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Protein folding and misfolding: neurodegenerative diseases-Judit Ovádi 2008-12-21 Offering all the latest in the study of neurodegenerative diseases, this book reviews the molecular events initiated by unfolded or misfolded proteins leading to conformational human diseases, especially those found in Parkinson’s and Alzheimer’s diseases.

Protein Misfolding in Neurodegenerative Diseases-Robert D. E. Sewell 2007-12-03 Research focused on protein folding, misfolding, and aggregation is leading to major advances across biochemistry and medicine. The elucidation of a folding code is proving to be of extreme importance in the postgenomic era, where a number of orphan genes have been identified for which no clear function has yet been established. This research is starting to shed light on the molecular and biochemical basis of a number of neurodegenerative diseases of dramatic impact. Protein Misfolding in Neurodegenerative Diseases: Mechanisms and Therapeutic Strategies addresses key issues concerning protein misfolding and aggregation in neurodegenerative diseases. Building on recent developments, including the recognition of protein misfolding as both a marker and a causal agent, the text presents the work of those who are actively pursuing more effective treatments, as well as preventative measures, and a possible cure. These include the use of molecular chaperones to control misfolding and novel pharmaceuticals, as well as the potential role of various inhibitors and NSAIDS. A Comprehensive Multifaceted Examination of the Complex Causal Agents Implicated in Protein Misfolding Divided into five sections, this groundbreaking text provides up-to-date accounts for Alzheimer’s, Parkinson’s, Huntington’s, Amyotrophic Lateral Sclerosis and Transmissible Spongiform Encephalitis. It also explores the highly likelihood that multiple factors, including oxidative stress, play a role in these complex diseases.

Protein Misfolding and Spreading Pathology in Neurodegenerative Diseases-Diana Fernandes Lázaro 2020-02-20

Protein Misfolding Diseases-Marina Ramirez-Alvarado 2010-12-01 An increasingly aging population will add to the number of individuals suffering from amyloid. Protein Misfolding Diseases provides a systematic overview of the current and emerging therapies for these types of protein misfolding diseases, including Alzheimer's, Parkinson's, and Mad Cow. The book emphasizes therapeutics in an amyloid disease context to help students, faculty, scientific researchers, and doctors working with protein misfolding diseases bridge the gap between basic science and pharmaceutical applications to protein misfolding disease.

Neurodegenerative Diseases-Uday Kishore 2013-05-15 This book highlights the pathophysiological complexities of the mechanisms and factors that are likely to be involved in a range of neuroinflammatory and neurodegenerative diseases including Alzheimer's disease, other Dementia, Parkinson Diseases and Multiple Sclerosis. The spectrum of diverse factors involved in neurodegeneration, such as protein aggregation, oxidative stress, caspases and secretase, regulators, cholesterol, zinc, microglia, astrocytes, oligodendrocytes, etc, have been discussed in the context of disease progression. In addition, novel approaches to therapeutic interventions have also been presented. It is hoped that students, scientists and clinicians shall find this very informative book immensely useful and thought-provoking.

Protein Misfolding Disorders-Claudio Hetz 2009 Neurodegenerative disorders such as Amyotrophic lateral sclerosis (ALS), Alzheimer’s disease (AD), Parkinson’s disease (PD), Prion-related disorders (PrD) and Huntington’s disease (HD) share a common neuropathology, primarily featuring the presence of abnormal protein inclusions containing specific misfolded proteins. These groups of diseases are now classified as Protein Misfolding Disorders. This book gives a comprehensive overview of the possible mechanisms involved in Protein Misfolding Disorders and possible therapeutic strategies to treat these diseases. The Ebook provides the most recent evidence addressing the role of cellular stress responses to neurological diseases, along with therapeutic strategies to alleviate ER stress in a disease context. -- Publisher.

Molecular Aspects of the Stress Response: Chaperones, Membranes and Networks-Peter Csermely 2007-08-09 This book makes a novel synthesis of the molecular aspects of the stress response and long term adaptation processes with the system biology approach of biological networks. Authored by an exciting mixture of top experts and young rising stars, it provides a comprehensive summary of the field and identifies future trends.

Protein Misfolding, Aggregation and Conformational Diseases-Vladimir N. Uversky 2007-11-24 Research indicates that most neurodegenerative diseases, systemic amyloidoses and many others, arise from the misfolding and aggregation of an underlying protein. This is the first book to discuss significant achievements in protein structure-function relationships in biochemistry, molecular biology and molecular medicine. The authors summarize recent progress in the understanding of the relationships between protein misfolding, aggregation and development of protein deposition disorders.

Bio-nanoimaging-Vladimir Uversky 2013-11-05 Bio-Nanoimaging: Protein Misfolding & Aggregation provides a unique introduction to both novel and established nanoimaging techniques for visualization and characterization of misfolded and aggregated protein species. The book is divided into three sections covering: - Nanotechnology and nanoimaging technology, including cryoelectron microscopy of beta(2)-microglobulin, studying amyloidogensis by FRET; and scanning tunneling microscopy of protein deposits - Polymorphisms of protein misfolded and aggregated species, including fibrillar polymorphism, amyloid-like protofibrils, and insulin oligomers - Polymorphisms of misfolding and aggregation processes, including multiple pathways of lysozyme aggregation, misfolded intermediate of a PDZ domain, and micelle formation by human islet amyloid polypeptide Protein misfolding and aggregation is a fast-growing frontier in molecular medicine and protein chemistry. Related disorders include cataracts, arthritis, cystic fibrosis, late-onset diabetes mellitus, and numerous neurodegenerative diseases like Alzheimer's and Parkinson's. Nanoimaging technology has proved crucial in understanding protein-misfolding pathologies and in potential drug design aimed at the inhibition or reversal of protein aggregation. Using these technologies, researchers can monitor the aggregation process, visualize protein aggregates and analyze their properties. Provides practical examples of nanoimaging research from leading molecular biology, cell biology, protein chemistry, biotechnology, genetics, and pharmaceutical labs Includes over 200 color images to illustrate the power of various nanoimaging technologies Focuses on nanoimaging techniques applied to protein misfolding and aggregation in molecular medicine

Using Old Solutions to New Problems-Marianna Kulka 2013-06-19 The medicinal use of plants, animals and microorganisms has been a part of human evolution and likely began before recorded history. Is it possible that this knowledge can be used to create powerful new drugs and solve some of the human health problems facing us today? This book is a collection of an expert team of agronomists, chemists, biologists and policy makers who discuss some of the processes involved in developing a naturally-sourced bioactive compound into a drug therapy. These experts define a natural compound and elucidate the processes required to find, extract and define a

naturally-derived bioactive molecule. Finally, they describe the necessity for understanding the fundamental mechanisms of disease before applying bioactive molecules in bioassay-guided drug discovery platforms.

Protein Folding Disorders Of The Central Nervous System-Ghiso Jorge A 2017-09-15 This exciting new book explores the dark side of the molecular protein assembly bringing an updated view of how failures in the homeostatic mechanisms that efficiently regulate protein folding leads to the accumulation of structurally abnormal pathogenic assemblies, encompassing an emerging group of diseases collectively known as "Protein Folding Disorders." This complex and diverse group of chronic and progressive entities are bridged together by their relationship to structural transitions in the native state of specific proteinaceous components, which for reasons poorly understood, convert into polymeric aggregates that generate poorly soluble tissue deposits and which are considered today the culprit of the disease pathogenesis in their respective diseases. Despite the diversity in the amino acid sequence of the different proteins involved in these heterogeneous disorders, all the pathologic conformers can trigger cascades of events ultimately resulting in cell dysfunction and death with devastating clinical consequences in many of the most precious aspects of human existence including personality, cognition, memory, and skilled movements. This book, which is composed of a compilation of chapters authored by outstanding and well-published scientists in the respective fields currently performing active investigations at world renowned universities and research centers, focuses on the growing number of diseases associated with protein misfolding in the central nervous system. Individual chapters are dedicated to the most common neurodegenerative diseases associated with protein aggregation/fibrillization focusing on the nature of the pathogenic species and the cellular pathways involved in the molecular pathogenesis of Alzheimer's, Parkinson's, and Huntington's diseases as well as in Amyotrophic Lateral Sclerosis, and Prion disorders. A group of contributions is centered on the current knowledge of the intracellular pathways and subcellular organelles affected by the different disease conditions, while others are focused in the emerging pathogenic role of misfolded subunits assembled into neurotoxic soluble oligomers, and in the novel notion of the transmissibility of the protein misfolded species, an innovative concept until recently only accepted for Prion diseases. Lastly, a different set of chapters is dedicated to the evaluation of novel therapeutic strategies for these devastating diseases. Contents: Misfolding, Aggregation, and Amyloid Formation: The Dark Side of Proteins (Agueda Rostagno and Jorge A Ghiso)Oligomers at the Synapse: Synaptic Dysfunction and Neurodegeneration (Emily Vogler, Matthew Mahavongtrakul, and Jorge Busciglio)Prion-Like Protein Seeding and the Pathobiology of Alzheimer's Disease (Lary C Walker)The Tau Misfolding Pathway to Dementia (Alejandra D Alonso, Leah S Cohen, and Viktoriya Morozova)The Biology and Pathobiology of α-Synuclein (Joel C Watts, Anurag Tandon, and Paul E Fraser)Impact of Loss of Proteostasis on Central Nervous System Disorders (Sentiljana Gumeni, Eleni N Tsakiri, Christina-Maria Cheimonidi, Zoi Evangelakou, Despoina Gianniou, Kostantinos Tallas, Eleni-Dimitra Papanagnou, Aimilia D Sklirou, and Ioannis P Trougakos)Protein Misfolding and Mitochondrial Dysfunction in Amyotrophic Lateral Sclerosis (Giovanni Manfredi and Hibiki Kawamata)Impact of Mitostasis and the Role of the Anti-Oxidant Responses on Central Nervous System Disorders (Sentiljana Gumeni, Eleni N Tsakiri, Christina-Maria Cheimonidi, Zoi Evangelakou, Despoina Gianniou, Kostantinos Tallas, Eleni-Dimitra Papanagnou, Aimilia D Sklirou, and Ioannis P Trougakos)Propagation of Misfolded Proteins in Neurodegeneration: Insights and Cautions from the Study of Prion Disease Prototypes (Robert C C Mercer, Nathalie Daude,

Leucine-Rich Repeat Kinase 2 (LRRK2)-Hardy J. Rideout 2017-03-28 This is the first book to assemble the leading researchers in the field of LRRK2 biology and neurology and provide a snapshot of the current state of knowledge, encompassing all major aspects of its function and dysfunction. The contributors are experts in cell biology and physiology, neurobiology, and medicinal chemistry, bringing a multidisciplinary perspective on the gene and its role in disease. The book covers the identification of LRRK2 as a major contributor to the pathogenesis of Parkinson's Disease. It also discusses the current state of the field after a decade of research, putative normal physiological roles of LRRK2, and the various pathways that have been identified in the search for the mechanism(s) of its induction of neurodegeneration.

Protein Misfolding and Disease-Peter Bross 2003 This volume presents a comprehensive review of the latest thinking about the molecular processes underlying conformational diseases, combined with a remarkable set of biochemical, genomic cellular, and chemical laboratory techniques for studying their genesis and pathologies. The authors apply their carefully refined methods to a variety of metabolic and neurodegenerative disorders, as well as to the aging process. The techniques presented are broadly applicable in many diverse disease contexts and may be used in both diagnosis and research on new treatment strategies. Use proven techniques for the study of diseases attributable to protein misfolding Understand conformational disease mechanisms in metabolic and neurodegenerative disorders Develop new treatment strategies Uncover new ideas and new angles of investigation.

Molecular Neurology-Stephen Waxman 2010-07-26 Why a book on molecular neurology? Molecular neuroscience is advancing at a spectacular rate. As it does so, it is revealing important clues to the pathogenesis and pathophysiology of neurological diseases, and to the therapeutic targets that they present. Medicines work by targeting molecules. The more specific the targeting, the more specific the actions, and the fewer the side effects. Molecular Neurology highlights, for graduate and MD-PhD students, research fellows and research-oriented clinical fellows, and researchers in the neurosciences and other biomedical sciences, the principles underlying molecular medicine as related to neurology. Written by internationally recognized experts, this well-illustrated and well-referenced book presents the most up-to-date principles and disease examples relevant to molecular neurology, and reviews the concepts, strategies, and latest progress in this field. This book will interest anyone studying the molecular basis of neurology, or developing new therapies in neurology. Describes the newest molecular aspects of neurological disorders Provides an introduction to neurological disorders for basic scientists Updates clinicians and clinical researchers on the most recent developments

TDP-43 and Neurodegeneration-Vijay Kumar 2021-10-23 Aggregates of the TAR DNA binding protein 43 (TDP-43), are hallmark features of the neurodegenerative diseases Amyotrophic Lateral Sclerosis (ALS) and frontotemporal dementia (FTD), with overlapping clinical, genetic and pathological features. TDP-43 and Neurodegeneration: From Bench to Bedside summarizes new findings in TDP-43 pathobiology and proteinopathies. The book summarizes TDP-43’s structure, function, biology, misfolding, aggregation, pathogenesis and therapeutics. It includes autophagy-mediated therapy, role of stress granule, novel genetic, cell culture-based models, systems biology for precision medicine, development of stem cells and mechanism-based therapies that can target ALS and other related neurodegenerative diseases. This book is written for neuroscientists, neurologists, clinicians, advanced graduate students, drug discovery researchers, as well as cellular and molecular biologists involved in ALS, motor neuron disease (MND) and other neurodegenerative disorders. Reviews TDP-43 structure, folding, function, and pathology Identifies TDP-43 role in ALS, FTP, and other neurodegenerative diseases Presents a systems and precision biology perspective of TDP-43 Discusses therapeutics of TDP-43 proteinopathies Translates bench research to application bedside

Frontiers in Protein Structure, Function, and Dynamics-Dev Bukhsh Singh 2020-07-02 This book discusses a broad range of basic and advanced topics in the field of protein structure, function, folding, flexibility, and dynamics. Starting with a basic introduction to protein purification, estimation, storage, and its effect on the protein structure, function, and dynamics, it also discusses various experimental and computational structure determination approaches; the importance of molecular interactions and water in protein stability, folding and dynamics; kinetic and thermodynamic parameters associated with protein-ligand binding; single molecule techniques and their applications in studying protein folding and aggregation; protein quality control; the role of amino acid sequence in protein aggregation; muscarinic acetylcholine receptors, antimuscarinic drugs, and their clinical significances. Further, the book explains the current understanding on the therapeutic importance of the enzyme dopamine beta hydroxylase; structural dynamics and motions in molecular motors; role of cathepsins in controlling degradation of extracellular matrix during disease states; and the important structure-function relationship of iron-binding proteins, ferritins. Overall, the book is an important guide and a comprehensive resource for understanding protein structure, function, dynamics, and interaction.

Dancing Protein Clouds: Intrinsically Disordered Proteins in the Norm and Pathology-Vladimir Uversky 2019-09-15 "Dancing protein clouds: Intrinsically disordered proteins in the norm and pathology" represents a set of selected studies on a variety of research topics related to intrinsically disordered proteins. Topics in this update include structural and functional characterization of several important intrinsically disordered proteins, such as 14-3-3 proteins and their partners, as well as proteins from muscle sarcomere; representation of intrinsic disorder-related concept of protein structure-function continuum; discussion of the role of intrinsic disorder in phenotypic switching; consideration of the role of intrinsically

disordered proteins in the pathogenesis of neurodegenerative diseases and cancer; discussion of the roles of intrinsic disorder in functional amyloids; demonstration of the usefulness of the analysis of translational diffusion of unfolded and intrinsically disordered proteins; consideration of various computational tools for evaluation of functions of intrinsically disordered regions; and discussion of the role of shear stress in the amyloid formation of intrinsically disordered regions in the brain. Provides some recent studies on the intrinsically disordered proteins and their functions, as well as on the involvement of intrinsically disordered proteins in pthogenesis of various diseases Contains numerous illustrative materials (color figures, diagrams, and tables) to help the readers to delve in the information provided Includes contributions from recognized experts in the field

Protein and Peptide Folding, Misfolding, and Non-Folding-Reinhard Schweitzer-Stenner 2012-02-08 Sheds new light on intrinsically disordered proteins andpeptides, including their role in neurodegenerative diseases With the discovery of intrinsically disordered proteins andpeptides (IDPs), researchers realized that proteins do notnecessarily adopt a well defined secondary and tertiary structurein order to perform biological functions. In fact, IDPs playbiologically relevant roles, acting as inhibitors, scavengers, andeven facilitating DNA/RNA-protein interactions. Due to theirpropensity for self-aggregation and fibril formation, some IDPs areinvolved in neurodegenerative diseases such as Parkinson's andAlzheimer's. With contributions from leading researchers, this text reviewsthe most recent studies, encapsulating our understanding of IDPs.The authors explain how the growing body of IDP research isbuilding our knowledge of the folding process, the binding ofligands to receptor molecules, and peptide self-aggregation.Readers will discover a variety of experimental, theoretical, andcomputational approaches used to better understand the propertiesand function of IDPs. Moreover, they'll discover the role of IDPsin human disease and as drug targets. Protein and Peptide Folding, Misfolding, and Non-Folding beginswith an introduction that explains why research on IDPs hassignificantly expanded in the past few years. Next, the book isdivided into three sections: Conformational Analysis of Unfolded States Disordered Peptides and Molecular Recognition Aggregation of Disordered Peptides Throughout the book, detailed figures help readers understandthe structure, properties, and function of IDPs. References at theend of each chapter serve as a gateway to the growing body ofliterature in the field. With the publication of Protein and Peptide Folding, Misfolding,and Non-Folding, researchers now have a single place to discoverIDPs, their diverse biological functions, and the many disciplinesthat have contributed to our evolving understanding of them.

Neurodegenerative Diseases and Metal Ions-Astrid Sigel 2006-07-11 About the Series... Metal Ions in Life Sciences links coordination chemistry and biochemistry in their widest sense and thus increases our understanding of the relationship between the chemistry of metals and life processes. The series reflects the interdisciplinary nature of Biological Inorganic Chemistry and coordinates the efforts of scientists in fields like biochemistry, inorganic chemistry, coordination chemistry, molecular and structural biology, enzymology, environmental chemistry, physiology, toxicology, biophysics, pharmacy, and medicine. Consequently, the volumes are an essential source for researchers active in these and related fields as well as teachers preparing courses, e.g., in Bioinorganic Chemistry. About this Book... Volume 1, devoted solely to the vital research area concerning the role of metal ions in neurodegenerative diseases, offers in 15 stimulating chapters an authoritative and timely view of this fascinating subject. Written by 41 internationally recognized experts, Neurodegenerative Diseases and Metal Ions highlights, supported by 130 illustrations, the recent progress made in understanding the role metal ions play in diseases like transmissible spongiform encephalopathies (Creutzfeldt-Jakob and related diseases), Alzheimer's, Parkinson's, Huntington's, Wilson's and Menkes' diseases, as well as in familial amyotrophic lateral sclerosis and others. The interplay between metal ions, catecholamines and the formation of reactive oxygen species resulting in oxidative stress is considered, as is the metalloneurochemistry of zinc and the neurotoxicity of aluminum, cadmium, lead, and mercury. The need for novel drugs which manipulate metal-centered neuropathology is emphasized.

Autophagy: Cancer, Other Pathologies, Inflammation, Immunity, Infection, and Aging-M. A. Hayat 2016-12-28 Autophagy: Cancer, Other Pathologies, Inflammation, Immunity, Infection, and Aging is an eleven volume series that discusses in detail all aspects of autophagy machinery in the context of health, cancer, and other pathologies. Autophagy maintains homeostasis during starvation or stress conditions by balancing the synthesis of cellular components and their deregulation by autophagy. This series discusses the characterization of autophagosome-enriched vaccines and its efficacy in cancer immunotherapy. Autophagy serves to maintain healthy cells, tissues, and organs, but also promotes cancer survival and growth of established tumors. Impaired or deregulated autophagy can also contribute to disease pathogenesis. Understanding the importance and necessity of the role of autophagy in health and disease is vital for the studies of cancer, aging, neurodegeneration, immunology, and infectious diseases. Comprehensive and forward-thinking, these books offer a valuable guide to cellular processes while also inciting researchers to explore their potentially important connections. Presents the most advanced information regarding the role of the autophagic system in life and death Examines whether autophagy acts fundamentally as a cell survivor or cell death pathway or both Introduces new, more effective therapeutic strategies in the development of targeted drugs and programmed cell death, providing information that will aid in preventing detrimental inflammation Features recent advancements in the molecular mechanisms underlying a large number of genetic and epigenetic diseases and abnormalities, including atherosclerosis and CNS tumors, and their development and treatment Includes chapters authored by leaders in the field around the globe—the broadest, most expert coverage available

Misbehaving Proteins-Regina Murphy 2007-10-12 This text provides an up-to-date collection of theoretical and experimental studies into protein folding, misfolding, aggregation, and stability. Additionally, issues faced during the development of protein products are illustrated. It contains an introductory chapter for readers new to the protein folding field. The book provides a thorough and clear discussion of computational approaches to understanding and modeling protein aggregation.

Autophagy: Biology and Diseases-Zheng-Hong Qin 2019-11-27 This book series consists of 3 volumes covering the basic science (Volume 1), clinical science (Volume 2) and the technology and methodology (Volume 3) of autophagy. Volume 1 focuses on the biology of autophagy, including the signaling pathways, regulating processes and biological functions. Autophagy is a fundamental physiological process in eukaryotic cells. It not only regulates normal cellular homeostasis, and organ development and function, but also plays an important role in the pathogenesis of a wide range of human diseases. Thanks to the rapid development of molecular biology and omic technologies, research on autophagy has boomed in recent decades, and more and more cellular and animal models and state-of the-art technologies are being used to shed light on the complexity of signaling networks involved in the autophagic process. Further, its involvement in biological functions and the pathogenesis of various diseases has attracted increased attention around the globe. Presenting cutting-edge knowledge, this book series is a useful reference resource for researchers and clinicians who are working on or interested in autophagy.

Molecular Chaperones in Health and Disease-Matthias Gaestel 2006-01-18 Molecular chaperones are involved in a wide variety of essential cellular processes in living cells. A subset of molecular chaperones have been initially described as heat shock proteins protecting cells from stress damage by keeping cellular proteins in a folding competent state and preventing them from irreversible aggregation. Later it became obvious that molecular chaperones are also expressed constitutively in the cell and are involved in complex processes such as protein synthesis, intracellular protein transport, post-translational modification and secretion of proteins as well as receptor signalling. Hence, it is not surprising that molecular chaperones are implicated in the pathogenesis of many relevant diseases and could be regarded as potential pharmacological targets. Starting with the analysis of the mode of action of chaperones at the molecular, cellular and organismic level, this book will then describe specific aspects where modulation of chaperone action could be of pharmacological and therapeutic interest.

Cellular Osmolytes-Laishram Rajendrakumar Singh 2017-05-16 This book provides essential information on improving protein folding/stability, which is a result of the balance between the intra-molecular interactions of protein functional groups and their interactions with the solvent environment. The protein folding solvent environment mainly consists of salts, small molecule compounds, metabolites, molecular chaperones and other chemical species. Therefore, subtle change in the composition of the environment will alter the protein folding process. The importance of the solvent environment in protein folding is precisely due to the fact that various disease-causing proteopathies can be reversed by manipulating the solvent environment of the malfolded proteins. Hostile environmental stresses represent one of the basic causes of such challenges in protein folding or misfolding. Since cells commonly encounter extreme environmental fluctuations, it is crucial that they equip themselves with strategies to circumvent the hostile environmental conditions. Nature has developed many strategies to ensure that the complex and challenging protein folding reaction occurs with

adequate efficiency and fidelity for the success of the organism. Among the strategies employed in a wide range of species and cell types is the elaboration of small organic molecules called osmolytes. Additionally, recent advances have also revealed that certain specific osmolytes might be key biomarkers of cancer, infectious diseases and vaccine flocculation. In fact, a large pool of data has been generated regarding their potential for the therapeutic intervention of neurodegenerative diseases and other metabolic disorders caused by protein aggregation or proteostasis failure. Reflecting the multiple applications of these small molecules in the health and other industries, this book combines contributions by respected leaders in the field and will help to inspire college students, basic researchers, and clinicians to translate these biological roles of osmolytes into clinical practice. It will also shed light on some important future prospects of osmolytes like their role as drug excipients and provide a deeper understanding of their mechanism of action in the prevention of neuro-degenerative diseases.

The Molecular and Cellular Basis of Neurodegenerative Diseases-Michael S. Wolfe 2018-03-29 The Molecular and Cellular Basis of Neurodegenerative Diseases: Underlying Mechanisms presents the pathology, genetics, biochemistry and cell biology of the major human neurodegenerative diseases, including Alzheimer’s, Parkinson’s, frontotemporal dementia, ALS, Huntington’s, and prion diseases. Edited and authored by internationally recognized leaders in the field, the book's chapters explore their pathogenic commonalities and differences, also including discussions of animal models and prospects for therapeutics. Diseases are presented first, with common mechanisms later. Individual chapters discuss each major neurodegenerative disease, integrating this information to offer multiple molecular and cellular mechanisms that diseases may have in common. This book provides readers with a timely update on this rapidly advancing area of investigation, presenting an invaluable resource for researchers in the field. Covers the spectrum of neurodegenerative diseases and their complex genetic, pathological, biochemical and cellular features Focuses on leading hypotheses regarding the biochemical and cellular dysfunctions that cause neurodegeneration Details features, advantages and limitations of animal models, as well as prospects for therapeutic development Authored by internationally recognized leaders in the field Includes illustrations that help clarify and consolidate complex concepts

Protein Homeostasis Diseases-Angel L. Pey 2020-02-13 Protein Homeostasis Diseases: Mechanisms and Novel Therapies offers an interdisciplinary examination of the fundamental aspects, biochemistry and molecular biology of protein homeostasis disease, including the use of natural and pharmacological small molecules to treat common and rare protein homeostasis disorders. Contributions from international experts discuss the biochemical and genetic components of protein homeostasis disorders, the mechanisms by which genetic variants may cause loss-of-function and gain-of-toxic-function, and how natural ligands can restore protein function and homeostasis in genetic diseases. Applied chapters provide guidance on employing high throughput sequencing and screening methodologies to develop pharmacological chaperones and repurpose approved drugs to treat protein homeostasis disorders. Provides an interdisciplinary examination of protein homeostasis disorders, with an emphasis on treatment strategies employing small natural and pharmacological ligands Offers applied approaches in employing high throughput sequencing and screening to develop pharmacological chaperones to treat protein homeostasis disease Gathers expertise from a range of international chapter authors who work across various biological methods and disease specific disciplines of relevance

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Prions-Claudio Soto 2005-12-20 Prion-related diseases, known as transmissible spongiform encephalopathies (TSEs), are infectious, fatal neurodegenerative disorders for which there is no cure, treatment, nor even a means for early diagnosis. The horrific advent of Mad Cow Disease -- transmitted to humans through eating meat from steers sickened by bovine spongiform encephalopathy

Protein Quality Control in Neurodegenerative Diseases-Richard I. Morimoto 2012-12-13 The health of the proteome depends upon protein quality control to regulate the proper synthesis, folding, translocation, and clearance of proteins. The cell is challenged constantly by environmental and physiological stress, aging, and the chronic expressions of disease associated misfolded proteins. Substantial evidence supports the hypothesis that the expression of damaged proteins initiates a cascade of molecular events that leads to Alzheimer's disease, Parkinson's disease, amyotrophic lateral sclerosis, Huntington's disease, and other diseases of protein conformation.

Protein Homeostasis-Richard I. Morimoto 2012 Proper expression, folding, transport, and clearance of proteins is critical for cell function. Chaperones and enzymes that posttranslationally assist newly synthesized proteins help ensure that they fold correctly or are degraded. Translocation machineries, proteasomes, and autophagic activities help to localize and degrade proteins as necessary. Stress and aging can cause such mechanisms to become dysfunctional or overloaded, resulting in the accumulation and aggregation of misfolded proteins a feature of numerous neurodegenerative conditions. Written and edited by experts in the field, this collection from Cold Spring Harbor Perspectives in Biology covers the entire spectrum of protein homeostasis in healthy cells and the diseases that result when control of protein production, protein folding, and protein degradation goes awry. The contributors examine the physical biochemistry of protein folding and the roles of the various cellular compartments in protein quality control, as well as approaches for ameliorating protein misfolding and aggregation diseases. Including discussions of specific disorders such as Alzheimer's disease, Huntington's disease, and prion diseases, this book is an essential reference for not only molecular and cellular biologists but also medical scientists wishing to understand the pathological consequences of and potential therapies for protein homeostasis deficiencies in common human diseases.

Folding for the Synapse-Andreas Wyttenbach 2010-10-22 Folding for the Synapse addresses the current view on how protein folding and misfolding, controlled by molecular chaperones, contribute to synapse function and dysfunction. Molecular chaperones have been studied in relation to de novo protein folding, but there is increasing awareness that chaperone function is required for the regulation of protein dynamics when functioning physiologically as an isolated moiety or part of a protein complex. This book will introduce both important concepts of folding machineries and give examples of the biological relevance of further chaperone functions.

Protein Folding, Misfolding and Aggregation-Victor Muñoz 2008-06-24 Protein folding and aggregation is the process by which newly synthesized proteins fold into the specific three-dimensional structures defining their biologically active states. It has always been a major focus of research in biochemistry and has often been seen as the unsolved second part of the genetic code. In the last 10 years we have witnessed a quantum leap in the research in this exciting area. Computational methods have improved to the extent of making possible to simulate the complete folding process of small proteins and the early stages of protein aggregation. Experimental methods have evolved to permit resolving fast processes of folding reactions and visualizing single molecules during folding. The findings from these novel experiments and detailed computer simulations have confirmed the main predictions of analytical theory of protein folding. In summary, protein folding research has finally acquired the status of a truly quantitative science, paving the way for more exciting developments in the near future. This unique book covers all the modern approaches and the many advances experienced in the field during the last 10 years. There is also much emphasis on computational methods and studies of protein aggregation which have really flourished in the last decade. It includes chapters in the areas that have witnessed major developments and are written by top experts including:computer simulations of folding, fast folding, single molecule spectroscopy, protein design, aggregation studies (both computational and experimental). Readers will obtain a unique perspective of the problems faced in the biophysical study of protein conformational behaviour in aqueous solution and how these problems are being solved with a multidisciplinary approach that combines theory, experiment and computer simulations. Protein Folding, Misfolding and Aggregation Classical Themes and Novel Approaches is essential reading for graduate students actively involved in protein folding research, other scientists interested in the recent progress of the field and instructors revamping the protein folding section of their biochemistry and biophysics courses.

Heat Shock Proteins in Neuroscience-Alexzander A. A. Asea 2019-10-30 The book Heat Shock Proteins in Neuroscience provides the most comprehensive review on contemporary knowledge on the role of HSP in signaling pathways relevant to a number of diseases. Using an integrative approach, the contributors provide a synopsis of novel mechanisms, signal transduction pathways. To enhance the ease of reading and comprehension, this book has been subdivided into various section including; Section I, reviews current progress on our understanding of Neurological Aspects of HSP; Section II, focuses on Aspects of HSP in Neurodegenerative Diseases and Disorders, Section III, emphasizes the importance of HSP in Multiple

Sclerosis; Section IV, reviews critical Aspects of HSP in Alzheimer's Disease and Section V, gives a comprehensive update of the Development of HSP-Based Therapies for Neurological Disorders. Key basic and clinical research laboratories from major universities, academic medical hospitals, biotechnology and pharmaceutical laboratories around the world have contributed chapters that review present research activity and importantly project the field into the future. The book is a must read for starters and professionals in the fields of Neurology and Neurosciences, Translational Medicine, Clinical Research, Human Physiology, Biotechnology, Cell & Molecular Medicine, Pharmaceutical Scientists and Researchers involved in Drug Discovery.

Oxidative Folding of Proteins-Matthias J Feige 2018-07-30 The formation of disulphide bonds is probably the most influential modification of proteins. These bonds are unique among post-translational modifications of proteins as they can covalently link cysteine residues far apart in the primary sequence of a protein. This has the potential to convey stability to otherwise marginally stable structures of proteins. However, the reactivity of cysteines comes at a price: the potential to form incorrect disulphide bonds, interfere with folding, or even cause aggregation. An elaborate set of cellular machinery exists to catalyze and guide this process: facilitating bond formation, inhibiting unwanted pairings and scrutinizing the outcomes. Only in recent years has it become clear how intimately connected this cellular machinery is with protein folding helpers, organellar redox balance and cellular homeostasis as a whole. This book comprehensively covers the basic principles of disulphide bond formation in proteins and describes the enzymes involved in the correct oxidative folding of cysteine-containing proteins. The biotechnological and pharmaceutical relevance of proteins, their variants and synthetic replicates is continuously increasing. Consequently this book is an invaluable resource for protein chemists involved in realted research and production.

Quality Control of Cellular Protein in Neurodegenerative Disorders-Uddin, Md. Sahab 2020-02-14 Protein misfolding and aggregation are hallmarks of several neurodegenerative proteinopathies. Though multiple factors like aging, oxidative stress, mitochondrial dysfunction, proteotoxic insults, genetic inconsistency, etc. are responsible for the dysfunction of the neuronal protein quality control system, targeting protein quality control has become an auspicious approach to halt the propagation of neurodegeneration. Quality Control of Cellular Protein in Neurodegenerative Disorders provides diverse aspects exploring the role of the protein quality control in neurodegenerative disorders and potential therapeutic strategies to combat the development and propagation of neurodegeneration. Featuring coverage on a broad range of topics such as molecular chaperones, protein misfolding, and stress signaling, this book is ideally designed for neurobiologists, neuropsychologists, neurophysiologists, medical professionals, neuropathologists, researchers, academicians, students, and practitioners engaged in studies of the protein quality control system in neuronal cells.

Fundamentals of Neurodegeneration and Protein Misfolding Disorders-Martin Beckerman 2015-11-06 This unique text introduces students and researchers to the world of misfolded proteins, toxic oligomers, and amyloid assemblages, and the diseases of the brain that result. During the past few years the connections between failures in protein quality control and neurological disorders have been reinforced and strengthened by discoveries on multiple fronts. These findings provide novel insights on how amyloidogenic oligomers and fibrils form, interconvert from one state to another, and propagate from cell to cell and region to region. Starting with protein folding and protein quality control basics, the reader will learn how misfolded proteins can cause diseases ranging from prion diseases to Alzheimer's disease and Parkinson's disease to Huntington's disease, amyotrophic lateral sclerosis and frontotemporal lobar degeneration. Authoritative but written in a clear and engaging style, Fundamentals of Neurodegeneration and Protein Misfolding Disorders addresses one of today's forefront areas of science and medicine. The text emphasizes the new groundbreaking biophysical and biochemical methods that enable molecular-level explorations and the conceptual breakthroughs that result. It contains separate chapters on each of the major disease classes. Special emphasis is placed on those factors and themes that are common to the diseases, especially failures in synaptic transmission, mitochondrial control, and axonal transport; breakdowns in RNA processing; the potential role of environmental factors; and the confounding effects of neuroinflammation. The book is ideal for use in teaching at the advanced undergraduate and graduate levels, and serves as a comprehensive reference for a broad audience of students and researchers in neuroscience, molecular biology, biological physics and biomedical engineering.

Biology of Serpins-James C. Whisstock 2011 Serpins are a group of proteins with similar structures that were first identified as a set of proteins able to inhibit proteases. The acronym serpin was originally coined because many serpins inhibit chymotrypsin-like serine proteases. This volume of Methods in Ezymology is split into 2 parts and comprehensively covers the subject.

Drug Discovery Approaches for the Treatment of Neurodegenerative Disorders-Adeboye Adejare 2016-09-20 Drug Discovery Approaches for the Treatment of Neurodegenerative Disorders: Alzheimer's Disease examines the drug discovery process for neurodegenerative diseases by focusing specifically on Alzheimer's Disease and illustrating the paradigm necessary to ensure future research and treatment success. The book explores diagnosis, epidemiology, drug discovery strategies, current therapeutics, and much more to provide a holistic approach to the discovery, development, and treatment of Alzheimer's Disease. Through its coverage of the latest research in targeted drug design, preclinical studies, and mouse and rat models, the book is a must-have resource for all pharmaceutical scientists, pharmacologists, neuroscientists, and clinical researchers working in this area. It illustrates why these drugs tend to fail at the clinical stage, and examines Alzheimer's Disease within the overall context of improving the drug discovery process for the treatment of other neurodegenerative disorders. Provides a compilation of chemical considerations required in drug discovery for the treatment of neurodegenerative disorders Examines different classes of compounds currently being used in discovery and development stages Explores in vitro and in vivo models with respect to their ability to translate these models to human conditions Distills the most significant information across multiple areas of Alzheimer's disease research to provide a single, comprehensive, and balanced resource

Protein Folding and Metal Ions-Cláudio M. Gomes 2016-04-19 The role of metal ions in protein folding and structure is a critical topic to a range of scientists in numerous fields, particularly those working in structural biology and bioinorganic chemistry, those studying protein folding and disease, and those involved in the molecular and cellular aspects of metals in biological systems. Protein Folding and Metal Ions: Mechanisms, Biology and Disease presents the contributions of a cadre of international experts who offer a comprehensive exploration of this timely subject at the forefront of current research. Divided into four sections, this volume: Provides case study examples of protein folding and stability studies in particular systems or proteins that comprise different metal ions of co-factors Reviews the proteins that shuttle metal ions in the cell to a particular target metalloprotein Illustrates how metal binding can be connected to pathological protein conformations in unrelated diseases, from cancer to protein deposition disorders such as Parkinson's disease Addresses protein redesign of metal-containing proteins by computational methods, folding simulation studies, and work on model peptides — dissecting the relative energetic contribution of metals sites to protein folding and stability Together, the 13 chapters in this text cogently describe the state of the science today, illuminate current challenges, propose future possibilities, and encourage further study in this area that offers much promise especially with regard to novel approaches to the treatment of some of the most challenging and tragic diseases.

Protein Chaperones and Protection from Neurodegenerative Diseases-Stephan N. Witt 2011-09-09 How protein chaperones protect cells from neurodegenerative diseases Including contributions from leading experts, Protein Chaperones and Protection from Neurodegenerative Diseases provides an in-depth exploration of how protein chaperones are involved in shielding cells from toxic aggregated or misfolded protein states that cause ALS, Parkinson's, and related diseases. Examining how different protein chaperones ameliorate the toxicity of proteins that are known to cause neurodegenerative damage, the book addresses both research and clinical perspectives on chaperone and anti-chaperone properties. The intersection of molecular chaperones and neurodegeneration is an intensely studied area, partly because of the potential for manipulating the expression of molecular chaperones to thwart the progression of debilitating diseases, and partly because of the ever-aging global population. Discussing the potential to harness the power of protein chaperones, and future directions for research, discovery, and therapeutics, this book is essential reading for scientists working in the fields of biochemistry, molecular medicine, pharmacology and drug discovery, biotechnology and pharmaceutical companies, advanced students, and anyone interested in this cutting-edge topic.