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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; 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: 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.

- See the Non-malignant CNS Rules when the primary site is optic nerve and the diagnosis is either optic glioma or pilocytic astrocytoma. The behavior for these tumors is non-malignant and coded 9421/1.
- **IMPORTANT FOR 2023 FORWARD: Beginning 1/1/2023, all cases diagnosed with pilocytic astrocytoma/juvenile pilocytic astrocytoma and new related terminology are to be reported with behavior /1. They will no longer be collected with malignant behavior (/3). See the Non-malignant CNS Rules.**

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.

Note 9: 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**.
- 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
- Infratentorial; below the tentorium cerebelli; either cerebellum or brainstem
- Intracranial; within the skull, within the cranium
- Intradural; between layers of the cerebral meninges; C700
- Intradural-extramedullary; within the spinal canal but outside of the nerves
- Intraspinal; occurring within the spinal column especially the vertebral canal; spinal nerve roots
- 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

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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/type/variant**
  Note: Component is only coded when the pathologist specifies the component as a second **carcinoma**.
- **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. 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. For example, gliomatosis cerebri is a term which is no longer recommended. Gliomatosis cerebri is now termed a "growth pattern" rather than a histologic type.
   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. For example, glioma NOS is an umbrella term for all gliomas and astrocytomas. Glioma NOS is not recommended because diagnostic methodology is able to determine a more specific diagnosis.
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. **Clarifications:**
   A. The following meningiomas are reportable: intraosseous, cavernous sinus and sphenoid wing.
   B. Multiple cerebral meningiomas are a single primary.
   C. Multiple brain tumors (same histology) are a single primary.
   D. Laterality is not used to determine multiple primaries.
   E. Timing is not used to determine multiple primaries.
   F. The brain (C710-C719) is a single primary site.
   G. Neurofibromatosis NOS, Neurofibromatosis 1 (NF1), Neurofibromatosis 2 (NF2), and schwannomatosis are genetic syndromes and not reportable neoplasms. People with this genetic syndrome do have a high risk of developing:
      i. Non-reportable non-malignant tumors occurring in skin and sites other than CNS AND
      ii. Reportable malignant tumors
4. 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.

New for 2023

1. Beginning with cases diagnosed 1/1/2023, pilocytic astrocytoma/juvenile pilocytic astrocytoma will **no longer be reported as malignant (/3)**. These neoplasms will continue to be reportable 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.
2. WHO 5th Ed CNS Tumors has assigned a new histology to 9421/3. Beginning with cases diagnosed 1/1/2023, for surveillance purposes, code 9421/3 will be valid for the following histology ***only:***
   A. High Grade astrocytoma with piloid features (HGAP)

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Reportability Criteria

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**)

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Directory of Sections and Tables

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

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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**
   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. **Scans**, 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.

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Table 1: WHO Grades for Select CNS Neoplasms

Note 1: CNS WHO classifications use a grading scheme that is a "malignancy scale" ranging across a wide variety of neoplasms rather than a strict histologic grading system that can be applied equally to all tumor types.
Note 2: See the SEER and COC Manuals for instructions on coding grade for CNS tumors.
Note 3: The table **does not** contain all neoplasms that may occur in the CNS.
Note 4: WHO does not provide Grades for all CNS and peripheral nerve neoplasms.

WHO Grade Definitions

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

WHO Grade II CNS Tumors: Non-malignant and Malignant

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

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

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

When a CNS tumor is stated to be WHO Grade II, code the corresponding ICD-O-3 code and behavior listed in ICD-O, ICD-O updates, or solid tumor rules. Code the WHO Grade in the appropriate SSDI data field.
Example: Astrocytoma, NOS, WHO Grade II code 9400/3

Table Instructions

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

Column 1 contains the **histology** term.
Column 2 contains the **WHO Grade** assigned **based** on the **molecular features** of the histology.

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

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Histology	WHO Grade
Atypical meningioma	2
Atypical teratoid/rhabdoid tumor	4
Central neurocytoma	2
Cerebellar liponeurocytoma	2
Chordoid glioma of third ventricle	2
Choroid plexus carcinoma	3
Choroid plexus papilloma	1
CNS embryonal tumor NOS	4
CNS embryonal tumor with rhabdoid features	4
Craniopharyngioma	1
Desmoplastic infantile astrocytoma and ganglioglioma	1
Diffuse astrocytoma, IDH-mutant	2
Diffuse midline glioma, H3K27M-mutant	4
Dysembryoplastic neuroepithelial tumor	1
Dysplastic gangliocytoma of cerebellum (Lhemitte-Duclos)	1
Ependymoma	2
Ependymoma, RELA fusion-positive ***Note:*** Tissue/pathology reports or CAP protocol/summary will specify whether this is WHO Grade 2 or 3	2 or 3
Extraventricular neurocytoma	2
Gangliocytoma	1
Glioblastoma, IDH-mutant	4
Glioblastoma, IDH-wildtype	4
Granular cell tumor	1
Hemangioblastoma	1

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Histology	WHO Grade
Malignant peripheral nerve sheath tumor (MPNST) ***Note:*** Tissue/pathology reports or CAP protocol/summary will specify whether this is WHO Grade 2, 3, or 4	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 ***Note:*** Tissue/pathology reports or CAP protocol/summary will specify whether it is WHO Grade 2 or 3	2 or 3
Perineuroma	1
Pilocytic astrocytoma ***Note:*** Collected as malignant /3 in North America	1
Pineal parenchymal tumor of intermediate differentiation ***Note****:* Tissue/pathology reports or CAP protocol/summary will specify WHO Grade 2 or 3	2 or 3
Pineoblastoma	4
Pineocytoma	1
Pituicytoma	1
Pleomorphic xanthroastrocytoma	2
Rosette-forming glioneuronal tumor	1
Schwannoma	1

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Histology	WHO Grade
Solitary fibrous tumor/hemangiopericytoma ***Note:*** Tissue/pathology reports or CAP protocol/summary will specify WHO Grade 1, 2, or 3	1, 2, or 3
Spindle cell oncocytoma	1
Subependymal giant cell astrocytoma	1
Subependymoma	1

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Section 2: Reportable Primary Sites and Histologies

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

Note 1: **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: 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

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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.

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

Continued on next page

2. **Spinal** sites: includes the **spinal meninges** and **all** structures **within** the meninges (Intradural, within/in the spinal meninges).
   A. **Spinal cord C720**
   B. **Spinal meninges C701** covers/encloses the spinal nerve roots and the spinal cord.
   C. 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

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Table 2: Reportable Primary Sites

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

Site Group	Reportable Subsite Terms and Code
Brain	Brain NOS **C719** Brain stem **C717** Cerebellum NOS **C716** Cerebrum **C710** Frontal lobe **C711** Occipital lobe **C714** Overlapping lesion of brain **C718** Parietal lobe **C713** Temporal lobe **C712** Ventricle NOS **C715**
Cranial Nerves	Abducent (cranial nerve VI) **C725** Accessory (cranial nerve XI) **C725** Acoustic (cranial nerve VIII) **C724** Cranial nerve NOS **C725** Facial (cranial nerve VII) C**725** Glossopharyngeal (cranial nerve IX) **C725** Hypoglossal (cranial nerve XII) **C725** Oculomotor (cranial nerve III) **C725** Olfactory (cranial nerve I) **C722** Optic (cranial nerve II) **C723** Trigeminal (cranial nerve V) **C725** Trochlear (cranial nerve IV) **C725** Vagus (cranial nerve X) **C725**

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Site Group	Reportable Subsite Terms and Code
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**
Peripheral Nerve and Autonomic Nervous System	Abdomen **C474** Autonomic nervous system NOS **C479** Head, face and neck **C470** Lower limb and hip **C472** Nerves of pelvis **C475** Overlapping lesion of peripheral nerves and autonomic nervous system **C478** Thorax **C473** Trunk NOS **C476** Upper limbs and shoulder **C471** Spinal Nerve NOS **C479**
Spinal Sites	Cauda equina/ **C721** Conus medullaris/filum terminale **C720** Meninges NOS **C709** Spinal cord **C720** Spinal meninges **C701**

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Table 3: Specific Histologies, NOS, and Subtypes/Variants

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.

Column 3 may contain NOS histologies which are part of a bigger histologic group. For example, sarcoma NOS 8800/3 (column 1) is a generic term that encompasses a number of soft tissue tumors, including chondrosarcoma 9220/3 (column 3). Chondrosarcoma is also a NOS because it has a subtype/variant, mesenchymal chondrosarcoma 9240. The subtype/variant is indented under the NOS (chondrosarcoma) in column 3. There is also a note in column 1 which calls attention to the fact that chondrosarcoma has a subtype/variant.

When subtypes/variants are indented under a NOS in Column 3, use coding rules for a NOS and a single subtype/variant. For example, chondrosarcoma **9220** and mesenchymal chondrosarcoma **9240** are a NOS and a subtype/variant, **NOT** two different subtypes.

Table begins on next page

Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Anaplastic ganglioglioma **9505**		
Astroblastoma **9430**	Astroblastoma, MN1-altered	
Astrocytoma NOS **9400**	Astrocytoma, IDH-mutant, grade 2 Diffuse astrocytoma IDH-mutant Diffuse astrocytoma IDH-wildtype Diffuse astrocytoma NOS	Anaplastic astrocytoma IDH-mutant/wildtype; anaplastic astrocytoma NOS **9401** Astrocytoma, IDH-mutant, grade 3 **9401** Astrocytoma, IDH-mutant, grade 4 **9445** Gemistocytic astrocytoma IDH-mutant **9411** Pleomorphic xanthroastrocytoma /anaplastic pleomorphic xanthroastrocytoma **9424**
Choriocarcinoma **9100**		
Choroid plexus carcinoma **9390**		
CNS embryonal tumor with rhabdoid features **9508**	Atypical teratoid/rhabdoid tumor Embryonal tumor with rhabdoid features	
CNS ganglioneuroblastoma **9490**		CNS embryonal tumor, NEC/NOS **9473**
CNS neuroblastoma **9500**	CAN neuroblastoma, FOXR2-activated CNS Tumor with BCOR internal tandem duplication	
Diffuse leptomeningeal glioneuronal tumor **9509*** ***Note 1***: Cases diagnosed prior to 1/1/2023 are coded 9509/1. See the non-malignant CNS rules. ***Note 2:*** Cases diagnosed 1/1/2023 forward are coded 9509/3	DLGNT	

Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
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Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Diffuse midline glioma H3 K27M mutant **9385***	Diffuse intrinsic pontine glioma Diffuse hemispheric glioma, H3 G34-mutant Diffuse pediatric-type high grade glioma, H3-wildtype and IDH-wildtype DIPG Infant-type hemispheric glioma	
Embryonal carcinoma **9070**		Yolk sac tumor **9071**
Embryonal tumor with multilayered rosettes C19MC-altered **9478***	Embryonal tumor with multilayered rosettes, NOS ETMR	
Ependymoma **9391** ***Note:*** The following terms are synonyms of ependymoma, RELA fusion-positive 9396, and are NOT subtypes/variants of it. They are all coded 9396. • Posterior fossa group A (PFA) ependymoma • Posterior fossa group B (PFB) ependymoma • Spinal ependymoma, MYCN-amplified • Supratentorial ependymoma, YAP1 fusion-positive • Supratentorial ependymoma, ZFTA fusion-positive	Clear cell ependymoma Posterior fossa ependymoma, NOS Spinal ependymoma, NOS Supratentorial ependymoma, NOS Tanycytic ependymoma	Anaplastic ependymoma **9392** Ependymoma, RELA fusion-positive **9396*** Posterior fossa group A (PFA) ependymoma Posterior fossa group B (PFB) ependymoma Spinal ependymoma, MYCN-amplified Supratentorial ependymoma, YAP1 fusion-positive Supratentorial ependymoma, ZFTA fusion-positive Papillary ependymoma **9393**
Epithelioid hemangioendothelioma **9133**		
Germinoma **9064**		

Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Glioblastoma NOS **9440**	Glioblastoma multiforme GBM Glioblastoma, IDH wild-type Epithelioid glioblastoma	Giant cell glioblastoma **9441** Glioblastoma IDH-mutant **9445*** Gliosarcoma **9442**
High-grade astrocytoma with piloid features 9421/3 Note: This term is reportable for cases diagnosed 1/1/2023 forward	HGAP	
Immature teratoma **9080**		Mixed germ cell tumor **9085** Teratoma with malignant transformation **9084**
Malignant meningioma **9530**	Anaplastic meningioma	Papillary/rhabdoid meningioma **9538**
Malignant peripheral nerve sheath tumor **9540**	Malignant melanotic nerve sheath tumor Malignant perineurioma MPNST MPNST with perineural differentiation	Epithelioid malignant peripheral nerve sheath tumor **9542**

Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
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Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Medulloblastoma NOS **9470**	Classic medulloblastoma Medulloblastoma, histologically defined	Anaplastic/large cell medulloblastoma **9474** Medulloblastoma described as one of the following **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***
Medulloepithelioma **9501**		
Meningeal melanoma **8720**		Meningeal melanomatosis **8728**
Neuroepithelial tumor, malignant 8000/3		
Oligoastrocytoma NOS **9382**	Anaplastic oligoastrocytoma NOS	
Oligodendroglioma NOS **9450** ***Note:*** Oligodendroglioma NOS is used when molecular markers cannot fully be determined	Oligodendroglioma 1p/19q-codeleted Oligodendroglioma IDH-mutant Oligodendroglioma IDH-mutant and 1p/19q-codeleted, grade 2	Anaplastic oligodendroglioma NOS **9451** IDH-mutant 1p/19q-codeleted IDH-mutant and 1p/19q-codeleted Oligodendroglioma, IDH-mutant and 1p/19q-codeleted, grade 3
Peripheral primitive neuroectodermal tumor **9364**	Ewing sarcoma pPNET	

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Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Pilocytic astrocytoma **9421** ***Note 1:*** ICD-O-3 lists a behavior code of /1. It is collected as /3 in North America for cases diagnosed prior to 1/1/2023. ***Note 2***: Beginning with cases diagnosed 1/1/2023 forward, pilocytic astrocytoma will no longer be reported with /3 behavior. Pilocytic astrocytoma and the related terms listed below are to be reported as a /1 • Diffuse astrocytoma, MTB- or MYBL1-alterd • Diffuse low-grade glioma, MAPK pathway- altered+		Pilomyxoid astrocytoma **9425**
Pineal parenchymal tumor of intermediate differentiation **9362**	Pineoblastoma	Papillary tumor of the pineal region **9395**
Pituitary adenoma/pituitary neuroendocrine tumor **8272/3**	PitNET	

Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
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Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Sarcoma NOS **8800** *Note 1:* **Chondrosarcoma 9220** has the following subtype/variant: Mesenchymal chondrosarcoma **9240** *Note 2:* **Leiomyosarcoma 8890** has the following subtypes/variants: Epithelioid leiomyosarcoma **8891** Myxoid leiomyosarcoma **8896**		Angiosarcoma **9120** Chondrosarcoma **9220** Mesenchymal chondrosarcoma **9240** Leiomyosarcoma/granular cell leiomyosarcoma/inflammatory leiomyosarcoma **8890** Epithelioid leiomyosarcoma **8891** Myxoid leiomyosarcoma **8896** Osteosarcoma **9180** Primary intracranial sarcoma, DICER1-mutant **9480** Undifferentiated pleomorphic sarcoma/malignant fibrous histiocytoma **8802**
Solitary fibrous tumor grade 3 **8815**	Hemangiopericytoma grade 3 Solitary fibrous tumor/Hemangiopericytoma grade 3 (CNS)	

*** These new codes were approved by the IARC/WHO Committee for ICD-O**

Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
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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

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

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Name and CN #	Exits Cranium Through	Site Code: Cranial Nerve	Site Code: Peripheral Nerve
Trochlear CN 4	Superior orbital fissure	Arises from the **dorsal brain stem**, loops around the brainstem and passes anteriorly within the **subarachnoid space**. It travels between the **superior cerebellar and posterior cerebral arteries** and through the **dura,** enters **cavernous sinus C725**	Enters the orbital fissure **C470**
Trigeminal CN 5 ***Note:*** Trigeminal 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	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 foremen ovale travels along the **mandibular** groove **C470**

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Name and CN #	Exits Cranium Through	Site Code: Cranial Nerve	Site Code: Peripheral Nerve
Abducent CN 6	Cranial meninges	Exits brainstem at junction of **pons** and the **medulla**, enters the **subarachnoid** space and runs upward between the pons and the **clivus** entering the **cavernous sinus C725**	**Dorello's canal** and travels to the tip of the **temporal bone**. Enters **orbit C470**
Facial CN 7	Internal acoustic meatus	**CN7** originates in the **pons,** along the posterior cranial fossa (**posterior cranial fossa** (the posterior cranial fossa is part of the intracranial cavity.) **C725**	Enters the temple through the internal auditory meatus and runs through the facial canal. **C470**
Acoustic or vestibulocochlear CN 8	Internal acoustic meatus	Originates in the **brain stem** (**medulla oblongata**) **between** the base of the brain (**pons**) and the **spinal cord C724** Both the **vestibular** branch and the **cochlear** branch are located in the **inner ear**	
Glossopharyngeal CN 9	Jugular foramin[a]	**Originates** in the anterior portion of the **medulla oblongata C725**	Jugular foramen Between the i**nternal 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
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Name and CN #	Exits Cranium Through	Site Code: Cranial Nerve	Site Code: Peripheral Nerve
Vagus CN 10	Jugular foramen	The vagus nerve originates from the **medulla** of the **brainstem**. **C725**	CN10 descends within the **carotid sheath** 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 **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 C473** Forms the anterior and posterior **gastric** nerves **C475**
Accessory CN 11	Jugular foramen	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 **medulla oblongata.** The fibers of the spinal accessory nerve coalesce to **form spinal rootlets, roots**, and finally the **spinal accessory nerv**e itself C**725**	The nerve **exits** the skull through the jugular foramen. It then runs along the internal **carotid** artery within the **neck C470** Reaches the sternocleidomastoid muscle and the trapezius **C476**
Hypoglossal CN 12	Hypoglossal canal	**CN12** starts in the **hypoglossal** nucleus of the **brainstem**, **C725**	**CN12** exits the hypoglossal canal, traveling **between** the **carotid** artery and **jugular** vein, ending under the **tongue C470**

Section 3: Additional Information to Complete the 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 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.

Table 5: Paired Sites

Use **Table 5** to identify sites for which laterality **must** be coded. Do **not** use this table to determine multiple primaries.

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

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
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(Excludes lymphoma and leukemia M9590 – M9993 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:

- 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**).

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.

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

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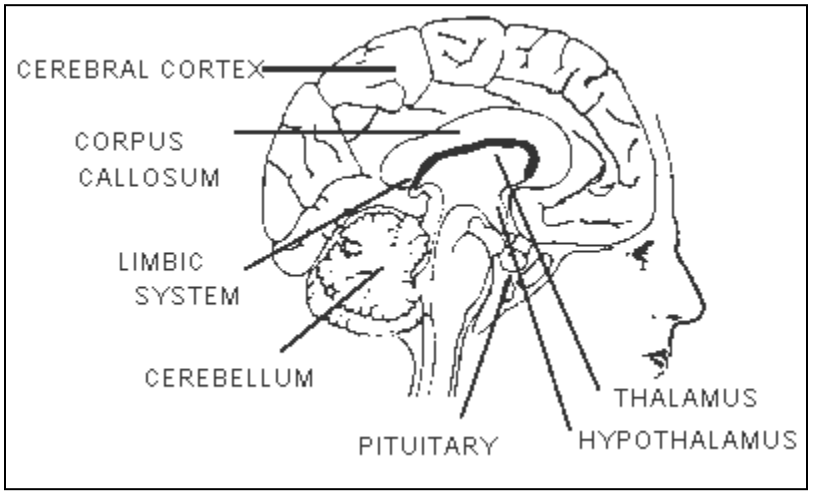
Illustrations



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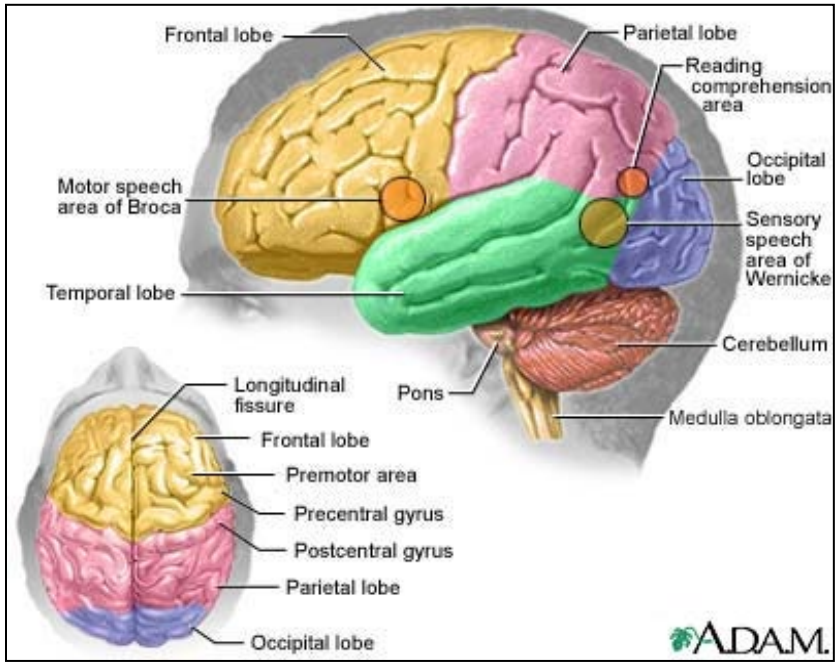

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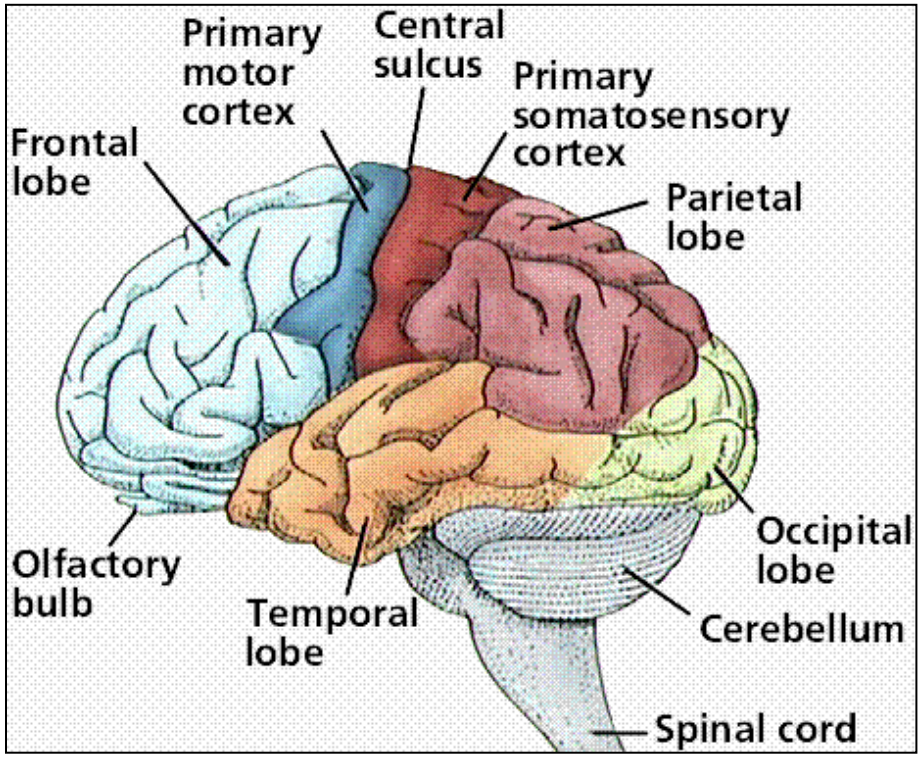

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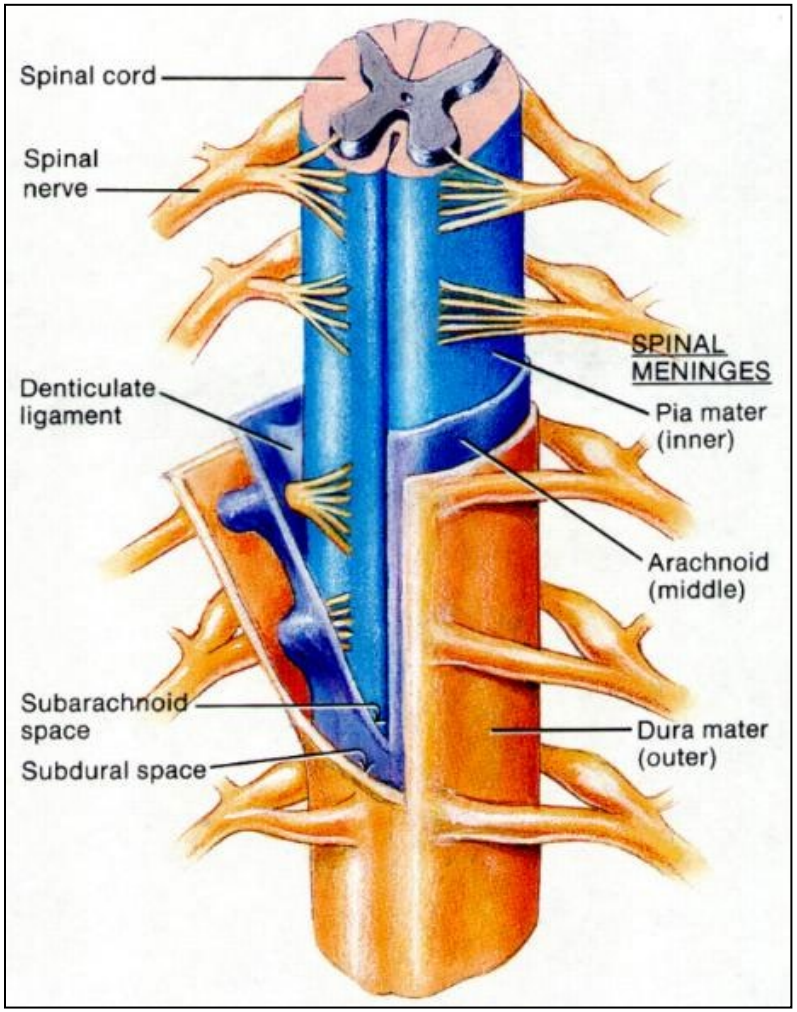

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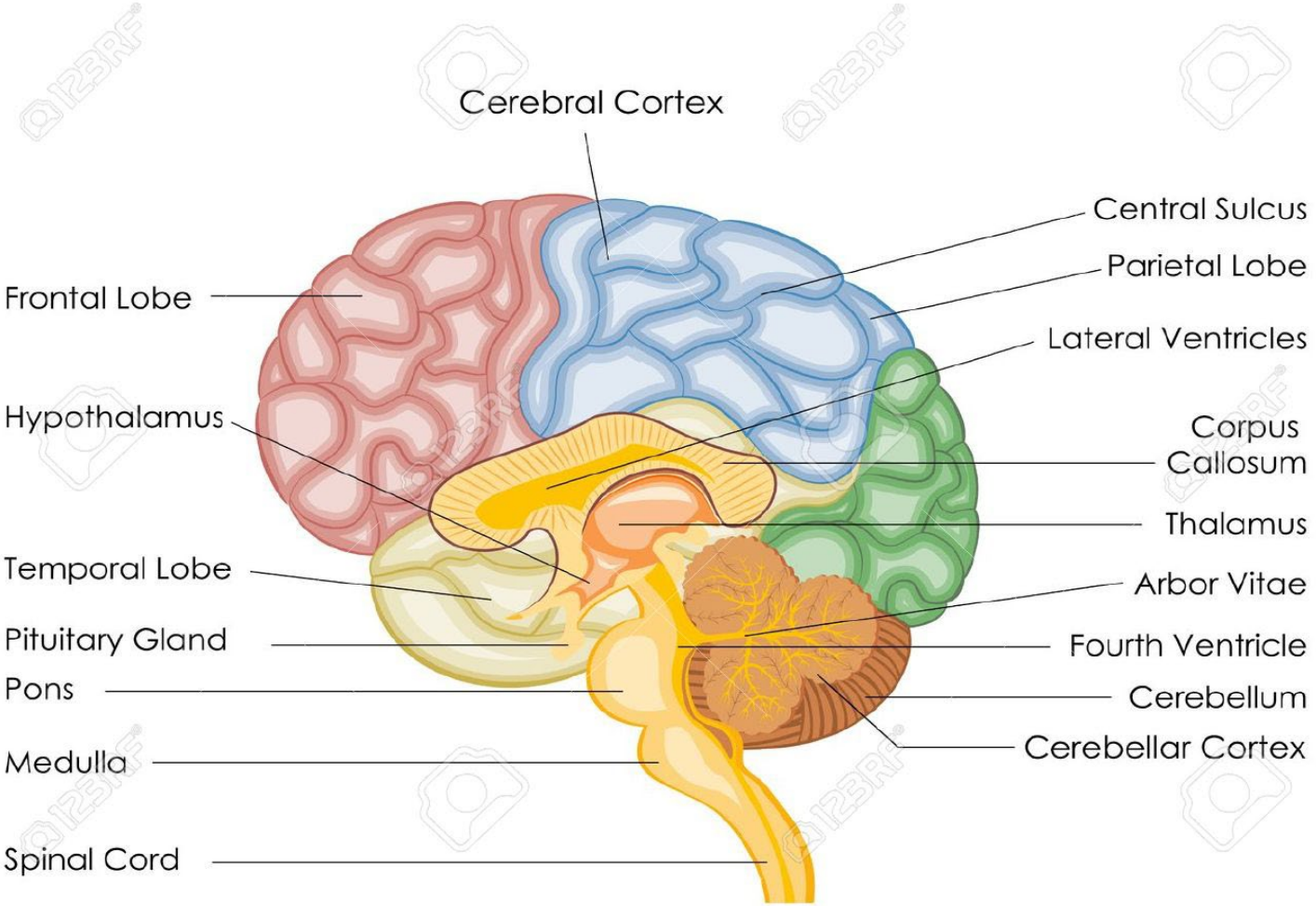

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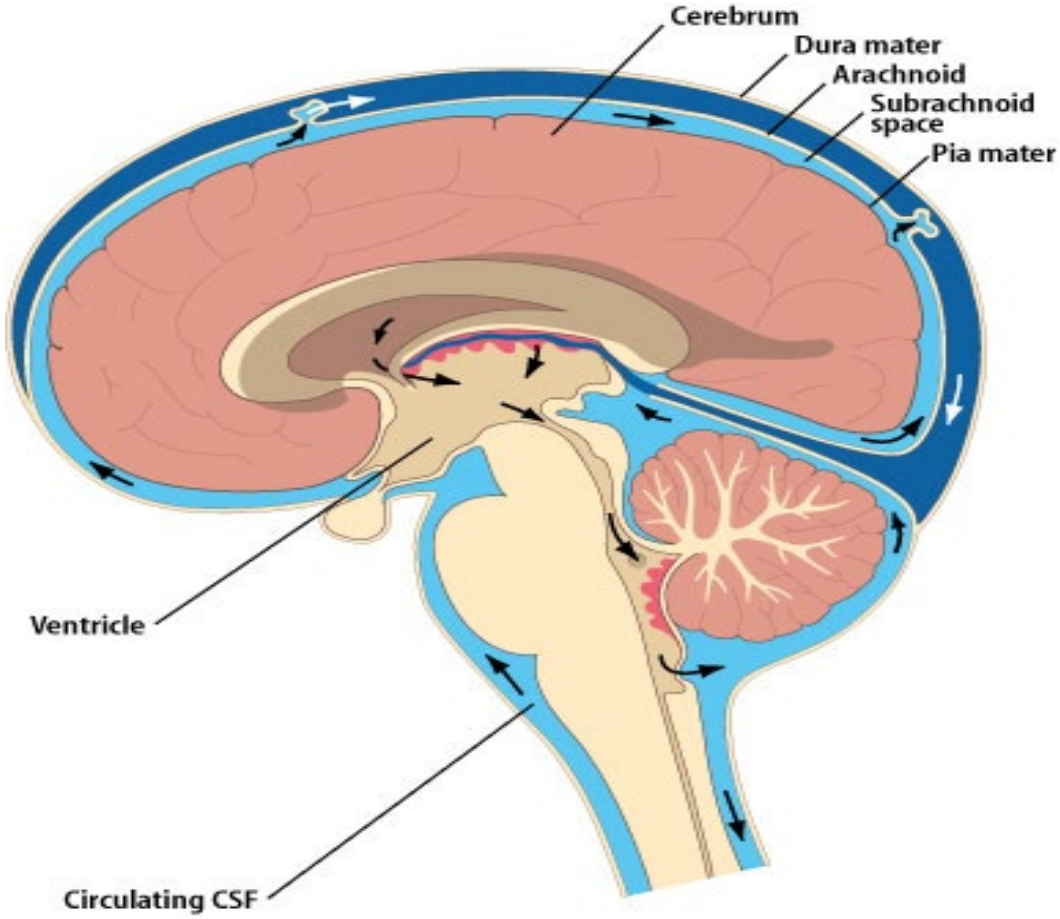

Malignant CNS and Peripheral Nerves Multiple Primary Rules
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Note 1: Non-malignant 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: Separate 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**[i] 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.

[i] **Prepare one abstract. Use the histology rules to assign the appropriate histology code.**

Malignant CNS and Peripheral Nerves Multiple Primary Rules
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Single Tumor

IMPORTANT: The **major difference** between **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 M2 Abstract a **single primary**[i] 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.
Note 4: A glioblastoma multiforme 9440 (GBM) diagnosed in residual tumor, regardless of timing, is not a new primary

Rule M3 Code a **single primary**[i] when a neoplasm is **originally diagnosed** as an **oligodendroglioma** 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**[i] (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: 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**.
- 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 2018. After months of active surveillance (watchful waiting), the **patient** becomes **symptomatic**. A **resection** is done in April 2019; the **resection** pathology is **anaplastic ganglioglioma 9505/3**. **Change behavior code** on the **original abstrac**t from /1 to /3. Do not **change date of diagnosis.**

Example 2: A November 15, 2017 **MRI** and **subsequent** stereotactic **biopsy** of lateral ventricle of brain show a **mature teratoma 9080/1**. The patient becomes symptomatic in 2018 and the neoplasm is resected on October 31, 2018. 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.

[i] **Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.**

Malignant CNS and Peripheral Nerves Multiple Primary Rules
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Multiple Tumors

Note 1: Multiple tumors may be a single primary or multiple primaries.
Note 2: Separate, non-contiguous tumors are always multiple primaries when:

- In the CNS (see **Table 2**) AND in a site other than the CNS
  Example: Patient has a malignant nerve sheath tumor of the right arm C471 and non-metastatic adenocarcinoma of the lung. The lung is not a CNS site. Abstract two primaries.
- In different CNS sites (see Rule M8)

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**[ii] 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.

Malignant CNS and Peripheral Nerves Multiple Primary Rules
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M6 Abstract **multiple primaries**[ii] when a patient has a glial tumor and is subsequently diagnosed with a **glioblastoma multiforme 9440** (GBM).

Note 1: Definition of a glial tumor: Any tumor arising from the CNS glial cells. The following is simply a list of all tumors which would be classified as glial.

- Astroblastoma **9430**
- Astrocytomas **9400** and all subtypes
  - Anaplastic astrocytoma IDH-mutant/wildtype; anaplastic astrocytoma NOS **9401**
  - Gemistocytic astrocytoma IDH-mutant **9411**
- Diffuse midline glioma H3 K27M Mutant **9385**
- Ependymoma **9391** and all subtypes
  - Anaplastic ependymoma **9392**
  - Ependymoma, RELA fusion-positive **9396**
  - Papillary ependymoma **9393**
- Glioblastoma **9440** and all subtypes **(this is a glial tumor; however do not apply this rule to a GBM followed by a GBM)**
  - Giant cell glioblastoma **9441**
  - Glioblastoma IDH-mutant **9445**
  - Gliosarcoma **9442**
- Oligodendroglioma and all subtypes **9450**
  - Anaplastic oligodendroglioma; IDH-mutant; 1p/19q-codeleted; IDH-mutant and 1p/19q-codeleted **9451**
- Pleomorphic xanthroastrocytoma **9424**

Note 2: This is a change from the 2007 Rules.

Note 3: 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

Note 4: This rule applies to multiple tumors. A glioblastoma multiforme (GBM) that is subsequently diagnosed in residual tumor from a glial tumor is a single primary. See previous rules.

Malignant CNS and Peripheral Nerves Multiple Primary Rules
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M7 Abstract a **single primary**[i] when there are separate, non-contiguous tumors in the brain (multicentric/multifocal) with the **same histology XXXX**. 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. Glioblastoma multiforme commonly exhibits multiple tumors which are described as **multifocal/multicentric**.

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.

Malignant CNS and Peripheral Nerves Multiple Primary Rules
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M8 Abstract **multiple primaries**[ii] 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
- 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 one of the cranial nerves **C722-C725 AND** any other part of the CNS
- Any two or more of the cranial nerves
  - **C722** Olfactory, **C723** Optic, **C724** Acoustic, **C725** Cranial nerves NOS
- Meninges of cranial or peripheral 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 M9 Abstract **multiple primaries**[ii] 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) **OR**
- A NOS histology in column 3 with an indented subtype/variant

Malignant CNS and Peripheral Nerves Multiple Primary Rules
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M11 Abstract **multiple primaries**[ii] 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.

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[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.**
[ii] **Prepare two or more abstracts. Use the histology rules to assign the appropriate histology code to each case abstracted.**

Malignant CNS and Peripheral Nerves Histology Rules
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

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

Priority Order for Using Documentation to Identify Histology

IMPORTANT NOTES

1. Code the histology diagnosed ***prior*** to **neoadjuvant treatment.**
   Note 1: Histology changes may occur following immunotherapy, chemotherapy, targeted therapy, and radiation therapy.
   Note 2: Neoadjuvant treatment is any tumor-related treatment given prior to surgical removal of the malignancy.

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

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

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

This is a hierarchical list of source documentation.

1. Pathology/tissue from **resection** of primary tumor
   A. Biomarkers
      Note 1: Biomarkers are emerging as an important part of cancer diagnosis and treatment. Some biomarkers are used to determine treatment rather than histologic type. The efficacy of identification of histologic type using biomarkers differs from primary site to primary site. When a histologic type is identified using a biomarker, code the identified histology. 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(s) and/or comment(s)
   C. Final diagnosis / synoptic report as required by CAP
   D. CAP protocol

2. Pathology/tissue from **biopsy** of primary tumor
   A. **Biomarkers**
      Note 1: Biomarkers are emerging as an important part of cancer diagnosis and treatment. Some biomarkers are used to determine treatment rather than histologic type. The efficacy of identification of histologic type using biomarkers differs from primary site to primary site. When a histologic type is identified using a biomarker, code the identified histology. 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 / synoptic report as required by CAP
   D. CAP protocol
   Note 1: Addendums and comments are given the second 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 third priority.
   Note 3: **Do not** use the microscopic or gross section of the pathology report for coding.
   Note 4: The CAP protocol is a checklist which
      - Provides guidelines for collecting the essential data elements for complete reporting of tumors and optimal patient care.
      - Allows physicians to check multiple histologies

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

4. Tissue/pathology from a metastatic site
   Note 1: Code the behavior /3
   Note 2: The tissue from a **metastatic s**ite 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. Treatment plan
   B. Documentation from Tumor Board
   C. Documentation in the medical record that **refers to original pathology, cytology, or scan(s)**
   D. 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, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Coding Histology

Note 1: The priority is to code the most specific histology. **DO NOT USE BREAST HISTOLOGY CODING RULES FOR THIS SITE.**
Note 2: Only use this section for one or more histologies within a single tumor.
Note 3: Do not use this section in place of the Histology Rules.

1. **Code** the **most specific** histology or **subtype/variant, regardless of whether it is described as:**
   A. The majority or predominant part of tumor
   B. The minority of tumor
   C. A component

   Example 1: Diagnosis for a single tumor is astrocytoma 9400 with the majority or predominant part of tumor being anaplastic astrocytoma IDH-mutant 9401. Code the subtype/variant: anaplastic astrocytoma IDH-mutant 9401.

   Example 2: Diagnosis for a single tumor is CNS ganglioneuroblastoma 9490 with minority of tumor being CNS embryonal tumor 9473. Code the subtype/variant: CNS embryonal tumor 9473.

   Example 3: Diagnosis for a single tumor is ependymoma 9391 with a component of anaplastic ependymoma 9392. Code the subtype/variant: anaplastic ependymoma 9392.

   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 <u>ONLY</u>** when there is a specific ICD-O code for the "NOS with ____ features" or "NOS with ____ differentiation".
   Note: Do not code differentiation or features when there is no specific ICD-O code.

3. Code the specific histology described by ambiguous terminology (list follows) ONLY when A or B is true:
   A. The only diagnosis available is one histology term described by ambiguous terminology
      - CoC and SEER require reporting of cases diagnosed only by ambiguous terminology
      - Case is accessioned (added to your database) based on ambiguous terminology and no other histology information is available/documented

   Example: Outpatient biopsy says probably desmoplastic medulloblastoma. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology desmoplastic medulloblastoma 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 astrocytoma consistent with gemistocytic astrocytoma IDH-mutant. The oncology consult says the patient has gemistocytic astrocytoma IDH-mutant in the occipital lobe. This is clinical confirmation of the diagnosis, code gemistocytic astrocytoma IDH-mutant. The case meets the criteria in bullet 1.

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

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

List of Ambiguous Terminology

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

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

Malignant CNS and Peripheral Nerves Histology Rules
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Single Tumor

Rule H1 Code the **reportable CNS tumor (Table 3** 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 malignant peripheral nerve sheath tumors.

Note 2: Tumors are reportable when they meet the behavior (/3) and histology 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.

Rule H2 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 H3 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 H4 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
- 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 H5 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 H6 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 H7 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
- 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 Multiple Tumors Abstracted as a Single Primary

Code the histology using the rule that fits the case.