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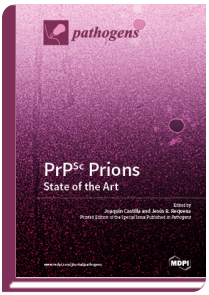
Special Issue Reprint

PrPSc Prions: State of the Art



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Prion diseases, or transmissible spongiform encephalopathies (TSEs), are a group of fatal and transmissible neurodegenerative disorders characterized by long incubation periods, misfolded prion protein (PrP) deposition, and usually spongiform vacuolation. These devastating diseases affect many mammals, with the best known examples being Creutzfeldt–Jakob disease (CJD), fatal familial insomnia (FFI), or Kuru in humans; and scrapie in sheep, bovine spongiform encephalopathy (BSE) in cattle, and chronic wasting disease (CWD) in cervids. Despite major achievements in research of TSEs, there are still many unresolved key issues that hamper the development of effective therapies. However, the last decade has been particularly prolific in advances in the prion field. Among others, prion propagation in vitro has been achieved, leading to new diagnostic methods; the basic architecture of infectious prions has been deciphered; new prion disease types have been described in humans and other animals; and prion disorders have emerged in places that had not previously reported the disorders, as is the case for CWD in Europe.

This Special Issue will focus on the state of the art of our knowledge of PrPSc: on what we know about its structure and propagation, the basis of strains and transmission barriers, the mechanisms of PrPSc toxicity, the possible function of PrPSc's properly folded precursor, PrPC and its evolutionary history, and recent technical breakthroughs in diagnostics and therapy development among other key aspects of PrPSc prion biology.

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