

Resources on Prions for Libraries

Books on Prions

(Publishers' descriptions)

Soto, Dr. Claudio, (2006). **Prions: the new biology of proteins.** CRC/Taylor & Francis. 9780849314421

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.

Kim, Kiheung, (2006). **The social construction of disease: From scrapie to prion.** Routledge. 041536051X

A historical exploration of scientific disputes on the causation of so-called 'prion diseases', this fascinating book covers diseases including Scrapie, Creutzfeldt-Jakob Disease (CJD) and Bovine Spongiform Encephalopathy (BSE). Firstly tracing the twentieth-century history of disease research and biomedicine, the text then focuses on the relations between scientific practice and wider social transformations, before finally building upon the sociologically informed methodological framework. Incisive and thought-provoking, The Social Construction of Disease provides a valuable contribution to that well-established tradition of social history of science, which refers primarily to the theoretical works of the sociology of scientific knowledge.

Mandell, Gerald L., R Gordon Douglas; John E Bennett; Raphael Dolin (Eds.) (2006). **Mandell, Douglas, and Bennett's principles and practice of infectious diseases.** (6th ed.) 2 vol. Elsevier/Churchill Livingstone. 0443066434 (set)

Authored by a "who's who" of experts from around the world, this 2-volume set encompasses all that is currently known about the epidemiology, aetiology, pathology, microbiology, immunology, and treatment of infectious agents. Completely revised and updated - and with a bold, new 4-colour format - it is a complete, essential compendium of knowledge on this vast and complex subject.

Kitamoto, T., (2005) **Prions: food and drug safety.** Springer. 4431255397

Prion diseases recently have attracted interest not only scientifically but also socially because of the bovine spongiform encephalopathy (BSE) epidemic and the outbreak of variant Creutzfeldt-Jakob disease (vCJD) in the United Kingdom. In 2004, the International Symposium of Prion Diseases for Food and Drug Safety was held October 31–November 2 in Sendai, Japan, where, 20 years earlier, arguments were first heard on whether the etiologic agent of transmissible spongiform encephalopathy was prions or scrapie-associated fibrils. This volume is a collection of current work on prion research that was presented at the 2004 symposium. Topics included range from basic research to clinical aspects of prion diseases, making the book a valuable resource for researchers and clinicians, and encouraging further developments by the next generation of researchers

Harris, David Alan (2004). **Mad cow disease and related spongiform encephalopathies.** Springer. 3540201076 #284 of Current topics in microbiology and immunology series. Bovine spongiform encephalopathy (BSE) has become the most publicly recognizable example of a group of fatal neurodegenerative diseases caused by proteinaceous infectious particles called prions. The contributors to this volume, all internationally recognized experts in their fields, provide an introduction to prion biology, followed by reviews of the latest information on BSE, vCJD, and chronic wasting disease, an animal prion disease that has recently emerged in North America.

Rabenau, Holger F., Rabenau, Jindrich Ciantl and Hans Wilhelm Doerr (2004). **Prions: a challenge for science, medicine and the public health system.** (2nd ed.) Karger. 3805576560

This second, completely revised and extended edition of *Prions: A Challenge for Science, Medicine and the Public Health System* is a comprehensive, up-to-date review of prions and prion-associated diseases. Leading scientists discuss the structure, molecular biology and origin of prions as well as strain variations and species barriers. Human prion diseases, prion inactivation and risks to public health are considered in detail. The new edition provides an update on basic findings of the last three years since publication of the previous edition and emphasizes practical aspects of fighting human and animal prion diseases. In addition, chapters on regulatory aspects of BSE and CJD as well as on veterinary measures have been included.

Prusiner, Stanley B. (2004). **Prion biology and diseases**. (2nd ed.) Cold Spring Harbor Laboratory Press. 0879696931

This volume is a new edition of the most authoritative book on Prion Biology, first published in 1999 and edited by the Nobel Prize–winning founder of the field. This expanded edition has been completely updated, and includes chapters on therapeutics, and diagnostic methods and approaches.

Yarn, Philip (2003). **The Pathological protein: mad cow, chronic wasting and other deadly prion diseases**. Springer-Verlag. 0387955089

Prions are an entirely new class of pathogens, and scientists are just beginning to understand them. This book tells the strange story of their discovery, and the medical controversies that swirl around them

Journals Covering Prion Research

(by discipline, from data in Web of Science and Journal Citation Reports)

Biochemistry & Molecular Biology

Best: Cell
Embo Journal
Journal of Biological Chemistry

Very Good: Biochemistry
Journal of Molecular Biology
Molecular & Cell Biology
Molecular Microbiology
Nature Structural & Molecular Biology
Nature Medicine
Trends in Biochemical Sciences

Good: Biochemical & Biophysical Research Communications
FEBS Letters

Biophysics

Best: Biochemical & Biophysical Research Communications
Very Good: FEBS Letters

Biotechnology & Applied Microbiology

Best: Journal of General Virology

Cell Biology

Best: Cell
Embo Journal
Very Good: Journal of Cell Biology
Molecular & Cell Biology
Nature Reviews Cell Biology
Nature Structural & Molecular Biology
Nature Medicine
Good: FEBS Letters

Clinical Neurology

Best: Annals of Neurology
Neurology
Very Good: Acta Neuropathologia

Developmental Biology

Best: Genes & Development

Genetics & Heredity

Best: Genetics
Very Good: Genes & Development
Nature Genetics
Good: Heredity

Immunology

Best: Journal of Immunology
Very Good: Journal of Experimental Medicine
Good: Journal of Neuroimmunology

Infectious Diseases

Best: Journal of Infectious Disease

Medicine, General & Internal

Best: Lancet
Very Good: New England Journal of Medicine
British Medical Bulletin

Medicine, Research & Experimental

Best: Nature Medicine
Journal of Clinical Investigation
Journal of Experimental Medicine

Microbiology

Best: Molecular Microbiology

Multidisciplinary Sciences

Best: Nature
PNAS
Science

Neuroscience

Best: Neuron
Annals of Neurology
Brain
Glia
Very Good: Brain Pathology
Experimental Neurology
Journal of Immunology
Journal of Neurovirology
Good: Acta Neuropathologia

Pathology

Best: American Journal of Pathology
Journal of Pathology

Very Good: Acta Neuropathologia
Brain Pathology

Good: Journal of Comparative Pathology

Veterinary Sciences

Best: Veterinary Record

Very Good: Journal of Comparative Pathology

Virology

Best: Journal of Virology

Very Good: Journal of General Virology

Good: Journal of Neurovirology

[Detailed information about how these rankings were derived is available upon request.]

Web Sites About Prions

Prion Diseases

Centers for Disease Control and Prevention, Department of Health and Human Services.

<http://www.cdc.gov/ncidod/dvrd/prions/>

This site provides links to information about human and animal prion diseases from the CDC and other government sources.

Prions and Transmissible Spongiform Encephalopathies

U.S. Food & Drug Administration, Center for Food Safety and Applied Nutrition

<http://www.cfsan.fda.gov/~mow/prion.html>

Information about prions and TSE's with links to additional documentation, as well as to the MMWR, PubMed and Agricola.

Prion Protein

Online Mendelian Inheritance in Man, from Johns Hopkins University and NCBI

<http://www.ncbi.nlm.nih.gov/entrez/dispomim.cgi?id=176640>

The catalog entry for the prion protein, including gene function, molecular genetics, population genetics, experimental information, history and references.

Transmissible Spongiform Encephalopathies Information Page

National Institute of Neurological Disorders and Stroke

<http://www.ninds.nih.gov/disorders/tse/tse.htm>

An information page about TSE's in humans, with links to studies recruiting patients, and to other organizations working on related topics.

CJD Surveillance

National Prion Disease Pathology Surveillance Center

<http://www.cjdsurveillance.com/index.html>

The Center monitors the occurrence of Creutzfeldt-Jakob Disease and other forms of prion disease in humans, and conducts research into animal prion diseases which may transmit to humans, such as Chronic Wasting Disease or Bovine Spongiform Encephalopathy.

Chronic Wasting Disease

Veterinary Services, Animal & Plant Health Inspection Services, USDA

<http://www.aphis.usda.gov/vs/naahps/cwd/>

An information page on CWD and diagnostics, with links to related USDA pages.

Chronic Wasting Disease

National Wildlife Health Center, USGS

http://www.nwhc.usgs.gov/disease_information/chronic_wasting_disease/index.jsp

A site on the role of the NWHC in combating CHW, with links to state and federal resources and factsheets.

Helping to Combat Chronic Wasting Disease

National Wildlife Health Center, USGS

http://www.nwhc.usgs.gov/publications/fact_sheets/pdfs/cwd/CWDsupport.pdf

Information about efforts especially in Wisconsin, from 2003.

Review Articles on Prions

(From Web of Science & Scopus)

Beekes, M., & P.A. McBride (2007). **The spread of prions through the body in naturally acquired transmissible spongiform encephalopathies.** *FEBS Journal* 274 (3) 588-605.

Transmissible spongiform encephalopathies are fatal neurodegenerative diseases that are caused by unconventional pathogens and affect the central nervous system of animals and humans. Several different forms of these diseases result from natural infection (i.e. exposure to transmissible spongiform encephalopathy agents or prions, present in the natural environment of the respective host). This holds true also for scrapie in sheep, bovine spongiform encephalopathy in cattle, chronic wasting disease in elk and deer, or variant Creutzfeldt-Jakob disease in humans, all of which are assumed to originate predominantly from peroral prion infection. This article intends to provide an overview of the current state of knowledge on the spread of scrapie, chronic wasting disease, bovine spongiform encephalopathy and variant Creutzfeldt-Jakob disease agents through the body in naturally affected hosts, and in model animals experimentally challenged via the alimentary tract. Special attention is given to the tissue components and spreading pathways involved in the key stages of prion routing through the body, such as intestinal uptake, neuroinvasion of nerves and the central nervous system, and centrifugal spread from the brain and spinal cord to peripheral sites (e.g. sensory ganglia or muscles). The elucidation of the pathways and mechanisms by which prions invade a host and spread through the organism can contribute to efficient infection control strategies and the improvement of transmissible spongiform encephalopathy diagnostics. It may also help to identify prophylactic or therapeutic approaches that would impede naturally acquired transmissible spongiform encephalopathy infections.

Caughey, B, & G.S. Baron (2006). **Prions and their partners in crime.** *Nature* 443 (7113) 803-810.

Prions, the infectious agents of transmissible spongiform encephalopathies (TSEs), have defied full characterization for decades. The dogma has been that prions lack nucleic acids and are composed of a pathological, self-inducing form of the host's prion protein (PrP). Recent progress in propagating TSE infectivity in cell-free systems has effectively ruled out the involvement of foreign nucleic acids. However, host-derived nucleic acids or other non-PrP molecules seem to be crucial. Interactions between TSE-associated PrP and its normal counterpart are also pathologically important, so the physiological functions of normal PrP and how they might be corrupted by TSE infections have been the subject of recent research.

Mabbot, N.A. & G.G. MacPherson (2006). **Prions and their lethal journey to the brain.** *Nature Reviews. Microbiology* 4 (3) 201-211 .

Prion diseases are neurodegenerative conditions that cause extensive damage to nerve cells within the brain and can be fatal. Some prion disease

agents accumulate first in lymphoid tissues, as they make their journey from the site of infection, such as the gut, to the brain. Studies in mouse models have shown that this accumulation is obligatory for the efficient delivery of prions to the brain. Indeed, if the accumulation of prions in lymphoid tissues is blocked, disease susceptibility is reduced. Therefore, the identification of the cells and molecules that are involved in the delivery of prions to the brain might identify targets for therapeutic intervention. This review describes the current understanding of the mechanisms involved in the delivery of prions to the brain.

True, H.L. (2006). **The battle of the fold: chaperones take on prions.** *Trends in Genetics* 22 (2): 110-117.

Protein conformational diseases, such as Alzheimer's, Parkinson's and Huntington's, affect a large portion of our aging population. Cells have evolved mechanisms for rescuing and recycling misfolded proteins, but these systems are not perfect. Chaperones can rescue misfolded proteins by breaking up aggregates and assisting in the refolding process. Proteins that cannot be rescued by refolding can be delivered to the proteasome by chaperones to be recycled. One class of 'misfolded' proteins, prions, appears to evade detection by this machinery and persist in a misfolded state. In fact, it seems that the prions usurp the refolding machinery and actually employ chaperones to propagate the prion state. Recent data has begun to uncover the mechanism behind this unique relationship.

Watts, J.C., A. Balachandran & D. Westaway (2006). **The expanding universe of prion diseases.** *PLOS Pathogens* 2 (3) 152-163.

Prions cause fatal and transmissible neurodegenerative disease. These etiological infectious agents are formed in greater part from a misfolded cell-surface protein called PrPC. Several mammalian species are affected by the diseases, and in the case of "mad cow disease" (BSE) the agent has a tropism for humans, with negative consequences for agribusiness and public health. Unfortunately, the known universe of prion diseases is expanding. At least four novel prion diseases - including human diseases variant Creutzfeldt-Jakob disease (vCJD) and sporadic fatal insomnia (sFI), bovine amyloidotic spongiform encephalopathy (BASE), and Nor98 of sheep - have been identified in the last ten years, and chronic wasting disease (CWD) of North American deer (*Odocoileus Specis*) and Rocky Mountain elk (*Cervus elaphus nelsoni*) is undergoing a dramatic spread across North America. While amplification (BSE) and dissemination (CWD, commercial sourcing of cervids from the wild and movement of farmed elk) can be attributed to human activity, the origins of emergent prion diseases cannot always be laid at the door of humankind. Instead, the continued appearance of new outbreaks in the form of "sporadic" disease may be an inevitable outcome in a situation where the replicating pathogen is host-encoded.

Wilson, J.F. (2005). **Why prion diseases are a mystery, and why they matter.** *Annals of Internal Medicine* 143 (10) 773-776.
(No abstract available.)

Ena, J. (2005). **Prions: Who should worry about them?** *Archives of Medical Research*. 36 (6): 622-627.

Prion diseases, also called transmissible spongiform encephalopathies (TSEs), are a family of neurodegenerative disorders affecting both humans and animals. They are caused by the accumulation of an abnormal form of a protein known as prion that results in neuronal death and a characteristic spongiform appearance of the brain tissue. Human prion diseases can be sporadic, acquired or hereditary. Acquired prion diseases have been linked to entering contaminated food into the human food chain, failure to completely disinfect or sterilize contaminated surgical instruments, patients receiving tissues and organs from infected donors, recipients of blood and other biological contaminated products, and potentially to cross infection in dental procedures. At present, there is unfortunately no efficient therapy that can be administered to clinically infected patients with prion diseases. Moreover, there are no simple diagnostic tests that can be used to show the agent of transmissible spongiform encephalopathy during the preclinical phase of the disease. Therefore, to prevent the spread of this emerging infectious agent it is necessary to implement several health control strategies and maintain surveillance for subclinical infections.