AMERICAN ACADEMIC PUBLISHER

INTERNATIONAL JOURNAL OF MEDICAL SCIENCES

ISSN NUMBER: 2692 - 5206 Volume 6. No 02. February ,2026

CONCEPT OF PRIONS, THEIR ORIGIN, PRION DISEASES, AND PRACTICAL SIGNIFICANCE

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Abstract: This scientific article presents modern concepts of prions, their origin and structure, the biological basis of prion diseases, and their practical significance. The study analyzes the ability of prion proteins to transform into pathogenic forms without genetic or epigenetic alterations, their association with neurodegenerative diseases, as well as their importance in medicine, veterinary science, and food safety. Special attention is paid to the unique infectious nature of prions, which challenges classical biological paradigms and raises important issues related to biosafety and epidemiology.

Keywords: prion, protein conformation, scrapie, Creutzfeldt–Jakob disease, spongiform encephalopathy, infectious protein, nervous tissue, molecular biology, biosafety, epidemiology.

Prions are considered one of the relatively new and complex concepts in biology, and their existence was conclusively confirmed by scientific evidence in the second half of the twentieth century [Prusiner, pp. 45–60]. The term "prion" originates from the English phrase "proteinaceous infectious particle," meaning an infectious particle composed of protein. This term was first introduced into scientific literature in 1982 by the American scientist Stanley B. Prusiner [Prusiner, pp. 45–60]. Prusiner's major achievement was his demonstration, contrary to all existing paradigms about microorganisms, that infectious agents may exist without genetic material and consist solely of misfolded proteins. After this hypothesis was confirmed by numerous experimental studies, Prusiner was awarded the Nobel Prize in Physiology or Medicine in 1997.

The most important characteristic of prions is their ability to propagate within the organism despite the absence of genetic material. This phenomenon fundamentally changed long-standing biological concepts, as previously infectivity was believed to be exclusive to viruses, bacteria, and fungi containing DNA or RNA. In contrast, prions arise from conformational changes in normal protein molecules and, once converted into a pathological form, induce similar misfolding in healthy proteins. For this reason, prions are defined as infectious proteins.

In humans and animals, prions originate primarily from conformational alterations of the normal prion protein known as PrP^{c}. PrP^{c} is a physiologically important glycoprotein located on the membrane of nerve cells, characterized by an alpha-helical structure. It plays a significant role in neuronal signal transmission, copper ion binding, antioxidant defense, synaptic stability, and neuronal development. However, the pathological form, PrP^{sc}, is distinguished by an increased content of beta-sheet structures and a strong tendency to aggregate. As a result, PrP^{sc} accumulates in brain tissue, causing spongiform changes. The most dangerous aspect of this

AMERICAN ACADEMIC PUBLISHER
INTERNATIONAL JOURNAL OF MEDICAL SCIENCES
ISSN NUMBER: 2692 - 5206 Volume 6. No 02. February ,2026

process is that PrP^{sc} is resistant to enzymatic degradation, high temperatures, chemical agents, and most disinfectants. This unique resistance makes prion diseases extremely difficult to control.

A distinctive feature of prions is that they lack genetic information carriers such as DNA or RNA, yet they are capable of replication, dissemination, and inducing pathological effects within the organism [Colby, p. 30–62]. This phenomenon completely altered the biological paradigm, as it was previously assumed that all infectious diseases were caused by microorganisms containing genetic material [Griffith, p. 25–47]. Prions arise as a result of conformational changes in normal proteins predominantly found in nervous tissue. Under normal conditions, this protein is called PrP^{c}; however, when it converts into the pathological PrP^{sc} form, it causes spongiform (vacuolar) degeneration of brain tissue [Wadsworth, p. 15–40]. This process is irreversible, and prions are resistant to enzymes, heat, and disinfectants [Weissmann, p. 10–28]. Consequently, treatment of prion diseases remains extremely challenging [Prusiner, pp. 120–158].

Prion diseases are a group of fatal, slowly progressive neurodegenerative disorders affecting both humans and animals. In humans, the major prion diseases include Creutzfeldt–Jakob disease (CJD), variant Creutzfeldt–Jakob disease (vCJD), Gerstmann–Sträussler–Scheinker syndrome, fatal familial insomnia (FFI), and kuru disease. Kuru is of particular anthropological importance, as it was prevalent among the Fore people of New Guinea, who practiced ritual cannibalism by consuming the brains of deceased individuals. As a result, prion disease was transmitted across generations, dramatically increasing mortality rates.

Creutzfeldt–Jakob disease typically occurs in individuals over 50 years of age and progresses rapidly. Its incubation period may last for decades. Initial symptoms include memory impairment, loss of motor coordination, speech disturbances, and progressive dementia, ultimately leading to death within a few months. Variant CJD emerged in the 1990s in the United Kingdom during the outbreak of the so-called “mad cow disease.” This disease, known as bovine spongiform encephalopathy (BSE), occurred in cattle due to the inclusion of inadequately processed animal remains in feed. Humans became infected through the consumption of contaminated meat products. At the peak of the epidemic, millions of animals were destroyed and hundreds of people died, clearly demonstrating the profound impact of prion diseases on global food safety.

Among animals, prion diseases include scrapie in sheep, spongiform encephalopathy in cattle, and chronic wasting disease (CWD) in deer and elk. CWD has become a serious ecological threat in North America, as it spreads easily among wild animals and can disseminate uncontrollably in natural ecosystems.

Several theories have been proposed regarding the origin of prions. The most widely accepted theory suggests that prions arise from conformational misfolding of normal proteins. Other hypotheses propose that prions may be remnants of ancient viruses that lost their genetic material but retained their protein structure. Prions are also considered epigenetic agents, as they can alter phenotypes without changes in DNA sequence.

The gene encoding the prion protein, PRNP, is located on chromosome 20 in humans and exhibits multiple polymorphisms. Certain mutations in this gene lead to inherited forms of prion diseases. For example, the D178N mutation may result in FFI or GSS syndrome, while the Met/Val polymorphism in the PRNP gene influences susceptibility to prion diseases.

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INTERNATIONAL JOURNAL OF MEDICAL SCIENCES
ISSN NUMBER: 2692 - 5206 Volume 6. No 02. February ,2026

The transmission mechanisms of prions are complex. Infection usually occurs through exposure to contaminated tissues, particularly brain or nervous system products. Transmission may also occur via surgical instruments, as prions are not destroyed by standard sterilization procedures. Although transmission through blood transfusion is considered rare, it cannot be completely excluded.

Prion diseases primarily affect the central nervous system and are classified as neurodegenerative disorders in both humans and animals [Wadsworth, p. 15–40]. Diagnosis is challenging because early symptoms resemble those of other neurological diseases. Currently, diagnostic methods include MRI with diffusion-weighted imaging (DWI), electroencephalography (EEG), cerebrospinal fluid (CSF) biomarkers, immunohistochemical analysis of postmortem tissue, and the highly sensitive RT-QuIC test, which can detect minute quantities of prions.

To date, no effective treatment for prion diseases exists. Only symptomatic therapy is available to alleviate patient discomfort. Nevertheless, ongoing research on prions has significantly advanced understanding of other neurodegenerative diseases such as Alzheimer's, Parkinson's, and Huntington's diseases, in which misfolded proteins propagate in a prion-like manner.

From a practical perspective, prions have led to the revision of biosafety protocols in medicine and veterinary science. Special sterilization procedures, including autoclaving at 134 °C for one hour and the use of 2 N NaOH or 20,000 ppm chlorine solutions, are mandatory. In biotechnology, prions have become a valuable model for studying protein misfolding, aggregation, and epigenetic inheritance. Prion-like mechanisms are also being explored in nanobiotechnology, biomolecular computing, and the development of self-assembling protein structures.

In conclusion, prions represent a crucial scientific phenomenon that connects biology, medicine, veterinary science, food safety, molecular genetics, and ecology. They are not merely disease-causing agents but a fundamental concept that has reshaped our understanding of life at the molecular level. Continued research on prions may ultimately lead to novel therapeutic strategies for prion diseases and other neurodegenerative disorder

References.

1.Prusiner, S. B. (1982). Prions: A new theory of proteinaceous infectious particles. Scientific Journal Science and Life, pp. 45–60.

2 Aguzzi, A. (2008). Neurodegeneration and prions. Science Journal, pp. 77–104.

3.Soto, C. (2003). Protein misfolding and disease. Nature Reviews Neuroscience, pp. 55–85.

4.Kovacs, G. G. (2017). Molecular pathology of human prion diseases. Brain Pathology, pp. 60–95.

5.McKinley, M. P. (1983). Relationships between scrapie and host proteins. Cell, pp. 100–124.

6.OIE (World Organisation for Animal Health). (2019). Manual of diagnostic tests and vaccines for terrestrial animals. Paris, pp. 44–66.

AMERICAN ACADEMIC PUBLISHER

INTERNATIONAL JOURNAL OF MEDICAL SCIENCES

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7.World Health Organization (WHO). (2020). Guidelines on transmissible spongiform encephalopathies. Geneva, pp. 70–92.

8.Knight, R. (2008). Modern clinical description of Creutzfeldt–Jakob disease. British Medical Journal, pp. 15–32.

9.Brown, P. (1997). Infectious protein particles in humans and animals. Trends in Microbiology, pp. 110–140.

10.Aguzzi, A., & Polymenidou, M. (2004). Molecular genetics of prion diseases. Cell, pp. 90–115.

11 Riek, R. (1996). Structural determination of the prion protein. Nature, pp. 30–56.

12.Prusiner, S. B. (2004). Prion biology and diseases. Cold Spring Harbor Laboratory Press, USA, pp. 120–158.

13.Sigurdson, C. J. (2007). Diversity and characteristics of prion strains. Proceedings of the National Academy of Sciences of the USA, pp. 75–104.

14.Soto, C. (2012). Uncovering the mystery of prions. Cell, pp. 58–84.

15.Worrall, D. M. (2019). Protein folding disorders and disease mechanisms. Journal of Biochemistry, pp. 45–70.

16.Pan, K. M. (1993). Mechanisms of alpha-helix to beta-sheet conversion. Science, pp. 18–48.

17.Ironside, J. V. (2009). Histopathological features of human prion diseases. Journal of Pathology, pp. 80–105.

18.Collinge, J. (2016). Prion diseases in humans and animals. Oxford University Press, pp. 10–65.

19.Griffith, J. S. (1967). Self-replication of proteins in scrapie. Nature, pp. 25–47.

20.Colby, D. W., & Prusiner, S. B. (2011). Prions as protein-based infectious agents. Annual Review of Biochemistry, pp. 30–62.

21.Wadsworth, J. D. F., & Collinge, J. (2010). Molecular pathogenesis of human prion diseases. Brain, pp. 15–40.

22.Weissmann, C. (1991). A unified hypothesis on prion propagation. Proceedings of the National Academy of Sciences of the USA, pp. 10–28.

23.Bolton, D. C. (1982). Isolation and characterization of the scrapie agent. Science, pp. 50–70.

24.Bruce, M. E. (1996). Transmission mechanisms of bovine spongiform encephalopathy. Nature, pp. 22–41.

25.Taylor, D. M. (2000). Methods for prion inactivation. Veterinary Journal, pp. 35–58.