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Nota di contenuto	Frontmatter -- Table of Contents -- Abbreviations -- Topic I: History -- 1. Historical Introduction -- 2. History of Kuru Research -- 3. History of Prion Research -- Topic II: Molecular Biology and Genetics -- 4. The Physical Nature of the Prion -- 5. Folding of the Recombinant Prion Protein -- 6. Structural Studies of Prion Proteins -- 7. Function of Cellular Prion Protein (PrPC) in Copper Homeostasis and Redox Signaling at the Synapse -- 8. The Scarpie Isoform of the Prion Protein PrPSe Compared to the Celluar Isoform PrPC -- 9. The Phylogeny of Mammalian and Nonmammalian Prion Proteins -- 10. Knockouts and Transgenic Mice in Prion Research -- 11. Transplantation as a Tool in Prion Research -- 12. Prion Strains -- Topic III: Portraits of Prion Diseases -- 13. Portrait of Kuru -- 14. Portrait of Creutzfeldt-Jakob Disease -- 15. Portrait of Variant Creutzfeldt-Jakob Disease -- 16. Portrait of Gerstmann-Sträussler-Scheinker Disease -- 17. Portrait of Fatal Familial Insomnia and Sporadic Fatal Insomnia -- 18. Portrait of Scrapie in Sheep and Goat -- 19. Portrait of Bovine Spongiform Encephalopathy in Cattle and Other Ungulates -- 20. Portrait of Prion Diseases in Zoo Animals -- 21. Portrait of Chronic Wasting Disease in

Deer Species -- 22. Portrait of Transmissible Mink Encephalopathy -- 23. Portrait of Transmissible Feline Spongiform Encephalopathy -- 24. Portrait of Experimental BSE in Pigs -- 25. Portrait of a Spongiform Encephalopathy in Birds and the Transmissibility of Mammalian Prior Diseases to Birds -- Topic IV: Pathology -- 26. Pathology and Genetics of Human Prion Diseases -- 27. The Pathology of Prion Diseases in Animals -- 28. Pathophysiology of Prion Diseases Following Peripheral Infection -- Topic V: Surveillance, Clinical Aspects and Diagnostics -- 29. Introduction of Surveillance for Human Prion Diseases -- 30. Clinical Findings in Human Prion Diseases -- 31. Methods for the Clinical Diagnosis of Human Prion Deseases -- 32. Introduction to Surveillance for Animal Prion Diseases -- 33. Clinical Findings in Bovine Spongiform Encephalopathy -- 34. Clinical Findings in Scrapie -- 35. Diagnosis of Bovine Spongiform Encephalopathy by Immunological Methods -- Topic VI: Epidemiology -- 36. Epidemiology and Risk Factors of Creutzfeldt-Jakob Disease -- 37. Creutzfeldt-Jakob Disease in Germany -- 38. The Epidemiology of Kuru -- 39. The Course of the BSE Epidemic - Retrospective Epidemiological Considerations -- 40. The Causes of the BSE Epidemic -- Topic VII: Transmissibility -- 41. The Experimental Transmissibility of Prions and Infectivity Distribution in the Body -- 42. Iatrogenic and "Natural" Transmissibility of Prion Diseases -- Topic VIII: Agent Inactivation -- 43. Inactivation in Practice - Risk Assessment and Validation for Food Gelatin -- 44. Chemical Disinfection and Inactivation of Prions -- 45. Thermal Inactivation of Prions -- Topic IX: Prevention -- 46. Prevention of Prion Diseases in the Production of Medicinal Products, Medical Devices, and Cosmetics -- 47. Prevention of the Transmission of Prion Diseases in Healthcare Settings -- 48. Precautionary Measures for Autopsies Performed in Cases of Suspected Prion Disease -- 49. Prevention of Prion Diseases in Research Laboratories -- Topic X: Risk Assessment -- 50. Evidence for a Link between Variant Creutzfeldt-Jakob Disease and Bovine Spongiform Encephalopathy -- 51. Risk Assessment of Transmitting Prion Diseases through Blood, Cornea, and Dura Mater -- 52. BSE Risk Assessment and Minimization -- 53. BSE Control - Internationally Recommended Approaches -- 54. Atypical Scrapie-Nor98 -- 55. Scrapie Control - Internationally Recommended Approaches -- 56. The PrP Genotype as a Marker for Scrapie Susceptibility in Sheep -- 57. Scrapie control at the National level: The Norwegian Example -- Backmatter

Sommario/riassunto

Dieses umfassende Werk in englischer Sprache richtet sich gleichermaßen an Studenten und Forscher. Es behandelt systematisch alle Aspekte der Prionenkrankheiten, von ihrer Geschichte, Mikrobiologie und Pathologie bis hin zu Übertragungswegen und möglicher Prävention. Das Buch beschreibt Krankheiten wie Creutzfeld-Jakob-Krankheit, Kuru, Rinderwahn (BSE) und Scrapie, unter besonderer Berücksichtigung der biochemischen, molekularbiologischen, genetischen und klinischen Aspekte. Eine genaue Beschreibung der Wirkung von Prionenkrankheiten im Bereich der Pharmazie, bei Blutprodukten, Desinfektion, chirurgischen Instrumenten und der Epidemiologie rundet das Werk mit einer Diskussion über präventive Maßnahmen ab.
This comprehensive work, aimed at both students and researchers alike, systematically covers all aspects of prion diseases (transmissible spongiform encephalopathies), from their history, microbiology and pathology to their transmissibility and prevention. The book describes diseases such as Creutzfeldt-Jakob disease, kuru, mad cow disease (BSE), chronic wasting disease and scrapie, highlighting their biochemical, molecular biological, genetic, and clinical aspects. A

detailed presentation of the impact of prion diseases in fields such as pharmaceutics, blood products, disinfection, surgical instruments and epidemiology concludes with a discussion of preventive measures. A renowned editorial team, representing the fields of medicine, veterinary medicine and molecular biology, brought together 80 internationally respected authors for this translation and new edition of the successful German publication, not only from relevant research fields, but also from industry and public health institutions. The book includes chapters by, among many other notable scientists, William J. Hadlow, who discovered the relationship between the human and animal forms of prion diseases and Michael P. Alpers, with 45 years of experience in Papua New Guinea investigating the first known human epidemic form, kuru, transmitted by endocannibalism. Further contributions from Gerald A. H. Wells, a veterinary pathologist who described BSE and recognised its similarity to scrapie, thus recording the first cases in 1986 of the most important animal epidemic of modern times, and Robert G. Will, a medical neurologist and epidemiologist who discovered the emergence of the variant form of Creutzfeldt-Jakob disease in 1996, underscore the strength of this author team. Carefully edited with numerous illustrations, this work offers a systematic approach committed to a clear presentation of the current knowledge of prion diseases. It aims to inspire and stimulate interdisciplinary cooperation, innovative research ideas and effective prevention.