

Contents

Please note that the contents of this volume have been split into two parts for publication in print: Part A and Part B.

List of abbreviations	xiii		
Foreword	xiv		
PART A			
ICD-O topographical coding of paediatric tumours	1		
ICD-O morphological coding: Introduction	5		
Haematolymphoid disorders	6		
CNS tumours	10		
Peripheral neuroblastic tumours	13		
Eye tumours	14		
Soft tissue and bone tumours	15		
Germ cell tumours	20		
Renal and male genital tumours	22		
Female genital tumours	23		
Breast tumours	24		
Digestive system tumours	25		
Endocrine tumours	27		
Head and neck tumours	28		
Thoracic tumours	29		
Skin tumours	30		
TNM staging of paediatric tumours	31		
1 Introduction to paediatric tumours	35		
2 Haematolymphoid disorders	39		
Introduction	40		
Myeloid neoplasms			
Myeloproliferative neoplasms			
Chronic myeloid leukaemia (CML)	41		
Myelodysplastic/myeloproliferative neoplasms			
Juvenile myelomonocytic leukaemia	45		
Myelodysplastic syndromes			
Refractory cytopenia of childhood (childhood myelodysplastic neoplasm with low blasts)	48		
Myelodysplastic syndrome with excess blasts (childhood myelodysplastic neoplasm with increased blasts)	51		
Acute myeloid leukaemia (AML)			
AMLs defined by differentiation	53		
AMLs with defining genetic abnormalities	56		
Myeloid neoplasms and proliferations associated with antecedent or predisposing conditions			
Myeloid neoplasms associated with germline predisposition	62		
Myeloid proliferations associated with Down syndrome	66		
Mast cell neoplasia			
Mastocytosis	69		
Lymphoid neoplasms			
Precursor lymphoid neoplasms			
B-lymphoblastic leukaemia/lymphoma	75		
T- and NK-lymphoblastic leukaemia/lymphoma	80		
Mature B-cell neoplasms			
Primary mediastinal large B-cell lymphoma	83		
Diffuse large B-cell lymphoma NOS	86		
EBV-positive diffuse large B-cell lymphoma	89		
Large B-cell lymphoma with <i>IRF4</i> rearrangement	91		
Paediatric-type follicular lymphoma	93		
		Paediatric nodal marginal zone lymphoma	96
		ALK-positive large B-cell lymphoma	99
		Lymphomatoid granulomatosis	101
		Plasmablastic lymphoma	103
		Mediastinal grey zone lymphoma	106
		Burkitt lymphoma	109
		High-grade B-cell lymphoma with 11q aberration	113
		Mature T/NK-cell neoplasms	
		Peripheral T-cell lymphoma NOS	115
		Aggressive NK-cell leukaemia	117
		Primary cutaneous T-cell neoplasms	
		Mycosis fungoides	119
		Primary cutaneous CD30-positive T-cell lymphoproliferative disorders	123
		Subcutaneous panniculitis-like T-cell lymphoma	126
		Anaplastic large cell lymphoma	
		ALK-positive anaplastic large cell lymphoma	129
		Hepatosplenic T-cell lymphoma	
		Hepatosplenic T-cell lymphoma	133
		EBV-positive lymphoproliferative diseases of childhood	
		Systemic EBV-positive T-cell lymphoma of childhood	135
		Hydroa vacciniforme lymphoproliferative disorder	138
		Hodgkin lymphoma	
		Classic Hodgkin lymphoma	141
		Nodular lymphocyte-predominant Hodgkin lymphoma	145
		Histiocytic and dendritic cell neoplasms	
		Langerhans cell histiocytosis and related disorders	148
		Immunodeficiency-associated lymphoproliferative disorders	
		Primary immunodeficiency-associated lymphoproliferative disorders	154
		Posttransplant lymphoproliferative disorders	158
		HIV-associated lymphoproliferative disorders	163
		3 CNS tumours	167
		Introduction	169
		Gliomas, glioneuronal tumours, and neuronal tumours	
		Introduction	172
		Paediatric-type diffuse low-grade gliomas	
		Diffuse astrocytoma, <i>MYB</i> - or <i>MYBL1</i> -altered	174
		Angiocentric glioma	177
		Polymorphous low-grade neuroepithelial tumour of the young	180
		Diffuse low-grade glioma, MAPK pathway-altered	183
		Paediatric-type diffuse high-grade gliomas	
		Diffuse midline glioma, H3 K27-altered	187
		Diffuse hemispheric glioma, H3 G34-mutant	193
		Diffuse paediatric-type high-grade glioma, H3-wildtype and IDH-wildtype	196
		Infant-type hemispheric glioma	200
		Circumscribed astrocytic gliomas	
		Pilocytic astrocytoma	202
		High-grade astrocytoma with piloid features	209
		Pleomorphic xanthoastrocytoma	213
		Subependymal giant cell astrocytoma	218
		Astroblastoma, <i>MN1</i> -altered	222
		Glioneuronal and neuronal tumours	
		Ganglioglioma	225
		Desmoplastic infantile ganglioglioma / desmoplastic infantile astrocytoma	230
		Dysembryoplastic neuroepithelial tumour	234

Diffuse glioneuronal tumour with oligodendro-		Phakomatous choristoma	370
glioma-like features and nuclear clusters	238	Melanocytic neoplasms	
Diffuse leptomeningeal glioneuronal tumour	241	Conjunctival junctional, compound, and subepi-	
Multinodular and vacuolating neuronal tumour	245	thelial naevi	372
Ependymal tumours		Inflamed juvenile conjunctival naevus	375
Introduction	248	Uveal neoplasms	
Supratentorial ependymoma	249	Diffuse choroidal neurofibroma and ganglioneuroma	378
Supratentorial ependymoma, <i>ZFTA</i> fusion-positive	252	Lisch nodule (iris hamartoma)	381
Supratentorial ependymoma, <i>YAP1</i> fusion-positive	255	Retinal and neuroepithelial tumours	
Posterior fossa ependymoma	257	Retinocytoma	383
Posterior fossa group A (PFA) ependymoma	260	Retinoblastoma	385
Posterior fossa group B (PFB) ependymoma	263	Medulloepithelioma	392
Spinal ependymoma	265	Optic nerve tumours	
Spinal ependymoma, <i>MYCN</i> -amplified	268	Pilocytic astrocytoma and other gliomas of the	
Myxopapillary ependymoma	271	optic nerve	395
Choroid plexus tumours		6 Soft tissue and bone tumours	397
Choroid plexus papilloma	274	Introduction	399
Atypical choroid plexus papilloma	277	Soft tissue tumours	
Choroid plexus carcinoma	279	Adipocytic tumours	
CNS embryonal tumours		Lipomatosis	402
Medulloblastoma: Introduction	282	Lipoblastoma/lipoblastomatosis	405
Medulloblastomas, molecularly defined		Liposarcoma	408
Medulloblastoma, WNT-activated	285	Fibroblastic and myofibroblastic tumours	
Medulloblastoma, SHH-activated and		Fasciitis/myositis	411
<i>TP53</i> -wildtype	288	Fibrodysplasia ossificans progressiva	415
Medulloblastoma, SHH-activated and		Fibroma of tendon sheath	417
<i>TP53</i> -mutant	291	Gardner fibroma	418
Medulloblastoma, non-WNT/non-SHH	293	Fibrous hamartoma of infancy	420
Medulloblastomas, histologically defined		Lipofibromatosis	422
Medulloblastoma, histologically defined	295	Inclusion body infantile digital fibromatosis	424
Other CNS embryonal tumours		Juvenile hyaline fibromatosis	
Introduction	302	(hyaline fibromatosis syndrome)	426
Atypical teratoid/rhabdoid tumour	303	Fibromatosis colli	428
Cribriform neuroepithelial tumour	308	Calcifying aponeurotic fibroma	430
Embryonal tumour with multilayered rosettes	310	Sinonasal angiofibroma	432
CNS neuroblastoma, <i>FOXO2</i> -activated	314	Plantar/palmar fibromatoses	435
CNS tumour with <i>BCOR</i> internal tandem duplication	317	Dermatofibrosarcoma protuberans	437
CNS embryonal tumour NEC/NOS	320	Desmoid fibromatosis	441
Pineal tumours		<i>EWSR1</i> :: <i>SMAD3</i> -rearranged fibroblastic tumour	443
Pineal tumours		Infantile fibrosarcoma	445
Introduction	322	Paediatric NTRK-rearranged spindle cell neoplasm	448
Pineoblastoma	323	Low-grade fibromyxoid sarcoma / sclerosing	
Melanocytic CNS tumours		epithelioid fibrosarcoma	451
Introduction	327	Inflammatory myofibroblastic tumour	454
Diffuse meningeal melanocytic neoplasms:		Low-grade myofibroblastic sarcoma	457
Melanocytosis and melanomatosis	328	So-called fibrohistiocytic tumours	
Tumours of the sellar region		Fibrous histiocytoma	459
Introduction	331	Plexiform fibrohistiocytic tumour	462
Pituitary endocrine tumours		Tenosynovial giant cell tumour	464
Pituitary adenoma / pituitary neuroendocrine tumour	332	Vascular tumours	
Pituitary blastoma	341	Capillary malformations	466
Craniopharyngiomas		Venous malformations	468
Adamantinomatous craniopharyngioma	343	Arteriovenous malformations	471
4 Peripheral neuroblastic tumours	349	Intramuscular vascular anomalies	474
Introduction	350	Lymphatic anomalies	477
Ganglioneuroma	351	Congenital haemangioma	479
Ganglioneuroblastoma, intermixed	353	Infantile haemangioma	481
Neuroblastoma	356	Haemangioma of placenta	483
Ganglioneuroblastoma, nodular (and other		Lobular capillary haemangioma (pyogenic	
composite neuroblastic tumours)	363	granuloma)	485
5 Eye tumours	365	Epithelioid haemangioma	487
Introduction	366	Tufted angioma and kaposiform haemangioen-	
Conjunctival neoplasms		dothelioma	489
Hamartomas		Papillary intralymphatic angioendothelioma and	
Epibulbar choristoma	367	retiform haemangioendothelioma	492
Epibulbar osseous choristoma	369	Pseudomyogenic haemangioendothelioma	494
		Kaposi sarcoma	497

Epithelioid haemangioendothelioma	500	Adamantinoma	640
Angiosarcoma	502	Osteofibrous dysplasia	642
Pericytic (perivascular) tumours		Fibrous dysplasia	644
Myofibroma and myopericytoma	504		
Glomus tumour and glomuvenous malformation	507	PART B	
Smooth muscle tumours		7 Germ cell tumours	647
EBV-associated smooth muscle tumour	509	Introduction	648
Skeletal muscle tumours		Germ cell tumours	
Rhabdomyoma	511	Non-invasive germ cell neoplasia	
Rhabdomyosarcomas	513	Intratubular germ cell neoplasia (male gonadal)	649
Ectomesenchymoma	517	Gonadoblastoma	652
Gastrointestinal stromal tumour		Germinoma-family tumours	
Paediatric gastrointestinal stromal tumour	519	Germinoma/dysgerminoma/seminoma	655
Peripheral nerve sheath tumours		Non-germinomatous germ cell tumours	
Schwannoma	522	Introduction	661
Neurofibroma	525	Mature cystic teratoma	663
Perineurioma	529	Extragenital teratoma	666
Hybrid nerve sheath tumour	531	Monodermal teratomas (female gonadal)	668
Granular cell tumour	533	gonadal) Immature teratoma (female gonadal)	671
Solitary circumscribed neuroma	535	Prepubertal-type testicular teratoma	673
Ectopic meningioma and meningotheelial hamartoma	537	Postpubertal-type teratoma	675
Benign triton tumour / neuromuscular choristoma	539	Embryonal carcinoma	679
Malignant peripheral nerve sheath tumour	541	Yolk sac tumour	681
Tumours of uncertain derivation		Fetus in fetu	684
Intramuscular/juxta-articular myxoma	545	Choriocarcinoma (non-gestational)	686
Superficial angiomyxoma	547	Malignant mixed germ cell tumours	688
Deep angiomyxoma	548		
Angiomatoid fibrous histiocytoma	550	8 Renal and male genital tumours	691
Clear cell sarcoma of soft tissue	552	Introduction	692
Alveolar soft part sarcoma	554	Renal tumours	
Extrarenal rhabdoid tumour	556	Nephroblastic and related tumours	
PEComa	559	Paediatric cystic nephroma	695
Synovial sarcoma	562	Nephroblastoma	697
Epithelioid sarcoma	565	Molecularly defined renal tumours	
Myoepithelial tumours of soft tissue	569	Renal cell carcinoma with MiT translocations	702
Phosphaturic mesenchymal tumour	572	ALK-rearranged renal cell carcinoma	706
Desmoplastic small round cell tumour	574	Eosinophilic solid and cystic renal cell carcinoma	708
Undifferentiated sarcomas	577	SMARCB1-deficient renal medullary carcinoma	710
Undifferentiated small round cell sarcomas of soft tissue and bone		Metanephric tumours	
Ewing sarcoma	579	Metanephric adenoma	712
Round cell sarcoma with <i>EWSR1</i> ::non-ETS fusions	582	Metanephric adenofibroma	714
<i>CIC</i> -rearranged sarcoma	585	Metanephric stromal tumour	716
Sarcoma with <i>BCOR</i> genetic alterations	587	Mesenchymal renal tumours	
Bone tumours		Ossifying renal tumour of infancy	718
Osteogenic tumours		Mesoblastic nephroma	720
Subungual exostosis	590	Clear cell sarcoma of the kidney	722
Bizarre parosteal osteochondromatous proliferation	592	Malignant rhabdoid tumour of the kidney	725
Osteoblastoma	594	Anaplastic sarcoma of the kidney	727
Osteoid osteoma	596	Ewing sarcoma	729
Chondromesenchymal hamartoma of the chest wall	598	Testicular tumours	
Osteosarcoma	600	Non-germ cell testicular tumours: Introduction	731
Chondrogenic tumours		Juvenile granulosa cell tumour of the testis	732
Chondroblastoma	608	9 Female genital tumours	735
Osteochondroma	611	Non-germ cell tumours of the female genital tract: Introduction	736
Chondromyxoid fibroma	613	Ovarian tumours	
Enchondroma and enchondromatosis	616	Sex cord stromal tumours	
Chondrosarcoma	618	Ovarian fibroma	737
Mesenchymal chondrosarcoma	623	Sclerosing stromal tumour	739
Other bone tumours		Juvenile granulosa cell tumour of the ovary	741
Vascular tumours of bone	625	Sex cord tumour with annular tubules	743
Aneurysmal bone cyst	628	Papillary cystadenoma	745
Giant cell tumour of bone	630	Sertoli-Leydig cell tumour	746
Non-ossifying fibroma	632	Gynandroblastoma	749
Notochordal tumours	634	Other ovarian tumours	
Simple bone cyst	638		

Small cell carcinoma of the ovary, hypercalcaemic type	751	13 Head and neck tumours	873
Lower female genital tract tumours		Introduction	874
Epithelial tumours		Benign tumours	
Mesonephric remnants and hyperplasia	753	Squamous papilloma and papillomatosis	875
Müllerian papilloma	755	White sponge naevus	877
Condyloma acuminatum	757	Congenital granular cell epulis	879
Peritoneal tumours		Central giant cell granuloma	881
Mesothelial tumours		Odontogenic tumours	883
Peritoneal inclusion cysts	759	Ossifying fibroma	888
10 Breast tumours	761	Sinonasal tract myxoma	890
Introduction	762	Nasal dermoid cyst	891
Breast tumours		Nasopharyngeal dermoid	893
Fibroepithelial tumours	764	Nasal chondromesenchymal hamartoma	894
Juvenile fibroadenoma	767	Pleomorphic adenoma	896
Juvenile papillomatosis	770	Malignant tumours	
11 Digestive system tumours	773	Mucoepidermoid carcinoma	899
Introduction	774	Acinic cell carcinoma	901
Liver tumours		Sialoblastoma	903
Epithelial tumours		Nasopharyngeal carcinoma	905
Hepatoblastoma	775	NUT carcinoma	908
Fibrolamellar variant of hepatocellular carcinoma	781	Melanotic neuroectodermal tumour of infancy	910
Paediatric hepatocellular carcinoma	784	14 Thoracic tumours	913
Mesenchymal tumours unique to the liver		Lung tumours	
Mesenchymal hamartoma	790	Fetal lung interstitial tumour	914
Calcifying nested stromal-epithelial tumour	793	Congenital peribronchial myofibroblastic tumour	916
Embryonal sarcoma of the liver	795	Pleuropulmonary blastoma	918
Hepatic congenital haemangioma Hepatic infantile haemangioma	797	Heart tumours	
Hepatic angiosarcoma	800	Cardiac rhabdomyoma	922
Pancreatic tumours	803	15 Skin tumours	925
Epithelial tumours		Introduction	926
Pancreatoblastoma	805	Hamartomas	
Pancreatic acinar cell carcinoma	809	Hamartomas	927
Solid pseudopapillary neoplasm	812	Epithelial neoplasms	
Gastrointestinal tumours		Squamous neoplasms	
Epithelial tumours		Angiokeratoma	932
Gastroblastoma	816	Epidermal naevi	934
Appendiceal neuroendocrine tumours	818	Pilomatricoma	938
12 Endocrine tumours	821	Melanocytic neoplasms	
Introduction	822	Naevi	
Thyroid tumours		Congenital naevi	940
Thyroid epithelial tumours		Junctional, compound, and dermal naevi Blue naevus	943
Follicular adenoma of the thyroid	823	Spitz naevus	947
Papillary thyroid carcinoma	827	Pigmented spindle cell naevus (Reed naevus)	950
Cribriform morular thyroid carcinoma	834	Melanoma	
Medullary thyroid carcinoma	837	Melanoma	952
Spindle epithelial tumour with thymus-like elements	841	16 Genetic tumour syndromes	957
Parathyroid tumours		Selected tumour predisposition syndromes: Introduction	958
Parathyroid endocrine tumours		Syndromes predisposing primarily to neural tumours	
Parathyroid adenoma	843	Neurofibromatosis type 1	962
Adrenal tumours		Neurofibromatosis type 2	966
Adrenocortical tumours		Tuberous sclerosis	971
Adrenal cortical tumours	846	Naevoid basal cell carcinoma syndrome (Gorlin syndrome)	975
Tumours of the adrenal medulla and extra-adrenal paraganglia		Retinoblastoma syndrome	979
Sympathetic paraganglioma	851	Syndromes predisposing primarily to endocrine tumours	
Parasympathetic paraganglioma (head and neck paraganglioma)	854	Von Hippel–Lindau syndrome	981
Pheochromocytoma	858	Hereditary pheochromocytoma–paraganglioma syndromes	984
Composite pheochromocytoma/paraganglioma	865	Syndromes predisposing primarily to Wilms tumours	
Neuroendocrine neoplasms		WAGR syndrome	987
Neuroendocrine tumours		Beckwith–Wiedemann and related overgrowth syndromes	989
Other neuroendocrine tumours	867		

Syndromes predisposing primarily to gastrointestinal tumours		Declaration of interests	1034
Familial adenomatous polyposis	991	IARC/WHO Committee for ICD-O	1036
Lynch syndrome	994	Sources	1037
Other syndromes		References	1055
Xeroderma pigmentosum	999	Subject index	1177
Rothmund–Thomson syndrome	1002	Previous volumes in the series	1191
Li–Fraumeni syndrome	1005		
<i>DICER1</i> syndrome	1008		
<i>BAP1</i> tumour predisposition syndrome	1012		
Constitutional mismatch repair deficiency syndrome	1014		
Rhabdoid tumour predisposition syndrome	1017		
Contributors	1021		