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EDITORIAL

Twist and Sheet: Variations on the Theme of Amyloid

Amyloidoses are protein deposition diseases in which normally soluble proteins or peptides polymerize to form insoluble assemblies. The deposition process can occur both intra- and extracellularly and may be anatomically localized or systemic. The results of amyloidosis are tissue injury and, often death. Among the more well known of the approximately 20 clinically defined amyloidoses are Alzheimer's disease and bovine spongiform encephalopathy (also known as "mad cow disease"). One of the defining features of all amyloid deposits is their ability to bind amyloidophilic dyes such as Congo red. This ability is a consequence of the structural organization of the abnormally assembled protein or peptide, which invariably involves extensive β -pleated sheet formation. The β -sheets can be organized into a variety of structures, including plates, ribbons, and fibers, often exhibiting twisted or coiled morphologies. The substructure of these assemblies may consist of either discrete subunits or a continuous β -sheet.

Recent studies have shown that many, if not all, amyloids share a common cross- β core structure. In addition, a surprisingly large number of proteins and peptides, under appropriate conditions, have been shown to form amyloid fibrils, suggesting that this capacity is intrinsic to most proteins. Study of fibrillogenesis thus may have relevance not only in developing therapies to treat amyloidoses, but also for elucidating fundamental mechanisms of protein folding and assembly.

In this special issue, we have provided a set of reviews, perspectives, and original articles that reflect the diversity of research foci in studies of the structural biology of amyloid. This diversity is reflected in the range of natural and synthetic proteins and peptides discussed, which include amyloid β -protein, tau, immunoglobulin light chain, α -synuclein, transthyretin, yeast prion, scrapie prion, lysozyme, islet amyloid polypeptide, and betabellin. An equally broad range of experimental questions and methodological approaches are covered. These include electron microscopic, X-ray diffraction, and NMR studies of amyloid structure, studies of the kinetics and thermodynamics of the fibrillogenesis process, the roles and effects of protein, proteoglycan, and chemical modulators of amyloid assembly, the biological activities of amyloid proteins and peptides, and the search for effective fibrillogenesis inhibitors.

No single volume can present the entire range of worthy and exciting research in an area and we regret that space limitations prevented inclusion of fine work from many laboratories. However, we trust that the articles we have selected will provide the reader with an enriching and stimulating exposure to key areas in amyloid research.

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