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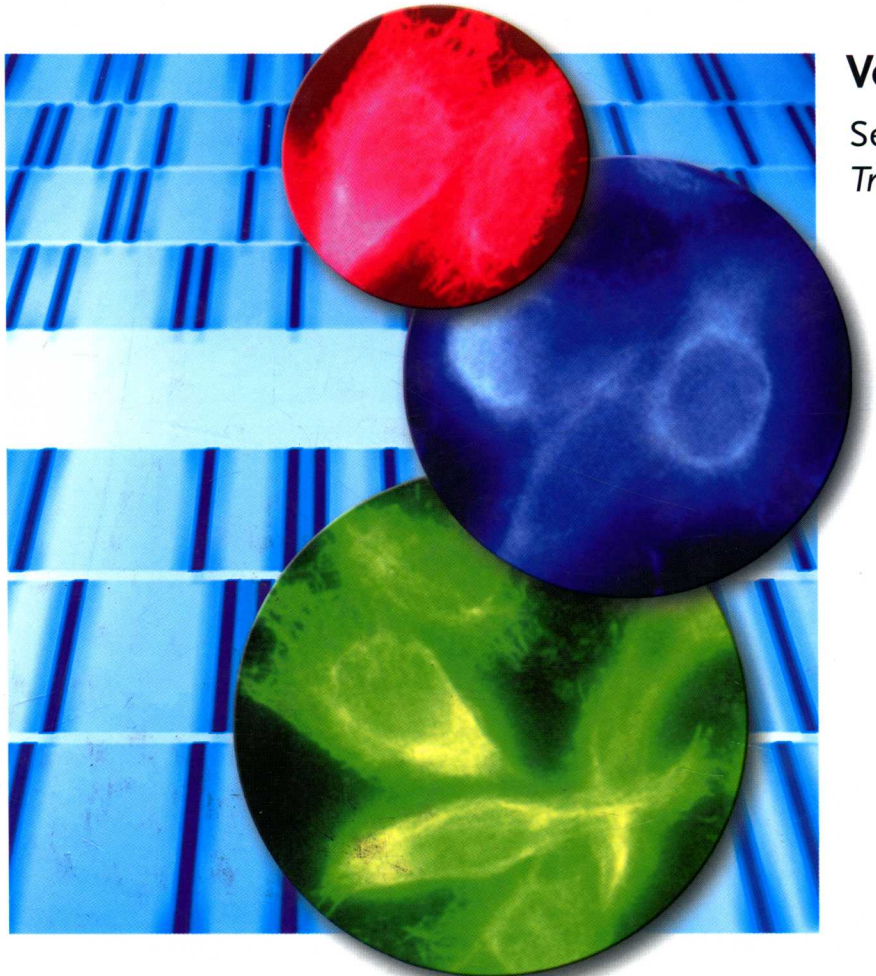
Encyclopedia of Molecular Cell Biology and Molecular Medicine

Edited by Robert A. Meyers



Volume 15

Second Edition

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Volume 15
Triplet Repeat Diseases to Zebrafish (*Danio rerio*) Genome and Genetics

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Encyclopedia of Molecular Cell Biology and Molecular Medicine

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.

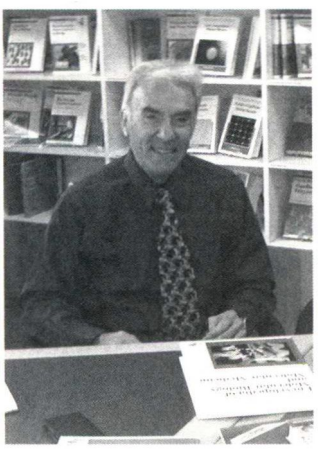
Approximately 250 article titles from the first edition are retained, but rewritten, half by new authors and half by returning authors. Approximately 80 articles on cell biology and 70 molecular biology articles have been added covering areas that have become prominent since preparation of the first 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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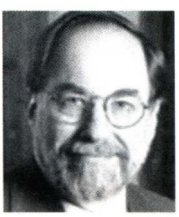
Nobel Prize in Physiology/Medicine for the discovery of restriction enzymes and their application to problems of molecular genetics



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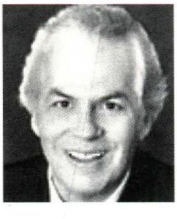
Nobel Prize in Chemistry for the development of crystallographic electron microscopy and his structural elucidation of biologically important nucleic acid-protein complexes



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Nobel Prize in Physiology/Medicine for the discoveries concerning prostaglandins and related biologically active substances

Phillip A. Sharp
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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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Color Plates

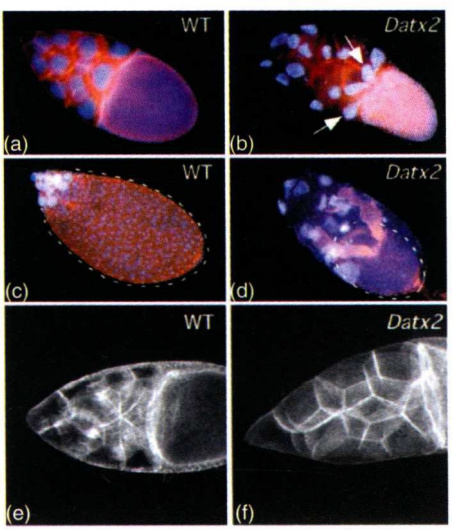

Fig. 4 (p. 20) Studies of the ataxin-2 ortholog in *Drosophila melanogaster* reveals a role for Drosophila ataxin-2 (*Datx2*) in actin filament formation during oogenesis. Egg chambers from normal (Wild-Type (WT)) and mutant flies with reduced expression (*Datx2*) of Drosophila ataxin-2 were stained with DAPI (blue) and phalloidin (red) to indicate nuclei and filamentous actin respectively (a–d). The egg chambers of WT flies prior to cytoplasmic transport (a) display well demarcated, separated but interconnected cells (blue) as expected, while the egg chambers of *Datx2* flies (b) contain irregularly arranged cells. After cytoplasmic transport, the egg chambers of WT flies (c) show one greatly enlarged oocyte (dashed line) with only a small section of compressed cells, while the egg chambers of *Datx2* flies (d) have failed to yield an enlarged oocyte, instead retaining dispersed and large adjacent cells. Confocal images of egg chambers prior to cytoplasmic transport stage reveal a prominent actin filament network in WT flies (e), but a remarkably transparent actin filament network in *Datx2* flies (f). The decreased density of the actin filament network underlies the cytoplasmic "dumping" defect in the *Datx2* flies. (From Satterfield, T.F., Jackson, S.M., Pallanck, L.J. (2002) A Drosophila homolog of the polyglutamine disease gene *SCA2* is a dosage-sensitive regulator of actin filament formation, *Genetics* **162**, 1687–1702, used with permission of *Genetics*.)

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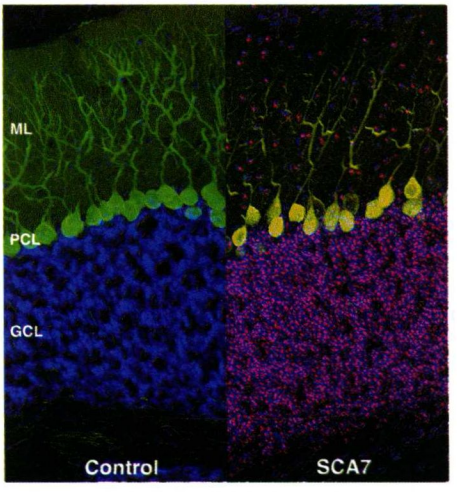

Fig. 5 (p. 24) Noncell autonomous Purkinje cell degeneration in a mouse model of spinocerebellar ataxia type 7 (SCA7). Confocal microscopy analysis of cerebellar sections from a SCA7 transgenic mouse (SCA7) created with an ataxin-7 CAG-92 containing murine prion protein expression vector and from an age- and sex-matched nontransgenic littermate (Control). Staining with an anti-ataxin-7 antibody (magenta), a calbindin antibody (green), and DAPI (blue) reveals a healthy, normal-appearing cerebellum characterized by properly oriented Purkinje cells with extensive dendritic arborization in the "Control" mice. However, SCA7 transgenic mice display pronounced Purkinje cell degeneration as evidenced by decreased dendritic arborization and displacement of Purkinje cell bodies. Interestingly, although numerous neurons in the granule cell layer (GCL) and the molecular layer (ML) display aggregates of ataxin-7, there is no accumulation of mutant ataxin-7 in the degenerating Purkinje cells due to lack of appreciable expression there. As the Purkinje cells degenerate without expressing the mutant protein, the degeneration is described as noncell autonomous. (Adapted from Garden, G.A., Libby, R.T., Fu, Y.H., Kinoshita, Y., Huang, J., Possin, D.E., Smith, A.C., Martinez, R.A., Fine, G.C., Grote, S.K., et al. (2002) Polyglutamine-expanded ataxin-7 promotes noncell-autonomous Purkinje cell degeneration and displays proteolytic cleavage in ataxic transgenic mice, *J. Neurosci.* **22**, 4897–4905, used with permission of the *Journal of Neuroscience*.)

Fig. 5 (p. 79) Two-photon optophysiology in the retina. Dye-filled "starburst" amacrine cell (a) in flat-mounted rabbit retina. Like many amacrine cells, this neuron bears no axon; it receives inputs and makes output synapses with its dendrites. Starburst cells are involved in the detection of image motion. Using (b) two-photon microscopy, light stimulus-evoked Ca^{2+} signals (green trace) were recorded in the dendritic tips of a starburst amacrine cell. Simultaneously, membrane voltage (black trace) was measured at the soma using a patch electrode (see schematic drawing). The light stimulus, a concentric sinusoidal wave, induced stronger responses when it was expanding (left) than when it was contracting (right).