Statement from Stanley B. Prusiner, M.D., about ‘Mad Cow’ disease in the United States - January 27, 2004

Thank you, I am pleased to be here to address the Food Safety Caucus of the House of Representatives of the United States Congress about Mad Cow disease. I appear here as a concerned citizen, a loving parent, a dedicated physician specializing in Neurology, an educator who is a Professor of Neurology at the University of California, and scientist-businessman who is the Founder of InPro Biotechnology. I am also an expert on prion diseases, one of which is Mad Cow disease or bovine spongiform encephalopathy, often-abbreviated BSE.

Both Federal and State Governments now find themselves embroiled over concerns over Mad Cow disease after Secretary of Agriculture, Ann Veneman, announced on December 23, 2003, that a 6.5-year-old cow from Mabton, Washington, had been diagnosed with Mad Cow disease.

I would like to discuss five points concerning Mad Cow disease and what I believe should be done in our country to combat this malady.

1. Prions cause Mad Cow disease: First, Mad Cow disease is caused by an infectious agent that is so small that it cannot even be seen with the most powerful microscopes. These small infectious agents are called prions. Although large aggregates of prions can be studied with electron microscopes, we still cannot see the individual prions. For more than a century, viruses that can be seen in the electron microscope were the smallest known microbes. But, prions are much smaller than viruses and this extremely small size makes prions extremely difficult to kill.

2. Prion disease is always fatal: Second, prions cause severe destruction of the brain. The prion diseases of humans and animals are 100% fatal. Indeed, everyone with prion disease eventually dies. A single prion is sufficient to initiate the multiplication process that results in hundreds of prions being made followed by thousands, then millions and finally billions. It is well documented that billions of prions destroy the brain and spinal cord.

From a wide variety of biomedical investigations, we know that prions from cattle can infect humans and destroy their brains. More than 150 teenagers and young adults in Europe have died of prion disease that they contracted after eating prion tainted beef or beef products.

3. Spontaneously induced prions: Third, prions arise spontaneously. This is an extremely important concept; furthermore, the ability to arise spontaneously is a feature that distinguishes prions from viruses. Any mammal is capable of producing prions spontaneously.
In humans, the most common form of prion disease results from the spontaneous formation of prions. Despite decades of looking for prions in the environment, there is no evidence for exposure to prions in spontaneous cases of prion disease.

The initial event in an epidemic of human prion disease referred to as kuru must have been a spontaneous case of prion disease. Once kuru prions arose spontaneously, they were propagated by ritualistic cannibalism that was practiced among New Guinea natives. While halting cannibalism of dead relatives resulted in the disappearance of kuru in a small population of natives, it did not eliminate the spontaneous formation of human prions. Similarly, stopping industrial cannibalism where cattle are fed the rendered offal of other cattle has diminished the number of cattle with BSE in Britain but will not prevent spontaneous prions from being formed. Thus, while changing feeding practices for cattle will stop the amplification of prions, it will not prevent the spontaneous formation of bovine prions.

As I said, prions can develop spontaneously within any mammal. We don’t know what triggers this process but there are several reasonable hypotheses, one or more of which may eventually explain the spontaneous formation of prions.

4. The Japanese solution: Fourth, I cannot understand as the father of two daughters and the uncle of a niece and nephew why our country remains unwilling to adopt the Japanese policy of testing every cow and bull destined for consumption by humans. I have difficulty explaining to these young people that the beef in Japan is safer than that in the U.S.

The United States has the same problem that the Japan has, but the Japanese test all of the cattle that they slaughter. This issue particularly troubles children when they learn that the time from exposure to prions until the onset of neurological disease can exceed 50 years. Some New Guinea natives developed kuru more than 50 years after ingesting prions during cannibalistic feasts.

5. Prion science is new: Fifth, the science of prions is still very young. Only 25 years ago, I discovered prions and named these unprecedented infectious agents. Thus, the naysayers, who continue to deny the existence of prions, should not surprise you. A chorus of naysayers has always accompanied big changes in scientific thinking. When Galileo wrote about the planets orbiting the sun, he was imprisoned. How dare he think that the earth was not the center of universe? From the time that Einstein proposed his special theory of relatively in 1905 until his death 50 years later, the naysayers scorned him almost daily. Each week, at least two or three letters arrived at his Princeton office that declared him insane and his theories impossible. Only his death terminated this non-sense! Philip Semelweiss, a Viennese obstetrician, was eventually admitted to an insane asylum. Semelweiss was ridiculed mercilessly for proposing that his
colleagues could prevent deadly bacterial infections in mothers after childbirth if they would only wash their hands between the deliveries of newborn infants. And few believed Alfred Wagener when he proposed continental drift as a mechanism to explain the shapes and positions of the landmasses on our planet.

I recited a few instances of “scientific heresy” to place the discovery of prions in some perspective for you. For much of my career, I faced a legion of scientists who vehemently argued that prions couldn’t exist! They yelled, “prions are nonsense. They are impossible!” Twenty-five years after my discovery of prions, there remain some people who are still unable or unwilling to comprehend the novel concepts of prion biology. The famous German physicist Max Planck encountered many naysayers when he and others set forth the principles of quantum mechanics. In frustration, Planck once remarked, “a new scientific truth does not win out by convincing its opponents, rather they eventually die off and a whole generation familiar with it grows up.”

In non-scientific terms, prions must be considered new, strange and scary microbes by any measure. Twenty-five years ago, there were no prions – now the biology of prions is taught in every medical school throughout the world. Prion biology is also taught in many high schools and most colleges. Moreover, the word “prion” appears in every dictionary.

Because the discovery of prions ushered in major changes in our thinking, your duties as Congresswomen and Congressmen have and will continue to be subject to much misinformation with respect to Mad Cow disease. But I hasten to add that this is inevitable when an entirely new field of science emerges.

Despite the fact that prions were once branded scientific heresy and are now considered orthodoxy by most scholars, the naysayers still exist. This means that you and your staff will hear some opinions that are not based on the body of scientific knowledge that has been accumulated over the past quarter century. Instead, you will hear views that ignore a constantly enlarging body of scientific information that has been verified by experimental studies.

**Concluding remarks:** In conclusion, from studies over the past half-century, we know that people should not eat prions, particularly prions of human or bovine origin. I want to reiterate that the problem of prion contamination in the food supply will not disappear. If we do nothing, confidence in the safety of food supply will only continue to erode. The sooner we face the problem of prion contamination, the more easily we shall be able to contain it. Only the Japanese solution of testing every slaughtered cow or bull will eliminate prions from the food supply and restore consumer confidence. Certainly, the citizens of the most prosperous and accomplished nation on our planet deserve to eat meat that is devoid of prions.