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-extradural; within the spinal canal but outside of the nerves
- Intraspinal; 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).
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**).
Information is presented in the general order in which a case is abstracted.

**Section 1: Behavior Code**
- **Priority Order** for Using Documentation to Assign Behavior
- **Table 1**: WHO Grades of Select CNS Neoplasms

**Section 2: Reportable Primary Sites and Histologies**
- **Priorities for Coding Primary Site**
- **Reportable Primary Site Groups**
- **Table 2**: Reportable Primary Sites
- **Table 3**: Reportability of Non-Malignant Cranial Nerve (CN) Tumors (defines which portions of the cranial nerves are reportable)
- **Table 4**: Non-Reportable Neoplasms
- **Table 5**: Histologic Types of Non-Malignant Intracranial (Brain and Gland) Tumors
- **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**
- **Conflicting information on Pathology report(s)**
- **Table 7**: Paired Sites
- **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

Jump to Multiple Primary Rules
Jump to Histology Coding Rules
D. Angiogram

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

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

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

**WHO Grade Definitions**

**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 4th Ed Tumors of CNS separates CNS entities into histologic groups. For example, diffuse astrocytic and oligodendrogial 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.
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

<table>
<thead>
<tr>
<th>Histology</th>
<th>WHO Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anaplastic (malignant) meningioma</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic astrocytoma, IDH mutant</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic ependymoma</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic ganglioglioma</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic oligodendroglioma IDH mutant and 1p/19q-codeleted</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic pleomorphic xanthroastrocytoma</td>
<td>3</td>
</tr>
<tr>
<td>Angiocentric glioma</td>
<td>1</td>
</tr>
<tr>
<td>Atypical choroid plexus papilloma</td>
<td>2</td>
</tr>
<tr>
<td>Atypical meningioma</td>
<td>2</td>
</tr>
<tr>
<td>Atypical teratoid/rhabdoid tumor</td>
<td>4</td>
</tr>
<tr>
<td>Central neurocytoma</td>
<td>2</td>
</tr>
</tbody>
</table>
Non-Malignant CNS Equivalent Terms and Definitions  
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

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

Jump to [Multiple Primary Rules](#)  
Jump to [Histology Coding Rules](#)  
Non-malignant CNS Solid Tumor Rules  
2023 Update
## 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>
<thead>
<tr>
<th>Histology</th>
<th>WHO Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medulloepithelioma</td>
<td>4</td>
</tr>
<tr>
<td>Meningioma</td>
<td>1</td>
</tr>
<tr>
<td>Myxopapillary ependymoma</td>
<td>1</td>
</tr>
<tr>
<td>Neurofibroma</td>
<td>1</td>
</tr>
<tr>
<td>Oligodendroglioma IDH mutant and 1p/19q-codeleted</td>
<td>2</td>
</tr>
<tr>
<td>Papillary glioneuronal tumor</td>
<td>1</td>
</tr>
<tr>
<td>Papillary tumor of the pineal region</td>
<td>2 or 3</td>
</tr>
<tr>
<td>Note: Tissue/pathology report or CAP will specify whether it is WHO Grade 2 or 3</td>
<td></td>
</tr>
<tr>
<td>Perineuroma</td>
<td>1</td>
</tr>
<tr>
<td>Pilocytic astrocytoma</td>
<td>1</td>
</tr>
<tr>
<td>Note: ICD-O-3 lists a behavior code of /1. US and Canada collect as /3.</td>
<td></td>
</tr>
<tr>
<td>Pineal parenchymal tumor of intermediate differentiation</td>
<td>2 or 3</td>
</tr>
<tr>
<td>Note: Tissue/pathology or CAP synoptic report will specify WHO Grade 2 or 3</td>
<td></td>
</tr>
<tr>
<td>Pineoblastoma</td>
<td>4</td>
</tr>
<tr>
<td>Pineocytoma</td>
<td>1</td>
</tr>
<tr>
<td>Pituicytoma</td>
<td>1</td>
</tr>
<tr>
<td>Pleomorphic xanthroastrocytoma</td>
<td>2</td>
</tr>
<tr>
<td>Rosette-forming glioneuronal tumor</td>
<td>1</td>
</tr>
<tr>
<td>Schwannoma</td>
<td>1</td>
</tr>
<tr>
<td>Solitary fibrous tumor/hemangiopericytoma</td>
<td>1, 2, or 3</td>
</tr>
<tr>
<td>Note: Tissue/pathology will specify WHO Grade 1, 2, or 3</td>
<td></td>
</tr>
<tr>
<td>Spindle cell oncocytoma</td>
<td>1</td>
</tr>
<tr>
<td>Subependymal giant cell astrocytoma</td>
<td>1</td>
</tr>
<tr>
<td>Subependymoma</td>
<td>1</td>
</tr>
</tbody>
</table>
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
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).
   - **Cerebral/cranial dura/meninges** C700. The cerebral meninges has three layers:
     - **Dura** mater is the *superficial* layer of meninges
       - Tightly adherent to skull
       - Contains folds and sinuses
       - Contacts *endosteu* which lines the bones of the skull
     - **Arachnoid** mater forms the middle of the three layers of meninges
     - **Pia** mater is the delicate vascular fibrous membrane which is adherent to the brain.
   - **Brain** C710-C719
   - **Craniopharyngeal duct** C752
   - **Pineal gland** C753
   - **Pituitary gland** C751

2. **Spinal** sites: includes the *spinal meninges* and all structures *within* the meninges (Intradural, within/in the spinal meninges).
   - **Spinal cord** C720
   - 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.

<table>
<thead>
<tr>
<th>Site Group</th>
<th>Reportable Subsite Terms and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Brain</strong></td>
<td>Brain NOS C719</td>
</tr>
<tr>
<td></td>
<td>Brain stem C717</td>
</tr>
<tr>
<td></td>
<td>Cerebellum NOS C716</td>
</tr>
<tr>
<td></td>
<td>Cerebrum C710</td>
</tr>
<tr>
<td></td>
<td>Frontal lobe C711</td>
</tr>
<tr>
<td></td>
<td>Occipital lobe C714</td>
</tr>
<tr>
<td></td>
<td>Overlapping lesion of brain and central nervous system C718</td>
</tr>
<tr>
<td></td>
<td>Parietal lobe C713</td>
</tr>
<tr>
<td></td>
<td>Temporal lobe C712</td>
</tr>
<tr>
<td></td>
<td>Ventricle NOS C715</td>
</tr>
<tr>
<td><strong>Cranial Nerves</strong></td>
<td>Abducent (cranial nerve VI) C725</td>
</tr>
<tr>
<td></td>
<td>Accessory (cranial nerve XI) C725</td>
</tr>
<tr>
<td></td>
<td>Acoustic (cranial nerve VIII) C724</td>
</tr>
<tr>
<td></td>
<td>Cranial nerve NOS C725</td>
</tr>
<tr>
<td></td>
<td>Facial (cranial nerve VII) C725</td>
</tr>
<tr>
<td></td>
<td>Glossopharyngeal (cranial nerve IX) C725</td>
</tr>
<tr>
<td></td>
<td>Hypoglossal (cranial nerve XII) C725</td>
</tr>
<tr>
<td></td>
<td>Oculomotor (cranial nerve III) C725</td>
</tr>
<tr>
<td></td>
<td>Olfactory (cranial nerve I C722)</td>
</tr>
<tr>
<td></td>
<td>Optic (cranial nerve II) C723</td>
</tr>
<tr>
<td></td>
<td>Trigeminal (cranial nerve V) C725</td>
</tr>
<tr>
<td></td>
<td>Trochlear (cranial nerve IV) C725</td>
</tr>
<tr>
<td></td>
<td>Vagus (cranial nerve X) C725</td>
</tr>
</tbody>
</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>
<thead>
<tr>
<th>Site Group</th>
<th>Reportable Subsite Terms and Code</th>
</tr>
</thead>
</table>
| Ill-Defined Sites Central Nervous System | Nervous system NOS **C729**  
Overlapping lesion of brain and central nervous system **C728** |
| Intracranial Duct and Glands        | Craniopharyngeal duct **C752**  
Pineal gland **C753**  
Pituitary gland **C751** |
| Meninges                           | Cerebral meninges **C700**  
Meninges NOS **C709**  
Spinal meninges **C701** |
| Spinal Sites                        | Cauda equina/ **C721**  
Conus medullaris/filum terminale **C720**  
Meninges NOS **C709**  
Spinal cord **C720**  
Spinal meninges **C701** |
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 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

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

### 2023 Update

<table>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Reportable Portions of CN</th>
<th>Non-Reportable Portions of CN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trochlear CN 4 C725</td>
<td>Superior orbital fissure</td>
<td>Arises from the <strong>dorsal brain stem</strong>, loops around the brainstem and passes anteriorly within the <strong>subarachnoid space</strong>. It travels between the <strong>superior cerebellar and posterior cerebral arteries</strong> and through the <strong>dura</strong>, enters <strong>cavernous sinus</strong>.</td>
<td>Enters the <strong>orbital fissure</strong>.</td>
</tr>
</tbody>
</table>
| Trigeminal CN 5 C725 | 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 **pons**. Upon leaving the pons it enters a **small fossa** posterior and inferolateral to the cavernous sinus called **Meckel's (trigeminal) cave**. | **Ophthalmic nerve branch** crosses the **pterygopalatine fossa**, inclines laterally on the back of the **maxilla**, and enters the **orbit** through the inferior orbital fissure where it is called the **infraorbital nerve**. It ends beneath the **quadatus labii superior**, and divides into multiple branches that spread to the side of the **nose**, the lower **eyelid**, and the upper **lip**. **Maxillary nerve** leaves foramen rotundum and traverses the **infraorbital groove** and **canal** in the floor of the **orbit**, and appears on the face at the infraorbital foramen. **Mandibular** nerve leaves via the foramen ovale travels along the **mandibular groove**.

---

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### 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>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Reportable Portions of CN</th>
<th>Non-Reportable Portions of CN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abducent CN 6 C725</td>
<td>Cranial meninges</td>
<td>Exits brainstem at junction of <strong>pons</strong> and the <strong>medulla</strong>, enters the <strong>subarachnoid</strong> space and runs upward between the pons and the <strong>clivus</strong> entering the <strong>cavernous sinus</strong>.</td>
<td><strong>Dorello's canal</strong> and travels to the tip of the <strong>temporal bone</strong>. Enters <strong>orbit</strong></td>
</tr>
<tr>
<td>Facial CN 7 C725</td>
<td>Internal acoustic meatus</td>
<td>CN7 originates in the <strong>pons</strong>, along the posterior cranial fossa (the posterior cranial fossa is part of the intracranial cavity)</td>
<td>Enters the <strong>temple</strong> through the <strong>internal auditory meatus</strong> and runs through the <strong>facial canal</strong>.</td>
</tr>
<tr>
<td>Acoustic or vestibulocochlear CN 8 C724</td>
<td>Internal acoustic meatus</td>
<td>Originates in the <strong>brain stem</strong> (<strong>medulla oblongata</strong> between the base of the brain (<strong>pons</strong>) and the <strong>spinal cord</strong>) Both the <strong>vestibular</strong> branch and the <strong>cochlear</strong> branch are located in the <strong>inner ear</strong></td>
<td></td>
</tr>
<tr>
<td>Glossopharyngeal CN 9 C725</td>
<td>Jugular foramen</td>
<td><strong>Originates</strong> in the anterior portion of the <strong>medulla oblongata</strong></td>
<td><strong>Jugular foramen</strong> Between the <strong>internal jugular vein</strong> and <strong>internal carotid artery</strong> Lies on the <strong>stylopharyngeus</strong> and <strong>middle pharyngeal constrictor muscle</strong> Passes under the <strong>hypoglossus muscle</strong> <strong>Palatine tonsil</strong> Extends to <strong>mucous glands</strong> of the <strong>mouth</strong>, and <strong>base</strong> of the <strong>tongue</strong></td>
</tr>
</tbody>
</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>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Reportable Portions of CN</th>
<th>Non-Reportable Portions of CN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vagus CN 10  C725</td>
<td>Jugular foramen</td>
<td>The vagus nerve originates from the medulla of the brainstem.</td>
<td>CN10 descends within the carotid sheath medial to the internal jugular vein at the root of the neck. The right vagus crosses in front of the subclavian artery and travels into the fat behind the blood vessels, reaching the thorax. It then inclines behind the hilum of the right lung and moves toward the esophagus. The nerve splits into the right and left vagus at the esophageal plexus forming the anterior and posterior gastric nerves.</td>
</tr>
<tr>
<td>Accessory CN 11 C725</td>
<td>Jugular foramen</td>
<td>The nerve enters the foramen magnum or lateral aspect of the medulla oblongata.</td>
<td>The spinal accessory nerve originates in the neurons of the upper spinal cord, specifically C1-C5/C6 spinal nerve roots. The fibers of the spinal accessory nerve coalesce to form spinal rootlets, roots, and finally the spinal accessory nerve itself. The nerve exits the skull through the jugular foramen. It then runs along the internal carotid artery within the neck and reaches the sternocleidomastoid muscle and the trapezius.</td>
</tr>
</tbody>
</table>

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Non-malignant CNS Solid Tumor Rules  
2023 Update
<table>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Reportable Portions of CN</th>
<th>Non-Reportable Portions of CN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypoglossal CN 12 C725</td>
<td>Hypoglossal canal</td>
<td>CN12 starts in the hypoglossal nucleus of the brainstem.</td>
<td>CN12 exits the hypoglossal canal, traveling between the carotid artery and jugular vein, ending under the tongue.</td>
</tr>
</tbody>
</table>

Non-Malignant CNS Solid Tumor Rules

2023 Update
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).

<table>
<thead>
<tr>
<th>Non-reportable Histology Term</th>
<th>Non-reportable Histology Code</th>
<th>Definitions and Sites</th>
</tr>
</thead>
<tbody>
<tr>
<td>Carcinomas</td>
<td>8010-8060, 8071-8671, 8940-8941</td>
<td>Brain C710-C719</td>
</tr>
<tr>
<td></td>
<td></td>
<td><strong>Site/histology edit</strong> carcinomas/brain</td>
</tr>
<tr>
<td>Carcinomas</td>
<td>8010-8671, 8940-8941</td>
<td>Cerebral meninges, spinal meninges, meninges NOS C700-C709</td>
</tr>
<tr>
<td></td>
<td></td>
<td><strong>Site/histology edit</strong> carcinomas/meninges</td>
</tr>
<tr>
<td>Carcinomas</td>
<td>8010-8671, 8940-8941</td>
<td>C721-C729 (Other central nervous system)</td>
</tr>
<tr>
<td></td>
<td></td>
<td><strong>Site/histology edit</strong> carcinomas/other CNS</td>
</tr>
<tr>
<td>Colloid cyst</td>
<td>No code</td>
<td>These tumors occur in the inner ear, the aortic body and other paraganglia respectively; these sites are <strong>not reportable</strong>.</td>
</tr>
<tr>
<td>Epidermoid tumor/cyst</td>
<td>No code</td>
<td></td>
</tr>
<tr>
<td>Glomus tympanicum, glomus jugulare</td>
<td>8690/1</td>
<td></td>
</tr>
<tr>
<td>Hygroma</td>
<td>9173/0</td>
<td>Occurs in hypothalamus</td>
</tr>
<tr>
<td>Hypothalamic hamartoma</td>
<td>No code</td>
<td></td>
</tr>
<tr>
<td>Neurofibromatosis, NOS</td>
<td>9540/1</td>
<td>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.</td>
</tr>
<tr>
<td>Neurofibromatosis, type 1 (NF1)</td>
<td>No code*</td>
<td>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.</td>
</tr>
<tr>
<td>Neurofibromatosis, type 2 (NF2)</td>
<td>No code*</td>
<td>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.</td>
</tr>
<tr>
<td>Neuroglial cyst</td>
<td>No code</td>
<td>Ventricles</td>
</tr>
<tr>
<td>Osteochondroma</td>
<td>9210/0</td>
<td>Originates in the cartilage around bone, site not reportable for non-malignant neoplasms</td>
</tr>
</tbody>
</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>
<thead>
<tr>
<th>Non-reportable Histology Term</th>
<th>Non-reportable Histology Code</th>
<th>Definitions and Sites</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rathke cleft cyst</td>
<td><strong>No code</strong></td>
<td>Sella turcica C751. This is a Rathke cleft CYST, not a Rathke cleft tumor.</td>
</tr>
<tr>
<td>Schwannomatosis</td>
<td><strong>No code</strong>*</td>
<td>A form of neurofibromatosis newly named/discovered</td>
</tr>
</tbody>
</table>

*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
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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

<table>
<thead>
<tr>
<th>Histology Term and Code</th>
<th>Most Common Intracranial Primary Site</th>
</tr>
</thead>
<tbody>
<tr>
<td>Angiocentric glioma 9431/1*</td>
<td>Cerebrum C710</td>
</tr>
<tr>
<td>Choroid plexus papilloma 9390/0</td>
<td>Intraventricular site (lateral/third ventricle C715 and IV ventricle C717)</td>
</tr>
<tr>
<td>(Capillary) hemangioblastoma 9161/1</td>
<td>Cerebellum C716, cerebrum (rare) C710</td>
</tr>
<tr>
<td>Craniopharyngioma 9350/1</td>
<td>Craniopharyngeal duct C752, pituitary gland, sella turcica C751</td>
</tr>
<tr>
<td>Dermoid cyst 9084/0</td>
<td>Pineal gland C753, suprasellar C719</td>
</tr>
<tr>
<td>Desmoplastic infantile astrocytoma and ganglioglioma 9412/1</td>
<td>Cerebrum/supratentorial brain NOS C710</td>
</tr>
<tr>
<td>Dysembryoplastic neuroepithelial tumor (DNT) 9413/0</td>
<td>Cerebrum C710, temporal lobe C712</td>
</tr>
<tr>
<td>Histology Term and Code</td>
<td>Most Common Intracranial Primary Site</td>
</tr>
<tr>
<td>------------------------</td>
<td>-------------------------------------</td>
</tr>
<tr>
<td>Dysplastic gangliocytoma 9493/0</td>
<td>Cerebellum C716</td>
</tr>
<tr>
<td>Juvenile xanthogranuloma 9749/1</td>
<td>Intraventricular C715</td>
</tr>
<tr>
<td>Meningioma (rare) 9530/0</td>
<td>Intraventricular C715</td>
</tr>
<tr>
<td>Myxopapillary ependymoma 9394/1</td>
<td>4th ventricle C717</td>
</tr>
<tr>
<td>Pilocytic astrocytoma/juvenile pilocytic astrocytoma 9421/1</td>
<td>Optic nerve C723</td>
</tr>
<tr>
<td>Pineocytoma 9361/1</td>
<td>Pineal gland C753</td>
</tr>
<tr>
<td>Pituicytoma 9432/1*</td>
<td>Pituitary gland C751, sella turcica C751, suprasellar C719</td>
</tr>
<tr>
<td>Pituitary adenoma 8272/0</td>
<td>Pituitary gland C751</td>
</tr>
<tr>
<td>Prolactinoma 8271/0</td>
<td>Pituitary gland C751</td>
</tr>
<tr>
<td>Subependymal giant cell tumor (SEGA) 9384/1</td>
<td>Lateral ventricles C715</td>
</tr>
<tr>
<td>Subependymoma 9383/1</td>
<td>Intraventricular site (lateral/third ventricle, rare C715 and IV ventricle C717)</td>
</tr>
</tbody>
</table>
Non-Malignant CNS Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

**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.

<table>
<thead>
<tr>
<th>NOS/Specific Histology Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants Histology Term and Codes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Angiocentric glioma 9431/1*</td>
<td>Angiocentric neuroepithelial tumor Monomorphous angiocentric glioma</td>
<td></td>
</tr>
<tr>
<td>Benign fibrous histiocytoma 8830/0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chondroma 9220/0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chordoid glioma of the third ventricle 9444/1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Choroid plexus papilloma 9390/0</td>
<td>Atypical choroid plexus papilloma 9390/1</td>
<td></td>
</tr>
<tr>
<td>Craniopharyngioma 9350/1</td>
<td>Adamantinomatous craniopharyngioma 9351/1 Papillary craniopharyngioma 9352/1</td>
<td></td>
</tr>
<tr>
<td>Desmoplastic infantile astrocytoma and ganglioglioma 9412/1</td>
<td>DIAG</td>
<td></td>
</tr>
</tbody>
</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>
<thead>
<tr>
<th>NOS/Specific Histology Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants Histology Term and Codes</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Diffuse astrocytoma, MYB- or MYBL1 altered 9421/1</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Note 1:</strong> Beginning 1/1/2023, diffuse astrocytoma, MYB- or MYBL1 altered is the preferred term for 9421/1.</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Note 2:</strong> Beginning 1/1/2023, pilocytic astrocytoma/juvenile pilocytic astrocytoma are coded 9421/1. Cases diagnosed prior to 1/1/2023 are coded 9421/3.</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Angiocentric glioma</strong></td>
<td>Diffuse low-grade glioma, MAPK pathway-altered</td>
<td></td>
</tr>
<tr>
<td><strong>Juvenile pilocytic astrocytoma</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Pilocytic astrocytoma</strong></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

| **Dysembryoplastic neuroepithelial tumor 9413/0** | | |
| **Note:** 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** | | |

| **Gangliocytoma 9492/0** | | |
| **Ganglioglioma 9505/1** | | |
| **Granular cell tumor of the sellar region 9582/0** | | |
| **Hemangioblastoma 9161/1** | Capillary hemangioblastoma | |
| **Hemangioma 9120/0** | Cavernous hemangioma 9121/0 | |
| **Juvenile xanthogranuloma 9749/1** | | |
| **Leiomyoma 8890/0** | | |
| **Lipoma 8850/0** | Hibernoma 8880/0 | |
| **Meningeal melanocytosis 8728/0** | Meningeal melanocytoma 8728/1 | |

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## Non-Malignant CNS Equivalent Terms and Definitions
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<table>
<thead>
<tr>
<th>NOS/Specific Histology Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants Histology Term and Codes</th>
</tr>
</thead>
</table>
| Meningioma 9530/0                    | Lymphoplasmacyte-rich meningioma  
Metaplastic meningioma  
Microcystic meningioma  
Secretory meningioma | Angiomatous meningioma 9534/0  
Atypical meningioma 9539/1  
Clear cell/chordoid meningioma 9538/1  
Fibrous meningioma 9532/0  
Meningothelial meningioma 9531/0  
Psammomatous meningioma 9533/0  
Transitional meningioma 9537/0 |
| Multinodular and vacuolating neuronal tumor 9509/0 | MVNT | |
| Note: 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. |
| Myofibroblastoma 8825/0               | Inflammatory myofibroblastic tumor 8825/1 |
| Myxopapillary ependymoma 9394/1      | | |
| Neurocytoma 9506/1                    | Central neurocytoma  
Cerebellar liponeurocytoma  
Extraventriculare neurocytoma  
Lipomatous medulloblastoma  
Medullocytooma  
Neurolipocytoma | |
| Neuroepithelial tumor, benign 8000/0 | Neuroepithelial tumor, NOS 8000/1 |
| Neurofibroma 9540/0                   | Atypical neurofibroma  
Plexiform neurofibroma 9550/0 |
| Optic glioma/pilocytic astrocytoma 9421/1 | | |

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Non-malignant CNS Solid Tumor Rules  
2023 Update
## 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>
<thead>
<tr>
<th>NOS/Specific Histology Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants Histology Term and Codes</th>
</tr>
</thead>
</table>
| Papillary glioneuronal tumor 9509/1 |          | Diffuse leptomeningeal glioneuronal tumor (see note 2)  
Rosette-forming glioneuronal tumor |          |
| Note 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 rows in column 1. |          |                                           |
| Note 2: Beginning with cases diagnosed 1/1/2023 forward, leptomeningeal glioneuronal tumor is coded 9509/3. See the Malignant CNS rules. |          |                                           |
| Paraganglioma 8693/1 |          |                                           |
| Perineurioma 9571/0 |          |                                           |
| Pineocytoma 9361/1 |          |                                           |
| Pituicytoma 9432/1* |          |                                           |
| Pituitary adenoma 8272/0 | | Corticotroph  
Gonadotroph adenoma  
Somatotroph adenoma  
Thyrotroph adenoma  
Null cell adenoma  
Plurihormonal and double adenomas |                                           |
| Polymorphous low-grade neuroepithelial tumor of the young 9413/0 | | PLNTY |                                           |
| Note: 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. |          |                                           |
| Prolactinoma 8271/0 |          |                                           |
| Rhabdomyoma 8900/0 |          |                                           |
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<tr>
<th>NOS/Specific Histology Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants Histology Term and Codes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Schwannoma 9560/0</td>
<td>Acoustic neuroma</td>
<td>Melanotic schwannoma 9560/1*</td>
</tr>
<tr>
<td></td>
<td>Cellular schwannoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Neurilemoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Neurinoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Plexiform schwannoma</td>
<td></td>
</tr>
<tr>
<td>Melanotic schwannoma 9560/1*</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Spindle cell oncocytoma 8290/0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Subependymal giant cell astrocytoma 9384/1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Subependymoma 9383/1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Teratoma 9080/1</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Jump to [Multiple Primary Rules](#)  
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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.
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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.

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

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.
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### 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.

<table>
<thead>
<tr>
<th>Original Histology and Code</th>
<th>Transformed Histology and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chondroma 9220/0</td>
<td>Chondrosarcoma 9220/3</td>
</tr>
<tr>
<td>Ganglioglioma 9505/1</td>
<td>Anaplastic ganglioglioma 9505/3</td>
</tr>
<tr>
<td>Hemangioma 9120/0</td>
<td>Angiosarcoma 9120/3</td>
</tr>
<tr>
<td>Hemangiopericytoma 9150/1</td>
<td>Anaplastic hemangiopericytoma 9150/3</td>
</tr>
<tr>
<td>Leiomyoma 8890/0</td>
<td>Leiomyosarcoma 8890/3</td>
</tr>
<tr>
<td>Lipoma 8850/0</td>
<td>Liposarcoma 8850/3</td>
</tr>
<tr>
<td>Osteoma 9180/0</td>
<td>Osteosarcoma 9180/3</td>
</tr>
<tr>
<td>Perineurioma 9571/0</td>
<td>Malignant perineurioma 9571/3</td>
</tr>
<tr>
<td>Rhabdomyoma 8900/0</td>
<td>Rhabdomyosarcoma 8900/3</td>
</tr>
<tr>
<td>Teratoma 9080/1</td>
<td>Immature teratoma 9080/3</td>
</tr>
<tr>
<td>Teratoma, mature 9080/0</td>
<td>Immature teratoma 9080/3</td>
</tr>
</tbody>
</table>
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Illustrations
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**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\(^1\) 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.

\(^1\) Prepare one abstract. Use the histology rules to assign the appropriate histology code.
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C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
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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\(^1\) 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\(^1\) (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:
     - Clinical
     - Radiographic
     - 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.
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- 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 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.

---

i Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.
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(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 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 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).
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Rule M7 Abstract multiple primaries 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 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 menigioma 9539/1 and fibrous menigioma 9532/0 are both subtypes of menigioma 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 when two or more separate/non-contiguous menigiomas 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 menigiomas that are either a NOS and subtype/variant, OR they are the same histology.
Rule M10  Abstract a single primary when there are separate/non-contiguous tumors in the brain (multicentric/multifocal) with the same histology. Tumors may be in any of the following locations and/or lateralties:
- Same laterality: In the same lobe; for example, two tumors in right temporal lobe C712 (same site code)
- Different lateralties 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)

**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 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:
  - 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
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Rule M12  Abstract multiple primaries\(^i\) when separate/non-contiguous tumors are on different rows in Table 6 in the Equivalent Terms and Definitions. Timing is irrelevant.
Note: Each row in the table is a distinctly different histology.

Rule M13  Abstract a single primary\(^\text{i}\) 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

\(^{i}\) 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.
\(^{\text{i}}\) Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.
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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
   - 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.
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- 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.

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**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
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.
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(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/documentated
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 meningothelial meningioma. The treatment plan says the
patient will receive the following treatment for meningothelial meningioma. Treatment plan confirms
meningothelial meningioma; code meningothelial 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
- Appears
- Comparable with
- Compatible with
- Consistent with
- Favor(s)
- Malignant appearing
- Most likely
- Presumed
- Probable
- Suspect(ed)
- Suspicious (for)
- Typical (of)

4. **Do not code** histology when described as:
   - Architecture
   - Foci; focus; focal
   - Pattern

Jump to [Equivalent Terms and Definitions](#) Jump to [Multiple Primary Rules](#)
Non-Malignant CNS Histology Rules
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
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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.
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 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.**