ENCR Recommendations 2024

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

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Contents

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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 18-12-2024. These recommendations should be applied to all tumours with an incidence date as of 1-1-2025, but may also be applied to earlier dates.

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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 its behaviour
- 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) schwannoma, 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 for registration purposes.

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)

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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 ENCR Recommendations for the Basis of Diagnosis for coding these tumours.

3. 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

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- 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')

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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

4. 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).
5. A numeric grade (1, 2, 3, 4) prevails over a descriptive grade (low grade, high grade)
6. In the absence of a numeric grade 'low grade' should be coded as 2 and 'high grade' should be coded as 3.
7. If a grade range is indicated, e.g. grade 2-3, the higher should be coded.
8. 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.
9. 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.

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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.

10. Progression to a higher grade should not be registered as a new primary
11. Progression of a tumour to a higher grade should be registered as a progression of that tumour
12. 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
    a) The incidence data is the date of the first confirmation of the low grade tumour (according to ENCR recommendations for the date of incidence)
    b) The grade registered at the incidence date is the grade of the low grade tumour
    c) 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)

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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

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Diagnosis	Grade	Morphology	Behaviour	CNS grade
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

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Diagnosis	Grade	Morphology	Behaviour	CNS grade
	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

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Diagnosis	Grade	Morphology	Behaviour	CNS grade
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

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