



**EXPLORABLE**  
*Think Outside The Box*

Published on *Explorable.com* (<https://explorable.com>)

[Home](#) > Prion as an Infectious Agent

---

## Prion as an Infectious Agent

Explorable.com 17.5K reads

### 1997 Nobel Prize Medicine

Stanley B. Prusiner was awarded the Nobel Prize in Medicine or Physiology of 1997 for his discovery of Prion as an infectious agent. In his elegant series of experiments, he was able to identify Prions as the cause behind some of the common neurologic dysfunctions in animals and in humans.

Prions are protein; they have no genetic material and it is normally found in most of the cells in our body. Every person in the planet has Prions, but does that mean we are all infected by a neurologic dysfunction?



**EXPLORABLE**  
*Quiz Time!*



**Quiz:**  
Psychology 101 Part 2



**Quiz:**  
Psychology 101 Part 2



**Quiz:**  
Flags in Europe

[See all quizzes =>](#)

## Scientific Context of the 1970s and 80s

The well known infectious agents in the 1970's and 1980's are only bacteria, viruses, fungi and parasites. All the well-known infectious agents during that time have genetic materials that enable them to propagate and infest the body. The genetic material of these agents were considered foreign to human bodies and that is the reason why human immune defense mechanisms act against these agents. Researchers conducted by Prusiner added another agent into the infectious agent category in the form of prions.

## Who is Stanley Prusiner?

Stanley Prusiner was born on the 28th of May 1942 in Des Moines. He developed his interest in scientific papers when he was a high school student in Walnut Hills High School. He then studied in the University of Pennsylvania where he majored in chemistry. He finished college a cum laude and earned his A.B. in 1964. He then earned his M.D. in 1968 in the University of Pennsylvania, School of Medicine. He underwent medical internship in the University of California from 1968 to 1969. He finished his residency in Neurology from the same university in 1974.

Prusiner then became professor in multiple fields such as neurology, biochemistry, virology and biophysics in the University of California. It was during his residency, due to an admitted patient dying of a “slow virus” infection called Creutzfeldt-Jakob disease, or CJD, when he developed an interest in identifying the molecular structure of this slow virus.

## Decades of Research to Identify Prion as an Infectious Agent

The mystery behind the slow virus that caused the death of his patient gave him the push to read more about CJD and other related diseases like Kuru in New Guinea and the scrapie disease of sheep. His first step was to purify a scrapie agent wherein he expected to find small viruses but he kept finding proteins in his results but no nucleic acid. He conducted other similar experiments but the results showed only proteins and a persistent absence of nucleic acids.

The crucial step in the understanding of the novel infectious agent was Prusiner’s isolation of the protein from the infectious material which he named prion, an acronym for “proteinaceous infectious particle.” He also found that infectivity of prions are significantly reduced by chemicals that destroy protein structure, and more importantly, by antibodies to prion proteins. It was also after the isolation of the prion protein that Prusiner was able to identify the gene coding for prion protein.

Furthermore, he found that the prion protein mRNA is a product of a single host gene which he noted to be present even in uninfected brain tissues of animals. He also found that prion proteins are present in white blood cells and especially abundant in the surface of nerve cells in the brain. This initially was a very contradicting result since he was able to find the gene that causes prion infection in an uninfected specimen.

To reconcile the contradicting results, Prusiner hypothesized that the prion proteins can exist in two different forms: a normal cellular protein, which explains the presence of the prion gene, mRNA and protein in uninfected specimen, and a pathologic protein form, which explains the diseases.

Prusiner indeed found that a structural difference exists between the prion proteins in normal tissue and the prion proteins in infected tissue. He added that for a normal prion protein to become pathologic, it involves a conformational change whereby the  $\alpha$ -helical content of the normal protein diminishes and the  $\beta$ -sheet content increases. This means that prions are not

totally infectious agents; prions are also normal proteins present in most of the cells in our bodies. Prions only become infectious agents if it undergoes the mentioned conformational changes.

## Clinical Relevance

Prion as an infectious agent <sup>[1]</sup> can be considered as a relatively new development in the field of medicine. They are considered inevitably fatal due the progressive destruction of the brains of the infected individual.

Normal and healthy prion proteins can be infected if it comes in close contact with an infected prion protein. The continuous infection of prion proteins forms a thread-like structure of prion aggregates that ultimately destroys nerve cells. Brain specimens that are infected with prions have a characteristic porous and spongy appearance due to the continuous death of nerve cells in the region.

Symptoms associated with a prion infection depend on the area of the brain that is infected by the agent. If the prion infects the cerebral cortex, memory and learning is impaired. If the infected region is the cerebellum, the infection manifests as gait and coordination impairment. If the infected region is the thalamus, arousal and sleep disturbances are observed.

There are three ways by which a person can be infected with a prion disease. First is by spontaneous mutation of the prion gene that causes the conformational change of harmless prions into infectious prions. Second is by heredity; mutations in the prion protein gene can be transmitted to an individual's offspring. Lastly, infection can be transmitted if prions from an infected person or animal are ingested. Prions cannot be transmitted through air, touch or other casual contact.

---

**Source URL:** <https://explorable.com/prion-as-an-infectious-agent?gid=20975>

### Links

[1] <http://www.nature.com/nm/journal/v10/n7s/full/nm1069>