Special Issue

Prion Diseases: A Model for Neurodegenerative Disorders

Message from the Guest Editor

Prion diseases or transmissible spongiform encephalopathies are invariably fatal neurodegenerative disorders of humans and animals. They are caused by prions, self-propagating proteinaceous infectious particles which consist of a misfolded and aggregation-prone isoform of the cellular prion protein (PrPC), termed PrPSc. Over the last decade, the concept of prion-like mechanisms in other neurodegenerative diseases evolved, based on the principle of seeding, spreading, and propagation of protein misfolding in the brains of affected individuals. These mechanisms were initially ascribed to prions and now have been well established for other neurodegenerative disorders, such as Alzheimer's or Parkinson's disease. Still, prion diseases are exceptional as their natural transmission is well documented while not observed, for example, in Alzheimer's or Parkinson's disease. Despite this difference, the structural characteristics and shared mechanisms of propagation or cell-to-cell transmission suggest potential common therapeutic targets and principles of diagnostic assays, which will be discussed in this Special Issue.

Guest Editor

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