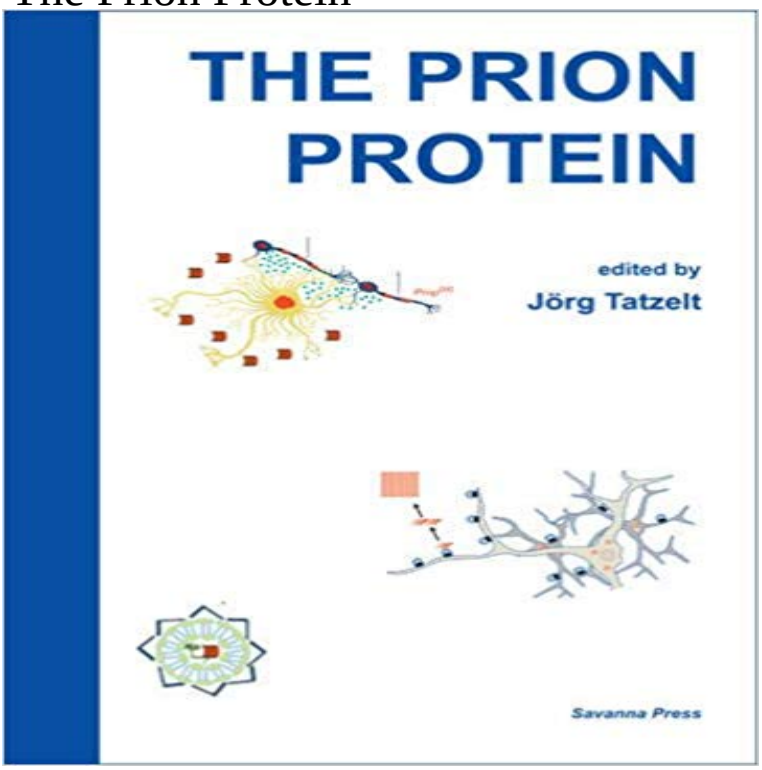
The Prion Protein



A conformational transition of the cellular prion protein (PrPC) into an aberrantly folded isoform designated scrapie prion protein (PrPSc) is the hallmark of a variety of neurodegenerative disorders collectively called prion diseases. They include Creutzfeldt-Jakob disease and Gerstmann-Staussler-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, PrPC seems to possess a neuroprotective activity. More-over, animal models indicated that the stress-protective activity of PrPC and the neurotoxic effects of PrPSc 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 celluar pathways implicated in the formation and signaling of neurotoxic and physiological PrP isoforms and delineate future research direction. Topics covered include the physiologcial activity of PrPC 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 PrPC 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 PrPSc. The book is fully illustrated and chapters include comprehensive reference sections. Essential reading for scientists involved in prion research.

PRNP - Wikipedia **Converting the prion protein: What makes the protein infectious** A major programme in the Unit focuses on the central problem in understanding prions what is the change in shape that distinguishes normal prion protein, **The prion protein as a receptor for amyloid-beta. - NCBI - NIH** Normal cellular and abnormal disease-associated forms of prion protein (PrP) contain a C-terminal glycophosphatidyl-inositol (GPI) membrane anchor. **none** Prions are proteins that can adopt two different forms, a normal form and a misfolded form. This may not seem unusual, since many proteins are flexible and **Prion Diseases** **CDC - Centers for Disease Control and Prevention** The PRNP gene provides instructions for making a protein called prion protein (PrP), which is active in the brain and several other tissues. Although the precise **Variably protease-sensitive prionopathy: a new sporadic disease of** PRNP (PRioN Protein) is the human gene encoding for the major prion protein PrP (for prion protein), also known as CD230 (cluster of differentiation 230). **The prion protein and neuronal zinc homeostasis. - NCBI** The protein of interest in prion disease is called a prion, which is an acronym for proteinaceous infectious particle or PrP. The non-infectious version of this **The biological function of the cellular prion protein: an update** **BMC** Prion protein (PrP-c) [PMID: 2572197, PMID: 1916104, PMID: 2908696] is a small glycoprotein found in high quantity in the brain of animals infected with certain **Physiology of the prion protein. - NCBI - NIH** FEBS J. 2011 Nov278(21):4002-14. doi: 10.1111/j.1742-4658.2011.08304.x. Epub 2011 Sep 15. The prion protein binds thiamine. Perez-Pineiro R(1), **Prions - PDB-101 - RCSB PDB** The functions of these normal prion proteins are still not completely understood. The abnormal folding of the prion proteins leads to brain **none** Ann Neurol. 2010 Aug68(2):162-72. doi: 10.1002/ana.22094. Variably protease-sensitive prionopathy: a new sporadic disease of the prion protein. Zou WQ(1) **The role of the prion protein membrane anchor in prion infection** Ablation of the cellular prion protein PrPC leads to a chronic demyelinating polyneuropathy affecting Schwann cells. Neuron-restricted **The prion protein is an agonistic ligand of the G protein** **- NCBI - NIH** Curr Protein Pept Sci. 2010 Mar11(2):166-79. Structure of the prion protein and its gene: an analysis using bioinformatics and computer simulation. Sakudo A(1) **none** Ablation of the cellular prion protein PrP(C) leads to a chronic demyelinating polyneuropathy affecting Schwann cells. Neuron-restricted **Prions and prion proteins. - NCBI - NIH** The basis of human prion diseases affecting the nervous system is accumulation of a disease-associated conformer (PrPSc) of the normal cellular prion protein **Prion proteins** Prion diseases are caused by conversion of a normal cell-surface glycoprotein (PrPC) into a conformationally altered isoform (PrPSc) that is **The prion protein family: diversity, rivalry, and dysfunction. - NCBI** Since then, the gene for this protein has been successfully cloned, and studies using transgenic mice have bolstered the prion hypothesis. The evidence in **PRNP prion protein [(human)] - NCBI** The misfolding of the cellular prion protein (PrPC) causes fatal neurodegenerative diseases. Yet PrPC is highly conserved in mammals, **PRNP gene - Genetics Home Reference** This work sheds light on the amyloid core structures underlying prion strains and how I138M, I139M, and S143N affect prion protein **The prion protein is an agonistic ligand of the G protein** **- Nature** **The prion protein gene in humans revisited: Lessons from a** Although the prion protein (PrP) is known to be the causative agent of the neurodegenerative transmissible spongiform encephalopathies, its normal cellular **The prion protein in human neuromuscular diseases. - NCBI** The prion protein family: diversity, rivalry, and dysfunction. members: Prnp which encodes PrP(C), the precursor to prion disease associated isoforms such as The prion hypothesis states that the infectious agent of prion diseases is an abnormally folded isoform of the prion protein (PrPSc) that replicates its abnormal **The Cellular Prion Protein (PrPC): Its Physiological Function and** **The Prion Protein Knockout Mouse - NCBI - NIH** There is no evidence for a nucleic acid in the prion, but diverse experimental results indicate that a host-derived protein called PrPSc is a component of the **Prion Protein Misfolding - NCBI - NIH** Prions are proteins that can adopt two different forms, a normal form and a misfolded form. This may not seem unusual, since many proteins are flexible and