Introduction

Note 1: This section includes the following primary sites: Peripheral nerves C470-C479; cerebral meninges C700; spinal meninges C701; meninges NOS C709; brain C710-C719; 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; craniohypophysial duct C752; pineal gland C753.

Note 2: Non-malignant intracranial and CNS tumors 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: There must be a histologic, cytologic, radiographic, or clinical diagnosis of a malignant neoplasm /3.

Note 5: Tumors from a number of primary sites metastasize to the brain. Do not use these rules for tumors described as metastases; report metastatic tumors using the rules for that primary site.

Note 6: Pilocytic astrocytoma/juvenile pilocytic astrocytoma is reportable in North America as a malignant neoplasm 9421/3.

Note 7: Tables and rules refer to ICD-O rather than ICD-O-3. The specific version is not specified to allow for updates. Use the currently approved version of ICD-O and all updates.

Note 8: 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.

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.
Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions

C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia – M9590-M9992 and Kaposi sarcoma M9140)

- Cerebrospinal fluid; CSF
- 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; spinal nerve roots
- Infratentorial; below the tentorium cerebelli; either cerebellum or brainstem
- Intracranial; inside the skull
- Intradural; with/between layers of meninges
- Site; topography
- Supratentorial; above the tentorium cerebelli; cerebrum
  - Cerebrum contains frontal lobe, parietal lobe, occipital lobe, and temporal lobe
- Tentorium cerebelli; cerebellar tentorium
- Tumor; mass; lesion; neoplasm when
  - These terms are used ONLY to determine multiple primaries
  - Do not use these terms for casefinding or determining reportability
- Type; subtype; variant
- WHO Grade 3 and WHO Grade 4; malignant; invasive; /3

Terms that are NOT Equivalent or Equal

This is a term that is **not equivalent**. There are no casefinding implications.

- **Component** is not equivalent to subtype/variant
  - **Note**: Component is only coded when the pathologist specifies the component as a second carcinoma.
Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
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Changes from 2007 MPH Rules

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

1. 2016 CNS WHO presents major restructuring of the diffuse gliomas, medulloblastomas and other embryonal tumors, and incorporates new entities that are defined by both histology and molecular features, including glioblastoma, IDH-wildtype and glioblastoma, IDH-mutant; diffuse midline glioma, H3 K27M-mutant, RELA fusion-positive ependymoma, medulloblastoma, WNT-activated and medulloblastoma, SHH-activated, and embryonal tumor with multilayered rosettes, C19MC-altered. The 2016 edition has added newly recognized neoplasms and has referred to some entities, variants and patterns as “not recommended” (previously called obsolete).
   A. It has been determined that these “not recommended” terms no longer have diagnostic and/or biological relevance.
   B. Terms which are not recommended are not included in the tables. When one of these terms are used, refer to the ICD-O and all updates for the correct histology code.
2. **Rule change:** The 2007 rules said a glioblastoma multiforme (GBM) following an astrocytic or glial tumor was a single primary (recurrence). In the 2018 Solid Tumor Rules, GBM subsequent to an astrocytic or glial tumor is a multiple primary.
   - GBM is now being collected as a new primary so it is possible to analyze the frequency with which these tumors recur in a more aggressive form (GBM)
3. **Clarification:** The following meningiomas are reportable: intraosseous, cavernous sinus and sphenoid wing.
4. **Clarification:** Multiple cerebral meningiomas are a single primary.
5. **Clarification:** Multiple brain tumors (same histology) are a single primary.
6. 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 **Table 3** in the Terms and Definitions.
CNS neoplasms must meet all three of the conditions below to be reported as malignant /3:

1. The behavior must be malignant /3:
   A. Pathology designates the behavior as malignant/invasive, /3 OR
   B. The tumor is a WHO Grade 3 or 4 (See Section 1: Table 1)
      Note 1: WHO Grade 2 tumors may be non-malignant or malignant.
      Note 2: Always code the behavior as designated by the pathologist.
      Note 3: 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.

2. The primary site must be reportable (See Section 2: Table 2) AND

3. The histology must be reportable (See Section 2: Table 3)
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**
   - A. Use the pathologist’s **description** of malignant/invasive behavior
   - B. Cases are reportable as malignant when pathology states a WHO Grade 3 or 4
      - i. WHO Grade 2 may be either non-malignant or malignant. Use the pathologist’s description of behavior (priority #1).
   - C. **Never change behavior** described by pathologist

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 pathologic diagnosis and behavior

   **Example:** Pathology report is not in the medical record. Oncology consult states biopsy was done in a different facility and cites the original pathology diagnosis including the behavior.
   - C. **Documentation** of behavior, no mention of original diagnosis

   **Example:** Pathology report is not in the medical record. **Physician documents** the behavior as malignant, or WHO Grade 3 or 4, but **does not cite/mention original** pathology report as source of behavior classification.

5. **Scan,** use behavior information from radiography in the following **priority order**:
   - A. MRI
   - B. CT
   - C. PET
   - D. Angiogram

6. When instructions 1-5 **do not apply**, use **Table 1** to determine behavior.
Table 1: WHO Grades for Select CNS Neoplasms

Note: See the SEER and COC Manuals for instructions on coding grade for CNS tumors.

1. WHO does not provide Grades for all CNS and peripheral nerve neoplasms
2. WHO Grade(s) 3 and 4 neoplasms are always malignant /3
3. Go to Instructions for Identifying and Assigning Behavior to determine whether WHO Grade 2 neoplasms are non-malignant or malignant.

Column 1 contains the histology term.
Column 2 contains the WHO Grade assigned based on the molecular features of the 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>
<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>
</tbody>
</table>
### Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions

C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753

(Excludes lymphoma and leukemia – M9590-M9992 and Kaposi sarcoma M9140)

<table>
<thead>
<tr>
<th>Histology</th>
<th>WHO Grade</th>
</tr>
</thead>
<tbody>
<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 (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>
</tbody>
</table>

**Note:** Tissue/pathology reports or CAP protocol/summary 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|

**Note:** Tissue/pathology reports or CAP protocol/summary will specify whether this is WHO Grade 2, 3, or 4

| Medulloblastoma (including all subtypes)                                 | 4         |
| Medulloepithelioma                                                       | 4         |
| Meningioma                                                               | 1         |
| Myxopapillary ependymoma                                                 | 1         |
| Neurofibroma                                                             | 1         |
| Oligodendroglioma IDH-mutant and 1p/19q deleted                          | 2         |
| Papillary glioneuronal tumor                                             | 1         |
| Papillary tumor of the pineal region                                     | 2 or 3    |

**Note:** Tissue/pathology reports or CAP protocol/summary will specify whether it is WHO Grade 2 or 3

| Perineuroma                                                              | 1         |
# Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions

C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753

(Excludes lymphoma and leukemia – M9590-M9992 and Kaposi sarcoma M9140)

<table>
<thead>
<tr>
<th>Histology</th>
<th>WHO Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pilocytic astrocytoma</td>
<td>1</td>
</tr>
</tbody>
</table>

*Note:* Collected as malignant /3 in North America

<table>
<thead>
<tr>
<th>Histology</th>
<th>WHO Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pineal parenchymal tumor of intermediate differentiation</td>
<td>2 or 3</td>
</tr>
</tbody>
</table>

*Note:* Tissue/pathology reports or CAP protocol/summary will specify WHO Grade 2 or 3

<table>
<thead>
<tr>
<th>Histology</th>
<th>WHO Grade</th>
</tr>
</thead>
<tbody>
<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>
</tbody>
</table>

*Note:* Tissue/pathology reports or CAP protocol/summary will specify WHO Grade 1, 2, or 3

<table>
<thead>
<tr>
<th>Histology</th>
<th>WHO Grade</th>
</tr>
</thead>
<tbody>
<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>
Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia – M9590-M9992 and Kaposi sarcoma M9140)

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:** Peripheral nerves are included in the Malignant CNS and Peripheral Nerve rules because:
- All malignant tumors are reportable, including peripheral nerve tumors AND
- The Malignant CNS and Peripheral Nerve rules contain the correct histologies and coding rules for tumors of peripheral nerves and meninges/dura.

**Note 2:** Peripheral nerves are:
- Extracranial/outside the cranium OR
- Extradural/outside the spinal dura

**Note 3:** Peripheral nerves are not included in the non-malignant CNS rules because benign tumors of peripheral nerves and/or meninges of those nerves are not reportable.

**Note 4:** The following 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
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.

Use the list 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 OR
   C. Physician’s documentation of primary site in the medical record

   Example: The patient had a biopsy done at another facility. 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
5. See Table 2: Reportable Primary Sites to confirm the primary site is reportable.
6. When the primary site is cranial nerve OR peripheral nerve, see Table 4: Coding Primary Site for Malignant Tumors of Cranial and Peripheral Nerves to determine whether the portion of the nerve is cranial or peripheral (different site codes).
Reportable Primary Site Groups

The three major groups of reportable sites are:
1. **Intracranial** (within the skull/cranium) AND
2. **Spinal** sites (spinal meninges and sites within the spinal meninges)
3. **Peripheral nerves** (extracranial or extraspinal nerves).

Terms and sites for these broad categories are:

### 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 4: Coding Primary Site for Malignant Tumors of Cranial and Peripheral Nerves**
   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 meninges** C701 covers/encloses the spinal nerve roots and the spinal cord.
   B. Spinal nerve roots:
      i. **Cervical** nerve, 8 pair, occipital nerve. Code to peripheral nerve of head, face, and neck C470
      ii. **Coccygeal** nerve, 1 pair. Code to cauda equina C721
iii. **Lumbar** nerve, 5 pair. Code to cauda equina C721
iv. **Sacral** nerve, 5 pair. Code to cauda equina C721
v. **Thoracic** nerve, 12 pair. Code to peripheral nerves of thorax C473

3. **Peripheral nerves**
   i. **Cervical** nerve, 8 pair, occipital nerve. Code to peripheral nerve of head, face, and neck C470
   ii. **Thoracic** nerve, 12 pair. Code to peripheral nerves of thorax C473
   iii. **Lumbar** nerve, 5 pair. Code to cauda equina C721
   iv. **Sacral** nerve, 5 pair. Code to cauda equina C721
   v. **Coccygeal** nerve, 1 pair. Code to cauda equina C721
### Table 2: Reportable Primary Sites

Use Table 2 to determine whether a primary site is reportable.

<table>
<thead>
<tr>
<th>Site Group</th>
<th>Reportable Subsite Terms and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Brain</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>Cranial Nerves</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>
<tr>
<td>Ill-Defined Sites Central Nervous System</td>
<td>Nervous system NOS C729</td>
</tr>
<tr>
<td></td>
<td>Overlapping lesion of brain and central nervous system C728</td>
</tr>
</tbody>
</table>
## Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions

**C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753**

(Excludes lymphoma and leukemia – M9590-M9992 and Kaposi sarcoma M9140)

<table>
<thead>
<tr>
<th>Site Group</th>
<th>Reportable Subsite Terms and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intracranial Duct and Glands</td>
<td>Craniopharyngeal duct <strong>C752</strong>&lt;br&gt;Pineal gland <strong>C753</strong>&lt;br&gt;Pituitary gland <strong>C751</strong></td>
</tr>
<tr>
<td>Meninges</td>
<td>Cerebral meninges <strong>C700</strong>&lt;br&gt;Meninges NOS <strong>C709</strong>&lt;br&gt;Spinal meninges <strong>C701</strong></td>
</tr>
<tr>
<td>Peripheral Nerve and Autonomic Nervous System</td>
<td>Abdomen <strong>C475</strong>&lt;br&gt;Autonomic nervous system NOS <strong>C479</strong>&lt;br&gt;Head, face and neck <strong>C470</strong>&lt;br&gt;Lower limb and hip <strong>C472</strong>&lt;br&gt;Overlapping lesion of peripheral nerves and autonomic nervous system <strong>C478</strong>&lt;br&gt;Thorax <strong>C473</strong>&lt;br&gt;Trunk NOS <strong>C476</strong>&lt;br&gt;Upper limbs and shoulder <strong>C471</strong>&lt;br&gt;Spinal Nerve NOS <strong>C479</strong></td>
</tr>
<tr>
<td>Spinal Sites</td>
<td>Cauda equina/conus medullaris/filum terminale <strong>C721</strong>&lt;br&gt;Meninges NOS <strong>C709</strong>&lt;br&gt;Spinal meninges <strong>C701</strong></td>
</tr>
</tbody>
</table>
Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
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<table>
<thead>
<tr>
<th>Table 3: Specific Histologies, NOS, and Subtypes/Variants</th>
</tr>
</thead>
</table>

Table 3 lists the more common histologies for CNS and Peripheral Nerves. For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

**Note:** Behavior codes are not listed because all histologies are **malignant** /3.

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

**Note:** All tumors are malignant/invasive /3.

<table>
<thead>
<tr>
<th>Specific and NOS Histology Codes</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anaplastic ganglioglioma 9505</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Astroblastoma 9430</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Astrocytoma NOS 9400</td>
<td>Diffuse astrocytoma IDH-mutant</td>
<td>Anaplastic astrocytoma IDH-mutant/wildtype; anaplastic astrocytoma NOS 9401</td>
</tr>
<tr>
<td></td>
<td>Diffuse astrocytoma IDH-wildtype</td>
<td>Gemistocytic astrocytoma IDH-mutant 9411</td>
</tr>
<tr>
<td></td>
<td>Diffuse astrocytoma NOS</td>
<td></td>
</tr>
<tr>
<td>Choriocarcinoma 9100</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Choroid plexus carcinoma 9390</td>
<td></td>
<td></td>
</tr>
<tr>
<td>CNS embryonal tumor with rhabdoid features 9508</td>
<td>Atypical teratoid/rhabdoid tumor</td>
<td></td>
</tr>
<tr>
<td>CNS ganglioneuroblastoma 9490</td>
<td>Embryonal tumor with rhabdoid features</td>
<td>CNS embryonal tumor 9473</td>
</tr>
<tr>
<td>CNS neuroblastoma 9500</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
### Specific and NOS Histology Codes

<table>
<thead>
<tr>
<th>Code</th>
<th>Definition</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>9385*</td>
<td>Diffuse midline glioma H3 K27M mutant</td>
<td></td>
<td>Yolk sac tumor 9071</td>
</tr>
<tr>
<td>9070</td>
<td>Embryonal carcinoma</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9478*</td>
<td>Embryonal tumor with multilayered rosettes C19MC-altered</td>
<td>Embryonal tumor with multilayered rosettes, NOS ETMR</td>
<td>Anaplastic ependymoma 9392 Ependymoma, RELA fusion-positive 9396* Papillary ependymoma 9393</td>
</tr>
<tr>
<td>9391</td>
<td>Ependymoma</td>
<td>Clear cell ependymoma</td>
<td>Anaplastic ependymoma 9392 Ependymoma, RELA fusion-positive 9396* Papillary ependymoma 9393</td>
</tr>
<tr>
<td>9133*</td>
<td>Epithelioid hemangioendothelioma</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9064</td>
<td>Germinoma</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9440</td>
<td>Glioblastoma NOS</td>
<td>Glioblastoma multiforme</td>
<td>Glioblastoma IDH-mutant 9445* Giant cell glioblastoma 9441 Gliosarcoma 9442</td>
</tr>
<tr>
<td>9080</td>
<td>Immature teratoma</td>
<td>Glioblastoma, IDH wild-type Epithelioid glioblastoma</td>
<td>Mixed germ cell tumor 9085 Teratoma with malignant transformation 9084</td>
</tr>
<tr>
<td>9530</td>
<td>Malignant meningioma</td>
<td>Anaplastic meningioma</td>
<td>Papillary/rhabdoid meningioma 9538</td>
</tr>
<tr>
<td>9540</td>
<td>Malignant peripheral nerve sheath tumor</td>
<td>Epithelioid malignant peripheral nerve sheath tumor Malignant perineurioma MPNST MPNST with perineural differentiation</td>
<td></td>
</tr>
<tr>
<td>Specific and NOS Histology Codes</td>
<td>Synonyms</td>
<td>Subtypes/Variants</td>
<td></td>
</tr>
<tr>
<td>---------------------------------</td>
<td>----------</td>
<td>------------------</td>
<td></td>
</tr>
<tr>
<td>Medulloblastoma NOS 9470</td>
<td>Classic medulloblastoma</td>
<td>Anaplastic/large cell medulloblastoma 9474 Medulloblastoma 9471 Desmoplastic SHH-activated and TP53-wildtype With extensive nodularity Nodular Medulloblastoma non-WNT/non-SHH; medulloblastoma group 3 or group 4 9477* Medulloblastoma SHH-activated and TP53-mutant 9476* Medulloblastoma WNT-activated 9475*</td>
<td></td>
</tr>
<tr>
<td>Medulloepithelioma 9501</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Meningeal melanoma 8720</td>
<td></td>
<td>Meningeal melanomatosis 8728</td>
<td></td>
</tr>
<tr>
<td>Oligoastrocytoma NOS 9382</td>
<td>Anaplastic oligoastrocytoma NOS</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Oligodendroglioma NOS 9450</td>
<td>Anaplastic oligodendroglioma NOS Oligodendroglioma 1p/19q-codeleted Oligodendroglioma IDH-mutant Oligodendroglioma IDH-mutant and 1p/19q-codeleted</td>
<td>Anaplastic oligodendroglioma 9451 IDH-mutant 1p/19q-codeleted IDH-mutant and 1p/19q-codeleted</td>
<td></td>
</tr>
<tr>
<td><strong>Note:</strong> Oligodendroglioma NOS is used when molecular markers cannot fully be determined</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Peripheral primitive neuroectodermal tumor 9364</td>
<td>Ewing sarcoma PpNET</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pilocytic astrocytoma 9421</td>
<td></td>
<td>Pilomyxoid astrocytoma 9425</td>
<td></td>
</tr>
<tr>
<td><strong>Note:</strong> ICD-O-3 lists a behavior code of /1. It is collected as /3 in North America.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pineal parenchymal tumor of intermediate differentiation 9362</td>
<td>Pineoblastoma</td>
<td>Papillary tumor of the pineal region 9395</td>
<td></td>
</tr>
<tr>
<td>Specific and NOS Histology Codes</td>
<td>Synonyms</td>
<td>Subtypes/Variants</td>
<td></td>
</tr>
<tr>
<td>---------------------------------</td>
<td>----------</td>
<td>------------------</td>
<td></td>
</tr>
<tr>
<td>Pleomorphic xanthroastrocytoma 9424</td>
<td>Anaplastic pleomorphic xanthroastrocytoma</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sarcoma NOS 8800</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Note 1:</strong> Chondrosarcoma 9220 has the following subtype/variant:</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mesenchymal chondrosarcoma 9240</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Note 2:</strong> Leiomyosarcoma 8890 has the following subtypes/variants:</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Epithelioid leiomyosarcoma 8891</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Myxoid leiomyosarcoma 8896</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Solitary fibrous tumor grade 3 8815</td>
<td>Hemangiopericytoma grade 3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Solitary fibrous tumor/Hemangiopericytoma grade 3 (CNS)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* These new codes were approved by the IARC/WHO Committee for ICD-O
### Table 4: Coding Primary Site for Malignant Tumors of Cranial and Peripheral Nerves

**Note 1:** Neoplasms arising in a cranial or spinal nerve are coded to the specific nerve in which it arises.

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

**Note 3:** Neoplasms, commonly meningiomas, arising in the dura/meninges of the spinal nerve roots are coded to the ICD-O site code spinal meninges C701.

**Note 4:** It is important to check the operative report to determine whether the surgery is intracranial or intradural.

Use Table 4 to determine whether a malignant cranial nerve tumor should be coded to a cranial nerve or to a peripheral nerve. The anatomic location of the tumor determines whether the primary site is a peripheral or a cranial nerve.

**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:** Portions of the nerve coded to cranial nerve

**Column 4:** Portions of nerve coded to peripheral nerve

<table>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Site Code: Cranial Nerve</th>
<th>Site Code: Peripheral Nerve</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cranial nerve NOS</td>
<td></td>
<td>Within cranium, unknown which nerve C725</td>
<td></td>
</tr>
<tr>
<td>Olfactory CN 1</td>
<td>Cribriform plate</td>
<td>Surface of the brain C722</td>
<td>Originates on the olfactory mucosa of nasal cavity, then travels through the cribriform plate of the ethmoid bone C470</td>
</tr>
<tr>
<td>Optic CN 2</td>
<td>Optic canal</td>
<td>All portions are covered by meninges/dura so are reportable as C723</td>
<td></td>
</tr>
<tr>
<td>Oculomotor CN 3</td>
<td>Superior orbital fissure</td>
<td>Originates in the midbrain C725</td>
<td>After exiting the superior orbital fissure, the nerve enters the orbit C470</td>
</tr>
</tbody>
</table>
### Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions

C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753

(Excludes lymphoma and leukemia – M9590-M9992 and Kaposi sarcoma M9140)

<table>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Site Code: Cranial Nerve</th>
<th>Site Code: Peripheral Nerve</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trochlear CN 4</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 C725</strong></td>
<td>Enters the orbital fissure <strong>C470</strong></td>
</tr>
</tbody>
</table>

**Note:** 
- Trochlear is derived from Latin *trigeminus* which means *born in threes* (tri) and *born at the same time* (germinal).
- As the name implies, the nerve **separates** into three branches; ophthalmic, maxillary, and mandibular.

| Trigeminal CN 5     | The ophthalmic, maxillary and mandibular branches leave the skull through three separate foramina. Superior orbital fissure. The foramen rotundum. 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 C725**. | **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 superius**, and divides into multiple branches that spread to the side of the **nose**, the lower **eyelid**, and the upper **lip C470** |
|                     |                                      |                          | **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 **C470** |
|                     |                                      |                          | **Mandibular nerve leaves** via the foramen ovale travels |

Jump to [Multiple Primary Rules](#)  
Jump to [Histology Coding Rules](#)
# Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions

C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia – M9590-M9992 and Kaposi sarcoma M9140)

<table>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Site Code: Cranial Nerve</th>
<th>Site Code: Peripheral Nerve</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abducent CN 6</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 C725</strong></td>
<td><strong>Dorello’s canal</strong> and travels to the tip of the <strong>temporal bone</strong>. Enters <strong>orbit C470</strong></td>
</tr>
<tr>
<td>Facial CN 7</td>
<td>Internal acoustic meatus</td>
<td>CN7 originates in the <strong>pons</strong>, along the posterior cranial fossa (<strong>posterior cranial fossa</strong>) (the posterior cranial fossa is part of the intracranial cavity.) C725</td>
<td>Enters the temple through the internal auditory meatus and runs through the facial canal. C470</td>
</tr>
<tr>
<td>Acoustic or vestibulocochlear CN 8</td>
<td>Internal acoustic meatus</td>
<td>Originates in the <strong>brain stem (medulla oblongata)</strong> between the base of the brain (<strong>pons</strong>) and the <strong>spinal cord C724</strong></td>
<td>Both the <strong>vestibular</strong> branch and the <strong>cochlear</strong> branch are located in the <strong>inner ear</strong></td>
</tr>
</tbody>
</table>
| Glossopharyngeal CN 9 | Jugular foramina | **Originates** in the anterior portion of the **medulla oblongata C725** | Jugular foramen
Between the **internal jugular vein** and **internal carotid** artery
Lies on the **stylopharyngeus** and middle **pharyngeal** constrictor muscle
Passes under the **hypoglossus** muscle
Palatine tonsil
Extends to mucous **glands** of the **mouth**, and **base** of the **tongue** C470 |
### Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions

**C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753**

(Excludes lymphoma and leukemia – M9590-M9992 and Kaposi sarcoma M9140)

<table>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Site Code: Cranial Nerve</th>
<th>Site Code: Peripheral Nerve</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vagus CN 10</td>
<td>Jugular foramen</td>
<td>The vagus nerve originates from the medulla of the brainstem. C725</td>
<td>CN10 descends within the <strong>carotid sheath</strong> medial to the internal jugular vein at the root of the neck C470. The right vagus crosses in front of the subclavian artery and travels into the fat behind the blood vessels, reaching the <strong>thorax</strong>. It then inclines behind the hilum of the <strong>right lung</strong> and moves toward the esophagus. The nerve splits into the <strong>right and left</strong> vagus at the <strong>esophageal plexus C473</strong>. Forms the anterior and posterior <strong>gastric nerves C475</strong></td>
</tr>
<tr>
<td>Accessory CN 11</td>
<td>Jugular foramen</td>
<td>The spinal accessory nerve originates in the neurons of the upper spinal cord, specifically C1-C5/C6 spinal nerve roots. The nerve enters the foramen magnum or lateral aspect of the <strong>medulla oblongata</strong>. The fibers of the spinal accessory nerve coalesce to form <strong>spinal rootlets, roots</strong>, and finally the <strong>spinal accessory nerve</strong> itself C725</td>
<td>The nerve exits the skull through the jugular foramen. It then runs along the internal carotid artery within the <strong>neck C470</strong>. Reaches the sternocleidomastoid muscle and the trapezius C476</td>
</tr>
<tr>
<td>Hypoglossal CN 12</td>
<td>Hypoglossal canal</td>
<td>CN12 starts in the <strong>hypoglossal nucleus of the brainstem, C725</strong></td>
<td>CN12 exits the hypoglossal canal, traveling between the <strong>carotid artery</strong> and <strong>jugular vein</strong>, ending under the <strong>tongue C470</strong></td>
</tr>
</tbody>
</table>
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 behavior 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.
Use Table 5 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 common for glioblastoma multiform and meningiomas.

**Note 2:** SEER allows laterality to be coded for sites other than those in the table.
Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia – M9590-M9992 and Kaposi sarcoma M9140)

Table 6: Non-malignant CNS Tumors with the Potential of Transforming to Malignant Behavior

The word “transformation” as used in this table means that:
  o Residual tumor becomes more aggressive /3 OR
  o 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).

Use Table 6 when directed to by the Multiple Primary Rules.

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>Hemangioendothelioma 9159/1</td>
<td>Anaplastic hemangioendothelioma 9159/3</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>
Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753
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Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions

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Malignant CNS and Peripheral Nerves Multiple Primary Rules
C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia – M9590-M9992 and Kaposi sarcoma M9140)

Note 1: Non-malignant intracranial and CNS tumors have a separate set of rules.
Note 2: Laterality is not used to determine multiple primaries for malignant CNS tumors.
Note 3: Timing is not used to determine multiple primaries for malignant CNS tumors.
Note 4: GBM following an astrocytic or glial tumor is a multiple primary.
Note 5: These rules are NOT used for tumor(s) described as metastases.
Note 6: 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 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.

Prepare one abstract. Use the histology rules to assign the appropriate histology code.
Malignant CNS Solid Tumor Rules 2018
Updated 8/16/2018

Single Tumor

| Rule M2 | Abstract a single primary 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 adjacent/contiguous site or subsites.  
| Note 3: | The tumor may have two or more histologic components. |

| Rule M3 | Code a single primary when a neoplasm is originally diagnosed as an oligodendrogloma and subsequently recurs in residual tumor tissue with different features such as a densely cellular tumor with pseudo palisading necrosis.  
| Note 1: | The pathology may state that the recurrence “looks like” or “has the appearance of” a glioblastoma multiforme (GBM). This is not a true GBM.  
| Note 2: | Record as a recurrence for those registrars who collect recurrence data. |

| Rule M4 | Abstract a single primary (the malignant) when a single tumor meets the following two criteria:  
| 1. | The original diagnosis was non-malignant /0 or /1 AND  
| o | 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: | 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 3: | There is no time requirement from initial diagnosis to resection.  
| Note 4: | Edit the original abstract as follows:  
| • | Do not change date of diagnosis. |
Malignant CNS and Peripheral Nerves Multiple Primary Rules
C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia – M9590-M9992 and Kaposi sarcoma M9140)

- 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.
- See the COC and SEER manuals for **instructions** on coding other data items such as Accession Year, Treatment and Sequence Number.

**Note 5:** 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.

This is the end of instructions for Single Tumor.

1 Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.
Multiple Tumors

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

IMPORTANT: The major difference between M4 and M5 is:
M4: 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 there are multiple CNS tumors, one of which is malignant /3 and the other is non-malignant /0 or /1.
- Original non-malignant tumor followed by malignant tumor
  - Patient had a resection of the non-malignant tumor (not the same tumor) OR
  - It is unknown/not documented if the patient had a resection
- Simultaneous non-malignant and malignant tumors
  - Abstract both the malignant and the non-malignant tumors

Note 1: The rules are hierarchical. Only use when previous rules do not apply.
Note 2: See Table 2 in the Equivalent Terms and Definitions for a listing of CNS sites.
Note 3: A non-malignant CNS tumor and a malignant CNS tumor are always multiple primaries (timing and primary sites are irrelevant). Prepare two abstracts; one for the non-malignant and another for the malignant tumor.

Rule M6  Abstract multiple primaries when a patient has a glial or astrocytic tumor and is subsequently diagnosed with a glioblastoma multiforme 9440 (GBM).

Note 1: This is a change from the 2007 Rules.
Note 2: Abstracting GBM as a new primary will allow analysis of:
- The number of tumors that recur as a more aggressive histology (GBM)
- The time interval between occurrence of the glial or astrocytic tumors and a GBM
- Which histologies are more likely to recur as a GBM
Malignant CNS and Peripheral Nerves Multiple Primary Rules
C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia – M9590-M9992 and Kaposi sarcoma M9140)

Rule M7  Abstract a single primary\(^1\) when there are separate, non-contiguous tumors in the brain (multicentric/multifocal). Tumors may be any of the following combinations:
- 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)
- In different lobes; for example, parietal lobe C713 and occipital lobe C714 (different site codes)

Example: The patient had a resection of an anaplastic astrocytoma 9401 in the right parietal lobe. Three months later the patient is diagnosed with a de novo anaplastic astrocytoma in the left parietal lobe. This is one primary because neither laterality nor timing are used to determine multiple primary status.

Note 1: Multiple sites/subsites and/or different lateralities imply either metastatic or multifocal/multicentric disease. The following histologies commonly have multiple tumors which are described as multifocal/multicentric:
- Glioblastoma multiforme
- Gliomatosis cerebri

Note 2: Metastases are never used to determine multiple primaries. Seeding metastasis is often noted for the following tumors:
- Glioblastoma multiforme
- pNET-medulloblastoma
- Oligodendroglioma

Note 3: Hereditary syndromes frequently exhibit multiple tumors including the following:
- Neurofibromatosis type 1 (NF1)
  - Malignant peripheral nerve sheath tumors (MPNST)
- Neurofibromatosis type 2 (NF2)
  - Anaplastic ependymomas
  - Meningiomas

Note 4: Most malignant neoplasms are single tumors with the exception of those listed in this rule.

Note 5: This is a change from/clarification to previous rules.

Rule M8  Abstract multiple primaries\(^2\) when multiple tumors are present in any of the following sites or subsites:
- Any lobe of the brain C710-C719 AND any other part of CNS
- Cerebral meninges C700 AND spinal meninges C701
- Cerebral meninges C700 AND any other part of CNS
- Any of the cranial nerves C721-C725 AND any other part of the CNS
- Meninges of cranial or peripheral nerves C709 AND any other part of the CNS
- Spinal meninges C701 AND any other part of CNS
Malignant CNS and Peripheral Nerves Multiple Primary Rules
C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia – M9590-M9992 and Kaposi sarcoma M9140)

Rule M9  Abstract multiple primaries\(^i\) when separate, non-contiguous tumors are two or more different subtypes/variants in Column 3, Table 3 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: Anaplastic astrocytoma IDH-mutant 9401 and gemistocytic astrocytoma IDH-mutant 9411 are both subtypes of astrocytoma NOS 9400/3 but are distinctly different histologies. Abstract multiple primaries.
- Different NOS: Papillary ependymoma 9393 is a subtype of ependymoma NOS 9391; gliosarcoma 9442 is a subtype of glioblastoma NOS 9440. They are distinctly different histologies. Abstract multiple primaries.

Rule M10  Abstract a single primary\(^i\) when separate, non-contiguous tumors are on the same row in Table 3 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)

Rule M11  Abstract multiple primaries\(^i\) when separate, non-contiguous tumors are on different rows in Table 3 in the Equivalent Terms and Definitions. Timing is irrelevant.
Note:  Each row in the table is a distinctly different histology.

Rule M12  Abstract a single primary\(^i\) when multiple tumors do not meet any of the above criteria.
Note:  Use this rule as a last resort. Please confirm that you have not overlooked an applicable rule.

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.
Malignant CNS and Peripheral Nerves Histology Rules
C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590-M9992 and Kaposi sarcoma M9140)

Note: Non-malignant CNS tumors have a separate set of rules.

Priority Order for Using Documentation to Identify Histology

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. Biomarkers
      Note 1: Biomarkers do not identify all histologic types.
      Note 2: Biomarkers are not listed because they change rapidly.
      Example: Biomarkers are required to diagnose medulloblastoma SHH-activated and TP53-wildtype. Biomarkers include genetic testing, FISH, MYCN.
   B. The addendum and/or comments
   C. Final diagnosis
   D. CAP protocol/summary
2. Pathology/tissue from biopsy
   A. Biomarkers
      Note 1: Biomarkers do not identify all histologic types.
      Note 2: Biomarkers are not listed because they change rapidly.
      Example: Biomarkers are required to diagnose medulloblastoma SHH-activated and TP53-wildtype. Biomarkers include genetic testing, FISH, MYCN.
   B. The addendum and/or comments
   C. Final diagnosis
   D. CAP protocol/summary
   Note 1: Addendums and comments 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

Jump to Equivalent Terms and Definitions
Jump to Multiple Primary Rules
Malignant CNS and Peripheral Nerves Histology Rules
C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753
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*Note*: The synoptic portion of CAP must be documented in one location. Most frequently, in the:
- The pathology final diagnosis
- Addendum to the path report

3. **Cytology** (most frequently cerebrospinal fluid)
4. Tissue/pathology from a metastatic site
   *Note 1*: Code the behavior /3
   *Note 2*: The tissue from a metastatic site often shows variations from the primary tumor. When it is the only tissue available, it is more accurate than a scan.

5. **Scan**: The following list is in priority order.
   A. MRI
   B. CT
   C. PET
   D. Angiogram

6. Code the histology documented by the physician when none of the above are available. Use the documentation in the following priority order
   A. Documentation from Tumor Board
   B. Documentation in the medical record that refers to original pathology, cytology, or scan(s)
   C. Physician’s reference to type of cancer (histology) in the medical record
      *Note 1*: Code the specific histology when documented.
      *Note 2*: Code the histology to malignant neoplasm NOS 8000 or as stated by the physician when nothing more specific is documented.
Malignant CNS and Peripheral Nerves Histology Rules
C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590-M9992 and Kaposi sarcoma M9140)

Coding Multiple Histologies

1. **Code** the histology when the:
   A. **Exact term is documented OR**
   B. **Histology is described as**
      - Subtype
      - Type
      - Variant

2. **Do not** code the histology when:
   A. The following modifiers are used as a descriptor:
      - Architecture
      - Differentiation
      *Note:* Only code differentiation when there is a specific code for the NOS with differentiation in Table 3 in the Equivalent Terms and Definitions, ICD-O and all updates.
      - Features (of)/with features of
      *Note:* Only code features when there is a specific code for the NOS with features in Table 3 in the Equivalent Terms and Definitions, ICD-O and all updates.
      - Foci; focus, focal
      - Major/majority of
      *Note:* Major/majority describes the greater amount of tumor.
      - Pattern(s)
      - Predominantly
      *Note:* Predominantly describes the greater amount of tumor.
   B. The following ambiguous terms are used as a descriptor:
      - Apparently
      - Appears
      - Comparable with
      - Compatible with
      - Consistent with
      - Favor(s)
Malignant CNS and Peripheral Nerves Histology Rules
C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590-M9992 and Kaposi sarcoma M9140)

- Malignant appearing
- Most likely
- Presumed
- Probable
- Suspect(ed)
- Suspicious (for)
- Typical (of)

Note 1: See SEER Program Manual and COC Manual. Ambiguous terminology is used to determine reportability.
Note 2: Histology described by ambiguous terminology is coded ONLY when a case is accessioned based on ambiguous terminology and no other histology information is availabledocumentoed.

Single Tumor

**Rule H1** Code malignant meningioma 9530 when the diagnosis specifically states malignant/invasive.
*Note:* Code the behavior designated in the pathology report. Do not code malignant /3 based on invasion of brain or skull. Atypical meningioma (non-malignant) may invade contiguous structures.

**Rule H2** Code the histology when only one histology is present.
*Note 1:* Use Table 3 to code histology. New codes, terms, and synonyms are included in Table 3 and coding errors may occur if the table is not used.
*Note 2:* When the histology is not listed in Table 3, 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 3, ICD-O or all updates.

**Rule H3** Code the subtype/variant when there is a NOS and a single subtype/variant of that NOS such as the following:
*Note:* All tumors are malignant/invasive /3.
- Astrocytoma 9400 and a subtype/variant of astrocytoma
- CNS ganglioneuroblastoma 9490 and a subtype/variant of CNS ganglioneuroblastoma
- Embryonal carcinoma 9070 and a subtype/variant of embryonal carcinoma
- Ependymoma 9391 and a subtype/variant of ependymoma
- Glioblastoma 9440 and a subtype/variant of glioblastoma
Malignant CNS and Peripheral Nerves Histology Rules
C470-C479, C700, C701, C709, C710-C719, C721-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590-M9992 and Kaposi sarcoma M9140)

- Immature teratoma 9080 and a subtype/variant of immature teratoma
- Malignant meningioma 9530 and a subtype/variant of malignant meningioma
- Malignant peripheral nerve sheath tumor 9540 and a subtype/variant of malignant peripheral nerve sheath tumor
- Medulloblastoma 9470 and a subtype/variant of medulloblastoma
- Meningeal melanoma 8720 and a subtype/variant of meningeal melanoma
- Oligodendroglioma 9450 and a subtype/variant of oligodendroglioma
- Pilocytic astrocytoma 9421 and a subtype/variant of pilocytic astrocytoma
- Pineal parenchymal tumor of intermediate differentiation 9362 and a subtype/variant of pineal parenchymal tumor of intermediate differentiation
- Sarcoma 8800 and a subtype/variant of sarcoma

Note: See Table 3 in the Equivalent Terms and Definitions to find NOS and subtypes/variants.

This is the end of instructions for Single Tumor

Multiple Tumors Abstracted as a Single Primary

Rule H4 Code malignant meningioma 9530 when the diagnosis specifically states malignant/invasive.

Note: Code the behavior designated in the pathology report. Do not code malignant /3 based on invasion of brain or skull. Atypical meningioma (non-malignant) may invade contiguous structures.

Rule H5 Code the histology when only one histology is present in all tumors.

Note 1: Use Table 3 to code histology. New codes, terms, and synonyms are included in Table 3 and coding errors may occur if the table is not used.

Note 2: When the histology is not listed in Table 3, 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 3, ICD-O or all updates.
Rule H6  Code the subtype/variant when all tumors are a NOS and a single subtype/variant of that NOS such as the following:
Note: All tumors are malignant/invasive /3.

- Astrocytoma 9400 and a subtype/variant of astrocytoma
- CNS ganglioneuroblastoma 9490 and a subtype/variant of CNS ganglioneuroblastoma
- Embryonal carcinoma 9070 and a subtype/variant of embryonal carcinoma
- Ependymoma 9391 and a subtype/variant of ependymoma
- Glioblastoma 9440 and a subtype/variant of glioblastoma
- Immature teratoma 9080 and a subtype/variant of immature teratoma
- Malignant meningioma 9530 and a subtype/variant of malignant meningioma
- Malignant peripheral nerve sheath tumor 9540 and a subtype/variant of malignant peripheral nerve sheath tumor
- Medulloblastoma 9470 and a subtype/variant of medulloblastoma
- Meningeal melanoma 8720 and a subtype/variant of meningeal melanoma
- Oligodendrogioma 9450 and a subtype/variant of oligodendrogioma
- Pilocytic astrocytoma 9421 and a subtype/variant of pilocytic astrocytoma
- Pineal parenchymal tumor of intermediate differentiation 9362 and a subtype/variant of pineal parenchymal tumor of intermediate differentiation
- Sarcoma 8800 and a subtype/variant of sarcoma

Note: See Table 3 in the Equivalent Terms and Definitions to find NOS and subtypes/variants.

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

Code the histology using the rule that fits the case.