

Adopt a Pathogen  
Bovine Spongiform Encephalopathy (BSE)  
Creutzfeldt Jakob Disease (CJD)

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# Chapter 1

## At a Glance

### 1.1 Morphology and Size

BSE and human variation called Creutzfeldt-Jakob disease is caused by the presence of abnormal prion  $\text{PrP}^{\text{Sc}}$ . Both the normal and abnormal prion are composed of 253 amino acids; the difference between the two arises when observing how the protein is folded. The normal prion has a function of folding other proteins, its abnormal counterpart does a similar thing but folds other similar proteins into themselves. This creates a replication loop similar to a virus.

**Note:-**

Since prions are simply proteins "sanitizing" prions is extremely difficult as the protein is not "Killed" until it is denatured. This process requires extreme sustained heat.

### 1.2 Taxonomic Data

Prions are oddly unique as they are a misfolded protein and the only occurrence of a protein replicating like a virus. Currently, the only known disease-causing prion is  $\text{PrP}^{\text{Sc}}$ . Similar to how viruses evolve into existence, prions can enter a disease-free population. This likelihood increases if organisms are frequently consuming themselves.

### 1.3 Reservoir

The process of infection occurs when a  $\text{PrP}^{\text{Sc}}$  abnormal protein enters an organism that produces  $\text{PrP}^{\text{C}}$  proteins. The abnormal protein proceeds to convert normal proteins into their counterparts. This affects the reservoir as prions can be present in an organism with neurons as  $\text{PrP}^{\text{C}}$  is a required protein with multiple functions within the neuron.

### 1.4 Mode of Transmission

Prions have been observed transmitting between neuron cells within an organism but an infected individual never becomes infectious. Prions are propagated to other individuals by the ingestion of tissue from an affected organism. In the meat industry, it was common for meat scraps to end up as feed for cattle. This created a mechanism for the spread of prions that cannot occur naturally.

### 1.5 Natural History of Disease

The first cases in North America were reported in 1993. This was shortly after the adoption of animal products into animal feed. After this relationship was discovered, both the US and Canada passed legislation banning animal tissues entering animal feed. As a result, an occasional case or outbreak will occur around once every five years.

## 1.6 Means of Diagnosis

Currently the only means of diagnosis is by brain biopsy. Often times the diagnosis is suspected by the symptoms not being explained by another more easily diagnose disease. At this point the is not cure, vaccine, or sanitizer to deal with prions.

## Chapter 2

# History and Burden

### 2.1 History of Discovery

This first case of BSE was diagnosed in 1986 in the United Kingdoms. This initial outbreak started with 2 infected cows that researchers believe became infected around 1970. As a result of the long onset of symptoms for this disease the presence of prions can go long undetected. Once the protien was identified as the cause the public became concerned with how many cows may have been infected during the time of 1970-1986. Because of the practices in the comercial beef production sector the outbreak had grown quite large.

### 2.2 History of Burden

In response to the intial outbreak the United Kingdoms implemented strong quarantine and clean up measures. Due to the resilience of the protien prions clean up efforts entailed to eliminating all cows found on farms where cases were reported. After this inital outbreak the United Kingdoms experiences the most human cross overs than any other country and continue to suffer the most burden globally. Since 1996 the UK has reported 178 instances of vCJD the second highest country is France with only 27 cases.

### 2.3 Current Global Burden Distribution

## Chapter 3

# Countermeasures

# Chapter 4

## Modeling