The Chaperonopathies Diseases with Defective Molecular Chaperones
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In the mid 1950's the Nobel Prize Laureate Christian B. Anfinsen from his research on the folding of ribonuclease A,[1] began to concentrate on the problem of the relationship between structure and function in enzymes. On the basis of studies on ribonuclease, he proposed that the information determining the tertiary structure of a protein resides in the chemistry of its amino acid sequence. He also elegantly showed that, after cleavage of disulfide bonds and disruption of tertiary structure, many proteins could spontaneously refold to their native forms.[2] However, in the following years several studies demonstrated that this assertion is not valid for all synthetized proteins thus suggesting that some proteins may violate the Anfinsen's dogma.[3] In fact, some highly specific 'steric chaperones' do convey unique structural (steric) information onto proteins, which cannot be folded spontaneously. This finding changed dramatically the way of studying protein synthesis and their maturation following the translation process. In addition, further studies demonstrated that impairment of the chaperone machinery might lead to various pathologies, which were extensively studied by prof. Alberto J.L. Macario and his collaborators who termed such conditions as "chaperonopathies".[4] However, only recently such diseases were classified and molecularly characterized.

This book describes chaperones with demonstrated chaperoning roles and other molecules related to them evolutionarily and/or functionally. Furthermore, it also focuses on conditions with chaperone malfunction and associated pathologies.

In the first two chapters the authors drive the readers in the fascinating world of chaperones by using a clear language and direct messages and thus making this book suitable also for those who are not strictly confident with biochemistry or molecular biology. From chapter 3 to 8 the authors classify chaperonopathies and describe the molecular mechanisms underlying specific diseases such as autoimmune diseases, chronic obstructive pulmonary disease, inflammatory bowel diseases and cancer. In the last chapter the authors face one of the most intriguing and challenging topics of chaperonology that is the extracellular chaperones. In particular several questions are still open and matter of scientific debate such as the molecular mechanism/s of chaperones' secretion and clinical/biological significance of chaperones in the extracellular space.

This book is a very useful tool not only for students but also for all those researchers working in the field or would like to receive an update regarding this very hot topic.

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