

Contents

Contributors	xi
Preface	
Ronald L. Nagel	xvii
Dedications	
Amoz Chernoff, Ronald L. Nagel	xix
I. MOLECULAR ASPECTS	
Staphylococcus Aureus V8-Protease Catalyzed Segment Exchange Reaction of α-chain of Hemoglobin S: A Semisynthetic Approach for the Preparation of Variants of α-Chain	
A. Seetharama Acharya, Youngnan J. Cho, and K. Subramonia Iyer	3
Surface Hydrophobicity and Solubility of Hb S	
Kazuhiko Adachi	21
The Three-Dimensional Structure of Sickle Hemoglobin Macrofibers	
David A. Bluemke, Bridget Carragher, Michael J. Potel, and Robert Josephs	31
Polymer Domains, Gelation Models, and Sickle Cell Crises	
Frank A. Ferrone, Soumen Basak, Anthony J. Martino, and Huan Xiang Zhou	47
Proton NMR Studies of the Molecular Basis for the Antisickling Activity of Non-Covalent Antisickling Compounds	
Chien Ho and Irina M. Russu	59
Continuity of Transition in Hemoglobin S Crystallization	
William A. McDade, Robert J. Vassar, and Robert Josephs	67
Molecular Motion of Hb S in Gel: Study of Dipolar Orientation	
Shiro Takashima	91
Effects of Methyl Acetyl Phosphate on Hemoglobin S: A Novel Acetylating Agent Directed Towards the DPG Binding Site	
Hiroshi Ueno, Lennette J. Benjamin, and James M. Manning	105
II. RHEOLOGICAL ASPECTS	
Rheological Studies of Erythrocyte-Endothelial Cell Interactions in Sickle Cell Disease	
Gilda A. Barabino, Larry V. McIntire, Suzanne G. Eskin, David A. Sears, and Mark Udden	113

Exponential Progress Curves and Shear in the Gelation of Hemoglobin S	
Robin W. Briehl and Garrott W. Christoph	129
Rheology of Sickle Cells and Its Role in Microcirculatory Dynamics	
S. Chien, A.A. Kaperonis, R.G. King, H.H. Lipowsky, E.A. Schmalzer, L.A. Sung, K.L.P. Sung, and S. Usami	151
Sickle Erythrocytes Adhere to Endothelial Cell Monolayers (ECM's) Exposed to Flowing Blood	
Eric F. Grabowski	167
Characterization of Cellular Determinants and Plasma Factors Responsible for Increased Adherence of Sickle Cells to Vascular Endothelium	
N. Mohandas and E. Evans	181
 III. CELLULAR ASPECTS	
Activation of Calcium-Dependent Potassium Channels in Deoxygenated Sickled Red Cells	
Robert M. Bookchin, Olga E. Ortiz, and Virgilio L. Lew	193
Activation of a K:Cl Cotransporter by Cell Swelling in HbAA and HbSS Red Cells	
Mitzy Canessa, Mary E. Fabry, Anda Spalvins, Nadia Blumenfeld, and Ronald L. Nagel	201
Formation of Light Irreversibly Sickled Cells	
Kazumi Horiuchi and Toshio Asakura	217
Studies on the Mechanism of Passive Cation Fluxes Activated by Deoxygenation of Sickle Cells	
Clinton H. Joiner	229
The Problem of Describing Gelation In Vivo	
Andrea Mozzarelli, James Hofrichter, and William A. Eaton	237
Survival Rates and Properties of Sickle Cell Anemia Red Cells Treated With Nitrogen Mustard	
Eugene F. Roth, Jr., Barry Wenz, Hyo Bok Lee, Mary E. Fabry, Henry Chang, Dhananjaya K. Kaul, Silvio Baez, and Ronald L. Nagel	245
 IV. PAINFUL CRISES AND CLINICAL ASPECTS	
Pathogenesis of Painful Sickle Cell Crises	
Samuel Charache and Isidore Zohoun	265
The Effect of Increased Fetal Hemoglobin Production on the Frequency of Vaso-Occlusive Crisis in Sickle Cell Disease	
George J. Dover and Samuel Charache	277
The Role of Fluctuating Populations of Blood Cells in the Acute Painful Episodes of Sickle Cell Disease	
Stephen H. Embury	287
An Animal Model for Sickle Cell Vaso-Occlusion: A Study Using NMR and Technetium Imaging	
M.E. Fabry, D.K. Kaul, L. Davis, J.C. Gore, M. Brown, and R.L. Nagel . . .	297

Frequency of Pain Crisis During Pregnancy	
Mabel Koshy, Laurence Burd, Louise Dorn, and Glenda Huff	305
Sickle Cell Vaso-Occlusion in an Animal Model: Intravital Microscopy and Radionuclide Imaging of Selective Sequestration of Dense Cells	
Joseph Kurantsin-Mills, Helena M. Jacobs, and Lawrence S. Lessin	313
Objective Indices of Sickle Cell Painful Crisis: Decrease in RDW and Percent Dense Cells and Increase in ESR and Fibrinogen	
Christine Lawrence and Mary E. Fabry	329
Magnetic Resonance Imaging, Percentage of Dense Cells, and Serum Prostanoids as Tools for Objective Assessment of Pain Crisis: A Preliminary Report	
Vipul N. Mankad, J. Powell Williams, Michael Harpen, Gesina Longenecker, Byron Brogdon, and Blaine Moore	337
Post-Transfusion Crises in Sickle Cell Anemia	
Paul F. Milner and Jerry E. Squires	351
Sickle Cell Painful Crises: A Multifactorial Event	
R.L. Nagel, M.E. Fabry, H.H. Billett, and D.K. Kaul	361
Intracellular Polymerization of Sickle Hemoglobin: Disease Severity and Therapeutic Goals	
Constance Tom Noguchi, Griffin P. Rodgers, and Alan N. Schechter	381
Is Sickle Cell Crisis a Valid Measure of Clinical Severity in Sickle Cell Anemia?	
Darleen R. Powars and Linda S. Chan	393
The Development of Non-Invasive Methods to Evaluate Sickle Cell Patients in the Steady State and in Crisis	
Griffin P. Rodgers, Constance Tom Noguchi, and Alan N. Schechter	403
Determinants of Vaso-Occlusive Severity in Sickle Cell Disease	
Martin H. Steinberg	413
Painful Crisis and Dense Echinocytes: Effects of Hydration and Vasodilators	
James A. Warth and Donald L. Rucknagel	429
Index	451