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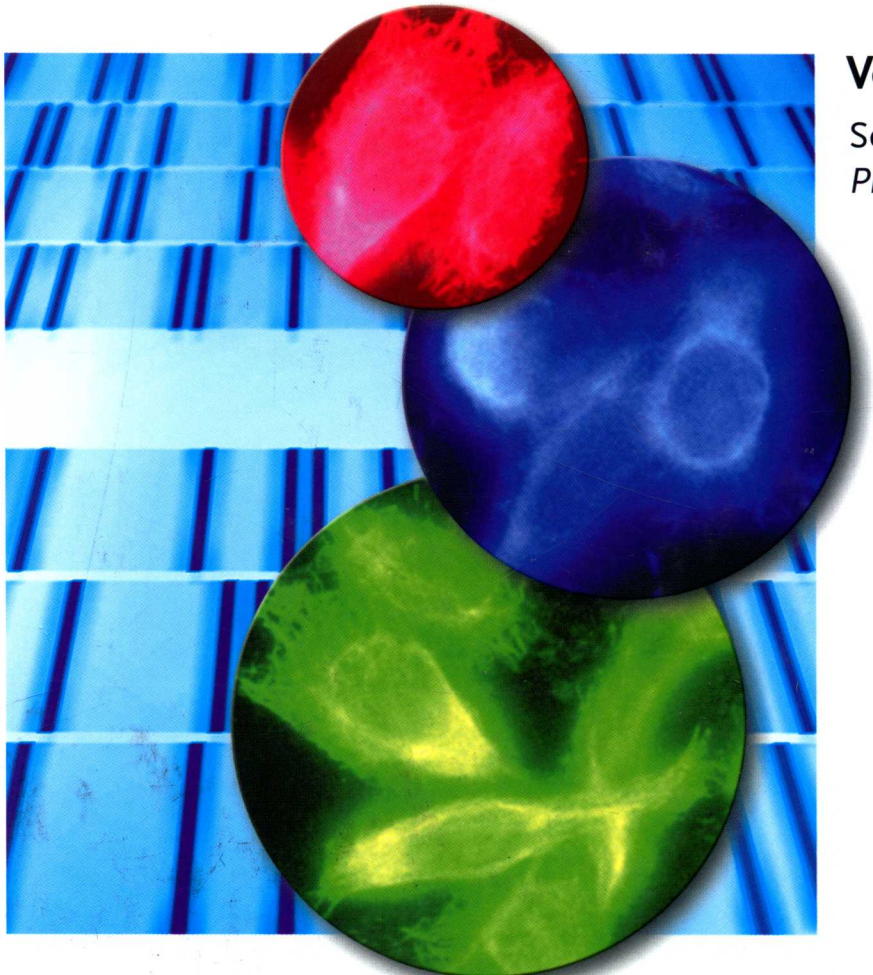
Encyclopedia of Molecular Cell Biology and Molecular Medicine

Edited by Robert A. Meyers



Volume 11

Second Edition

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Second Edition

Volume 11
Proteasomes to Receptor, Transporter and Ion Channel Diseases

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Preface

The *Encyclopedia of Molecular Cell Biology and Molecular Medicine*, which is the successor and second edition of the *Encyclopedia of Molecular Biology and Molecular Medicine* (VCH Publishers, Weinheim), covers the molecular and cellular basis of life at a university and professional researcher level. The first edition, published in 1996–97, was very successful and is being used in libraries around the world. This second edition will almost double the first edition in length and will comprise the most detailed treatment of both molecular cell biology and molecular medicine available today. The Board Members and I believe that there is a serious need for this publication, even in view of the vast amount of information available on the World Wide Web and in text books and monographs. We feel that there is no substitute for our tightly organized and integrated approach to selection of articles and authors and implementation of peer review standards for providing an authoritative single-source reference for undergraduate and graduate students, faculty, librarians, and researchers in industry and government.

Our purpose is to provide a comprehensive foundation for the expanding number of molecular biologists, cell biologists, pharmacologists, biophysicists, biotechnologists, biochemists, and physicians, as well as for those entering molecular cell biology and molecular medicine from majors or careers in physics, chemistry, mathematics, computer science, and engineering. For example, there is an unprecedented demand for physicists, chemists, and computer scientists who will work with biologists to define the genome, proteome, and interactome through experimental and computational biology.

The Board Members and I first divided the entire study of molecular cell biology and molecular medicine into primary topical categories and further defined each of these into subtopics. The following is a summary of the topics and subtopics:

- *Nucleic Acids:* amplification, disease genetics overview, DNA structure, evolution, general genetics, nucleic acid processes, oligonucleotides, RNA structure, RNA replication and transcription.
- *Structure Determination Technologies Applicable to Biomolecules:* chromatography, labeling, large structures, mapping, mass spectrometry, microscopy, magnetic resonance, sequencing, spectroscopy, X-ray diffraction.
- *Biochemistry:* carbohydrates, chirality, energetics, enzymes, biochemical genetics, inorganics, lipids, mechanisms, metabolism, neurology, vitamins.

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- *Proteins, Peptides, and Amino Acids:* analysis, enzymes, folding, mechanisms, modeling, peptides, structural genomics (proteomics), structure, types.
- *Biomolecular Interactions:* cell properties, charge transfer, immunology, recognition, senses.
- *Cell Biology:* developmental cell biology, diseases, dynamics, fertilization, immunology, organelles and structures, senses, structural biology, techniques.
- *Molecular Cell Biology of Specific Organisms:* algae, amoeba, birds, fish, insects, mammals, microbes, nematodes, parasites, plants, viruses, yeasts.
- *Molecular Cell Biology of Specific Organs or Systems:* excretory, lymphatic, muscular, nervous, reproductive, skin.
- *Molecular Cell Biology of Specific Diseases:* cancer, circulatory, endocrinal, environmental stress, immune, infectious, neurological, radiational.
- *Pharmacology:* chemistry, disease therapy, gene therapy, general molecular medicine, synthesis, toxicology.
- *Biotechnology:* applications, diagnostics, gene-altered animals, bacteria and fungi, laboratory techniques, legal, materials, process engineering, nanotechnology, production of classes or specific molecules, sensors, vaccine production.

We then selected some 400 article titles and author or author teams to cover the above topics. Each article is designed as a self-contained treatment which begins with a keyword section including definitions, to assist the scientist or student who is unfamiliar with the specific subject area. The Encyclopedia includes more than 3000 key words, each defined within the context of the particular scientific field covered by the article. In addition to these definitions, the glossary of basic terms found at the back of each volume, defines the most commonly used terms in molecular cell biology. These definitions, along with the reference materials (the genetic code, the common amino acids, and the structures of the deoxyribonucleotides) printed at the back of each volume, should allow most readers to understand articles in the Encyclopedia without referring to a dictionary, textbook, or other reference work. There is, of course, a detailed subject index in Volume 16 as well as a cumulative table of contents and list of authors, as well as a list of scientists who assisted in the development of this Encyclopedia.

Each article begins with a concise definition of the subject and its importance, followed by the body of the article and extensive references for further reading. The references are divided into secondary references (books and review articles) and primary research papers. Each subject is presented on a first-principle basis, including detailed figures, tables and drawings. Because of the self-contained nature of each article, some articles on related topics overlap. Extensive cross-referencing is provided to help the reader expand his or her range of inquiry.

The articles contained in the Encyclopedia include core articles, which summarize broad areas, directing the reader to satellite articles that present additional detail and depth for each subject. The core article Brain Development is a typical example. This 45-page article spans neural induction, early patterning, differentiation, and wiring at a molecular through to cellular and tissue level. It is directly supported, and cross-referenced, by a number of molecular neurobiology satellite articles, for example, Behavior Genes, and further supported by other core presentations, for example,

Developmental Cell Biology; Genetics, Molecular Basis of, and their satellite articles. Another example is the core article on Genetic Variation and Molecular Evolution by Werner Arber. It is supported by a number of satellite articles supporting the evolutionary relatedness of genetic information, for example, Genetic Analysis of Populations.

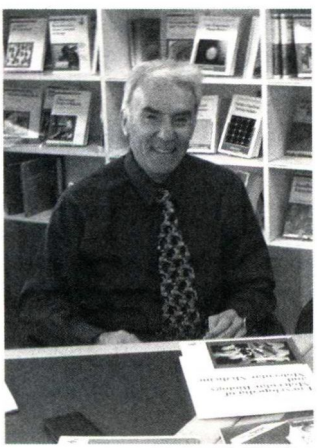
Approximately 250 article titles from the first edition are retained, but rewritten, half by new authors and half by returning authors. Approximately 80 articles on cell biology and 70 molecular biology articles have been added covering areas that have become prominent since preparation of the first edition. Thus, we have compiled a totally updated single source treatment of the molecular and cellular basis of life.

Finally, I wish to thank the following Wiley-VCH staff for their outstanding support of this project: Andreas Sendtko, who provided project and personnel supervision from the earliest phases, and Prisca-Maryla Henheik and Renate Dötzer, who served as the managing editors.

July 2005

Robert A. Meyers
Editor-in-Chief

Editor-in-Chief



Robert A. Meyers

Dr. Meyers earned his Ph.D. in organic chemistry from the University of California Los Angeles, was a post-doctoral fellow at California Institute of Technology and manager of chemical processes for TRW Inc. He has published in *Science*, written or edited 12 scientific books and his research has been reviewed in the *New York Times* and the *Wall Street Journal*. He is one of the most prolific science editors in the world having originated, organized and served as Editor-in-Chief of three editions of the *Encyclopedia of Physical Science and Technology*, the *Encyclopedia of Analytical Chemistry* and two editions of the present *Encyclopedia of Molecular Cell Biology and Molecular Medicine*.

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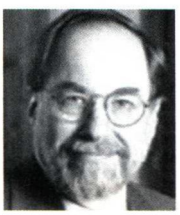
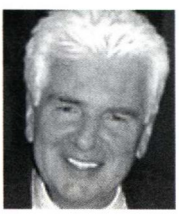
Editorial Board





Paul Greengard
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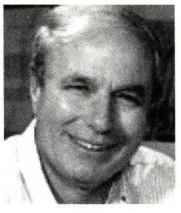
Nobel Prize in Physiology/Medicine for the discoveries concerning signal transduction in the nervous system



Avram Hershko
Technion – Israel Institute of Technology, Haifa, Israel
Nobel Prize in Chemistry for the discovery of ubiquitin-mediated protein degration

Robert Huber
Max Planck Institute of Biochemistry, Martinsried, Germany
Nobel Prize in Chemistry for the determination of the three-dimensional structure of a photosynthetic reaction centre

Aaron Klug
MRC Laboratory of Molecular Biology Cambridge, United Kingdom
Nobel Prize in Chemistry for the development of crystallographic electron microscopy and his structural elucidation of biologically important nucleic acid-protein complexes

Stanley B. Prusiner
University of California, San Francisco, USA
Nobel Prize in Physiology/Medicine for the discovery of Prions – a new biological principle of infection

Bengt Samuelsson
Karolinska Institute, Stockholm, Sweden
Nobel Prize in Physiology/Medicine for the discoveries concerning prostaglandins and related biologically active substances

Phillip A. Sharp
Massachusetts Institute of Technology, Cambridge, USA
Nobel Prize in Physiology/Medicine for the discoveries of split genes

Alexander Varshavsky
California Institute of Technology, Pasadena, USA
Lasker Award for the discovery and the recognition of the significance of the ubiquitin system of regulated protein degradation

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Director of the RIKEN Genomic Science Center

Shigeyuki Yokoyama
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Nobel Prize in Physiology/Medicine for the discoveries concerning the specificity of the cell mediated immune defence

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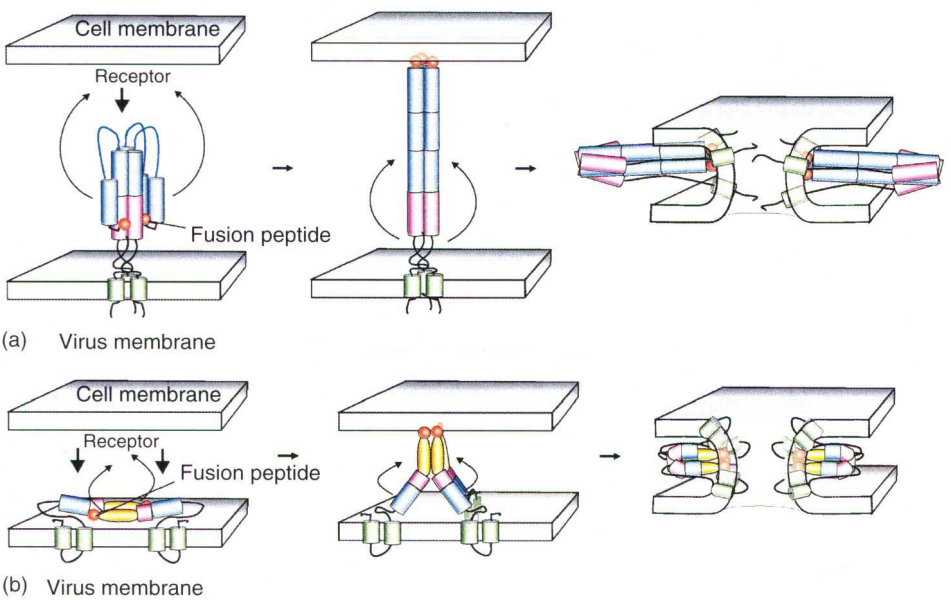

Fig. 3 (p. 77) Models for membrane fusion mediated by class I (a) and class II (b) viral fusion proteins involving a two-step mechanism. In the first step, the fusion peptides are exposed and are inserted in the target membrane. In the second step, the C-terminal part of the proteins undergoes another conformational change, resulting in bending/pulling that results in fusion. Red balls depict the fusion peptides, green cylinders the transmembrane domains. Note that the cellular receptors and the viral receptor–binding proteins are not shown. The colors indicate different domain of the proteins. Note that in class I proteins the colored boxes indicate α-helices, whereas in class II proteins they define folded domains that are not helical. Arrows indicate the presumed movements.

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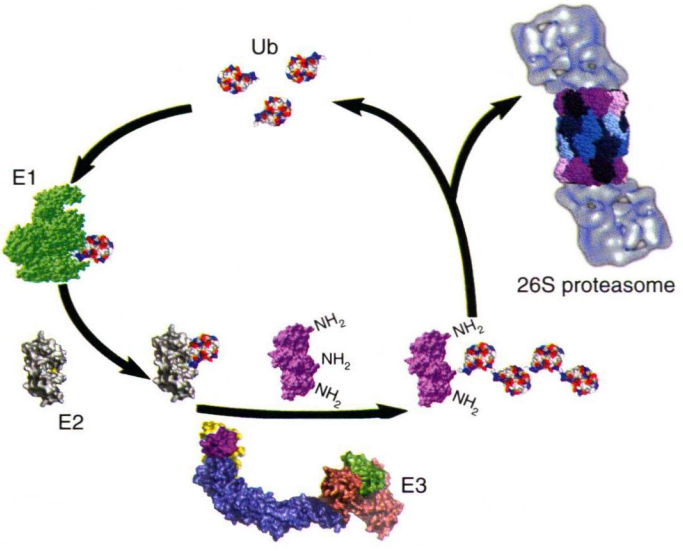

Fig. 3 (p. 8) Schematic representation of the ubiquitin-proteasome pathway. Ubiquitin molecules are activated by an E1 enzyme (shown in green at one-third scale) in an ATP-dependent reaction, transferred to a cysteine residue (yellow) on an E2 or Ub carrier protein and subsequently attached to amino groups (NH_2) on a substrate protein (lysozyme shown in purple) by an E3 or ubiquitin ligase, (the multicolored SCF complex). Note that chains of Ub are generated on the substrate, and these are recognized by the 26S proteasome depicted in the upper right at one-twentieth scale.

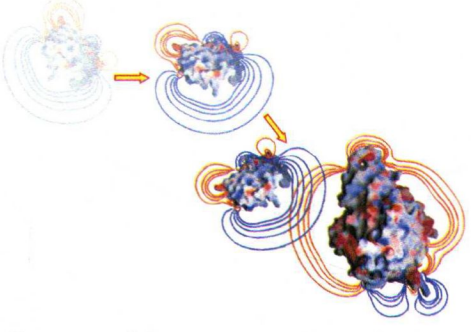

Fig. 4 (p. 150) Schematic illustration of the association of fasciculin (on the left) with acetylcholinesterase. Blue and red contour lines indicate regions of positive and negative electrostatic potential, respectively. Figure courtesy of D. Sept and A. Elcock. See Elcock, A.H., Gabdoulline, R.R., Wade, R.C., McCammon, J.A. (1999) Computer simulation of protein-protein association kinetics: acetylcholinesterase-fasciculin, *J. Mol. Biol.* **291**, 149–162 for more details.