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Prion Diseases

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Overview

Prion diseases or transmissible spongiform encephalopathies (TSEs) are a family of rare progressive neurodegenerative disorders that affect both humans and animals. They are distinguished by long incubation periods, characteristic spongiform changes associated with neuronal loss, and a failure to induce an inflammatory response.

The causative agents of TSEs are believed to be prions. The term “prions” refers to abnormal, pathogenic agents that are transmissible and are able to induce abnormal folding of specific normal cellular proteins called prion proteins that are found most abundantly in the brain. The functions of these normal prion proteins are still not completely understood. The abnormal folding of the prion proteins leads to brain damage and the characteristic signs and symptoms of the disease. Prion diseases are usually rapidly progressive and always fatal. [Source: CDC](#)

Resources

Centers for Disease Control and Prevention (CDC)

- [Prion Diseases](#)
- [Guidelines for Infection Control in Dental Health-Care Settings — 2003 , page 36](#)

National Institute of Allergy and Infectious Diseases (NIAID)

- [Prion Diseases](#)

National Institute of Neurological Disorders and Stroke (NINDS)

- [Creutzfeldt-Jakob Disease Information Page](#)

Fact Sheets & Information

MedlinePlus

- [Prion Disease](#)

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