

CONTENTS

.....	ix
.....	xxi

Clinical Evaluation of the Patient

1. Initial Approach to the Patient: History and Physical Examination.....	3
<i>Marshall A. Lichtman and Linda J. Burns</i>	
2. Examination of Blood Cells.....	11
<i>David H. Byan</i>	
3. Examination of The Marrow.....	27
<i>David H. Byan</i>	
4. Consultative Hematology.....	41
<i>Sanjay S. Brar and Stanley L. Schrier</i>	

The Organization of the Lymphohematopoietic Tissues

5. Structure of the Marrow and the Hematopoietic Microenvironment.....	53
<i>David P. Dave and Mark J. Koury</i>	
6. The Organization and Structure of Lymphoid Tissues.....	85
<i>Steven D. Douglas and Michael A. Caligiuri</i>	

Special Hematology

7. Hematology of the Fetus and Newborn.....	99
<i>James Paik and George B. Segel</i>	
8. Hematology during Pregnancy.....	119
<i>Walter P. Hines</i>	
9. Hematology in Older Persons.....	129
<i>William B. Erhler, Andrew S. Artz, and Bindu Kanapuru</i>	

Molecular and Cellular Hematology

10. Genetic Principles and Molecular Biology.....	145
<i>David B. Asch</i>	
11. Genomics.....	155
<i>James D. Wertzman and Elaine R. Mardis</i>	
12. Cytogenetics.....	165
<i>Walter B. Cairns</i>	
13. Cytogenetics and Genetic Abnormalities.....	173
<i>James D. Godley, Madina Sukhanova, Gordana Raca, and Michelle M. Le Beau</i>	
14. Mechanism of Hematologic Neoplastic Cells.....	191
<i>David E. Willen, Annie L. Hsieh, and Chi V. Dang</i>	
15. Signaling Mechanisms: Relevance to the Hematopoietic System.....	203
<i>David C. Reed</i>	

16. Cell-Cycle Regulation and Hematologic Disorders.....	213
<i>Yun Dai, Prithviraj Bose, and Steven Grant</i>	
17. Signal Transduction Pathways.....	247
<i>Kenneth Kaushansky</i>	
18. Hematopoietic Stem Cells, Progenitors, and Cytokines.....	257
<i>Kenneth Kaushansky</i>	
19. The Inflammatory Response.....	279
<i>Jeffrey S. Warren and Peter A. Ward</i>	
20. Innate Immunity.....	293
<i>Bruce Beutler</i>	
21. Dendritic Cells and Adaptive Immunity.....	307
<i>Madhav Dhodapkar, Crystal L. Mackall, and Ralph M. Steinman</i>	

PART V

Therapeutic Principles

22. Pharmacology and Toxicity of Antineoplastic Drugs.....	315
<i>Benjamin Izar, Dustin Dzube, James M. Cleary, Constantine S. Mitsiades, Paul G. Richardson, Jeffrey A. Barnes, and Bruce A. Chabner</i>	
23. Hematopoietic Cell Transplantation.....	353
<i>Andrew R. Rezvani, Robert Lowsky, and Robert S. Negrin</i>	
24. Treatment of Infections in The Immunocompromised Host.....	383
<i>Lisa Beutler and Jennifer Babik</i>	
25. Antithrombotic Therapy.....	393
<i>Gregory C. Connolly and Charles W. Francis</i>	
26. Immune Cell Therapy.....	409
<i>Carolina Berger and Stanley R. Riddell</i>	
27. Vaccine Therapy.....	421
<i>Katayoun Rezvani and Jeffrey J. Mollndrem</i>	
28. Therapeutic Apheresis: Indications, Efficacy, and Complications.....	427
<i>Robert Weinstein</i>	
29. Gene Therapy for Hematologic Diseases.....	437
<i>Hua Fung and Stanton Gerson</i>	
30. Regenerative Medicine: Multipotential Cell Therapy for Tissue Repair.....	447
<i>Jakub Tolar, Mark J Osborn, Randy Daughters, Anannya Banga, and John Wagner</i>	

PART VI

The Erythrocyte

31. Structure and Composition of the Erythrocyte.....	461
<i>Narla Mohandas</i>	
32. Erythropoiesis.....	479
<i>Josef T. Prchal and Perumal Thiagarajan</i>	
33. Erythrocyte Turnover.....	495
<i>Perumal Thiagarajan and Josef Prchal</i>	
34. Clinical Manifestations and Classification of Erythrocyte Disorders.....	503
<i>Josef T. Prchal</i>	

35. Aplastic Anemia: Acquired and Inherited	513
<i>George B. Segel and Marshall A. Lichtman</i>	
36. Pure Red Cell Aplasia	539
<i>Neal S. Young</i>	
37. Anemia of Chronic Disease	549
<i>Tomas Ganz</i>	
38. Erythropoietic Effects of Endocrine Disorders	559
<i>Xylina T. Gregg</i>	
39. The Congenital Dyserythropoietic Anemias	563
<i>Achille Iolascon</i>	
40. Paroxysmal Nocturnal Hemoglobinuria	571
<i>Charles J. Parker</i>	
41. Folate, Cobalamin, and Megaloblastic Anemias	583
<i>Ralph Green</i>	
42. Iron Metabolism	617
<i>Tomas Ganz</i>	
43. Iron Deficiency and Overload	627
<i>Tomas Ganz</i>	
44. Anemia Resulting from Other Nutritional Deficiencies	651
<i>Ralph Green</i>	
45. Anemia Associated with Marrow Infiltration	657
<i>Vishnu VB Reddy and Josef T. Prchal</i>	
46. Erythrocyte Membrane Disorders	661
<i>Theresa L Coetzer</i>	
47. Erythrocyte Enzyme Disorders	689
<i>Wouter W. van Solinge and Richard van Wijk</i>	
48. The Thalassemias: Disorders of Globin Synthesis	725
<i>David J. Weatherall</i>	
49. Disorders of Hemoglobin Structure: Sickle Cell Anemia and Related Abnormalities	759
<i>Kavita Natrajan and Abdullah Kutlar</i>	
50. Methemoglobinemia and Other Dyshemoglobinemias	789
<i>Archana M. Agarwal and Josef T. Prchal</i>	
51. Fragmentation Hemolytic Anemia	801
<i>Kelty R. Baker and Joel Moake</i>	
52. Erythrocyte Disorders as a Result of Chemical and Physical Agents	809
<i>Paul C. Herrmann</i>	
53. Hemolytic Anemia Resulting from Infections with Microorganisms	815
<i>Marshall A. Lichtman</i>	
54. Hemolytic Anemia Resulting from Immune Injury	823
<i>Charles H. Packman</i>	
55. Alloimmune Hemolytic Disease of the Fetus and Newborn	847
<i>Ross M. Fasano, Jeanne E. Hendrickson, and Naomi L. C. Luban</i>	
56. Hypersplenism and Hyposplenism	863
<i>Jaime Caro and Srikanth Nagalla</i>	
57. Primary and Secondary Erythrocytoses	871
<i>Josef T. Prchal</i>	
58. The Porphyrias	889
<i>John D. Phillips and Karl E. Anderson</i>	
59. Polyclonal and Hereditary Sideroblastic Anemias	915
<i>Prem Ponka and Josef T. Prchal</i>	

PART VII**Neutrophils, Eosinophils, Basophils, and Mast Cells**

60. Structure and Composition of Neutrophils, Eosinophils, and Basophils	925
<i>C. Wayne Smith</i>	
61. Production, Distribution, and Fate of Neutrophils	939
<i>C. Wayne Smith</i>	
62. Eosinophils and Related Disorders	947
<i>Andrew J. Wardlaw</i>	
63. Basophils, Mast Cells, and Related Disorders	965
<i>Stephen J. Galli, Dean D. Metcalfe, Daniel A. Arber, and Ann M. Dvorak</i>	
64. Classification and Clinical Manifestations of Neutrophil Disorders	983
<i>Marshall A. Lichtman</i>	
65. Neutropenia and Neutrophilia	991
<i>David C. Dale and Karl Welte</i>	
66. Disorders of Neutrophil Function	1005
<i>Niels Borregaard</i>	

PART VIII**Monocytes and Macrophages**

67. Structure, Receptors, and Functions of Monocytes and Macrophages	1045
<i>Steven D. Douglas and Anne G. Douglas</i>	
68. Production, Distribution, and Activation of Monocytes and Macrophages	1075
<i>Steven D. Douglas and Anne G. Douglas</i>	
69. Classification and Clinical Manifestations of Disorders of Monocytes and Macrophages	1089
<i>Marshall A. Lichtman</i>	
70. Monocytosis and Monocytopenia	1095
<i>Marshall A. Lichtman</i>	
71. Inflammatory and Malignant Histiocytosis	1101
<i>Kenneth L. McClain and Carl E. Allen</i>	
72. Gaucher Disease and Related Lysosomal Storage Diseases	1121
<i>Ari Zimran and Deborah Elstein</i>	

PART IX**Lymphocytes and Plasma Cells**

73. The Structure of Lymphocytes and Plasma Cells	1137
<i>Natarajan Muthusamy and Michael A. Caligiuri</i>	
74. Lymphopoiesis	1149
<i>Christopher S. Seet and Gay M. Crooks</i>	
75. Functions of B Lymphocytes and Plasma Cells in Immunoglobulin Production	1159
<i>Thomas J. Kipps</i>	
76. Functions of T Lymphocytes: T-Cell Receptors for Antigen	1175
<i>Fabienne McClanahan and John Gribben</i>	
77. Functions of Natural Killer Cells	1189
<i>Giorgio Trinchieri, Richard W. Childs, and Lewis L. Lanier</i>	
78. Classification and Clinical Manifestations of Lymphocyte and Plasma Cell Disorders	1195
<i>Yvonne A. Efebera and Michael A. Caligiuri</i>	

- 79.** Lymphocytosis and Lymphocytopenia 1199
Sumithira Vasu and Michael A. Caligiuri
- 80.** Immunodeficiency Diseases 1211
Hans D. Ochs and Luigi D. Notarangelo
- 81.** Hematologic Manifestations of Acquired Immunodeficiency Syndrome 1239
Virginia C. Broudy, Robert D. Harrington
- 82.** Mononucleosis Syndromes 1261
Robert F. Betts

PART X**Malignant Myeloid Diseases**

- 83.** Classification and Clinical Manifestations of the Clonal Myeloid Disorders 1275
Marshall A. Lichtman
- 84.** Polycythemia Vera 1291
Jaroslav F. Prchal and Josef T. Prchal
- 85.** Essential Thrombocythemia 1307
Philip A. Beer and Anthony R. Green
- 86.** Primary Myelofibrosis 1319
Marshall A. Lichtman and Josef T. Prchal
- 87.** Myelodysplastic Syndromes 1341
Rafael Bejar and David P. Steensma
- 88.** Acute Myelogenous Leukemia 1373
Jane L. Liesveld and Marshall A. Lichtman
- 89.** Chronic Myelogenous Leukemia and Related Disorders ... 1437
Jane L. Liesveld and Marshall A. Lichtman

PART XI**Malignant Lymphoid Diseases**

- 90.** Classification of Malignant Lymphoid Disorders 1493
Robert A. Baiocchi
- 91.** Acute Lymphoblastic Leukemia 1505
Richard A. Larson
- 92.** Chronic Lymphocytic Leukemia 1527
Farrukh T. Awan and John C. Byrd
- 93.** Hairy Cell Leukemia 1553
Michael R. Grever and Gerard Lozanski
- 94.** Large Granular Lymphocytic Leukemia 1563
Pierluigi Porcu and Aharon G. Freud
- 95.** General Considerations for Lymphomas: Epidemiology, Etiology, Heterogeneity, and Primary Extranodal Disease .. 1569
Oliver W. Press and Marshall A. Lichtman
- 96.** Pathology of Lymphomas 1587
Randy D. Gascoyne and Brian F. Skinnider
- 97.** Hodgkin Lymphoma 1603
Oliver W. Press
- 98.** Diffuse Large B-Cell Lymphoma and Related Diseases 1625
Stephen D. Smith and Oliver W. Press
- 99.** Follicular Lymphoma 1641
Oliver W. Press
- 100.** Mantle Cell Lymphoma 1653
Martin Dreyling

- 101.** Marginal Zone B-Cell Lymphomas 1663
Pier Luigi Zinzani and Alessandro Broccoli
- 102.** Burkitt Lymphoma 1671
Andrew G. Evans and Jonathan W. Friedberg
- 103.** Cutaneous T-Cell Lymphoma (Mycosis Fungoides and Sézary Syndrome) 1679
Larisa J. Geskin
- 104.** Mature T-Cell and Natural Killer Cell Lymphomas 1693
Neha Mehta, Alison Moskowitz, and Steven Horwitz
- 105.** Plasma Cell Neoplasms: General Considerations 1707
Guido Tricot, Siegfried Janz, Kalyan Nadiminti, Erik Wendlandt, and Fenghuang Zhan
- 106.** Essential Monoclonal Gammopathy 1721
Marshall A. Lichtman
- 107.** Myeloma 1733
Elizabeth O'Donnell, Francesca Cottini, Noopur Raje, and Kenneth Anderson
- 108.** Immunoglobulin Light-Chain Amyloidosis 1773
Morie A. Gertz, Taimur Sher, Angela Dispenzieri, and Francis K. Buadi
- 109.** Macroglobulinemia 1785
Steven P. Treon, Jorge J. Castillo, Zachary R. Hunter, and Giampaolo Merlini
- 110.** Heavy-Chain Disease 1803
Dietlind L. Wahner-Roedler and Robert A. Kyle

PART XII**Hemostasis and Thrombosis**

- 111.** Megakaryopoiesis and Thrombopoiesis 1815
Kenneth Kaushansky
- 112.** Platelet Morphology, Biochemistry, and Function 1829
Susan S. Smyth, Sidney Whiteheart, Joseph E. Italiano Jr., Paul Bray, and Barry S. Collier
- 113.** Molecular Biology and Biochemistry of the Coagulation Factors and Pathways of Hemostasis 1915
Mettine H. A. Bos, Cornelis van 't Veer, and Pieter H. Reitsma
- 114.** Control of Coagulation Reactions 1949
Laurent O. Mosnier and John H. Griffin
- 115.** Vascular Function in Hemostasis 1967
Katherine A. Hajjar, Aaron J. Marcus, and William Muller
- 116.** Classification, Clinical Manifestations, and Evaluation of Disorders of Hemostasis 1985
Marcel Levi, Uri Seligsohn, and Kenneth Kaushansky
- 117.** Thrombocytopenia 1993
Reyhan Diz-Küçükaya and José A. López
- 118.** Heparin-Induced Thrombocytopenia 2025
Adam Cuker and Mortimer Poncz
- 119.** Reactive Thrombocytosis 2035
Kenneth Kaushansky
- 120.** Hereditary Qualitative Platelet Disorders 2039
A. Koneti Rao and Barry S. Collier
- 121.** Acquired Qualitative Platelet Disorders 2073
Charles S. Abrams, Sanford J. Shattil, and Joel S. Bennett
- 122.** The Vascular Purpuras 2097
Doru T. Alexandrescu and Marcel Levi

- 123.** Hemophilia A and Hemophilia B 2113
Miguel A. Escobar and Nigel S. Key
- 124.** Inherited Deficiencies of Coagulation Factors II, V, V+VIII, VII, X, XI, and XIII 2133
Flora Peyvandi and Marzia Menegatti
- 125.** Hereditary Fibrinogen Abnormalities 2151
Marguerite Neerman-Arbez and Philippe de Moerloose
- 126.** von Willebrand Disease 2163
Jill Johnsen and David Ginsburg
- 127.** Antibody-Mediated Coagulation Factor Deficiencies 2183
Sean R. Stowell, John S. (Pete) Lollar, and Shannon L. Meeks
- 128.** Hemostatic Alterations in Liver Disease and Liver Transplantation 2191
Frank W.G. Leebeek and Ton Lisman
- 129.** Disseminated Intravascular Coagulation 2199
Marcel Levi and Uri Seligsohn
- 130.** Hereditary Thrombophilia 2221
Saskia Middeldorp and Michiel Coppens
- 131.** The Antiphospholipid Syndrome 2233
Jacob H. Rand and Lucia Wolgast
- 132.** Thrombotic Microangiopathies 2253
J. Evan Sadler

- 133.** Venous Thrombosis 2267
Gary E. Raskob, Russell D. Hull, and Harry R. Buller
- 134.** Atherothrombosis: Disease Initiation, Progression, and Treatment 2281
Emile R. Mohler III and Andrew I. Schafer
- 135.** Fibrinolysis and Thrombolysis 2303
Katherine A. Hajjar and Jia Ruan

PART XIII

Transfusion Medicine

- 136.** Erythrocyte Antigens and Antibodies 2329
Marion E. Reid and Christine Lomas-Francis
- 137.** Human Leukocyte and Platelet Antigens 2353
Myra Coppage, David Stroncek, Janice McFarland, and Neil Blumberg
- 138.** Blood Procurement and Red Cell Transfusion 2365
Jeffrey McCullough, Majed A. Refaai, and Claudia S. Cohn
- 139.** Preservation and Clinical Use of Platelets 2381
Terry Gernsheimer and Sherrill Slichter

- Index** 2393