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## Preface

Prion diseases represent one of the most exciting and intriguing fields in biomedical sciences, with three Nobel Prizes awarded so far—D. Carleton Gajdusek in 1976, Stanley B. Prusiner in 1997, who coined the term “prion” in 1982 [1], and Kurt Wuthrich in 2002. I became interested in the prion field in 1977, when I read an article, entitled “Epidemiology of Creutzfeldt-Jakob disease in England and Wales,” by Walter B. Matthews, in the *Journal of Neurology Neurosurgery and Psychiatry* [2], where for the first time I was confronted by “scrapie,” a term previously unknown to me. On immediately searching this key word, I found the Nobel Lecture by Gajdusek [3]. At that time, I was a student with an interest in electron microscopy, which I began to learn from Prof. Michal Karasek (1937–2009), Dr. Iwona Giryn (deceased 2011), and Dr. Barbara Mirecka (deceased 1992). My skills were expanded through a British Council Fellowship at the MRC and AFRC Neuropathogenesis Unit, under Dr. Peter Gibson. These research training experiences paved the way to a Fogarty Fellowship in the Laboratory of Central Nervous System Studies, under the direction of D. Carleton Gajdusek, a Nobel laureate, and Dr. C. J. Gibbs Jr., at the National Institute of Neurological Disorders and Stroke, of the National Institutes of Health. I developed and printed there 17,000 electron micrographs. Professionally, this period was the most exhilarating time of my life.

The proposal of this book came as a little surprise from Wolfgang Walz, the Editor of this series at Springer. I then approached several of my expert colleagues and we started to write. Based on my professional experience, this book is somewhat biased toward morphological approaches. Chapters by Diane Ritchie and James Ironside, by Martin Jeffrey and Gilian McGovern, and by Frank Bastian and me provide in-depth coverage of different aspects of neuropathology, immunohistochemistry, electron microscopy, and immune-gold electron microscopy of prion diseases. These colleagues are the best in the world in this research area. It is noteworthy that a substantial portion of the book is devoted to electron microscopy, which has become merely a shadow of the importance it played in the biomedical sciences when I started my career, now being replaced by molecular biology.

The next section provides the reader with detailed information about the clinical description of prion diseases and the detection of prion protein and biomarkers, written by Richard Knight; Byron Caughey, Christina D. Orru, Bradley R. Groveman, Matilde Bongianni, Andrew G. Hughson, Lynne D. Raymond, Matteo Manca, Allison Kraus, Gregory J. Raymond, Michele Fiorini, Maurizio Pocchiari, and Gianluigi Zanusso; Elizaveta Katorcha and Ilya Baskakov; and Joanna Gawinecka, Matthias Schmitz, and Inga Zerr.

Separate chapters cover kuru. Among the authors, two were privileged to see real kuru—Shirley Lindenbaum and David Asher. The chapter by Pedro Piccardo and Luisa Gregori provides a broad overview of prion research in nonhuman primates. This chapter is unique as higher primates such as chimpanzees are no longer used in experimental research. There are not many in the world, who studied kuru in its natural environment. For me, I could study only one of the last kuru brains.

Finally, we return to molecular biology, which is now the ultimate answer to any scientific question, in contrast to what Gajdusek used to say: “Molecular biology became molecular technology and because of this is no longer a science.” To this group belong the exquisite chapters by Giuseppe Legname on synthetic prions, Abigail Diack and Jean Manson, and Glenn C. Telling and Julie Moreno on transgenic mouse models of prion disease. I am more than happy to include their chapters.

Last but not least, I would like to thank Michael Alpers for his invaluable discussions of kuru, Ewa Skarżyńska for her enormous help in handling the multiple tasks associated with this book, and Anna Rakovsky, from Springer Verlag, for her unending patience.

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