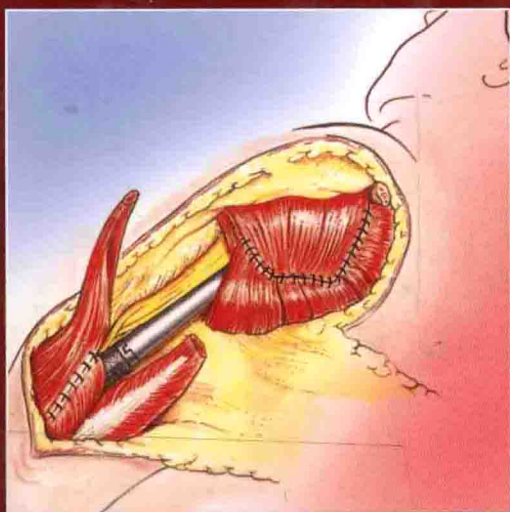


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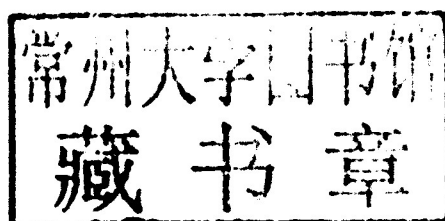
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**JAMES C. WITTIG & JACOB BICKELS**

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# OPERATIVE TECHNIQUES IN ORTHOPAEDIC SURGICAL ONCOLOGY

## **Martin M. Malawer, MD, FACS**

Professor (Clinical Scholar) of Orthopaedics  
Professor of Pediatrics (Hematology and  
Oncology)

Georgetown University School of Medicine  
Washington, District of Columbia

Professor of Orthopaedic Surgery  
Director of Orthopedic Oncology  
George Washington University  
Washington, District of Columbia

Consultant (Pediatric and Surgery Branch)  
National Cancer Institute, National  
Institutes of Health  
Bethesda, Maryland

## **James C. Wittig, MD**

Associate Professor of Orthopedic Surgery  
Chief of Orthopedic Oncology and  
Sarcoma Program  
Mount Sinai Medical Center  
New York, New York

Chief, Orthopedic Oncology  
Department Orthopedic Surgery  
Director, Skin and Sarcoma Division  
John Theuer Cancer Center  
Hackensack University Medical Center  
Hackensack, New Jersey

## **Jacob Bickels, MD**

Professor, Orthopedic Surgery  
National Unit of Orthopedic  
Oncology  
Tel-Aviv Sourasky Medical Center  
Tel Aviv, Israel



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## Dedication

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William F. Enneking, MD, Professor of Orthopaedic Surgery, University of Florida School of Medicine, Gainesville, Florida

Kenneth C. Francis, MD, Clinical Professor of Orthopaedic Surgery, New York University School of Medicine, New York, NY, and former Chief of the Bone Tumor Service at Memorial Sloan-Kettering Cancer Center, New York, New York

Ralph C. Marcove, MD, Professor of Orthopaedic Surgery, Columbia University College of Physicians and Surgeons, New York, NY, and former Chief of the Bone Tumor Service at Memorial Sloan-Kettering Cancer Center, New York, New York

*William F. Enneking, MD*, was one of the original pioneers and academic researchers who laid the foundation for the basic biology and understanding of bone and soft-tissue sarcomas. He held the Eugene L. Jewett Professorship in Orthopaedic Surgery at the University of Florida from 1977 until his retirement. Before Bill Enneking, bone cancer studies were confused and lacked direction. His personal innovations advanced the field in concept and in practice. His teaching has resulted in a score of clinical and laboratory oncologists who dominate the field in both practice and research activities. Dr. Enneking's dedication, patience, and intellect have provided the scientific basis for the new discipline of Orthopaedic Oncology.

Prior to the early 1970s almost all bone sarcomas were treated by amputation. Two surgeons in the United States, both in New York City, began the specialty of limb-sparing surgery as we know it today. They were *Kenneth C. Francis and Ralph C. Marcove*. Dr. Francis was the Clinical Professor of Orthopaedic Surgery at New York University School of Medicine. He was also the first Orthopaedic surgeon to be the Chief of the Bone Tumor service at Memorial Sloan-Kettering Cancer Center. His first, and only, Fellow was Ralph Marcove, who ultimately became Chief of the Bone Tumor service at Memorial Sloan-Kettering Cancer Center as well. The two men independently performed and developed the original techniques of distal femoral resections, total femoral prosthetic replacements, shoulder girdle resections, scapular resections, and pelvic resections.

In 1973, Dr. Francis performed the first distal femoral resection at the New York University Hospital, and reconstructed the defect with a long-stemmed Walldius prosthesis. This, to the author's knowledge, was the first such surgery performed in the United States, if not the world.

This book is dedicated to the knowledge, insightfulness, teaching, and surgical skills of these three men. *All three surgeons were my mentors*. I have attempted to build upon their tradition of excellence, determination, and innovation in preparing the current and future generations of Orthopaedic Oncology surgeons.

Martin M. Malawer, MD, FACS  
2011



# FOREWORD

In the past two decades significant progress has occurred, in the management of patients with musculoskeletal cancers, that has improved both the survival and the quality of life of afflicted patients. Changes in the management of these patients have mirrored trends in the entire field of oncology. The most significant change has been improvement in the surgical techniques for the resection of musculoskeletal cancers based on a detailed understanding of the anatomic features of each particular tumor site, as well as an appreciation of the natural biology that affects the local spread of these tumors. *Operative Techniques in Orthopaedic Surgical Oncology* provides a detailed description of important changes in the surgical approach to these patients. Amputation, once the mainstay of treatment for patients with bony and soft-tissue extremity sarcomas, has now largely been replaced by limb-sparing surgery using innovative approaches to cancer resection and the advent of new reconstruction techniques that can restore function in ways not possible even a decade ago. Although debilitating amputations are still required for some patients with locally extensive cancers, most patients with these tumors can look forward to surgical procedures that will maximize their functional outcome.

The sophistication of many of these limb-sparing surgical approaches has resulted in a shift in the expertise required to perform these procedures and, increasingly, specialists in the management of musculoskeletal tumors have arisen to provide these patients with the benefits of these advances. A second change in the management of these patients has been the introduction of combined modality treatment utilizing the concerted application of surgery, radiation therapy and chemotherapy in a carefully integrated fashion to maximize survival and quality of life. The use of local radiation therapy has had a profound impact on the ability to achieve local control. Cooperation between surgeons and radiation therapists often results in the tailoring of surgical procedures to maximize the combined application of these two effective treatment modalities. Although impact on overall survival has not been demonstrated due to the addition of radiation therapy, important advances in improving the quality of life of patients receiving this combined-modality treatment have been evident.

A third change impacting on the survival of patients with musculoskeletal cancers has been the aggressive resection of metastatic deposits. Surgery remains the most effective treatment for adult patients with limited metastatic cancer, and durable disease-free and overall survival can be achieved by the vigorous resection of metastases arising from these cancers. Although the use of adjuvant chemotherapy has had dramatic impact on the treatment of many musculoskeletal cancers in children, the impact of chemotherapy on adults remains a controversial issue. Although transient responses can be seen in adults with many types of soft-tissue sarcomas, they are very rarely curative and the development of more effective systemic treatments for patients with soft-tissue sarcomas remains a major challenge in the future treatment of patients with this disease.

The state-of-the-art surgical techniques described in this text, applied in the context of integrated cancer therapy, can provide great benefit to patients with musculoskeletal cancers.

Steven A. Rosenberg, MD, PhD  
Chief, Surgery Branch  
National Cancer Institute  
National Institutes of Health  
Bethesda, MD



# CONTRIBUTORS

**Adesegun Abudu, FRCS**  
Royal Orthopaedic Hospital Oncology  
Service  
Northfield, Birmingham, United Kingdom

**Aharon Amir, MD**  
Attending Surgeon  
Department of Plastic Surgery  
Tel-Aviv Sourasky Medical Center  
Tel-Aviv, Israel

**Jacob Bickels, MD**  
Head, Service for the Management of  
Metastatic Bone Disease  
Attending Surgeon, National Unit of  
Orthopedic Oncology  
Tel-Aviv Sourasky Medical Center  
Professor of Orthopedic Surgery  
Sackler School of Medicine, Tel-Aviv  
University  
Tel-Aviv, Israel

**Loretta B. Chou, MD**  
Professor of Orthopaedic Surgery  
Stanford University  
Chief, Foot and Ankle Service  
Lucile Packard Children's Hospital at  
Stanford  
Palo Alto, California

**Ernest U. Conrad III, MD**  
Professor of Orthopaedics  
University of Washington  
Director, Bone Tumor Clinic  
Children's Hospital and Regional Medical  
Center  
Seattle, Washington

**Jeffrey J. Eckardt, MD**  
Director, Orthopaedic Oncology  
UCLA Santa Monica Orthopaedic Center  
Santa Monica, California

**Steven Gitelis, MD**  
Professor and Vice Chairman of  
Orthopaedic Surgery  
Director, Section of Orthopaedic Oncology  
Rush University Medical Center  
Chicago, Illinois

**Robert Grimer, FRCS**  
Consultant, Orthopaedic Surgeon  
Royal Orthopaedic Hospital  
Northfield, Birmingham, United Kingdom

**Eyal Gur, MD**  
Director, Unit of Microsurgery  
Department of Plastic Surgery  
Tel-Aviv Sourasky Medical Center  
Senior Lecturer  
Sackler School of Medicine  
Tel-Aviv University  
Tel-Aviv, Israel

**Yvette Ho**  
Research Assistant  
Washington Musculoskeletal Tumor Center  
Washington Cancer Institute  
Washington, District of Columbia

**Lee Jeys, MB, ChB, MSc, FRCS**  
Consultant, Orthopaedic Surgeon  
Specialist in Hip, Knee, and Oncology  
Surgery  
Midland Hip & Knee Clinic  
Royal Orthopaedic Hospital  
Northfield, Birmingham, United Kingdom

**Norio Kawahara, MD, PhD**  
Clinical Professor  
Department of Orthopaedic Surgery  
Kanazawa University School of Medicine  
Ishikawa, Japan

**Kristen Kellar-Graney, MS**  
Tumor Biologist and Clinical Research  
Coordinator  
Washington Cancer Institute  
Washington, District of Columbia

**Piya Kiatsevi, MD**  
Orthopaedic Oncology Unit  
Institute of Orthopaedics  
Lerdsin Hospital  
Bangkok, Thailand

**Yehuda Kollender, MD**  
Attending Surgeon, National Unit of  
Orthopedic Oncology  
Tel-Aviv Sourasky Medical Center  
Senior Lecturer  
Sackler School of Medicine, Tel-Aviv  
University  
Tel-Aviv, Israel

**Jennifer Lisle, MD**  
Assistant Professor of Orthopedics,  
Rehabilitation, and Pediatrics  
University of Vermont College of Medicine  
Vermont Children's Hospital at Fletcher  
Allen Health Care  
Burlington, Vermont

**Martin M. Malawer, MD, FACS**  
Professor (Clinical Scholar) of  
Orthopaedics  
Professor of Pediatrics (Hematology and  
Oncology)  
Georgetown University Medical Center  
Washington, District of Columbia

Professor of Orthopaedic Surgery  
Director of Orthopedic Oncology  
George Washington University  
Washington, District of Columbia

Consultant (Pediatric and Surgery Branch)  
National Cancer Institute, National  
Institutes of Health  
Bethesda, Maryland

**Isaac Meller, MD**  
Director, National Unit of Orthopedic  
Oncology  
Tel-Aviv Sourasky Medical Center  
Professor of Orthopedic Surgery  
Sackler School of Medicine, Tel-Aviv  
University  
Tel-Aviv, Israel

**Benjamin J. Miller, MD**  
Rush Orthopaedic Oncology  
Rush University Medical Center  
Chicago, Illinois

**Hideki Murakami, MD**  
Lecturer of Orthopaedic Surgery  
Department of Orthopaedic Surgery  
Kanazawa University School of Medicine  
Ishikawa, Japan

**Gregory P. Nicholson, MD**  
Associate Professor of Orthopaedic Surgery  
Rush University Medical Center  
Chicago, Illinois

**Tamir Pritsch, MD**  
Department of Orthopaedic Surgery  
Tel Aviv Sourasky Medical Center  
Tel Aviv, Israel

**Amir Sternheim, MD**  
Orthopedic Oncology  
Washington Cancer Institute  
Washington, District of Columbia

**H. Thomas Temple, MD**  
Professor of Orthopaedics and Pathology  
Vice Chair and Chief, Oncology Division  
Director, University of Miami Tissue Bank  
University of Miami Leonard M. Miller  
School of Medicine  
Miami, Florida

**Daria Brooks Terrell, MD**  
Attending Physician  
Department of Orthopaedic Oncology  
Washington Hospital Center  
Washington, District of Columbia  
Consultant (Pediatric and Surgery Branch)  
National Cancer Institute, National  
Institutes of Health  
Bethesda, Maryland

**Katsuro Tomita, MD**  
Professor of Orthopaedic Surgery  
Department of Orthopaedic Surgery  
Kanazawa University School of Medicine  
Ishikawa, Japan



**Walter W. Virkus, MD**

Associate Professor of Orthopaedic Surgery  
Associate Attending Surgeon (Orthopedic  
Surgery)  
Rush University Medical Center  
Chicago, Illinois

**Jason Weisstein, MD, MPH, FACS**

Assistant Professor of Orthopaedics and  
Sports Medicine  
Co-Director, Northwest Tissue Center  
University of Washington  
Seattle, Washington

**James C. Wittig, MD**

Associate Professor of Orthopedic Surgery  
Chief of Orthopedic Oncology and  
Sarcoma Program  
Mount Sinai Medical Center  
New York, New York

Chief, Orthopedic Oncology  
Department Orthopedic Surgery  
Director, Skin and Sarcoma Division  
John Theuer Cancer Center  
Hackensack University Medical Center  
Hackensack, New Jersey

**Yehuda Wolf, MD**

Director, Department of Vascular Surgery  
Tel-Aviv Sourasky Medical Center  
Professor of Surgery  
Sackler School of Medicine, Tel-Aviv  
University  
Tel-Aviv, Israel

**Arik Zaretski, MD**

Attending Surgeon  
Department of Plastic Surgery  
Tel-Aviv Sourasky Medical Center  
Tel-Aviv, Israel



# PREFACE

This is the third book in a series that documents and details the progress and innovations made in surgical techniques in the field of Orthopaedic Oncology. In 1992, Drs. Sugarbaker and Malawer published *Musculoskeletal Surgery for Cancer: Principles and Techniques*. This book was comprised of 30 chapters in black and white, describing in detail the new procedures in Orthopaedic Oncology. This text was and is used internationally as a standard text in the field of Orthopaedic Oncology; it was translated into Chinese, Spanish, Russian, and Portuguese.

In 2001, Malawer and Sugarbaker published a completely revised and updated textbook, *Musculoskeletal Cancer Surgery: Treatment of Sarcomas and Allied Diseases*. One of the first surgical texts published in color, including full color illustrations and schematics. This book was based on over 50 years of combined surgical and oncological experience. The 2001 text has recently been translated into Chinese by one of the outstanding medical publishing houses in China, Shanghai Scientific and Technical Publishers, and is widely used by the many of new and developing orthopaedic oncologists at some of the leading universities and teaching hospitals.

This book, *Operative Techniques in Orthopaedic Surgical Oncology*, is the natural progression as a representative text of the current state of Orthopaedic Oncology today. The coauthors of this text are Drs. James C. Wittig and Jacob Bickels. This text represents a movement in the field toward Orthopaedic Oncology standing on its own as a true subspecialty in the field of Orthopaedics. They and Dr. Malawer represent over 60 years of surgical experience dedicated to the treatment of bone and soft tissue sarcomas.

This book is a reprint of Part 4, Oncology, of *Operative Techniques in Orthopaedic Oncology*, Volume 2, edited by Sam W. Wiesel. It consists of 4 sections, and 42 chapters in total: Section 1, Surgical Management; Section 2, Shoulder Girdle and Upper Extremities; Section 3, Spine and Pelvis; and Section 4, Lower Extremities. In addition, for the first time, there are 25 surgical videos, edited and correlated with most chapters. Many of these videos have been shown at various national and international meetings. Thus, taken together with the text, the surgeon is given the most complete visual as well as didactic information to date.

The purpose of this text, as with the previous ones, is to illustrate and detail the surgical techniques, indications, and anatomy of each procedure. From the Preface of the previous edition, it was stated that “surgery is a visual field, and that the surgeon works in three dimensions.” That concept holds just as true today, and has only been enhanced by digital, three-dimensional, navigational, and other real-time imaging techniques. The prior Preface continues on to say:

*Therefore the majority of contents of this book are accompanied by photographs and illustrations of the surgical procedure, as well as preoperative studies that the authors feel are uniquely important. The surgical descriptions, anatomic depictions, and the significance of each imaging study to each operative procedure are emphasized . . . It was the purpose of the authors to present [this] data in a simple visual format.*

The authors of the current text hope that this book is helpful to all surgeons undertaking the care of sarcoma patients. Additionally, that this text both reinforces and builds upon the previous foundation of techniques in the field of Orthopaedic Oncology.

Martin M. Malawer, MD



Preface to original textbook Malawer M and Sugarbaker P. *Musculoskeletal Cancer Surgery: Treatment of Sarcomas and Allied Diseases*. Kluwer Academic Press; 2001.

Musculoskeletal cancer surgery has undergone dramatic changes within the past two decades. Limb-sparing surgery is the hallmark of the surgical advances developed by this specialty. The role of the orthopaedic oncology surgeon in the 1970s was to perform high level amputations. This is what distinguished the orthopaedic cancer surgeon from the general orthopaedic surgeon. The development of limb-sparing surgery, in conjunction with the dynamic advances in imaging and chemotherapy, created the specialty of musculoskeletal (orthopaedic) oncology. The operative procedures performed today barely existed two decades ago. Today, approximately 90–95% of all bone and soft-tissue sarcomas can be treated by limb-sparing surgery. The aim of this book is to present in a concise, organized and well-illustrated format, the surgical techniques involved with limb-sparing procedures of the entire musculoskeletal system, including the upper and lower extremities as well as the pelvic and shoulder girdle.

These techniques are a combination of surgical procedures that used to be considered of interest only to the general surgeon, vascular surgeon, plastic surgeon and orthopaedic surgeon. The development of these multiple-treatment strategies and techniques has created the field of musculoskeletal cancer surgery as we know it today.

The surgical experience of the two authors, Martin M. Malawer, MD, and Paul H. Sugarbaker, MD, spans more than two decades each. The combined experience of Dr Malawer, Professor of Orthopaedic Surgery at George Washington University, Children's Hospital National Medical Center, and the Washington Cancer Institute Department of Orthopaedic Oncology, and Dr Sugarbaker, who developed several of the techniques described in this book while at the National Cancer Institute, Bethesda, Maryland, forms the foundation for this book. The techniques described in these pages have been developed by both authors over the past two decades. The aim of this textbook is to illustrate well a step-by-step approach to limb-sparing procedures of the musculoskeletal system. These techniques, although not unique, are described specifically from the authors' experience, which began in the early 1970s and continues today. Dr. Malawer has extensively reported different techniques for limb-sparing resections and endoprosthetic reconstructions. The technique of allograft replacement was developed in the early 1970s and utilized widely until the 1980s. Such techniques are not utilized by the authors but are well described elsewhere.

Surgery is a visual field, and the surgeon works in three dimensions. Therefore, the majority of the contents of this book are accompanied by photographs and illustrations of the surgical procedure as well as preoperative studies that the authors feel are uniquely important. The surgical descriptions, anatomic depictions, and the significance of each imaging study to each operative procedure are emphasized.

Since the mid-1970s the two authors have provided surgical care for approximately 4000 patients with benign, malignant and metastatic lesions, in children and adults. Detailed files, including operative photographs, pathology slides, Kodachrome slides and slides of the significant imaging studies for all of the major cases have been kept on each operative procedure. This collection is housed at the Center for Orthopaedic Oncology and Musculoskeletal Research at the Washington Cancer Institute and is available to American and international surgeons and oncologists for study and research purposes.

Despite the complexity of limb-sparing surgical procedures and the multiple imaging studies, it was the purpose of the authors to present these data in a simple visual format. The bibliography is limited in most chapters, because the techniques described were developed by the authors themselves. The general chapters are well-referenced with the most up-to-date citations.

This book contains 36 chapters, divided into four sections. It addresses the basic pathology, surgical technique and management of all extremity and pelvic and shoulder girdle tumors as well as abdominal and truncal sarcoma surgery.

Chapter 1 includes a discussion of bone and soft-tissue sarcomas, including their epidemiology, radiographic characteristics and pathology. Biopsy techniques are discussed in Chapter 2. In Chapter 3, chemotherapy is discussed, outlining the chemotherapeutic agents that are effective in the treatment of bone and soft-tissue sarcomas. Dr. Dennis Priebe details the chemotherapy strategy that has evolved over the past 20 years. The experience of isolated-limb perfusion, a new technique used to treat sarcomas, is described in Chapter 4. Dr Brian Fuller chief of the Radiation Oncology Branch at the National Cancer Institute, in Chapter 5, describes the most current and comprehensive strategy for radiation therapy for

extremity sarcomas. Chapter 6 discusses the use of cryosurgery in the treatment of certain bone tumors. This technique was developed by Dr Ralph Marcove at Memorial Sloan-Kettering Cancer Center and has been continued by the authors. Chapters 7 and 8 summarize the management of abdominal and pelvic sarcomas. This author, Dr Paul Sugarbaker, has uniquely developed many techniques over the past 25 years. Chapters 9, 10 and 11 provide overviews of the treatment of tumors of the shoulder girdle, pelvic girdle and metastatic bone tumors, respectively. These chapters lay the foundation for the specific limb-sparing procedures in the remaining portion of the textbook.

Section Two describes in detail the techniques for muscle group resections of the lower extremity including gluteus maximus, adductor muscle group, quadriceps muscle group, posterior thigh and popliteal space. Section Three discusses the surgical amputations utilized in orthopaedic oncology surgery. These tend to be high-level amputations with which orthopaedic and general surgeons tend not to be familiar. Forequarter amputation and the various types of hemipelvectomy are described. Dr Sugarbaker developed the technique of an anterior myocutaneous flap for patients with massively contaminated pelvic and buttock structures that cannot be resected by the standard posterior flap hemipelvectomy. These operative procedures were often deemed difficult and radical, but are still used today to cure certain patients who cannot be treated by limb-sparing surgeries. Phantom pain, a complication following an amputation for cancer patients, is discussed in Chapter 24. Due to the neuropathic effects of the chemotherapeutic agents, as well as the young age of these patients, this problem occurs frequently. Dr LeeAnn Rhodes described treatment considerations for phantom limb pain.

Section Four describes in detail the limb-sparing procedures around the pelvis, proximal and distal femur, proximal tibia, fibula, proximal humerus and scapula. Although these procedures are performed at many centers throughout the world today, the techniques, morbidity and complications vary tremendously. The techniques as described by Dr Malawer, which he has perfected over the past 20 years, are presented in detail. The required staging studies and unique anatomic considerations, from a surgeon's viewpoint, are outlined for each anatomic site, with special considerations for the evaluation of the imaging studies. The Appendix includes two chapters of interest to the musculoskeletal surgeon. Appendix A is a description of an abdominoinguinal incision developed by Dr. Constantine Karakousis for resection of pelvic tumors. This is a combined intra- and extraperitoneal approach. Canine osteosarcomas are discussed by Dr Charles Kuntz. Osteosarcomas in dogs are extremely common, and the techniques and basis of limb-sparing surgical techniques in dogs offer an excellent orthopaedic animal model and are of interest to musculoskeletal cancer surgeons.

Martin M. Malawer, MD





# ACKNOWLEDGMENTS

Halstead once stated that “the operating room was the Laboratory for the surgeon.” This is as true today as it was a century ago. The surgical techniques described in this textbook were developed over a course of now 30 years in the treatment of over 6,000 patients. We would like to acknowledge our patients for their belief in us as surgeons and in our ability to restore the function to their extremities in lieu of an amputation. For those patients who did require an amputation, we acknowledge the tremendous will that they have had.

Surgery and surgical training is a three-way communication between patients, fellows, and senior surgeons. This spoken and unspoken dialogue has been the foundation of many of these concepts, ideas, and motivations presented in this textbook. We would like to acknowledge all of the residents and fellows that we have trained.

This textbook would not exist without the outstanding artistic skills of Joyce Hurwitz. Mrs. Hurwitz has spent 30 years in close cooperation with the senior authors, illustrating multiple publications, presentations, posters, and textbooks. Her utmost knowledge of surgical anatomy, combined with her interest and motivation in participating within the scientific community, sets her apart as a truly gifted individual.

Finally, the authors would like to acknowledge the hard work and dedication of all of the contributors. We are especially appreciative of the long hours and days that our Senior Research Assistant, Kristen Kellar-Graney, has spent, without which this book never would have been completed. It goes without saying that a text of this magnitude took many long and hard hours of work by the authors but, more important, time away from our wives and families, whom we greatly appreciate and acknowledge.

Martin M. Malawer, MD, FACS  
2011

To my beloved and inspiring family: Shelly, Yarden, Tom, Nimrod, and Dan

JB



# RESIDENCY ADVISORY BOARD

The editors and the publisher would like to thank the resident reviewers who participated in the reviews of the manuscript and page proofs. Their detailed review and analysis was invaluable in helping to make certain this text meets the needs of residents today and in the future.

**Daniel Galat, MD**

Dr. Galat is a graduate of Ohio State University College of Medicine and the Mayo Clinic Department of Orthopaedic Surgery residency program.

He is currently serving at Tenwek Hospital in Kenya as an orthopedic surgeon.

**Lawrence V. Gulotta, MD**

Fellow in Sports Medicine/Shoulder Surgery  
Hospital for Special Surgery  
New York, New York

**Dara Chafik, MD, PhD**

Southwest Shoulder, Elbow and Hand Center  
PC  
Tucson, Arizona

**Gautam Yagnik, MD**

Attending Physician  
Orthopaedic Surgery  
DRMC Sports Medicine  
DuBois, Pennsylvania

**Gregg T. Nicandri, MD**

Assistant Professor  
Department of Orthopaedics (SMD)  
University of Rochester  
School of Medicine and Dentistry  
Rochester, New York

**Catherine M. Robertson, MD**

Assistant Clinical Professor  
UCSD Orthopaedic Surgery—Sports Medicine  
San Diego, California

**Jonathan Schoenecker, MD**

Assistant Professor  
Departments of Orthopaedics, Pharmacology  
and Pediatrics  
Vanderbilt University  
Nashville, Tennessee





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## INDEX I-1



## OVERVIEW

- An understanding of the basic biology and pathology of bone and soft tissue tumors is essential for appropriate planning of their treatment.
- This chapter reviews the unique biologic behavior of soft tissue and bone sarcomas, which provides the basis for their staging and resection and the use of appropriate adjuvant treatment modalities.
- A detailed description of the clinical, radiographic, and pathological characteristics for the most common sarcomas is presented.

## EPIDEMIOLOGY

- Soft tissue and bone sarcomas are a rare and heterogeneous group of tumors. These neoplasms represent less than 1% of all adult and 15% of pediatric malignancies.
- As of 2006, the annual incidence in the United States, which remains relatively constant, was approximately 6000 to 7000 soft tissue sarcomas and 2750 bone sarcomas.
- In 2006, the overall mortality rate was 30% for soft tissue sarcomas and 45% for bone sarcomas.
- In the U.S., the 5-year survival rates for osteosarcoma and Ewing sarcoma were comparable among 15- to 29-year-olds, about 60% for the most recent era. Survival rates for chondrosarcoma exceeded 90% in the most recent era. The U.S. bone cancer mortality was highest for males and females 15 to 19 years of age.

## RISK FACTORS

- Risk factors for soft tissue and bone sarcomas include previous radiation therapy, exposure to chemicals (eg, vinyl chloride, arsenic), immunodeficiency, prior injury (scars, burns), chronic tissue irritation (foreign body implants, lymphedema, chronic infection), neurofibromatosis, Paget disease, bone infarcts, and genetic cancer syndromes (eg, hereditary retinoblastoma, Li-Fraumeni syndrome, Gardner syndrome, Rothmund-Thomson syndrome, Werner syndrome, Bloom syndrome), Marfucci syndrome, Ollier disease, multiple osteochondromatosis, and hereditary multiple exostoses. In most patients, however, no specific etiology can be identified.
- In the past two decades, both survival and quality of life of patients with soft tissue and bone sarcomas have improved dramatically as a result of the multimodality treatment approach. Limb-sparing surgery, used in combination with chemotherapy and radiation therapy, can achieve cure in the majority of patients with soft tissue and bone sarcomas, and resection is performed in lieu of amputation in more than 90% of all patients.
- The three most common soft tissue sarcomas are malignant fibrous histiocytoma (MFH), liposarcoma, and leiomyosarcoma.
- The most common bone sarcomas are osteosarcoma, chondrosarcoma, and Ewing sarcoma.

## PATHOPHYSIOLOGY AND BIOLOGIC BEHAVIOR

- Sarcomas originate primarily from elements of the mesodermal embryonic layer.
- Soft tissue sarcomas are classified according to the adult tissue that they resemble.
  - Similarly, bone sarcomas usually are classified according to the type of matrix production: osteoid-producing sarcomas are classified as osteosarcomas, and chondroid-producing sarcomas are classified as chondrosarcomas.
- Tumors arising in bone and soft tissues have characteristic patterns of biologic behavior because of their common mesenchymal origin and anatomic environment. Those unique patterns form the basis of the staging system and current treatment strategies.
  - Histologically, sarcomas are graded as low, intermediate, or high grade. The grade is based on tumor morphology, extent of pleomorphism, atypia, mitosis, matrix production, and necrosis, with the two main factors being mitotic count and spontaneous tumor necrosis.
  - Tumor grade represents the tumor's biologic aggressiveness and correlates with the likelihood of metastases. Low-grade lesions metastasize in fewer than 15% of patients. High-grade lesions metastasize in over 20% of patients.
- Sarcomas form a solid mass that grows centrifugally, with the periphery of the lesion being the least mature.
  - In contradistinction to the true capsule that surrounds benign lesions, which is composed of compressed normal cells, sarcomas usually are enclosed by a reactive zone, or pseudocapsule. This pseudocapsule consists of compressed tumor cells and a fibrovascular zone of reactive tissue with a variable inflammatory component that interacts with the surrounding normal tissues.
  - The thickness of the reactive zone varies according to the histogenic type and grade of malignancy. High-grade sarcomas have a poorly defined reactive zone that may be locally invaded by the tumor (**FIG 1A**).
- Tumor foci within the reactive zone are called *satellite lesions*.
- High-grade, and occasionally low-grade, may break through the pseudocapsule to form metastases, termed *skip metastases*, within the same anatomic compartment in which the lesion is located. By definition, these are locoregional micrometastases that have not passed through the circulation (**FIG 1B**).
  - This phenomenon may be responsible for local recurrences that develop in spite of apparently negative margins after a resection.
  - Although low-grade sarcomas regularly interdigitate into the reactive zone, they rarely form tumor skip nodules beyond that area (**FIG 1C,D**).
- Sarcomas respect anatomic borders. Local anatomy influences tumor growth by setting natural barriers to extension. In