

# Contents

<b>List of abbreviations</b>	<b>ix</b>		
<b>Foreword</b>	<b>x</b>		
<b>1 Soft tissue tumours</b>	<b>1</b>		
WHO classification of soft tissue tumours	2		
TNM staging of tumours of soft tissues	4		
TNM staging of gastrointestinal stromal tumours	5		
Introduction	6		
Adipocytic tumours			
Lipoma	13		
Lipomatosis	16		
Lipomatosis of nerve	18		
Lipoblastoma and lipoblastomatosis	20		
Angiolipoma	23		
Myolipoma of soft tissue	25		
Chondroid lipoma	27		
Spindle cell lipoma and pleomorphic lipoma	29		
Hibernoma	31		
Atypical spindle cell / pleomorphic lipomatous tumour	34		
Atypical lipomatous tumour / well-differentiated liposarcoma	36		
Dedifferentiated liposarcoma	39		
Myxoid liposarcoma	42		
Pleomorphic liposarcoma	45		
Myxoid pleomorphic liposarcoma	47		
Fibroblastic and myofibroblastic tumours			
Nodular fasciitis	49		
Proliferative fasciitis and proliferative myositis	51		
Myositis ossificans and fibro-osseous pseudotumour of digits	53		
Ischaemic fasciitis	55		
Elastofibroma	57		
Fibrous hamartoma of infancy	59		
Fibromatosis colli	61		
Juvenile hyaline fibromatosis	63		
Inclusion body fibromatosis	65		
Fibroma of tendon sheath	67		
Desmoplastic fibroblastoma	69		
Myofibroblastoma	71		
Calcifying aponeurotic fibroma	74		
<i>EWSR1-SMAD3</i> -positive fibroblastic tumour (emerging)	76		
Angiomyofibroblastoma	78		
Cellular angiofibroma	80		
Angiofibroma of soft tissue	82		
Nuchal-type fibroma	84		
Acral fibromyxoma	86		
Gardner fibroma	88		
Palmar fibromatosis and plantar fibromatosis	90		
Desmoid fibromatosis	93		
Lipofibromatosis	96		
Giant cell fibroblastoma	98		
Dermatofibrosarcoma protuberans	100		
Solitary fibrous tumour	104		
Inflammatory myofibroblastic tumour	109		
Low-grade myofibroblastic sarcoma	112		
Superficial CD34-positive fibroblastic tumour	114		
Myxoinflammatory fibroblastic sarcoma	116		
Infantile fibrosarcoma	119		
Adult fibrosarcoma	122		
Myxofibrosarcoma	124		
Low-grade fibromyxoid sarcoma	127		
		Sclerosing epithelioid fibrosarcoma	130
		So-called fibrohistiocytic tumours	
		Tenosynovial giant cell tumour	133
		Deep fibrous histiocytoma	137
		Plexiform fibrohistiocytic tumour	139
		Giant cell tumour of soft tissue	141
		Vascular tumours	
		Haemangiomas	
		Synovial haemangioma	143
		Intramuscular angioma	145
		Arteriovenous malformation/haemangioma	147
		Venous haemangioma	149
		Anastomosing haemangioma	150
		Epithelioid haemangioma	152
		Lymphangioma and lymphangiomatosis	154
		Tufted angioma and kaposiform haemangioendothelioma	156
		Retiform haemangioendothelioma	159
		Papillary intralymphatic angioendothelioma	161
		Composite haemangioendothelioma	163
		Kaposi sarcoma	166
		Pseudomyogenic haemangioendothelioma	169
		Epithelioid haemangioendothelioma	172
		Angiosarcoma	176
		Pericytic (perivascular) tumours	
		Glomus tumour	179
		Myopericytoma, including myofibroma	182
		Angioleiomyoma	186
		Smooth muscle tumours	
		Leiomyoma	188
		EBV-associated smooth muscle tumour	190
		Inflammatory leiomyosarcoma	193
		Leiomyosarcoma	195
		Skeletal muscle tumours	
		Rhabdomyoma	198
		Embryonal rhabdomyosarcoma	201
		Alveolar rhabdomyosarcoma	205
		Pleomorphic rhabdomyosarcoma	209
		Spindle cell / sclerosing rhabdomyosarcoma	211
		Ectomesenchymoma	214
		Gastrointestinal stromal tumour	216
		Chondro-osseous tumours	
		Soft tissue chondroma	222
		Extraskelatal osteosarcoma	224
		Peripheral nerve sheath tumours	
		Schwannoma	226
		Neurofibroma	232
		Perineurioma	237
		Granular cell tumour	240
		Dermal nerve sheath myxoma	243
		Solitary circumscribed neuroma	245
		Ectopic meningioma and meningotheial hamartoma	247
		Benign triton tumour / neuromuscular choristoma	249
		Hybrid nerve sheath tumour	252
		Malignant peripheral nerve sheath tumour	254
		Malignant melanotic nerve sheath tumour	258
		Tumours of uncertain differentiation	
		Intramuscular myxoma	261
		Juxta-articular myxoma	264
		Deep (aggressive) angiomyxoma	266
		Atypical fibroxanthoma	268
		Angiomatoid fibrous histiocytoma	271
		Ossifying fibromyxoid tumour	274

Myoepithelioma, myoepithelial carcinoma, and mixed tumour	277	Osteoclastic giant cell-rich tumours	
Pleomorphic hyalinizing angiectatic tumour of soft parts	280	Aneurysmal bone cyst	437
Haemosiderotic fibrolipomatous tumour	282	Giant cell tumour of bone	440
Phosphaturic mesenchymal tumour	284	Non-ossifying fibroma	447
NTRK-rearranged spindle cell neoplasm (emerging)	287	Notochordal tumours	
Synovial sarcoma	290	Benign notochordal cell tumour	449
Epithelioid sarcoma	294	Conventional chordoma	451
Alveolar soft part sarcoma	297	Dedifferentiated chordoma	454
Clear cell sarcoma of soft tissue	300	Poorly differentiated chordoma	456
Extraskeletal myxoid chondrosarcoma	303	Other mesenchymal tumours of bone	
Desmoplastic small round cell tumour	306	Chondromesenchymal hamartoma of chest wall	458
Extrarenal rhabdoid tumour	309	Osteofibrous dysplasia	460
PEComa	312	Adamantinoma of long bones	463
Intimal sarcoma	315	Simple bone cyst	467
Undifferentiated sarcoma	318	Fibrocartilaginous mesenchymoma	470
		Fibrous dysplasia	472
		Lipoma and hibernoma of bone	475
<b>2 Undifferentiated small round cell sarcomas of bone and soft tissue</b>	<b>321</b>	Leiomyosarcoma of bone	478
WHO classification of undifferentiated small round cell sarcomas of bone and soft tissue	322	Undifferentiated pleomorphic sarcoma	480
Ewing sarcoma	323	Bone metastases	483
Round cell sarcoma with <i>EWSR1</i> -non-ETS fusions	326	Haematopoietic neoplasms of bone	
<i>CIC</i> -rearranged sarcoma	330	Solitary plasmacytoma of bone	486
Sarcoma with <i>BCOR</i> genetic alterations	333	Primary non-Hodgkin lymphoma of bone	489
		Langerhans cell histiocytosis	492
		Erdheim-Chester disease	495
		Rosai-Dorfman disease	498
<b>3 Bone tumours</b>	<b>337</b>		
WHO classification of bone tumours	338	<b>4 Genetic tumour syndromes of soft tissue and bone</b>	<b>501</b>
TNM staging of tumours of bone	339	Introduction	502
Introduction	340	Enchondromatosis	506
Chondrogenic tumours		Li-Fraumeni syndrome	510
Subungual exostosis	345	McCune-Albright syndrome	514
Bizarre parosteal osteochondromatous proliferation	348	Multiple osteochondromas	517
Periosteal chondroma	351	Neurofibromatosis type 1	520
Enchondroma	353	Rothmund-Thomson syndrome	522
Osteochondroma	356	Werner syndrome	525
Chondroblastoma	359		
Chondromyxoid fibroma	362	<b>Contributors</b>	<b>528</b>
Osteochondromyxoma	365		
Synovial chondromatosis	368	<b>Declaration of interests</b>	<b>535</b>
Central atypical cartilaginous tumour / chondrosarcoma, grade 1	370	<b>IARC/WHO Committee for ICD-O</b>	<b>536</b>
Secondary peripheral atypical cartilaginous tumour / chondrosarcoma, grade 1	373	<b>Sources</b>	<b>537</b>
Central chondrosarcoma, grades 2 and 3	375		
Secondary peripheral chondrosarcoma, grades 2 and 3	379	<b>References</b>	<b>544</b>
Periosteal chondrosarcoma	381		
Clear cell chondrosarcoma	383	<b>Subject index</b>	<b>598</b>
Mesenchymal chondrosarcoma	385		
Dedifferentiated chondrosarcoma	388	<b>Previous volumes in the series</b>	<b>607</b>
Osteogenic tumours			
Osteoma	391		
Osteoid osteoma	394		
Osteoblastoma	397		
Low-grade central osteosarcoma	400		
Osteosarcoma	403		
Parosteal osteosarcoma	410		
Periosteal osteosarcoma	414		
High-grade surface osteosarcoma	417		
Secondary osteosarcoma	419		
Fibrogenic tumours			
Desmoplastic fibroma of bone	422		
Fibrosarcoma of bone	424		
Vascular tumours of bone			
Haemangioma of bone	426		
Epithelioid haemangioma of bone	428		
Epithelioid haemangioendothelioma of bone	431		
Angiosarcoma of bone	434		