

Prion Disease Fact Sheet

1. **What is a Prion?** - The word "*prion*" is an abbreviation for a "*proteinaceous infectious particle*." Specifically, a prion is a malformed protein capable of causing other normal prion proteins upon contact to also miss-fold (change their 3-D shape) and, thereby, self-replicate in chain reaction like process.

a. The key to understanding prion disease is an understanding that the 3-D shape of all proteins is critical to their normal function. The abnormally shaped prion protein serves no function and is not easily removed. Prions accumulate exponentially in the brain cell causing progressive brain damage, gait and balance disturbances, difficulty swallowing, weight loss, behavioral changes, and death.

b. When viewed at autopsy, stained tissue slides from the brain are filled with characteristic microscopic holes created by the excess accumulation of the abnormal prion protein. Dr. Stanley B. Prusiner received the 1997 Noble Prize in Medicine for his discovery of prions and the diseases they cause.

2. **How is a Prion Disease different from Viral and Bacterial Diseases?** - Viruses and bacteria are microorganisms that contain genetic material. They do not generate spontaneously. In contrast, Prion Disease is caused by a change in shape of a cellular protein. The resulting pathogenic prion proteins begin to recruit and change normal proteins into an abnormal shape. In older humans, this occurs at a rate of 1/1,000,000 human cases per year. The abnormal prion protein can also come from an outside source.

3. **Where are normal Prion proteins found?** - Normal prion protein is widely distributed throughout the body, but has its highest concentrations in brain, nerve, and related tissue.

4. **What is a Prion Disease?** - Prion diseases or transmissible spongiform encephalopathies (TSE's) are a family of rare progressive neurological disorders that affect both humans and animals. They have long incubation periods (period of time between infection and observable disease), and characteristic spongiform (sponge-like) changes in the brain. These diseases are now grouped together according to whether they are sporadic, inherited, or acquired.

- a. The first TSE was identified in sheep in 1759. Since afflicted sheep would scrape their sides along the fences of their pens the condition was commonly referred to as scrapie. Under a microscope, the brains of infected animals were sponge-like in appearance.
- b. In the 1920's, two separate reports appeared describing a neurological disease in humans presenting with brain dysfunction similar to that seen in scrapie. That human disease became known as Creutzfeldt - Jakob disease (CJD).
- c. In 1956 and 1957, another form of TSE called Kuru reached epidemic proportions through the practice of cannibalism among the Fore people of Papua New Guinea. 2
- d. In the mid-1980's, Great Britain experienced an outbreak of TSE among cattle. The brains were sponge-like in all afflicted cattle, and the symptoms included exaggerated behaviors - hence, the term "mad cow disease" was coined for Bovine Spongiform Encephalopathy (BSE). In 1996, human disease associated with BSE was reported in Great Britain. This disease is known as variant CJD (vCJD).
- e. Recently, there has been concern over the rising incidence of a prion disease in deer and elk in North America called Chronic Wasting Disease (CWD).

5. Should I be concerned about Prion Diseases?

- a. At this time there is no vaccine and/or effective cure for prion-associated diseases in any species, including humans.
- b. Prion diseases are rapidly progressive and uniformly fatal.
- c. There is much information that is unknown about the various types of the disease.
- d. Prion diseases generated increased public health concern after an outbreak of BSE occurred in several European countries and subsequent scientific evidence clearly indicated its transmission to humans.
- e. CWD prions are different in that they are transmitted animal to animal by contaminated feces, urine, and saliva. Deer habitat is therefore expected to remain infectious for years.
- f. To date, there has been no known human illness associated with CWD infected deer or elk.

6. For more information: <http://www.cdc.gov/ncidod/dvrd/prions/>

This fact sheet provides general information. Please contact your physician and/or veterinarian for specific clinical information related to you or your animal.

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