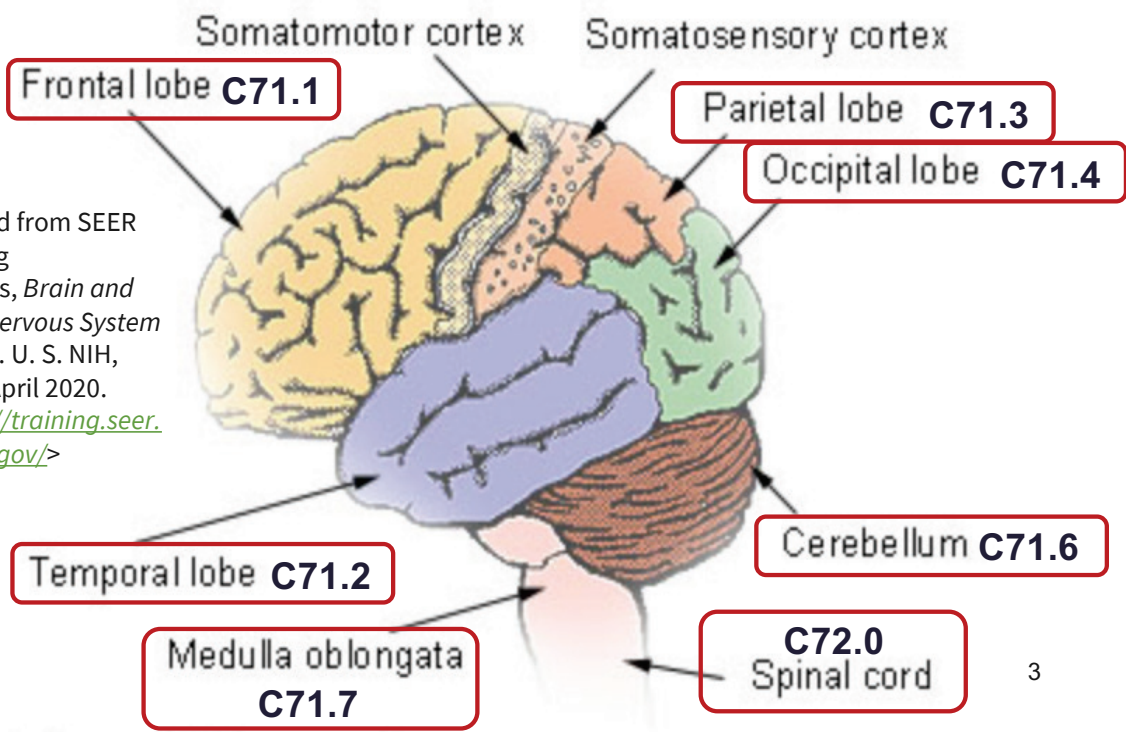
CNS Tumors

Let's see if your brain is up to this!

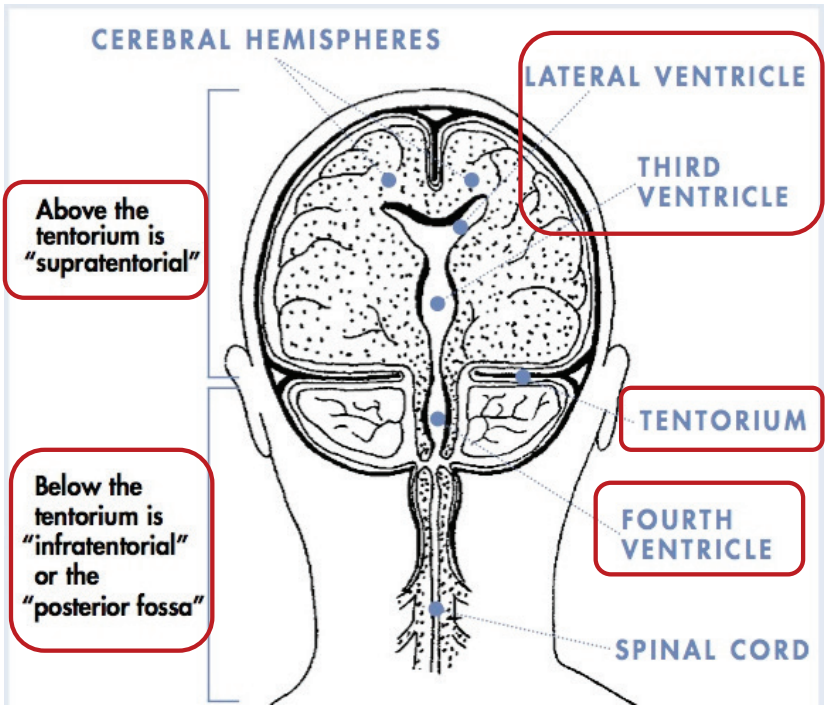
Section One

CNS ANATOMY

Parts of the Brain & CNS



Adapted from SEER Training Modules, *Brain and Other Nervous System Tumors*. U. S. NIH, NCI. 3 April 2020. <*https://training.seer.cancer.gov/*>

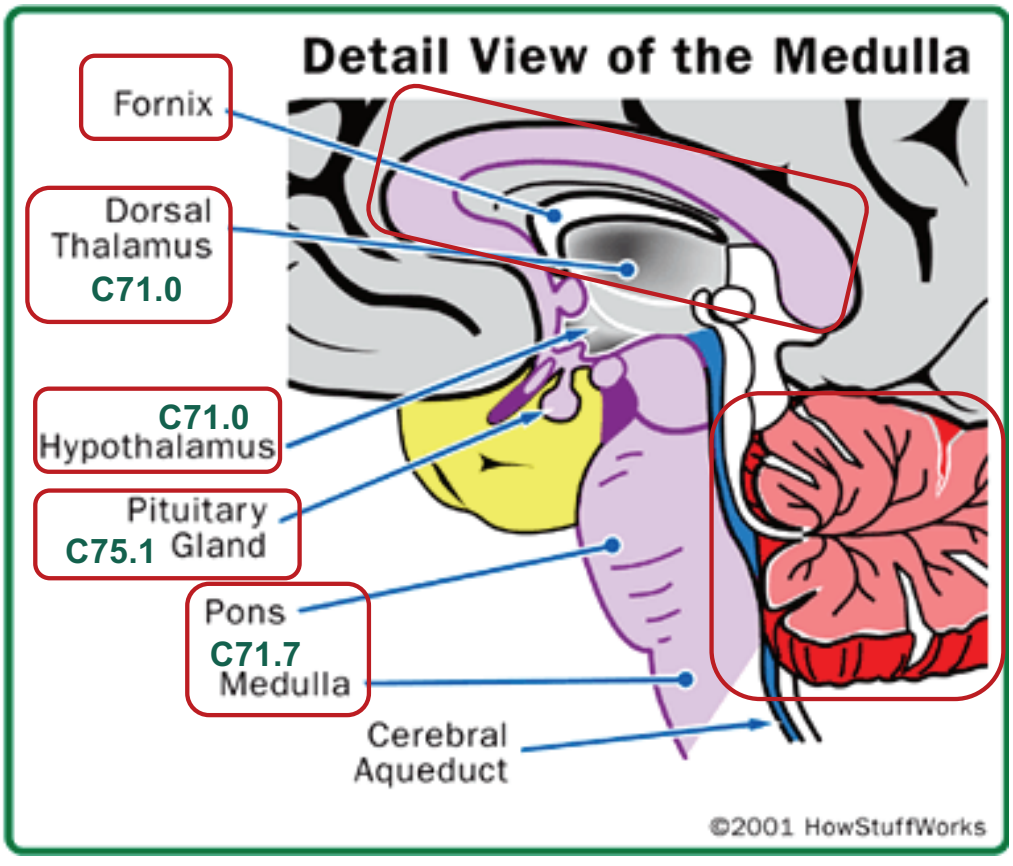


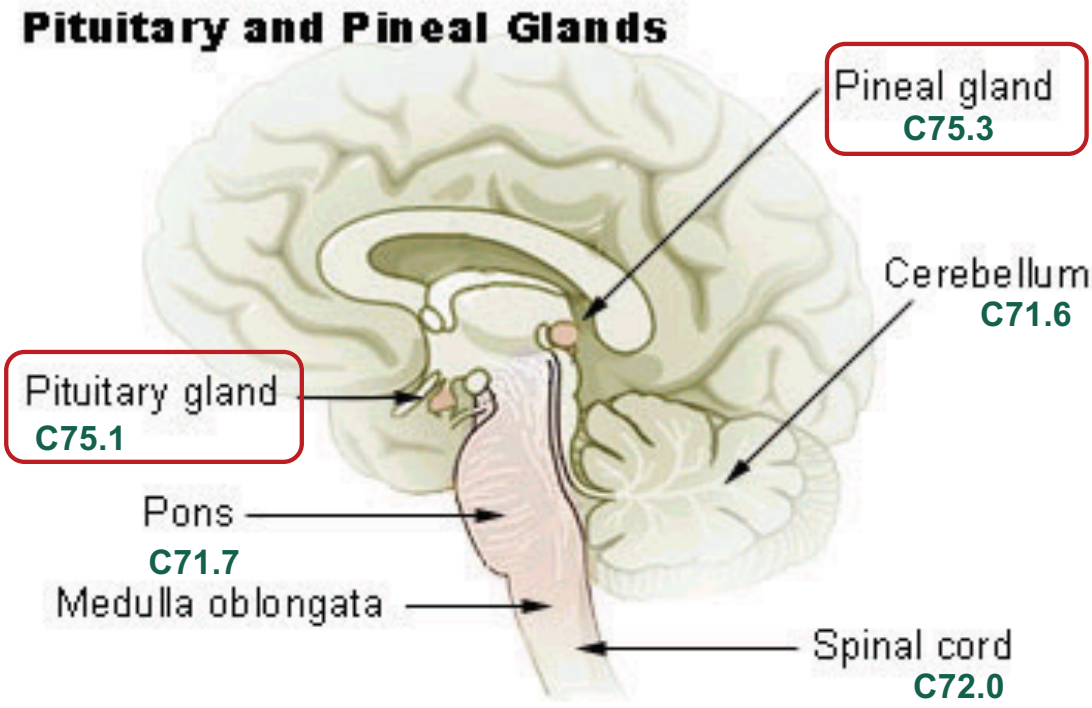
www.abta.org/primer2.htm

What is Where?

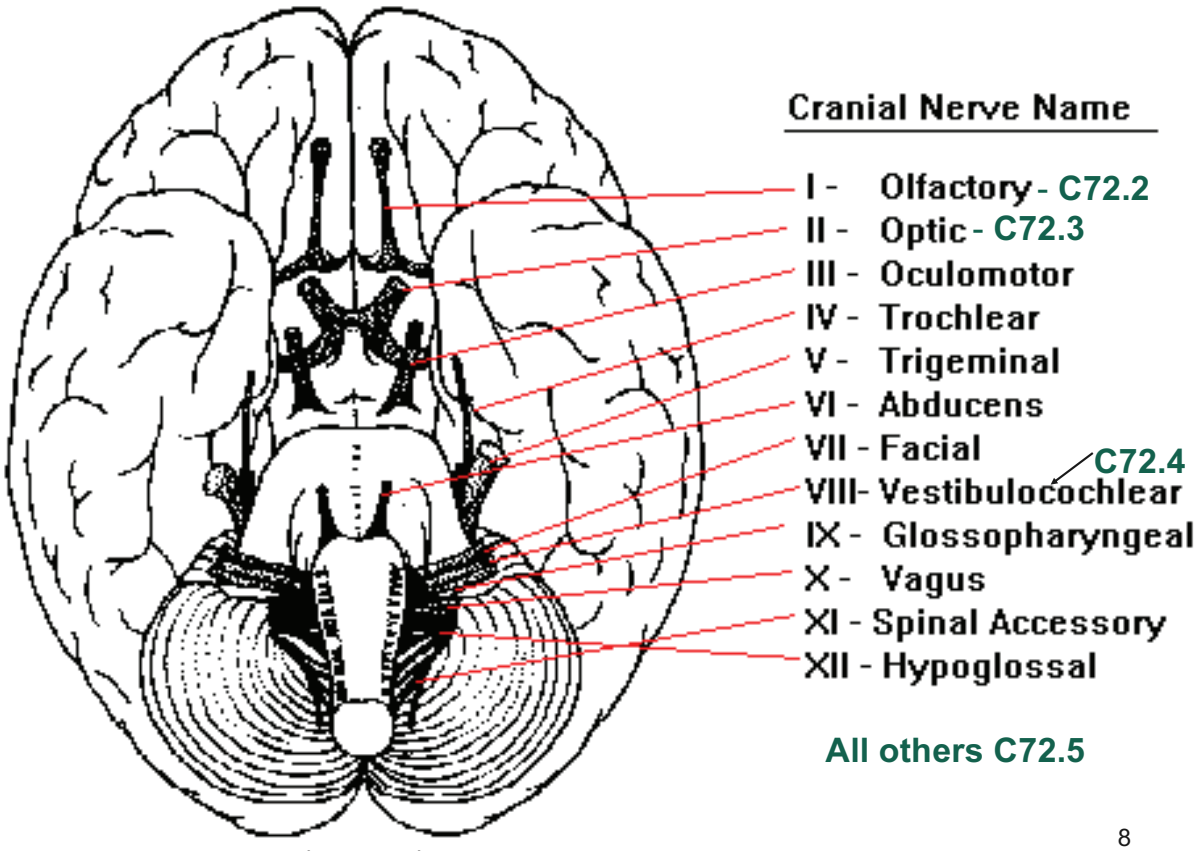
Supratentorial
- Cerebrum
- Lateral ventricles
- Third ventricle
- Choroid plexus
- Hypothalamus
- Pineal gland
- Pituitary gland
- Optic nerve

Infratentorial
(Posterior fossa)
- Cerebellum
- Tectum
- Fourth ventricle
- Brain stem
  - Pons
  - Medulla

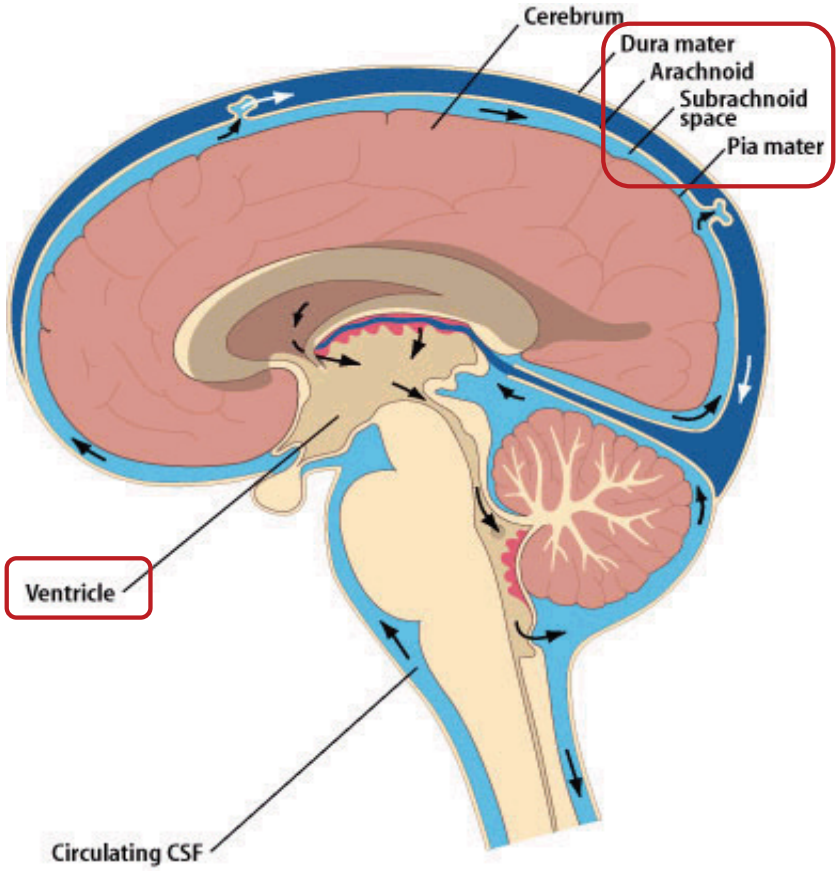




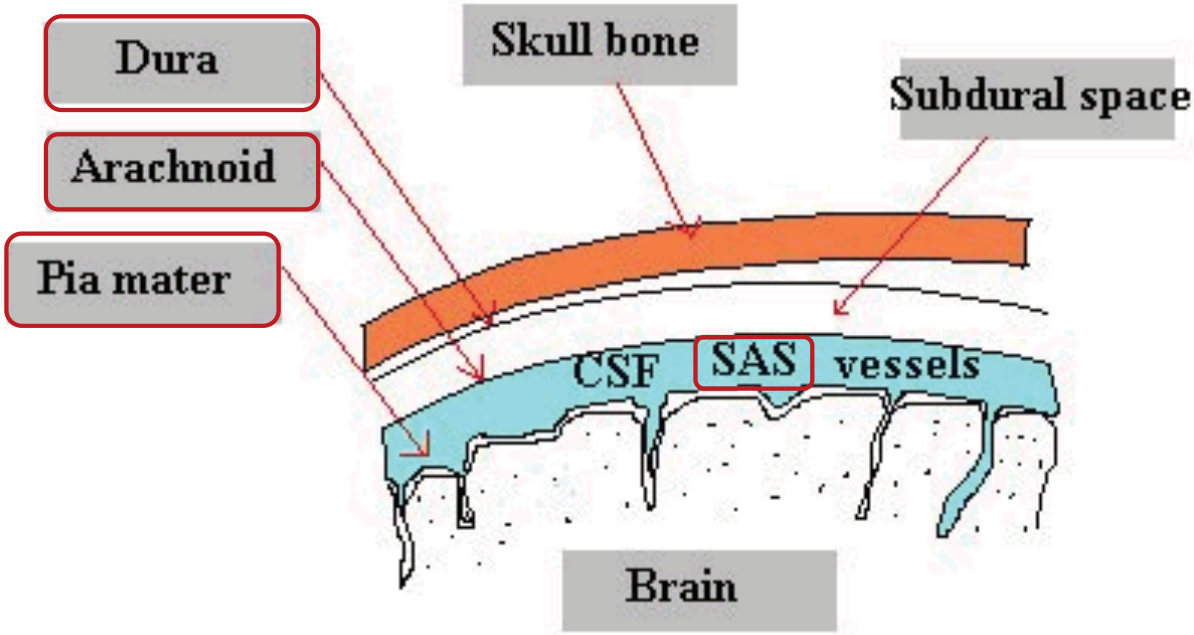
www.training.seer.cancer.gov/module anatomy/unit6 3 endo glnds1 pituitary.html



faculty.washington.edu/chudler/cranial.html

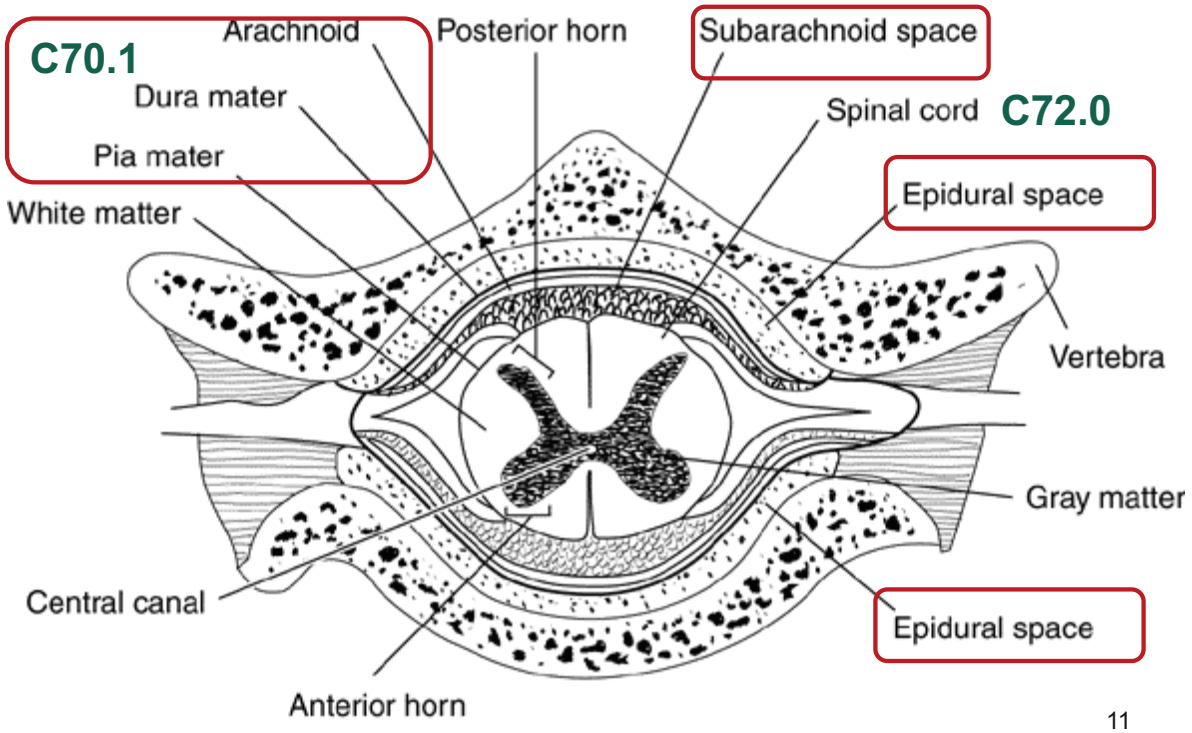


www.cardioliving.com/consumer/Stroke/Hemorrhagic_Stroke.shtm

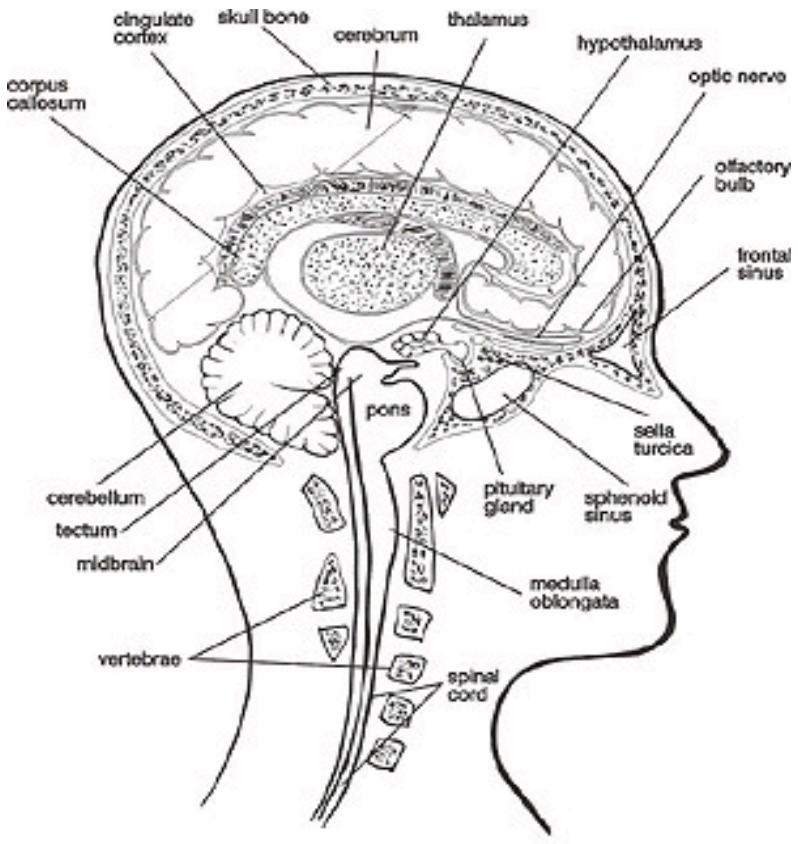


www.angelfire.com/wa/wafshaf50/CSF.html

SPINAL CORD



www.merck.com/pubs/mmanual/figures/182fig1.htm



Cross Section of the Brain

ABTA
Brain
Primer

Section Two

CASE SCENARIO

Case Scenario

2/2018 MRI Brain: 1.2 cm mass in Rt parietal region and abutting interhemispheric falx; most likely meningioma

3/2018 H&P: Pt presented w/ Lt hand tremor; MRI showed meningioma. Plan: Surveillance until symptoms appear

4/2019 MRI Brain: Minimal enlargement of Rt parietal meningioma, now 1.5 cm

12/2019 H&P: Stable meningioma; recent progressive lethargy, short term memory problems and confused speech. Plan: Imaging

Case Scenario, cont.

12/2019 CT Head: Lt frontal lobe mass extending into corpus callosum; surrounding edema. MRI brain: 5.1 cm cystic/solid mass centered in Lt frontal lobe and involving genu of corpus callosum; stable Rt parietal meningioma

1/2020 Op Note: Lt frontal craniotomy for excision of brain tumor; Tumor in Lt frontal lobe w/ extension to corpus callosum, achieved a gross total resection.

Path: Anaplastic oligodendroglioma, IDH-mutant and 1p/19q-codeleted, WHO grade III

Section Three

SOLID TUMOR RULES MALIGNANT CNS

Primary Sites

Topography Code	Primary Site(s)
C470-C479	Peripheral nerves
C700	Cerebral meninges
C701	Spinal meninges
C709	Meninges NOS
C710-C719	Brain
C720	Spinal cord
C721	Cauda equina
C722	Olfactory nerve
C723	Optic nerve
C724	Acoustic nerve
C725	Cranial nerve NOS
C728	Overlapping lesion of brain and central nervous system
C729	Nervous system NOS
C751	Pituitary gland
C752	Craniopharyngeal duct
C753	Pineal gland

STR Introduction

- Non-malignant (/0, /1) have separate set of rules
- Latest revision July 2019 for tumors dx'd 1/1/2018 and forward
- MUST have histology, cytology, radiology, or clinical dx of malignant (/3) behavior
- If mets found in brain from other primary site, do NOT use these rules
- North America ONLY: (juvenile) pilocytic astrocytoma = 9421/3 (WHO = 9421/1)
- Do not code MPs based on biomarkers
- See Hd/Nk rules for coding paragangliomas

Equivalent or Equal Terms

- And; with (for ≥ 2 histo in 1 tumor)
- Cerebrospinal fluid; CSF
- Dura; meninges
- Extradural; not w/in meninges; w/in cranium; w/in skull but not w/in cerebral meninges
- Extramedullary; outside medulla oblongata; C700
- Infratentorial; below tentorium cerebelli; cerebellum or brainstem
- Intracranial; within the skull, within the cranium
- Intradural; between layers of cerebral meninges; C700
- Intradural-extramedullary; w/in the spinal canal but outside of the nerves

Not Equivalent or Equal

- **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 MP/H

- 2016 CNS WHO presents
  - Major restructuring
  - New entities defined by histology AND molecular features
    - EX: GBM, IDH-wild-type (9440) vs GBM, IDH-mutant (9445)
  - "Not recommended" (aka obsolete) terms removed from histology tables
    - EX: Glioma NOS is an umbrella term for all gliomas and astrocytomas, and is not listed in the tables - new methods allow for better description

Changes From 2007 MP/H

- Rule change: GBM occurring after glial or astrocytic tumor = NEW primary
  - So they can know how often this happens
  - Primary GBMs develop w/o evidence of a less malignant precursor lesion – de novo
  - Secondary GBMs progress from low-grade diffuse astrocytoma or anaplastic astrocytoma
    - Differ in genetic signposts, better prognosis

Clarifications

- Intraosseous meningiomas & meningiomas of cavernous sinus & sphenoid wing ARE REPORTABLE
- Mult. cerebral meningiomas = single primary
- Mult. brain tumors (same histo) = single primary
- Laterality NOT used to determine mult primaries
- Timing NOT used to determine mult primaries
- Brain (C710-C719) is a single primary site
- NF, NF1, NF2, & schwannomatosis NOT reportable (Genetic syndromes - Code for NF in ICD-O-3 is **NA** 1/1/18+)
- New histologies denoted with an * in Table 3

Reportability Criteria

Must meet THREE conditions to be reported as malignant /3:

1. Behavior must be /3
   - Path designates as malignant/invasive, /3 OR
   - Tumor is WHO Grade 3 or 4 (Table 1)
   - WHO Grade 2 may be non-malignant or malignant
2. Primary site must be reportable (Table 2) AND
3. Histology must be reportable (Table 3)

Directory of Sections & Tables

- **Section 1: Behavior code**
  - Priority Order for using documentation to assign behavior
  - Table 1: WHO grades for CNS neoplasms
- **Section 2: Reportable primary sites & histologies**
  - Priorities for coding primary site
  - Reportable primary site groups
  - Table 2: Reportable primary sites
  - Table 3: Specific histo, NOS, & subtypes/variants
  - Table 4: Coding primary site for CNS & peripheral
- **Section 3: Additional info to complete abstract**
  - Conflicting Information on Path Reports
  - Table 5: Paired sites
  - Table 6: Non-malignant CNS tumors with potential to transform to malignant

Priorities for Assigning Behavior

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

1. Pathology from <u>resection</u>
   - Path describes malignant
   - WHO grade 3 or 4
     - WHO grade 2 can be malignant or non-malignant
   - NEVER change behavior described by pathologist
2. Pathology from <u>bx</u>
3. Cytology (CSF)
4. Physician's documentation (no path)
   - Tumor board > Documentation of original pathologic dx and behavior > Documentation of and behavior w/o mention of original dx
5. Radiology: MRI > CT> PET > Angiogram
6. When instructions 1-5 do not apply, use Table 1

Use these in priority order.
Stop at the **first** one that fits.

Table 1: WHO Grades for Select CNS Neoplasms

Histology	WHO Grade
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
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

Priority for Coding Primary Site

- Notes
  - Op report will distinguish between intracranial and intraspinal
  - Use NOS site code only when specific site is unknown
- Resection: Op > Path report
- Biopsy: Op > Path report
- Resection and/or biopsy done, but no op or path report available
  - Tumor board
  - MD statement original dx from op or path OR
  - MD statement of primary site
- Imaging (no resection): MRI > CT > PET > Angiogram
- See Table 2: Reportable sites, to confirm site is reportable
- See Table 4: When primary site is cranial or peripheral nerve

Reportable Primary Sites and Their ICD-O Codes

Reportable Primary Site Groups		
Intracranial	**Spinal sites**	**Peripheral nerves**
(within the skull/cranium)	(spinal meninges and sites w/in the spinal meninges, intradural)	(extracranial or extraspinal nerves)
Cerebral meninges C700	Spinal meninges C701	
Brain C710—710	Spinal cord C720	
Cranial nerves C722—C729	[Spinal nerve roots +	Peripheral nerves]
Intracranial glands Craniopharyngeal duct C752 Pineal gland C753 Pituitary gland C751	Cervical nerve (8 pair), occipital nerve C470 Coccygeal nerve (1 pair) C721 Lumbar nerve (5 pair) C721 Sacral nerve (5 pair) C721 Thoracic nerve (12 pair) C473	

Table 2: Reportable Primary Sites

Site Group	Reportable Subsite Terms and Code
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 **C475** Autonomic nervous system NOS **C479** Head, face and neck **C470** Lower limb and hip **C472** 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/conus medullaris/filum terminale **C721** Meninges NOS **C709** Spinal meninges **C701**

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

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
CNS neuroblastoma 9500		
Diffuse midline glioma H3 K27M mutant **9385***		
Embryonal carcinoma 9070		Yolk sac tumor 9071
Embryonal tumor with multilayered rosettes C19MC-altered **9478***	Embryonal tumor with multilayered rosettes, NOS ETMR	
Ependymoma 9391	Clear cell ependymoma Tanycytic ependymoma	Anaplastic ependymoma 9392 Ependymoma, RELA fusion-positive **9396*** Papillary ependymoma 9393
Epithelioid hemangioendothelioma **9133**		
Germinoma **9064**		

Table 4: Coding Primary Site for Malignant Tumors of Cranial and Peripheral Nerves

- Neoplasms arising in a **cranial** or **spinal** nerve are coded to the specific nerve in which they arise
- Neoplasms, commonly meningiomas, arising in the **dura/meninges** of:
  1. An **intracranial** nerve (cranial nerve within the skull) are coded to **cerebral meninges** C700*
  2. The **spinal nerve roots** are coded to the ICD-O site code **spinal meninges** C701*
- **Check** the **operative report** to determine whether the surgery is **intracranial** or **intradural**

*C700 and C701 are **not** listed anywhere in Table 4.

Table 4: Coding Primary Site for Malignant Tumors of Cranial & Peripheral Nerves

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**

Table 5: Paired Sites

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**

NEW for 2018: *Laterality is no longer a factor in determining multiple primaries.*

Table 6 Non-Malignant CNS Tumors with Potential to Transform to /3

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**

Malignant CNS: M rules

Unknown if Single or Multiple Tumors

M1 **SP** when not possible to determine if there is a single or multiple tumors

Single Tumor

M2 **SP** when there is a single tumor

M3 **SP** when original tumor is oligodendroglioma and subsequently recurs in residual tumor tissue w/ different features (**new rule for 2018**)

Malignant CNS: M rules, cont.

M4 SP (the malignant) when:

- Original tumor is /0 or /1 AND FCOT was active surveillance (no resection) and
- Subsequent tumor resection is malignant /3 (**new rule for 2018**)
- Diagnosis was:
  - Clinical
  - Radiographic
  - Stereotactic biopsy

🔎 The original tumor and the resected tumor must be the SAME tumor to use this rule.

Stereotactic Biopsy & M4

20 Local excision of tumor, lesion, or mass, excisional biopsy
21 Subtotal resection of tumor, lesion or mass in brain
22 Resection of tumor in spinal cord or nerve
[***SEER Note:*** Assign code 20 for stereotactic biopsy of brain tumor]

SEER Program Manual 2018

20 Local excision of tumor, lesion or mass; excisional biopsy
21 Subtotal resection of tumor, lesion or mass in brain
22 Resection of tumor of spinal cord or nerve

STORE 2018 Appendix B

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

Practice Rules?

4/15/2016: Patient in E.R. post MVA, complaining of headaches. MRI brain showed incidental 8mm right-sided meningioma. Neurosurgeon consult recommends observation.
9/3/2019: Recent MRI brain shows formerly suspected meningioma now enlarged to 1.8cm. Excision of tumor shows path of papillary meningioma

# Primaries	**1**	Dx Date	**04/15/2016**
M Rule used	**M4**	Histology	**9538/3**

Malignant CNS: M rules, cont.

Multiple Tumors

M5 **MP** when multiple CNS tumors, one is /3 and another is /0 or /1; simultaneous or metachronous

M6 **MP** when glial tumor, other than GBM, is followed by GBM 9440 (**change from the 2007 MP/H rules**)

M7 **SP** when separate, non-contiguous tumors in the brain C71.X with the same histology XXXX/3 (**change from/clarification to the 2007 MP/H rules**)

M8 MP when multiple tumors present in any of the following sites or subsites

Tumor in Site 1 AND	Tumor in Site 2 (not mets!)
Any lobe of the brain C710-C719	Any other part of CNS
Cauda equina C721	Any other part of CNS
Cerebral meninges C700	Spinal meninges C701
Cerebral meninges C700	Any other part of CNS
Any one of the cranial nerves C722-C725	Any other part of 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	Any other part of CNS
Spinal cord C720	Any other part of CNS
Spinal meninges C701	Any other part of CNS

Malignant CNS: M rules, *Multiple Tumors, cont.*

M9 **MP** when separate, non-contiguous tumors are ≥ 2 subtypes/variants in column 3, Table 3

M10 **SP** when separate, non-contiguous tumors are on the same row in Table 3 (excludes different subtypes of same NOS)

M11 **MP** when separate, non-contiguous tumors are on different rows in Table 3

M12 **SP** when multiple tumors do not meet any of the previous criteria

Table 3: Rules M9 – M11

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Anaplastic ganglioglioma **9505**		
Astroblastoma **9430**		
Astrocytoma NOS **9400**	Diffuse astrocytoma IDH-mutant Diffuse astrocytoma IDH-wildtype Diffuse astrocytoma NOS	Anaplastic astrocytoma IDH-mutant/wildtype; anaplastic astrocytoma NOS **9401** 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 9473

M10, Same row = SP
- Same histo
  - Synonyms or
  - Col. 1 + Col. 2
- Col. 1 + 1 sub/var Col. 3
- Col. 2 + 1 sub/var Col. 3

M11: Different rows = MP

M9: Different subtypes = MP

Priority Order for using Documentation to Identify Histology

Code histo prior to neoadjuvant Tx; do not change histo to make the case applicable to staging

1. Pathology/Tissue from **resection**
   a. *Biomarkers*
   b. Addendum/comment
   c. Final dx/synoptic report
   d. CAP protocol
2. Pathology/Tissue from **biopsy**
   Same list as a – d above
3. Cytology (CSF)
4. Tissue/path from mets
5. Scan: MRI > CT > PET > Angiogram
6. MD documentation when above N/A
   a. Treatment plan
   b. Tumor board
   c. Medical record (original path, cytol, imaging)
   d. Doctor's reference

Coding Histology (Single Tumor)

- Code **most specific** histology or subtype/variant regardless of whether it is described as:
  - Majority or predominant part of tumor
  - Minority part of tumor
  - A component
- Code histo described as differentiation or features/features of ONLY when there is a specific ICD-O code for the "NOS with _____ features" or "NOS with _____ differentiation"
- Use Ambiguous Terms ONLY when
  - Case accessioned based on ambiguous terminology
  - NOS and more specific histo described by ambiguous term and the more specific histo is confirmed by a physician OR the patient is being treated for the more specific histo
- Do **NOT** code histo when described as architecture, focal, or pattern

Examples of Coding Most Specific Histology

1. Astrocytoma 9400 w/ majority of tumor being anaplastic astrocytoma IDH-mutant 9401. anaplastic astrocytoma IDH-mutant 9401
2. CNS ganglioneuroblastoma 9490 w/ minority of tumor being CNS embryonal tumor 9473. CNS embryonal tumor 9473
3. Ependymoma 9391 w/ a component of anaplastic ependymoma 9392.

   anaplastic ependymoma 9392

Malignant CNS: H Rules Single (*Multiple*) Tumor*(s)*

H1: Code the reportable CNS tumor (Table 3) when patient has NF1, NF2, or Schwannomatosis

H2 (*H5*): Code malignant meningioma 9530 when dx specifically states malignant/invasive

H3 (*H6*): Code the histology when a single histology is present

H4 (*H7*): Code the subtype/variant when NOS and a single subtype/variant of that NOS

(Rule number for multiple tumors is in *italics* and parentheses.)

Section FOUR

SOLID TUMOR RULES NON-MALIGNANT CNS

Primary Sites

Topography Code	Primary Site(s)
C700	Cerebral meninges
C701	Spinal meninges
C709	Meninges NOS
C710-C719	Brain
C720	Spinal cord
C721	Cauda equina
C722	Olfactory nerve
C723	Optic nerve
C724	Acoustic nerve
C725	Cranial nerve NOS
C728	Overlapping lesion of brain and central nervous system
C729	Nervous system NOS
C751	Pituitary gland
C752	Craniopharyngeal duct
C753	Pineal gland

STR Introduction

- Malignant (/3) have separate set of rules
- Latest revision July 2019 for tumors dx'd 1/1/2018 and forward
- Non-malignant CNS neoplasms are reportable for cases diagnosed 1/1/2004 and forward
- North America ONLY: (juvenile) pilocytic astrocytoma = 9421/3 (WHO = 9421/1)
  - When primary site is **optic nerve**, behavior is /1
- Do not code MPs based on biomarkers
- See Hd/Nk rules for coding paragangliomas

CNS Equivalent or Equal Terms

- And; with (for ≥ 2 histo in 1 tumor)
- Atypical; uncertain behavior /1
- Cerebrospinal fluid; CSF
- Dermoid; dermoid cyst
- Dura; meninges
- Extradural; not w/in meninges; w/in cranium; w/in / skull but not w/in cerebral meninges
- Extramedullary; outside medulla oblongata; C700
- Intracranial; within the skull, within the cranium
- Intradural; between layers of cerebral meninges; C700

CNS Equivalent or Equal Terms, cont.

- Intradural-extramedullary; w/in the spinal canal but outside of the nerves
- Intraspinal; occurring w/in the spinal column especially the vertebral canal; spinal nerve roots
- Majority; major; predominantly; > 50%
- Non-malignant; /0; /1 (uncertain, borderline malignancy, LMP, uncertain malignant potential)
- Site; topography
- Tumor; mass; lesion; neoplasm (only to determine MPs)
- Type; subtype; variant

Not Equivalent or Equal

- **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**

Clarifications

- Intraosseous meningiomas & meningiomas of cavernous sinus & sphenoid wing ARE REPORTABLE
- Mult. cerebral meningiomas = single primary
- Mult. brain tumors (same histo) = single primary
- Bilat optic nerve gliomas/pilocytic astro = single primary
- Laterality NOT used to determine mult primaries
- Timing NOT used to determine mult primaries
- Brain (C710-C719) is a single primary site
- NF, NF1, NF2, & schwannomatosis NOT reportable (Genetic syndromes - Code for NF is **NA** 1/1/18+)
- New histologies denoted with an * in Table 3

Reportability Criteria

- Must meet THREE conditions to be reported as non-malignant /0 or /1:
  1. Behavior must be /0 or /1 on pathology **OR**
     - Tumor is WHO Grade I **AND**
  2. Primary site must be reportable (Tables 3 and 4) **AND**
  3. Histology must be reportable (Tables 5 and 6)

Directory of Sections & Tables

- **Section 1: Behavior code**
  - Priority Order for using documentation to assign behavior
  - Table 1: WHO grades for CNS neoplasms
- **Section 2: Reportable primary sites & histologies**
  - Priorities for coding primary site
  - Reportable primary site groups
  - Table 2: Reportable primary sites
  - Table 3: Reportable Cranial Nerve Tumors
  - Table 4: Non-reportable Neoplasms
  - Table 5: Histologic Types
  - Table 6: Reportable specific & NOS histo

Directory of Sections & Tables

- **Section 3: Additional info to complete abstract**
  - Conflicting Information on Path Reports
  - Table 7: Paired sites
  - Table 8: Non-malignant CNS tumors with potential to transform to malignant

Priorities for Assigning Behavior

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

1. Pathology from resection
   - Use pathologist's description of behavior
     - Never change the behavior assigned by the pathologist
   - WHO grade 1
     - WHO grade 2 can be malignant or non-malignant
     - Use the pathologist's description of behavior
2. Pathology from bx
3. Cytology (CSF)
4. Physician's documentation (no path)
   - Tumor board > Documentation of original pathologic dx and behavior > Documentation of and behavior w/o mention of original dx
5. Radiology: MRI > CT> PET > Angiogram
6. When instructions 1-5 do not apply, use Table 1

Use these in priority order. **Stop** at the **first** one that fits.

Table 1: WHO Grades for Select CNS Neoplasms

Histology	WHO Grade
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
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

Reportable Primary Sites and Histologies

Non-Malignant Meningiomas		
Location	**ICD-O**	**Notes**
Intraosseous	C700	Dura layer of meninges contacts the endosteum of skull bones
Sphenoid Wing	C700	Arise in meninges covering sphenoid wing bone Can be very invasive spreading to dura of frontal, temporal, and orbital regions
Cavernous sinus	-	Between the endosteal and meningeal layers of the dura (no ICD-O code for cavernous sinus)
	C725	Cranial nerves passing through the sinus (trochlelar and abducens(t))
	C700	Meninges covering the cranial nerve

Cavernous sinus hemangiomas are reportable.
Code primary site to cerebral meninges C700.

Priority for Coding Primary Site

- Notes
  - Op report will distinguish between intracranial and intraspinal
  - Use NOS site code only when specific site is not known
  - See Table 2 to confirm site is reportable
  - See Table 3 when primary site is cranial or peripheral nerve
  - See Table 4 for primary site/histology combos that are NR
  - See Table 5 when primary site is brain or intracranial glands
- Resection: Op > Path report
- Biopsy: Op > Path report
- Resection and/or biopsy done, but no op or path report available
  - Tumor board
  - MD statement original dx from op or path OR
  - MD statement of primary site
- Imaging (no resection): MRI > CT > PET > Angiogram

Reportable Primary Sites and Their ICD-O Codes

Reportable Primary Site Groups	
Intracranial	**Spinal sites**
(within the skull/cranium)	(spinal meninges and sites w/in the spinal meninges, intradural)
Cerebral meninges C700	Spinal meninges C701
Brain C710—710	Spinal cord C720
Cranial nerves C722—C729	
Intracranial glands Craniopharyngeal duct C752 Pineal gland C753 Pituitary gland C751	

Table 2: Reportable Primary Sites

Site Group	Reportable Subsite Terms and Code
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 Except for **this** shaded row which is ONLY in the Malignant CNS/PN rules, the table is identical in both sets of rules.	Abdomen **C475** Autonomic nervous system NOS **C479** Head, face and neck **C470** Lower limb and hip **C472** 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/conus medullaris/filum terminale **C721** Meninges NOS **C709** Spinal meninges **C701**

Table 3: Reportability of Non-Malignant Cranial Nerve Tumors

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**.

Table 4: Non-Reportable Neoplasms

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**	
Fibermoma	**No code**	
Glomus tympanicum, glomus jugulare	**8690/1**	These tumors occur in the inner ear, the aortic body and other paraganglia respectively; sites for which non-malignant tumors are **not reportable**
Hygroma	**9173/0**	
Hypothalamic hamartoma	**No code**	Occurs in hypothalamus

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

Table 5: Histologic Types of Non-Malignant Intracranial Tumors

Histology Term and Code	Most Common 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	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**
Pilocytic astrocytoma/juvenile pilocytic astrocytoma 9421/1	Posterior fossa **C719,** cerebrum **C710**
Pineocytoma 9361/1	Pineal gland **C753**
Pituicytoma 9432/1	Pituitary gland **C751**, sella turcica **C751,** suprasellar **C719**

Table 6: Specific Histologies/NOS, Synonyms, & Subtypes/Variants

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 8860/0		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** Transitional meningioma **9537/0**
Myofibroblastoma 8825/0		Inflammatory myofibroblastic tumor **8825/1**

Table 7: Paired Sites

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**

***New for 2018**: Laterality is no longer a factor in determining multiple primaries.*

Table 8 Non-Malignant CNS Tumors with Potential to Transform to /3

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**

Shaded terms = mesenchymal tumors (see next slide)

Mesenchymal Tumors

- Mesenchymal, non-meningothelial tumors
  - Mesenchymal tumors originate from mesodermal tissue that forms various connective tissues
  - In CNS sites, they most commonly arise from the meninges rather than CNS parenchyma
  - Code to the CNS site in which they arise
    - Example: Hemangioma 9120/0 in the cerebral meninges ⟹ Code primary site to C70.0

Non- Malignant CNS: M rules

Unknown if Single or Multiple Tumors

M1 **SP** when not possible to determine if there is a single or multiple tumors

Single Tumor

M2 **SP** when there is a single tumor

M3 **SP** (the malignant) when original tumor is /0 or /1 AND FCOT was active surveillance (no resection) and subsequent tumor resection is malignant (**new rule for 2018**)

Stereotactic Biopsy & M3

20 Local excision of tumor, lesion, or mass, excisional biopsy
21 Subtotal resection of tumor, lesion or mass in brain
22 Resection of tumor in spinal cord or nerve
[***SEER Note:*** Assign code 20 for stereotactic biopsy of brain tumor]

SEER Program Manual 2018

20 Local excision of tumor, lesion or mass; excisional biopsy
21 Subtotal resection of tumor, lesion or mass in brain
22 Resection of tumor of spinal cord or nerve

STORE 2018 Appendix B

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

M3 Notes

- Single tumor is **always** a single primary
  - Malignant behavior is reported
- Use malignant CNS rules to code histo
  - Resection path is more accurate than clinical, radiographic, or bx info
- No time requirement from initial dx to resection
- Edit original abstract as follows
  - Do not change date of dx
  - For cases that have been abstracted, change behavior to /3
  - Report all data changes for cases that have already been submitted to the central registry

Non- Malignant CNS: M rules cont.

Single Tumor

M4 **SP** when benign tumor /0 transforms to an uncertain/borderline tumor /1 (must be same histo or NOS and subtype/variant)

- Don't change behavior or date of dx on original abstract (both /0 and /1 are non-malignant)
- A single tumor is always a single primary

Non- Malignant CNS: M rules cont.

Multiple Tumors

M5 **MP** when malignant tumor /3 occurs after a non-malignant tumor /0 or /1 AND non-malignant tumor was resected or unknown if resected

- Use malignant rules for the second tumor

M6 **SP** when bilateral acoustic neuromas/ vestibular schwannomas 9560/0 or optic glioma/pilocytic astrocytoma 9421/1

- Tumors can be simultaneous or the contralateral tumor can be diagnosed at any time following the original diagnosis

M7 MP when multiple tumors present in any of the following sites or subsites

Tumor in Site 1 AND	Tumor in Site 2
Any lobe(s) of the brain **C710-C719**	Any other part of CNS
Cauda equina **C721**	Any other part of CNS
Cerebral meninges **C700**	Spinal meninges **C701**
Cerebral meninges **C700**	Any other part of CNS
Any the cranial nerve(s) C722-C725	Any other part of CNS
Meninges of cranial nerves	Any other part of CNS
Spinal cord C720	Any other part of CNS
Spinal meninges C701	Any other part of CNS

Non- Malignant CNS: M rules *Multiple Tumors* cont.,

M8 **MP** when separate, non-contiguous tumors are ≥ 2 subtypes/variants in column 3, Table 6

M9 **SP** when separate, non-contiguous **meningiomas** arise in the cranial meninges
- Same histo or NOS and single subtype/variant
- Laterality doesn't matter

M10 **SP** when separate, non-contiguous tumors in the **brain** with the same histology (XXXX)
- Laterality doesn't matter
- Lobe of brain doesn't matter
- **Change from/clarification to the 2007 MP/H rules**

Non- Malignant CNS: M rules *Multiple Tumors* cont.,

M11 **SP** when separate, non-contiguous tumors are on the **same** row in column 3, Table 6

M12 **MP** when separate, non-contiguous are on **different** rows in column 3, Table 6

M13 **SP** when multiple tumors do not meet any of the previous criteria

Table 6: Rules M8, M11, & M12

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		
Leiomyoma 8890/0		
Lipoma 8860/0		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** Transitional meningioma **9537/0**
Myofibroblastoma 8825/0		Inflammatory myofibroblastic tumor **8825/1**

M11, Same row = SP
- Same histo
  - Synonyms or
  - Col. 1 + Col. 2
- Col. 1 + 1 sub/var Col. 3
- Col. 2 + 1 sub/var Col. 3

M12: Different rows = MP

M8: Different subtypes = MP

Priority Order for using Documentation to Identify Histology

Code histo prior to neoadjuvant Tx; do not change histo to make the case applicable to staging

1. Pathology/Tissue from **resection**
   a. Addendum/comment
   b. Final dx/synoptic report
   c. CAP protocol
   d. Biomarkers
2. Pathology/Tissue from **biopsy**
   Same list as a – d above
3. Cytology (SF)
4. Tissue/path from mets
5. Scan: MRI > CT > PET > Angiogram
6. MD documentation when above N/A
   a. Treatment plan
   b. Tumor board
   c. Medical record (original path, cytol, imaging)
   d. Doctor's reference

Coding Histology (Single Tumor)

- Code **most specific** histology or subtype/variant regardless of whether it is described as:
  - Majority or predominant part of tumor
  - Minority part of tumor
  - A component
- Code histo described as differentiation or features/features of ONLY when there is a specific ICD-O code for the "NOS with ____ features" or "NOS with ____ differentiation"
- Use Ambiguous Terms ONLY when
  - Case accessioned based on ambiguous terminology
  - NOS and more specific histo described by ambiguous term and the more specific histo is confirmed by a physician OR the patient is being treated for the more specific histo
- Do NOT code histo when described as architecture, focal, or pattern

Examples of Coding Most Specific Histology

1. Choroid plexus papilloma 9390/0 w/ majority of tumor being atypical choroid plexus papilloma 9390/1.

   atypical choroid plexus papilloma 9390/1

2. Meningioma 9530/0 w/ minority of tumor being atypical meningioma 9539/1.

   atypical meningioma 9539/1

3. Schwannoma 9560/0 w/ a component of melanotic schwannoma 9560/1 .

   melanotic schwannoma 9560/1

Non-Malignant CNS: H Rules Single (*Multiple*) Tumor*(s)*

H1 (*H5*): Code meningioma 9530/0 when dx is:

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

H2 (*H7*): Code the reportable CNS tumor (Table 6) when patient has NF1, NF2, or Schwannomatosis

(Rule number for multiple tumors is in in *italics* and parentheses.)

Non-Malignant CNS: H Rules *Single* (*Multiple*) Tumor*(s)*

H3 (*H8*): Code the histology when a single histology is present

H4 (*H9*): Code the subtype/variant when NOS and a single subtype/variant of that NOS

H6: Code meningioma 9530/1 when multiple meningiomas of uncertain behavior

(Rule number for multiple tumors is in in *italics* and parentheses.)

Case Scenario: Primary Site and Histology

How many primaries?	**2**
Which rule?	**Non-malignant rule M7** **Malignant rule M5**
Primary Site 1:	**Cerebral meninges C70.0**
Histology 1:	**Meningioma 9530/0**
Primary Site 2:	**Frontal lobe C71.1**
Histology 2	**Anaplastic oligodendroglioma, IDH-mutant and 1p/19q-codeleted 9451/3**

Section Five

STAGING
SEER SUMMARY 2018 & EOD

Brain Lymphoma, Histiocytic Tumors, Germ Cell Tumors

- Site codes C70._, C71._, C72.4-5 & C72.8-9, C75._ moved from Ch. 80 (Lymphoma) to Chapter 72 (Brain and Spinal Cord)

Code	Lymphoma	Code	Germ Cell
9702	ALCL, ALK neg	9100	Chorioca
9714	ALCL, ALK +	9070	Embryonal
9680	DLBCL	9064	Germinoma
9712	ILBCL	9080	Immature teratoma
9699	MALT/dura	9085	Mixed germ cell
	Histiocytic	9084	Teratoma w/malig transformation
9750	Erdheim-Chester	9071	Yolk sac
9755	Histiocytic sarcoma		*Per WHO CNS 4th Ed*
9571	Langerhans cell histiocytosis		*Grade Table? SSDI for CNS probably "9"*

Ambiguous Terms EOD and SS2018

- Use the lists of ambiguous terms to interpret the intent of the clinician ONLY when further documentation is not available and/or there is no specific statement of involvement in the medical record. The physician's definitions/ descriptions and choice of therapy have priority over these lists because individual clinicians may use these terms differently.

Ambiguous Terms

Same list in EOD and SS18

Involved

Adherent
Apparent(ly)
Appears to
Comparable with
Compatible with
Consistent with
Contiguous/continuous with
Encroaching upon*
Extension to, into, onto, out onto
Features of
Fixation to a structure other than primary**
Fixed to another structure**
Impending perforation of
Impinging upon
Impose/imposing on
Incipient invasion
Induration
Infringe/infringing
Into*
Intrude
Most likely
Onto*
Overstep
Presumed
Probable
Protruding into (unless encapsulated)
Suspected
Suspicious
To*
Up to

Ambiguous Terms

Same list in EOD and SS18

Not Involved

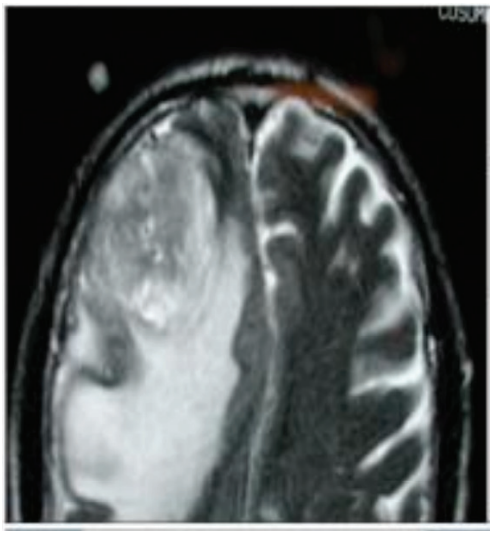
Abuts
Approaching
Approximates
Attached
Cannot be excluded/ruled out
Efface/effacing/effacement
Encased/encasing
Encompass(ed)
Entrapped
Equivocal
Extension to without invasion/involvement of
Kiss/kissing
Matted (except for lymph nodes)
Possible
Questionable
Reaching
Rule out
Suggests
Very close to
Worrisome

BRAIN SS18 and EOD Notes

- Assign SS18 code 8 and EOD Primary tumor code 050 for benign or borderline brain tumors
  - If other /0 or /1 tumors collected, use 9/999
- SS18 Codes 0, 3, and 4 are not applicable
- Midline shift ≠ crossing the midline
  - It must state tumor crosses midline
- Discontiguous spread, including circulating cells in cerebrospinal fluid (CSF), is coded in EOD Mets
- ICD-O-3 codes C71.0 and C71.9 include both supratentorial and infratentorial subsites

Midline Shift

- Confined space within cranium – not much "give" when tumor grows or swelling occurs
- Midline is a shift of the brain past its center line
  - Pushes midline out of alignment (off to the side)
  - Sign of intracranial pressure



Source: Medscape

BRAIN SS18 and EOD Notes

C71.0-C71.9

Infratentorial sites

- All subsites for codes C716-C717
- Hypothalamus (C710)
- Pallium (C710)
- Posterior cranial fossa (C719)
- Thalamus (C710)

Supratentorial sites

- All subsites for codes C711-C715
- Primary site C710 (excluding hypothalamus, pallium, thalamus)
- Anterior cranial fossa (C719)
- Corpus callosum (C718)
- Middle cranial fossa (C719)
- Tapetum (C718)
- Suprasellar (C719)

SS18 & EOD Primary Tumor Codes

SS18	EOD	Description
8	050	Benign or borderline brain tumor
1	100	Confined to brain, NOS; (Localized NOS included in SS2018)
		Confined to ventricles
		Confined to meninges, NOS (to be added to SS18 in v2)
		Infratentorial tumor confined to: Brain stem or meninges of brain stem (one side)
		Medulla oblongata | Midbrain (mesencephalon) | Pons
		Cerebellum or meninges of cerebellum (one side or midline)
		Lateral lobes | Median lobe of cerebellum | Vermis
		Hypothalamus Infratentorial tumor Both cerebellum & brain stem involved w/tumor on one side Supratentorial tumor confined to
		Frontal lobe | Occipital lobe | Parietal lobe | Temporal lobe
		Tumor invades or encroaches upon ventricular system

SS18 & EOD Primary Tumor Codes, cont.

SS18	Code	Description
2	500	Bone (skull) Contralateral hemisphere Corpus callosum (including splenium) Major blood vessel(s) Meninges (e.g. dura) Nerves (cranial, NOS) Spinal cord/canal Supratentorial tumor extends infratentorially to involve: Brain Stem, Cerebellum, Hypothalamus Pallium, Posterior cranial fossa, Thalamus Infratentorial tumor extends supratentorially to involve: Anterior cranial fossa Cerebrum (cerebral hemisphere) (excluding hypothalamus, pallium, thalamus) Corpus callosum, Middle cranial fossa Suprasellar brain, Tapetum Tumor crosses the midline

SS18 & EOD Primary Tumor Codes, cont.

SS18	EOD	Description
7	700	Circulating cells in cerebral spinal fluid (CSF) Nasal cavity Nasopharynx Other direct extension outside CNS Posterior pharynx Further contiguous extension
U	800	No evidence of primary tumor
9	999	Unknown if extension or mets (SS18) Unknown; extension not stated; Primary tumor cannot be assessed; Not documented in patient record; DCO

SS18 and EOD RLN & Mets Codes

REGIONAL LYMPH NODES		
SS18	**EOD**	**Description**
N/A	**888**	Not applicable. Info not collected for this schema

METS		
SS18	**EOD**	**Description**
-	**00**	No distant mets; unknown if distant mets
7	**10**	Distant lymph node(s)
	70	Mets within CNS & CSF pathways "Drop" mets Mets outside the CNS Extra-neural mets Carcinomatosis Distant mets WITH or WITHOUT distant LN Distant mets, NOS
U	**99**	Death certificate only

Case Scenario: EOD and SS2018 Fields

Primary 1: 9530/0		**Primary 2: 9451/3**	
EOD Primary Tumor	**050**	EOD Primary Tumor	**500**
EOD Regional Nodes	**888**	EOD Regional Nodes	**888**
EOD Mets at Dx	**00**	EOD Mets at Dx	**00**
SS2018	**8**	SS2018	**2**

Section 6

GRADE & SSDI (NO SSDI FOR INTRACRANIAL GLANDS C75X)

Grade Fields Common Instructions

- Assign highest grade from primary tumor during specified time frame (c, p, *yc*, *yp*)
- *Multiple tumors w/ different grades abstracted as a single primary, assign highest grade*
- Codes 1-4 take priority over A-D, L and H
- Assign WHO grade from AJCC 8th edition Table 72.2 when grade not documented in record
- *For benign tumors ONLY, assign grade 1 for all histologies*

*Blue (italics) font represents **proposed** v2.0 updates.*

Grade Clinical

- Cannot be blank
- Assign grade 9 when
  - Grade from primary tumor not documented
  - Clinical workup not done (incidental finding during surgery for another condition)
  - Grade checked N/A on CAP Protocol and no other grade information available
- If only 1 grade available, and cannot determine if clinical or pathological, assume it is grade clinical and assign 9 for grade pathological and blank for grade post therapy

Grade Pathological

- Cannot be blank
- Use grade clinical as follows
  - Behavior
    - Same for clinical and pathological dx and clinical grade is higher
    - *Clinical dx is invasive and pathological dx is in situ*
  - Surgical resection of primary tumor
    - Performed
      - No grade documented on surgical resection
      - No residual tumor
      - *Not performed, but positive microscopic confirmation of distant mets (pM1) during clinical timeframe*

*Blue (italics) font represents **proposed** v2.0 updates.*

Grade Pathological, cont.

- Assign grade 9 when
  - Grade from primary tumor not documented
  - No resection of primary site *(exception when pM1 found during clinical workup)*
  - Neoadjuvant therapy is followed by resection (yp)
  - Clinical case only
  - Grade checked N/A on CAP Protocol and no other grade information available
  - Only 1 grade available, and cannot determine if clinical or pathological, assume it is grade clinical and assign 9 for grade pathological and blank for grade post therapy

*Blue (italics) font represents **proposed** v2.0 updates.*

Grade Post-Therapy (*yc*, yp)

- Leave blank when:
  - No neoadjuvant therapy
  - Clinical or pathological case only
  - Only 1 grade available, and unknown if c, p, yc, or yp
- Code 9 when:
  - *Microscopic exam (yc)* / Surgical resection (yp) performed after neoadjuvant therapy, and
    - Grade from primary not documented
    - No residual tumor
  - Grade checked N/A on CAP Protocol and no other grade info available

*Blue (italics) font represents **proposed** v2.0 updates.*

Grade ID Table 24 – Chap 72: Brain, CNS, Intracranial Gland

CODE	Grade Description	
1	WHO Grade I	Text describing meanings
2	WHO Grade II	included in table for code 1-4
3	WHO Grade III	
4	WHO Grade IV	
L	Stated as “low grade” NOS	
H	Stated as “high grade” NOS	
A	Well differentiated	
B	Moderately differentiated	
C	Poorly differentiated	
D	Undifferentiated, anaplastic	
9	Unknown; can’t assess	

Case Scenario: Grade Fields

Primary 1: 9530/0

Grade Clinical	1
Grade Pathological	9
Grade Post-therapy	-

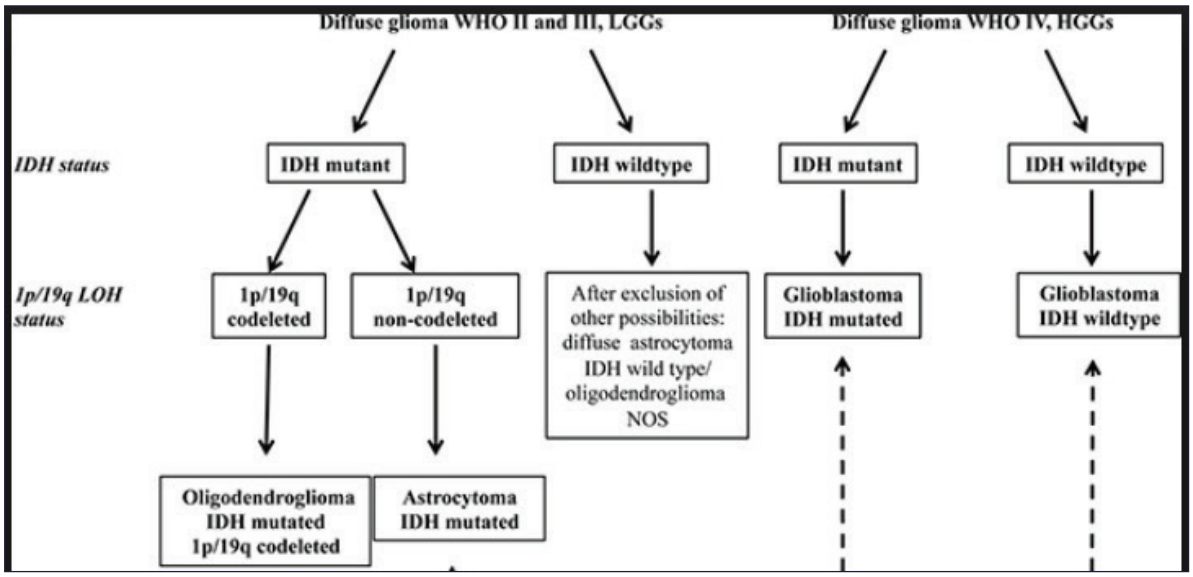
Primary 2: 9451/3

Grade Clinical	9
Grade Pathological	3
Grade Post-therapy	-

Biomarkers for Brain Tumors (source: www.abta.org)

Biomarker	Type of Tumor	Use of Test
MGMT	ANAPLASTIC astrocytoma, glioma, oligodendroglia, oligoastrocytoma Glioblastoma	Predictive (chemo response?)
IDH1/IDH2	Mainly low grade gliomas	Diagnostic, Prognostic
1p/19q	Oligoastrocytoma,Oligodendroglioma	Prognostic
BRAF	Astrocytoma, WHO grade 1/2 Pilocytic astrocytoma WHO grade 1	Diagnostic (pediatric gliomas) Predictive (immuno?)
EGFR, EGFRvIII	Glioblastoma, GBM	Prognostic
PTEN	Astrocytoma Glioblastoma, GBM	Diagnostic
TERT	Astrocytoma, WHO grade 2/3 Glioblastoma, GBM Oligodendroglioma	Prognostic
ATRX	Astrocytoma, WHO grade 2/3 Secondary glioblastoma	Prognostic
Akt3	Glioblastoma, GBM	Predictive (resistant to tx?)

Molecular Characterization of Diffuse Gliomas (WHO 2016)



Oncotarget. 2017; 8:57134-57148

SSDI Brain: Molecular Markers

- Assign 85 if histology is not 9400/3, 9401/3, 9440/3, 9450/3, 9451/3, 9471/3 or 9478/3
- MD statement of subtype can be used for this data item
- Only 1 code is applicable for each tumor
- Codes distinguish clinically important subtypes

SSDI: Brain Molecular Markers

CODE	Description
01	Diffuse astrocytoma, IDH-mutant (9400/3)
02	Diffuse astrocytoma, IDH-wildtype (9400/3)
03	Anaplastic astrocytoma, IDH-mutant (9401/3)
04	Anaplastic astrocytoma, IDH-wildtype (9401/3)
05	Glioblastoma, IDH-wildtype (9440/3)
06	Oligodendroglioma, IDH-mutant and 1 p/19 q co-deleted (9450/3)
07	Anaplastic oligodendroglioma, IDH-mutant and 1p/19q co-deleted (9451/3)
08	Medulloblastoma, SHH-activated and TP53-wildtype (9471/3)
09	Embryonal tumor w/multi-layered rosettes, C19MC-altered (9478/3)
85	N/A; histo not 9400/3, 9401/3, 9440/3, 9450/3, 9451/3, 9471/3, 9478/3
86	Benign or borderline tumor
87	Test ordered, results not in chart
88	N/A; Info not collected in this case
99	Not documented in record; no micro confirmation; not assessed /unk

2 SSDI: Chromosome 1p and Chromosome 19q LOH

- MD statement of chromosome 1p/19q deletion/LOH can be used
- Molecular test performed on tumor to ID genetic material normally found on the:
  - Short arm (p) of chromosome 1
  - Long arm (q) of chromosome 19

  Often performed @ same time and reported on single report
- Heterozygous = normal cells have 2 copies of each chromosome (1 from each parent)
- LOH = abnormal state reflecting loss of the
  - Entire short arm of chromosome 1
  - Entire long arm of chromosome 19
- LOH also termed [whole] arm loss, gene deletion, 1p/19q fragment analysis, and allelic loss

2 SSDI: Chromosome 1P (19q): Loss of Heterozygosity (LOH)

Same codes for both SSDI

CODE	Description
0	Chromosome 1p (19q) deletion/LOH not identified/not present
1	Chromosome 1p (19q) deletion/LOH identified/present
6	Benign or borderline tumor
7	Test ordered, results not in chart
8	N/A; Info not collected in this case
9	Not documented in record; no micro confirmation; not assessed /unknown if assessed

- LOH results in failure of tumor suppression
- Sensitivity to chemotherapy agents, such as lomustine, procarbazine, and vincristine, is increased with either 1p or 19q LOH

SSDI: Methylation of MGMT

- MD statement can be used
- MGMT is an enzyme that repairs DNA Methylation of the MGMT gene reduces the production of the MGMT enzyme
  - Can't repair DNA damaged by chemo → could indicate prolonged survival
- If MGMT gene is methylated, patient is more likely to respond to alkylating agents (temozolmide [Temodar] and the nitrosoureas)
- MGMT also called MGMT promoter methylation, methylation status

SSDI: MGMT (Methylation of 06-methylguanine-methyltransferase

CODE	Description
0	MGMT methylation absent/not present, unmethylated MGMT
1	MGMT methylation present, low level; hypomethylated; partial methylated
2	MGMT methylation present, high level; hypermethylated
3	MGMT methylation present, level unspecified
6	Benign or borderline tumor
7	Test ordered, results not in chart
8	N/A; Info not collected in this case
9	Not documented in record; no micro confirmation; not assessed /unk

MGMT is enzyme that repairs DNA which is BAD in tumors because the DNA repair may allow them to repair damage that chemo does to the cancer cells

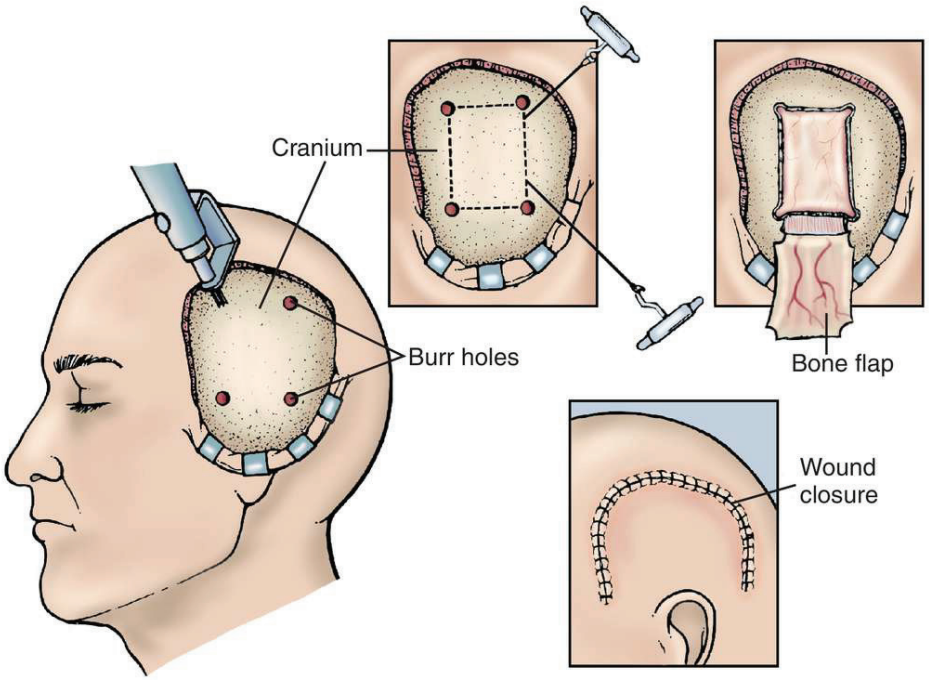
Case Scenario: SSDI Fields

Primary 1: 9530/0		**Primary 2: 9451/3**	
Brain molecular markers	**86**	Brain Molecular Markers	**07**
LOH 1p	**6**	LOH 1p	**1**
LOH 19q	**6**	LOH 19q	**1**
Methylation MGMT	**6**	Methylation MGMT	**9**

Section 7

TREATMENT

Craniotomy



Mosby's Medical Dictionary, 9th edition. © 2009, Elsevier

Brain Surgery Codes

- Surgery codes apply to:
  - Meninges C70.0–C70.9
  - Brain C71.0–C71.9
  - Spinal Cord, Cranial Nerves and Other Parts of Central Nervous System C72.0–C72.9
- Histology Exclusions:
  - M-9727, 9732, 9741-9742, 9762-9809, 9832, 9840-9931, 9945-9946, 9950-9967, and 9975-9992
- Notes:
  - Do not code laminectomies for spinal cord primaries

Stereotactic Brain/CNS Biopsy

CoC Facilities

- **Intent** is for diagnosis
  - Code in Surg Dx/Stg Proc
    - No STORE instruction to code stereotactic bx to 20 in surgery codes
- **Intent** is treatment
  - Code to 20 (excisional biopsy) in surgery field since removing the tumor

SEER registries

- Code to 20 in surgery codes per SEER manual

Stereotactic radiosurgery is not surgery

- Code in Radiation Treatment/Boost Modality

Brain Surgery Codes

Code	Description	Notes
00	None; no surgery of primary site; autopsy ONLY	
10	Tumor destruction, NOS (No specimen sent to path)	Code stereotactic radiosurgery (SRS), Gamma knife, Cyber knife, or Linac radiosurgery in RT fields
20	Local excision of tumor, lesion or mass; excisional bx	Excisional bx; SEER note: Includes stereotactic bx of brain tumor
21	Subtotal resection of tumor, lesion or mass in brain	Visible/known tumor left behind; “Debulking” (< full removal of tumor)
22	Resection of tumor of spinal cord or nerve	

Brain Surgery Codes

Code	Description	Notes
Codes 30 - 55 are not applicable for spinal cord or spinal nerve primary sites		
30	Radical, total, gross resection of tumor, lesion or mass in brain	Entire brain tumor resected – all macroscopic tumor removed; Total resection – all tumor removed to microscopic level; Resection of some normal brain tissue to ensure clean margins
40	Partial resection of lobe of brain	Use when the surgery cannot be coded as 20-30; < lobectomy, but wider margins than 30
55	Gross total resection of lobe of brain (lobectomy)	Used primarily for seizure disorders; uncommon for brain tumors
90	Surgery, NOS	
99	Unknown if surgery performed; DCO	

Chemo

- Intrathecal chemotherapy
  - Drugs directly injected into CSF via spinal injection or Ommaya reservoir
- Interstitial chemotherapy
  - Administered directly to involved tissue
  - Polymer wafers soaked in chemo agent inserted into tumor bed after resection
    - Gliadel wafers (carmustine/BCNU)

Chemo

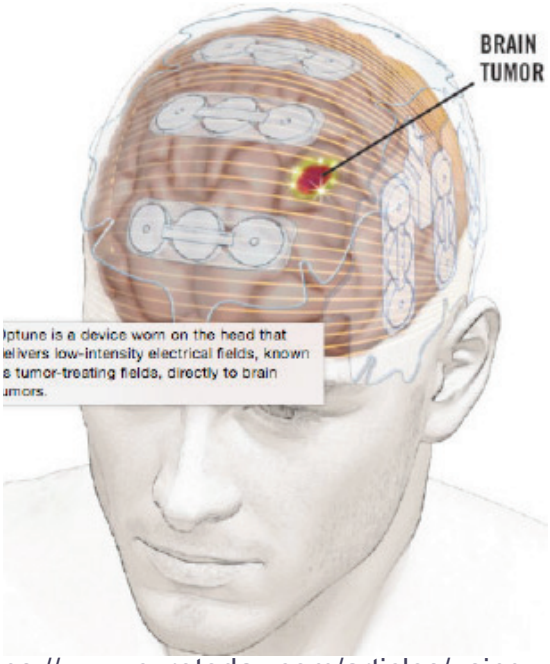
- GBM and high-grade gliomas
  - Temodar aka temozolomide
- Oligodendroglioma with 1p19q
  - Gleostine (lomustine), Matulane (procarbazine), Vincasar (vincistrine)

Targeted Therapy Brain Tumor

- Anti-angiogenesis therapy
  - Bevacizumab (Avastin, Mvasi)
  - Code as immunotherapy
- NTRK fusion genetic change
  - Larotrectinib (Vitrakvi)
  - Code as chemotherapy

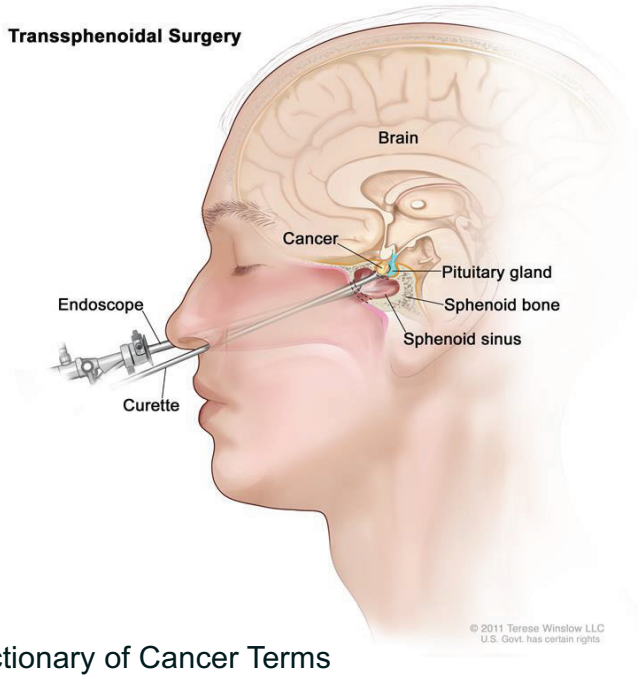
Other Treatment – Brain Tumor

- Optune: low-intensity alternating electric fields (Tumor Treating Fields/TTF)
- FDA-approved for GBM
  - New dx GBM? Surgery + Temodar + Optune
  - Recurrent GBM? Optune alone?
- Other Tx code “1”



https://www.curetoday.com/articles/using-cuttingedge-technology-to-treat-glioblastoma

Pituitary Surgery (Hypophysectomy)



Reminder: If hypophysectomy done for hormone manipulation, code it as “30, endocrine surgery” under Hematologic Transplant & Endocrine Procedures

Source: NCI Dictionary of Cancer Terms

Surgical Procedure Primary (C75.1-3 *see All Other Sites*)

- 30 Simple/partial removal primary site
- 40 Total surgical removal primary site
- 50 Surgery stated as "debulking"
- 60 Radical surgery
  - Partial or total removal WITH other organs
- 90 Surgery, NOS
- 99 Unk if surgery

Pituitary Tumor

- Prolactin producing
  - Parlodel (bromocriptine)
  - Dostinex (cabergoline)
  - Code as hormone
- Growth-hormone secreting
  - Sandostatin (octreotide)
    - Code as hormone ONLY IF physician states it is being prescribed to shrink tumor – could be used for side-effects of secreting adenomas
  - Somavert (pegvisomant)
  - Not listed in SEER RX