

# Contents

|                                                                                                                             |     |
|-----------------------------------------------------------------------------------------------------------------------------|-----|
| <b>Genetics of Prion Disease</b> .....                                                                                      | 1   |
| Sarah Lloyd, Simon Mead, and John Collinge                                                                                  |     |
| <b>Atypical Prion Diseases in Humans and Animals</b> .....                                                                  | 23  |
| Michael A. Tranulis, Sylvie L. Benestad, Thierry Baron,<br>and Hans Kretzschmar                                             |     |
| <b>Chronic Wasting Disease</b> .....                                                                                        | 51  |
| Sabine Gilch, Nandini Chitoor, Yuzuru Taguchi, Melissa Stuart,<br>Jean E. Jewell, and Hermann M. Schätzl                    |     |
| <b>Transgenic Mouse Models and Prion Strains</b> .....                                                                      | 79  |
| Glenn C. Telling                                                                                                            |     |
| <b>Neuroprotective and Neurotoxic Signaling by the Prion Protein</b> .....                                                  | 101 |
| Ulrike K. Resenberger, Konstanze F. Winklhofer, and Jörg Tatzelt                                                            |     |
| <b>Prion Seeded Conversion and Amplification Assays</b> .....                                                               | 121 |
| Christina D. Orrú and Byron Caughey                                                                                         |     |
| <b>Prion Protein and Its Conformational Conversion:<br/>A Structural Perspective</b> .....                                  | 135 |
| Witold K. Surewicz and Marcin I. Apostol                                                                                    |     |
| <b>Molecular Dynamics as an Approach to Study Prion<br/>Protein Misfolding and the Effect of Pathogenic Mutations</b> ..... | 169 |
| Marc W. van der Kamp and Valerie Daggett                                                                                    |     |
| <b>Chemical Biology of Prion Protein: Tools to Bridge<br/>the In Vitro/Vivo Interface</b> .....                             | 199 |
| Ralf Seidel and Martin Engelhard                                                                                            |     |

**The PrP-Like Proteins Shadoo and Doppel** ..... 225  
David Westaway, Nathalie Daude, Serene Wohlgemuth, and Paul Harrison

**Fungal Prions: Structure, Function and Propagation** ..... 257  
Mick F. Tuite, Ricardo Marchante, and Vitaly Kushnirov

**Index** ..... 299