

# Preface

ATP, the universal energy source of life, is in large part synthesized in the mitochondria by the oxidative phosphorylation (OxPhos) process. After identification of the mitochondrial metabolic pathways and the complexes of OxPhos, i.e., respiratory chain complexes I–IV and ATP synthase (complex V) in the second half of the last century, bioenergetic research concentrated on bacterial OxPhos enzymes, which contain fewer protein subunits but fulfill the same catalytic activity. Mitochondria came again into focus after identification of new mitochondrial functions like apoptosis, heat generation via uncoupling proteins and passive proton leak, formation and degradation of reactive oxygen species (ROS), and their involvement in numerous “mitochondrial diseases.” This book focuses on new developments in mitochondrial morphology, biogenesis, and evolution, as well as in structure and regulatory functions of the enzyme complexes of OxPhos by experts in their fields. Particular emphasis is given to the oxygen consuming enzyme of the respiratory chain, cytochrome *c* oxidase, representing a controlling step of OxPhos. The book also opens up a new field of research, the physiological regulation of mitochondrial energy synthesis. In addition, many newly identified mitochondrial diseases based on nuclear-encoded genes and proteins are reviewed.

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