

Chronic wasting disease

Chronic wasting disease is a fatal disorder of deer and elk that causes deterioration of the nervous system and significant weight loss. It was first noticed in 1967 in deer near Fort Collins, Colorado. The disease has now been detected in animals in the Midwest and Rocky Mountain regions of the United States as well as in some Canadian provinces, but its extent is extremely difficult to gauge accurately and is probably expanding. Known locations where the disorder currently occurs include Colorado, Illinois, Kansas, Minnesota, Montana, Nebraska, New Mexico, Oklahoma, South Dakota, Wisconsin, and Wyoming as well as Alberta and Saskatchewan.

An infection caused by proteins

Chronic wasting disease is the latest example of a prion disease to raise concern that it may, like mad cow disease, have the potential to move from animals to people. Prion (pronounced PREE-awn) diseases are a bizarre group of disorders that develop when a protein normally present in nerve cells acquires an abnormal three-dimensional structure. The abnormal shape gradually induces the same abnormality in the protein throughout the brain, causing increasing damage and, ultimately, death.

Other prion diseases include mad cow disease in cattle, Creutzfeldt-Jakob (pronounced CROYTZ-felt YAH-cawb) disease in humans, and scrapie (pronounced SCRAY-pee) in sheep and goats.

The abnormally folded proteins that initiate the destruction can arise spontaneously for unknown reasons, as happens with most Creutzfeldt-Jakob cases. In chronic wasting disease and some other prion diseases, the prions function as infectious agents and can transmit the disorder from one individual to another. This property is the source of the term “prion,” shorthand for “proteinaceous infectious particle.”

Jumping from species to species

Some prion diseases have traveled from one species to another, a process called “jumping the species barrier.” Many experts believe that mad cow disease developed in the United Kingdom primarily from use of cattle feed that contained byproducts from scrapie-infected sheep. Other experts believe that the disorder was spread by animal byproducts in feed but that it originated in cattle. Consumption of beef containing residue from brain or spinal cord or use of other products derived from “mad cows” in turn caused a human disorder called variant Creutzfeldt-Jakob disease (vCJD). The human disorder has affected more than 100 individuals in Europe, chiefly in the United Kingdom.

The mad cow epidemic has generated concern that chronic wasting disease could spread from deer and elk to farm animals or to human beings. Although there is no evidence that either type of transmission has taken place, several states and one Canadian province have initiated programs to eliminate affected animals. It is not yet known how chronic wasting disease spreads from animal to animal; current theories of transmission focus on nose-to-nose contact, saliva and waste products.

Because of these uncertainties, some experts urge caution in butchering of deer and elk or consumption of their meat. The state of Colorado, for example, urges hunters not to shoot for personal consumption any animal that appears sick or abnormal in any way, to wear gloves while butchering, and not to consume any part of the brain and spinal cord or any meat from an animal that may have the disorder. Other states suggest similar precautions. The World Health Organization, the health agency of the United Nations, recommends against using any part of an animal that may have chronic wasting disease in human or animal food. Many U.S. states offer testing programs, in most cases voluntary, to determine if a carcass is infected.

Searching for answers

Chronic wasting disease experts from around the world convened in Denver in August 2002 to share information about the best strategies for understanding and dealing effectively with the disorder. One closely watched study is testing whether the disorder can infect squirrel monkeys, which are, like other primates, close biological relatives of human beings. Results of this study, which are anticipated in a few years, are expected to provide the best indication to date on whether the disorder can infect humans. Mad cow disease has proved highly infective to a variety of species in animal studies. However, molecular studies so far suggest that it would be difficult for chronic wasting disease to jump to humans.

At the 55th Annual Meeting of the American Academy of Neurology in April 2003, researchers from the Colorado Department of Public Health and the University of Colorado reported results of two autopsies in individuals whose illness had raised concern that they might have contracted a prion disease from exposure to deer or elk meat. One individual lacked evidence of prion disease but did have Alzheimer pathology. The other had abnormal prion proteins but turned out also to have a gene mutation associated with an inherited form of Creutzfeldt-Jakob disease. The researchers concluded that neither of these cases offered evidence linking prion disease in deer or elk to human prion disorders.

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For more information, please see:

A June 2004 article from the U.S. Centers for Disease Control and Prevention (CDC) Emerging Infectious Disease series, posted at:
<http://www.cdc.gov/ncidod/EID/vol10no6/03-1082.htm>

A list of contact information for state wildlife agencies is provided by the Chronic Wasting Disease Alliance at:
<http://www.cwd-info.org/index.php/fuseaction/links.main>

The Alzheimer's Association, the world leader in Alzheimer research, care and support, is dedicated to finding prevention methods, treatments and an eventual cure for Alzheimer's.

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