

# CONTENTS

## ENZYMES AND APOLIPOPROTEINS IN THE DIAGNOSIS OF DISEASE

|                                                                                               |    |
|-----------------------------------------------------------------------------------------------|----|
| Lipoprotein Lipase: Gene Variants and Coronary Atherosclerosis. ....                          | 1  |
| R.K. Mattu and D.J. Galton                                                                    |    |
| Apolipoproteins in the Diagnosis of Disease: Cerebral and Peripheral<br>Atherosclerosis. .... | 13 |
| R. Fellin and G.B. Vigna                                                                      |    |

## STRUCTURAL AND FUNCTIONAL RELATIONSHIPS IN APOLIPOPROTEINS

|                                                                                                                                                                            |    |
|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------|----|
| Functional Properties of Discoidal Apoprotein/Phospholipid Complexes: Cholesterol Efflux from<br>Macrophages and Conversion by Lecithin-Cholesterol Acyl Transferase. .... | 21 |
| M. Rosseneu, A. Devreese, B. Vanloo and J. Baert                                                                                                                           |    |
| Lipid Transport by Apolipoprotein AI: Structure and Function. ....                                                                                                         | 31 |
| E.L. Gong, A.V. Nichols and A.R. Tall                                                                                                                                      |    |
| Characterization of Lipoprotein Particles Containing Apolipoprotein A-IV. ....                                                                                             | 43 |
| G.R. Castro, N. Duverger, P. Duchateau and J.C. Fruchart                                                                                                                   |    |
| Structural and Functional Properties of Apo A-I Variants in<br>Reconstituted HDL. ....                                                                                     | 53 |
| A. Jonas, A. von Eckardstein, W.W. Mantulin and G. Assmann                                                                                                                 |    |
| Structural Domains in Human Serum Low Density Lipoprotein<br>Apolipoprotein B100. ....                                                                                     | 67 |
| L.P. Aggerbeck                                                                                                                                                             |    |
| Crystal Structures of the Common Apolipoprotein E Variants:<br>Insights into Functional Mechanisms. ....                                                                   | 81 |
| M.R. Wardell, C.W. Wilson, D.A. Agard, R.W. Mahley and K.H. Weisgraber                                                                                                     |    |

|                                                                                                                                  |    |
|----------------------------------------------------------------------------------------------------------------------------------|----|
| Analysis of Apolipoprotein E Function in Adrenal Cells: Effects on Cholesterol Metabolism and Signal Transduction Pathways. .... | 97 |
| D.L. Williams, M.M. Prack, M. Nicosia, J.T. Gwynne and M.E. Reyland                                                              |    |

## ANIMAL MODELS IN THE APOLIPOPROTEIN MUTANT RESEARCH

|                                                                                                                                            |     |
|--------------------------------------------------------------------------------------------------------------------------------------------|-----|
| Spontaneous Hypercholesterolemia in Pigs. ....                                                                                             | 107 |
| A.D. Attie, R.J. Aiello, S.T. Cooper and P. Uelmen                                                                                         |     |
| The Apolipoprotein E Cys-142 Mutant: Role in Dominant Inheritance of Type III Hyperlipoproteinemia and Expression in Transgenic Mice. .... | 113 |
| S.C. Rall Jr, Y. Horie and S. Fazio                                                                                                        |     |

## RECOMBINANT APOLIPOPROTEINS AS DRUGS

|                                                                                                                    |     |
|--------------------------------------------------------------------------------------------------------------------|-----|
| Apolipoproteins as "Drugs": Current Status. ....                                                                   | 121 |
| C.R. Sirtori                                                                                                       |     |
| Non-Mammalian Host System for the Expression of Mammalian Apolipoproteins. ....                                    | 135 |
| S.L. Sturley, D.G. Gretch, P.D. Friesen, M.R. Culbertson, N. Beckage and A.D. Attie                                |     |
| Recombinant Human Proapolipoprotein A-I: Experimental Strategies for the Production of an Authentic Molecule. .... | 145 |
| N. Mogilevsky, F. Varsalona, J.-P. Guillaume, C. Roobol and A. Bollen                                              |     |
| The Role of HDL Apolipoproteins in Mediating HDL Receptor Binding and Intracellular Cholesterol Efflux. ....       | 165 |
| A.J. Mendez and J.F. Oram                                                                                          |     |

## MOLECULAR ABNORMALITIES OF APOLIPOPROTEINS

|                                                                                        |     |
|----------------------------------------------------------------------------------------|-----|
| The Metabolism of Truncated Forms of Apolipoprotein B in Hypobetalipoproteinemia. .... | 175 |
| G. Schonfeld                                                                           |     |

|                                                                                                                                                                    |     |
|--------------------------------------------------------------------------------------------------------------------------------------------------------------------|-----|
| Apolipoprotein B: Genetic Variants Provide Insight Into<br>Structure and Function.....                                                                             | 183 |
| P. Talmud, A. Dunning and S. Humphries                                                                                                                             |     |
| The Mutations of Apolipoprotein B Gene Causing Hypobetalipoproteinemia:<br>Molecular and Functional Abnormalities in Apo B-87 <sub>Padova</sub> .....              | 195 |
| C. Gabelli, C. Bilato, S. Martini, G.E. Tennyson, H.B. Brewer Jr,<br>G. Crepaldi and G. Baggio                                                                     |     |
| Causes Underlying the Reduced Response to Simvastatin Treatment in<br>Hypercholesterolemic Patients. ....                                                          | 207 |
| A. Corsini, M. Mazzotti, T. Zanelli, P. Uboldi, D. Sommariva, F. Pazzucconi,<br>C.R. Sirtori, A. Gaddi, G.C. Descovich, A. Granata, R. Fumagalli and A.L. Catapano |     |
| Molecular Genetics of the Familial Chylomicronemia Syndrome.....                                                                                                   | 231 |
| S. Santamarina-Fojo                                                                                                                                                |     |
| Rare Mutations in the Apo E Gene Associated with Dominant Mode of<br>Inheritance of Familial Dysbetalipoproteinemia (FD). ....                                     | 241 |
| L.M. Havekes, M. Mulder, P.de Knijff, A. van den Maagdenberg<br>and R.R. Frants                                                                                    |     |
| Identification of Common Genetic Polymorphisms that Determine Plasma<br>Levels of Apo AI and HDL-C.....                                                            | 247 |
| S. Humphries, V. Gudnason, H. Paul-Hayase, N. Saha and M.Y. Rosseneu                                                                                               |     |
| Structure-Function Relationships of Apolipoprotein A-I Variants. ....                                                                                              | 257 |
| A. von Eckardstein, A. Jonas, G. Castro, G.-C. Fruchart, H. Funke<br>and G. Assmann                                                                                |     |
| Sex-Related Differences in the Phenotypic Expression of the Apolipoprotein<br>A-I <sub>Milano</sub> mutant. ....                                                   | 267 |
| G. Franceschini, L. Calabresi, G. Gianfranceschi and C.R. Sirtori                                                                                                  |     |
| Human Apolipoprotein A-IV.....                                                                                                                                     | 279 |
| H.B. Brewer Jr and P. Lohse                                                                                                                                        |     |