

# Diagnosis of Musculoskeletal Tumors and Tumor-like Conditions

Clinical, Radiological and Histological  
Correlations - The Rizzoli Case Archive

Piero Picci  
Marco Manfrini  
Davide Maria Donati  
Marco Gambarotti  
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Daniel Vanel  
Angelo Paolo Dei Tos  
*Editors*

*Second Edition*

 Springer

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*Editors*

Piero Picci  
I.S.G. Italian Sarcoma Group  
Bologna  
Italy

Davide Maria Donati  
Istituto Ortopedico Rizzoli  
Bologna  
Italy

Alberto Righi  
Department of Pathology  
IRCCS Istituto Ortopedico Rizzoli  
Bologna  
Italy

Angelo Paolo Dei Tos  
Department of Medicine  
University of Padua School of Medicine  
Padua  
Italy

Marco Manfrini  
IIIrd Clinic of Orthopaedic Oncology  
Istituto Ortopedico Rizzoli  
Bologna  
Italy

Marco Gambarotti  
Department of Pathology  
IRCCS Istituto Ortopedico Rizzoli  
Bologna  
Italy

Daniel Vanel  
Istituto Rizzoli  
Bologna  
Italy

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*This book is dedicated to  
Prof. Mario Campanacci,  
Prof. Mario Mercuri,  
Dr. Gaetano Bacci, and  
Dr. Marco Alberghini  
who strongly supported the dissemination  
of knowledge of musculoskeletal tumors.*

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## Preface

This is a second book based on the data of the Rizzoli case archive, after the one published in 2014 with the title *Atlas of Musculoskeletal Tumors and Tumorlike Lesions: The Rizzoli Case Archive*.

In this new one, not only the epidemiological data are updated with the addition of about 7000 new lesions diagnosed in 5 years between 2013 and 2017, but also some chapters were added and/or modified based on the more recent discoveries in terms of clinical, histological, and molecular investigations.

In particular, small blue round cell tumors are deeply analyzed and presented, as the whole spectrum of vascular lesions, with the introduction of new entities, as for the presentation of a new classification for those sarcomas of bone in the past simply diagnosed as “malignant fibrous histiocytoma” or “fibrosarcoma.” More space is also dedicated to soft tissue lesions and also to those more rare entities not discussed in the previous book.

What presented reflects the experience of the Rizzoli Orthopedic Institute in over 100 years of treatment of musculoskeletal tumors and tumorlike lesions. The first treated case dated September 28, 1900, and the archive contains the original material (clinical charts, imaging, paraffin blocks, and histological slides) of more than 47,000 cases (about 32,000 bone lesions and 15,000 soft tissue lesions).

Each single entity is presented multidisciplinary, with the pertinent clinical, radiological, and histological correlations. The treatment is briefly reported for each entity. Other separate chapters analyze the more recent biomolecular findings useful for diagnosis, prognosis, and treatment.

The text reflects the improvements in knowledge of musculoskeletal tumors as presented during the yearly international course held at the Rizzoli Institute.

This course, promoted by Prof. Mario Campanacci since 1970, has seen the participation as guest professors of the major international experts in musculoskeletal lesions:

D. Dahlin (Rochester) 1974, 1984

W.F. Enneking (Gainesville) 1984, 1989–1990, 1992–1994, 1998, 2004

N. Jaffe (Houston) 1984

D. Springfield (Gainesville, New York, Boston) 1995, 1997, 2000, 2002–2007

J.M. Mirra (Los Angeles) 1996, 2008

H. Mankin (Boston) 1999  
 A.L. Schiller (Boston) 2002  
 D. Vanel (Villejuif-Bologna) 2003, 2005, 2007–2018  
 P.C.W. Hogendoorn (Leiden) 2006–2008  
 N. Athanasou (Oxford) 2008  
 M.C. Gebhardt (Boston) 2008–2017  
 F.H. Sim (Rochester) 2010–2011  
 M.I. O'Connor (Jacksonville) 2011–2013  
 J.M. Coindre (Bordeaux) 2011  
 M.J. Klein (New York) 2012–2017  
 N. Fabbri (New York) 2013–2017  
 J.H. Healey (New York) 2013–2014  
 A.P. Dei Tos (Treviso) 2013–2019  
 S. Cammelli (Bologna University) 2016–2019  
 R. Grimer (Birmingham) 2017–2019  
 R. Windhager (Wien) 2018–2019

*Rizzoli collaborators to the Annual Course of the last years:*

Patrizia Bacchini	Pathology
Alberto Bazzocchi	Radiology
Maria Serena Benassi	Biology
Stefania Benini	Biology
Franco Bertoni	Pathology
Roberto Biagini	Orthopaedics
Giuseppe Bianchi	Orthopaedics
Stefano Boriani	Orthopaedics
Laura Campanacci	Orthopaedics
Roberto Casadei	Orthopaedics
Marco Colangeli	Orthopaedics
Massimiliano De Paolis	Orthopaedics
Davide Maria Donati	Orthopaedics
Costantino Errani	Orthopaedics
Nicola Fabbri	Orthopaedics
Stefano Ferrari	Oncology
Andrea Ferraro	Orthopaedics
Tommaso Frisoni	Orthopaedics
Marco Gambarotti	Pathology
Alessandro Gasbarrini	Orthopaedics
Claudia Hattinger	Biology
Marco Manfrini	Orthopaedics
Emanuela Palmerini	Oncology
Piero Picci	Oncology
Alberto Righi	Pathology
Eugenio Rimondi	Radiology
Pietro Ruggieri	Orthopaedics
Katia Scotlandi	Biology
Massimo Serra	Biology

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Eric Staals	Orthopaedics
Daniel Vanel	Radiology
Licciana Zanella	Biology

Bologna, Italy  
Bologna, Italy  
Bologna, Italy  
Bologna, Italy  
Bologna, Italy  
Padua, Italy

Piero Picci  
Marco Manfrini  
Davide Maria Donati  
Marco Gambarotti  
Alberto Righi  
Daniel Vanel  
Angelo Paolo Dei Tos



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## Preface of the First Book

The Rizzoli Orthopedic Institute has a proud and dynamic history, with several very famous orthopedic surgeons over the past 120 years. Dr. Mario Campanacci was especially concerned with the diagnosis and therapy of bone and soft tissue tumors, particularly the malignant bone tumors, two of which are very common and often lethal to children, namely Osteosarcoma and Ewing's sarcoma. Beginning in the early 1970s he pioneered the use of chemotherapy in Europe for these two tumors. He organized a team of surgeons and oncologists, who helped make an amazing reversal for most of these unfortunate patients dying of lung metastases within 2 years, to the majority being cured. The Rizzoli Orthopedic Institute has an independent Department dedicated to The Treatment of Malignant Bone Tumors and they now achieve, for osteosarcoma and Ewing's sarcoma an over 70–75% cure rate for their patients (by cure is meant the total eradication of the cancerous tumor).

Prior to the era of modern chemotherapy in the treatment of malignant tumors of bone, most patients died, even if they were diagnosed within days of arriving at a hospital, and even if an amputation was quickly applied as a desperate measure. The reason they died despite appropriate rapid diagnosis and local ablative surgery, is that about 90% of these patients already had microscopic lung metastases, impossible to see on their initial admission to the hospital by standard radiology studies. Months later, however, the tiny, microscopic seeds of metastases to the lungs would grow to grossly visible proportions, and within months, the patient would succumb to death by suffocation. Thanks to the Rizzoli and other Research Institutes throughout the world, not only are most patients with Ewing's and Osteosarcoma now cured, but as amazingly the majority can be cured without even the need for amputation. It is now possible to maintain their limb simply by removing the tumor area en-bloc, with a margin of uninvolved tissue, and using a prosthetic replacement, after a course of pre-operative chemotherapy. The Rizzoli Institute also stands at the cornerstone of these superb surgical advancements in prosthetic replacement techniques. As an effect of these outstanding results the Rizzoli Institute Bone Tumor Treatment Department is now, not only the premier treatment center for such tumors in Europe, treating some 80% of all such malignant tumor patients in Italy, but it may well be the largest and most renowned such treatment center in the world.

Another of the main goals of the Rizzoli has always been centered around the education of young doctors to become some of the finest Orthopedic Surgeons, Oncologists, Pathologists and Radiologists in the world with

respect to all aspects of Orthopedics, and especially for the specialty in the Bone Tumor field. Bone tumors are very rare, representing only about 1% of all benign and malignant tumors. It is extremely important that major bone tumor centers such as the Rizzoli exist, where patients afflicted by very rare tumors can be sent and where the physicians who will, or are directly caring for these patients obtain the necessary training and experience. A standard community hospital serving a local population of some 100,000 individuals will only see about 1 patient per year with a malignant bone tumor. In my opinion, to begin to understand how to accurately diagnose (with over 95% accuracy) and treat these tumors adequately requires a physician to have personally seen at least 500 such patients. In a general hospital that could take 500 physician years, well beyond the lifetime of any ordinary person I know. But at the Rizzoli it is possible for diagnosticians, treating physicians and student doctors to be involved with some 500 bone tumor patients in 2–3 years.

Which now brings us to the “Rizzoli Syllabus” which you now hold in your hands. This is a truly remarkable primer for students and even trained physicians to study the essentials of bone tumor diagnosis and treatment of virtually all of the benign and malignant tumor entities of bone. And for the first time in my experience, this syllabus also includes considerations of basic biology concepts of Giant Cell Tumor, Chondrosarcoma, Osteosarcoma and several other important basic science oriented topics. Many of these topics have been pioneered in the Rizzoli Research Institute under Dr. Piero Picci's direction. In addition, very important principals of Staging and Radiology have been added to The Syllabus, vital to an overall understanding of the treatment and diagnosis of tumors of the bone.

I have been involved with writing my own syllabi for The UCLA Orthopedic residents in years past, and I have seen a number of other syllabi over the years, but for a Bone Tumor Syllabus, this is by far the best I have ever seen published. It is informative, it is accurate, it is concise, and it is beautifully illustrated. The authors are to be highly commended for their efforts and dedication to teaching, from which a new generation of highly competent Bone Tumor Specialists will emerge.

Los Angeles, CA, USA

Joseph M. Mirra

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