SEER*Educate Workshop Exercises

Brain & Other Sites Histology Coding

12/2023: Carolyn Callaghan, BS, CTR

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CEs for the 2023 SEER*Educate Workshop Cases

The 2023 SEER Workshop exercises are in three modules.

1. NCRA 2023-196, 5 LAMN/HAMN coding exercises, 2.5 CEs
2. NCRA 2023-194, Brain (site/histo/behavior) coding exercises, 2.5 CEs
3. NCRA 2023-195, Other Sites (site/histo/behavior) coding exercises, 3.0 CEs

To earn the CE for each module, you must score a minimum of 70% on each case. No partial credit is given for these modules. We recognize these are challenging cases and most people need to retake one or more cases.

If time permits, we suggest that you wait at least a day before retrying any cases so that you can better assess whether the information in the rationale has helped you to use the available coding guidelines to arrive at the preferred answer.

To view the CE report for this series, go to Reports, View Excel Reports, CE Reports – Current Years. The last report in that section is 2023 SEER Workshop - CE Hours Earned.

Change the start date to 8/1/2023 to ensure that any early results are reflected in the report. If you have completed a case multiple times, the report should reflect your best results. If you have scored less than 70% on a case, it should reflect that as well so you can identify which cases you need to retake.

You must successfully complete the cases by May 30, 2024 to earn the CEs for these cases. The CEs for the cases are separate from the CEs for this presentation.

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Acknowledgements and Disclaimers

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- Acknowledge Seattle registry staff for beta testing all new and changed material in SEER*Educate
- The opinions and views expressed in this presentation are solely the author's and do not reflect the view of the National Cancer Institute (NCI).

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Brain & Other Sites Histology Coding

General Solid Tumor Rules Issues

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Brain & Other Sites Histologies

- Brain histology has become more complicated over time as biomarkers are becoming a critical histology component for many of these tumors.
- Priority Order for Using Documentation to Identify Histology must be used to determine which source should be used.
  - The Priority Order (Malignant CNS and Peripheral Nerves Histology Rules) provides a hierarchical list of source documentation used to code histology. What has the highest priority for both pathology/tissue from resection AND pathology/tissue from biopsy?
  - **Biomarkers!**
    - "Note 1: Biomarkers are emerging as an important part of cancer diagnosis and treatment. Some biomarkers are used to determine treatment rather than histologic type. The efficacy of identification of histologic type using biomarkers differs from primary site to primary site. When a histologic type is identified using a biomarker, code the identified histology. Biomarkers do not identify all histologic types."

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Brain & Other Sites Histologies

Reminders for coding Brain histology:

- Use the H Rules to code the histology. There may be a specific H Rule applicable to the case at hand.
  - Do not just jump to the ICD-O-3.2.
- When the applicable H Rule doesn't provide the specific histology for the case at hand, remember there is a **priority order** for histology coding sources!
  - #1: Use the site/schema-specific table to code histology. For example, Table 3 for Malignant CNS. The H Rules remind you, "Use Table 3 to code histology. New codes, terms, and synonyms are included in Table 3 and coding errors may occur if the table is not used."
  - #2: Use the ICD-O-3.2 and all updates ***WHEN*** the histology is not listed in the site/schema-specific table (i.e., the histology is not included in Table 3). The ICD-O-3.2 is not the first priority!
  - #3: SINQ or Ask a SEER Registrar is the last resort when the histology code is not found in the site/schema-specific table or the ICD-O-3.2 (including all updates).

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Brain & Other Sites Histologies

- Other Sites histology have new rules. These were updated in the 2023 Solid Tumor Rules revision. We no longer follow the 2007 MPH General Rules for these sites.
  - All Solid Tumor Rules sites now follow the same set of general rules, including the ambiguous terminology rule (Rule 3A-B). Ensure you are not coding a more specific histology described by ambiguous terminology UNLESS the case meets the Solid Tumor Rules criteria!
- Use the Priority Order list for Other Sites to determine which source to code histology from (i.e., addendum dx, synoptic report, etc.).
- Nineteen site-specific histology tables were added to Other Sites. These site-specific tables must be used to appropriately code histology for the case at hand.
  - The Site or Site Group Histology-Specific Table instructions in the Other Sites Equivalent Terms and Definitions confirm, "In place of adding numerous site-based histology rules to the 2023 revision, the histology tables will include additional coding instructions and notes to assign the correct ICD-O code when appropriate. Follow the H rules and refer to the tables if directed."

IMPORTANT: It is important to refer to these tables when determining a histology code as the notes may provide coding guidance.

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Brain & Other Sites Histologies

Reminders for coding Other Sites histology (cont.):

- Use the H Rules to code the histology. There may be a specific H Rule applicable to the case at hand.
  - Do not just jump to the ICD-O-3.2.
  - Remember to review the Notes for the H Rules, the Notes may help with histology coding decisions.
- When the applicable H Rule doesn't provide the specific histology for the case at hand, we should follow the **priority order** we discussed for Brain!
  - #1: Use the site/schema-specific table to code histology. Many of the H Rules for Other Sites refer one to the applicable Tables; the rules will tell you to see or use Tables 3-21.
  - #2: Use the ICD-O-3.2 and all updates ***WHEN*** the histology is not listed in the site/schema-specific table (i.e., the histology is not included in Tables 3-21). The ICD-O-3.2 is not the first priority because there are site-specific histologies that may apply to some Other Sites, but not others.
  - #3: SINQ or Ask a SEER Registrar is the last resort when the histology code is not found in the site/schema-specific table or the ICD-O-3.2 (including all updates).

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Brain & Other Sites Histologies

Reminders for coding Other Sites histology (cont.):

- Let's look at some reasons why the H Rule Notes and Tables are critical for histology coding:

Single tumor: Invasive and In Situ Components

Rule H8 Code the **invasive histology** when both invasive and in situ components are present.
Note 1: Use Tables 3-21, ICD-O, and all ICD-O updates to determine if the term containing both invasive and in situ histologies has a specific ICD-O code.
Example: Intraductal papillary mucinous neoplasm with associated carcinoma has both in situ (intraductal) and associated invasive carcinoma and has an ICD-O code of 8453/3
Note 2: When the term is not listed in Tables 3-21, ICD-O, and ICD-O updates, ignore the in situ term.

Rule H12 Code the histology when only **one histologic type** is identified.
Note 1: Do not code terms that do not appear in the histology description.
Example: Do not code squamous cell carcinoma non-keratinizing unless the words "non-keratinizing" actually appear in the diagnosis.
Note 2: Some histologies are compound terms meaning two or more histology types are combined into a single ICD-O code. Use Tables 3-21, ICD-O, and all ICD-O updates to determine if the term containing multiple histologies has a specific code.
Example: Myxoid pleomorphic liposarcoma has more than one histology listed in the term and is coded 8854/3 per ICD-O-3.2
Note 3: If histology is papillary carcinoma of thyroid, continue through the rules.

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SEER*Educate Brain Histology Exercises

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Brain Histology – Cases/Issues Not Discussed

Type of Issues	Case #	Accuracy Rate	Helpful Hints
Primary Site	01	39%	**Primary Site should be coded to the site the tumor arose in.** • Code the specific brain/CNS subsite the tumor arose in. • Use default or NOS sites like brain, NOS (C719) when the primary tumor arose in a structure included in site C719 per the ICD-O-3 (e.g., posterior fossa = C719). • Use the ICD-O-3 (hardcopy manual) for the primary site list PLUS the primary site instructions in the SEER and Solid Tumor Rules Manuals to help code primary site (e.g., Table 2 for Malignant CNS).
	07	82.6%	
	09	88.2%	
	12	89.1%	
	14	88.2%	
Behavior	01	89.3%	• **All** WHO Grade 3 and 4 tumors are malignant /3. • WHO Grade I tumors are **always** non-malignant, use the schema-specific histology tables and/or ICD-O-3.2 to determine whether /0 or /1 applies. • WHO Grade 2 tumors can be non-malignant or malignant, use the default behavior per the schema-specific histology tables and/or ICD-O-3.2. • Use the default behavior provided by the Solid Tumor Rules (schema-specific histology tables), the ICD-O-3.2, or the SINQ/AASR response when no other information is available.
	14	88.2%	
2023 Histology Update	04	88.1%	High grade astrocytoma with piloid features (HGAP) is a new malignant histology for 2023.
Histology	12	88.2%	Specific clinical diagnoses can be coded in the absence of a histologic diagnosis; use the site-specific histology table to help identify histology.

CASES CUT FOR TIME!

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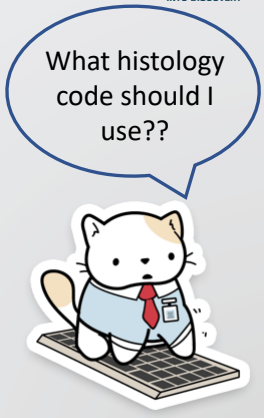
Brain Histology – Cases/Issues Discussed

General Types of Issues	Case #	Frequency (Preferred Histology)
Integrated Diagnoses	02	76.9%
	03	78.1%
	06	79.3%
	13	28.6%
NOS & More Specific Biomarker Histologies	01	43.9%
	05	86.4%
	08	85.8%
SINQ Only Histologies	11	50.1%
	15	56.7%
Ambiguous Terminology	09	72.3%
	14	82.5%



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Brain Histology – Integrated Diagnosis Scenarios

- Brain 02: Final Diagnosis = Glial/Glioneuronal neoplasm. Integrated Diagnosis = **Diffuse low-grade glioma, MAPK pathway altered = 9421/1.**
- Brain 03: Final Diagnosis = Diffuse glioma with focal high-grade features. Integrated Diagnosis = **Oligodendroglioma, IDH-mutant and 1p/19q-codeleted, CNS WHO grade 3 = 9451/3.**
- Brain 06: Integrated Diagnosis = **Astrocytoma, IDH-mutant, CNS WHO grade 4 = 9445/3.**
- Brain 13: Final Diagnosis = High grade neuroepithelial tumor. Integrated Diagnosis = **CNS tumor with BCOR-ITD = 9500/3.**

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Brain Histology – Integrated Diagnosis Scenarios

Case	Final Diagnosis	Integrated Diagnosis	Resulting Morphology
02	~~Glial/Glioneuronal Neoplasm = 9413/0 (per SINQ 20190105)~~	(Included in Addendum Dx) Diffuse low-grade glioma, MAPK pathway altered = 9421/1	Diffuse low-grade glioma, MAPK pathway altered **9421/1**
03	~~Diffuse glioma with focal high-grade features = 9380/3~~	Oligodendroglioma, IDH-mutant and 1p/19q-codeleted, CNS WHO grade 3 = 9451/3	Oligodendroglioma, IDH-mutant and 1p/19q-codeleted, CNS WHO grade 3 **9451/3**
06	~~N/A~~	Astrocytoma, IDH-mutant, CNS WHO grade 4 = 9445/3	Astrocytoma, IDH-mutant, CNS WHO grade 4 **9445/3**
13	~~High grade neuroepithelial tumor = 8000/3 (Neuroepithelial tumor, malignant)~~	CNS tumor with BCOR-ITD = 9500/3	CNS tumor with BCOR-ITD **9500/3**

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Brain Histology – Integrated Diagnosis Scenarios

- Cases 03 & 06: Why don't NOS histologies apply?
  - Integrated Diagnoses: Oligodendroglioma, IDH-mutant and 1p/19q-codeleted, CNS WHO grade 3 and Astrocytoma, IDH-mutant, CNS WHO grade 4 (AKA: Astrocytoma, IDH-mutant, grade 4). Table 3 confirms:

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Oligodendroglioma NOS **9450** ***Note:*** Oligodendroglioma NOS is used when molecular markers cannot fully be determined	Oligodendroglioma 1p/19q-codeleted Oligodendroglioma IDH-mutant Oligodendroglioma IDH-mutant and 1p/19q-codeleted, grade 2	Anaplastic oligodendroglioma NOS **9451** IDH-mutant 1p/19q-codeleted IDH-mutant and 1p/19q-codeleted Oligodendroglioma, IDH-mutant and 1p/19q-codeleted, grade 3
Astrocytoma NOS **9400**	Astrocytoma, IDH-mutant, grade 2 Diffuse astrocytoma IDH-mutant Diffuse astrocytoma IDH-wildtype Diffuse astrocytoma NOS	Anaplastic astrocytoma IDH-mutant/wildtype; anaplastic astrocytoma NOS **9401** Astrocytoma, IDH-mutant, grade 3 **9401** Astrocytoma, IDH-mutant, grade 4 **9445** Gemistocytic astrocytoma IDH-mutant **9411** Pleomorphic xanthroastrocytoma /anaplastic pleomorphic xanthroastrocytoma **9424**

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Brain Histology – Integrated Diagnosis Scenarios

- Case 13: **Morphology code is 9500/3 – CNS tumor with BCOR-ITD.**
- There are a few issues related to this case complicating histology coding:
  1. CNS tumor with BCOR-ITD (BCOR internal tandem duplication) is a new synonym or related term for morphology 9500/3 for cases diagnosed 1/1/2023 and later.
  2. SINQ 20180102 does not apply to a 2023 diagnosis of CNS tumor with BCOR-ITD (or a high grade neuroepithelial tumor with BCOR-ITD). While SINQ instructs one to code this as 8005/3, this SINQ only applies to diagnosis years 2018-2022.
  3. There is a typo in the 2023 ICD-O-3.2 Implementation Guidelines, but not in the 2023 Solid Tumor Rule update. The ICD-O-3.2 states, "CNS tumor with ***BCCR*** internal tandem duplication," but the Solid Tumor Rules has the correct term and states, "CNS tumor with ***BCOR*** internal tandem duplication."

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Brain Histology – NOS & Biomarker Scenarios

- Brain 01: Final Diagnosis = Posterior fossa ependymoma. Addendum Diagnosis (based on biomarkers, DNA Methylation Microarray Analysis) = **Ependymoma, posterior fossa group A = 9396/3.**
- Brain 05: Final Diagnosis = Supratentorial Ependymoma. Addendum Diagnosis (based on biomarkers) = **Supratentorial ependymoma, YAP1 fusion-positive = 9396/3.**
- Brain 08: Final Diagnosis = Diffuse astrocytic glioma, at least CNS WHO Grade 3. Addendum Diagnosis #2 (based on two sets of biomarkers) = **Glioblastoma, IDH wild-type, CNS WHO Grade 4 = 9440/3.**

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Brain Histology – NOS & Biomarker Scenarios

Case	Final Diagnosis	Specific Biomarker Diagnosis	Resulting Morphology
01	~~Posterior fossa ependymoma = 9391/3~~	(Included in Addendum Dx) Ependymoma, posterior fossa group A = 9396/3	Ependymoma, posterior fossa group A **9396/3**
05	~~Supratentorial Ependymoma = 9391/3~~	(Included in Addendum Dx) Supratentorial ependymoma, YAP1 fusion-positive = 9396/3	Supratentorial ependymoma, YAP1 fusion-positive **9396/3**
08	~~Diffuse astrocytic glioma, at least CNS WHO Grade 3 = 9400/3 (Astrocytic glioma per ICD-O-3.2)~~	(Included in Addendum Dxs) Glioblastoma, IDH wild-type, CNS WHO Grade 4 = 9440/3	Glioblastoma, IDH wild-type, CNS WHO Grade 4 **9440/3**

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Brain Histology – NOS & Biomarker Scenarios

- Case 01: **Morphology code is 9396/3 – Ependymoma, posterior fossa group A.**
  - The Addendum Dx states, "Ependymoma, posterior fossa group A by DNA Methylation Microarray Analysis (see interpretation)." Table 3 confirms:

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Ependymoma **9391** *Note:* The following terms are synonyms of ependymoma, RELA fusion-positive 9396, and are NOT subtypes/variants of it. They are all coded 9396. • Posterior fossa group A (PFA) ependymoma • Posterior fossa group B (PFB) ependymoma • Spinal ependymoma, MYCN-amplified • Supratentorial ependymoma, YAP1 fusion-positive • Supratentorial ependymoma, ZFTA fusion-positive	Clear cell ependymoma Posterior fossa ependymoma, NOS Spinal ependymoma, NOS Supratentorial ependymoma, NOS Tanycytic ependymoma	Anaplastic ependymoma **9392** Ependymoma, RELA fusion-positive **9396*** Posterior fossa group A (PFA) ependymoma Posterior fossa group B (PFB) ependymoma Spinal ependymoma, MYCN-amplified Supratentorial ependymoma, YAP1 fusion-positive Supratentorial ependymoma, ZFTA fusion-positive Papillary ependymoma **9393**

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Brain Histology – SINQ Only Scenarios

- Brain 11: Final Diagnosis = Glioneuronal neoplasm, favor low grade. Addendum Diagnosis (is the Integrated Diagnosis) = Neuroepithelial tumor with PATZ1 fusion, not elsewhere classified (NEC).
  - Documented NGS findings in the Integrated Diagnosis and the Addendum Comment confirm, "NGS: Positive for an EWSR1-PATZ1 fusion."
  - The Addendum Comment provides the same diagnosis, "The combined findings support a diagnosis of neuroepithelial tumor with PATZ1 fusion."
  - **SINQ 20220006 = 8000/1 (Neuroepithelial tumor with PATZ1 fusion)**
- Brain 15: Final Diagnosis = Papillary neoplasm. Diagnosis Comment = "The resulting diagnosis is a primary papillary epithelial tumor of the sella (PPETS)."
  - Subsequent 05/28/2023 Neurosurgery note confirms, "Rsxn path proved PPETS."
  - **SINQ 20220037 = 8000/0 (Primary papillary epithelial tumor of sella, PPETS)**

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Brain Histology – SINQ Only Scenarios

Case	Histology Term	User Selected Morphologies	SINQ Morphology
11	Neuroepithelial tumor with PATZ1 fusion, not elsewhere classified (NEC) (NGS: EWSR1-PATZ1 fusion)	9413/0: Low-grade glial/glioneuronal neoplasm per SINQ 20190105 9490/0: Ganglioneuroma 9503/3: Neuroepithelioma, NOS 9509/1: Papillary glioneuronal tumor	Neuroepithelial tumor with PATZ1-EWSR1 fusion, not elsewhere classified **8000/1**

SINQ 20220006:

- Question: Histology/Brain and CNS: How is histology coded for a 2021 diagnosis of "neuroepithelial tumor with PATZ1-EWSR1 fusion, not elsewhere classified" found during a right thalamic mass resection?
- Answer: **Assign 8000/1**. Neuroepithelial tumor with PATZ1-EWSR1 fusion, not elsewhere classified, is not recognized as a distinct entity at this time. It is not listed in ICD-O-3.2 or in the 5th edition of the WHO CNS classification.

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Brain Histology – SINQ Only Scenarios

Brain 11 Selected Path:

ADDENDUM DIAGNOSIS

A) Brain, occipital, left, resection:
Integrated diagnosis: Neuroepithelial tumor with PATZ1 fusion, not elsewhere classified (NEC)
~~Histological classification: Glioneuronal neoplasm~~
WHO Grade: Not applicable
Molecular information:
- IDH: Negative for IDH1 (R132H) mutation by immunohistochemistry and IDH1/2 sequencing (exon 4)
- ATRX: Retained nuclear immunoreactivity
- p53: Increased immunoreactivity, 10-50% of neoplastic cells
- BRAF: Negative for BRAF (V600E) mutation by immunohistochemistry and negative for BRAF mutations and fusions by sequencing
- FGFR: Negative for mutations, amplifications, and fusions involving the tested regions of FGFR1, FGFR2, FGFR3, andFGFR4 by sequencing
- Methylation array: Neuroepithelial tumor with PATZ1 fusion (methylation class score 0.99999)
- NGS: Positive for an EWSR1-PATZ1 fusion. See COMMENT

ADDENDUM COMMENT

. . . Methylation array profiling demonstrated a significant match (0.99999) for the category neuroepithelial tumor with PATZ1 fusion, and unmethylated MGMT promoter. NGS analysis confirmed the presence of an EWSR1-PATZ1 fusion. The combined findings support a diagnosis of neuroepithelial tumor with PATZ1 fusion . . . Neuroepithelial tumor with PATZ1 fusion is a recently described provisional entity which is not currently listed in the 2021 WHO Classification of Tumours of the Central Nervous System and therefore is not assigned a CNS WHO grade.

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Brain Histology – Ambiguous Term Scenarios

- Brain 09: 04/07/2023 ROS Final Diagnosis = Embryonal tumor, consistent with medulloblastoma, SHH-activated. Diagnosis comment only confirmed, "this tumor is SHH-activated." Clinical correlation supporting specific histology = 05/10/2023 Neurosurgeon Note: **"Pt w/ sonic hedgehog-activated medulloblastoma." = 9471/3**
  - SHH = Sonic hedgehog. SHH-activated = sonic hedgehog-activated.
- Brain 14: 04/10/2023 Final Diagnosis = Extensively sclerotic low grade glioma, FGFR1-mutatnt and PIK3CA-mutant, consistent with rosette-forming glioneuronal tumor, CNS WHO grade 1. Diagnosis comment discussed the biomarker results without a definitive diagnosis; the comment only notes, "RGNTs frequently have co-occurrence of PIK3CA mutations with FGFR1 mutation." Clinical correlation supporting specific histology = 05/02/2023 Neurosurgery Note: **"RGNT s/p subtotal resection." = 9509/1**
  - RGNT = Rosette-forming glioneuronal tumor.

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Brain Histology – Ambiguous Term Scenarios

Case	NOS Histology	Ambiguous Term Used	Specific Histology	Resulting Morphology
09	~~Embryonal tumor = 9473 (CNS embryonal tumor, NEC/NOS)~~	Consistent with	Medulloblastoma, SHH-activated = 9471	Medulloblastoma, SHH-activated (per ICD-O-3.2 table) **9471/3**
14	~~Low-grade glioma = 9380/3~~	Consistent with	Rosette-forming glioneuronal tumor, CNS WHO grade 1 = 9509/1	Rosette-forming glioneuronal tumor **9509/1**

Coding Histology, Rule 3B, bullet 1: Code the specific histology described by ambiguous terminology when there is a NOS histology and a more specific (subtype/variant) described by ambiguous terminology:

- Specific histology is clinically confirmed by a physician (attending, pathologist, oncologist, etc.).

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Brain Histology – Ambiguous Term Scenarios

- Case 09: **Morphology code is 9471/3 – Medulloblastoma, SHH-activated.**
  - The Solid Tumor Rules, Table 3, does not provide an exact match to the case at hand.
    - Remember, the Solid Tumor Rules (including the tables) have the first coding priority, but the Solid Tumor Rules manual is not the only source.

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Medulloblastoma NOS **9470**	Classic medulloblastoma Medulloblastoma, histologically defined	Anaplastic/large cell medulloblastoma **9474** Medulloblastoma described as one of the following **9471** Desmoplastic SHH-activated and TP53-wildtype With extensive nodularity Nodular Medulloblastoma non-WNT/non-SHH; medulloblastoma group 3 or group 4 **9477*** Medulloblastoma SHH-activated and TP53-mutant **9476*** Medulloblastoma WNT-activated **9475***

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Brain Histology – Ambiguous Term Scenarios

- Case 09: **Morphology code is 9471/3 – Medulloblastoma, SHH-activated.**
  - When the Solid Tumor Rules don't provide the histology, move on to the ICD-O-3.2 table and all updates. The ICD-O-3.2 table does provide a histology for a diagnosis of "Medulloblastoma, SHH-activated."
    - Note: This histology is included in the ICD-O-3.2 table implemented in 2021. This histology has NOT been removed or made obsolete for 2022 or 2023.

9470/3	Preferred	Medulloblastoma, NOS
9470/3	Related	Classic medulloblastoma
9470/3	Related	Melanotic medulloblastoma
9471/3	Preferred	Desmoplastic nodular medulloblastoma
9471/3	Synonym	Circumscribed arachnoidal cerebellar sarcoma
9471/3	Synonym	Desmoplastic medulloblastoma
9471/3	Related	Medulloblastoma with extensive nodularity
9471/3	Related	Medulloblastoma, SHH-activated and TP53-wildtype
9471/3	Related	Medulloblastoma, SHH activated, NOS
9472/3	Preferred	Medullomyoblastoma

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SEER*Educate Other Sites Histology Exercises

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Other Sites Histology – Cases/Issues Not Discussed

Type of Issues	Case #	Accuracy Rate	Helpful Hints
Primary Site (Other issues may be discussed)	05	85.9%	**Primary Site should be coded to the site the tumor arose in.** • Use the ICD-O-3 (hardcopy manual) for the primary site list, the SEER Program Coding Staging Manual instructions. • Use all the information available to code the primary site. • Code the site of origin, not the site of metastatic tumor. • Code the site of origin, not the biopsy site. Primary tumors may involve more than one site and a biopsy-site may not represent the origin (e.g., intrahepatic bile duct tumors are C221, even if the diagnosis is made on a "liver" biopsy). • Code the overlapping site code (e.g., C168) when there is a single tumor overlapping multiple subsites and there is no indication of the subsite of origin
	06	54.5%	
	09	83.5%	
	16	84%	
	18	64.5%	
General Coding Histology Rule	11	54.9%	**Remember to use the General Coding Histology rules!** Coding Histology rule 2: Code the histology described as differentiation ONLY when there is a specific ICD-O code for the "NOS with ____ differentiation". • Carcinoma with neuroendocrine differentiation = 8574 per the ICD-O-3.2
Histology	05	96.3%	**Discussion of histology is not required.** • All frequencies are greater than 90%.
	08	96.4%	
	14	93.3%	
	20	92.6%	

CASES CUT FOR TIME!

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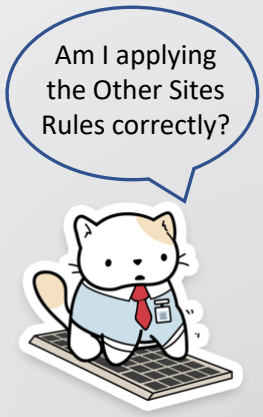
Other Sites Histology – Cases/Issues Discussed

General Types of Issues	Case #	Frequency (Preferred Histology)
Site-Specific Tables	01	76.2%
	06	51.2%
	10	89.3%
	12	38.5%
	16	83.2%
Mixed Histologies	02	70.8%
	03	32.7%
Specific H Rules	04	44.3%
	07	61%
	19	54.8%
Ambiguous Terminology	09	63.8%
Neoadjuvant Treatment	13	33.2%
	18	65.3%
p16 Histologies	15	44.9%
	17	71.7%



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Other Sites Histology – Site-Specific Tables Scenarios

- Other Sites 01: Resection Final Diagnosis & Synoptic Report = Primary Site: C570 (Fallopian tube), Histology: **High grade serous carcinoma = 8461/3.**
  - Rule H12 (single histology) & Site-Specific Table = Table 15: Fallopian Tube Histologies
- Other Sites 06: Liver Bx Final Diagnosis = Adenocarcinoma. PE & Med-Onc Note = Intrahepatic cholangiocarcinoma. Primary Site: C221 (Intrahepatic bile ducts), Histology: **Adenocarcinoma = 8140/3.**
  - Rule H12 (single histology) & Site-Specific Table = Table 9: Liver and Intrahepatic Bile Duct Histologies
- Other Sites 10: Hemithyroidectomy Synoptic Report = Primary Site: C739 (Thyroid), Histology: **Follicular carcinoma, encapsulated angioinvasive type = 8339/3.**
  - Rule H15 (NOS & subtype/variant) & Site-Specific Table = Table 12: Thyroid Histologies
- Other Sites 12: Diagnosis Comment & Synoptic Report = Primary Site: C621 (Descended testis), Histology: **Adult-type teratoma with a somatic-type malignancy = 9084/3.**
  - Rule H21 (combination code) & Site-Specific Table = Table 4: Testis Histologies
- Other Sites 16: Whipple Final Diagnosis = Primary Site: C241 (Ampulla of Vater; Intra-ampullary (SINQ 20210051)), Histology: **Pancreatobiliary-type carcinoma = 8163/3.**
  - Rule H15 (NOS & subtype/variant) & Site-Specific Table = Table 7: Small Intestine and Ampulla of Vater Histologies

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Other Sites Histology – Site-Specific Tables Scenarios

- Other Sites 06: Liver Biopsy Final Diagnosis: Adenocarcinoma (NOS). Primary Site: C221 (Intrahepatic bile ducts) based on the subsequent Med-Onc Note.
  - Pathology diagnosis = Adenocarcinoma
  - Clinical diagnosis = Cholangiocarcinoma
  - Applicable H Rule: Rule H12 – Code the histology when only one histologic type is identified.
  - Applicable Site-Specific Table: Table 9: Liver and Intrahepatic Bile Duct Histologies

Coding notes for Cholangiocarcinoma: Intrahepatic cholangiocarcinomas are almost exclusively adenocarcinomas and often diagnosed by cytology. Additional diagnostic molecular tests and clinical collaboration are needed to define a diagnosis of cholangiocarcinoma. Clinicians often indicate a clinical diagnosis of cholangiocarcinoma without pathologic confirmation. Per histology coding rules, pathology and cytology have priority over clinical/physician diagnosis. If the diagnosis of cholangiocarcinoma is made on a resected specimen, then code this histology.

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Other Sites Histology – Site-Specific Tables Scenarios

- Other Sites 06 (cont.): Liver Biopsy Final Diagnosis: Adenocarcinoma (NOS). Primary Site: C221 (Intrahepatic bile ducts) based on the subsequent Med-Onc Note.
  - Pathology diagnosis = Adenocarcinoma **= 8140/3 per ICD-I-O-3.2**
  - Clinical diagnosis = Cholangiocarcinoma
  - Applicable Site-Specific Table: Table 9: Liver and Intrahepatic Bile Duct Histologies

Specific or NOS Terms and Code	Synonyms	Subtypes/Variants
Carcinoma, undifferentiated **8020/3**		
Cholangiocarcinoma **8160**	Bile duct adenocarcinoma/carcinoma Intrahepatic cholangiocarcinoma (iCCA) Large duct intrahepatic cholangiocarcinoma Small duct intrahepatic cholangiocarcinoma	
Combined hepatocellular carcinoma and cholangiocarcinoma **8180**	Hepatocholangiocarcinoma Mixed hepatobiliary carcinoma Mixed hepatocellular-cholangiocarcinoma	
Hepatoblastoma **8970/3**		

No entry for Adenocarcinoma, NOS

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Other Sites Histology – Site-Specific Tables Scenarios

- Other Sites 12: Resection Primary Site: C621 (Descended testis), Resection Diagnosis Comment = "Adult-type teratoma with a somatic-type malignancy."
  - The Final Diagnosis and Synoptic Report do not disprove the Diagnosis Comment; they only further describe the "somatic-type malignancy."
    - Final Diagnosis of "Teratoma with embryonic-type neuroectodermal tumor" is the same as "Teratoma with somatic-type malignancy" because the embryonic-type neuroectodermal tumor is a somatic-type malignancy.
    - A teratoma with somatic-type malignancy is a teratoma with a distinct component resembling histologies seen in other organs or tissues (e.g., a sarcoma).
    - An embryonic-type neuroectodermal tumor is a primitive neuroectodermal tumor, and the WHO Blue Book (Urinary and Male Genital Tumors, 5th Ed.) confirms an embryonic-type neuroectodermal tumor is a relatively common somatic-type malignancy seen in this type of tumor.
  - This is both a mixed histology tumor (Teratoma PLUS somatic-type malignancy) and a specific histology code for the testis. Both tumor components must be considered when coding the histology.

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Other Sites Histology – Site-Specific Tables Scenarios

- Other Sites 12 (cont.): Resection Primary Site: C621 (Descended testis), Resection Diagnosis Comment = "Adult-type teratoma with a somatic-type malignancy." **= 9084/3**
  - Applicable H Rule: Rule H12 vs. Rule H21 – Code the single histology vs. Code a combination code from Table 2 or Table 4.
  - Applicable Site-Specific Table: Table 4: Testis Histologies

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Germ cell tumor, NOS **9064** *Note*: The following teratomas are **not reportable**: Teratoma, prepubertal type 9084/0 Teratoma, mature, prepubertal type 9084/0	Germ cell neoplasia in situ **9064/2** Intratubular germ cell neoplasia **9064/2** Intratubular malignant germ cells **9064/2**	Choriocarcinoma **9100** Embryonal carcinoma **9070** Spermatocytic seminoma/Spermatocytic tumor with sarcomatous differentiation **9063** Yolk sac tumor/Yolk sac tumor, prepubertal **9071** Teratoma with malignant transformation/Teratoma with somatic-type malignancy **9084**

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Other Sites Histology – Mixed Histology Scenarios

- Other Sites 02: Resection Final Diagnosis & Synoptic Report = Primary Site: C541 (Endometrium), Histology: **Mixed serous carcinoma and endometrioid carcinoma = 8323/3.**
  - Rule H21 (combination code/mixed histologies) & Table 2: Mixed and Combination Codes
- Other Sites 03: Prostate Core Bx Final Diagnosis = Primary Site: C619 (Prostate), Histology: Two-component carcinoma: **Acinar adenocarcinoma & Small cell neuroendocrine carcinoma = 8045/3.**
  - Rule H21 (combination code/mixed histologies) & Table 2: Mixed and Combination Codes

➢ Both cases follow the same general steps once the distinct or specific histologies comprising these tumors are identified. In both cases, the histologies under consideration are **NOT** an NOS and a more specific histology.

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Other Sites Histology – Mixed Histology Scenarios

- Other Sites 03: Prostate Core Bx Final Diagnosis = Primary Site: C619 (Prostate), Histology: "Summary findings: Two-component carcinoma; Acinar adenocarcinoma, 3+4=7, Grade group 2; Small cell neuroendocrine carcinoma; These components are separate and mixed in different cores."
  - Applicable H Rule: Rule H21 – Code a combination code when there are multiple specific histologies AND the combination is listed in Table 2.
    - The two specific histologies under consideration are:
      - Acinar adenocarcinoma, 8140/3
      - Small cell neuroendocrine carcinoma, 8041/3
  - Applicable Histology Table: Table 2: Mixed and Combination Codes because Rule H21 refers one to this table first!
    - Note: Rule H21 also indicates the Site-Specific Table (e.g., Table 3) can also be used when the Site-Specific Table includes the mixed histology code. Table 3 does not include this mixed histology tumor.

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Other Sites Histology – Mixed Histology Scenarios

- Other Sites 03 (cont.): Prostate Core Bx Histologies:
  - Acinar adenocarcinoma
  - Small cell neuroendocrine carcinoma

= 8045/3

  - Applicable Histology Table: Table 2: Mixed and Combination (Row 1)

Required Histology Terms	Histology Combination Term and Code
Small cell carcinoma/neuroendocrine tumor (NET) **AND** **At least one** of the following: • Adenocarcinoma and any subtype/variant of adenocarcinoma • Adenosquamous carcinoma • Large cell carcinoma and any subtype/variant of large cell carcinoma (includes large cell neuroendocrine carcinoma) • Squamous cell carcinoma and any subtype/variant of squamous cell carcinoma • Non-small cell carcinoma	Combined small cell carcinoma **8045**

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Other Sites Histology – Mixed Histology Scenarios

- Other Sites 03 (cont.): Histology = 8045/3 (Combined small cell carcinoma):
  - Why don't other histologies from Table 2 apply?
    - 8154/3: Mixed acinar-endocrine/neuroendocrine carcinoma (Frequency 13.3%)
    - 8244/3: Mixed adenoneuroendocrine carcinoma/combined carcinoid and adenocarcinoma (Frequency 2.8%)
  - These histologies are not clearly applicable to the prostate; they are predominantly applicable to gastrointestinal, biliary, and pancreatic primaries (confirmed by the WHO Blue Books).
  - These specific histologies can be found in the Site-Specific Tables for Stomach, Colon, Esophagus, Small Intestine, Anus, Liver and Bile Duct, Gallbladder, and Pancreas.
    - It seems when these histologies can apply, these specific mixed histology codes are ALSO included in the Site-Specific tables.
    - These histologies are **NOT** listed for the prostate (Table 3: Prostate Histologies).

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Other Sites Histology – Specific H Rule Scenarios

- Other Sites 04: Resection Final Diagnosis = Primary Site: C170 (Duodenum), Histology: **Adenomatous polyp with invasive adenocarcinoma = 8210/3.**
  - Rule H13 (Code 8210 when the final diagnosis is adenocarcinoma in a polyp). Specific histology provided by the H Rule.
- Other Sites 07: Resection Final Diagnosis = Primary Site: C739 (Thyroid), Histology (Dx Comment): **Poorly differentiated thyroid carcinoma with anaplastic carcinoma = 8021/3.**
  - Rule H16 (Code anaplastic carcinoma of thyroid (8021) when other thyroid histologies are present in a **single** tumor. There was a single 5 cm tumor in this case. Specific histology provided by the H Rule.
- Other Sites 19: Resection Final Diagnosis = Primary Site: C250 (Head of pancreas), Histology: **Invasive adenocarcinoma arising in an intraductal oncocytic papillary neoplasm (IOPN) = 8455/3.**
  - Rule H8, Note 1 & 2: Code the invasive histology when both invasive and in situ components are present WHEN the Site-Specific Table (Tables 3-21) does not include a specific ICD-O code for the tumor with both invasive and in situ histologies.
  - Additional use of Table 11: Pancreas Histologies is needed to determine the specific histology code.

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Other Sites Histology – Specific H Rule Scenarios

- Other Sites 19: Resection Final Diagnosis = Primary Site: C250 (Head of pancreas), **Histology: Invasive adenocarcinoma, arising in an intraductal oncocytic papillary neoplasm (IOPN) = 8455/3.**
  - Applicable H Rule: Rule H8, Note 1 & 2 plus Table 11: Pancreas Histologies.
  - Rule H8: Code the invasive histology when both invasive and in situ components are present.
    - Note 1: Use Tables 3-21, ICD-O, and all ICD-O updates to determine if the term containing both invasive and in situ histologies has a specific ICD-O code.
    - Note 2: When the term is not listed in Tables 3-21, ICD-O, and ICD-O updates, ignore the in situ term.

➢ In other words: Do NOT ignore the in situ when there is a specific code for the in situ and invasive histology. This is a change from 2007!

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Other Sites Histology – Specific H Rule Scenarios

- Other Sites 19 (cont.): Histology: Invasive adenocarcinoma, arising in an intraductal oncocytic papillary neoplasm (IOPN).
  - Don't ignore the in situ tumor component (intraductal oncocytic papillary neoplasm (IOPN)) because there is a SPECIFIC histology code for this combined invasive and in situ tumor.
    - Morphology 8140/3 (Invasive adenocarcinoma) was a common response for this case. Are Rule H8, Notes 1 and 2 being ignored?
  - Following the instructions in Note 1, use Table 11: Pancreas Histologies to determine if this is an invasive plus in situ tumor: **= 8455/3**

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Intraductal oncocytic papillary neoplasm 8455	Intraductal oncocytic papillary neoplasm with associated invasive carcinoma 8455/3 Intraductal oncocytic papillary neoplasm, NOS 8455/2	

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Other Sites Histology – Ambiguous Term Scenario

- Other Sites 09: Primary Site: C402 (Femur). **Histology: Ewing Sarcoma = 9364/3.**
- There are three histology statements to take into account:
  1. 04/15/2023 Biopsy Final Diagnosis: "Round cell sarcoma, see Comment."
  2. 04/15/2023 Biopsy Diagnosis Comment: "Overall based on the morphology and immunophenotype the findings are compatible with a round cell sarcoma, favor Ewing sarcoma."
  3. 05/01/2023 Med-Onc Note: "Ewing sarcoma, Lt distal femur."
- While ambiguous terminology (favor) describes the more specific type of round cell sarcoma, the histology is coded to Ewing sarcoma for this case.

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Other Sites Histology – Ambiguous Term Scenario

- Other Sites 09 (cont.): Histology: Round cell sarcoma, favor Ewing sarcoma.
- Coding Histology Rule 3B: Code the specific histology described by ambiguous terminology **ONLY** when 3A or 3B is true.
  - 3B: There is a NOS histology and a more specific (subtype/variant) described by ambiguous terminology and the specific histology is clinically confirmed by a physician (attending, pathologist, oncologist, etc.).
- The Med-Onc confirmed the specific histology described by ambiguous terminology meeting the criteria in Rule 3B.
- Histology is based on the Med-Onc clinical confirmation: **Ewing Sarcoma = 9364/3**

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Other Sites Histology – Ambiguous Term Scenario

- Other Sites 09 (cont.): Why doesn't histology 9366 (Round cell sarcoma with EWSR1-non ETS fusions) apply?
  - This was a frequent response and prompted disagreement with preferred answer emails from users.
- Ewing sarcoma is a type of round cell sarcoma.
  - While round cell sarcoma has a specific histology code, Ewing sarcoma is a specific type of round cell sarcoma.
  - The WHO Blue Book defines Ewing sarcoma as, "a small round cell sarcoma showing gene fusions involving one member of the FET family of genes (usually EWSR1) and a member of the ETS family of transcription factors."
    - In other words, Ewing sarcoma usually has an EWSR1-ETS fusion.
- Path proved this Ewing sarcoma has an EWSR1 fusion (gene rearrangement), but there is NO mention of a fusion with a non-ETS gene.

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Other Sites Histology – Neoadjuvant Rx Scenarios

- Other Sites 13: Primary Site = C492 (Soft tissue, thigh). **Histology = 8900 (Rhabdomyosarcoma) – The histology prior to neoadjuvant therapy based on the primary tumor diagnostic biopsy.**
  - 04/01/2023 Rt thigh mass, Biopsy Final Diagnosis = Rhabdomyosarcoma. Biopsy Diagnosis Comment = ". . .findings which do not distinguish clearly between alveolar and embryonal subtypes."
  - 08/01/2023 Rt thigh mass, Resection Final Diagnosis & Synoptic Report = Treated alveolar rhabdomyosarcoma. Clinical History = "Radical resection right thigh rhabdomyosarcoma, pt s/p neoadjuvant chemotherapy."
- Other Sites 18: Primary Site = C168 (Overlapping lesion of stomach). **Histology = 8154 (Mixed neuroendocrine non-neuroendocrine neoplasm (MiNEN)) – The histology from the primary tumor resection after neoadjuvant therapy based on diagnostic mets cytology.**
  - 04/01/2023 Pleural fluid, Lt thoracentesis, Final Diagnosis = Positive for malignancy. Malignant cells are consistent with adenocarcinoma.
  - 04/11/2023 Med-Onc Note: "Plan is for curative intent rx w/ neoadjuvant chemo, followed by resection."
  - 09/15/2023 Total gastrectomy, Resection Final Diagnosis & Synoptic Report = Mixed neuroendocrine non-neuroendocrine neoplasm (MiNEN), post-treatment ypT3 disease.

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Other Sites Histology – Neoadjuvant Rx Scenarios

- Other Sites 13 & 18: Should we code the histology before or after neoadjuvant treatment?
- Priority Order for Using Documents to Identify Histology includes Important Notes and an Exception:
  1. Code the histology diagnosed prior to neoadjuvant treatment.

     Note 1: Histology changes may occur following immunotherapy, targeted therapy, and radiation therapy.

     Note 2: Neoadjuvant treatment is any tumor-related treatment given prior to surgical removal of the malignancy.
- ***Exception:*** If the initial diagnosis is based on histology from FNA, smears, cytology, or from a regional or metastatic site, and neoadjuvant treatment is given and followed by resection of primary site which identifies a different or specific histology, code the histology from the primary site.

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Other Sites Histology – Neoadjuvant Rx Scenarios

- Other Sites 13 & 18 (cont.): Should we code the histology before or after neoadjuvant treatment?
- Other Sites 13 – The initial diagnosis was based on a **primary tumor tissue biopsy** (Rt thigh mass biopsy).
  - This doesn't meet the ***Exception*** because it was not a cytology and it was not a metastatic site biopsy.
  - The primary tumor tissue biopsy only diagnosed "Rhabdomyosarcoma" (NOS) **= 8900/3**
- Other Sites 18 – The initial diagnosis was based on **metastatic cytology** (pleural effusion). A primary tumor resection followed neoadjuvant treatment.
  - This does meet the Exception because it was both a cytology and a mets!
  - The primary tumor resection following treatment proved "Mixed neuroendocrine non-neuroendocrine neoplasm (MiNEN)" **= 8154/3**

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Other Sites Histology – p16 Histology Scenarios

- Other Sites 15: Excision Final Diagnosis = Primary Site: C538 (Overlapping lesion of cervix uteri), Histology: **Invasive adenocarcinoma, p16 diffusely positive = 8483/3.**
  - Rule H12 (single histology) & Site-Specific Table = Table 17: Uterine Cervix Histologies
- Other Sites 17: Biopsy Final Diagnosis = Primary Site: C211 (Anal canal), Histology: **"highly suspicious for invasive well differentiated (p16 negative) squamous cell carcinoma." = 8086/3**
  - Rule H12 (single histology) & Site-Specific Table = Table 8: Anus Histologies

➢ For sites where p16 test results can be used to code a specific HPV-related histology, there will be a NOTE before the Site-Specific Table indicating this.

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Other Sites Histology – Site-Specific Tables Scenarios

- Other Sites 15: Excision Final Diagnosis = Histology: Invasive adenocarcinoma, p16 diffusely positive. **= 8483/3**
  - Applicable H Rule: Rule H12 – Code the histology when only one histologic type is identified.
  - Applicable Site-Specific Table: Table 17: Uterine Cervix Histologies

Uterine Cervix Coding Notes

- In situ carcinoma of cervix (/2), any histology, is not reportable
- p16 is a valid test to determine HPV status and can be used to code HPV associated and HPV independent histologies

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Adenocarcinoma NOS **8140/3**		Adenocarcinoma, HPV-associated **8483/3** Adenocarcinoma, HPV-independent **8484/3** Adenocarcinoma, HPV-independent, gastric type **8482/3** Adenocarcinoma, HPV-independent, clear cell type **8310/3** Adenocarcinoma, HPV-independent, mesonephric type **9110/3**

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Brain & Other Sites Histologies – In Summary

- Follow the Solid Tumor Rules for coding histology.
  - The Other Sites schema now follows the same general rules as the rest of the Solid Tumor Rules schemas, but there are changes for 2023 and we need to follow the rules to code the appropriate histology.
- Remember, there are priority orders for coding histology!
  - There's a priority order for using documents to identify the histology (e.g., for Brain, the biomarkers have the highest priority).
  - There's a separate priority for coding histology once the highest priority source is identified (e.g., biomarkers, primary tumor pathology, etc.).:
    - Use the Site/Schema-Specific Histology tables first! If the applicable histology is listed, we don't need to look further.
    - Use the ICD-O-3.2 table and all its updates (i.e., the annual NAACCR Implementation Guidelines) next.
    - If no histology can be identified in either of these sources, use SINQ, followed by Ask a SEER Registrar. Sometimes the rules do not lead one to a histology code and we need to ask the standard setter for clarification.
- When questions arise while coding histology, we need to submit them to the standard setter.
  - Areas of confusion will remain confusing unless we speak up!

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Questions?

Histology codes and rules continue to evolve and change.

Don't be discouraged - we can keep up with the changes and improve our coding accuracy and consistency!

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Thank You!

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