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## Preface

The transmissible spongiform encephalopathy or prion diseases are a group of invariably fatal neurodegenerative disorders with sporadic, familial and acquired etiologies that affect a variety of animal species including humans.

It is now generally accepted that prion diseases arise through the aberrant misfolding of the host-encoded cellular prion protein. Protein misfolding, although not unique amongst neurodegenerative disorders, can in the case of prion diseases be seeded by the misfolded protein, a property which imparts the transmissible nature of prion diseases in the absence of a conventional infectious agent. It is this feature, in conjunction with difficulties associated with decontamination of infected surgical instruments and material, which can be attributed to the level of community interest and public health concerns despite the diseases' low prevalence.

It has been over 30 years since a novel infectious particle was proposed to be the causative agent of the prototypic prion disease, scrapie. Proving the existence of a transmissible protein or 'prion' and understanding prion disease transmission and pathogenesis have led to the development of many innovative methodologies. Understanding the unique nature of the disease has led to a truly multidisciplinary approach, which unites biochemical and biophysical approaches with in vitro and in vivo models and animal biologists and veterinarians with biomedical researchers and clinicians.

This book brings together protocols from each of these disciplines and highlights the contribution each discipline has made to our understanding of the nature of prion disease.

In addition to contributing to our understanding of prion disease, these methods may also find application to the newly emerging and so-called 'prion-like' properties observed in other protein misfolding neurodegenerative diseases as highlighted in the final method which describes a method for detecting the intercellular prion-like conversion and transmission of Cu/Zn superoxide dismutase (SOD1).

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Prions

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