SECOND EDITION

Pediatric Psycho-Oncology

A Quick Reference on the Psychosocial Dimensions of Cancer Symptom Management

LORI S. WIENER, MARYLAND PAO, ANNE E. KAZAK, MARY JO KUPST, AND ANDREA FARKAS PATENAUDE

OXFORD

Pediatric Psycho-Oncology

APOS CLINICAL REFERENCE HANDBOOKS

Psycho-Oncology: A Quick Reference on the Psychosocial Dimensions of Cancer Symptom Management, 2nd edition, Jimmie C. Holland, Mitch Golant, Donna B. Greenberg, Mary K. Hughes, Jon A. Levenson, Matthew J. Loscalzo, William F. Pirl

Pediatric Psycho-Oncology: A Quick Reference on the Psychosocial Dimensions of Cancer Symptom Management, 2nd edition, Lori Wiener, Maryland Pao, Anne E. Kazak, Mary Jo Kupst, Andrea Farkas Patenaude

Geriatric Psycho-Oncology: A Quick Reference on the Psychosocial Dimensions of Cancer Symptom Management, Jimmie C. Holland, Talia Weiss Wiesel, Christian J. Nelson, Andrew J. Roth, Yesne Alici

Pediatric Psycho-Oncology

A Quick Reference on the Psychosocial Dimensions of Cancer Symptom Management

SECOND EDITION

Edited by

Lori Wiener, PhD, DCSW
Maryland Pao, MD
Anne E. Kazak, PhD, ABPP
Mary Jo Kupst, PhD
Andrea Farkas Patenaude, PhD
Robert J. Arceci, MD, PhD

OXFORD
UNIVERSITY PRESS

OXFORD
UNIVERSITY PRESS

Oxford University Press is a department of the University of Oxford. It furthers the University's objective of excellence in research, scholarship, and education by publishing worldwide.

Oxford New York
Auckland Cape Town Dar es Salaam Hong Kong Karachi
Kuala Lumpur Madrid Melbourne Mexico City Nairobi
New Delhi Shanghai Taipei Toronto

With offices in
Argentina Austria Brazil Chile Czech Republic France Greece
Guatemala Hungary Italy Japan Poland Portugal Singapore
South Korea Switzerland Thailand Turkey Ukraine Vietnam

Oxford is a registered trademark of Oxford University Press in the UK and certain other countries.

Published in the United States of America by
Oxford University Press
198 Madison Avenue, New York, NY 10016

Library of Congress Cataloging-in-Publication Data
Quick reference for pediatric oncology clinicians
Pediatric psycho-oncology : a quick reference on the psychosocial dimensions of cancer symptom management/edited by Lori Wiener, Maryland Pao, Anne E. Kazak, Mary Jo Kupst, Andrea Farkas Patenaude; with the American Psycho-Social Oncology Society.—2nd edition.
p. ; cm.
Preceded by: Quick reference for pediatric oncology clinicians/senior editors, Lori Wiener, Maryland Pao. c2009. Includes bibliographical references and index.
ISBN 978–0–19–933511–4 (alk. paper)
I. Wiener, Lori, editor. II. Pao, Maryland, editor. III. Kazak, Anne E., editor. IV. Kupst, Mary Jo, editor. V. Patenaude, Andrea Farkas, editor. VI. American Psychosocial Oncology Society. VII. Title.
[DNLM: 1. Child. 2. Neoplasms—psychology. 3. Age Factors. 4. Family Relations. 5. Neoplasms—therapy. 6. Social Support. QZ 275]
RC281.C4
618.92′994—dc23
2014022636

9 8 7 6 5 4 3 2 1
Printed in the United States of America
on acid-free paper

This book is dedicated to all children and families affected by pediatric cancer. Those we have worked with have taught us what we are now sharing with others in this volume.
LW
MP
AEK
MJK
AFP
RJA

Contents

Foreword

I vividly recall, as a second-year medical school student, accompanying my clinical advisor to visit a 2-year-old child who had been hospitalized with acute leukemia. My advisor was a relatively newly appointed faculty member in pediatric hematology-oncology—a field very much in its infancy at that time. The suffering of the child and family was apparent even to me, a novice medical student, and I wondered how anyone could pursue a career focusing on children with such a catastrophic disease. Although I was touched by the sensitive way in which my advisor interacted with the child and his family, I was still frightened by the likely life-threatening illness that this child faced and how he and his family would cope with its consequences.

Only a few years later, as a newly minted intern in pediatrics at the same children's hospital where my clinical advisor had trained me, I found myself drawn to the care of the very children toward whom I had such a worried reaction as a young medical student. In the few short years that had intervened, incredible progress had been made in the care of children with acute leukemia. The role of combination chemotherapy and the prospect for preventing central nervous system relapse with prophylactic radiation therapy was being defined and providing hope. I found myself drawn to the excitement of working in a field of medicine that was changing rapidly and where research could impact patient outcomes in meaningful ways. Having been educated in a medical school that had defined the biopsychosocial model of care, I also appreciated the importance of combining psychosocial care with medical treatment. In many ways, this challenging area of medicine brought together all my interests and passions—basic and clinical research, challenging multisystem medical disease management, along with interdisciplinary psychosocial care and intervention. I don't think I ever questioned whether a child diagnosed with cancer should receive psychosocial support; I believed that this was necessary for every child and family to have comprehensive care.

Over the decades that have followed my early entry into the field of pediatric oncology, I have witnessed, over and over again, the impact of serious disease on the child, adolescent, and young adult. In tandem, the impact of a serious childhood disorder on the parents, siblings, grandparents, relatives, and the community of friends is also abundantly apparent. It is implausible to think that any child or family facing the diagnosis of cancer will be unaffected emotionally and psychologically. Regardless of one's background, resources, social strata, professional status or knowledge, every child and every family requires help and support. No amount of inner strength or resilience can cope with the impact of cancer and its treatment without psychosocial support. From my perspective, psychosocial support is as important as medical care and treatment—both are needed to successfully

manage catastrophic disease. I was fortunate to witness this over and over again during the many years that I worked at the National Cancer Institute where we formed a team with social workers, psychologists, recreation therapists, and others to bring our respective skills and knowledge into a compassionate unison on behalf of the children and families we treated. I have long viewed the management of fears and feelings of children, adolescents—no matter how resistant—and young adults, about their illness and its consequences, and the ability to support the child within the family constellation and community not as ideal care (even though it is) but as the standard of care for the child with cancer.

Coping with cancer requires the ability to face the prospect of death along with numerous physical and emotional alterations. Learning how to cope with these challenges is a prerequisite to survival and should be part of every program of care for children with cancer (and I believe adults with cancer as well).

Since I began, much research has been brought to bear on many important aspects of caring for children with cancer including developmental and educational needs, impact on family, and the adverse psychological and social sequelae of cancer and its therapy. Many psychosocial specialists and researchers have dedicated their lives and careers to caring for children and families with cancer and catastrophic diseases. Their work has contributed as much to the quality survival of children as has the medical care that has been administered. Every pediatric oncologist must be conversant in the psychosocial dimensions of cancer management delineated in this book. I am honored to provide a personal introduction and affirmation of this important book.

Philip A. Pizzo, MD
David and Susan Heckerman Professor of Pediatrics,
Microbiology, and Immunology
Stanford University School of Medicine
Stanford, California
February, 2014

Preface

We are pleased to provide the second edition of this comprehensive reference book written to provide pediatric oncologists, pediatric nurse practitioners and nurses, psychologists, psychiatrists, and social workers with an overview of the many facets that comprise excellent psychosocial care for children and adolescents living with cancer and their family members. It is not meant to be an exhaustive reference; rather, we aim to provide helpful recommendations and best-practice models for management of the emotional reactions of children undergoing cancer treatment and of the psychological, cognitive, and social outcomes for children with cancer and of the emotional burdens of pediatric cancer care on siblings and parents.*

This book is unique in covering a broad range of topics in pediatric psychosocial oncology in a handbook format geared toward answering specific clinical questions. An international group of experts in each area have written chapters to help clinicians review, anticipate, and respond to emotional issues that often arise in the context of treating pediatric cancer patients. The chapters may also help pediatric oncologists and nurses recognize when it may be best to refer patients to their behavioral health colleagues. We hope it will be useful for those who are establishing pediatric oncology services or adding psychosocial components to existing clinics to recognize the range of ways in which psychological services are important for the provision of comprehensive care to children and families.

All the chapters have been updated since the first edition, and they cover a wide range of topics including psychological aspects of particular pediatric cancers and their treatments; how to talk to a child and family at critical times during the disease course; new information in the area of genomics and genetic testing; individual, family, educational, psychological and psychiatric interventions; and how to assist the international family. This second edition includes additional clinically relevant chapters on stem cell transplantation, social media, and electronic psychosocial interventions. Each chapter recognizes the necessity of embracing an interdisciplinary approach to ensure that each child with cancer has the best opportunity for a satisfying, productive life and, when cure is not possible, that the period of time prior to, at the time of, and following death occurs with as much dignity as possible for the child and family.

We have assembled the handbook in concise, practical, and highly readable brief chapters. The "bullet point" format is to facilitate a quick read. We anticipate most clinicians will turn to specific chapters for guidance on managing the psychosocial care of their patients. Some readers, however, may read this book more comprehensively. Such readers may find that similar issues such as diagnosis, treatment completion, and survivorship are addressed in multiple chapters. Each chapter stands on its own, but we have

cross-referenced places in other chapters where similar issues are dealt with in depth to avoid unnecessary repetition.

We recognize that not every child and family will have access to a full range of psychosocial services. We have described state-of-the-art care as practiced in cancer centers that have the greatest resources, and we acknowledge that providers elsewhere will know how best to adapt these principles to their own circumstances, possibly through developing referral connections to trusted mental health colleagues in their community.

We hope this book conveys and extends the warm spirit of collegiality and mutual respect that exists in many centers and regions between pediatric oncology clinicians and mental health professionals who have productively worked together over the past 30 years.

We recognize that children are cared for by adults with many different relationships to them. We use the term *parents* to refer to all adult caregivers who have parenting responsibilities.

The pharmacological dosage information in this handbook has been carefully reviewed. However, the drug manufacturers' current indications, dosage information, and drug interaction warnings should be consulted prior to prescribing any drug listed in this work.

Lori Wiener, PhD, DCSW
Maryland Pao, MD
Anne E. Kazak, PhD, ABPP
Mary Jo Kupst, PhD
Andrea Farkas Patenaude, PhD
Robert J. Arceci, MD, PhD

Acknowledgments

The editors thank those who support access of psychosocial support services to all patients and families coping with cancer.

We especially acknowledge the support of Vicki Sardi-Brown and Peter Brown who established the Mattie Miracle Cancer Foundation, which focuses on improving psychosocial care and on the development of psychosocial standards of care for children with cancer.

Jimmie C. Holland, MD, is widely recognized as the founder of "psychooncology". She is also the driving force behind the creation of the first and second editions of *Pediatric Psycho-Oncology: A Quick Reference on the Psychosocial Dimensions of Cancer Symptom Management*. While Dr. Holland's recent research focus has been geriatric oncology, she has consistently challenged clinicians and researchers to investigator and focus on the needs of children with cancer. We will be forever be indebted to Dr. Holland for her insight, wisdom, perseverance, and commitment to the needs of children with cancer and their families and for helping to pave the road for the field of Pediatric Psychooncology to develop, expand, and thrive.

Contributors

Allistair Abraham, MD
Attending, Division of Blood and Marrow Transplantation
Center for Cancer and Blood Disorders
Children's National Health System
Assistant Professor of Pediatrics
The George Washington University Medical Center
Washington, D.C.

Robert J. Arceci, MD, PhD
Director
Children's Center for Cancer and Blood Disorders
Director of the Ron Matricaria Institute of Molecular Medicine
Phoenix Children's Hospital
Department of Child Health
University of Arizona College of Medicine
Phoenix, Arizona

Sarah R. Brand, PhD
Instructor of Psychology
Harvard Medical School
Staff Psychologist
Department of Psychosocial Oncology and Palliative Care
Dana Farber Cancer Institute
Boston, Massachusetts

Joanna Breyer, PhD
Assistant Professor of Psychiatry
Harvard Medical School
Pediatric Psychosocial Unit
Dana-Farber Cancer Institute
Boston, Massachusetts

Ronald T. Brown, PhD
President
University of North Texas at Dallas
Dallas, Texas

William M. Bukowski, PhD
Research Chair in Psychology
Concordia University
Montreal, Quebec

Anu Chittenden, MS, CGC
Genetic Counselor
Center for Cancer Genetics and Prevention
Dana-Farber Cancer Institute
Boston, Massachusetts

Todd Dalberg, DO
Division of Pediatric Hematology-Oncology
Oregon Health and Science University
Doernbecher Children's Hospital
Portland, Oregon

Brian P. Daly, PhD
Assistant Professor of Psychology
Drexel University
Philadelphia, Pennsylvania

Mary Lynn Dell, MD, DMin
Department of Psychiatry
Nationwide Children's Hospital
Columbus, Ohio

Jeffrey S. Dome, MD, PhD
Professor of Pediatrics
George Washington University School of Medicine and Health Sciences
Thomas Willson and Lenore Williams McKnew Professor of Pediatric Oncology
Chief, Divisions of Hematology and Oncology
Children's National Medical Center
Washington, D.C.

Dennis Drotar, PhD
Professor of Pediatrics
University of Cincinnati
Clinical Psychologist, Behavioral Medicine and Clinical Psychology
Cincinnati Children's Hospital
Cincinnati, Ohio

Jennifer S. Ford, PhD
Clinical Psychologist
Department of Psychiatry and Behavioral Sciences
Memorial Sloan-Kettering Cancer Center
New York, New York

Sarah Friebert, MD
Director of Pediatric Palliative Care
Akron Children's Hospital
Akron, Ohio

Debra L. Friedman, MD
Associate Professor of Pediatrics
Vanderbilt University School of Medicine
Vanderbilt-Ingram Cancer Center
Monroe Carell Jr. Children's Hospital at Vanderbilt
Nashville, Tennessee

Terry J. Fry, MD
Center for Cancer Research
National Cancer Institute
National Institutes of Health
Bethesda, Maryland

Martha A. Grootenhuis, PhD
Pediatric Psychosocial Department
Emma Children's Hospital
Academic Medical Center
University of Amsterdam
Amsterdam, the Netherlands

Daniel H. Grossoehme, DMin, MS
Chaplain
Division of Pulmonary Medicine
Assistant Professor of Pediatrics
University of Cincinnati
Cincinnati, Ohio

Kristina K. Hardy, PhD
Neuropsychology Division
Children's National Medical Center
The George Washington University School of Medicine
Washington, D.C.

Pamela S. Hinds, PhD, RN, FAAN
Department of Pediatrics
The George Washington University
Department of Nursing Research and Quality Outcomes
Children's National Medical Center
Washington, D.C.

Marilyn J. Hockenberry, PhD, RN, PNP-BC, FAAN
Bessie Baker Professor of Nursing
Duke University School of Nursing
Durham, North Carolina

Lisa M. Horowitz, PhD, MPH
Staff Scientist and Clinical Psychologist
National Institute of Mental Health
National Cancer Institute
Bethesda, Maryland

Mary Jo Kupst, PhD
Emeritus Professor of Pediatrics
Medical College of Wisconsin
Milwaukee, Wisconsin

Ernest R. Katz, PhD
Adjunct Professor of Clinical Pediatrics and Psychology
Keck School of Medicine and the Dornsife College
University of Southern California
University Center of Excellence in Developmental Disabilities
Children's Hospital of Los Angeles
Founder, School Transition and Re-Entry Service
Children's Center for Cancer and Blood Diseases
Los Angeles, California

Anne E. Kazak, PhD, ABPP
Center for Healthcare Delivery Science
Nemours Pediatric Health System
Wilmington, Delaware
Professor
Department of Pediatrics
Sidney Kimmel Medical School of Thomas Jefferson University
Philadelphia, Pennsylvania

Julia A. Kearney, MD
Child Psychiatrist and Pediatrician
Department of Psychiatry and Behavioral Sciences
Memorial Sloan-Kettering Cancer Center
New York, New York

Elliot J. Krane, MD
Professor of Anesthesiology
Perioperative and Pain Medicine
Stanford University Medical Center
Professor of Pediatrics
Lucile Salter Packard Children's Hospital
Stanford, California

Marguerite S. Lederberg, MD
Psychiatrist
Department of Psychiatry and Behavioral Sciences
Memorial Sloan-Kettering Cancer Center
New York, New York

Jennifer M. Levine, MD, MSW
Assistant Professor of Pediatrics
Columbia University Medical Center
New York, New York

Avi Madan-Swain, PhD
Associate Professor of Pediatrics
University of Alabama at Birmingham
Children's Hospital of Alabama
Birmingham, Alabama

David Malkin, MD
Professor of Pediatrics and Medical Biophysics
University of Toronto
The Hospital for Sick Children
Toronto, Ontario

D. Richard Martini, MD
Division Chief of Behavioral Health
Department of Pediatrics
University of Utah School of Medicine
Director of Psychiatry and Behavioral Health
Primary Children's Medical Center
Salt Lake City, Utah

Hilary McClafferty, MD, FAAP
Assistant Director of the Integrative Medicine Fellowship
Arizona Center for Integrative Medicine
University of Arizona
Tucson, Arizona

Anna T. Meadows, MD
Emeritus Professor of Pediatrics and Medicine
University of Pennsylvania School of Medicine
The Children's Hospital of Philadelphia
Philadelphia, Pennsylvania

Melinda Merchant, MD, PhD
Pediatric Oncology Branch Clinical Director
Center for Cancer Research
National Cancer Institute
Bethesda, Maryland

Travis Mickelson, MD
Assistant Professor of Pediatrics
University of Utah
Salt Lake City, Utah

Robert B. Noll, PhD
Professor of Pediatrics
University of Pittsburgh School of Medicine
Children's Hospital of Pittsburgh
Pittsburgh, Pennsylvania

Diana E. Nora, BS
National Institute of Mental Health
National Cancer Institute
Bethesda, Maryland

Kevin C. Oeffinger, MD
Director, Adult Long-Term Follow-Up Program
Memorial Sloan-Kettering Cancer Center
New York, New York

Ahna L. H. Pai, PhD
Associate Professor of Pediatrics
University of Cincinnati
Clinical Psychologist, Behavioral Medicine and Clinical Psychology
Cincinnati Children's Hospital
Cincinnati, Ohio

Maryland Pao, MD
Clinical Director and Deputy Scientific Director
National Institute of Mental Health
Bethesda, Maryland

Andrea Farkas Patenaude, PhD
Associate Professor of Psychology
Harvard Medical School
Director, Psychology Research and Clinical Services
Center for Cancer Genetics and Prevention
Dana-Farber Cancer Institute
Boston, Massachusetts

Sean Phipps, PhD
Chair & Member, Department of Psychology
St. Jude Children's Research Hospital
Memphis, Tennessee

Rebecca A. Prengler, MD
National Institute of Mental Health
National Cancer Institute
Bethesda, Maryland

Maria E. Radulovic, CMI-Spanish
Department of Social Work
National Institutes of Health
Bethesda, Maryland

Steven K. Reader, PhD
Nemours Center for Healthcare Delivery Science
Assistant Professor
Department of Pediatrics
Sidney Kimmel Medical School of Thomas Jefferson University
Philadelphia, Pennsylvania
Division of Behavioral Health
Nemours Alfred I. duPont Hospital for Children
Wilmington, Delaware

Cheryl C. Rodgers, PhD, RN, CPNP
Assistant Professor of Nursing
Duke University School of Nursing
Durham, North Carolina

Marilyn A. Rosen, MLS
University of Rochester Medical Center
Rochester, New York

Julia H. Rowland, PhD
Director, Office of Cancer Survivorship
National Cancer Institute
Bethesda, Maryland

Olle Jane Z. Sahler, MD
Professor of Pediatrics, Psychiatry, Medical Humanities, and Oncology
University of Rochester Medical Center
Rochester, New York

Lisa A. Schwartz, PhD
Psychologist
University of Pennsylvania School of Medicine
The Children's Hospital of Philadelphia
Philadelphia, Pennsylvania

Jaclyn Schienda, ScM, CGC
Genetic Counselor
Center for Cancer Genetics and Prevention
Dana-Farber Cancer Institute
Boston, Massachusetts

Aziza Shad, MD
Amey Distinguished Professor of Neuro-Oncology and Childhood Cancer
Georgetown University Lombardi Comprehensive Cancer Center
Georgetown University Hospital
Washington, D.C.

Nirali N. Shah, MD
Pediatric Oncology Branch
Center for Cancer Research
National Cancer Institute
National Institutes of Health
Bethesda, Maryland

Charles A. Sklar, MD
Director
Long-Term Follow-Up Program
Department of Pediatrics
Memorial Sloan-Kettering Cancer Center
New York, New York

Giselle Saulnier Sholler, MD, MSc
Department of Pediatric Hematology-Oncology
Helen DeVos Children's Hospital
Grand Rapids, Michigan

Barbara Sourkes, PhD
Professor of Pediatrics
Stanford University Medical Center
Lucile Salter Packard Children's Hospital
Stanford, California

Rachel A. Tunick, PhD
Instructor of Psychology
Harvard Medical School Staff Psychologist
Department of Psychiatry and Children's Hospital Primary Care Center
Boston Children's Hospital
Boston, Massachusetts

Katherine E. Warren, MD
National Cancer Institute
Pediatric Oncology Branch
Bethesda, Maryland

Alan S. Wayne, MD
Professor of Pediatrics
Keck School of Medicine
University of Southern California
Director
Children's Center for Cancer and Blood Diseases
Head, Division of Hematology, Oncology and Blood & Marrow Transplantation
Children's Hospital Los Angeles
Los Angeles, California

Lori Wiener, PhD, DCSW
Co-Director
Behavioral Science Core
Director
Psychosocial Support and Research Program
Pediatric Oncology Branch,
Center for Cancer Research
National Cancer Institute,
National Institutes of Health
Bethesda, Maryland

Matthew Wright, MD
Chief of Hospital Medicine
Western Regional Medical Center
Goodyear, Arizona

Lonnie Zeltzer, MD
Professor of Pediatrics,
Anesthesiology, and Psychiatry and Biobehavioral Sciences
David Geffen School of Medicine
University of California
Los Angeles, California
UCLA Mattel Children's Hospital
UCLA Jonsson Comprehensive Cancer Center
Los Angeles, California

Introduction

Jimmie C. Holland

It is a pleasure to write the Purpose and Overview for the second edition of the *Pediatric Oncology Handbook* relating to the psychiatric and psychosocial care of the child with cancer. The first edition was published in 2009 by the American Psychosocial Oncology Society. This second edition is being published by the Oxford University Press as part of its series of handbooks for psychiatric and psychosocial care of the adult, child, and elder with cancer. Because these handbooks depend strongly on tables and rapid learning formats, they are ideal not only for the busy clinic staff but also for teaching students, residents, and fellows in pediatric oncology and its subspecialties. Every child at every visit presents psychological and social issues that, if handled well, result in enhanced trust, reduced distress, and greater adherence to treatment. If handled poorly, the converse too often results in greater distress, distance from the staff, and poorer adherence. The consequences are too great to be neglected as we progress to truly patient-centered care.

The psychological care of children with cancer has for far too long lacked both standards of quality and clinical practice guidelines to assure that pediatric staff take care of the whole child with the optimal tools available. This volume goes a long way to assuring that this goal is reached. The editors are five outstanding clinical investigators in pediatric psycho-oncology. They have chosen authors to write on specific critical areas of clinical care: how to talk to parents and children; psychological and social problems by site of cancer; management of common psychological and physical symptoms; psychotherapeutic approaches; cognition problems, their impact and treatment; school issues; spiritual and religious issues. Taken together, these chapters contribute greatly to the development of the first practice guidelines for psychosocial care of children with cancer. This is a goal that we can all celebrate as a benchmark of progress.

Jimmie C. Holland, MD Wayne E. Chapman
Chair in Psychiatric Oncology Department of Psychiatry and
Behavioral Sciences
Memorial Sloan-Kettering Cancer Center, New York,
New York
February, 2014

Section I

Cancers of Childhood

Chapter 1

Leukemias and Lymphomas

Nirali N. Shah and Alan S. Wayne

Leukemia is the most common pediatric cancer diagnosis, comprising approximately 25% of all childhood malignancies.[1] Lymphomas account for about 15% of pediatric cancer. In general, cure rates for leukemias and lymphomas in childhood exceed 75%. However, the outcome varies between subtypes and prognostic groups, and hematologic malignancies remain the most common cause of cancer-related mortality in pediatrics.[2]

Although leukemias and lymphomas are highly curable in pediatrics, given the intensive nature of therapy that these children receive, they are at risk for a multitude of acute and late effects associated with both the therapy and the underlying disease (Table 1.1).[3–5] Diagnosis and treatment confer significant physical, neurocognitive, psychological, and social risks that can have life-long impact. Accordingly, children and adolescents with leukemia and lymphoma should receive systematic and serial neurocognitive and psychosocial assessments during and after treatment as part of comprehensive multidisciplinary care. This chapter will focus specifically on these particular issues.

Epidemiology and Treatment

Acute lymphoblastic leukemia (ALL) accounts for about 75% and acute myelogenous leukemia (AML) about 20% of pediatric leukemia, whereas chronic myelogenous leukemia (CML) and juvenile myelomonocytic leukemia (JMML) are infrequent (Figure 1.1),[6,7] with Hodgkin disease slightly more prevalent than non-Hodgkin lymphoma (NHL). There are three predominant subtypes of NHL in pediatrics, each occurring with approximate equal frequency: lymphoblastic (LBL), large cell, and Burkitt lymphoma.

Treatment regimens, which are specific to the disease subtype and risk-adapted based on clinicopathologic prognostic features, in most cases consist of combination chemotherapy (Table 1.2). Initial treatment phases are commonly more intensive, with the goal of inducing and consolidating a remission. Such "induction" and "consolidation" chemotherapy is usually administered in an inpatient setting. Individuals with ALL require long-term low-dose outpatient chemotherapy ("maintenance" or "continuation"). The duration of therapy is also subtype specific and may be quite prolonged (e.g., 2–3 years for ALL and LBL). Central nervous system (CNS) involvement is common in ALL, AML, LBL, and Burkitt lymphoma, and CNS-directed

Table 1.1 Common Acute and Late Effects for Patients Diagnosed with and Treated for Leukemia or Lymphoma

Effect	Risk Factors	Monitoring
Cardiomyopathy	Anthracyclines (e.g., doxorubicin) (cumulative dose) Cyclophosphamide (high-dose) Radiation (thoracic) Younger age	Electrocardiogram Echocardiogram
Pulmonary dysfunction	Bleomycin (cumulative dose) Busulfan Radiation (thoracic) Stem cell transplantation	Oxygen saturation Pulmonary function tests Imaging studies
Endocrinopathies	Alkylators (e.g., cyclophosphamide) (cumulative dose) Radiation (CNS, glandular) Corticosteroids Stem cell transplantation	Growth Sexual development & fertility Hormone levels
Altered body appearance and composition (e.g., alopecia, Cushing's, obesity), Metabolic syndrome	Corticosteroids (cumulative dose) Systemic chemotherapy Stem cell transplantation Radiation Reduced physical activity High caloric intake Female sex	Growth and development Body mass index Lipid profile
Infection (e.g., opportunistic, bacterial, fungal, viral)	Systemic chemotherapy Corticosteroids Stem cell transplantation Hodgkin lymphoma Central venous catheter	Blood counts Surveillance for infection Prophylactic interventions for prevention of infectious complications during periods of highest risk
Cytopenias with need for transfusion and associated complications (e.g., transfusion-transmitted infection; iron overload)	Systemic chemotherapy Stem cell transplantation	Blood counts Surveillance for bleeding
Neurocognitive dysfunction, Neuropsychologic dysfunction, Leukoencephalopathy	Intrathecal chemotherapy (e.g., Methotrexate, Cytarabine, Corticosteroids) Radiation (CNS) Female sex Younger age Environmental/Familial/Social	Neurodevelopmental, Psychosocial, Cognitive, Educational, and Vocational assessments

(continued)

Table 1.1 (Continued)

Effect	Risk Factors	Monitoring
CNS hemorrhage, Stroke	L-asparaginase Hyperleukocytosis Radiation (CNS)	Neurologic examination Imaging studies
Neuropathies	Vinca alkaloids (cumulative dose) Cranial nerve infiltration	Neurologic examination
Ophthalmologic: Cataracts, Glaucoma, Visual impairment	Corticosteroids (cumulative dose) Stem cell transplantation Radiation (eye) Cranial nerve infiltration Orbital infiltration	Visual acuity Ophthalmologic examination
Osteopenia, Osteonecrosis	Corticosteroids (cumulative dose) Stem cell transplantation Caucasians Adolescents	Bone mineral density Imaging studies
Secondary malignancy	Alkylators (e.g., cyclophosphamide) (cumulative dose) Epipodophyllotoxins (e.g., etoposide) Stem cell transplantation Radiation	Complete blood count Imaging studies

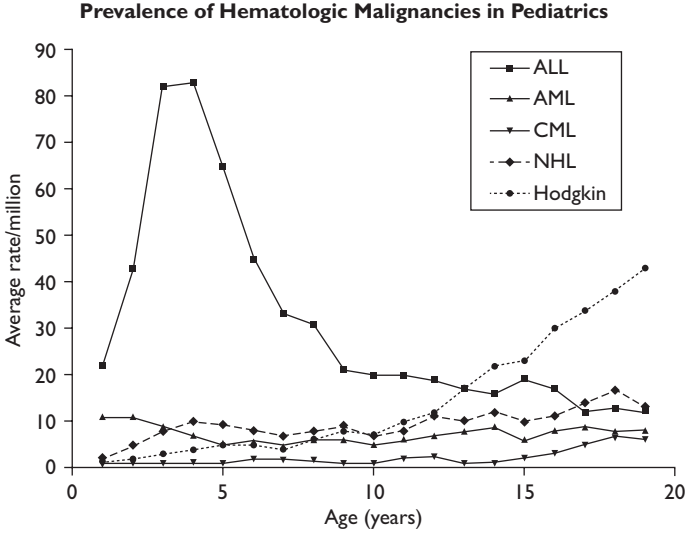

Figure 1.1 Age-Related Prevalence. Data from: Gloeckler L, Percy C, Bunin GR. Introduction. In: Cancer Incidence and Survival among Children and Adolescents: United States SEER Program 1975–1995, NIH Publication 1999; (99-4649): 1–16.

Table 1.2 General Approach to Treatment: Treatment Is Stratified Based on Diagnostic Subtype and Clinicopathologic Prognostic Risk Factors

Diagnosis	Therapy
ALL & LBL	Treatment phases (excluding mature B-cell) • Induction chemotherapy • Consolidation/Re-induction chemotherapy • Maintenance chemotherapy, total treatment duration 2–3 years • CNS treatment: intrathecal chemotherapy; +/- XRT if CNS involvement Mature B-ALL: as per Burkitt lymphoma regimens
AML	Treatment phases (excluding promyelocytic) • Induction chemotherapy • Consolidation chemotherapy • CNS treatment: intrathecal chemotherapy; +/- XRT if CNS involvement • Matched-sibling donor SCT (higher risk subtypes) Promyelocytic: Include all-*trans*-retinoic acid during induction and maintenance
JMML	Allogeneic SCT
CML	Tyrosine kinase inhibitors (e.g., imatinib mesylate) Allogeneic SCT
Burkitt lymphoma	Chemotherapy CNS treatment: intrathecal chemotherapy; +/- XRT if CNS involvement
Large cell lymphoma	Chemotherapy
Hodgkin lymphoma	Chemotherapy +/- local XRT

therapy is required for all individuals with those diagnoses. Allogeneic stem cell transplantation (SCT) is commonly utilized for individuals with the highest risk features (e.g., relapse). Radiation therapy (XRT) is infrequently employed in pediatric hematologic malignancies. Its use is reserved for local control of active disease involving the CNS or testes, certain cases of lymphoma, and for pretransplant conditioning (total body irradiation).

Neurocognitive and Psychosocial Concerns

Because disease prevalence is age related (Figure 1.1), the risks of long-term side effects, or late effects, are influenced by the state of organ development at the time of diagnosis and during treatment. Patients with leukemia and lymphoma are at particularly high risk for long-standing neurocognitive and neuropsychologic dysfunction due, in part, to the intensive therapy that these children receive. This is especially notable in those patients receiving CNS-directed therapy.[8–10] More recent retrospective evaluations

of cohorts of patients treated for childhood ALL have demonstrated impairment rates of up to 60% across various neurocognitive domains in adult survivors, likely related to a combination of intensive intrathecal methotrexate and CNS radiation therapy.[10,11] Girls and younger children are at greatest risk of neurocognitive toxicities (Table 1.3).[8,9] As these impairments are often persistent and significant, ongoing monitoring of neurocognitive and psychological function is needed well into adulthood.[11] (See Chapter 18, Cognitive Sequelae.)

Table 1.3 Common Neurocognitive and Psychosocial Effects

Effect	Intervention
Neurocognitive dysfunction • IQ • Memory • Executive functions • Problem solving • Organizational skills • Visual perceptual and motor function • Information processing	Educational approaches (e.g., individualized education plans, accommodations, special services, assistive technologies, targeted learning and "re-training" approaches, selective environment) Compensatory techniques (e.g., organizational tools, environmental supports, occupational therapy) Vocational training
Neuropsychological dysfunction • Attention, concentration Risky behaviors Adjustment (school, work, relationships) • Social isolation • Adherence	Mental health specialist, neuropsychologist Behavioral therapy Behavioral therapy Support groups Social work and/or school counselor support Psychosocial service consultation for psychotherapy, cognitive behavioral therapy Pharmacologic approaches
Psychiatric manifestations • Anxiety • Depression • Post-traumatic stress	Mental health specialist Individual and group therapy Pharmacologic approaches
School absenteeism	Homebound schooling School re-entry program
Financial concerns • Healthcare expenses • Unemployment • Parental loss of work • Lack of insurance	Social work assessment Referral to local and national resources
Healthcare concerns • Complex multidisciplinary care needs • Inadequate insurance	Coordinated oncologic care followed by cancer survivorship care Social work support

The diagnosis and treatment of leukemias and lymphomas not only impacts the affected child, but also parents, siblings, extended family members, and their communities.[12] (See Chapter 24, Impact of Cancer on Family and Siblings.) In addition to the direct psychological effects that the diagnosis of cancer confers, there are substantial psychosocial consequences of the demands of treatment and associated toxicities (Tables 1.1 and 1.3).[13] Specific to ALL therapy, for instance, the use of steroids may lead to a Cushingoid appearance with altered body image, in addition to a sequale of medical complications including but not limited to the development of obesity, metabolic syndrome, and osteoporosis.

Prolonged school absenteeism can be anticipated during induction and consolidation chemotherapy and after stem cell transplantation, and the ability to arrange for intermittent school attendance is often restricted due to concerns about infection during periods of neutropenia and especially after bone marrow transplant. Homebound schooling can be arranged, but may further contribute to the sense of isolation due to reduced direct interaction with peers. Videoconferencing and other innovative strategies of communication that allow improved integration of the affected child into the school system may prove especially effective in patients with leukemia or lymphoma. (See Chapter 27, School and Peer Relationships, and Chapter 28, School and Academic Planning.) Maintenance therapy for ALL, which usually begins 9–12 months after the start of treatment, is often a good time for children to re-enter the school system and return to their usual routine.

Adolescents and young adults (AYA) are at particularly high risk for adverse consequences, for both medical and psychosocial issues.[14] The AYA population generally has worse outcomes, and may be at higher risk of relapse.[15] A specific emphasis on medication adherence, which may be especially challenging during the long periods of ALL maintenance therapy and also posttransplant, is an important consideration for which innovative strategies of monitoring may be indicated.[16] (See Chapter 16, Psychosocial Issues for Transplant Patients and Donors, and Chapter 17, Medication Adherence.) After completion of therapy, the AYA population often has significant barriers to obtaining healthcare during survivorship follow-up and monitoring. Specific attention to obtaining resources and medical insurance is needed to optimize their long-term care and overall quality of health. (See Chapter 33, Pediatric Cancer Survivors.)

Summary

Although leukemias and lymphomas are highly curable in pediatrics, diagnosis and treatment confer significant physical, psychological, and social risks that can have life-long impact.

- Children and adolescents with leukemia and lymphoma should receive systematic and serial neurocognitive and psychosocial assessments during and after treatment as part of comprehensive multidisciplinary care.

- Long-term follow-up is essential to monitor for possible relapse, treatment- and disease-associated toxicities, and secondary malignancies.
- Recommended guidelines for healthcare providers and patients/families are available from a number of sources (e.g., http://www.survivorshipguidelines.org).

References

1. Gloeckler L, Percy C, Bunin GR. Introduction. In: *Cancer Incidence and Survival among Children and Adolescents: United States SEER Program 1975-1995*. NIH Publication 99-4649. Bethesda, MD: National Institute of Health; 1999: 1–16.
2. Gloeckler Ries L, Mortality CC. In: Reis LAG SM, Gurney JG, et al. eds. *Cancer Incidence and Survival among Children and Adolescents: United States SEER Program 1975-1995. National Cancer Institute, SEER Program*. NIH Pub No. 99-4649. Bethesda, MD: National Cancer Institute; 1999: 165–170.
3. Oeffinger KC, Mertens AC, Sklar CA, et al. Chronic health conditions in adult survivors of childhood cancer. *N Engl J Med*. 2006;355(15):1572–1582.
4. Pui CH, Cheng C, Leung W, et al. Extended follow-up of long-term survivors of childhood acute lymphoblastic leukemia. *N Engl J Med* 2003;349(7):640–649.
5. Leung W, Hudson MM, Strickland DK, et al. Late effects of treatment in survivors of childhood acute myeloid leukemia. *J Clin Oncol*. 2000;18(18):3273–3279.
6. Pui CHe. *Childhood Leukemias*. 3rd ed. Cambridge, UK: Cambridge University Press; 2012.
7. Smith MA, Gloeckler Ries LA, Gurney JG, Ross J. Leukemia. In: Reis LAG SM, Gurney JG, et al., eds. *Cancer Incidence and Survival among Children and Adolescents: United States SEER Program 1975-1995*. National Cancer Institute, SEER Program. NIH Pub. No. 99-4649. Bethesda, MD: National Cancer Institute; 1999: 17–34.
8. Butler RW, Mulhern RK. Neurocognitive interventions for children and adolescents surviving cancer. *J Pediatr Psychol*. 2005;30(1):65–78.
9. Nathan PC, Patel SK, Dilley K, et al. Guidelines for identification of, advocacy for, and intervention in neurocognitive problems in survivors of childhood cancer: a report from the Children's Oncology Group. *Arch Pediatr Adolesc Med*. 2007;161(8):798–806.
10. Duffner PK, Armstrong FD, Chen L, et al. Neurocognitive and neuroradiologic central nervous system late effects in children treated on pediatric oncology group (POG) P9605 (standard risk) and P9201 (lesser risk) acute lymphoblastic leukemia protocols (ACCL0131): a methotrexate consequence? A report from the children's oncology group. *J Pediatr Hematol Oncol*. 2014;36(1):8–15.
11. Krull KR, Brinkman TM, Li C, et al. Neurocognitive outcomes decades after treatment for childhood acute lymphoblastic leukemia: a report from the st jude lifetime cohort study. *J Clin Oncol*. 2013;31(35):4407–4415.
12. Buchbinder D, Casillas J, Krull KR, et al. Psychological outcomes of siblings of cancer survivors: a report from the Childhood Cancer Survivor Study. *Psychooncology* 2011;20(12):1259–1268.
13. Zebrack BJ, Zeltzer LK, Whitton J, et al. Psychological outcomes in long-term survivors of childhood leukemia, Hodgkin's disease, and

non-Hodgkin's lymphoma: a report from the childhood cancer survivor study. *Pediatr.* 2002;110(1 Pt 1):42–52.

14. Zebrack B, Isaacson S. Psychosocial care of adolescent and young adult patients with cancer and survivors. *J Clin Oncol.* 2012;30(11):1221–1226.
15. Keegan TH, Tao L, Derouen MC, et al. Medical care in adolescents and young adult cancer survivors: what are the biggest access-related barriers? *J Cancer Surviv.* 2014;8(2):282–292.
16. Rohan JM, Drotar D, Alderfer M, et al. Electronic monitoring of medication adherence in early maintenance phase treatment for pediatric leukemia and lymphoma: Identifying patterns of nonadherence. *J Pediatr Psychol.* 2013; Epub ahead of print.

Chapter 2

Neuroblastoma

Giselle Saulnier Sholler

Neuroblastoma is the most common malignant solid tumor of childhood, and may be diagnosed as early as the prenatal period (by fetal ultrasound) or as late as young adulthood (Table 2.1). Most patients are from 3 months of age to 6 years.[1,2] Children may present with symptoms related to an abdominal mass, back or bone pain with limping, malaise, anemia, and occasionally orbital ecchymoses or skin nodules. Diagnosis is by surgical biopsy, or by bone marrow biopsy and urine sample for tumor markers (catecholamines). The child's treatment, which depends on the age, stage, and tumor biology, may range from biopsy only followed by observation for regression (low risk) to surgery, high dose chemotherapy, stem cell transplant, and immune therapies for advanced disease (high risk). Children diagnosed at less than one year of age usually have early stage disease with good outcome (survival may exceed 80–90%) and require minimal therapy. Children diagnosed after 12–18 months of age often have disease that has spread and require extensive therapy with lower chance of survival.[3,4]

Therapy

Large studies have shown that patients treated on clinical trials tend to have better survival. Most children will be eligible for such trials. These clinical trials (often called "protocols"), which include standard therapies as well as a novel component, are designed to improve survival. Sometimes protocols involve a randomization between standard therapy and standard therapy plus experimental therapies. During the protocol consent discussion between physicians and family, the parents will require time and professional help in order to understand the therapies and side effects (Tables 2.2 and 2.3). Parents should be informed that, if they decline to consent, they will be given the best known therapy with full medical staff support. Treatment of side effects is also an important component of treatment (Table 2.4). In some instances, in disorders with a particularly poor prognosis, discussions of palliative care are initiated at the time of diagnosis.

Table 2.1 Specific Neuroblastoma Syndromes

Perinatal neuroblastoma	• Neuroblastoma may be detected as a suprarenal mass by fetal ultrasound before birth, or in the newborn. • Current practice is to inform parents that most of these infants do not need treatment and to observe with repeated radiological scans.
Stage IVS (IV = Four and S = Special)	• Infants have small primary adrenal tumors but with liver infiltration and/or cells in the marrow cavity and/or in pale skin nodules. • Encourage parents that these infants do well in more than 90% of cases. • This unique kind of tumor shrinks over about 6 months without treatment, although rare cases may need surgical intervention or chemotherapy because an enlarged liver may compromise breathing. • Explain to parents that some patients have disease that may progress initially, and "watching" the tumor shrink may be stressful.
Opsoclonus-Myoclonus (neurological syndrome of 'dancing' eye movements and/or muscle jerks with dis-coordination almost always associated with early Stage I-II tumors)	• It is very likely their child will survive; this neurological syndrome can improve with chemotherapy and immunotherapy with prednisone and gammaglobulin, but some children can have life-long neurological complications.
Spinal Cord compression by tumor	• If treated rapidly, the nerve damage improves over time with surgery, radiation, or chemotherapy. • Patients may have residual weakness or paralysis.
Horner's Syndrome (unilateral facial paralysis from neck tumor)	• This mild asymmetry of the face may be permanent.
Profuse diarrhea secondary to increased hormone (VIP) secretion	• This syndrome abates with chemotherapy.

Table 2.2 Common Questions Asked by Parents During the Diagnostic Period

Question	Parent Education and Management Suggestions
What caused the tumor?	• Neuroblastoma occurs in all populations throughout the world. • Provide parents information that the cause is currently unknown. • Recent studies have linked an inherited gene, ALK, with neuroblastoma in approximately 7% of patients. Children may be tested for this.
What about environment/ diet?	• Environmental studies have not found a cause.

(continued)

Table 2.2 (Continued)

Question	Parent Education and Management Suggestions
Was my child born with the disease?	• There is evidence that occasionally neuroblastoma may start in the fetus, but no links have been found to maternal health.
Could the cancer have been found earlier?	• Timing of diagnosis has not been shown to change the outcome. • High-risk neuroblastoma is a rapidly arising tumor in children greater than one year at diagnosis. It is often not present long before diagnosis.
What will the diagnostic testing be like for my child?	• Diagnostic testing by several different radiologic procedures are required. These often require sedation so the child will be able to remain still. The child may be hungry and irritable beforehand and when he or she wakes up. Biopsy of the tumor and bone marrow will be done under sedation and may require pain medication afterwards.
Will my child need surgery?	• The child may require a central venous line placement for chemotherapy. • Surgery to remove all or part of a tumor may be needed. The child's pain will be treated hour by hour with medicine and he or she will be up and about usually in 1–3 days depending on the extent of surgery.
Will I be able to continue working?	• The impact of intensive treatments should be discussed with parents, particularly with regard to support available to the family and employment options. • One parent may be required to take time away from work to be with the child throughout treatment. • Offer to call, write letters to employers, and assist with family medical leave forms. • Assist with communication with the child's school. It may be possible for the child to keep up with schoolwork in hospital if hospital educators are present.
What about my other child(ren)?	• Suggest the parent(s) ask family and friends for help, shared parenting if possible (weekdays, weekends, week nights). • Encourage the involvement, understanding, and frequent presence of siblings. • Refer siblings to sibling support programs within the hospital if available or within the community.
Will my child die?	• At diagnosis, the therapy given is expected to cure the child. • Support with encouragement that survival is the goal and therapies continually improve. • It is important to remain aware that some tumors will be resistant to therapy and not all children will survive; this will only be known months to years after the initial diagnosis.

Table 2.3 Concerns Surrounding Initiation of Treatment

Concern	Parent Education and Management Suggestions
Will the chemotherapy cause immediate sickness, vomiting and/or pain?	• Provide information that many children play and have no side effects as the drugs infuse. • Supportive medicines are given to prevent nausea before it occurs and can be adjusted for each child as needed. • Nausea or pain may develop over days, but will be treated to minimize or omit their symptoms.
What will happen when we go home?	• Children will not be discharged until it is considered safe. Some patients may require IV nutrition and/or pain medication, which may be able to be given at home. • Some children will require daily subcutaneous shots with G-CSF to help boost the immune system. Nurses can help teach parents how to administer injections. • Due to risk of infections, lifestyle adjustments will be necessary. Fevers can be life threatening and parents need to be instructed to call their physician immediately if a fever occurs. • Most children will resume normal activities, including eating, and many return to school as their energy rebounds.
Should I make my child eat? Should he/she follow a certain diet or take vitamins?	• Certain vitamins may interact with chemotherapy and it is important to encourage parents to discuss all supplements with their physician before taking them. • Chemotherapy may change the child's taste preferences. Encourage parents to have their child try different foods to find those he/she likes to eat. Encourage children to help choose foods to eat and eating times.
What should I be concerned about at home?	• Caution that the parent should call the medical-care team at any time for any fever or any new symptoms. • Parents should call if they are worried about their child for any reason.
How quickly will the tumor shrink?	• Most children may have fewer symptoms from the cancer as early as the first few weeks after chemotherapy, suggesting improvement. • Radiology scans and other repeat tests are generally scheduled 2–3 months later, when future therapies need to be determined and scheduled.

Table 2.4 Common Chemotherapy Side Effects

Side Effect	Parent Education and Management Suggestions
Vincristine: Constipation Peripheral neuropathy Pain	• Parents should keep track of stool frequency and consistency. • Therapy with prescribed laxative medications should be given. • Numbness, tingling, or mild weakness in the hands or feet is possible and usually resolves over time. • Pain is reversible and will subside over several days to a week if Vincristine is decreased or omitted. • Inform parents that Tylenol should not be given unless they have spoken with the child's physician and only after a temperature check, since it may mask the presence of fever and infection. • Opiates may be prescribed if pain is significant.
Platinum: Hearing Loss	• Hearing loss can increase over time with recurrent exposures of this drug. • Patients will be monitored with hearing tests to prevent significant loss, but some children will need hearing aids.
Retinoic Acid	• Side effects such as dry, chapped lips can be treated topically.

Bone Marrow Transplant (BMT)

Bone Marrow Transplant includes very high dose chemotherapy with stem cell rescue for Stage IV and high-risk Stage III patients, given as one or two transplants successively (Table 2.5).

Therapeutic Options for the Child Who Relapses

There is currently no standard therapy for children who relapse. Explore options with the family taking into consideration those that are most suitable for the child and family's expectations:

- New chemotherapy with standard drugs.
- Experimental therapies, including Phase I and II trials, should also be considered if part of a clinical trial.
- Radiation therapy to sites of symptomatic masses or to bone disease to help with pain and symptoms.
- Nonstandard holistic therapies chosen from outside of the medical center (see Chapter 21, Integrative Oncology).
- No therapy with curative goal—Supportive care to manage symptoms and promote quality time with family including family trips and time for increased sibling involvement (see Chapter 11, Pain, and Chapter 30, Integrating Palliative Care).

Table 2.5 Concerns Surrounding BMT

Concern	Parent Education and Management Suggestions
Fear of "high dose" therapy	• Children will experience similar side effects as with standard chemotherapy, although older patients may have more nausea-vomiting during therapy. Other side effects, such as mucositis, may be more severe.
Fear of the early weeks after stem cell infusion when the child has very low blood cells	• Patients will be given almost daily transfusions, antibiotics, and will be given IV nutrition as well as opiates for mucositis mouth pain. • The child may prefer to rest in bed most of the day, but many patients feel better in the third week. • Gently inform parents that fatalities are rare but may occur.
Distress with isolation in specially filtered room, boredom, inability to have visitors, exhaustion	• Shared parent-family "shifts" may help to give caregivers a break. • A planned daily schedule (washing up, mouth care, out-of-bed-to-chair, TV time, book time, craft time, etc.) is helpful. • Daily phone calls or e-mails to and from family and friends are helpful. • Creating a free, personalized webpage through programs such as Care Pages or Caring Bridge can help families keep others abreast of their child's progress.
Antineuroblastoma antibody therapy posttransplant	• Side effects of pain and numbness during infusion will be treated aggressively.

Symptom Relief for the Child Who Is Dying

- Children with neuroblastoma may experience worsening bone pain, fever-infections, massive liver enlargement, lung infiltration, or sometimes bony growths on scalp and trunk. This may occur over weeks to months.
- Reassure family that aggressive pain and symptom control will be provided and involvement with a hospice team is recommended.
- Decision for continuation of transfusions is individual. Transfusions can help alleviate some side effects such as fatigue from anemia and bleeding due to low platelet counts, but it may become difficult and painful to transport child to the hospital and the decision to stop transfusions is acceptable. A balance of the advantages and disadvantages are usually different for each family.
- Communicate openly with parents and encourage honest discussions with relatives, siblings, and the patient, if age appropriate.
- Encourage sibling involvement, even if distressing, to alleviate future survival guilt.
- Integrate palliative care principles. (See Chapter 30, Integrating Palliative Care).

References

1. American Cancer Society. *Cancer Facts and Figures*. Atlanta, GA: Author; 2008.
2. Bernstein ML, et al., A population-based study of neuroblastoma incidence, survival, and mortality in North America. *J Clin Oncol*. 1992: 10(2):323–329.
3. Brodeur GM, et al., Revisions of the international criteria for neuroblastoma diagnosis, staging, and response to treatment. *J Clin Oncol*. 1993: 11(8):1466–1477.
4. Park JR, Eggert A, Caron H. Neuroblastoma: biology, prognosis, and treatment. *Pediatr Clin North Am*. 2008;55(1):97–120.

Chapter 3

Wilms Tumor

Jeffrey S. Dome

Pediatric kidney tumors comprise approximately 7% of cancers in children and adolescents.[1] The vast majority are Wilms tumor, though other tumor types such as renal cell carcinoma, clear cell sarcoma of the kidney, malignant rhabdoid tumor, congenital mesoblastic nephroma, and renal sarcomas are occasionally encountered. Wilms tumor is historically important because it was one of the first malignancies to be treated with a multidisciplinary approach involving surgeons, pediatric oncologists, and radiation oncologists. With multimodality therapy, cure rates have risen to 85–90%.[2] Additionally, biological studies of Wilms tumor have provided a paradigm for our understanding cancer genetics.[2] Although the majority of patients with Wilms tumor are cured, patients and families face significant psychosocial challenges at diagnosis, during treatment, and after completion of therapy (Table 3.1). The total care of a patient involves a multidisciplinary approach that includes family support measures to optimize the quality of life.

Diagnosis of Wilms Tumor

- The average age of a child at the time of Wilms tumor diagnosis is about 3.5 years, although older children and adults may develop the disease.[3]
- In North America, there is a slight predisposition to Wilms tumor in girls compared to boys.
- The tumor presents as an abdominal mass and is often discovered incidentally by parents giving their child a bath or by a healthcare provider during a routine physical exam.
- Children with Wilms tumor usually appear healthy but may have abdominal pain, fever, or hematuria.
- Physical examination reveals high blood pressure in about 25% of patients.
- Once an abdominal mass is detected, patients undergo an abdominal ultrasound and computed-tomography (CT) scan.
- The most common sites of spread for Wilms tumor are the lymph nodes, lung, and liver.
- Wilms tumor can also migrate through blood vessels and grow into the renal vein, inferior vena cava, and, occasionally, the heart.