1. Record Nr. UNINA9910437824903321

Titolo	Prions and diseases . Volume 2 Animals, humans and the environment / / Wen-Quan Zou, Pierluigi Gambetti, editors
Pubbl/distr/stampa	New York, : Springer, 2013
ISBN	1-283-91187-6 1-4614-5338-0
Descrizione fisica	1 online resource (296 p.)
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Disciplina	616.83
Soggetti	Prion diseases Prions
Lingua di pubblicazione	Inglese
Formato	Materiale a stampa
Livello bibliografico	Monografia
Note generali	Description based upon print version of record.
Nota di bibliografia	Includes bibliographical references and index.
Nota di contenuto	Bovine Spongiform Encephalopathy -- Classical and Atypical Scrapie in Sheep and Goats -- Sporadic Human Prion Disease -- Environmentally-acquired Transmissible Spongiform -- Prions in the Environment -- The Spectrum of Tau Pathology in Human Prion Disease -- Risk of Transmission of Creutzfeldt-Jakob Disease by Blood Transfusion -- Species Barriers in Prion Disease -- Transgenic Mice Modelling -- Prion Transmission Studies in Transgenic Mice -- Alternative Models of Prion Diseases -- Diagnosis of Prion Disease: Conventional Approaches -- Quaking-induced Conversion (QuIC) Assays for the Detection and Diagnosis of Prion Diseases -- Overview on Treatment of Prion Diseases and Decontamination of Prions -- Immunomodulation.
Sommario/riassunto	Transmissible spongiform encephalopathies (TSE), now broadly known as prion diseases, have been recognized for nearly 300 years in animals and almost 100 years in humans. However, the nature of the transmissible agent had largely remained a mystery until Stanley Prusiner discovered the infectious isoform of the prion protein (PrP), named prion or scrapie PrP (PrPSc), in 1982. The subsequent modern studies with protein chemistry and molecular biology in cell culture, transgenic animals, and cell-free systems, including the revolutionary protein-misfolding cyclic amplification (PMCA), have greatly advanced

our understanding of the pathogenesis of prion diseases and facilitated the identification of new prion diseases in animals and humans. In Prions and Prion Diseases, more than 60 leading researchers and clinicians worldwide provide an up-to-date development in many aspects of these unique infectious pathogens and their associated diseases. Volume II features a variety of animal and human prion diseases, including the newly-identified atypical forms of bovine spongiform encephalopathy and scrapie in animals, and variably protease-sensitive prionopathy in humans, prions in the environment, Tau pathology in human prion disease, transmission of the disease by blood transfusion, mammalian and non-mammalian models, conventional and advanced diagnoses, prion-specific antibodies, as well as decontamination of prions and development of therapeutics of prion diseases, such as the application of immunomodulation. This volume provides up-to-date knowledge about the etiology, pathogenesis, classification, histopathological, and clinical aspects of the highly publicized animal and human prion diseases.