



NPCR CNS CODING GUIDELINES 2007



- Always use the **behavior code** listed in the ICD-O-3 unless otherwise directed by a pathologist.
Exception: Piloid/ Pilocytic/ Juvenile Astrocytomas **are reportable** as 9421/3 per NAACCR and SEER. (An ICD-O-3 correction was issued for this to assure that all ICD-O-3 manuals were corrected from a behavior of /1 to /3 for this histology.)
- **Meningiomas** are always coded to meninges (C70.-) unless specifically directed otherwise by a pathologist.
 - ◆ **Intraparenchymal meningiomas** are exceedingly rare.
 - ◆ Meningioma can also occur as a tumor of the **choroid plexus** in rare cases.
- **Solitary fibrous tumor** is a rare, usually dural based lesion of cranium or spinal canal; occasionally occurring in the lateral ventricle or spinal cord. They should be coded to meninges (70.-) unless specifically directed otherwise by a pathologist.
- **Nerve Sheath Tumors**
 - ◆ Malignant: all tumors are reportable. Always code to nerve of origin (C47.- or (C72.-).
 - ◆ Nonmalignant: reportable for intracranial segment of cranial nerves only. Always code to the nerve of origin, (C72.2, .3, .4, & .5)
 - ◆ **Neurofibroma, neurilemmoma and neuroma** are always peripheral nerve and thus can be either cranial nerve or nerve root.
 - ◆ **Pacinian tumor** (M9507/0) is a non-malignant peripheral nerve tumor. The only reportable pacinian tumors are those arising intracranially.
- **Germ cell tumors:** Intracranially, these tumors are usually located in the **pineal gland** (C75.3) and suprasellar region (C71.9 Brain, NOS), and posterior 3rd ventricle (C71.5). Code to site of origin.
 - ◆ A **teratoma** (M908-) is always a germ cell tumor. It may be malignant or non-malignant. The only nonmalignant teratomas that are reportable are those occurring intracranially.
- **Vascular tumors**
 - ◆ Malignant: all tumors are reportable
 - ◆ Nonmalignant: reportable for blood vessels of brain and spinal cord only
 - ❖ Code to CNS site of occurrence, not blood vessel.

Chordomas (9370-9372) are malignant tumors so ALL chordomas are reportable. These tumors usually start in the bone at the back of the skull (C41.0 bones of skull) or at the lower end of the spinal column (C41.2 vertebral column). 35% occur at the base of the skull. Intracranially, the tumors occur at the clivus (bones of skull: C41.0), and occasionally in the parasellar and sellar area (C71.9 Brain, NOS). All chordomas should be **coded to the bone of origin** unless otherwise directed by a pathologist.

Chondrosarcoma is a malignant tumor of cartilage cells so ALL chondrosarcomas are reportable. When these tumors develop in the skull base, they usually arise in the parasellar area, cerebellopontine angle, or paranasal sinuses. These tumors may also arise in the clivus. Clivus chordomas and chondrosarcomas may extend into the sella tursica, the clinoids, the nasopharynx, the posterior fossa, the foramen magnum, and may affect C1-2, the cranio-cervical junction. These tumors rarely may occur in the segmental spine arising from the vertebrae and commonly occur in the sacrum. These tumors should be coded to the bone of origin such as skull base/clivus (bones of skull: **C41.0**), spine: C3-L5 (**C41.2**), or sacrum (**C41.4**).

Chondromas (M9220/0 & M9221/0) are rare, slowly growing nonmalignant tumors which are only reportable if the primary tumor is in an intracranial site. In the cranial region, this includes the bones of the skull base and paranasal sinuses. These tumors should be coded to bones of skull (**C41.0**). Reportability of these tumors is an area of “Unresolved Issues”. Expert neurosurgeons and neuropathologists believe that they should be reported and included in analysis of CNS tumors.

Paragangliomas are rare nonmalignant tumors. The only reportable paragangliomas are those arising intracranially. Paraganglia are located in several areas along the cervical nerves.

Carotid body tumors or **chemodectoma** comprise the majority of head and neck paragangliomas. These are coded to carotid body (**C75.4**).

In the ICD-O, **aortic body** and **other paraganglia** are in the same section as paragangliomas and are coded **C75.5**.

Glomus tumors arise from paraganglionic tissue in glomus bodies. These tumors are also coded to Aortic body and other paraganglia (**C75.5**).

Reportability of non-malignant intracranial paragangliomas is another area of “Unresolved Issues”. Again, expert neurosurgeons and neuropathologists believe that they should be collected and reported with CNS tumors.

HISTOLOGIES THAT ARE SITE-SPECIFIC

- **Choroid plexus tumors**
 - ◆ Located in the ventricular system
 - ❖ Code to ventricle (**C71.5**) unless otherwise directed by a pathologist.
- **Pituitary adenoma (M8272/0) & Pituitary carcinoma (M8272/3)**
 - ◆ Always code to pituitary (**C75.1**) unless otherwise directed by a pathologist.
- **Craniopharyngiomas (M9350/1)**
 - ◆ All craniopharyngiomas are non-malignant
 - ◆ Very few of these tumors actually arise in the craniopharyngeal duct. Most are either suprasellar (**C71.9 Brain, NOS**), or in the 3rd ventricle (**C71.5**).

- **Pineal Parenchymal tumors:** Always code to Pineal gland (**C75.3**) unless otherwise directed by a pathologist. This includes:
 - ◆ Pineocytomas (M9361/1)
 - ◆ Pineoblastomas (M9362/3)
 - ◆ Mixed pineocytoma-pineoblastoma (M9362/3)
 - ◆ Pineal astrocytomas (M9400/3)

A **dermoid** (M9084) is usually a mal-developmental tumor that can be either malignant or nonmalignant. The only nonmalignant dermoids that are reportable are those occurring intracranially.

A **desmoid tumor** is a nonmalignant fibrous tumor that does not occur intracranially, but is found on the neck. These tumors are not reportable.

A **myxoma** (M 9562) never occurs intracranially. However, something very similar to it does occur and is usually a scarred over meningioma.

THE FOLLOWING HISTOLOGIES SHOULD BE EXCLUDED FROM ALL BRAIN AND CNS (C70.0-72.9 AND C75.1-75.3) SITES:

- 8041/3 SMALL CELL CARCINOMA
- 8070/3 SQUAMOUS CELL CARCINOMA, NOS
- 8130/3 PAPILLARY TRANSITIONAL CELL CARCINOMA
- 8360 MULTIPLE ENDOCRINE ADENOMAS
- 8370 ADRENAL CORTICAL ADENOMA (these are sometimes coded to C75.1 (pituitary) site, but should be coded to C74.0 Adrenal)
- 8410 SEBACEOUS ADENOMA
- 8700 PHEOCHROMOCYTOMA (these sometimes are coded to C71.x and C72.x sites, but should be coded to C74.1 Medulla of adrenal gland)
- 8726 MAGNOCELLULAR NEVUS
- 8832/0, /3 DEMATOFIBROMA/SARCOMA
- 8891 EPITHELIAL LEIOMYOMA
- 8894 ANGIOMYOMA/MYOSARCOMA
- 8940/0 PLEOMORPHIC ADENOMA
- 9000 BRENNER TUMOR
- 9050 MESOTHELIOMA
- 9160 ANGIOFIBROMA, NOS
- 9520/3 OLFACTORY NEUROGENIC TUMOR