

# The Prion Protein

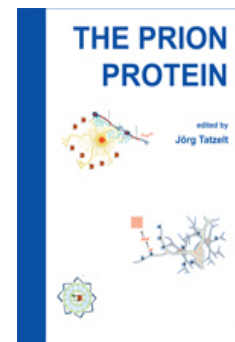
Edited by: **Jörg Tatzelt**

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**Published:** January 2010. **Pages:** 86

**ISBN:** Book: 978-1-912530-07-6 £159, \$319

**Published by:** Caister Academic Press [www.caister.com](http://www.caister.com)



A conformational transition of the cellular prion protein (PrP(C)) into an aberrantly folded isoform designated scrapie prion protein (PrP(Sc)) is the hallmark of a variety of neurodegenerative disorders collectively called prion diseases. They include Creutzfeldt-Jakob disease and Gerstmann-Stäussler-Scheinker syndrome in humans, scrapie in sheep, bovine spongiform encephalopathy (BSE) in cattle and chronic wasting disease (CWD) in free-ranging deer. In contrast to the deadly properties of misfolded PrP, PrP(C) seems to possess a neuroprotective activity. More-over, animal models indicated that the stress-protective activity of PrP(C) and the neurotoxic effects of PrP(Sc) are somehow interconnected.

In this timely book, leading scientists in the field have come together to highlight the apparently incongruous activities of different PrP conformers. The articles outline current research on cellular pathways implicated in the formation and signaling of neurotoxic and physiological PrP isoforms and delineate future research direction. Topics covered include the physiological activity of PrP(C) and its possible role as a neurotrophic factor, the finding that aberrant PrP conformers can cause neurodegeneration in the absence of infectious prion propagation, the requirement of the GPI anchor of PrP(C) for the neurotoxic effects of scrapie prions, the pathways implicated in the formation and neurotoxic properties of cytosolically localized PrP, the impact of metal ions on the processing of PrP, and the role of autophagy in the propagation and clearance of PrP(Sc). The book is fully illustrated and chapters include comprehensive reference sections.

Essential reading for scientists involved in prion research.

**Chapter 1.** Prion Neurotoxicity: Insights from Prion Protein Mutants (*Isaac H. Solomon, Jessie A. Schepker and David A. Harris*)

**Chapter 2.** Prion Protein: Orchestrating Neurotrophic Activities (*Vilma R. Martins, Flavio H. Beraldo, Glauca N. Hajj, Marilene H. Lopes, Kil Sun Lee, Marco A. Prado and Rafael Linden*)

**Chapter 3.** Autophagy, Prion Infection and their Mutual Interactions (*Andreas Heiseke, Yasmine Aguib and Hermann M. Schatzl*)

**Chapter 4.** Prion Protein and Metal Interaction: Physiological and Pathological Implications (*Neena Singh, Dola Das, Ajay Singh and Maradumane L. Mohan*)

**Chapter 5.** Targeting of the Prion Protein to the Cytosol: Mechanisms and Consequences. (*Margit Miesbauer, Angelika S. Rambold, Konstanze F. Winklhofer and Jörg Tatzelt*)

**Chapter 6.** The Role of GPI-anchored PrPC in Mediating the Neurotoxic Effect of Scrapie Prions in Neurons (*Helois E. Radford and Giovanna R. Mallucci*)

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