

# Contents

|                                                                          |           |                                                                               |            |
|--------------------------------------------------------------------------|-----------|-------------------------------------------------------------------------------|------------|
| <b>List of abbreviations</b>                                             | <b>x</b>  | Sellar neurocytoma                                                            | 100        |
| <b>Foreword</b>                                                          | <b>xi</b> | Other sellar tumours                                                          |            |
| <b>ICD-O topographical coding</b>                                        | <b>1</b>  | Meningiomas                                                                   | 103        |
| <b>ICD-O morphological coding</b>                                        | <b>4</b>  | Chordoma                                                                      | 105        |
| Pituitary tumours                                                        | 5         | <b>3 Thyroid tumours</b>                                                      |            |
| Thyroid tumours                                                          | 7         | Introduction                                                                  | 108        |
| Parathyroid tumours                                                      | 9         | Developmental abnormalities                                                   |            |
| Adrenal gland tumours                                                    | 10        | Thyroglossal duct cyst                                                        | 113        |
| Adrenal medulla and extra-adrenal paraganglia tumours                    | 11        | Other congenital thyroid abnormalities                                        | 116        |
| Pancreatic neuroendocrine neoplasms                                      | 12        | Follicular cell-derived neoplasms                                             |            |
| Neuroendocrine neoplasms in non-endocrine organs                         | 13        | Benign tumours                                                                |            |
| Mesenchymal and stromal tumours                                          | 14        | Thyroid follicular nodular disease                                            | 119        |
| Haematolymphoid tumours                                                  | 16        | Follicular thyroid adenoma                                                    | 122        |
| Germ cell tumours                                                        | 17        | Follicular thyroid adenoma with papillary architecture                        | 126        |
| <b>TNM staging of endocrine and neuroendocrine tumours</b>               | <b>18</b> | Oncocytic adenoma of the thyroid gland                                        | 129        |
| Carcinomas of the thyroid gland                                          | 18        | Low-risk neoplasms                                                            |            |
| Carcinomas of the adrenal cortex                                         | 20        | Non-invasive follicular thyroid neoplasm with papillary-like nuclear features | 132        |
| Tumours of the pancreas                                                  | 21        | Thyroid tumours of uncertain malignant potential                              | 135        |
| Well-differentiated neuroendocrine tumours of the gastrointestinal tract | 22        | Hyalinizing trabecular tumour                                                 | 138        |
| <b>1 Introduction to endocrine and neuroendocrine tumours</b>            | <b>28</b> | Malignant neoplasms                                                           |            |
| <b>2 Pituitary tumours</b>                                               | <b>33</b> | Follicular thyroid carcinoma                                                  | 142        |
| Introduction                                                             | 34        | Invasive encapsulated follicular variant of papillary thyroid carcinoma       | 147        |
| Anterior pituitary neuroendocrine tumours (PitNETs)                      |           | Papillary thyroid carcinoma                                                   | 150        |
| PitNETs of PIT1 lineage                                                  |           | Oncocytic carcinoma of the thyroid gland                                      | 161        |
| Somatotroph PitNET/adenoma                                               | 40        | High-grade follicular cell-derived non-anaplastic thyroid carcinoma           | 165        |
| Mammotroph PitNET/adenoma                                                | 45        | Anaplastic thyroid carcinoma                                                  | 170        |
| Lactotroph PitNET/adenoma                                                | 48        | Thyroid C cell-derived carcinoma                                              |            |
| Thyrotroph PitNET/adenoma                                                | 53        | Medullary thyroid carcinoma                                                   | 174        |
| Mature plurihormonal PIT1-lineage PitNET/adenoma                         | 55        | Mixed medullary and follicular cell-derived carcinomas                        |            |
| Immature PIT1-lineage PitNET/adenoma                                     | 57        | Mixed medullary and follicular cell-derived thyroid carcinoma                 | 179        |
| Acidophil stem cell PitNET/adenoma                                       | 59        | Salivary gland-type carcinomas of the thyroid gland                           |            |
| Mixed somatotroph–lactotroph PitNET/adenoma                              | 62        | Mucoepidermoid carcinoma                                                      | 182        |
| PitNETs of TPIT lineage                                                  |           | Secretory carcinoma                                                           | 184        |
| Corticotroph PitNET/adenoma                                              | 65        | Thyroid tumours of uncertain histogenesis                                     |            |
| PitNETs of SF1 lineage                                                   |           | Sclerosing mucoepidermoid carcinoma with eosinophilia                         | 186        |
| Gonadotroph PitNET/adenoma                                               | 70        | Cribiform morular thyroid carcinoma                                           | 189        |
| PitNETs without distinct lineage differentiation                         |           | Thymic tumours within the thyroid                                             |            |
| Null cell PitNET/adenoma                                                 | 73        | Thymoma                                                                       | 192        |
| Plurihormonal PitNET/adenoma                                             | 75        | Spindle epithelial tumour with thymus-like elements                           | 194        |
| Multiple PitNETs                                                         |           | Intrathyroidal thymic carcinoma                                               | 196        |
| Multiple synchronous PitNETs/adenomas of distinct lineages               | 77        | Embryonal thyroid neoplasms                                                   |            |
| Metastatic PitNETs                                                       |           | Thyroblastoma                                                                 | 199        |
| Metastatic PitNET                                                        | 79        | <b>4 Parathyroid tumours</b>                                                  | <b>201</b> |
| Other anterior pituitary tumours                                         |           | Introduction                                                                  | 202        |
| Adamantinomatous craniopharyngioma                                       | 82        | Parathyroid hyperplasia                                                       | 204        |
| Papillary craniopharyngioma                                              | 86        | Parathyroid adenoma                                                           | 207        |
| Pituitary blastoma                                                       | 89        | Parathyroid lipoadenoma                                                       | 213        |
| Posterior pituitary and hypothalamic neoplasms                           |           | Atypical parathyroid tumour                                                   | 215        |
| Pituicyte-derived tumours                                                |           | Parathyroid carcinoma                                                         | 218        |
| Pituicytomas                                                             | 92        | <b>5 Adrenal gland tumours</b>                                                | <b>223</b> |
| Neuronal tumours                                                         |           | Introduction                                                                  | 224        |
| Gangliocytoma and mixed gangliocytoma–PitNET/adenoma                     | 97        | Lesions in the adrenal cortex                                                 |            |
|                                                                          |           | Adrenal rests                                                                 | 226        |

|                                                                |            |                                                              |            |
|----------------------------------------------------------------|------------|--------------------------------------------------------------|------------|
| Adrenal cysts                                                  | 228        | <b>9 Mesenchymal and stromal tumours</b>                     | <b>357</b> |
| Adrenal myelolipoma                                            | 230        | Introduction                                                 | 358        |
| Adrenal cortical tumours                                       |            | Lipocytic tumours                                            |            |
| Congenital adrenal hyperplasia                                 | 232        | Lipoma                                                       | 359        |
| Adrenocortical nodular disease                                 | 236        | Fibrous tumours                                              |            |
| Adrenal cortical adenoma                                       | 240        | Solitary fibrous tumour                                      | 360        |
| Adrenal cortical carcinoma                                     | 246        | Vascular tumours                                             |            |
| Sex cord stromal and other tumours                             |            | Benign vascular tumours                                      | 362        |
| Adrenal sex cord stromal tumour                                | 254        | PEComa                                                       | 364        |
| Adenomatoid tumour                                             | 256        | Glomangioma and glomus tumour                                | 366        |
| Adrenal melanoma                                               | 258        | Angiosarcoma                                                 | 368        |
| <b>6 Adrenal medulla and extra-adrenal paraganglia tumours</b> | <b>261</b> | Smooth muscle tumours                                        |            |
| Introduction                                                   | 262        | Leiomyoma                                                    | 371        |
| Neuroblastic tumours                                           |            | Leiomyosarcoma                                               | 373        |
| Neuroblastoma                                                  | 265        | Skeletal muscle tumours                                      |            |
| Ganglioneuroblastoma, intermixed                               | 271        | Rhabdomyosarcomas                                            | 376        |
| Ganglioneuroblastoma, nodular                                  | 273        | Peripheral nerve sheath tumours                              |            |
| Ganglioneuroma                                                 | 275        | Schwannoma                                                   | 379        |
| Paraganglioma and pheochromocytoma                             |            | Neurofibroma                                                 | 381        |
| Pheochromocytoma                                               | 277        | Granular cell tumour                                         | 383        |
| Sympathetic paraganglioma                                      | 285        | Malignant peripheral nerve sheath tumour                     | 385        |
| Parasympathetic paraganglioma                                  | 288        | <b>10 Haematolymphoid tumours</b>                            | <b>387</b> |
| Composite paraganglion tumours                                 |            | Introduction                                                 | 388        |
| Composite pheochromocytomas and                                |            | IgG4-related disease                                         | 389        |
| paragangliomas                                                 | 292        | Extranodal marginal zone lymphoma of                         |            |
| <b>7 Pancreatic neuroendocrine neoplasms</b>                   | <b>295</b> | mucosa-associated lymphoid tissue                            | 392        |
| Introduction                                                   | 296        | Diffuse large B-cell lymphoma NOS                            | 395        |
| Neuroendocrine neoplasms                                       |            | Langerhans cell histiocytosis                                | 397        |
| Neuroendocrine tumours                                         |            | Rosai–Dorfman disease                                        | 400        |
| Non-functioning (non-syndromic) pancreatic                     |            | Erdheim–Chester disease                                      | 402        |
| neuroendocrine tumours                                         | 300        | Follicular dendritic cell sarcoma                            | 404        |
| Insulinoma                                                     | 305        | <b>11 Germ cell tumours</b>                                  | <b>407</b> |
| Glucagonoma                                                    | 309        | Introduction                                                 | 408        |
| Somatostatinoma                                                | 312        | Germ cell tumours                                            |            |
| Gastrinoma                                                     | 315        | Germ cell tumours in endocrine organs                        | 409        |
| VIPoma                                                         | 317        | Teratomas with endocrine differentiation                     | 412        |
| Serotonin-producing tumours                                    |            | <b>12 Metastasis</b>                                         | <b>415</b> |
| with and without carcinoid syndrome                            | 320        | Introduction                                                 | 416        |
| Other pancreatic neuroendocrine tumours with                   |            | Metastasis to the pituitary gland                            | 417        |
| ectopic hormone production                                     | 322        | Metastasis to the thyroid gland                              | 419        |
| Neuroendocrine carcinomas                                      |            | Metastasis to the adrenal gland                              | 422        |
| Pancreatic neuroendocrine carcinoma                            | 325        | <b>13 Genetic tumour syndromes associated with endocrine</b> | <b>425</b> |
| Mixed neuroendocrine–non-neuroendocrine neoplasms              |            | lesions and tumours                                          |            |
| Mixed ductal adenocarcinoma–neuroendocrine                     |            | Introduction                                                 | 426        |
| neoplasms                                                      | 328        | Multiple endocrine neoplasia type 1                          | 429        |
| Mixed acinar–neuroendocrine carcinoma                          | 330        | Multiple endocrine neoplasia type 2                          | 435        |
| <b>8 Neuroendocrine neoplasms in non-endocrine organs</b>      | <b>333</b> | Multiple endocrine neoplasia type 4                          | 440        |
| Introduction                                                   | 334        | Multiple endocrine neoplasia type 5 (MAX-related             |            |
| Neuroendocrine neoplasms                                       |            | tumours)                                                     | 444        |
| Neuroendocrine tumours                                         |            | Hyperparathyroidism–jaw tumour syndrome                      | 448        |
| Neuroendocrine tumours                                         | 337        | Von Hippel–Lindau syndrome                                   | 452        |
| Neuroendocrine carcinomas                                      |            | SDH-deficient tumour syndromes                               | 457        |
| Small cell neuroendocrine carcinoma                            | 339        | Neurofibromatosis type 1                                     | 463        |
| Large cell neuroendocrine carcinoma                            | 342        | Carney complex                                               | 466        |
| Merkel cell carcinoma                                          | 346        | McCune–Albright syndrome                                     | 471        |
| Mixed neuroendocrine neoplasms                                 |            | DICER1 syndrome                                              | 474        |
| Mixed neuroendocrine–non-neuroendocrine                        |            | Glucagon cell hyperplasia and neoplasia                      | 478        |
| neoplasms                                                      | 348        | MAFA-related familial insulinomatosis                        | 480        |
| Paraganglioma-like neuroendocrine neoplasms                    |            | Syndromic familial follicular cell–derived thyroid           |            |
| Composite gangliocytoma/neuroma and                            |            | tumours                                                      | 482        |
| neuroendocrine tumour                                          | 351        | Non-syndromic familial follicular cell–derived               |            |
| Cauda equina neuroendocrine tumour (previously                 |            | thyroid tumours                                              | 488        |
| paraganglioma)                                                 | 354        |                                                              |            |

|                                |     |
|--------------------------------|-----|
| Contributors                   | 493 |
| Declaration of interests       | 499 |
| IARC/WHO Committee for ICD-O   | 500 |
| Sources                        | 501 |
| References                     | 507 |
| Subject index                  | 587 |
| Previous volumes in the series | 595 |