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Clinical Grade for CNS Primaries
Using the WHO Grading System for Selected Tumors of the CNS

Mary Trimble, CTR

If we have a diagnostic biopsy of a tumor, we can code the clinical grade regardless of the primary site. What is odd about Central Nervous System (CNS) tumors (Figures 1 and 2) is that we can code a clinical grade in the absence of a diagnostic biopsy or histologic confirmation for these primaries. For any other primary, we must have histologic or cytologic confirmation in order to code grade at all. So, what makes the Central Nervous System different? How is it we are able to code clinical grade without any histologic confirmation of the tumor? The World Health Organization (WHO) Classification of Tumors of the Central Nervous System, 4th Edition, released in 2016, provides the WHO grading system of CNS tumors as a uniform way to categorize CNS tumors.

Figure 1
Lobes of the Cerebellum

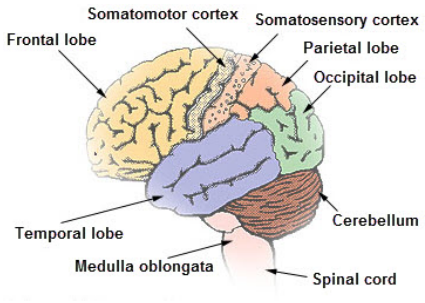
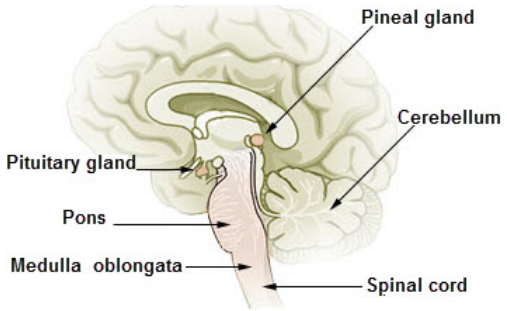


Figure 2
Pituitary and Pineal Glands



American Joint Committee on Cancer (AJCC) Manual, 8th Edition - Table 72.2

This four-tiered grade scale helps to classify select CNS histologies based on proliferative potential and the risk of spread or recurrence. In other words, the WHO grade is a malignancy scale where the higher the WHO grade, the more aggressive the clinical course expected for the tumor." This WHO classification can be seen as part of a larger clinical definition for select histologies and can therefore be used to assign grade without histologic confirmation.

There are several pitfalls we need to avoid to improve our coding of this new field. Most importantly, it is critical to remember that a default WHO grade can only be coded for the histologies listed in Table 1, which reflects the information from Table 72.2 of the AJCC Manual, 8th Edition. The WHO grade for many common CNS histologies is provided on the table. A simple way to improve our coding accuracy of the Clinical Grade field is to check these tables if we aren't certain the default WHO grade can be coded for the case we're working on. For example, Glioma, NOS is a fairly common clinical diagnosis made for brain tumors. However, Glioma, NOS is not listed in Table 72.2, so clinical grade for this case would be coded to 9. It may be helpful to have a version of the table printed out and readily available to aid in quick verification.

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Table 1 • AJCC Manual 8th Edition • WHO Grading System Table 72.2

Tumor Type	Grade I	Grade II	Grade III	Grade IV
Anaplastic astrocytoma			X	
Anaplastic ependymoma			X	
Anaplastic ganglioglioma			X	
Anaplastic meningioma			X	
Anaplastic oligodendroglioma			X	
Anaplastic pleomorphic xanthoastrocytoma			X	
Angiocentric glioma	X			
Atypical choroid plexus papilloma		X		
Atypical meningioma		X		
Atypical teratoid/rhabdoid tumor				X
Central neurocytoma		X		
Cerebellar liponeurocytoma		X		
Chordoid glioma of the third ventricle		X		
Chordoid meningioma		X		
Choroid plexus carcinoma			X	
Choroid plexus papilloma	X			
Clear cell meningioma		X		
CNS embryonal tumor				X
CNS ganglioneuroblastoma				X
CNS neuroblastoma				X
Craniopharyngioma	X			
Desmoplastic infantile ganglioglioma/astroctyoma	X			
Diffuse astrocytoma		X		
Dysembryoplastic neuroepithelial tumor (DNET)	X			
Embryonal tumor with multilayered rosettes				X
Ependymoma		X		
Extraventricular neurocytoma		X		
Gangliocytoma	X			
Ganglioglioma	X			
Glioblastoma				X
Granular cell tumor	X			
Hemangioblastoma	X			
Malignant peripheral nerve sheath tumor (MPNST)		X	X	X
Medulloblastoma				X
Medulloepithelioma				X
Meningioma	X			
Myxopapillary ependymoma	X			
Neurofibroma	X			
Oligodendroglioma		X		
Papillary glioneuronal tumor (PGNT)	X			
Papillary meningioma			X	
Papillary tumor of the pineal regiona		X	X	
Paraganglioma	X			
Perineurioma	X	X	X	
Pilocytic astrocytoma	X			
Pilomyxoid astrocytoma		X		
Pineal parenchymal tumor of intermediate differentiation		X	X	
Pineoblastoma				X
Pineocytoma	X			
Pituicytoma	X			
Pituitary adenoma	X			
Pleomorphic xanthoastrocytoma		X		
Rhabdoid meningioma			X	
Rosette-forming glioneuronal tumor of the foruth ventricle (RGNT)	X			
Schwannoma	X			
Solitary fibrous tumor/hemangiopericytoma	X	X	X	
Spindle cell oncocytoma	X			
Subependymal giant cell astrocytoma	X			
Subependymoma	X			

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Differential Diagnosis

It is important we remember that we need a single clinical diagnosis in order to use the table. In other words, Table 72.2 cannot be used to determine a default grade when multiple CNS histologies are listed in a differential diagnosis. However, when a differential diagnosis includes a statement that one histology is favored over the others mentioned, then we can use the table to determine the default grade. Even if all the histologies in the differential diagnosis have the same default grade indicated on the table, we cannot use the table. In this situation we have to code clinical grade to 9.

Timeframe Consideration

Findings from the pathological timeframe can never be used to retroactively code clinical timeframe data items. Clinical grade must be assigned during the clinical timeframe and that timeframe is over as soon as soon as first course treatment begins.

Stereotactic biopsy

Stereotactic biopsies are another special circumstance. Just because we code a "stereotactic biopsy" as surgery does not mean it necessarily meets the pathological timeframe requirements set out by the AJCC. A diagnostic biopsy is still a clinical procedure, even if SEER instructs us to code it as surgery. On the other hand, a stereotactic biopsy can qualify as falling in the pathological timeframe if the intent is to grossly remove the tumor during that procedure.

Scenarios

- A patient is diagnosed with a meningioma, NOS on imaging. The clinical grade is coded to 1 since meningioma, NOS has a WHO grade of I per Table 72.2.
- A patient is clinically diagnosed with a meningioma, NOS on imaging. A resection is done and patient is diagnosed pathologically with atypical meningioma with WHO grade of II. Clinical grade is coded to 1, because even though the histology was confirmed to be atypical meningioma, that diagnosis was not made during the clinical timeframe. Clinically, the patient had a histology assigned a WHO grade of I per Table 72.2, so that is what we code clinical grade to.
- A patient is diagnosed with a glioma, NOS on imaging. A resection is done and patient is found to have a glioblastoma, WHO grade IV. Clinical grade should be coded to 9 because the histologic confirmation of a WHO Grade IV tumor was made during the pathological timeframe, not the clinical timeframe, and a default WHO grade is not listed for a glioma, NOS in Table 72.2.
- A patient is clinically diagnosed with a brain tumor on MRI. The radiologist states the mass is most consistent with an oligodendroglioma vs. ependymoma. Oligodendroglioma is subsequently confirmed on resection. Clinical grade should be coded to 9. Even though both histologies in the differential diagnosis are found on Table 72.2 and both are WHO grade II tumors, the clinical grade can only be coded if a definitive diagnosis was made during the clinical timeframe. In this case, the oligodendroglioma wasn't confirmed until the resection. Therefore WHO grade II cannot be coded in the Clinical Grade field.

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Conclusion

Grade fields describe how much the tumor cells look like healthy cells when viewed on imaging or under the microscope. Cells from low-grade tumors (grades I and II) look more normal and generally grow more slowly than cells from high-grade tumors (grades III and IV).

CNS tumors are categorized and graded for clinical and research purposes according to the WHO classification scheme which separates tumors by histological type and predicted biological behavior. Unlike most other tumors, CNS neoplasms are not AJCC staged and, therefore, grading takes on greater importance in predicting the tumor's clinical behavior. As such, WHO grade plays a major role in estimating clinical outcomes and guiding treatment decisions.

Bottom-line, we need to remember the rules related to establishing clinical vs. pathologic timeframes and make sure we keep that AJCC Table 72.2 handy in order to improve our coding of clinical and pathologic grade fields for CNS tumors so researchers and clinicians can accurately assess their impact on diagnostic and clinical practice.

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