Report to the Boards of Health

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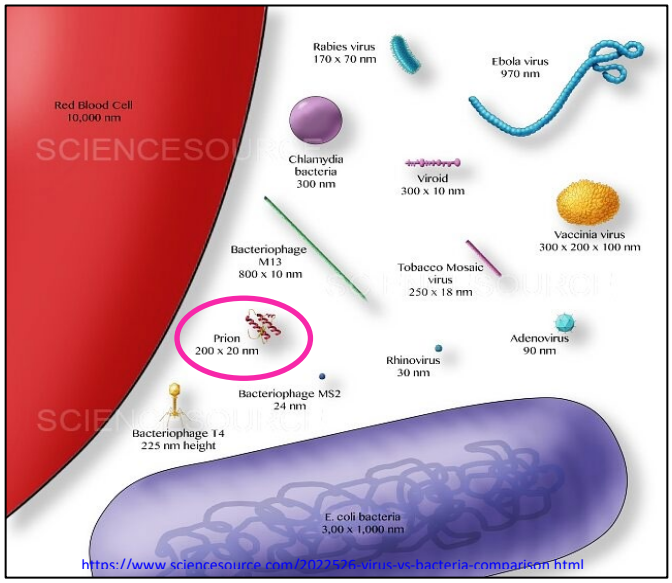
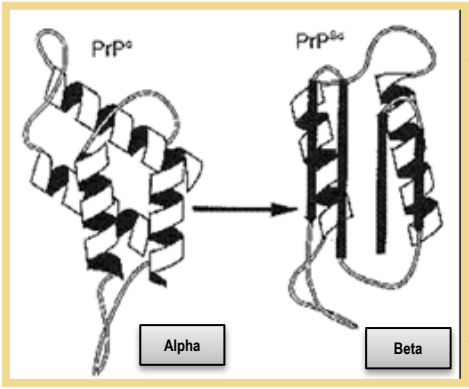
Mid-Michigan District Health Department, Wednesday, January 24, 2024
Central Michigan District Health Department, Wednesday, January 24, 2024
District Health Department 10, Friday, January 26, 2024

Prion Disease

A prion is an abnormal protein that can be infectious (spread to others) and be pathogenic (cause illness). Prions are not living organisms like bacteria or parasites and are different from a virus as they do not have any genetic material and do not replicate. Prions affect the brain and can trigger normal proteins to become abnormal and turn into prions. Prions do not trigger an immune response. There is no known treatment for prion diseases making these diseases fatal.





- The normal alpha structure (left) of a protein change to a beta configuration (right) to become a prion.
- When the abnormal beta protein comes in contact with a normal protein, it converts it to the abnormal beta form. Only the shape of the protein changes-the building blocks in the protein stay the same.
- Normal alpha proteins are soluble and easily broken down.
- Abnormal beta proteins are insoluble in all but the strongest solvents, and resistant to being broken down, heat, normal sterilization processes, and sunlight.

Prion diseases can affect both humans and animals. It is possible but rare that prion disease will spread to humans by infected meat products. There are six forms of disease in humans. The most common overall is Creutzfeldt-Jakob disease (CJD). Three of the human prion diseases are genetically inherited (run in families) and three are not inherited.

- Inherited Human Prion Diseases
    1. Gerstmann-Sträussler-Scheinker disease (GSS) is extremely rare and typically occurs around age 40.
    2. Fatal insomnia (FI) is also rare, causing difficulty sleeping. There is also a sporadic form of the disease that is not inherited.
    3. Familial Creutzfeldt Jakob Disease (CJD), is the less common form of this disease, accounting for 5-15% of CJD cases.

- Acquired Human Prion Diseases
    1. Sporadic, or classic Creutzfeldt-Jakob (CJD), develops suddenly without any known risk factors or cause, and typically starts around the age of 65. The duration of illness is approximately 4 to 5 months before death. The sporadic, or classic, for accounts for about 85% of CJD cases.
    2. Variant CJD, or vCJD, is an infectious type of the disease that is related to bovine spongiform encephalopathy (BSE, or "mad cow disease"). Eating meat or tissue from cows with BSE causes vCJD disease in humans. This form of the disease usually affects younger people and is fortunately very rare.
    3. Kuru was seen in areas of New Guinea. It was caused by eating human brain tissue contaminated with infectious prions. Because of increased awareness about the disease and how it is transmitted, kuru is now rare.

CJD has also been spread by contaminated tissue products and medical equipment. These types of cases are referred to as iatrogenic. Between the 1960s and 1980s, cases of CJD developed after the use of human growth hormone (hGH) derived from human pituitary glands. The pituitary glands were taken from donors that had unknowingly died of CJD. CJD has similarly been spread from dura mater grafts, corneal transplant, exposure to contaminated neurosurgical instruments, and blood transfusion where the donor had vCJD.

There are several different prion diseases in animals. Prion disease in cattle is called bovine spongiform encephalopathy (BSE, or "mad cow disease"). Prion disease in sheep, and less often goats, is called scrapie. Members of the cat family can get feline spongiform encephalopathy (FSE). Transmissible mink encephalopathy (TME) affects ranched mink. Chronic wasting disease (CWD) is a prion disease that affects cervids including deer, elk, and moose. In addition, there are also several exotic animals, such as cheetah, eland, gemsbok, greater kudo, moufflon, mule deer, nyala, ocelot, and puma that have been infected with prion disease when fed feed contaminated with BSE contaminated parts.

Prion diseases are classified as transmissible spongiform encephalopathy (TSE). They are neurodegenerative disorders which means they cause breakdown and disfunction of the neurologic system. This usually occurs rapidly and is fatal within a year of the onset of symptoms. Common signs and symptoms of CJD include dementia, behavioral changes, problems with speech and balance, and impaired concentration, memory, and judgement. Mood changes such as lack of interest in things and depression are common. Sometimes inappropriate happiness, anxiety, or unstable mood can be seen but this is less common. Sleep disturbances, mainly increased sleep, but also insomnia, are common. Some have psychotic features, especially visual hallucinations. There may be sudden, brief involuntary twitching or jerking of muscles especially if startled. Abnormal reflexes, spastic muscles, slow movements, poor muscle tone and rigid muscles may also be seen. In the end stages, many lose the ability to talk. Muscle become spasmed and some develop seizures. There is no effective treatment and supportive and comfort care is recommended.

Brain MRI is the most helpful test for diagnosis. Electroencephalograms (EEG) usually show characteristic abnormal brain waves. Cerebrospinal fluid protein markers may also be helpful. Definitive diagnosis can only be made by taking a sample of brain tissue during a biopsy or after death. The National Prion Disease Pathology Surveillance Center https://case.edu/medicine/pathology/divisions/national-prion-disease-pathology-surveillance-center/about-us at Case Western Reserve University School of Medicine Pathology Department operates the nation's clinical reference lab for prion disease and performs cerebrospinal fluid as well as genetic testing. They also offer a free-of-charge brain MRI consultation program.

CJD has been recognized since the early 1920s. It is a reportable disease in Michigan, and it is monitored nationally.

Reported cases of CJD in Michigan									
2014	**2015**	**2016**	**2017**	**2018**	**2019**	**2020**	**2021**	**2022**	**2023**
7	18	14	13	11	15	11	15	17	27

National Prion Disease Pathology Surveillance Center Cases Examined[1]					
Updated quarterly. Last updated on: September 2nd, 2023					
Year	**Prion Disease**	**Sporadic**	**Genetic**	**Iatrogenic**	**vCJD**
2014	208	185	21	1	1[2]
2015	262	243	19	0	0
2016	277	248	29	0	0
2017	266	247	18	0	0
2018	221	202	18	1	0
2019	281	259	22	0	0
2020	253	228	24	1	0
2021	248	224	22	0	0
2022	228	206	21	0	0
2023	114	101	8	0	0

[1]Listed based on the year of death or, if not available, on the year of referral; [2]Disease possibly acquired in a Middle Eastern or Eastern European country; Source: https://case.edu/medicine/pathology/divisions/national-prion-disease-pathology-surveillance-center/surveillance/tables-cases-examined

Some History of Prion Diseases:

Kuru appeared in an isolated group of remote Papua New Guinea known as South Foré in the early 1900s. This small group practiced a ritualistic form of cannibalism that involved family members eating their loved ones after death. They felt this helped them to take in the spirit of their ancestors and it also allowed them to ingest a rich source of protein which was difficult to get in the area. Typically, the women and children ate the brain, and the men ate the muscle tissue. Only the gallbladder was left uneaten. Kuru is believed to have been transmitted among the people through this cannibalism. As women and children ate the most infectious parts of the body, it is not surprising they were affected with kuru about 5 times more often than men. The people of the South Foré believed kuru was the result of sorcery and that victims were chosen because of some real or imaginary faults. There were many elaborate ways that the families of kuru victims tried to identify and subsequently kill (called "tukabu", or ritualistic form of vendetta) a suspected sorcerer if they could not bribe or intimidate him to release a victim from the kuru spell. Between 1957 and 1968, over 1,100 of the South Foré died from kuru. Since the 1970s, kuru has essentially disappeared since the end of cannibalism.

Bovine spongiform encephalopathy (BSE, or "mad cow disease") was first found in 1986 in the United Kingdom. It peaked in the UK in January 1993, with more than 1,000 cases being reported weekly. Millions of cattle in the UK were slaughtered in an effort to stop the spread BSE. The UK banned the use of meat and bone meal products from ruminants (animals such as cows, sheep, goats, and buffalos) in cattle feed. In 1989, the USDA banned the importation of live ruminants and most ruminant products from countries that were known to have BSE. In 1997, Canada and the FDA of the United States instituted a ban on feeding ruminant meat, bone meal, and other ruminant proteins, back to ruminants. Additionally in 1997, the U.S. extended its ruminant import ban to all of Europe regardless of BSE status.

Despite these efforts, on May 20, 2003, BSE was diagnosed in a cow in Alberta Canada. Canada had a prior case of BSE in 1993, in a single cow imported from the UK. As of 2023, seven cases of BSE have been found in the U.S. in total. The first case was detected in 2003 in Washington State in a dairy cow imported from Canada and confirmed as classical BSE. The other five cases were confirmed as atypical BSE forms. Atypical BSE can happen

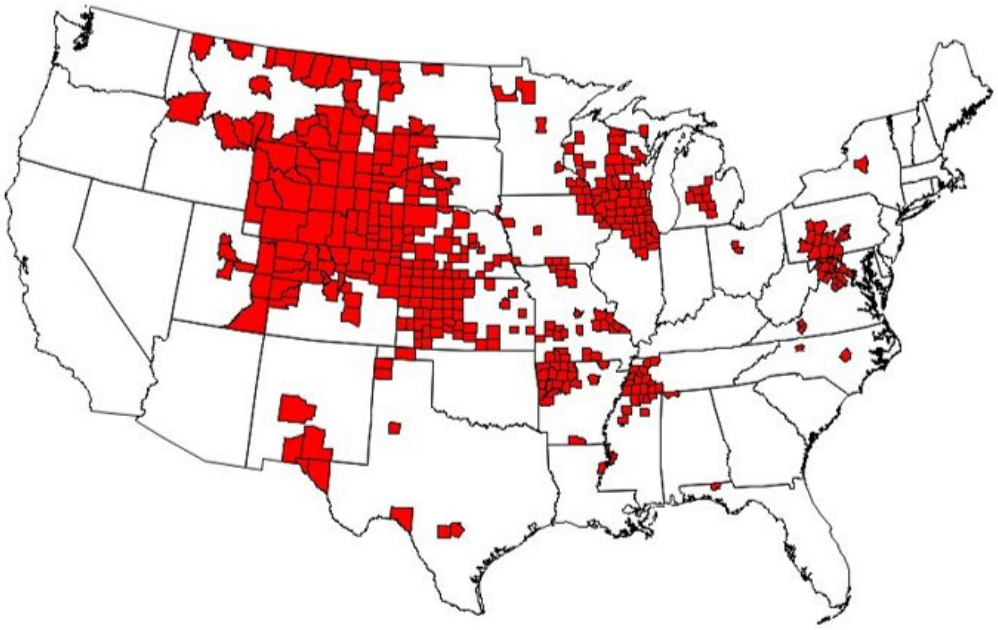
rarely and spontaneously in any cattle populations, particularly in older cattle. The second case was in 2005 in a 12-year-old beef cow in Texas. The third, in 2006, was a 10-year-old beef cow in Alabama. The fourth, in 2012, was a 10-year-old dairy cow in California. The fifth case, in 2017, was an 11-year-old beef cow in Alabama. The sixth case, in 2018, was a 6-year-old mixed breed beef cow in Florida. Case seven was announced by the USDA May of 2023 in an approximately five-year-old or older beef cow in South Carolina.

The first confirmed case of variant Creutzfeldt Jakob Disease (vCJD) (human form of BSE) was diagnosed in the UK in March 1996. vCJD occurs by eating affected cattle products, primarily brain and spinal tissue, infected with BSE. There may be people that are more susceptible to getting vCJD. The incubation period for vCJD, the time from infection to signs of illness, is likely to be many years or decades so a person who develops vCJD could have eaten an infected product or products many years earlier. In the UK, the variant form of CJD mainly affected young people with the average age at death 28 years. This is much younger than the classic form of CJD which usually affects people at an average age of 65 years. The length of illness for vCJD is much longer, lasting on average 14 months, while classic CJD lasts 4 to 5 months. As of 2019, 232 people worldwide are known to have become sick and died with vCJD.

Scrapie was first recognized in sheep in Europe more than 250 years ago and has since been reported throughout the world. Only Australia and New Zealand are considered free of scrapie. The first case of scrapie in the United States was diagnosed in 1947 in a Michigan flock. The flock owner had been importing sheep of British origin through Canada. Since then, scrapie has been diagnosed in more than 1,000 flocks in the United States.

Chronic wasting disease was first recognized in 1967 as a clinical wasting syndrome leading to chronic weight loss and death in mule deer in a wildlife research facility in northern Colorado. In 1978 it was identified as a transmissible spongiform encephalopathy (TSE) in both captive herds and in free ranging deer and elk. There is no known relationship between CWD and any other TSE diseases of animals or people. There has been no documented case of CWD being transferred to humans and at this time it is not felt that CWD causes disease in humans.

Chronic Wasting Disease Among Free-Ranging Cervids* by County
United States, November 2023



www.Michigan.gov/CWD
Counties with previous positive detections of CWD (as of 12/2023)

- **Upper Peninsula**
  - Dickinson
- **Lower Peninsula**
  - Clinton
  - Eaton
  - Gratiot
  - Hillsdale
  - Ingham
  - Ionia
  - Isabella
  - Jackson
  - Kent
  - Midland
  - Montcalm
  - Ogemaw

As of November 2023, there were 414 counties in 31 states with reported CWD in free-ranging cervids.
This map is based on the best-available information from multiple sources, including state wildlife agencies and the United States Geological Survey (USGS). https://www.cdc.gov/prions/cwd/occurrence.html
*Include white-tailed deer, moose, elk, etc.

Recommendations:

1. Though rare, be aware of prion diseases and the impact they can play on human and animal health as well as their public health impact. https://www.cdc.gov/prions/index.html.
2. Help prevent the spread of chronic wasting disease in our deer population. Find more information at https://www.canr.msu.edu/chronic-wasting-disease/news and https://cwd-info.org/cwd-the-basics/ .

Sources

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