

2018 SEER Solid Tumor Manual

2018 KCR SPRING TRAINING

Eight Groups are Revised for 2018

Head & Neck – Still in DRAFT, presented earlier

Colon and Rectum ([2018 FINAL, included in this presentation](#))

Lung ([2018 FINAL included in this KCR 2018 Spring training update](#))

Breast ([2018 FINAL, included in this Spring training update](#))

Kidney (2018 FINAL, not included in this presentation)

Urinary Sites ([2018 FINAL included in this KCR 2018 Spring training update](#))

Non-malignant CNS ([2018 Draft included in this KCR 2018 Spring training update](#))

Malignant CNS and Peripheral Nerves ([2018 Draft included in this KCR 2018 Spring training update](#))

Solid Tumor Rules - 2018

The Multiple Primary rules are based primarily on these things:

Topography (anatomic site of tumor)

Histology

Behavior code

Number of tumors

Time between occurrence of first tumor and subsequent tumor(s)

2018 Changes to MP/H rules – Bottom Line

Lots of significant changes to coding histology

- **New codes, terms, and instructions** in the Histology update table. Many are site-specific.
- If in situ and invasive tumors are present at the same time, **code the invasive histology**
- If multiple histologies are mentioned, **code the most specific subtype of variant, even if it is not the majority of the tumor**. Use only modifiers such as type, subtype, or variant to code histology. Do not code histology based on terms such as features, differentiation, architecture, or any terms modified by ambiguous terms.

Using the Histology coding rules

Use the ICD-O-3 Histology Code and Behavior update for 2018, along with the ICD-O-3 publication by WHO, to accurately code histology.

The Priority of documents to be used for coding Histology vary by site:

- Use tissue/pathology for CNS tumors, benign or malignant, if available. For all other sites, use the most specific pathology from either the resection or the biopsy. Use the most specific histology from biomarkers first.
- Radiology reports are prioritized by site: Breast – use mammogram first; for all other sites, the order to use US, CT, or MRI may differ – use the site specific priority order listed for that site
- Clinical documents for diagnoses also differ in their priority by site. Use the site specific priority listed

Primary sites are now listed in each group of Solid Tumor rules.

Head and Neck now includes trachea (C33.9), and bones of the skull, face, and mandible (C41.0-C41.1), and external auditory canal (C44.2)

Colon now includes rectosigmoid junction and rectum (C19.9, C20.9)

Malignant CNS now includes peripheral nerves (C47.0-C47.9)

Very few changes to the Multiple Primary Rules themselves:

- New instructions regarding 'recurrence' of colorectal tumors at the anastomotic site. Previously, all of these were considered new primaries.
- Glioblastoma multiforme is now considered a new primary, if it occurs after an astrocytoma or glial tumor. Previously this was NOT a new primary, but progression of disease.
- Added a new rule for meningiomas, to disregard laterality. Previously, they were being over-counted.

Timing rule clarification

- Time interval between subsequent tumor means after a disease free interval – if a recurrence occurs within the time interval, then the 'clock starts over'
- No other timing rules changed

Solid Tumor Rules

What we will cover:

- New Lung Multiple Primary and Histology coding rules
- New Urinary system MP/H rules
- New MP/H rules for non-malignant CNS tumors
- New MP/H rules for malignant tumors of the CNS and Peripheral Nerves

Remember: The malignant and non-malignant CNS tumors are currently in draft form and may change slightly in the final version!

Lung: 2018 Solid Tumor Rules

Lung Solid Tumor Rules

Separate sections for:

Major Changes from the 2007 MPH rules

Equivalent Terms

Terms that are NOT Equivalent

Table 1: Coding Primary Site

Table 2: Combination/ Mixed Histology Codes

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

Illustrations

Multiple Primary Rules

Histology Coding Rules

Lung: Major Changes from 2007 rules

The WHO 4th Ed Tumors of the Lung uses a new classification of adenocarcinoma, resulting in significant changes to the 2004 WHO blue book for lung cancers. The term, bronchioloalveolar carcinoma (BAC) has been discontinued and the new preferred term is mucinous adenocarcinoma (8253).

New and changed histology codes are included in the Lung rules, Table 3.

- In situ and minimally invasive terms and codes have been added
- Some terms have been assigned a new histology code
- Some codes have been assigned a different preferred term

Many new histology codes and terms have been introduced in 2018. Careful review of the Lung Solid Tumor Rules, and Table 3, are essential for correct coding of 2018+ lung cancers.

Lung: Major Changes from 2007 rules

The following new terms and codes have been added for lung only:

- Mucinous carcinoma/adenocarcinoma 8253/3
 - **Microinvasive mucinous** carcinoma/adenocarcinoma 8257/3
 - **Minimally invasive** mucinous carcinoma/adenocarcinoma 8257/3
- Mucinous carcinoma/adenocarcinoma in situ 8253/2
- Non-mucinous carcinoma/adenocarcinoma 8140/3
 - Non-mucinous carcinoma/adenocarcinoma in situ 8250/2
 - **Lepidic adenocarcinoma**/ adenocarcinoma, lepidic predominant 8250/3
 - Non-mucinous carcinoma/adenocarcinoma, **minimally invasive** 8256/3

Lung: Equivalent and NOT Equivalent Terms

Equivalent terms:

- Adenocarcinoma, carcinoma
- Simultaneous, at the same time, concurrent, prior to first course treatment
- Squamous cell carcinoma, SCC, epidermoid carcinoma
- Type, subtype, variant

NOT Equivalent terms

- Bilateral does not always mean a multiple primary will be abstracted
- **Bronchus is not always mainstem bronchus** – when only bronchus is mentioned, code to the lobe in which the bronchial tumor is located
- Mucin-producing/mucin-secreting carcinoma (8481) is not the same as mucinous carcinoma (8253)
- Multilocular is not the same as multinodular
- **'Component' is not equivalent to subtype/variant**

Lung: Coding Primary Site

Terminology	Laterality	Topography and code
Apex, Pancoast tumor, superior lobe or bronchus, upper lobe	Either	Upper lobe C34.1
Lung base, lower lobe, lower lobe bronchus	Either	Lower lobe C34.3
Bronchus, NOS, bronchogenic, pulmonary, Lung NOS	Either	Lung, NOS C34.9
Bronchus intermedius, Carina, Hilus, perihilar	Either	Mainstem bronchus, C34.0
Lingula of lung	Left only	Upper lobe C34.1
Lobar bronchus	Either	Use designated lobe or C34.9, if unknown
Middle lobe, Middle lobe bronchi	Right only	Middle lobe C34.2
Overlapping lesion of lung	Either	Overlapping lesion C34.8

Lung: Combination/Mixed Histologies Table 2

Review the terms used in the diagnosis; if the terms match those listed in column 1, then use the combination code listed in column 2

Do NOT use Table 2 when:

- Tumors are both invasive and in situ; Use the invasive histology code
- One of the histologies is described as 'differentiation' or 'features'
- One of the histologies is an NOS term and another is a subtype/variant; Use the histology coding rules here instead of Table 2

Lung: Combination/Mixed histologies Examples from Table 2

Required ICD-O Terms	Combination Histology and Code
Adenocarcinoma, NOS AND Squamous cell carcinoma, NOS (cannot be any subtypes or variants of adeno or SCC)	Adenosquamous carcinoma 8560
Giant cell carcinoma AND Spindle cell carcinoma	Sarcomatoid carcinoma 8033
Epithelial carcinoma AND Myoepithelial carcinoma	Epithelial-myoepithelial carcinoma 8562

Lung: Histologies and Subtypes/Variants Example from Table 3

Histology Term and Code (may be specific term or NOS term)	Synonyms for Histology Term	Subtypes/ variants and Histology code
<i>Squamous cell carcinoma 8070</i>	Epidermoid carcinoma Squamous carcinoma Squamous cell epithelioma	Basaloid carcinoma/basaloid squamous cell carcinoma 8083 Keratinizing squamous cell carcinoma 8071 Non-keratinizing squamous cell carcinoma 8072

Lung – MP Rules

M1. Unknown if Single or Multiple Tumors – Abstract as Single Primary

M2. Single Tumor – A single tumor is always a Single Primary

Multiple Tumors –

- M3. Multiple primary if separate tumors are present in topography codes that differ at the second or third character (example: C34.0 and C33.9)
- M4. Multiple primaries if diagnosed more than 3 years apart
 - **NOTE: The time frame means clinically disease free for more than 3 years. If a patient has a recurrence within the 3 years, the 'clock' starts over, and the 3 year interval is computed from the date of last known recurrence. If recurrence is unknown, compute time from date of diagnosis.**
- M5. Multiple primaries when there is at least one tumor that is small cell carcinoma, or a small cell variant, and another tumor that is non-small cell carcinoma, or a non-small cell carcinoma variant.

Lung – MP Rules

Multiple Tumors – (cont.)

- M6. Multiple primaries if separate tumors are different subtypes in column 3 of Table 3. The tumors may be subtypes of the same or different NOS histologies.
- M7. Single primary if multiple tumors are in the same row in Table 3, **but they must be the same behavior.**
- M8. Multiple primaries if separate tumors are described in different rows of Table 3, first or second column.
- M9. Single primary when there are simultaneously:
 - A single tumor in one lung and multiple tumors in the other lung
 - Multiple tumors in both lungs
 - Multiple tumors in the same lung [Exception: this could be multiple primaries if pathology proves the tumors are of different histologies or the physician state unequivocally that the tumors are different primaries.]

Lung – MP Rules

Multiple Tumors – (cont.)

- M10. Single tumor when an in situ tumor is diagnosed after an invasive tumor in the same lung.
- M11. Multiple primary when there is a single tumor in each lung.
- M12. Single primary when an invasive tumor is diagnosed less than or equal to 60 days after in situ tumor in the same lung.
- M13. Multiple primaries when an invasive tumor occurs more than 60 days after an in situ tumor of the same lung
- M14. Single primary if none of the previous rules apply.

Lung – Histology Rules

Priority list for using documentation to code histology

1. Use the most specific tissue diagnosis; **may be from biopsy or resection**
 - **Biomarkers *change***
 - **The most specific is the subtype or variant term that may used for histology coding**
 - **Use tissue reports in this order: Addenda, Comments, Final diagnosis, CAP report**
2. Cytology report from primary site
3. Tissue or cytology from a metastatic site
4. Radiology – a) CT b) PET c) MRI d) Chest x-ray
5. Physician documentation

Lung – Histology Rules

Terminology to determine subtypes and variants:

DO NOT USE: NOS code, or specific histology or subtype/variant if described as:

DO USE:

Subtype

Type

Variant

Architecture

Differentiation

Features of

Foci, focus, focal

Majority – Use H instructions, most specific histology, even if not the majority of the tumor

Predominantly

Pattern

Any subtype or variant modified by an ambiguous term

Lung – Histology Rules

Single tumor –

H1. Code mucinous adenocarcinoma as follows:

- 8253/3 when behavior is invasive or unknown
- 8257/3 when microinvasive or minimally invasive
- 8253/2 when pre-invasive or in situ

H2. Code non-mucinous adenocarcinoma as follows:

- 8250/3 when invasive, lepidic or behavior is unknown
- 8256/3 when microinvasive or minimally invasive
- 8250/2 when pre-invasive or in situ

Lung – Histology Rules

Single tumor (cont.) –

- H3. Code the histology using Tables 3 when only 1 histologic type is identified
- H4. Code the invasive histology when both invasive and in situ elements are present
- H5. Code the subtype or variant when both a subtype and an NOS histology are identified
- H6. Code the combination code when the combination is listed in Table 2
- H7. Code adenocarcinoma with mixed subtypes (8255) ONLY when there is no combination code for the histologies within the tumor, or there are 2 or more subtypes of the same variant

Lung – Histology Rules

Multiple tumors –

- H8. Code mucinous adenocarcinoma as follows:
 - 8253/3 when behavior is invasive or unknown
 - 8257/3 when microinvasive or minimally invasive
 - 8253/2 when pre-invasive or in situ
- H9. Code non-mucinous adenocarcinoma as follows:
 - 8250/3 when invasive, lepidic or behavior is unknown
 - 8256/3 when microinvasive or minimally invasive
 - 8250/2 when pre-invasive or in situ

Lung – Histology Rules

Single tumor (cont.) –

H10. Code the histology using Tables 3 when only 1 histologic type is identified

H11. Code the invasive histology when both invasive and in situ elements are present

H12. Code the subtype or variant when both a subtype and an NOS histology are identified

H13. Code the combination code when the combination is listed in Table 2

H7. Code adenocarcinoma with mixed subtypes (8255) ONLY when there is no combination code for the histologies within the tumor, or there are 2 or more subtypes of the same variant

Urinary Sites: 2018 Solid Tumor Rules

Urinary Sites Solid Tumor Rules

Separate sections for:

Multifocal Tumors of Urinary Sites

Major Changes from 2007 MP/H rules

Equivalent Terms

Terms that are NOT Equivalent

Primary Site Coding for Urinary Organs and Urothelial Carcinoma

Table 1: Primary site/Topography codes

Table 2: Specific histologies, NOS Terms and Variants and Subtypes

Table 3. Non-reportable urinary tumors

Illustrations

Multiple Primary Rules

Histology Coding Rules

Urinary Sites Solid Tumor Rules

Introduction

- Multifocality in urothelial carcinomas is a common finding. The origin of the field effect concept has not been conclusively proven.

Major Changes from 2007 MP/H rules - No significant new histology terms or codes for 2018

Priority for coding Primary Site, when multiple sites are involved

- Non-invasive or in situ urothelial carcinoma or subtypes ONLY
 - Code Bladder, overlapping lesion (C67.8) when the bladder and one or both ureters ONLY are involved by a **single tumor**
 - Code Bladder, NOS (C67.9) when **multiple tumors** are present in the bladder or both bladder and ureter(s)
 - Code Urothelial system, NOS (C68.9) when **multiple organs** of the urinary system are involved
- Invasive urothelial carcinoma
 - Code Bladder, overlapping lesion (C67.8) when a **single tumor** overlaps sub-sites of the bladder
 - Code Bladder, NOS (C67.9) when **multiple tumors** are present in the bladder and the sub-site is unknown
 - Code Urothelial system, NOS (C68.9) when **multiple organs** of the urinary system are involved

Urinary Sites Solid Tumor Rules

Table 1 Lists the Topography codes with the preferred site description and synonyms

Table 2 Contains specific Histology and NOS terms, as well as Subtypes and Variants

Histology , Specific term or NOS term and Code	Synonyms for Histology Term	Subtypes/ variants and Histology code
Urothelial carcinoma 8120	Clear cell urothelial carcinoma Infiltrating urothelial carcinoma with divergent differentiation Infiltrating urothelial carcinoma with glandular differentiation Infiltrating urothelial carcinoma with squamous differentiation Infiltrating urothelial carcinoma with trophoblastic differentiation Lipid-rich urothelial carcinoma Nested urothelial carcinoma Plasmacytoid urothelial carcinoma	Giant cell urothelial carcinoma 8031/3 Lymphoepithelioma-like urothelial carcinoma 8082/3 Micropapillary urothelial carcinoma 8131/3 Papillary urothelial carcinoma in situ 8130/2 invasive 8130/3 Poorly differentiated carcinoma 8020/3 Sarcomatoid urothelial carcinoma 8122/3

Urinary Table 3. Neoplasms NOT Reportable

Histology	Synonyms
Benign perivascular epitheloid tumor 8714/0	Benign PEComa
Papillary urothelial neoplasm of low malignant potential 8130/1	
Paraganglioma 8693/1	Extra-adrenal pheochromocytoma
Urothelial dysplasia	
Urothelial papilloma 8120/0	

Urinary – MP Rules

M1. Unknown if Single or Multiple Tumors – Abstract as Single Primary

M2. Single Tumor – A single tumor is always a Single Primary

Multiple tumors

M3. Multiple primaries when there are tumors in both the left and the right renal pelvis and no other urinary sites are involved

M4. Multiple primaries when there are tumors in both the left and the right ureter and no other urinary sites are involved

M5. Multiple primaries when there are separate tumors with different subtypes or variants in Column 3 of Table 2

M6. Single primary when there are separate tumors are in the same row in Table 2

M7. Multiple primaries when there are multiple tumors with histology codes that are on different rows of Table 2

Urinary – MP Rules

Multiple tumors – (cont.)

M8. [Single primary when patient has a recurrence of in situ urothelial carcinoma](#)

M9. Single primary when a non-invasive urothelial tumors are diagnosed in the bladder and one or both ureters.

M10. [Single primary when an in situ tumor is diagnosed after an invasive in the same urinary sites.](#)

M11. Single tumor when an invasive tumor of the same histology occurs within 60 days of an in situ tumor in the same site

M12. Multiple primaries when an invasive tumor occurs more than 60 days after an in situ tumor of the same histology

M13. Single primary when there is an NOS histology and a subtype/variant of that histology

Urinary – MP Rules

M14. Single primary when a patient has multiple occurrences of invasive urothelial carcinoma of the bladder

M15. Multiple primaries if diagnosed more than 3 years apart

- **NOTE: The time frame means clinically disease free for more than 3 years. If a patient has a recurrence within the 3 years, the 'clock' starts over, and the 3 year interval is computed from the date of last known recurrence. If recurrence is unknown, compute time from date of diagnosis.**

M16. Single primary when multifocal urothelial carcinomas are diagnosed simultaneously in two or more of these sites:

- Renal pelvis
- Ureter
- Bladder
- Urethra/prostatic urethra

M17. Abstract as a single primary when tumors do not meet any of the criteria above

Urinary– Histology Rules

Priority list for using documentation to code histology

1. Use the most specific tissue diagnosis; **may be from biopsy or resection**
 - Biomarkers
 - **The most specific is the subtype or variant term that may used for histology coding**
 - **Use tissue reports in this order: Addenda, Comments, Final diagnosis, CAP report**
2. Cytology report from primary site
3. Tissue or cytology from a metastatic site
4. Physician documentation
5. Radiology – CT, MRI

Urinary – Histology Rules

Single tumor –

H1. Code the histology using Table 1 (or ICD-O-3) when only 1 histologic type is identified

H2. Code the invasive histology when in situ and invasive histologies occur in the same tumor

H3. Code the subtype/variant when an NOS and a subtype/variant of the NOS term is present

H4. Code mixed small cell carcinoma 8045 when a single tumor contains small cell neuroendocrine carcinoma, or one of its subtypes, and any other type of carcinoma

Urinary – Histology Rules

Single tumor –

H5. Code a mixture of urothelial carcinoma and another type of carcinoma as follows:

- **Adenocarcinoma and urothelial – code 8120** – adenocarcinoma is coded for a urinary site only when it is purely adenocarcinoma and not mixed with any other type
- **Squamous cell carcinoma – code 8120** – Squamous cell carcinoma is coded for a urinary site only when it is purely squamous cell carcinoma and not mixed with any other type
- Clear cell (8310), Endometrioid (8380), Sarcoma (8800), or any specific sarcoma – **code to the non-urothelial histology**

Urinary – Histology Rules

Multiple tumors –

H6. Code the histology using Table 2 (or ICD-O-3) when only 1 histologic type is identified

H7. Code the invasive histology when there are invasive and in situ tumors

H8. Code the subtype/variant when an NOS and a single subtype/variant of that NOS is present

H9. Code mixed small cell carcinoma 8045 when a the diagnosis is small cell neuroendocrine carcinoma, or one of its subtypes, and any other type of carcinoma

Malignant CNS and Peripheral Nerves: 2018 Solid Tumor Rules

Malignant CNS and Peripheral Nerves Solid Tumor Rules

Separate sections for:

Introduction

Changes from 2007 MP/H rules

Reportability Criteria

Section 1. Behavior Code includes

- Instructions for identifying and assigning behavior code
- Tables 1: WHO Grades for select CNS neoplasms

Section 2. Reportable Primary Sites and Histologies

- Reportable site terms
- Table 2: Reportable primary site terms and ICD-O site codes
- Table 3: Reportable specific and NOS histologies, synonyms, subtypes/variants, and ICD-O codes
- Table 4: Coding primary site for malignant tumors of cranial and peripheral nerves

Malignant CNS and Peripheral Nerves Solid Tumor Rules

Section 3. Additional information, including

- Table 5: Paired sites
- Table 6: Non-malignant CNS tumors with potential to transform to malignant behavior

Illustrations

Multiple Primary Rules

Histology Coding Rules

Malignant CNS and Peripheral Nerves

Introduction

Central nervous system (**CNS**) **includes** the following **primary sites**: Peripheral nerves, cerebral meninges; spinal meninges; meninges NOS; brain; spinal cord; cauda equina; olfactory nerve; optic nerve; acoustic nerve; cranial nerve NOS; overlapping lesion of brain and central nervous system; nervous system, nervous system NOS; pituitary gland, craniopharyngeal duct and pineal gland.

Pilocytic juvenile astrocytoma is reportable in the **US** as a **malignant** neoplasm 9421/3. It is reportable in **Canada** as **non-malignant** 9421/1.

Juvenile pilocytic astrocytoma was malignant in ICD-O-1 and ICD-O-2 with the histology and behavior 9421/3. The behavior code was changed to borderline (9421/1) with ICD-O-3.

Intraosseous meningiomas and meningiomas of the cavernous sinus and sphenoid wing are reportable.

Malignant CNS and Peripheral Nerves

Changes from the 2007 MPH rules

2016 CNS WHO presents major restructuring of the diffuse gliomas, medulloblastomas and other embryonal tumors, and incorporates new entities that are defined by both histology and molecular features, including glioblastoma, IDH-wildtype and glioblastoma, IDH-mutant; diffuse midline glioma, H3 K27M-mutant; RELA fusion-positive ependymoma; medulloblastoma, WNT-activated and medulloblastoma, SHH-activated; and embryonal tumor with multilayered rosettes, C19MC-altered.

Rule change: The 2007 rules said a glioblastoma multiforme (GBM) following an astrocytic or glial tumor was a single primary (recurrence).

- In the 2018 Solid Tumor rules, GBM subsequent to an astrocytic or glial tumor is a new (multiple) primary

Malignant CNS and Peripheral Nerves

Reportability criteria

Malignant CNS neoplasms must meet **three criteria/conditions** to be **reportable**:

1. The behavior must be reportable **AND**
 - a. Malignant, invasive, /3 **OR**
 - b. WHO Grade 3 or 4 (See Section 1, **Table 1**)
 - **Note 1: Always** code the behavior as designated 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.
2. The primary site must be reportable (See Section 2, **Table 2**) **AND**
3. The histology must be reportable (See Section 2, **Table 3**)

Malignant CNS and Peripheral Nerves

Section 1. Behavior code

Instructions for using source documentation to determine behavior are in priority order, with 1 having the highest priority.

Priority Order for Using Information to Assign Behavior

1. Pathology: Tissue from a resection
 - A. Use the pathologist's **description** of **malignant** behavior
 - B. Cases are reportable as malignant when pathology states a WHO Grade 3 or 4
 - C. **Never change behavior** described by pathologist
 - D. When there are **discrepancies** in behavior, use the documents in the following **priority** order:
 - i. **Addendum** or **comments** on pathology report , ii. **Final** diagnosis, iii. **CAP** report
2. Pathology: Tissue from a biopsy
3. Cytology (usually cerebrospinal fluid)
4. Physician's documentation
5. Scans: MRI, CT, PET, Angiogram
6. If none of the above are available, use Table 1. WHO Grade for select CNS neoplasms

Malignant CNS: Table 1. WHO Grade

1. WHO does not provide Grades for all CNS and peripheral nerve neoplasms.

2. WHO **Grade 3** and **4** neoplasms are always malignant

Table 1 contains histology terms and the WHO Grade based on molecular features, e.g.,

Histology	WHO grade
Anaplastic meningioma	3
Anaplastic astrocytoma, IDH-mutant	3
Angiocentric glioma	1
Atypical teratoid/rhabdoid tumor	4
Central neurocytoma	2
Choroid plexus carcinoma	3

Malignant CNS: Section 2. Reportable sites and Histologies

Major groups of reportable sites are:

Intracranial (within the skull/cranium)

- Cerebral meninges (C70.0)
- Brain (C71.0-C71.9)
- Cranial nerves (C72.0-C72.9)
- Intracranial glands (craniopharyngeal duct C75.2, pineal gland C75.3, pituitary gland C75.1)

Spinal sites (spinal meninges and sites within the spinal meninges)

- Spinal meninges C70.1
- Spinal nerve roots C47.0, C47.3, C72.0, C72.1)

Peripheral nerves (extracranial and extraspinal nerves) (C47.0-C47.9)

Malignant CNS: Section 2. Reportable sites and Histologies

Table 2 displays each of the reportable topographies from the previous slide with their terms and codes.

Table 3 includes reportable specific and NOS histologies, synonyms, and subtypes/variants, e.g.,

Specific or NOS Histology	Synonyms	Subtypes/Variants
Anaplastic ganglioma 9505		
Angiosarcoma 9120		
Astroblastoma 9430		
Astrocytoma, NOS 9400	Diffuse astrocytoma IDH – mutant Diffuse astrocytoma NOS Diffuse astrocytoma IDH – wild type	Anaplastic astrocytoma IDH – mutant 9401 Gemistocytic astrocytoma IDH – mutant 9411 Anaplastic astrocytoma IDH –wild type 9401

Malignant CNS: Section 2. Reportable sites and Histologies

Table 4 displays each of the reportable topographies for cranial and peripheral nerves and their topography codes.

Name and CN#	Exits cranium through	Site code – cranial nerve	Site code – peripheral nerve
Olfactory CN1	Cribriform plate	Surface of the brain C72.2	Originates in mucosa of nasal cavity, travels to ethmoid bone C47.0
Optic CN2	Optic canal	Intradural, all portions are reportable C72.3	
Oculomotor CN3	Superior orbital fissure	Originates in the midbrain C72.5	After exiting fissure, enters the orbit C47.0

Malignant CNS: Section 3. Additional instructions

Table 5 identifies paired sites for which laterality must be coded.

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: Midline tumors are limited to glioblastoma multiforme, meningiomas, lymphomas, and epidermoid cysts.

Table 6. Non-malignant tumors that may transform to malignant (new primary)

Original histology	Transformed histology
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 – MP Rules

M1. Unknown if Single or Multiple Tumors – Abstract as Single Primary

M2. Single Tumor – A single tumor is always a Single Primary

M3. Single primary when original diagnosis is oligodendroglioma and subsequently recurs with different features (path may state 'looks like GBM')

M4. Single primary when a single tumor is diagnosed clinically or radiographically or by stereotactic biopsy as non-malignant and no resection is performed as first course treatment; then a resection is done when symptoms appear and the pathology from the resection is malignant. Change the behavior code on the original abstract to /3. There is no time limit for this rule. This is another example of a single tumor is always a single primary.

Malignant CNS – MP Rules

Multiple tumors

M5. Multiple primaries when there is an invasive tumor and a non-malignant tumor

M6. Multiple primaries when there are tumors in both

- Brain and any other part of the CNS
- Cerebral meninges and spinal meninges
- Cranial nerves and any other part of the CNS
- Meninges of cranial or peripheral nerves and any other part of the CNS
- Peripheral nerves and any other part of the CNS

M7. Multiple primaries when there are multiple tumors with histology codes that are on different rows of Table 3, first, second or third column

M8. Multiple primaries when there are separate tumors with different subtypes or variants in Column 3 of Table 3

Malignant CNS – MP Rules

Multiple Tumors – (cont.)

- M9. Single primary when there are separate tumors in
 - Same lobe
 - Different lateralities
 - Different lobes

Multiple site involvement implies metastases or multicentric disease in these histologies: glioblastoma multiforme, gliomatosis, PNET medulloblastoma, ependymoma, oligodendroglioma, and hereditary syndromes (NF1, NF2)

- M10. Multiple primaries when there are multiple tumors with histology codes that differ at the first, second, or third digit
- M11. Single primary if none of the previous rules apply.

Malignant CNS– Histology Rules

Priority list for using documentation to code histology

1. Use the pathology from a resection. **Use tissue reports in this order: Addenda, Comments, Final diagnosis, CAP report**

Use the pathology from a biopsy. **Use tissue reports in this order: Addenda, Comments, Final diagnosis, CAP report**

2. Cytology report from primary site
3. Tissue or cytology from a metastatic site
4. Radiology – MRI, CT, PET
5. Physician documentation

Malignant CNS – Histology Rules

Terminology to determine subtypes and variants:

DO NOT USE: NOS code, or specific histology or subtype/variant if described as:

DO USE:

Subtype

Type

Variant

Architecture

Differentiation

Features of

Foci, focus, focal

Majority

Predominantly

Pattern

Any subtype or variant modified by an ambiguous term

Malignant CNS – Histology Rules

Single tumor –

H1. Code the histology using Tables 3 when only 1 histologic type is identified

H2. Code the subtype or variant when both a subtype and an NOS histology are identified

H3. Code the histology with the numerically higher code

Multiple tumors

H4. Code the histology using Tables 3 when only 1 histologic type is identified

H5. Code the subtype or variant when both a subtype and an NOS histology are identified

H6. Code the histology with the numerically higher code

Non-Malignant CNS Neoplasms: 2018 Solid Tumor Rules

Non-Malignant CNS Solid Tumor Rules

Separate sections for:

Introduction

Reportability Criteria

Section 1. Behavior Code includes

- Instructions for identifying and assigning behavior code
- Tables 1: WHO Grades for select CNS neoplasms

Section 2. Reportable Primary Sites and Histologies

- Reportable primary site terms and definitions
- Priorities for coding primary site
- Table 2: Reportable primary sites
- Table 3: Reportability of non-malignant cranial nerve (CN) tumors
- Table 4: Non-reportable neoplasms
- Table 5: Histologic terms of non-malignant intracranial tumors

Non-Malignant CNS Solid Tumor Rules

Section 3. Additional information, including

- Conflicting information on path report
- Table 6: Specific histologies, NOS terms and synonyms, and subtypes/variants
- Table 7: Paired sites
- Table 8: Non-malignant CNS tumors with potential to transform to malignant behavior

Illustrations

Multiple Primary Rules

Histology Coding Rules

Non-Malignant CNS

Section 1. Behavior code

Instructions for using source documentation to determine behavior are in priority order, with 1 having the highest priority.

Priority Order for Using Information to Assign Behavior

1. Pathology: Tissue from a biopsy or resection
 - A. Use the tissue report with the **highest grade** (biopsy or resection)
 - B. Use the pathologist's **description of borderline or benign** behavior
 - C. Cases may be reportable when pathology states a WHO Grade 1
 - D. **Never change behavior** described by pathologist
 - E. When there are **discrepancies** in behavior, use the documents in the following **priority** order: i. **Addendum or comments** on pathology report, ii. **Final diagnosis**, iii. **CAP report**
2. Cytology (usually cerebrospinal fluid)
3. Physician's documentation
4. Scans: MRI, CT, PET, Angiogram
5. If none of the above are available, use Table 1. WHO Grade for select CNS neoplasms

Non-Malignant CNS: Table 1. WHO Grade

1. Use non-malignant CNS rules for all WHO Grade 1 (always non-malignant)
2. WHO **Grade 3** and **4** neoplasms are always malignant; go to the malignant CNS rules

Table 1 contains histology terms and the WHO Grade based on molecular features, e.g.,

Histology	WHO grade
Anaplastic meningioma	3
Anaplastic astrocytoma, IDH-mutant	3
Angiocentric glioma	1
Atypical teratoid/rhabdoid tumor	4
Central neurocytoma	2
Choroid plexus carcinoma	3

Non-Malignant CNS: Table 2. Reportable primary sites

C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753

Note 1: Sphenoid wing meningiomas are reportable – code C70.0

Note 2: Cavernous sinus neoplasms are reportable; they arise in either the meninges or the cranial nerves (there is no site code for the cavernous sinus).

Major groups of reportable sites are:

Intracranial (within the skull/cranium)

- Cerebral meninges (C70.0)
- Brain (C71.0-C71.9)
- Cranial nerves (C72.0-C72.9)
- Intracranial glands (craniopharyngeal duct C75.2, pineal gland C75.3, pituitary gland C75.1)

Spinal sites (spinal meninges and sites within the spinal meninges)

- Spinal meninges (C70.1, C70.9)
- Spinal cord, Cauda equina (C72.0, C72.1)

Non-Malignant CNS: Table 2. Reportable primary sites

Olfactory nerve (C72.2)

Optic nerve (C72.3)

Acoustic nerve (C72.4)

Cranial nerves (C72.5)

Other parts of the CNS

Nervous system, NOS (C72.9)

Overlapping lesion of brain and CNS (C72.8)

Non-Malignant CNS: Table 3: Reportable sites and Histologies

Table 3 displays each of the reportable topographies for non-malignant cranial nerves

Name and CN#	Exits cranium through	Reportable portion – cranial nerve	Non-reportable portion – cranial nerve
Olfactory CN1	Cribriform plate	Surface of the brain C72.2	Originates in olfactory mucosa of nasal cavity, travels to ethmoid bone C47.0
Optic CN2	Optic canal	Intradural, all portions are reportable C72.3	
Oculomotor CN3	Superior orbital fissure	Originates in the dorsal brain stem C72.5	After exiting fissure, enters the orbit C47.0

Non-Malignant CNS: Table 4: Non-reportable neoplasms

Non-reportable histology	Histology code	Definition and sites
Carcinomas	8010-8060, 8071-8671, 8940-8941	Brain C71.0-C71.9 Site/histology edit carcinoma/brain
Glomus tympanicum, Glomus jugulare	8690/1	Occur in inner ear, aortic body, or other paraganglia; non-reportable sites
Hypothalamic hamartoma	No code	Occurs in hypothalamus
Neurofibromatosis, type 1 (NF1)	No code	Genetic disease that produces non—malignant tumors in many sites. The brain and CNS tumors spawned by NF1 are reportable but the genetic disease is not.

Non-Malignant CNS: Table 5. Histologic types

1. Use Table 5 to code primary site
2. Sites listed are the most common sites for that specific histology

Histology and code	Most common site
Angiocentric glioma 9431/1	Cerebrum C71.0
Choroid plexus papilloma 9390/0	Intraventricular site (C71.3, C71.5, C71.7)
Craniopharyngioma 9350/1	Pituitary gland 75.1
Dermoid cyst 9084/0	Pineal gland C75.3 or suprasellar C75.2
Pineocytoma 9361/1	Pineal gland C75.3
Subependymoma 9383/1	Lateral ventricle C71.5

Non-Malignant CNS: Table 6. Specific Histologies, Synonyms, Subtypes/Variants

Table 6 includes reportable specific and NOS histologies, synonyms, and subtypes/variants, e.g.,

Specific or NOS Histology	Synonyms	Subtypes/Variants
Angiocentric glioma 9431/1	Angiocentric neuroepithelial tumor Monomorphous angiocentric glioma	
Central neurocytoma 9506/1	Cerebellar liponeuroneurocytoma Extraventricular neurocytoma	
Chondroma 9220/0		
Choroid plexus papilloma 9390/0		Atypical choroid plexus papilloma 9390/1

Non-Malignant CNS: Table 7. Paired sites

Table 7 identifies paired sites for which laterality must be coded.

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: Cerebellum C71.6 – laterality may be coded but is not required

Table 8. Non-malignant tumors that may transform to malignant (new primary)

Original histology	Transformed histology
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

Non-Malignant CNS – MP Rules

M1. Unknown if Single or Multiple Tumors – Abstract as Single Primary

M2. Single Tumor – A single tumor is always a Single Primary

M3. Single primary when a single tumor is diagnosed clinically or radiographically or by stereotactic biopsy as non-malignant and no resection is performed as first course treatment. Then a resection is done when symptoms appear and the pathology from the resection is malignant. Change the behavior code on the original abstract to /3.

There is no time limit for this rule. This is another example of a single tumor is always a single primary.

M4. Single primary when a benign tumor transforms to a borderline (/1) tumor with the same histology as the original tumor, or a subtype of variant of that histology.

Non-Malignant CNS – MP Rules

Multiple tumors

M5. Multiple primaries when a non-malignant tumor transforms into a malignant tumor AND the patient **had a resection, or it is unknown if a resection occurred.**

M6. Single primary when the patient has bilateral acoustic neuromas, vestibular schwannomas or optic nerve gliomas.

M7. Multiple primaries when separate tumors are described in different rows of Table 6, first or second column.

M8. Multiple primaries when separate tumors are described as different subtypes in Column 3 of Table 6.

M9. Single primary when an NOS tumor and a subtype/variant of the NOS is in the same CNS site code (same second, third and fourth character). See Table 6.

Non-Malignant CNS – MP Rules

Multiple tumors (con't.)

M10. Single primary when two or more separate meningiomas arise in the cranial meninges (laterality is irrelevant).

M11. Multiple primaries when there are tumors in both

- Brain and any other part of the CNS
- Cerebral meninges and spinal meninges
- Cranial nerves and any other part of the CNS
- Meninges of cranial or peripheral nerves and any other part of the CNS

M12. Single primary when there are multiple tumors in the brain

M13. Multiple primaries when there are multiple tumors with histology codes that are different at the first, second or third digit

M14. Single primary when tumors do not meet any of the above criteria

Non-Malignant CNS– Histology Rules

Priority list for using documentation to code histology

1. Use the pathology from a resection. **Use tissue reports in this order: Addenda, Comments, Final diagnosis, CAP report**

Use the pathology from a biopsy. **Use tissue reports in this order: Addenda, Comments, Final diagnosis, CAP report**

2. Cytology report from primary site
3. Tissue or cytology from a metastatic site
4. Radiology – MRI, CT, PET
5. Physician documentation

Non-Malignant CNS – Histology Rules

Terminology to determine subtypes and variants:

DO NOT USE: NOS code, or specific histology or subtype/variant if described as:

DO USE:

Subtype

Type

Variant

Architecture

Differentiation

Features of

Foci, focus, focal

Majority

Predominantly

Pattern

Any subtype or variant modified by an ambiguous term

Non-Malignant CNS – Histology Rules

Single tumor –

H1. Code meningioma (9530/0) when the diagnosis is Meningioma, NOS, Metaplastic meningioma, Microcystic meningioma, Secretory meningioma, or Lymphoplasmacyte-rich meningioma

H2. Code the reportable CNS tumor if the patient has a tumor along with NF1, NF2, or schwannomatosis (e.g., NF1: plexiform neurofibroma). Do not code the NF1 or NF2; only the tumors that result from the neurofibromatosis.

H3. Code the histology when only 1 histologic type is identified

H4. Code the subtype or variant when both a subtype and an NOS histology are identified

H5. Code the histology with the numerically higher code

Non-Malignant CNS – Histology Rules

Multiple tumors –

H6. Code meningioma (9530/1) when there are single or multiple Meningiomas of uncertain behavior

H7. Code meningioma (9530/0) when the diagnosis is Meningioma, NOS, Metaplastic meningioma, Microcystic meningioma, Secretory meningioma, or Lymphoplasmacyte-rich meningioma

H8. Code the reportable CNS tumor if the patient has a tumor along with NF1, NF2, or schwannomatosis (e.g., NF1: plexiform neurofibroma). Do not code the NF1 or NF2; only the tumors that result from the neurofibromatosis.

H9. Code the histology when only 1 histologic type is identified

H10. Code the subtype or variant when both a subtype and an NOS histology are identified

H11. Code the histology with the numerically higher code