Prion Diseases

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With the rise in reports of Chronic Wasting Disease (CWD) in Tennessee deer, I have been asked several questions lately if CWD can be transferred to cattle. This is considered a prion disease, and there are several misconceptions concerning this disease. I thought this would be an opportunity to briefly cover prion diseases that affect humans and animals.

Before we move on any further, I want to also make sure everyone understands this misnomer. Some will refer to Johne's disease as chronic wasting disease. Johne's is a bacterial disease of cattle that is fecal-oral transmitted to a calf less than 48 hours old. Johne's is not the same as Chronic Wasting Disease of deer.

What is a Prion Disease?

Prion diseases or transmissible spongiform encephalopathies (TSEs) are a family of rare progressive neurodegenerative disorders that affect both humans and animals. They are distinguished by long incubation periods, characteristic changes associated with neuronal loss (brain & spinal cord tissue), and a failure to induce inflammatory response.

The causative agents of TSEs are believed to be prions. The term "prions" refers to abnormal, pathogenic agents that are transmissible and are able to induce abnormal folding of specific normal cellular proteins called prion proteins that are found most abundantly in the brain. The abnormal folding of the prion proteins leads to brain damage and the characteristic signs of the disease. Prion diseases are usually rapidly progressive and always fatal.

Chronic Wasting Disease

Chronic wasting disease (CWD) is a prion disease that affects deer, elk, reindeer, sika deer and moose. It has been found in some areas of North America, including Canada and the United States, Norway and South Korea. It may take over a year before an infected animal develops symptoms, which can include drastic weight loss (wasting), stumbling, listlessness and other neurologic symptoms. CWD can affect animals of all ages and some infected animals may die without ever developing the disease. CWD is fatal to animals and there are no treatments or vaccines.

To date, there have been no reported cases of CWD infection in people. However, animal studies suggest CWD poses a risk to some types of non-human primates, like monkeys, that eat meat from CWD-infected animals or come in contact with brain or body fluids from infected deer or elk. These studies raise concerns that there may also be a risk to people. Since 1997, the World Health Organization has recommended that it is important to keep the agents of all known prion diseases from entering the human food chain.

Bovine Spongiform Encephalopathy

BSE is an unusual prion disease in that the time between an animal's exposure to the disease and the onset of clinical signs normally ranges from four to five years. Animals with BSE may show a number of different signs including nervous or aggressive behavior, abnormal posture, lack of co-ordination or difficulty in rising from a lying position, decreased milk production, and weight loss despite an increased appetite. These symptoms may last for a period of two to six months before the animal dies.

It is believed that the spread of this disease in cattle was caused by feeding protein products made from infected cattle or sheep. As of October 26, 2009, a regulation issued by FDA in April 2009 came into effect establishing an enhanced BSE-related feed ban in the United States. This enhanced ban will further harmonize BSE feed control measures in the U.S. In addition, FDA continues to enforce its important 1997 mammalian-to-ruminant feed ban through its BSE inspection and BSE feed testing programs.

Creutzfeldt-Jakob disease

Creutzfeldt-Jakob disease (CJD) is a rare, degenerative, fatal brain disorder. It affects about one person in every one million per year worldwide; in the United States there are about 350 cases per year. CJD usually appears in later life and runs a rapid course. Typical onset of symptoms occurs at about age 60, and about 70 percent of individuals die within one year.
There are three types of CJD.

- *Sporadic CJD*
  The disease appears even though the person has no known risk factors for the disease. this is by far the most common type of CJD and accounts for at least 85 percent of cases.
- *Hereditary CJD*
  The person may have a family history of the disease and test positive for a genetic mutation associated with CJD. About 10 to 15 percent of cases of CJD in the United States are hereditary.
- *Acquired CJD*
  The disease is transmitted by exposure to human brain or nervous system tissue, usually through certain medical procedures. There is no evidence that CJD is contagious through casual contact with someone who has CJD. Since CJD was first described in 1920, fewer than one percent of cases have been acquired CJD.

What about our cattle?

Because CWD is a prion disease, concerns have lingered that (CWD) in deer and other cervids could eventually could spread to cattle. So far though, there has been no strong evidence that CWD can cross species barriers to affect cattle through typical environmental exposure, or for that matter, humans who consume meat from infected cervids. New results from a long-term exposure trial now support the belief that the lethal prion disease is highly unlikely to spread to cattle.

In a 10-year trial, researchers exposed groups of cattle to high levels of CWD prions. The researchers used two methods and in different locations in Colorado, Wyoming and at the USDA's National Animal Disease Center in Ames, Iowa. One group of 12 calves received an initial oral dose of brain material from CWD-infected mule deer, with five un-inoculated cattle serving controls. Two other groups of cattle spent 10 years housed in outdoor pens with CWD-infected mule deer or elk, with continuous exposure through feed, water and direct contact.

At the end of the trial, the researchers euthanized and examined all the surviving cattle. None of the cattle showed any sign of CWD or any neurological pathology during the trial period or at necropsy.

As with other transmissible spongiform encephalopathy (TSE) diseases, CWD can develop slowly, with exposed animals remaining healthy for years before showing clinical signs. Thus, a long-term trial using environmental exposure similar to what occurs in the field, helps answer critical questions about the potential for the disease to cross species barriers.

In a separate study, a mixture of brain/CSF from CWD infected cervids was injected directly into the brain and spinal cords of cattle. Two calves out of 14 did develop clinical signs, but this was not considered a significant amount. This rate of transmission after intracranial inoculation suggests that risk of transmission through other routes is low.

CWD is a disease that certainly should be kept on the radar as research continues to the transmission of the disease. If you have any questions concerning CWD, please contact your veterinarian, Extension agent, or myself at, lstrick5@utk.edu, or 865-974-3538.

References:

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