

Recommendations for coding Tumours of the Brain and Central Nervous System

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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 malignancy
- Aetiological and clinical syndromes associated with certain benign tumours may be of especial interest (meningiomas, pituitary tumours...)
- 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.

WHO GRADE (MALIGNANCY SCALE)

1. The recording of grade is an important, although not indispensable element in CNS tumour typing. It is essential to the interpretation of data on clinical outcomes. Using the new WHO classification of brain tumours resolves a great many of the problems of determining tumour grade, since in most cases tumour grade is implicit in the diagnostic category.

GRADE I (e.g. pilocytic astrocytoma). Tumours with a low proliferative potential, a frequently discrete nature, and a possibility of cure following surgical resection alone.

GRADE II Generally infiltrating tumours low in mitotic activity, but with a potential to recur. Some tumour types tend to progress to lesions with higher grades of malignancy (e.g. well-differentiated astrocytomas, oligodendrogliomas and ependymomas).

GRADE III Histological evidence of malignancy, generally in the form of mitotic activity, clearly expressed infiltrative capabilities, and anaplasia.

GRADE IV Mitotically-active, necrosis-prone neoplasms, generally associated with a rapid pre- and postoperative evolution of the disease.

2. These definitions are not the same as those proposed for the general grading of tumours via the 6th digit of the morphology code of ICD-O (page xxviii of ICD-O, Second Edition), which relates primarily to degree of differentiation.

HOWEVER

For malignant tumours of the central nervous system (site codes C70 - C72, C75.1- C75.3) the grade should be recorded as the sixth digit of the ICD-O M code, according to the definition in Section 1.

3. The following table details the available grades:

**WHO GRADING SYSTEM (MALIGNANCY SCALE) OF CNS TUMOURS
AND ICD-O BEHAVIOUR CODE**

Tumour Type	Grade				ICD-O behaviour code
	I	II	III	IV	
Astrocytic tumours					
Subependymal giant cell	*				1
Pilocytic	*				3
Low grade		*			3
Pleomorphic xanthoastrocytoma		*	*		3
Anaplastic			*		3
Glioblastoma				*	3
Oligodendrogliomas					
Low grade		*			3
Anaplastic			*		3
Oligo-astrocytomas					
Low grade		*			3
Anaplastic			*		3
Ependymal tumours					
Subependymoma	*				1
Myxopapillary	*				1
Low grade		*			3
Anaplastic			*		3
Choroid plexus tumours					
Papilloma	*				0
Carcinoma			*	*	3
Neuronal/glial tumours					
Gangliocytoma	*				0
Ganglioglioma	*	*			1
Anaplastic ganglioglioma			*		3
Desmoplastic infantile ganglioglioma	*				0
Dysembryoplastic neuroepithelial tumour	*				-
Central neurocytoma	*				0
Pineal tumours					
Pineocytoma		*			1
Pineocytoma/pineoblastoma			*	*	-
Pineoblastoma				*	3
Embryonal tumours					
Medulloblastoma				*	3
Other PNETs				*	3
Medulloepithelioma				*	3
Neuroblastoma				*	3
Ependymoblastoma				*	3
Cranial and spinal nerve tumours					
Schwannoma	*				0
Malignant peripheral nerve sheath tumour			*	*	3
Meningeal tumours					
Meningioma	*				0
Atypical meningioma		*			1
Papillary meningioma		*	*		1
Haemangiopericytoma		*	*		3
Anaplastic meningioma			*		3

Footnote: " - " = no specific histology or malignancy code

UNUSED ICD-O CODES

The European Network of Cancer Registries working group recommends that cancer registries no longer use certain morphology codes, which correspond to diagnostic terms considered to

be obsolete. When these terms are encountered, the appropriate code (and diagnostic synonym) is as follows:

SUPPLEMENTARY INDEX*

9505/0	Dysembryoplastic neuroepithelial tumour (DNET)
9505/0	Desmoplastic infantile ganglioglioma
9505/3	Anaplastic (malignant) ganglioglioma
9361/1	Mixed/transitional pineal tumour
8726/1	Melanocytoma
9390/3	Choroid plexus carcinoma
9506/0	Central neurocytoma
9530/1	Atypical meningioma
9540/3	Malignant peripheral nerve sheath tumour
9470/3	Melanotic medulloblastoma (WHO blue book)
9470/3	Lipomatous medulloblastoma (Kleihues et al.)
8963/3	Atypical teratoid/rhabdoid tumour (Kleihues et al.)

- Terms not appearing in the ICD-O index
Some codes are 'matrix codes' - i.e. already exist but without the behaviour code specified

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