

Contents

List of Contributors	xiii	3. Classification and Diagnosis of Anemia in Children.....	32
About the Editors	xvii	PHILIP LANZKOWSKY	
Preface to the Sixth Edition.....	xxi	Classification and Diagnosis	32
Preface to the Fifth Edition	xxiii	Further Reading and References.....	41
Preface to the Fourth Edition	xxv	4. Lymphadenopathy and Diseases of the Spleen.....	42
Preface to the Third Edition.....	xxvii	PHILIP LANZKOWSKY	
Preface to the Second Edition.....	xxix	Lymphadenopathy	42
Preface to the First Edition.....	xxxi	Diseases of the Spleen.....	47
Introduction: Historic Perspective	xxxiii	Splenomegaly.....	48
		Further Reading and References.....	50
1. Diagnostic Molecular and Genomic Methodologies for the Hematologist/Oncologist.....	1	5. Anemia During the Neonatal Period	51
VIJAY G. SANKARAN		PHILIP LANZKOWSKY	
Clinical Molecular and Genomic Methodologies	1	Hemorrhage	51
Interpreting and Evaluating the Results from Clinical Genetic Testing	5	Hemolytic Anemia.....	55
Further Reading and References.....	6	Failure of Red Cell Production	65
2. Hematologic Manifestations of Systemic Illness.....	7	Anemia of Prematurity.....	65
LAWRENCE C. WOLFE		Physiologic Anemia.....	66
Hematologic Manifestations of Diseases Related to Various Organs	7	Diagnostic Approach to Anemia in the Newborn	66
Chronic Illness.....	13	Further Reading and References.....	68
Lead Intoxication	23	6. Iron-Deficiency Anemia.....	69
Nutritional Disorders.....	23	PHILIP LANZKOWSKY	
Bone Marrow Infiltration	23	Prevalence.....	69
Further Reading and References.....	31	Etiologic Factors	69
		Clinical Features.....	73

Diagnosis.....	74	Membrane Defects.....	137
Stages of Iron Depletion.....	78	Paroxysmal Nocturnal Hemoglobinuria.....	147
Differential Diagnosis.....	79	Enzyme Defects.....	151
Treatment.....	81	Further Reading and References.....	157
Further Reading and References.....	83		
7. Megaloblastic Anemia.....	84	10. Extracorporeal Hemolytic	
PHILIP LANZKOWSKY		Anemia.....	159
Vitamin B ₁₂ (Cobalamin) Deficiency.....	84	LAWRENCE C. WOLFE	
Folic Acid Deficiency.....	93	Immune Hemolytic Anemia.....	159
Clinical Features of Cobalamin		NonImmune Hemolytic Anemia.....	164
and Folate Deficiency.....	97	Further Reading and References.....	165
Diagnosis of Cobalamin and Folate			
Deficiency.....	98	11. Hemoglobinopathies.....	166
Treatment.....	100	JANET L. KWIATKOWSKI	
Further Reading and References.....	101	Sickle Cell Disease.....	166
8. Bone Marrow Failure.....	102	Thalassemias.....	186
ADRIANNA VLACHOS AND JEFFREY M. LIPTON		Management of the Acutely Ill Thalassemic	
Aplastic Anemia.....	102	Patient.....	193
Acquired Aplastic Anemia.....	102	Further Reading and References.....	195
Inherited Bone Marrow Failure Syndromes.....	112		
Fanconi Anemia.....	113	12. Polycythemia.....	197
Dyskeratosis Congenita.....	119	TSEWANG TASHI AND JOSEF T. PRCHAL	
Congenital Aplastic Anemias of Unknown		Polycythemia (Erythrocytosis) in the	
Inheritance.....	120	Newborn.....	197
Diamond–Blackfan Anemia (Congenital		Polycythemia in Childhood.....	200
Pure Red Cell Aplasia).....	121	Polycythemia Vera.....	201
Transient Erythroblastopenia of Childhood.....	125	Primary Familial and Congenital	
Congenital Dyserythropoietic Anemia.....	126	Polycythemia.....	203
Sideroblastic Anemias (Mitochondrial		Congenital Polycythemia Due to Altered	
Diseases with Bone Marrow Failure		Hypoxia Sensing with Normal P ₅₀	203
Syndromes).....	127	Congenital Polycythemia Due to Altered	
Severe Congenital Neutropenia and		Hypoxia Sensing with Decreased P ₅₀	204
Kostmann Syndrome.....	130	Other Causes of Polycythemia.....	205
Reticular Dysgenesis.....	132	Diagnostic Approach.....	206
Further Reading and References.....	132	Further Reading and References.....	207
9. General Considerations of Hemolytic			
Diseases, Red Cell Membrane, and		13. Disorders of White Blood Cells.....	209
Enzyme Defects.....	134	MARY ANN BONILLA AND JILL S. MENELL	
LIONEL BLANC AND LAWRENCE C. WOLFE		Quantitative Disorders of Leukocytes.....	209
Clinical Features of Hemolytic Disease.....	134	Decreased Production or Intrinsic	
Laboratory Findings.....	134	Defects of Neutrophils.....	212

Increased Destruction or Extrinsic Defects of Neutrophils.....	218	Thrombotic Disorders in Newborns	320
Immune Neutropenia	218	Antithrombotic Agents.....	321
NonImmune Neutropenia	220	Antithrombotic Therapy in Special Conditions	330
Eosinophils.....	222	Further Reading and References	332
Lymphocytes	228	16. Lymphoproliferative Disorders	334
Disorders of Leukocyte Function	229	DAVID T. TEACHEY	
Disorders of Adhesion and Chemotaxis	230	Lymphoproliferative Disorders	334
Disorders of Neutrophil Granules	232	Further Reading and References	347
Disorders of Oxidative Metabolism	234	17. Myelodysplastic Syndromes and Myeloproliferative Disorders	348
Neutrophil Production and Destruction in Newborn Infants.....	237	INGA HOFMANN AND TAREK M. ELGHETANY	
Neutrophil Function in the Newborn	237	Myelodysplastic Syndromes.....	348
Further Reading and References	238	Juvenile Myelomonocytic Leukemia	355
14. Disorders of Platelets	239	Myeloproliferative Neoplasms.....	359
CATHERINE MCGUINN AND JAMES B. BUSSEL		CML, BCR-ABL1 Positive.....	360
Thrombocytopenia in the Newborn	239	Essential Thrombocythemia and Polycythemia Vera.....	364
Inherited Thrombocytopenias.....	249	Primary Myelofibrosis.....	364
Immune Thrombocytopenic Purpura (ITP)	254	Further Reading and References	365
Other Causes of Thrombocytopenia.....	261	18. Acute Lymphoblastic Leukemia.....	367
Thrombocytosis	265	WILLIAM L. CARROLL AND TEENA BHATLA	
Qualitative Platelet Disorders	267	Incidence of ALL	367
Acquired Platelet Disorders	273	Etiology	367
Nonthrombocytopenic Purpura	277	Clinical Features of ALL.....	368
Further Reading and References	277	Prognostic Factors.....	376
15. Disorders of Coagulation.....	279	Treatment.....	377
SUCHITRA S. ACHARYA AND SUSMITA N. SARANGI		Infant Leukemia	379
Hemostatic Disorders	279	Philadelphia-Positive ALL	380
Hemostasis in the Newborn.....	282	Down Syndrome and ALL	383
Detection of Hemostatic Defects.....	284	Relapse in Children with ALL	383
Acquired Coagulation Factor Disorders	288	Future Drugs in ALL Therapy	387
Inherited Coagulation Factor Disorders	290	Further Reading and References	388
von Willebrand Disease	301	19. Acute Myeloid Leukemia.....	390
Rare Coagulation Factor Disorders (FII, V, VII, X, XI, XIII, Fibrinogen Deficiencies)....	305	ARLENE REDNER AND RACHEL KESSEL	
Thrombotic Disorders.....	306	Incidence and Epidemiology	390
Venous Thrombosis	308	Etiology and Predisposing Conditions	390
Arterial Thrombosis	311	Clinical Features.....	391
Antiphospholipid Antibody Syndrome	313		
Hereditary Thrombotic Disorders	316		

Diagnosis.....	391	23. Central Nervous System	
Classification of AML.....	393	Malignancies.....	453
Treatment.....	397	DEREK R. HANSON AND MARK P. ATLAS	
Novel Therapeutic Approaches.....	401	Epidemiology.....	453
Acute Promyelocytic Leukemia.....	402	Pathology.....	453
AML Special Subgroups.....	404	Clinical Manifestations.....	455
Further Reading and References.....	405	Genetic Syndromes Associated with	
		Pediatric Brain Tumors.....	456
20. Histiocytosis Syndromes.....	407	Diagnostic Evaluation.....	458
ROBERT J. ARCECI		Treatment.....	459
Langerhans Cell Histiocytosis.....	407	Astrocytomas.....	461
Other Histiocytic Disorders.....	420	Medulloblastoma.....	464
Macrophage Mediated Disorders.....	420	Relapsed Medulloblastoma.....	466
Further Reading and References.....	428	Brain Stem Tumors.....	467
		Ependymomas.....	468
21. Hodgkin Lymphoma.....	429	Optic Glioma.....	469
DEBRA L. FRIEDMAN		Craniopharyngiomas.....	470
Etiology and Epidemiology.....	429	Intracranial Germ Cell Tumors.....	471
Risk Factors.....	429	Malignant Brain Tumors in Infants and	
Biology.....	430	Children Less Than 3 Years of Age.....	471
Pathology.....	430	Further Reading and References.....	472
Clinical Presentation.....	432	24. Neuroblastoma.....	473
Diagnostic Evaluation and Staging.....	434	JULIE R. PARK AND ROCHELLE BAGATELL	
Prognostic Factors.....	435	Epidemiology.....	473
Treatment.....	436	Predisposition.....	473
Long-Term Complications.....	438	Pathology and Biology.....	473
Follow-Up Evaluations.....	440	Clinical Features.....	474
Further Reading and References.....	440	Diagnosis and Staging.....	476
		Treatment.....	478
22. Non-Hodgkin Lymphoma.....	442	Prognosis, Risk Stratification, and Therapy.....	479
MARY S. HUANG AND HOWARD WEINSTEIN		Post-Therapy Monitoring.....	487
Introduction.....	442	Special Treatment Considerations.....	488
Incidence and Epidemiology.....	442	Neuroblastoma in the Adolescent and	
Pathologic Classification.....	445	Young Adult.....	489
Clinical Features.....	445	Further Reading and References.....	489
Diagnosis.....	446	25. Renal Tumors.....	491
Staging.....	446	ANNE B. WARWICK AND JEFFREY S. DOME	
Prognosis.....	447	Wilms' Tumor.....	491
Management.....	447	Nephroblastomatosis.....	502
Lymphoblastic Lymphoma.....	448	Congenital Mesoblastic Nephroma.....	503
B Lineage NHL.....	449	Clear Cell Sarcoma of the Kidney.....	503
Further Reading and References.....	452		

Rhabdoid Tumor of the Kidney	503	29. Germ Cell Tumors.....	555
Renal Cell Carcinoma.....	503	THOMAS A. OLSON	
Further Reading and References.....	504	Incidence	555
26. Rhabdomyosarcoma and Other		Pathology	555
Soft-Tissue Sarcomas	505	Clinical Features.....	557
CAROLYN FEIN LEVY AND LEONARD H. WEXLER		Diagnostic Evaluation	558
Incidence and Epidemiology	505	Tumor Markers	558
Pathologic and Genetic Classification	506	Staging	559
Clinical Features.....	506	Treatment.....	560
Diagnostic Evaluation	509	Further Reading and References.....	567
Staging	511	30. Hepatic Tumors	569
Prognosis	513	LOUIS B. RAPKIN AND THOMAS A. OLSON	
Treatment.....	515	Incidence	569
Follow-up After Completion of Therapy	520	Epidemiology	569
Recurrent Disease	521	Pathology	571
Future Perspectives	521	Clinical Features.....	571
Further Reading and References.....	522	Diagnostic Evaluation	572
27. Malignant Bone Tumors.....	524	Staging	573
NOAH FEDERMAN, ELIZABETH A. VAN DYNE AND		Treatment.....	573
NICHOLAS BERNTHAL		Follow-up	575
Osteosarcoma.....	524	Prognosis	575
Ewing Sarcoma Family of Tumors.....	533	Further Reading and References.....	576
Other Bone Tumors.....	539	31. Hematopoietic Stem Cell	
Further Reading and References.....	542	Transplantation.....	577
28. Retinoblastoma	544	INDIRA SAHDEV AND HISHAM ABDEL-AZIM	
ANN LEAHEY		Allogeneic Stem Cell Transplantation.....	577
Incidence	544	HSC Sources, Collection, and Manipulation ...	583
Classification	544	Engraftment	589
13q Deletion Syndrome	546	Complications of HSCT	589
Risk for Second Malignant Neoplasms.....	546	Late Sequelae of Stem Cell Transplantation....	601
Pathology	546	Further Reading and References.....	603
Clinical Features.....	546	32. Management of Oncologic	
Diagnostic Procedures	547	Emergencies	605
Treatment.....	548	JASON L. FREEDMAN AND SUSAN R. RHEINGOLD	
Posttreatment Management	552	Metabolic Emergencies.....	605
Future Perspectives	553	Cardiothoracic Emergencies.....	610
Further Reading and References.....	553	Abdominal Emergencies.....	611

Renal Emergencies	613	Obesity	672
Neurologic Emergencies	614	Second Malignant Neoplasms.....	673
Endocrine Emergencies	616	Psychosocial Issues.....	674
Treatment-Associated Emergencies	617	Further Reading and References.....	675
Further Reading and References.....	618		
33. Supportive Care of Patients with Cancer.....	620	35. Psychosocial Aspects of Cancer for Children and Their Families	676
ANURAG K. AGRAWAL AND JAMES FEUSNER		MEG TIPPY	
Management of Infectious Complications.....	620	Time of Diagnosis	676
Infection Prophylaxis	627	Treatment Initiation.....	680
Recognition and Management of Nausea and Vomiting	631	Illness Stabilization.....	681
Mucositis	633	During Relapses and Recurrences.....	683
Pain Management.....	634	Treatment Outcomes: The Unsuccessful Course	683
Nutritional Status of the Oncology Patient.....	640	Treatment Outcomes: The Successful Course	685
Utilization of Hematopoietic Growth Factors...	642	Conclusion.....	687
Management of Acute Radiation Side Effects ..	644	Further Reading and References.....	687
Management of CVCs.....	647		
Posttreatment Immunizations	652	 	
Palliative Care	652	36. Blood Banking Principles and Practices	688
Further Reading and References.....	654	EDWARD C.C. WONG AND NAOMI L.C. LUBAN	
34. Evaluation, Investigations, and Management of Late Effects of Childhood Cancer	656	Introduction.....	688
JULIE I. KRYSZAL AND JONATHAN D. FISH		Donor Selection and Collection.....	688
Musculoskeletal System.....	656	Component Preparation.....	688
Cardiovascular System	659	Blood Component Modifications	692
Respiratory System	661	Special Pediatric Population Needs.....	695
Central Nervous System	663	Administration	700
Endocrine System	664	Summary	708
Genitourinary System.....	669	Further Reading and References.....	708
Ocular System.....	670		
Auditory System	671	Appendix 1:	
Gastrointestinal System	671	Hematological Reference Values	709
Immunologic System	672	Appendix 2:	
		Biological Tumor Markers	729
		Index	733