Record Nr. UNINA9910437839403321 Prions and diseases . Volume 1 Physiology and pathophysiology // **Titolo** Wen-Quan Zou, Pierluigi Gambetti, editors Pubbl/distr/stampa New York, : Springer, 2013 **ISBN** 1-283-91173-6 1-4614-5305-4 Edizione [1st ed. 2013.] Descrizione fisica 1 online resource (244 p.) Altri autori (Persone) ZouWen-Quan GambettiPierluigi Disciplina 579.29 Soggetti **Prions** Prion diseases 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 Transmissible Spongiform Encephalopathy: from the High Middle Ages to Daniel Carlton Gajdusek -- The Rich Chemistry of the Copper and Zinc Sites in Cellular Prion Protein -- Role of Cellular Prion Protein in the Amyloid-ß Oligomer Pathophysiology of Alzheimer's Disease --Cellular Prion Protein and Cancers -- Insoluble Cellular Prion Protein --Protein Misfolding Cyclic Amplification -- Cofactor Involvement in Prion Propagation -- Prion Protein Conversion and Lipids -- New Perspectives on Prion Conversion: Introducing a Mechanism of Deformed Templating -- Infectious and Pathogenic Forms of Prion Protein -- Cellular Mechanisms of Propagation and Clearance --Molecular Mechanisms Encoding Quantitative and Qualitative Traits of Prion Strains -- Modeling the Cell Biology of Prions -- Prion Strain Interference -- Introduction to Yeast and Fungal Prions -- Yeast Prions are Pathogenic, in-register Parallel Amyloids.

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 I highlights the association of the cellular prion protein (PrPC) with copper and zinc, the potential roles of PrPC in Alzheimer's disease and cancers, insoluble PrPC, PMCA, molecular and cellular mechanisms of PrPSc formation and clearance, possible cofactors involved in the conversion of PrPC into PrPSc, infectious and pathogenic forms of PrP, cell biology of prions, prion strains and their interference, as well as yeast prions and their inheritable and structural traits. This unique volume covers history from the high Middle Ages to the TSE era of Daniel Carleton Gajdusek, followed by the prion era of Stanley Prusiner whose extraordinary discovery opened a new chapter in prion research. Volume I will take you through the fascinating chronicle of prions in mammals, yeast, and fungi.