

# Preface

This volume of the Subcellular Biochemistry series is the result of the long-standing research interest of the Editor in the molecular mechanism underlying Alzheimer's disease and other amyloid diseases, indicated also by the earlier book in the series (Volume 38), devoted to Alzheimer's disease. The broad coverage within the present amyloidogenesis book represents an attempt to collate current knowledge relating to the proteins and peptides involved in most of the known amyloid diseases, together with some amyloid/fibril-forming proteins and peptides that are not involved in diseases. Thus, the range of topics included is comprehensive and furthermore it was thought appropriate to include both basic science and clinical presentation of the subjects, in most cases. Accordingly, the book has been divided into the following sections: I. Introduction, II. Basic Science (12 chapters), III. Clinical Science: the Cerebral and Systemic Diseases (9 Chapters). Internationally-based authoritative authors who are actively involved in their field of study have been selected as contributors to the book.

The book starts with an Introductory chapter, by Nathaniel Milton and myself, that also includes a technical survey of the many laboratory-based microscopical, analytical and biophysical, biochemical and cellular approaches used for the study of protein/peptide oligomerization, aggregation and fibrillogenesis. Some emphasis is given to the application of transmission electron microscopy for the assessment of these events, from the molecular (oligomer) level through the prefibril, mature fibril and fibril aggregates to the amyloid plaque level, a long-time interest of the Editor. A glance at many recently published studies across the whole amyloidogenesis field shows that this technique, along with others, generates useful information. In general, however, it must be emphasized that most groups use a combination of techniques to study amyloidogenesis and that the correlation of data from different approaches is a significant strength. In Chap. 2 Andrew Smith covers "*Fibril Formation by Short Synthetic Peptides*", which sets the scene from the point of view and underlying importance of  $\beta$ -sheet formation. Chap. 3, by Nuria Benseny-Cases, Oksana Klementieva and Josep Cladera, deals with "*In vitro Oligomerization and Fibrillogenesis of Amyloid-beta Peptides*", followed by Chap. 4 with the complementary Alzheimer topic, "*Tau Fibrillogenesis In vitro*", by Nitin Chaudhary and Ramakrishnan Nagaraj.

In Chap. 5 Jan Stöhr writes on “*Prion Protein Aggregation and Fibrillogenesis In vitro*”, providing the basic science behind the prion diseases. In Chap. 6, Katerina Paleologu and Omar El-Agnaf present the fundamentals of “ *$\alpha$ -Synuclein Aggregation and Modulating Factors*”, the principal cause of Parkinson’s disease.  $\alpha$ -Synuclein is also involved in a number of other neurodegenerative diseases. The topic “*Pathological Self-aggregation of  $\beta_2$ -Microglobulin: a Challenge for Protein Biophysics*” is contributed by Gennaro Esposito, Alessandra Corazza and Vittorio Bellotti in Chap. 7, followed in Chap. 8 by the consideration of the “*Islet Amyloid Polypeptide—Aggregation and Fibrillogenesis In vitro and its Inhibition*”, by Janine Seeliger and Roland Winter. The “*Mechanisms of Transthyretin Aggregation and Toxicity*” is dealt with by Robert Gasperini, David Klaver, Xu Hou, Marie-Isabel Aguilar and David Small in Chap. 9. The principal protein involved in Huntington’s disease, huntingtin, is covered in Chap. 10 by Yuri Lyubchenko, Alexey Krasnoslobodtsev and Sorin Luca, under the title “*Fibrillogenesis of Huntingtin and other Glutamine Containing Proteins*”. In Chap. 11 Moritz Lassé, Juliet Gerrard and Grant Pearce write on “*Aggregation and Fibrillogenesis of Proteins not Associated with Disease—A Few Case Studies*”. Two chapters on the inhibition of fibrillogenesis then complete the basic science section of the book. Alagari Srinivasan contributes “*Experimental Inhibition of Peptide Fibrillogenesis by Synthetic Peptides, Carbohydrates and Drugs*” in Chap. 12 and in Chap. 13 Suresh Kumar, Edward J. Okello and myself deal with “*Experimental inhibition of fibrillogenesis and neurotoxicity by amyloid-beta ( $A\beta$ ) and other disease-related peptides/proteins by plant extracts and herbal compounds*”.

Then there is the clinical section of the book, firstly with some chapters dealing with the cerebral amyloid diseases. In Chap. 14 Vanessa De-Paukla, Marcia Radanovic, Breno Diniz and Orestes Forlenza write on “*Alzheimer’s Disease*”, linking strongly with Chaps. 3 and 4. In Chap. 15 Keizo Sugaya contributes “*Modeling the Polyamine Aggregation Pathway in Huntington’s Disease: from Basic Studies to Clinical Application*”. “*Parkinson’s Disease*” then appears in Chap. 16, from Timothy Mhyre, James Boyd, Robert Hamill and Kathleen A. Maguire-Zeiss. The two following chapters then deal with the human and animal prion diseases. Beata Sikorska and Pawel Liberski write on “*Human Prion Diseases: from Kuru to Variant Creutzfeldt-Jakob Disease*” in Chap. 17, and in Chap. 18 Otto Windl and Mike Dawson write on the “*Animal Prion Diseases*”. Then follow several chapters on the systemic/peripheral amyloid diseases. Chap. 19 on “ *$\beta_2$ -Microglobulin Amyloidosis*” is by Dorthe Corlin and Niels Heegaard. Jennifer Pinney and Helen Lachmann then write in Chap. 20 on “*Systemic AA Amyloidosis*”, and in Chap. 21 Takamura Nagasaka deals with “*Familial Amyloidotic Polyneuropathy and Transthyretin*”. Finally, in Chap. 22, Giovanni Palladini and Raymond Comenzo write on “*The Challenge of Systemic Immunoglobulin Light-chain Amyloidosis (AL)*”.

No multi-author book can ever cover all possible relevant topics indicated by the book title. A few subjects were lost from my initial chapter list, owing to the lack of commitment by some of the initially commissioned authors, but the 22 chapters

included here cover the most relevant areas of amyloidogenesis and amyloid disease. Nevertheless, it is hoped that the overall content will be of interest and useful to both biomedical scientists and clinicians researching and involved in the treatment of amyloid disease.

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