

**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Introduction**

**Note 1:** Central nervous system (CNS) **includes** the following **primary sites**: Cerebral meninges C700; spinal meninges C701; meninges NOS C709; brain C710-C719; spinal cord C720; cauda equina C721; olfactory nerve C722; optic nerve C723; acoustic nerve C724; cranial nerve NOS C725; overlapping lesion of brain and central nervous system C728; nervous system NOS C729; pituitary gland C751; craniopharyngeal duct C752; pineal gland C753.

**Note 2:** **Malignant CNS** neoplasms have a separate set of rules.

**Note 3:** 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

- Tumors diagnosed 01/01/2007 through 12/31/2017: Use 2007 MPH Rules
- Tumors diagnosed 01/01/2018 and later: Use 2018 Solid Tumor Rules
- The original tumor diagnosed before 1/1/2018 and a subsequent tumor diagnosed 1/1/2018 or later **in the same primary site**: Use the 2018 Solid Tumor Rules.

**Note 4:** Non-malignant central nervous system (CNS) neoplasms (previously called benign and borderline) are **reportable** for cases **diagnosed 1/1/2004** and later.

**Note 5:** **Pilocytic astrocytoma/juvenile pilocytic astrocytoma:**

- For cases diagnosed prior to 1/1/2023, these neoplasms are reportable in North American as malignant 9421/3 for all CNS sites with the exception of the optic nerve:
  - WHO Classification Tumors of the Central Nervous System and IARC designate pilocytic astrocytoma as a synonym for optic glioma
  - When the primary site is optic nerve and the diagnosis is either optic glioma or pilocytic astrocytoma, the behavior is non-malignant and coded 9421/1
  - Beginning with cases diagnosed 1/1/2023 forward, pilocytic astrocytoma/juvenile pilocytic astrocytoma are to be reported as 9421/1 for **all** CNS sites.

**Note 6:** Tables and rules refer to **ICD-O rather than ICD-O-3**. The version is not specified to allow for updates. Use the currently approved version of ICD-O.

**Note 7:** For those sites/histologies which have recognized **biomarkers**, the biomarkers may aid in the identification of histologic type. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.

**Note 8:** See the Head and Neck Rules for coding paragangliomas.

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**Equivalent or Equal Terms**

These terms can be used interchangeably:

- And; with  
*Note:* “And” and “with” are used as synonyms when **describing multiple histologies** within a **single tumor**.
- Atypical; uncertain behavior /1
- Cerebrospinal fluid; CSF
- Dermoid; dermoid cyst
- Dura; meninges
- Extradural; not within the meninges; within the cranium; within the skull but not within the cerebral meninges
- Extramedullary; outside medulla oblongata C700
- Intracranial; within the skull, within the cranium
- Intradural; between layers of the cerebral meninges; C700
- Intradural-extramedullary; within the spinal canal but outside of the nerves
- Intraspidal; occurring within the spinal column especially the vertebral canal
- Non-malignant is synonymous with:
  - /0 Benign
  - /1 Uncertain whether benign or malignant; borderline malignancy; low malignant potential; uncertain malignant potential
  - WHO Grade 1
- Site; topography
- Subarachnoid space; between the arachnoid mater and the pia mater of spinal meninges
- Tumor; mass; tumor mass; lesion; neoplasm
  - The terms tumor, mass, tumor mass, lesion, and neoplasm are **not** used in a **standard manner** in clinical diagnoses, scans, or consults. **Disregard** the terms unless there is a **physician’s statement** that the term is a non-malignant tumor/neoplasm
  - These terms are used **ONLY** for determining multiple primaries
  - **DO NOT USE** these terms for **casefinding** or **determining reportability**
- Type; subtype; variant

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**Terms that are NOT Equivalent or Equal**

These terms are **not equivalent**. There are no casefinding implications.

- **Component** is not equivalent to **subtype/type/variant**  
*Note:* Component is only coded when the pathologist specifies the component as a second **non-malignancy**.
- **Phenotype** is not equivalent to **subtype/type/variant**
- **WHO Grade** is not equivalent to **tumor grade**

**Changes from 2007 MPH Rules**

These changes are effective with cases diagnosed 1/1/2018 and later.

1. **Clarifications:**

- The following meningiomas are reportable: **Intraosseous, cavernous sinus, and sphenoid wing.**
- Multiple cerebral meningiomas (same histology or NOS and subtype/variant) are a single primary.
- Multiple brain tumors (same histology) are a single primary.
- Bilateral optic nerve gliomas/pilocytic astrocytomas are a single primary.
- Laterality is not used to determine multiple primaries.
- Timing is not used to determine multiple primaries.
- The brain C710-C719 is a single primary site.
- Neurofibromatosis NOS, Neurofibromatosis 1 (NF1), Neurofibromatosis 2 (NF2), and schwannomatosis are familial tumor syndromes and are not reportable conditions. People with NF1 and NF2 have a high risk of developing reportable and non-reportable tumors. Tumors associated with NF1 and NF2 are reportable when they meet the behavior (/0 or /1), site (within the CNS), and histology reportability requirements (see Reportability Criteria).

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2. The 2016 World Health Organization (WHO) Classification of Tumors of the Central Nervous System (CNS) contains newly recognized histologies and subtypes/variants. New codes/terms are identified by asterisks (\*) in Tables 5 and 6 in the Terms and Definitions.

**New for 2023**

Beginning with cases diagnosed 1/1/2023, pilocytic astrocytoma/juvenile pilocytic astrocytoma will **no longer be reported as malignant (9421/3)**. These neoplasms will continue to be reportable as a benign CNS tumor with a behavior of /1. Historically, pilocytic astrocytomas were coded as 9421/3 from 1976-2000. Beginning in 2001 with the release of ICD-O-3, WHO changed the behavior to /1, however, the standard setters opted to continue collecting these cases using malignant /3 behavior. This practice will continue through 12/31/2022.

**Reportability Criteria for Non-Malignant CNS Neoplasms**

CNS neoplasms must meet **all three** criteria/conditions below to be reported as non-malignant:

1. **The behavior must non-malignant /0 or /1.**

- A. Pathology designates the tumor as non-malignant (/0 or /1) **OR**
- B. Diagnostic imaging definitively states the tumor as non-malignant (/0 or/1) **OR**
- C. The tumor is a WHO Grade I (See [Section 1: Table 1](#))

*Note 1:* Always code the behavior code reported by the **pathologist**.

*Note 2:* **Never report** a **malignant /3** behavior code for a **meningioma** based on **tumor extension** to brain, skin of scalp, or other regional organs/tissue. Non-malignant CNS tumors can extend to the regional tissue and bone. Tumor extension is usually equivalent to atypical meningioma 9539/1.

2. **The primary site must be reportable** (See [Section 2: Table 3](#) and [Table 4](#))
3. **The histology must be reportable** (See [Section 2: Table 5](#) and [Table 6](#)).

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**Directory of Sections and Tables**

**Information is presented in the general order in which a case is abstracted.**

**Section 1: Behavior Code**

- A. **Priority Order** for Using Documentation to Assign Behavior
- B. **Table 1:** WHO Grades of Select CNS Neoplasms

**Section 2: Reportable Primary Sites and Histologies.**

- A. **Priorities for Coding Primary Site**
- B. **Reportable Primary Site Groups**
- C. **Table 2:** Reportable Primary Sites
- D. **Table 3:** Reportability of Non-Malignant Cranial Nerve (CN) Tumors (defines which portions of the cranial nerves are reportable)
- E. **Table 4:** Non-Reportable Neoplasms
- F. **Table 5:** Histologic Types of Non-Malignant Intracranial (Brain and Gland) Tumors
- G. **Table 6:** Specific Histologies, NOS, and Subtypes/Variants

*Note:* It is important to understand that non-malignant neoplasms do occur within the brain tissue.

**Section 3: Additional Information to Complete Abstract**

- A. **Conflicting information on Pathology report(s)**
- B. **Table 7:** Paired Sites
- C. **Table 8:** Non-malignant CNS Tumors with the Potential of Transforming to Malignant Behavior

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**Section 1: Behavior Code**

**Note:** Behavior determines which set of CNS rules should be used: malignant or non-malignant.

**Instructions** for using source **documentation** to determine behavior are in **priority order**. Start with Instruction 1 and **STOP** when you reach the instruction that applies to the case being abstracted. For example, when the **resection pathology** specifies behavior, use Instruction 1. **Do not** code the behavior using Instructions 2-5.

**Priority Order for Using Documentation to Assign Behavior**

1. Pathology: Tissue from **resection** in the following priority order:
  - A. Use the pathologist’s description of behavior  
*Note:* **Never change behavior** described by pathologist
  - B. Cases are reportable as non-malignant when pathology states a WHO Grade 1
  - C. WHO Grade 2 may be either non-malignant or malignant. Use the pathologist’s description of behavior (priority #1a)
2. Pathology: Tissue from **biopsy**
3. Cytology (usually cerebrospinal fluid)
4. Physician’s documentation (no pathology report) in the following priority order:
  - A. Tumor Board
  - B. Documentation of original diagnosis/tumor behavior  
*Example:* **Biopsy or resection** was done at a **different institution**; pathology report is not in the medical record. **Oncology consult** states **biopsy was done** in a different facility and **cites pathology from biopsy** including the **behavior** as benign, borderline, non-malignant or WHO Grade 1.
  - C. Documentation of behavior, no mention of original diagnosis  
*Example:* **Biopsy or resection** was done at a **different institution**; pathology report is not in the medical record. **Physician documents** the **behavior** as benign, borderline, non-malignant, or WHO Grade 1, but **does not cite/mention original** pathology report as source of behavior classification.
5. **Scans:** Use behavior information from imaging in the following **priority order**:
  - A. MRI
  - B. CT
  - C. PET

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D. Angiogram

6. When above instructions **do not apply**, use **Table 1** below to determine behavior.

**Table 1: WHO Grades of Select CNS Neoplasms**

**Note 1:** CNS WHO classifications use a grading scheme that is a "malignancy scale" ranging across a wide variety of neoplasms rather than a strict histologic grading system that can be applied equally to all tumor types.

**Note 2:** See the SEER and CoC Manuals for instructions on coding grade for CNS tumors.

**Note 3:** The table **does not** contain all neoplasms that may occur in the CNS.

**WHO Grade Definitions**

WHO Grade	Definition
<b>WHO Grade I</b>	Circumscribed tumors of low proliferative potential associated with the possibility of cure following resection
<b>WHO Grade II</b>	Infiltrative tumors with low proliferative potential with increased risk of recurrence
<b>WHO Grade III</b>	Tumors with histologic evidence of malignancy, including nuclear atypia and mitotic activity, associated with an aggressive clinical course
<b>WHO Grade IV</b>	Tumors that are cytologically malignant, mitotically active, and associated with rapid clinical progression and potential for dissemination

**WHO Grade II CNS Tumors: Non-malignant and Malignant**

Histologic grading is a means of predicting the biological behavior of a neoplasm. In the clinical setting, tumor grade is a key factor influencing the choice of therapies. WHO grade is based on histological features and is one component of a set of criteria used to predict response to therapy and outcomes.

WHO 4<sup>th</sup> Ed Tumors of CNS separates CNS entities into histologic groups. For example, diffuse astrocytic and oligodendroglial tumors are all malignant (/3). Some of the histologies that fall into this group have a WHO Grade II. They are malignant tumors and the grade predicts if the tumor is likely to transform into a more aggressive histology, determine treatment, and the prognosis.

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For the most part, benign CNS tumors (/0) and uncertain if benign or malignant CNS tumors (/1) are WHO Grade I. They may be given a WHO Grade II based on histologic features, clinical findings, and genetic testing. Grade II CNS tumors tend to be slow growing but may spread into nearby tissue or recur. This does not necessarily indicate the tumor is invasive. It indicates the tumor may progress to a higher behavior. The grade also guides treatment, follow-up, and prognosis.

When a CNS tumor is stated to be WHO Grade II, code the corresponding ICD-O-3 code and behavior listed in ICD-O, ICD-O updates, or solid tumor rules. Code the WHO Grade in the appropriate SSDI data field.

**Example:** Astrocytoma, NOS, WHO Grade II code 9400/3

**Table Instructions**

1. Use **non-malignant** CNS rules for **all WHO Grade I** (always non-malignant).
2. Go to the **malignant** CNS rules for **all WHO Grade 3 and 4**.
3. Go to **Section 1: Behavior Code** to determine whether **WHO Grade 2** neoplasms are non-malignant or malignant. Use the appropriate set of rules.

**Column 1** contains the **histology** term

**Column 2** contains the **WHO Grade** assigned **based on the histology and molecular features** of that histology

Histology	WHO Grade
Anaplastic (malignant) meningioma	3
Anaplastic astrocytoma, IDH mutant	3
Anaplastic ependymoma	3
Anaplastic ganglioglioma	3
Anaplastic oligodendroglioma IDH mutant and 1p/19q-codeleted	3
Anaplastic pleomorphic xanthroastrocytoma	3
Angiocentric glioma	1
Atypical choroid plexus papilloma	2
Atypical meningioma	2
Atypical teratoid/rhabdoid tumor	4
Central neurocytoma	2



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<b>Histology</b>	<b>WHO Grade</b>
Cerebellar liponeurocytoma	2
Chordoid glioma of third ventricle	2
Choroid plexus carcinoma	3
Choroid plexus papilloma	1
CNS embryonal tumor NOS	4
CNS embryonal tumor with rhabdoid features	4
Craniopharyngioma	1
Desmoplastic infantile astrocytoma and ganglioglioma	1
Diffuse astrocytoma, IDH mutant	2
Diffuse midline glioma, H3K27M-mutant	4
Dysembryoplastic neuroepithelial tumor	1
Dysplastic gangliocytoma of cerebellum (D=Lhemitte-Duclos)	1
Ependymoma	2
Ependymoma, RELA fusion-positive	2 or 3
<i>Note:</i> Tissue/pathology reports or CAP report will specify whether this is WHO Grade 2 or 3	
Extraventricular neurocytoma	2
Gangliocytoma	1
Glioblastoma, IDH mutant	4
Glioblastoma, IDH wildtype	4
Granular cell tumor	1
Hemangioblastoma	1
Malignant peripheral nerve sheath tumor (MPNST)	2, 3, or 4
<i>Note:</i> Tissue/pathology reports or CAP report will specify whether this is WHO Grade 2, 3, or 4	
Medulloblastoma (including all subtypes)	4

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<b>Histology</b>	<b>WHO Grade</b>
Medulloepithelioma	4
Meningioma	1
Myxopapillary ependymoma	1
Neurofibroma	1
Oligodendroglioma IDH mutant and 1p/19q-codeleted	2
Papillary glioneuronal tumor	1
Papillary tumor of the pineal region <i>Note:</i> Tissue/pathology report or CAP will specify whether it is WHO Grade 2 or 3	2 or 3
Perineuroma	1
Pilocytic astrocytoma <i>Note:</i> ICD-O-3 lists a behavior code of /1. US and Canada collect as /3.	1
Pineal parenchymal tumor of intermediate differentiation <i>Note:</i> Tissue/pathology or CAP synoptic report will specify WHO Grade 2 or 3	2 or 3
Pineoblastoma	4
Pineocytoma	1
Pituicytoma	1
Pleomorphic xanthroastrocytoma	2
Rosette-forming glioneuronal tumor	1
Schwannoma	1
Solitary fibrous tumor/hemangiopericytoma <i>Note:</i> Tissue/pathology will specify WHO Grade 1, 2, or 3	1, 2, or 3
Spindle cell oncocytoma	1
Subependymal giant cell astrocytoma	1
Subependymoma	1

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**Section 2: Reportable Primary Sites and Histologies**

After determining behavior code, the next step is to confirm both the primary site and the histology are reportable.

*Note 1:* The following non-malignant meningiomas are reportable:

- **Intraosseous**

*Note:* The **dura** layer of the meninges **contacts** the **endosteum** of the bones of the skull. The primary site for intraosseous meningioma is cranial meninges C700.

- **Sphenoid wing**

*Note 1:* Sphenoid wing meningiomas arise in the **cranial meninges** C700 which **covers** the bony structure called the sphenoid wing.

*Note 2:* The term “sphenoid wing meningioma” is used to identify the **location** of the meningioma because sphenoid wing meningiomas may be very **invasive**, spreading to the dura of the frontal, temporal and orbital regions.

- **Cavernous sinus**

*Note 1:* Cavernous sinus is located between the endosteal and meningeal layers of the dura.

*Note 2:* There is **no ICD-O site** code for cavernous sinus because tumors do not arise in the sinus itself; the primary sites are:

- The **cranial nerves** passing through the sinus (trochlear, abducent **C725**) **OR**
- The cerebral **meninges/dura C700** covering the cranial nerve

*Note 2:* Cavernous sinus hemangiomas are reportable. Code primary site cerebral meninges C700.

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**Priorities for Coding Primary Site**

- Note 1:** Always check the **operative report(s)** which will have information on whether the surgery or biopsy was **intracranial** (inside the cranium/skull) or **intraspinal** (within the dura/meninges covering the spinal cord)
- Note 2:** Code **the specific primary site**. Use an NOS site code only when a specific site is not known.
- Note 3:** See **Table 2: Reportable Primary Sites** to confirm the primary site is reportable.
- Note 4:** When the primary site is cranial nerve **OR** cranial nerve meninges, see **Table 3: Reportability of Non-Malignant Cranial Nerve (CN) Tumors** to confirm the site of the tumor is intracranial (reportable) or extracranial (not reportable)
- Note 5:** See **Table 4: Non-Reportable Neoplasms** for site/histology combinations and histologies that are not reportable.
- Note 6:** When the primary site is brain or intracranial glands, see **Table 5: Histologic Types of Non-Malignant Intracranial (Brain and Gland) Tumors** to confirm site/histology combinations.

Use the list below in **hierarchical order**:

1. **Resection**
  - A. Operative report(s)
  - B. Pathology report(s)
2. **Biopsy**
  - A. Operative report(s)
  - B. Pathology report(s)
3. Resection and/or biopsy performed, but operative report(s) and pathology are not available (minimal information):
  - A. Tumor Board
  - B. Code from physician's documentation of **original diagnosis** from operative or pathology report
  - C. Physician's **documentation of primary site** in the medical record

**Example:** The operative and pathology reports are not in the medical record. The **attending documents** that the patient had a biopsy of a **cerebral meningioma** at a different facility. Code the primary site cerebral meninges C700.
4. For cases diagnosed by imaging (**no pathology/resection or biopsy**) use information from scans in the following priority order:
  - A. MRI
  - B. CT
  - C. PET
  - D. Angiogram

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**Reportable Primary Site Groups**

The two major **groups** of reportable sites are:

1. **Intracranial** (within the skull/cranium)
2. **Spinal sites** (spinal meninges and sites within the spinal meninges)

**Reportable Primary Sites and their ICD-O Codes**

1. Intracranial (intra means inside or within; intracranial is inside the skull).
  - A. **Cerebral/cranial dura/meninges C700**. The cerebral meninges has three layers:
    - i. **Dura mater** is the **superficial** layer of meninges
      - Tightly adherent to skull
      - Contains folds and **sinuses**
      - Contacts **endosteum** which lines the bones of the skull
    - ii. **Arachnoid** mater forms the middle of the three layers of meninges
    - iii. **Pia** mater is the delicate vascular fibrous membrane which is adherent to the brain.
  - B. **Brain C710-C719**
  - C. **Cranial nerves C722-C729**. See [Table 3: Reportability of Non-Malignant Cranial Nerve \(CN\) Tumors](#)
  - D. **Intracranial glands C751-C753**
    - i. Craniopharyngeal duct C752
    - ii. Pineal gland C753
    - iii. Pituitary gland C751
2. **Spinal sites**: includes the **spinal meninges** and **all** structures **within** the meninges (Intradural, within/in the spinal meninges).
  - A. Spinal cord **C720**
  - B. The spinal meninges **C701** covers/encloses the spinal cord.

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**Table 2: Reportable Primary Sites**

**Column 1** lists the reportable primary site term.

**Column 2** lists the site code for the reportable primary site.

Site Group	Reportable Subsite Terms and Code
Brain	Brain NOS <b>C719</b> Brain stem <b>C717</b> Cerebellum NOS <b>C716</b> Cerebrum <b>C710</b> Frontal lobe <b>C711</b> Occipital lobe <b>C714</b> Overlapping lesion of brain and central nervous system <b>C718</b> Parietal lobe <b>C713</b> Temporal lobe <b>C712</b> Ventricle NOS <b>C715</b>
Cranial Nerves	Abducent (cranial nerve VI) <b>C725</b> Accessory (cranial nerve XI) <b>C725</b> Acoustic (cranial nerve VIII) <b>C724</b> Cranial nerve NOS <b>C725</b> Facial (cranial nerve VII) <b>C725</b> Glossopharyngeal (cranial nerve IX) <b>C725</b> Hypoglossal (cranial nerve XII) <b>C725</b> Oculomotor (cranial nerve III) <b>C725</b> Olfactory (cranial nerve I <b>C722</b> ) Optic (cranial nerve II) <b>C723</b> Trigeminal (cranial nerve V) <b>C725</b> Trochlear (cranial nerve IV) <b>C725</b> Vagus (cranial nerve X) <b>C725</b>

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Site Group	Reportable Subsite Terms and Code
Ill-Defined Sites Central Nervous System	Nervous system NOS <b>C729</b> Overlapping lesion of brain and central nervous system <b>C728</b>
Intracranial Duct and Glands	Craniopharyngeal duct <b>C752</b> Pineal gland <b>C753</b> Pituitary gland <b>C751</b>
Meninges	Cerebral meninges <b>C700</b> Meninges NOS <b>C709</b> Spinal meninges <b>C701</b>
Spinal Sites	Cauda equina <b>C721</b> Conus medullaris/filum terminale <b>C720</b> Meninges NOS <b>C709</b> Spinal cord <b>C720</b> Spinal meninges <b>C701</b>

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**Table 3: Reportability of Non-Malignant Cranial Nerve (CN) Tumors**

This table is used to determine whether non-malignant cranial nerve tumors and cranial nerve meningiomas are **reportable**. When cranial nerves **exit** the **intracranial** space, they become **peripheral nerves (non-reportable)**.

**Note 1:** A neoplasm arising in a **cranial** nerve is coded to the **specific nerve** in which it **arises**.

**Note 2:** Neoplasms, commonly meningiomas, **arising** in the **dura/meninges** of an **intracranial** nerve are coded to **cerebral meninges C700**.

**Note 3:** It is important to check the **operative report** to determine whether the surgery is **intracranial** or **intradural**.

**Note 4:** This table is used for **non-malignant** neoplasms **ONLY**.

**Column 1:** The **proper name** for the cranial nerve (CN) and the **cranial nerve number**

**Column 2:** The point at which the nerve exits the cranium

**Column 3:** **Reportable** portions of the cranial nerve. Of note, malignant neoplasms are reportable for all parts of the cranial nerves

**Column 4:** **Non-reportable** portions of the cranial nerve

<b>Name and CN #</b>	<b>Exits Cranium Through</b>	<b>Reportable Portions of CN</b>	<b>Non-Reportable Portions of CN</b>
Cranial nerve NOS <b>C725</b>		<b>Within cranium</b> , unknown which nerve	
Olfactory CN 1 <b>C722</b>	Cribriform plate	Surface of the <b>brain</b>	Originates on the <b>olfactory mucosa</b> of <b>nasal cavity</b> , then travels through the <b>cribriform plate</b> of the <b>ethmoid bone</b>
Optic CN 2 <b>C723</b>	Optic canal	<b>Always reportable:</b> CN2 is unique because it is intradural, covered with the meninges/dura and <b>all portions</b> are <b>reportable</b> .	
Oculomotor CN 3 <b>C725</b>	Superior orbital fissure	Originates in the <b>midbrain</b> .	After exiting the superior orbital fissure, the nerve enters the <b>orbit</b> .



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**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

Name and CN #	Exits Cranium Through	Reportable Portions of CN	Non-Reportable Portions of CN
Trochlear CN 4 <b>C725</b>	Superior orbital fissure	Arises from the <b>dorsal brain stem</b> , loops around the brainstem and passes anteriorly within the <b>subarachnoid space</b> . It travels between the <b>superior cerebellar and posterior cerebral arteries</b> and through the <b>dura</b> , enters <b>cavernous sinus</b> .	Enters the <b>orbital fissure</b> .
Trigeminal CN 5 <b>C725</b>  <i>Note:</i> Trigeminal is derived from Latin trigeminus which means <b>born in threes</b> (tri) and born <b>at the same time</b> (germinal). As the name implies, the nerve <b>separates</b> into <b>three branches</b> ; ophthalmic, maxillary, and mandibular	The ophthalmic, maxillary and mandibular branches leave the skull through three separate foramina, the superior orbital fissure, the foramen rotundum and the foramen ovale.	CN5 originates in the <b>pons</b> . Upon leaving the pons it enters a <b>small fossa</b> posterior and inferolateral to the cavernous sinus called <b>Meckel's (trigeminal) cave</b> .	<ul style="list-style-type: none"> <li>• <b>Ophthalmic nerve branch</b> crosses the <b>pterygopalatine fossa</b>, inclines laterally on the back of the <b>maxilla</b>, and enters the <b>orbit</b> through the inferior orbital fissure where it is called the <b>infraorbital nerve</b>. It ends beneath the <b>quadatus labii superius</b>, and divides into multiple branches that spread to the side of the <b>nose</b>, the lower <b>eyelid</b>, and the upper <b>lip</b></li> <li>• <b>Maxillary nerve</b> leaves foramen rotundum and traverses the <b>infraorbital groove and canal</b> in the floor of the <b>orbit</b>, and appears on the face at the infraorbital foramen.</li> <li>• <b>Mandibular nerve</b> leaves via the foremen ovale travels along the <b>mandibular groove</b></li> </ul>

**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

Name and CN #	Exits Cranium Through	Reportable Portions of CN	Non-Reportable Portions of CN
Abducent CN 6 <b>C725</b>	Cranial meninges	Exits brainstem at junction of <b>pons</b> and the <b>medulla</b> , enters the <b>subarachnoid</b> space and runs upward between the pons and the <b>clivus</b> entering the <b>cavernous sinus</b> .	<b>Dorello's canal</b> and travels to the tip of the <b>temporal bone</b> . Enters <b>orbit</b>
Facial CN 7 <b>C725</b>	Internal acoustic meatus	<b>CN7</b> originates in the <b>pons</b> , along the posterior cranial fossa (the posterior cranial fossa is part of the intracranial cavity)	Enters the <b>temple</b> through the <b>internal auditory meatus</b> and runs through the <b>facial canal</b> .
Acoustic or vestibulocochlear CN 8 <b>C724</b>	Internal acoustic meatus	Originates in the <b>brain stem (medulla oblongata)</b> between the base of the brain ( <b>pons</b> ) and the <b>spinal cord</b>  Both the <b>vestibular</b> branch and the <b>cochlear</b> branch are located in the <b>inner ear</b>	
Glossopharyngeal CN 9 <b>C725</b>	Jugular foramen	<b>Originates</b> in the anterior portion of the <b>medulla oblongata</b>	<b>Jugular foramen</b> Between the <b>internal jugular vein</b> and <b>internal carotid artery</b> Lies on the <b>stylopharyngeus</b> and <b>middle pharyngeal constrictor muscle</b> Passes under the <b>hypoglossus muscle</b> <b>Palatine tonsil</b> Extends to <b>mucous glands</b> of the <b>mouth</b> , and <b>base of the tongue</b>

**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

Name and CN #	Exits Cranium Through	Reportable Portions of CN	Non-Reportable Portions of CN
Vagus CN 10 <b>C725</b>	Jugular foramen	The vagus nerve originates from the <b>medulla</b> of the <b>brainstem</b> .	CN10 descends within the <b>carotid sheath</b> medial to the internal jugular vein at the root of the <b>neck</b> . The right vagus crosses in front of the subclavian artery and travels into the <b>fat behind the blood vessels</b> , reaching the <b>thorax</b> . It then inclines <b>behind the hilum</b> of the <b>right lung</b> and moves toward the esophagus. The nerve splits into the <b>right and left vagus</b> at the <b>esophageal plexus</b> forming the anterior and posterior <b>gastric nerves</b>
Accessory CN 11 <b>C725</b>	Jugular foramen	The nerve enters the foramen magnum or lateral aspect of the <b>medulla oblongata</b> .	<p>The spinal accessory nerve <b>originates</b> in the neurons of the upper spinal cord, specifically <b>C1-C5/C6 spinal nerve roots</b>.</p> <p>The fibers of the spinal accessory nerve coalesce to <b>form spinal rootlets, roots</b>, and finally the <b>spinal accessory nerve</b> itself.</p> <p>The nerve <b>exits</b> the skull through the jugular foramen. It then runs along the internal <b>carotid artery</b> within the <b>neck</b> and reaches the <b>sternocleidomastoid</b> muscle and the <b>trapezius</b>.</p>

**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

Name and CN #	Exits Cranium Through	Reportable Portions of CN	Non-Reportable Portions of CN
Hypoglossal CN 12 C725	Hypoglossal canal	CN12 starts in the <b>hypoglossal</b> nucleus of the <b>brainstem</b> ,	CN12 exits the hypoglossal canal, traveling <b>between</b> the <b>carotid</b> artery and <b>jugular</b> vein, ending under the <b>tongue</b> .

**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Table 4: Non-Reportable Neoplasms**

Use **Table 4** for **non-malignant neoplasms ONLY**. The table identifies **histology/site** combinations which are **not reportable**. This table was created from WHO with the cooperation of the Central Brain Tumor Registry of the United States (CBTRUS).

<b>Non-reportable Histology Term</b>	<b>Non-reportable Histology Code</b>	<b>Definitions and Sites</b>
Carcinomas	<b>8010-8060, 8071-8671, 8940-8941</b>	Brain C710-C719 <b>Site/histology edit</b> carcinomas/brain
Carcinomas	<b>8010-8671, 8940-8941</b>	Cerebral meninges, spinal meninges, meninges NOS C700-C709 <b>Site/histology edit</b> carcinomas/meninges
Carcinomas	<b>8010-8671, 8940-8941</b>	C721-C729 (Other central nervous system) <b>Site/histology edit</b> carcinomas/other CNS
Colloid cyst	<b>No code</b>	
Epidermoid tumor/cyst	<b>No code</b>	
Glomus tympanicum, glomus jugulare	<b>8690/1</b>	These tumors occur in the inner ear, the aortic body and other paraganglia respectively; these sites are <b>not reportable</b> .
Hygroma	<b>9173/0</b>	
Hypothalamic hamartoma	<b>No code</b>	Occurs in hypothalamus
Neurofibromatosis, NOS	<b>9540/1</b>	Genetic disease that produces non-malignant tumors in the skin, brain, CNS, and other sites. The brain and CNS tumors spawned by NF, NOS are reportable, the genetic disease is not.
Neurofibromatosis, type 1 (NF1)	<b>No code*</b>	Genetic disease that produces non-malignant tumors in the skin, brain, CNS, and other sites. The brain and CNS tumors spawned by NF1 are reportable, the genetic disease is not.
Neurofibromatosis, type 2 (NF2)	<b>No code*</b>	Genetic disease that produces non-malignant tumors in the skin, brain, CNS, and other sites. The brain and CNS tumors produced by NF2 are reportable, the genetic disease is not.
Neuroglial cyst	<b>No code</b>	Ventricles
Osteochondroma	<b>9210/0</b>	Originates in the cartilage around bone, site not reportable for non-malignant neoplasms

**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

Non-reportable Histology Term	Non-reportable Histology Code	Definitions and Sites
Rathke cleft cyst	<b>No code</b>	Sella turcica C751. This is a Rathke cleft CYST, not a Rathke cleft tumor.
Schwannomatosis	<b>No code*</b>	A form of neurofibromatosis newly named/discovered

**\*The code which is currently in ICD-O-3 for neurofibromatosis will not be used in ICD-O updates or in future ICD-O editions**

**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Table 5: Histologic Types of Non-Malignant Intracranial (Brain and Glands) Tumors**

Because non-malignant brain and gland tumors are **less common**, this table identifies histologies which occur in the brain C710-C719 and the glands within the cranium C751-C753. These histologies also appear in Table 6.

**IMPORTANT: This table does not list ALL PRIMARY SITES that are possible for a histology. It only includes sites where the histology can occur INTRACRANIALY.**

Use **Table 5** to:

- **Code primary site** when the instructions in **Section 2: Reportable Primary Sites and Histologies** do not apply
- **Confirm** that a **histology** can/should be coded to brain or intracranial glands

**Column 1** contains **histology terms and codes** that occur in the **brain, ventricles** of the brain, and **intracranial glands**

**Column 2** contains the **site code** for the most common **intracranial** primary site(s) for that specific histology

<b>Histology Term and Code</b>	<b>Most Common Intracranial Primary Site</b>
<b>Angiocentric glioma 9431/1*</b>	Cerebrum <b>C710</b>
<b>Choroid plexus papilloma 9390/0</b>	Intraventricular site (lateral/third ventricle <b>C715</b> and IV ventricle <b>C717</b> )
<b>(Capillary) hemangioblastoma 9161/1</b>	Cerebellum <b>C716</b> , cerebrum (rare) <b>C710</b>
<b>Craniopharyngioma 9350/1</b>	Craniopharyngeal duct <b>C752</b> , pituitary gland, sella turcica <b>C751</b>
<b>Dermoid cyst 9084/0</b>	Pineal gland <b>C753</b> , suprasellar <b>C719</b>
<b>Desmoplastic infantile astrocytoma and ganglioglioma 9412/1</b>	Cerebrum/supratentorial brain NOS <b>C710</b>
<b>Dysembryoplastic neuroepithelial tumor (DNT) 9413/0</b>	Cerebrum <b>C710</b> , temporal lobe <b>C712</b>

**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

<b>Histology Term and Code</b>	<b>Most Common Intracranial Primary Site</b>
<b>Dysplastic gangliocytoma 9493/0</b>	Cerebellum <b>C716</b>
<b>Juvenile xanthogranuloma 9749/1</b>	Intraventricular <b>C715</b>
<b>Meningioma (rare) 9530/0</b>	Intraventricular <b>C715</b>
<b>Myxopapillary ependymoma 9394/1</b>	4 <sup>th</sup> ventricle <b>C717</b>
<b>Pilocytic astrocytoma/juvenile pilocytic astrocytoma 9421/1</b>	<b>Optic nerve C723</b>
<b>Pineocytoma 9361/1</b>	Pineal gland <b>C753</b>
<b>Pituicytoma 9432/1*</b>	Pituitary gland <b>C751</b> , sella turcica <b>C751</b> , suprasellar <b>C719</b>
<b>Pituitary adenoma 8272/0</b>	Pituitary gland <b>C751</b>
<b>Prolactinoma 8271/0</b>	Pituitary gland <b>C751</b>
<b>Subependymal giant cell tumor (SEGA) 9384/1</b>	Lateral ventricles <b>C715</b>
<b>Subependymoma 9383/1</b>	Intraventricular site (lateral/third ventricle, rare <b>C715</b> and IV ventricle <b>C717</b> )



**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
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**Table 6: Specific Histologies, NOS, and Subtypes/Variants**

Use this table to identify **reportable** histologies, **including specific, NOS**, and the **subtype/variant** of that NOS when referenced in the Multiple Primary and Histology coding rules.

This table includes the more common non-malignant histologies which occur in the CNS.

**Column 1** contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants**.

**Column 2** contains **synonyms** for the specific or NOS term. Synonyms have the **same** histology **code** as the specific or NOS term.

**Column 3** contains **subtypes/variants** of the **NOS** histology. Subtypes/variants **do not** have the **same** histology code as the NOS term.

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants Histology Term and Codes
<b>Angiocentric glioma 9431/1*</b>	Angiocentric neuroepithelial tumor Monomorphous angiocentric glioma	
<b>Benign fibrous histiocytoma 8830/0</b>		
<b>Chondroma 9220/0</b>		
<b>Chordoid glioma of the third ventricle 9444/1</b>		
<b>Choroid plexus papilloma 9390/0</b>		Atypical choroid plexus papilloma <b>9390/1</b>
<b>Craniopharyngioma 9350/1</b>		Adamantinomatous craniopharyngioma <b>9351/1</b> Papillary craniopharyngioma <b>9352/1</b>
<b>Desmoplastic infantile astrocytoma and ganglioglioma 9412/1</b>	DIAG	

**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

<b>NOS/Specific Histology Term and Code</b>	<b>Synonyms</b>	<b>Subtypes/Variants Histology Term and Codes</b>
<b>Diffuse astrocytoma, MYB- or MYBL1 altered 9421/1</b>  <i>Note 1:</i> Beginning 1/1/2023, diffuse astrocytoma, MYB- or MYBL1 altered is the preferred term for 9421/1.  <i>Note 2:</i> Beginning 1/1/2023, pilocytic astrocytoma/juvenile pilocytic astrocytoma are coded 9421/1. Cases diagnosed prior to 1/1/2023 are coded 9421/3.	Angiocentric glioma Diffuse low-grade glioma, MAPK pathway-altered Juvenile pilocytic astrocytoma Pilocytic astrocytoma	
<b>Dysembryoplastic neuroepithelial tumor 9413/0</b>  <i>Note:</i> DNET and PLNTY have the same ICD-O code but are distinctly different histologies. Because they are different, they are on separate rows in column 1.	DNET	
<b>Gangliocytoma 9492/0</b>		Dysplastic cerebellar gangliocytoma/Lhermitte-Duclos disease <b>9493/0</b>
<b>Ganglioglioma 9505/1</b>		
<b>Granular cell tumor of the sellar region 9582/0</b>		
<b>Hemangioblastoma 9161/1</b>	Capillary hemangioblastoma	
<b>Hemangioma 9120/0</b>	Cavernoma	Cavernous hemangioma <b>9121/0</b> Venous hemangioma <b>9122/0</b>
<b>Juvenile xanthogranuloma 9749/1</b>		
<b>Leiomyoma 8890/0</b>		
<b>Lipoma 8850/0</b>		Hibernoma <b>8880/0</b>
<b>Meningeal melanocytosis 8728/0</b>		Meningeal melanocytoma <b>8728/1</b>

**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants Histology Term and Codes
<b>Meningioma 9530/0</b>	Lymphoplasmacyte-rich meningioma Metaplastic meningioma Microcystic meningioma Secretory meningioma	Angiomatous meningioma <b>9534/0</b> Atypical meningioma <b>9539/1</b> Clear cell/chordoid meningioma <b>9538/1</b> Fibrous meningioma <b>9532/0</b> Meningothelial meningioma <b>9531/0</b> Psammomatous meningioma <b>9533/0</b> Transitional meningioma <b>9537/0</b>
<b>Multinodular and vacuolating neuronal tumor 9509/0</b>  <i>Note:</i> MVNT and papillary glioneuronal tumor have the same ICD-O code but are distinctly different histologies based on behavior codes. Because they are different, they are on separate rows in column 1.	MVNT	
<b>Myofibroblastoma 8825/0</b>		Inflammatory myofibroblastic tumor <b>8825/1</b>
<b>Myxopapillary ependymoma 9394/1</b>		
<b>Neurocytoma 9506/1</b>	Central neurocytoma Cerebellar liponeurocytoma Extraventricular neurocytoma Lipomatous medulloblastoma Medullocytoma Neurolipocytoma	

**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
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NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants Histology Term and Codes
<p><b>Neuroepithelial tumor, benign 8000/0</b></p> <p><i>Note:</i> Neuroepithelial tumor is a rare tumor specific to children. These neoplasms have numerous subtypes which are not easily identified so a specific type may not be provided in the pathology report. WHO has not proposed an ICD-O code for this entity. The current option is to assign code 8000. Because these tumors are different, they are on a separate row.</p>	<p>Neuroepithelial tumor, NOS 8000/1</p>	
<p><b>Neurofibroma 9540/0</b></p>	<p>Atypical neurofibroma</p>	<p>Plexiform neurofibroma 9550/0</p>
<p><b>Optic glioma/pilocytic astrocytoma 9421/1</b></p>		
<p><b>Papillary glioneuronal tumor 9509/1</b></p> <p><i>Note 1:</i> MVNT and papillary glioneuronal tumor have the same ICD-O code but are distinctly different histologies based on behavior codes. Because they are different, they are on rows in column 1.</p> <p><i>Note 2:</i> Beginning with cases diagnosed 1/1/2023 forward, leptomenigeal glioneuronal tumor is coded 9509/3. See the Malignant CNS rules.</p>	<p>Diffuse leptomenigeal glioneuronal tumor (see note 2)  Rosette-forming glioneuronal tumor</p>	
<p><b>Paraganglioma 8693/1</b></p>		
<p><b>Perineurioma 9571/0</b></p>		
<p><b>Pineocytoma 9361/1</b></p>		
<p><b>Pituicytoma 9432/1*</b></p>		

**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

<b>NOS/Specific Histology Term and Code</b>	<b>Synonyms</b>	<b>Subtypes/Variants Histology Term and Codes</b>
<b>Pituitary adenoma 8272/0</b>	Corticotroph Gonadotroph adenoma Somatotroph adenoma Thyrotroph adenoma Null cell adenoma Plurihormonal and double adenomas	
<b>Polymorphous low-grade neuroepithelial tumor of the young 9413/0</b>  <i>Note:</i> DNET and PLNTY have the same ICD-O code but are distinctly different histologies. Because they are different, they are on separate rows in column 1.	PLNTY	
<b>Prolactinoma 8271/0</b>		
<b>Rhabdomyoma 8900/0</b>		
<b>Schwannoma 9560/0</b>	Acoustic neuroma Cellular schwannoma Neurilemoma Neurinoma Plexiform schwannoma	Melanotic schwannoma <b>9560/1*</b>
<b>Solitary fibrous tumor Grade 1 8815/0</b>	Hemangiopericytoma Grade 1	Solitary fibrous tumor/hemangiopericytoma Grade 2 <b>8815/1*</b>
<b>Spindle cell oncocytoma 8290/0</b>		
<b>Subependymal giant cell astrocytoma 9384/1</b>		
<b>Subependymoma 9383/1</b>		
<b>Teratoma 9080/1</b>		

**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Section 3: Additional Information to Complete Abstract**

**Conflicting Information on Pathology Report(s)**

The **classification** of brain tumors is a **subjective** matter because definitive criteria have not been established/accepted. Pathologists may **disagree** on the histology or behavior.

Follow the steps below for coding the histology and/or behavior of tumor when there are discrepancies:

- When possible, get advice from the **pathologist**
- When possible, contact **attending** physician
- When possible, consult with **registry advisor**
- If none of those **options** are **available**, code the histology and grade from **the most dependable source** (see [Priority List for Coding Histology](#)).

The following are examples of how conflicting information occurs in single and in multiple pathology reports

- **Single pathology report:**
  - **Multiple pathologists** within the institution review the slides
  - Slides are sent for **outside review** and the information from the **consulting** lab **conflicts** with the **original** pathology report
- **Multiple pathology reports:** The **first report** is from a biopsy and the **second report** is from a resection. Code the histology and/or behavior from the resection.

**Non-Malignant CNS Equivalent Terms and Definitions**  
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**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Table 7: Paired Sites**

Use **Table 7** to identify sites for which laterality **must** be coded. Do **not** use this table to determine multiple primaries.

<b>Paired Sites and Codes</b>
Acoustic nerve <b>C724</b>
Cerebral meninges <b>C700</b>
Cerebrum <b>C710</b>
Cranial nerves <b>C725</b>
Frontal lobe <b>C711</b>
Occipital lobe <b>C714</b>
Olfactory nerve <b>C722</b>
Optic nerve <b>C723</b>
Parietal lobe <b>C713</b>
Temporal lobe <b>C712</b>

**Note 1:** Midline tumors are most common for the following histologies: glioblastoma multiforme, meningiomas, lymphomas (usually located near the midline), and epidermoid cysts (which can cross the midline via the subarachnoid space).

**Note 2:** SEER allows laterality to be coded for sites other than those in the table.

**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Table 8: Non-Malignant CNS Tumors with the Potential of Transforming to Malignant Behavior**

The word “**transformation**” as used in this table means that:

- Residual tumor becomes **more aggressive /3 OR**
- The tumor **recurs** as a **more aggressive /3 histology**

The table identifies non-malignant tumors that have the **potential of transforming** to a malignant tumor (**new primary**).

**Column 1** is the **non-malignant** ICD-O histology term and code.

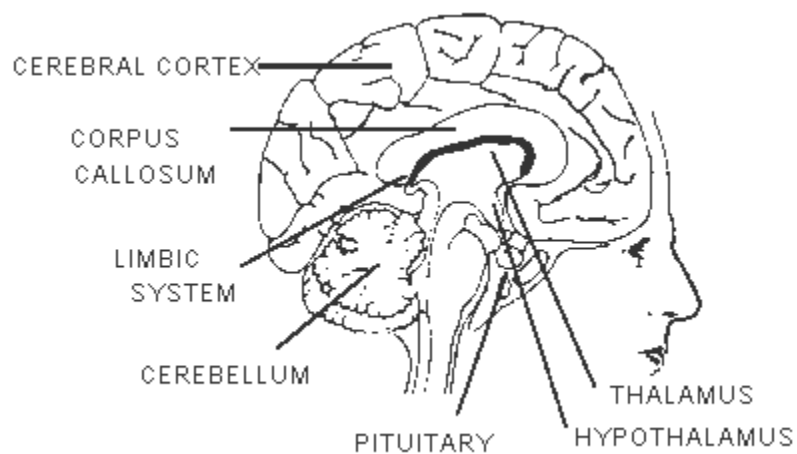
**Column 2** is the **malignant /3** ICD-O histology term and code to which the non-malignant tumor can transform.

<b>Original Histology and Code</b>	<b>Transformed Histology and Code</b>
Chondroma <b>9220/0</b>	Chondrosarcoma <b>9220/3</b>
Ganglioglioma <b>9505/1</b>	Anaplastic ganglioglioma <b>9505/3</b>
Hemangioma <b>9120/0</b>	Angiosarcoma <b>9120/3</b>
Hemangiopericytoma <b>9150/1</b>	Anaplastic hemangiopericytoma <b>9150/3</b>
Leiomyoma <b>8890/0</b>	Leiomyosarcoma <b>8890/3</b>
Lipoma <b>8850/0</b>	Liposarcoma <b>8850/3</b>
Osteoma <b>9180/0</b>	Osteosarcoma <b>9180/3</b>
Perineurioma <b>9571/0</b>	Malignant perineurioma <b>9571/3</b>
Rhabdomyoma <b>8900/0</b>	Rhabdomyosarcoma <b>8900/3</b>
Teratoma <b>9080/1</b>	Immature teratoma <b>9080/3</b>
Teratoma, mature <b>9080/0</b>	Immature teratoma <b>9080/3</b>

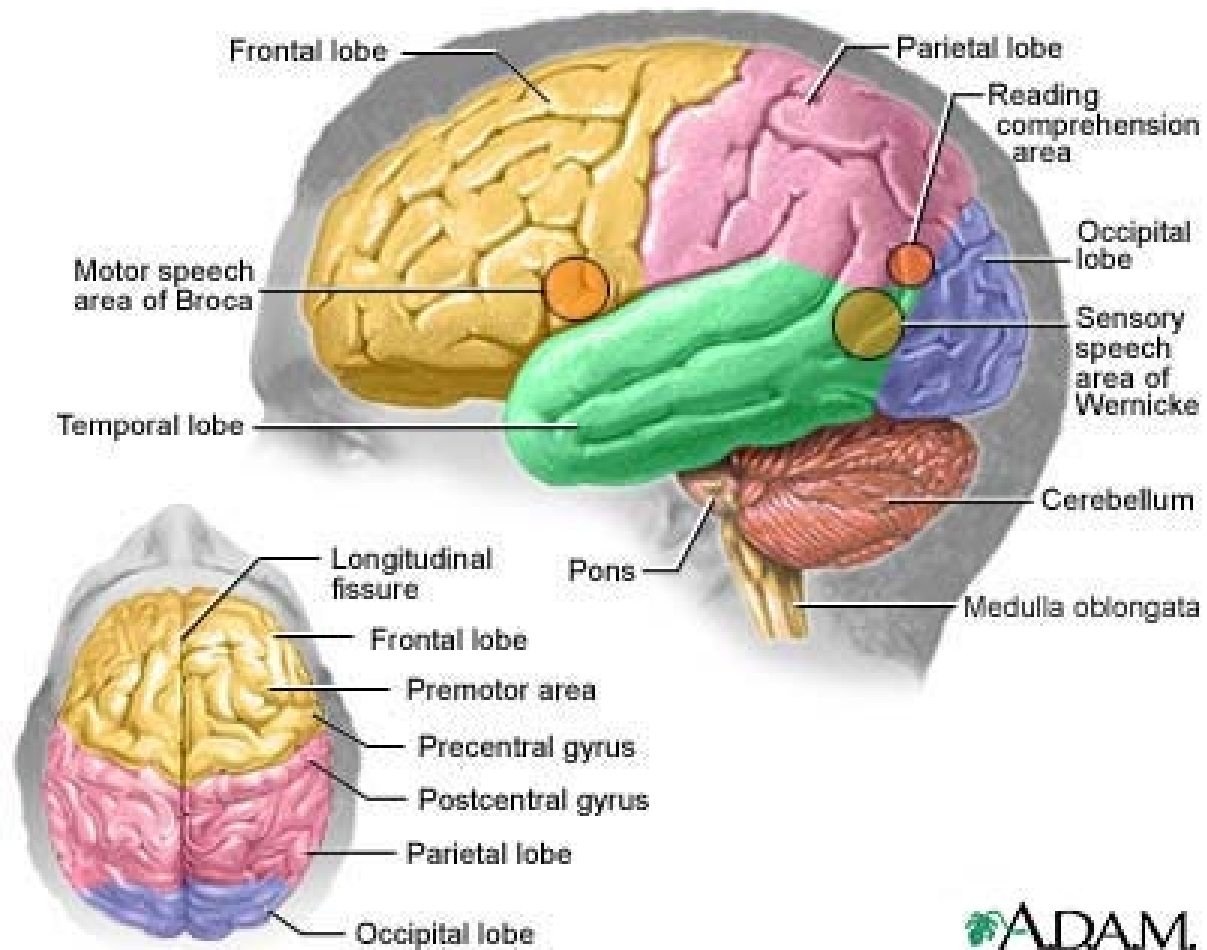


**Non-Malignant CNS Equivalent Terms and Definitions**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Illustrations**



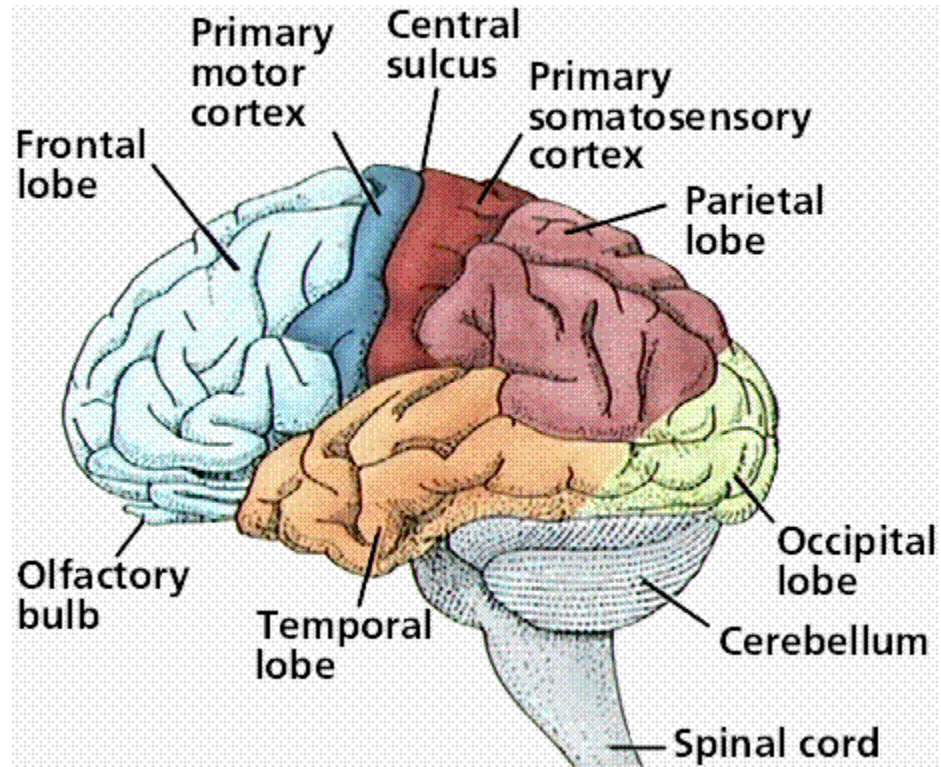
**Non-Malignant CNS Equivalent Terms and Definitions**  
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 ADAM.

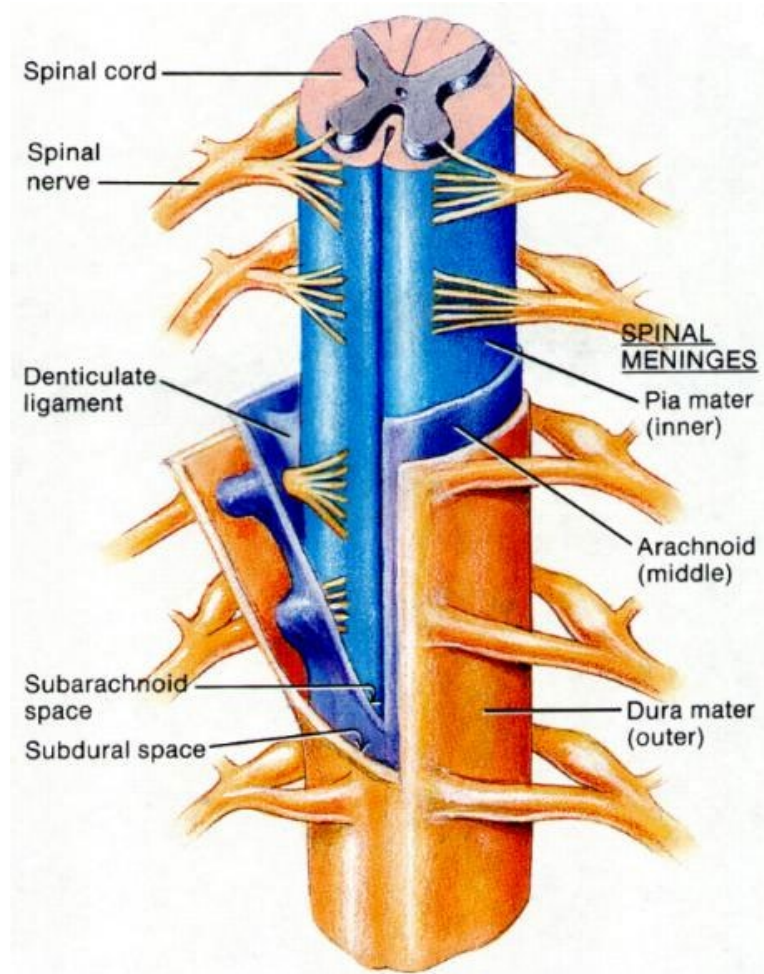
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**Non-Malignant CNS Equivalent Terms and Definitions**  
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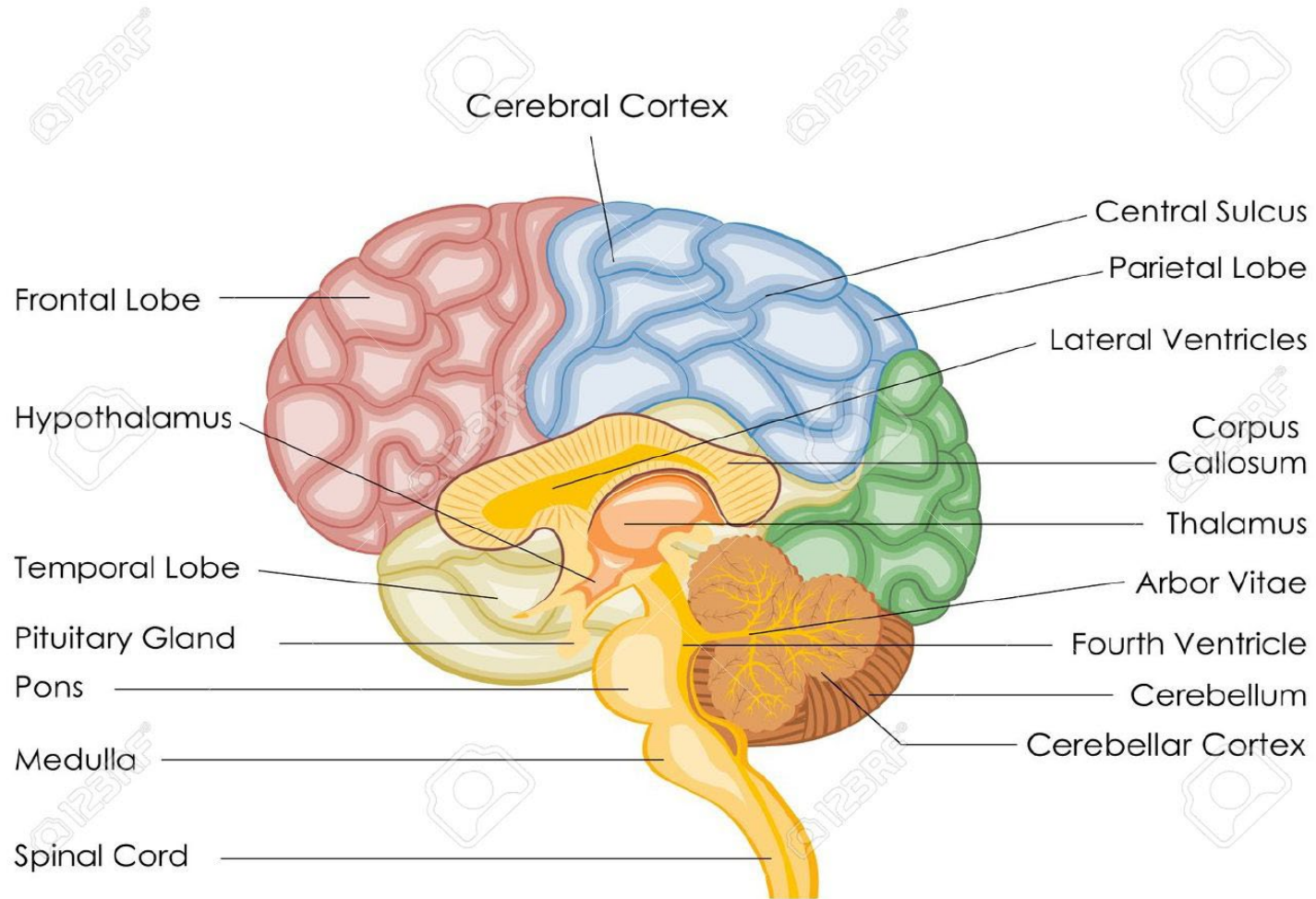


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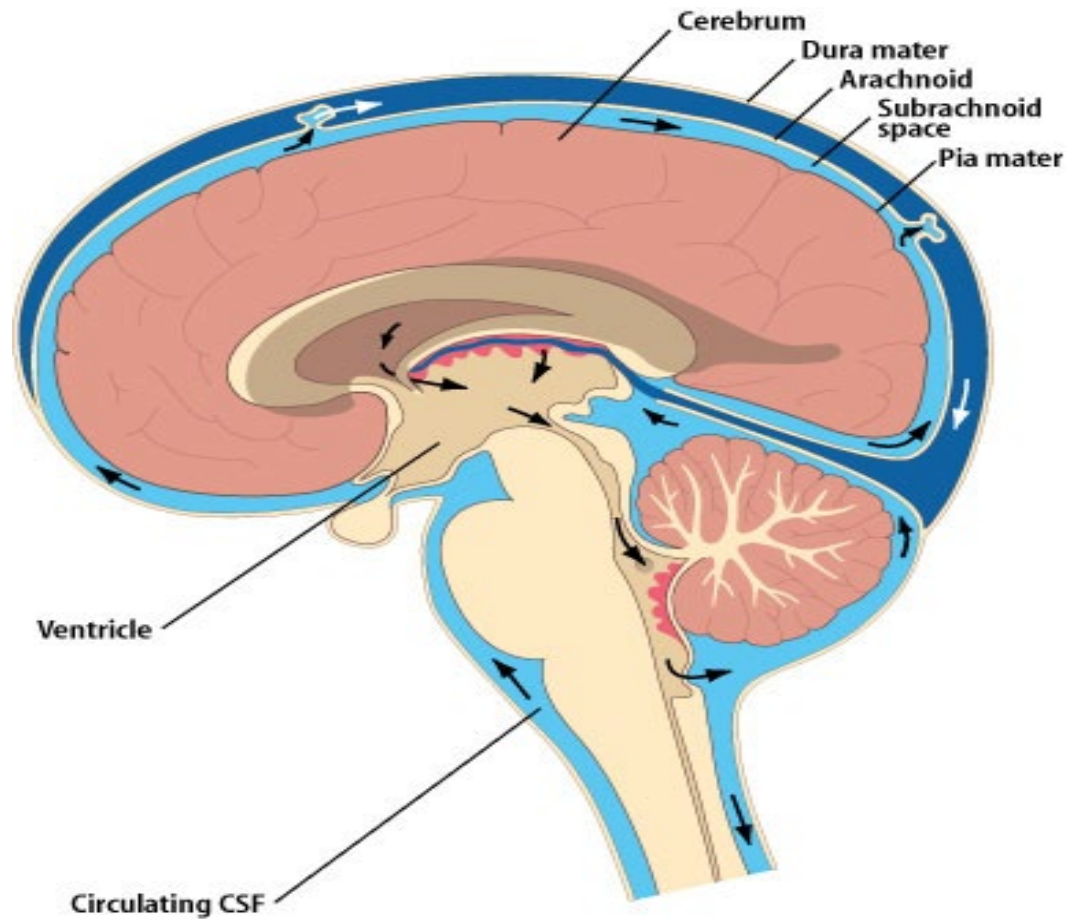
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**Non-Malignant CNS Equivalent Terms and Definitions**  
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**Non-Malignant CNS Equivalent Terms and Definitions**  
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**Non-Malignant CNS Multiple Primary Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Note 1:** **Timing** is **not used** to determine multiple primaries.

**Note 2:** **Laterality** is **not used** to determine multiple primaries.

**Note 3:** **Malignant** central nervous system (CNS) tumors have a separate set of rules.

**Note 4:** 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

- Tumors diagnosed 01/01/2007 through 12/31/2017: Use 2007 MPH Rules
- Tumors diagnosed 01/01/2018 and later: Use 2018 Solid Tumor Rules
- The original tumor diagnosed before 1/1/2018 and a subsequent tumor diagnosed 1/1/2018 or later **in the same primary site**: Use the 2018 Solid Tumor Rules.

**Unknown if Single or Multiple Tumors**

**Rule M1** Abstract a **single primary**<sup>i</sup> when it is not possible to determine if there is a **single** tumor or **multiple** tumors.

**Note 1:** Use this rule only after all information sources have been exhausted

**Note 2:** Examples of cases with minimal information include

- Death certificate only (DCO)
- Cases for which information is limited to pathology report only
- Outpatient biopsy with no follow-up information available
- Multiple pathology reports which do not specify whether a single tumor or multiple tumors have been biopsied and/or resected

**This is the end of instructions for Unknown if Single or Multiple Tumors.**

<sup>i</sup> Prepare one abstract. Use the [histology rules](#) to assign the appropriate histology code.

**Non-Malignant CNS Multiple Primary Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Single Tumor**

**IMPORTANT:** The **major difference** between **M3** and **M5** is:

**M3: No resection** as first course of treatment **AND** when the same tumor is subsequently **resected**, **pathology proves malignant** behavior

**M5: Tumor resected** as first course of treatment. Subsequent tumor (**recurrence** or **de novo**) is **malignant**

**Rule M2** Abstract a **single primary**<sup>i</sup> when there is a **single tumor**.

*Note 1:* A single tumor is **always** a single primary.

*Note 2:* The tumor may overlap onto or extend into an adjacent/contiguous site or subsite.

*Note 3:* The tumor may have multiple histologic components.

*Note 4:* The tumor may be diagnosed as borderline /1 on scans and subsequent pathology proves a benign /0 tumor.

*Example:* Patient has a MRI with a diagnosis of a benign /0 tumor. The pathology report from the subsequent surgery shows a borderline /1 tumor. This is a single tumor and a **single primary**. (It is the same tumor as seen on the MRI; better information is provided by the pathology report.)

**Rule M3** Abstract a **single primary**<sup>i</sup> (the malignant) when a single tumor meets the following two criteria:

1. The original diagnosis was non-malignant /0 or /1 **AND**

- First course treatment was active surveillance (no tumor resection). Diagnosis was:
  - o Clinical
  - o Radiographic
  - o Stereotactic biopsy

2. Subsequent resection pathology is malignant /3

*Note 1:* This is a **new rule** which **clarifies** that a single tumor is **always** a single primary and the malignant behavior is reported.

*Note 2:* Use the Malignant CNS and Peripheral Nerves Rules to code histology.

*Note 3:* The **resection pathology** is **more accurate** than clinical, radiographic or stereotactic biopsy information. While stereotactic biopsy provides a pathologic specimen, it is small and may not have included the malignant portion of tumor.

*Note 4:* There is **no time requirement** from initial diagnosis to resection.

*Note 5:* Edit the original abstract as follows:

- Do **not** change **date of diagnosis**.
- For cases which have been abstracted, **change behavior** code on original abstract from /0 or /1 to /3.
- **Report** all data changes for cases which have been submitted to the central registry.



**Non-Malignant CNS Multiple Primary Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

- See the COC and [SEER manuals](#) for instructions on coding other data items such as Accession Year, Treatment and Sequence Number.

**Note 6:** The physician may have staged the non-malignant tumor at the time of original diagnosis (benign). The physician will also stage the malignant tumor. Staging and determining multiple primaries are done for different reasons. Staging determines which treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).

**Example 1:** A patient is diagnosed by MRI with **ganglioglioma 9505/1** in July 2009. After months of active surveillance (watchful waiting), the patient becomes symptomatic. A resection is done in April 2010; the resection pathology is **anaplastic ganglioglioma 9505/3**. Change behavior code on the original abstract from /1 to /3. Do not change date of diagnosis.

**Example 2:** A November 15, 2016 MRI and subsequent stereotactic biopsy of lateral ventricle of brain show a **mature teratoma 9080/1**. The patient becomes symptomatic in 2017 and the neoplasm is resected on October 31, 2017. The resection pathology diagnoses **immature teratoma 9080/3**. Change behavior code on the original abstract. Do not change date of diagnosis.

**Rule M4** Abstract a **single primary**<sup>i</sup> when a single **benign** tumor /0 **transforms** to an **uncertain/borderline** tumor /1. Timing is irrelevant. The tumors are:

- The **same histology OR**
- A **NOS** and a **subtype/variant** of that NOS

**Note 1:** Do not change the date of diagnosis **OR** the behavior code on the original abstract.

**Note 2:** This is a single tumor; single primary

**Note 3:** Both behaviors /0 and /1 are non-malignant; it is not necessary to change the original abstract.

**Note 4:** The physician may stage both tumors. Staging and determining multiple primaries are done for different reasons. Staging determines which treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).

**Note 5:** For registries that collect recurrence data, document the transformed tumor as a recurrence.

**Example 1:** A choroid plexus papilloma NOS **9390/0 transforms** to an atypical choroid plexus papilloma **9390/1**. This is a **single primary**.

**Example 2:** A meningioma **9530/0 transforms** to an atypical meningioma **9539/1**.

**This is the end of instructions for Single Tumor.**

<sup>i</sup> Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.

**Non-Malignant CNS Multiple Primary Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Multiple Tumors**

**Note 1:** Multiple tumors may be a single primary or multiple primaries.

**Note 2:** Separate, non-contiguous tumors are always multiple primaries when:

- In the CNS (see [Table 2](#)) AND in a site other than the CNS

**Example:** Patient has a non-malignant meningioma of the cerebral meninges and adenocarcinoma of the lung. The lung is not a CNS site. Abstract two primaries.

- In different CNS sites (see Rule M7)

**IMPORTANT:** The major difference between **M3** and **M5** is:

**M3:** No resection as first course of treatment AND when the same tumor is subsequently resected, pathology proves malignant behavior

**M5:** Tumor resected as first course of treatment. Subsequent tumor (recurrence or de novo) is malignant

**Rule M5** Abstract **multiple primaries**<sup>ii</sup> when a **malignant** tumor /3 occurs **after** a **non-malignant** tumor /0 or /1 **AND:**

- The patient had a **resection** of the non-malignant tumor **OR**
- It is **unknown/not documented** whether a resection was done

**Note:** Abstract the second tumor (malignant) using the Malignant CNS rules.

**Rule M6** Abstract a **single primary**<sup>i</sup> when the patient has **bilateral:**

- **Acoustic neuromas/ vestibular schwannomas 9560/0**
- **Optic gliomas/pilocytic astrocytomas 9421/1**

**Note 1:** The bilateral tumors may appear simultaneously (at the same time) **OR** the contralateral tumor may be diagnosed at any time following the original diagnosis.

**Note 2:** WHO and IARC designate pilocytic astrocytoma as a synonym for optic glioma. When the primary site is optic nerve, the behavior is non-malignant.

**Note 3:** When the bilateral tumors are diagnosed at different times, the physician **may stage each tumor** because staging and determining multiple primaries are done for different reasons. Staging determines which course of treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).

**Non-Malignant CNS Multiple Primary Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Rule M7** Abstract **multiple primaries**<sup>ii</sup> when multiple tumors are present in any of the following sites:

- Any lobe(s) of the brain **C710-C719 AND** any other part of CNS
- Cauda equina **C721 AND** any other part of CNS
- Cerebral meninges **C700 AND** spinal meninges **C701**
- Cerebral meninges **C700 AND** any other part of CNS
- Any cranial nerve(s) **C722-C725 AND** any other part of the CNS
- Meninges of cranial nerves **C709 AND** any other part of the CNS
- Spinal cord **C720 AND** any other part of CNS
- Spinal meninges **C701 AND** any other part of CNS

**Rule M8** Abstract **multiple primaries**<sup>ii</sup> when separate, non-contiguous tumors are **two** or more different **subtypes/variants** in Column 3, **Table 6** in the Equivalent Terms and Definitions. Timing is irrelevant.

*Note:* The tumors may be subtypes/variants of the **same** or **different** NOS histologies.

- **Same NOS:** Atypical meningioma 9539/1 and fibrous meningioma 9532/0 are both subtypes of meningioma NOS 9530 but are distinctly different histologies. Abstract multiple primaries.
- **Different NOS:** Melanotic schwannoma 9560/1 is a subtype of schwannoma NOS 9560/0; papillary craniopharyngioma 9352/1 is a subtype of craniopharyngioma 9350/1. They are distinctly different histologies. Abstract multiple primaries.

**Rule M9** Abstract a **single primary**<sup>i</sup> when two or more separate/non-contiguous **meningiomas** arise in the cranial meninges. Laterality is irrelevant and may be any of the following combinations:

- The same laterality (left or right) of the cranial meninges
- Bilateral (both left and right) cranial meninges
- The midline **AND** in either the right or left cranial meninges

*Note:* This rule applies **ONLY** to **meningiomas** that are either a **NOS** and **subtype/variant**, OR they are the **same histology**.

**Non-Malignant CNS Multiple Primary Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Rule M10** Abstract a **single primary**<sup>i</sup> when there are separate/non-contiguous tumors in the brain (multicentric/multifocal) with the **same histology XXXX**. Tumors may be in any of the following locations and/or lateralities:

- Same laterality: In the same lobe; for example, two tumors in right temporal lobe **C712** (same site code)
- Different lateralities of the same lobe; for example, left and right frontal lobes **C711** (same site code)
- Different lobes; for example, parietal lobe **C713** and occipital lobe **C714** (different site codes)

**\*Exception 1:** MVNT and papillary glioneuronal tumor have the same ICD-O code but are distinctly different histologies based on behavior codes. Because they are different, they are on separate rows in Table 6.

**\*Exception 2:** DNET and PLNTY have the same ICD-O code but are distinctly different histologies. Because they are different, they are on separate rows in Table 6.

**Note 1:** Metastases are never used to determine multiple primaries. Seeding metastasis is often noted in ependymomas.

**Note 2:** This is a **change** from/**clarification** to **previous** rules.

**Note 3:** These rules are hierarchical. Use this rule **ONLY** when the previous rules do not apply.

**Note 4:** An example of a non-malignant brain tumor that may be multi-focal/multi-centric is hemangioblastoma **9161/1**.

**Note 5:** The physician may **stage each tumor** because staging and determining multiple primaries are done for different reasons. Staging determines which course of treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).

**Rule M11** Abstract a **single primary**<sup>i</sup> when separate/non-contiguous tumors are on the **same row** in **Table 6** in the Equivalent Terms and Definitions. Timing is irrelevant.

**Note:** The same row means the tumors are:

- The same histology\* (same four-digit ICD-O code) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3). NOS and subtype/variants are:
  - o Choroid plexus papilloma **9390/0** and a subtype/variant of choroid plexus papilloma
  - o Craniopharyngioma **9350/1** and a subtype/variant of craniopharyngioma
  - o Gangliocytoma **9492/0** and a subtype/variant of gangliocytoma
  - o Lipoma **8850/0** and a subtype/variant of lipoma
  - o Meningeal melanocytosis **8728/0** and a subtype/variant of meningeal melanocytosis
  - o Meningioma **9530/0** and a subtype/variant of meningioma

**Non-Malignant CNS Multiple Primary Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

- o Myofibroblastoma **8825/0** and a subtype/variant of myofibroblastoma
- o Neurofibroma **9540/0** and a subtype/variant of neurofibroma
- o Schwannoma **9560/0** and a subtype/variant of schwannoma
- o Solitary fibrous tumor WHO Grade 1 **8815/0** and a subtype/variant of solitary fibrous tumor WHO Grade 1

**\*Exception 1:** MVNT and papillary glioneuronal tumor have the same ICD-O code but are distinctly different histologies based on behavior codes. Because they are different, they are on separate rows in Table 6.

**\*Exception 2:** DNET and PLNTY have the same ICD-O code but are distinctly different histologies. Because they are different, they are on separate rows in Table 6.

**Rule M12** Abstract **multiple primaries**<sup>ii</sup> when separate/non-contiguous tumors are on **different rows** in **Table 6** in the Equivalent Terms and Definitions. Timing is irrelevant.

**Note 1:** Each row in the table is a **distinctly different** histology.

**Note 2:** MVNT and papillary glioneuronal tumor have the same ICD-O code but are distinctly different histologies based on behavior codes. Because they are different, they are on different rows in Table 6.

**Note 3:** DNET and PLNTY have the same ICD-O code but are distinctly different histologies. Because they are different, they are on separate rows in Table 6.

**Note 4:** 8000 is considered a different row **ONLY** when the diagnosis is neuroepithelial tumor. If the diagnosis is cancer, NOS, do not consider 8000 to be a separate row from other histologies for the purpose of the table rules.

- Example 1: A tumor is diagnosed as 8000/1 Neuroepithelial tumor, NOS. Later, a separate tumor is diagnosed as Hemangioma 9120/0. These are considered separate rows.
- Example 2: A tumor has a provisional diagnosis of 8000/0 and further diagnosis is done. A subsequent tumor in another lobe of the brain is diagnosed as myofibroblastoma 8825/0. These are not considered separate rows.

**Rule M13** Abstract a **single primary**<sup>i</sup> when the tumors do not meet any of the above criteria.

**Note:** These rules are hierarchical. Use this rule **ONLY** when the previous rules do not apply.

**This is the end of instructions for Multiple Tumors**

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<sup>i</sup> Prepare one abstract. Use the histology coding rules to assign the appropriate histology code. For registries collecting recurrence data: When a subsequent tumor is “single primary,” record that subsequent tumor as a recurrence.

<sup>ii</sup> Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.

**Non-Malignant CNS Histology Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

- Note 1:** Non-malignant central nervous system (CNS) neoplasms (previously called benign and borderline) are **reportable** for cases **diagnosed 1/1/2004** and later.
- Note 2:** **Malignant** central nervous system (CNS) tumors have a **separate** set of **rules**.
- Note 3:** These rules are **not used** for tumor(s) or neoplasm(s) described as **metastatic/metastasis**.
- Note 4:** For rules specifying a NOS and a **subtype/variant** of the NOS, the NOS may be the preferred/most common term **OR** any of the **synonyms** for the NOS.

**Priority Order for Using Documentation to Identify Histology**

**IMPORTANT NOTES**

1. Code the histology diagnosed **prior** to **neoadjuvant treatment**.

**Note 1:** Histology changes may occur following immunotherapy, chemotherapy, targeted therapy, and radiation therapy.

**Note 2:** Neoadjuvant treatment is any tumor-related treatment given prior to surgical removal of the malignancy.

**Exception:** If the initial diagnosis is based on histology from **FNA, smears, cytology**, or from a regional or metastatic site, and neoadjuvant treatment is given and followed by resection of primary site which identifies a different or specific histology, code the histology from the primary site.

2. Code the histology using the following priority list and the Histology Rules. Do not change histology in order to make the case applicable to staging.

The priority list is used for single primaries (including multiple tumors abstracted as a single primary).

**This is a hierarchical list of source documentation:**

1. **Pathology/tissue** from **resection**
  - A. The addendum and/or comments
  - B. Final diagnosis / synoptic report as required by CAP
  - C. CAP protocol
  - D. Biomarkers
    - Biomarkers do not identify all histologic types.

**Non-Malignant CNS Histology Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

- Biomarkers are not listed because they change rapidly.

**Example:** BRAF will help determine whether a tumor is a pilocytic astrocytoma or a non-pilocytic astrocytoma.

**Note 1:** Addendums and comments on the pathology report are given the highest priority because they often contain information about molecular testing, genetic testing, and/or special stains which give a more specific diagnosis.

**Note 2:** The pathologist's diagnosis is always the most reliable, so the final diagnosis is the second priority.

**Note 3:** **Do not** use the microscopic or gross section of the pathology report for coding.

**Note 4:** The CAP protocol is a checklist which

- Provides guidelines for collecting the essential data elements for complete reporting of tumors and optimal patient care.
- Allows physicians to check multiple histologies

2. Pathology/tissue from **biopsy**

- A. The addendum and/or comments
- B. Final diagnosis / synoptic report as required by CAP
- C. CAP protocol
- D. Biomarkers

- Biomarkers do not identify all histologic types.
- Biomarkers are not listed because they change rapidly.

**Example:** BRAF will help determine whether a tumor is a pilocytic astrocytoma or a non-pilocytic astrocytoma.

**Note 1:** Addendums and comments on the pathology report are given the highest priority because they often contain information about molecular testing, genetic testing, and/or special stains which give a more specific diagnosis.

**Note 2:** The pathologist's diagnosis is always the most reliable, so the final diagnosis is the second priority.

**Note 3:** **Do not** use the microscopic or gross section of the pathology report for coding.

**Note 4:** The CAP protocol is a checklist which

- Provides guidelines for collecting the essential data elements for complete reporting of tumors and optimal patient care.
- Allows physicians to check multiple histologies

3. **Cytology** (most frequently spinal fluid)

4. **Radiography:** The following list is in priority order.

- A. MRI
- B. CT
- C. PET
- D. Angiogram

**Non-Malignant CNS Histology Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

5. **Clinical Diagnosis:** Code the histology documented by the physician when none of the above are **available**. **Priority** for using documentation:
- A. Treatment plan
  - B. Documentation from Tumor Board
  - C. References to pathology diagnosis
  - D. Physician's reference to type of cancer (histology) in the medical record
- Note:* Code the **specific** histology when documented.



**Non-Malignant CNS Histology Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Coding Histology**

**Note 1:** The priority is to code the most specific histology. **DO NOT USE BREAST HISTOLOGY CODING RULES FOR THIS SITE.**

**Note 2:** Only use this section for one or more histologies within a single tumor.

**Note 3:** Do not use this section in place of the Histology Rules.

1. **Code the most specific histology or subtype/variant, regardless of whether it is described as:**

- A. The majority or predominant part of tumor
- B. The minority of tumor
- C. A component

**Example 1:** Diagnosis for a single tumor is choroid plexus papilloma 9390/0 with the majority or predominant part of tumor being atypical choroid plexus papilloma 9390/1. Code the subtype/variant: atypical choroid plexus papilloma 9390/1.

**Example 2:** Diagnosis for a single tumor is meningioma 9530/0 with minority of tumor being atypical meningioma 9539/1. Code the subtype/variant: atypical meningioma 9539/1.

**Example 3:** Diagnosis for a single tumor is schwannoma 9560/0 with a component of melanotic schwannoma 9560/1. Code the subtype/variant: melanotic schwannoma 9560/1.

**Note:** When the most specific histology is described as differentiation or features, see #2.

2. Code the histology described as **differentiation** or **features/features of ONLY** when there is a specific ICD-O code for the “NOS with \_\_\_ features” or “NOS with \_\_\_ differentiation”.

**Note:** Do not code differentiation or features when there is no specific ICD-O code.

3. Code the specific histology described by ambiguous terminology (list follows) **ONLY** when A or B is true:

- A. The only diagnosis available is **one histology** term described by ambiguous terminology
  - CoC and SEER require reporting of cases diagnosed only by ambiguous terminology
  - Case is accessioned (added to your database) based on ambiguous terminology and no other histology information is available/documented

**Non-Malignant CNS Histology Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Example:** Outpatient biopsy says probably central neurocytoma. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology central neurocytoma. The case meets the criteria in #3A.

- B. There is a **NOS histology and a more specific** (subtype/variant) described by ambiguous terminology
- Specific histology is clinically confirmed by a physician (attending, pathologist, oncologist, etc.) **OR**
  - Patient is receiving treatment based on the specific histology described by ambiguous term

**Example 1:** The pathology diagnosis is craniopharyngioma consistent with papillary craniopharyngioma. The oncology consult says the patient has papillary craniopharyngioma of the sellar region. This is clinical confirmation of the diagnosis, code papillary craniopharyngioma. The case meets the criteria in bullet 1.

**Example 2:** The pathology diagnosis is meningioma consistent with meningotheial meningioma. The treatment plan says the patient will receive the following treatment for meningotheial meningioma. Treatment plan confirms meningotheial meningioma; code meningotheial meningioma. The case meets the criteria in bullet 2.

**If the specific histology does not meet the criteria in #3B, then code the NOS histology.**

List of Ambiguous Terminology

Apparently	Most likely
Appears	Presumed
Comparable with	Probable
Compatible with	Suspect(ed)
Consistent with	Suspicious (for)
Favor(s)	Typical (of)
Malignant appearing	

4. **Do not code** histology when described as:

- Architecture
- Foci; focus; focal
- Pattern

**Non-Malignant CNS Histology Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Single Tumor**

**Rule H1** Code meningioma **9530/0** when the diagnosis is any of the following:

- Benign meningioma
- Lymphoplasmacyte-rich meningioma
- Meningioma with no mention of behavior
- Metaplastic meningioma
- Microcystic meningioma
- Secretory meningioma
- Two meningioma subtypes (See [Table 6](#))

**Note:** **Do not report** a **malignant /3** meningioma based on:

- **Invasion** of the **skull bone**
- Tumor **extension through the foramina at the base of the skull**
- Do not report a malignant /3 meningioma based on tumor extension to brain

**Rule H2** Code the **reportable CNS tumor** ([Table 6](#) in the Equivalent Terms and Definitions) when a patient has any of the following:

- Neurofibromatosis type 1 (**NF1**)
- Neurofibromatosis type 2 (**NF2**)
- **Schwannomatosis**

**Note 1:** **Do not code** NF1 or NF2 as neurofibromatosis. NF1, NF2, and schwannomatosis are genetic syndromes which have a high risk of developing reportable and non-reportable tumors. **ONLY** abstract **reportable tumors** such as:

- Plexiform neurofibroma (usually NF1)
- Tumors of brain and spinal cord, meningiomas of cranial or spinal dura, glioma (usually NF2)

**Note 2:** Tumors are reportable when they meet the behavior (/0 or /1), site (within the CNS), and histology reportability requirements (see [Reportability Criteria](#)).

**Note 3:** Schwannomatosis is a newer term for a distinct subtype/variant of the genetic diseases NF1 and NF2.

**Example:** Patient presents with vestibular schwannoma (acoustic neuroma). Genetic testing proves the patient has NF2. Report the acoustic nerve neuroma.

**Non-Malignant CNS Histology Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Rule H3** Code the histology when only **one histology** is present.  
*Note 1:* Use [Table 6](#) to code histology. New codes, terms, and synonyms are included in [Table 6](#) and coding errors may occur if the table is not used.  
*Note 2:* When the histology is **not listed** in [Table 6](#) use the **ICD-O** and all **updates**.  
*Note 3:* Submit a question to [Ask a SEER Registrar](#) when the histology code is not found in [Table 6](#), ICD-O or all updates.

**Rule H4** Code the **subtype/variant** when there is a **NOS** and a **single subtype/variant** of that NOS, such as the following:

- Choroid plexus papilloma **9390/0** and a subtype/variant of choroid plexus papilloma
- Craniopharyngioma **9350/1** and a subtype/variant of craniopharyngioma
- Gangliocytoma **9492/0** and a subtype/variant of gangliocytoma
- Lipoma **8850/0** and a subtype/variant of lipoma
- Meningeal melanocytosis **8728/0** and a subtype/variant of meningeal melanocytosis
- Meningioma **9530/0** and a subtype/variant of meningioma
- Myofibroblastoma **8825/0** and a subtype/variant of myofibroblastoma
- Neurofibroma **9540/0** and a subtype/variant of neurofibroma
- Schwannoma **9560/0** and a subtype/variant of schwannoma
- Solitary fibrous tumor WHO Grade 1 **8815/0** and a subtype/variant of solitary fibrous tumor WHO Grade 1

*Note:* Use [Table 6](#) in the Equivalent Terms and Definitions to identify the NOS and subtypes/variants of the NOS.

**This is the end of instructions for Single Tumor.**

**Code the histology according to the rule that fits the case.**

**Non-Malignant CNS Histology Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Multiple Tumors Abstracted as a Single Primary**

**Rule H5** Code meningioma **9530/0** when there are multiple meningiomas with the following diagnosis:

- Benign meningioma
- Lymphoplasmacyte-rich meningioma
- Meningioma with no mention of behavior
- Metaplastic meningioma
- Microcystic meningioma
- Secretory meningioma

**Note:** **Do not report** a **malignant** /3 meningioma based on tumor extension/tumor expansion such as:

- **Invasion** of the skull bone
- Tumor **extension through the foramina at the base of the skull**
- Tumor **extension to brain**

**Rule H6** Code meningioma **9530/1** when there are multiple meningiomas of uncertain behavior.

**Note 1:** This is a rare condition that is usually associated with neurofibromatosis type 2 (NF2) and other genetic disorders.

**Note 2:** Use this code only for meningiomas with uncertain behavior; **do not use** this code for **multiple benign or malignant** meningiomas.

**Note 3:** It is **not necessary** for **all tumors** to be **biopsied** to use this code.

**Rule H7** Code the **reportable CNS tumor** (**Table 6** in the Equivalent Terms and Definitions) when a patient has any of the following:

- Neurofibromatosis type 1 (NF1)
- Neurofibromatosis type 2 (NF2)
- **Schwannomatosis**

**Note 1:** Only report tumors such as:

- Plexiform neurofibroma (usually NF1)
- Tumors of brain and spinal cord, meningiomas of cranial or spinal dura, glioma (usually NF2)

**Note 2:** Tumors are reportable when they meet the behavior code, site, and histology reportability requirements (see **Reportability Criteria**). Do not code neurofibromatosis.

**Note 3:** NF1 is a genetic disorder causing lesions in the skin, nervous system and skeleton.

**Non-Malignant CNS Histology Rules**  
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**  
**(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)**

**Note 4:** NF2 is a central form of the genetic disease and produces CNS tumors such as vestibular tumors, meningiomas, and ependymomas. When tumors meet the behavior code, site, and histology reportability requirements (see [Reportability Criteria](#)) those tumors are reportable.

**Note 5:** Schwannomatosis is a newer term for a distinct subtype/variant of the genetic diseases NF1 and NF2.

**Example:** Patient presents with bilateral vestibular schwannoma (acoustic neuroma). Genetic testing proves the patient has NF2. Report the acoustic nerve neuromas 9560/0.

**Rule H8** Code the histology when only **one histology** is present in all tumors.

**Note 1:** Use [Table 6](#) to code histology. New codes, terms, and synonyms are included in [Table 6](#) and coding errors may occur if the table is not used.

**Note 2:** When the histology is **not listed** in [Table 6](#) use the **ICD-O** and all **updates**.

**Note 3:** Submit a question to [Ask a SEER Registrar](#) when the histology code is not found in [Table 6](#), ICD-O or all updates.

**Rule H9** Code the **subtype/variant** when there is a **NOS** and a **single subtype/variant** of that NOS present in all tumors, such as the following:

- Choroid plexus papilloma **9390/0** and a subtype/variant of choroid plexus papilloma
- Craniopharyngioma **9350/1** and a subtype/variant of craniopharyngioma
- Gangliocytoma **9492/0** and a subtype/variant of gangliocytoma
- Lipoma **8850/0** and a subtype/variant of lipoma
- Meningeal melanocytosis **8728/0** and a subtype/variant of meningeal melanocytosis
- Meningioma **9530/0** and a subtype/variant of meningioma
- Myofibroblastoma **8825/0** and a subtype/variant of myofibroblastoma
- Neurofibroma **9540/0** and a subtype/variant of neurofibroma
- Schwannoma **9560/0** and a subtype/variant of schwannoma
- Solitary fibrous tumor WHO Grade 1 **8815/0** and a subtype/variant of solitary fibrous tumor WHO Grade 1

**Note:** Use [Table 6](#) in the Equivalent Terms and Definitions to identify the NOS and subtypes/variants.

**This is the end of instructions for Multiple Tumors Abstracted as a Single Primary.**

**Code the histology according to the rule that fits the case.**

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