**Recommendations for coding tumours of the central nervous system (CNS)**

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# Background

While for most solid tumours the behaviour (benign, borderline, in situ, malignant) of the tumour can be used to differentiate between malignant (‘cancer’) and non-malignant, this is not the case for neoplasms of the central nervous system (CNS), as the clinical behaviour of a CNS tumour is not in accordance with the pathological behaviour. Therefore, as of the 11th edition of the International Classification of Diseases (ICD11), all neoplasms of the central nervous system are grouped in the same category, irrespective of the behaviour of the tumour.

In 1998, the first ENCR recommendations for coding of tumours of the CNS were published. For the first time, it was recommended to European cancer registries to include all CNS tumours irrespective of the behaviour. Since 1998, the classification of CNS tumours has evolved, taking into account many developments in the diagnostics, including molecular diagnostics, in CNS tumour classification. To align to these new developments the 1998 ENCR recommendations required an update.

# Aims of the Recommendation

The aim of these recommendations is to register CNS tumours in a standardized way in all European cancer registries, in order to be able to generate comparable cancer statistics (incidence, prevalence, survival, mortality) of CNS tumours for all European countries. These cancer statistics are essential for the detection of cancer inequalities in Europe and impose plans to diminish these inequalities.

# Entering into Force

The new Recommendations for coding tumours of the central nervous system (CNS) is published on the website on ##-##-2024. These recommendations should be applied to all tumours with an incidence date as of 1-1-2024, but may also be applied to earlier dates.

# Tumours to be registered

It is recommended that cancer registries include in their database all intracranial and intraspinal neoplasms irrespective of their behaviour (benign/uncertain/malignant). The principal reasons are:

* It is difficult to distinguish benign from malignant tumours by symptoms alone
* All brain and spinal tumours are capable of producing severe clinical effects, irrespective of malignancy
* Aetiological and clinical syndromes associated with certain benign tumours may be of especial interest (meningiomas, pituitary tumours, etc.)
* Certain tumours - notably astrocytomas - progress from low grade (benign) to high grade (malignant) during their clinical course

Certain ‘tumours’ such as benign vascular lesions of meninges (haemangiomas), and cysts may, however, be excluded. Reporting of brain and spinal lesions may or may not include benign/uncertain neoplasms, according to the comparisons being made.

Multiple tumours

Multiple CNS tumours are common in neurofibromatosis, but may also occur in other cases. Neurofibromatosis is a group of conditions in which tumours grow in the nervous system. Type 2 neurofibromatosis (NF2) is a genetic condition and causes tumours of the brain, spinal cord and peripheral nerves, including (vestibular) schannoma, meningioma and ependymoma. Many NF2 patients have multiple tumours.

According to international rules one tumour should be registered per organ and per morphological group (see paragraph **4.4** **Multiple primary neoplasms** in ICD-O 3). For tumours in the central nervous system the tumour sites in table 1 are considered as separate organs. Table 2 gives an overview of the tumour types that should be considered as different.

1. A new primary CNS tumour should be registered for each different site (table 1) and/or each different morphology (table 2)
2. If there are multiple tumours in the same site and of the same tumour type, only the first should be registered

Table 1: Tumour sites to be considered as different

|  |  |
| --- | --- |
| Site | Topography codes |
| Meninges | C70 |
| Cerebrum | C71.0-C71.4 |
| Ventricles | C71.5 |
| Cerebellum/brain stem | C71.6-C71.7 |
| Spinal cord/cauda equina | C72.0-C72.1 |
| Left cranial nerves | C72.2-C72.5 (left) |
| Right cranial nerves | C72.2-C72.5 (right) |
| Pituitary gland/craniopharyngeal duct | C75.1-C75.2 |
| Pineal gland | C75.3 |

Table 2: Tumour types to be considered as different

|  |  |
| --- | --- |
| Tumour type | Morphology codes |
| Gliomas | 9380, 9381, 9382, 9384, 9385, 9400-9460 |
| Glioneuronal and neuronal tumours | 9412, 9413, 9492, 9493, 9505, 9506, 9509 |
| Ependymal tumours | 9383, 9391-9394, 9396 |
| Choroid plexus tumours | 9390 |
| Embryonal tumours | 9470-9478, 9490, 9500, 9501, 9508 |
| Pineal tumours | 9360, 9361, 9362, 9395 |
| Nerve sheath tumours | 9540-9571 |
| Meningiomas | 9530-9539 |
| Melanomas | 8720-8790 |
| Germ cell tumours | 9060-9110 |
| Cauda equina NET/paraganglioma | 8680, 8693 |

Clinical diagnosis only

A considerable proportion is CNS tumours is not pathologically confirmed. See the recommendations for the basis of diagnosis for coding these tumours. If a grade is mentioned in the imaging report, the grade should also be registered.

1. Grade (if indicated in the imaging report) should also be registered in the case of a clinical diagnosis (see paragraph “CNS tumour grading”).

Topography

**Cranial and spinal nerves**

It is not always possible to distinguish between extracranial and intracranial tumours of the cranial and spinal nerves. For tumours of the cranial and spinal nerves the following rules apply:

* All malignant tumours have to be registered
* Cauda equina neuroendocrine tumours (previously paraganglioma) have to be registered
* If the site (intracranial/extracranial) of schwannoma of a cranial nerve is unknown, intracranial may be assumed and the tumour should be registered
* If the site (intracranial/extracranial) of neurofibroma, perineuroma of a cranial nerve is unknown, the tumour should not be registered
* A tumour of the ‘spinal nerve root’ or an ‘intradural’ spinal tumour is considered intraspinal
* All benign *intracranial* tumours (schwannoma, neurofibroma, perineuroma) of the cranial nerves have to be registered
* The registration of benign *intraspinal* tumours (schwannoma, neurofibroma, perineuroma) of the spinal nerves is optional; these tumours should not be included in cancer incidence
* If the site (intracranial/extracranial) of schwannoma, neurofibroma, perineuroma of the spinal nerves is unknown, the tumour should not be registered

**Overlapping sites of the brain**

For tumours of overlapping sites of the frontal, temporal, parietal or occipital lobe of the brain, topography code C71.0 (‘supratentorial brain’ or ‘hemisphere’) is recommended.

Laterality

For tumours of the cerebrum prognosis and/or treatment may be different for tumours in the midline or in the hemispheres. The same may be true for tumours that extent into both hemispheres. It is recommended to code laterality for tumours of the brain and cranial nerves. Laterality does not have to collected for tumours of the meninges (C70), cerebellum (C71.6), brain stem (C71.7), spinal cord/cauda equina (C72.0-1), overlapping and unspecified nervous system (C72.8-9), pituitary gland (C75.1), craniopharyngeal duct C75.2) and the pineal gland (C75.3). The following combinations are valid:

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
|  | Left (1) | Right (2) | Midline (3) | Bilateral (4) |
| C71.0 | Left hemisphere | Right hemisphere | Midline structures of the cerebrum\* | Left and right hemisphere |
| C71.1 | Left frontal lobe | Right frontal lobe | - | Left and right frontal lobe |
| C71.2 | Left temporal lobe | Right temporal lobe | - | - |
| C71.3 | Left parietal lobe | Right parietal lobe | - | - |
| C71.3 | Left occipital lobe | Right occipital lobe | - | Left and right occipital lobe |
| C71.5 | Left lateral ventricle | Right lateral ventricle | Third ventricle | - |
| C71.8 | - | - | Corpus callosum | Butterfly glioma |
| C71.9 | Left brain | Right brain | Brian, midline | Left and right brain |
| C72.2\*\* | Left olfactory nerve | Right olfactory nerve | - | - |
| C72.3\*\* | Left optic nerve | Right optic nerve | Optic chiasm | - |
| C72.4\*\* | Left acoustic nerve | Right acoustic nerve | - | - |
| C72.5\*\* | Left cranial nerve, NOS | Right cranial nerve, NOS | - | - |

\* These include thalamus, hypothalamus, basal ganglia, etc.

\*\* Bilateral primary tumours of the cranial nerves have to be registered as two tumours (see section ‘multiple tumours’)

Morphology

Morphology should be registered according to the latest version of the WHO Classification of CNS Tumours.

**Cytogenetic and molecular aberrations**

Given the important role of molecular diagnostics in CNS tumour classification registries are recommended to actively search for the results of those molecular diagnostics and include them in the classification.

Behaviour code for CNS tumours

In ICD-O all neoplasms are registered with a behaviour code at the 5th digit, where /0 is used for benign tumours, /1 for tumours with uncertain malignant potential and /3 for malignant tumours. In the 2021 WHO Classification of CNS Tumours behaviour is no longer used. For the sake of comparisons over time a behaviour code should still be registered as shown in Appendix 2.

CNS tumour grading

Grade is one of the most important prognostic factors for CNS tumours (equivalent to ‘stage’ for most other solid cancers) and it is therefore an essential variable for cancer registries

1. For tumours of the central nervous system ‘CNS WHO grade’ should be registered as the sixth digit of the morphology. For many decades CNS tumour grading has differed from the grading of other, non-CNS neoplasms. For example, ‘anaplastic’ is coded as CNS WHO grade 3 (e.g. anaplastic astrocytoma= 9401/33), while is it grade 4 for carcinomas (e.g. anaplastic thyroid carcinoma = M8021/34).
2. A numeric grade (1, 2, 3, 4) prevails over a descriptive grade (low grade, high grade)
3. In the absence of a numeric grade ‘low grade’ should be coded as 2 and ‘high grade’ should be coded as 3.
4. If a grade range is indicated, e.g. grade 2-3, the higher should be coded.
5. If grade is not mentioned in the (pathology) report the grade should be coded as shown in table 1. Code as ‘unknown’ (9) if no or more than one grade is shown in this table.
6. In case of a discrepancy between the grade in table 1 and the pathology report it is recommended to seek advice from a coding expert of the registry or at <https://encr.eu/ask-an-expert>.

Progression

Low grade CNS tumours may progress to a higher grade over time. For example, a grade 2 astrocytoma may progress to an anaplastic astrocytoma (grade 3). Such a progression should not be registered as a new primary, but should be recorded as a progression.

1. Progression to a higher grade should not be registered as a new primary
2. Progression of a tumour to a higher grade should be registered as a progression of that tumour
3. If a pathologically confirmed CNS tumour was preceded by a CNS tumour of a lower grade, the tumour of the lower grade should be registered as the incident one, even if there is no pathology
   1. The incidence data is the date of the first confirmation of the low grade tumour (according to ENCR recommendations for the date of incidence)
   2. The grade registered at the incidence date is the grade of the low grade tumour
   3. If there was only a radiological diagnosis of the low grade tumour, the same morphology is assumed as for the (pathologically confirmed) high grade tumour

Example:

A tumour is notified to the registry by a pathology laboratory with the diagnosis ‘anaplastic oligodendroglioma’ (date: 21-07-2022). At review of the medical file it appears that on 14-3-2020 the patient was diagnosed with a ‘low grade glioma’ on MRI, followed by a wait & see policy.

1. incidence date 14-3-2020
2. CNS grade 2 (low grade)
3. morphology 9450/3 (oligodendroglioma)
4. 2. and 3. combined 9450/32 (low grade oligodendroglioma)
5. date of progression 21-07-2022
6. morphology at progression 9451/33 (anaplastic oligodendroglioma)

# References

David N Louis, Arie Perry, Pieter Wesseling, Daniel J Brat, Ian A Cree, Dominique Figarella-Branger, Cynthia Hawkins, H K Ng, Stefan M Pfister, Guido Reifenberger, Riccardo Soffietti, Andreas von Deimling, David W Ellison. The 2021 WHO Classification of Tumors of the Central Nervous System: a summary. Neuro-oncology 2021 Aug 2;23(8):1231-1251.

# Appendix 1: Working Group Members

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# Appendix 2: Morphology codes, behaviour codes and CNS grade

| Diagnosis | Grade | Morphology | Behaviour | CNS grade |
| --- | --- | --- | --- | --- |
| **Adult-type diffuse gliomas** | | | | |
| Astrocytoma, IDH-mutant | grade 2/low grade | 9400 | 3 | 2 |
| grade 3/anaplastic | 9401 | 3 | 3 |
| grade 4 | 9445 | 3 | 4 |
| unknown | 9400 | 3 | 9 |
| Astrocytoma, IDH-wildtype | grade 2/low grade | 9400 | 3 | 2 |
| grade 3/anaplastic | 9401 | 3 | 3 |
| unknown | 9400 | 3 | 9 |
| Astrocytoma, not otherwise specified | grade 2/low grade | 9400 | 3 | 2 |
| grade 3/anaplastic | 9401 | 3 | 3 |
| unknown | 9400 | 3 | 9 |
| Oligodendroglioma/oligoastocytoma, IDH-mutant and 1p/19q-codeleted | grade 2/low grade | 9450 | 3 | 2 |
| grade 3/anaplastic | 9451 | 3 | 3 |
| unknown | 9450 | 3 | 9 |
| Oligodendroglioma, not otherwise specified | grade 2/low grade | 9450 | 3 | 2 |
| grade 3/anaplastic | 9451 | 3 | 3 |
| unknown | 9450 | 3 | 9 |
| Oligoastocytoma, not otherwise specified | grade 2/low grade | 9382 | 3 | 2 |
| grade 3/anaplastic | 9382 | 3 | 3 |
| unknown | 9382 | 3 | 9 |
| Glioblastoma, IDH-wildtype |  | 9440 | 3 | 4 |
| Glioblastoma, not otherwise specified |  | 9440 | 3 | 4 |
| **Paediatric-type diffuse low-grade gliomas** | | | | |
| Diffuse astrocytoma, MYB- or MYBL1-altered |  | 9421 | 1 | 1 |
| Angiocentric glioma |  | 9431 | 1 | 1 |
| Polymorphous low-grade neuroepithelial tumour of the young |  | 9413 | 0 | 1 |
| Diffuse low-grade glioma, MAPK pathway-altered |  | 9421 | 1 | 1 |
| Diffuse midline glioma, H3 K27-altered |  | 9385 | 3 | 4 |
| Diffuse hemispheric glioma, H3 G34-mutant |  | 9385 | 3 | 4 |
| Diffuse paediatric-type high-grade glioma, H3-wildtype and IDH-wildtype |  | 9385 | 3 | 4 |
| Infant-type hemispheric glioma |  | 9385 | 3 | 4 |
| **Circumscribed astrocytic gliomas** | | | | |
| Pilocytic astrocytoma |  | 9421 | 1 | 1 |
| High-grade astrocytoma with piloid features |  | 9421 | 3 | 3,4 |
| Pleomorphic xanthoastrocytoma | grade 2/low grade | 9424 | 3 | 2 |
| grade 3/anaplastic | 9424 | 3 | 3 |
| unknown | 9424 | 3 | 9 |
| Subependymal giant cell astrocytoma |  | 9384 | 1 | 1 |
| Chordoid glioma |  | 9444 | 1 | 2 |
| Astroblastoma, MN1-altered | grade 2/low grade | 9430 | 3 | 2 |
| grade 3/anaplastic | 9430 | 3 | 3 |
| grade 4 | 9430 | 3 | 4 |
| unknown | 9430 | 3 | 9 |
| **Unspecified gliomas** |  |  |  |  |
| Glioma, not otherwise specified | grade 2/low grade | 9380 | 3 | 2 |
| grade 3/anaplastic | 9380 | 3 | 3 |
| grade 4 | 9380 | 3 | 4 |
| unknown | 9380 | 3 | 9 |
| Gliomatosis cerebri | grade 2/low grade | 9381 | 3 | 2 |
| grade 3/anaplastic | 9381 | 3 | 3 |
| grade 4 | 9381 | 3 | 4 |
| unknown | 9381 | 3 | 9 |
| **Glioneuronal and neuronal tumours** | | | | |
| Gangliocytoma |  | 9492 | 0 | 1 |
| Ganglioglioma | grade 1 | 9505 | 1 | 1 |
| grade 2 | 9505 | 1 | 2 |
| grade 3/anaplastic | 9505 | 3 | 3 |
| unknown | 9505 | 1 | 9 |
| Desmoplastic infantile ganglioglioma |  | 9412 | 1 | 1 |
| Desmoplastic infantile astrocytoma |  | 9412 | 1 | 1 |
| Dysembryoplastic neuroepithelial tumour |  | 9413 | 0 | 1 |
| Diffuse glioneuronal tumour with oligodendroglioma-like features and nuclear clusters |  |  |  |  |
| Papillary glioneuronal tumour |  | 9509 | 1 | 1 |
| Rosette-forming glioneuronal tumour |  | 9509 | 1 | 1 |
| Myxoid glioneuronal tumour |  | 9509 | 1 | 1 |
| Diffuse leptomeningeal glioneuronal tumour | grade 2 | 9509 | 3 | 2 |
| grade 3/anaplastic | 9509 | 3 | 9 |
| unknown | 9509 | 3 | 9 |
| Multinodular and vacuolating neuronal tumour |  | 9509 | 0 | 1 |
| Dysplastic cerebellar gangliocytoma (Lhermitte-Duclos disease) |  | 9493 | 0 | 1 |
| Central neurocytoma |  | 9506 | 1 | 2 |
| Extraventricular neurocytoma |  | 9506 | 1 | 2 |
| Cerebellar liponeurocytoma |  | 9506 | 1 | 2 |
| Glioneuronal tumour, not otherwise specified | grade 1 | 9509 | 1 | 1 |
| grade 2 | 9509 | 1 | 2 |
| grade 3/anaplastic | 9509 | 3 | 3 |
| unknown | 9509 | 1 | 9 |
| **Ependymal tumours** | | | | |
| Supratentorial ependymoma | grade 2 | 9391 | 3 | 2 |
| grade 3/anaplastic | 9392 | 3 | 3 |
| unknown | 9391 | 3 | 9 |
| Supratentorial ependymoma, ZFTA fusion-positive | grade 2 | 9396 | 3 | 2 |
| grade 3/anaplastic | 9396 | 3 | 3 |
| unknown | 9396 | 3 | 9 |
| Supratentorial ependymoma, YAP1 fusion-positive | grade 2 | 9396 | 3 | 2 |
| grade 3/anaplastic | 9396 | 3 | 3 |
| unknown | 9396 | 3 | 9 |
| Posterior fossa ependymoma | grade 2 | 9391 | 3 | 2 |
| grade 3/anaplastic | 9392 | 3 | 3 |
| unknown | 9391 | 3 | 9 |
| Posterior fossa group A (PFA) ependymoma | grade 2 | 9396 | 3 | 2 |
| grade 3/anaplastic | 9396 | 3 | 3 |
| unknown | 9396 | 3 | 9 |
| Posterior fossa group B (PFB) ependymoma | grade 2 | 9396 | 3 | 2 |
| grade 3/anaplastic | 9396 | 3 | 3 |
| unknown | 9396 | 3 | 9 |
| Spinal ependymoma | grade 2 | 9391 | 3 | 2 |
| grade 3/anaplastic | 9392 | 3 | 3 |
| unknown | 9391 | 3 | 9 |
| Spinal ependymoma, MYCN-amplified | grade 2 | 9396 | 3 | 2 |
| grade 3/anaplastic | 9396 | 3 | 3 |
| unknown | 9396 | 3 | 9 |
| Myxopapillary ependymoma |  | 9394 | 1 | 2 |
| Subependymoma |  | 9383 | 1 | 1 |
| **Choroid plexus tumours** | | | | |
| Choroid plexus papilloma |  | 9390 | 0 | 1 |
| Atypical choroid plexus papilloma |  | 9390 | 1 | 2 |
| Choroid plexus carcinoma |  | 9390 | 3 | 3 |
| **Embryonal tumours** | | | | |
| Medulloblastoma, WNT-activated |  | 9475 | 3 | 4 |
| Medulloblastoma, SHH-activated and TP53-wildtype |  | 9471 | 3 | 4 |
| Medulloblastoma, SHH-activated and TP53-mutant |  | 9476 | 3 | 4 |
| Medulloblastoma, non-WNT/non-SHH |  | 9477 | 3 | 4 |
| Medulloblastoma, histologically defined |  | 9470 | 3 | 4 |
| Atypical teratoid/rhabdoid tumour |  | 9508 | 3 | 4 |
| Cribriform neuroepithelial tumour |  | 9473 | 3 | 4 |
| Embryonal tumour with multilayered rosettes |  | 9478 | 3 | 4 |
| CNS neuroblastoma, FOXR2-activated |  | 9500 | 3 | 4 |
| CNS tumour with BCOR internal tandem duplication |  | 9500 | 3 | 4 |
| CNS embryonal tumour NEC/NOS |  | 9473 | 3 | 4 |
| **Pineal tumours** | | | | |
| Pineocytoma |  | 9631 | 1 | 1 |
| Pineal parenchymal tumour of intermediate differentiation | grade 2 | 9362 | 3 | 2 |
| grade 3 | 9632 | 3 | 3 |
| unknown | 9362 | 3 | 9 |
| Pineoblastoma |  | 9362 | 3 | 4 |
| Papillary tumour of the pineal region | grade 2 | 9395 | 3 | 2 |
| grade 3 | 9395 | 3 | 3 |
| unknown | 9395 | 3 | 9 |
| Desmoplastic myxoid tumour of the pineal region, SMARCB1-mutant |  |  |  |  |
| **Cranial and paraspinal nerve tumours** | | | | |
| Schwannoma |  | 9560 | 0 | 1 |
| Neurofibroma |  | 9540 | 0 | 1 |
| Plexiform neurofibroma |  | 9550 | 0 | 1 |
| Perineurioma |  | 9571 | 0 | 1 |
| Hybrid nerve sheath tumours |  | 9563 | 0 | 1 |
| Malignant melanotic nerve sheath tumour |  | 9540 | 3 | 9 |
| Malignant peripheral nerve sheath tumour |  | 9540 | 3 | 9 |
| Cauda equina neuroendocrine tumour (previously paraganglioma) | grade 1 | 8693 | 3 | 1 |
| unknown | 8693 | 3 | 9 |
| **Meningioma** | | | | |
| Meningioma, not otherwise specified |  | 9530 | 0 | 1 |
| Microcystic meningioma |  | 9530 | 0 | 1 |
| Secretory meningioma |  | 9530 | 0 | 1 |
| Lymphoplasmacyte-rich meningioma |  | 9530 | 0 | 1 |
| Metaplastic meningioma |  | 9530 | 0 | 1 |
| Meningothelial meningioma |  | 9531 | 0 | 1 |
| Fibrous meningioma |  | 9532 | 0 | 1 |
| Psammomatous meningioma |  | 9533 | 0 | 1 |
| Angiomatous meningioma |  | 9534 | 0 | 1 |
| Transitional meningioma |  | 9537 | 0 | 1 |
| Chordoid meningioma |  | 9538 | 1 | 2 |
| Clear cell meningioma |  | 9538 | 1 | 2 |
| Atypical meningioma |  | 9539 | 1 | 2 |
| Anaplastic (malignant) meningioma |  | 9530 | 3 | 3 |
| Rhabdoid meningioma |  | 9538 | 3 | 3 |
| Papillary meningioma |  | 9538 | 3 | 3 |