disease.
- If CWD could be transmitted to humans, the resulting clinical presentation, pathology and epidemiological characteristics are unknown.
- Epidemiological methodologies have limited power to detect rare events. The neurological study of >500,000 records over a 22 year period would detect an event occurring at the rate of approximately 30-50 cases per million.

CONCLUSIONS

- No increase in the incidence or any unusual clinical or pathological presentations of human TSE cases in Colorado has been detected.
- There was no detectable difference in deaths from CJD or related neurological diagnoses in counties with endemic CWD compared to the rest of the state.
- Results consistent with no link between CWD and human TSE, but do not preclude a rare event that may be difficult to detect.