

# Contents

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

<b>List of abbreviations</b>	xiii	Paediatric nodal marginal zone lymphoma	96
<b>Foreword</b>	xiv	ALK-positive large B-cell lymphoma	99
<b>PART A</b>		Lymphomatoid granulomatosis	101
<b>ICD-O topographical coding of paediatric tumours</b>	1	Plasmablastic lymphoma	103
<b>ICD-O morphological coding: Introduction</b>	5	Mediastinal grey zone lymphoma	106
Haematolymphoid disorders	6	Burkitt lymphoma	109
CNS tumours	10	High-grade B-cell lymphoma with 11q aberration	113
Peripheral neuroblastic tumours	13	Mature T/NK-cell neoplasms	
Eye tumours	14	Peripheral T-cell lymphoma NOS	115
Soft tissue and bone tumours	15	Aggressive NK-cell leukaemia	117
Germ cell tumours	20	Primary cutaneous T-cell neoplasms	
Renal and male genital tumours	22	Mycosis fungoides	119
Female genital tumours	23	Primary cutaneous CD30-positive T-cell lymphoproliferative disorders	123
Breast tumours	24	Subcutaneous panniculitis-like T-cell lymphoma	126
Digestive system tumours	25	Anaplastic large cell lymphoma	
Endocrine tumours	27	ALK-positive anaplastic large cell lymphoma	129
Head and neck tumours	28	Hepatosplenic T-cell lymphoma	
Thoracic tumours	29	Hepatosplenic T-cell lymphoma	133
Skin tumours	30	EBV-positive lymphoproliferative diseases of childhood	
<b>TNM staging of paediatric tumours</b>	31	Systemic EBV-positive T-cell lymphoma of childhood	135
<b>1 Introduction to paediatric tumours</b>	35	Hydroa vacciniforme lymphoproliferative disorder	138
<b>2 Haematolymphoid disorders</b>	39	Hodgkin lymphoma	
Introduction	40	Classic Hodgkin lymphoma	141
Myeloid neoplasms		Nodular lymphocyte-predominant Hodgkin lymphoma	145
Myeloproliferative neoplasms		Histiocytic and dendritic cell neoplasms	
Chronic myeloid leukaemia (CML)	41	Langerhans cell histiocytosis and related disorders	148
Myelodysplastic/myeloproliferative neoplasms		Immunodeficiency-associated lymphoproliferative disorders	
Juvenile myelomonocytic leukaemia		Primary immunodeficiency-associated lymphoproliferative disorders	154
Myelodysplastic syndromes	45	Posttransplant lymphoproliferative disorders	158
Refractory cytopenia of childhood (childhood myelodysplastic neoplasm with low blasts)	48	HIV-associated lymphoproliferative disorders	163
Myelodysplastic syndrome with excess blasts (childhood myelodysplastic neoplasm with increased blasts)	51	<b>3 CNS tumours</b>	167
Acute myeloid leukaemia (AML)		Introduction	169
AMLS defined by differentiation	53	Gliomas, glioneuronal tumours, and neuronal tumours	
AMLS with defining genetic abnormalities	56	Introduction	172
Myeloid neoplasms and proliferations associated with antecedent or predisposing conditions		Paediatric-type diffuse low-grade gliomas	
Myeloid neoplasms associated with germline predisposition	62	Diffuse astrocytoma, MYB- or MYBL1-altered	174
Myeloid proliferations associated with Down syndrome	66	Angiocentric glioma	177
Mast cell neoplasia		Polymorphous low-grade neuroepithelial tumour of the young	180
Mastocytosis	69	Diffuse low-grade glioma, MAPK pathway-altered	183
Lymphoid neoplasms		Paediatric-type diffuse high-grade gliomas	
Precursor lymphoid neoplasms		Diffuse midline glioma, H3 K27-altered	187
B-lymphoblastic leukaemia/lymphoma	75	Diffuse hemispheric glioma, H3 G34-mutant	193
T- and NK-lymphoblastic leukaemia/lymphoma	80	Diffuse paediatric-type high-grade glioma, H3-wildtype and IDH-wildtype	196
Mature B-cell neoplasms		Infant-type hemispheric glioma	200
Primary mediastinal large B-cell lymphoma	83	Circumscribed astrocytic gliomas	
Diffuse large B-cell lymphoma NOS	86	Pilocytic astrocytoma	202
EBV-positive diffuse large B-cell lymphoma	89	High-grade astrocytoma with piloid features	209
Large B-cell lymphoma with <i>IRF4</i> rearrangement	91	Pleomorphic xanthoastrocytoma	213
Paediatric-type follicular lymphoma	93	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-glioma-like features and nuclear clusters	238	Phakomatous choristoma	370
Diffuse leptomeningeal glioneuronal tumour	241	Melanocytic neoplasms	
Multinodular and vacuolating neuronal tumour	245	Conjunctival junctional, compound, and subepithelial naevi	372
<b>Epidual tumours</b>		Inflamed juvenile conjunctival naevus	375
Introduction	248	<b>Uveal neoplasms</b>	
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	<b>Retinal and neuroepithelial tumours</b>	
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	<b>Optic nerve tumours</b>	
Spinal ependymoma, <i>MYCN</i> -amplified	268	Pilocytic astrocytoma and other gliomas of the optic nerve	395
Myxopapillary ependymoma	271		
<b>Choroid plexus tumours</b>			
Choroid plexus papilloma	274	<b>6 Soft tissue and bone tumours</b>	397
Atypical choroid plexus papilloma	277	Introduction	399
Choroid plexus carcinoma	279	Soft tissue tumours	
<b>CNS embryonal tumours</b>		Adipocytic tumours	
Medulloblastoma: Introduction	282	Lipomatosis	402
Medulloblastomas, molecularly defined		Lipoblastoma/lipoblastomatosis	405
Medulloblastoma, WNT-activated	285	Liposarcoma	408
Medulloblastoma, SHH-activated and		<b>Fibroblastic and myofibroblastic tumours</b>	
<i>TP53</i> -wildtype	288	Fasciitis/myositis	411
Medulloblastoma, SHH-activated and		Fibrodysplasia ossificans progressiva	415
<i>TP53</i> -mutant	291	Fibroma of tendon sheath	417
Medulloblastoma, non-WNT/non-SHH	293	Gardner fibroma	418
Medulloblastomas, histologically defined		Fibrous hamartoma of infancy	420
Medulloblastoma, histologically defined	295	Lipofibromatosis	422
Other CNS embryonal tumours		Inclusion body infantile digital fibromatosis	424
Introduction	302	Juvenile hyaline fibromatosis	
Atypical teratoid/rhabdoid tumour	303	(hyaline fibromatosis syndrome)	426
Cribiform neuroepithelial tumour	308	Fibromatosis colli	428
Embryonal tumour with multilayered rosettes	310	Calcifying aponeurotic fibroma	430
CNS neuroblastoma, <i>FOXR2</i> -activated	314	Sinonasal angiofibroma	432
CNS tumour with <i>BCOR</i> internal tandem duplication	317	Plantar/palmar fibromatoses	435
CNS embryonal tumour NEC/NOS	320	Dermatofibrosarcoma protuberans	437
<b>Pineal tumours</b>		Desmoid fibromatosis	441
Pineal tumours		<i>EWSR1::SMAD3</i> -rearranged fibroblastic tumour	443
Introduction	322	Infantile fibrosarcoma	445
Pineoblastoma	323	Paediatric NTRK-rearranged spindle cell neoplasm	448
<b>Melanocytic CNS tumours</b>		Low-grade fibromyxoid sarcoma / sclerosing epithelioid fibrosarcoma	451
Introduction	327	Inflammatory myofibroblastic tumour	454
Diffuse meningeal melanocytic neoplasms:		Low-grade myofibroblastic sarcoma	457
Melanocytosis and melanomatosis	328	<b>So-called fibrohistiocytic tumours</b>	
<b>Tumours of the sellar region</b>		Fibrous histiocytoma	459
Introduction	331	Plexiform fibrohistiocytic tumour	462
Pituitary endocrine tumours		Tenosynovial giant cell tumour	464
Pituitary adenoma / pituitary neuroendocrine tumour	332	<b>Vascular tumours</b>	
Pituitary blastoma	341	Capillary malformations	466
Cranioopharyngiomas		Venous malformations	468
Adamantinomatous craniopharyngioma	343	Arteriovenous malformations	471
<b>4 Peripheral neuroblastic tumours</b>	<b>349</b>	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 composite neuroblastic tumours)	363	Lobular capillary haemangioma (pyogenic granuloma)	485
<b>5 Eye tumours</b>	<b>365</b>	Epithelioid haemangioma	487
Introduction	366	Tufted angioma and kaposiform haemangioendothelioma	489
Conjunctival neoplasms		Papillary intralymphatic angioendothelioma and retiform haemangiomyxoma	492
Hamartomas		Pseudomyogenic haemangiomyxoma	494
Epibulbar choristoma	367	Kaposi sarcoma	497
Epibulbar osseous choristoma	369		

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

Small cell carcinoma of the ovary, hypercalcaemic type	751	<b>13 Head and neck tumours</b>	<b>873</b>
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
<b>10 Breast tumours</b>	<b>761</b>	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	
<b>11 Digestive system tumours</b>	<b>773</b>	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		
Mesenchymal tumours unique to the liver		<b>14 Thoracic tumours</b>	<b>913</b>
Mesenchymal hamartoma	790	Lung tumours	
Calcifying nested stromal-epithelial tumour	793	Fetal lung interstitial tumour	914
Embryonal sarcoma of the liver	795	Congenital peribranchial myofibroblastic tumour	916
Hepatic congenital haemangioma	797	Pleuropulmonary blastoma	918
Hepatic infantile haemangioma	800	Heart tumours	
Hepatic angiosarcoma	803	Cardiac rhabdomyoma	922
Pancreatic tumours			
Epithelial tumours		<b>15 Skin tumours</b>	<b>925</b>
Pancreatoblastoma	805	Introduction	926
Pancreatic acinar cell carcinoma	809	Hamartomas	
Solid pseudopapillary neoplasm	812	Hamartomas	927
Gastrointestinal tumours		Epithelial neoplasms	
Epithelial tumours		Squamous neoplasms	
Gastroblastoma	816	Angiokeratoma	932
Appendiceal neuroendocrine tumours	818	Epidermal naevi	934
<b>12 Endocrine tumours</b>	<b>821</b>	Pilomatrixoma	938
Introduction	822	Melanocytic neoplasms	
Thyroid tumours		Naevi	
Thyroid epithelial tumours		Congenital naevi	940
Follicular adenoma of the thyroid	823	Junctional, compound, and dermal naevi	943
Papillary thyroid carcinoma	827	Blue naevus	945
Cribiform morular thyroid carcinoma	834	Spitz naevus	947
Medullary thyroid carcinoma	837	Pigmented spindle cell naevus (Reed naevus)	950
Spindle epithelial tumour with thymus-like elements	841	Melanoma	
Parathyroid tumours		Melanoma	952
Parathyroid endocrine tumours			
Parathyroid adenoma	843	<b>16 Genetic tumour syndromes</b>	<b>957</b>
Adrenal tumours		Selected tumour predisposition syndromes: Introduction	958
Adrenocortical tumours		Syndromes predisposing primarily to neural tumours	
Adrenal cortical tumours	846	Neurofibromatosis type 1	962
Tumours of the adrenal medulla and extra-adrenal paraganglia		Neurofibromatosis type 2	966
Sympathetic paraganglioma	851	Tuberous sclerosis	971
Parasympathetic paraganglioma (head and		Naevi basal cell carcinoma syndrome (Gorlin	
neck paraganglioma)	854	syndrome)	975
Phaeochromocytoma	858	Retinoblastoma syndrome	979
Composite phaeochromocytoma/paraganglioma	865	Syndromes predisposing primarily to endocrine tumours	
Neuroendocrine neoplasms		Von Hippel–Lindau syndrome	981
Neuroendocrine tumours		Hereditary phaeochromocytoma–paraganglioma	
Other neuroendocrine tumours	867	syndromes	984
		Syndromes predisposing primarily to Wilms tumours	
		WAGR syndrome	987
		Beckwith–Wiedemann and related overgrowth	
		syndromes	989

Syndromes predisposing primarily to gastrointestinal tumours		<b>Declaration of interests</b>	1034
Familial adenomatous polyposis	991	IARC/WHO Committee for ICD-O	1036
Lynch syndrome	994		
Other syndromes		<b>Sources</b>	1037
Xeroderma pigmentosum	999	<b>References</b>	1055
Rothmund–Thomson syndrome	1002	<b>Subject index</b>	1177
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	<b>Previous volumes in the series</b>	1191
<b>Contributors</b>	<b>1021</b>		