Central Nervous System Tumours/ Forewords and Introductions/ ICD-O coding of CNS tumours

Headings

ICD-O topographical coding of central nervous system tumours

The ICD-O topography codes for the main anatomical sites covered in this volume are as follows [[Fritz A, Percy C, Jack A, et al., editors. International classification of diseases for oncology (ICD-O). 3rd ed. 1st rev. Geneva (Switzerland): World Health Organization; 2013.]]:

C70 Meninges

- C70.0 Cerebral meninges
- C70.1 Spinal meninges
- C70.9 Meninges, NOS

C71 Brain

- C71.0 Cerebrum
- C71.1 Frontal lobe
- C71.2 Temporal lobe
- C71.3 Parietal lobe
- C71.4 Occipital lobe
- C71.5 Ventricle, NOS
- C71.6 Cerebellum, NOS
- C71.7 Brain stem
- C71.8 Overlapping lesion of brain
- C71.9 Brain, NOS

C72 Spinal cord, cranial nerves, and other parts of central nervous system

- C72.0 Spinal cord
- C72.1 Cauda equina
- C72.2 Olfactory nerve
- C72.3 Optic nerve
- C72.4 Acoustic nerve
- C72.5 Cranial nerve, NOS
- C72.8 Overlapping lesion of brain and central nervous system
- C72.9 Nervous system, NOS

C75 Other endocrine glands and related structures

C75.1 Pituitary gland

C75.2 Craniopharyngeal duct

C75.3 Pineal gland

ICD-O morphological coding: Introduction

The ICD-O coding system uses a topography (T) code and a morphology (M) code together, but these are presented in separate lists for ease of use. Behaviour is coded /0 for benign tumours; /1 for unspecified, borderline, or uncertain behaviour; /2 for carcinoma in situ and grade III intraepithelial neoplasia; /3 for malignant tumours, primary site; and /6 for malignant tumours, metastatic site. Behaviour code /6 is not generally used by cancer registries. For various reasons, the ICD-O morphology terms may not always be identical to the entity names used in the WHO classification, but they should be sufficiently similar to avoid confusion. The designation “NOS” (“not otherwise specified”) is provided to make coding possible when subtypes exist but exact classification may not be possible in small biopsies or certain other scenarios. Therefore, it is usual to have “NOS” even when a more specific alternative term is listed in ICD-O.

ICD-O coding of central nervous system tumours

ICD-O-3.2 ICD-O label (subtypes are indicated in grey text, with the label indented);

Please note that the WHO classification of tumour types is more readily reflected in the table of contents

Gliomas, glioneuronal tumours, and neuronal tumours

Adult-type diffuse gliomas

Astrocytoma, IDH-mutant

9400/3 Astrocytoma, IDH-mutant, grade 2

9401/3 Astrocytoma, IDH-mutant, grade 3

9445/3 Astrocytoma, IDH-mutant, grade 4

Oligodendroglioma, IDH-mutant and 1p/19q-codeleted

9450/3 Oligodendroglioma, IDH-mutant and 1p/19q-codeleted, grade 2

9451/3 Oligodendroglioma, IDH-mutant and 1p/19q-codeleted, grade 3

9440/3 Glioblastoma, IDH-wildtype

Paediatric-type diffuse low-grade gliomas

9421/1 Diffuse astrocytoma, *MYB*- or *MYBL1*-altered†

9431/1 Angiocentric glioma

9413/0 Polymorphous low-grade neuroepithelial tumour of the young†

9421/1 Diffuse low-grade glioma, MAPK pathway–altered†

Paediatric-type diffuse high-grade gliomas

9385/3 Diffuse midline glioma, H3 K27–altered†

9385/3 Diffuse hemispheric glioma, H3 G34–mutant†

9385/3 Diffuse paediatric-type high-grade glioma, H3-wildtype and IDH-wildtype†

9385/3 Infant-type hemispheric glioma†

Circumscribed astrocytic gliomas

9421/1 Pilocytic astrocytoma

9421/3* High-grade astrocytoma with piloid features

9424/3 Pleomorphic xanthoastrocytoma

9384/1 Subependymal giant cell astrocytoma

9444/1 Chordoid glioma

9430/3 Astroblastoma, *MN1*-altered†

Glioneuronal and neuronal tumours

9505/1 Ganglioglioma

9492/0 Gangliocytoma

9412/1 Desmoplastic infantile ganglioglioma

9412/1 Desmoplastic infantile astrocytoma

9413/0 Dysembryoplastic neuroepithelial tumour

n/a Diffuse glioneuronal tumour with oligodendroglioma-like features and nuclear clusters (provisional entity)

9509/1 Papillary glioneuronal tumour

9509/1 Rosette-forming glioneuronal tumour

9509/1 Myxoid glioneuronal tumour†

9509/3* Diffuse leptomeningeal glioneuronal tumour

9509/0* Multinodular and vacuolating neuronal tumour

9493/0 Dysplastic cerebellar gangliocytoma (Lhermitte–Duclos disease)

9506/1 Central neurocytoma

9506/1 Extraventricular neurocytoma

9506/1 Cerebellar liponeurocytoma

Ependymal tumours

9391/3 Supratentorial ependymoma, NOS†

9396/3 Supratentorial ependymoma, *ZFTA* fusion–positive†

9396/3 Supratentorial ependymoma, *YAP1* fusion–positive†

9391/3 Posterior fossa ependymoma, NOS†

9396/3 Posterior fossa group A (PFA) ependymoma†

9396/3 Posterior fossa group B (PFB) ependymoma†

9391/3 Spinal ependymoma, NOS†

9396/3 Spinal ependymoma, *MYCN*-amplified†

9394/1 Myxopapillary ependymoma

9383/1 Subependymoma

Choroid plexus tumours

9390/0 Choroid plexus papilloma

9390/1 Atypical choroid plexus papilloma

9390/3 Choroid plexus carcinoma

Embryonal tumours

Medulloblastomas, molecularly defined

9475/3 Medulloblastoma, WNT-activated

9471/3 Medulloblastoma, SHH-activated and *TP53*-wildtype

9476/3 Medulloblastoma, SHH-activated and *TP53*-mutant

9477/3 Medulloblastoma, non-WNT/non-SHH

Medulloblastomas, histologically defined

9470/3 Medulloblastoma, histologically defined

9471/3 Desmoplastic nodular medulloblastoma

9471/3 Medulloblastoma with extensive nodularity

9474/3 Large cell medulloblastoma

9474/3 Anaplastic medulloblastoma

Other CNS embryonal tumours

9508/3 Atypical teratoid/rhabdoid tumour

n/a Cribriform neuroepithelial tumour (provisional entity)

9478/3 Embryonal tumour with multilayered rosettes

9500/3 CNS neuroblastoma, *FOXR2*-activated†

9500/3 CNS tumour with *BCOR* internal tandem duplication†

9473/3 CNS embryonal tumour, NEC/NOS

Pineal tumours

9361/1 Pineocytoma

9362/3 Pineal parenchymal tumour of intermediate differentiation

9362/3 Pineoblastoma

9395/3 Papillary tumour of the pineal region

n/a Desmoplastic myxoid tumour of the pineal region, *SMARCB1*-mutant (provisional entity)

Cranial and paraspinal nerve tumours

9560/0 Schwannoma

9540/0 Neurofibroma

9550/0 Plexiform neurofibroma

9571/0 Perineurioma

9563/0 Hybrid nerve sheath tumour

9540/3 Malignant melanotic nerve sheath tumour

9540/3 Malignant peripheral nerve sheath tumour

8693/3 Cauda equina neuroendocrine tumour (previously paraganglioma)

Meningioma

9530/0 Meningioma

Mesenchymal, non-meningothelial tumours involving the CNS

Fibroblastic and myofibroblastic tumours

8815/1 Solitary fibrous tumour

Vascular tumours

9121/0 Cavernous haemangioma

9131/0 Capillary haemangioma

9123/0 Arteriovenous malformation

9161/1 Haemangioblastoma

Skeletal muscle tumours

8910/3 Embryonal rhabdomyosarcoma

8920/3 Alveolar rhabdomyosarcoma

8901/3 Rhabdomyosarcoma, pleomorphic-type

8912/3 Spindle cell rhabdomyosarcoma

Tumours of uncertain differentiation

n/a Intracranial mesenchymal tumour, FET::CREB fusion–positive (provisional entity)

9367/3 *CIC*-rearranged sarcoma

9480/3 Primary intracranial sarcoma, *DICER1*-mutant†

9364/3 Ewing sarcoma

Chondrogenic tumours

9240/3 Mesenchymal chondrosarcoma

9220/3 Chondrosarcoma

9243/3 Dedifferentiated chondrosarcoma

Notochordal tumours

9370/3 Chordoma

Melanocytic tumours

Diffuse meningeal melanocytic neoplasms

8728/0 Meningeal melanocytosis

8728/3 Meningeal melanomatosis

Circumscribed meningeal melanocytic neoplasms

8728/1 Meningeal melanocytoma

8720/3 Meningeal melanoma

Haematolymphoid tumours involving the CNS

CNS lymphomas

9680/3 Primary diffuse large B-cell lymphoma of the CNS

9766/1 Lymphomatoid granulomatosis

9766/1 Lymphomatoid granulomatosis, grade 1

9766/1 Lymphomatoid granulomatosis, grade 2

9766/3 Lymphomatoid granulomatosis, grade 3

9712/3 Intravascular large B-cell lymphoma

Miscellaneous rare lymphomas in the CNS

9699/3 MALT lymphoma of the dura

9671/3 Lymphoplasmacytic lymphoma

9690/3 Follicular lymphoma

9714/3 Anaplastic large cell lymphoma (ALK+/ALK–)

9702/3 T-cell lymphoma

9719/3 NK/T-cell lymphoma

Histiocytic tumours

9749/3 Erdheim–Chester disease

9749/3 Rosai–Dorfman disease†

9749/1 Juvenile xanthogranuloma†

9751/1	Langerhans cell histiocytosis
9755/3	Histiocytic sarcoma

Germ cell tumours

9080/0	Mature teratoma
9080/3	Immature teratoma
9084/3	Teratoma with somatic-type malignancy
9064/3	Germinoma
9070/3	Embryonal carcinoma
9071/3	Yolk sac tumour
9100/3	Choriocarcinoma
9085/3	Mixed germ cell tumour

Tumours of the sellar region

9351/1	Adamantinomatous craniopharyngioma
9352/1	Papillary craniopharyngioma
9432/1	Pituicytoma
9582/0	Granular cell tumour of the sellar region
8290/0	Spindle cell oncocytoma
8272/3	Pituitary adenoma / pituitary neuroendocrine tumour (PitNET)†
8273/3	Pituitary blastoma

These morphology codes are from the International Classification of Diseases for Oncology, third edition, second revision (ICD-O-3.2) [[International Association of Cancer Registries (IACR) [Internet]. Lyon (France): International Agency for Research on Cancer; 2021. International Classification of Diseases for Oncology (ICD-O) – ICD-O-3.2; updated 2021 Jan 25. Available from: http://www.iacr.com.fr/index.php?option=com_content&view=category&layout=blog&id=100&Itemid=577.]]. Behaviour is coded /0 for benign tumours; /1 for unspecified, borderline, or uncertain behaviour; /2 for carcinoma in situ and grade III intraepithelial neoplasia; /3 for malignant tumours, primary site; and /6 for malignant tumours, metastatic site. Behaviour code /6 is not generally used by cancer registries.

This classification is modified from the previous WHO classification, taking into account changes in our understanding of these lesions.

n/a, not available (provisional entity).

* Codes marked with an asterisk were approved by the IARC/WHO Committee for ICD-O at its meeting in May 2021.

† Labels marked with a dagger have undergone a change in terminology of a previous code.