

PRIONS

The New Biology of Proteins

Prion-related diseases, also known as transmissible spongiform encephalopathies (TSEs), are a group of infectious, fatal neurodegenerative disorders for which there is presently no cure, treatment, nor even a means for early diagnosis. The recent and horrific advent of Mad Cow Disease, which had been transmitted to humans through eating meat from steers sickened by bovine spongiform encephalopathy (BSE), brought prion-related diseases well-deserved international attention. Exceptionally dramatic, these diseases progressively and inexorably destroy the cognitive, motor, and sensorial skills that are the essence of human beings.

Prions: The New Biology of Proteins provides a well-organized overview of what is presently known about prion-related diseases. This comprehensive work reviews the clinical symptoms, epidemiology, and neuropathology of the disease. It focuses on evidence supporting the idea that TSEs result from an entirely novel disease mechanism, involving transmission by replication of the misfolding of a single protein in the absence of nucleic acids. Following this hypothesis, the book examines the structure, conversion, and mechanism of prion propagation and details its cellular biology. It explores the transmission of the disease, discusses the challenges involved with diagnosis, and considers the various therapeutic avenues that are presently being explored.

A cohesive volume that explores and integrates the pioneering work of many researchers, this book is authored by Claudio Soto, an internationally renowned researcher whose prolific and innovative work has led to an increased understanding of the heretical biology of prions and the development of novel strategies for the treatment and diagnosis of neurodegenerative diseases. As protein misfolding diseases are his specialty, Soto also looks at the phenomenon from a wider perspective. He examines other diseases that display folding aberrations, considers just how commonly such aberrations occur in nature, and asks readers to open their minds to consider the impact of prions on broader areas of biology, public health, and biotechnological strategies.



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