Prions

Current Progress in Advanced Research

Second Edition

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Preface

It is now more than 30 years since the discovery of proteinaceous infectious particles known as prions. Prior to their discovery, Tikvah Alper (Alper *et al.*, Nature *214*, 764–766, 1967) and John Stanley Griffith (Griffith, *Nature 215*,1043–1044, 1967) performed a series of elegant studies that implied the infectious agent responsible for transmissible spongiform encephalopathy was of protein origin. Crucially, the infectious agents causing scrapie and Creutzfeldt–Jakob disease were found to be resistant to high levels of ultraviolet radiation, suggesting that the agents lack nucleic acids. In 1982, Dr Stanley Prusiner finally isolated the infectious agent responsible for scrapie, and found that the major component of the factor was a specific protein (Prusiner, *Science 216*, 136–144, 1982). It was Prusiner who originally coined the word prion, which is short for 'proteinaceous infectious particle'. The discovery of prions initiated further intensive research into the biology of these infectious agents. However, it was several years before the protein-only hypothesis was widely accepted within the scientific community. Nonetheless, subsequent investigations confirmed the prion hypothesis put forward by Prusiner, for which he was awarded the Nobel Prize in 1997.

This book is the second edition of a previous book entitled 'Prions: Current Progress in Advanced Research' and is now fully updated with recent findings in the rapidly advancing field of prion research. Since the publication of the first edition of this book in 2013, numerous research groups have employed advanced technologies to gain further insight into the biology of these infectious agents. Significant progress has been made in understanding these prion agents, including the identification of new strains of bovine spongiform encephalopathy (BSE), chronic wasting disease (CWD) as well as abnormal isoforms of the prion protein (PrP^{Sc}) types associated with Creutzfeldt–Jakob disease (CJD). Moreover, the need to detect prions in easily accessible biological samples (e.g. blood or cerebrospinal fluid) for early and specific diagnosis of neurodegenerative diseases has led to the development of highly sensitive assays for disease-related proteins, such as the real-time quaking-induced conversion (RT-QuIC) method.

The aim of this book is to update students, scientists and engineers with recent progress in advanced research into the biology of prions. This book comprises 10 chapters, Chapter 1 being the introduction. The following four chapters (Chapters 2–5) deal with fundamental aspects of prion biology, including functions of the cellular isoform of prion protein (PrP^{C}) and molecular mechanisms of prion diseases. The next two chapters (Chapters 6 and 7) focus on clinical aspects of human prion diseases and current approaches for effective inactivation methods. The last part of the book (Chapters 8 and 9) summarizes animal prion

diseases, including BSE, scrapie and CWD. In the final chapter, Professor Onodera and colleagues discuss the likely future direction of research in this field.

It was a great honour to have been given the opportunity to work with the eminent scientists as chapter contributors, together with my co-editor Professor Takashi Onodera. I would like to thank Professor Onodera and all the contributors to this book for their commitment and enthusiasm during compilation of the respective chapters. Finally, I wish to thank Annette Griffin and the other editorial staff at Caister Academic Press for their professionalism and dedication.

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