Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Introduction

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

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

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

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

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

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

- These neoplasms are reportable in North American as malignant 9421/3 for all CNS sites with the exception of the optic nerve:
  - WHO Classification Tumors of the Central Nervous System and IARC designate pilocytic astrocytoma as a synonym for optic glioma
  - When the primary site is optic nerve and the diagnosis is either optic glioma or pilocytic astrocytoma, the behavior is non-malignant and coded 9421/1

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

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

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

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Equivalent or Equal Terms

These terms can be used interchangeably:

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

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Terms that are NOT Equivalent or Equal

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

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

Changes from 2007 MPH Rules

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

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

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

Reportability Criteria for Non-Malignant CNS Neoplasms

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

1. **The behavior must non-malignant /0 or /1.**
   A. Pathology designates the tumor as non-malignant (/0 or /1) **OR**
   B. The tumor is a WHO Grade I (See **Section 1: Table 1**)
   Note 1: **Always** code the behavior code reported by the **pathologist.**
   Note 2: **Never report** a **malignant** /3 behavior code for a **meningioma** based on **tumor extension** to brain, skin of scalp, or other regional organs/tissue. Non-malignant CNS tumors can extend to the regional tissue and bone. Tumor extension is usually equivalent to atypical meningioma 9539/1.
2. **The primary site must be reportable** (See **Section 2**: **Table 3** and **Table 4**)
3. **The histology must be reportable** (See **Section 2: Table 5** and **Table 6**).

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Directory of Sections and Tables

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

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Section 1: Behavior Code

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

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

Priority Order for Using Documentation to Assign Behavior

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

D. Angiogram

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

Table 1: WHO Grades of Select CNS Neoplasms

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

WHO Grade Definitions

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

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

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

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

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

Section 2: Reportable Primary Sites and Histologies

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

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

- **Intraosseous**
  Note: The **dura** layer of the meninges **contacts** the **endosteum** of the bones of the skull. The primary site for intraosseous meningioma is cranial meninges C700.
- **Sphenoid wing**
  Note 1: Sphenoid wing meningiomas arise in the **cranial meninges** C700 which **covers** the bony structure called the sphenoid wing.
  Note 2: The term "sphenoid wing meningioma" is used to identify the **location** of the meningioma because sphenoid wing meningiomas may be very **invasive**, spreading to the dura of the frontal, temporal and orbital regions.
- **Cavernous sinus**
  Note 1: Cavernous sinus is located between the endosteal and meningeal layers of the dura.
  Note 2: There is **no ICD-O site** code for cavernous sinus because tumors do not arise in the sinus itself; the primary sites are:
  - The **cranial nerves** passing through the sinus (trochlear, abducent **C725**) **OR**
  - The cerebral **meninges/dura C700** covering the cranial nerve

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

Priorities for Coding Primary Site

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

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

1. **Resection**
   A. Operative report(s)
   B. Pathology report(s)
2. **Biopsy**
   A. Operative report(s)
   B. Pathology report(s)
3. Resection and/or biopsy performed, but operative report(s) and pathology are not available (minimal information):
   A. Tumor Board
   B. Code from physician's documentation of **original diagnosis** from operative or pathology report
   C. Physician's **documentation of primary site** in the medical record
   Example: The operative and pathology reports are not in the medical record. The **attending documents** that the patient had a biopsy of a **cerebral meningioma** at a different facility. Code the primary site cerebral meninges C700.
4. For cases diagnosed by imaging (**no pathology/resection or biopsy**) use information from scans in the following priority order:
   A. MRI
   B. CT
   C. PET
   D. Angiogram

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Reportable Primary Site Groups

The two major groups of reportable sites are:

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

Reportable Primary Sites and their ICD-O Codes

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

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Table 2: Reportable Primary Sites

Column 1 lists the reportable primary site term.
Column 2 lists the site code for the reportable primary site.

Site Group	Reportable Subsite Terms and Code
Brain	Brain NOS **C719** Brain stem **C717** Cerebellum NOS **C716** Cerebrum **C710** Frontal lobe **C711** Occipital lobe **C714** Overlapping lesion of brain and central nervous system **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**
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: Reportability of Non-Malignant Cranial Nerve (CN) Tumors

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

Note 1: A neoplasm arising in a **cranial** nerve is coded to the **specific nerve** in which it **arises**.
Note 2: Neoplasms, commonly meningiomas, **arising** in the **dura/meninges** of an **intracranial** nerve are coded to **cerebral meninges** C700.
Note 3: It is important to check the **operative report** to determine whether the surgery is **intracranial** or **intradural.**
Note 4: This table is used for **non-malignant** neoplasms **ONLY.**

Column 1: The **proper name** for the cranial nerve (CN) and the **cranial nerve number**
Column 2: The point at which the nerve exits the cranium
Column 3: **Reportable** portions of the cranial nerve. Of note, malignant neoplasms are reportable for all parts of the cranial nerves
Column 4: **Non-reportable** portions of the cranial nerve

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

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Name and CN #	Exits Cranium Through	Reportable Portions of CN	Non-Reportable Portions of CN
Trochlear CN 4 **C725**	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.**	Enters the **orbital fissure**.
Trigeminal CN 5 **C725** ***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, the superior orbital fissure, the foramen rotundum and the foramen ovale.	CN5 originates in the **pons**. Upon leaving the pons it enters a **small fossa** posterior and inferolateral to the cavernous sinus called **Meckel's (trigeminal) cave**.	• **Ophthalmic nerve branch** crosses the **pterygopalatine fossa**, inclines laterally on the back of the **maxilla**, and enters the **orbit** through the inferior orbital fissure where it is called the **infraorbital** nerve. It ends beneath the **quadatus labii superius**, and divides into multiple branches that spread to the side of the **nose**, the lower **eyelid**, and the upper **lip** • **Maxillary** nerve leaves foramen rotundum and traverses the **infraorbital groove** and **canal** in the floor of the **orbit**, and appears on the face at the infraorbital foramen. • **Mandibular** nerve leaves via the foremen ovale travels along the **mandibular** groove

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Name and CN #	Exits Cranium Through	Reportable Portions of CN	Non-Reportable Portions of CN
Abducent CN 6 **C725**	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.**	**Dorello's canal** and travels to the tip of the **temporal bone**. Enters **orbit**
Facial CN 7 **C725**	Internal acoustic meatus	**CN7** originates in the **pons,** along the posterior cranial fossa (the posterior cranial fossa is part of the intracranial cavity)	Enters the **temple** through the **internal auditory meatus** and runs through the **facial canal**.
Acoustic or vestibulocochlear CN 8 **C724**	Internal acoustic meatus	Originates in the **brain stem** (**medulla oblongata**) **between** the base of the brain (**pons**) and the **spinal cord** Both the **vestibular** branch and the **cochlear** branch are located in the **inner ear**	
Glossopharyngeal CN 9 **C725**	Jugular foramen	**Originates** in the anterior portion of the **medulla oblongata**	**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**

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

Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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

Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Table 4: Non-Reportable Neoplasms

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

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

Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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

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

Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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

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

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

Use **Table 5** to:

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

Column 1 contains **histology terms and codes** that occur in the **brain**, **ventricles** of the brain, and **intracranial glands**
Column 2 contains the **site code** for the most common **intracranial** primary site(s) for that specific histology

Histology Term and Code	Most Common Intracranial Primary Site
Angiocentric glioma 9431/1*	Cerebrum **C710**
Choroid plexus papilloma 9390/0	Intraventricular site (lateral/third ventricle **C715** and IV ventricle **C717**)
(Capillary) hemangioblastoma 9161/1	Cerebellum **C716**, cerebrum (rare) **C710**
Craniopharyngioma 9350/1	Craniopharyngeal duct **C752,** pituitary gland, sella turcica **C751**
Dermoid cyst 9084/0	Pineal gland **C753**, suprasellar **C719**
Desmoplastic infantile astrocytoma and ganglioglioma 9412/1	Cerebrum/supratentorial brain NOS **C710**
Dysembryoplastic neuroepithelial tumor (DNT) 9413/0	Cerebrum **C710**, temporal lobe **C712**
Dysplastic gangliocytoma 9493/0	Cerebellum **C716**
Meningioma (rare) 9530/0	Intraventricular **C715**
Myxopapillary ependymoma 9394/1	4th ventricle **C717**

Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Histology Term and Code	Most Common Intracranial Primary Site
Pilocytic astrocytoma/juvenile pilocytic astrocytoma 9421/1	**Optic nerve C723**
Pineocytoma 9361/1	Pineal gland C**753**
Pituicytoma 9432/1*	Pituitary gland **C751**, sella turcica **C751,** suprasellar **C719**
Pituitary adenoma 8272/0	Pituitary gland **C751**
Prolactinoma 8271/0	Pituitary gland **C751**
Subependymal giant cell tumor (SEGA) 9384/1	Lateral ventricles **C715**
Subependymoma 9383/1	Intraventricular site (lateral/third ventricle, rare **C715** and IV ventricle **C717**)

Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Table 6: Specific Histologies, NOS, and Subtypes/Variants

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

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

Column 1 contains specific and NOS histology terms.
- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

Column 2 contains **synonyms** for the specific or NOS term. Synonyms have the **same** histology **code** as the specific or NOS term.
Column 3 contains **subtypes/variants** of the **NOS** histology. Subtypes/variants **do not** have the **same** histology code as the NOS term.

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

Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants Histology Term and Codes
Dysembryoplastic neuroepithelial tumor 9413/0	DNT	
Gangliocytoma 9492/0		Dysplastic cerebellar gangliocytoma/Lhermitte-Duclos disease **9493/0**
Ganglioglioma 9505/1		
Granular cell tumor of the sellar region 9582/0		
Hemangioblastoma 9161/1	Capillary hemangioblastoma	
Hemangioma 9120/0		Cavernous hemangioma **9121/0**
Leiomyoma 8890/0		
Lipoma 8850/0		Hibernoma **8880/0**
Meningeal melanocytosis 8728/0		Meningeal melanocytoma **8728/1**
Meningioma 9530/0	Lymphoplasmacyte-rich meningioma Metaplastic meningioma Microcystic meningioma Secretory meningioma	Angiomatous meningioma **9534/0** Atypical meningioma **9539/1** Clear cell/chordoid meningioma **9538/1** Fibrous meningioma **9532/0** Meningothelial meningioma **9531/0** Psammomatous meningioma **9533/0** Transitional meningioma **9537/0**
Myofibroblastoma 8825/0		Inflammatory myofibroblastic tumor **8825/1**
Myxopapillary ependymoma 9394/1		
Neurocytoma 9506/1	Central neurocytoma Cerebellar liponeurocytoma Extraventriculare neurocytoma Lipomatous medulloblastoma Medullocytoma Neurolipocytoma	

Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants Histology Term and Codes
Neurofibroma 9540/0	Atypical neurofibroma	Plexiform neurofibroma **9550/0**
Optic glioma/pilocytic astrocytoma 9421/1		
Osteoma 9180/0		
Papillary glioneuronal tumor 9509/1*	Diffuse leptomeningeal glioneuronal tumor Rosette-forming glioneuronal tumor	
Paraganglioma 8693/1		
Perineurioma 9571/0		
Pineocytoma 9361/1		
Pituicytoma 9432/1*		
Pituitary adenoma 8272/0	Corticotroph Gonadotroph adenoma Somatotroph adenoma Thyrotroph adenoma Null cell adenoma Plurihormonal and double adenomas	
Prolactinoma 8271/0		
Rhabdomyoma 8900/0		
Schwannoma 9560/0	Cellular schwannoma Neurilemoma Neurinoma Plexiform schwannoma	Melanotic schwannoma **9560/1***
Solitary fibrous tumor Grade 1 8815/0	Hemangiopericytoma Grade 1	Solitary fibrous tumor/hemangiopericytoma Grade 2 **8815/1***

Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants Histology Term and Codes
Spindle cell oncocytoma 8290/0		
Subependymal giant cell astrocytoma 9384/1		
Subependymoma 9383/1		
Teratoma 9080/1		

Section 3: Additional Information to Complete Abstract

Conflicting Information on Pathology Report(s)

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

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

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

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

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

Table 7: Paired Sites

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

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 most common for the following histologies: glioblastoma multiforme, meningiomas, lymphomas (usually located near the midline), and epidermoid cysts (which can cross the midline via the subarachnoid space).
Note 2: SEER allows laterality to be coded for sites other than those in the table.

Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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

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

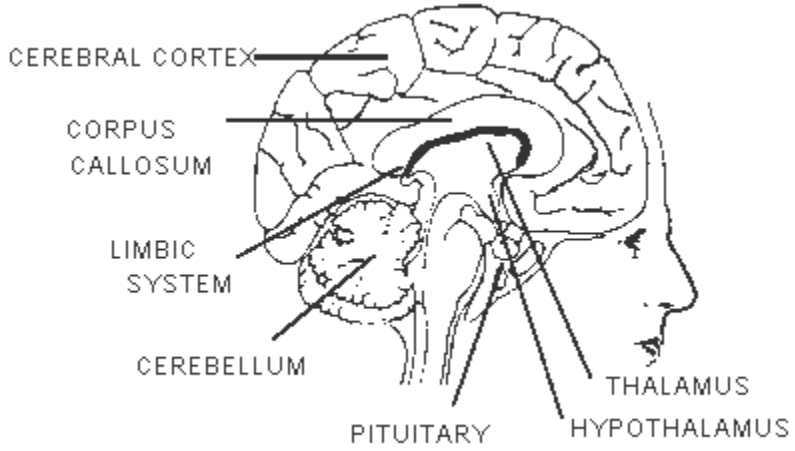
- Residual tumor becomes more aggressive /3 **OR**
- The tumor **recurs** as a **more aggressive /3 histology**

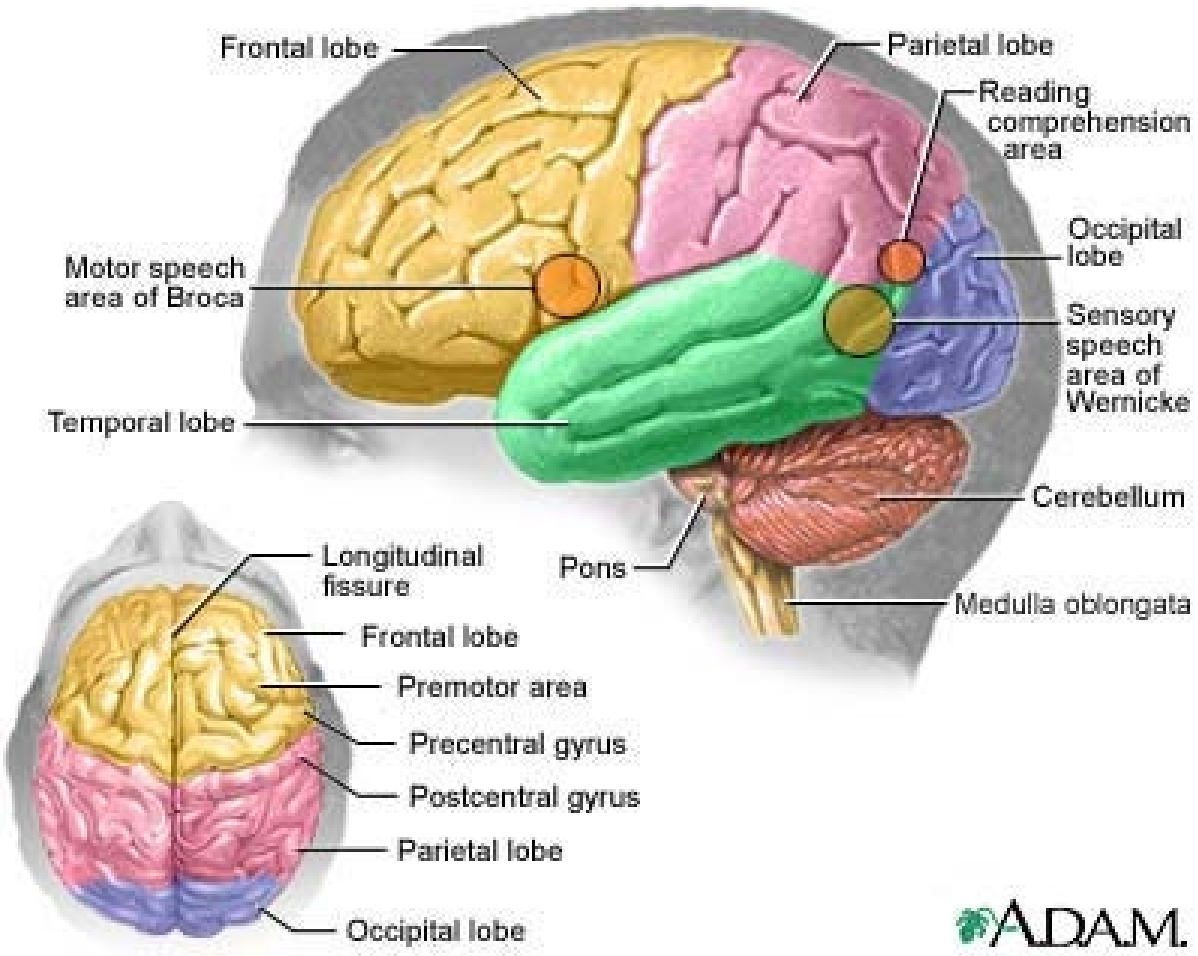
The table identifies non-malignant tumors that have the **potential** of **transforming to** a malignant tumor (**new primary**).

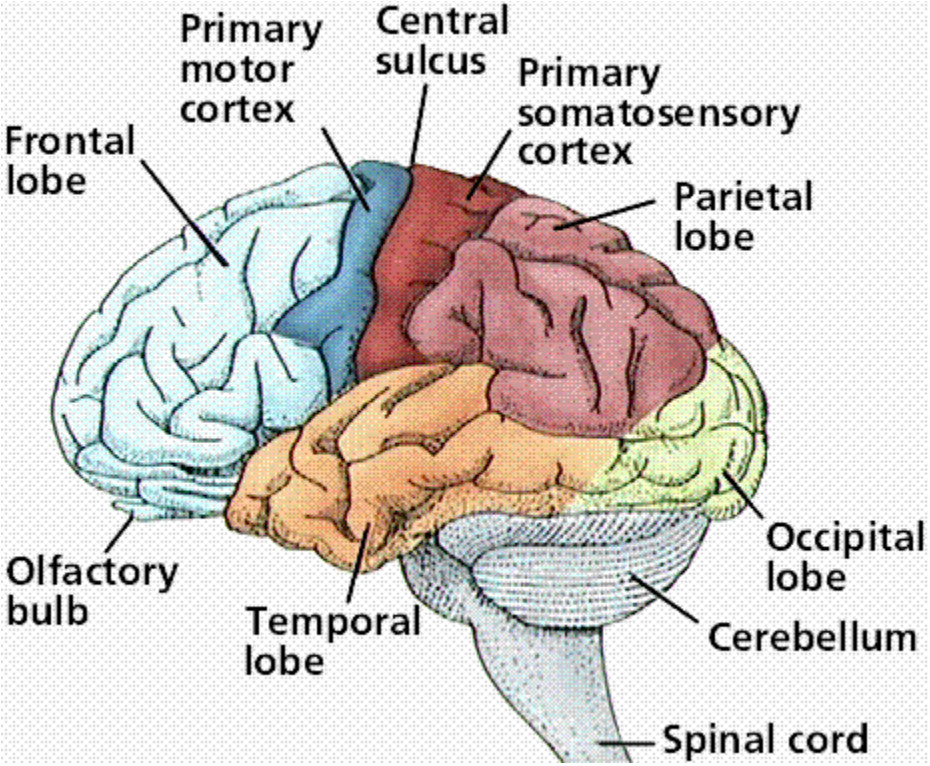
Column 1 is the **non-malignant** ICD-O histology term and code.
Column 2 is the **malignant** /3 ICD-O histology term and code to which the non-malignant tumor can transform.

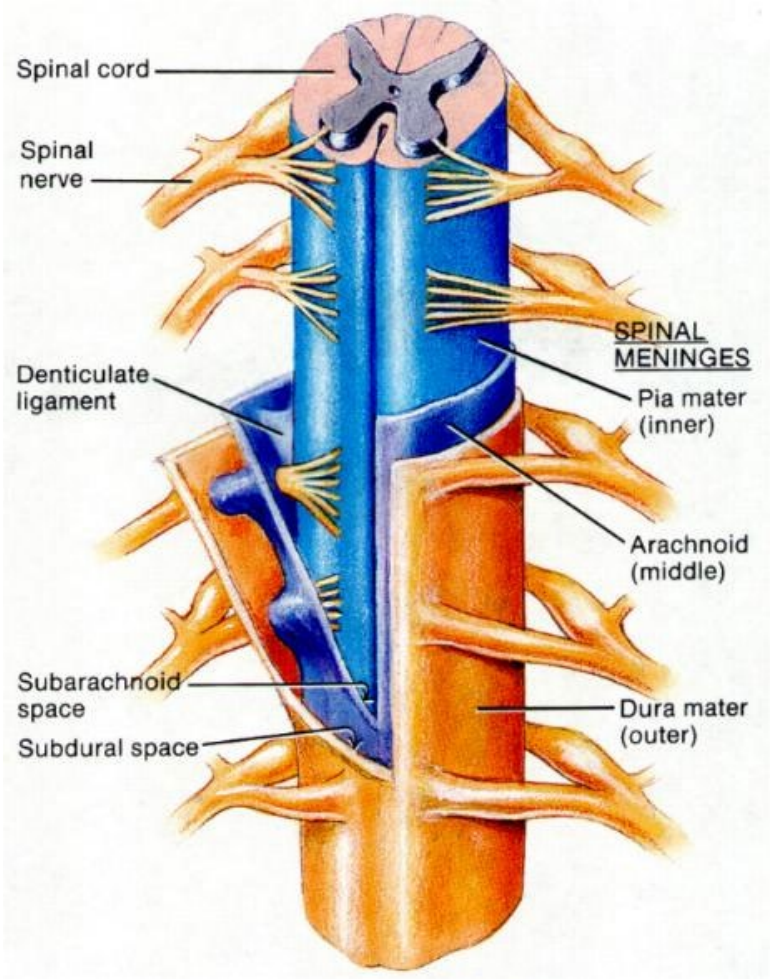
Original Histology and Code	Transformed Histology and Code
Chondroma **9220/0**	Chondrosarcoma **9220/3**
Ganglioglioma **9505/1**	Anaplastic ganglioglioma **9505/3**
Hemangioma **9120/0**	Angiosarcoma **9120/3**
Hemangiopericytoma **9150/1**	Anaplastic hemangiopericytoma **9150/3**
Leiomyoma **8890/0**	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**

Illustrations

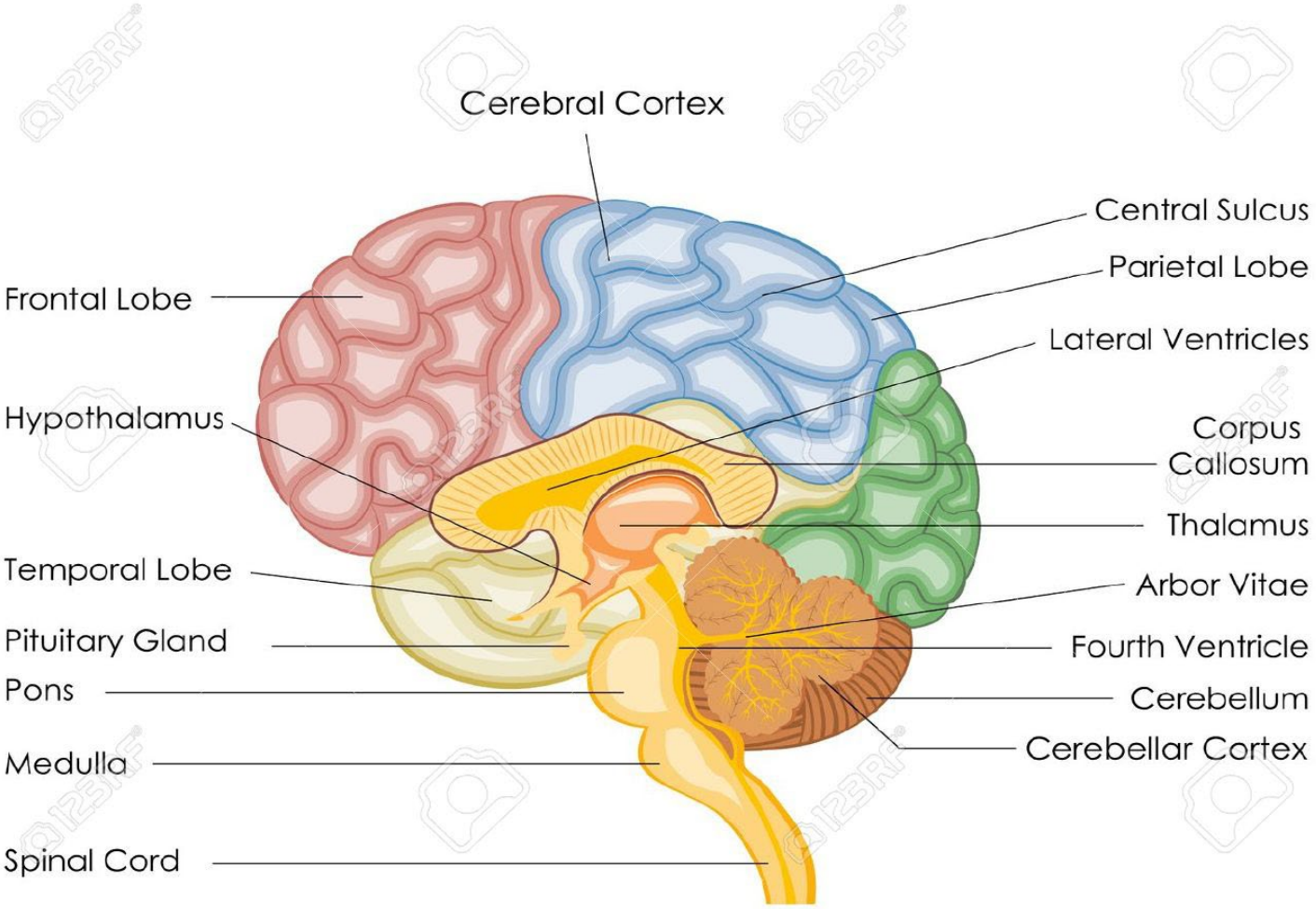




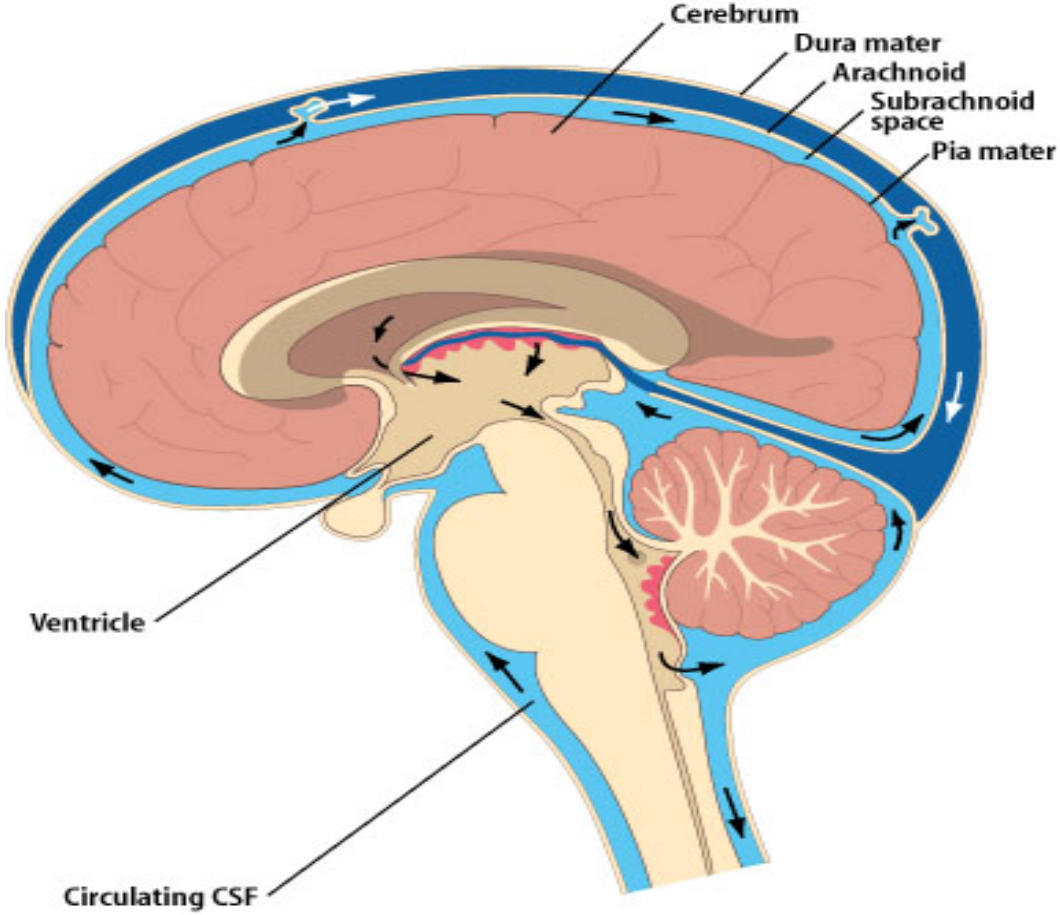



Spinal cord
Spinal nerve
SPINAL MENINGES
Denticulate ligament
Pia mater (inner)
Arachnoid (middle)
Subarachnoid space
Dura mater (outer)
Subdural space

Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)



Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)



Non-Malignant CNS Multiple Primary Rules
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Note 1: **Timing** is **not used** to determine multiple primaries.
Note 2: **Laterality** is **not used** to determine multiple primaries.
Note 3: **Malignant** central nervous system (CNS) tumors have a separate set of rules.
Note 4: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

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

Unknown if Single or Multiple Tumors

Rule M1 Abstract a **single primary**[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.**

Non-Malignant CNS Multiple Primary Rules
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Single Tumor

IMPORTANT: The **major difference** between **M3** and **M5** is:
M3: No resection as first course of treatment **AND** when the same tumor is subsequently **resected**, **pathology proves malignant** behavior
M5: Tumor resected as first course of treatment. Subsequent tumor (**recurrence** or **de novo)** is **malignant**

Rule M2 Abstract a **single primary**[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 an adjacent/contiguous site or subsite.
Note 3: The tumor may have multiple histologic components.
Note 4: The tumor may be diagnosed as borderline /1 on scans and subsequent pathology proves a benign /0 tumor.
Example: Patient has a MRI with a diagnosis of a benign /0 tumor. The pathology report from the subsequent surgery shows a borderline /1 tumor. This is a single tumor and **a single primary**. (It is the same tumor as seen on the MRI; better information is provided by the pathology report.)

Rule M3 Abstract a **single primary**[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: Use the Malignant CNS and Peripheral Nerves Rules to code histology.
Note 3: The **resection pathology** is **more accurate** than clinical, radiographic or stereotactic biopsy information. While stereotactic biopsy provides a pathologic specimen, it is small and may not have included the malignant portion of tumor.
Note 4: There is **no time requirement** from initial diagnosis to resection.
Note 5: Edit the original abstract as follows:
- Do **not** change **date of diagnosis**.
- For cases which have been abstracted, **change behavior** code on original abstract from /0 or /1 to /3.
- **Report** all data changes for cases which have been submitted to the central registry.

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

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

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

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

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

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

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

Note 2: This is a single tumor; single primary

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

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

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

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

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

This is the end of instructions for Single Tumor.

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

Non-Malignant CNS Multiple Primary Rules
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Multiple Tumors

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

- In the CNS (see **Table 2**) AND in a site other than the CNS
  Example: Patient has a non-malignant meningioma of the cerebral meninges and adenocarcinoma of the lung. The lung is not a CNS site. Abstract two primaries.
- In different CNS sites (see Rule M7)

IMPORTANT: The **major difference** between **M3** and **M5** is:
M3: No resection as first course of treatment **AND** when the same tumor is subsequently **resected**, **pathology proves malignant** behavior
M5: Tumor resected as first course of treatment. Subsequent tumor (**recurrence** or **de novo)** is **malignant**

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

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

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

Rule M6 Abstract a **single primary**[i] when the patient has **bilateral**:

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

Note 1: The bilateral tumors may appear simultaneously (at the same time) **OR** the contralateral tumor may be diagnosed at any time following the original diagnosis.
Note 2: WHO and IARC designate pilocytic astrocytoma as a synonym for optic glioma. When the primary site is optic nerve, the behavior is non-malignant.
Note 3: When the bilateral tumors are diagnosed at different times**,** the physician **may stage each tumor** because staging and determining multiple primaries are done for different reasons. Staging determines which course of treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).

Non-Malignant CNS Multiple Primary Rules
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule M7 Abstract **multiple primaries**[ii] when multiple tumors are present in any of the following **sites**:

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

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

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

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

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

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

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

Non-Malignant CNS Multiple Primary Rules
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule M10 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 in any of the following locations and/or lateralities:

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

Note 1: Metastases are never used to determine multiple primaries. Seeding metastasis is often noted in ependymomas.
Note 2: This is a **change** from/**clarification** to **previous** rules.
Note 3: These rules are hierarchical. Use this rule ONLY when the previous rules do not apply.
Note 4: An example of a non-malignant brain tumor that may be multi-focal/multi-centric is hemangioblastoma **9161/1**.
Note 5: The physician may **stage each tumor** because staging and determining multiple primaries are done for different reasons. Staging determines which course of treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).

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

Note: The same row means the tumors are:

- The same histology (same four-digit ICD-O code) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3). NOS and subtype/variants are:
  - Choroid plexus papilloma **9390/0** and a subtype/variant of choroid plexus papilloma
  - Craniopharyngioma **9350/1** and a subtype/variant of craniopharyngioma
  - Gangliocytoma **9492/0** and a subtype/variant of gangliocytoma
  - Lipoma **8850/0** and a subtype/variant of lipoma
  - Meningeal melanocytosis **8728/0** and a subtype/variant of meningeal melanocytosis
  - Meningioma **9530/0** and a subtype/variant of meningioma
  - Myofibroblastoma **8825/0** and a subtype/variant of myofibroblastoma
  - Neurofibroma **9540/0** and a subtype/variant of neurofibroma
  - Schwannoma **9560/0** and a subtype/variant of schwannoma
  - Solitary fibrous tumor WHO Grade 1 **8815/0** and a subtype/variant of solitary fibrous tumor WHO Grade 1

Non-Malignant CNS Multiple Primary Rules
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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

Rule M13 Abstract a **single primary**[i] when the tumors do not meet any of the above criteria.
Note: These rules are hierarchical. Use this rule **ONLY** when the previous rules do not apply.

This is the end of instructions for Multiple Tumors

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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 coding rules to assign the appropriate histology code to each case abstracted.

Non-Malignant CNS Histology Rules
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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

Priority Order for Using Documentation to Identify Histology

IMPORTANT NOTES

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

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

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

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

This is a hierarchical list of source documentation:

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

- Biomarkers are not listed because they change rapidly.

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

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

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

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

Note 4: The CAP protocol is a checklist which

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

2. Pathology/tissue from **biopsy**
   A. The addendum and/or comments
   B. Final diagnosis / synoptic report as required by CAP
   C. CAP protocol
   D. Biomarkers
      - Biomarkers do not identify all histologic types.
      - Biomarkers are not listed because they change rapidly.

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

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

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

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

   Note 4: The CAP protocol is a checklist which
      - Provides guidelines for collecting the essential data elements for complete reporting of tumors and optimal patient care.
      - Allows physicians to check multiple histologies

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

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

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

Non-Malignant CNS Histology Rules
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Coding Histology

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

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

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

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

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

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

2. Code the histology described as **differentiation** or **features/features of <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) <u>ONLY</u> 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 central neurocytoma. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology central neurocytoma. The case meets the criteria in #**3A.**

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

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

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

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

List of Ambiguous Terminology

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

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

Non-Malignant CNS Histology Rules
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Single Tumor

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

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

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

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

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

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

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

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

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

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

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

Non-Malignant CNS Histology Rules
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule H3 Code the histology when only **one histology** is present.

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

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

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

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

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

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

This is the end of instructions for Single Tumor.

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

Multiple Tumors Abstracted as a Single Primary

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

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

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

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

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

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

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

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

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

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

Note 1: Only report tumors such as:

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

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

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

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

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

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

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

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

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

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

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

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

Note: Use **Table 6** in the Equivalent Terms and Definitions to identify the NOS and subtypes/variants.

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

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