

## *PRIONS*



All of us have heard of “Mad Cow Disease” but how many have heard of prions? Prions are “proteinaceous infectious particles” thought to be responsible for mad cow disease as well as other diseases. Are they bacteria, viruses, or parasites? No, they are simply proteins which were never felt to be infectious like other organisms until Dr. Stanley Prusiner came along.

### **DR. PRUSINER**

Dr. Stanley Prusiner is an American Neurologist and biochemist currently the director of the Institute for Neurodegenerative Diseases at the University of California, San Francisco. He won the Nobel Prize in Medicine in 1997 for proposing that prions were the cause of bovine spongiform encephalopathy (BSE) or Mad Cow Disease as well as other diseases.

## TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHIES (TSE)

BSE is only one of the brain diseases felt to be caused by prions. Perhaps one of the oldest known TSE diseases caused by prions is “Scrapie”, the term used by Scottish shepherds for several centuries to explain the very abnormal behavior of their sheep.

### A Few Of The Other TSE's Are:

1. **Kuru** – Acquired in the past in New Guinea because of the practice of cannibalism.
2. **Creutzfeldt – Jakob Disease (CJD)** – Initially of unknown cause but can be passed on through contaminated meat or contaminated surgical instruments or tissue implants. CJD can also be inherited through a genetic mutation.
3. **Fatal Familial Insomnia** – Caused by an inherited mutation in a gene that makes the prion. It strikes families of people in midlife and disrupts their sleep cycle and eventually leads to coma and death.
4. **Chronic Wasting Disease** – This occurs in wild deer and elk in the Western United States.



## PRIONS

Prions are proteins that are unique in that they reproduce without any DNA and become infectious. When these abnormal proteins enter the brain of a human or animal they change normal proteins into these abnormal proteins and eventually cause large areas of the brain to stop functioning and the brain becomes full of holes like a sponge. Since prions are not alive like a bacteria or virus, they can be present in tissue for decades and they cannot be destroyed by boiling, alcohol, autoclaving, formaldehyde, or radiation. Thus there are no effective treatments for prion diseases and all are fatal.

## MAD COW DISEASE

Bovine Spongiform Encephalopathy (BSE) or “Mad Cow Disease” first appeared in the United Kingdom in 1986. Since that time there have been nearly 200,000 confirmed cases in cattle in the UK; BSE has also been identified in 20 European countries, Japan, Israel, and Canada. There is one reported case of BSE in the United States, probably from imported cattle from Canada that were exposed to contaminated feed.



## VARIANT CREUTZFELDT-JAKOB DISEASE

Variant Creutzfeldt-Jakob disease (CJD) is the human result of Mad Cow Disease. It first appeared in 1994 and probably resulted from the consumption of contaminated beef. Averaging about 10 to 15 cases a year, there is still concern that prions may be incubating in apparently healthy people and years from now we may see many new cases of CJD; the possibility of infecting healthy persons through surgical instrumentation also exists, since the prion is resistant to all current forms of sterilization.



AL SEAR Los Angeles Times

**ROOMING BUSINESS**

Starting with a few stools and tables, Bob McCord's Sagebrush Cantina grew within a dozen years to seat more than 800. He regularly provided free Thanksgiving dinners to the elderly.

*Bob McCord, 1938 - 2008*

## His Sagebrush Cantina gave Calabasas a boost

By VALERIE J. NELSON  
*Times Staff Writer*

**B**ob McCord, whose Sagebrush Cantina restaurant, open since 1974, became a popular destination that fueled the growth of Calabasas and its historic old town, has died. He was 69.

McCord died Monday at his Burbank home of Creutzfeldt-Jakob disease, a rare brain disorder, the restaurant announced.

"Bob is one of the pioneers

**'Bob is one of the pioneers . . . who has added to the color and success of our community.'**

— DENNIS WASHBURN,  
*a Calabasas councilman*

While on location in Arizona, he and three colleagues went up in private plane that crashed and killed two men. Two days passed before McCord, who had a broken back and ankle, was rescued. He used settlement money from the accident to start the cantina.

Janiro said the restaurant would remain open, but he would not comment on whether it would be sold.

A circus fanatic, McCord showcased some of his artifacts at the cantina. He owned Ferris wheels, carousels, steam

### SUMMARY

As of today, prion diseases are rare, with an average world-wide occurrence of one case per million people per year. Even though all scientists have not accepted the theory of prions as the cause of spongy brains, until a better theory or a slow-acting virus is found, it appears that Dr. Prusiner has discovered a new infectious agent for us to deal with.

References:

- US Food and Drug Administration (FDA)
- Genetic Science Learning Center
- Centers for Disease Control (CDC)
- The New England Journal of Medicine
- Microbe World
- National Institute of Health (NIH)